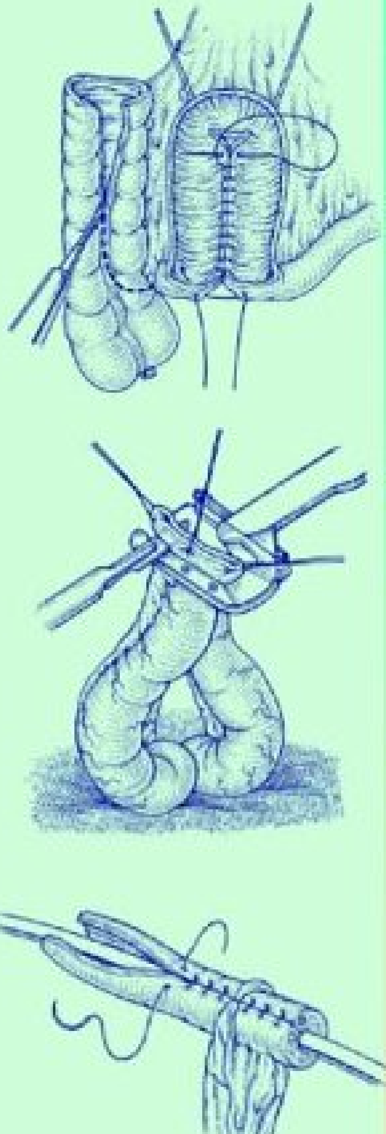

SECOND EDITION

Operative Pediatric Urology

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Churchill Livingstone

Operative Pediatric Urology

Second Edition

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CHURCHILL LIVINGSTONE

London • Edinburgh • New York • Philadelphia • St Louis • Sydney • Toronto 2002

CHURCHILL LIVINGSTONE
An imprint of Harcourt Publishers Limited

First published 1990
Second Edition 2002

ISBN 0-443-05358-8

British Library Cataloguing in Publication Data
A catalogue record for this book is available from the British Library

Library of Congress Cataloging in Publication Data
A catalog record for this book is available from the Library of Congress

Note
Medical knowledge is constantly changing. As new information becomes available, changes in treatment, procedures, equipment and the use of drugs become necessary. The editors/authors/contributors and the publishers have taken care to ensure that the information given in this text is accurate and up to date. However, readers are strongly advised to confirm that the information, especially with regard to drug usage, complies with the latest legislation and standards of practice.

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Preface

Welcome to the second edition of *Operative Pediatric Urology*, first edited by J. David Frank and Herbert Johnston in 1990. Some chapters have remained the same, some have undergone major revision and new chapters particularly relating to continent diversion and the Mitrofanoff procedure have been added.

We have omitted chapters on laparoscopic and most endoscopic procedures because in our view they are better learnt using videos rather than pictorial representations. As in the first edition, urological procedures that are technically similar for all ages, such as renovascular surgery and the surgery for renal calculi, have not been included. An exhaustive description of every means of correction of the various lesions discussed would have led to the production of a multivolume encyclopedia. For that reason we have again asked expert contributors to describe the methods they have found to be most successful and therefore to be recommended to others.

An essential contribution to this book is that of our principal artist Philip Wilson who has again produced wonderful drawings often based upon inadequate information from the relevant surgeon! He was helped on this occasion by Gillian Oliver. We are truly indebted to them both.

As always, the production of textbooks undergoes a lengthy gestation and this edition is no exception. We have worked with various commissioning editors and even had a change of publishers before finally reaching term. Ultimate delivery was carried out by Sue Hodgson and Kim Benson. We are grateful to them and their colleagues for their endurance and continued enthusiasm. We would also like to thank Mrs Carole Bisouth for all her help in the production of the manuscripts.

It was with great sadness that we learnt of John Duckett's untimely and unexpected death during the preparation of this book. We have taken the liberty of retaining his chapter on hypospadias surgery that he so kindly updated for us as the best way of paying our respects to a truly great figure in pediatric urology.

All three editors have a good reason for being involved in this textbook. J. D. F. had the pleasure of editing the first edition with Herbie, enjoying his sense of humor, the occasional round of golf and his and Dorothy's wonderful hospitality. J.P.G. and H.M.S. both served as fellows with Herbie and look upon their time with him as the seminal event in their career in pediatric urology. Again, their friendship with Herbie and Dorothy has remained as important today as it was nearly thirty years ago.

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Acknowledgements

Figure 5.2 adapted from Kaplan GW 1976 Posterior urethra. In: Kelalis EP, King LR (eds) Clinical pediatric urology, WB Saunders, Philadelphia, with permission.

Figures 5.3a and b adapted from Retik AB 1991 Management of posterior urethral valves. In: Glenn JF (ed) Urologic surgery, 4th edn, JB Lippincott, Philadelphia, with permission.

Figure 5.5 adapted from Rich MA, Keating MA, Snyder HM III, Duckett JW 1990 Low transurethral incision of single system intravesical ureteroceles in children. Journal of Urology 144: 120, with permission.

Chapter 1

Pyeloplasty

J. David Frank

Introduction

A patient presenting with a pelviureteric junction (PUJ) obstruction may be asymptomatic following an antenatal diagnosis of the hydronephrosis or symptomatic in the older child. The diagnosis of obstruction and the decision of when to operate is easier to make in this latter group of patients. It is outside the scope of this book to discuss in detail the management of the antenatally diagnosed group. The conservative management of the majority of these patients is recommended, but there remains a group of these patients with diminished or diminishing renal function who will require surgery (Ransley et al 1990).

Diagnosis and investigations

The main symptom of a PUJ obstruction is usually flank pain, often associated with nausea or vomiting. Occasionally, a urinary tract infection or haematuria is the presenting feature. These patients should be investigated with a renal ultrasound examination. If this confirms a hydronephrosis without a dilated ureter, the diagnosis should be confirmed by either a MAG 3 renal scan and/or an intravenous urogram. In patients with a massive hydronephrosis, the stasis within the renal pelvis may be sufficiently severe to give a false impression of obstruction on a MAG 3 scan. In this situation, an intravenous urogram may help, as the visualization of well-preserved calyces in spite of a dilated renal pelvis will confirm that conservative management is appropriate. If the renal ultrasound is normal but the patient continues to get intermittent loin pain, an intermittent PUJ obstruction should be excluded by repeating the ultrasound during an attack of pain.

If the kidney is very poorly functioning, it may be difficult to decide whether it is obstructed and, if it is, on the exact site of obstruction. In this situation, an antegrade pyelogram with or without pressure studies may be required. The decision of whether to proceed to a pyeloplasty or a nephrectomy of a poorly functioning kidney, i.e. one with less than 10% of overall renal function, is difficult. It is probably better to be conservative and perform a pyeloplasty in children less than 1 year of age in the hope that there will be some recovery of renal function (King et al 1984). All children under the age of 1 year should undergo a cystogram to exclude vesico-ureteric reflux. If reflux is found, a bladder catheter should be used after the pyeloplasty to diminish the risk of an anastomotic leak.

Surgical technique

There are many different techniques available to repair a PUJ obstruction. The decision of which type to use should be taken at the time of surgery after the PUJ has been exposed. The dismembered pyeloplasty (Anderson 1963) is suitable for the majority of patients. There is occasionally a dependent PUJ with a long narrow ureteric segment and an Anderson–Hynes pyeloplasty may leave the surgeon short of ureteric length. A Culp–De Weerd pyeloplasty (Culp & De Weerd 1951) is then more suitable.

Surgical approach

Three approaches may be used: the loin, the anterior extraperitoneal and the lumbotomy. All kidneys can be approached by the loin with excellent exposure, but it is a painful incision postoperatively. The anterior extraperitoneal approach is excellent in younger children with a reasonably large renal pelvis, but access is more difficult in the older or obese child. The main advantage of this technique is speed of access and excellent postoperative healing of the scar. The lumbotomy incision is now being used by many surgeons. Its proponents emphasize the relatively pain-free postoperative period, thus allowing early discharge from hospital. Whichever approach is favored, it is important to stay extraperitoneal: any tear in the peritoneum allows the small bowel to herniate through the defect and obscure the operative field. If bilateral PUJ obstructions are being dealt with, a bilateral anterior extraperitoneal approach that avoids the necessity of re-towelling and re-positioning the patient, or a bilateral lumbotomy, may be used. The following operative description applies to the loin approach.

Dismembered pyeloplasty

The skin incision is made just below and in line with the twelfth rib. The external and internal oblique muscles are divided by cutting diathermy, and the transversus abdominis is split in the line of its fibers. The peritoneum is gently pushed medially, and a self-retaining Denis Browne retractor is inserted.

The renal fascia is opened, and the renal pelvis and PUJ are exposed using blunt and sharp dissection. In the majority of patients, a PUJ is best approached anteriorly, but it is sometimes easier to rotate the kidney, retracting the lower pole forwards and upwards to approach the PUJ from behind. If the kidney is severely hydronephrotic, it may be easier to deflate the renal pelvis first by aspirating it. Full mobilization of the kidney with delivery into the wound is usually unnecessary. The adventitia covering the renal pelvis and PUJ is incised laterally (Fig. 1.1). A 6/0 Vicryl or PDS suture is placed in the ureter just below the narrowed segment. Further sutures are placed in the renal pelvis to mark the upper and lower margins of the proposed line of excision. The excess renal pelvis is then resected. It should be trimmed in a straight line without jagged margins but with a slight angulation towards the kidney at the inferior part of the incision in order to prevent angulation of the anastomosis. The adventitia of the upper 2–3 cm of the ureter is freed laterally (Fig. 1.2). The ureter is spatulated by incising its posterolateral margin using fine scissors and avoiding its medial blood supply. A length of 1–2 cm of spatulation is normally adequate.

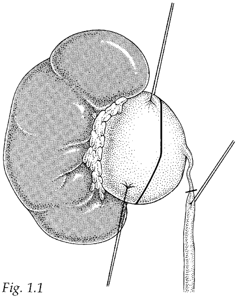


Fig. 1.1

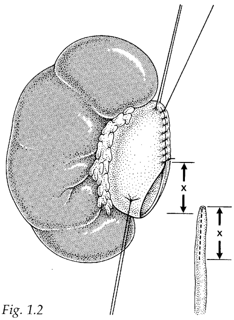
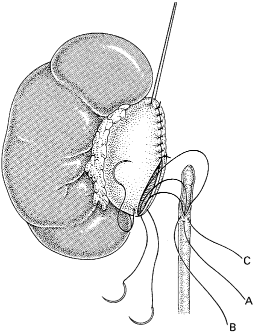


Fig. 1.2

The upper part of the renal pelvis is closed with a running 6/0 or 7/0 Vicryl or PDS suture until the length of the open renal pelvis equals the length of the spatulated ureter. The ureter is then anastomosed to the renal pelvis (Fig. 1.3). The first three sutures are of critical importance. The first stitch is an everting suture approximating the lower margin of the renal pelvis to the lower margin of the spatulated ureter (A). A further interrupted everting suture is then placed on either side of this initial stitch (B and C). The importance of these stitches lies in the fact that this is the narrowest part of the anastomosis. The posterior aspect of the anastomosis is then completed using 6/0 or 7/0 Vicryl or PDS (Fig. 1.4).

A fine catheter is passed temporarily through the anastomosis, and the anterior closure is performed using a running 6/0 or 7/0 suture (Fig. 1.5). The fine catheter prevents the inadvertent picking up of the posterio[r] wall during this final stage of the anastomosis. The catheter is removed just before the anastomosis is completed (Fig. 1.6). If the ureteric length is too long to fit exactly into th[e] opened renal pelvis, it is trimmed nea[r] its upper margin (Fig. 1.7). For infant pyeloplasties, loupe magnification wil[l] be found helpful.



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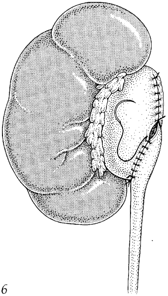

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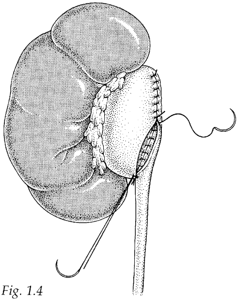
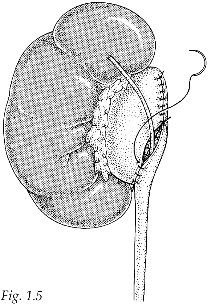


Fig. 1.4



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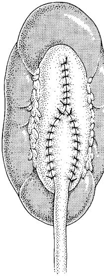


Fig. 1.7

Drainage

Normally, no stent or nephrostomy catheter is used. If, for some reason, the anastomosis is technically difficult, the tissues are friable or it is a single kidney, a double-J stent should be used. These have traditionally been removed cystoscopically approximately 6–8 weeks after surgery. The newer stents produced by Angiomed – 'the blue stent' – can be left knotted after passing through the skin. These can then be removed without requiring a cystoscopy. The results of unstented pyeloplasties using fine sutures and loupe magnification have been excellent (Hussain & Frank 1994).

The Culp–De Weerd pyeloplasty

There is occasionally a shortage of ureteric length with a dependent type of PUJ obstruction. A dismembered pyeloplasty will then leave the surgeon in difficulties, so in this situation a Culp–de Weerd pyeloplasty should be performed.

The exposure of the kidney is as described. The incision on the renal pelvis is carried from the mid-part of the renal pelvis superiorly, curving around and down the anterolateral margin of the upper ureter for approximately 1–2 cm (Fig. 1.8). The length of the flap and the length of the incision of the ureter must be equal. It is important that the base of the flap is of adequate width so that the length-to-width ratio is no more than 3–1.

Whether the flap is fashioned in a longitudinal manner (Scardino 1967) or in a spiral fashion depends on the shape of the renal pelvis. A long renal pelvis allows a vertical flap to be formed, whereas a more rectangularly shaped pelvis is better suited to a spirally shaped flap (Fig. 1.9). The flap is rotated inferiorly and sutured to the ureter (Fig. 1.10). The initial sutures – A, B and C – are as for a dismembered pyeloplasty. These sutures are placed to join the apex of the flap to the lower margin of the ureteric opening.

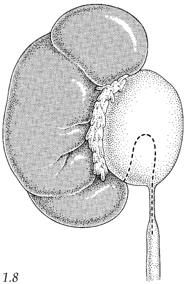
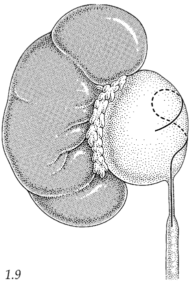


Fig. 1.8



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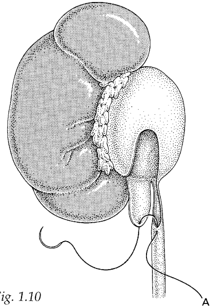

Fig. 1.10

A continuous 6/0 or 7/0 Vicryl or PDS suture is then used to complete the anastomosis (Figs 1.11–1.13). Again, a ureteric splint is used temporarily to avoid picking up the posterior wall.

Drainage and postoperative management are the same as for a dismembered pyeloplasty.

Lower pole vessels

A PUJ obstruction is sometimes associated with the presence of aberrant lower pole vessels. Whether these vessels are the cause of the obstruction or impinge on the PUJ secondarily is a matter of debate. When repairing an obstructed PUJ in these circumstances, a dismembered type of pyeloplasty should be performed so that the vessels can be placed posterior to the anastomosis.

Management of a PUJ obstruction of the upper or lower moiety of a duplex kidney

An obstruction of the PUJ of one moiety of a duplex kidney may affect either moiety but more commonly the lower. If there is poor function on an isotope scan, this condition is best treated by a heminephro-ureterectomy (see Chapter 16). The surgical management of either an upper or lower moiety PUJ obstruction must be individualized and will depend upon the exact anatomy found at the time of surgery. This anatomy may be best

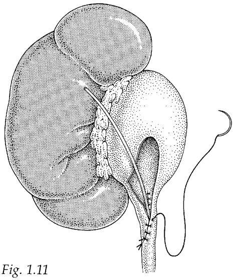

Fig. 1.11

defined using an antegrade pyelogram. The most frequent method of repair is an end-to-side or side-to-side anastomosis, as illustrated (Figs 1.14 and 1.15). Drainage with a double-J stent may be required, particularly when the ureter to be anastomosed to the renal pelvis is very narrow. Anastomotic techniques are otherwise the same as for a standard pyeloplasty.

Postoperative management

The patients normally receive one injection of an antibiotic at the time of injunction of anesthesia. The drain is removed on the second or third postoperative day unless there is drainage of urine. Patients with a double-J stent are normally sent home on the third postoperative day, to be

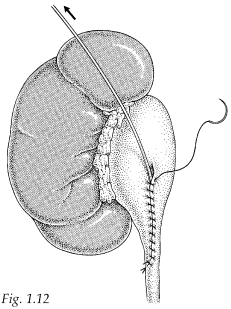

Fig. 1.12

re-admitted in 6–8 weeks for removal of the stent, cystoscopically if a traditional stent has been used. If one of the more modern stents has been used, the patient is sent home with the stent knotted, to be removed a week postoperatively.

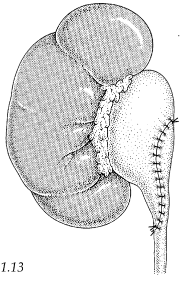

Fig. 1.13

If no stent has been used and the patient leaks urine postoperatively for more than 48 hours, a bladder catheter should be inserted. The urinary drainage will then normally cease. If, however, it continues, a double-J stent should be inserted. In a review of 130 patients, Homsy et al (1980) found that, with or without a urinary diversion, the leakage of urine for 7 days postoperatively was compatible with a successful surgical result.

Follow-up studies using a MAG 3 renal scan or intravenous pyelogram should be performed at 3 months.

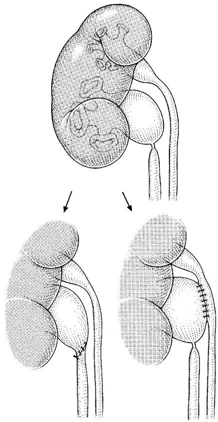

Fig. 1.14

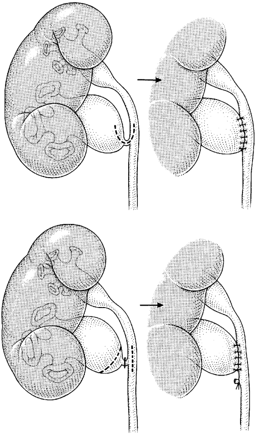

Fig. 1.15

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Chapter 2

Ureteric reimplantation for vesico-ureteric reflux

S. Joseph Cohen

Introduction

In the 1950s, the problem of vesico-ureteric reflux with urinary infection and resultant pyelonephritis became recognized, and many surgical operations to correct reflux were devised. At the time when the efficacy of operative techniques had been proven, physicians showed that a similar control of the situation could be obtained by long-term antimicrobial therapy accompanied by the practice of double micturition to reduce the residual urine volume. Extensive controlled double-blind studies are being carried out to compare these two methods, and there are staunch supporters of each.

It is the purpose of this chapter to illustrate the surgical techniques of reimplantation of the ureters and not to be involved in this controversy.

Preoperative investigations

Vesico-ureteric reflux was previously only demonstrable by micturating cysto-urethrography (MCUG). This procedure is invasive, exposes the child to a fair amount of radiation and is often uncomfortable, but it is still nevertheless the most effective method of demonstrating the condition. In addition, it is the best way to demonstrate the outflow tract, which is essential in order to differentiate primary reflux from reflux secondary to such outflow obstructions as urethral valves, strictures, duplications, polyps, etc. The newer methods of indirect and direct isotope renography and ultrasonography are being improved and may in the future supersede MCUG as the most effective way of making the diagnosis.

Radionucleide isotope scans have superseded the intravenous urogram as an essential preoperative investigation for patients with vesico-ureteric reflux. The DMSA static renal scan gives the best information about differential renal function and the presence or absence of renal scars. If there is any doubt about the drainage of the kidney, in particular whether there is an associated pelviureteric junction obstruction, the MAG 3 scan should be used.

The fact that vesico-ureteric reflux has been demonstrated does not necessarily mean that reimplantation is mandatory; far from it, for the minor degrees of reflux (stages 1 and 2 of Dwoskin & Perlmutter 1973) are best treated by non-surgical means. In grade 5, where there is gross reflux and severe reflux nephropathy, the use of either surgical or medical treatment is unable to reverse the renal pathology and its inevitable consequences.

Surgical techniques

Since the early 1950s, many surgical techniques have been devised for the correction of vesico-ureteric reflux. The earliest was probably that of Hutch (1963) followed by those of Politano & Leadbetter (1958), Paquin (1959), Glenn & Anderson (1967), Gregoir & Van Regemorter (1964) and Cohen (1975). The techniques are mainly divisible into two groups: first, those in which the dissection of the ureter is mainly or entirely intravesical, and second, those in which the dissection is entirely extravesical, of which the Gregoir technique is the best example.

The endoscopic injection of Teflon submucosally below and behind the ureteric orifice has recently been introduced by O'Donnell & Puri (1984). This technique is simple and the long-term results are very satisfactory. It also obviates long stays in hospital. The only drawback is that some countries have prohibited the use of Teflon in this situation. Consequently, other urologists have been encouraged to find alternative injectable materials such as Collagen and Microplastique. These have also proved to be efficacious. Time alone will prove whether these simple procedures are as safe and effective as the more extensive surgical operations listed above.

The operation

Endoscopy should always precede opening of the bladder in order to determine the size, shape and number of the ureteric orifices, to show that there is no outflow obstruction and to confirm that acute cystitis is absent. The latter should persuade the operator to postpone surgery until the inflammation has subsided.

Cohen trans-trigonal technique

A low, transverse, suprapubic incision is made (Fig. 2.1). Its position is such that it will be, in later life, hidden in the pubic hair and thus not prevent young girls wearing bikinis.

After the skin and subcutaneous tissue have been incised, the rectus sheath is exposed. The bladder is approached either by a true Pfannenstiel incision, i.e. a transverse division of the rectus sheath with separation of the rectus muscles, or by dividing the sheath vertically between the recti and then separating them. When this has been done, vesical stay sutures are inserted, and between them the bladder is opened vertically or by a T incision. A Denis Browne ring retractor is inserted as illustrated in Figure 2.2. A swab inserted under the apical retractor blade facilitates the exposure. The ureteric orifices are carefully examined with regard to site, size, number, shape and the length of tunnels. A fine infant feeding tube is inserted up the ureter

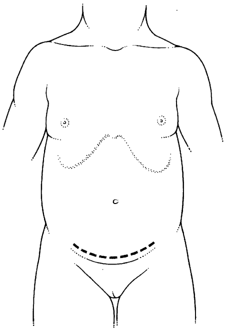


Fig. 2.1

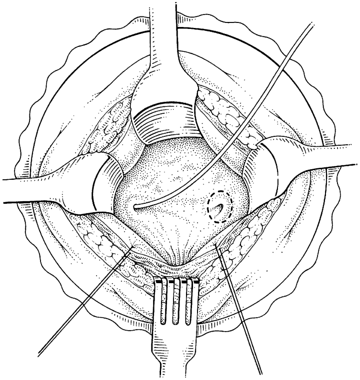


Fig. 2.2

patients, the vas deferens may lie close to this point, and accidental damage must be carefully avoided.

The ureter is now free and ready for reimplantation. In the majority of cases, the opposite ureter is freed in the same way, and reimplantation may commence. The dissection of the ureter may have enlarged its vesical hiatus to such an extent that it should be narrowed with one, two or three 3/0 or 4/0 Dexon sutures in order to prevent the subsequent formation of a bladder diverticulum.

and fixed by a fine Dexon or Vicryl Rapide suture, as shown on the patient's right side in the figure. A circumferential incision is then made around the orifice, as shown on the patient's left side.

The dissection of the ureter is now carried out. The incision is deepened, carefully dividing the muscle fibers that fix the ureter to the bladder trigone (Fig. 2.3). It is wise to commence just below the orifice and, once the plane between the ureter and bladder musculature has been found, this is progressively developed around the ureter until the latter has been completely freed. Great care must be taken not to dissect too close to the ureter for fear of damaging its blood supply or its musculature. The peritoneum, which almost surrounds the ureter, must be carefully teased away using a small pledget. In male

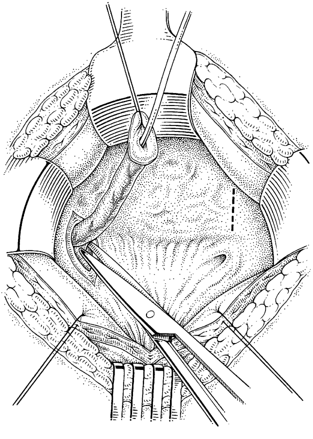


Fig. 2.3

The submucosal tunnel is then constructed; a point is selected above and perhaps a little lateral to the opposite ureteric orifice. An incision in the mucosa is made, and, after inserting its closed blades, a pair of scissors is advanced under the mucosa by an opening and closing movement (Fig. 2.4). This is continued until the ureter is reached. The tunnel should be a gentle curve, a prolongation of the entrance of the ureter into the bladder. Care must be taken as one approaches the midline for it is here that the mucosa is easily buttonholed. This can be best avoided by placing a pair of Allis forceps just lateral to the commencement of the tunnel: by gently retracting these laterally, the posterior bladder wall can be straightened and the problem averted. The new tunnel must be wide enough comfortably to hold the ureter, and long enough to prevent reflux. The minimal length is 2–3 times the ureteric diameter.

The ureter is now gently threaded through its new tunnel (Fig. 2.5). This is best accomplished by passing a pair of forceps into the tunnel, grasping the stay suture and drawing the ureter into place, taking care not to twist or kink it in the process.

The cuff of the ureter is then sutured into position (Fig. 2.6). First, a 3/0 Dexon suture is inserted through the full thickness of the cuff and also through a full thickness of bladder muscle. This prevents the ureter from retracting. Next, three or four 5/0 Dexon sutures are inserted, joining the bladder and ureteric mucosae. The incision in the mucosa of the original orifice is closed with fine Dexon sutures. Many surgeons do not believe in retaining this cuff of ureteric mucosa, but if it has not been devascularized, traumatized or the site of stenosis, this author believes that it is worth preserving. If any doubt exists, the terminal part of the ureter should be excised.

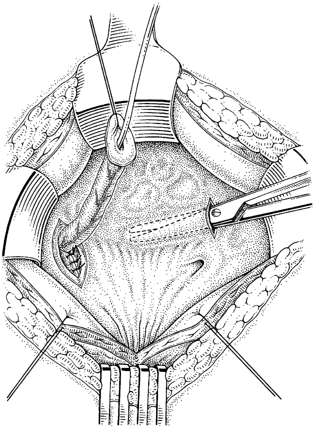


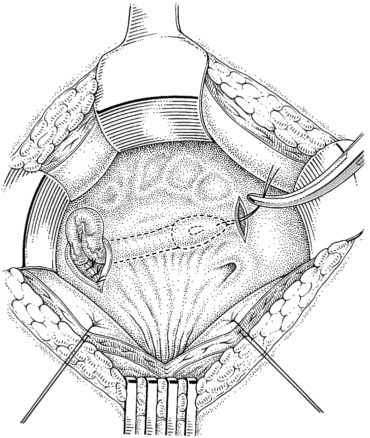
Fig. 2.4

Bilateral reimplantation

The dissection of both ureters having been carried out as mentioned above, the two ureters are held and assessed in terms of which would sit most comfortably in the upper tunnel. If both seem equally well suited, this author tends to use the upper tunnel for the ureter that has the more severe degree of reflux. The second tunnel is then constructed by the same technique as above. Its position is below the tunnel already made, and it goes from the ureteric entrance to the orifice of the opposite side. The ureter is threaded through in the same manner and similarly fixed in its new position. In the very young, where the intertrigonal distance is short, the tunnel may be extended through this orifice to a more lateral point and fixed there. Some surgeons have used a single tunnel for both ureters and have had no trouble with this, but the two ureters may theoretically adhere to one another, and this author still prefers to use separate tunnels.

Bladder closure

Having made sure that the ureters are lying comfortably in their new tunnels and are well vascularized to their tips, the swab is removed from the bladder; fine infant feeding tubes are inserted up the reimplanted ureters, and bladder closure is carried out. The mucosa should be closed with a continuous fine Dexon or Vicryl Rapide suture (4/0 or 5/0). The



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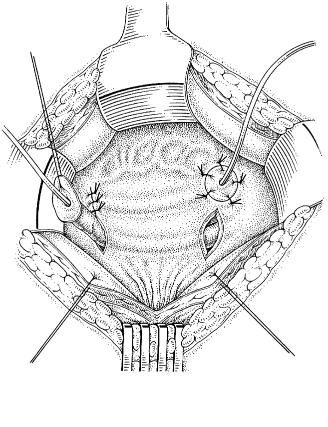

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bladder muscle is closed with interrupted 2/0 or 3/0 Dexon or Vicryl Rapide sutures, a Malecot catheter being left in situ for bladder drainage. In girls, one may use a Foley transurethral catheter. A drain is inserted retropubically and the muscles and fasciae closed with 2/0 or 3/0 Dexon or Vicryl Rapide sutures. The skin is closed with a continuous subcuticular 4/0 Dexon suture.

Reimplantation in duplex systems

Ureteric duplication may be complete or incomplete. Where it is complete, the lower moiety ureter is the one that is laterally placed and often incompetent. The two ureters are often very closely adherent and may even share a common musculature and blood supply. This necessitates the dissection of the two ureters as a single unit, without attempting to separate one from the other. The technique is therefore exactly the same as that illustrated in Figures 2.4 and 2.5.

Conclusions

Reimplantation of the ureters is recognized as a reliable method of correcting vesico-ureteric reflux. Its efficacy has been proven over the last three decades, and one should expect to achieve success in virtually all cases. The complication rate is also continuously being reduced, complications being expected in only 1–4% of cases. Obstructive complications are more common in those cases in which there was originally gross dilatation of the ureters or where the bladder was thickened and trabeculated owing to a neurogenic abnormality or previous outflow obstruction.

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Chapter 3

Ureteric tailoring and psoas hitch reimplantation for uretero-vesical obstruction

Joseph H. Johnston

Indications

The technique described here is particularly applicable to the obstructed megaureter. In this condition, which is most commonly unilateral, left-sided and affecting the male, there is a functional interference with the transmission of peristalsis in the terminal ureter. The obstructive ureteric segment remains of normal calibre while the ureter above becomes dilated, lengthened and tortuous, and the kidney shows hydronephrotic changes.

The operative procedure can also be employed when there is a fibrotic uretero-vesical obstruction secondary to such infravesical obstructions as posterior urethral valves. The orthotopic ureterocele may be similarly managed, usually combined with the excision of the ureterocele. The object of the operation is to relieve the obstructive effects on the renal parenchyma, the consequences of which are commonly exacerbated by infection or by stone formation in the dilated system; at the same time, subsequent vesico-ureteric reflux must be prevented.

Preoperative measures

Infrequently, in the acutely obstructed case or when there is severe infection, preliminary drainage of the system is needed. This can usually be obtained by the percutaneous insertion of a catheter into the renal pelvis under ultrasonographic guidance.

Surgical technique

An oblique, muscle-cutting incision is made in the appropriate iliac fossa, centered on the anterior superior iliac spine (Fig. 3.1). The dilated, tortuous ureter (left in the illustration) is exposed extraperitoneally.

The ureter is divided at the entrance to the bladder, with ligature of its distal extremity, and is straightened by the release of tortuosity as far as the ureteric blood supply allows (Fig. 3.2). The resulting redundant portion of the ureter is excised.

The lowest 5–7 cm of the remaining ureter generally needs to be narrowed. This is best performed by passing a 10F catheter into the ureter. The catheter is then enclosed, through the

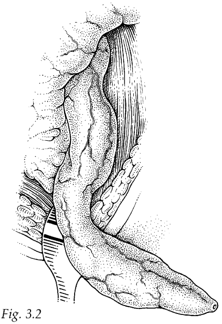

Fig. 3.2

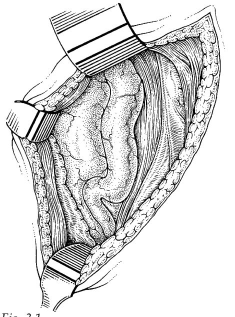

Fig. 3.1

ureteric wall, by Babcock or Hendren forceps (Fig. 3.3a). The ureter outside the grasping forceps is resected (Fig. 3.3b). It is important that the excised wedge of ureteral wall should be from its 'anti-mesenteric' aspect so that the mainly longitudinally running blood vessels are preserved.

The narrowed segment of ureter is closed, using interrupted 4/0 Vicryl Rapide sutures (Fig. 3.4). The temptation to produce a normal ureteric caliber must be resisted in order to avoid jeopardizing the blood supply. There is no need for extensive ureteric remodeling: only that portion which is to form the new intravesical ureter need be narrowed.

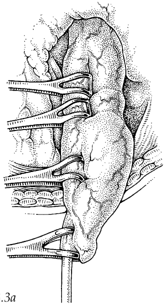

Fig. 3.3a

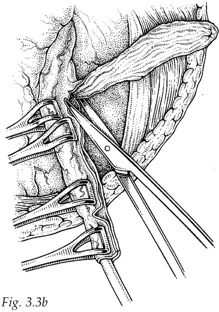

Fig. 3.3b

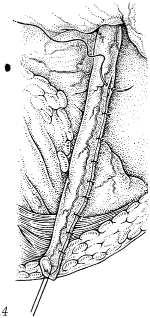

Fig. 3.4

The bladder is opened through an oblique incision on its superolateral aspect, and a submucosal tunnel is fashioned between the incision and the region of the original ureteric orifice (Fig. 3.5). In order to avoid subsequent angulation of the ureter at the new vesical hiatus during bladder filling, the musculature at the mouth of the tunnel is sutured, with three Vicryl Rapide stitches, to the psoas muscle lateral to the external iliac vessels.

The narrowed segment of ureter is passed through the submucosal tunnel, and its extremity is sutured posteriorly to the bladder musculature and circumferentially to the mucosal edges of the tunnel (Fig. 3.6). One should aim for the length of the new intravesical ureter to be five times its width. Ureteric splinting may or may not be considered necessary.

The bladder is closed with suprapubic drainage, and the wound is sutured (Fig. 3.7).

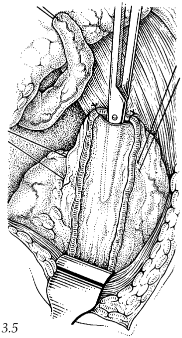

Fig. 3.5

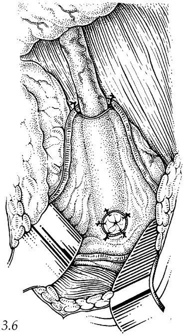

Fig. 3.6

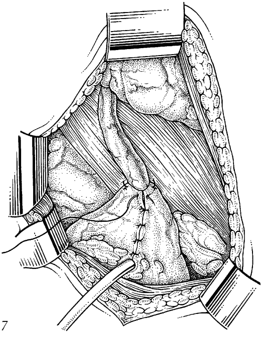

Fig. 3.7

Comments and results

Angulations of the ureter commonly exist in a tortuous megaureter, but they are not, of themselves, obstructive, and any which remain after the reimplantation operation straighten spontaneously with the growth of the patient. Secondary kinking at the pelviureteric junction can, however, persist and may necessitate pelviureteroplasty.

The technique described here can be relied upon to give highly satisfactory results, especially in cases of obstructed megaureter in which kidney function is generally well preserved and ureteric peristalsis remains active or at least recoverable, in spite of complications. The psoas hitch procedure cannot be performed bilaterally. When both ureters require reimplantation, this aspect of the technique must be omitted or, alternatively, one ureter may be reimplanted as described and the other divided near the bladder, brought to the contralateral side retroperitoneally and anastomosed end-to-side to the reimplanted ureter.

Chapter 4

Ureteric reimplantation of the megaureter

Clare E. Close and Michael E. Mitchell

Indications

The most efficient drainage of the kidney is accomplished by a peristaltic ureter that tapers gently from renal pelvis to bladder, runs a straight course into a compliant non-obstructed bladder and is without reflux or obstruction at the uretero-vesical junction. The surgical goal of megaureter repair is to restore, as closely as possible, that normal anatomy and physiology. This can only be successfully achieved when the underlying conditions of the ureter, bladder or urethra that led to the ureteric dilatation have been identified and treated. This is a very simple but important concept that is sometimes forgotten.

Megaureters may be caused by primary distal ureteric obstruction (obstructive megaureter) or may develop secondary to vesico-ureteric reflux (refluxing megaureter). Gross dilatation of the ureter can also occur unrelated to any *ongoing* reflux or obstruction (non-refluxing, non-obstructed megaureter). Surgical repair is often indicated in cases of refluxing or obstructing megaureter as both conditions can lead to infection and/or renal damage. The non-refluxing, non-obstructing megaureter is usually a stable condition; poor drainage can, however, occur because of decreased peristaltic action with poor coaptation of the ureteric walls. Occasionally, some patients with non-refluxing, non-obstructing megaureter undergo ureteric tailoring to decrease urinary stasis and reduce the risk of infection.

The presence of a refluxing megaureter should alert the surgeon to possible lower tract problems that must be addressed prior to megaureter repair. Accurate preoperative evaluation of lower urinary tract function in these patients is difficult because of the extreme compliance of the upper tracts. Attempted bladder filling during micturating cysto-urethrography or urodynamic investigations results in upper tract filling and an inaccurate measurement of bladder capacity and compliance. The diagnosis of a neurogenic bladder or even obstructing urethral valves may therefore be easily missed.

In the refluxing state, the dilated floppy ureter protects the bladder from pressure work. Repair of the megaureter removes this 'pop-off' mechanism, and the bladder capacity is reduced. If functional or anatomic bladder obstruction persists undetected, the bladder will generate high voiding pressures. This scenario can cause relative obstruction at the uretero-vesical junction or a recurrence of the reflux, thus leading to progressive renal damage from the retrograde transmission of the high voiding pressures. Even after the correction of the anatomic obstruction, any change in lower tract dynamics that occurs with the removal of the 'pop-off' mechanism can lead to reimplantation complications. If the refluxing megaureter exists secondary to posterior urethral valves, valve ablation in the first weeks of life will allow the return of normal bladder dynamics. These children can often be managed without surgical megaureter repair as the reflux resolves spontaneously over time with valve ablation alone.

After lower ureteric tailoring, the capacity of the renal pelvis and upper ureter remains elevated. The upper tract volume drains after voiding, resulting in a persistent post-voiding residual volume. Some patients can be managed with simple double voiding. Other children with bladder abnormalities arising from functional or anatomic obstruction are at high risk of developing urinary retention. Urodynamic evaluation should be performed on suspect patients, and, if necessary, clean intermittent catheterization should be initiated. These children must be followed closely after megaureter repair to avoid recurrence and renal damage.

Operative technique

Hendren's excisional tapering

Megaureter repair by dismembered ureteroplasty has traditionally been carried out via an intravesical approach. The classic excisional tapering has been the favored procedure, as described by Hendren in 1969. The bladder is approached through a modified Pfannenstiel incision (a transverse suprapubic skin incision and a midline vertical incision through the rectus fascia). The bladder is opened anteriorly, avoiding the bladder neck and keeping the bladder dome intact to facilitate superior retraction on the bladder. The placement of a ring retractor with blades in the bladder flattens the trigone and posterior bladder wall, maximizing exposure and access to the intramural ureter.

An 8F feeding tube is placed into the ureter and sewn into position. The bladder mucosa around the ureteric orifice is sharply incised and the ureter mobilized transmurally. The distal end of the ureter is now free, and the dissection is continued extravesically. The ureter is mobilized from the retroperitoneum to the level of the iliac vessels. Care is taken to preserve the ureteric blood supply medially. As the tortuous ureter is mobilized, its redundancy will become apparent, and an appropriate distal segment should be resected. A 12F catheter is inserted into the ureter and straight Allis clamps are applied over the catheter on the mesenteric side of the ureter (Fig. 4.1). The redundant ureter in front of the clamp row is then excised longitudinally. The defect is closed in two running layers of a fine resorbable monofilament suture. A locking stitch is used on the inner layer to facilitate a watertight closure. Interrupted sutures are used distally to allow length adjustment after tunnelling.

The reimplantation of the tapered ureter should take account of three basic principles: (1) a submucosal tunnel of adequate length, (2) good detrusor backing, and (3) secure anchoring of the distal ureter. The bladder mucosa surrounding the original hiatus should be dissected from the detrusor to allow a secure re-approximation of the detrusor. In the Politano–Leadbetter reimplantation technique, a neo-ureterocystotomy is made cephalad and medial to the original hiatus (Fig. 4.2). Alternatively, in the Cohen technique, the original hiatus is reduced in size, and the ureter is advanced across the bladder base (Fig. 4.3). In both techniques, an adequate-length submucosal tunnel is created, and the tapered ureter is brought through the tunnel, care being taken to avoid kinking or twisting it along the course. The end of the ureter can be folded back as a 270 degree nipple as an added anti-reflux measure (Fig. 4.4). The nipple increases the effective tunnel length and secures the configuration of the orifice. The end of the ureter is anchored at the 6 o'clock position with a full-thickness suture into the bladder wall, and the ureteric meatus is then sutured to the bladder mucosa with fine Vicryl sutures. A feeding tube is left in the ureter and placed on drainage to protect the ureteral suture line.

Ureteral plication and folding

Non-excisional tapering of the distal ureter is promoted by some as an alternative to resection. Kalicinski's method (Kalicinski et al 1977) involves folding the redundant ureter. The ureter is mobilized as in the above method. The redundant length and any stenotic segments of the ureter are resected. A catheter is placed in the ureter, and mattress sutures are placed longitudinally along the catheter for a distance of 5–10 cm. The redundant width is then folded over the catheter and secured along its length with a running suture (Fig. 4.5). The reimplantation is carried out as previously described.

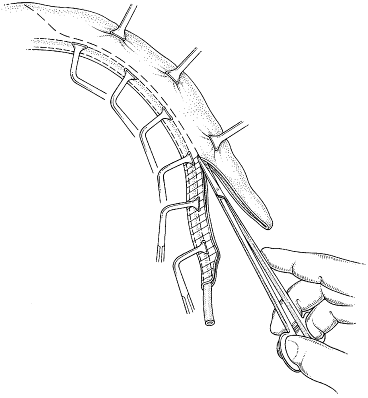

Fig. 4.1

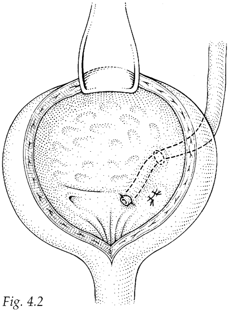

Fig. 4.2

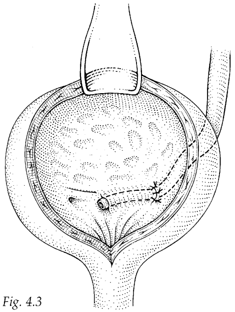

Fig. 4.3

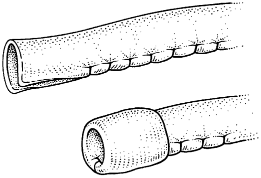

Fig. 4.4

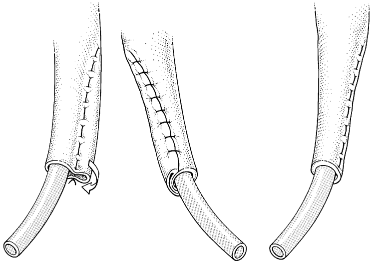

Fig. 4.5

Starr (1979) describes a slightly different method of tapering by plication. Lembert sutures are placed 1 cm apart to fold the ureter on itself over a catheter. The plication suture line is further reinforced with intervening simple sutures (Fig. 4.6).

Extravesical reimplantation

The extravesical approach to ureteric reimplantation has recently been modified for application to the megaureter (McLorie et al 1994). A catheter should be placed into the bladder at the onset of the case to allow adjustment of the bladder volume during the dissection. The catheter can be left in the sterile field or can be connected to a saline bag that can be raised and lowered to adjust the bladder volume.

The ureter is mobilized from the retroperitoneum and transected at the bladder. The distal stump is ligated or oversewn. Tapering of the ureter can be accomplished by excision or folding. The bladder is filled, and a detrusorotomy is made along the path of the proposed submucosal tunnel. Care should be taken to keep the mucosal layer intact. The lateral margins of the detrusor should be dissected from the mucosa to allow a good approximation of the detrusor

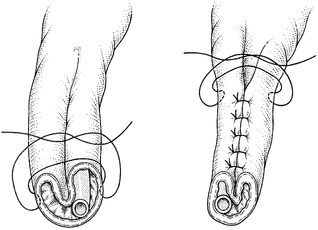

Fig. 4.6

over the ureter. A small opening in the mucosa is made at the distal end of the detrusorotomy. The ureter is anastomosed to the bladder mucosa with interrupted Vicryl sutures. The ureter is then anchored at the 6 o'clock position with a horizontal mattress suture that goes through the bladder, coming out through the small cystotomy, catching the seromuscular layer of the anterior ureteral wall and passing back through the cystostomy lip and up through the full bladder wall again (Fig. 4.7). This stitch pulls the ureter into the bladder and securely anchors it. The detrusor is then closed over the ureter with interrupted sutures to form the tunnel. The ureter is also anchored proximally with a simple interrupted suture as it enters the tunnel.

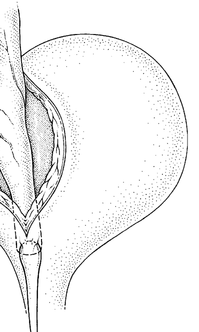

Fig. 4.7

Early and late complications

Historically, the refluxing megaureters have presented the most difficult surgical challenge. Transmural scarring of the distal ureter secondary to associated urinary tract infections puts these ureters at risk of recurrence of reflux after repair. The rigidity of the scarred ureteric wall prevents its collapse within the submucosal tunnel and results in persistent reflux. Additional resistance against reflux may be achieved by nippling the end of the ureter. A recognition and treatment of ongoing bladder outlet obstruction is critical for a successful repair. If reflux recurs or persists, it is feasible to re-do the reimplantation, although the rate of failure is still high as repeat operation further jeopardizes the blood supply to the distal ureter, and chronic bladder wall abnormalities persist as a threat to the reimplantation.

Uretero-vesical junction stenosis occurs as a result of transmural scarring and ischemic insult to the distal ureter. This complication can occur at any time after repair. Meticulous attention at the preservation of the intrinsic blood supply of the ureter is necessary to avoid this complication. It is also important not to narrow the distal ureter too aggressively in the tapering procedure, and to leave the repair stented in the immediate postoperative period. If the obstruction occurs at the ureteric orifice, the distal ureteric tunnel can be unroofed endoscopically. This can be safely done after the antegrade placement of a ureteric wire or catheter to indicate the course of the submucosal ureter. Ureteric stenosis proximal to the ureteric orifice is often amenable to balloon dilatation or endoscopic incision. Open re-operation can be carried out, but again the risk of further insult to the ureter is high.

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Chapter 5

Endoscopic surgery of urethral valves and ureteroceles

David A. Diamond and Alan B. Retik

Endoscopic surgery of posterior urethral valves

The optimum initial surgical approach to posterior urethral valves is endoscopic ablation. Whereas in certain clinical settings other surgical approaches may seem preferable, endoscopic ablation affords a precise surgical solution without the complexity of urinary diversion or the uncertainty of anatomically blind techniques of valve ablation.

Endoscopic valve ablation entails three essential steps – assessment of the urethral caliber, endoscopic delineation of the anatomy, and disruption of the obstructing circular ring of valve tissue.

Assessment of urethral caliber

The majority of newborn urethras will accept a 7.5F or 8F instrument comfortably. A number 10F instrument can be used in older infants and children. The limiting factors anatomically are the caliber of the urethral meatus and the fossa navicularis. In order to determine the maximum capacity of the urethra, pediatric Campbell's sounds are gently passed into the anterior urethra only. Ideally, the urethra is calibrated to one size larger than the instrument to be used. One should never overdilate the urethra.

Delineation of anatomy

The anatomy is usually most easily defined by carefully passing the endoscope under direct vision into the bladder and working from the bladder neck distally. The bladder neck may be exceedingly high, requiring the passage of the endoscope almost directly upwards to enter the bladder. The endoscope is then very gradually withdrawn with the irrigation flowing. Just distal to bladder neck, which appears as a proximal lip posteriorly, one visualizes prominent urethral folds radiating upward from the verumontanum (Fig. 5.1a). Emanating distally and laterally from the lower portion of the verumontanum are the sail-like folds of the type I posterior urethral valves, which join anteriorly at the 12 o'clock position (Fig. 5.1b). These are usually seen most clearly at the 5 and 7 o'clock positions. Distal to the valve leaflets, one may visualize the external sphincter.

Visualizing posterior urethral valves is best done with the bladder full and the end of the cystoscope at the external sphincter. The bladder is pressed by hand from above with a drainage valve open on the cystoscope to create a urinary flow. This fills the valve leaflets, which should snap into an obstructing position. An exception to this approach may be the rarer type III valve presenting as a narrow diaphragm in the posterior urethra. The passage of the endoscope through the aperture in the diaphragm often disrupts this type of valve.

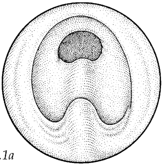
Fig. 5.1a

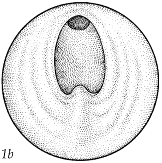
Fig. 5.1b

One should not begin ablation until the anatomy of the valve leaflets is quite clear. In addition, the first observer to perform the endoscopic examination should perform valve fulguration rather than switching off midway during the procedure.

Valve ablation

A variety of transurethral techniques for posterior urethral valve ablation have been described over the years, but the mainstay of management remains direct vision valve ablation with the infant resectoscope loop or endoscopic electrode. The alternative approaches proposed for valve ablation have been designed for settings in which direct endoscopic ablation is impossible because of either the diminutive size of the infant urethra or the lack of availability of miniature endoscopic equipment. Beginning with D.I. Williams' use of the crochet hook, these techniques have included the perineal urethrostomy approach with the electric auroscope (Johnston 1966), Fogarty balloon catheter valve ablation of the valves (Diamond & Ransley 1987) and the Whitaker insulated hook for valve ablation (Whitaker & Sherwood 1986). With the availability of improved endoscopic equipment of diminishing caliber, alternative approaches to valve ablation have rarely become necessary.

The optimal anatomic location on the valve leaflet for ablation has been the subject of some debate. Some have argued that the most significant element of obstruction is the anterior fusion of the valves. Thus, ablation at the 12 o'clock position is critical.

Others argue that valve fulguration at the 5 and 7 o'clock positions is optimal because it is there that the leaflets are best visualized and most safely fulgurated. A superior result with either approach has never been demonstrated. It is our preference to ablate the valve leaflets at the 5 and 7 o'clock positions. Having said this, it is quite likely, as some authors have stated, that effectively ablating the valves in one location will disrupt the ring-like obstruction and be curative.

The infant resectoscope affords a safe and straightforward approach to valve ablation. After examining the bladder and urethra with the cystoscope, the resectoscope is introduced. The resectoscope sheath is somewhat larger than the 10F cystoscope and should be introduced gently after urethral calibration. It should be passed into the anterior urethra only with its obturator, at which point the working element is placed to direct the instrument into the bladder under vision. The visibility with the resectoscope is usually slightly inferior to that with the cystoscope as the flow of irrigation is restricted. Thus, landmarks should be re-established.

One then positions the resectoscope just distal to the valve leaflets. The resectoscope loop is used to hook the leaflets and draw them into the end of the resectoscope sheath. Pure cutting current (set at 20–30 W) is then used to destroy the valve tissue. The leaflets are often most easily visualized at the 5 and 7 o'clock positions with the zero degree lens (Fig. 5.2). The instrument may, however, also be rotated 180 degrees to visualize and ablate the obstructing valve tissue at 12 o'clock. One distinct advantage of the resectoscope loop technique is its ability to distinguish inconsequential leaflets, which do not produce enough resistance to be 'hooked' by the loop. If the fossa navicularis does not accommodate the resectoscope sheath comfortably, one should use the 8.5F or 10F cystoscope and 3F electrode.

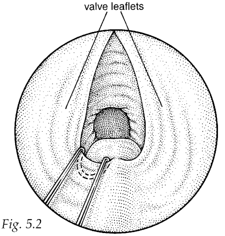

Fig. 5.2

Electrode ablation is performed by engaging the medial edge of the leaflet with the Bugbee electrode, gently pushing toward the bladder and briefly applying current. The cautery current should be set at 25 W on pure cut. By applying the electrode at the 5 and 7 o'clock positions, a segment of the leaflet is ablated, leaving the valve incompetent (Fig. 5.3). The remnant leaflets flutter with the expressed urine flow. Fulguration should be repeated if residual obstructive tissue is noted.

An alternative means of valve ablation is to use a small Bugbee-type electrode made from a 3F ureteric catheter (Retik 1991). This is especially helpful with small infants because it may be passed through or alongside the 8F infant cystoscope and still permit the flow of irrigant during the procedure in order to maintain adequate visualization. The wire stylet is cut distally to protrude from the end of the catheter, and the proximal end is clamped to the cautery electrode. The plastic

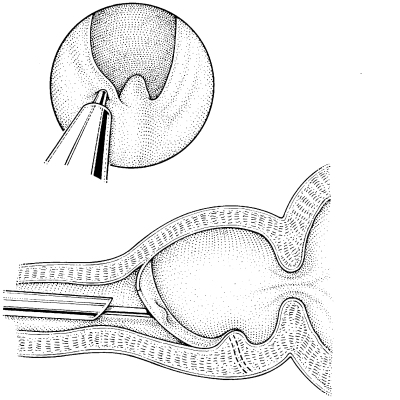

Fig. 5.3a

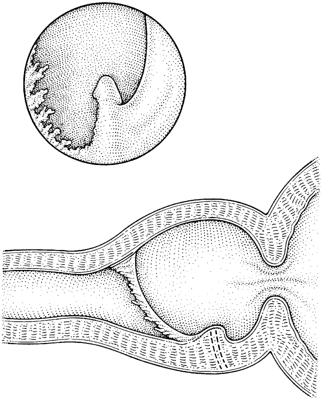

Fig. 5.3b

ureteric catheter itself acts as the insulator. The catheter is advanced through the cystoscope and up to the valve before the wire is advanced. The wire is advanced 2 or 3 mm and pushed just into the valve leaflet. Fulguration is carried out as with the larger Bugbee electrode. Several applications of current in short bursts may be required to ablate the valves effectively. In the process of ablating valve tissue, one should err on fulgurating too little rather than too much, thus avoiding the risk of injury to the prostatic urethra and external sphincter.

Following valve ablation, a small catheter is traditionally left indwelling for 24 hours but is not always necessary.

Transurethral puncture of ureteroceles

The transurethral puncture and decompression of ureteroceles – previously reserved for the emergency drainage of an infected system – has recently achieved credibility as an initial procedure for the elective treatment of an obstructing ureterocele in an infant or child (Blyth et al 1993). We reserve transurethral puncture for ureteroceles (either orthotopic or ectopic) that are associated with a functional renal moiety or obstructing the normal contralateral collecting system.

The transurethral puncture of a ureterocele is performed with a 3F cauterizing electrode, either a Bugbee or the wire stylet of a 3F whistle-tip ureteric catheter. Either may be passed through the working channel of a 7.5F–10F panendoscope.

One should first clearly define the anatomic relationship of the ureterocele to the bladder neck and ureteric orifices. The anatomy of the orthotopic ureterocele is quite straightforward. The ureterocele will be entirely intravesical and typically lie well away from the contralateral ureteric orifice (Fig. 5.4a). An ectopic ureterocele is likely to be more complex anatomically. It may extend to or beyond the bladder neck and may extend laterally across the trigone to abut the contralateral ureteric orifice (Fig. 5.4b). The orifice of the ipsilateral lower pole moiety tends to lie on the proximal aspect of the ureterocele, often difficult to visualize endoscopically. For both orthotopic and ectopic ureteroceles, the goal is to effectively decompress the hydronephrotic system while attempting to preserve whatever flap-valve mechanism may exist, thus minimizing the likelihood of reflux into the ureterocele.

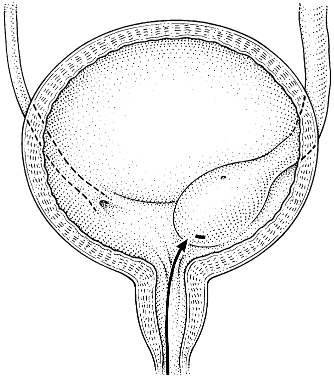

Fig. 5.4a

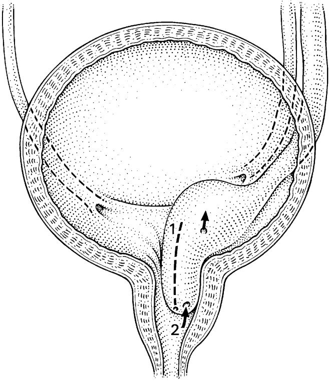

Fig. 5.4b

The management of the orthotopic ureterocele is straightforward. One makes a puncture using a pure cutting current at the distal aspect of the ureterocele just above its junction with bladder (Fig. 5.5). As the Bugbee electrode is passed through the ureterocele wall, it may be moved slightly laterally in either direction to enlarge the opening. One can readily see the ureterocele deflate. In some cases, manual compression of the flank over the hydronephrotic moiety is helpful in distending the ureterocele prior to puncture, and this may demonstrate a jet of urine following decompression.

For the ectopic ureterocele, a similar technique is used as the puncture is made at the base of the ureterocele at its junction with bladder wall in a clearly intravesical location. Because the ureterocele may extend distally beyond bladder neck, the location of

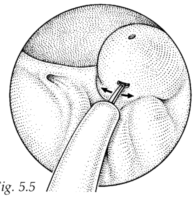

Fig. 5.5

the incision must be very clearly visualized. On occasion, a ceco-ureteroecele will require an additional puncture distal to the bladder neck to provide adequate drainage and avoid an obstructing lip. This should if possible be avoided as the risk of subsequent urinary incontinence, particularly in the female, is significant.

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Chapter 6

Temporary urinary diversion (ureterostomy and vesicostomy)

Harry P. Koo and David A. Bloom

Introduction

Pediatric urinary diversion has undergone appreciable change in recent years. In the past, supravesical diversion was commonly used in a variety of chronic conditions. Today, the number of patients requiring supravesical diversion has declined significantly because of percutaneous nephrostomy, cutaneous vesicostomy and an improved understanding of urinary tract dysfunction. Nonetheless, in selected patients, non-intubated temporary urinary diversion remains a useful temporizing therapeutic strategy.

Temporary urinary diversion in infants and children may be intubated or non-intubated, the choice depending on the pathology involved and on the expected duration of the required drainage. Intubated diversion is usually reserved for patients requiring only a brief period of diversion. Non-intubated diversion is performed for patients requiring lengthy diversion and is associated with lower rates of infection and stone formation than are seen with intubated diversions.

Ureterostomy

Indications for ureterostomy are now few. Percutaneous nephrostomy offers a good alternative when weeks or months of diversion are necessary. For longer periods, cutaneous ureterostomy remains a viable option. Cutaneous ureterostomy is advisable only with ureters that are substantially and chronically dilated. The mobilization of ureters of normal caliber to skin level risks ischemic necrosis, this also being the case with acute dilatation, in which the vascularity tends to be attenuated. Ureterostomies should be fashioned proximally or distally. Exteriorization of the mid-ureter is avoided because the blood supply is relatively tenuous and the options for subsequent reconstruction are limited.

Proximal ureterostomy

Proximal ureterostomies for temporary diversion maximize the free drainage of massively dilated, atonic upper renal tracts. They are constructed as loop, Sober or ring ureterostomies. The advantage claimed for the Sober or ring techniques is that when the ureterostomies are formed bilaterally, the bladder is not totally defunctionalized. A properly performed proximal ureterostomy does not sacrifice ureteric length or disrupt the periureteric vasculature, and it permits renal biopsy at the time of diversion.

Surgical technique

The child is placed in a modified flank position. A transverse subcostal incision is made so that the stoma can be incorporated into the wound near the mid-axillary line. The abdominal muscles are divided in the line of the skin incision. To expose the ureter, a plane is developed between the peritoneum and the retroperitoneum. A markedly dilated ureter that adheres to the peritoneum may resemble an infant's small intestine. The ureter is usually tortuous, and the division of the flimsy avascular adhesions uniting adjacent loops enables a tension-free delivery to skin level without distal dissection.

For a loop ureterostomy, the lumen is opened by a longitudinal incision, and the muscle layers are loosely approximated beneath the loop of the ureter (Fig. 6.1). The stoma is matured by approximating the edges of the ureterotomy to the skin using fine interrupted synthetic absorbable sutures; the skin on each side of the stoma is closed with interrupted absorbable sutures (Fig. 6.2).

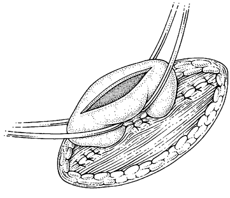

Fig. 6.1

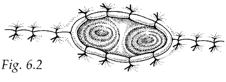

Fig. 6.2

To construct a Sober ureterostomy, the ureteric loop is divided after mobilization at a level at which the proximal limb lies comfortably at skin level. The distal limb is anastomosed to a corresponding aperture in the distal renal pelvis or proximal ureter. The proximal limb is sutured to the skin as a stoma (Fig. 6.3). In the reversed Sober procedure, the distal limb of the ureteric loop forms the stoma while the proximal limb is anastomosed to it in an 'en-Y' formation. This formation theoretically enhances onward urinary drainage yet still adequately decompresses the upper urinary tract.

For a ring ureterostomy, both ureters are longitudinally incised toward the base of the loop and are then anastomosed side-to-side (Fig. 6.4). The skin stoma is formed as for a loop ureterostomy. With this formation, the ureter is not divided, thus lessening the risk of ischemic necrosis.

The patency of the stoma and proximal ureter is then reassessed using a catheter. Stenting of the ureter is, however, generally unnecessary. Drainage of the anastomosis is provided externally via a 1/4 inch Penrose tube or internally via a percutaneous 5F infant feeding tube, entering the ureter distal to the suture line and running upwards to the renal pelvis.

Distal ureterostomy

Distal ureterostomies for longer-term diversion are constructed as terminal stomas by detachment of the ureter from the bladder. The level and bilaterality of a cutaneous ureterostomy is of little concern in infants as the urine drains into the diaper. In older children, however, a distal ureterostomy is preferred to facilitate the application and maintenance of a urinary appliance. For bilateral distal ureterostomies, both ureters may be diverted to a single site, either by transuretero-ureterostomy or via a double-barreled stoma.

Surgical technique

The patient is placed in the supine position, and the bladder is decompressed with a urethral catheter to facilitate pelvic exposure. A transverse lower abdominal incision is made, and the ureter is approached extraperitoneally. The ureter is mobilized as far distally as possible, taking care to avoid any disruption of the overlying gonadal vessels and of vessels that feed the uterus or vas deferens. The ureter is transected

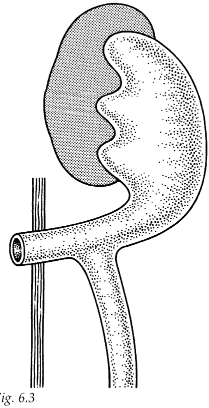

Fig. 6.3

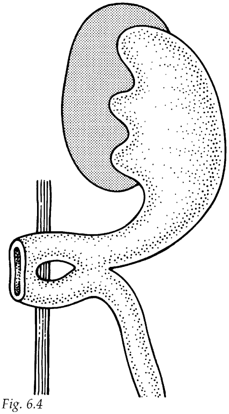

Fig. 6.4

adjacent to the bladder, and the stump is ligated with an absorbable suture if there is vesico-ureteric reflux. The stump is left open to drain if there is an obstructive anomaly.

The proximal ureter is brought through the lateral aspect of the incision and anchored to the fascia of the external oblique muscle with interrupted synthetic absorbable sutures. To reduce the risk of stomal stenosis, a V-shaped flap is fashioned from the edge of the skin incision to be incorporated into the spatulated ureter. The edges of the ureter are approximated to the skin using interrupted 5/0 synthetic absorbable sutures (Figs 6.5–6.7). Stenting of the ureter is not necessary.

When a bilateral cutaneous ureterostomy is warranted as a temporizing measure, a transuretero-ureterostomy with a single stoma is a useful technique. The more dilated ureter is brought to the skin, the contralateral ureter being drawn through the retroperitoneal space and anastomosed in an end-to-side fashion (Figs 6.8 and 6.9).

Closure of distal ureterostomy

The take-down of an end-cutaneous ureterostomy may necessitate the reconstruction of the distal ureter. A catheter is introduced into the ureter and sutured to the skin. A standard Pfannenstiel incision is made that incorporates the original incision. The ureter is freed from the abdominal wall and mobilized proximally, and the stoma and adjacent fibrotic ureter are excised.

Reconstruction may involve transuretero-ureterostomy, a uretero-neocystostomy with or without a psoas hitch, or reimplantation into an intestinal reservoir. Ureteric tapering may be required.

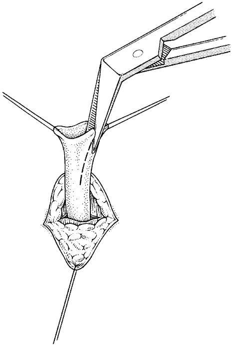

Fig. 6.5

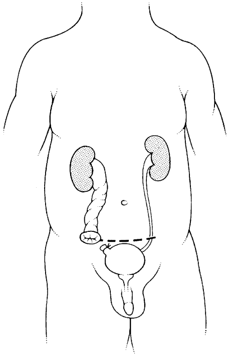

Fig. 6.8

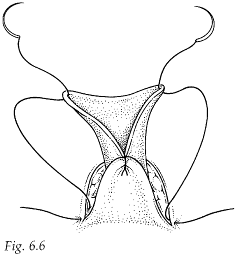

Fig. 6.6

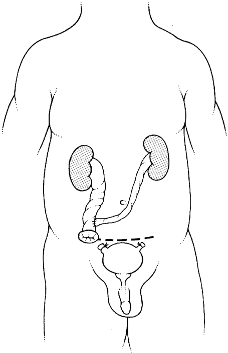

Fig. 6.9

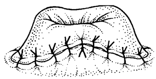

Fig. 6.7

Complications

Complications of a cutaneous ureterostomy are more common in patients with normally sized ureters or acutely dilated ureters that do not have the rich blood supply and redundancy of a chronically dilated ureter. In well-selected patients, complications of loop and ring ureterostomies are rare. Ischemic necrosis of the stomal limb may complicate Sober procedures and if, extensive, necessitate the abandonment of the ureterostomy and the establishment of nephrostomy drainage. Ureteric ischemia and stomal stenosis is the principal complication of distal ureterostomy. For minor degrees of stenosis, intubation or stomal revision can be performed. If extensive necrosis occurs, an alternative form of urinary diversion is sometimes necessary.

Vesicostomy

Vesicostomy in children is an effective form of a temporary diversion for a variety of conditions in which drainage at bladder level effectively decompresses the entire urinary tract. Vesicostomy may be considered for ill infants in whom reconstruction must be delayed and in neonates with posterior urethral valves, prune belly syndrome or neurogenic bladder associated with myelodysplasia or severe vesico-ureteric reflux.

Surgical technique

With the child in a supine position, a 2.5 cm midline transverse incision is made half way between the lower edge of umbilicus and the upper edge of the symphysis pubis, exposing the rectus fascia (Fig. 6.10). A transverse rectus fascial incision is made, exposing the rectus muscles, which are retracted laterally. The inferior fascial edge is incised vertically for 1 cm and the edges excised, leaving a triangularly shaped fascial defect (Fig. 6.11).

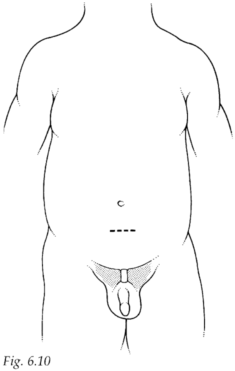

Fig. 6.10

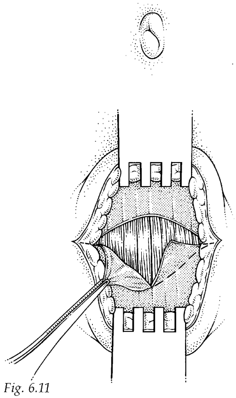

Fig. 6.11

The bladder is filled via a urethral catheter, and the perivesical fascia is incised to expose the bladder. The peritoneum is reflected superiorly with blunt dissection. Progressive traction sutures are placed on the anterior wall of the bladder for downward traction (Fig. 6.12a) until the obliterated umbilical arteries and urachus are identified (Fig. 6.12b).

The bladder is opened by excising the urachus (Fig. 6.13). The detrusor edge is anchored to the triangularly shaped fascia with interrupted 3/0 synthetic absorbable sutures (Fig. 6.14). The lateral fascial defects are closed around the bladder to achieve a 24F lumen at the internal stoma. Too wide a stoma will

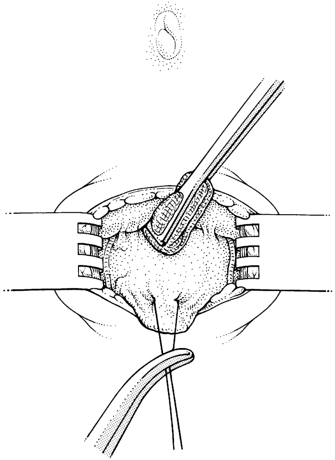

Fig. 6.12b

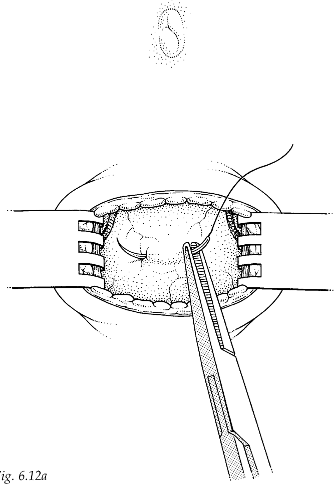

Fig. 6.12a

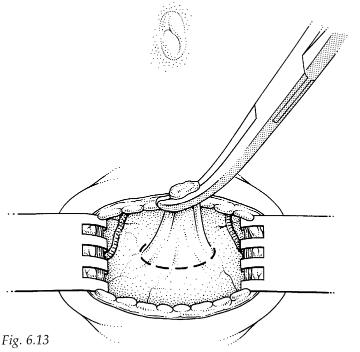

Fig. 6.13

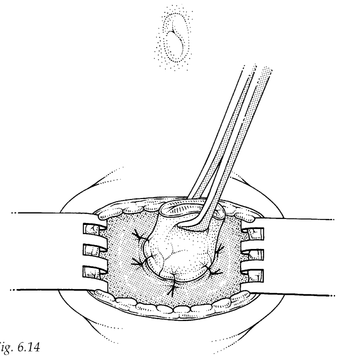

Fig. 6.14

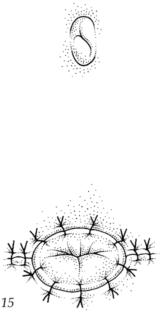

Fig. 6.15

permit prolapse. The cutaneous stoma is created by suturing the full thickness of the bladder to the subcuticular layer with 5/0 synthetic absorbable sutures (Fig. 6.15). The final stoma is carefully calibrated to 24F. Postoperative edema will temporarily narrow the stoma, so a catheter is left in place for 48 hours. The stoma is dilated with a soft eye-dropper at home for a few months.

Closure of vesicostomy

The vesical epithelial tract is dissected free from skin and fascia, whereupon it is excised. The bladder is closed in two layers with synthetic absorbable sutures. The fascia and skin are approximated, and the bladder is drained with a urethral catheter. In infants with posterior urethral valves, the valve ablation should be performed at the time of vesicostomy closure to allow urine to flow through the urethra.

Complications

The main complications of cutaneous vesicostomy are stomal stenosis and bladder prolapse. By a careful adherence to the technique, and by understanding the variations in bladder pathology, the rate of complications can be significantly reduced.

The most important considerations in the technical success of a temporary cutaneous vesicostomy in children are the following:

1. The urachal portion of the bladder should be identified as the site for the stoma.
2. A flush stoma of an appropriate caliber should be created.
3. In children with posterior urethral valves and myelodysplasia, the bladders are thick walled and unlikely to prolapse with increased abdominal pressure, so a 22F–24F caliber is optimal.
4. In prune belly syndrome, the stoma should be made larger (28F–30F) as stenosis is more likely.
5. In severe vesico-ureteric reflux, the bladder is of normal thickness and very likely to lead to prolapse, so an 18F–20F caliber is preferable.

Further Reading

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Augmentation and substitution cystoplasty

A.R. Mundy and Jeremy G. Noble

Introduction

There are two types of cystoplasty: augmentation cystoplasty in which the bladder is enlarged, and substitution cystoplasty in which it is replaced. These are both uncommon procedures outside specialized units and are usually used only as a last resort for an otherwise intractable lower urinary tract problem for which the only alternative is some form of urinary diversion. The most common indication in pediatric practice for both procedures is detrusor dysfunction caused by spina bifida, and the most common problem after both is voiding imbalance, often giving exactly the same symptoms as the patient had beforehand. For this reason, it is easy to ascribe a poor result to refractory disease rather than to poor selection or inadequate surgery, but it is in fact poor selection of the patient and inadequate surgery, resulting from a failure to understand the principles of cystoplasty, that are responsible for most poor results.

Indications

The indications for both procedures in pediatric practice are similar, the main difference being one of degree. The usual indications are detrusor dysfunction, the small contracted bladder and, for substitution cystoplasty, selected cases of lower urinary tract malignancy.

Detrusor dysfunction

There are three main types of detrusor dysfunction for which cystoplasty may be appropriate: refractory detrusor instability, detrusor hyperreflexia and poor compliance.

Refractory detrusor instability and detrusor hyperreflexia are here taken to mean the occurrence of involuntary detrusor contractions on urodynamic testing that have not responded to the full range of conservative treatment (Mundy 1985 a,b). 'Instability' is the term used in the absence of an overt neuropathy; 'hyperreflexia' is used when the child has an overt neuropathy to account for the vesico-urethral dysfunction.

Poor compliance may be seen on urodynamic testing in children with small-capacity bladders whatever the cause, in some children with detrusor instability as a consequence of outflow obstruction (past or present), particularly in relation to posterior urethral valves, and in children with neuropathic dysfunction (Mundy 1985c).

Augmentation cystoplasty is appropriate for the patient with poor bladder compliance in the absence of neuropathy (Mundy & Stephenson 1985), for refractory detrusor instability (Mundy 1985 a, b, Mundy & Stephenson 1985) and for minor degrees of neuropathic dysfunction showing poor compliance, detrusor hyperreflexia or both, usually when the thickness of the bladder wall is relatively normal. Substitution cystoplasty is indicated when poor compliance is associated with gross thickening of the bladder wall and a marked reduction in capacity, as well as in severe degrees of neuropathic dysfunction (Stephenson & Mundy 1985).

In children without neuropathic dysfunction, age is obviously an important factor when considering surgical intervention, common sense dictating that cystoplasty should be considered only in teenagers with intractable symptoms who are past the age at which a natural spontaneous resolution of their symptoms could be expected. The most common 'non-neuropathic' group comprises children with a bladder dysfunction that looks 'neuropathic' on investigation of the lower urinary tract but shows no clinical evidence of an overt neuropathy, particularly when the lower tract abnormality appears to be causing upper tract complications.

The small contracted bladder

In pediatric practice, this is usually related to detrusor dysfunction, but one occasionally sees children with small contracted bladders for no apparent reason or following some previous surgical misadventure.

Other indications

These include conditions such as previously inadequately treated lower urinary tract malignancy, usually rhabdomyosarcoma, bladder exstrophy when there is insufficient bladder tissue for a simple closure, and other situations that have left the bladder in a similar state. The latter group – of children with a whole variety of underlying conditions in whom a small capacity bladder is the common denominator – is collectively the largest group in this category and overall the second largest group of patients undergoing cystoplasty.

In practice, by far the largest group of children to be considered for cystoplasty comprises those with congenital cord lesions, particularly spina bifida, and here it is important to remember that, in addition to the bladder dysfunction, there is almost invariably a degree of sphincter dysfunction that will also need attention. The decision of which type of cystoplasty should be performed depends on the bladder capacity, the thickness of the bladder wall and the presence or absence of vesico-ureteric junction obstruction as a consequence of the bladder wall abnormality. Those children with a reasonable bladder capacity, a relatively normal bladder wall thickness and no upper tract abnormalities (the vast majority) will be treated by augmentation cystoplasty, and those with small, thick-walled bladder and vesico-ureteric junction obstruction will require substitution.

In adults, the situation is entirely different. Augmentation cystoplasty for detrusor instability or hyperreflexia is still fairly common, but substitution cystoplasty is more common, and becoming increasingly so, as an alternative to ileal conduit urinary diversion after cystectomy for bladder cancer.

Assessment with a view to cystoplasty

The minimum requirements for a typical patient are an intravenous urogram or a high-quality ultrasound scan of the upper urinary tract and a video urodynamic study (VUD). A cysto-urethroscopy is often helpful, particularly in those who have had previous surgery, when it should be regarded as mandatory.

The VUD provides the urodynamic diagnosis and an assessment of its severity, whereas the intravenous urogran or ultrasound scan shows whether there are secondary upper tract changes. If there are, and these are not caused by reflux (which will be shown on the VUD), a MAG 3 renal scan will show whether or not there is obstruction. A repeat scan after a week or so of indwelling catheterization will disclose whether this is caused by outflow obstruction (which is usually the case), in which case the upper tract changes will resolve with catheterization, or by obstruction of the intramural ureters as they pass through a thickened and fibrotic bladder wall (which is rare), in which case the upper tract obstruction will persist.

Cysto-urethroscopy allows the measurement of bladder capacity, the exclusion of or need for treatment of outflow obstruction and a further assessment of any intravesical pathology in patients with anything other than neuropathic dysfunction (Mundy & Stephenson 1984, Mundy 1985a), the severity of which may sway the surgeon towards substitution rather than augmentation cystoplasty.

Problems associated with cystoplasty

Apart from the usual complications of any major operation, there are three particular problems associated with cystoplasty that must be anticipated (Turner-Warwick 1976). The first two are voiding imbalance and diverticulization of the cystoplasty segments. The choice of procedure, the operative technique and the further management are all influenced by these two factors. The third problem associated with cystoplasty results from the presence of an intestinal segment within the urinary tract, this being discussed at the end of the chapter.

Voiding imbalance

This is almost universal after either type of cystoplasty if bladder neck compliance is unimpaired. It is less common in girls than in boys because bladder neck incompetence is a common incidental abnormality in girls even in the absence of neuropathy. In neuropathy, bladder neck incompetence is very common.

Whether or not the bladder neck is incompetent, there is in both sexes a balance to be considered between the effectiveness of bladder emptying and the resistance to bladder emptying afforded by the sphincter mechanism. The effectiveness of bladder emptying is the sum of its contractile force and the ability (if any) to empty by straining. Sphincteric resistance to emptying may be active, and in neuropathy the active component may be enhanced, or simply the passive resistance of a bladder outflow may be devoid of any contractile activity.

As the specific intention of a cystoplasty is to make involuntary detrusor contractions ineffectual and thus stop them leading to involuntary voiding, an inefficiency of voluntary voiding should be regarded as an inevitable consequence of the procedure rather than as a true complication, a term implying that it is not routinely to be expected.

In the past, various sphincter-balancing procedures were employed to make voiding more efficient after cystoplasty as and when the problem presented itself (Turner-Warwick 1976). Nowadays, clean intermittent self-catheterization (CISC) has almost entirely replaced sphincter-balancing procedures, and such is the frequency of post-cystoplasty voiding dysfunction that it is wise to anticipate the problem and ensure that the child (or carer) is willing to perform CISC before the operation is undertaken. Indeed, the author would regard it as an essential part of the preoperative preparation and would not usually proceed with the cystoplasty if the child or carer had not been shown to be both willing and able to perform CISC beforehand.

Diverticulization of cystoplasty segment

This tends to occur after both types of cystoplasty unless steps are taken to prevent it. In both instances, it is usually caused by a contraction of the suture line by which the cystoplasty segment is sewn in place, such that an 'hour-glass' deformity develops. As described below, this is usually overcome by an almost complete bisection of the bladder before inlaying the cystoplasty segment in the augmentation procedure (Mundy & Stephenson 1985) and by making sure not to leave any more of the bladder than the trigone and bladder neck in substitution cystoplasty (Turner-Warwick 1976, Mundy 1986).

Preoperative preparation

Patients for either procedure are admitted to hospital 2 days before operation. The preoperative regimen is the same for both procedures and consists of a gentle mechanical bowel preparation and prophylactic antibiotics.

1. *Day 2 preoperatively*:
   Low-residue diet
   Oral magnesium sulphate 5–10 ml three times daily

2. *Preoperative day*:
   Clear fluids only
   Oral magnesium sulphate 5–10 ml three times per day
   Enema if constipated; rectal wash-out otherwise

3. *Day of operation*:
   Rectal wash-out
   Parenteral gentamycin, ampicillin and metronidazole at the induction of anesthetic and for at least three doses postoperatively up to a maximum of 5 days of treatment.

There is sometimes, particularly in children with impaired renal function, a tendency to dehydration preoperatively, so intravenous hydration will be necessary to prevent this.

Augmentation cystoplasty

The technique described here is the one that the author uses routinely and almost exclusively using ileum (Bramble 1982, Mundy 1985 a, b, Mundy & Stephenson 1985). In many children, it is easier to use the sigmoid colon to get a patch down into the pelvis on a vascular pedicle. In young children, this is usually safe, but in adolescents, as in adults, colonic anastomoses are more prone to complications than ileal anastomoses, which, coupled with the possible higher risk of a carcinoma developing in colon than in ileum when it is in contact with urine over the long term (Leadbetter et al 1979, Nurse & Mundy 1989a), makes an ileocystoplasty preferable.

Occasionally, for various reasons, there is a deficiency of usable length of small or large intestine so stomach must be used. Gastrocystoplasty is quite commonly used in various pediatric centers on a more routine basis for reasons based on a hypothetical metabolic argument that the author finds specious. It is an unnecessarily complicated procedure with an unnecessarily high incidence of postoperative dysuria, which makes it less suitable when ileum or sigmoid colon is freely available.

Technique

A midline, lower abdominal or Pfannenstiel incision may be used, but the author prefers a Cherney incision (Cherney 1941) as this provides a wider and easier access to the pelvis than a Pfannenstiel incision but is a more cosmetic alternative to the lower midline approach.

A curved skin incision, as in the Pfannenstiel approach, is made deeply and down to the rectus sheath, which is then similarly incised to expose the pyramidalis muscles and the underlying recti. The pyramidales are then reflected off the recti with the distal part of the rectus sheath down to the anterior aspect of the pubis, exposing the rectus tendons as they insert into the pubis. The rectus tendons are then divided (indicated by the dotted line in Fig. 7.1), leaving just enough inferiorly for later closure, and the lateral fascial attachments of the recti to the deep aspect of the inguinal ligaments are then divided to allow

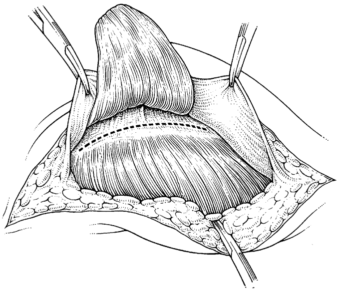

Fig. 7.1

the recti to be lifted up (Fig. 7.2). A ring retractor can then be placed to hold the incision open.

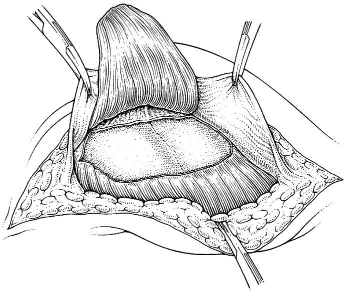

Fig. 7.2

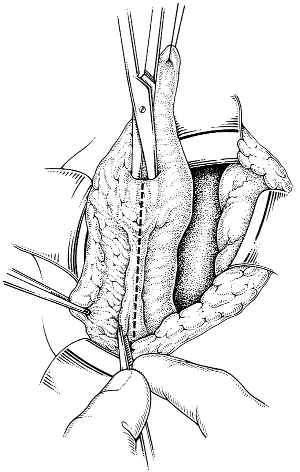

Fig. 7.3

The retropubic space is widely opened to expose the pelvic floor all the way round the front and both lateral aspects of the bladder. The urachus is defined as it leaves the dome of the bladder, ligated and divided. The peritoneum just deep to this point is then opened to expose the dome of the bladder with its peritoneal covering.

Traction on the ligature that secures the urachus at the dome of the bladder and blunt dissection on either side will open up the plane of cleavage between the fascia on the lateral aspect of the bladder wall (anterior to the dotted line in Fig. 7.3) and the fascia and overlying peritoneum on the dome of the bladder (posterior to the dotted line in Fig. 7.3), where these two fascial layers join to run together as a single sheet to the brim of the pelvis along the line of the iliac vessels. The development of this plane of cleavage down along the lateral margin of the bladder to the ureter and superior vesical pedicle on each side exposes the maximal circumference of the bladder and allows the large veins that sometimes run in the fascia on the lateral wall to be secured without much bleeding. This separation of the fascial planes before dividing the bladder wall may be regarded by some as rather finicky and unnecessary. Such surgeons simply divide the bladder wall around its maximal circumference and accept whatever bleeding occurs.

When the maximal circumference of the bladder has been exposed, the bladder wall can be incised around its maximal circumference from a point 2 cm or so in front of the ureteric orifice and about 1 cm from the bladder neck on one side to a similar point on the other side. This is most easily achieved by starting on the dome and working down one side at a time (Fig. 7.4) using a diathermy point. (Ureteric catheters are shown in these figures for orientation; they are also necessary in practice.) In this way, the bladder is completely bisected except for a bridge of about 1 cm on either side of the bladder neck region (Fig. 7.5).

In Fig. 7.5, the circumference of the bisected bladder is being measured with a length of tubing. The cut edge of the bladder is then measured (which should be the same on both sides except that the bladder wall tends to contract down asymmetrically during the course of the procedure), and hemostasis is secured, particularly at the two ends of the incision in the bladder wall, where a few significant vessels are likely to be encountered.

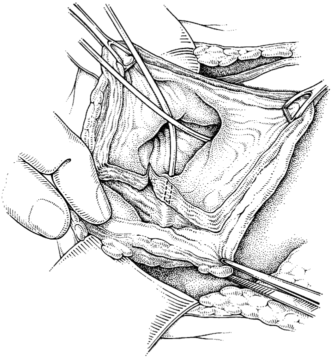

Fig. 7.4

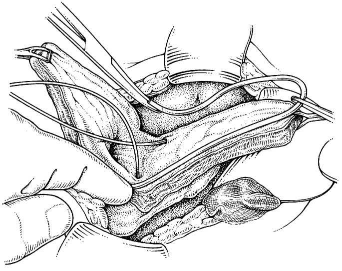

Fig. 7.5

The next stage is to define and isolate a section of ileum on its vascular pedicle (Fig. 7.6). A convenient section of terminal ileum is selected about 25 cm or so proximal to the ileocecal valve in order not to interfere with that part of the ileum with the most important absorptive function. A convenient section of the terminal ileum is selected so that it will drop down easily into the pelvis. It must be equal in length to the measured maximal circumference and have a well-defined vascular arcade supplying it. The ileum on either side is reconstituted in the usual way.

Sometimes, although the mid-section of the ileal segment will reach easily to the bisected bladder, the natural upward curve of the two ends of the segment prevents them reaching down to the two ends of the bladder bisection without tension. In this situation, the mesenteric vessels and the proximal arcades at each side of the isolated segment must be ligated and divided in such a way as to allow the two ends of the ileum to drop down sufficiently. The ileal segment is then opened on its anti-mesenteric aspect to produce a patch (Fig. 7.7).

The ileal patch is now inset into the bisected bladder and sewn in place. There is a tendency for the margins of the bladder to contract during the course of the procedure, producing an overlap between the ileal edge and the bladder edge during the anastomosis of the patch. To prevent this overlap developing, each suture line is halved and then quartered with stay sutures, each quarter then being sewn up individually. The posterior suture line is dealt with first, from inside the bladder, using a continuous 3/0 Vicryl stitch picking up the full thickness of the bladder and ileal walls, starting at the apex of the bladder incision in front of the ureter and working up on

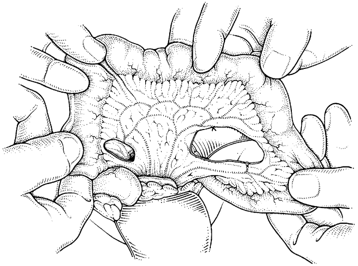

Fig. 7.6

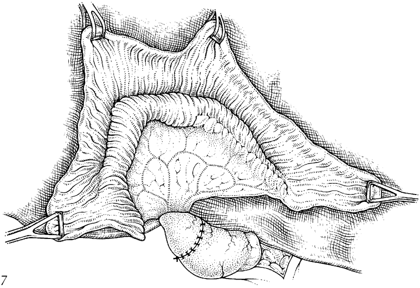

Fig. 7.7

each side to the dome, locking to each of the 'quartering' stay stitches in turn as they are encountered. Figure 7.8 shows stay stitches at each end of the bladder (A), at the halfway mark (B) and halfway between on each side (C); the second quarter of the left side of the posterior suture line is nearing completion.

When the posterior half of the anastomosis has been completed (Fig. 7.9), the ileal patch is flipped over and the procedure repeated, this time from outside the bladder, to sew the anterior edge of the patch to the margin of the anterior half of the bladder with stay sutures as before (Fig. 7.10). At the end of the procedure, a wound drain is left into the retropubic space and a suprapubic catheter that is large enough not to be blocked by the ileal mucus in the urine is left in the bladder, this being brought out through the bladder wall rather than through the suture line between the bladder and the ileum. Finally, the omentum is brought down to cover the suture line.

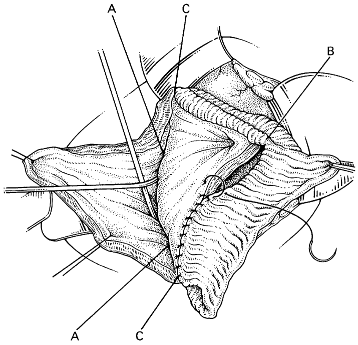

Fig. 7.8

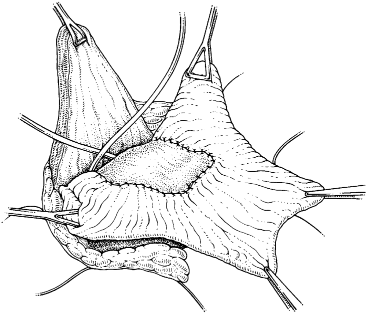

Fig. 7.9

Postoperative management

The wound drain is removed when it stops draining, and the suprapubic catheter is clamped on about the eighth postoperative day, by which time the child should normally be active. The parenteral antibiotic regimen is continued for as long as is felt necessary (usually about 3–5 days) and then changed for a low-dose prophylactic antibiotic such as trimethoprim, assuming that the urine is sterile at that time.

When the suprapubic catheter is first clamped and the child starts to void spontaneously, a voided volume chart should be kept, noting the time and volume passed and, on two or three occasions, the residual urine volume. Assuming that the child is voiding satisfactorily, the catheter is removed after 24 hours, and the patient is discharged home the next day, by which time the suprapubic drain site should have closed. Any excessive urine leakage from this site is dealt with by 24 hours of indwelling urethral catheterization. Children being managed by intermittent self-catheterization may need to keep an indwelling urethral catheter for a day or so after the removal of the suprapubic catheter in order to allow the suprapubic site to heal completely before changing to intermittent catheterization.

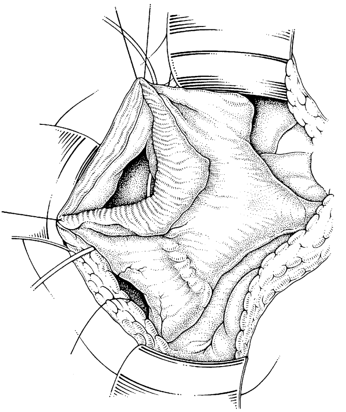

Fig. 7.10

Three months or thereabouts postoperatively, the child should be reassessed by VUD or more simply by pre- and post-voiding ultrasound scans to look for any voiding imbalance. If an imbalance is present but not causing any symptoms or other problems (such as recurrent urinary tract infection), it can probably just be ignored. If, on the other hand, the child is suffering undue frequency or recurrent urinary infection, CISC should be instituted. If the child is already on CISC, the technique clearly needs attention.

Substitution cystoplasty

Substitution cystoplasty is used much less commonly but for a wider range of indications than augmentation cystoplasty in children. In some instances, the child will have incompetence of the bladder neck and urethral sphincter mechanisms in addition to the bladder problem, particularly when this is the result of a congenital anomaly or previous surgery.

In the absence of such sphincteric incompetence, bowel contractility is rarely troublesome and may in fact be useful as it facilitates spontaneous voiding. In the presence of sphincteric incompetence, particularly in neuropathy, bowel contractility may be troublesome and cause incontinence so this must be eliminated as far as is possible. This is achieved by the procedure of so-called 'de-tubularization'.

In the presence of a normally innervated and normally functioning sphincter mechanism, an unmodified ('straight') gut segment may be used for bladder substitution. Otherwise, the gut segment is 'de-tubularized' to form a pouch, recognizing that, with a pouch, CISC will be required much more commonly to achieve complete bladder emptying. It should be pointed out that this view is controversial. Many authors feel that a substitution cystoplasty should always be de-tubularized, although the evidence for this is scant except in neuropathy, in which case the authors would, in any case, agree (see below for a further discussion).

The procedure described here first is a 'straight substitution' as all but the final stages are the same as for a de-tubularized cystoplasty. The final staging for a de-tubularized cystoplasty will be addressed later in the chapter.

A straight substitution cystoplasty is often called a ceco-cystoplasty. Indeed, many surgeons use just the cecum for this purpose. The point of a straight cystoplasty, or indeed any cystoplasty, is to use a segment of adequate capacity so that contractile activity is largely irrelevant. Thus, this author uses the whole of the right colon and the proximal half of the transverse colon (as well as the last few centimetres of the terminal ileum), so the procedure is more accurately, albeit more clumsily, termed an ileoceco-colo-cystoplasty.

A long midline abdominal incision is used. During the mobilization of the right side of the colon, the middle colic artery will need to be clearly exposed, so the upper end of the incision will need to be high enough to allow this. A point half way between the umbilicus and the xiphisternum, or a little above this, is usually sufficient.

The incision is deepened down to the peritoneum, exposing the urachus and the obliterated umbilical ligaments, which are ligated together just below the umbilicus, where they lie side by side. Above this point, the peritoneum is opened in the midline. Below this, the peritoneum and the underlying fascia are incised just lateral to the obliterated umbilical artery on each side down as far as the point at which the artery is crossed by the vas or the round ligaments. The peritoneal incision is then extended across from one side to the other along the line of the posterior margin of the bladder. This part of the incision is then deepened to separate the posterior bladder wall, between two lateral pedicles, from the seminal vesicles in the male and the anterior vaginal wall in the female.

The retropubic space is then widely opened all the way round the front and including both anterolateral aspects of the bladder, and down to the pelvic floor to expose the anterolateral aspects of the lateral pedicles of the bladder, which, having already had their posteromedial aspects exposed by the procedure described in the previous paragraph, are thereby clearly defined.

The first stage of the procedure in some circumstances, but not often in pediatric practice, is to perform a subtotal cystectomy, starting with the ligation and division of the lateral pedicles. There are four parts of the lateral pedicle, which are, from above down, the obliterated umbilical artery, the rest of the superior vesical pedicle, the ureter and the upper part of the inferior vesical pedicle, which sweeps up from below the ureter on to the lateral wall of the bladder. These parts should be ligated and divided individually.

The aim of the subtotal cystectomy is to excise all of the bladder except the subureteric part of the trigone. Many surgeons like to leave the ureters in place to avoid the need for subsequent ureteric reimplantation into the cystoplasty, but this almost invariably leaves more of the bladder behind than one might wish, with a tendency to diverticularization, and in any case interference with vesico-ureteric function by dissecting close to them usually causes bilateral reflux.

Having ligated the 4 components of the lateral pedicle as described above (Fig. 7.11), this is best achieved by first splitting the bladder open in the sagittal plane with diathermy (to reduce bleeding) from the bladder neck anteriorly to the interureteric bar posteriorly. The bladder neck and trigone are then clearly exposed on the inside of the bladder, and each half of the bladder neck and trigone is excised along the margins of the trigone (dotted line Fig. 7.12), again with

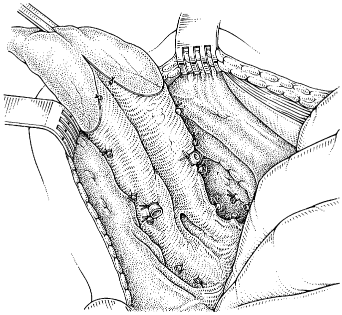

Fig. 7.11

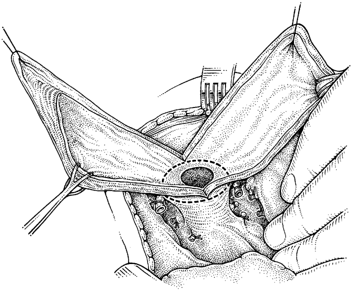

Fig. 7.12

diathermy (Figs 7.12 and 7.13). There are usually a few bleeding points that need to be secured at the anterior aspect of the bladder neck and at the bases of the lateral pedicles. If the plane between the interureteric region of the bladder base and the seminal vesicles/anterior vaginal wall has not previously been clearly defined, there may be some troublesome bleeding points in this area as well, particularly in girls. These points should be well controlled at this stage as they will be impossible to control with any ease after the cystoplasty segment has been sewn in place.

The right side of the colon is then mobilized on its vascular pedicle. The right colon is chosen because it is the most mobile segment by virtue of its vascular pedicle, of which the most important component is the marginal artery. This means that a tension-free anastomosis to the bladder outflow can be guaranteed. The mobilization of the right colon begins with an incision in the peritoneum of the right paracolic gutter, which is continued around the cecum and terminal ileum below and the hepatic flexure above. This peritoneal incision includes the underlying connective tissue bands that hold the mesocolon on to the posterior abdominal wall, particularly the phrenocolic ligament, which runs on the surface of Gerata's fascia to the hepatic flexure. As the mobilization continues, the right colon can be retracted medially, allowing a further separation of the right mesocolon and the vessels contained in it from the posterior abdominal wall (Fig. 7.14).

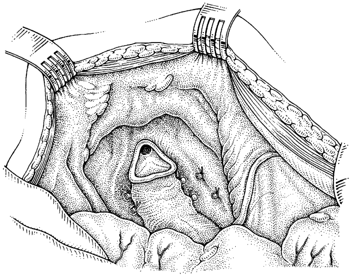

Fig. 7.13

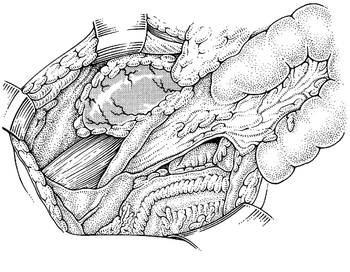

Fig. 7.14

As the peritoneal incision is extended around the hepatic flexure, the right lateral limit of the attachment of the omentum to the transverse colon will be encountered. The omentum will need to be separated from the transverse colon in this area over a length of about 7–10 cm, which, assuming that the underlying phrenocolic ligament has been completely divided, will then allow the right colon and the first part of the transverse colon to be retracted medially to expose the middle colic vessels on their posterior aspects (Fig. 7.15, left). An adequate reflection of the omentum will also expose these vessels on their anterior or peritoneal aspect, but the vascular anatomy is more easily appreciated from the posterior aspect, and a clean exposure of the posterior aspect of the middle colic artery at this stage has the additional advantage of ensuring adequate mobilization of the colonic segment from the underlying duodenum and pancreas, thereby reducing the risk of damage to these structures and to the superior mesenteric vessels.

The key to the vascular supply of the colonic segment is the marginal artery, which, at this stage of the operation, can be seen to be supplied by the ileocolic artery below (Fig. 7.15, right), the right branch of the middle colic artery above and the right colic artery nearer the ileocolic end. Figure 7.15 shows the posterior view of these vessels. The size of the middle colic, right colic and ileocolic arteries varies from individual to individual, but that of the marginal artery is constant. It always runs close to the medial aspect of the colon as far as the right branch of the middle colic artery, at which point it runs anatomically up to the bifurcation of the main trunk of the middle colic and down the left branch to continue as the marginal artery again, hence the importance of an adequate exposure of the middle colic artery.

In most cases, the best place to divide the colon is between the two branches of the middle colic artery, the most suitable place to divide the blood supply being at the right branch of the middle colic artery, just below the bifurcation. This will give a colonic segment, supplied by the marginal artery and its arcades and fed by the ileocolic artery (Fig. 7.16), of sufficient length to reach down to the bladder remnant with predictable ease (Fig. 7.17) and of sufficient

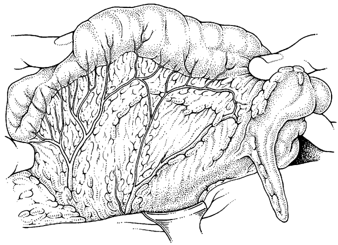

Fig. 7.15

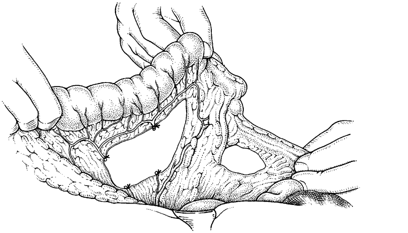

Fig. 7.16

capacity to serve as a substitute bladder (Fig. 7.18). Figure 7.18 shows the cystoplasty segment filled with water to illustrate its capacity.

At the ileocecal end, the mesenteric vessels are identified, ligated and divided in the usual way to give an ileal tail of about 5–7 cm in length. The isolated ileoceco-colonic segment is then retracted posteriorly to allow the restoration of bowel continuity by a two-layer anastomosis of the ileum to the transverse colon in front of the isolated cystoplasty segment. The cystoplasty segment is then washed out with saline to clear it of any fecal material. If the ileocecal valve is sufficiently competent to act as an anti-reflux mechanism, or if an anti-reflux mechanism is thought to be unnecessary or undesirable, the ureters are anastomosed to the ileal tail. This is often the case in adults undergoing a cystoplasty after a cystectomy for bladder cancer, for example, or, for the latter reason, in children with grossly dilated ureters. It is very difficult to achieve anti-reflux prevention when the ureters are grossly dilated without simultaneously causing obstruction. It is for this reason that anti-reflux prevention is undesirable with such ureters.

The left ureter is mobilized to above the pelvic brim and then brought across the front of the great vessels, deep to the sigmoid mesocolon, to join to the right ureter side-to-side just above the right common iliac vessels. The cystoplasty segment is then rotated through 180 degrees to put the colonic end into the pelvis and the ileal end into its correct relationship to the common ureteric orifice, which is then anastomosed to the ileum with one layer of continuous 3/0 or 4/0 Vicryl sutures (Fig. 7.19). The surrounding peritoneum is then tacked to the ileum to position the ureteroileal anastomosis retroperitoneally.

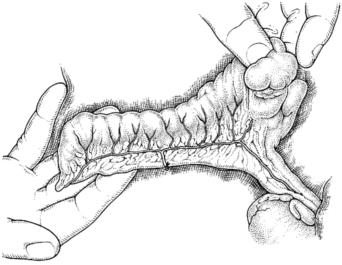

Fig. 7.17

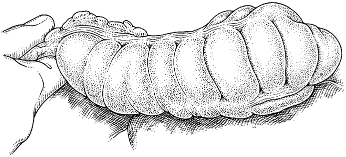

Fig. 7.18

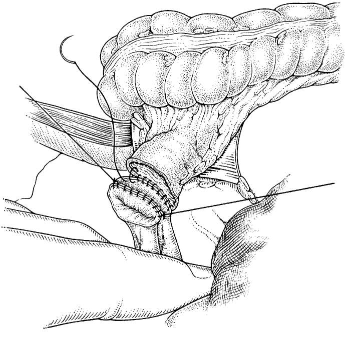

Fig. 7.19

If an anti-reflux ureteric reimplantation is thought to be desirable, it is safest not to rely on the ileocecal valve but to perform an extramural tunneled reimplantation into one of the tenia coli – either of both ureters into two separate sites or of one ureter with a transuretero-uterostomy of the other ureter at a higher level.

This leaves the anastomosis of the colonic end of the cystoplasty segment to the bladder remnant. This is achieved by starting two continuous sutures in the posterior midline and then running one around each side, locking the running stitch at two or three points to prevent a purse-string constriction of the anastomosis (Fig. 7.20). Each bite of the stitch should pick up the full thickness of the colonic wall and both the outer fascial layer of the bladder remnant and the urothelium. In this way, the urothelium is splayed out to the outer fascial layer of the bladder remnant, providing a fixed bladder base, and direct mucosal apposition between the two parts of the anastomosis is assured.

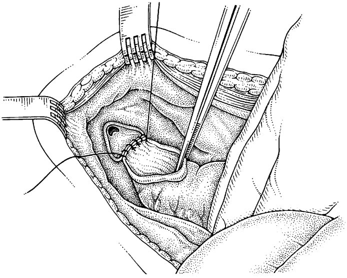

Fig. 7.20

Finally, the defect between the cystoplasty and the right posterolateral wall of the pelvis should be closed to prevent the small bowel prolapsing through it. This is achieved by tacking the peritoneum of the side wall of the pelvis, where it was incised during the course of the subtotal cystectomy, to the colonic segment, taking care not to damage any of the blood vessels in the vascular pedicle.

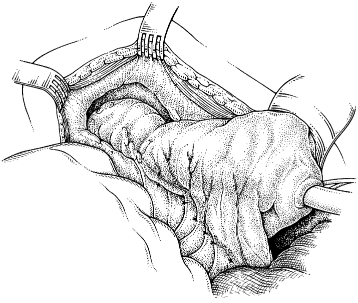

Fig. 7.21

At the end of the procedure, a large-bore Foley catheter is left in the cystoplasty bladder through a stump of the appendix (Fig. 7.21) or some other convenient site. A wound drain is left to drain the uretero-ileal anastomosis and the pelvis.

The omentum is wrapped around the cystoplasty segment and its upper and lower anastomoses and held in place with a few stitches (Fig. 7.22), the wound then being closed in the usual way.

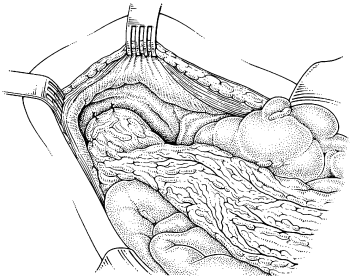

Fig. 7.22

De-tubularization: the ileocecal pouch

Troublesome colonic activity is sometimes found after the type of substitution cystoplasty described above, and this may be a cause of incontinence if the child's sphincter mechanisms are unable to control it. This is particularly so in children with neuropathic sphincter weakness or when a sphincter ablation and implantation of an artificial sphincter has been performed. Colonic activity may in any case be excessive in neuropathy.

For these reasons, it is wise to anticipate problems in children with neuropathic dysfunction, or those in whom the sphincter is congenitally inadequate for either structural or functional reasons, and to modify the cystoplasty technique to ensure as far as possible a low-pressure system. This is achieved in one of two ways. The first involves isolating a segment with a longer than usual ileal component that is nearly as long as the ceco-colonic part (Fig. 7.23) and then opening up the ileal part and all but the last few centimetres of the colonic part (Fig. 7.24), thereby dividing the circularly orientated smooth muscle in the segment that is thought to be responsible for most of the troublesome contractile activity. The ileal segment is then sewn as a patch into the divided colonic segment to produce an ileocecal pouch (Fig. 7.25).

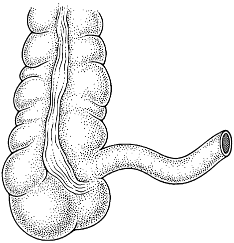

Fig. 7.23

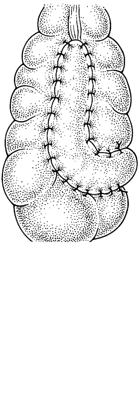

Fig. 7.25

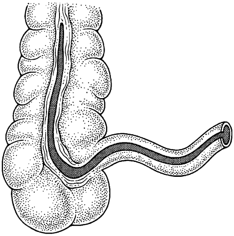

Fig. 7.24

The ureters are then reimplanted into the cecum directly (as described above), and the distal colonic end is sutured to the bladder remnant in the usual way (Fig. 7.26).

This technique is particularly suitable when the right colon tends to lie in a higher than usual position on the posterior abdominal wall, as regularly happens in spina bifida. In the more usual anatomic arrangement, with a normally sited right colon, de-tubularization can be achieved by opening the colon, mobilized in the way detailed above, along its anterior border and then dropping the distal half over the proximal half and sewing the two together.

Postoperative management

This is the same as for augmentation cystoplasty, the only difference being the increased likelihood of voiding difficulties. Although it is more common with substitution than augmentation cystoplasty, any voiding difficulty is managed in the same way.

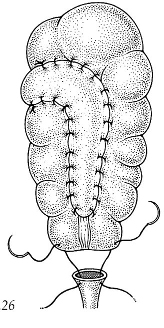

Fig. 7.26

Alternatives to cystoplasty

One occasionally needs an alternative to the cystoplasty techniques described above. As alluded to earlier, the stomach may be used in patients with a short gut syndrome, usually the result of cloacal exstrophy, intestinal disease or previous surgery.

In an attempt to avoid the complications of incorporating a gut segment into the urinary tract, various surgeons have tried alternative techniques, such as autoaugmentation, in which the bladder wall muscle is incised or excised to allow the bladder mucosa to prolapse through as a large diverticulum, thereby restoring a normal capacity at low pressure. The results of this are very satisfactory in the short term but very poor in the long term.

To try to overcome these long-term bad results yet still maintain the urothelial lining, thereby avoiding most of the disadvantages of intestinocystoplasty, autoaugmentation has been combined with the replacement of the bladder muscle layer with a de-epithelialized gut segment, usually sigmoid colon. The hope is that the transitional epithelium exposed in autoaugmentation will 'take' on to the de-epithelialized sigmoid colonic segment. The long-term results of this technique are not yet available.

The question more commonly concerns not whether there is an alternative cystoplasty technique that would be more appropriate, but whether there is some alternative to cystoplasty that would be more appropriate. This alternative has traditionally been ileal conduit urinary diversion. Nowadays, continent diversion is the alternative more readily considered, although ileal conduit urinary diversion still has its place and should not be overlooked. This is not the place to discuss continent urinary diversion, but it should be considered in any patient being considered for a cystoplasty in whom urethral catheterization would be difficult or impossible, or when the patient has a restriction of mobility such that access to toileting facilities is not readily available. Both of these considerations apply principally to the wheelchair-bound spina bifida patient. Continent diversion is described elsewhere in this volume (See Chapter 9).

Problems associated with cystoplasty caused by the incorporation of an intestinal segment into the urinary tract

Gut continues to function as gut after transposition into the urinary tract, therefore continuing to contract, to secrete mucus and to absorb water and electrolytes.

Gut contractility has already been discussed above with reference to de-tubularization. It has been suggested that de-tubularization abolishes gut contractility, but this is not the case. Gut will always be contractile, although this contractility may be temporarily reduced after surgery. If the gut segment used for a cystoplasty has a sufficiently high capacity, it will be acontractile while it holds a volume sufficient to give the patient a useful degree, that is 3–4 hours' worth, of continence. The aim of de-tubularization is not therefore to abolish contraction but to maximize bladder capacity for a gut segment of a given size in order to keep functional capacity as a relatively small fraction of total bladder capacity and thus reduce the likelihood of significant contractile activity in normal circumstances of use.

Problems from mucus secretion are rare in patients voiding spontaneously and to completion. Mucus may build up when voiding is inefficient, but mucus retention is most commonly seen in patients using CISC to void, particularly when the catheter is of a caliber less than 12F. When mucus is a problem, it can be dissolved by instillations of 30 ml 20% acetyl cysteine. It is useful to use regular wash-outs of a cystoplasty segment whether or not the patient has problems from mucus when a small-caliber catheter is used for CISC, in order to prevent problems (such as infection or stone formation; see below) occurring.

Fluid absorption or secretion by the cystoplasty segment does not usually cause problems (although fluid secretion may contribute to nocturnal enuresis in some patients), but electrolyte shifts may cause problems of metabolic acidosis and, with colonic segments, hypokalemia (because of potassium secretion). Many patients with a cystoplasty have metabolic acidosis, usually with respiratory compensation, but this is usually detectable only by arterial blood gas analysis. Only 16% have hyperchloremia (Nurse & Mundy 1989b). This acidosis may be symptomatic and require oral bicarbonate treatment. Mostly, however, it is asymptomatic, but it may nonetheless lead to skeletal demineralization and an impairment of growth in growing children. Growing children should therefore receive prophylactic oral bicarbonate as a matter of course. In older children and those who are asymptomatic, bicarbonate is unnecessary.

Another major potential problem after cystoplasty is bacterial colonization of the neo-bladder. About 50% of patients with ileocystoplasties and 75% of patients with colo-cystoplasties develop chronic or recurrent bacteriuria with a mixed growth of organisms. This is often asymptomatic and probably does not require treatment in its own right, although it may give rise, in association with mucus, to stone formation (see below). This bacteriuria is, however, associated with a high concentration of urinary nitrosamines, which are known carcinogens. The presence of nitrosamines is in turn associated with metaplastic changes in the cystoplasty and the bladder remnant, particularly with colo-cystoplasty (Nurse & Mundy 1989a). This association is obviously worrying as it suggests that there is a potential for long-term malignant change in a cystoplasty similar to that seen after a ureterosigmoidostomy (Leadbetter et al 1979). A recent review of over 2000 continent urinary diversions and neo-bladders failed to identify any cases of malignancy, but the follow-up period was only about 5 years. Given that the latent period of adenocarcinoma in ureterosigmoidostomy can be over 20 years, the apparently favorable results of Roland and Reagan's series should be interpreted with caution.

Several animal and human studies have suggested that there is an increased risk of developing cancer in augmented bladders. High rates of transitional cell metaplasia and carcinoma have been found in ileocystoplasties and gastrocystoplasties in rats, and there is some evidence that the potential risk is increased in gastrocystoplasty and colo-cystoplasty compared with ileal augmentation or substitution. The significance of these results in children undergoing augmentation or substitution cystoplasty remains to be seen, but the long-term follow-up of all patients should clearly be regarded as mandatory.

Stones are increasingly being found in patients with cystoplasties and continent diversions. In about 50% of patients, they are found incidentally; the rest present with symptomatic infection or a deterioration in urinary continence in roughly equal proportions. Stones occur in about 6% of patients with augmentation and substitution cystoplasties overall, being very much more common in those who use CISC to void (10–17%) than in those who void spontaneously (2%). Mucus and bacteriuria must clearly be at least contributory, but stasis seems to be the most important cause of stone formation as stones occur even more readily in continent diversions emptied by CISC from above. It is for this reason (to reduce urinary stasis and thereby stone formation and other complications of stasis) that regular wash-outs are advised in all patients using CISC to empty a cystoplasty.

The rupture of the augmented or substitute bladder has been widely reported in the pediatric literature, but this author has only ever seen two cases in a series of over 500 patients. It is difficult to explain away this discrepancy, but rupture does seem to be far more common in those people who are 'overcontinent' using CISC to empty. If the bladder does not have a natural tendency to leak at capacity, rupture seems that much more likely. The importance of identifying bladder rupture cannot be overemphasized: it has a rather insidious and undramatic presentation, but the condition carries a high mortality.

Other complications are unusual. Additional natural and metabolic disturbances have been reported, but urodynamic problems are much more likely to be caused by contractile activity in the gut segment or by sphincter weakness, whereas incontinence and diarrhea are much more likely to result from an interference with ileocecal valve function or simply from a disruption of the patient's normal bowel routine by the experience of surgery.

Finally, it should be pointed out that many of these children are female and will hope to become pregnant at some stage in their later lives. This does not appear to be a problem. In our own series of 245 females who underwent augmentation or substitution cystoplasty, 27 have undergone 34 pregnancies. Of these, 28 were normal vaginal deliveries that occurred without complication. Of the 6 patients who underwent a cesarean section, 4 interventions resulted from obstetric indications. In short, if the patient is able to became pregnant, a normal vaginal delivery can be expected, a cesarean section usually being indicated only on obstetric grounds.

Conclusion

Augmentation and substitution cystoplasty are useful and important techniques in the management of a wide range of lower urinary tract problems. To achieve the best from them requires careful patient selection, appropriate preoperative investigation, attention to detail in the performance of the procedure and careful long-term follow-up.

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Chapter 8

Gastric augmentation

Michael E. Mitchell and Michael C. Carr

Indications

The indications for gastrocystoplasty are similar to those for other types of augmentation cystoplasty: detrusor dysfunction with a small, poorly compliant bladder. In selected cases, bladder replacement with stomach may be necessary. The advantages of gastrocystoplasty are particularly apparent in patients with chronic acidosis secondary to renal insufficiency and in those patients with major complex problems.

Augmentation cystoplasty has been described using ileum, cecum or sigmoid colon as isolated flaps or in curious shapes, combinations and quantities. This has allowed for the creation of a large and compliant urinary reservoir, such as in the Mainz, Koch, Indiana or LeBag procedure. The long-term application of some of these procedures has been reported, but many of these applications are relatively new (Mitchell & Piser 1987, Fisch et al 1992).

Following successes with gastrocystoplasties in a canine model, it became clear that stomach could be utilized to augment the bladder in the pediatric population (Adams et al 1988). A net excretion of chloride ions occurs which supports a main buffer system of the urine (ammonium chloride). This permits the secretion of acid without the use of titratable acids that potentially leads to the depletion of the buffer systems. Ammonia resorption does not occur either, as has been observed when large and small bowel are used in the urinary tract. In a partial renal failure model, gastrocystoplasty actually protected dogs with chronic renal failure from the acidosis of chronic ammonium chloride loss (Kennedy et al 1988). A patient with chronic acidosis and renal insufficiency could theoretically be in part protected by gastrocystoplasty, whereas large and small bowel in the same capacity would only serve to increase the acidotic state. This has proved to be the case clinically. The presence of acid in the urine may also contribute to a reduced rate of infection. In the rat model, however, metaplastic changes have been noted, although the potential for malignant degeneration is still uncertain.

The use of stomach has proved to be invaluable in reconstructive procedures for patients with complex problems. We have used it in patients in whom no other type of bowel was available for urinary reconstruction, including those with previous irradiation and those with an absence of large bowel or a reduced length of small bowel (the cloacal exstrophy group). Stomach has also been selected for patients with metabolic acidosis, as this condition would be exacerbated by the addition of large or small bowel because of chloride or ammonia resorption. In addition, the aciduria could prove beneficial for patients with chronic and recurrent infections.

Despite the successes with gastric augmentation and bladder substitution, the ideal augmentation would *not* introduce either bowel or gastric *mucosa* into the urinary tract. Metabolic complications, urolithiasis, mucus production, spontaneous perforation and even malignant degeneration would thus be eliminated. A urothelium-lined bladder with gastrointestinal tract muscle augmentation of the detrusor muscle was attempted as early as 1955. Other de-mucosalized intestinal segments have also been used in several other studies. Another surgical attempt to preserve transitional epithelium is detrusorrhaphy or autoaugmentation. The detrusor muscle can, however, potentially be replaced with fibrous tissue or whatever adheres to the urothelium, which may result in an inadequate bladder volume or poor compliance. An autoaugmentation gastrocystoplasty or a de-mucosalized gastric flap procedure uses a full-thickness urothelial graft in conjunction with a raw inner surface of incorporated stomach muscle/submucosa.

Operative technique

Patient selection for gastrocystoplasty is no different from that for any other bladder reconstruction operation. Patient selection was initially limited to those who had a deficiency of bowel or patients with a metabolic acidosis. The procedure is, however, well suited to patients who use small catheters or who, in general, would tolerate mucus and highly viscous urine poorly, and for those with problems with chronic urinary infection.

Rigorous bowel preparation is not required. Most patients are placed on a liquid diet 48 hours before surgery in addition to receiving a bottle of magnesium citrate 24 hours prior to operation. Mechanical and antibiotic bowel preparations have been used in patients anticipating major reconstruction and those who have had multiple previous abdominal procedures. Recently, because of restrictions on hospital stays, patients have been admitted on the day of surgery. When possible, all patients have preoperative video urodynamics, a renal ultrasound, and either an intravenous pyelogram or renal scan

(MAG 3 lasix renogram or DMSA scan). Every effort is made to sterilize the urine prior to the procedure. In children, we have not felt it necessary to obtain an upper gastrointestinal series, but this may be prudent in adults.

A long midline incision is made from the symphysis pubis to the xyphoid process. A self-retaining ring retractor is very useful in this procedure, which demands adequate exposure deep in the pelvis and high in the epigastrium. In a case of gastrocystoplasty, the bladder is initially explored through the lower part of the incision and opened in the sagittal plane in the midline from the bladder neck (anteriorly) to the trigone (posteriorly). The upper part of the incision and peritoneum usually need to be opened only after bladder preparation has been completed. Ureteric reimplantation is, if necessary, performed at this time. It is also a good time to develop a plane around the bladder neck should this be required (for example, for the implantation of an artificial sphincter or for a sling procedure). Pediatric feeding tubes are left in the ureters to facilitate identification and prevent injury during the augmentation procedure. The bladder is packed with a moist sponge and the upper abdomen explored.

The stomach is brought into the surgical field. It is helpful to use large Babcock clamps to perform this maneuver (making sure that the Babcock clamps do not compromise or injure the gastroepiploic arteries [GEAs]). The GEAs are then carefully evaluated. Common observations at this point will be that the GEA along the left inferior margin of the greater curvature is either very thin or will apparently dive into the stomach wall. It is thus usually preferable to base the wedge gastric flap on the *right* GEA. The greater omentum is then incised parallel to the GEA several centimetres inferior to this vessel; the electrocautery and bipolar electrode can be used to do this very effectively. Larger vessels may, however, need to be ligated. Although some of the omental vessels are quite significant, and it may seen preferable to base the wedge flap on these, we have not done so to date as this would necessitate bringing the blood supply to the flap *anteriorly* to the abdominal contents.

The triangular gastric wedge is next selected, the apex of the wedge being close to, but not including, the lesser curvature of the stomach. The length along the greater curvature is usually about 9–15 cm, but this depends upon the patient's age and size, and the anticipated final bladder volume. The position of the wedge will depend on the anticipated blood supply. If the more constant right GEA pedicle is used, the wedge is usually taken closer to the cardia of the stomach. The rule of thumb is that the largest possible wedge should be taken with the longest blood supply without altering gastric physiology.

About a third to a half of the stomach is usually removed. The short arteries from the GEA to the stomach flap are kept intact. The others along the anticipated pedicle are carefully ligated in place and divided (Fig. 8.1a). Great care must be taken during this portion of the procedure to make sure that the GEA is protected and not injured. It is sometimes helpful gently to apply papaverine solution to the artery along its entire length to prevent spasm. The GEA at the distal end of the wedge is then divided. The 90 mm

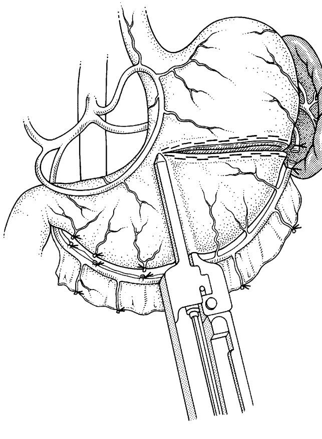

Fig. 8.1a

gastrointestinal anastomosis stapler is used to resect the wedge. The staples prevent blood loss from the flap and stomach during the repair of the stomach. The wedge flap is wrapped in a moist sponge and placed in the pelvis, with a moist sponge over the pedicle to protect it during the gastric repair.

The repair of the stomach is made in two layers. An outer layer of interrupted 3/0 silk sutures in the muscularis and serosa and an inner layer of through-and-through running 3/0 Monocryl sutures are used. The posterior interrupted silk sutures are placed before the gastric lumen is opened. The staples may be left in the posterior suture lines. A nasogastric tube is then positioned across the suture line, this being left in place until bowel motility returns (usually on postoperative day 4 or 5). The wedge-shaped gastric flap is then brought with its blood supply through the mesentery of the transverse colon and through the root of the small bowel mesentery (Fig. 8.1b). It therefore courses in a retroperitoneal position with its blood supply on the right side. The surgeon should ensure that there is no twisting of this pedicle in the process of placing the flap on the bladder. The wedge-shaped flap should reach well down into the pelvis without any tension on the blood supply. If a further length of pedicle is required, this is secured by dissecting proximally at the gastroduodenal angle.

The wedge flap is then opened to form a parallelogram. All the staples are removed. The posterior apex is sutured into the area of the posterior bladder wall close to the trigone (Fig. 8.1c). Running, locking through-and-through 3/0 Monocryl sutures are used to sew the back wall in place. A second layer of 3/0 Vicryl sutures through the muscularis and serosa then ensure a watertight anastomosis. A Malecot catheter is usually placed before the anterior segment of the augmentation is sutured in place, the size of this catheter depending on the age of the patient. It is usually preferable to use one no larger than 14F gauge. This tube is usually brought out through native bladder if possible, but it may exit through the flap if fixed to the abdominal wall like a gastrostomy tube. The ureteric catheters are left in place during the posterior anastomosis and may then be removed if reimplantation or tapering has not been performed. The anterior portion of the augmentation is then completed, and the bladder is tested to make sure that the suture lines are watertight. Any leaks must be repaired at this point.

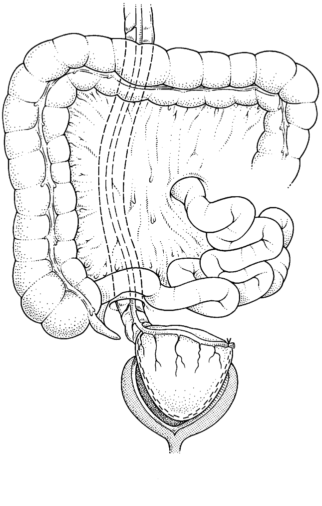

Fig. 8.1b

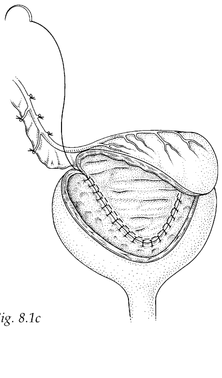

Fig. 8.1c

We tend not to drain gastrocystoplasty patients. The omentum, which is still intact, is placed over the anterior portion of the bowel after the windows in the retroperitoneum have been closed with a running Vicryl suture. With the omentum tucked over the bowel and behind the augmented bladder, the bladder resides close to its original position. A careful check should be made that there is no tension on the pedicle of the flap.

Total bladder replacement with the wedge flap

Total bladder replacement with stomach is possible using the wedge flap. Following the removal of the staples, the apex of the wedge is sutured to the urethra (Fig. 8.2). This may be feasible in the patient with an intact urethra (bladder replacement). The urethra may be sutured to the apex of the flap, which can be tubularized to provide a nice bladder neck.

Several options are available for patients requiring the construction of a catheterizable reservoir. A distal ureter may be used as a catheterizable channel. The ureter can be tunnelled between the mucosa and the muscularis of the reservoir wall to provide continence. This is generally possible only in the cloacal exstrophy population. The appendix has been used in a similar manner. The construction of a bladder tube, using a strip of the gastric flap, has also been used successfully (Fig. 8.3a). The tube is nippled into the reservoir to provide continence. This has resulted in a channel that is catheterizable and dry (Fig. 8.3b). Our preference, however, is to use a tunnel technique for dryness, with the construction of a gastric reservoir.

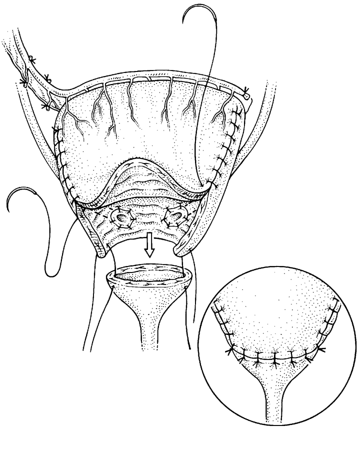

Fig. 8.2

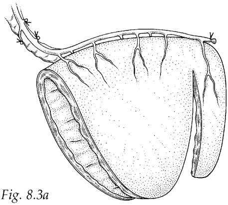

Fig. 8.3a

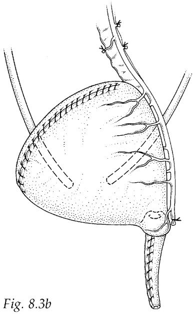

Fig. 8.3b

In bladder replacement, it is usually most convenient to open the wedge completely and suture the posterior tip (which usually is the anterior apex of the wedge) onto the urethra. Subsequently, with the gastric segment open, ureteric reimplantation is easily performed in much the same way that a tunnelled reimplant would be performed in the bladder. A plane between the mucosa and muscularis of the gastric flap is easily defined with sharp dissection. The anterior portion apex is then sewn to the anterior portion of the urethra, and the lateral aspects of the wedge are then closed using a two-layer closure of running 3/0 Monocryl and Vicryl. A suprapubic tube (a Malecot catheter) may be brought out through the gastric tissue, but if this is done, it is advisable to suture the wall of the gastric reservoir to the anterior abdominal wall as with a gastrostomy. This prevents intraperitoneal leakage when the tube is removed. A catheterizable stoma should be constructed flush with adjacent tissue and in such a manner that catheterization can be performed without difficulty. A small V flap of skin and spatulation of the catheterizable channel works well. It should be remembered that if catheterization is difficult in the operating room, it will be impossible outside it.

De-mucosalized gastric flap procedure

The augmentation of a bladder after detrusorrhaphy with a de-mucosalized gastric flap is similar to a gastrocystoplasty. The bladder is approached initially. It is helpful to have a Foley catheter in the bladder connected to a reservoir of irrigation saline that can be raised or lowered to fill or empty the bladder as desired. The bladder is filled to a pressure of approximately 20 cm of water and the detrusor incised in the midline sagittal plane. Great care is taken to preserve the transitional epithelium when finding the plane between the transitional epithelium and the muscularis. It is best to use blunt and sharp dissection with the bladder full (Fig. 8.4a). If no holes are made in the epithelium, this is fairly easy. Once a hole has been made and the bladder decompressed, it becomes very difficult to dissect the plane between the transitional epithelium and the bladder muscularis. We usually remove the muscularis from between two-thirds and three-quarters of the anterior and lateral wall of the bladder. The bladder is emptied by lowering the reservoir and is then gently packed with moist sponges. An alternative technique is to use a large balloon catheter (Helmstein balloon) to facilitate this dissection. This can be left in place following surgery to promote the healing of the epithelium to the seromuscular flap.

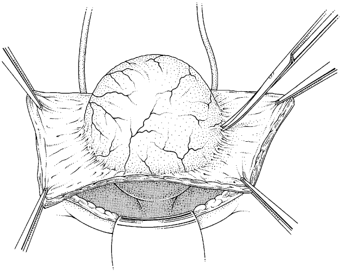

Fig. 8.4a

A gastric flap is obtained. Before it is brought into the pelvis, however, the mucosa is removed. The staples are removed, four quadrant sutures for traction being used so that the flap is opened as a parallelogram. A vascular loop is placed gently around the pedicle with enough traction that the blood supply to the pedicle is temporarily interrupted. It takes approximately 10–15 minutes sharply to dissect the mucosa from the muscularis layer. Potts tenotomy scissors are used for this. The proper plane is easily found and tends to have a somewhat foamy appearance (Fig. 8.4b). Once the mucosa has been

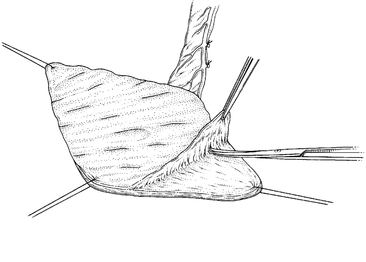

Fig. 8.4b

removed, the vascular loop is loosened and bleeding vessels are electrocauterized using bipolar cautery. Most of the bleeding will be in the midline where perforators follow the gastroepiploic pedicle.

The de-mucosalized flap is brought into the pelvis as with any augmentation, being laid directly on top of the outside of the bladder epithelium, raw surface to raw surface. Either a Penrose drain or a suction drain is placed between the transitional epithelium and the flap, and is brought out through the abdominal wall. This will be removed in approximately 48 hours. The de-mucosalized gastric flap is sutured in place using a running, locking 3/0 Vicryl suture as a single layer closure (Fig. 8.4c). In an effort to maintain contact between the transitional epithelium and the de-mucosalized gastric flap, 'the bladder is kept partially filled for approximately 5–7 days. Alternatively, a large balloon catheter may be used. Subsequently, the patient is placed on either intermittent catheterization or the intermittent clamping of a suprapubic tube. If a suprapubic tube is used, it is brought through the intact bladder wall rather than through the healing transitional epithelium and de-mucosalized gastric flap.

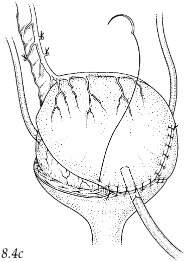

Fig. 8.4c

Postoperative care

The management of the gastrocystoplasty patient is similar to that of any patient with an intestinal augmentation. Mucus is generally not a problem, but irrigation of the bladder should initially be routinely performed. All patients are maintained on intravenous antibiotics and then converted to chronic acid suppression for at least 2 months after surgery. H_2-blockade, usually using ranitidine (1 mg/kg every 8 hours), is continued for 1–2 months postoperatively. If the patient then develops dysuria or perineal pain, the ranitidine is re-started until the symptoms resolve or the hematuria improves.

The removal of the suprapubic tube is dependent on the ability of the patient to empty the bladder. A trial period of at least 1 week of continuous clamping is usually wise while the patient begins to void or intermittent catheterization is reinstituted. Finally, patients with anuria, or dilute urine, may rarely require irrigation with phosphate buffer if a low pH and irritation are observed. Infection is less common with the gastrocystoplasty patient, but asymptomatic infection is usually not treated unless the organism is felt to be significant for potential stone formation or pyelonephritis. Serum electrolyte levels should be periodically followed.

Early and late complications

Our initial experience of 80 patients with stomach augmentation or replacement of the bladder has been very encouraging. With an average follow-up of 4 years, all but three patients have shown either stable or improved renal function. All but seven patients are dry, and urodynamic studies have demonstrated a marked improvement in compliance. The gastrin level has not been elevated, and no patient has experienced duodenal or gastric ulceration. Early complications have included a gastric bleed in one patient that did not require transfusion, a perivesical extravasation in one patient, and an obstruction of a ureter reimplanted into the stomach in another. Delayed complications have included bowel obstruction in 7 patients, spontaneous bladder perforation in 3 and hematuria and/or dysuria in 28 patients.

The so-called hematuria/dysuria syndrome is unique to the use of stomach in the urinary tract (Nguyen et al 1993). The symptoms are usually intermittent and self-limiting and improve with time. A spectrum of severity is seen, ranging from intermittent dysuria and tea-coloured urine to the rare occurrence of ulcer formation and bladder perforation. The majority of patients are controlled with the intermittent use of H_2-blockers, sodium bicarbonate bladder irrigation or oral bicarbonate therapy. A few children have required short-term oral omeprazole (an anti-secretory compound that suppresses gastric acid secretion).

The symptoms are more pronounced in adult patients and those children with sensate urethras in whom voiding is attempted or if there is wetness. The cause is believed to be hydrochloric acid production by the gastric segment and a reduced buffering effect by the urine. Symptoms have, however, been seen even with urine acidity in the normal range. There is a risk of developing a peptic ulcer in the gastric portion of a defunctionalized gastrocystoplasty. Anuric states (i.e. renal failure or urinary diversion) should be treated with basic bladder irrigation solutions such as phosphate-buffered saline or bicarbonate to prevent this.

Patients with gastrocystoplasties who lose large amounts of hydrochloric acid, sodium and potassium in the urine may become severely dehydrated and alkalotic. This usually occurs only when patients have significant salt loss from the bowel or kidney such as with severe diarrhea or vomiting, salt-losing nephropathy or, rarely, hypergastrinemia. The ensuing hyponatremic hypochloremic alkalosis requires intravenous saline volume replacement and is usually easily corrected. It may be that excessive salt loss related to an increased serum gastrin level relates to reduced acid production in the stomach secondary to the gastric resection. This can sometimes be reversed by taking carbonic acid (soda pop) with meals. The majority (77%) of the patients on clean intermittent catheterization have had at least one positive urine culture.

Our follow-up of patients who have undergone the de-mucosalized gastric flap procedure is limited to 10 patients, all of whom have tolerated the procedure well. Postoperative urodynamic studies will be necessary in order to demonstrate an improvement in bladder capacity and compliance, but our early dynamic data are very encouraging. For those patients who do not have a problem with recurrent urinary tract infection or have underlying renal insufficiency, this procedure may be an excellent alternative that avoids some of the complications of the traditional gastrocystoplasty.

Our experience with using the stomach for urinary reconstruction now extends to more than 80 patients treated at this institution, their diagnoses including neurogenic bladder, bladder exstrophy, exstrophy of the cloaca, urethral valves and ectopic ureter. We have performed augmentation and bladder replacement in both adults and children, although most of our experience is with the younger patients. Stomach can be used totally to replace the bladder, but older male patients may experience urethral irritation. The advantages are little mucus, reduced infection and the ability to tunnel ureters easily or use the stomach as a conduit with a catheterizable stoma, which make this a very appealing technique. We feel that stomach is, in many clinical situations, clearly advantageous over other tissues, particularly large and small bowel.

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Chapter 9

Continent urinary diversion

Christopher R.J. Woodhouse

Introduction

The lower urinary tract consists of a reservoir, a passage (or conduit) to the surface and a continence mechanism. The bladder and the urethra with its sphincters comprise much the most efficient system and should be abandoned only if they are hopelessly diseased. The reconstruction of this natural system is covered in Chapters 7 and 20.

Great ingenuity has gone into devising operations for urinary diversion. Historically, three distinct phases can be seen. From the begining of modern surgery in the second half of the nineteenth century, the urine was diverted into the colon, continence depending on the integrity of the anal sphincter. The ureterosigmoidostomy was the most commonly used technique, but there were several variations designed to limit the infective complications.

The second phase of development began in 1950 with the introduction of the ileal conduit. It is ironic that the management of the complications of ureterosigmoidostomy was just beginning to be understood at the time when its replacement was devised.

The third phase, continent urinary diversion, first became practicable with the adaption of the Kock pouch for the storage of urine (Kock et al 1978). Since then, a large number of different continent diversions have been devised, many differing from each other only in small details.

In a child requiring urinary diversion, it is most important to consider both medical and social factors. A suitable system should be tailored to the patient rather than blindly selecting the surgeon's favorite from a limited repertoire. Although incontinent cutaneous urinary diversions with a bag are seldom used in children, they still have an occasional indication.

For a continent diversion, a reservoir, a conduit and a continence mechanism may be chosen from the lists in Table 9.1.

Table 9.1 Tissues that may be used in the construction of continent urinary diversions.

Reservoir	Conduit	Control system
Stomach	Appendix	Mitrofanoff
Ileum	Ileum	Kock
Cecum	Ureter	Ileocecal valve
Colon	Fallopian tube	Anal sphincter
	Skin tube	
	Tubed stomach	

Indications for continent urinary diversion

As mentioned above, the natural lower urinary tract (the bladder, the urethra and the sphincters) is incomparably better than any continent diversion, so every effort should be made to preserve it or reconstruct it. In children, there are few indications for continent diversion, although many components of the relevant techniques may be used for other reconstructions.

Exstrophy

Modern reconstructive techniques allow about 80% of children to void normally for at least their early years. Even in the worst group, augmentation and an artificial sphincter should allow the retention of the bladder. Continent diversion is indicated only as a salvage method if reconstruction fails. In cloacal exstrophy, it may not be possible to reconstruct the bladder, but, on the other hand, neither may there be enough intestine to make a continent diversion.

Neuropathic bladder

A combination of augmentation and intermittent clean self-catheterization (ICSC) should provide a safe and continent reservoir in neuropaths. It should be remembered that those who spend all of their day time in a wheelchair, especially females, find urethral ICSC inconvenient. They may require a suprapubic continent stoma and occasionally a complete continent diversion.

Neoplasia

In spite of steady advances in the conservative therapy of pelvic rhabdomyosarcoma, many patients still require an anterior exenteration. Continent diversion is the reconstruction of choice.

Previous inappropriate diversion

Even now, children occasionally present having had an inappropriate cutaneous diversion. Most, fortunately, still have a defunctioned bladder in situ. Undoing the diversion, if necessary with reconstruction, is the best management. A few children will have had a cystectomy and therefore require the conversion of their cutaneous diversion to one that is continent.

Salvage procedures

Although lower urinary tract reconstruction has good results in the best hands, there are still some failures. These unfortunate children, who have often had many operations, may be salvaged by continent diversion. Although such a solution has its own complications, it is still preferable to an ileal conduit. Many children who were incontinent or had a bag have asked for a continent diversion; the author has never known a child with a working continent diversion ask for a bag.

The reservoir

A low-pressure, compliant reservoir may be made from 40 cm of small intestine or 20 cm of large intestine. The largest volume and lowest pressure for the smallest surface area is obtained by dividing the circular muscle and re-configuring the intestine as closely to a sphere as possible.

The choice of intestine is controlled primarily by that which is available and by the likely consequences of its removal from the gastrointestinal tract. As a urinary reservoir, one type of intestine seems to be as good as another, except that the jejunum is too metabolically active to store urine and should never be used. In children with a shortage of intestine or with impaired renal function, the stomach is a good choice for bladder reconstruction. It has been rarely used for continent diversion and will not be considered further.

Most children will have normal intestine and will not have had abdominal radiation. Fortunately, the remaining intestine has power to adapt when a substantial length is removed. Nonetheless, in those for whom chronic diarrhea would be a disaster (particularly myelomeningocele patients), the distal ileum, ileocecal valve and cecum should be preserved (for a review of the gastrointestinal consequences, see Schultz-Lampel & Thuroff 1991).

The conduit

The choices of conduit and continence mechanism are, to a certain, extent linked. For suprapubic diversion, three broad approaches may be considered:

1. the tunnelling of a narrow conduit, typically the appendix, into the reservoir wall between the muscle and mucosal layers (the Mitrofanoff principle);
2. the intussusception of a broad conduit, typically the ileum, to form a flutter valve (the Kock nipple);
3. use of the ileocecal valve with or without augmentation (the Indiana approach).

Aside from these, the anal sphincter still has a role when used with a low-pressure reservoir such as the Mainz sigma II or the Kock–Gonheim.

In terms of the first consideration, a narrow tube – the appendix, the fallopian tube or the ureter – must be used as the conduit. A very satisfactory narrow tube can, however, be made by tailoring an appropriate length of ileum if nothing else is available. As far as the second point is concerned, essentially the only choice is the ileum. For the third approach, only the terminal ileum can be used as a conduit, the ileocecal valve inevitably being removed from the the gastrointestinal tract.

Preoperative assessment

By the time continent diversion is being planned, the surgeon is likely to know the child and parents quite well. Nonetheless, it is important to take the time to discuss the extent of the surgery, the possible complications and the long-term consequences with the whole family. The implications of lifelong ICSC and the need for follow-up must be considered. This author has found it invaluable to have a clinical nurse specialist, a booklet and an explanatory video to help.

If the anus is to be used, particularly careful counseling is required. Sphincter function should be tested by asking the patient to retain a semi-solid enema using porridge or something similar. The offensive smell of the urine/feces mixture must also be emphasized. In the long term, the high incidence of carcinoma may be critical in the decision (see below).

Apart from the routine blood profiles, the following are most useful for planning surgery and in follow-up:

1. a urine culture
2. electrolyte levels, including chloride and bicarbonate
3. the glomerular filtration rate (preferably by chromium EDTA clearance)
4. an intravenous urogram.

Patients with spina bifida have a surprisingly poor respiratory reserve. In doubtful cases and especially those with thoracic lesions, respiratory function tests are essential.

Immediately before surgery, the child will need a bowel preparation. It is important that this preparation and the starvation on the night before do not leave the child dehydrated or, when the renal function is already impaired, precipitate acute renal failure. To prevent these, body weight should be closely monitored, and a saline drip can be started on the night before surgery.

The operation

Anesthesia

In this author's hands, a continent diversion takes up to 4 hours. It may involve an extensive laparotomy, mobilization of the whole of the gastrointestinal tract, isolation of a bowel segment and anastomosis as well as the reconstruction. It is not the job of the surgeon to tell the anesthetist how to give the anesthetic, but it is prudent to warn of the extent of the procedure, to prepare the patient adequately and to cross-match at least 3 units of blood. Aside from the bleeding, the fluid loss is considerable. A broad-spectrum antibiotic and metronidazole are started with induction and continued for 48 hours.

There is a prolonged paralytic ileus and a high incidence of chest complications postoperatively, high-grade spina bifida patients being particularly at risk. I have found that these problems are easier to manage if a gastrostomy tube is placed at operation and a nasogastric tube is avoided. Early mobilization and physiotherapy are essential. I do not routinely use intravenous hyperalimentation unless there is some clear indication for it.

Incision

The patient is positioned supine on the table. This author uses a midline incision. If there is any possibility that the umbilicus will be used for the stoma, it must be given a wide berth. All adhesions are taken down: a limited dissection of the previously operated abdomen nearly always results in inadvertent tears of the intestine, poor exposure or both. After a full exploration, a preliminary selection of the components to be used for the diversion can be made.

The conduit and continence system

The Mitrofanoff system The appendix is the best choice for a Mitrofanoff conduit. It commonly admits a 12F catheter and has a good vascular pedicle. The wall is compliant and thin enough to allow easy submucosal implantation.

The appendix is mobilized with a 3–4 cm cuff of cecum to allow lengthening of the conduit should it be too short (Fig. 9.1). The co-lateral supply of the cecum from the appendicular vessels is tenuous. Mobilization of the pedicle should be kept to a minimum. The distal tip is resected.

If the appendix is not available, the next best choice for the conduit is a segment of tailored ileum. It is most convenient to use the system originally described by Yang but usually attributed to Monti (Gerharz & Woodhouse 1998). A 2.5 cm length of ileum is isolated on a good pedicle (Fig. 9.2a). It is opened longitudinally, not on the anti-mesenteric border but one-quarter of the way around (along the line AB–CD in Fig. 9.2a). Having been opened longitudinally in this way, the ileum is closed transversely over a 16F catheter with interrupted absorbable sutures (Fig. 9.2b and c).

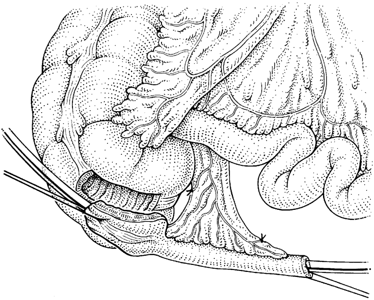

Fig. 9.1

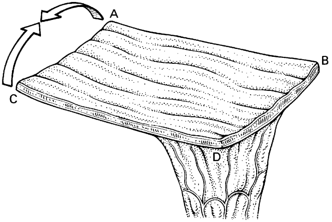

Fig. 9.2b

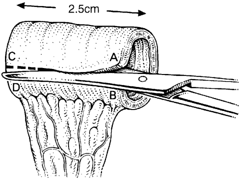

Fig. 9.2a

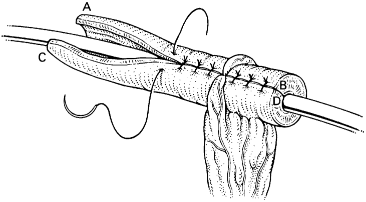

Fig. 9.2c

This produces a tube 7 cm long with a pedicle close to one end (Fig. 9.2d). The end remote from the pedicle is implanted into the reservoir and the other end brought out on to the skin. If the patient is very fat, two Monti tubes can be made and sutured together end-to-end (Fig. 9.2e) to increase the length available to be brought through the abdominal wall.

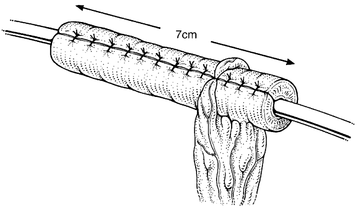

Fig. 9.2d

Providing the Monti tube is implanted in a submucosal tunnel, continence is as reliable as with any other Mitrofanoff tube. If the implant is into any form of trough that must be closed over the Monti tube, there is a danger that the suture line will overlie the sutures of the tube (Fig. 9.2f top) and cause an internal fistula and incontinence. This complication can be avoided if the Monti tube can be made to lie in the trough with its suture line against the back wall (Fig. 9.2f bottom). Alternatively, a gastrointestinal anastomosis (GIA)

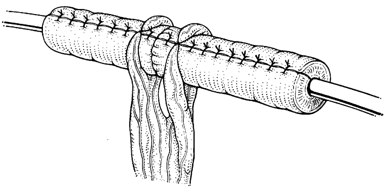

Fig. 9.2e

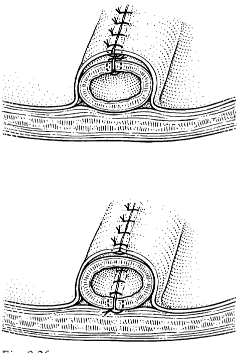

Fig. 9.2f

staple gun is applied to tailor a 7 cm length of ileum (Fig. 9.2g and h). Before firing the gun, the catheter should be taken out and re-inserted in each direction to be sure that catheterization will be possible. The gun is then fired with the catheter in position.

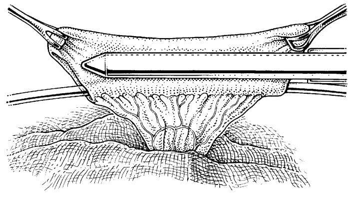

Fig. 9.2g

The ureter has properties similar to those of the appendix. A ureter that is completely normal may admit only an 8F catheter, but it can usually be dilated at least to 10F. Although this may provide a good conduit in the short term, this author has been disappointed with the long-term results and no longer recommends the use of normal ureters. A ureter that has been anastomosed to a conduit or has been obstructed will admit a 12F or 14F catheter without difficulty and has proved durable for long-term ICSC.

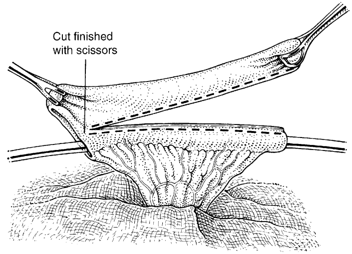

Fig. 9.2h

The ureter has several potential blood supplies. If the bladder is to be incorporated into the reservoir, it is likely to have at least one ureter attached to it with sufficient vascularity to maintain its viability. Similarly, if an ileal loop is being incorporated into a continent diversion, part of a ureter vascularized from the loop can be used. In a pelvis that has been spared previous dissection, the ureter can be mobilized on its artery, which is a branch of the internal iliac artery.

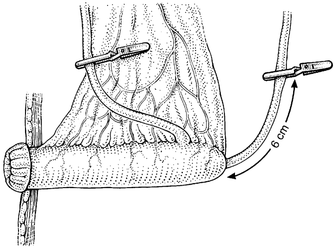

Fig. 9.3

If an ileal loop is being converted to a continent diversion, the ureter must have an adequate blood supply from the loop. When the ureter has been sufficiently mobilized, a bulldog clip should be applied at the proposed site of resection to check that both portions are independently viable (Fig. 9.3). If the blood supply is shown to be adequate, the ureter is divided.

The fallopian tube is a more delicate structure and is of adequate calibre only in its lateral two thirds. Long-term results with it have been poor, and it is difficult to justify its use in children who would otherwise be fertile. If it is to be used, it is mobilized on the ovarian pedicle.

A narrow tube can be made of skin or a short conduit lengthened by a skin tube.

Construction of the reservoir

A complete reservoir can be made from 20 cm of large bowel or 40 cm of small bowel (Fig. 9.4a).

In constructing a reservoir from scratch, it is convenient to use the cecum and ascending colon, but any large bowel or ileum can be used. It is essential to have a reservoir that will ultimately have a low pressure. This is best achieved by de-tubularization and re-configuration as close to a spherical shape as is possible. For large bowel, a U form is suitable. For ileum, more complex re-configuration is needed to produce a sphere.

Colonic reservoirs The length of colon to be used is marked out and its blood supply defined (Fig. 9.4b and c). The right colon and cecum are best mobilized on the ileocecal artery with or without the right colic. The transverse colon is usually supplied by a complex of two or more vessels referred to as the middle colic. The sigmoid colon usually has a complex of vessels arising from the inferior mesenteric artery; the site of division from the upper rectum must be chosen with care as the anastomosis between

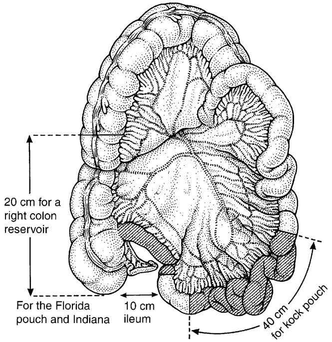

Fig. 9.4a

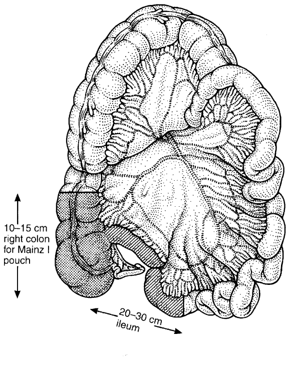

Fig. 9.4b

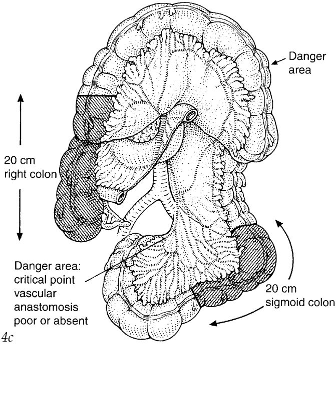

Fig. 9.4c

the sigmoid and rectal systems can be poor.

The blood supply of the descending colon is the most vulnerable, and anastomoses in this area have a high risk of fistula formation. This author uses descending colon only as a last resort.

Ileal reservoirs The blood supply of the ileum is varied and excellent, and it is seldom difficult to select a suitable segment. Although the normal ileum has great adaptive power, it is best to avoid using the terminal 15 cm of ileum, which are very important for bile salt and vitamin B_{12} absorption.

Bowel mobilization Once an area has been selected, the vessels in the mesentery are ligated and divided. Although this may be carried out in the standard way with clamps and ties, small metal clips (Ligaclips) are often quicker and neater. Soft bowel clamps are applied at the resection sites, and abdomen is carefully protected with towels. The bowel is divided.

Gastrointestinal continuity is then restored (Fig. 9.5a–f). This anastomosis may be made in the standard way with two layers of sutures. Again, however, metal staples are often easier. The two lengths of bowel are tacked together over 5 cm on their anti-mesenteric borders. A side-to-side anastomosis is made with a GIA staple gun. The open end is often closed with a T-55 or T-90 gun. The defect in the mesentery is closed to prevent an internal hernia.

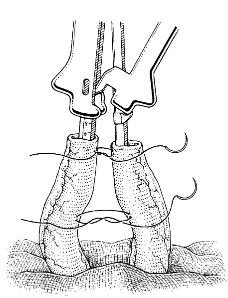

Fig. 9.5a

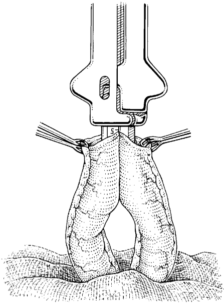

Fig. 9.5b

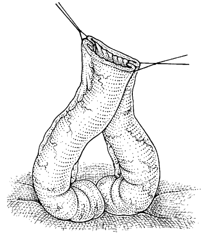

Fig. 9.5c

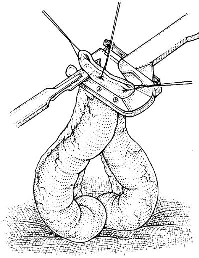

Fig. 9.5d

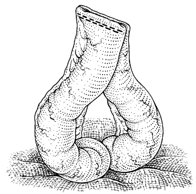

Fig. 9.5e

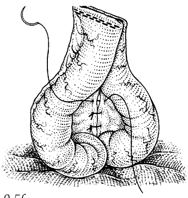

Fig. 9.5f

Re-configuration of the reservoir segment The isolated bowel segment is copiously washed out with warm physiological saline.

A U-shaped pouch is most easily made with absorbable staples. The bowel is held in a U configuration with Babcock clamps (Fig. 9.6a). A GIA gun with polyglycolic acid staples is positioned with one arm down each limb of the U (Fig. 9.6b). With tension applied in either direction via the Babcock clamps to stretch out the bowel, the gun is closed and fired. This will complete almost half of the anastomosis (Fig. 9.6c).

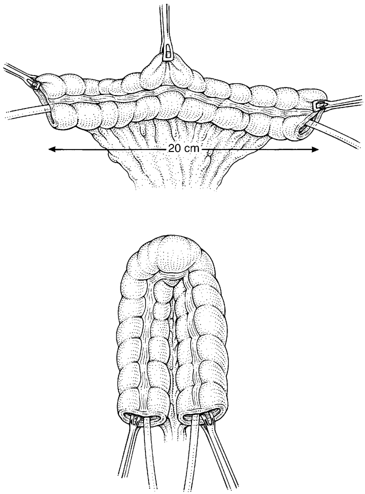

Fig. 9.6a

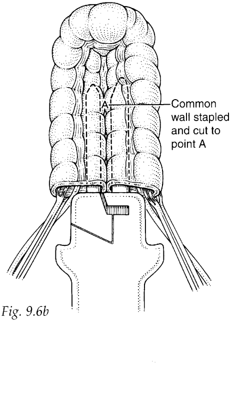

Fig. 9.6b

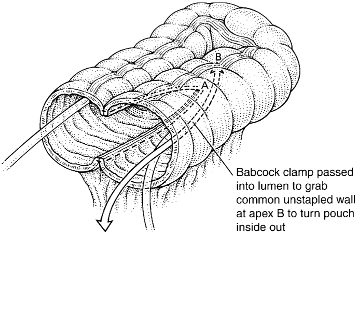

Fig. 9.6c

The reservoir is then turned inside out to expose the upper common wall of the 'reservoir to be'. Another stapler is positioned in a direction opposite to that of the first (Fig. 9.6d). When this is fired, there will be only a short length of undivided common wall between the two staple lines (Fig. 9.6e). This is carefully divided with scissors, the two resulting defects being closed with continuous 2/0 Vicryl (Fig. 9.6f).

The ureters and conduit are anastomosed to the reservoir with inverting Vicryl before the open end is closed. This author uses a size 18F Foley catheter in the reservoir as well as a 12F or 14F Jacques catheter in the conduit.

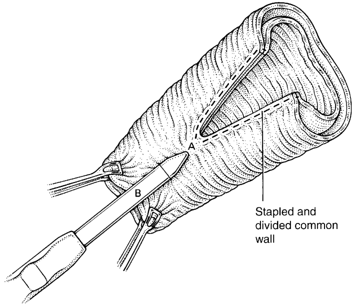

Fig. 9.6d

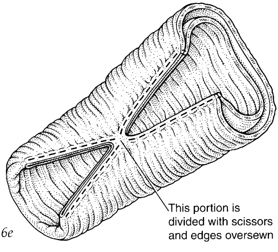

Fig. 9.6e

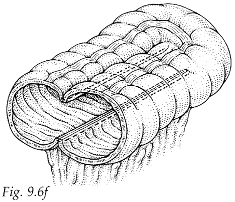
Fig. 9.6f

Ileum requires a more complex reconstruction, and several variations have been described. At least two folds are required, but there is little to choose between the variations. For the first fold, the bowel is tacked together in the shape of a U or an S. The bowel is then opened along its anti-mesenteric border. The 'back wall' is then sutured with continuous 2/0 Vicryl. At this stage, it is best to anastomose the ureters and the conduit (Fig. 9.7a and b). To close the reservoir, a U is folded from bottom-to-top (Fig. 9.7c) and an S is folded from

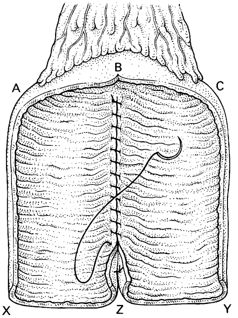

Fig. 9.7b

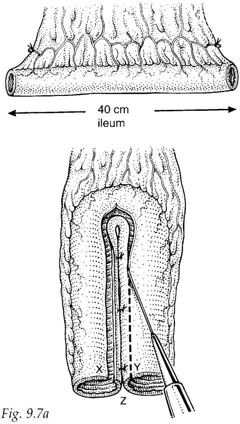

Fig. 9.7a

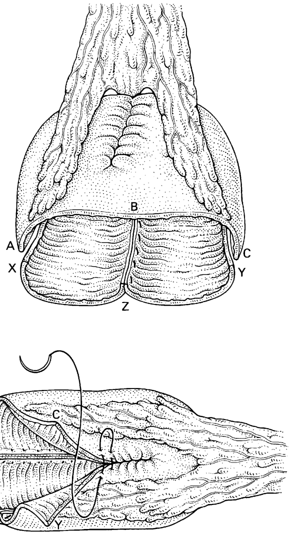

Fig. 9.7c

side-to-side (Fig. 9.7d). Closure is completed using continuous inverting Vicryl. The same drains are used.

When there is an existing ileal loop to be used, it is mobilized on its pedicle and opened longitudinally. It is then augmented with further bowel sutured on face-to-face (Fig. 9.8). An existing colonic loop is treated in the same way. If the original loop is large enough, especially if it was formed with later conversion in mind, further augmentation may not be necessary.

Although any bowel can be used to form the pouch, care must be taken in the arrangements in girls. The pedicles must be so positioned that the gravid uterus can enlarge without obstruction and without kinking them. It is best to position the pouch and pedicles on one side of the abdomen and as posteriorly as possible.

Formation of the Mitrofanoff tunnel

As the outer end of the implantation must lie immediately under the cutaneous stoma site, the position of the tunnel on the reservoir must be chosen with care. An area of the reservoir that will lie anteriorly and conveniently for the chosen cutaneous site is selected.

The Mitrofanoff system depends for continence on the construction of a tunnel about 5 cm long through which the chosen conduit passes. The great versatility of this approach is that virtually any conduit can be tunnelled into virtually any reservoir. The tunnel may be made either between the mucosa and the muscle of the reservoir, or by wrapping the full thickness of the reservoir wall around the conduit, which is similar to the

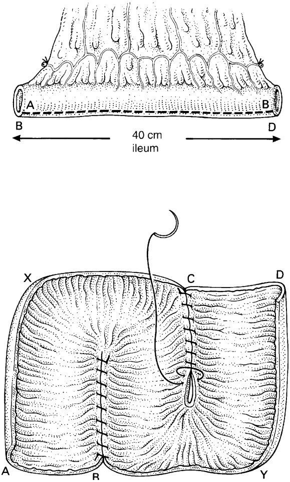

Fig. 9.7d

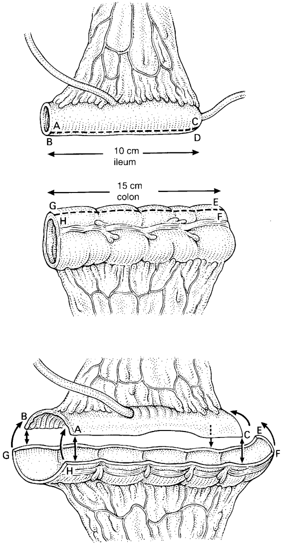

Fig. 9.8

Nissen fundal plication used to prevent gastro-oesophageal reflux. The former is better with a thick-walled reservoir such as colon and a conduit with little fat in the mesentery; the latter is used for thinner-walled reservoirs and fatter mesenteries.

For a tunnel, this author usually approaches from the outside of the reservoir. The muscle layer is incised over about 5 cm, if possible in one of the tenia coli (Fig. 9.9a). It is very important to form a 'shoulder' at the outer end in order not to constrict the conduit when the tunnel is closed. The muscle is mobilized as two lateral flaps, being careful to preserve the vessels that lie in the layer between muscle and mucosa.

A small opening is then made in the mucosa at the inner end of the tunnel. The conduit is anastomosed end-to-side with the mucosal opening using 4/0 Vicryl (Fig. 9.9b). The muscle layer is then closed with non-absorbable sutures such as 3/0 black silk. It is best to do this closure with the catheter in the conduit (Fig. 9.9c and d). Ease of catheterization should be checked at each stage.

For a wrap, the conduit is anastomosed end-to-side with the reservoir. The full thickness of the reservoir wall is then sutured around the conduit with 3/0 black silk sutures to form a 5 cm tunnel (Fig. 9.10a and b). If the mesentery is very bulky, some of its fat may be teased off, or small windows may be made in it through which the reservoir wall may be sutured.

The Kock nipple

The Kock nipple has been widely used for continent diversion, but this author's experience with it has been disappointing. There seems to be a long learning curve for its construction

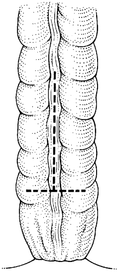
Fig. 9.9a

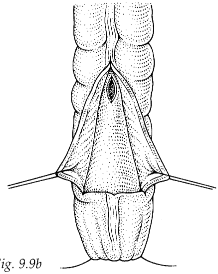
Fig. 9.9b

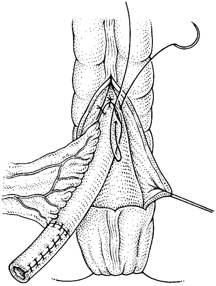
Fig. 9.9c

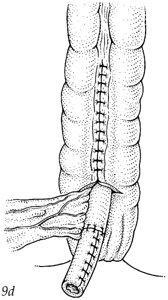
Fig. 9.9d

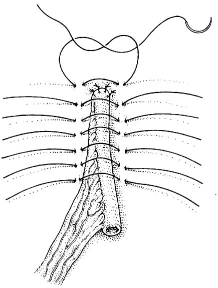
Fig. 9.10a

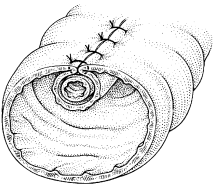
Fig. 9.10b

so that it is unsuitable for the surgeon who does only a few continent diversions. It also appears that the pressure within the nipple (analagous to the urethral pressure profile) is only slightly higher than the pressure in the reservoir, so that the system is only just continent. Nonetheless, more patients have probably had a Kock valve than any other system, if only because it is the oldest technique and has been used by the surgeon with the largest personal experience of continent diversion in adults.

The Kock pouch is a complete continent diversion consisting of two nipples (one for continence and one to prevent ureteric reflux) in a reservoir. It is formed from a continuous 72 cm length of ileum. The term 'hemi-Kock' is applied to a reservoir with the anti-reflux nipple used as an orthotopic bladder replacement anastomosed to the membraneous urethra. The nipple may, however, be used with any reservoir, the technique of its formation being the same. In this description, it is assumed that a complete Kock pouch is to be made.

The chosen 72 cm length of ileum is isolated from the gastrointestinal tract. The 16 cm at either end will form the nipple and are marked out as shown in Fig. 9.4a above. The mesentery is dissected off the ileum over about 7–8 cm, and the bleeding points are controlled with diathermy. The ileum is then intussuscepted using a pair of Babcock clamps (Fig. 9.11). The intussusception, which has a distressing tendency to undo itself, is maintained in three ways.

1. The nipple is fixed with metal staples. The main published descriptions recommend three rows of staples from a T-55 staple gun. The tissue pin must first be removed from the cartridge in order to avoid the formation of a fistula. This author has, however, found this to be inadequate and prefers to use a 7 cm GIA stapler from which the manufacturer has removed the knife.
2. The nipple must be fixed to the inside of the reservoir. For this, the mucosa of the reservoir and the adjacent mucosa of the nipple mu[st] be extensively scarified with diathermy. The two are then fixed together with a further row of staples from the T-55 stapler or w[ith] non-absorbable GIA staples.
3. The serosal surfaces of the outer outer end of the nipple are fixed with a ring of polyglycolic acid mesh.

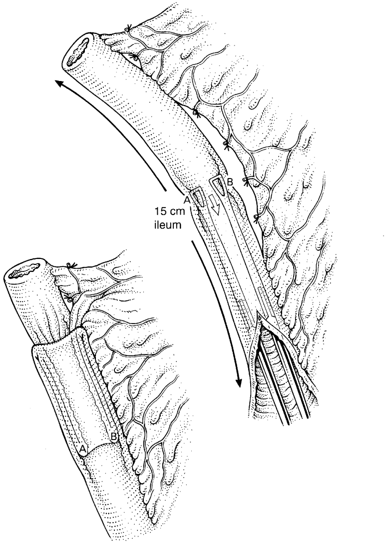

Fig. 9.11

The ileocecal valve

The ileocecal valve was used in the early 1950s in attempts to make a continent diversion. Although the initial reports were quite encouraging, it soon became clear that continence was, at best, tenous. The main reason was that the reservoir was made of straight right colon and was of too high a pressure. Second, the valve itself was not always as continent as was supposed. The technique has now been extensively modified and is known as the Indiana. It must, almost by definition, be a complete system of right colonic reservoir and continence valve.

Ten centimetres of terminal ileum and 20 cm of right colon are isolated from the gastrointestinal tract. The right colon is incised on its anti-mesenteric border. The reservoir will later be formed by folding the colon and closing transversely.

In the meantime, the continence mechanism is formed. The ileum is tapered over a 12F or 14F catheter in very much the same way as described above. The tapering is continued right down to the ileocecal valve. (Fig. 9.12a–c).

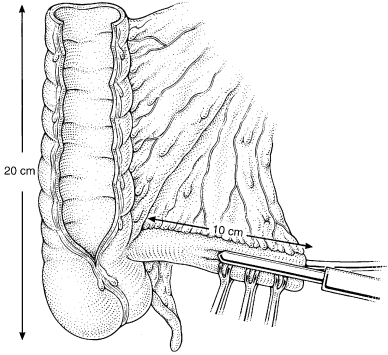

Fig. 9.12a

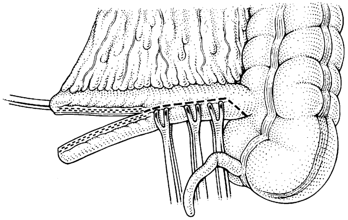

Fig. 9.12b

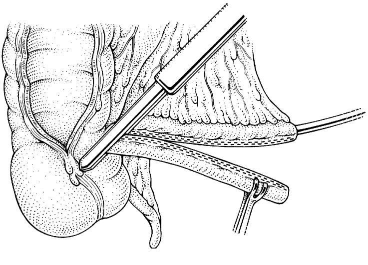

Fig. 9.12c

The valve is then reinforced by imbrocating the cecal wall around the ileum. In the original description, this was done with three or four non-absorbable sutures (Fig. 9.12d and e), but this may not always be sufficient to maintain continence. Sufficient sutures can therefore be used to create a tunnel 4–5 cm long, similar to that of a Nissen fundal-plication. In doing this, it is essential to check that catheterization does not become obstructed, particularly at ileocecal valve level. It must be said that if this full tunnel is made, there is little difference from the Mitrofanoff principle.

As the ileocecal valve is of dubious power on its own, several other systems have been devised to augment it.

Mainz I pouch In this system, the reservoir is formed from 20–30 cm of terminal ileum and 10–15 cm of right colon (Fig. 9.13a). The ileal segment is opened on its anti-mesenteric border and formed into a U shape. The colonic segment is also opened on its anti-mesenteric border (Fig. 9.13b). The reservoir will eventually be

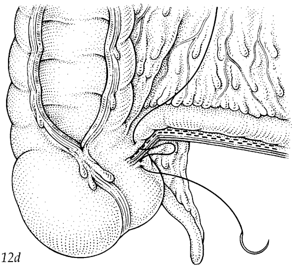

Fig. 9.12d

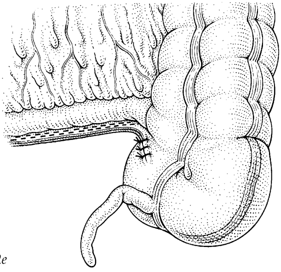

Fig. 9.12e

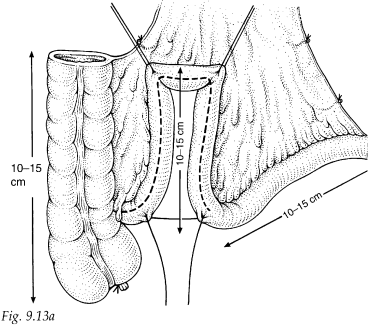

Fig. 9.13a

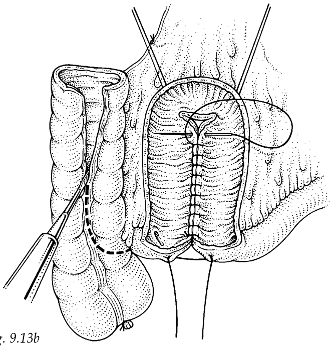

Fig. 9.13b

formed by suturing these two segments together face-to-face (Fig. 9.13c).

The continence system is formed by augmenting the ileocecal valve with a Kock nipple. The mesentery is detached from about 7 cm of the most proximal segment of ileum. Two Babcock forceps are passed through the ileocecal valve and into this most proximal ileal segment (Fig. 9.13d).

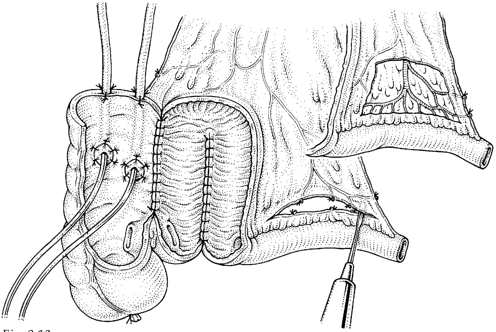

Fig. 9.13c

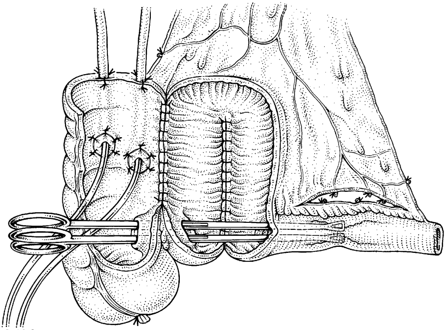

Fig. 9.13d

The ileum is then intussuscepted through the valve. The nipple thus formed is fixed with staples as for the Kock nipple (Fig. 9.13e–g). The reservoir is then closed with absorbable sutures. The open end of the proximal ileum forms the stoma.

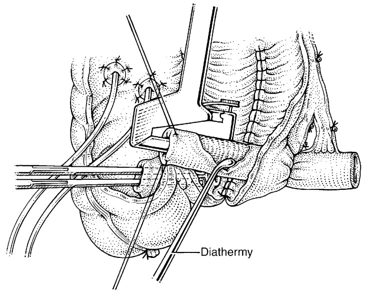

Fig. 9.13e

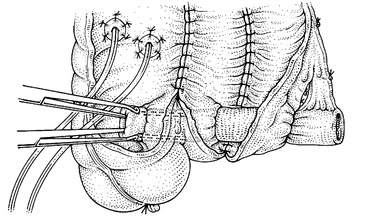

Fig. 9.13f

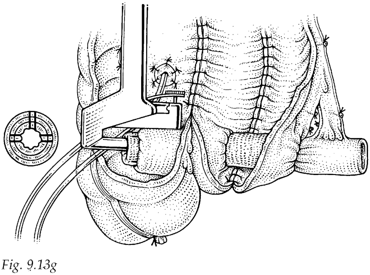

Fig. 9.13g

The Florida pouch This too is a complete continent reservoir system. The reservoir is formed from 20 cm of de-tubularized right colon. The ileocecal valve is reinforced with a Kock nipple formed from terminal ileum. In this, it is almost identical to the Mainz system except in the relative lengths of ileum and colon used (Fig. 9.14a and b).

Comparison of passive continence systems

There has been no formal comparison of the many systems available to create passive continence. The choice between the three broad approaches described above must be made according first to that which is available in the patient and then by the surgeon's own preference. Within the broad principles, the differences between the various named systems is often too small to alter the result. Whichever is used, the surgery must, however, be carried out with the greatest care and attention to detail.

The author's own experience is that the Mitrofanoff produces reliable continence in nearly all patients, having been less successful with the Kock nipple or the ileocecal valve. Personal work has shown that the pressure within the lumen of a Mitrofanoff tube is much higher than that in the lumen of a Kock nipple, corresponding to more reliable continence. Other authors have reported good continence with the other systems, although not in comparative trials.

Anastomosis of the ureters

The ureters are anastomosed last. The site is selected when it has been seen how the reservoir will lie in the abdomen. The ureters may be buried in submucosal tunnels in the standard manner unless they are grossly dilated and poorly peristaltic. If there is an inadequate length on one side, a transuretero-ureterostomy is carried out. The anastomosis can be protected with splints. If the ileocecal valve is a part of the reservoir it is convenient to use it to prevent reflux. The ureters are anastomosed end-to-end to the tail of the ileum.

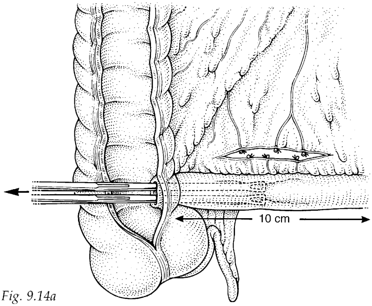

Fig. 9.14a

There has been much debate at clinical meetings over the need to prevent reflux in these low-pressure reservoirs. The general view is that this is desirable but not essential: that experimental information which is available in the literature tends to support the need to prevent reflux. Nonetheless, obstruction is far more damaging than reflux to the kidneys.

Formation of the continent cutaneous stoma

The long-term success of the procedure depends on this part of the operation more than any other. If the conduit cannot be catheterized, the whole system is useless.

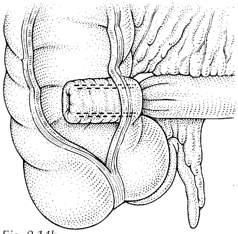

Fig. 9.14b

The site of the stoma is selected entirely for the patient's convenience. For the normally mobile child, this author puts it as low as possible, preferably below the normal 'bikini line'. There is great enthusiasm in the United States for the use of the umbilicus, which invariably provides a good passage, at least down to the rectus sheath. Although this is not an option for the exstrophy patient, it is an excellent general choice.

In patients in wheelchairs, there is a tendency for the spine to become more twisted with time, the abdomen becoming more compressed. Eventually, with increasing age and obesity, most of the lower abdomen becomes hidden from the patient's field of vision. In this group, it is usually better to put the stoma close to the xiphisternum.

The distal end of the conduit is brought through the abdominal wall obliquely to make the track as straight as possible. There must be no sharp bends between the skin and the pouch. The cutaneous end is spatulated and a triangular skin flap (2–3 cm) sutured into the spatulation (Fig. 9.15a). This allows the minimum of mucosa to be visible, forms a neat funnel for catheterization and may contribute to the prevention of stenosis. If the conduit is too short, a rectangular skin flap is raised. It may be rolled over the catheter to make a tube and sutured to the spatulated end of the conduit to lengthen it (Fig. 9.15b and c).

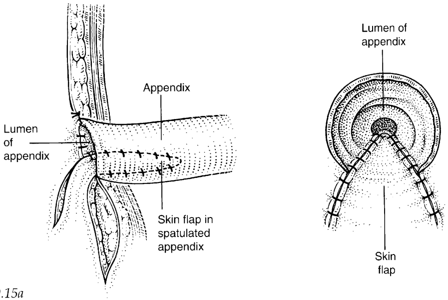

Fig. 9.15a

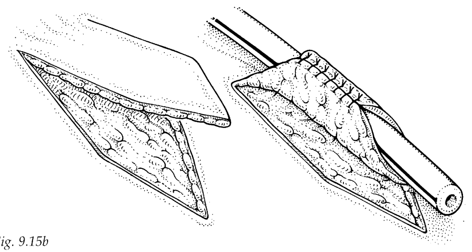

Fig. 9.15b

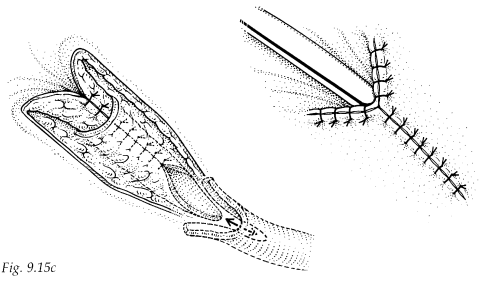

Fig. 9.15c

If the umbilicus is to be used, its deep attachment to the rectus sheath must first be divided. A stay suture is then placed on its cranial aspect a few millimetres from its apex. An incision is made around a half of its circumference so that an inferiorly based flap is formed. This flap is sutured into the spatulated end of the conduit (Fig. 9.16).

Whatever the type of stoma, this author uses an absorbable suture that generates the minimum of local inflammation: 5/0 PDS is suitable.

It has not been this author's practice to bring the stoma to the position of the normal external urinary meatus in either sex. The female urinary meatus is a difficult target for self-catheterization, especially in those with imperfect manual dexterity. In the male, although some surgeons indeed have no difficulty in constructing an easily catheterizable neo-urethra, this author's own results have been disappointing. In patients who have never remembered voiding through the urethra and who are in any event going to self-catheterize, it appears best to put the stoma in the most accessible site.

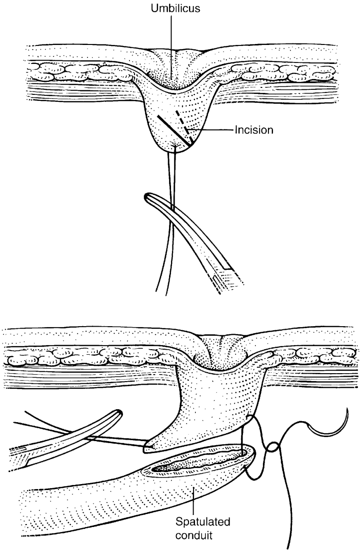

Fig. 9.16

Postoperative care

At the end of the operation, there will be a great many abdominal tubes and drains. It will be helpful to the recovery room nurses to have a diagram and for each tube to be labelled; those which may legitimately transmit urine should be distinguished from the others (Fig. 9.17).

Patients require a careful monitoring of fluid balance, which generally means the assignment of a special nurse. There is a heavy and continued fluid loss in the first 24 or 48 hours, so blood pressure, pulse, central venous pressure and urine output must be carefully watched. The most common problem is underfilling of the circulation.

The gastrostomy (instead of nasogastric) tube is a great boon. Patients may drink as soon as they wish as the fluid will come out via the gastrostomy. The balloon of the Foley catheter is occasionally passed into the duodenum so that drainage stops and the patient feels nauseated. If this complication is suspected, the ballon is deflated completely and 1–2 ml of air is then put back once the duodenum has disengaged itself and flow been re-established.

Prophylactic antibiotics are stopped after 48 hours regardless of the patient's temperature. If there is a fever at this time, blood and urine are sent for culture after 24 hours and different antibiotics started on the basis of the results. There is no point in continuing the prophylactic antibiotics in the face of a rising fever because they are obviously not doing any good. Antibiotics should only be used 'blindly' if the patient is very ill.

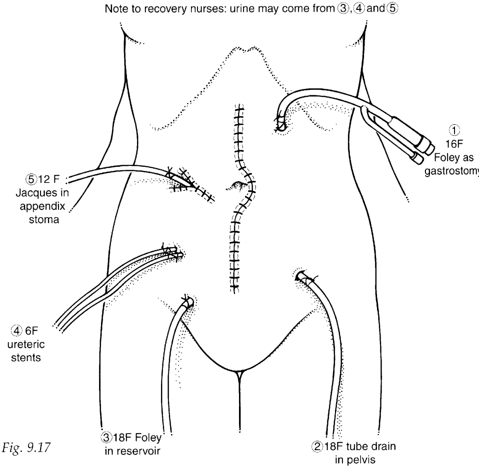

Fig. 9.17

The catheters in the pouch (one through the conduit and one as a 'vesicostomy') must be irrigated twice a day so that they do not bcome blocked with mucus. Ureteric stents are removed on the 7th day. The 'vesicostomy' is clamped on the 10th day and removed on the 11th if no untoward event has occurred. The gastrostomy tube is removed after the 10th day, just before the patient is discharged.

The patient stays at home with a catheter in the stoma on free drainage until 6 weeks post-operation. The mucus usually needs to be washed out twice daily with sterile saline, but only twice weekly once self-catheterization has been established.

The patient is then re-admitted to lea self-catheterization and to expand the pouch. This seldom takes more than 1 days. The reservoir catheter is clampe for increasing periods, starting at 2 hours. When it can tolerate 400 ml without pain, the catheter is removed and self-catheterization started. The catheter should initially be passed every 2 hours, but with increasing confidence this is done 3-hourly and then 4-hourly by day, the catheter being left on continuous drainage at night.

The patient goes home to practise thi regime. After a week, it should becom apparent whether the pouch has sufficient sensation to alert the patien to the need for catheterization. If so, the patient catheterizes according to need day and night. If there is insufficient sensation, catheterization must be carried out by the clock every 4–6 hours. The capacity will eventually reach 600–800 ml, when the catheter can be left out at night. I still advise patients to catheterize at least every 4–6 hours by day to lessen the risk of infection. Most patients sleep through the night.

Before final discharge, the patient is shown the area of the abdominal wall where the pouch lies superficially. Should he or she be unable to catheterize and go into acute retention, the pouch may be aspirated with a needle and syringe at this site. Patients are also advised to wear a bracelet or necklace describing their diversion in case they should be involved in an accident.

Complications

Early

It will be evident from the extensive surgery that there will be a significant complication rate. It is difficult to put a figure on the incidence as both the children and the surgery are highly variable, but major complications occur in about 10% of patients.

In the gastrointestinal tract, almost every patient will have a paralytic ileus for several days. At what point this becomes a complication is a matter of opinion, but the gastrostomy tube certainly makes its management much easier. Chest complications are particularly common in spina bifida patients. Vigorous physiotherapy should be used from the 1st postoperative day.

In the reservoir, the continous drainage should minimize the risk of leaks. The mucus that is produced in large quantities by the intestinal mucosa is liable to block the catheters. For the first few days the urine should obviously drain through the ureteric stents. However, it sometimes by-passes the stents and drains into the reservoir. Therefore, careful attention must be paid to the reservoir drains to prevent them blocking.

When the stents are removed, the ureteric anstomoses may occlude. Any fever or renal pain at this time should be investigated by ultrasound. It is rare for obstruction to be complete, and it usually seems to be self-limiting. A nephrostomy tube is occasionally needed and even more rarely the antegrade positioning of a double-J stent across the anastomosis.

Late complications and general consequences of continent diversion

The problems related to the removal of so much intestine from the gastrointestinal tract are important but beyond the scope of this chapter. In otherwise normal children, they are, in practice, few and far between.

The storage of urine in intestinal reservoirs is fraught with problems, more of these becoming apparent with longer experience.

Storage and emptying In the short term, it has been shown that continent diversions can store urine and can be emptied by ICSC. There are, however, no long-term results. Although various forms of enterocystoplasty have been carried out for 60 years or more, it is only in the past 15 years that they have been done in any great number. It is already apparent that there is a constant need for review and surgical revision. In general, once continent, they remain continent, although this author has had one Kock nipple that became wet after 7 years.

This author's own experience with the Mitrofanoff continence system, much in line with that of others, is that no more than 5% of patients are incontinent. Once established, continence is maintained, only 1 of more than 100 patients in a personal series developing late, complete, incontinence. Incontinence occurring early is always the result of faulty implantation and requires surgical revision. Late incontinence (or minor leakage) may be caused by 'instability' of the reservoir and may respond to anticholinergic drugs. The author has twice had to augment a reservoir with further bowel for intractable instability.

The opposite problem is more common: difficulty with catheterization, usually caused by stenosis at skin level, occurs in up to 30% of patients, usually in the first year. Although most initially respond to simple dilatation and catheterization for 10 days, three-quarters will eventually need re-formation of the stoma with a new skin flap. The emergency treatment for a failure of catheterization is transabdominal puncture and drainage under ultrasound control.

Reservoir rupture The incidence of spontaneous rupture varies between different units and is said to be more common in reservoirs containing some colon. When it does occur, the consequences are devastating. There may be a delay in diagnosis, although the history of sudden abdominal pain and diminished or absent urine drainage should make it obvious. The patient rapidly becomes very ill with generalized peritonitis. A 'pouchogram' is commonly normal. Diagnosis is best made by ultrasonography and the peritoneocentesis of fluid collections.

If the condition is diagnosed early, catheterization and broad-spectrum antibiotics may lead to recovery. If the patient fails to respond rapidly on this regime, or if there has been a delay in diagnosis, laparotomy should be performed at once. Up to a quarter of patients die, usually through delayed or inadequate management. Patients and their families should be warned of this possible complication and advised to return to hospital at once for any symptoms of acute abdomen, especially if the reservoir stops draining its usual volume of urine.

Pregnancy When carrying out a reconstruction in females, it is essential to bear future pregnancy in mind. Pregnancies after any type of removal of diversion or lower tract reconstruction with bowel are so far rare. Published experience suggests that they may be complicated and require the joint care of obstetrician and urologist. Particular problems include upper tract obstruction and changes in continence as the uterus enlarges.

Metabolic changes Metabolic changes are common when urine is stored in intestinal reservoirs and must be carefully monitored. In experimental studies of the handling of electrolytes instilled into intestinal reservoirs, all patients are found to absorb sodium and potassium to a variable extent. A third of all patients (but 50% of those with an ileocecal reservoir) have hyperchloremia. Some authors have found all patients to have abnormal blood gases, the majority having metabolic acidosis with respiratory compensation. The findings are unrelated to renal function or the time since the reservoir was constructed.

In 183 patients of all ages at St Peter's Hospitals, London, who underwent any form of enterocystoplasty, hyperchloremic acidosis was found in 25 (14%) and borderline hyperchloremic acidosis in 40 (22%). The incidence was lower in reservoirs using ileum as the only bowel segment compared with those containing some colon (9% versus 16%). When arterial blood gases were measured in 29 children from this group, a consistent pattern was not found, although the majority were normal.

There are conflicting reports on the incidence of metabolic abnormalities from other units, albeit often considering a smaller number of cases. The incidence of hyperchloremic acidosis ranges from zero to 52%. The length of follow-up, the parameters tested, the size and type of reservoir and its continence all seem to affect the metabolic outcome. It is prudent to monitor patients for metabolic abnormalities and to treat them when these are found.

Renal function In the follow-up data available so far, continent diversion seems not to have affected renal function. When function has improved after such surgery, it is likely to be the result of eliminating obstruction or a high bladder storage pressure. Experiments in animals tend to support this clinical observation.

In the longer term, the renal deterioration that has been found has been related to obstruction, reflux and stone formation. These complications occur at the same rate as is found in patients with ileal conduits.

Infection and stones Stones are a particular problem in reservoirs containing metal staples. The incidence in the Kock pouch is about 17%. In those who form stones, the recurrence rate is 22%. Otherwise, stones in reservoirs are associated with infection and retained mucus. In this author's patients, 28% have troublesome urinary infections, and a further 20% have an occasional infection. Renal stones have developed in 1.6% of patients of all ages and reservoir stones in 12%. A literature review suggests that, in patients with all types of reservoir, renal stones occur in about 7% and pouch stones in 28–52% over a follow-up of up to 4 years. On analysis, nearly all are triple phosphate stones.

Although many of these incidence figures refer to reservoirs based on the bladder, two points are clear. The more that stones are looked for in bowel reservoirs, the more they are found. Second, the cause is infection, with or without retained mucus. In this author's own practice, the incidence of stones appears to have fallen since greater attention has been paid to avoiding the known predisposing factors. In particular, patients must be taught to wash-out their reservoir vigorously once or twice a week to avoid the retention of mucus.

The significance of infection on its own is uncertain. It has been said that such infection is harmless except in the presence of high-grade reflux, when 60% of patients suffer renal damage. In ileal conduits, however, persistent infection, especially with *Escherichia coli*, has been associated with progressive renal scarring, early evidence suggesting that the same will occur in continent reservoirs.

Growth The most worrying consequence of enterocystoplasty in children has been a delayed growth in height. In 60 children with any form of enterocystoplasty at Great Ormond Street and St Peter's Hospitals, London, a delay in linear growth was found in 20%, whereas the growth in weight was normal. There were no metabolic or infective complications to account for this finding. Further follow-up has shown that catch-up growth occurs. Enterocystoplasty does not affect ultimate height.

Cancer It is the constant fear of surgeons who carry out these operations on the young that history will repeat itself: that the bowel segments will develop neoplasia in the same way that the ureterosigmoidostomy does. Animal evidence suggests that the fecal and urinary streams must be mixed for neoplasia to occur. However, if it is chronic mixed bacterial infection, rather than the feces *per se*, that is required, the bowel urinary reservoir may be at risk.

In patients with colonic and ileal cystoplasties, a high level of nitrosamines (similar to that found in ureterosigmoidostomy patients) has been found in the urine of most patients examined. Biopsies of the ileal and colonic segments have shown changes similar to those which have been found in ileal and colonic conduits and in ureterosigmoidostomies. More severe histological changes and higher levels of nitrosamines correlate with heavy mixed bacterial growth on urine culture.

Single cases of pouch neoplasia have been reported: in a review of the literature, 14 cases were identified. Special features can be found in nearly all the cases. Ten patients, for example had been reconstructed for tuberculosis, 4 tumors were not adenocarcinomas 1 patient had a pre-existing carcinoma, and 6 patients were over 50 years old. The cancers reported in enterocystoplasties occurred at a mean of 18 years from formation. This is a few years earlier than the mean time at which malignant neoplasms are seen in ureterosigmoidostomies. The incidence to date has been too small to warrant special screening.

Although there is some doubt surrounding the role of nitrosamines in the causation of ureterosigmoid neoplasia, cancer must remain a cause for great vigilance in the follow-up of patients. It is interesting to note that no cases have been reported from units routinely performing a large number of these operations.

Variations on the ureterosigmoidostomy

The disadvantages of the ureterosigmoidostomy in children have in general been overwhelming. In particular, the high risk of carcinoma in patients followed into adult life has caused the procedure to be abandoned. Reconstruction of the bladder is a more attractive alternative.

It is, however, becoming apparent that the complication rate is high with continent diversion. Many of the complications seem to be the same in both type and frequency as those seen in ureterosigmoidostomy. If it were not for the cancer risk, there would be little to choose between the two approaches. Should it turn out that there is a significant risk of cancer in all bowel reservoirs, the balance of advantage would change.

Modified ureterosigmoidostomy, particularly the Mainz II, has been reported in recent series to have good results. Two things can be said in favor of even the traditional ureterosigmoidostomy. The first is that there is a sphincter so that patients can 'void' spontaneously. The second is that those with an established and continent ureterosigmoidostomy never want to change it for a different system, even if they do develop cance

In this author's unit, all such patients have a flexible colonoscopy once a year. The incidence of all types of neoplasia is 22% at 30-year follow-up, all cases having been identified while the tumor was still localized. The tumors have been excised and the urinary tracts have usually, at the patient's insistence, been reconstructe with another ureterosigmoidostomy.

Although it is currently difficult to recommend the ureterosigmoidostom for children in the Western world, there appears to be the beginning of a renaissance for this procedure. In poorer parts of the world, the system remains popular. It will not be considered here in detail. Surgeons who are familiar with the techniques of continent diversion should, however, have no difficulty in constructing the new variations on the ureterosigmoidostomy.

Two recent variations have attempted to resolve the known drawbacks of ureterosigmoidostomy, in particular the poor continence. The problem wit the classical ureterosigmoidostomy lie not with the anal sphincter but with the rectum, its storage pressure being much too high. The new variations produce a low-pressure reservoir.

In the Kock–Ghoneim system, the urine is stored in the lower rectum, which is augmented with ileum. This produces a reservoir of good capacity and low pressure. The stool passes from the sigmoid colon into the rectu through an intussuscepted Kock nipple at the colorectal junction. The confinement of the urine within the rectum is said to reduce the risk of hyperchloremic acidosis and possibly of carcinoma. The main disadvantage of this operation is the need for a temporary colostomy.

In the sigma-rectum pouch described by Hohenfellner (Mainz II), the lower sigmoid and upper rectum are opened longitudinally and closed together side-by-side in a U formation. This reconfiguration also produces a low-pressure, high-volume reservoir (Fig. 9.18).

The two systems may be combined. The nipple in the colon is retained from the Kock–Ghoneim method, but the reservoir is enlarged by the formation of a Hohenfellner sigma-rectum pouch rather than by using isolated ileum.

Short-term results

The surgical results of the ureterosigmoidostomy are well known. The introduction of an anti-reflux nipple greatly reduces the incidence of upper tract infection. Subsequent radiological testing of the anti-reflux mechanism has proved difficult: even when watery contrast put into the rectum does not reflux, gas can sometimes be seen in the renal pelvis, indicating that some reflux is occurring.

The general improvements in intestinal and urological surgery, anesthetics and antibiotics have considerably reduced the short-term complication rate. The new variations have been carried out in such small number that a true idea of early complications cannot yet be formed. They are, however, unlikely to be different from those of the more conventional operation.

Long-term results

Psychosocial aspects The effluent of mixed urine and feces has a very offensive smell. It is not a suitable

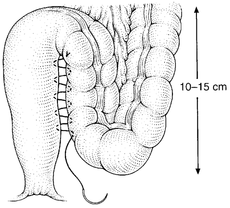

Fig. 9.18a

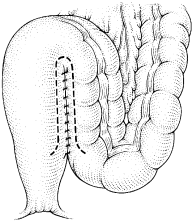

Fig. 9.18b

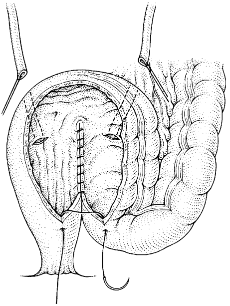

Fig. 9.18c

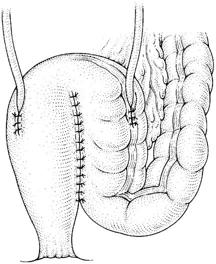

Fig. 9.18d

diversion for children from socially deprived families for whom no private lavatory is available. Likewise, the misery of incontinence is worsened by ostracism owing to the smell.

Even with a traditional ureterosigmoidostomy made in childhood, continence usually develops. Those who fail to become dry are given a different diversion; thus, incontinence is rare in long-term follow-up. Both of the new methods provide continence in almost 100% of cases.

Renal consequences The renal and metabolic problems were a cause of the poor reputation of the ureterosigmoidostomy. Although the metabolic problems are better understood and antibiotics have lessened the consequences of infection, the long-term effects on the kidneys are still worrying. It is unlikely that the new systems will be any different.

Histological changes The presence of urine within the intact sigmoid colon does not cause major histologic changes on conventional examination. At laparotomy, however, the bowel is seen to be dilated, and the wall is thin. On colonoscopy, the ureteric orifice usually looks like a small cherry 3 or 4 mm in diameter, sometimes being a flat opening that can be hard to see.

On the hematoxylin and eosin staining of biopsies, normal colonic epithelium is seen abutting normal transitional cell epithelium with inflammatory cell infiltrate. The orifice may become sufficiently prominent and inflamed to earn the title of 'inflammatory polyp'. Even in longstanding anastomoses, the junction between the two epithelial types is abrupt.

Histochemistry does, however, show differences from normal. The significance of these changes is unknown. They could be an innocent change in colonic mucosa caused by the presence of urine, or they could indicate that urine, almost invariably, produces a premalignant change of long latency. It is not seen in colonic conduits and has not been looked for in intestinal reservoirs.

Ureterosigmoid cancer The most important change that occurs in ureterosigmoidostomies is neoplastic. The first case of carcinoma of a ureterosigmoidostomy was reported in 1929, but little interest was taken in this complication until about 25 years ago. Only 18 adenomas or adenocarcinomas were recorded in the literature up until 1970. Presumably few patients with ureterosigmoidostomies lived long enough to develop the tumors.

The tumor arises at the anastomosis or very close to it in 90% of cases. In patients diverted for benign disease, 95% of tumors are adenomatous (adenomas or adenocarcinomas), but one transitional cell and one lymphomatous tumor have been reported. The most important etiological factor, both experimentally and clinically, is the mixture of urine and feces at the anastomotic site. Urine can be found throughout the colon, but tumors do not occur as a result.

It is not known which component of feces is required to precipitate the malignant change. There is some evidence to suggest that it lies in the bacterial flora, especially the anerobes. If this is the case, there is probably a similar (albeit lesser) risk in enterocystoplasties of all types. If the carcinogen is formed from a component found only in feces (such as bile acids), the risk is effectively unique to ureterosigmoidostomies.

Aside from the risk of cancer, the complications of modified ureterosigmoidostomy are likely to be the same as for continent diversions, except that stones should not be a problem.

If the final analysis is based on the cancer risk, no informed decision can be made at this time. It is, however very easy to endoscope the rectum and very difficult to endoscope the reservoir of a continent diversion.

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Chapter 10

The 'VQZ' plasty for catheterizable stomas

Philip G. Ransley

The 'VQZ' (V-Quadrilateral-Z) plasty for the cutaneous opening into a Mitrofanoff or antegrade colonic enema channel provides several significant advantages over the simple flush stoma or the V flap technique. First, it is discreet and does not attract attention. Second, there is no exposed mucosa, which, apart from its unsightly appearance, may secrete mucus and bleed onto clothing. The mucocutaneous junction is a very long suture line that is not circumferential, thereby reducing the risk of stenosis.

The V flap (Fig. 10.1) is created by a sharp incision through the skin and subcutaneous tissue. It is important to site the flap so that the *base* of the triangle rather than its tip is at the desired stoma site. The perpendicular axis of the V flap should be angled slightly caudally compared with the line of the emerging appendix. This facilitates the later formation and positioning of the Q flap.

The V flap is elevated, and a generous opening is made in the anterior abdominal wall musculature, through which the appendix is delivered (Fig. 10.2) Having delivered the appendix, it is generally advisable to stop working on the stoma site and to proceed instead with the closure of the abdominal wound, which is completed before the VQZ stoma is constructed, thereby ensuring that the stoma is not altered by changes in skin tension with wound closure.

After wound closure has been completed, the appendix is drawn out of the wound under slight tension and incised along its anti-mesenteric border. The V flap is laid into the gap and sutured into position using 5/0 polyglycolic acid sutures. It should be noted that the suture line continues right up to the angle of the V flap on the superior border but stops 5–10 mm short on the inferior margin (Fig. 10.3a,b).

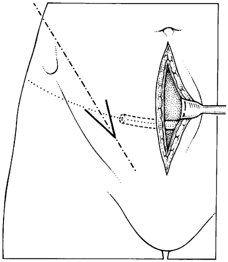

Fig. 10.1

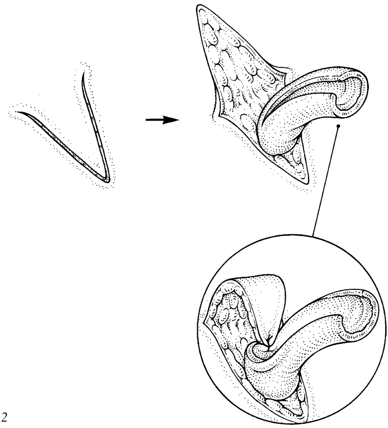

Fig. 10.2

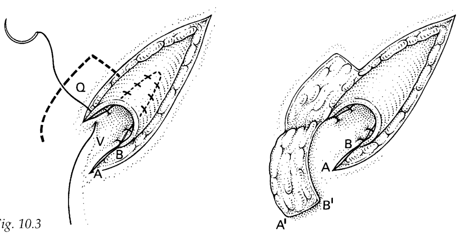

Fig. 10.3

Having completed the V flap insertion, the Q flap is now created from the superior margin of the abdominal wall defect created by V flap formation (Figs 10.3 and 10.4). The lateral end of the incision is 're-curved' slightly to allow for easier rotation. The flap is elevated and then rotated so that the medial end of the flap A'B' addresses the free margin of the V flap, AB (see Fig. 10.3).

The margins of the Q flap are sutured to the V flap and to the appendix using interrupted 5/0 polyglycolic acid sutures. The opening on the abdominal wall is now completely surrounded by skin with no exposed mucosa. The suture line is long and not circumferential at any one level.

Figure 10.5 shows the standard Z plasty used to close the skin defect overlying the emerging appendix and stoma site. In a virgin case, this is straightforward, but with a scarred abdominal wall it may require modification, usually in the form of an additional medial Z-plasty to relieve the tension.

Following an initially conservative incision of the inferior margin, the point X is sutured to X' (Fig. 10.5) using a relatively stout suture, such 3/0 PDS, as this may be under some tension. The incision may then be extended in whatever direction is appropriate to accommodate the movement Y–Y'. The initial suture X–X' must be onto abdominal wall skin and not the root of the Q flap (Fig. 10.6). Subcutaneous sutures of 3/0 PDS are used to accommodate the tension, and the skin margins are approximated with 6/0 PDS.

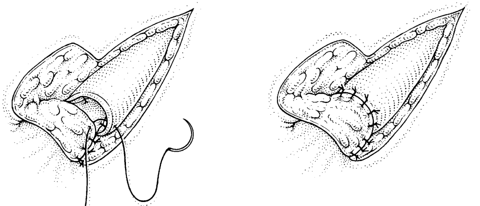

Fig. 10.4

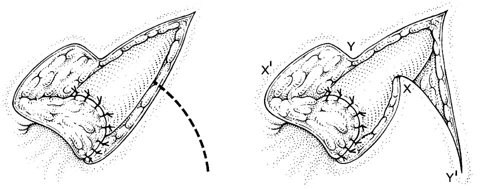

Fig. 10.5

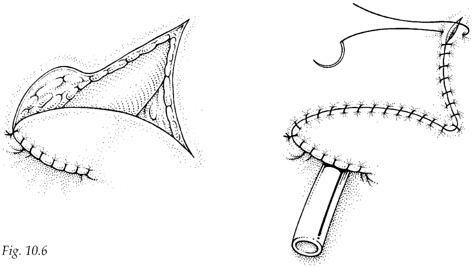

Fig. 10.6

The technique may be modified when the appendix stoma is incorporated in an extended Pfannenstiel or 'big smile' incision in the lower abdominal wall (Fig. 10.7). Note that the V flap is created by an oblique incision in the inferior margin of the wound so that, following the suture of the V flap to the appendix, a free margin is left on both sides rather than just on one side, as in the classical technique.

In creating the Q flap, the parallel incisions are made not at right angles to the skin margin but angled laterally to allow the inverted flap to follow the oblique line of the emerging appendix. The free margin X–Y is sutured to the free border of the appendix, and the progressive suturing of A–X to A′–X′ and B–Y to B′–Y′ rolls the Q flap inwards. The shaded areas of skin are excised before completing wound closure.

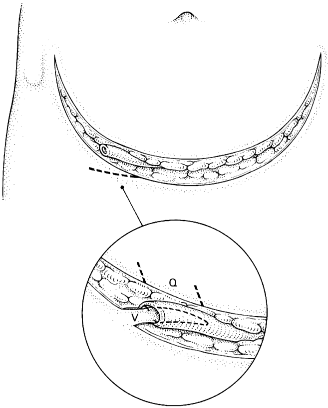

Fig. 10.7a

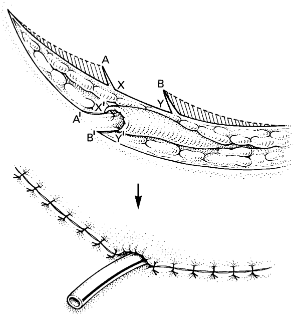

Fig. 10.7b

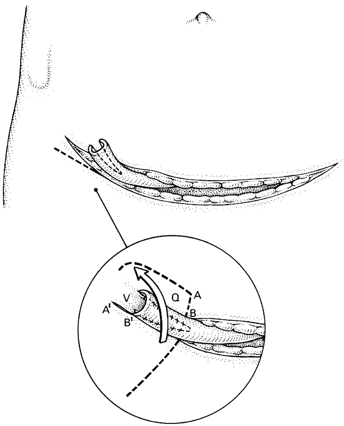

Fig. 10.8

Figure 10.8 illustrates the use of the VQZ technique in a Pfannenstiel type of incision where the appendix emerges at the lateral point of the incision. The V flap is formed from the lower skin margin as in Fig. 10.7 above, but the formation of the Q flap and its anastomosis becomes almost identical to the process used in the classical operation (see Figs 10.4 and 10.5 above).

Considerable experience with the technique is necessary successfully to convert an existing flush stoma into a concealed VQZ form (Fig. 10.9), but the effort is greatly appreciated by patients. It is best to harvest the V flap from the inferior border first before completing the circumcision and mobilization of the stoma. A quite extensive undermining of the surrounding skin is usually necessary as it is often tethered by previous scars. Once the skin has been freed from the muscle, a Q flap is manufactured from the upper border, construction proceeding as in Figs 10.3 and 10.4 above. Skin closure can be difficult and requires imagination, but the effort is worthwhile.

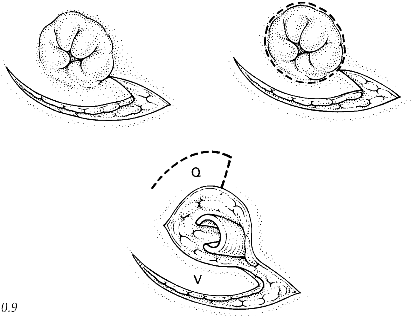

Fig. 10.9

Chapter 11

Persistent cloaca

Alberto Peña

Introduction

With persistent cloaca, which is encountered only in the female, there is an orifice on the perineum leading to a mucosa-lined channel in which the urinary, genital and alimentary systems terminate.

Embryology

The cloaca is a normal stage in embryogenesis, recognizable at the 5th week. Distally, it opens into the tailgut, while proximally lie the openings of the hindgut and the allantois. It is separated from the exterior of the embryo by the cloacal membrane. On either side of the cloaca, folds of mesoderm arise that fuse with each other from above downwards to form the urorectal septum. By the 7th week of embryogenesis, the cloaca is thus divided into the rectum posteriorly and the urogenital sinus anteriorly, and the cloacal membrane is divided into the urogenital and anal membranes.

A persistent cloaca is presumed to arise from an arrest of the formation of the urorectal septum; when the unstable cloacal membrane dehisces, the primitive cloaca then opens on the exterior. The relevant embryology is discussed by Pohlman (1911), Johnson et al (1972) and Gray & Skandalakis (1972).

History and incidence

The earliest mention of patients with persistent cloaca in the English literature are those of Nordenfelt (1926) and Major (1929). They described patients who died shortly after birth. In 1926, Pennock & Stark published a paper entitled 'Persistence of cloaca', but what they described was two patients with imperforate anus and a recto-urethral fistula.

Interestingly, in large series of anorectal malformations, such as the 162 cases of Ladd & Gross (1934), the 62 cases of Santulli (1952) and the 147 cases of Trusler & Wilkinson (1962), there was no mention of persistent cloaca. Table 11.1 summarizes the comparative frequency of persistent cloaca. We can say that roughly between 1% and 19.5% of anorectal malformations in females are persistent cloacas, and that 10–40% of all female anomalies correspond to this malformation. A careful chronological analysis of the number in the different series allows one to assume that the real frequency may be a little higher and may depend on the index of suspicion.

The striking difference in frequency in the incidence of persistent cloaca shown in Table 11.1 between this author and previous ones may represent errors in diagnosis. Most of the series shown present a high incidence of 'rectovaginal fistula'. In the author's experience, rectovaginal fistulas are almost non-existent defects. In addition, many teenage patients are referred to this author for consultation with a history of being born with a 'rectovaginal fistula', having been operated on by another surgeon. Patients have actually been born with an undiagnosed cloaca, undergoing a repair of only the rectal component of the malformation, leaving them with an untouched urogenital sinus. Many of these patients were recorded in the literature as 'rectovaginal fistulas' when they actually possessed a persistent cloaca. As surgeons and pediatricians are becoming more aware of the existence of this defect, the number of cases diagnosed early seems to be increasing. On the other hand, this author's

Table 11.1 Comparative frequency of persistent cloaca

	Anorectal malformations	Persistent cloaca
Both sexes		
Louw et al (1971)	287	6
Stephens & Smith (1971)	260	8
Nixon & Puri (1977)	86	1
Peña (work in progress)	940	183 (19.5%)
Santulli et al (1971)	1166	23
Stephens & Smith (series quoted by authors, 1971)	1098	27
Total	3276	112
Females		
Peña (work in progress)	441	183 (41.5%)
Synder (1966)	42	5
Palken et al (1972)	88	10
Bill et al (1975)	46	8
Total	319	70

frequency merely reflects the tertiary nature of his institution and cannot be taken as representative of the real incidence of this defect since the centre receives a large number of referrals of complex anomalies from other hospitals.

Anatomy, clinical findings and diagnosis

Based on the author's experience, persistent cloaca must be considered to be a spectrum of malformations with a common denominator or pattern consisting of the presence of one functioning external opening through which urine, genital secretions and feces may be passed (Fig. 11.1). An understanding of the concept of a 'spectrum' (Fig. 11.2) will prove particularly useful when discussing treatment of the malformation.

Although the external appearance of all these girls may at first glance be very similar (see Fig. 11.1a), the real internal anatomy, as seen through a panendoscope or when widely exposed using a posterior sagittal operative approach, varies significantly from one case to another (Fig. 11.2). One important clinical clue to the diagnosis is the presence of rather small-looking genitalia in a girl with an imperforate anus. The importance of an early suspicion cannot be overemphasized since the lack of a correct diagnosis may cause the surgeon to believe that he is dealing with a case of a girl with a simple type of imperforate anus with a vaginal fistula; the surgeon may subsequently repair only the rectal component of the malformation. The consequence of this is a girl with a pulled-through rectum and a persistent genitourinary sinus, the repair of which will be more difficult than a primary reconstruction including a repair of both the rectum and the genitourinary tract.

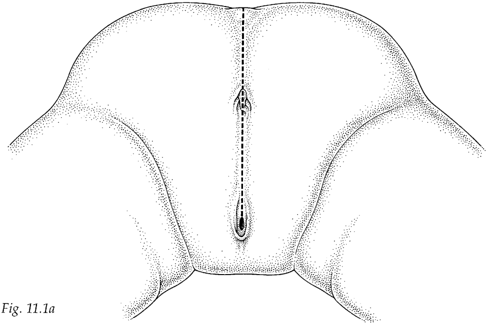

Fig. 11.1a

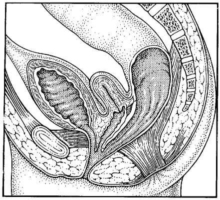

Fig. 11.1b

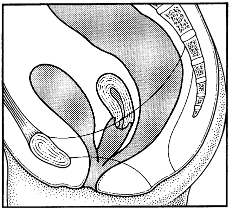

Fig. 11.2a

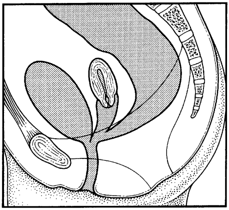

Fig. 11.2b

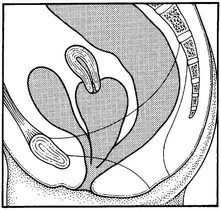

Fig. 11.2c

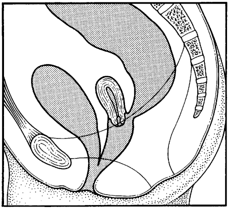

Fig. 11.2d

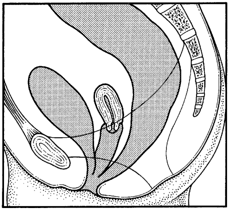

Fig. 11.2e

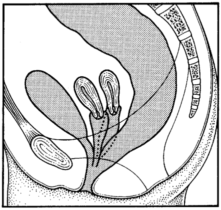

Fig. 11.2f

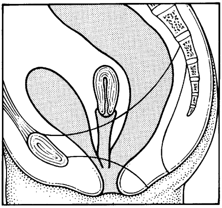

Fig. 11.2g

As can be seen in Fig. 11.2, the single channel or persistent cloaca communicates with the rectum, vagina and urethra at different levels. The vagina is septate or double in 44% of cases in the author's experience (Fig. 11.2f), a fact that must be kept in mind by the surgeon when interpreting the radiologic or endoscopic findings.

Contrast X-ray studies must always include good lateral films to identify the three visceral components of the cloaca (Fig. 11.3). Precise interpretation is frequently difficult. It is, however, since the advent of the posterior sagittal approach, not so important to determine the precise and minute detail of the local anatomy since the approach to repairing it will initially be the same. The combination of panendoscopy plus a 'cloacagram' (the injection of contrast material through the single perineal hole) has proved useful in determining the basic characteristics of most cloacas. This must be complemented by contrast injection through the distal stoma of the colostomy.

There is, however, a much more important evaluation to be carried out initially, which is the urological investigation, since these patients usually suffer from severe urological disturbances; in fact, genitourinary defects account for most deaths. Studies may reveal renal agenesis, severe vesico-ureteric reflux or pelviureteric obstruction. When severe sacral anomalies are present, one must also expect to deal with a neuropathic bladder.

It is common to find an associated hydrometrocolpos (see Fig. 11.2c); in fact, when a cloaca is diagnosed or suspected in a patient with a lower abdominal mass, the surgeon can assume that an obstructed vagina is part of the malformation. A very distended vagina may compress the trigone and interfere with the emptying of the ureters into the bladder (Cook &

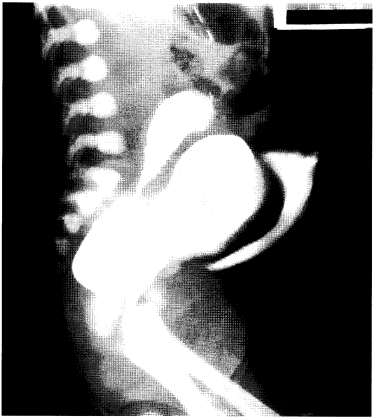

Fig. 11.3

Marshall 1964). This should be taken into consideration during the neonatal management of these patients. Cutaneous ureterostomies can be avoided by simple drainage of the vagina. When the vagina is not large enough to reach the abdomen above the area of the bladder, it can be drained by a tube vaginostomy. When the vagina is large enough to reach the abdominal wall above the bladder, drainage can be achieved by a formal exteriorization of the vaginal dome or by tubeless vaginostomy.

Rare types of cloaca include complex defects. The rectum may be located behind the urinary tract and open between the urinary tract and the vagina; the ureters may open ectopically, most frequently into the vagina. Eight patients from the author's series had a very unusual form of cloaca called posterior cloaca (Fig. 11.2). The main characteristic of this defect is that the patient has a normal or minimally abnormal anus and the urogenital sinus opens into the anterior rectal wall or immediately anterior to the rectum. This specific type of defect may require a special technical modification to carry out the repair properly. Even more complex defects may include a large, wide cloaca, communicating with the intestine and ureters and without any recognizable bladder, vagina or rectum.

Important prognostic factors are the length of the common channel (the cloaca), the size of the vagina, the condition of the sacrum and the quality of the muscles. The common cloacal channel varies in length between 1 and 7 cm (see Fig. 11.2a and b). It has been this author's experience that the presence of a common channel length of more than 3 cm is associated with significant technical difficulty in mobilizing the vagina sufficiently to reach the labia. The surgeon is then obligated to try an alternative maneuver to fill the gap between vagina and perineum, as will be discussed later. Common channels of less than 3 cm usually allow a primary suture between vagina and perineum.

The size of the vagina is also relevant: a small vagina is more difficult to dissect and mobilize, but it is also more likely to become devascularized. A large vagina allows the surgeon an easier mobilization and also the possibility of using a series of technical maneuvers aimed at reconstructing the vagina.

A complete sacrum usually indicates that the pelvic innervation is normal and therefore that the prognosis in terms of bowel and urinary continence is good. One missing sacral vertebra does not seem to have any adverse effect on fecal and urinary continence.

The quality of the sphincter muscles can usually be predicted by looking at the patient's perineum. A marked midline groove and a distinct recognizable anal dimple are usually associated with a good external anal sphincter and levator muscle.

Treatment

Establishing therapeutic priorities when dealing with a newborn with this malformation is the key to avoiding neonatal death. One must remember that there are two real emergencies requiring urgent treatment: intestinal obstruction and a urinary problem. Therefore a urologic evaluation is mandatory and must be carried out even before the colostomy is performed, particularly if the baby is showing signs of sepsis. Thus, the surgeon will be in the position to plan a procedure to decompress the colon and the urinary or genital tract simultaneously.

Before 1953, there were, in the English literature, isolated reports of cloacal deformities, but the babies were either born dead or died shortly after birth. In 1958, Sieber & Klein reported two cases; one died, and the other was referred to Orvar Swenson who performed the first simultaneous urethral, vaginal and rectal reconstruction in 1953.

Gough, in 1959, reviewed 18 cases from the Great Ormond Street experience; 11 patients died. Therapeutic attempts included colostomy, urinary diversion and abdominoperineal pull-through for the rectal part of the defect. No mention was made of the management of the genitourinary sinus. Bock & Madsen published a case report in 1971 of a patient who underwent an abdominoperineal pull-through for the rectum only, the genitourinary sinus being left untouched.

In 1973, Raffensperger & Ramenofsky, on the basis of their experience with 15 patients treated in different ways, proposed that the urinary, genital and gastrointestinal tracts should all be separated from one another in the newborn period. Two of their patients received a vaginal abdominoperineal pull-through in the newborn period. This paper represents the first series illustrating the multiple complications that must be expected when dealing with persistent cloacas.

Cheng et al (1974) reported their experience with five cases, two of whom died. They recommended a multistage approach, including a colostomy during the newborn period, followed by a rectal pull-through between the ages of 10 and 15 months and a genitoplasty later in life. This paper includes a good literature review of 50 cases previously published, emphasizing the high incidence of associated anomalies of the genital (77%) and urinary (63%) systems.

Kay & Tank (1977) published a review of five patients seen at their center and 34 documented cases; they also emphasized the high frequency of coexisting anomalies of the urinary tract (33%) and other systems and organs. They proposed a right transverse colostomy in the newborn period, followed in cases of high vaginal opening by rectal and vaginal pull-through when the patient weighed 25 lb (13 kg). In cases of low vaginal opening, they preferred a vaginoplasty by a posterior flap, to be carried out when the child was 10–12 years old.

Mollitt et al (1981) published an important paper pointing out the importance of long-term follow-up in these patients since they can suffer from an obstructed vagina or uterus, with severe symptoms later in life.

Hendren (1977, 1980, 1982, 1986) published four papers that together represent the largest, most ambitious and knowledgeable personal experience ever published on the subject. By reading them, one can see that the author has really been exposed to the spectrum of malformations; many of his cases were referred to him as secondary problems and therefore present additional and more complicated therapeutic challenges. His series demonstrates the magnitude of the effort required to treat these cases and that it must only be carried out by knowledgeable, experienced, meticulous and skilled pediatric surgeons also trained in pediatric urology.

Skin flaps may be useful to allow the suture of the vagina to the skin, and vaginal tubularization may be needed in cases of large vaginas (Kimura et al 1985). Bowel interposition may save some apparently 'non-useful' vaginas (Harrison et al 1983).

Many of the previously proposed treatments may work for certain kinds of persistent cloacas but will not be useful for others. This can only be acknowledged by understanding the concept of the spectrum of this group of defects. Thus, a simple perineal approach could conceivably be enough to repair a simple low defect (see Fig. 11.2 above); unfortunately, this variant is very unusual.

A posterior sagittal approach may be sufficient to repair most variants of this defect (see Fig. 11.2a, b, e and f above). A posterior sagittal approach and laparotomy would be indicated for the repair of a high rectal opening, such as the one shown in Fig. 11.2d above, in order to allow the mobilization the bowel. The same approach is necessary to repair a defect with a long common channel, such as the one shown in Fig. 11.2b; in this case, the justification for the laparotomy is the long gap existing between vagina and perineum. Different combinations of defects (other than those shown in Fig. 11.2a–f above) must also be expected.

Posterior sagittal anorectovagino-urethroplasty

Since 1982, this author (Peña 1985, 1989, 1992, 1995, Peña & de Vries 1982) has been treating persisting cloacas with an ambitious operation repairing the rectal, vaginal and urethral components in a single procedure called posterior sagittal anorectovagino-urethroplasty (PSARVUP). The basic steps when dealing with a new persistent cloaca case include:

1. establishing therapeutic priorities in the newborn in order to treat the most pressing or life-threatening defect (usually urinary obstruction);
2. the construction of a completely diverting descending colostomy (double barrel). In addition, when necessary, vaginal and/or bladder decompression must be performed simultaneously;
3. PSARVUP on an elective basis when the child is older than 6 months.

This discussion will be limited to the repair of the cloaca itself, granted that the urinary problem must be treated on an individual basis before, during or after PSARVUP, depending on the circumstances.

The main object of the posterior sagittal operation is to attain, when feasible, in a single operation:

1. fecal continence
2. urinary continence
3. normal sexual function.

In this author's experience, 77% of cloacas can be reconstructed through the posterior approach without opening the abdomen, whereas 23% of cloacas will require a laparotomy to complete the repair, either because the rectum is very high or because the vagina is located very high and requires a more complex reconstruction. The operation consists of a posterior sagittal incision with full exposure of the entire malformation, the separation of the three visceral components and the construction of a neo-urethra using the single cloacal channel, and vaginal and rectal pull-through with preservation of the entire striated muscle mechanism (levator muscle complex and external sphincter) (Fig. 11.4).

The patient is placed in a prone position with the pelvis elevated. No attempt is made to place a urinary catheter preoperatively since it is difficult to direct it from outside.

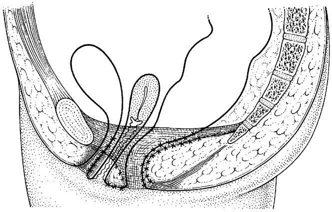

Fig. 11.4a

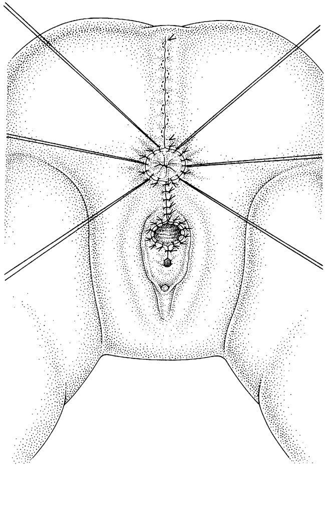

Fig. 11.4b

The incision runs from the mid-sacrum down to the cloacal orifice, dividing all the muscle structures behind the viscera (Fig. 11.5a). The external sphincter and muscle complex must be divided, including both their posterior and anterior aspects. The surgeon must remain strictly in the midline during the entire operation. To achieve this, a symmetric contraction of the striated muscles on both sides of the midline when electrically stimulated must be appreciated. One must preserve a very fine midline sagittal fascia that will avoid herniation of fat into the operative field. Staying in the midline guarantees avoiding any neurological damage since all nerves run paramedially and are terminal at the midline.

Needle-tip cautery is used through the entire operation, scrupulous hemostasis being essential to be able to recognize the important muscle and visceral structures. Deep to the skin, the longitudinal fibers of the external sphincter, also called parasagittal fibers, are identified. These run from the coccyx to the anal site. Deeper in the muscle, one will find more fat, which separates the sphincter muscle fibers from the levator muscle at the cephalad part of the incision. For more information related to the anatomic and functional details of these muscles, the reader is referred to previous publications by the present author.

Two sharp Weitlaner retractors provide excellent exposure, but they must be placed superficially in order not to crush the muscles. The coccyx is split in the midline, a right-angle clamp is introduced through the split coccyx into the presacral space, and the levator muscle is divided in the midline. After this, a visceral structure is found; its external appearance does not allow one to elucidate whether it is rectum, vagina or cloacal channel (Fig. 11.5a). This structure must therefore be opened in

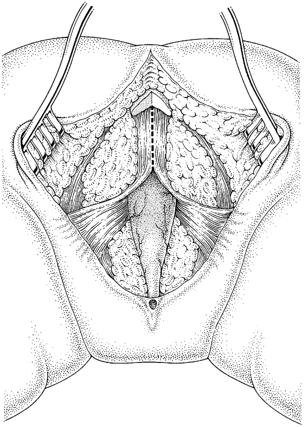

Fig. 11.5a

the midline, holding its wall edges apart by 5/0 silk stitches (Fig. 11.5b). By looking inside, one may determine the precise anatomic variant with which one is dealing (Fig. 11.5c).

It is at this point that one can predict the specific type of technical variant necessary to correct the specific malformation. In this author's experience, whenever the rectum is identified through this approach, a laparotomy will not be necessary since, with an adequate technique, the bowel can be pulled down to the skin without tension. If the rectum is not seen through this approach, a laparotomy will be required to bring it down.

The vagina usually presents a more difficult problem. High, small vaginas are the most challenging; large, low vaginas are easy to deal with, and one can expect much better results from such cases. It is sometimes difficult to predict, even with the malformation exposed, whether or not a laparotomy will be required for vaginal construction. One may have to start trying to dissect and mobilize the vagina, only to find after a while that a laparotomy is indeed needed since the vagina is too high to be pulled down.

Once the malformation has been completely exposed (Fig. 11.5c) the first goal is to achieve the separation of the rectum from the vagina. It is important to remember that the two structures share a common wall immediately above the point of union.

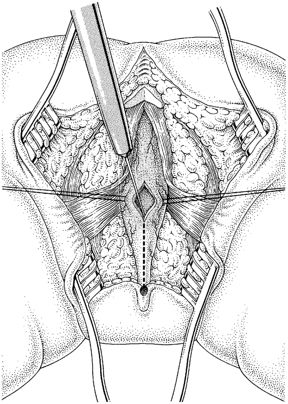

Fig. 11.5b

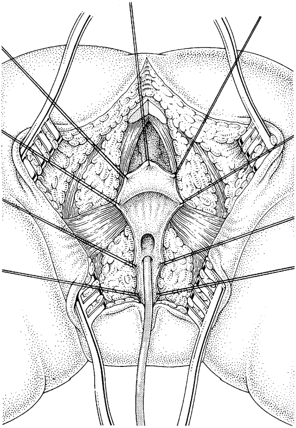

Fig. 11.5c

A plane of dissection should be created between the rectum and vagina. This maneuver is relatively simple but requires a meticulous technique (Fig. 11.6a). To avoid tissue damage, many 6/0 silk mucosal stitches are used for traction. The injection of 1:100 000 epinephrine solution sometimes allows an easier separation of the structures.

Dealing with the urogenital component of this defect requires more sophisticated technical maneuvers. Traditionally, the vagina was separated from the urethra and bladder, the old common channel was reconstructed as a neo-urethra, and the vagina was then mobilized and pulled down, to be placed immediately behind the urethra (Fig. 11.6b and c). This basic approach is still used in a significant number of cases. Lately, however, this author has been using a new and very advantageous maneuver consisting of the total mobilization of the urogenital sinus. The separation of vagina from urinary tract is still recommended in malformations with a common channel longer than 3 cm, whereas a total urogenital mobilization is recommended in malformations with a shorter common channel. Each type of approach will be described separately.

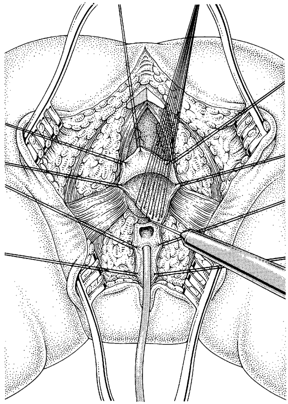

Fig. 11.6a

Separation of vagina from urinary tract

The separation of vagina from the urinary tract is a difficult maneuver and requires more skill since both structures contain friable tissue (Fig. 11.6b). It is convenient to remain closer to the vaginal lumen than to the urethral side in order to protect the urinary sphincter and avoid urinary fistulae. This full exposure of the area and its correlation with the results obtained has allowed the present author to clarify an important anatomic misconception related to the

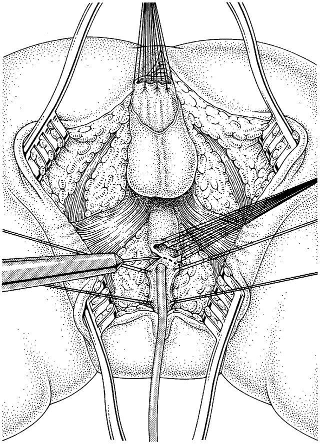

Fig. 11.6b

striated urinary sphincter. This structure has been considered to be, and depicted as, a well-limited ring muscle band that must be protected to attain urinary continence. A meticulous dissection and search has not allowed this author to identify any similar structure in cases that were subsequently demonstrated to be urinary continent. The urinary sphincter therefore seems to be represented in these cases by a continuum of striated muscle existing all along the common channel that will function as a neo-urethra. This muscle is easily identifiable by electrical stimulation. Urinary continence depends upon the presence of a good sacrum. In addition, the length of the common channel seems to have a significant prognostic value since only 19% of patients with a common channel shorter than 3 cm required intermittent catheterization to empty the bladder, whereas 69% of those with a common channel longer than this required the same maneuver.

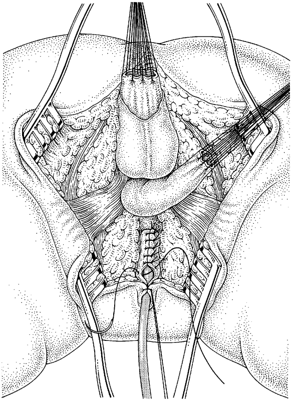

Fig. 11.6c

A vaginal septum, frequently found, can be easily resected during the operation. A very large vagina may require tapering in order to create an introitus proportionate to the size of the girl's perineum. Once the vagina has been separated from the urinary tract and mobilized enough to reach the perineum, the neo-urethra must be constructed using the common cloacal channel (Fig. 11.6c).

Total urogenital mobilization

The author has found total urogenital mobilization to be a highly advantageous maneuver, mainly because it avoids the tedious and dangerous separation of the vagina from the urinary tract. The goal is to mobilize both urinary tract and vagina as a block down to the perineum enough to achieve a wide vaginal opening. This is carried out after the rectum has been separated from the urogenital sinus as previously described (Figs 11.7 and 11.8). The additional advantages of performing this maneuver include an excellent blood supply and the elimination of the risk of vaginal strictures and urethrovaginal fistula.

To achieve this, multiple 6/0 silk stitches are placed in the vaginal edge as well as across the common channel a few millimeters cephalad to the location of the clitoris (Fig. 11.9). The common channel is divided distal to the series of 6/0 silk stitches, and a dissection is carried on between the pubic bone and the anterior wall of the common channel. This dissection is very easy because there is a pre-established plane of dissection.

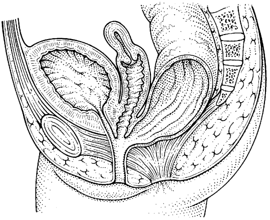

Fig. 11.7

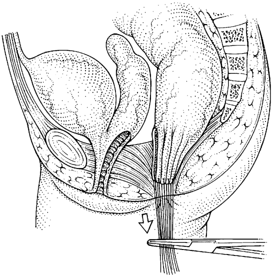

Fig. 11.8

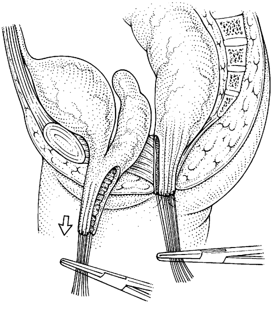

Fig. 11.9

Simultaneously, a circumferential dissection is carried out around the anterior wall of the common channel as well as the bladder, the lateral wall of the bladder and the lateral wall of vagina as a block. This total mobilization of the urogenital sinus takes about 80% less time than the traditional mobilization of vagina and urinary tract. The mobilization must continue until the vagina can comfortably reach the perineum in order to establish a wide vaginal opening (Fig. 11.10).

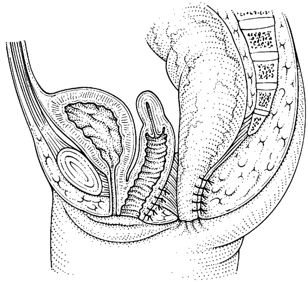

Fig. 11.10

The reason why this maneuver is not performed in all cases of persistent cloaca is simply that there is a limit to the mobilization of these structures in this fashion. Therefore, longer common channels require the traditional approach and most likely a laparotomy. Fortunately, most cloacas belong to the category that can be treated this way.

The vaginal wall is then sutured to the perineal skin using separate stitches of a long-term absorbable synthetic material. The perineal body must then be reconstructed; for this, the anterior and posterior limits of the external sphincter and muscle complex are determined using the electrical stimulator. The anterior limit of the muscle complex is re-approximated using interrupted 5/0 Vicryl stitches (Fig. 11.11a). The rectum will be located in front of the levator muscles and within the limits of the muscle complex and external sphincter. The width of the rectum cannot be larger than the muscle complex width. In cases with a larger rectum, it is imperative to tailor it to achieve the goal, this being required in more than 50% of cases. The amount of rectum resected depends on the size discrepancy between the size of the rectum and the space available. The tapering is carried out, resecting part of the posterior rectal wall and closing

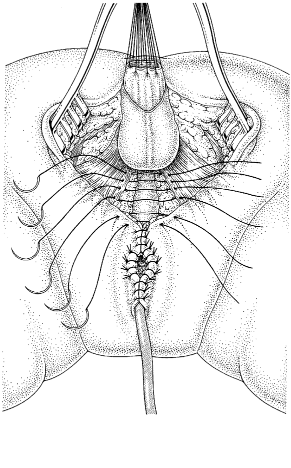

Fig. 11.11a

with two layers of interrupted stitches. The levator muscle is sutured behind the rectum. The sutures that bring together both posterior edges of the muscle complex must be anchored to the posterior rectal wall to avoid rectal prolapse (Fig. 11.11b).

The anoplasty is achieved with 16 stitches of interrupted 5/0 long-term absorbable sutures (Fig. 11.11c). A Foley catheter is left in place in cases with a common channel of 2 cm or shorter, whereas a suprapubic cystostomy tube is left in place in cases with a longer common channel.

Anal dilatation is started 2 weeks after the operation. This is performed twice daily by the mother, and every week the surgeon must pass the next larger-sized dilator. The goal is to reach a 12 Hegar dilator in newborns or young infants, a 15 Hegar in 1-year-old patients and a 16 or 17 Hegar in school-age children before considering the closure of the colostomy. This process usually takes between 2 and 3 months. After the colostomy has been closed, the last-sized dilator must be passed daily and a gradual tapering in frequency can be carried out over a 6-month period.

We have not dilated the vagina postoperatively. The vaginal introitus of these girls frequently looks rather narrow, but endoscopic examination 3 months after surgery usually shows a very satisfactory vagina. A few of our patients have already reached puberty, and we have observed that the oestrogen secretion enlarges the vagina even more.

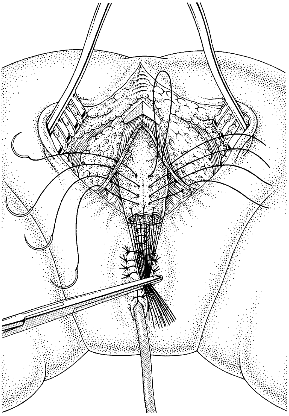

Fig. 11.11b

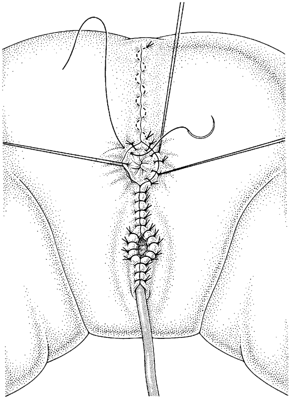

Fig. 11.11c

Special technical maneuvers for vaginal reconstruction

When patients have a common channel longer than 3 cm, they usually require a special maneuver for the vaginal reconstruction. This author gave up using skin flaps because they have a high tendency to retract and scar down. At the present time, the special maneuvers for vaginal reconstruction include:

1. vaginal flap
2. vaginal switch
3. vaginal replacement with a piece of intestine.

Vaginal flap The vaginal flap can be used in patients who have a common channel of approximately 3–4 cm associated with a large vagina. In addition to the routine vaginal mobilization, one can mobilize the vagina laterally as well as posteriorly, and one can then tailor a flap from the vaginal dome to create a posterior vaginal wall. This maneuver, together with a total urogenital mobilization, may prove to be enough for an adequate reconstruction.

Vaginal switch The vaginal switch maneuver is used in cases with a long (more than 3 cm) common channel associated with two large hemivaginas (hydrocolpos) (Fig. 11.12). These patients require a laparotomy, and once these two large hemivaginas have been separated from the bladder, the surgeon may find that the distance between both uteri is longer than the vertical dimension of the vagina. The vessels that provide the blood supply for these vaginas enter laterally, so one cannot mobilize this type of vagina down to the perineum without sacrificing its blood supply. The author has thus been using a maneuver called the vaginal switch. One of the hemiuteri is resected, the blood supply is sacrificed on the same side, both hemivaginas are tubularized after the resection of the vaginal septum, and the dome of the vagina on the same side as the resected hemiuterus is switched down to the perineum (Fig. 11.13). This has proved to be an excellent maneuver. The blood supply of the hemivagina whose hemiuterus has been preserved usually provides enough circulation to maintain the viability of the vagina that was sutured down.

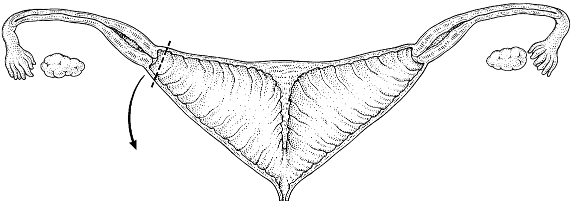

Fig. 11.12

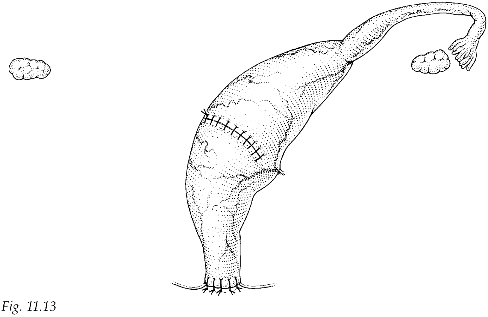

Fig. 11.13

Vaginal replacement with intestine

When using a vaginal replacement with intestine, the author prefers to use sigmoid colon. Sometimes, however, the presence of a sigmoid colostomy however, prevents the use of the sigmoid colon so small bowel must be used. It is important to determine the point of the terminal ileum that has the longest mesentery, which usually corresponds to a point located approximately 15 cm from the ileocecal valve. At this point, the small bowel is divided (Fig. 11.14). The arcades of the blood supply of that piece of small bowel are studied and divided accordingly in order to achieve enough length preserving a good blood supply, and the bowel is pulled down to the perineum.

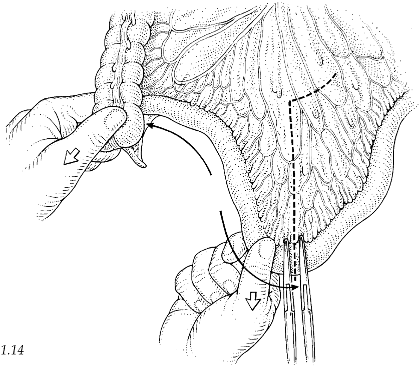

Fig. 11.14

Once it has been confirmed that the blood supply and bowel length is adequate, the upper part of the bowel is divided and the continuity of the small bowel re-established (Figs 11.15 and 11.16). Some patients with persistent cloaca do not have a uterus,

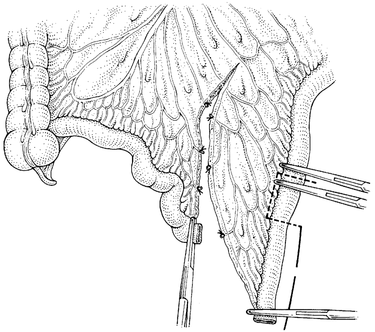

Fig. 11.15

so the piece of bowel used for vaginal replacement must be closed in its upper portion. Some other patients have a very small vagina and require a vaginal replacement. In such cases, one has to suture the upper part of the piece of bowel that is used to replace the vagina to the little vaginal cuff of the original vagina (Fig. 11.16).

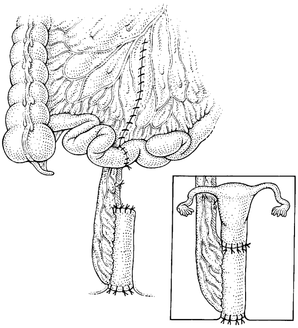

Fig. 11.16

Re-operations for persistent genitourinary sinus

The most frequent indication for the re-operation of persistent cloaca is a school-age or teenage patient who was born with this defect and was operated on with conventional techniques prior to the advent of PSARVUP; the most common method of treatment was a rectal pull-through, leaving intact the urogenital sinus. The patient may or may not have had good fecal and/or urinary control. In such circumstances, treatment consists of creating a protective right transverse colostomy, followed by a secondary PSARVUP, the colostomy being closed in a third stage.

The reason for recommending a right transverse colostomy rather than a descending one, as we do for primary cases, is that most patients who have previously undergone a pull-through procedure have had a ligation of the inferior mesenteric or left colic artery; the entire left colon is therefore supplied by the middle colic vessels and by the new blood supply formed between the perianal area and the rectum. Thus, a descending colostomy may compromise the blood supply of the distal colon.

In such cases, the rectum is usually found to be mislocated either anteriorly or lateral to the muscle complex and external sphincter. The external sphincter and muscle complex are usually found intact. The operation consists of a full midline sagittal incision, dividing the external sphincter and muscle complex. Multiple 5/0 silk traction stitches are placed at the mucocutaneous junction of the anus. A meticulous dissection is carried out around the rectum, staying as close as possible to the bowel serosa and leaving intact all the striated muscle in the vicinity. For the identification of such muscle, we use the electrical stimulator. Once the rectum has been completely mobilized, the genitourinary sinus is exposed as in the second part of the original primary cloacal repair (see Fig. 11.6b above). We then proceed with the repair of the vagina and the urinary tract as previously described. The perineal body is also repaired.

Many of these patients have had a previous abdominoperineal operation and therefore have colon pulled down to the perineum. To preserve the blood supply of the colon, the previous surgeon has been obliged to mobilize a piece of mesentery with it, which subsequently leaves a bulging mass surrounding the colon, interfering with a good muscle–bowel contact and thus being disadvantageous for bowel continence. In these cases, we resect the mesentery that surrounds the rectum along the last few centimeters, leaving the blood supply of the bowel to be provided by its transmural vessels. In this author's experience, no cases of bowel ischemia have so far been recorded. The rectum is then relocated anteriorly to the levator muscle, within the limits of the muscle complex and at the center of the external sphincter.

Some patients who were previously suffering from urinary incontinence have re-gained urinary control, the explanation being that they were previously urinating into a large vagina and subsequently dribbling urine; once we successfully separated the vagina from the urinary tract, the patients were able to urinate and have control. In other cases, with a poor sacrum and a primary neuropathic bladder, the separation of vagina from the urinary tract was also advantageous for the patient since she was able intermittently to catheterize her bladder because the new urethral orifice was easily visible. Patients with a genitourinary sinus or persistent cloaca cannot usually practice intermittent catheterization.

In some cases of re-operation we find, as in primary cases, a long gap between the vagina and perineum so that it is impossible to mobilize the vagina sufficiently without risking de-vascularization. Here, we take advantage of the fact that many of these patients have a colon with mesentery pulled down as a rectum, and we may therefore elect to use the most distal part of this bowel, preserving its mesentery, to fill the gap between vagina and perineum. The proximal bowel is then mobilized, pulled down posterior to the vagina and anastomosed to the skin within the limits of the external sphincter. By doing this, we do not have to open the abdomen.

Secondary operations for the treatment of fecal and urinary incontinence

Patients with poor sacrums and demonstrated neuropathic bladders are not good candidates for a re-operation using the posterior sagittal approach. On the other hand, patients with a good sacrum, a good-looking perineum, evidence of a large vagina and urinary incontinence must be re-operated on because of the reasons discussed above.

Fecal incontinence in these patients may be the result of a mislocated rectum. The ideal candidate for re-operation for the treatment of such incontinence is a patient with a good sacrum, good muscles and evidence of a completely mislocated rectum. The rectum is most frequently located too anteriorly to the external sphincter, and the muscles remain intact. In this author's experience, re-operation carried out in patients with a poor sacrum gives rather poor results.

Results

One hundred and seventy patients underwent a primary or secondary repair for a persistent cloaca. Of these, 37 who underwent a primary repair were recently reviewed. Twenty-one have a common channel shorter than 3 cm and 16 a longer common channel. Nineteen per cent of the patients with a short common channel require intermittent catheterization to empty this bladder, whereas 69% of the patients with a longer common channel need such a maneuver. Of those patients with a short common channel, 67.2% have enjoyed voluntary bowel movements, whereas 66.5% of those with a long common channel have done so. Urethrovaginal fistula, occurred in 7.3% of all cases of persistent cloaca, another 7.3% suffering from a narrow introitus that required a revision. Two patients had complete, acquired vaginal atresia caused by de-vascularization, which required re-operation.

The complications already mentioned here have been avoided during the last few years because of the advent of the new maneuvers described above.

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Chapter 12

Genitoplasty for congenital adrenal hyperplasia

Giacomo Passerini-Glazel

Introduction

The most common biochemical anomaly that leads to congenital adrenal hyperplasia is 21-hydroxdroxylase deficiency. It is inherited as a recessive disorder, and there is, in approximately half the patients, an associated salt loss. There is an excessive conversion of 17 α-hydroxyprogesterone to weak adrenal androgens, which in turn cause virilization. All these female pseudohermaphrodites have a normal 46XX karyotype and normal development of the ovaries, uterus and upper vagina.

Anatomy

The degree of virilization is variable and can be mild, with little more than an enlarged clitoris and fused labial folds; in severe forms, however, the appearance is almost that of a normal cryptorchid male. In the mild form, the vagina extends down to the urogenital sinus and is easily accessible surgically. In the severe form, the vagina connects to the posterior urethra via a small slit-like orifice opening in the area that would be the verumontanum in the male. Most children show an intermediate degree of virilization.

Indications for surgery

As these children are otherwise normal females and will want to have a normal sex life and enjoy the possibility of becoming pregnant, it is important that the external genitalia are corrected as perfectly as possible. Surgery should provide a good cosmetic and functional result. Thus, the phallic size should be reduced, labia minora and majora created and the vagina properly positioned with an adequate opening for later menstruation and sexual function. The two surgical procedures commonly required are a cliteroplasty and vaginoplasty. The optimum age for surgery is still under debate, but this author feels that enlarged phallus should be reduced in size by 2 months of age in order to reduce parental concern and ensuing psychological problems. If the vaginal insertion is low, a simple inverted U-shaped skin flap can be inserted into the vagina with ease, a procedure that can be performed early in life.

Controversy exists over the best age for surgery for those with a high insertion of the vagina. Some surgeons carry out this procedure at 3–5 years of age, whereas others delay surgery until after puberty. This author prefers to carry out the clitero- and vaginoplasty as a single stage in the severely virilized patients in the first few months of life or soon after puberty. Exposure to maternal oestrogens in the first few months of life produces a larger and thicker vagina, thus making the surgery easier. At puberty, there is the normal increase in oestrogen production, allowing an increase in vaginal size, which leads to easier dissection.

Preoperative preparation

The diagnosis is confirmed by a normal female karyotype and abnormal plasma steroid levels. It is helpful to know where the vagina inserts into the genitourinary sinus, so a micturating cystourethrogram should be performed in order to delineate the vagina. In those patients with a high insertion, a membrane occasionally covers the vagina, and therefore no vagina is seen on radiological examination. In these patients, an initial endoscopy will allow the opening of the vagina to be visualized and a high insertion to be confirmed.

Surgery should only be undertaken when the patient is biochemically stable. Extra steroids should be given before surgery. The help of a pediatric endocrinologist should be enlisted both pre- and postoperatively.

Surgical technique

The skin is prepared and the child draped in a lithotomy position. If the virilization is minimal, an inlay technique using a Fortunoff flap may be all that is required. An inverted U-shaped flap is made with the base sited where the posterior vaginal lip is desired. A thick flap of skin is then dissected. The posterior wall of the urogenital sinus is visualized and incised vertically until the vaginal opening is found. The incision is carried into the posterior wall so that the vagina is opened widely. The apex of the flap is then sutured to the posterior wall of the distal vagina, and the vaginal lips are reconstructed with labial skin.

This technique can only be used for the distal insertion of a vagina into the urogenital sinus. If an attempt is made to carry out this technique when the vaginal opening is too proximal, a significant female hypospadias will be created, which can lead to problems of discomfort with voiding and recurrent urinary infection.

Cliteroplasty and vaginoplasty for severe virilization

A Fogarty catheter is placed into the vagina endoscopically, and the balloon is inflated. A Foley catheter is then placed into the bladder. The operation begins with an inverted Y incision at the level of the scrotal-like labial folds (Fig. 12.1). The bulbospongiosus muscle is incised in the midline (Fig. 12.2). An injection of epinephrine into the tissues around the urogenital sinus will help to prevent bleeding during the dissection. The perineal body is freed from its surrounding connections. The urogenital sinus is progressively freed with sharp dissection, displacing it from the superficial and deep transverse perineal muscles and the pararectal fibres of the levator ani. Further sharp and blunt dissection is continued to allow the creation of an adequate

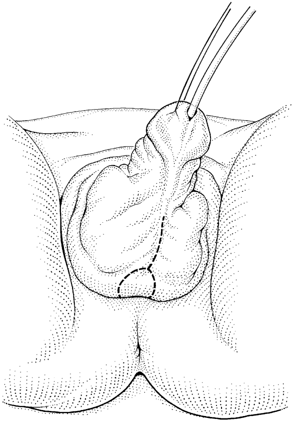

Fig. 12.1

space around the vagina. An inadequate dissection at this point may well lead to subsequent vaginal stenosis. An assistant can help by retracting the bulbar portion of the urogenital sinus using a wet swab under the tip of the forefinger.

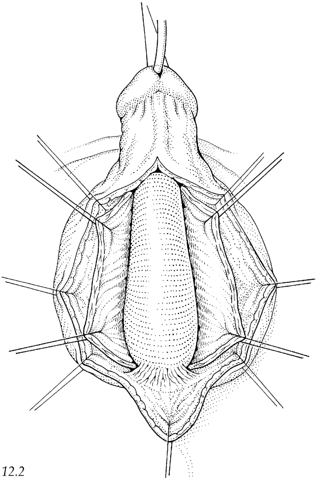

Fig. 12.2

Further dissection of the vagina is continued in order adequately to visualize the urethrovaginal confluence. This confluence can be confirmed by gently pulling on the Fogarty catheter, when the inflated balloon will be felt by the surgeon's finger. Further dissection is then carried out until the posterior wall of the vagina has been adequately exposed and the balloon can be visualized. A vertical incision is then made close to the urethrovaginal confluence (Fig. 12.3). At this point, the balloon of the Fogarty catheter is often burst. The incision is then completed with a transverse incision 5 mm from the junction of the urethra.

The transverse incision in the vagina is then extended laterally, and the vagina is dissected free from its junction with the urethra. There is unfortunately no easy plane of dissection between the vagina and the urethra since they share a common wall. The inside view obtained through the transverse incision can help with this dissection. The dissection must be continued until the vagina has been completely freed and adequately mobilized. The opening in the urethra is then closed with interrupted 5/0 or 6/0 Vicryl sutures (Fig. 12.4).

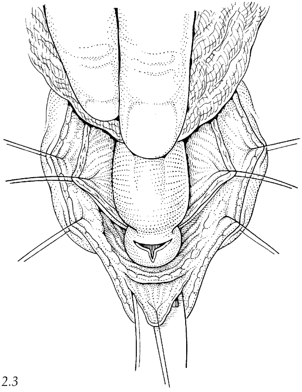

Fig. 12.3

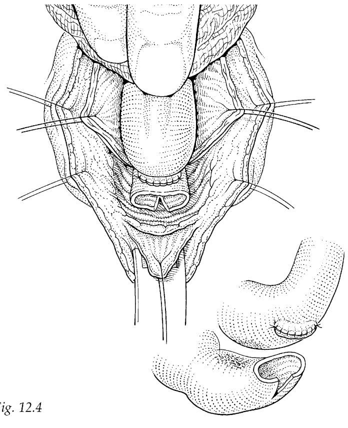

Fig. 12.4

A moist swab is then placed in the cavity and the cliteroplasty is then undertaken. A circumcision is made around the coronal region, and the phallic skin is mobilized and retracted. The dissection around the corpora is then continued in order to free up the neurovascular bundle dorsally and the urogenital sinus ventrally (Fig. 12.5). Further mobilization of the urogenital sinus is carried out until it has been completely mobilized from the glans and can be carried ventrally. The dissection of the corpora is then continued until they can be separated from the glans. The corpora are then transfixed, ligated and excised at the point at which the clitoris will need to be situated (Fig. 12.6a). The glans, which is normally too large, will need to be trimmed to give the appearance of a clitoris. This trimming is performed by removing a wedge from the glans ventrally, thus leaving the neurovascular connections to the glans intact (Fig. 12.6b). The glans is then reconstituted using interrupted mattress 4/0 sutures (Fig. 12.6c).

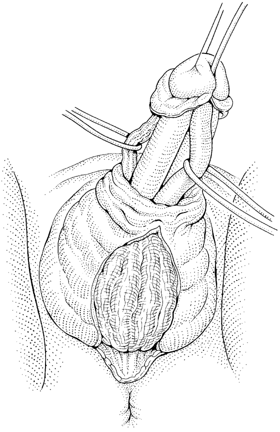

Fig. 12.5

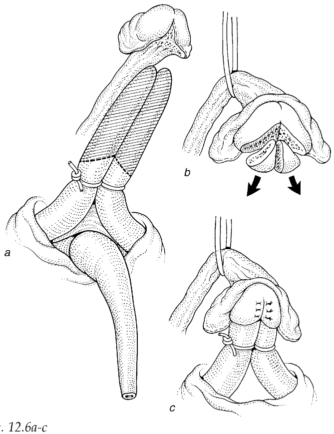

Fig. 12.6a-c

The phallic skin is then pulled up and incised ventrally and dorsally (Fig. 12.7), creating two skin flaps. The urogenital sinus is incised dorsally (Fig. 12.8). The proximal part of this incision is carried out in a V shape and finished where the urethral meatus needs to be positioned. The small proximal wedge shape of the

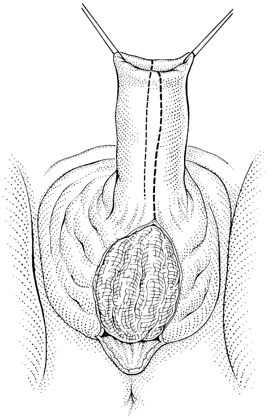

Fig. 12.7

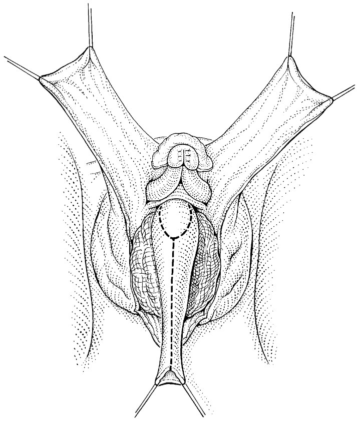

Fig. 12.8

urogenital sinus can then be sutured to the glans (Fig. 12.9). The two skin flaps are then moved dorsally, the medial edge being sutured around the glans and then sutured on each side to the divided urogenital sinus, leaving a large mucocutaneous flap that can be trimmed if it is too long (Figs 12.10

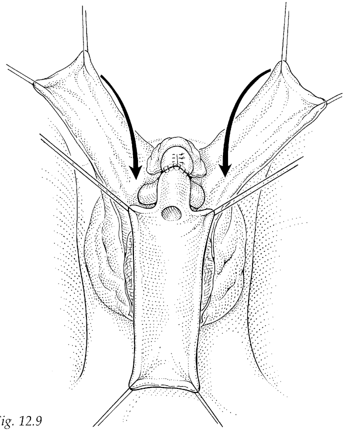

Fig. 12.9

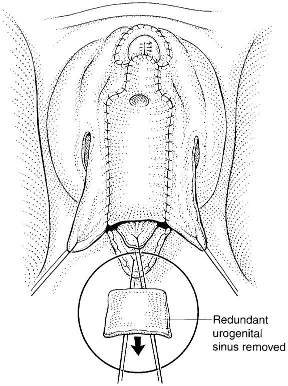

Fig. 12.10

and 12.11). This flap is then sutured to the ventral wall of the vagina (Figs 12.12 and 12.13). The posterior Fortunoff flap is sutured to the dorsal wall of the vagina.

If there is any excess fat in the labial folds, this can be excised at this point in order to create cosmetically pleasing labia majora (Fig. 12.14). The fatty tissue from one side should be discarded, but that from the other side can be placed between the urogenital sinus and urethra above and the reconstructed vagina below, thus helping to prevent the occurrence of a fistula. This should also help to produce a gentle curve in the anterior vulval region between the urethral and vaginal openings. The lateral suturing of the labia is then carried out using fine adsorbable continuous sutures (Fig. 12.15).

A small vaginal pack can be left in the vagina in order to control hemorrhage and keep the vagina open while healing occurs. This, together with the urethral catheter, should be removed after approximately 1 week.

Patients will occasionally have a very small vagina with a high insertion, which will make its dissection from the perineum extremely difficult. In such cases, a transtrigonal approach has been found to be helpful. The anterior bladder wall is opened, and the trigone is incised from above the interureteric bar down to the bladder neck (Fig. 12.16). With dissection, the vagina can be easily visualized.

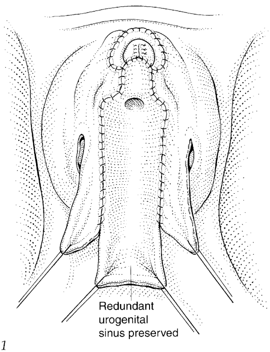

Fig. 12.11

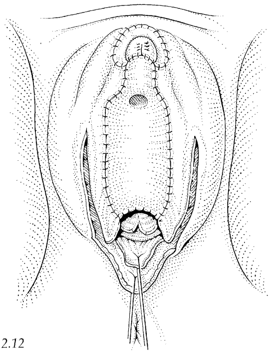

Fig. 12.12

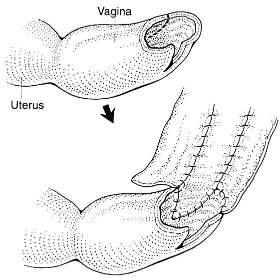

Fig. 12.13

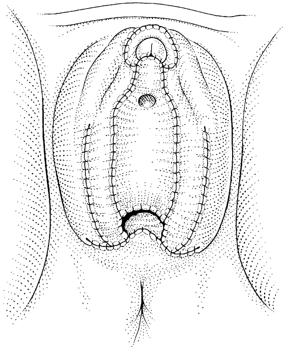

Fig. 12.15

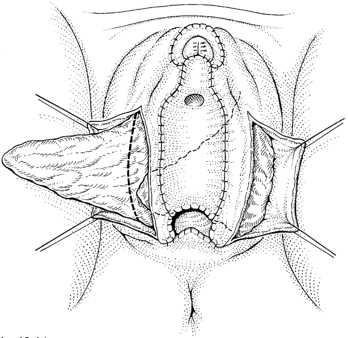

Fig. 12.14

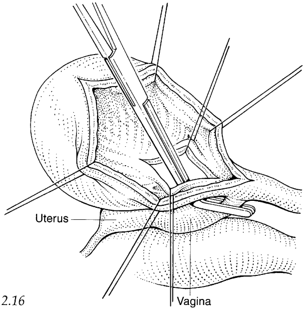

Fig. 12.16

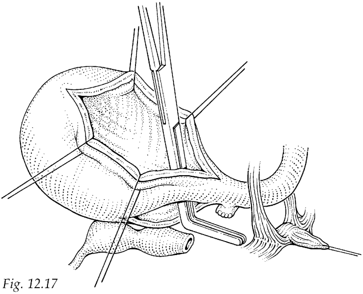

Fig. 12.17

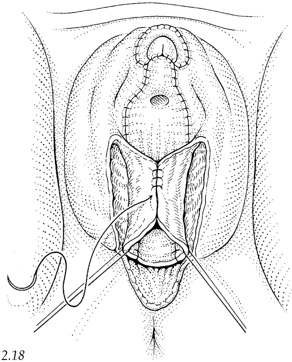

Fig. 12.18

It then needs to be divided, but the stump of the urethra will also require closing, which can be technically difficult (Fig. 12.17). The suturing is therefore started with a stitch in the ventral wall of the urethra, this then being closed using a continuous suture as the vagina is divided from the urethra. This stitch enables the closure to be carried out reasonably easily.

The mucocutaneous flap that has previously been created is closed distally to form a tube (Fig. 12.18). This tube can then be anastomosed to the small proximal vagina. A right-angled clamp is passed through the retrovesical space, and, taking hold of the holding stitch left in the distal part of the neo-vagina created from the mucocutaneous skin (Fig. 12.19), the tube is pulled into the peritoneum and can be anastomosed to the vagina through the transtrigonal incision. The bladder is then closed, leaving a Foley catheter in situ, and the perineal wound is closed as for any high vagina.

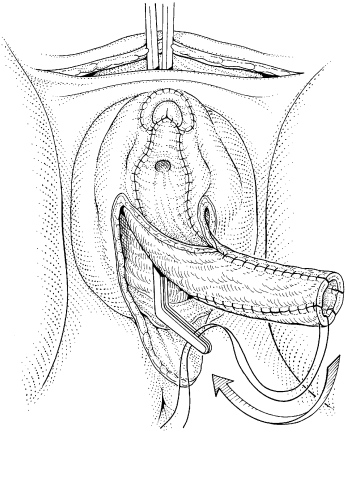

Fig. 12.19

Chapter 13

Hypospadias repair

John W. Duckett

Introduction

Since hypospadias occurs in 1 in 300 male births, its repair is a very common pediatric urological procedure. In general, upper urinary tract anomalies are not common, and investigation is not required. The more severe the placement of the meatus, the more likely it is that a large prostatic utricle will be present. This entity generally does not cause a problem except for the placement of a catheter through the urethra. Nine per cent of hypospadias patients have undescended testes, and 9% have inguinal hernias or hydroceles. The presence of other associated anomalies, such as a cardiac defect, imperforate anus or trachea-esophageal fistula, will of course require more extensive urological investigation.

After 20 years' experience of repairing hypospadias, the present author is confining his diagrams and remarks to those techniques used at the Children's Hospital of Philadelphia. The classification used is the location of the meatus after the release of chordee. Thus, anterior hypospadias contributes about 50% of cases, middle hypospadias 30% of cases and posterior hypospadias 20% of cases.

Techniques used for anterior hypospadias are meatal advancement and glanuloplasty MAGPI, extended MAGPI, the Mathieu procedure and the onlay transverse preputial island flap. Approaches used for middle hypospadias are the onlay island flap and transverse preputial island flap urethroplasty. Techniques used for posterior hypospadias are the onlay island flap, the island flap tubularized and the extended 'horseshoe' island flap tube including a re-do onlay island flap using adjacent penile skin. Buccal graft urethroplasty has replaced bladder mucosa and extragenital skin free grafts. The final group is complex re-do hypospadias repair. A variety of techniques, including bladder mucosal free-graft urethroplasty, are utilized.

Meatal advancement and glanuloplasty

The most common lesion seen in hypospadias is a subcoronal meatus with a blind-ending distal groove (Fig. 13.1a). The urinary stream is deflected ventrally by 45 degrees. In about one-third of cases, the meatus is very stenotic. A dorsal preputial hood is present with a median raphe deviating off to one side, causing penile torsion to the left. The skin on the ventrum is frequently very thin and tethered. There is apparent chordee in many of these cases, but, after skin release, an artificial erection will reveal a straight penis. It is inappropriate to judge the presence of chordee prior to the release of skin tethering.

To flatten out the skin bridge distal to the meatus, a vertical incision is made into the glanular groove for about 1 cm (Fig. 13.1b and c). This creates a diamond-shaped defect that is closed transversely with 6/0 or 7/0 Vicryl. This effectively opens the meatus, flattens the groove and advances the dorsal meatal edge into the glans. For a prominent bridge distal to the meatus, a wedge resection is preferred.

The glanuloplasty portion is formed by elevating the ventral meatal edge forward (Fig. 13.1d–f). Two holding stitches are placed on the glanular wings, which are brought ventrally. Redundant skin in the midline is excised so that the glanular tissue can be approximated in the normal conical anatomy. Vicryl Rapide sutures are used for the closure of the glans. A sleeve re-approximation for skin cover is generally accomplished, but if there is a significant skin deficiency, Byars' flaps must be utilized, as depicted in Fig. 13.2c and d below.

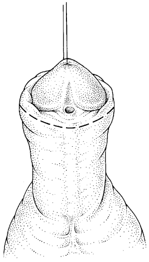
Fig. 13.1a

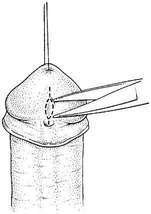
Fig. 13.1b

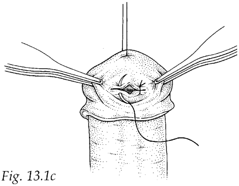
Fig. 13.1c

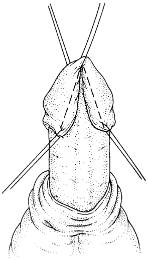
Fig. 13.1d

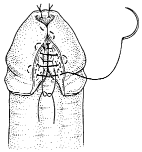
Fig. 13.1e

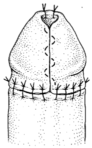
Fig. 13.1f

A Vaseline gauze dressing is applied, which is removed the following day. No stent is required, nor is a catheter for diversion. The preferred age for operation between 6 months and 1 year.

Mathieu procedure (parameatal-based flap)

This technique is utilized for the distal meatus that is about 1 cm from the corona but is wide open and fixed so that it cannot be mobilized onto the glans with a MAGPI technique. These cases must not have any chordee. The ventral skin must be thick so that the parameatally based flap will have an adequate blood supply for mobilization and extension. The subcutaneous tissue should be left attached to the central flap to assure this.

The dotted lines in (Fig. 13.2a) outline the skin flaps. The glans wings are approximated together to create a conical glans configuration (Fig. 13.2c). Skin coverage for this technique is often very difficult. Depicted in the diagram is Byars' ventral rotation (Fig. 13.2c and d), which is most often utilized. It is because of the skin coverage that we generally prefer an onlay island flap technique for such a meatal configuration.

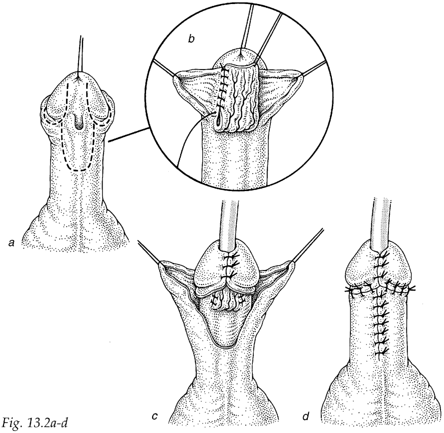

Fig. 13.2a-d

Onlay island flap

This procedure is recommended for cases in which the proximal meatus has a distal urethral plate of good quality. A strip of urethral plate is outlined about 6–8 mm in width out to the tip of the penis, extended on either side of the glanular groove (Fig. 13.3a). The proximal urethra is cut back to good spongiosal tissue (Fig. 13.3b). An onlay of a preputial island flap is then accomplished (Fig. 13.3c and d),

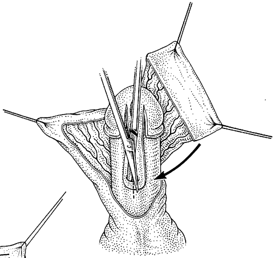

Fig. 13.3d

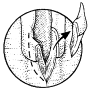

Fig. 13.3b

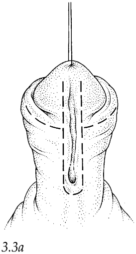

Fig. 13.3a

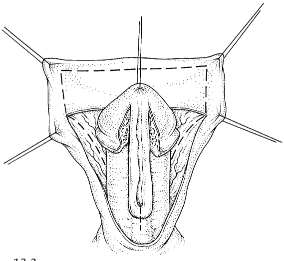

Fig. 13.3c

making a parallel running suture line from the edge of the meatus out to the tip of the glans, rotating the half of an island flap over to the other side with a parallel suture line running up the other side (Fig. 13.3e).

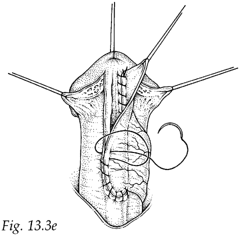

Fig. 13.3e

Around the proximal meatus, interrupted sutures are used and fashioned in such a way that the proximal opening is calibrated at 12–14F. In this modification, it is important to trim the onlay flap to make sure that redundancy does not exist. This is assisted with calibration using *bougies-à-boule*. By extending the meatus out to the tip of the glans in a similar way to the Mathieu technique, glanular wings can be brought around, forming a normal conical configuration (Fig. 13.3f). Skin cover is achieved in the usual fashion.

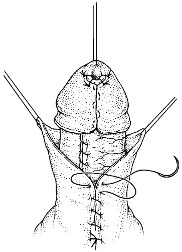

Fig. 13.3f

This technique has been utilized more and more in recent years for the proximal meatus in which the urethral plate is well formed. It is surprising how many times, after a careful dissection of the penile skin, the penis will be perfectly straight on artificial erection, leaving the urethral plate intact. If only a small bend exists, dorsal tucks will alleviate this. In addition, this assures a better vascularized urethroplasty than the meatal-based flap provides.

Transverse preputial island flap urethroplasty with glans channel

This technique is universally useful for one-stage procedures in which chordee release is carried out at the same time as urethroplasty. It is reserved for a urethral replacement of approximately 2.5–5 cm in length by having a vascularized neo-urethra. Healing proceeds without the need for re-vascularization from the skin cover as is required in free skin grafts. The circumferential incision around the corona (Fig. 13.4a and b) should be made as proximally as possible in order to leave a good glans cap of tissue. The urethra and shaft skin are dissected free of the corpora cavernosa to release chordee tethering. It is sometimes necessary to take dorsal tucks on either side of the neurovascular bundle in order completely to correct a ventral bend to the shaft.

Once the shaft is free of chordee, as demonstrated by artificial erection, the proximal native urethra is brought to a comfortable position on the shaft and fixed with 7/0 Vicryl sutures to the tunica albuginea. The skin of the urethral meatus should be excised back to good spongiosal tissue (Fig. 13.4c).

The mobilization of the transverse preputial island flap (as depicted in

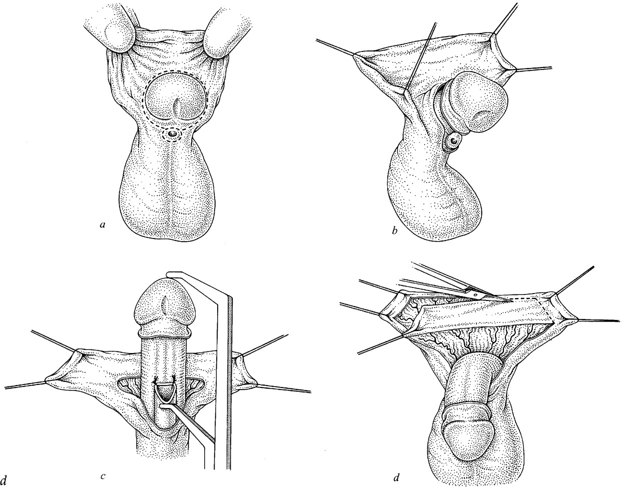

Fig. 13.4a–d

Fig. 13.4d–f) is carried out in such a way as to develop a midline pedicle that is mobilized all the way down to the base of the penis. This can be done with lateral dissections so that the base of the pedicle is narrowed and will not rotate the penis as it is spiralled around to the ventrum. The plane for this dissection is very delineated, leaving most of the vasculature to the flap itself. Since the distal extent of the penile skin will be excised, it is not a problem to have adequately vascularized penile skin for coverage.

The inner transverse preputial island flap is tubularized and fashioned in such way as to create a 12F calibre neo-urethra (Fig. 13.4f). The edges are rolled inwards with Lembert-type sutures. The ends of the tube are held by interrupted sutures, and the middle portion may be stitched by a running suture if desired. This permits the trimming off of the ends in order to tailor the urethroplasty.

The pedicle and tube are spiralled around to the ventrum (Fig. 13.4g and h). The proximal anastomosis is carried out in an open fashion so that a careful approximation of the edges against the fixed proximal urethra may be accomplished (Fig. 13.4g). The anastomosis is covered by the pedicle. The opposite edge of the pedicle may be tethered and may thus cause torsion. This should be relieved by careful dissection, avoiding disturbing the vasculature to the pedicle.

A glans channel is made by dissecting flat against the tunica albuginea of the corpora out to the tip of the glanular groove so that a generous channel sized 16–18F is created. Glanular tissue itself must be removed to make this channel adequate in size. It is not enough to stretch the channel since it will contract back down and compress the blood supply of the pedicle as it lies in the glans channel (Fig. 13.4h). This is the most common cause of technical error in the procedure and can lead to the disastrous consequence of a strictured glanular urethra.

Once the neo-urethra is in the appropriate position, skin cover is obtained by splitting the dorsal preputial skin and bringing around flaps to the ventrum as depicted in Fig. 13.4i.

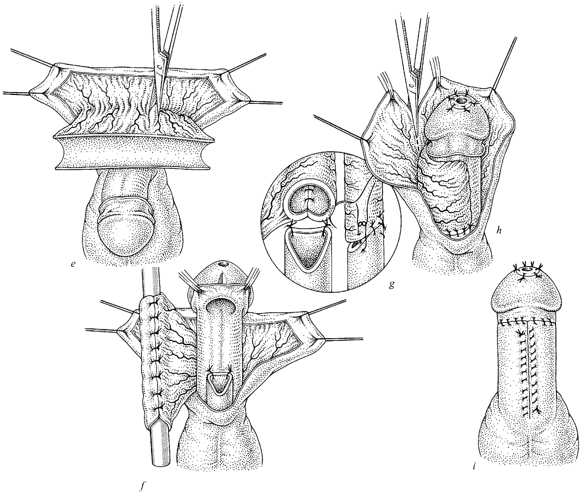

Fig. 13.4e–i

Horseshoe preputial/penile extended island flap for severe cases with marked curvature

In severe cases of penoscrotal hypospadias, the glans may be tethered with a short urethral plate to the scrotum or perineal meatus (Fig. 13.5a). A more extended preputial tubed urethroplasty is then preferred over a two-stage procedure. Figure 13.5b shows the lines of incision around the urethra and urethral plate. The penile and preputial skin is mobilized, with an elongation of the preputial area in the form of a horseshoe (Fig. 13.5c and d) using this technique. A 5–7 cm vascularized inner preputial flap can be elevated and then tubularized using a

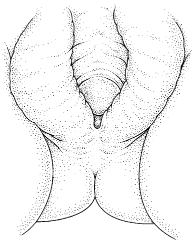

Fig. 13.5a

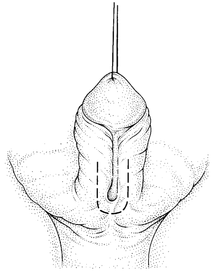

Fig. 13.5b

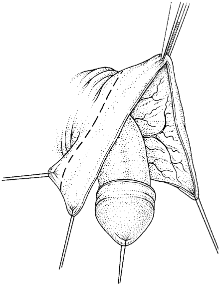

Fig. 13.5c

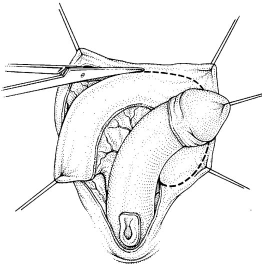

Fig. 13.5d

combination of interrupted and continuous fine PDS sutures (Fig. 13.5e). Interrupted sutures at the ends of the tubularization allow later excision as necessary to achieve the correct length of tube.

A buttonhole of the pedicle in the midline permits this to be brought around the ventrum with a bi-pedicle flap (Fig. 13.5f). The urethra can be stretched and trimmed as necessary so that there is no redundancy. The bi-pedicled flap for a long urethroplasty is shown in Fig. 13.5g. It is important to calibrate the width of the tubed urethra to no more than 10F or 12F so that a diverticulum does not form because of a redundancy of skin. Skin closure is carried out by rotating dorsal penile skin around the ventrum (Byars' flaps). A urethral dripping stent is left in situ for 2 weeks.

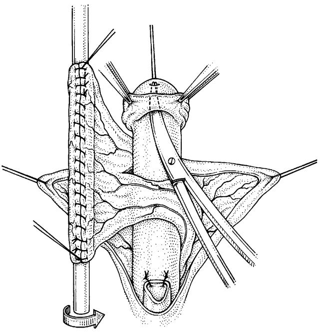

Fig. 13.5f

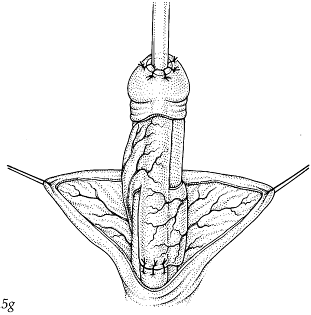

Fig. 13.5g

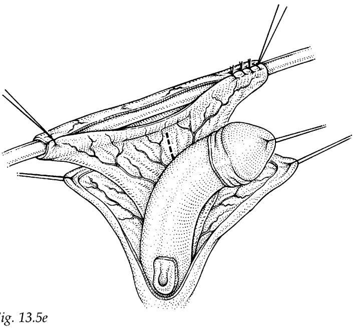

Fig. 13.5e

Correction of congenital curvature using a tunica albuginea plication

In the majority of patients with hypospadias, an artificial erection after adequate skin mobilization will reveal that the penis is straight without any curvature. In approximately 10–15% of patients, particularly those with a severe degree of hypospadias, an erection will, however, show persisting curvature of the corpora (Fig. 13.6a). In this situation, the chordee can be corrected using dorsal plication sutures.

Buck's fascia is elevated by lateral-to-medial dissection on either side of the midline in order to avoid damage to the neurovascular bundle. Two transverse parallel incisions approximately 4–7 mm apart and 5 mm long are made through the tunica albuginea opposite the point of greatest curvature (Fig. 13.6b).

Non-absorbable prolene sutures (approximately 5/0 in infants) are used to approximate the tunica using buried stitches. There is some diminution of penile length using this procedure, but the actual amount of length lost is extremely small and does not appear to be a major problem (Fig. 13.6c).

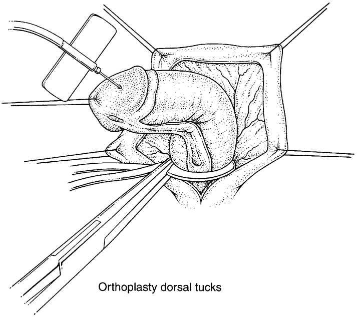

Fig. 13.6a

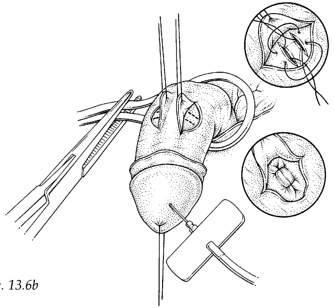

Fig. 13.6b

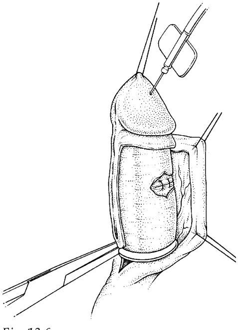

Fig. 13.6c

Points of technique and management

Postoperative follow-up Patients are seen 4–6 days after surgery to remove the dressing and again after 10–14 days to remove the stent. Two weeks after that, the patient returns for an evaluation of the meatus and proximal anastomosis with a *bougie-à-boule* probing. This is not considered to be a dilatation but a calibration to ensure that there are no narrow areas. If the proximal anastomosis cannot be traversed at this visit, no dilatation or stretching is carried out unless the patient is truly obstructed. The patient returns 6 weeks later for an additional calibration and is seen again at 1 year, 3 year and 6 year intervals.

Magnification Optical loupes or the Optivisor have been found to be quite satisfactory during surgery. The optical microscope provides too much magnification to be helpful.

Age This author prefers to operate on children at 6–18 months of age, most commonly between 6 and 12 months.

Sutures Polyglycolic acid or PDS 6/0 or 7/0 sutures are normally used on the internal structures such as the neo-urethra, whereas 7/0 Vicryl is our preferred suture for the skin and meatus.

Instruments Opththalmic instruments such as Castroviejo needle holders and forceps (0.5 mm), iris scissors and fine plastic needle holders are essential.

Diversion A 6F silastic stent is used that goes all the way through the repair and into the bladder for 1 cm. A 5/0 prolene stitch is used to suture the stent to the meatus and glans. About 3–4 cm are left protruding from the tip of the penis, and urine drips continuously into the diaper. A stent will occasionally become plugged, but the patient will void around it without difficulty. Home care of the stent is very easy. For the older child, we prefer this drainage so much that we will put him into diapers for the 2 weeks that the stent is in place. This is tolerated satisfactorily as long as the child understands the reason ahead of time.

Dressing In the past, the author used silastic foam dressings but these are no longer available (although Cavi-care dressings are available in the United Kingdom). Therefore, a Telfa pad over the repair with a bio-occlusive dressing such as Op-Site or Tegederm will suffice. The dressing is left for only 3–4 days and is removed in the doctor's office or at home. No further dressing is left after that.

Outpatient surgery All of our patients are now operated on as day cases. The parents are very happy to take the children home, but if they come from some distance, we will keep them in a nearby hotel for a day or two for observation. The patient and parents are well versed ahead of time in terms of what to expect in home care. Tylenol (similar to a paracetamol elixir) or Tylenol plus codeine usually suffices for discomfort. B & O suppositories (a mixture of belladona and opium, not available in the United Kingdom) are used, divided into thirds for bladder spasms as necessary at home. In addition, we give Septra (Septrin), one teaspoon a day, for the suppression of infection in the open drainage tube. This is continued twice a day for 3 days after the removal of the tube to cover any residual infection. We have not had problems with urinary tract infections using this policy of open drainage.

Chapter 14

Tubularized incised plate hypospadias repair

Warren T. Snodgrass

Distal hypospadias

Today, it appears nearly all cases of distal hypospadias can be corrected by tubularizing the urethral plate, most often utilizing a dorsal midline relaxing incision as depicted in Fig. 14.1. This approach simplifies decision-making and improves the final cosmetic appearance, as the neo-meatus has a vertically orientated, slit-like configuration. The preferred age for surgery is 3–6 months in otherwise healthy patients.

The penis is de-gloved via a circumcising incision made 2 mm proximal to the hypospadiac meatus (Fig. 14.1a). Artificial erection tests are carried out for ventral curvature, which, when present, is straightened by dorsal plication. Next, parallel incisions separate the glans wings from the urethral plate (Fig. 14.1b). In most cases, the plate at this stage is too narrow simply to tubularize into a neo-urethra of sufficient diameter.

The key step in the procedure is a midline relaxing incision, which widens the urethral plate and thereby makes tubularization possible without the use of skin flaps (Fig. 14.1c). This incision extends deeply through the epithelium and underlying connective tissues that comprise the urethral plate to near the corpora cavernosa. A 6F stent is passed into the bladder for postoperative urinary diversion, the plate then being tubularized using 7/0 Vicryl (Fig. 14.1d). Care is taken to avoid suturing the neo-urethra too far distally in order to avoid meatal stenosis, as the neo-meatus should be generously sized and oval in configuration. A dartos pedicle covers the repair (Fig. 14.1e).

Glansplasty begins at the corona and proceeds distally, usually with a total of three stitches (Fig. 14.1e and f). The glans is sutured to the meatus at 5 and 7 o'clock using 7/0 Vicryl. The mucosal collar is then approximated in the midline, and shaft skin coverage is achieved using a variation of Byar's flaps to mimic the median raphe. The stent is maintained for approximately 1 week.

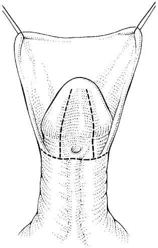
Fig. 14.1a

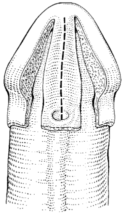
Fig. 14.1b

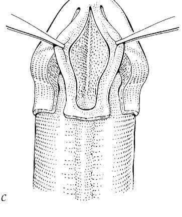
Fig. 14.1c

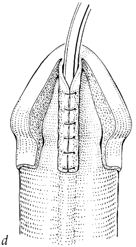
Fig. 14.1d

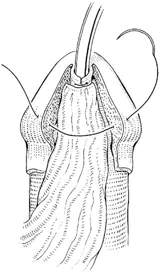
Fig. 14.1e

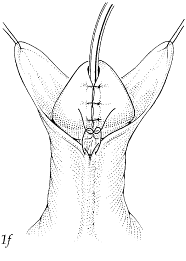
Fig. 14.1f

Proximal hypospadias

Many cases of proximal hypospadias can also be repaired by tubularizing the urethral plate. The main limiting factor is severe ventral curvature, which leads to transection of the plate to facilitate straightening, and, since the degree of curvature cannot be reliably assessed until the penis is de-gloved, it is appropriate always to begin hypospadias repair with an incision that preserves the urethral plate (Fig. 14.2a). Following this U-shaped incision, the remainder of the operation proceeds as described above (Fig. 14.2b–f). Advantages of the tubularized incised plate repair over onlay or transverse preputial island flaps include the avoidance of circumferential anastomoses, the remote likelihood of subsequent diverticulum formation and the superior appearance of the neo-meatus. When the plate cannot be tubularized, we prefer a staged repair over the use of tubularized preputial flaps.

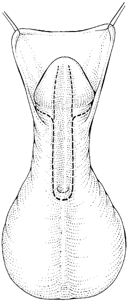

Fig. 14.2a

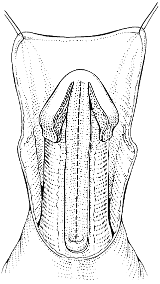

Fig. 14.2b

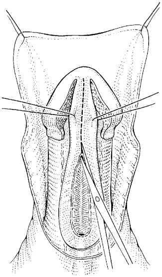

Fig. 14.2c

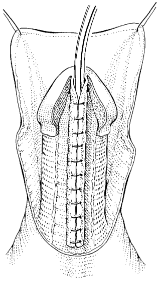

Fig. 14.2d

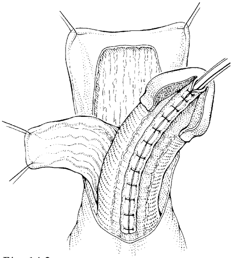

Fig. 14.2e

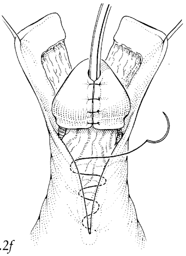

Fig. 14.2f

Chapter 15

Hypospadias free grafts

Andrew J. Kirsch

Introduction

In the correction of both primary and secondary hypospadias, it is preferable to use vascularized preputial or penile skin. When genital skin is unavailable or insufficient, it may be necessary to choose extragenital tissues in order to complete a successful repair. During these difficult salvage procedures, graft alternatives include extragenital skin, bladder mucosa and buccal mucosa.

Although free skin grafts have had a good initial success rate, late complications such as graft shrinkage and stricture formation have prompted reconstructive surgeons to reconsider mucosal biomaterials in an attempt to improve clinical outcome following urethral reconstruction. When using free grafts of any type, the urethral plate should when possible be preserved and onlay grafting, rather than tubularization, performed. It should be recognized, however, that fistula formation occurs more often, and stricture formation less often, when using onlay techniques. The greater success of treating fistulas over strictures may justify this approach.

Indications for surgery

Indications for the use of mucosal grafts include patients with tissue loss owing to prior hypospadias repairs, severe hypospadias variants, epispadias, urethral strictures, and chordee without hypospadias.

Recipient site

Regardless of the technique chosen for urethral reconstruction, the proper preliminary preparation of the recipient site cannot be emphasized too strongly. Residual chordee is frequently encountered and is most often caused by scarring from previous operative procedures. A thorough excision of all ischemic skin and scar tissue is needed to ensure a straight penis and a healthy, well-vascularized recipient site for the neo-urethra.

All remaining urethral tissue from previous repairs should also be sacrificed if there is any question of its viability. During the initial stages of healing, a graft is nourished by the passive imbibing of plasma and tissue fluids, whereas in the later stages an actual penetration of new capillaries into the graft occurs (Converse & Brauer 1964). In a compromised recipient field, as is often the case with a re-operation, bladder mucosa and buccal mucosa may offer a better alternative to free skin grafts.

Bladder mucosal grafts

Memmelaar (1947) first described a one-stage repair of penoscrotal hypospadias utilizing bladder mucosa. Marshall & Spellman (Marshall 1955, Marshall & Spellman 1957) reported their results with a two-stage technique, also utilizing bladder mucosa, for the construction of a neo-urethra; however, the rate of anastomotic stricture formation and fistula was high, and the method was largely abandoned. With improved surgical techniques, these problems have been re-addressed and the complications reduced to an acceptable range. The renewed enthusiasm for this type of procedure is understandable since it avoids the use of extragenital skin grafts altogether and utilizes only urothelium, with which the urologist is familiar.

Surgical technique

The bladder is catheterized and distended with fluid. The anterior wall of the bladder is identified through a Pfannenstiel incision, and sutures are placed to aid in retraction. Using electrocautery or a scalpel, the detrusor is incised to within a few muscle bundles of the underlying bladder mucosa (Fig. 15.1). The use of tenotomy scissors in dividing the remaining muscle fibers exposes the bulging mucosa. Accidental puncturing of the bladder mucosa may be closed with a figure-of-eight absorbable suture; if possible, this area should not be included in the graft.

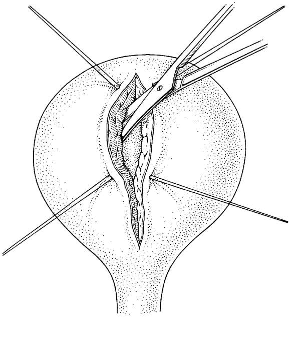

Fig. 15.1

The length of the graft should be approximately 1.5 times the urethral defect to allow for shrinkage during handling (Fig. 15.2). After removing the graft, the bladder is closed over a suprapubic tube to provide urinary diversion. No attempt need be made to cover the denuded area of mucosa since it readily re-epithelializes.

The graft is then tubularized over a red rubber or feeding tube (Fig. 15.3), with the mucosal surface facing inwards, using an absorbable microsuture in a running fashion. The graft should be moistened with saline during its tubularization; delicate smooth forceps are recommended for handling to avoid puncturing the mucosa. The penis is de-gloved, and the proximal urethral opening is circumscribed and freed from the surrounding skin (Fig. 15.4). Partial urethrectomy may be required to remove hypoplastic or scarred urethral tissue.

It the penis is straight with no residual chordee and there is good skin coverage of the penile shaft, the recipient bed can be created by tunneling under the shaft skin from

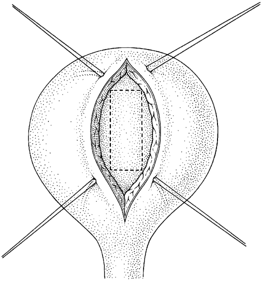

Fig. 15.2

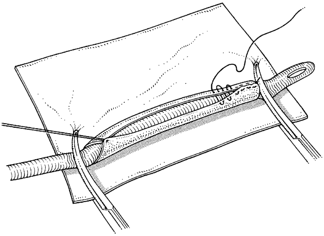

Fig. 15.3

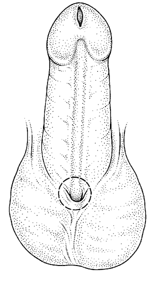

Fig. 15.4

the area of the proximal meatus through the glans itself (Fig. 15.5). This will automatically surround the neo-urethra with a well-vascularized recipient bed. If the skin of the shaft has to be mobilized to release residual chordee or correct a deformity of the penile shaft, it should be incised with a Z-shaped incision to aid in re-establishing the contour of the shaft once the graft has been put in place (Fig. 15.6).

The graft is then laid on the recipient site with the suture line against the corpora and anastomosed to the native urethra using absorbable sutures (6/0 or 7/0) in a running fashion. After incising the glans and creating glans wings, the external urethral meatus is fashioned at the tip of the penis. As an adjunct, the tunica vaginalis can be mobilized and brought over the neo-urethra to provide well-vascularized tissue around the graft (Fig. 15.7). The skin can then be closed in Z-plasty fashion over both the neo-urethra and the buffering layer of tunica vaginalis. The neo-urethra is stented with an indwelling silastic stent, which is sewn in place (Fig. 15.8). A dry sterile pressure dressing is applied to the

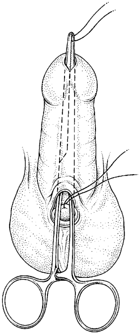

Fig. 15.5a

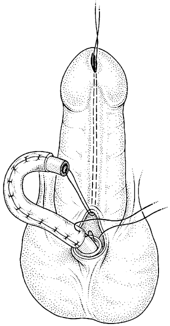

Fig. 15.5b

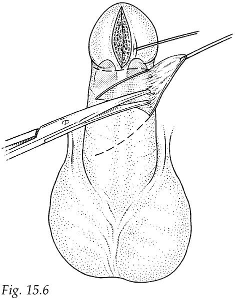

Fig. 15.6

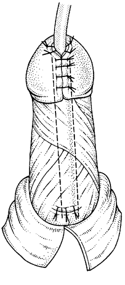

Fig. 15.7

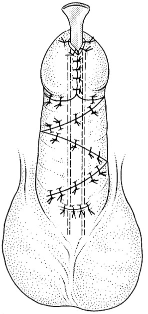

Fig. 15.8

grafted neo-urethra. Urinary diversion is provided by the urethral catheter (and/or suprapubic catheter in selected cases) for approximately 10 days.

When the urethral plate can be preserved without residual chordee, the mucosal graft can be trimmed to the appropriate size to cover the urethral defect. The onlay graft is then sutured in place over a stenting catheter and if possible covered with a blanket of tunica vaginalis or some other buffering vascularized layer (Fig. 15.9).

Buccal mucosal grafts

The use of buccal mucosa for urethral reconstruction is not new. In 1941, Humby used both bladder and buccal mucosa for urethral replacement in hypospadias repair. During this pre-antibiotic era, the risk of infection prompted Humby preferentially to use bladder mucosa.

Currently, buccal mucosal grafts appear to offer several advantages over bladder mucosal grafts. Besides the avoidance of an abdominal scar, the inherent complications of using bladder mucosa, such as urothelial hyperplasia, graft eversion, prolapse and distal stenosis, appear to be obviated with buccal mucosal grafts. In Duckett's review (Duckett et al 1995) of 18 patients undergoing buccal mucosal grafting (16 tubes), with 6–17 months follow-up, meatal stenosis was seen in 5 cases (28%), fistula in 1 (6%), stricture in 1 (6%) and meatal eversion in none (0%). In contrast, Woodard's review (Woodard et al 1995) of 21 patients, followed for 3–25 months, revealed anastomotic stricture in 8 cases (80%), stricture formation being more common in tubes (78%) than onlays (9%). In that series, fistula formation was seen in 6 cases (12%), occurring more in onlays than tubes. Importantly, meatal stenosis and eversion were not noted.

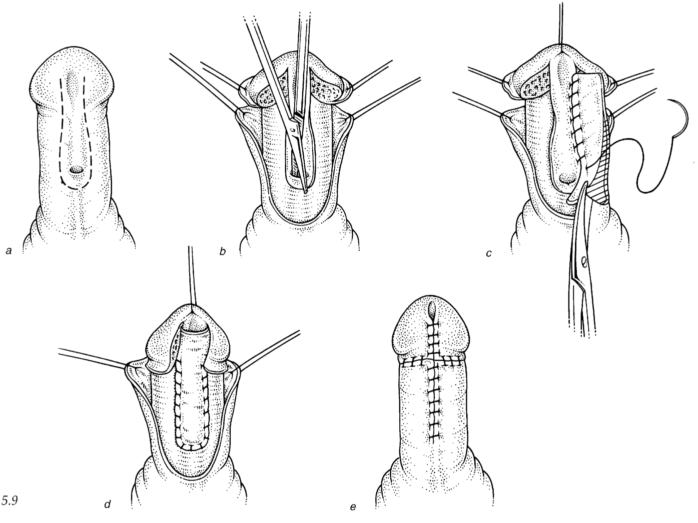

Fig. 15.9

Long-term follow-up and reports from other centers will determine whether the individual technique or the inherent properties of the mucosa contribute to the morbidity associated with buccal mucosal grafts. Nonetheless, the ease of harvesting the tissue and the low incidence of meatal problems associated with buccal mucosa favors their continued use over other biomaterials.

Surgical technique

When preparing to harvest buccal mucosa, no specific antibiotic mouth rinse is required. Either nasotracheal or endotracheal tubes may be utilized, although the former are preferable when larger-size grafts are required. A dental retractor is used to expose the inner cheek (Fig. 15.10; see Chapter 13). The identification and avoidance of Stenson's (parotid) duct below the second to third molar is mandatory. Either the inner cheek or the labial mucosa, or a combination of the two, may be used. The mucosa is injected with lidocaine mixed with epinephrine (1:100 000) for hemostasis. Gauze placed into the pharynx prevents blood entering the trachea or esophagus.

The buccal mucosa does not contract significantly after harvesting and may therefore be cut equal in size to the measured urethral defect. An appropriately sized rectangular incision is made in the buccal mucosa using knife and cautery down to the level of the buccinator muscle. The graft therefore includes both the buccal mucosa and its lamina propria. After cleaning the graft of muscle bundles and fat, it is placed in saline until needed. The mucosal defect may be closed with absorbable sutures, or, if the defect is too large, it may be left to heal by secondary intention. Either method results in excellent healing within a week, with little postoperative discomfort.

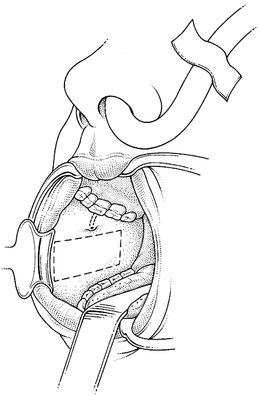

Fig. 15.10

The graft is then tubularized over an appropriately sized catheter, utilizing a cork board for retraction. A running absorbable suture, with several interrupted sutures placed at each end, allows for additional tapering of the neo-urethra if required. The graft is then laid on the recipient bed with the suture line against the corpora, being anastomosed and covered in the same manner as with a bladder mucosal graft. Just as with bladder mucosa, when an onlay technique is utilized, the graft can be trimmed to size and sutured over a stenting catheter (Fig. 15.11). In general, when using any mucosal graft (tubularized or onlay), a stenting catheter is best left in place for 7–10 days.

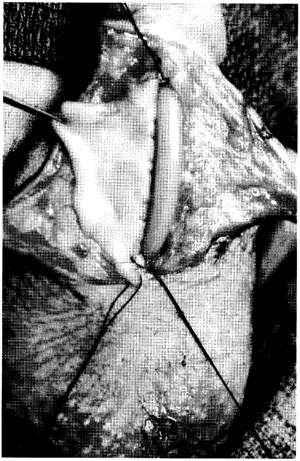

Fig. 15.11

Conclusion

From a technical viewpoint, urethral reconstruction following failed prior operations is a challenging endeavor. The surgeon not only needs to be familiar with the alternative available tissues, but also faces the obstacle of a poor tissue bed to receive the graft. Free grafts from extragenital sites, such as the bladder and oral cavity, to create a neo-urethra remain a viable option in these difficult situations (Kinkead *et al.* 1994). The techniques utilized in free graft urethral reconstruction should be familiar to surgeons contemplating re-operative urethral reconstruction.

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Chapter 16

Surgery of upper urinary tract duplication

J. David Frank

Introduction

This chapter is concerned with the techniques of upper and lower pole heminephro-ureterectomy in cases of ureteric duplication. Conservational surgery avoiding heminephrectomy is also discussed, but vesico-ureteric reflux with double ureters, the local management of an ectopic ureterocele and pelviureteric obstruction affecting the lower hemikidney are discussed elsewhere.

The management of duplication anomalies of the upper urinary tract depends on many factors. These include the presence of a ureterocele, when an endoscopic incision may be all that is required, the percentage function of the upper or lower pole and the presence or absence of lower pole reflux.

Upper pole heminephro-ureterectomy

The kidney is exposed by the surgeon's preferred method. The kidney is mobilized and delivered into the wound. The ligation and division of an upper pole vessel is often required before adequate mobilization of the upper pole can be achieved. The upper pole ureter, which is usually very dilated, is easily visible. The lower pole ureter is normally quite small and found posterolateral to the upper pole ureter. The ureters are mobilized (Fig. 16.1), and the lower pole ureter is separated from the upper pole, which is then mobilized superiorly to confirm that it is passing to the upper pole of the kidney. The upper pole ureter is divided between stay sutures. The plane between the renal pedicle and the upper pole ureter is developed, and the upper pole ureter is then passed behind the vessels (Fig. 16.2). This is then used for traction while further dissection of the upper pole is carried out.

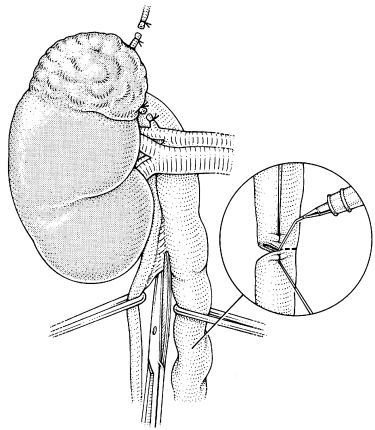

Fig. 16.1

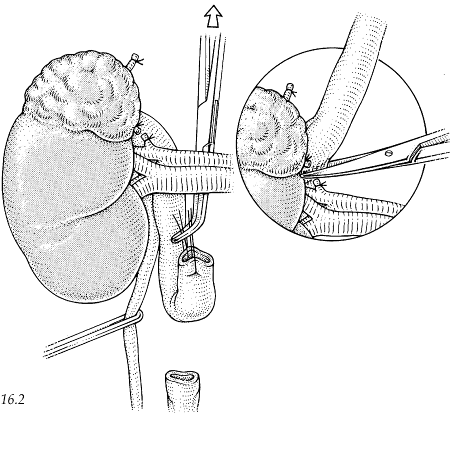

Fig. 16.2

Vessels supplying the upper pole are demonstrated by hylar dissection and divided between ligatures. If there is any doubt concerning the distribution of an artery, it may be necessary to clamp it with a vascular clamp temporarily and observe the effect on the kidney tissue. The renal capsule is incised (Fig. 16.3) above the demarcation line on the anterior and posterior aspects of the upper pole and dissected downwards. The redundant capsule is useful in the later closure of the upper extremity of the remaining lower hemikidney. If the upper kidney is dysplastic and severely scarred, it may not be possible to separate the renal capsule from it.

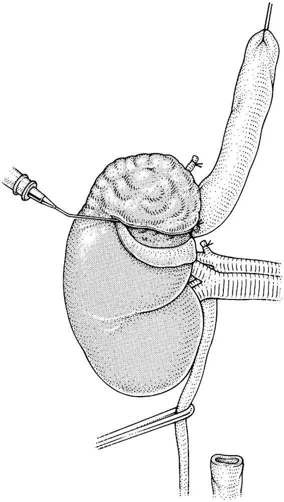

Fig. 16.3

Using the upper pole ureter for traction, the dissection is carried into the renal sinus of the upper pole. The junction between the two hemikidneys is normally easily palpable between the thumb and forefinger. Separation of the hemikidneys (Fig. 16.4) is carried out using a diathermy point electrode on a high setting for coagulation. This helps to coagulate the small vessels while providing a good cutting current. It is better to divide the kidney erring on the diseased side, cutting into the calyces of the upper hemikidney and then trimming these off together with their related parenchyma secondarily.

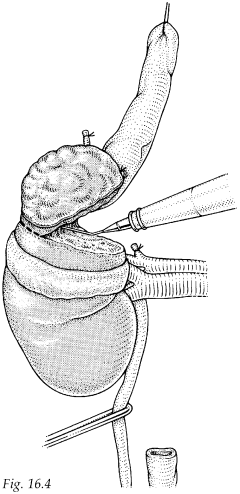

Fig. 16.4

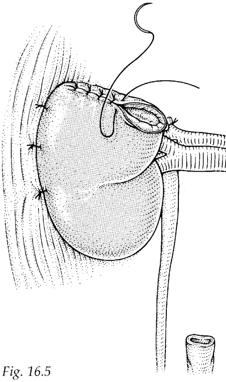

Fig. 16.5

Any major vessels should be underrun with 4/0 Vicryl Rapide. The upper pole is closed using Vicryl Rapide sutures, picking up the reflected capsule of the upper pole. Tying the sutures (Fig. 16.5) opposes the cut surfaces. The hemikidney is now suspended by a narrow pedicle, and, in order to avoid subsequent torsion, the renal capsule should be sutured to the adjacent musculofascial tissues on the posterior abdominal wall.
If closure of the upper pole of the lower hemikidney as described above is impossible, the lower pole may, after the control of any hemorrhage, be left open, and healing will rapidly occur.

The distal upper pole ureter is dissected as far down as can be comfortably visualized. If there is no reflux, the ureter can be divided with cutting diathermy. If, however, reflux exists, the ureter should be ligated using an absorbable suture. The two ureters normally share a common sheath near their entry into the bladder, so the upper pole ureter should be dissected off the lower pole ureter until the common sheath is encountered.

Ureterectomy from the upper hemikidney

When upper heminephrectomy has been performed as the definitive treatment for ectopic ureterocele or for the relief of urinary incontinence when an ectopic ureter opens onto the vulva or vagina, the ureter need be excised only as low as can be obtained within the limits of the abdominal heminephrectomy incision. The same recommendations apply when an ectopic ureter opens into the urethra in either sex and there is no urethro-ureteric reflux. When urethro-ureteric reflux exists, however, incomplete ureterectomy produces what is effectively a urethral diverticulum, which may lead to persistent urinary infection or even to calculus formation in the ureteric stump. Nevertheless, because of difficulties in operative exposure and the risk of harming the vesico-urethral sphincters and, in the male, the ejaculatory apparatus, it is often preferable to carry out an incomplete ureterectomy in the first instance and await events.

When an excision of the stump of an ectopic ureter opening into the urethra or into a cystically dilated seminal vesicle proves to be essential, the preferred exposure is transvesical. Through a lower abdominal incision, the bladder is opened via its anterior wall in routine fashion. A vertical incision is made through the vesicotrigonal wall midway between the ureteric orifices (Fig. 16.6). The ureteric stump and, in the male, an involved seminal vesicle are readily mobilized to their terminations and excised (Fig. 16.7).

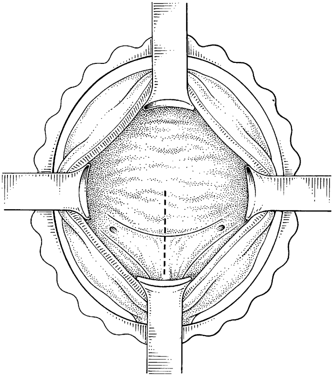

Fig. 16.6

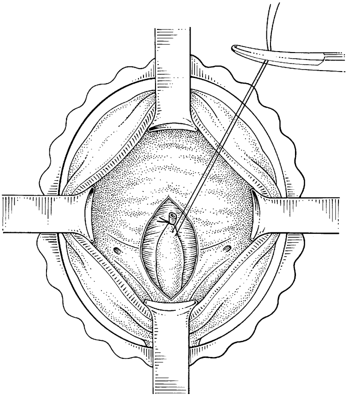

Fig. 16.7

Lower pole heminephrectomy

This procedure is required when vesico-ureteric reflux of the lower pole of a duplex kidney has produced irreparable destruction of the lower hemikidney and the child has presented with symptomatic urinary infections. A cystogram will show a grossly dilated lower pole refluxing ureter associated with extremely poor function of the lower pole on a DMSA scan.

The technique of a lower pole heminephrectomy (Fig. 16.8) is not dissimilar to that of the upper pole technique except that the hydronephrotic calyces of the lower pole excavate into the parenchyma of the upper pole. It is not infrequent therefore that when the lower pole is excised, it is found that the calyces of the lower pole have been cut across. These calyces then need to be dissected out of the upper pole renal parenchyma using sharp dissection. This is not an easy technique, but if it is not performed, a urinoma may result.

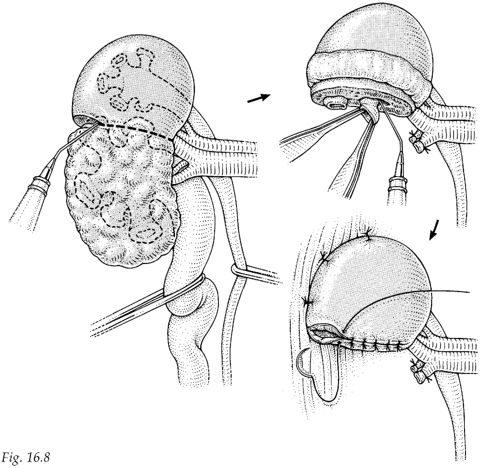

Fig. 16.8

Ureterectomy from the lower hemikidney

Complete ureterectomy is often not needed as leaving a small refluxing stump rarely leads to persistent infection provided that the lower pole ureter has been ligated as close as possible to the common wall of the two ureters. If, however, the patient presents with recurrent infection, a complete ureterectomy may be required as a secondary procedure. A separate lower abdominal incision is required; an oblique, muscle-cutting incision in the iliac fossa, as described for megaureter (see Chapter 3), is appropriate.

The blood supplies of the twin ureters are closely interrelated, careful dissection being needed to define, ligate and divide the vessels going to the ureter that is being removed in order to avoid endangering its fellow. Some 2–5 cm above the bladder, the two ureters fuse to share a common wall; attempts to separate them will almost certainly damage the ureter being retained.

The refluxing ureter should be dissected to and divided at the point of fusion. The stump is then laid open to the level of the bladder musculature, leaving the common wall intact. The resulting vesical aperture is closed with one or two Vicryl sutures (Fig. 16.9), care being taken to avoid occluding or kinking the remaining ureter. Redundant fringes from the incised ureteric stump may be excised, but the removal of its mucosa is not necessary. Drainage of the wound for a few days is advisable because temporary leakage from the bladder is possible.

Conservational surgery in cases of ureteric duplication

Upper ureteropyelostomy

This procedure is indicated as an alternative to upper pole heminephrectomy when the upper pole is shown on a DMSA scan to possess useful function. When this occurs, an intravenous urogram should be performed as this operation is only possible if the lower pole renal pelvis is extrarenal. If the lower pole renal pelvis is almost entirely intrarenal, it is better to carry out a double-barreled reimplantation of the two ureters into the bladder. If there is a reasonable extrarenal lower pole renal pelvis, the dilated ureter from the upper hemikidney is divided and anastomosed end-to-side to the pelvis of the lower pole (Fig. 16.10). Internal drainage to the exterior is usually not needed, but a soft drain should be left to the region of the anastomosis.

Lower pole pelviureterostomy

This technique is applicable, as an alternative to heminephrectomy, to cases of reflux to the lower hemikidney when the parenchyma has sufficient function to be of overall value to the patient but anti-reflux surgery is considered inadvisable because the refluxing ureter is severely dilated. The pelvis of the lower hemikidney is divided and anastomosed end-to-side to the ureter draining the upper hemikidney (Fig. 16.11). Since the latter is ordinarily a very narrow tube, meticulous suturing is essential in order to avoid its stenosis or devascularization. A transanastomotic splint brought to the exterior and nephrostomy drainage of the lower hemikidney are advisable. The lower end of the refluxing ureter is managed as discussed above.

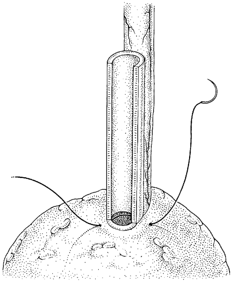

Fig. 16.9

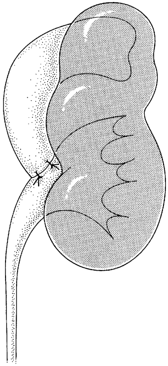

Fig. 16.10

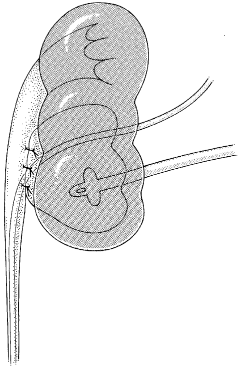

Fig. 16.11

Partial nephrectomy for other conditions

Once the segment to be removed has been identified, either the capsule can be incised with a scalpel and peeled back (it may be adherent if the renal segment is diseased) or cautery may be used to incise the renal parenchyma. Classic teaching is that blunt dissection with the back of a knife handle can be used to separate the parenchyma without disrupting the major blood vessels, which can then be ligated.

As mentioned, if the renal parenchyma is atrophied, it is divided with cautery. If there is considerable renal parenchyma to be divided, the Cavitron Ultrasonic Surgical Aspirator is very effective in dividing the kidney. The parenchyma is separated without dividing the vessels, which minimizes blood loss.

After the partial resection of a kidney with a single collecting system, closure of the transected calyx or infundibulum is required. A watertight closure with fine absorbable sutures (5/0 or 6/0 polyglycolic acid) is recommended. The renal capsule is closed over the cut surface of the kidney. If the capsule is not available, a patch of peritoneum is used.

Ureteropyelostomy in partial duplication

A flank incision is made, the kidney is left in situ, and the ureters are liberated from the renal sinus up to their point of junction. The dilated upper pole ureter is excised exactly where it opens into the lower pole ureter. It is important not to leave any ureter that could act as a diverticulum. The excess abnormal ureter is removed, and the incision in the remaining ureter is closed with interrupted polyglycolic acid sutures. A large end-to-side anastomosis between the upper ureter and the renal pelvis is created so that the end result mimics a bifid renal pelvis without functional obstruction.

Operation for reflux in complete ureteric duplication

As both ureters are bound together in a common sheath at their lower end, they are treated en bloc as a single unit as the two elements may be devascularized in an attempt to separate them. Any of the procedures to correct reflux may be used, either by intravesical advancement or by extravesical uretero-neocystostomy. The classic Cohen re-implantation is most widely used.

Principles and justification

Indications The indications for partial nephrectomy for benign disease have diminished over the years. Treatment for urolithiasis has evolved from an open surgical approach to the use of endoscopic techniques and extracorporeal lithotripsy. The early recognition and treatment of vesico-ureteric reflux and urinary tract infection has lessened the risk of severe segmental renal damage.

The most common indication for partial removal of the kidney in a child is complete ureteric duplication with a non-functioning segment, usually involving the upper pole. These anomalies are increasingly being diagnosed in the antenatal period. The upper pole moiety becomes dilated because of obstruction secondary to ureterocele or ectopic insertion of the ureter.

The evaluation of these patients consists of cystography, renal ultrasonography and radionuclide scans. This will in most cases provide the correct diagnosis and enable the clinician to decide upon the proper therapy. There are several treatment alternatives, including endoscopic incision of the obstructing ureterocele, ipsilateral uretero-ureterostomy and pyelo-ureterostomy. The merits of each approach continue to be debated, but, for non-functioning upper pole segments, partial nephrectomy with segmental ureterectomy is the favored approach.

Other less common indications for segmental resection of the kidney are trauma, renal cysts and calyceal diverticula. The last two problems are now frequently managed with endoscopic or percutaneous techniques.

Preoperative management The delineation of the anatomy of duplication anomalies can be readily accomplished in most cases by preoperative imaging. If the anatomy remains unclear, antegrade pyelography, retrograde ureterography or puncture of the ureterocele with the retrograde injection of contrast medium can be performed. When the child is brought to the operating theatre, endoscopic examination should be performed to assess the lower urinary tract.

Anesthesia General anesthesia is used in all cases. Most centers now use caudal or epidural anesthesia to assist in pain control after the operation. An intercostal nerve block can also be placed during the operation to assist in pain management.

Partial nephrectomy for duplication anomaly

In most cases, the dilated upper pole ureter is readily identified. Caution must be applied in that the lower pole ureter may also be abnormal; division of the ureter is not undertaken until the origin from the upper pole has been assured. After transection of the ureter, the collecting system is decompressed. This allows the junction between the upper and lower renal segment to be readily identified. In most patients, the ureters can easily be separated at the renal level. With careful dissection, the upper pole ureter is freed from its mate and transposed above the renal hilar vessels. The maneuver allows access into the renal sinus between the two collecting systems. Traction on the upper pole ureter should be minimized to avoid injury to the renal vessels.

The author prefers to incise the renal parenchyma with cautery in those cases in which it is dysplastic and/or atrophied. If a branch of the renal artery is clearly seen to enter the parenchymal segment being removed, it can be ligated and divided. If there is any question over the region supplied by an upper pole arterial branch, a vascular clamp can be placed and the line of demarcation observed. Removal of the cystic non-functional upper pole segment can generally be accomplished with minimal blood loss. After the upper pole has been excised, suture ligation of the bleeding points can be performed. Gentle manual compression of the parenchyma is all that is necessary to control bleeding. Mattress sutures are placed to approximate the cut edges of the renal parenchyma. Perirenal fat can be placed in this defect if it is too bulky to fold over.

When the upper resection is complete, the ureter is traced distally. Care should be taken to avoid devascularization of the lower ureter. The plane of dissection should be very close to the upper pole ureter. If the ureters are closely adherent, a strip of the upper pole ureter can be left attached to the lower pole ureter, but this is rarely needed. The ureter is removed as low as possible via the flank incision. This will be below the level of the iliac vessels in most infants and small children. The ureter is then transected, and a catheter is passed into the stump of the ureter. This is irrigated with antibiotic solution, and the catheter is aspirated. The ureter is left open except in rare cases of reflux into the upper pole moiety. Complete excision of the distal ureter or very low ligation is more appropriate in these patients. A drain is placed both in the renal bed and near the region where the ureter is transected in the pelvis.

Chapter 17

Bladder surgery for ureterocele reconstruction

Howard M. Snyder

Introduction

The type of surgery to be described in this chapter (see Fig. 17.1) is rarely needed for single-system ureteroceles as endoscopic puncture nowadays usually produces excellent decompression with preservation of a flap valve to the collapsed ureterocele, which avoids reflux. Associated contralateral reflux is less frequent, an endoscopic procedure generally sufficing. Today, open surgery usually is necessitated by the persistence of associated reflux after the adequate endoscopic incision of a ureterocele in a duplex renal unit.

The advantage of previous endoscopic decompression of the ureterocele is that the attached ureter decreases sufficiently in size that it may later be reimplanted into the bladder with an anticipation of approximately the same success rate that would be achieved for any ureteral reimplant. Poor function of the upper pole is not of significance. If there is no obstruction and no reflux, the reimplanted ureter is generally seen at follow-up to do very well

There are rare cases of ureteroceles in duplex systems in which the amount of associated upper pole ureteral dilatation is minor. In situations such as this, a primary operation without endoscopic puncture is feasible and appropriate.

With time, it has become recognized that the presence of a ureterocele distorts the trigone and bladder neck. This is more true for ectopic than intravesical ureteroceles. It is likely that the disturbance of the trigone is the reason for the high incidence of associated reflux with ureteroceles, which can be summarized as being approximately 50% into the ipsilateral lower pole, 25% into the contralateral side ureter and approximately 10% into the ureterocele itself. The distortion of the bladder outlet is also likely to be the reason for the approximately 5% long-term incidence of urinary incontinence that will follow the decompression of an ectopic ureterocele by upper pole partial nephrectomy with no surgery at the bladder level. Thus, reconstruction of the trigone and bladder neck are very important to master in the care of children with ureteroceles.

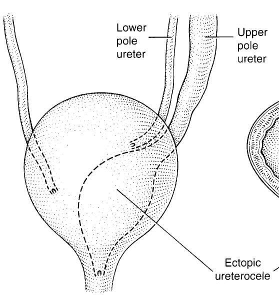

Fig. 17.1a

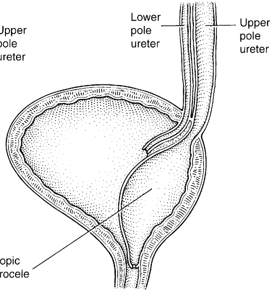

Fig. 17.1b

Operation for intravesical ureterocele

This type of surgery is usually straightforward. The only change from the practices of the distant past is that nowadays the entire ureterocele is virtually always excised, with a detrusor reconstruction. In the past, if there was good backing, the back wall of the ureterocele would at times be left in place and surgery carried out on that surface. If the excision of an intravesical ureterocele is required, the amount of surgery on the trigone usually requires the reimplantation of ureters from both sides.

Bladder operation for ectopic ureterocele

As mentioned above, this is usually carried out after endoscopic puncture and is driven by associated vesico-ureteric reflux. This can be anticipated to be needed in approximately one-third of patients with an ectopic ureterocele and approximately one-quarter of pat with an intravesical ureterocele.

The abdomen is opened by a rout Pfannenstiel incision, and the bla is vertically incised utilizing a De Browne retractor for exposure (Fig. 17.2). A pack in the dome of bladder draws the trigone up int field of surgery. The ureterocele i

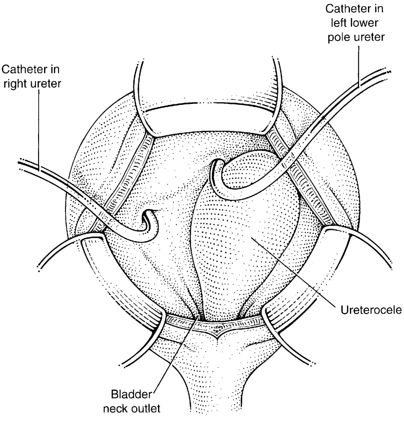

Fig. 17.2

opened between stay sutures (Fig. 17.3). Care is taken to open the distal end of the ureterocele to the bottom of its extension into the bladder neck or urethra (Fig. 17.4). This facilitates the removal of the distal portion of the ureterocele. All but a small rim of the side wall of the ureterocele is excised.

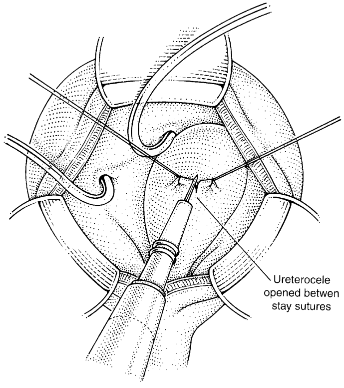

Fig. 17.3

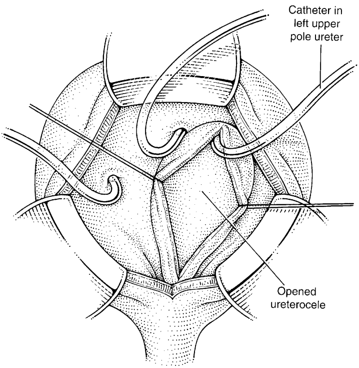

Fig. 17.4

Epinephrine 1:100 000 is injected in the wall of the ureterocele to facilitate hemostasis and assist in dissecting the back wall of the ureterocele to remove its mucosa. Dissection usually begins with the bladder neck and urethral extension (Fig. 17.5). The guide to back wall excision is the residual lateral walls of the ureterocele at their point of insertion into the bladder neck or urethra. This reflects the extent of potential bladder neck and urethral distortion by the ureterocele and provides a guide to reconstruction.

Once the more difficult excision of the mucosa of the ureterocele extending into the bladder neck and urethra has been accomplished, the remainder of the excision of the ureterocele back wall proceeds easily, exposing the detrusor for later reconstruction (Fig. 17.6). As the ureterocele is completely mobilized, the upper pole and lower

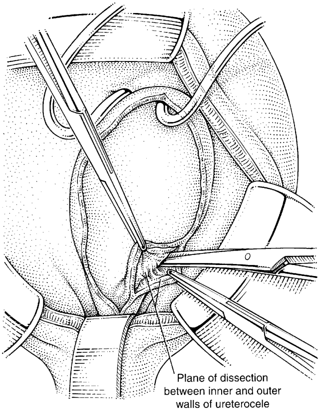

Fig. 17.5

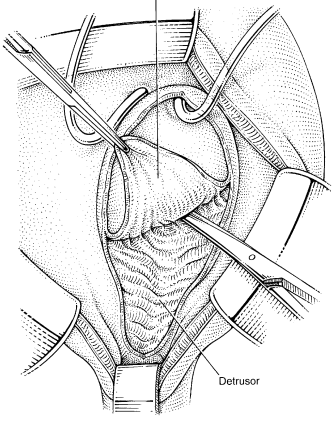

Fig. 17.6

pole ureter on the ipsilateral side are mobilized together to prepare for their later reimplantation (Fig. 17.7).

The keeling reconstruction of the bladder neck and urethra is the key to the successful functional reconstruction of the bladder neck and urethra after ureterocele excision. This technique, which was suggested by Douglas Stephens, reconstructs the annular function of the bladder neck and urethra to the functional integrity that would have been anticipated had there been no ureterocele present in the first place. The difficulty with this step is the small diameter of the bladder neck and urethra in the child. A hernia retractor or small malleable retractor used to elevate the front wall of the bladder neck is useful both in ureterocele excision and in the reconstructive steps to be taken (Fig. 17.8).

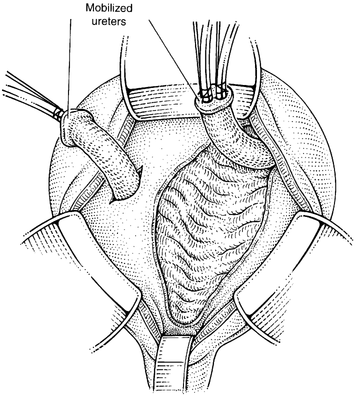

Fig. 17.7

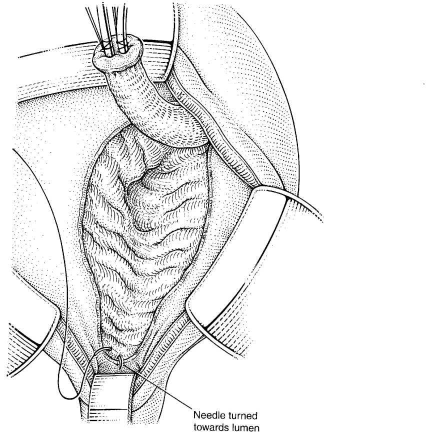

Fig. 17.8

A small tightly curved non-cutting needle with Dexon or Vicryl is used. The first stitch is placed as distally as possible. After taking a solid bite of detrusor or bladder neck tissue, the needle is turned toward the lumen, which makes it easier to retrieve (Fig. 17.8). This is done on each side with no concern about crossing sutures (Fig. 17.9). After the first stitch is tied, it is held cephalad as this increases the distal exposure of the bladder neck and urethral tissue that must be approximated in the reconstruction. Subsequent sutures are placed lateral to the first stitch and at least as distal if not more so (Fig. 17.10).

The goal here is to reconstruct the bladder neck and urethral tissue until the mucosal edges that mark the lateral extent of the ureterocele are drawn to a perfect apposition. This keeling reconstruction will complete the reconstruction of the continence mechanism of the bladder neck and urethra with the elimination of any distortion previously caused by the ureterocele.

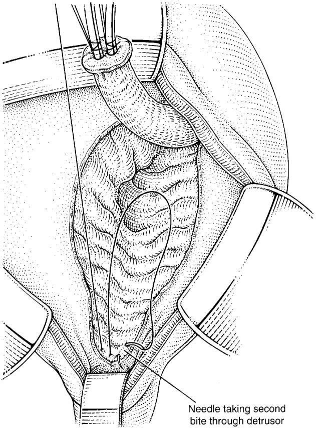

Fig. 17.9

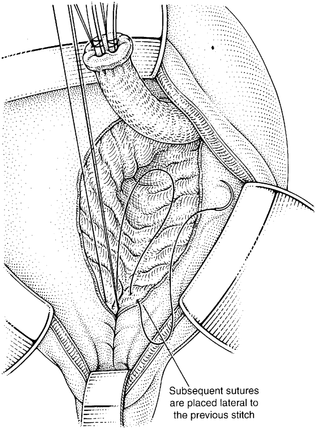

Fig. 17.10

So far, in a 20 year personal experience utilizing this technique of bladder outlet reconstruction in ureteroceles, there has not been a patient who failed to void spontaneously immediately after surgery, and there has been no problem with urinary incontinence. Longer follow-up into adulthood will, however, be required to confirm these good results.

Next, the trigonal distortion created by the ureterocele is reconstructed by drawing the detrusor together again following the limits defined by the lateral walls of the ureterocele (Fig. 17.11). As this reconstruction takes place, it becomes readily evident why it is not necessary to preserve much of the wall of the ureterocele for the mucosal repair of the bladder lining.

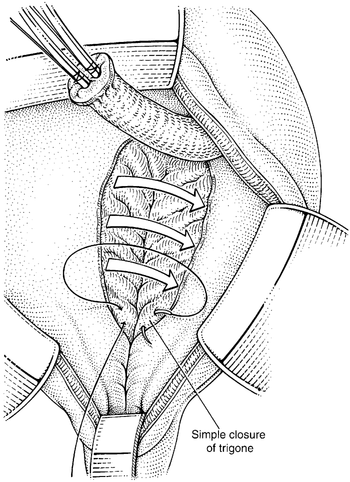

Fig. 17.11

A routine Cohen cross-trigonal reimplant is then completed, usually for the ureters on both sides (Figs 17.12 and 17.13). If the contralateral ureter is at some distance from the ureterocele, a simple Gil–Vernet type advancement is occasionally carried out. If either ureter appears to be marginally dilated, a Starr imbrication is carried out to ensure that a rate of at least 5:1 of length to diameter of the ureter is achieved for the reimplantation. This is usually not necessary after the endoscopic puncture of a ureterocele has resulted in good decompression.

Postoperative care

Any abnormal ureter or ones that have been tailored are stented. A small-caliber urethral Foley catheter is utilized for 48–72 hours. Suprapubic tubes are no longer employed. It is important to warn the family that there is, after this type of bladder neck and urethral surgery, more dysuria and frequency than is seen with a simple reimplant. At the Children's Hospital of Philadelphia, we have found that the slow infusion of morphine directly into the bladder takes advantage of the opiate receptors that exist in the bladder and thus helps substantially to reduce a child's dysuria. Frequency however, continues. Families can generally be reassured that the voiding pattern will return to normal in 10–14 days.

If a ureteric stent has been utilized, a low-pressure study via the stent to ensure free drainage around the stent into the bladder is carried out at about 14 days postoperatively before stent removal. A micturating cystourethrogram is usually performed 6 months after surgery, antibiotic prophylaxis being continued until that point.

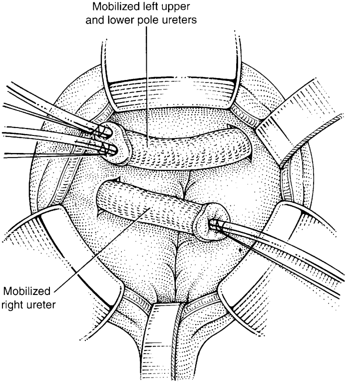

Fig. 17.12

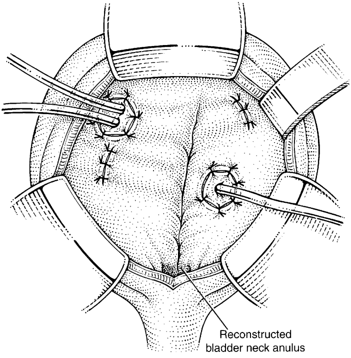

Fig. 17.13

Chapter 18

Osteotomy in bladder exstrophy

Gordon A. McLorie and Antoine E. Khoury

Bladder exstrophy involves the abdominal wall and the bony pelvic ring, along with the urinary tract, including the bladder, urethra and genitalia. The defect has been successfully repaired in infancy for several decades by surgeons around the world (Gearhart & Jeffs 1989, Sponseller et al 1995). The closure of the abdominal wall is usually performed shortly after birth and includes the repair of the urinary bladder. The primary goals of early therapy involve the abdominal wall, pelvis and urinary bladder, this treatment setting the stage for the subsequent challenges of rehabilitation: the provision of suitable bladder and urethral tissue that allows the preservation of the upper urinary tract in addition to the obvious defect of continence, along with suitable functional genitalia in both males and females.

Authors who have achieved most success in this long-term management have utilized some form of pelvic osteotomy in the initial stages of the repair. Although infants can be closed successfully during the first 48 hours of life without osteotomy, better continence can be achieved with greater certainty when the pelvic deformity is corrected to create a more complete pelvic ring. At the time of closure in infancy, the urogenital diaphragm, with its somatic musculature in the anterior urogenital triangle, can be positioned to provide a more circumferentially supportive area of somatic musculature. This enhances the ability to control voiding spontaneously and to achieve a 'stop and start' urinary stream.

It also appears that pelvic osteotomy may provide distinct advantages in repeat surgical procedures to achieve the closure of prolapsed and failed initial repairs. Ansel (1979) is credited with confirming that closure can be achieved early in infancy without osteotomy. However, only 8 out of his 28 patients were closed in this manner, the remaining 19 having had bony reconfiguration as part of their repair.

Our enthusiasm for the use of pelvic osteotomy stems from the early successes reported at our institution by Jeffs (1978) in early closure and his subsequent successes in achieving urinary continence. The initial procedure to fashion the pelvic ring involved bilateral posterior osteotomies. In an attempt to refine the methodology, we have studied the malformation by means of a model that we created from a three-dimensional computed tomography study of a child with as yet unclosed exstrophy (McKenna et al 1994). The model was then reconstructed in plaster and was used to replicate the various means of osteotomy. The configuration of the pelvis and the areas of thickest and thinnest bone, as well as the manner in which each different osteotomy method achieved its objective, that is, a circumferential closure of the pelvic ring with a minimal pubic diastasis, were studied in this manner. Sponseller et al (1995) have also carried out clinical studies using age-matched controls and three-dimensional computed tomography scans to define the geometric components of the defect, along with the manner of successful closure.

All proponents of osteotomy as an adjunct to the primary and/or secondary closure of bladder exstrophy have agreed that this procedure is a factor in the success of both abdominal wall and bladder closure, and has an implied benefit in the improvement in urinary continence. The decrease in the pubic diastasis must also have some beneficial effect on genital reconstruction, although there are few data available to support this hypothesis.

There are four different primary approaches to pelvic osteotomy: bilateral posterior osteotomy (Jeffs 1978), anterior diagonal mid-innominate (Toronto) osteotomy (McKenna et al 1994), anterior innominate (Baltimore) osteotomy (Gearhart et al 1996) and osteotomy of the superior pubic ramus (Sponseller et al 2000). The proponents of all the different methods stress the safety and efficacy of the orthopedic component of these repairs. The posterior and anterior iliac approaches result in a change of the alignment and rotation of the hips, but this appears to have little functional significance. There were some early reported neurologic complications following the anterior innominate procedure, but these resolved in all patients, and further reports suggest that these problems have not been encountered in subsequent cases.

Each of the osteotomy methods has different advantages and disadvantages, but all require a degree of collaboration between the orthopedic and urologic surgeons. Common to all is a need for various methods of postoperative immobilization to ensure bony healing, although this aspect is possibly a still greater challenge in those children who undergo closure of an exstrophic bladder without the assistance of osteotomy. We shall review the various methods of osteotomy and discuss some of the unique aspects attributable to each.

Posterior iliac osteotomy

This technique was the first, proposed in the mid-1950s, and was adopted by urologic surgeons wh
o achieved the first consistent successes in repair of infants (Fig. 18.1). Although initially carried out at a separate time from the abdominal wall and bladder closure, it was soon accepted that the two approaches could be combined simultaneously. A distinct disadvantage of the posterior approach is, however, the need to operate on children in the prone position for the orthopedic component.

The child is prepared and draped in a completely circumferential manner and placed in the prone position, the two incisions being made over the sacro-iliac joints (Fig. 18.1). Both plates of the ileum must be divided completely, a feature of both the anterior and posterior osteotomies. Some authors have suggested the division of the sacrotuberous ligament to be an important feature in that it allows a more medial rotation of the anterior aspect of the pelvis, but other supporters of the posterior technique do not feel that the sacrotubular ligaments play an important role in the repair. Subsequent to the osteotomy, the wounds are closed, and the child is turned to the supine position for the abdominal and bladder closure.

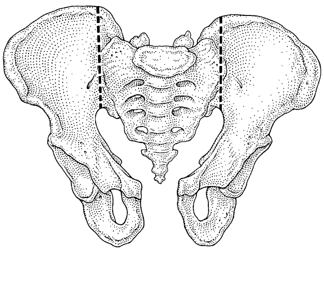

Fig. 18.1

Recent studies (Sponseller et al 1995) have compared the rotation and alignment of the hip joints along with gait analysis in patients who have had this form of osteotomy with that seen in patients who have had either no osteotomy or a division of the superior pubic ramus. They found that the latter groups showed few differences, whereas the anterior osteotomy population had more normal findings with respect to their hip alignment.

Postoperative fixation is most often accomplished by gallows traction in which the lower limbs are suspended to allow flexion in internal rotation of the hips, and the sacrum is elevated off the bed. This traction is maintained for 3–6 weeks. In infants and children under 24 months of age, skin traction may be employed for the suspension device. In older children, pins through the distal femurs are a better means of maintaining traction, the knees then being placed in a flexed position with the calves resting in slings.

Combined anterior innominate and vertical iliac (Baltimore) osteotomy

Reservations about several orthopedic aspects of posterior iliac and anterior innominate osteotomy alone led Sponseller & Gearhart to explore an alternative approach, combining the two via a purely anterior approach. Their reservations included the inability of the posterior approach to facilitate adequate medial rotation of the pelvic fragments without tension, and the observation that an asymmetric displacement of the fragments resulted in inconsistent asymmetry and overlapping of the bone plates. Sponseller & Gearhart also sought an osteotomy approach that would allow proper remodeling of the pelvic bones yet maintain the patient in the supine position, thus avoiding the need to turn and reposition the patient during the operative procedure. Therefore, in 1996, they reported the use of a new combined osteotomy in 32 patients with classic bladder exstrophy. Recently, Sponseller et al (2000) have reported their long-term experience with 85 patients with exstrophy using this combined technique, showing excellent results.

To perform this combined pelvic osteotomy procedure, the infant or child is placed in the supine position with the body circumferentially prepared, draped and exposed from the nipples distally to the foot. A small towel is placed beneath the sacrum, and the bladder is covered with sterile absorbent towels. Oblique incisions are made just inferior to the anterosuperior iliac spine, as described for the Salter osteotomy. The ilium is exposed through the tensor sartorius interval. The subperiosteal iliac wing exposure should continue posterior to the lateral border of the sacroiliac joints and inferior to the pectineal tubercle. The notch is carefully exposed subperiosteally. Anterior innominate osteotomy is achieved by pulling a Gigli saw interiorly to create an osteotomy from the upper edge of the sciatic notch to a point halfway between the anterosuperior and anteroinferior spines, which leaves enough bone in the distal fragments to achieve fixation with pins and to correct the anterior segment (ischiopubic) deformity (Fig. 18.2).

Posterior vertical osteotomy is performed to correct the external rotation of the iliac wings, which is done by an osteotomy just anterolateral to the beginning of the sciatic notch. A small bone rongeur is placed in the cancellous bone created by the first osteotomy and used to create a closing wedge osteotomy from this point vertically parallel to the sacroiliac joint. The posterior cortex of this osteotomy is left intact. This procedure is performed gradually so that the ilium does not crack. The cancellous chips from this osteotomy are left in place to promote healing.

The iliac wing should be very malleable but not completely free.

Two threaded fixator pins are placed in the inferior fragment, one just posterolateral to the anteroinferior iliac spine and one posterior to it. Both pins should enter the ilium anterolaterally and exit posteromedially. The tips of the pins should be visible on the inside of the ilium in the inferior fragment. The two superior pins enter the ilium superiorly and remain within the two cortices of the ilium. X-rays are taken to verify satisfactory pin placement before the bladder, posterior urethral and abdominal closure. Following soft tissue closure and the completion of the urological procedure, the external fixator is placed to buttress the pelvis in the corrected position. The usual construction is to have two long bars bridging the inferior pins and the two shorter bars connecting the superior to the inferior set of pins.

The patient is maintained in a supine stationary position until the osteotomy heals. This involves only minimal skin traction in a nearly horizontal position for approximately 4 weeks in infants and 6 weeks in older children, especially re-closure patients. A flat X-ray is obtained 10 days postoperatively. If the diastasis is not completely corrected initially, it can be gradually corrected at the beside in the succeeding days by a gradual approximation of the inferior fixator bars. Meticulous pin care by the nursing staff is required while the child is supine for 4–6 weeks.

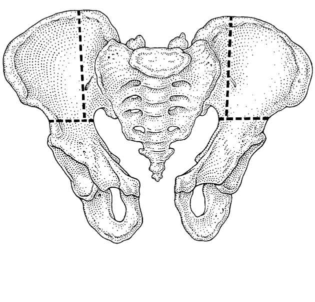

Fig. 18.2

Anterior pubic ramus osteotomy

This osteotomy is much more easily accomplished since it is carried through the anterior abdominal wall incision and involves much less time and surgical exposure. It does not, however, achieve a re-configuration of the entire bony pelvis, and comparative studies have shown minimal change in this respect from the unchanged exstrophy pelvis (Sponseller et al 2000). The operative technique includes the exposure of the superior ramus of the pubis lateral to the insertion of the rectus muscle, after incising the lateral border of the rectus sheath (Fig. 18.3). The periosteum of the superior ramus is elevated, and a spatula is inserted along this space into the obtruder foramen in order to protect the obtruder nerve. The optimal site is medial to the insertion of the inguinal ligaments, the superior ramus being completely divided with scissors, a chisel or, in older children, an oscillating saw. The procedure is performed bilaterally, on each superior ramus.

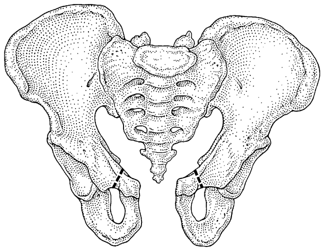

Fig. 18.3

This technique certainly frees the rectus muscles so that they close more easily, but tense strapping or wrapping of the lower pelvis is required to ensure healing. We feel that this method of postoperative immobilization compromises the postoperative healing of the repair, since the constant presence of urine may lead to maceration of the skin and postoperative wound infection. The coexistence of external dressings that apply the tension necessary to maintain the tissues in a closed position along with the presence of urine and/or stool does not provide the best milieu for care by either nursing staff or parents in the early stages of the postoperative period.

Anterior oblique mid-innominate osteotomy

This technique appears to us to fulfil the greatest number of goals, including applicability to both primary and repeat repairs, adaptability to all ages and sizes, the ability to facilitate all degrees of pubic diastasis and an avoidance of the need for external fixation. This technique can also be performed through either a single anterior operative site or two separate anterior incisions.

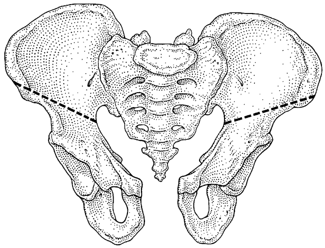

Fig. 18.4

The technique evolved from our model reconstruction from the three-dimensional computed tomography of an exstrophy patient's pelvis (see above). This model revealed that the transverse (Baltimore) osteotomy was excessively close to the tri-radiate cartilage of the acetabulum and through the thickest bone of the ileum. Our technique goes through the thinnest bone of the ileum and, upon rotation, leaves the greatest area of articulating bone, thus allowing firmer and more rapid dense fixation (Fig. 18.4). Our model also demonstrated extensive overriding of the fragments with a posterior osteotomy and showed that the superior pubic rami division did not allow sufficient flexibility of the entire pelvis, as is needed in older children. Our technique also has applicability in younger infants, a controversial issue, because it does not rely on external fixation as pins are not well maintained in the thin tables of bone in infancy.

The technique involves the mobilization of the anterior skin flaps until the anterior superior iliac spines have been identified. Bilateral separate skin incisions may be chosen for this exposure. The incision is made in the periosteum approximately 1 cm posterior to the iliac spine. The periosteum is then raised in a direction aimed towards the greater sciatic notch. A diagonal incision is made through both tables of the ileum using osteotomies or a reciprocating saw. It is imperative to divide ridges of cartilage at the level of the greater sciatic notch. When the technique has been correctly completed, the two pieces of the ileum are freely mobile and the pelvis rotates easily to allow a tension-free closure of the rectus muscles down to the site of their insertion into the symphysis pubis bilaterally. Postoperative immobilization is most effectively accomplished, and Gallows traction is employed in a manner similar to that used in the posterior osteotomy approach.

Summary

As we have outlined, there are several methods of performing an osteotomy during the closure of the abdominal wall and to facilitate the immediate and long-term reconstruction of the patient with bladder exstrophy. We believe that this adjunctive therapy is an advantage in all children, including infants, even though we recognize the controversy surrounding the need to perform osteotomy in infant closure. The cosmetic effect in infancy is facilitated by a midline closure instead of extensive flaps. An anterior exposure allows an easier operative exposure without the need to turn the patient during anesthesia. Long-term data suggest, but certainly cannot yet conclusively confirm, the benefits of osteotomy and the outcomes with respect to urinary continence and genital appearance and function. Accordingly, we recommend that osteotomy be considered as an adjunct to the initial closures or, when necessary, to repeat closures in all cases of bladder exstrophy.

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Chapter 19

Surgery of classic bladder exstrophy and epispadias

John P. Gearhart

Newborn exstrophy closure

With modern prenatal ultrasound techniques, the prenatal diagnosis of bladder exstrophy is not difficult. Important observations include:

1. an absence of bladder filling;
2. a low-set umbilicus;
3. a wide pubic ramus;
4. diminutive genitalia;
5. a lower abdominal mass that increases in size as the pregnancy progresses.

The prenatal diagnosis of bladder exstrophy allows for optimal prenatal management, including delivery in a pediatric center prepared to handle this complex malformation and appropriate prenatal counseling of the parents as to the complex nature of this congenital malformation and their fetus.

After delivery, the infant and the exstrophied bladder must be evaluated in the newborn nursery by a surgeon who is experienced in dealing with this condition. The potential for success must be carefully evaluated at birth before an operation of this magnitude is undertaken. A good cardiopulmonary and general physical assessment can be carried out in the first few hours of life. A renal ultrasound should be obtained to provide evidence of renal structure and drainage, even in the first few hours of life before the child undergoes closure of the exstrophy defect. If the child does not undergo immediate closure, the bladder should be covered with a plastic membrane and the bladder irrigated with sterile saline each time the diaper is changed until the child undergoes surgical repair.

Babies born with bladder exstrophy should have the bladder closed within the first 48 hours of life unless there are medical contraindications to surgery. The presence of the maternal hormone relaxin makes the pelvic bones flexible, and the pubic symphysis can often be approximated without pelvic osteotomy. If there is a wide pubic diastasis greater than 4 cm or if the baby is more than 72 hours old, it is, however, unlikely that closure can be undertaken without the use of pelvic osteotomy. If there is any doubt in the surgeon's mind about the suitability of closure without osteotomy, an osteotomy should be performed. Therefore, perioperative monitoring of the baby and appropriate cautions to maintain normal body temperature are essential as this is a lengthy operation. If an osteotomy is performed, a blood transfusion will often be needed.

The baby is prepared from the nipples to the knees anteriorly and from the tip of the scapula to the popliteal fossa posteriorly, the legs being wrapped in sterile towels. This is important so that pressure can be placed on the greater trochanters in order to bring the pelvic bones together easily whether or not osteotomy is performed.

The operation

The bladder is examined under anesthesia, and pressure is placed on the bladder to make sure that it will indent into the pelvis before the initial incision is made (Fig. 19.1). The ureteric orifices should be identified, as should the prostatic utricle and ejaculatory ducts. The penis should be carefully examined for the length and depth of the urethral groove and the amount of ventral foreskin, especially if a combined closure is to be undertaken.

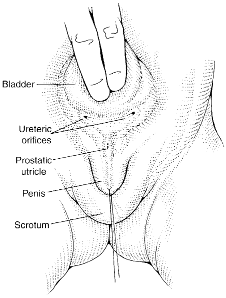


Fig. 19.1

Figure 19.2 shows the initial incision around the umbilicus and down to the urethral plate in both the male and the female. Transection of the urethral plate is now rarely carried out because of the widespread application of the Cantwell–Ransley repair and the increased incidence of posterior urethral problems after closure with para-exstrophy skin flaps. The umbilical vessels are tied off posteriorly at their junction with the umbilicus.

Introduction

The primary objectives of the modern surgical management of classic bladder exstrophy are:

1. a secure initial closure;
2. the reconstruction of a functioning and cosmetically acceptable penis in the male and external genitalia in the female;
3. urinary continence with the preservation of renal function.

These objectives can currently best be achieved with newborn primary bladder and posterior urethral closure, early epispadias repair and, finally, bladder neck reconstruction and ureteric reimplantation when the bladder reaches an appropriate volume for an outlet procedure if the child is motivated, ready to be dry and ready to participate in a postoperative voiding program. Occasionally, in very select patients, bladder closure and epispadias repair can be combined and bladder neck repair performed when an adequate bladder capacity has been reached and the child is ready to be continent.

Complete male epispadias and female epispadias, which are variants of the exstrophy condition, are similar to classic bladder exstrophy except that, in complete male epispadias and female epispadias, the bladder is closed and bladder closure is not required. In addition, the surgical objectives of the repair of epispadias include the achievement of urinary continence, the preservation of the upper urinary tracts and the reconstruction of cosmetically acceptable genitalia. The surgical management of incontinence in epispadias is virtually identical to that of a closed classic bladder exstrophy.

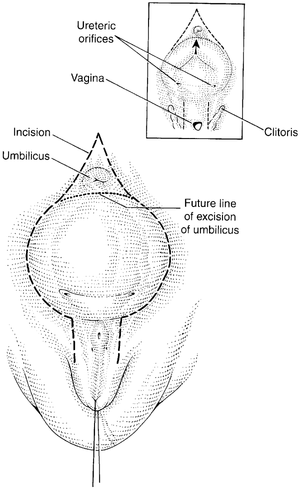


Fig. 19.2

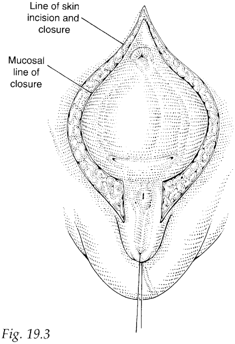


Fig. 19.3

Figure 19.3 depicts the completed skin incision around the umbilicus and down onto the urethral plate and external genitalia. The development of the retropubic space from the area of the umbilical insertion downwards facilitates the separation of the bladder from the rectus sheath and muscle. Figure 19.4 shows the incision of the remnants of the suspensory ligaments,

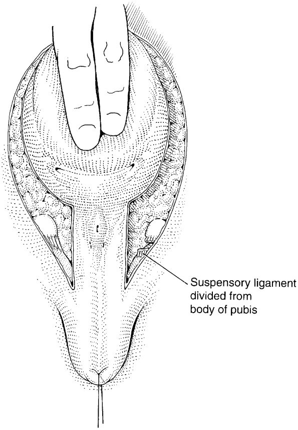


Fig. 19.4

and Fig. 19.5 depicts the development of the retropubic space between the bladder, rectus fascia and muscle. Also seen is the division of the filamentous attachments between the bladder and the underside of the rectus muscle. Figure 19.6 shows the final incision and the subperiosteal detachment of the crura from the pubic bone. The corpora are more easily visualized and the urogenital diaphragm fibers have now come into view.

Dissection then proceeds along the corporal bodies to free them from the urethral plate so that the posterior urethral plate can be sunk deeply into the pelvis at the time of abdominal wall and pelvic closure. Dissection should travel cephalad along the urethral plate up under the prostate gland in order to release any tethering and to allow placement of the posterior vesico-urethral unit deeply into the pelvis.

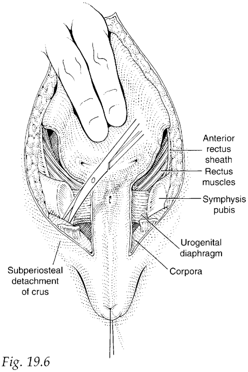


Fig. 19.6

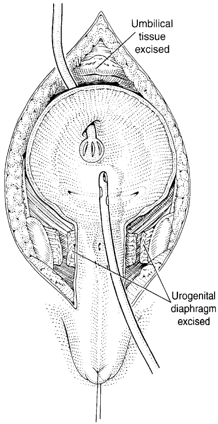


Fig. 19.7

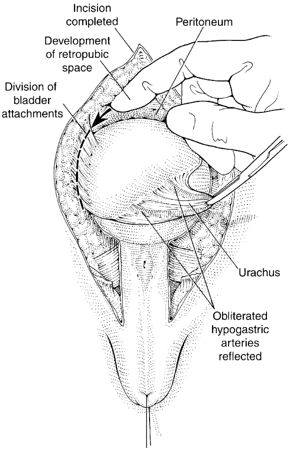


Fig. 19.5

Figure 19.7 depicts the completed transection of the urogenital diaphragm fibers from the pubic ramus and the levator in the male and the front wall of the vagina in the female in order to allow further placement of the posterior vesico-urethral unit into the pelvis. This figure also shows the completed incision of the urogenital diaphragm fibers from the pubis and from the levators. A suprapubic tube has been placed through the dome of the bladder, and the peritoneum has been swept off the dome so that the closed bladder can be placed into the pelvis behind the peritoneum.

After the bladder has been adequately freed from all the surrounding structures, the suprapubic tube is sutured to the dome of the bladder, and two ureteric stents are brought out through small stab wounds in the lateral aspect of the bladder (Fig. 19.8).

The posterior urethra is closed with 4/0 or 5/0 polyglycolic acid sutures. The bladder is then closed in two layers with running 3/0 polyglycolic or polygalactane sutures on the inside and interrupted Lembert sutures of 3/0 polyglycolic acid or polygalactane on the outside (Fig. 19.9). The posterior urethral closure is performed over a 12F catheter. This catheter is removed at the end of the urethral closure.

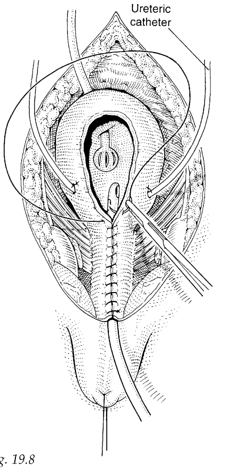


Fig. 19.8

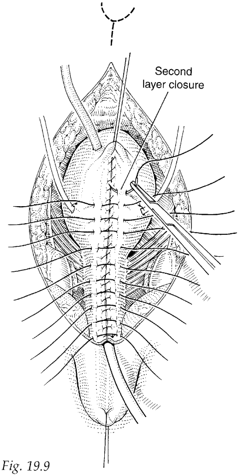


Fig. 19.9

Figure 19.10 depicts the pelvis, which has been brought together with a single horizontal mattress suture of size 2 nylon. This is tied anteriorly to the pubic bones. The rectus fascia is closed with figure-of-eight sutures of 2/0 Vicryl. The suprapubic tube and stents are brought out through the neo-umbilicus, which is reconstructed in the manner of Hanna by taking a flap and sewing this down to the anterior rectus fascia. When the suprapubic tube and stents are removed in 4 weeks, a normal umbilicus should result. Once the rectus fascia closure has been completed, it is often possible to place another suture of 2/0 nylon in the area of the junction of the rectus fascia fibers onto the superior aspect of the pubis.

In the older child with a failed exstrophy closure, the pelvic bones are brought together by medial compression of the intrafragmentary pins, which easily hold the pelvis together for suturing.

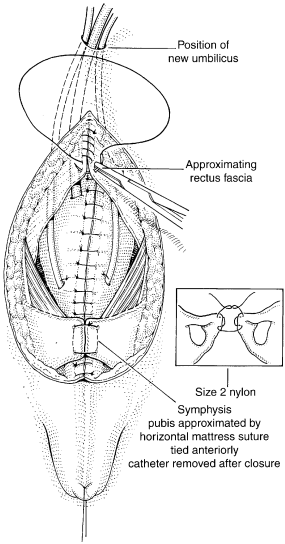


Fig. 19.10

Postoperative care of the newborn exstrophy closure

Ureteric stents are maintained for 3–4 weeks as long as they are draining well in order to keep the bladder closure dry during healing. The suprapubic tube is left in for 4 full weeks. If an osteotomy has been used, the child has been placed in modified Buck's traction for 4 weeks during the healing phase. A urine culture is sent from the suprapubic tube, and if the bladder urine is sterile, the ureteric stents are removed and an ultrasound is obtained. If the ultrasound shows only minimal-to-mild hydronephrosis, the suprapubic tube is clamped.

Residual urine volumes are checked by the nursing staff and if the volumes are low, the suprapubic tube is removed within 24 hours. Stent and suprapubic tube removal is usually not undertaken until after the removal of the intrafragmentary pins and the external fixation device. Patients are maintained on daily suppressive antibiotics, and at 2 months after closure an ultrasound of the urinary tract is performed to exclude major hydroureteronephrosis. At 4 months of age, a follow-up ultrasound is obtained, and if that is normal, another ultrasound is not taken until 12 months after the initial closure.

If the initial residual urine volumes are high when the suprapubic tube is clamped, the child is taken to the operating room for urethral calibration and cystoscopy. If at any time during the postoperative phase the child has a febrile urinary tract infection or hydronephrosis is noted on postoperative ultrasound scans, immediate cystoscopy is undertaken.

The Cantwell–Ransley second-stage urethroplasty for exstrophy

Many surgical techniques have been described for reconstruction of the penis and urethra in patients with classic bladder extrophy. The most current methods of epispadias repair in bladder exstrophy are the Cantwell–Ransley repair, the modified Cantwell–Ransley repair and the penile disassembly technique as described by Mitchell in 1996. Since the Cantwell–Ransley repair enjoys the most popularity worldwide and is the author's preference, it will be described in detail.

Regardless of the surgical technique chosen for reconstruction of the penis in bladder exstrophy, four key concerns must be addressed to ensure a functional and cosmetically pleasing penis. These concerns are correction of dorsal chordee, urethral reconstruction, penile skin closure and glandular reconstruction. Although it is possible to achieve some penile lengthening with release of chordee at the time of the initial closure, it is often necessary to perform formal penile lengthening with release of chordee at the time of urethroplasty in exstrophy patients.

Data by Silver et al (1997) clearly show that it is more an attempt at apparently lengthening the penis than a true lengthening because the anterior corporal bodies in exstrophy patients have 50% less length than those in age-matched controls. Certainly, all remnants of the suspensory ligaments and old scar tissue from the initial bladder closure must be excised. In addition, further dissection of the corpora cavernosa from the inferior pubic ramus can be achieved. It is often surprising how little is accomplished in freeing the corporal bodies from the pubis at the time of initial exstrophy closure. In the modern applications of staged reconstruction in bladder exstrophy, epispadias repair is performed under testosterone stimulation when the child is between 6 months and 1 year of age.

The operation

Skin incisions outlining the urethral strip are carried around the ventrum of the penis at the subcoronal level (Fig. 19.11a). A meatoplasty, the IPGAM, which is the reverse of the meatoplasty carried out in the MAGPI (meatal advancement and glansplasty) distal hypospadias repair, is performed. A vertical incision in the urethral groove is made almost to the glans (Fig. 19.11b and c). The incision is closed transversely, widening the urethra and positioning the meatus ventrally (Fig. 19.11d).

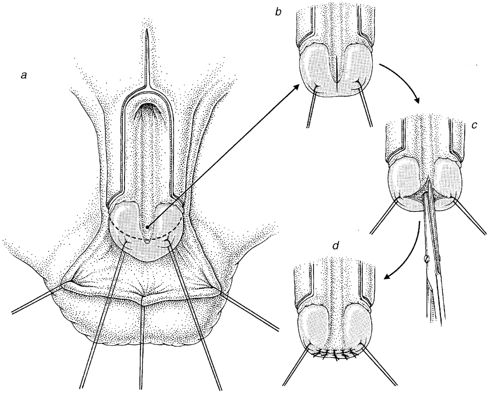


Fig. 19.11a-d

The penile skin is mobilized from the corporal bodies (Fig. 19.12). The dorsal nerves, as shown in the figure, are normally only seen in older children. The ventral view shows the extent of the corporal mobilization (Fig. 19.13).

Care must be taken not to dissect the urethral plate too widely from the subcutaneous tissue so that its blood supply is preserved. This is achieved by keeping dissection close to the corpora, while at the same time being careful not to injure the dorsal neurovascular bundle. The incisions outlining the urethral strip are carried out on to the glans (Fig. 19.14). The glans wings must be adequately mobilized to allow ventral placement of the new urethra and to achieve a conically appearing glans penis.

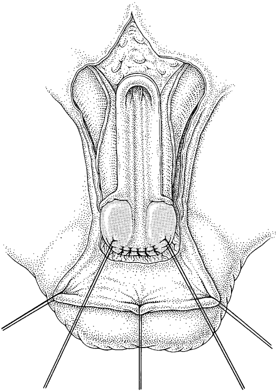


Fig. 19.12

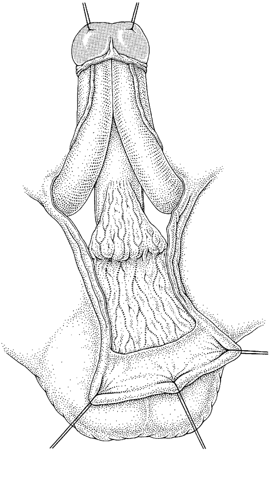
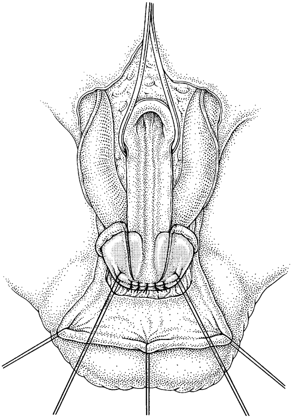
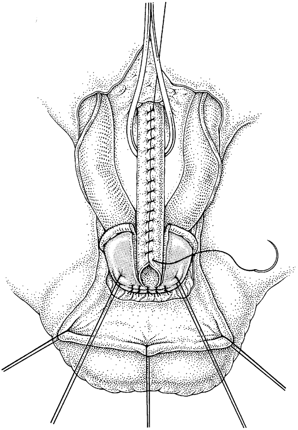


Fig. 19.13



ig. 19.14



g. 19.15

The neo-urethra is tubularized with continuous inverting Dexon or Vicryl sutures (Fig. 19.15). If the closure is watertight a fistula is less likely to develop. The common area of fistulization is where the urethra exits between the two proximal corporal bodies.

Figure 19.16 shows the Ransley technique to correct intrinsic chordee. This procedure is not often required in young infants undergoing epispadias repair, being most commonly required in older boys when dorsal chordee, which is usually caused by corporal disproportion, develops with growth. Transverse incisions are made dorsally through the tunica albuginea at the point of maximum upward curvature of each corpus (Fig. 19.16a). The incision must be wide enough to divide completely the short dorsal tunica albuginea. As this is done, the transverse incision will open into a diamond (Fig. 19.16b), effectively lengthening the dorsal portion of the tunica albuginea and correcting the intrinsic chordee. Two diamond-shaped openings in the corpora are now anastomosed with two continuous 5/0 PDS watertight sutures (Fig. 19.16c and d). This rolls the corporal bodies medially and creates a communication between the erectile tissue in each corpus (Fig. 19.16d). Note that the urethra is positioned ventral to the anastomosis. Two sets of incisions sometimes need to be made to correct the intrinsic dorsal chordee effectively.

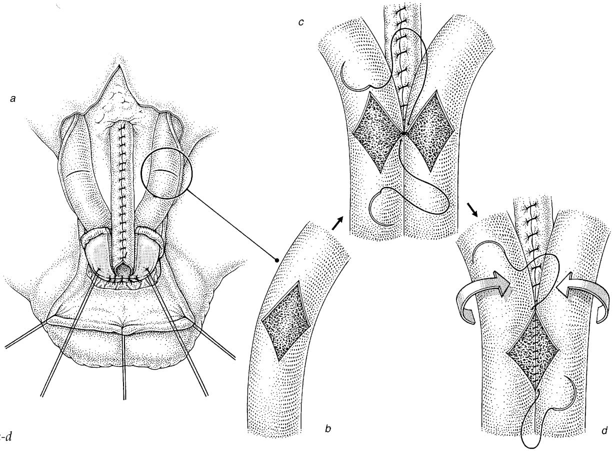


Fig. 19.16a-d

The glansplasty is completed in two layers, the second layer being a subcuticular closure of the glans to minimize scarring (Fig. 19.17). Note the ventral position of the meatus, providing the correct direction for the urethra. As the corporal bodies are rotated medially, the dorsal

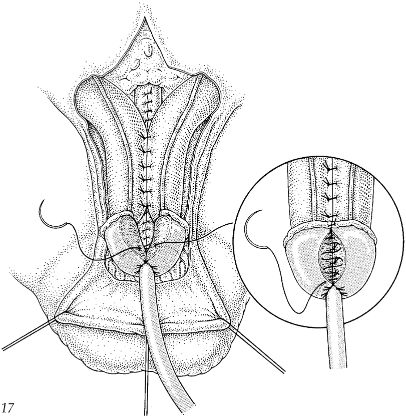


Fig. 19.17

neurovascular bundle comes to line in a normal position. Skin closure can be accomplished by using the ventral foreskin as an only or as reverse Byars' flaps (Figs 19.18–19.20).

Combined staged closure

The modern version of staged closure of bladder exstrophy has yielded consistently good cosmetic and functional results, and the utilization of osteotomy has improved the potential for successful initial closure and later continence. In an effort to decrease the cost and morbidity associated with multiple operative procedures, and hopefully effect continence, there has been a recent interest in performing single-staged reconstructions or combining procedures in appropriately selected patients. Using this technique, first described by Gearhart & Jeffs in 1991 for failed exstrophy closures, and recently reported by Grady and Mitchell for newborn patients, results have been reported in groups of boys undergoing single-stage reconstruction with closure and epispadias repair in infancy. In the author's opinion, this technique should be limited to boys of an older age, greater than 4–6 months, because recent experimental evidence indicates that newborn bladders differ from those of older infants in terms of the level of maturity of their muscle, connective tissue and neural components.

This author believes that these patients should be carefully selected, especially newborns, because of the above reasons. Otherwise, boys presenting after failed initial closure and/or primary closure who are greater than 4–6 months of age may be candidates

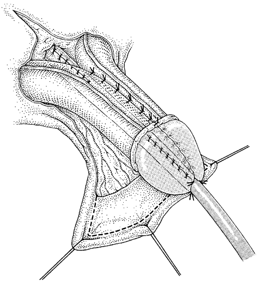


Fig. 19.18

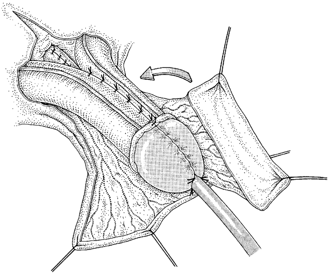


Fig. 19.19

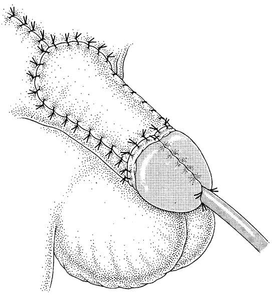


Fig. 19.20

for a combination of epispadias repair and bladder closure. Children should be carefully selected, based on phallic size, the length and depth of the urethral groove, the size of the bladder template in delayed primary closures, and perivesical and urethral scarring in children who have undergone a prior failed closure. This is not a technique for the occasional exstrophy surgeon as the complications are real and the procedure technically demanding. Figure 19.21 shows a classic bladder exstrophy with a nice deep, long urethral groove suitable for a combined bladder closure and epispadias repair.

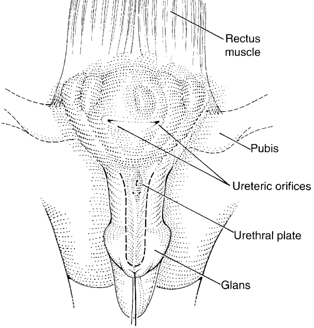


Fig. 19.21

Combined bladder closure and epispadias repair is very similar in its origin to the beginning of closure of bladder exstrophy alone. An evaluation of the bladder template is performed by inverting the bladder into the abdomen with a sterile gloved finger. This allows an evaluation of the extent of the bladder plate, which may be larger than noted on visual inspection of the abdominal wall defect. The extent of bladder polyposis and scarring is also noted. In addition, the urethral plate is examined for its length and its depth, and the presence of scarring from any prior attempt at repair.

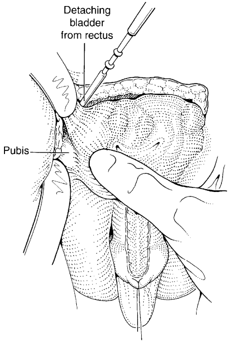


Fig. 19.22

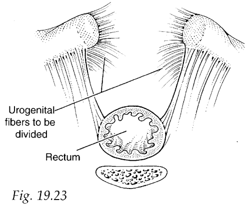


Fig. 19.23

All patients receive preoperative testosterone to enhance penile size, and testosterone cream to the urethral plate and bladder neck if prior surgery has occurred. The operative procedure begins as with a standard closure of the bladder, posterior urethra and abdominal wall, with only a few variations (Fig. 19.22) Great care must be used to divide any remnants of the urogenital diaphragm fibers that remain if there has been a previous closure (Fig. 19.23). It is surprising how frequently these fibers are intact in patients undergoing reclosure after a failed initial procedure. Once adequate dissection has been performed, the entire posterior urethra bladder complex can be moved posteriorly into the pelvis.

An incision is made around the base of the penis, and the penis is sharply de-gloved (Fig. 19.24). A plane is then established between the tunica vaginalis, tunica albuginea and urethral plate in a circumferential manner around the corporal body (Fig. 19.25)

Once the plane between the urethral plate and the corporal bodies has been established, the dissection is taken cephalad behind the prostate up to the level of the urogenital diaphragm and pelvic floor (Fig. 19.26). Once the bladder and posterior urethra have been adequately dissected, attention can be given to the dissection of the penis, corporal bodies and urethral plate. This part of the procedure progresses much as in a standard epispadias repair and exstrophy closure.

After the bladder and posterior urethra have been well dissected, the urethral plate is isolated from the corporal bodies, the suprapubic tube is brought out through the dome of the bladder, and two ureteral stents are brought out through stab wounds in the lateral wall of the bladder (Fig. 19.27). The bladder is closed in two layers with running 3/0 Vicryl sutures in the internal layer and Lembert sutures of 3/0 Vicryl in the external layer (Figs 19.28 and 19.29). The IPGAM procedure is performed as in a standard epispadias repair (Fig. 19.30).

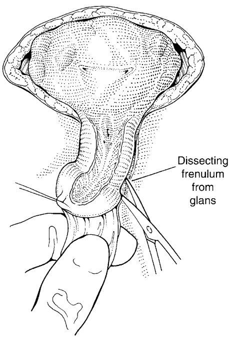


Fig. 19.24

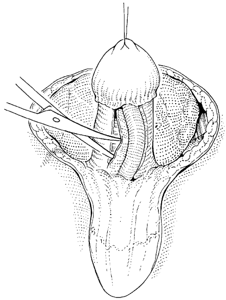


Fig. 19.25

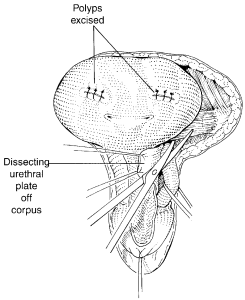


Fig. 19.26

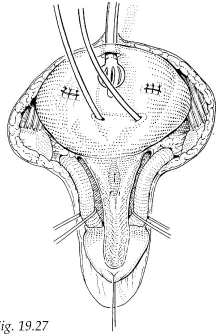


Fig. 19.27

The suprapubic tube and stents are brought out through a neo-umbilicus in the abdominal wall, just above the bladder exstrophy closure (Figs 19.31 and 19.32).

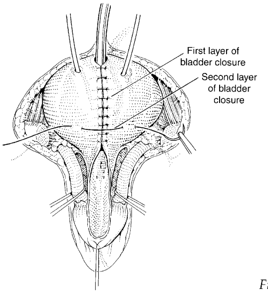


Fig. 19.28

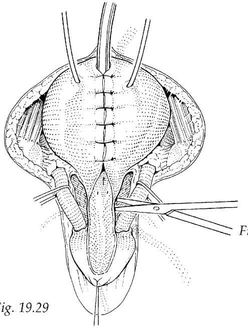


Fig. 19.29

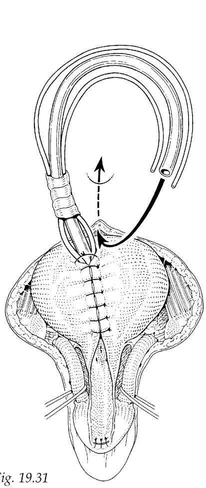


Fig. 19.31

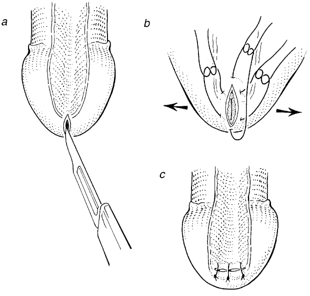


Fig. 19.30a-c

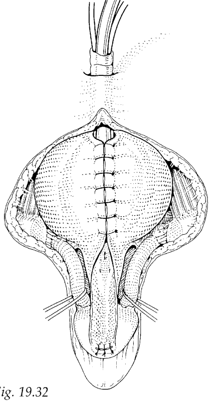


Fig. 19.32

The urethra is then closed with running 5/0 Vicryl sutures from the proximal prostatic urethra to the tip of the penis. Note that the urethral plate has been dissected from the corporal bodies and left attached only for the distal 1 cm inside the glans (Fig. 19.33). A size 2 nylon suture in a horizontal mattress fashion is placed in the pubis and used to bring the pelvis together (Figs 19.33 and 19.34). Approximation of the pubic tubercle allows the corporal bodies to significantly rotate medially so that they can be easily brought over the closed urethral plate (Fig. 19.34). The glans is then closed with two layers of subcuticular 5/0 Vicryl and 6/0 Vicryl in the epithelium of the glans (Fig. 19.35).

After glans reconstruction has been completed, the penile shaft is closed with appropriate skin coverage using reverse Byars' flaps or, in some cases, a Nesbitt turnover flap. A drain is placed next to the bladder closure, and the abdominal wall closure is completed. An 8F silastic stent is used to stent the urethra for 10 days (Fig. 19.36). The external fixating device from the osteotomy is then attached to the intrafragmentary pins and tightened.

External fixation of the pelvis is maintained for 4 weeks in children undergoing primary closure and for 6 weeks in children undergoing reclosure of the bladder. Follow-up is much the same as with a standard exstrophy closure, with the monitoring of residual urine volumes and upper tract imaging by ultrasound prior to suprapubic tube removal. While some of the author's patients have achieved long-term continence following the above procedure, most require bladder neck reconstruction when bladder capacity is deemed adequate and the child is able to participate in a postoperative voiding program.

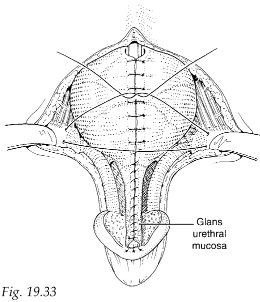


Fig. 19.33

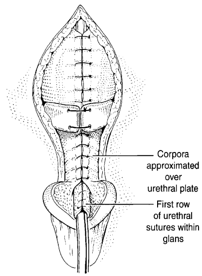


Fig. 19.34

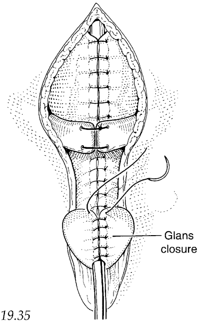


Fig. 19.35

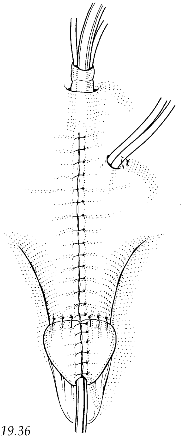


Fig. 19.36

Female epispadias

Female epispadias is a rare congenital anomaly occurring in 1 in 484 000 live female patients. In its most severe degree, the urethra is cleft along its entire length, the sphincteric mechanism is incompetent, and the patient is incontinent. The genital defect is characterized by a bifid clitoris. The mons is depressed in shape and coded by a smooth, glabrous area of skin. Beneath this area, there may be a moderate amount of subcutaneous tissue and fat, or the skin may be closely applied to the anterior/inferior surface of the symphysis pubis. The labia are usually poorly developed and terminate anteriorly at the corresponding half of the bifid clitoris, where there may be a rudiment of a labia minora (Fig. 19.37). Because there is no outlet resistance, the bladder is small and the wall is thin. Surgical objectives for the repair of female epispadias parallel those seen in the male:

1. the achievement of urinary continence;
2. preservation of the upper urinary tracts;
3. reconstruction of functionally and cosmetically acceptable external genitalia.

Operative technique

With the patient in the lithotomy position, the female epispadias opening with incontinence is apparent. The two halves of the clitoris lie widely apart and the roof of the urethra is cleft between the 9 o'clock and 3 o'clock positions.

The smooth mucosa of the urethra tends to blend cephalad with the thin glabrous skin over the mons. The urethral incision is begun at the cephalad extent of the vertical incision at the base of the mons and brought inferiorly to the full thickness of the urethral wall at the 9 o'clock and 3 o'clock positions (Fig. 19.38).

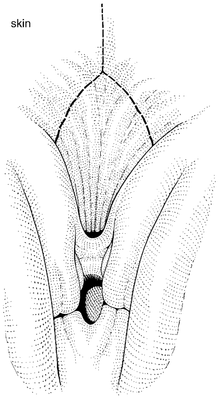


Fig. 19.38

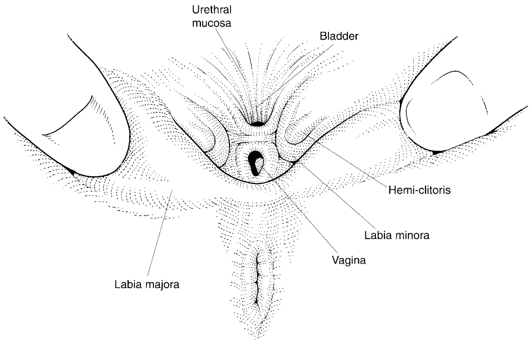


Fig. 19.37

Sutures can be placed in the urethra at this juncture and can be used to retract the cephalad extent of the urethra downwards so the roof of the urethra is excised to a level near the bladder neck (Fig. 19.39). One will often find the dissection under the symphysis pubis at this juncture (Fig. 19.40). A running closure of the urethra is performed over a 10F Foley catheter at this point (Fig. 19.41). Suturing is begun near the bladder neck and progress downwards until narrowing of the urethra is accomplished (Fig. 19.42).

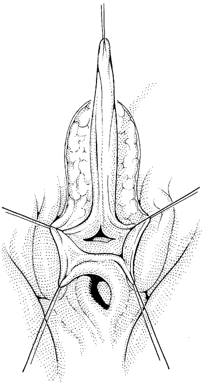


Fig. 19.39

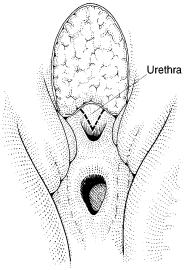


Fig. 19.40

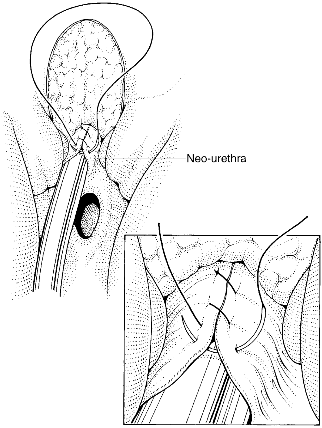


Fig. 19.41

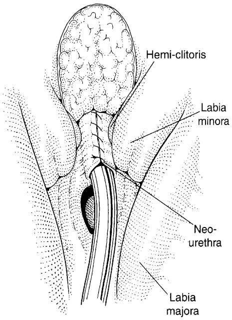


Fig. 19.42

Attention is then given to denuding the medial half of the bifid clitoris and the labia minora so that a proper genital coaptation can be obtained (Fig. 19.43). With this completed, the fat in the mons and subcutaneous tissues can be used to cover the suture line and obliterate the space in front of the pubic symphysis (Fig. 19.44).

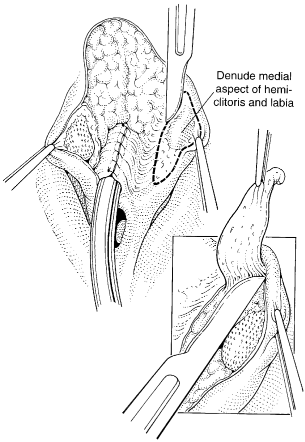


Fig. 19.43

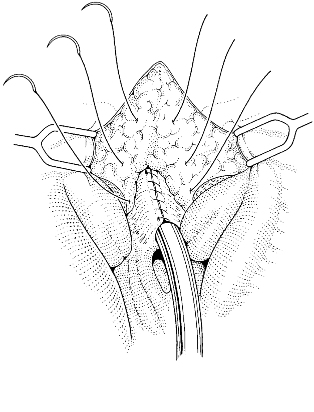


Fig. 19.44

The two halves of the clitoris and labia minora are brought together using interrupted sutures of 6/0 polyglycolic acid (Fig. 19.45). The corpora may be partly detached from the anterior ramus of the pubis to aid in the urethral closure. In addition, bringing these tissues together may contribute by adding resistance to the urethral closure. The mons closure is further aided by mobilizing the subcutaneous tissues laterally and bringing them medially to fill any depression that remains (Fig. 19.46).

The subcutaneous layer is closed with 4/0 polyglycolic acid sutures in an interrupted fashion. The skin is closed with a running subcuticular suture of 4/0 polyglycolic acid. Interrupted sutures of 6/0 polyglycolic acid are used to close the area, bringing the midline together as a clitoral hood (Figs 19.47 and 19.48).

Postoperative care

A 10F Foley catheter is left indwelling for 5 days. A pressure dressing utilizing an Elastoplast bandage is placed over the reconstructed external genitalia, left in place for 48 hours and then removed. Should the patient undergo simultaneous bladder neck reconstruction, a Foley catheter is not left in the urethra, but the patient is placed in the supine position for the abdominal part of the procedure.

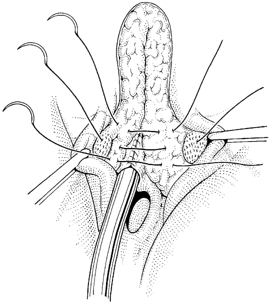


Fig. 19.45

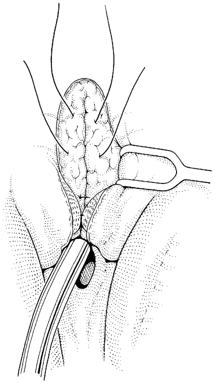


Fig. 19.46

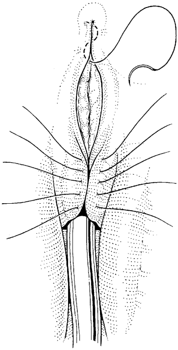


Fig. 19.47

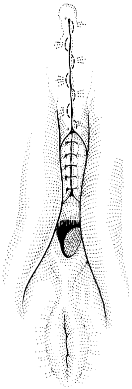


Fig. 19.48

Bladder neck reconstruction for incontinence

Bladder neck reconstruction remains a formidable challenge for the surgeon. Many techniques have been used in the past to achieve urinary continence. However, the timing of bladder neck surgery, the most reliable procedure, the need for bladder augmentation and the optimal treatment of the child with a previously failed incontinence procedure remains controversial. This section will deal with bladder neck reconstruction for the two most common causes of congenital urinary incontinence: classic bladder exstrophy and complete epispadias.

Bladder neck reconstruction in classic bladder exstrophy

Successful bladder neck reconstruction in bladder exstrophy patients begins with the initial closure. This should ideally be undertaken within 72 hours of birth, but later closure, when well performed, can lead to continence. In a recent review of our patients, a successful initial closure of the bladder was the most important factor leading to eventual continence. Patients who developed dehiscence or prolapse and required reclosure did not achieve continence as often as those with an initial primary closure.

Bladder neck reconstruction in complete epispadias

The attainment of satisfactory urinary continence in epispadias remains a surgical challenge. Not only is the bladder wall small and thin, but the trigonal urethral canal is also severely defective. The trigone is poorly developed, and the vesical neck is open, with patulous, laterally placed ureteral orifices. A vast majority of these children also have associated vesico-ureteric reflux, as in the exstrophy group. The rarity of this entity compared with classic bladder exstrophy makes a large series of these patients difficult to assemble.

Timing of bladder neck reconstruction

In both the exstrophy and the epispadias group, the author tends to perform cystoscopy and gravity cystogram at the 1 year anniversary of the initial bladder closure or, in the epispadias group, at the time of epispadias repair. Other means of estimating bladder capacity in the crying child on an X-ray table may be impossible. The cystogram will measure bladder capacity and also determine whether reflux is present. It was previously felt that if the bladder capacity was 60 cm^3 or higher and the child was ready to participate in a postoperative voiding program, bladder neck reconstruction could be planned. New data from Chan et al (2001) have, however, found that, in a select group of exstrophy patients who, underwent bladder closure, epispadias repair and bladder neck reconstruction at our institution by one or two surgeons, a median bladder capacity of 85 cm^3 was found to be more common in the group who were completely dry after bladder neck reconstruction. Most of these children were 4–5 years of age and were ready emotionally, maturationally and intellectually to participate in a postoperative voiding program.

Types of reconstruction

Surgical procedures designed to establish continence formerly gave rather discouraging results in both males and females. In the past few years, however, reports have shown that the modified Young–Dees–Leadbetter procedure offers the best probability of continence in these patients (Chan et al, 2001). In addition, the addition of a Marshall–Marchett–Kranz (Marshall et al 1949) bladder neck suspension certainly has improved the overall continence rate. Intraoperative pressure profilometry has confirmed the latter maneuver and increases both urethral closure pressure and continence length (Gearhart et al 1986).

Several differences exist between the classic Young–Dees–Leadbetter procedure (Leadbetter 1964) and the modified Young–Dees–Leadbetter procedure. The muscle flaps used in the modified Young–Dees–Leadbetter procedure not only are smaller, but are, more importantly, not incised transversely at their cephalad extent as described in the original Young–Dees–Leadbetter procedure. This author feels that if these flaps are incised medially at the cephalad border where they join the floor of the bladder, there is a significant risk of denervation and ischemia, which will harm the bladder neck repair. The basic premise of a modified Young–Dees–Leadbetter procedure is to create a mucosa-lined tube inside a muscular funnel that narrows from its junction with the floor of the bladder as it extends caudally.

Surgical technique

A U-shaped incision is made very low on the bladder neck and is extended into a midline cystotomy (Fig. 19.49). With a low U incision, the bladder can retract in a cephalad manner, with easy access to the bladder neck reconstruction area (Fig. 19.50). Ureteric reimplantation is performed in a transtrigonal manner, not only to correct reflux, but also to move the ureters away from the bladder neck so that sufficient trigonal tissue is available for bladder neck reconstruction (Fig. 19.51). Alternatively, the ureters can be directed more cephalad, thus allowing even greater access to the trigonal area (Fig. 19.52).

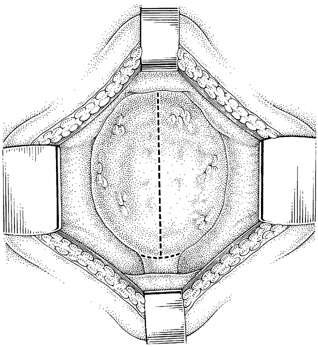


Fig. 19.49

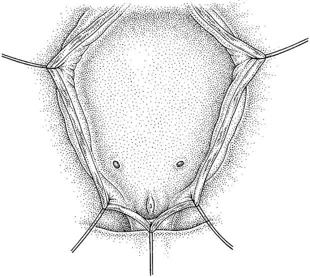


Fig. 19.50

The continence procedure is begun by outlining a strip of mucosa approximately 15 mm in width by 30 mm in length that extends from the mid-trigone to the prostatic or posterior urethra (Fig. 19.53). The bladder muscle lateral to the mucosal strip is denuded of mucosa by sharp dissection. Epinephrine-soaked sponges placed on this area are helpful in controlling the bleeding.

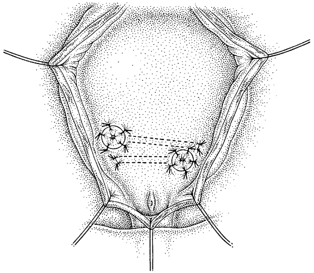


Fig. 19.51

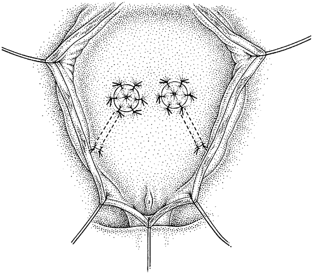


Fig. 19.52

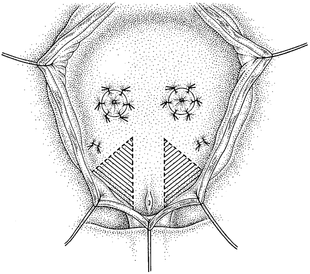


Fig. 19.53

Multiple small incisions into the bladder muscle in the area of the denuded lateral triangles provide a lengthening and flattening of the bladder neck area and allow the bladder to retract into the abdomen to a more cephalad position (Figs 19.54 and 19.55). This means that less bladder capacity can be taken during the bladder neck reconstruction.

At this juncture, the actual rolling of the bladder neck into a tube is ready to begin. The first suture is placed in the area of the new bladder neck, utilizing 3/0 Vicryl or Dexon (Fig. 19.56).

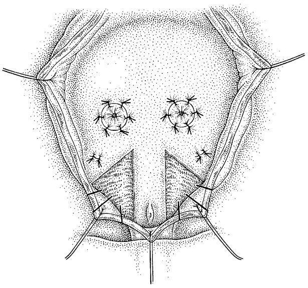


Fig. 19.54

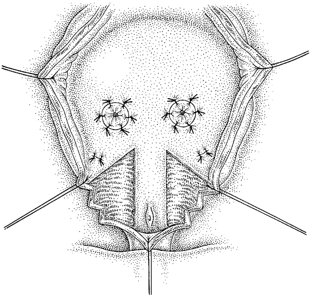


Fig. 19.55

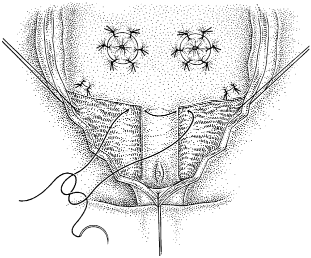


Fig. 19.56

This stitch goes through the muscle into the mucosa, across the midline, into the mucosa and out again through the muscle, being tied with a knot facing the inside the bladder. With cephalad retraction on the stitch, the more distal areas of the mucosa and the bladder neck are easily brought up into the field. At this point, multiple interrupted sutures of 4/0 Vicryl or Dexon that bring some muscle off the mucosa to the midline are inserted (Figs 19.57 and 19.58) These sutures are placed quite close together.

While the innermost aspect of the bladder neck reconstruction is being performed, an 8F plastic feeding tube is left in place through the neo-urethra. After the neo-urethral tube has been constructed, the adjacent denuded muscle flaps are overlapped and sutured firmly in place in a double-breasted muscle closure of the bladder neck (Fig. 19.58). These sutures are of the horizontal mattress type. While the second denuded muscle flap is being brought across the midline, the sutures are intentionally left long (Fig. 19.59).

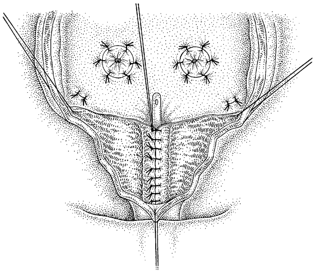


Fig. 19.57

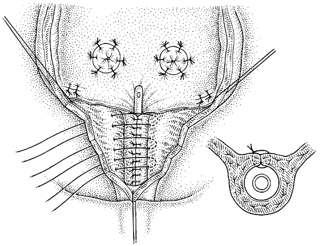


Fig. 19.58

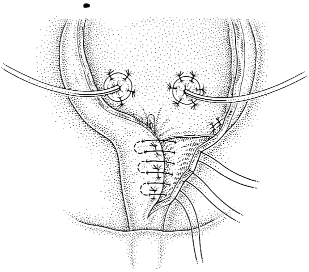


Fig. 19.59

The last suture at the new bladder neck and the suture more distal to this are going to be used for suspending the bladder neck. It is ideal to leave these sutures long enough for later bladder neck suspension. Most of the bladder neck reconstruction has now been completed, except for the suspension of the newly constructed bladder neck (Fig. 19.60).

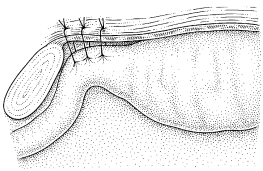


Fig. 19.61

After being tubularized, the bladder neck is suspended in a Marshall–Marchett–Kranz manner through the rectus fascia to increase both the continence length and the urethral closing pressure (Fig. 19.61). Pressure profilometry routinely reveals a closure pressure of greater than 100 ml water and a 2.5 cm continence length (Gearhart et al, 1986). No indwelling stents are routinely left through the bladder neck reconstruction area. If it is a re-operative reconstruction, however, a small 6F stent is employed. Ureteric stents are left in place for 10–21 days, and a suprapubic tube remains in place for a full 3 weeks.

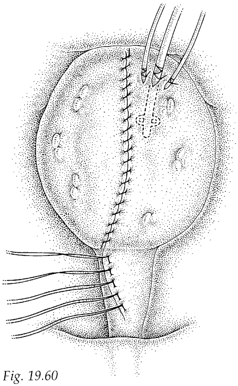


Fig. 19.60

A key factor to remember when performing this surgery is that a very radical dissection of the bladder, bladder neck and posterior urethra is required, not only from the pelvis, but also from the posterior aspect of the pubic bar to provide enough mobility for bladder neck reconstruction and later bladder neck suspension. Should visualization of the posterior urethra become a problem, the inner symphyseal bar can be cut, thus providing a widened field of exposure.

At the end of the 3 week drainage period, the suprapubic tube is clamped, and a residual voiding trial is attempted. If residual urine volumes are high during the voiding trial, the child is taken to the operating room and a gentle cystoscopy to 'find in way' is performed. An 8F catheter is then passed into the bladder through the urethra and left in place for 4–5 days. This is then removed, the suprapubic tube is clamped again, and a voiding trial is undertaken. Typically, this cystoscopy, followed by a short period of urethral drainage, will dilate the neo-urethra and bladder neck just enough to allow voiding. The ureteric stents are removed, but prior to removal of the suprapubic tube, ultrasound is obtained to document adequate drainage of the upper tracts.

The initial bladder capacity is usually quite small as a result of the operative procedure. However, if the child has an initial dry interval of 15–20 minutes, the long-term chance of success is quite good. Severe urinary frequency is often a problem after bladder neck reconstruction. Urine cultures are obtained prior to suprapubic tube removal to ensure the sterility of the urine, and suppressive chemotherapy is continued for 4 months postoperatively. If cultures are negative and urinary frequency is bothersome, low-dose oxybutinin or imipramine 3–4 times daily will often control the symptoms. After 4–6 weeks this dosage can usually be lessened and the drug later discontinued. An ultrasound is obtained 4–6 months postoperatively. The urine must be kept sterile, and the patient must be monitored by follow-up ultrasound. A cystogram in a well child is deferred for a year or more.

Gradually, over a period of 3–18 months, the dry interval lengthens. The voiding volume increases, the awareness of filling improves, and detrusor function may be voluntary with complete control. Nocturia and enuresis frequently persist after day time control has been achieved, but this will improve with increased capacity and maturity in sleep pattern.

Suppressive antibiotic therapy is continued for 6–12 months or beyond, if necessary, while satisfactory drainage, storage and bladder function are being established.

Parent and patient education are continually required, as are personal supervision by and encouragement from the surgeon. The timing of achievement of bladder control in the normal child is variable and subject to many psychological and physiological environmental influences. The child born with congenital incontinence must contend with the anatomic deficiency, the surgical repair, physiological differences and marked psychological and environmental influences. With cooperation and time, however, urinary continence is an achievable goal in a majority of patients born with congenital urinary incontinence.

Failed bladder neck reconstruction

Although the modified Young–Dees–Leadbetter procedure is very reliable when well performed in an appropriate patient, children are occasionally seen in whom an attempt has been made and has failed. Urinary diversion at this juncture is not recommended. The author believes that all such patients can be made dry if the surgeon persists and is not easily discouraged. In order to make the child dry, one must have a bladder of adequate volume, the pressure in the bladder must not be too high with uninhibited contractions, and there must be enough resistance in the urethra and bladder neck to exceed the pressure in the bladder during normal activity. In order to void, the child must be able to generate enough pressure in the bladder by detrusor activity or straining to exceed the outlet resistance. The child must also be able to empty the bladder by intermittent catheterization if needed postoperatively. These goals can usually be met in most patients with the reconstructive techniques that are available today.

When presented with a child who has had a failed bladder neck reconstruction, a complete work-up is in order, including urodynamic studies, cystography, cystoscopy and upper tract evaluation. If there is low outlet resistance, this must be increased. If the bladder volume and compliance are decreased, the bladder capacity must be augmented. Whenever a patient presents with this type of incontinence problem, we make it very clear to the family that it may be necessary to catheterize intermittently, either temporarily or, in some cases, permanently. This is especially true if there is a small bladder capacity, which must be augmented at the time of re-operative bladder neck reconstruction. If, however, the bladder capacity is adequate, a repeat modified Young–Dees–Leadbetter procedure is the operation of choice.

With a small-capacity bladder, bladder augmentation along with reimplantation of the ureters either into a bladder patch or a selected bowel segment may be in order. As with other reconstructive procedures, the bowel segment chosen should be de-tubularized prior to augmenting the bladder. Our results utilizing the artificial urinary sphincter in patients who have failed bladder neck reconstruction have not been rewarding. In the child who has had multiple failed bladder neck procedures, the treatment of choice is transection of the bladder neck above the ejaculatory ducts, bladder augmentation and the establishment of a continent urinary stoma.

Conclusion

Surgical repair of the exstrophy–epispadias complex requires appropriate patient selection and surgical experience as well as long-term, careful follow-up for success. The purpose of this chapter is to provide the pediatric urologist/surgeon with the basic surgical principles needed for reconstruction in this interesting group of anomalies.

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Chapter 20

Bladder neck reconstruction for urinary incontinence

João L. Pippi Salle

Introduction

The treatment of an incompetent bladder neck remains a difficult challenge with respect to the persistence of incontinence and difficulties with postoperative catheterization. Several techniques have been described in attempting to overcome these problems. We previously described a procedure for the treatment of neurogenic urinary incontinence using an anterior bladder wall flap to lengthen the urethra (Pippi Salle et al 1994, Rink et al 1994). This results in an intravesical neo-urethra with a flap-valve mechanism. Because the continuity of the posterior mucosal wall is maintained, the lengthened urethra has a smooth wall, which allows easy catheterization. Further modifications to the original technique have allowed its use in other conditions, such as bladder exstrophy and epispadias, if augmentation and intermittent catheterization are anticipated (Pippi Salle et al 1997).

Preoperative assessment includes a complete urodynamic evaluation, paying special attention to the determination of bladder compliance and urethral resistance. In cases of diminished urethral resistance, it may be useful to occlude the bladder neck for a more accurate evaluation of vesical compliance and capacity. These are important factors to take into consideration when deciding to perform a simultaneous bladder augmentation.

Patient selection is of crucial importance in order to delineate the strategy of the surgery. A successful outcome is related to the continuing commitment of the patient or, in younger patients, of their parents. Preoperative assessment includes a determination of patient motivation and intelligence, as well as the degree of manual dexterity to perform clean intermittent catheterization. Patients severely handicapped and with difficulty in accessing the perineum for clean intermittent catheterization may require the fashioning of a catheterizable stoma in the abdominal wall to enable intermittent bladder emptying. In addition, patients with normal bladder and urethra innervation may be reluctant to perform catheterization because of pain. This group of patients should also receive an abdominal catheterizable (Mitrofanoff) stoma.

Surgical technique

As most patients will require bladder augmentation, the bowel should be cleaned and ready for use as necessary. Antibiotic prophylaxis is indicated. The patient is placed in a hyperextended position, and a midline or a Pfannenstiel incision is made (depending on the need for enterocystoplasty). A wide-based anterior bladder wall flap measuring at least 5 cm in length is outlined, starting at the urethra. The base of the flap should be 2.5 cm wide, narrowing progressively to the top, which should measure approximately 1.5 cm wide. Stay-stitches in corners of the flap facilitate exposure and orientation, the view being from inside the bladder (Fig. 20.1).

In the absence of vesico-ureteric reflux (the most common scenario), fashioning of the flap should allow the creation of free mucosa beyond the distal aspect of the flap, which will later be used to cover the neo-urethra. This can be performed by cutting only the muscular layer of the distal flap and carrying the dissection upwards in the submucosal plane, similar to the approach used in extravesical ureteric reimplantation (Fig. 20.2). Laterally, the flap is cut full thickness, and its

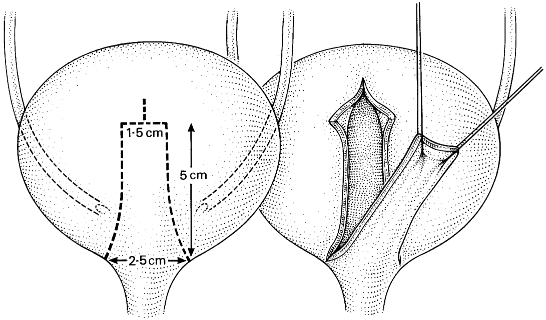

Fig. 20.1

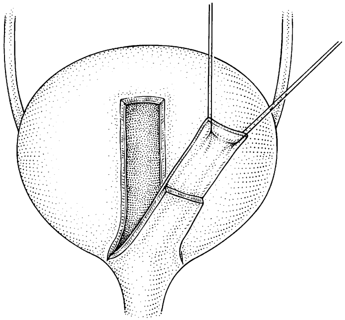

Fig. 20.2

lateral mucosal edges are excised to avoid an overlapping of the suture lines later in the procedure. This excision allows the construction of a rectangular mucosal area attached to the flap, which has a larger part of the denuded detrusor in its base, the most dangerous place for fistula formation. If reimplantation is necessary, the flap is outlined without the need to fashion distal free mucosa. The distal part of the flap can be incised to full thickness (Fig. 20.3).

Two parallel incisions, approximately 0.7 cm apart, are made in the posterior bladder wall, extending all the way from the urethra to the bladder neck

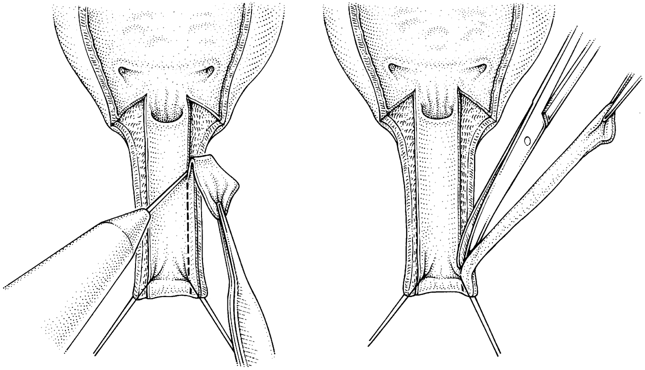

Fig. 20.3

(Fig. 20.4). If ureteric reimplantation is necessary, the ureters are at this point disconnected and reimplanted transversely and superiorly to the posterior mucosal incisions.

The posterior mucosa is elevated laterally on each side of the intact mucosal strip (Fig. 20.5). The mucosal edges of the flap are anastomosed to the posterior bladder mucosal strip with absorbable sutures in an onlay fashion. A second muscular–muscular layer is then used. It is easiest to alternate running the sutures on each side, especially in the proximal portion of the anastomosis, in order to improve exposure (Fig. 20.6).

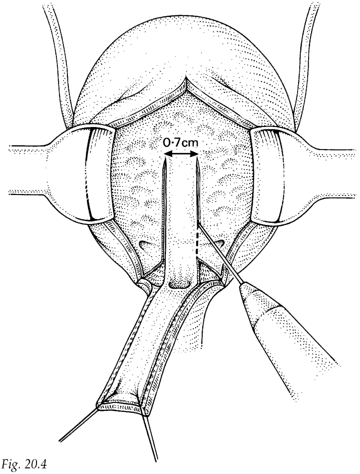

Fig. 20.4

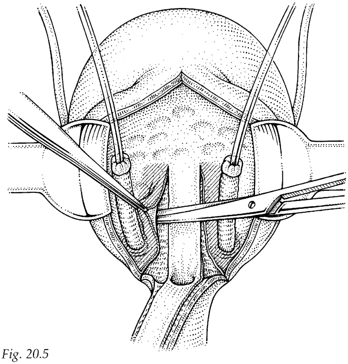

Fig. 20.5

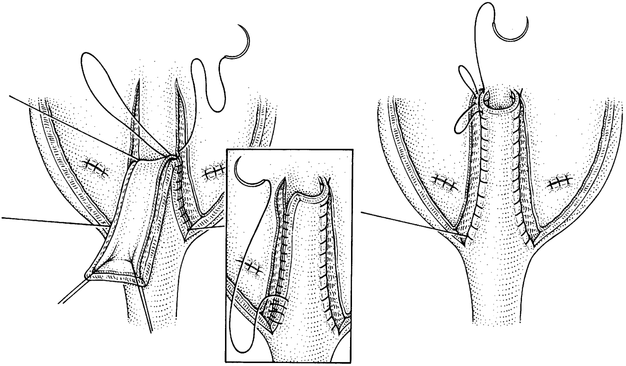

Fig. 20.6

The previously elevated lateral mucosal edges are approximated over the neo-urethra with absorbable sutures (Fig. 20.7). If reimplantation has not been carried out, the previously fashioned flap with

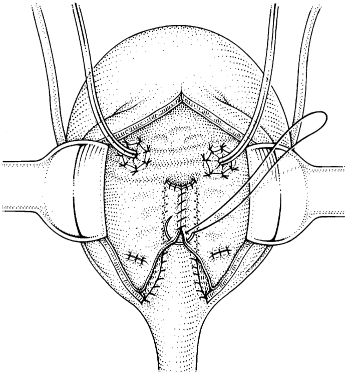

Fig. 20.7

redundant mucosa is flipped over to cover the neo-urethra and improve bladder closure (Fig. 20.8). The lateral edges of the anterior bladder wall are then approximated over the neo-urethra, creating a flap-valve mechanism (Figs 20.9 and 20.10). We try to ensure that no tension is created over the neo-urethra with this approximation. It may be necessary to leave part of the neo-urethra outside the bladder in order to do this. This is preferable because, if tension on the urethra is created by approximation, this will result in the ischemia of the whole flap, the necrosis of the intravesical urethra and the loss of the flap-valve mechanism. If part of the neo-urethra is left outside, it can be covered with a small portion of omentum.

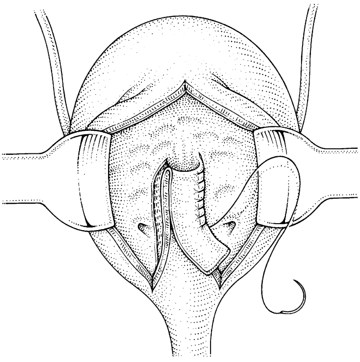

Fig. 20.8

When augmentation is necessary, this can be carried out in two ways: the bladder anterior wall edges can be

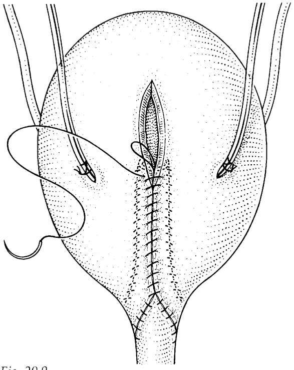

Fig. 20.9

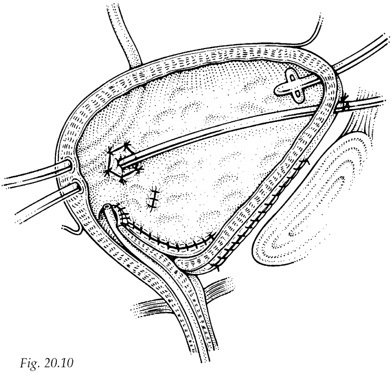

Fig. 20.10

approximated over the neo-urethra as previously described, or the augmented bowel itself can be sutured all the way down, covering the total neo-urethra and thus creating a valve mechanism with the intestine itself.

Modifications of this procedure have been used for the surgical treatment of incontinence after the primary closure of exstrophy or incontinent epispadias. Because a midline scar is present, a laterally based flap is created and sutured to the posterior wall as described above (Fig. 20.11). This approach has also been successfully used in cases of failed Young–Dees–Leadbetter bladder neck repair. Other variations of the technique have been used when a very wide and large urethra (prolapsing

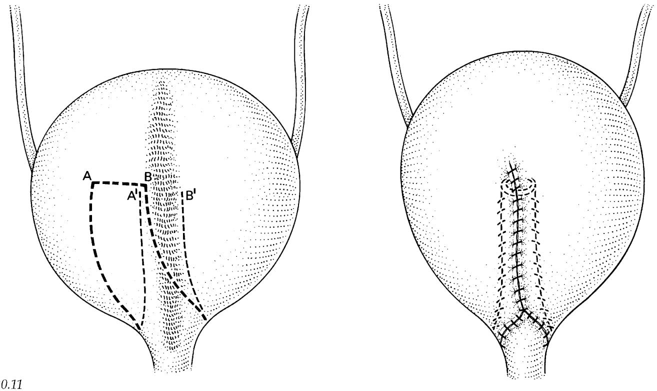

Fig. 20.11

exstrophy) has been present. In such cases, a Y-shaped flap is constructed, creating the whole urethra in a single onlay-tube procedure. Both arms of the flap will comprise the intravesical urethra. The base of the Y flap will become the extravesical urethra (Fig. 20.12).

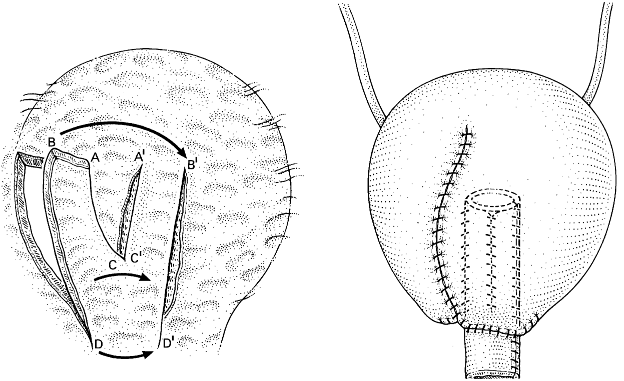

Fig. 20.12

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Chapter 21

Wilms' tumor

Richard D. Spicer and J. David Frank

Introduction

The management of childhood cancer changed dramatically with the advent of radiotherapy and later chemotherapy, and nowhere is this better demonstrated than in the treatment of patients with Wilms' tumor (nephroblastoma). Surgical treatment alone led to a survival of only 20%, but the addition of other treatment modalities has led to an overall survival of greater than 85%. Nephrectomy thus plays only a part in the overall management of these patients, the precise treatment strategy varying between different national collaborative groups, although common features include the use of vincristine for low-stage tumors and actinomycin D and doxorubicin for those of higher stages. A small subgroup is now being defined that may require no adjunctive therapy. Radiotherapy may be used for lymph node involvement and post-surgical residual disease. All patients should be referred to a specialist center (Pritchard et al 1995).

There are three broad approaches to treatment, all of which give very similar long-term results:

1. The National Wilms Tumor Study (NWTS), carried out in the United States, advocates initial nephrectomy and postoperative adjunctive therapy (D'Angio et al 1976).
2. The International Society of Paediatric Oncology (SIOP) uses preoperative chemotherapy in all cases with delayed nephrectomy. Diagnosis is by imaging alone with no initial biopsy. This approach is used in France and many other European countries (Tournade et al 1993).
3. Preoperative chemotherapy is given in all cases but with histological confirmation of the diagnosis by needle biopsy before treatment. This approach has been used in Sweden and Canada (McLorie et al 1991).

The United Kingdom national group (UKCCSG) is currently conducting a randomized trial comparing strategies 1 and 3.

Before operating on a child with a Wilms' tumor, the following questions should have been answered:

1. Is the mass a Wilms' tumor? The diagnostic accuracy with imaging alone is approximately 95%.
2. Is the disease unilateral or bilateral? Imaging should detect 93% of contralateral tumors.
3. Is there any evidence of tumor thrombus in the renal vein, inferior vena cava or right atrium? In skilled hands, ultrasound is extremely accurate for detecting caval extension.
4. Is there any evidence of pulmonary metastases on chest X-ray? Computerized tomography (CT) scanning is indicated in cases of diagnostic doubt.
5. Is the tumor operable? This is a subjective clinical judgment, but imaging will add to the clinical assessment.

Clinical presentation

The majority of children present with an otherwise asymptomatic abdominal mass, but hematuria is a feature in 15%, fever in 20% and abdominal pain in 10%. A minority have features of an associated syndrome such as aniridia, hemihypertrophy or genital abnormalities. Hypertension occurs in 10%, so the blood pressure should always be measured.

Investigation

Chest and abdominal X-rays and abdominal ultrasound should be performed in all cases, as should urine microscopy; serum electrolytes, full blood count, serum alphafetoprotein and urinary catecholamine levels should also be measured.

A **plain abdominal X-ray** should be taken, principally looking for calcification.

A **posteroanterior chest X-ray** film is initially taken, with appropriate lateral films to elucidate suspicious lesions. Small pulmonary metastases visible on CT scanning but not plain X-ray do not constitute a reason for upstaging to stage IV in the UKCCSG protocol.

Ultrasound will usually demonstrate a solid lesion, although cystic areas may be seen. The liver should be scanned for metastases and tumor extension, indicating inoperability. The renal vein and inferior vena cava should be scanned with great care: the renal vein is often not visible, especially with large tumors, but every effort should be made to determine whether or not there is caval extension. In expert hands, and particularly if Doppler ultrasound is used, the diagnostic accuracy approaches 100%. If caval thrombus is demonstrated, its upper extent should be accurately documented; if it is not clearly visible, echocardiography should be requested. Cavography has been superseded by modern ultrasound.

CT scanning is routinely carried out in some centers, but there are only three situations in which CT provides clinically essential information:

1. a suspicious but not diagnostic lesion on chest X-ray;
2. the suspicion of a lesion in the contralateral kidney;
3. in helping in the assessment of operability (e.g. extension into the diaphragm or liver).

Contrast CT examinations will confirm a functioning contralateral kidney.

Arteriography is not indicated except in the preoperative planning of a pre-treated patient with bilateral tumors and in the management of a tumor in a horseshoe kidney.

MRI scanning may give superior anatomical information to CT in some circumstances, particularly in the case of intracaval extension of tumor.

Using these investigations, the tumor can then be appropriately staged (Table 21.1).

Differential diagnosis

The diagnostic accuracy with modern imaging in a specialist center is 95%. The malignant lesion with which it is most commonly confused is the neuroblastoma. The chief benign lesion to be distinguished is xanthogranulomatous pyelonephritis, which may be clinically suspected in children with anemia, fever, leukocytosis and pyuria; the diagnosis of xanthogranulomatous pyelonephritis can be made accurately and reliably on ultrasound by an experienced radiologist. Contralateral renal hypertrophy is usual with this condition. Renal cell carcinoma is rare but should be considered in patients over 5 years of age. Mesoblastic nephroma is three times more common than Wilms' tumor in the neonate.

Table 21.1 Staging of Wilms' tumor (National Wilms Tumor Study-3)

Stage I	Tumor limited to kidney, completely excised. Capsular surface infact; no tumor rupture; no residual tumor apparent beyond margins of resection
Stage II	Tumor extends beyond kidney but is completely excised. Regional extension of tumor; vessel infiltration; *tumor biopsies or local spillage of tumor confined to flank. No residual tumor apparent at or beyond margins of excision
Stage III	Residual non-hematogenous tumor confined to abdomen. * Lymph node involvement of hilus, periaortic chains or beyond; diffuse peritoneal contamination by tumor spillage or peritoneal implants of tumor; tumor extends beyond surgical margins either microscopically or macroscopically; tumor not completely removable because of local infiltration into vital structures
Stage IV	Tumor deposits beyond stage III, i.e. lung, liver, bone and brain
Stage V	Bilateral renal involvement at diagnosis

**Differences between UK and International Society of Paediatric Oncology (SIOP) staging:*

1. Biopsy does not upstage from stage I in UK.
   Biopsy upstages to stage III in SIOP.
2. Regional node involvement does not upstage from stage II in SIOP.

Surgical management

The surgical management of a patient with a Wilms' tumor will be influenced by the size of the tumor, the presence of metastases, the presence of thrombus within the inferior vena cava or right atrium and any evidence of bilateral disease. Many national protocols now use preoperative chemotherapy in all patients. The traditional teaching in the United States has been that a tumor should if possible be primarily excised. Wilms' tumors are, however, extremely sensitive to chemotherapy, and there are four absolute indications for pre-treatment of a tumor prior to surgery, as described below.

The tumor is extremely large and it is thought that excision will be technically difficult It is then wiser to carry out a needle biopsy to confirm the diagnosis and pre-treat such a patient prior to nephrectomy. These large tumors shrink rapidly, allowing safe and relatively easy surgery some weeks later. Even if there is no great decrease in size, the vascularity of the tumor and its tendency to intraoperative rupture is always reduced. The days of heroic surgery that puts the patient's life at risk are over.

The presence of extensive inferior vena caval or right atrial tumor thrombus If tumor thrombus is found to extend into the inferior vena cava but is mobile and below the diaphragm, it is reasonable to proceed to nephrectomy with extraction of the tumor thrombus. If, however, there is extension of the thrombus above the diaphragm or into the right atrium, the patient may either be pre-treated or the tumor removed with the thrombus using right heart bypass surgery (Daum et al 1994). There have been many reports in the literature of successful surgery using this latter technique. There have, however, also been reports of the effective pre-treatment of such patients with chemotherapy, and the authors favor this more conservative approach, which allows safe resection using the same techniques at a later date. What must be avoided at all costs is an attempt to remove intravascular thrombus without adequate proximal and distal vascular control. Tumor embolization under such conditions can lead to an operative death.

Bilateral tumors (or tumor in a solitary kidney) At least 4% of patients with Wilms' tumors will have bilateral disease, and this may not be suspected preoperatively. Thus, when carrying out a laparotomy for Wilms' tumor, the apparently normal kidney should be examined first as the presence of bilateral disease will influence further surgical management.

In the past, the majority of patients with bilateral disease have undergone nephrectomy of the kidney with major tumor involvement and excision biopsy or partial nephrectomy on the contralateral side. Total excision of the tumor may not be possible, and in some children with bilateral disease, residual tumor will be left behind in spite of repeated surgical procedures. The 2 year survival rate is, however, extremely good, at 87% overall or 83% of those patients left with residual disease (Kay 1985).

These tumors occur in a young age group, and nephroblastomatosis is often present in both kidneys. The principles of modern treatment are to preserve as much functioning renal parenchyma as possible by optimal preoperative chemotherapy, avoiding nephrectomy until all conservative measures fail. After initial biopsy, chemotherapy is continued for as long as there is an objective response, as noted by ultrasound and/or CT scanning. If there is no response or a less than 50% reduction of tumor size, a second-look laparotomy is performed at the end of 3 months. This laparotomy has the intention of maximum conservation of renal parenchyma with either partial nephrectomy, enucleation or excision biopsy of the tumor.

Before the tumor is removed, chemotherapy is continued according to protocol, and no radiotherapy is given. If nephrectomy seems inevitable, definitive surgery is deferred, a repeat biopsy is performed and second-line chemotherapy is given. At a suitable time, depending on the continued shrinkage of the tumor, a third laparotomy is performed and definitive surgery carried out. Evidence is emerging that the enucleation of pretreated tumors may be as good as partial nephrectomy, with the advantage of better renal conservation.

Renal transplantation has been found to be an effective treatment for patients who have needed bilateral nephrectomy, but this should be deferred until 2 years after the remission of the disease with the patient on interim dialysis.

Biopsy This should be done using a cutting needle (e.g. Temno) and by a posterior loin approach (not transperitoneally), preferably with ultrasound guidance.

Stage IV disease Nephrectomy should be deferred until the pulmonary metastases have regressed with chemotherapy.

Surgical technique

The aims of surgery are:

1. an initial examination of the apparently normal kidney to exclude

bilateral disease (may be omitted if CT scan is done preoperatively);
2. early ligation of the renal artery and vein prior to mobilization of the kidney;
3. nephrectomy without capsular breech (tumor rupture).

Procedure

The surgeon usually stands on the same side of the table as the tumor is to be found, but some prefer to stand on the contralateral side for all or part of the procedure. The patient is placed in a supine position with a roll or sandbag under the lumbar region, and a generous transverse upper abdominal muscle cutting incision is made (Fig. 21.1). The peritoneum is opened. The liver is inspected for metastases, and the colon over the apparently normal kidney is reflected medially. The renal fascia is then incised, the anterior and posterior surfaces of the kidney being carefully inspected (Fig. 21.2). This step may be omitted if a preoperative CT scan has

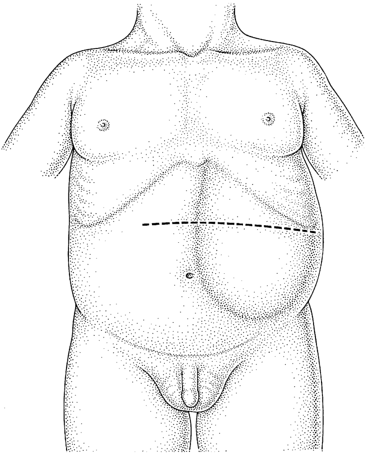

Fig. 21.1

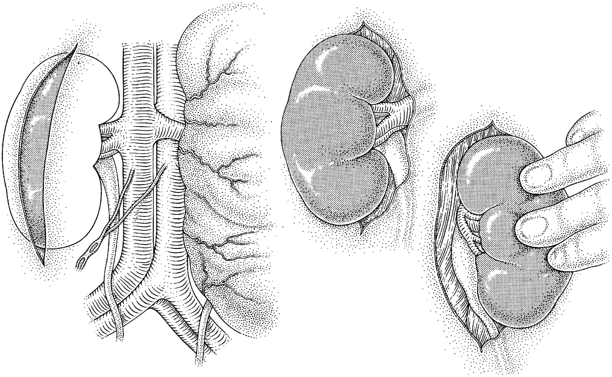

Fig. 21.2

been done. If the kidney is tumor free, a contralateral nephrectomy is undertaken. If contralateral tumor is found, a biopsy is taken and management continued according to the bilateral Wilms' tumor protocol.

The colon on the side of the tumor is mobilized medially by incising the lateral peritoneum (Fig. 21.3). If there is tumor invasion of the mesocolon or colon, they may be excised in continuity. In right-sided tumors, the duodenum will need to be separated from the tumor with care.

The ureter and gonadal vessels are identified as they pass over the pelvic brim and are ligated and divided separately (Fig. 21.4).

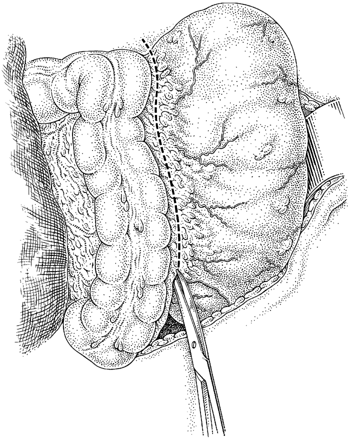

Fig. 21.3

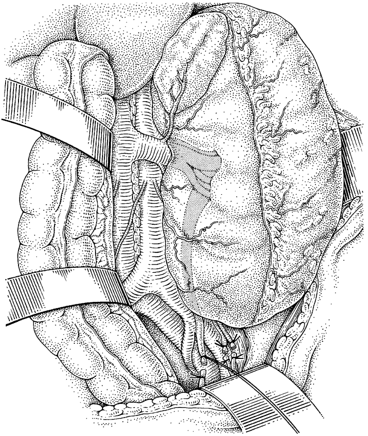

Fig. 21.4

The ureter is lifted up, and a plane is developed behind it. Mobilization is then continued by sharp dissection along the aorta, taking some lymph nodes along with the specimen (Fig. 21.5). Further hilar and para-aortic nodes should be sampled for histology, but radical node dissection is not indicated.

When the renal pedicle is reached, the vein overlies the artery. The vein is palpated to exclude the presence of tumor thrombus within it. Provided that it is clear, division of the artery and vein can then be undertaken (Figs 21.6 and 21.7). It is preferable to ligate the artery first, but occasionally, with an extremely large tumor, it is difficult to visualize the artery because it lies posterior to the vein. In these circumstances, the vein may be divided first. Care, however, must be taken because venous congestion of the tumor will occur, and there is a danger of capsular rupture.

A sling can normally be passed around the vein, and it can be lifted either superiorly or inferiorly out of the way of the artery, allowing the latter to be ligated and divided. Alternatively, the artery may be approached posteriorly by rotating the kidney medially. Although the importance of early vascular control has historically always been emphasized, in practice if the vessels cannot be safely visualized and controlled at an early stage, further mobilization of the superior and posterior attachments should be carried out to enable this.

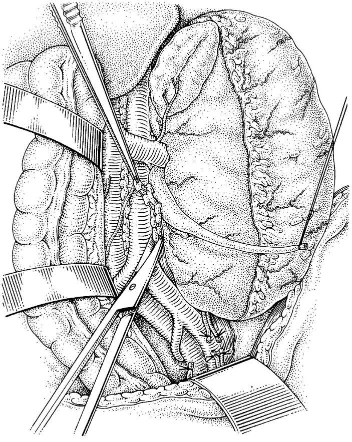

Fig. 21.5

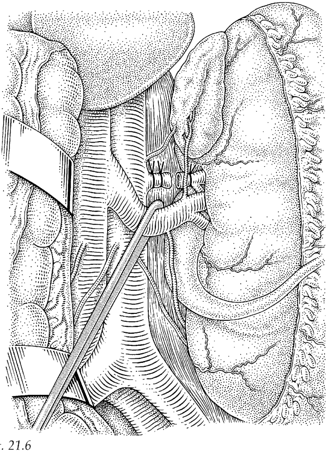

Fig. 21.6

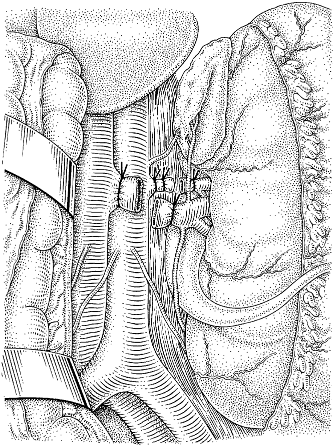

Fig. 21.7

On the right side, a plane is developed between tumor and liver capsule. The adrenal gland is usually excised en bloc with an upper pole tumor, but it may be left in situ if normal kidney adjoins the adrenal gland. The adrenal gland has a very rich blood supply, and great care must be taken to secure the adrenal veins individually if removing the gland (Fig. 21.8).

Caval extension If preoperative ultrasound has shown that there is tumor extension into the inferior vena cava adequate control must be obtained by placing slings around the inferior vena cava above and below the level of the tumor thrombus as well as around both renal veins. If access to the inferior vena cava is difficult anteriorly, it may be conveniently

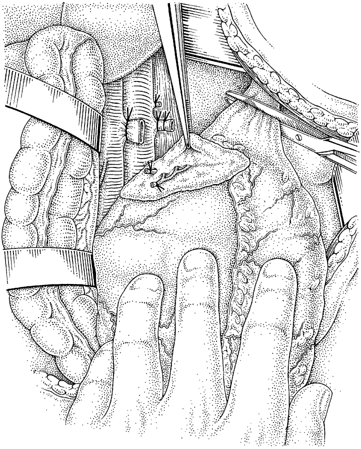

Fig. 21.8

approached from the right side after 'Kocherization' of the duodenum. After venotomy, the thrombus is removed; attachment to the intima may be quite firm, requiring dissection to free it using a probe or Watson Cheyne dissector. The venotomy is then closed with a 6/0 monofilament suture (Fig. 21.9).

The tumor can now be freed from the posterior abdominal wall using sharp and blunt dissection (Fig. 21.10). Hemostasis of the tumor bed is secured using bipolar diathermy. No drainage is required. The tumor should be sent fresh to the histopathologist with sutures to mark the renal vein and any crucial resection margins. Lymph nodes are sent in formalin.

Partial nephrectomy This should be reserved for bilateral tumors or tumors arising in a solitary kidney. Although partial nephrectomy has been performed for unilateral tumors, it is applicable in lower than 5% of cases, and there is a 20% recurrence rate (McLorie et al 1991). Thus, in the authors' opinion, it is difficult to justify it for unilateral tumors in patients with a normal contralateral kidney.

Postoperative care

Continuous nasogastric suction and intravenous fluid therapy are given. Chemotherapy according to protocol is prescribed, initially usually a single dose of vincristine: this was traditionally given intraoperatively, but some surgeons prefer to defer the dose until ileus has resolved.

Complications

Surgical mortality should be less than 1% (Pritchard et al 1995) and is either early, caused by bleeding or tumor embolus, or late, arising from adhesions. Morbidity includes prolonged ileus and small bowel intussusception.

Metastases

Pulmonary or liver metastases persisting after chemotherapy justify resection if they are sufficiently localized.

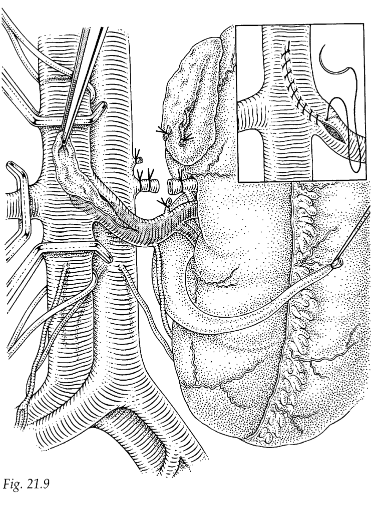

Fig. 21.9

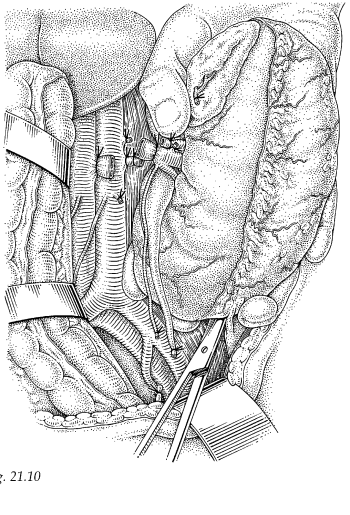

Fig. 21.10

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Chapter 22

Standard orchidopexy techniques and microvascular orchidopexy

Edwin A. Smith and John R. Woodard

Introduction

Surgical re-positioning of the undescended testis to a scrotal location is one of the most common procedures performed by the pediatric urologist. Although the details of caring for this problem continue to evolve, the goals of management remain constant. The purpose of orchidopexy includes promoting spermatogenic and androgenic function, providing cosmetic and psychological benefit, and improving testicular accessibility for the early detection of neoplastic change. An accurate preoperative physical examination, appropriate surgical timing and the selection of the surgical approach are aspects of treatment that must be familiar to the surgeon.

Preoperative assessment

Because the findings at physical examination are the initial determinants of the operative approach, the examination must be accurate. A palpable undescended testis should be differentiated from a non-palpable one. If the testis is palpable, its position is noted, and the distinction between a retractile testis and a truly undescended testis is made. The definition of these points requires the patient to be relaxed in a warm, comfortable environment and to be examined by warm hands to avoid cremasteric retraction. A sitting, cross-legged position may also help to eliminate the cremasteric reflex. With a firm sweeping motion from the anterior superior iliac spine forwards to the external ring, an attempt is made to locate and displace the testis as distally as possible.

A retractile testis may reside at any level along the line of descent but can be manipulated to the dependent portion of the scrotum, where it will briefly remain after it has been released. Intervention is not necessary for the retractile testis. Hormonal therapy may be useful if it is unclear whether the testis is retractile, but we do not use hormonal therapy as a method of managing the undescended testis. A testis that cannot be brought to the lower scrotum without tension represents an undescended testis. Radiographic imaging techniques have not been found to be useful for preoperative assessment. When orchidopexy is planned, re-examination following anesthetic induction should be performed. A non-palpable testis occasionally becomes palpable, and on rare occasions a retractile testis is diagnosed in this manner.

Timing of surgery

The optimal timing for orchidopexy is based on two factors: the age beyond which spontaneous descent is unlikely to occur, and the age beyond which the development of irreversible germ cell damage does occur. At birth, approximately 30% of preterm infants and 3–4% of term infants display cryptorchidism (Scorer and Farrington 1971). During the first few months, further descent may occur, so that at 9 months the incidence is 0.8%, which is the same incidence as found in postpubertal males (Kaplan 1993).

The point at which testicular injury becomes significant has been more difficult to delineate. Indeed, there is evidence that the germinal epithelium of the cryptorchid testis is intrinsically defective (Cortes et al 1995). Distinct, acquired changes are, however, recognized by standard histology at 18 months of age and are noted by electron microscopy even at 1 year of age (Mininberg et al 1982). We therefore currently recommend that orchidopexy be performed between 9 and 12 months of age. An associated clinically evident hernia would, of course, mandate earlier intervention.

Selection of surgical technique

The surgical approach is based on the position of the testis as determined by physical examination. For the palpable testis, a standard inguinal approach is nearly always successful, but the surgeon should be familiar with the Fowler–Stephens orchidopexy when the spermatic vessels are unexpectedly short.

The expanding role of laparoscopy has added new considerations to management. Diagnostic laparoscopy may be performed for the non-palpable testis to confirm its presence and determine its anatomic position. The possible findings at laparoscopy should be understood, and the surgeon should be equipped with a knowledge of the management options. These possibilities should also be discussed with the child's parents, obtaining an agreement for orchiectomy if indicated. Therapeutic laparoscopy may be undertaken to remove the testis if it is not suitable for orchidopexy, to accomplish the first stage of a staged orchidopexy or even to complete the orchidopexy.

Palpable undescended testis

Inguinal approach

A transverse groin incision measuring 2–4 cm is made overlying the inguinal canal. By positioning the incision within a skin crease along one of Langer's lines at this level, access to the inguinal canal is afforded and cosmesis is optimized. The incision is deepened through Scarpa's fascia to expose the external oblique aponeurosis, the external ring and often the emerging testis (Fig. 22.1). The external oblique aponeurosis forming the roof of the canal is incised using a scalpel, and the scissors are used to extend the incision along the course of the external oblique fibers down through the external ring. The ilioinguinal nerve is observed during this maneuver, and care must be taken to avoid injuring it. The cord structures and testis are elevated from the floor of the canal, leaving the ilioinguinal nerve behind. The course of the vas, which may loop below the body of the testis, is observed. With assurance that the vas is protected, the

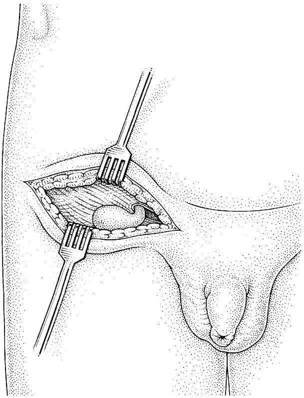

Fig. 22.1

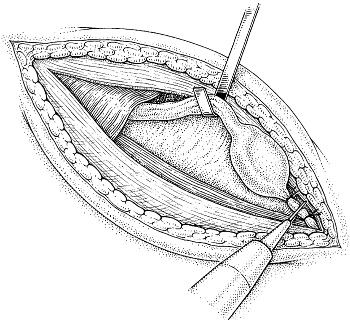

Fig. 22.2

gubernacular attachments are divided below the testis (Fig. 22.2).

The cremasteric muscle fibers are separated from the cord and divided; the tunica vaginalis is opened exposing the testis, and the testicular and epididymal appendages are removed. Probing of the tunica vaginalis proximally will usually reveal a patent processus. Downward traction on the testis at this point allows a general assessment of cord length. If a Fowler–Stephens orchidopexy is necessary, the decision should be made prior to dissecting the processus vaginalis. If a standard orchidopexy is feasible, the processus is divided just above the testis and carefully dissected to the level of the internal ring, where it is suture-ligated (Fig. 22.3). A further separation of the cord structures from the peritoneum

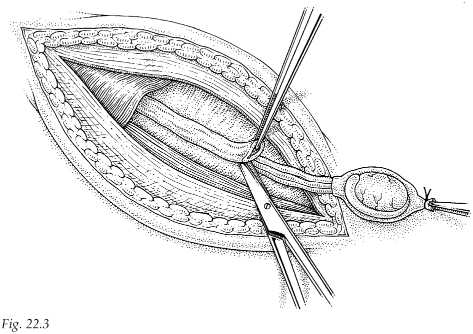

Fig. 22.3

just above the internal ring may then be accomplished. The lateral spermatic fascia, which is now evident extending up through the internal ring lateral to the cord, is divided (Fig. 22.4).

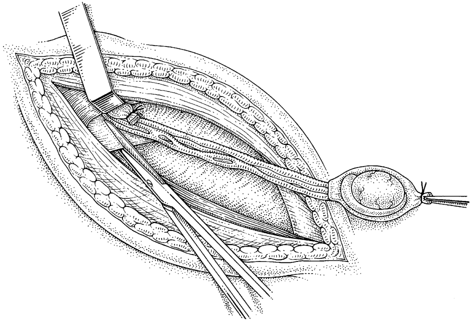

Fig. 22.4

When these measures have been completed, the testis will generally readily reach the scrotum. Additional maneuvers should not be expected to provide a great deal more cord length but may be beneficial. The tranversalis fascia forming the floor of the canal may be opened just below the internal ring. This will expose the inferior epigastric vessels, under which the cord may be transposed to provide a more direct course toward the scrotum (Fig. 22.5). Division of the epigastric vessels is unnecessary. The spermatic vessels may also be dissected away from the peritoneum to a higher level retroperitoneally to provide greater length.

Rarely, these methods are exhausted, leaving the vas and vessels skeletonized with failure to achieve a scrotal testis. At this point, it may be necessary to resort to a staged orchidopexy, bringing the testis down as far as is possible with fixation to the pubic tubercle with non-absorbable sutures, or to placement in a high dartos pouch. Wrapping the testis and cord in a silastic sheath may facilitate the dissection at the second stage, which is accomplished no sooner than 6 months later.

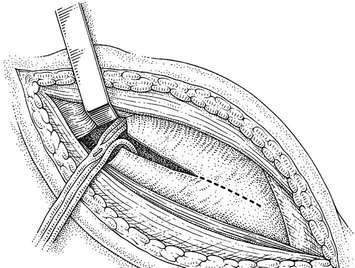

Fig. 22.5

When an adequate cord length is obtained, the path from the external ring to the scrotum is next developed bluntly by the surgeon's finger. An incision is then made transversely on the mid-anterior scrotal wall and deepened to expose the dartos fascia (Fig. 22.6). A subdartos pouch is developed bluntly with gentle spreading of a hemostat in this plane

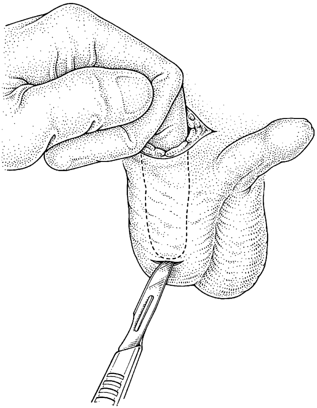

Fig. 22.6

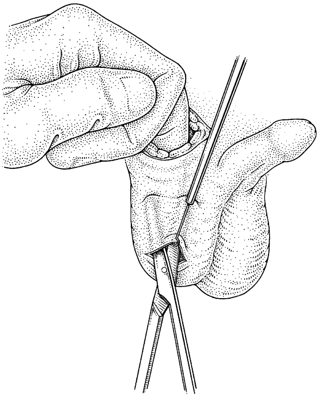

Fig. 22.7

(Fig. 22.7). A hemostat is passed from the scrotal incision up to the inguinal canal, where the tunica vaginalis is grasped and the testis is delivered to the subdartos pouch (Fig. 22.8). The transinguinal course of the cord structures should be inspected to ensure that torsion of the cord has not occurred during these manipulations.

There is controversy regarding the placement of fixation sutures through the tunica albuginea. The dartos fascia may be closed behind the testis with the inclusion of a bite of tunica vaginalis to maintain the testis in the scrotal position. Alternatively, the suture may be passed superficially through the tunica albuginea and through the dartos fascia to fixate the testis more assuredly. The scrotal incision is then closed using a simple inverted absorbable suture.

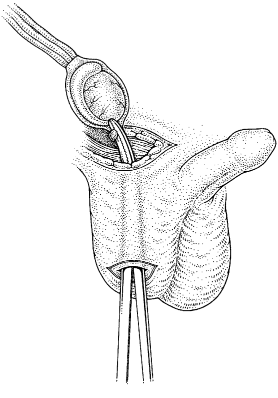

Fig. 22.8

Finally, the course of the cord is again observed, and the ilioinguinal nerve is returned to the inguinal canal. The roof of the canal (external oblique aponeurosis) is reconstituted over the cord with an interrupted suture. The subcutaneous tissue is re-approximated, and the skin is closed in a subcuticular fashion (Fig. 22.9).

Two-stage orchidopexy

As suggested by the preceding discussion, a staged orchidopexy is generally not planned from the outset but rather resorted to when all of the usual surgical maneuvers have failed to produce a scrotal testis. If a silastic sheath is used, it is tailored to surround the testis and cord up to the level of the internal ring. The sides of the sheath are sutured together using non-absorbable sutures, and the wound is copiously irrigated with antibiotic solution. The wound is closed by re-approximating the external oblique and the more superficial layers in the usual fashion. The possibility of infection with testicular loss remains present, and for this reason we have been reluctant to employ this technique.

The second stage is performed 6 months to a year later. The old inguinal incision may be used unless it was originally inconveniently placed. The incision is deepened through the subcutaneous tissue to the external oblique. If a silastic sheath was used, the external oblique may be entered parallel to its fibers and incised down through the external ring, thereby exposing the protected cord. The testis must then be dissected free from the pubic tubercle and the sheath slipped off over the testis. Further descent is then achieved by dissection at the level of the internal ring and more proximally retroperitoneally. The internal ring must be opened. Additional length is generally gained by a division of the lateral spermatic fascia and mobilization of the vessels within the retroperitoneum. These measures are accomplished most safely by exposing the peritoneum just above the internal ring and opening it anteriorly. The scar that has formed around the cord structures at the internal ring is thereby avoided, and the course of the vas and vessels beneath the posterior peritoneum can be identified.

The plane of dissection beneath the posterior peritoneum is then developed. The peritoneal edges may subsequently be gathered together and ligated or alternatively closed with a running suture. A small Dever retractor is placed beneath the posterior peritoneum to facilitate dissection of the retroperitoneum.

Re-do orchidopexy (Fig. 22.10)

When a silastic wrap has not been used, the external oblique is identified and dissected down to the level of the external ring. The testis is identified and separated from the pubic tubercle. During the dissection of the testis, no effort is made to clear the adherent fibrofatty tissue, and care is taken to avoid injury to the epididymis and vas. A traction suture may be placed through the mid-pole of the testis to facilitate this dissection. Using a hemostat, the area just medial and lateral to the cord structure within the

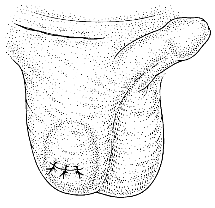

Fig. 22.9

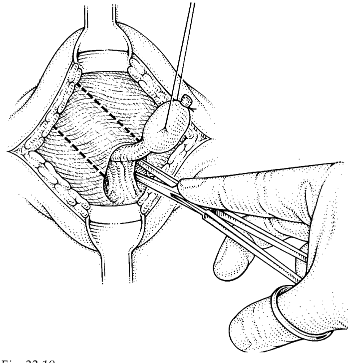

Fig. 22.10a

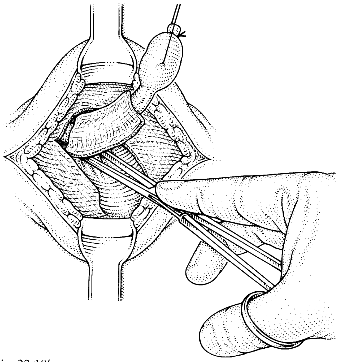

Fig. 22.10b

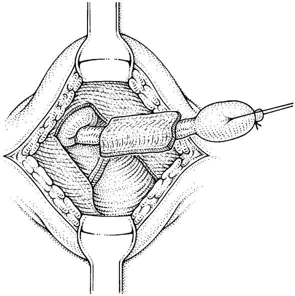

Fig. 22.10c

canal is developed. The external oblique is then incised along the medial and lateral aspects of the inguinal canal, leaving a strip of fascia overlying the cord structures. This method avoids the dense scar that is often present between the cord structures and the posterior surface of the external oblique where it was previously closed (Cartwright et al 1993). The parallel incisions are connected with a transverse incision above the level of the internal ring. The proximal dissection is then undertaken as described above.

Non-palpable undescended testis

Laparoscopy may be performed for the unilateral non-palpable undescended testis but has its greatest benefit in the management of bilateral non-palpable testes. In the case of bilaterality, the possibility of intersex should be considered first and excluded by rectal examination, pelvic ultrasound and karyotyping. If a genetic male is proven and müllerian structures are absent, anorchia is then excluded by human chorionic gonadotropin stimulation test. Laparoscopy is then undertaken.

In the case of unilateral or bilateral non-palpable undescended testis, a spectrum exists for the quality of the gonad and for its anatomic position, and a surgical response should be anticipated for each scenario. When a blindly ending vas and vessels are clearly seen, a diagnosis of a 'vanished testis' is made, and no further therapy is indicated. An atrophic testis that has no spermatogenic potential is removed if the position and quality of its contralateral mate are secure. When the vas and vessels enter the internal inguinal ring, or a 'peeping testis' is seen that pops in and out of the internal ring, an inguinal approach is indicated and a standard orchidopexy will usually be successful. For a more proximally positioned testis, a Fowler–Stephens orchidopexy via an inguinal or abdominal approach, or microvascular surgery, will be necessary. In experienced hands, laparoscopy may also be used interventionally to remove an atrophic testis, perform the first stage of a Fowler–Stephens orchidopexy or mobilize the vessels to the level of the inferior pole of the kidney to facilitate the subsequent inguinal approach. More recently, completion of the orchidopexy entirely laparoscopically has been described.

Fowler–Stephens orchidopexy

The Fowler–Stephens orchidopexy relies on the dual vascular supply to the testis from the spermatic artery and the vasal artery. The technique is applicable when the spermatic vessels are too short to permit descent of the testis, a collateral supply between the vessels can be demonstrated and the vas deferens loops below the testis and is of adequate length. The spermatic vessels may then be divided and the testis brought down to the scrotum, suspended by the vas and its vessels (Fowler & Stephens 1959). As suggested earlier, this maneuver must be premeditated to allow the protection of the collateral vascular arcade. The first stage of the Fowler–Stephens orchidopexy may be accomplished by ligating the spermatic vessels laparoscopically. This allows collateral vasculature to hypertrophy, possibly improving the success rate when the second stage with testicular mobilization is performed approximately 6 months later (Ransley et al 1984, Bloom 1991).

When the procedure is performed as a single stage, the inguinal canal is entered as described and the tunica vaginalis opened as it invests the testis. The looping vas deferens and vascular arcade between the spermatic vessels and vasal vessels are identified. The posterior aspect of the processus vaginalis must be perserved as a peritoneal flap to protect the underlying communications between the vessels. The processus is closed at the level of the internal ring without more distal dissection. The spermatic vessels will be divided above the level of the internal ring and above their point of convergence with the vas. By placing a vascular clamp on the spermatic vessels at this level, the effect on the testis is assessed (Fig. 22.11). The tunica albuginea may also be incised and the quality of bleeding noted if the adequacy of the testicular blood supply remains in question. With testicular vascularity confirmed, the spermatic vessels are ligated and divided (Fig. 22.12). To straighten the vas further it may be necessary to divide one or two of the vascular arcades crossing the loop. The testis is then brought to the scrotum and sutured in place using the subdartos technique.

Extended inguinal approach

Whereas some have routinely employed laparoscopy for the non-palpable testis, others have continued to address this problem via an inguinal approach. In the majority of cases, the testis will reside just inside the inguinal ring or distal to this point, and the inguinal incision will be sufficient for a successful orchidopexy. Placing the incision at the level of the internal ring will facilitate access to the peritoneal cavity if intra-abdominal exploration is needed. A useful landmark for judging the position of the internal inguinal ring is the mid-point between the anterior superior iliac spine and the pubic tubercle. The inguinal canal is entered as described, and the ilioinguinal nerve, consistently present regardless of the status of the testis, is protected. If the testis or processus are found within the canal, the processus is opened and the length of the cord and vessels assessed. Either a standard orchidopexy or a Fowler–Stephens procedure is employed.

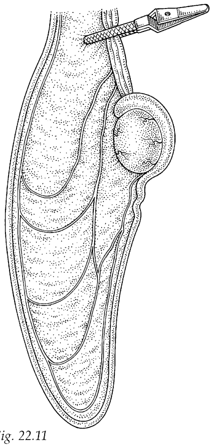

Fig. 22.11

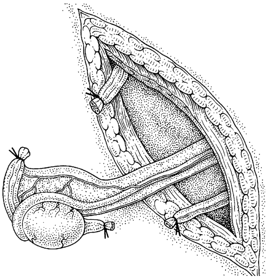

Fig. 22.12

Failure to locate the testis demands an intraperitoneal search. This is accomplished by the superior retraction of the internal oblique, as its arching fibers form the internal ring, and identification of the peritoneum. The peritoneum is then entered and the lower quadrant of the abdomen is searched. Once the testis has been located, the decision to bring it down or remove it must be made based on the quality of the testis, the likelihood of successful orchidopexy, as determined by vasal and vascular anatomy, and the status of the contralateral testis. If the testis is not initially located, the search must not end until the region from lower pole of the kidney to the level of the internal ring and either the testis or a blind ending vas and vessels are found. The search is often facilitated by following the course of the vas distally. A high intra-abdominal testis will require either a Fowler–Stephens procedure or microvascular autotransplantation.

Transabdominal orchidopexy

Because of the general agreement for early orchidopexy and the ability to achieve wide retroperitoneal access in the young child through an inguinal approach, the transabdominal approach is an infrequently employed open technique. It retains some usefulness in the older child with an intra-abdominal testis that has escaped intervention or for the child with prune belly syndrome, either in concert with other reconstructive surgery or as a separate procedure.

A midline incision from the umbilicus to the symphysis is used and carried down between the rectus muscles. The peritoneal cavity is entered, and the intestine is packed away to allow exposure of the posterolateral aspect of the bladder. The testis is usually found in this location, the vas being mobilized on a wide pedicle of peritoneum. The spermatic vessels are mobilized, and a Fowler–Stephens procedure is selected if necessary. When adequate descent of the testis has been achieved, a scrotal incision is made and a dartos pouch developed. The scrotal wall is then elevated up to the external ring with the forefinger, and a hemostat is pushed down through the transversalis fascia at the internal ring and onto the fingertip at the scrotal wall. The testis is then brought through this channel and positioned in the subdartos pouch.

Microvascular autotransplantation

Testicular autotransplantation represents a more controversial option for the management of the high undescended testis. The possibility of testicular atrophy with the application of conventional orchidopexy methods and the impressive success rates with autotransplantation have generated some enthusiasm (Bukowski et al 1995). Detractors have suggested extensive proximal mobilization of the vessels, as is now possible laparoscopically, which improves the likelihood that conventional methods of orchidopexy will suffice. Furthermore, the merit of investing extensive time and effort into the preservation of a testis that is unlikely to support spermatogenesis has been questioned. The utility of this approach is perhaps most clear in the case of bilateral intra-abdominal testes when autotransplantation may more reliably place at least one testis with a full blood supply in a scrotal position. The procedure obviously carries the contingency of the operator possessing expertise in microvascular techniques.

A transverse groin incision is made at the level of the internal ring (Fig. 22.13). The inguinal canal is opened, and access to the peritoneum is gained at the internal ring. The testis is then delivered from the abdomen, and the peritoneal defect is closed. Under

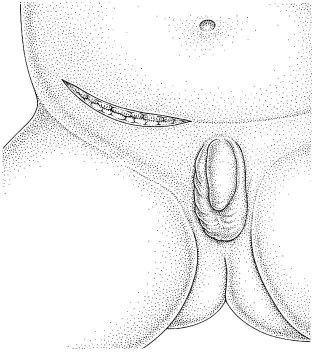

Fig. 22.13

loupe magnification, the spermatic vessels are dissected retroperitoneally towards their origin. The dissection should be far enough proximally to be above the confluence of the pampiniform veins, thereby allowing for a single-vessel venous anastomosis. The vas is mobilized on a wide strip of peritoneum in a Fowler–Stephens fashion.

The inferior epigastric vessels are first identified just below the internal ring, and the transversalis fascia forming the floor of the inguinal canal is incised to provide full exposure (Fig. 22.14). The vessels are then carefully dissected beneath the transversus abdominus up toward the lateral border of the rectus muscle. The side branches are ligated at some distance from the vessels as their luminal caliber may later be found to be appropriate for anastomosis. With a length of 8–9 cm dissected, the vessels are ligated distally, clamped proximally with microvascular clamps and divided (Fig. 22.15). The vessel ends are then washed with 10 units/ml heparin saline.

The subdartos pouch is formed, and a path is opened from the inguinal canal to the scrotum with a finger. After the closure of the peritoneal defect at the internal ring, the previously dissected spermatic vessels are ligated as close to their point of origin as possible and divided. The ends are washed with heparin saline and, if back-bleeding occurs, microvascular clamps are applied. The testis is then transferred to the subdartos pouch.

The operating microscope at a power of × 25 is required for the anastomosis. The vessels' ends are cleared of extraneous adventitial tissue, and the disparity of luminal caliber is assessed.

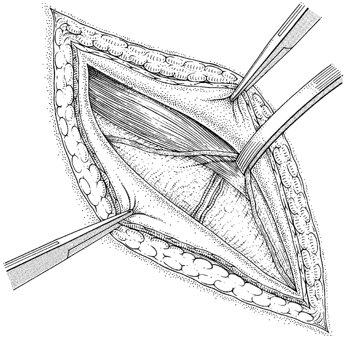

Fig. 22.14

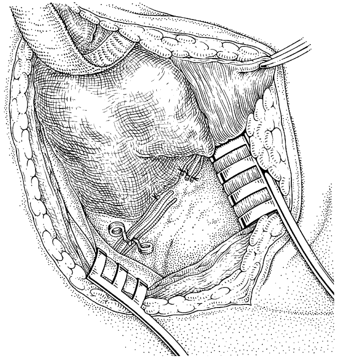

Fig. 22.15

The discrepancy between the venous ends is usually not great, and a direct circular anastomosis is possible (Fig. 22.17). A mismatch between the arteries is usually noted because of both a larger diameter and a thicker vessel wall of the inferior epigastric artery. This discrepancy may be addressed by spatulation of the spermatic artery (Fig. 22.18), end-to-side anastomosis or the use of a reverse vein stepdown graft. The latter may be harvested from a side branch of the inferior epigastric vein. The anastomoses are performed in an interrupted fashion using 10/0 monofilament nylon with a 50 μm needle.

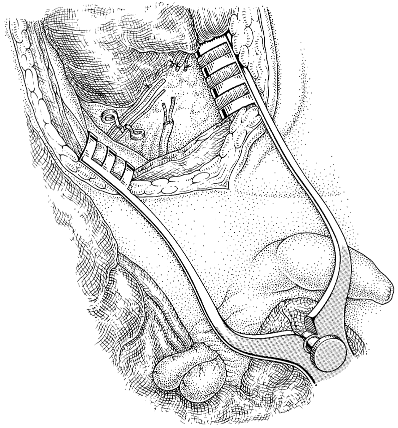

Fig. 22.16

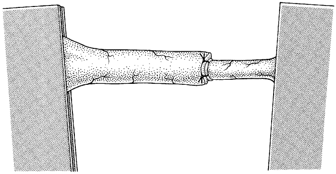

Fig. 22.17

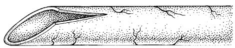

Fig. 22.18

Observation of the anastomosis with patency maintained for greater than 20 minutes correlates with a good long-term outcome (Fig. 22.19). Any evidence of vascular compromise during this interval should prompt a revision of the anastomosis. The testis is then fixed in place, the scrotal incision is closed, and the layers of the inguinal incision are re-approximated. The child is restricted to bedrest for 24 hours and then discharged home.

Indications for orchiectomy

The prime goal in the management of cryptorchidism is to provide the testis with a normal scrotal position, which is usually achievable. Removal rather than preservation should, however, be recognized as the preferred management in certain conditions. This decision is made when there is a normal scrotal contralateral testis and the benefit of orchidopexy is compromised by one of the following conditions:

1. The undescended testis occupies a high position such that it would be difficult to bring down.
2. The quality of the testis is poor and unlikely to support spermatogenesis.
3. Significant adnexal anomalies coexist, such as epididymal gonadal dissociation, vasal atresia or a short vas deferens that will not allow testicular descent even with division of the vessels.

There is also no recognized benefit of orchidopexy in the postpubertal patient with unilateral cryptorchidism.

Complications

Intraoperative

When a palpable testis is encountered and a standard transinguinal orchidopexy is employed, the expectation of a perfect result is realistic and nearly always achievable. The most common mistake is a failure to achieve an adequate position for the testis. A second operation with a more complete retroperitoneal dissection usually succeeds.

Injury to structures within the inguinal canal including the ilioinguinal nerve, vas deferens and spermatic vessels constitutes the second category of intraoperative complications. The ilioinguinal nerve is most vulnerable to transection when the external oblique is opened, the most common point of injury being at the external ring. Entrapment of the nerve should be avoided during closure of the external oblique. The vas may be injured during the dissection of the processus vaginalis. Twisting the processus vaginalis followed by suture ligation is a common method of controlling the hernia sac at the internal ring. This carries the risk of gathering the vas in the closure, thereby producing obstruction. If complete transection of the vas is recognized, microsurgical repair may be undertaken.

Vascular compromise should be avoided by a meticulous handling of the spermatic vessels as there is no recourse for the testis that is found to have atrophied when examined postoperatively. The Fowler–Stephens method carries an approximately 15–20% risk of testicular atrophy, a detail that should be included in preoperative counseling. This risk is possibly lowered by staging the ligation of the vessels and subsequent orchidopexy.

Postoperative

Scrotal swelling caused by edema is nearly always present to some degree. If there is continued expansion of the scrotal compartment, active bleeding or bowel herniation may be responsible. These conditions demand exploration. Delayed retraction of the testis has been described. This possibility and the well-known increased risk of testicular malignancy provide motivation for the yearly monitoring of the patient through the pediatric years. As an adult, the patient is counseled regarding fertility, and a semen analysis is performed if the patient desires. The risk of malignancy is reiterated, and testicular self-examination is taught.

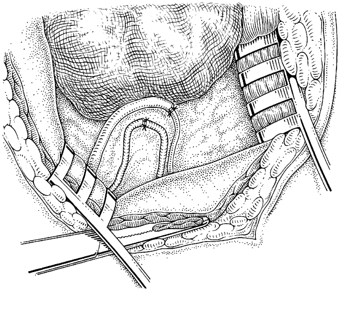

Fig. 22.19

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Chapter 23

Circumcision, urethral prolapse, penile torsion, buried penis, webbed penis and megalourethra

Jack S. Elder

Circumcision

Phimosis refers to the condition in which the prepuce cannot be completely retracted. In the newborn, phimosis is physiologic because of adhesions between the foreskin and the glans. By 4 years of age, more than 90% of boys demonstrate complete retraction of the foreskin. If the foreskin is retracted slightly and the phimotic ring is broken, secondary phimosis may occur, with cicatrix formation at the site of the phimotic ring. In these boys, a pale white scar may be visualized at the tip of the foreskin, preventing preputial retraction.

In boys who are uncircumcised, there are potential health consequences. For example, the risk of urinary tract infection during the first year of life is approximately 10 times higher than in circumcised boys (Wiswell 1992, American Academy of Pediatrics 1999). The increased risk results from a colonization of the glans and inner foreskin by urinary pathogens. In uncircumcised men with poor hygiene, the risk of penile cancer is significantly greater, but the risk of penile cancer is extremely low if good hygiene is maintained. Balanoposthitis also is more likely to occur in uncircumcised boys. In some adult men who are uncircumcised, the frenulum tears during intercourse. On the other hand, some men who undergo circumcision in adulthood have indicated that sensitivity of the glans is diminished following this procedure.

In the United States, circumcision is the most commonly performed operation (Ross 1996). Although it is in many boys undertaken as a religious preference, the procedure is usually carried out for social reasons.

Indications for circumcision include secondary phimosis, congenital anomaly predisposing to urinary tract infection (i.e. hydronephrosis or vesico-ureteric reflux), balanoposthitis and febrile urinary tract infection. Contraindications to circumcision in the neonate include hypospadias, chordee without hypospadias, dorsal hood deformity, webbed penis and microphallus. In addition, many neonates with a large hydrocele or hernia are more likely to develop secondary phimosis and a buried penis if a circumcision is performed.

The techniques for circumcision utilized in the newborn are different from those used on older boys. In the newborn, a Plastibell or Gomco clamp is generally employed. In ritual circumcisions, a Mogen clamp is used. In boys less than 2–3 years of age, the Plastibell technique may be effective, but in most cases formal incision of the foreskin with suturing of the skin edges is necessary.

Proper postoperative care is essential to having a successful outcome. In some boys, penile adhesions develop and can be prevented by being certain to retract the penile skin off the glans 2–3 times daily and applying a liberal amount of antibiotic ointment to the operative site for 10 days.

Surgical technique

The foreskin is retracted to expose the glans. A 5/0 polypropylene traction suture is placed through the glans (Fig. 23.1). The foreskin is then pulled over the glans and a circumferential incision is made along the line of the coronal sulcus.

The foreskin is then retracted, and the distal aspect of the frenulum is incised transversely to eliminate any glanular tilt. A second circumferential incision

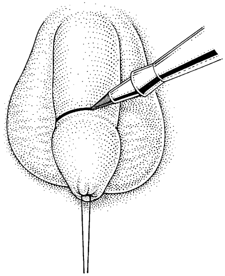

Fig. 23.1a

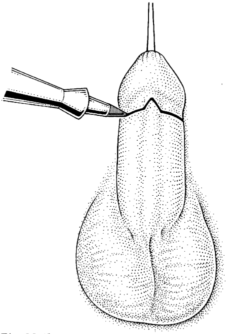

Fig. 23.1b

is made 1.5 cm proximal to the coronal sulcus to leave enough of the mucosal skin for suturing (Figs 23.2 and 23.3).

The two incisions are connected by dividing the dorsal skin with scissors (Fig. 23.4a). The foreskin is then carefully excised circumferentially (Fig. 23.4b). Bleeding vessels should be carefully cauterized. Alternatively, they may be ligated with 5/0 Vicryl. Dorsal and ventral sutures of 5/0 polyglycolic acid are placed. The wound on each half of the penis is then closed with a running 6/0 fast-absorbing Vicryl (Figs 23.5 and 23.6). By not tying this suture too tightly through the skin, the wound will heal with a smooth surface resembling a newborn circumcision. Alternatively, the skin may be closed with a subcuticular stitch.

Comment

The use of unipolar diathermy coagulation of vessels has been regarded by some as dangerous during penile surgery. The method can, however, be safely used for hemostasis as an alternative to vessel ligation provided that the penile shaft is kept in contact with the body wall while the current is applied. Alternatively, bipolar diathermy may be used safely.

Each surgeon has a preferred dressing; a Xeroform gauze applied to the wound is generally sufficient. The child may be bathed 48 hours after the procedure.

Newborn techniques

There are two primary techniques utilized for most boys undergoing neonatal circumcision: the Plastibell technique and the Gomco clamp. During ritual circumcision, the Mogen clamp is employed. Local anesthesia generally is preferred. A dorsal penile block or ring block at the penile base is performed with 1–2 cm^3 of 0.25% bupivacaine or 1% lidocaine.

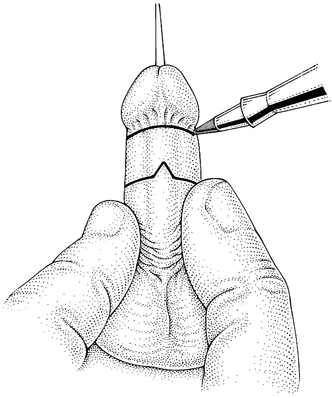

Fig. 23.2

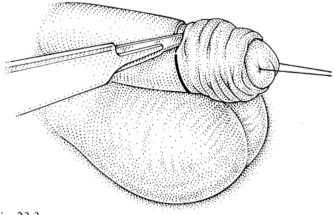

Fig. 23.3a

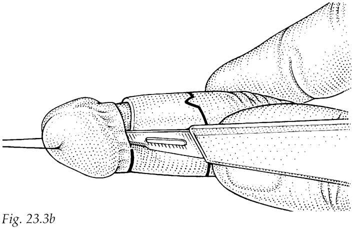

Fig. 23.3b

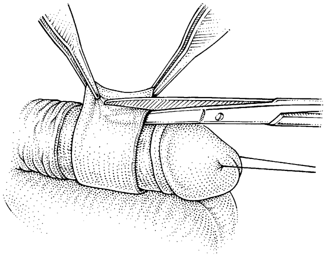

Fig. 23.4a

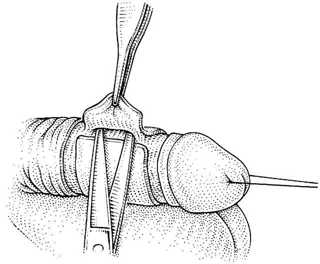

Fig. 23.4b

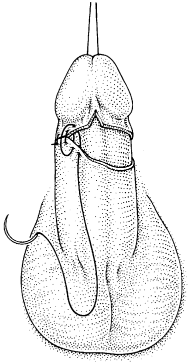

Fig. 23.5

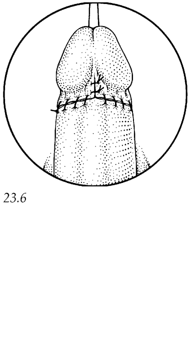

Fig. 23.6

The phimotic ring is stretched with a clamp and the foreskin is retracted slightly. A dorsal slit is then performed by placing a straight hemostat on the prepuce at the 12 o'clock position. The hemostat is then removed and the crushed line is incised (Fig. 23.7). The remainder of the prepuce is swept off the glans. Eventually the frenulum is incised, allowing complete foreskin mobilization. Another dorsal slit is made until the coronal sulcus is reached. The appropriate size Plastibell is selected. This plastic ring comes in five sizes: 1.1, 1.2, 1.3, 1.5 and 1.7 cm diameter.

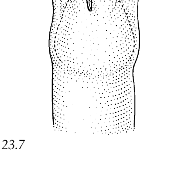

Fig. 23.7

The foreskin is then drawn up over the Plastibell until an optimal fit is achieved (Fig. 23.8). An 0 silk suture is tied in the groove (Fig. 23.9) and the handle is broken off. The Plastibell should fall off in 3–10 days.

In the Gomco technique, the appropriate size Gomco bell is selected. After making the dorsal slit, the Gomco bell is placed on the glans. The prepuce is pulled over the bell and through the plate and yoke portions of the Gomco clamp until the appropriate amount of skin can be removed. The prepuce is cut next to the top of the plate. The entire clamp, bell and excised prepuce are removed. If necessary, an interrupted 5/0 Vicryl suture is used to control any bleeding that may occur.

Complications

The reported complication rate is 0.2% to 5% (Baskin et al 1996), the most common early complication being bleeding, which is usually minor. Frequently bleeding can be managed with the application of gelfoam, silver nitrate or an epinephrine solution. Occasionally, a few Vicryl sutures must be placed. If epinephrine is used, it should be diluted to 1: 100 000 to eliminate the risk of ischemic effects on the penis. Care must be used if silver nitrate is employed not to damage adjacent tissue or the underlying urethra.

Infection occurs in fewer than 1% of circumcisions. An inappropriate amount of skin may at times be excised, and penile shortening or dehiscence may result. If insufficient skin is excised, secondary phimosis may occur. More commonly, inadequate skin excision simply results in an unsatisfactory cosmetic result. A buried penis results when excess shaft skin and inadequate

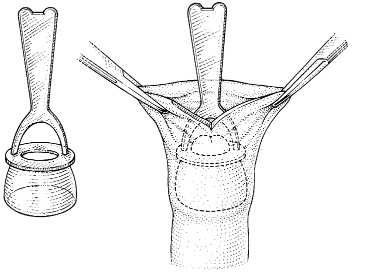

Fig. 23.8

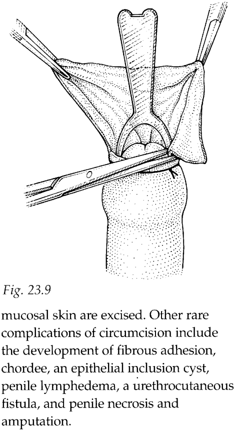

Fig. 23.9

mucosal skin are excised. Other rare complications of circumcision include the development of fibrous adhesion, chordee, an epithelial inclusion cyst, penile lymphedema, a urethrocutaneous fistula, and penile necrosis and amputation.

Urethral prolapse

Urethral prolapse presents as an interlabial mass (Elder 1998). It has the appearance of erythematous, inflamed mucosa protruding and surrounding the urethral meatus. The prolapsed appearance represents an eversion of the urethral mucosa. The lesion is not obstructive.

This condition occurs almost exclusively in black girls between 1 and 9 years of age, with an average age of 4 years. The most common sign is bloody spotting on the underwear or diaper, although dysuria or perineal discomfort may occasionally occur. Sexual abuse is suspected in some cases before the diagnosis of urethral prolapse is established (Anveden-Hertberg et al 1995).

Some think that urethral prolapse results from a poor attachment between the smooth muscle layers of the urethra in association with episodic increases in intra-abdominal pressure. No known relationship exists between external genital trauma and urethral prolapse.

After the diagnosis has been made, conventional therapy includes the application of estrogen cream 2 or 3 times daily for 1–2 weeks as well as sitz baths. If the prolapsed urethra persists for several months, it usually results in continued bloody spotting, and formal excision is necessary. In this setting, postoperative catheterization is unnecessary, and the procedure can be performed on an outpatient basis. If the child is undergoing examination under anesthesia for suspected sexual abuse, excision of the prolapsed urethra at the time is recommended.

Other causes of interlabial masses include paraurethral cyst, imperforate hymen, prolapsed ectopic ureterocele, sarcoma botryoides (rhabdomyosarcoma) and uterovaginal prolapse.

Surgical technique

In the acute phase, a Vicryl ligature can be tied around the base of the prolapse over an indwelling Foley catheter until the redundant tissue sloughs off (Fig. 23.10). In chronic cases, the edematous mucosa must be excised, generally with diathermy, as indicated in Fig. 23.11. After resection of the redundant tissue, the edges of urethral mucosa are sutured to the surrounding vulva mucosa using a 5/0 Vicryl suture (Fig. 23.12). An indwelling catheter is unnecessary.

Penile torsion

Penile torsion refers to a rotational defect of the penile shaft. It almost always occurs in a counterclockwise direction, that is, to the left side. In most cases, penile development is otherwise normal, and the condition goes unrecognized until circumcision or until the foreskin is retracted.

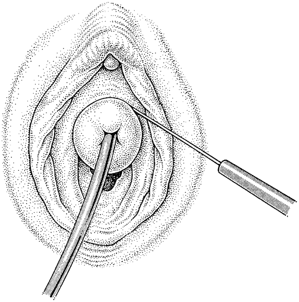

Fig. 23.11

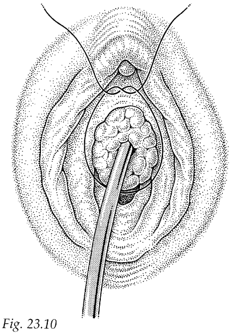

Fig. 23.10

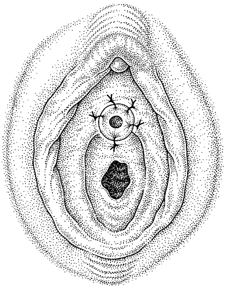

Fig. 23.12

Penile torsion can also occur in association with hypospadias or a dorsal hood preputial deformity without a urethral abnormality. In most cases of penile torsion, the medium raphe spirals around the penile shaft (Elder 1998). In general, the defect has primarily cosmetic significance, and correction is unnecessary if the rotation is less than 60 degrees from the midline.

Although the glans may be directed more than 90 degrees from the midline, the orientation of the corporal bodies and the corpus spongiosum at the base of the penis is usually normal.

Surgical technique

The foreskin is completely retracted to visualize the glans. A circumferential incision is made 1.5 cm proximal to the coronal sulcus (Fig. 23.13). The penis is completely de-gloved, allowing it to rotate clockwise to orient the meatus vertically (Fig. 23.14). The redundant foreskin is excised. In closing the skin, stay sutures (5/0 polyglycolic acid) should be placed between the dorsal penile skin in the midline and the mucosal skin on the right side of the glans, and between the ventral penile skin in the midline and the mucosal skin on the left side of the glans to correct the rotational defect (Fig. 23.15). The skin is closed with running 6/0 or 5/0 fast-absorbing Vicryl sutures (Fig. 23.16).

In more severe cases, in which the severity of torsion is more than 90 degrees, the base of the penile shaft may need to be mobilized by dividing the suspensory ligament of the penis. Next, the penile shaft is rotated clockwise until it is straight and anchored laterally to the pubic symphysis with a 4/0 or 5/0 braided non-absorbable suture. The skin is then closed as described above.

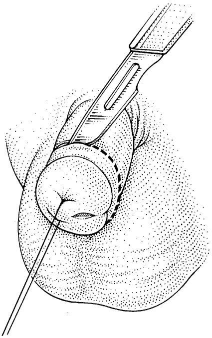

Fig. 23.13

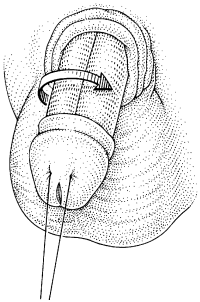

Fig. 23.14

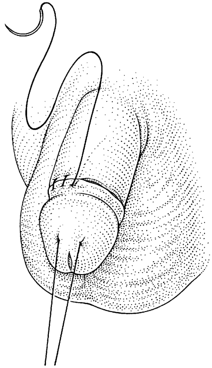

Fig. 23.15

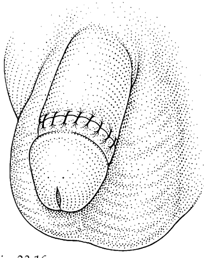

Fig. 23.16

Concealed (buried) penis

The term 'inconspicuous penis' refers to a penis that appears to be small. Several entities are included in this term, including concealed penis, webbed penis and trapped penis, in which the penis is normal in size, and micropenis, in which the penis is abnormally small (Elder 1998, Bergeson et al 1993).

A concealed penis is a normally developed penis that becomes camouflaged by the suprapubic fat pad. This anomaly may be congenital or iatrogenic following circumcision. In infants and young children, the condition has been speculated to result from inelasticity of the dartos fascia, which normally allows the penile skin to slide freely on the deep layers of the penile shaft.

On inspection, the contour of the penile shaft and the glans cannot be seen (Fig. 23.17). Careful palpation, however, allows one to determine that the penile shaft is actually normal in size and concealed rather than being a microphallus or a penis that has undergone injury during circumcision. It is important to determine whether the glans can be exposed when retracting the skin covering the glans. If so, it remains the surgeon's judgment whether correction is warranted.

Lesser degrees of concealed penis presenting in infancy generally improve spontaneously with growth, and in such cases an expectant attitude may be appropriate. In more severe cases, operative correction is needed, preferably being carried out around 6–12 months of age.

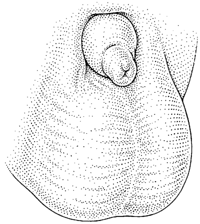

Fig. 23.17a

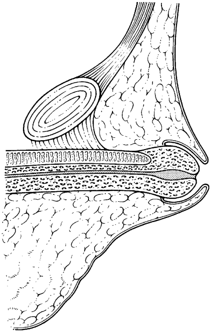

Fig. 23.17b

Surgical technique

The foreskin is pulled back off the glans, and a transverse traction stitch using a 5/0 polypropylene suture is placed. A circumferential incision is made 1.5 cm proximal to the coronal sulcus (Fig. 23.18). The penis is completely de-gloved (Fig. 23.19). The dermis of the upper scrotum and suprapubic skin are anchored to Buck's fascia at the proximal aspect of

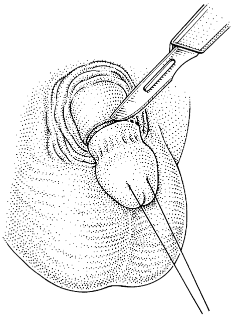

Fig. 23.18

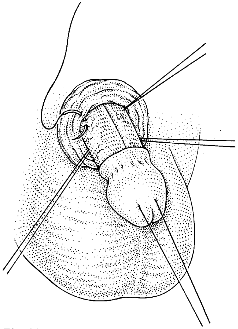

Fig. 23.19

the penile shaft with 5/0 Ethibond, Ticron or Tevdek (braided non-absorbable) sutures to prevent the penis retracting into the suprapubic fat pad in the future (Fig. 23.20). Usually, one suture is placed in each quadrant. The redundant foreskin is sharply excised. The skin is closed with interrupted 5/0 polyglycolic acid and running 6/0 fast-absorbing Vicryl (Fig. 23.21).

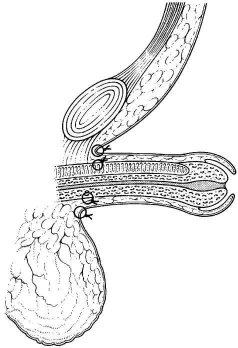

Fig. 23.20

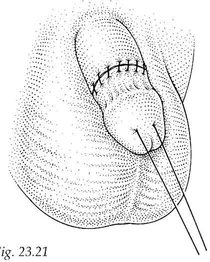

Fig. 23.21

Webbed penis

Webbed penis is a condition in which the scrotal skin extends onto the ventrum of the penis, generally to the coronal sulcus. When this condition is congenital, the penis, the urethra and the remainder of the scrotum are typically normal, the deformity representing an abnormality of the attachment between the penis and scrotum (Elder 1998). Although the webbed penis is usually asymptomatic, the cosmetic appearance is often unacceptable and may lead to difficulty during sexual intercourse in adulthood.

Surgical technique

In some cases, the web may be divided longitudinally to the base of the penis (Fig. 23.22). The skin is then resutured with interrupted 5/0 Vicryl sutures (Fig. 23.23).

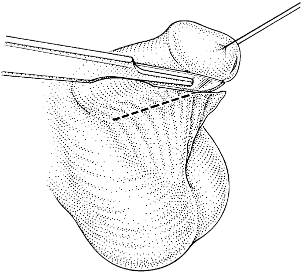

Fig. 23.22

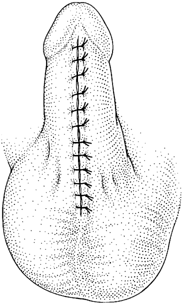

Fig. 23.23

In other cases, a circumferential incision is made 1.5 cm proximal to the coronal sulcus, Byars' preputial skin flaps are transferred to the ventral surface of the penis, and redundant foreskin is excised. The scrotum may be anchored to the base of the penis to prevent a recurrence of the webbed appearance. In rare cases, the distal urethra is hypoplastic, necessitating urethral reconstruction.

Megalourethra

Congenital megalourethra is seen in two forms: the fusiform and the scaphoid.

In the fusiform type, there is a significant deficiency or total absence of erectile tissue in the corpora cavernosa and corpus spongiosum. The penis is flabby, with redundant wrinkled skin, and is excessively long. Other genitourinary and anorectal anomalies commonly coexist. No treatment for this form of megalourethra is appropriate during childhood. In survivors to adult life, the insertion of a penile prosthesis may be possible to allow sexual activity.

With scaphoid megalourethra, the corpora cavernosa are normal, but there is a lack of spongy tissue in the corpus spongiosum of varying severity, which leads to differing degrees of dilatation of the penile urethra. The penis shows dorsal curvature with ventral swelling, which is particularly evident during micturition. In addition, the penis usually is longer than average. There is occasionally a congenital fistula in the thin urethral floor. This condition commonly occurs in boys with prune belly syndrome. Even in boys in whom the abdominal wall is normal and the testes are descended, the characteristic dysmorphic anomalies of the urinary tract frequently are present. Consequently, urologic investigation is indicated.

Treatment of the scaphoid megalourethra is necessary not only for cosmetic reasons, but also because the large urethra predisposes to urinary stasis, leading to urinary tract infection, as well as incontinence resulting from incomplete urethral emptying during micturition.

An acquired form of megalourethra occurs following the use of the transverse preputial island flap (Duckett). This complication occurs in approximately 10% of boys and typically results from a flap that is too long or too wide, a delayed meatal stenosis or a postoperative blow-out of the suture line.

Surgical technique

A circumferential incision is made 1.5 cm proximal to the coronal sulcus (Fig. 23.24). The penis is de-gloved, exposing the large urethra (Fig. 23.25). The ventral wall of the megalourethra is incised longitudinally, and

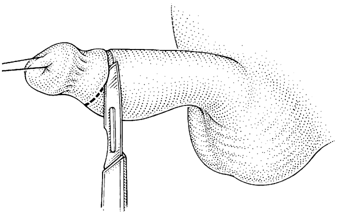

Fig. 23.24

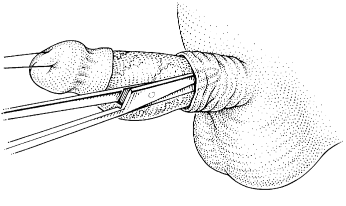

Fig. 23.25

redundant urethral tissue is excised from each side, leaving a strip of appropriate width between the proximal and distal urethral orifices (Fig. 23.26). The urethral strip is tubularized using a running layer of fine polydiaxanone sutures (Fig. 23.27). A second layer of imbricating sutures using a 6/0 polyglycolic acid suture helps to achieve a watertight closure (Fig. 23.28). The retracted penile skin is then drawn forward and sutured to the mucosal skin of the penis (Fig. 23.29). A urethral drip stent is left in place for 10–14 days postoperatively.

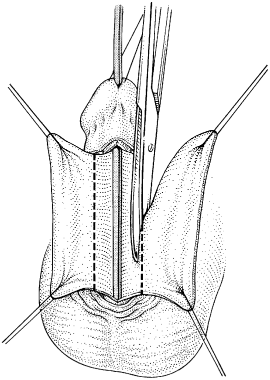

Fig. 23.26

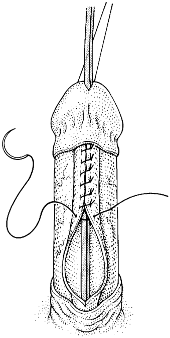

Fig. 23.27

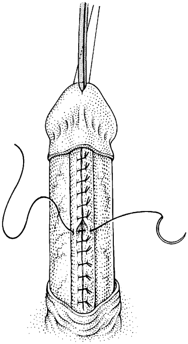

Fig. 23.28

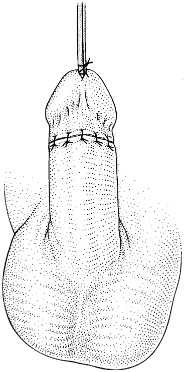

Fig. 23.29

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