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Pediatric Surgery

Second Edition

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Editors

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To our families for their love and patience.

Preface to the Second Edition

It is 13 years since the first edition of this book was published in 2006. In the intervening years, tremendous advances have been made in the management of children with surgical disorders. Minimally invasive surgical techniques are now routinely employed for most intracavity procedures in infants and children. Robot-assisted laparoscopy is evolving rapidly in the pediatric surgical field, and robotic technology is now available in many major children's hospitals around the world.

The second edition of *Pediatric Surgery* has been thoroughly revised and updated. It contains 74 chapters from 110 contributors from five continents. This edition contains 15 new chapters on key topics including thyroidectomy, empyema, gynecomastia, laparoscopy, stomas for small and large bowel, rectal biopsy, rectal prolapse, bariatric surgery, portal hypertension, liver transplantation, soft tissue tumors, ovarian tumors, laparoscopic and robotic urology, and dialysis. Each chapter has been written by internationally renowned leaders in their respective fields. Many younger surgeons were selected as coauthors, who will become the next generation of leaders in pediatric surgery and pediatric urology.

The main aim of the new edition, as with the first edition, is to provide a comprehensive description of operative techniques for various surgical conditions in infants and children. The most unique feature of the book is the generous use of high-quality color illustrations to clarify and simplify various operative techniques. The text is organized in a systematic manner providing a step-by-step, detailed, practical guide to operative approaches in the management of congenital and acquired surgical conditions in children. The book is intended for pediatric surgeons, pediatric urologists, trainees in pediatric surgery and pediatric urology, and general surgeons with interest in pediatric surgery.

The first edition of the book was very successful, accepted worldwide as a reference book for the operative management of childhood surgical disorders, and was translated into multiple languages including Chinese, Russian, and Turkish. We hope that the substantially revised and updated second edition of the book will continue to act as a reference book for pediatric surgeons all over the world.

We wish to thank most sincerely all the contributors for their outstanding work in the preparation of this innovative operative *Pediatric Surgery* atlas. We wish to express our gratitude to the editorial staff of Springer, for all their help during preparation and publication of this book. We wish to thank Dr. Hiroki Nakamura for help with the galley proofs of the book.

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Thyroglossal Duct Cyst

1

Michael E. Höllwarth

The median cervical cyst is a remnant of the thyroglossus duct, which runs from the pyramidal lobe of the thyroid gland to the foramen caecum in the dorsal part of the tongue. Embryologically, the thyroid diverticulum develops in a caudal direction from the foramen caecum after formation of the tongue. The thyroid gland descends to the neck in the same period of gestation when the hyoid bone develops from the second branchial arch. The thyroglossal duct may pass in front, behind, or through the body of the hyoid bone in the middle of the neck, and islands of thyroid tissue may be found scattered along the tract. At no time during embryogenesis does the thyroglossal duct contact the body surface; the original cysts thus never open to the skin. A fistula can only develop secondarily, such as following spontaneous perforation or surgical incision of an infected cyst.

Thyroglossal duct remnants are slightly more common than branchial cleft anomalies (55% *vs* 45%) and are the most common tumors of the anterior cervical region. They are usually located in the midline at the level of the hyoid bone or somewhat below it. Because of its connection with the foramen caecum of the tongue, the lesion typically moves upward with swallowing, like the thyroid gland, but unlike the thyroid, it also moves upward with tongue protrusion. In contrast, dermoid cysts or lymph nodes from the same location do not change their position with either act. In rare cases, the duct or cysts may contain either papillary cancer or squamous cell carcinoma. Thus, histologic evaluation is always indicated.

Ultrasound examination may be helpful, both to ascertain the presence of a normally situated, normal-size thyroid gland and to confirm the cystic nature of the mass under consideration. In cases of a suppurative infection, the appropriate treatment is incision and drainage in combination with antibiotics, followed by excision once the acute inflammation has settled (Figs. 1.1, 1.2, 1.3 and 1.4).

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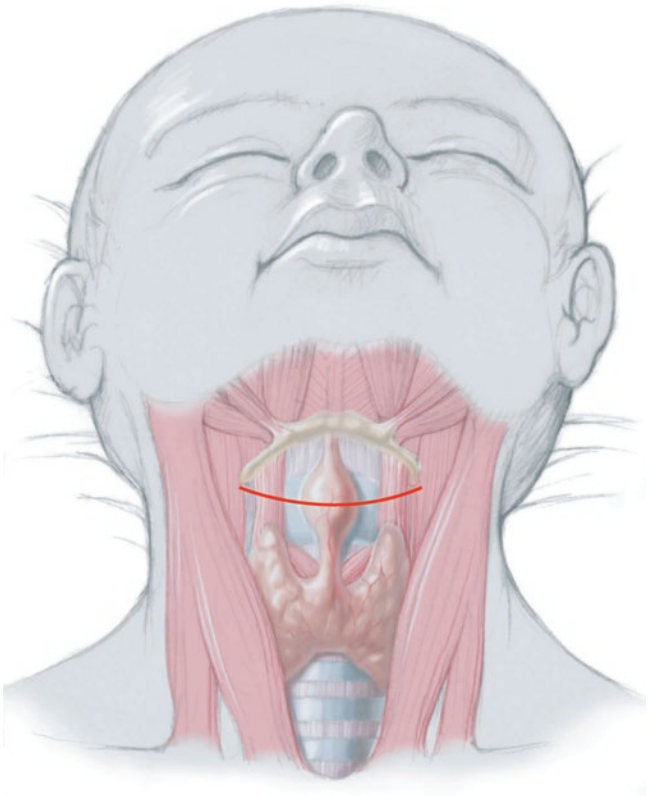

Fig. 1.1 Following induction of general anaesthesia with endotracheal intubation, the neck is hyperextended by placing a sandbag or towel roll beneath the shoulders. A horizontal skin incision is made over the cyst. In case of a fistula, the cutaneous orifice is circumcised in a horizontally oriented, elliptical fashion. Subcutaneous tissue, platysma, and cervical fascia are divided, exposing the capsule of the cyst. In cases with a previous history of inflammation, these layers may be fibrose and lack a clear demarcation from each other and from the cyst wall. The cyst is carefully separated from the surrounding tissue by blunt and sharp dissection

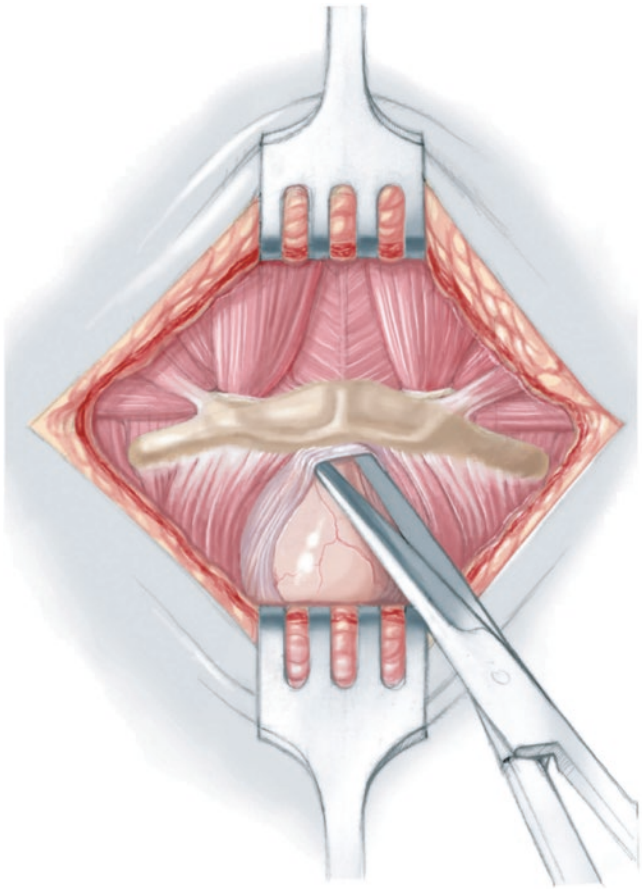

Fig. 1.2 The duct is attached to the cyst, running in a cephalic direction between the sternohyoid muscles to the body of the hyoid bone. It is usually not possible to recognize whether the duct perforates the hyoid body or passes across its anterior or posterior surface. The central part of the hyoid bone is freed from the muscles attached to its upper and lower margin. The thyrohyoid membrane is carefully dissected off the posterior aspect with scissors

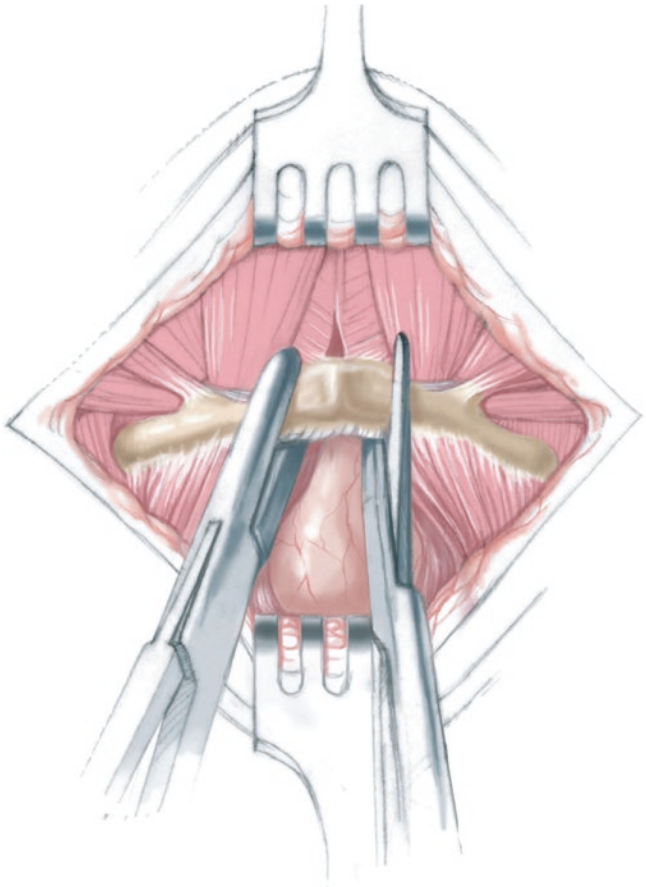

Fig. 1.3 The exposed hyoid bone is then stabilized with strong Kocher forceps on one side, clearly lateral to the median line, and the central segment is excised with strong Mayo scissors

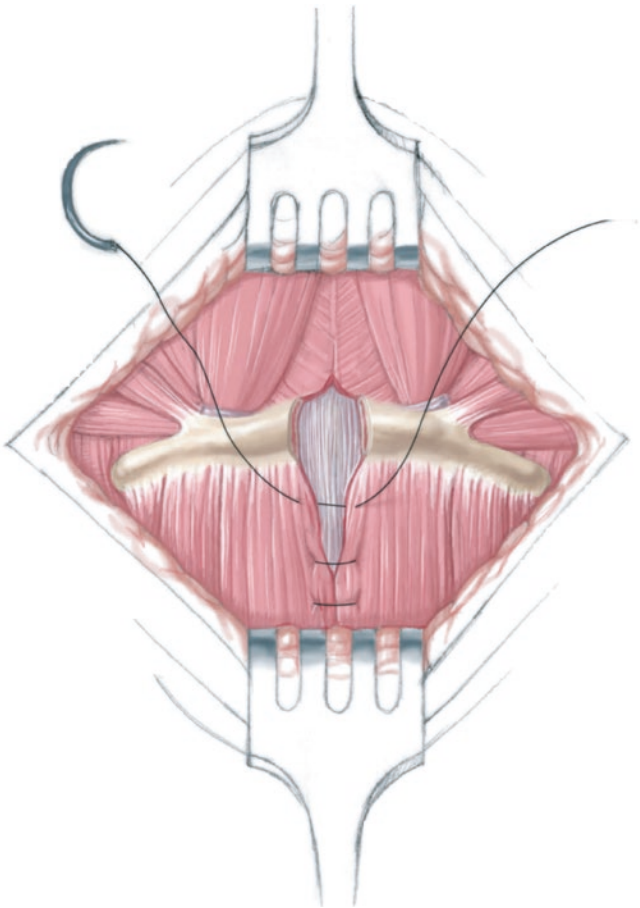

Fig. 1.4 If the duct extends beyond the posterior aspect of the hyoid bone, it is followed upward and divided close to the base of the tongue with a 5/0 absorbable transfixation ligature. If the floor of the mouth is entered accidentally, the mucosa of the tongue is closed with interrupted plain absorbable sutures. Often, however, no duct structures are found behind the hyoid bone, in which case some of the midline connective tissue is excised in the cranial direction to make sure that no duct epithelium is left behind. The lateral segments of the hyoid bone are left separated, but the anterior neck muscles are approximated in the midline with absorbable 4/0 sutures. Platysma and subcutaneous fat are closed with absorbable 5/0 sutures, and the skin is closed with either interrupted subcuticular absorbable 6/0 stitches or a continuous subcuticular nonabsorbable 4/0 suture, which can be removed 3–4 days later. A drain is usually not necessary, except in cases requiring extensive dissection, as may occur if the cyst is previously infected or recurrent

1.1 Results and Conclusions

Complete excision of the thyroglossal cyst consists of removal of the cyst, the entire tract, and the midportion of the hyoid bone through which the tract passes. If this principle is followed, recurrence is extremely unlikely, but if the central part of the hyoid bone is untouched, the cyst will recur. Though the procedure is easily performed in native tissue, dissection of a previously infected cyst is much more difficult, so postponement of the surgical procedure is not recommended once the diagnosis has been made.

Suggested Reading

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Branchial Cysts and Sinuses

2

Michael E. Höllwarth

During the fourth to eighth weeks of gestation, four pairs of branchial arches and their intervening clefts and pouches are formed. Congenital branchial cysts and sinuses are remnants of these embryonic structures that have failed to regress completely. Treatment of branchial remnants requires knowledge of the related embryology. The first arch, cleft, and pouch form the mandible, the maxillary process of the upper jaw, the external ear, parts of the Eustachian tube, and the tympanic cavity. Anomalies of the first branchial pouch are rare. Sinuses typically have their external orifice inferior to the ramus of the mandible. They may traverse the parotid gland, and run in close vicinity to the facial nerve in the external auditory canal. Cysts are located anterior or posterior to the ear or in the submandibular region. They must be distinguished from preauricular cysts and sinuses, which are ectodermal remnants from an aberrant development of the auditory tubercles, tend to be bilateral, and are localized anterior to the tragus of the ear. These sinuses are blind, ending in close vicinity of the external auditory meatus.

The most common branchial cysts and sinuses derive from the second branchial pouch, which forms the tonsillar fossa and the palatine tonsils. The external orifice of the sinus can be located anywhere along the middle to lower third of the anterior border of the sternocleidomastoid muscle. The sinus penetrates the platysma and runs parallel to the common carotid artery, crosses through its bifurcation, and most commonly exits internally in the posterior tonsillar fossa. A complete sinus may discharge clear saliva. A cyst, as a remnant of the second branchial pouch, presents as a soft mass deep to the upper third of the sternocleidomastoid muscle. The depth distinguishes it from cystic hygromas, which are located in the subcutaneous plane.

The third arch forms the inferior parathyroid glands and the thymus, whereas the fourth arch migrates less far down and develops into the superior parathyroid glands. Sinuses of the third arch open externally in the same region as those of the second one, but run upward behind the carotid artery to the piriform fossa. Cystic remnants may compress the trachea and cause stridor. Sinuses and cysts of the fourth branchial arch and cleft are extremely rare. Remnants of both the third and fourth arches most commonly present as inflammatory, lateral neck masses, more often on the left side. The cyst may evoke a false impression of acute thyroiditis. CT scans of the neck help to identify the origin of such lesions. In an acute suppurative phase, external pressure onto the mass may result in laryngoscopically visible evacuation of pus into the piriform fossa.

Cystic remnants present commonly in adolescence and adulthood, whereas sinuses and fistulas are usually seen in infancy and early childhood. In principle, regardless of the patient's age, clinical manifestation should be taken as an indication for elective excision before complications—mainly of an inflammatory nature—supervene (Figs. 2.1, 2.2, 2.3, 2.4 and 2.5).

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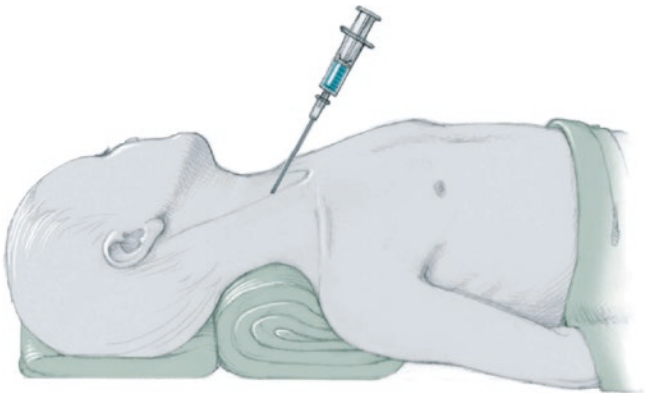

Fig. 2.1 For excision of the most common second branchial pouch remnant, the patient is placed in a supine position. Following induction of general anaesthesia with endotracheal intubation, the head is turned to the side. A sandbag is placed beneath the shoulders to expose the affected side. Instillation of methylene blue into the orifice aids identification of the sinus during dissection. Some surgeons introduce a lacrimal duct probe into the orifice to guide dissection of the tract

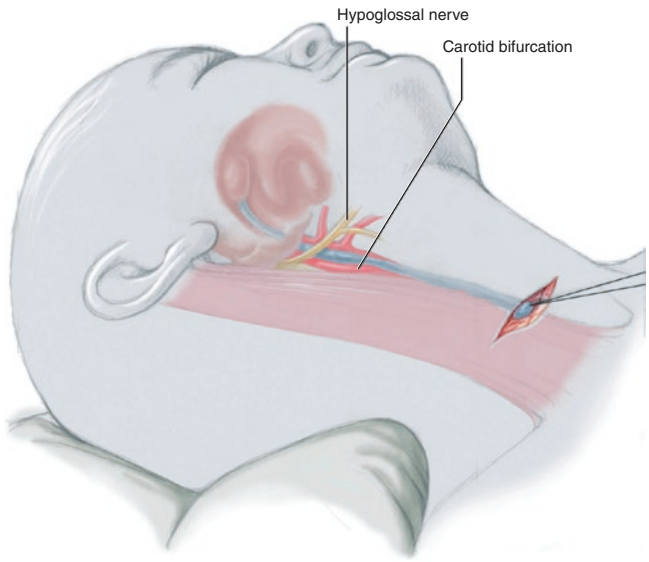

Fig. 2.2 In patients with a branchial cyst, the incision is made over the cyst along the Langer's lines. An elliptical incision is made around the sinus. A traction suture is applied to it just underneath the skin for manipulation during further dissection

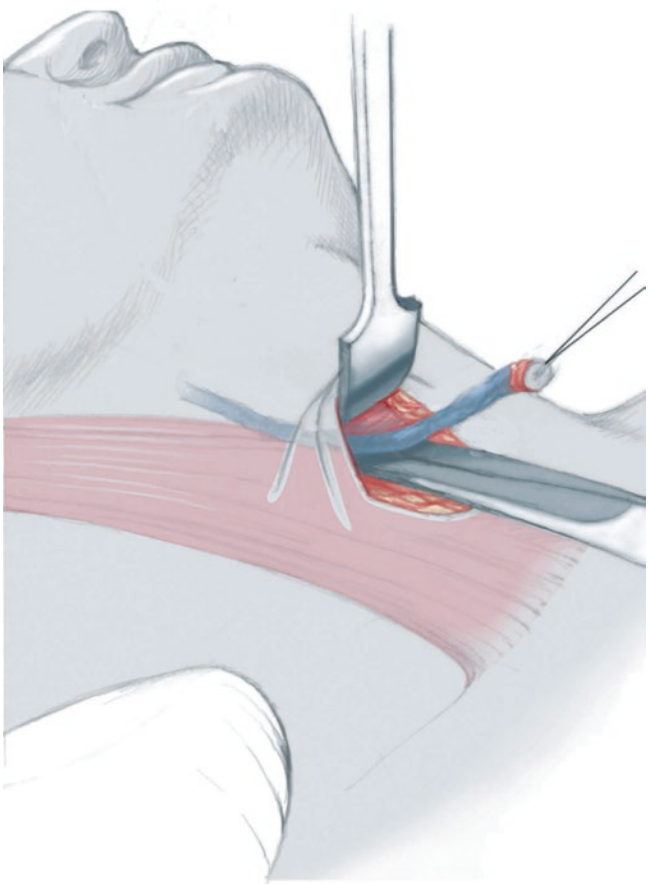

Fig. 2.3 Subcutaneous tissue and platysma are divided until the sinus tract is reached; the tract is easily palpable when the traction suture is gently tensed. Mobilization of the sinus continues in a cephalad direction as far as possible with gentle traction. The operation can usually be done through a single elliptical incision by keeping traction on the sinus tract; the anaesthetist places a gloved finger to push the tonsillar fossa downwards. Dissection then continues through the carotid bifurcation to the tonsillar fossa. Close contact with the sinus is obligatory to avoid any injury to the arteries or the hypoglossal nerve. Close to the tonsillar fossa, the sinus is ligated with a 5/0 absorbable transfixation suture and is divided

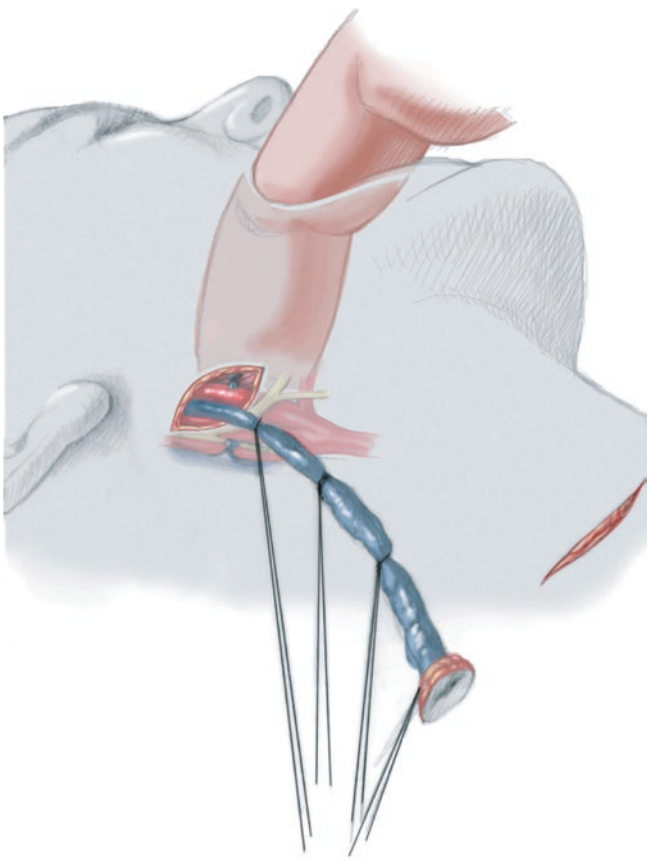

Fig. 2.4 In adolescents, a second transverse (stepladder) incision, made approximately 4–5 cm above the first, may be necessary to completely excise the sinus tract. Both incisions are closed with absorbable interrupted fine subcutaneous (5/0) and subcuticular (6/0) sutures

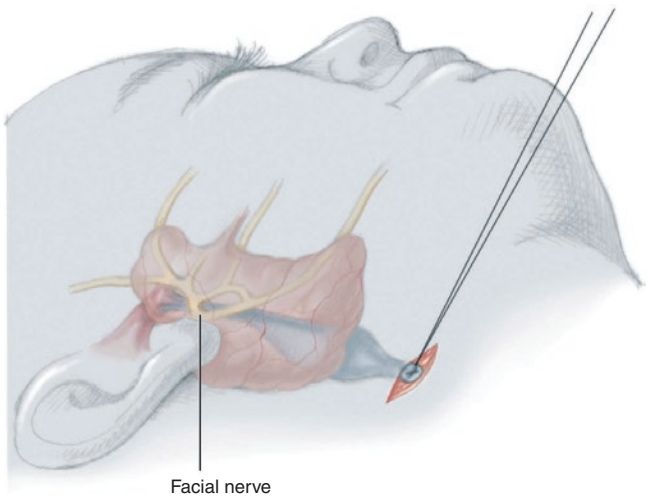

Fig. 2.5 For first branchial pouch remnants, the opening of the fistula is circumcised with an elliptical skin incision. Careful dissection liberates the subcutaneous layer of the embryological remnant, which is now transfixed with a stay suture. This suture is used for traction on the duct, which facilitates its identification on subsequent dissection into the depth towards the auditory canal. Because the tract is in intimate contact with the parotid gland and may be very close to the facial nerve, dissection must stay close to the tract, and electrocoagulation—exclusively bipolar—must be used sparingly. A neurosurgical nerve stimulator may be employed to identify and preserve fine nerve fibers. The opening of the fistula to the external ear canal should be included into the resection to avoid recurrence. The subcutaneous tissue is approximated using 5/0 absorbable sutures, followed by interrupted subcuticular absorbable 6/0 sutures

2.1 Results and Conclusions

Recurrences are most likely due to proliferation of residual epithelium from cysts or sinuses. The surgical procedure should thus be performed electively soon after diagnosis. Infected cysts and sinuses are treated with antibiotics until the inflammatory signs subside, unless abscess formation mandates incision and drainage. Repeated infections render identification of the tissue layers much more difficult. Surgery after infections of remnants of the first branchial pouch carries an increased risk of facial nerve injury. To avoid damage to vital vascular and nerve structures, it is important to keep dissection close to the sinus tract.

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Lymphatic Malformations

3

James K. Wall and Craig T. Albanese

Lymphatic malformations describe a broad range of lymphatic lesions, including common cystic lymphatic lesions, lymphangiomatoses, lymphangiectasias, and lymphedema. Cystic lymphatic malformations are congenital lesions that result in a complex of multiple cysts lined with lymphatic vascular endothelium. The incidence is estimated at 1 in 5000 live births. Lymphatic malformations have been reported in almost every type of tissue in the body with the notable exception of the central nervous system. Most are found in areas rich in lymphatic channels, including the head, neck, and axillary regions. Traditionally, vascular malformations involving the lymphatic vessels have been called by a variety of terms, including cystic hygroma, lymphangioma, or hemangio-lymphangioma. Given the benign nature of these anomalies and their wide-ranging manifestations, the preferred all-encompassing term is "lymphatic malformations."

3.1 Classification and Diagnosis

Cystic lymphatic malformations are the most common lymphatic malformation. They are benign and tend to enlarge slowly over time. They typically present as a soft, spongy, nontender mass noticeable in infancy. Rapid enlargement is associated with infection, hemorrhage, or trauma. Modern classification is focused on characterizing the cystic component as macrocystic, microcystic, or mixed, and the extent as superficial versus deep and localized versus diffuse. Both characteristics are clinically useful in determining management.

Diagnosis is made based on a combination of history, physical exam, and diagnostic imaging. The best imaging modalities for lymphatic malformations are ultrasonography, MRI, and contrast lymphography. Ultrasound is valuable in differentiating lymphatic malformations from other vascular malformations, which contain blood flow. Ultrasound is further able to characterizing the size and extent of superficial localized lesions. MRI offers an additional advantage of axial imaging, which can further identify the extent of large, diffuse lesions and highlight the precise relationship between malformations and adjacent anatomic structures. Lymphography with both conventional contrast and specialized MRI sequences has been described. These modalities are most helpful in looking for a source of lymphatic leak in cases of ongoing chylous output from cysts, effusions, or ascites.

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3.2 Treatment Goals and Options

Large lymphatic malformations involving the head and neck identified in utero have the potential to create airway obstruction upon delivery. Very large cervical lesions that deviate and partially occlude the fetal airway have been managed by elective ceasarean delivery and either intubation or resection while the fetus is still attached to the placenta, called the *ex utero, intrapartum treatment* (*EXIT*) procedure. Unlike head and neck teratomas, lymphatic malformations are typically soft and less likely to significantly compress surrounding structures. The prenatal tracheoesophageal displacement index may be useful in predicting the need for surgical airway management at birth.

The morbidity of lymphatic malformations after birth is associated with their type – macrocystic or microcystic. While the macrocystic lymphangiomas usually displace surrounding tissues, the microcystic forms growth within the nearby structures, especially within muscles. Thus the local extent and effects on surrounding anatomic structures is different. Macroglossia is commonly seen in microcystic lymphangiomas and can have profound effects on ventilation and feeding, the teeth and the upper and lower jawbones causing craniofacial disfigurement. Proptosis can result in permanent vision loss in up to 40% of cases. Over time, cystic lymphatic lesions can develop bleeding, infections, and cutaneous vesicles. Recurrent infections and chronic wounds are common with superficial lesions.

Goals of treatment for lymphatic malformations in childhood are to minimize symptoms and accomplish realistic cosmetic goals. These benefits must be weighed against the risks of disfigurement and disability.

Lymphatic malformations are amenable to multiple therapeutic modalities, including sclerotherapy, surgery, and medical therapy. The symptoms, location, extent, and characteristics of the lesion determine the best individual therapy or combination of therapies. Sclerotherapy is ideally suited for superficial macrocystic lesions, but deeper macrocystic lesions in the chest and abdomen have also been treated successfully with sclerotherapy. The preferred approach to sclerotherapy utilizes ultrasound guidance with aspiration followed by injection of a sclerosing agent. Multiple agents have been reported, including ethanol, doxycycline, bleomycin, OK-432, and sodium tetradecyl sulfate. There is no level I or II evidence to guide the use of specific agent(s), dwell times, or size criteria for sclerotherapy. Complications associated with sclerotherapy include nerve damage, systemic toxicity, and skin necrosis. Macrocystic lesions tend to spread along fascial planes and around neurovascular structures. Surgical resection is possible due to the fact that they growth circumscript and are well bordered, but injuries to surrounding structures and nerves, especially in the neck and supraclavicular region, must be carefully avoided. Sclerotherapy is recommended for small recurrent cysts.

3.3 Surgical Resection

Surgical resection is typically required for the treatment of microcystic lesions. These lesions can encase major structures, and great care must be taken to avoid significant vascular and nerve damage. Involvement of neck structures, jawbones, the mouth floor muscles and the tongue are very difficult to operate without mutilating local structures. The lesions can be well vascularized and transfusion may be required. Staged resections for large tumors have been advocated but bring additional risks associated with a scarred resection bed. Because this is not a malignant lesion, it is seldom necessary to sacrifice essential local structures.

Since microvascular lesions tend to infiltrate tissue planes, they are more likely to bleed, and have a high rate of recurrence. Any residual cystic tissue will increase the likelihood of recurrence. Recurrence is reported to range from 15 to 50% after surgical resection. Recurrence is associated with incomplete resection, often due to preservation of critical structures. Intraoperative rupture decreases the likelihood of complete resection, which averages 50%.

Complicated microvascular lymphangiomas in the head and neck region have limited surgical options. Recently, Sirolimus, a mammalian target of Rapamycin, has shown to be efficacious and well tolerated in children with these malformations.

3.3.1 Preoperative Planning

General anaesthesia is used and blood is made available if the lesion appears vascular on preoperative screening. If lesions are close to important motor nerves, one may use a nerve stimulator and interdict use of musculoskeletal blocking agents. Preoperative planning will usually demonstrate a safe plane of attack and may set expectations with regard to a complete excision or a debulking operation. Loupe magnification is often helpful, as is a bipolar cautery when working close to nerves or vital structures. A first-generation cephalosporin is used perioperatively.

3.3.2 Operative Procedure

The series of intraoperative surgical steps illustrated in Figs. 3.1, 3.2, 3.3, 3.4, 3.5, and 3.6 demonstrate the resection of a microcystic cervical lesion that is not amenable to sclerotherapy.

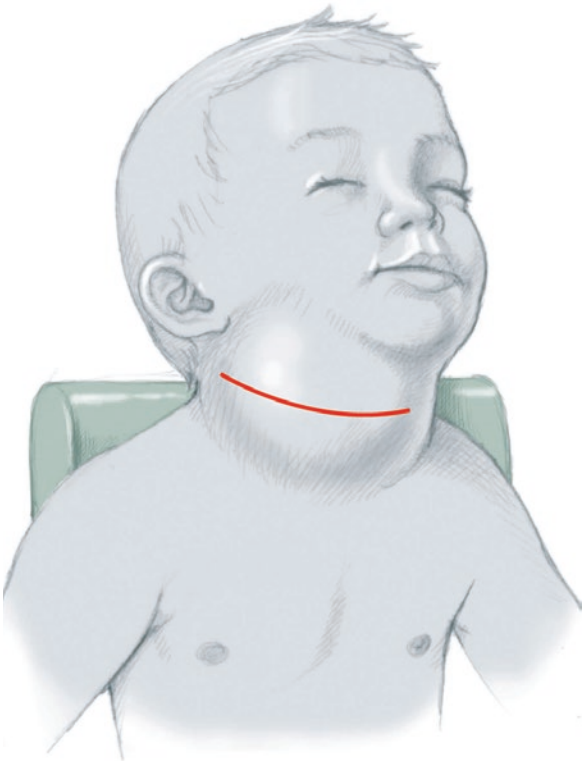

Fig. 3.1 For the most common (cervical) lesions, a transverse skin crease incision extending the length of the mass is placed in Langer's lines

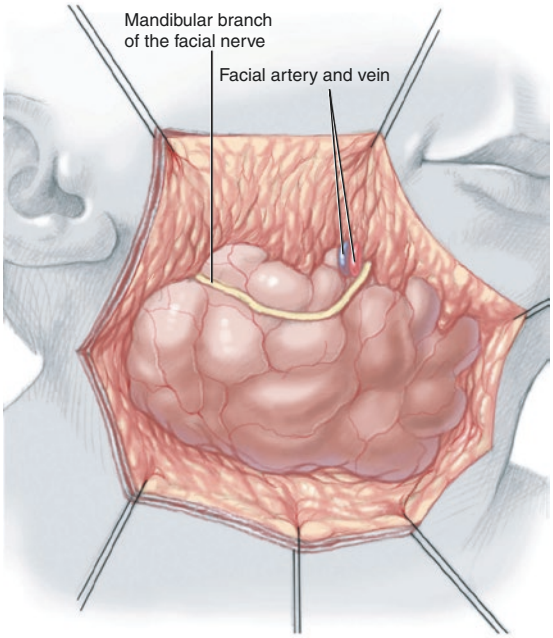

Fig. 3.3 Dissection of cervical lesions begins at the superior margin of the mass, near the ramus of the mandible. Upward reflection of the facial artery and vein allow the precise visualization necessary to preserve the marginal mandibular branch of the facial nerve. Bipolar cautery may be used and optical magnification is often helpful

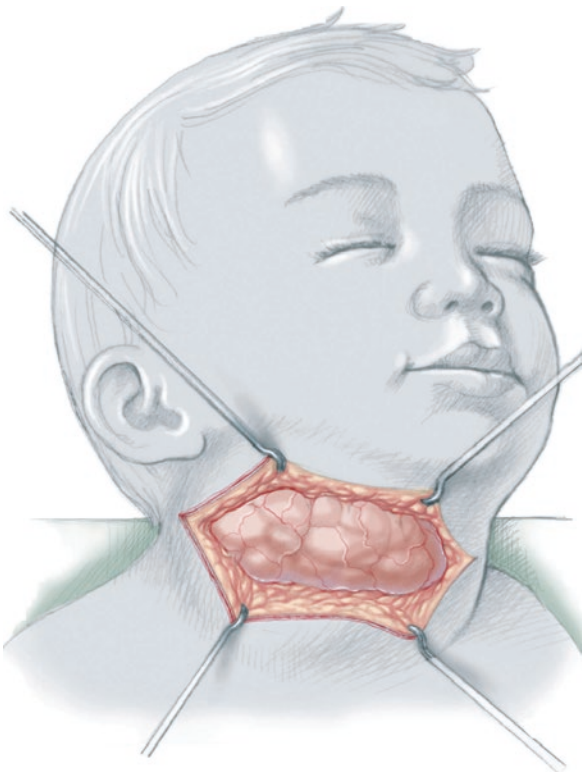

Fig. 3.2 If the lymphatic malformation demonstrates dermal infiltration, an ellipse of skin is removed. Otherwise, generous sub-platysmal skin flaps are raised. The external jugular vein and ansa cervicalis are not considered essential and may be sacrificed

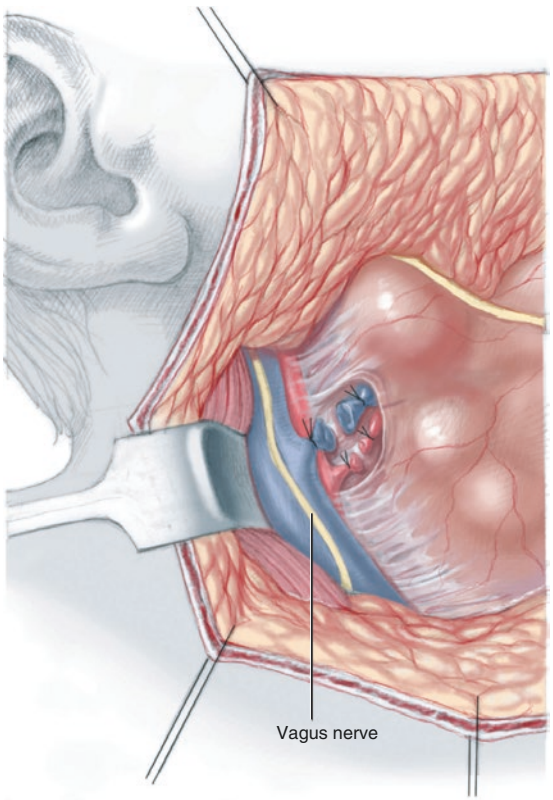

Fig. 3.4 The dissection proceeds medially, lifting the cyst from the surrounding alveolar tissue. It may be necessary to divide the middle thyroid vein and artery as the carotid sheath is approached. Deep dissection frequently involves the contents of the carotid sheath and sometimes the following nerves: vagus, spinal accessory, hypoglossal, sympathetic trunk, phrenic, and the brachial plexus

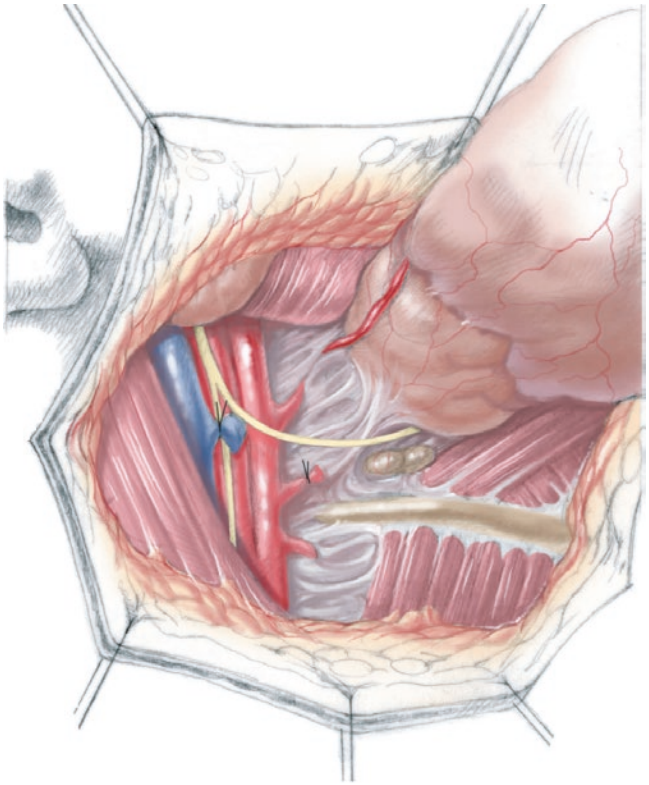

Fig. 3.5 Care is taken to preserve the hypoglossal nerve as it passes through the bifurcation of the carotid artery. The mass must then be freed from the hyoid bone and submandibular gland. It is rarely necessary to remove the submandibular gland en bloc with the mass, sacrificing the facial artery. The mass may be adherent to the brachial plexus in the floor of the anterior triangle or the spinal accessory nerve as it courses through the posterior triangle. Extension of the lymphatic malformation under the clavicle may lead to axillary or mediastinal involvement (requiring sternotomy if the lesion proceeds deeply). Combined masses may be delivered either above or below the clavicle

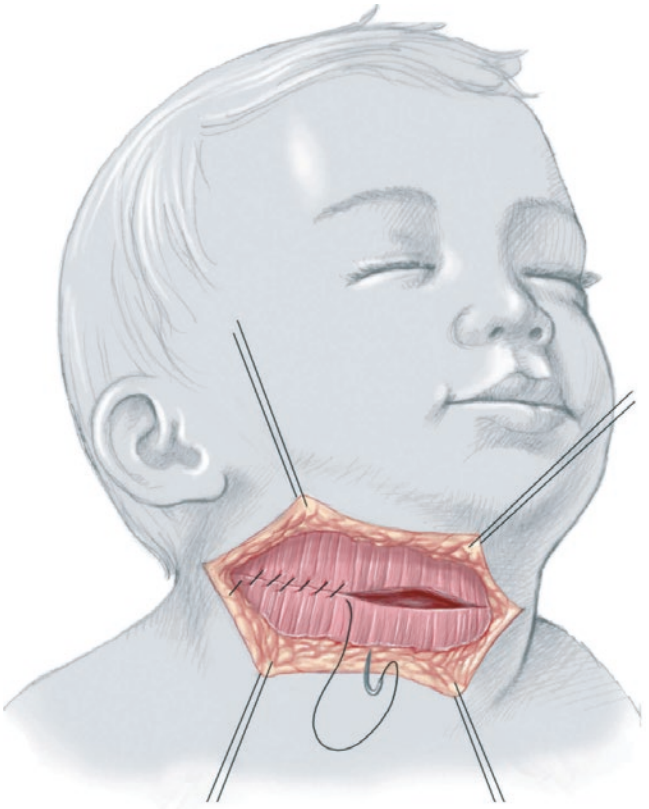

Fig. 3.6 The platysma is reapproximated with fine absorbable sutures, and the skin is closed with subcuticular sutures of similar material. Closed suction drainage is used for most lesions

3.4 Postoperative Care and Complications

Feeding resumes when the infant is awake and alert. Extensive intraoral dissection may temporarily impair swallowing and delay the onset of oral feeds. Drain removal may take days or weeks and is dictated by the daily drainage volume. Antibiotics are administered daily for 1 to 3 days.

In cases of partial resection, recurrence typically occurs within a year of surgery. Lymph leaks and nerve injuries are minimized by the use of bipolar diathermy. Rarely, lymph leaks may require re-exploration when drains are inadequate or are removed early.

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Tracheostomy

4

Andrew Dias, Sheila Hayes, Siobhan Fitzgerald, and John Russell

4.1 Indications and Anatomic Considerations

The indications for paediatric tracheostomies are divided into three main categories: upper airway obstruction, assisted ventilation, and pulmonary toilet. The most common indications have changed over the years, from upper airway obstruction secondary to infectious disorders to the current most common indication of prolonged ventilation. Paediatric tracheostomies place a significant amount of stress on the child and the carers, making social and verbal development more challenging, so alternatives to tracheostomy should be explored first. Currently, the indication to tracheotomise a child is generally ruled by the anticipation of long-term cardiopulmonary compromise or by the presence of a fixed upper airway obstruction that is unlikely to resolve for a significant period. The goal is an orderly, well-timed procedure with an experienced surgeon and the best personnel and equipment possible.

Basic anatomic differences from an adult neck should be borne in mind. The dome of the pleura extends into the neck and is thus vulnerable to injury. The trachea is pliable and can be difficult to palpate. It can be easily retracted to a great extent with little pull, and care must be taken to distinguish it from the carotid vessels. The paediatric neck is short, so the operative field is confined. Finally, the cricoid can be injured if it is not correctly identified.

4.2 Preoperative Planning

A key element of planning for tracheostomy is selection of the appropriate tube size. The tube should be small enough to allow the child to verbalise but not so small that insufflation leaks cause hypoventilation. Both diameter and length should be considered. Using a tube with too large a diameter may injure the tracheal mucosa by compromising its vascular supply. The tube should be long enough to allow adequate air entry with easy suctioning and clearance of secretions. A tube that is too short may result in accidental decannulation or formation of a false passage. If the tube is too long, the end may damage the carina or lie within the right main bronchus, thereby occluding the left bronchus. Paediatric tracheostomy tubes are cuffless except in larger children and adolescents. The diameter of the tracheostomy tube can be estimated on the basis of the size (corresponding to the inner diameter) of the child's endotracheal tube.

Most tracheostomies are performed under general anaesthesia (except in an absolute emergency), with an intubated patient. The patient's neck is extended with a shoulder roll, and the head is stabilized with a ring under the occiput.

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4.3 Operative Procedure

Figures 4.1, 4.2, 4.3, 4.4, 4.5, and 4.6 illustrate the steps in performing a successful paediatric tracheostomy.

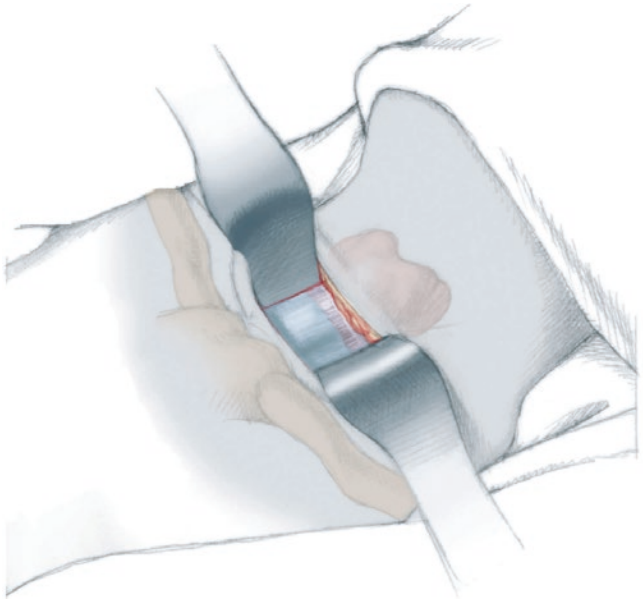

Fig. 4.1 The tracheostomy begins with a horizontal incision after local anaesthetic with adrenaline for infiltration is administered midway between the cricoid cartilage and the sternal notch. The incision is extended to the subcutaneous tissues and platysma muscle (superficial cervical fascia). Excess subcutaneous fat is removed with bipolar cautery. Right-angled retractors better expose the operative site. Two atraumatic forceps are used to grasp the investing layer of deep cervical fascia on either side of the midline, which is opened vertically. Next, the strap muscles enclosed in the muscular part of the pretracheal layer of deep cervical fascia are divided along the midline raphe. The visceral part of the pretracheal layer of deep cervical fascia is exposed and divided with bipolar cautery to expose the thyroid isthmus. The isthmus is either divided with bipolar cautery or, if possible, retracted superiorly. It is imperative to keep the operative field dry at all times to avoid complications

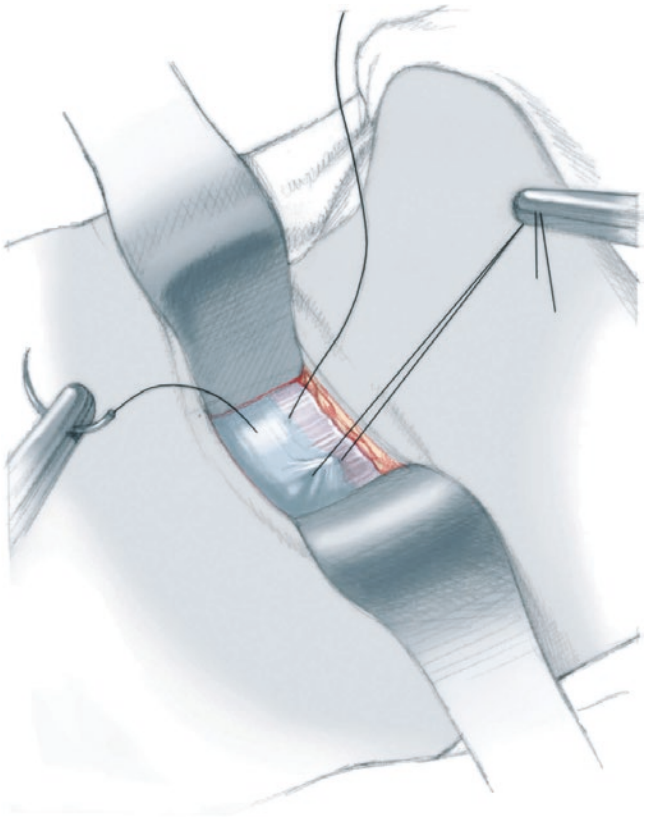

Fig. 4.2 The trachea is now exposed. The proposed tracheostomy cannula should be opened and its outer diameter visually checked against the exposed trachea. The pretracheal fascia is scored vertically with bipolar cautery to coagulate any vessels on the trachea in the midline. A suture of 4/0 nonabsorbable monofilament or its equivalent is placed on either side of the scored midline anterior trachea. Each suture incorporates one or two tracheal rings. These sutures are not tied to the tracheal wall but at their ends; they are left 6–8 cm in length. On completion of the case, these sutures are labelled and taped securely to the anterior chest wall, so they can be used to locate the tracheal incision in the event of tracheal cannula dislodgment. These sutures have the added benefit of holding open the edges of the tracheal incision for ease of placement of the tracheostomy cannula at operation

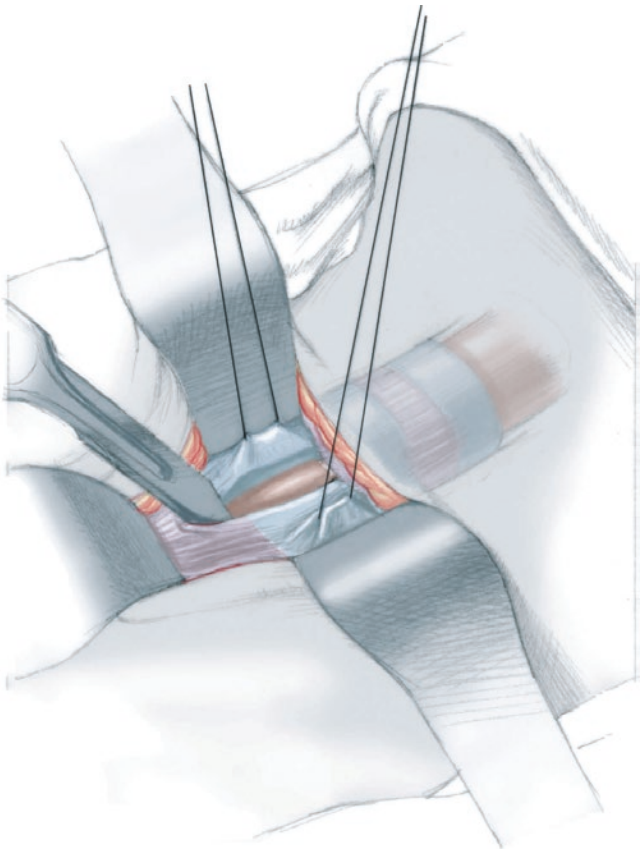

Fig. 4.3 The surgeon should request that the endotracheal tube be loosened and prepared for removal. The lateral retractors are removed and the trachea is elevated with the stay sutures. A number 15 blade is used to enter the trachea through a vertical incision along the score mark. Two or three tracheal rings (second and third or third and fourth) are divided. Suction should be available in case blood or secretions obscure the surgeon's view. The tip of the cannula to be inserted should be lubricated with a water-soluble surgical lubricant and positioned over the incision, poised for insertion when the endotracheal tube is withdrawn. The surgeon then asks the anaesthesiologist to withdraw the endotracheal tube just enough to clear the superior end of the lumen so that the tracheostomy cannula can be inserted and directed caudally towards the carina. If the tracheostomy cannula does not fit easily into the trachea, it should be removed and the endotracheal tube should be advanced beyond the tracheal incision so that ventilation is not compromised. This situation can occur if the diameter of the tracheal lumen has been overestimated. In this case, a smaller tube should be used. As soon as the cannula is in place, the obturator is removed and the anaesthesiologist should disconnect the ventilator hose from the endotracheal tube and reconnect it to the tracheostomy cannula. The anaesthesiologist should administer several deep breaths to the patient to confirm correct placement of the cannula, using carbon dioxide return and bilateral breath sounds to make sure that the patient can be ventilated correctly. Flexible endoscopy through the tracheostomy cannula confirms correct length of the tube away from the carina or bronchus

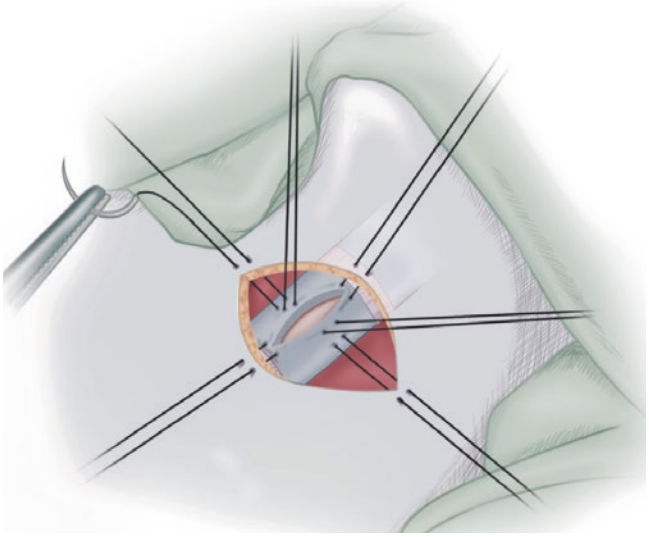

Fig. 4.4 The senior author favours a modified starplasty, a design based on the geometry of a three-dimensional Z-plasty. Horizontal skin flaps are sutured to the vertical tracheal incision with four sutures (superior, inferior, and one on either side of the tracheal incision) for further safety. Starplasty facilitates insertion of the tube and reduces the incidence of major complications, including pneumothorax, subcutaneous emphysema, infection of the stomal site, death from accidental decannulation, absence of stenosis, and anterior wall collapse. The drawback appears to be a higher incidence of persistent tracheocutaneous fistula following decannulation

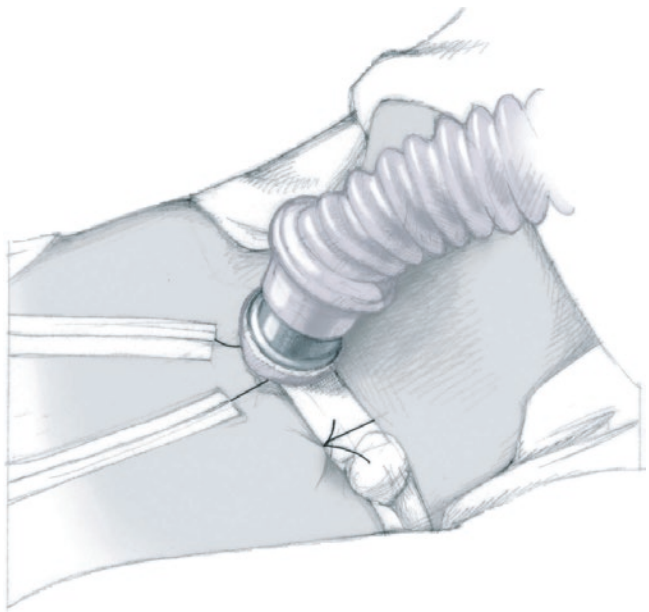

Fig. 4.5 The endotracheal tube can now be removed completely. The cervical wings of the body of the cannula are now secured to the patient. For each wing, two sutures with 3/0 nylon or its equivalent are used, one superior and one inferior. These are passed through the skin of the neck and the edge of the wing of the cannula. The two stay sutures are now labelled and taped to the anterior chest wall in such a fashion that their ends are readily accessible in case they are needed to reinsert the cannula in an emergency. Finally, the neck ties are passed through the two openings in the wings of the cannula and tied around the neck to further secure the tracheostomy cannula. Trachi-Dress (Kapitex Healthcare; Wetherby, West Yorkshire, UK). placed under the wings of the cannula with antibiotic ointment completes the procedure. Postoperative chest radiography is obtained to exclude any pneumothorax

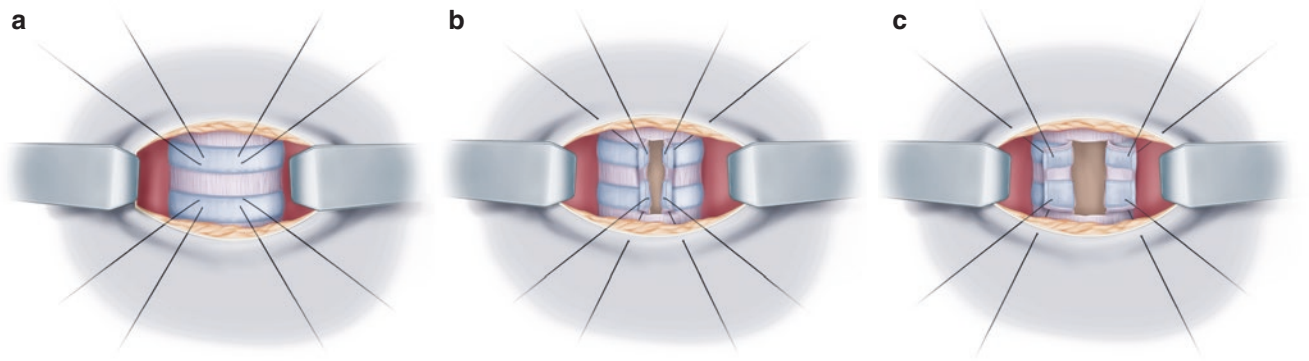

Fig. 4.6 (**a**–**c**) In small newborns and infants the tube may easily slip out of the trachea. It can be difficult to bring it back in place as fast as needed, and one might slip unnoticed along the anterior wall of the trachea into the ventral mediastinum. In order to prevent this complication and allowing an easy reintubation it is useful to prepare a doorflap of the ventral tracheal wall and to fix the two flaps to the lateral skin. Of course, when the tracheostomy is not needed anymore, the two flaps must be freed from the skin and sutured together to restore the ventral tracheal wall, usually an easy-to-perform manoeuvre

4.4 Postoperative Care

In our institution, all fresh tracheostomies are managed in the paediatric intensive care unit. This ensures that they are continuously monitored and observed, avoiding early potentially life-threatening complications such as accidental tube dislodgement or obstruction. Once the first tube change has occurred and a safe tract has been established, the stay sutures are removed and the child may transfer to a ward. Emergency equipment and supplies must accompany the child at all times. These should include a tracheostomy tube of the same size and a tube a size smaller, should the need for an emergency tube change occur. To keep the tracheostomy tube patent, suctioning is performed as clinically indicated. The suction catheter used is half the internal diameter of the tracheostomy tube; it is advanced to 0.5 cm past the end of the tracheostomy tube. The suction pressure recommended for infants and children is 80–120 mm Hg.

After a tracheostomy has been formed, there is a loss of filtering, warming, and humidification of inspired air. Lack of humidity may lead to thicker secretions and increased risk of mucus plugging, which can block the tracheostomy tube. In the initial postoperative period, humidification is replaced by the use of a heated humidity system. Children then progress to the use of a heat and moisture exchanger (HME). Normal saline nebulisers are administered to liquefy and loosen secretions. The tracheostomy tube is secured with cotton twill ties. Velcro tube holders are not recommended for children less than 7 years of age, and are used in older children only after a risk assessment. The tracheostomy stoma and skin under the ties are cleaned daily or as required, as mucus or exudate lying on the skin can contribute to skin breakdown. The area is observed for redness, excoriation, granulation tissue, and dryness. The frequency of a tube change may vary from weekly to monthly depending on the type of tube used. Tracheostomy tube changes are initially performed by healthcare staff, but parents or other caregivers can assume this responsibility after completing a tracheostomy training programme.

Children with a tracheostomy may have an altered ability to vocalise, as most of the airflow bypasses the upper airway. Early involvement of the speech and language therapist allows the use of aids appropriate to the child's needs and abilities to promote communication. If deemed suitable, a speaking valve can be introduced. The presence of a tracheostomy tube does not preclude oral feeding. An assessment of the child's swallow is performed by the speech and language therapist.

4.5 Complications

Early complications of paediatric tracheostomy include pneumomediastinum. In infants, the apex of the lung extends into the root of the neck. Dissection in the midline of the neck prevents violation of the pleural space. Identifying vascular anomalies preoperatively and dividing the thyroid isthmus with electrocautery avoids haemorrhage. Stay sutures and starplasty are ideal in preventing accidental decannulation and creation of a false passage anterior to the trachea during emergent reinsertion of the tracheostomy tube. Tube blockage is avoided by frequent suctioning and awareness of the problem. Subcutaneous emphysema can be avoided by using a tight-fitting tube in the trachea and making sure that the wound is not closed too tightly.

Late complications include local infection, excessive granulation, and difficult decannulation. Decannulation problems in children can be due to various factors. The lower respiratory tract must be free of infection and excessive secretions when decannulation is contemplated. Microlaryngoscopy and bronchoscopy must be performed to reveal any stomal granulations in the trachea or any suprastomal collapse obstructing the lumen, as well as to evaluate the mobility of the vocal cords. Psychological dependence on the tube is not common. Accidental decannulation becomes more of a risk as a young child develops the manual dexterity to remove the tracheostomy tube. Subglottic stenosis is generally caused by inserting the tube too high and too close to the cricoid. The incidence of persistent tracheocutaneous fistula is 20–40%. Finally, performing decannulation as early as possible improves the chances of normal speech and language development.

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Thyroidectomy in Children

5

Tom R. Kurzawinski and Paolo De Coppi

Thyroid surgery is an uncommon procedure in children and should be performed only in pediatric regional centres, with appropriate expertise and facilities to diagnose and treat thyroid problems. To best treat these children, a large, multidisciplinary team should discuss each case before the indication for surgery is made. This team should include a paediatric endocrinologist, a paediatric or general surgeon specialised in thyroid surgery and with experience in operating on children, a paediatric oncologist, a nuclear physician, a radiologist, a pathologist, and a clinical geneticist.

5.1 Indications for Surgery

Indications for thyroid surgery in children include both benign and malignant conditions:

Benign disease:

- Thyrotoxicosis (Graves' disease, toxic adenoma)
- Symptomatic multinodular goitre causing dysphagia, stridor, pain or discomfort

Malignant disease:

- Solitary nodules suspected of being malignant
- Thyroid cancer (papillary, follicular, medullary)
- Prophylactic thyroidectomy for multiple endocrine neoplasia type 2 (MEN 2)

5.2 Types of Operations

Similar to adult practice, partial and subtotal thyroidectomy has been abandoned in children. It has been observed that leaving thyroid tissue behind increases the risk of recurrences, and reoperations carry much higher risks of complications. Therefore, the following types of surgery are now recommended:

- **Lobectomy and isthmusectomy** is performed for solitary thyroid nodules that are either causing compression symptoms and/or thyrotoxicosis or are suspected of being malignant.
- **Near-total thyroidectomy** is the operation of choice in thyrotoxicosis and large, bilateral, multinodular goitre. In this case, almost all thyroid tissue is removed; a small fragment of thyroid with capsule (2–3 mm^3) is left to preserve attached parathyroid.
- **Total thyroidectomy** is indicated for treating thyroid cancer and as a prophylactic measure in children with MEN 2.

For thyroid cancer, lymph node surgery is also performed. Central lymphadenectomy aims to remove lymph nodes adjacent to the thyroid (level VI) and should be performed for tumours bigger than 1 cm. Unilateral or bilateral modified neck dissection with preservation of nonlymphatic structures is performed in children with thyroid cancer and palpable lymph nodes in the carotid chain.

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5.3 Diagnosis and Preoperative Planning

Examination of the neck is very important to assess the size of the goitre, its retrosternal extension, nodularity, and the presence of lymphadenopathy. Thyroid function tests (serum thyroid-stimulating hormone [TSH], estimated free T4 and/or total T4) are essential to confirm normal function. Serum thyroglobulin for well-differentiated thyroid cancers and calcitonin for medullary carcinoma/hyperplasia and thyroid antibodies sometimes can be helpful. Serum calcium and vitamin D levels should be measured and corrected if deficient. Calcitonin levels should be assessed to screen for medullary thyroid cancer, which accounts for 3–5% of paediatric thyroid cancers.

The thyroid can easily be imaged in children using neck ultrasound and chest x-ray. Ultrasound is invaluable in assessing thyroid nodules and lymph nodes (size, cystic or solid nature, number, blood flow, microcalcifications) and in guiding a biopsy needle. Ultrasound characteristics suggestive of malignancy include microcalcifications, indistinct margins, and a variable echotexture, but ultrasonographic appearance alone cannot reliably distinguish between benign and malignant lesions. Nuclear scans provide information about thyroid function and expression of specific receptors. Positron emission tomography (PET) could be helpful in managing malignant disease. CT or MRI scans are rarely necessary; they are almost exclusively confined to cases in which malignancy is suspected.

Fine-needle aspiration cytology is pivotal in assessing potentially malignant thyroid nodules or enlarged lymph nodes. Older children can tolerate it without anaesthesia. Solitary thyroid nodules in children carry a much higher risk of cancer than in the adult population, and negative cytology should not always obviate the need for surgical excision and full histological assessment if a high level of clinical suspicion exists.

Preoperative laryngoscopy is indicated to confirm the normal movement of the vocal cords, particularly in children who have had previous neck surgery.

Thyroid surgery carries significant potential morbidity, and consent for the operation should include clear information about risks such as bleeding (1%), infection (<1%), recurrent laryngeal nerve injury (1%), hypocalcaemia (2–5%), and poor cosmesis (keloid).

5.4 Operative Procedure

5.4.1 Positioning of the Patient

The child should be positioned supine on the operating table with a sandbag between the shoulders and the head ring. Extending the neck improves the access, and head up tilt reduces venous pressure.

5.4.2 Skin Incision

A 15-blade knife should be used to create a skin crease incision between the sternomastoid muscles. The incision should be placed between the lower one third and the upper two thirds of the distance between the chin and the manubrium of the sternum. This level should give adequate access to the whole thyroid gland, especially to the upper poles and vessels. The length of the incision depends on the size of the goitre; it can be extended during the procedure. Dissection of upper and lower flaps by developing a subplatysma plane should be performed using McIndoe scissors, avoiding damage to the anterior jugular veins and cutaneous nerves. The upper flap is raised to the level of the thyroid cartilage. Bipolar diathermy is used to control bleeding.

5.4.3 Thyroidectomy

Figures 5.1, 5.2, 5.3, 5.4, 5.5, 5.6, and 5.7 illustrate the steps in performing a successful paediatric thyroidectomy.

In children, drains are rarely necessary after thyroid surgery. A small Surgicel® patch (Ethicon, Somerville, NJ) can be placed in the thyroid bed. Before closing the wound, check hemostasis, viability of the parathyroids, and integrity of the nerve. Stop all bleeding points by a combination of bipolar diathermy and 5.0 PDS sutures. Closure of the wound starts with approximation of the strap muscles with absorbable (i.e., Vicryl®, Ethicon) stitches, leaving a small gap at the bottom that will allow blood to escape into superficial spaces to prevent compression in case of postoperative bleeding. Close the platysma and subcuticular layers with 4.0 Vicryl® sutures and skin with absorbable subcuticular 5.0 Monocryl® (Ethicon).

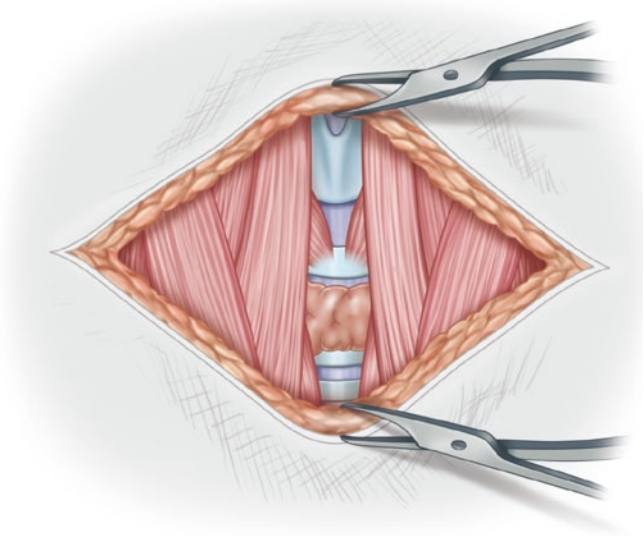

Fig. 5.1 The Joll's self-retaining retractor should be placed, and the bloodless midline plane between strap muscles (raphe) should be divided longitudinally. The strap muscles are separated from the thyroid and retracted laterally using artery clips. In younger children, the thymus could still be large and can extend from the mediastinum upwards to the neck. When necessary, the thymus should be mobilised and retracted downwards to allow safe dissection of the thyroid and surrounding structures

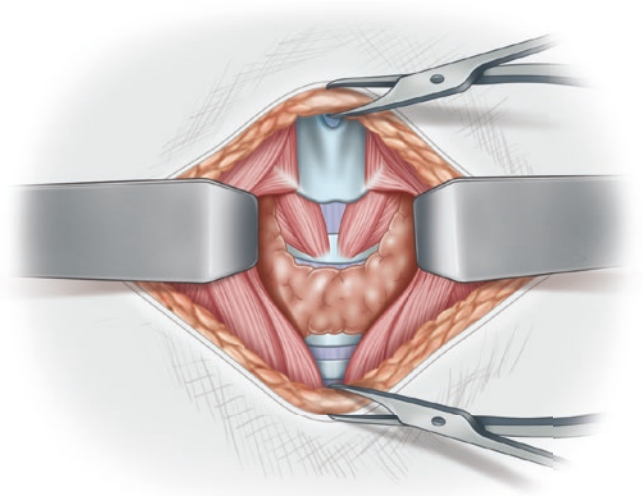

Fig. 5.2 The lead surgeon should stand opposite to the lobe that is to be dissected and should change position during the procedure if total or subtotal thyroidectomy is to be performed. The contralateral side of the thyroid should be pulled towards the surgeon and gently dissected laterally towards the carotid artery, which should be retracted gently by the assistant. Smaller vessels are divided with diathermy, and the middle thyroid vein is divided between ties, using 4/0 or 3/0 absorbable sutures

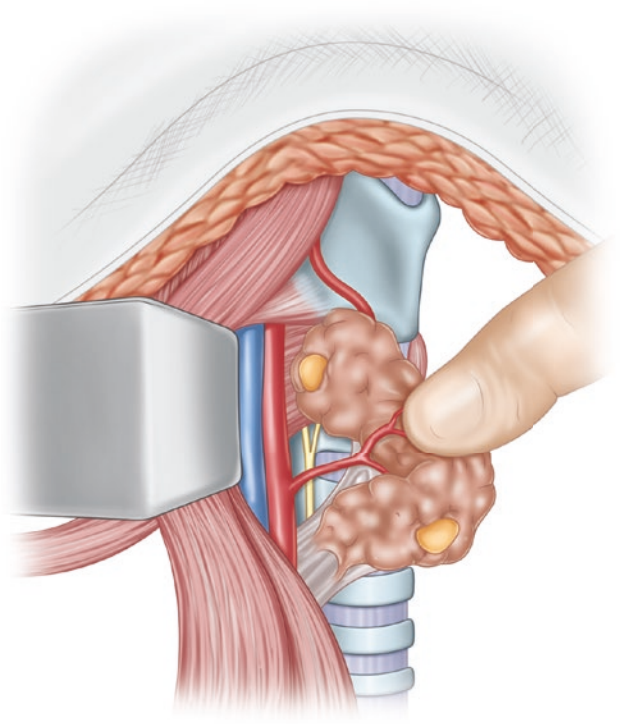

Fig. 5.3 If better access to the upper pole vessels is needed, the upper lobe of the sternothyroid muscle can be divided. The vessels should be carefully dissected and divided between ties, with careful identification of the external branch of the laryngeal nerve, which lies close and can be damaged

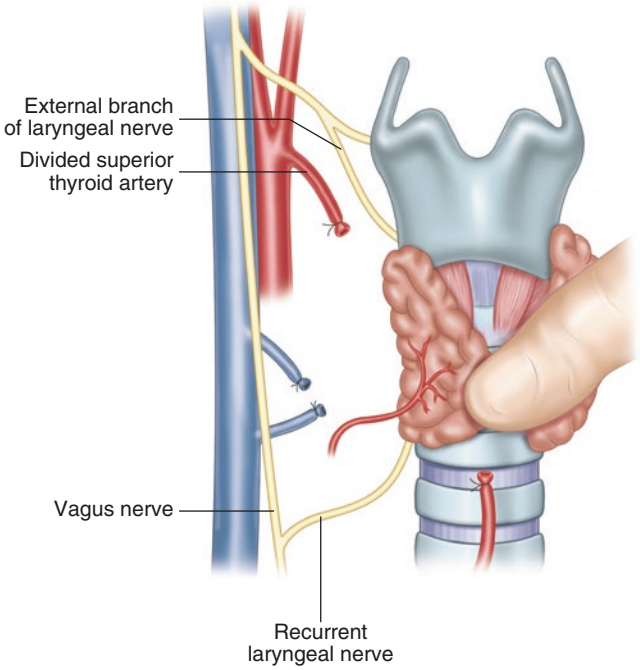

Fig. 5.4 The thyroid is pulled upwards and to the middle. The strap muscles and carotid artery are retracted laterally, identifying branches of the inferior thyroid artery and recurrent laryngeal nerve. This part of the operation should be performed gently, avoiding unnecessary stretching and diathermy close to the nerve. The recurrent laryngeal nerve usually lies in the tracheo-oesophageal groove, either behind, between, or in front of branches of the inferior thyroid artery. It is usually easy to recognise by its white colour, and it has vessels running on its surface. Remember that sometimes (17%), the nerve bifurcates early, before entering cricopharyngeal muscle, and it is important to identify this variant and preserve all branches. On the right side, the laryngeal nerve could be nonrecurrent, with a medial-to-lateral course rather than a vertical course

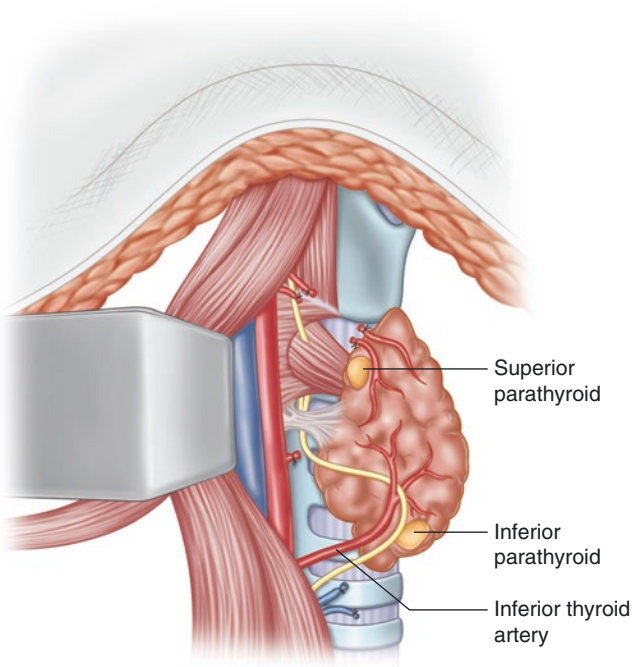

Fig. 5.5 Before proceeding with further dissection, it is of paramount importance to trace the whole length of the recurrent laryngeal nerve all the way behind the thyroid to its entry into the larynx behind the cricothyroid joint. Superior and inferior parathyroid glands are identified, and the small arterial branches of the inferior thyroid artery are divided between ties, preserving blood supply to the superior parathyroid gland. The blood vessels to the lower pole are divided, preserving the inferior parathyroid on its vascular pedicle. Anatomical positions of parathyroids are variable, but they should be always identified and dissected gently from thyroid capsule using bipolar diathermy. If the tiny blood vessels that supply the parathyroid glands are disrupted, their colour will change to dark blue or black, necessitating their auto-grafting. In this scenario, the parathyroid should be preserved in a gauze embedded in ice-cold saline until the thyroidectomy has been completed. Parathyroid should then be cut up into small pieces and implanted into small pockets in the sternomastoid muscle

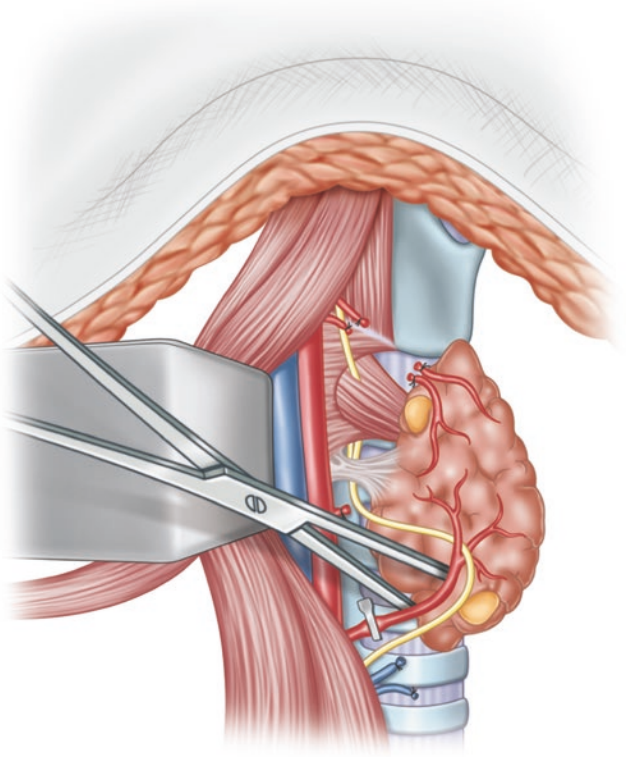

Fig. 5.6 Recurrent laryngeal nerve at its entry into the larynx is covered by fibres of lateral thyroid (Berry) ligament, which should be dissected by creating a tunnel parallel to the nerve with artery clips and divided between ties. Almost always, there is a small arterial vessel retracting behind the nerve, which can cause troublesome bleeding. The use of bipolar diathermy should be avoided at any cost during this part of the dissection

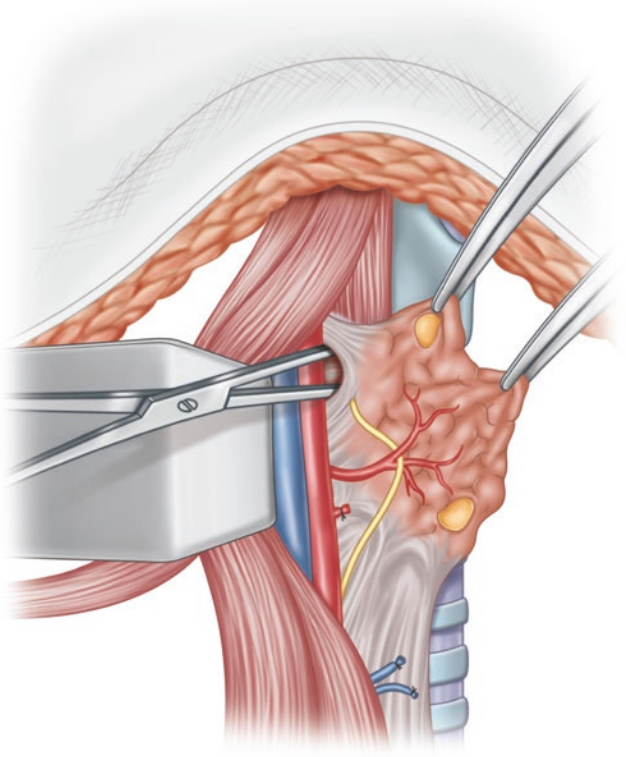

Fig. 5.7 Once the nerve is isolated and dissected from the gland, the thyroid lobe can be detached from the trachea with sharp dissection and diathermy. It is important to dissect and remove the pyramidal lobe. To perform hemithyroidectomy, divide the thyroid at the isthmus and oversew the capsule of the contralateral lobe using 3/0 absorbable sutures (i.e., Vicryl). If total thyroidectomy is planned, continue with the same procedure on the other side. The surgeon should move to the other side of the patient to maintain a good view of the operative field

5.5 Postoperative Care

5.5.1 Follow-Up and Surveillance

Immediately after surgery, children should be nursed in a semisitting position with routine observations (blood pressure, pulse, and respiratory rate) checked frequently. Neck swelling or excessive drain discharge must be reported urgently to the surgical team.

All children will require life-long surveillance after thyroid surgery, especially for thyroid cancer. Paediatric thyroid cancer is rare. (The incidence of differentiated thyroid cancer is 1 per 1,000,000 in children less than 10 years of age.) It is a treatable disease with an excellent prognosis. Treatment with radioactive iodine is recommended for most children after total thyroidectomy for papillary and follicular cancer. Radioiodine ablation should be carried about 4 weeks after surgery, with a diagnostic scan 6 months later. Regular review should include neck palpation, serum thyroglobulin, and ultrasound with cytology if necessary. In case of a cancer, follow-up ultrasound and TSH-suppressed thyroglobulin (Tg) level assessment is performed 6 months after initial therapy and at least annually thereafter, although it may be preferable to observe patients every 6 months for at least 5 years after diagnosis of more advanced initial or metastatic cancer. Assessment of free T4, T3, and TSH levels is indicated every 6 months, and 1–2 months after dosage changes. Calcitonin is a marker of recurrent disease in medullary cancer.

5.5.2 Management of Complications

5.5.2.1 Dyspnoea or Stridor

Dyspnoea can be potentially life-threatening. It may be due to recurrent laryngeal nerve injury or bleeding into the neck (~1%). Immediate intervention at the bedside may be required. If there is obvious neck swelling with stridor, the wound should be opened by cutting the skin and deeper stitches, the haematoma should be evacuated, and the patient should be transferred to the operating theatre for further management. Emergency intubation is difficult; if necessary, it should be performed by a senior anaesthetist.

5.5.2.2 Hypocalcaemia

Hypocalcaemia occurs as a result of parathyroid damage. Temporary hypocalcaemia after hemithyroidectomy is rare, but it is common if total thyroidectomy and lymph node surgery have been performed. Permanent hypocalcaemia occurs in 1–3% of these cases. Serum calcium levels should be checked every 12 h postoperatively, and symptomatic hypocalcaemia (tingling, cramps, tetany +ve Chvostek +ve Trousseau signs) should be treated urgently with IV calcium infusion (1.7 mL/kg calcium gluconate 10%). Prolonged hypocalcaemia requires treatment with oral calcium and vitamin D3.

5.5.2.3 Hypothyroidism

Thyroxine replacement is often necessary. Lobectomy causes hypothyroidism in about 10–20% of children. Maintaining normal thyroid function depends on the volume of remaining thyroid tissue and its functional capacity. The decision to start thyroxine should be based on symptoms and TSH levels during follow-up. After near-total thyroidectomy for benign disease, patients should start thyroxine before discharge from the hospital. The initial dose depends on the patient's age and weight and should be titrated to achieve a euthyroid state. In the past, after total thyroidectomy for cancer, the initial replacement was with short-acting liothyronine (T3), to reduce the time of withdrawal necessary to render the patient hypothyroid before treatment with radioactive iodine. This practice has been now replaced with continous replacement with thyroxine and two injections of Thyrogen. Once treatment with radioactive iodine is completed, lifelong thyroxine (T4) with an aim to suppress the TSH to undetectable levels is continued. TSH can stimulate tumour growth and its suppression is very important.

5.5.2.4 Voice Change

Voice change is caused by injury to the recurrent or superior laryngeal nerves and can be temporary (neuropraxia) or permanent. The risk of injuring a single recurrent laryngeal nerve, which causes a hoarse and croaky voice, is about 1%. Injury to both nerves is extremely rare and may require tracheostomy and laryngeal surgery. Injury to the external branch of the superior laryngeal nerve occurs in 2–5% of thyroidectomies and causes voice fatigue and weakness. Patients with abnormal-sounding voice after surgery should be reassured that these changes are usually temporary. Laryngoscopy should be performed if changes persist.

Suggested Reading

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Oesophageal Atresia

6

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Oesophageal atresia is defined as an interruption in the continuity of the oesophagus, with or without fistula to the trachea. The anomaly results from an insult occurring in the fourth week of gestation, the time when the separation of trachea and oesophagus by folding of the primitive foregut normally takes place. Familial cases affecting siblings or offspring suggest genetic factors. Most cases occur sporadically, however, without evidence for either hereditary or specific environmental teratogenic causes. The incidence is approximately 1:4500 live births, with a slight male preponderance (59%). Associated malformations are obvious or easily detected in 40–60% of cases and may be found in up to 80% by a meticulous search for structural and numerical anomalies in the skeletal system. At least 18 different syndromes have been reported in association with oesophageal atresia. The best known is probably the VATER or VACTERL association of anomalies (**V**ertebral-**A**nal-**C**ardiac-**T**racheal-**E**sophageal-**R**enal-**L**imb).

The earliest symptom of oesophageal atresia is polyhydramnios in the second half of pregnancy. Polyhydramnios is an nonspecific manifestation of swallowing disorders or of disturbance of fluid passage through the uppermost part of the intestinal tract of the fetus. Prenatal ultrasound may further reveal forward and backward shifting of fluid in the upper pouch, and in cases without a lower fistula, a paucity of fluid in the stomach and small intestine. Postnatal presentation is characterized by drooling of saliva and cyanotic attacks. If passage of a 12F feeding tube into the stomach is not possible, oesophageal atresia is almost certain. Immediate oro-oesophageal or naso-oesophageal insertion of a Replogle tube as soon as the diagnosis is established is mandatory to prevent continuous or intermittent aspiration of saliva. The baby should be nursed propped up in order to prevent aspiration of gastric contents into the tracheobronchial tree.

Prior to surgery, the type of atresia should be determined (Figs. 6.1–6.20). Air below the diaphragm on a plain x-ray film including neck, chest, and abdomen provides evidence of a commonly seen lower tracheo-oesophageal fistula. In most of these cases (type 3b/C or 3c/D), a primary anastomosis between the oesophageal segments is possible. In contrast, a gasless abdomen indicates that a pure oesophageal atresia without a lower fistula is present, and a long distance between the segments is to be expected (type 1/−, 2/A or 3a/B). A Replogle tube maximally advanced into the upper pouch helps to estimate its approximate length.

Additional malformations are looked for. Every neonate is checked for visible anomalies such as anal atresia or limb malformations. Thoraco-abdominal radiography may reveal duodenal or lower intestinal atresia, a diaphragmatic hernia, and/or skeletal anomalies. Ribs and vertebrae must be counted and carefully examined for deformations. Usage of contrast medium is rarely indicated. Cardiologic assessment, including echocardiography, forms part of the routine preoperative workup in order to recognize associated congenital cardiac abnormalities, which may influence anaesthetic management, and the presence of right-sided aortic arch, which is of importance for the surgeon. Abdominal ultrasound is performed routinely to search for urinary tract anomalies.

The baby is nursed in the intensive care unit (ICU). Immediate surgery is rarely required, so all the above-mentioned investigations can be performed step by step. Intubation and ventilation are necessary only in cases of respiratory distress, severe pneumonia, or severe associated malformations that demand respirator therapy. The endotracheal tube should be positioned beyond a distal tracheo-oesophageal fistula to avoid insufflation of gas into the stomach, inducing a risk of rupture, especially in babies with a high gastrointestinal atresia.

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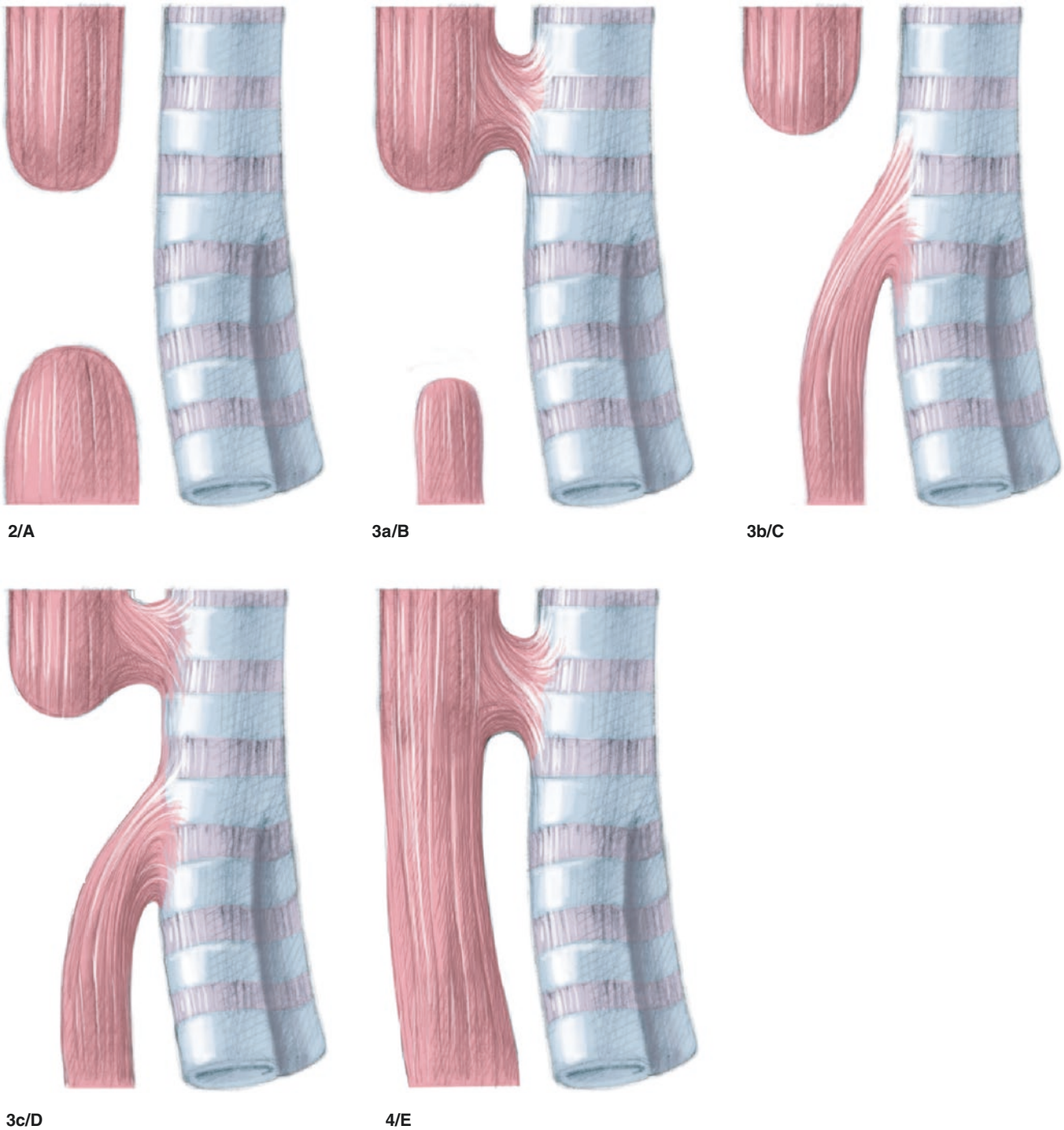


Fig. 6.1 (**a**–**e**) Classifications usually take their orientation from the concurrence and type of tracheo-oesophageal fistula. The commonly used systems are those described by Vogt (numbers ± lower case letters) and Gross (capital letters). The most frequent type of oesophageal atresia (3b according to Vogt, C by Gross) affects over 85% of the patients and consists of a blind-ending upper pouch with a fistula between trachea and lower oesophagus. Vogt's extremely rare type 1, characterized by a more or less total lack of the oesophagus, is not included in Gross's classification. Type 2/A (7%) corresponds to pure atresia without a fistula. The distance between the two segments is usually too long—the same as in type 3a/B (2%)—with a fistula to the upper oesophageal pouch. Patients with type 3c/D oesophageal atresia (3%) have an upper and a lower pouch fistula. Some authors classify an isolated tracheo-oesophageal fistula without atresia—an H-type fistula—as type 4/E (3%), although it belongs to a different spectrum because the oesophagus is patent. In Gross's classification, congenital oesophageal stenosis constitutes type F

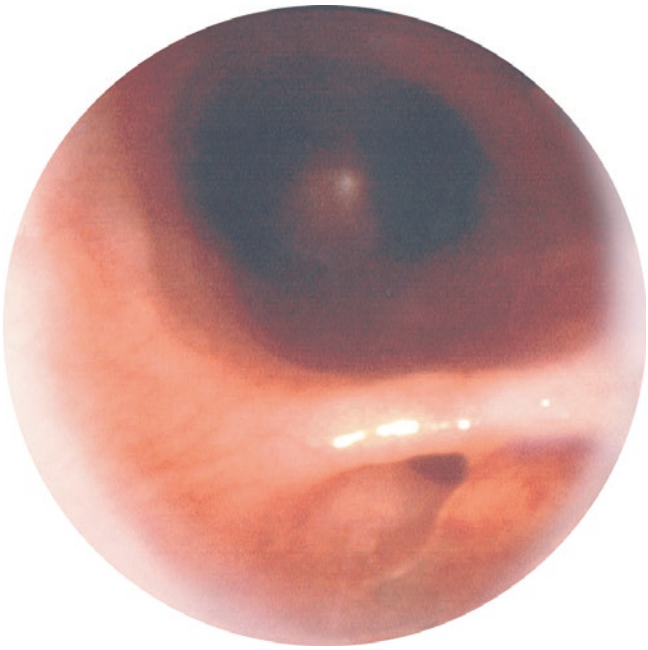


Fig. 6.2 Surgical repair is performed under general anaesthesia with endotracheal intubation. The endotracheal tube is advanced close to the tracheal bifurcation, and the infant is ventilated manually with rather low inspiration pressures and small tidal volumes. These measures serve to avoid overinflation of the stomach as well as to stabilize the trachea throughout the intervention. The Replogle tube is initially kept in place to easily identify the upper pouch intraoperatively. Broad-spectrum antibiotic prophylaxis is administered on induction. We routinely start with a tracheobronchoscopy using a rigid, 3.5-mm endoscope. The trachea and main bronchi are briefly inspected, and the the fistula to the oesophagus is localized, usually about 5–7 mm above the carina. Exceptionally, it may be found at the carina or even in the right main bronchus, indicating a short lower segment, and most likely a long oesophageal gap. The next step is to look for an upper fistula. The dorsal (membranous) region of the tracheal wall is inspected carefully up to the cricoid cartilage. Small upper fistulas are easily missed. To avoid this pitfall, irregularities of the dorsal wall are gently probed with the tip of a 3F ureteric catheter passed through the bronchoscope. If a fistula is present, the ureteric catheter will glide into it

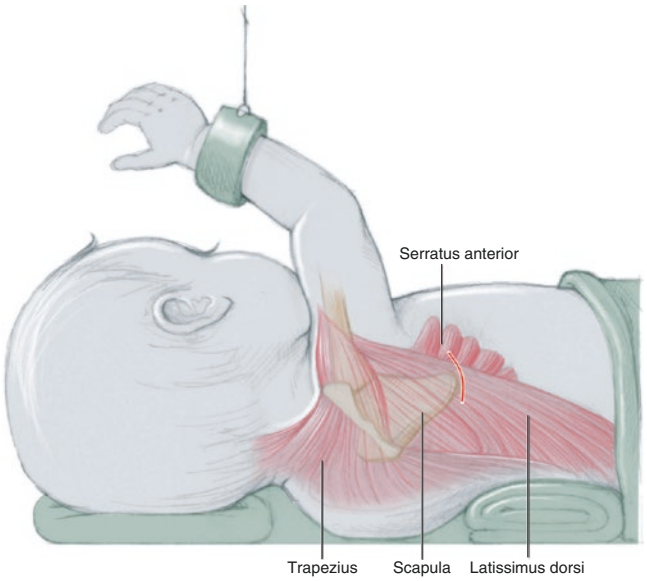


Fig. 6.3 The standard approach for repair of oesophageal atresia is a right laterodorsal thoracotomy. If a right aortic arch is diagnosed preoperatively, a left-sided thoracotomy is recommended. If an unsuspected right descending aorta is encountered during surgery, the procedure can be continued in most cases, however, establishing the anastomosis on the right of the aortic arch. The baby is positioned on the left side, stabilized with sandbags, and fixed to the table with adhesive bands. The right arm is abducted without undue tension. Mild anteversion helps to reduce the risk of traction injury to the brachial plexus. The elbow is flexed to 90°, and the forearm is best tied to a transverse bar mounted over the head of the child with soft slings. Care must be taken that no part of the body is submitted to pressure during the procedure. Exposed sites must be well padded. Soft pillars may be placed between the knees and beneath the feet, or the limbs can be wrapped with cotton wool, which also protects against heat loss. A folded towel under the left side of the chest improves exposure and facilitates access in particular to the deeper structures. A slightly curved skin incision is placed 1 cm below the tip of the scapula from the midaxillary line to the angle of the scapula. Some surgeons prefer a vertical skin incision in the midaxillary line for cosmetic reasons. A major advantage in neonates is the possibility of employing a muscle-sparing technique owing to their soft and mobile tissue layers. Only small flaps of skin and subcutaneous tissue are raised around the incision. The latissimus dorsi muscle is mobilized by cutting through the anterior fascial attachment. It is then lifted off the thoracic wall and retracted posteriorly, together with the thoracodorsal nerve, which runs on its deep surface following the posterior axillary line. When the latissimus muscle is retracted, the border of the serratus anterior muscle is mobilized along its origin from the tip of the scapula to the sixth rib and retracted up and forwards simultaneously with the scapula

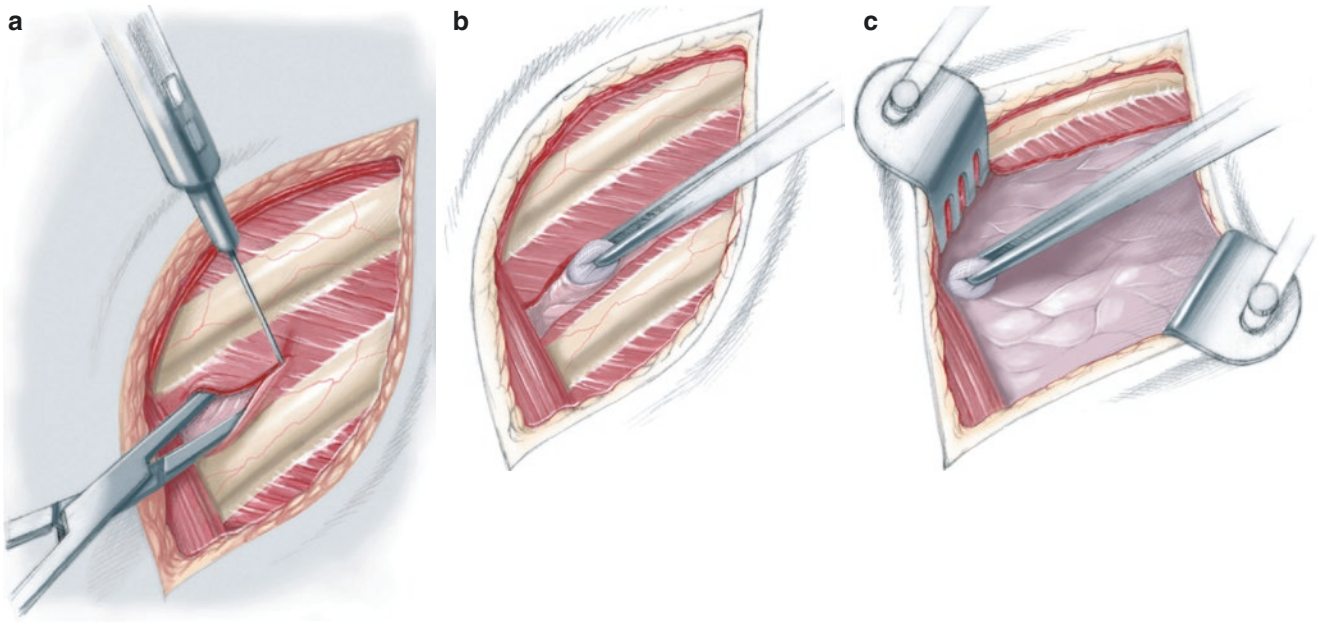
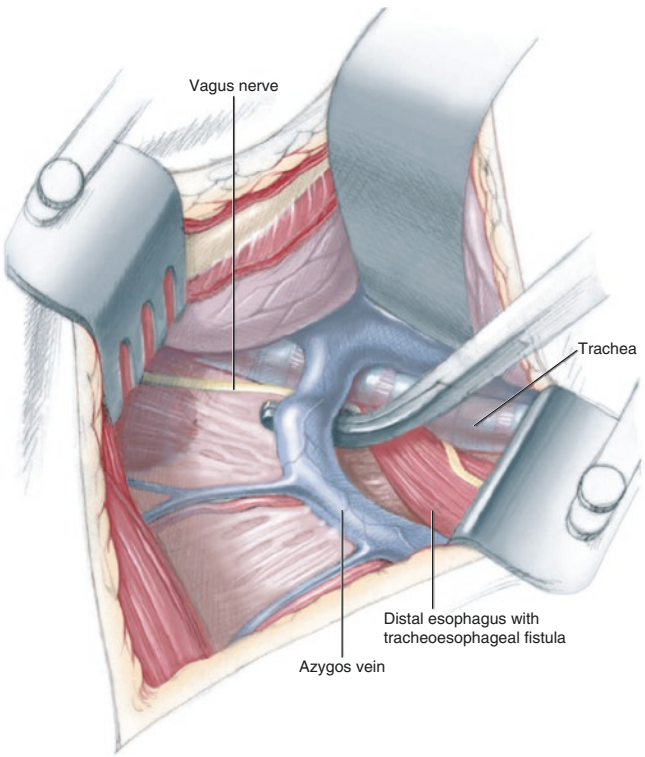


Fig. 6.4 (**a–c**) The intercostal muscles are divided along the upper border of the fifth rib. When the parietal pleura is exposed in one spot, a tiny, moist cotton swab mounted on an artery forceps is used to sweep it off the thoracic wall for an extrapleural approach. As soon as possible, a rib spreader is inserted and opened stepwise with care. For continuation of the pleural stripping towards the dorsal mediastinum, the use of two soft pledgets is recommended—one to hold the already reflected pleura under mild tension by pressing it towards the dorsal mediastinum, and the other to proceed with the dissection. An inadvertent tear in the pleura can be closed with a fine (6/0) monofilament absorbable suture

Fig. 6.5 The azygos vein is mobilized with right-angled forceps and divided between two ligatures (4/0 Vicryl). The right vagus nerve is identified, which runs along the lateral border to the upper pouch and accompanies the tracheo-oesophageal fistula toward the lower oesophagus, which is usually rather thin and hypoplastic. Extreme care must be taken to avoid any trauma to the delicate tissue. Handling and squeezing the oesophageal wall with forceps should be restricted to an absolute minimum, and the surgeon also should aim for preservation of all vagal fibres supplying the lower oesophagus. Denudation invariably entails a significant motility disorder and may cause severe gastro-oesophageal reflux



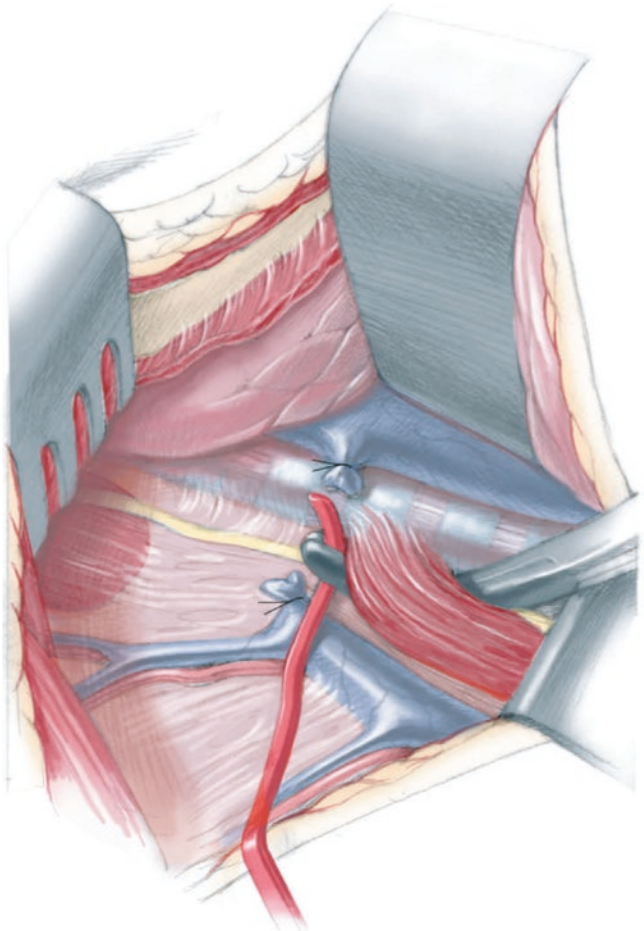


Fig. 6.6 Right-angled forceps are passed behind the distal oesophagus and a vascular sling is placed around it in order to pull it away from the trachea, facilitating the identification of the tracheo-oesophageal fistula, which is now freed from surrounding tissue

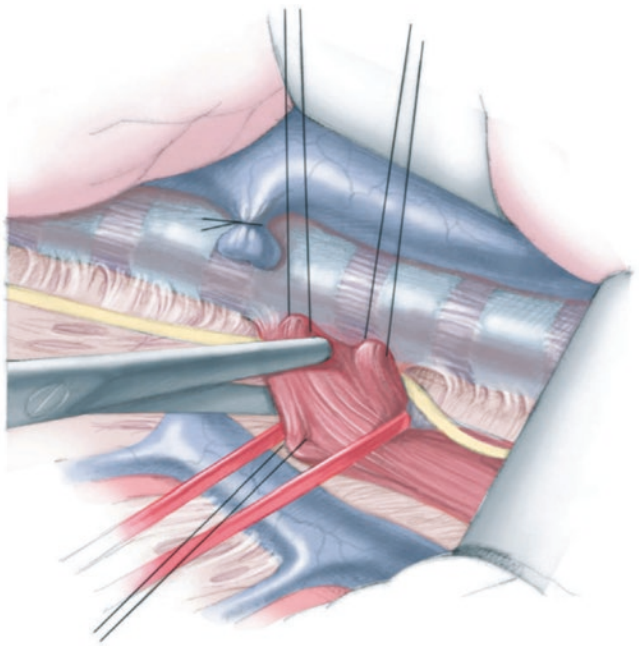


Fig. 6.7 Traction sutures are then placed at the tracheal and oesophageal ends of the fistula, and one additional stay suture nearby holds the lower oesophagus

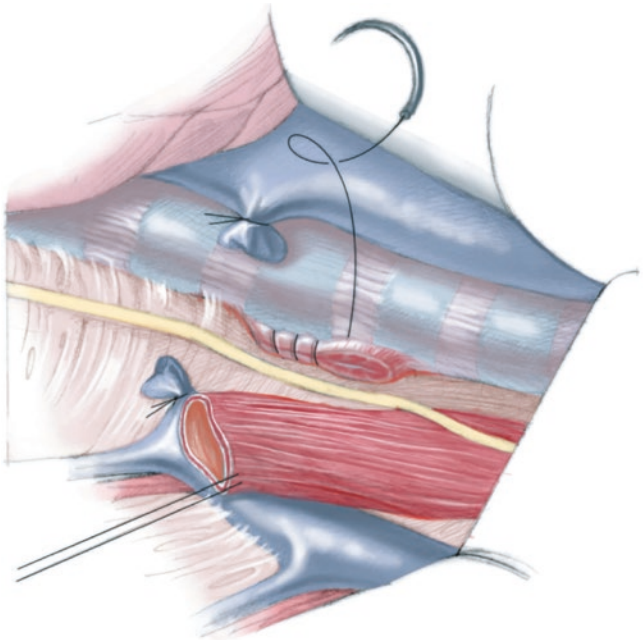


Fig. 6.8 At this stage, the fistula is divided and closed with a continuous absorbable monofilament 6/0 suture. Some authors prefer interrupted stitches. The level of division must be as close to the trachea as possible without risking a narrowing of the airway. Because most fistulas run obliquely upwards, a small residual pouch frequently remains in the trachea. The fistula closure is tested for an air leak by watching out for air bubbles during forceful ventilation after filling warm saline solution into the chest. At this stage, it is advisable to temporarily relieve the lung from the continuous retraction and achieve (through careful ventilation cycles) a full expansion of all collapsed areas

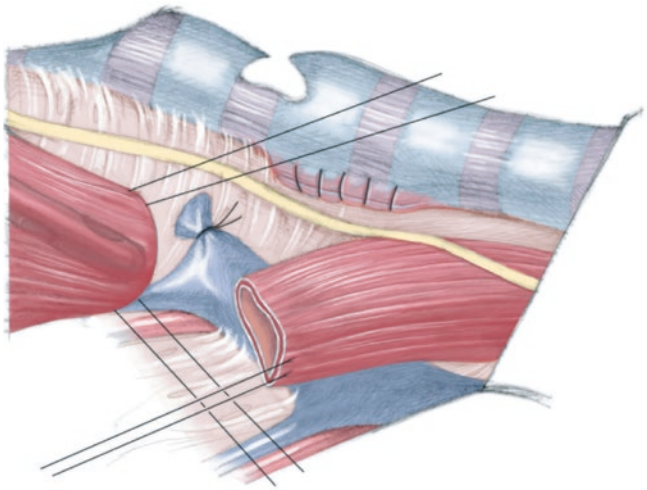


Fig. 6.9 The upper pouch is often retracted into the neck. Asking the anaesthetist to push on the Replogle tube serves to advance the upper pouch into the operative field. Traction sutures are placed on either side of the pouch to assist mobilization. Dissection of the oesophagus from the trachea is most challenging because they are adherent to each other by an intervening layer of firm connective tissue. Sharp scissor dissection is required, taking extreme care to avoid any accidental penetration into either organ. Anterior and lateral aspects of the upper pouch are easily freed using pledgets. If an upper fistula is encountered, it is transected close to the oesophagus and closed on both sides with interrupted monofilament absorbable 6/0 sutures. Unlike the lower oesophagus, the upper pouch has an excellent blood supply and can be dissected up to the thoracic inlet if necessary. Thus, if a large gap exists, further dissection of the upper oesophagus is preferable to extensive mobilization of the lower segment, which involves the risks of ischaemia and subsequent dysmotility. After the upper oesophageal pouch is mobilized, both segments are approximated to see whether an end-to-end anastomosis is possible

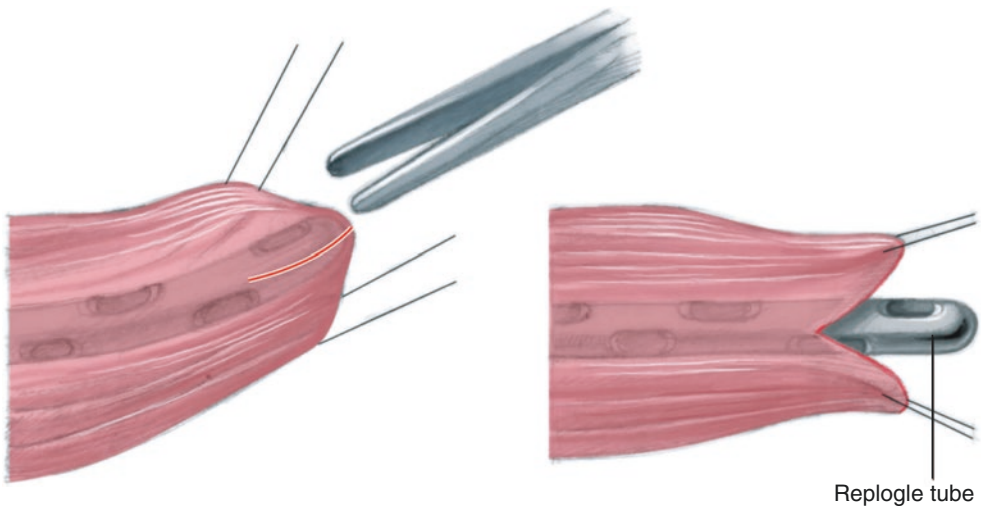


Fig. 6.10 Opening of the upper pouch for the anastomosis should be well centred at its lowermost point. This is best achieved by incising the pouch exactly over the tip of the fully advanced Replogle tube. An asymmetric opening results in an uncentred anastomosis, potentially leading to lateral preanastomotic out-pouching. The upper pouch is opened by a horizontal incision, which results in a fish-mouth–shaped aperture, adapted to the diameter of the lower oesophagus

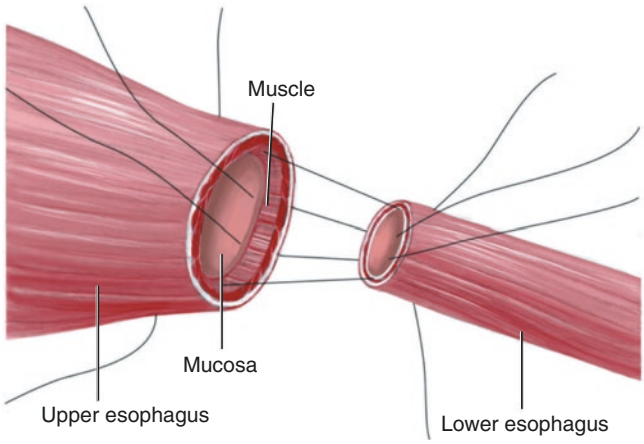


Fig. 6.11 The end-to-end anastomosis is fashioned with interrupted absorbable 6/0 sutures. The first two stitches are placed on either side. The posterior wall needs two or three additional sutures. Meticulous care must be given to take sufficiently large "bites" of both muscular tissue and the mucosal layer, which tends to retract upwards as soon as the upper pouch is opened. Once all posterior wall sutures are placed, the oesophageal segments are gently pulled together, and the sutures are tied on the mucosal surface. Thereafter, a 5F Silastic feeding tube—the connection hub of which has been cut off—is sutured with the cut end to the tip of the Replogle tube, which is then withdrawn by the anaesthetist until the feeding tube appears outside the mouth. The distal end of the feeding tube is passed into the stomach. The tube serves for postoperative gastrointestinal decompression and early feeding, and also functions as a transanastomotic splint for drainage of saliva

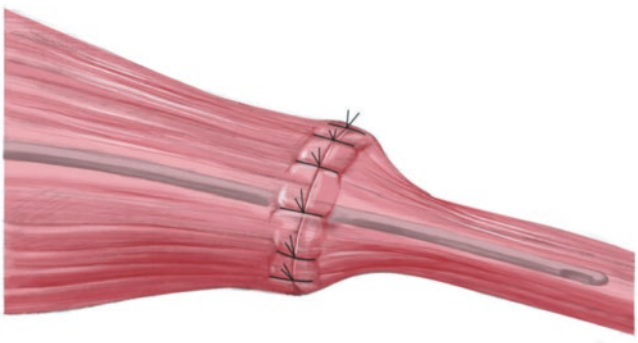


Fig. 6.12 The anterior aspect of the anastomosis is completed in a similar way as described above, with three or four stitches, this time tying the knots on the outside of the oesophageal wall. Thoracoscopic esophageal atresia repair has recently been performed in some very experienced centres. This procedure certainly requires great experience in endoscopic surgery, and late results comparing endoscopic with thoracoscopic procedures are not yet available

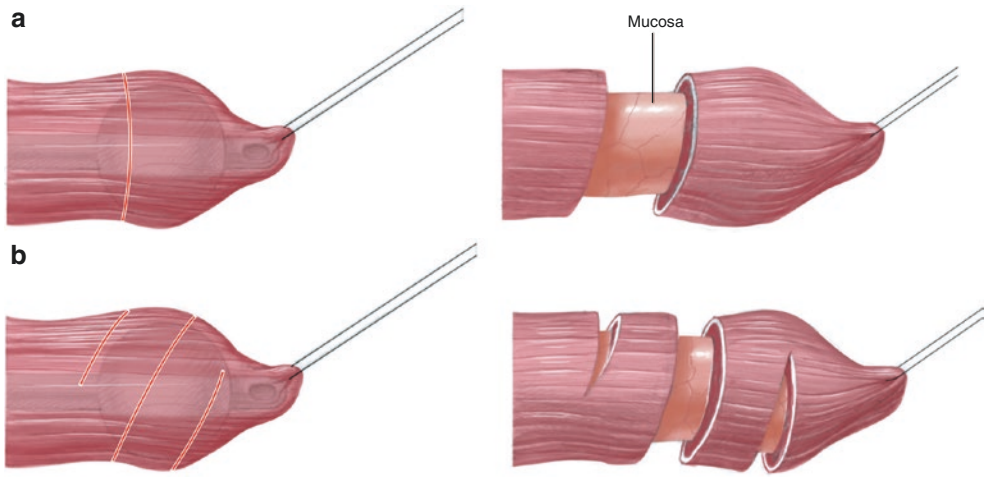


Fig. 6.13 (**a, b**) The goal of a tension-free end-to-end anastomosis can be achieved with this technique in most cases of oesophageal atresia with a distal fistula. If the tension appears to be too much despite mobilization of the upper pouch up to the thoracic inlet, further length may be gained with a circular myotomy in the upper pouch (Livaditis et al. 1972). This is achieved by introduction of a 8F balloon catheter into the upper pouch transorally, which is transfixed at the lower end of the pouch with a 4/0 monofilament traction suture; the balloon is blown up until it fills the pouch. The muscle layer is then divided above the balloon approximately 1 cm cranial to the future anastomotic line, in either a circular or a spiral fashion. The mucosal layer of the upper pouch is rather thick, so mucosal tears usually can be avoided with careful dissection. The upper pouch can be lengthened by 5–10 mm by this method, which may suffice to create an anastomosis without undue tension. Development of a pseudodiverticulum after circular myotomy (out-pouching of the mucosa through the established gap in the muscle layer) has been described

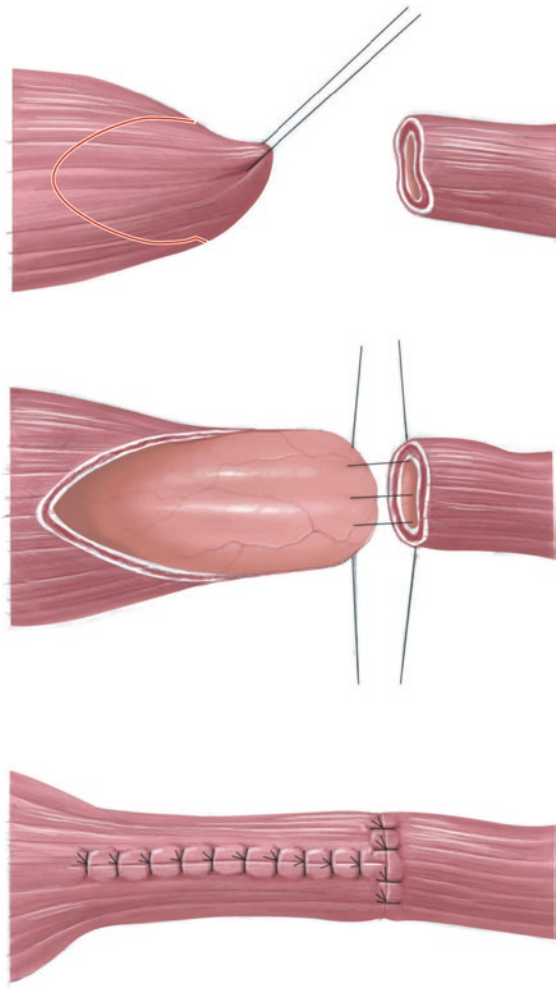


Fig. 6.14 Another way to reduce inappropriate tension on the anastomosis is to fashion a mucosal-muscular flap from a larger upper oesophagus. A right-angled incision is made in one half of the upper pouch. The flap thus created is turned by 90° so that the vertical cut surface faces downwards. It is then rolled into a tube. However, the gain in length results in a reduction in diameter

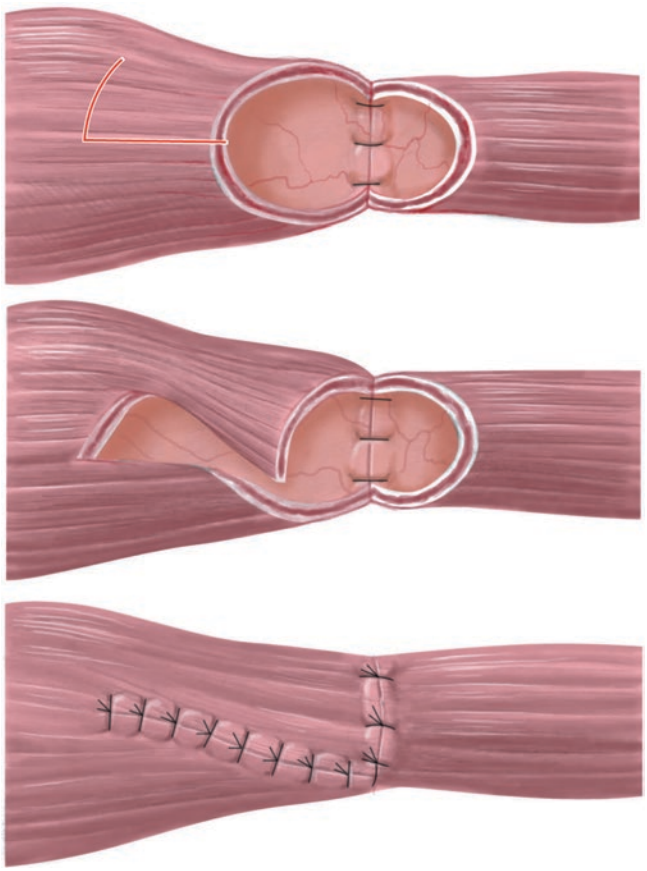


Fig. 6.15 If a satisfactory dorsal wall anastomosis can be established but undue tension arises in the anterior half, a right-angled flap in the corresponding part of the upper pouch without tubularization may bridge the gap and result in a safe anastomosis. The thoracic cavity is irrigated with normal saline. A soft drain is introduced via a separate intercostal stab incision, and the tip is placed near the anastomosis. Before closure, the lungs are fully expanded by forced ventilation until all collapsed regions are well aerated again. Most authors prefer to approximate the ribs with two or three pericostal sutures, but the ribs may be left as they are after the procedure, and they will reapproximate spontaneously within 1–2 weeks. This strategy may avoid the typical costal synostosis after suturing the ribs together. The latissimus dorsi and serratus anterior muscles are allowed to fall back into their original positions and are sutured to their fascial insertion sites with one or two 3/0 absorbable sutures each. The subcutaneous fat is readapted with 5/0 absorbable sutures, including the corium. This technique approximates the skin perfectly in most cases, so that separate skin sutures are not necessary. The incision is simply approximated with adhesive strips. If the wound margin adaptation is unsatisfactory, a continuous subcuticular monofilament 5/0 suture is applied, which is pulled out after a few days

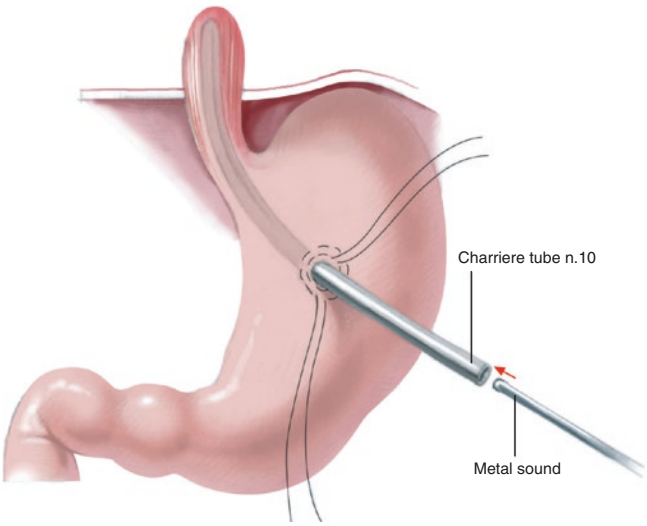


Fig. 6.16 An airless abdomen on thoraco-abdominal x-ray leads to suspicions of oesophageal atresia without a lower fistula (10%). A primary end-to-end anastomosis is not possible in these cases, owing to the long distance between the oesophageal pouches. The decision is now either to plan an oesophageal replacement or to try to preserve the patient. In any case, a primary gastrostomy is essential for enteral feeding in all patients with long-gap oesophageal atresia. It is also used for estimation of the length of the gap. Regarding the preservation of the patients own oesophagus, there are three strategies possible. The often-used first strategy is to await spontaneous growth, which is more pronounced in the upper stump. Experience tells us that it takes 8–12 weeks (on the average) until a safe anastomosis is feasible. Secondly, one can attempt to promote elongation of the upper oesophageal segment by regular longitudinal stretching. Approximation may be further accelerated by additional bougienage of the lower pouch. The latter is our preferred method, permitting anastomosis of the two segments after 3–5 weeks. If mechanical elongation of the lower pouch is planned, the gap is assessed in the following way: a 8F or 10F feeding tube is cut approximately 10–13 cm from its distal end, and a 70° curved metal sound is introduced into the feeding tube up to its tip. This assembly is passed into the lower oesophageal pouch via the stomach. At the same time, the anesthetist introduces a radio-opaque device into the upper pouch. Both probes are pushed toward each other under fluoroscopic control, and the distance between the maximally approximated oesophageal stumps is gauged. Usually it corresponds to four or more vertebral bodies. The feeding tube with the metal probe is kept in the stomach for the stretching procedures, and longitudinal stretching of both oesophageal stumps is performed twice daily for 3–5 min under mild sedation. Gentle pressure is used in the lower pouch, with more forceful pressure in the upper pouch. Leaving the manoeuvre in the same experienced hands throughout has saved us from ever causing a perforation

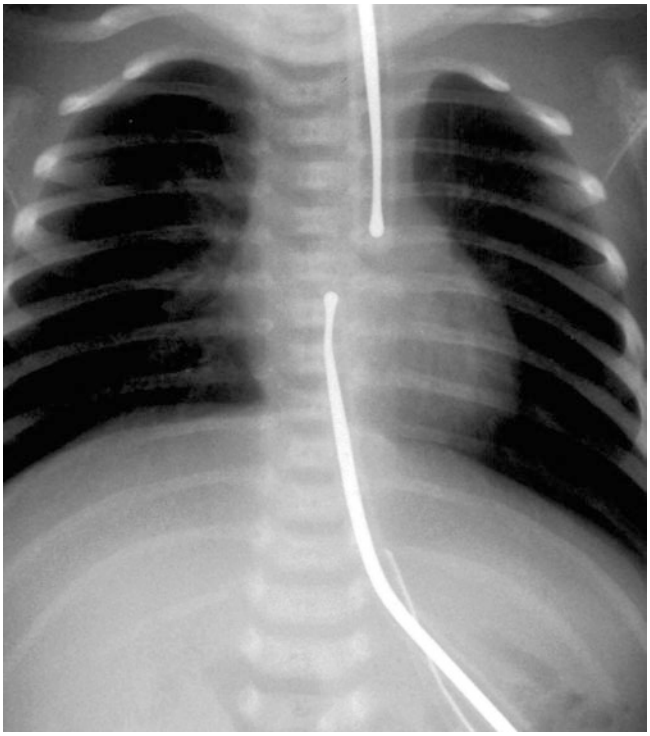


Fig. 6.17 The progress of elongation is evaluated by weekly fluoroscopic calibration and radiographic documentation. Distinct overlapping of the segments, which is necessary for end-to-end anastomosis without tension, is achieved within 3–5 weeks. It must be mentioned that before beginning the procedure, it is necessary to confirm with a pediatric endoscope that a short lower esophageal pouch exists. In rare cases, there may be instead of a lower esophageal stump only a nubbin that cannot be elongated by bougienage

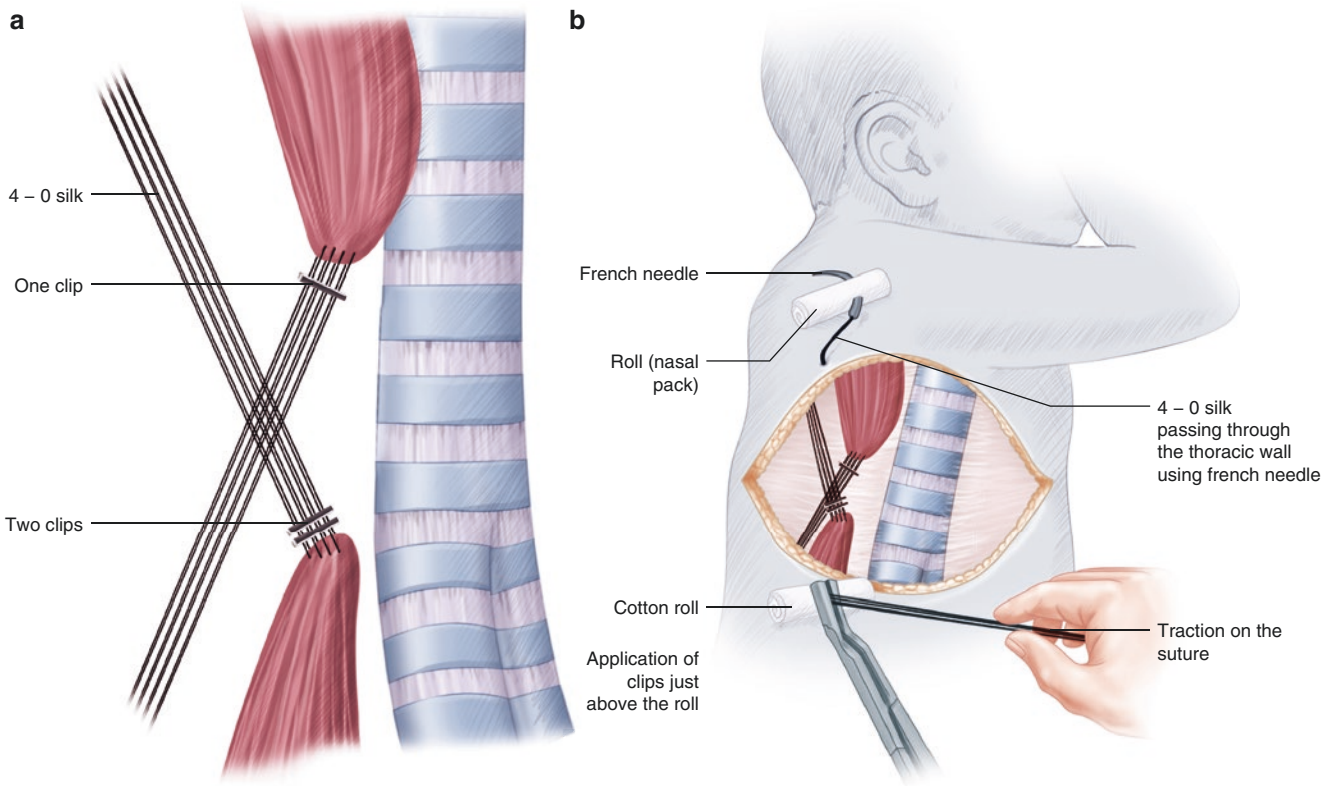


Fig. 6.18 (**a**, **b**), The third strategy was recently introduced by Foker (but is based on the old olive traction method of Rehbein). During thoracotomy, four tissue-pledgeted 5/0 or 4/0 traction sutures are placed extramucosally in the upper and lower segments and brought out to the skin below and above the incision. Daily external traction on these sutures allows the segments to be brought together within 14 (± 2.9) days, and the anastomosis is performed by a second thoracotomy. Recently, even a thoracoscopic application of the pledgets was proposed. A recent survey of 88 international surgeons showed that 39% are using the Foker technique, but 24% of those were not satisfied with the results

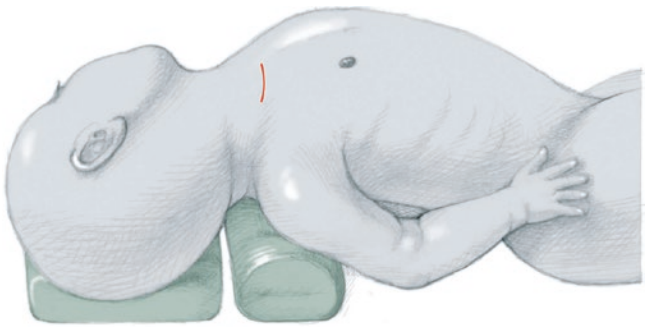


Fig. 6.19 H-type fistulas without atresia account for about 3% of the tracheo-oesophageal anomalies. The presentation is usually more protracted and sometimes is delayed beyond the first year of life. Typical symptoms are choking episodes during feeding, together with cyanotic spells. Diagnosis is made either by contrast oesophagogram or tracheobronchoscopy. If an H-type fistula is confirmed, a 3F ureteric catheter is passed across the fistula during bronchoscopy. Most H-type fistulas can be approached from the neck because they are usually situated at or above the level of the second thoracic vertebra. For the cervical repair, the child is placed supine on the operating table. The head is turned to the left and a folded towel or a sandbag is placed underneath the shoulders to hyperextend the neck. This position maximally expands and exposes the right cervical area. The incision follows a suitable skin crease, approximately 1 cm above the medial third of the right clavicle. After dividing the platysma, the medial border of the sternomastoid muscle is retracted posteriorly. The dissection proceeds medially to the carotid artery, and it may be necessary to divide the middle thyroid vein and the inferior thyroid artery to reach the trachea and oesophagus, which are situated medial and posterior to the thyroid lobes and isthmus. Palpation of the tracheal cartilages and the feeding tube in the oesophagus facilitates anatomical orientation. The recurrent laryngeal nerve runs upwards in the groove between the trachea and oesophagus, close to the fistula. It must be clearly identified and protected from any injury

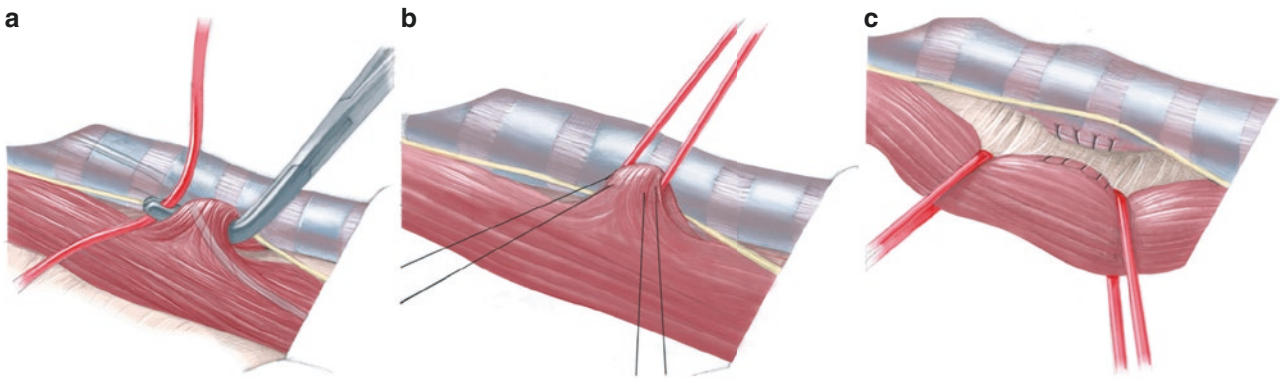


Fig. 6.20 (**a–c**) The plane between the oesophagus and trachea is carefully developed. The ureteric catheter in the fistula aids its identification. Right-angled forceps are used to dissect the fistula, and a small vascular sling is passed around it. Two stay sutures are placed on the oesophageal side of the fistula, which is divided after withdrawal of the ureteric catheter. A transfixation monofilament absorbable 6/0 suture is employed to close the tracheal side of the fistula and the oesophagus in interrupted technique. The wound is closed in layers with absorbable suture material, finishing with interrupted subcuticular absorbable 6/0 sutures. At the end of the operation, the motility of the vocal cords should be reassessed

6.1 Results and Conclusions

The first successful primary repair of an oesophageal atresia was achieved by Cameron Haight in 1941, but mortality remained high in the following decades. The outcome was influenced by birth weight, severity of additional malformations, and the development of aspiration pneumonia due to delayed diagnosis. Today, the diagnosis is usually established immediately after birth, and pneumonia can be prevented by continuous suction of the upper pouch. Survival of premature infants has significantly improved with progress in neonatal intensive care, so severe associated anomalies have become the main factor determining outcome.

Basic surgical management has become uniform for the most common type of oesophageal atresia with a lower fistula, but the best strategy for babies with long-gap oesophageal atresia remains controversial. Some authors (including our team) prefer to restore the native oesophagus whenever possible, even at the price of severe gastro-oesophageal reflux, whereas others favour more generous indications for oesophageal substitution, either with colon or stomach.

The overall prognosis of patients with oesophageal atresia is good, but recurrent dysphagia, secondary problems of gastro-oesophageal reflux, and an increased incidence of recurrent respiratory tract infection—possibly due to repeated minimal aspirations during sleep—are common sequelae. The distal oesophagus frequently suffers from delayed clearance due to disturbed motility. The impairment of propulsive peristalsis may be part of the malformation pattern, but it also may be iatrogenically worsened by damage of vagal nerve fibres during dissection of the distal oesophageal pouch. Nevertheless, severe swallowing problems with dysphagia are rare, but impaction of foreign bodies (most often pieces of bread, meat, or fruit) may be partially attributable to the motility disorder.

An anastomotic stricture can be the result of an anastomosis fashioned under high tension, impaired perfusion, and/or an anastomotic leak, or it may be caused by continuous acid exposure due to gastro-oesophageal reflux. Clinically, delayed clearance of acid reflux is probably of greater importance, as the incidence of gastro-oesophageal reflux disease exceeds 40% in patients with oesophageal atresia.

Atypically shaped cartilaginous C-rings and a wide intercartilaginous membrane within the region of the former fistula may be underlying causes of tracheomalacia, another common complication, with an incidence of about 20%. The anterior-posterior diameter of the trachea is reduced, and it may collapse completely with strained inspiration and expiration. The anomaly rarely causes serious problems and usually resolves with age and growth. Sometimes, however, severe respiratory distress with near-miss events may occur. Continuous monitoring and urgent treatment are then indicated. Aortopexy under bronchoscopic control is currently the most commonly used surgical method. It resolves the problem in many cases, unless the weak tracheal segment is too long. Recently, tracheoscopic stabilization with a self-expanding or balloon-expandable stent has been advocated. The ideal stent has yet to be found, however, and long-term results are awaited.

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Gastro-oesophageal Reflux and Hiatus Hernia

7

Keith E. Georgeson and Michael E. Höllwarth

Gastro-oesophageal reflux (GER) is a physiological phenomenon that occurs in persons of all age groups, more frequently after the ingestion of liquid food. Therefore, most infants spit up milk after feedings, sometimes in a spectacular fashion. This postprandial regurgitation is rarely associated with any serious consequences to the baby and is usually outgrown by 1–2 years of age.

Pathologic gastro-oesophageal reflux disease (GERD) is defined when reflux of gastric contents causes troublesome symptoms and/or complications. In infants and young children, it is associated with complications such as failure to thrive, recurrent apnea, and recurrent aspiration of gastric contents. In older children, GERD may cause oesophagitis and Barrett oesophagus; it also can present with reactive airways disease and chronic sinusitis.

7.1 Diagnosis

A careful history is invaluable in eliciting the symptoms of GER in children. Frequent regurgitation, failure to thrive, constipation, recurrent epigastric or retrosternal pain, and extreme irritability are all potential signals of GER. The workup should include an upper gastrointestinal study to rule out reflux-associated stenosis, the presence of a hiatus hernia, or other anatomic causes of regurgitation. For a long time, a 24-h pH probe study was considered the gold standard for detecting pathological GER in children, but pH monitoring can detect only acidic refluxes, and a negative pH probe study does not rule out symptomatic GER due to the common occurrence of nonacidic reflux in paediatric patients. Thus, today multiple intraluminal esophageal impedance recording with pH monitoring (MII-pH) allows the detection of both acidic and nonacidic refluxes. Upper gastrointestinal endoscopy with biopsy and histology is occasionally useful in defining the presence of oesophagitis secondary to GER. Gastric emptying studies are usually not useful in the workup for GER in children.

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7.2 Treatment

Proton pump inhibitors (PPIs) are useful as a first therapeutic modality for most children with pathologic GER, but it must be remembered that although PPIs significantly reduce the acidity of refluxes, they do not necessarily reduce the total number of refluxes. Long-term treatment of children with PPIs cannot be recommended, unless surgical procedures are not indicated. Patients who suffer from reflux-associated oesophageal stenosis should be treated with PPIs and esophageal bougienage before surgical therapy is considered, unless the patient is experiencing life-threatening symptoms.

A number of new technical methods have been introduced in the last few years for treatment of reflux in adults. Transoral endoscopic folding of the stomach anteriorly around the oesophagus has been proven effective in randomized control trials and has been shown to complement current surgically and medically available options in children, especially in complicated patients.

Antireflux surgery is indicated as antireflux surgery in all patients with inadequate response to medical management or in children who cannot be weaned from medical management. Antireflux surgery is also appropriate in neurologically impaired children, in children with complications of peptic oesophagitis and Barrett's oesophagus, and in children with reflux-associated hiatus hernia (Figs. 7.1, 7.2, 7.3, 7.4, 7.5, 7.6, 7.7 and 7.8).

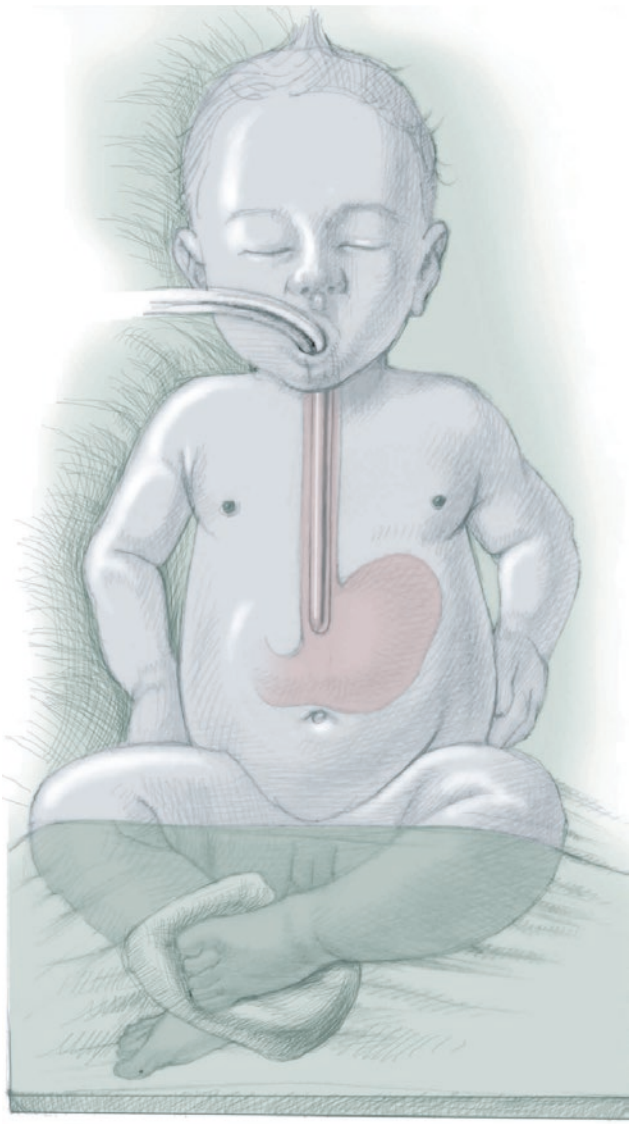

Fig. 7.1 The intubated patient is positioned at the end of the operating table. The knees are flexed and the feet cushioned. The patient is taped to the table so he or she will not slide when placed in the reversed Trendelenburg position. The operator stands at the end of the table, which is positioned low enough for easy manipulation of the laparoscopic instruments. A large Maloney bougie is passed through the mouth into the stomach. The dilator should be large enough to fully distend the distal oesophagus for safer peri-oesophageal dissection. The patient's head should be positioned so that the anaesthetist has access to withdraw and advance the dilator as needed throughout the course of the operative procedure. The patient is prepped from nipples to groin

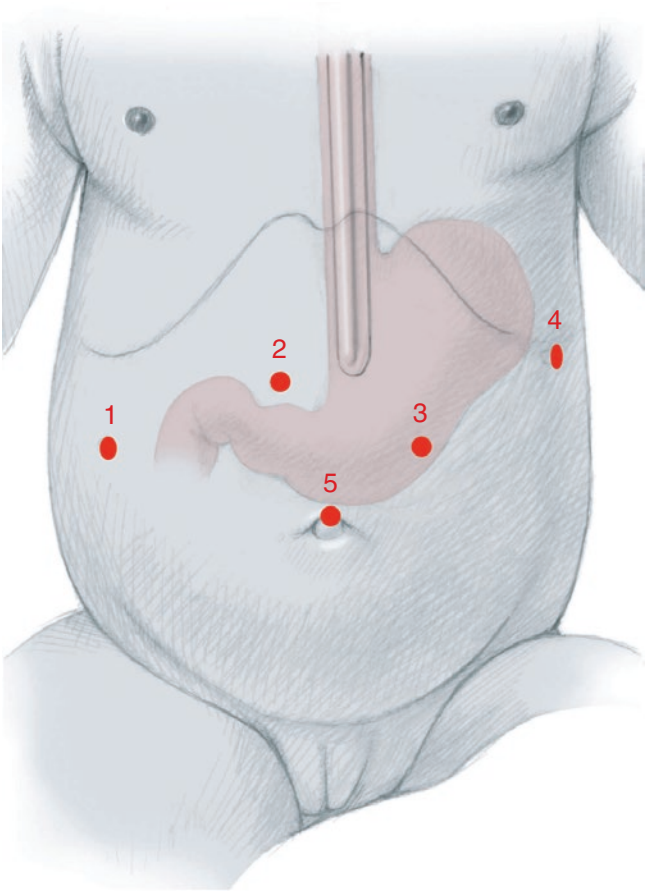

Fig. 7.2 Five trocars are inserted in the abdominal wall. Unlike adult patients, who have a precisely defined position for each trocar, children have more variation in body habitus and liver position, so the placement of the trocars must be tailored to the individual patient. The initial trocar placement is through the centre of the umbilicus in the midline. Each trocar site should be infiltrated with a local anaesthetic prior to the placement of the trocar. The incision in the umbilicus should be the same size as the trocar. The umbilical scar does not stretch well. An extremely tight trocar will cause ischaemic injury to the skin of the umbilicus if the skin incision is not large enough to accommodate the trocar. The incision should be made through the central portion of the umbilicus. The peritoneal cavity is usually easily entered through the umbilicus because the layers of the abdomen are scarred together at this point. Once the peritoneal cavity is opened with a no. 11 blade, a curved mosquito clamp is introduced into the peritoneal cavity with the tip upward, pointing away from the abdominal viscera. The clamp is pushed inward to dilate the umbilical opening, allowing easier access to the peritoneal cavity. A radially expanding disposable 5-mm trocar with a fitted Veress needle is then passed into the peritoneal cavity while pulling upward on the umbilical skin. The trocar should be advanced at a 30° angle and its tip kept as close to the parietal peritoneum of the anterior abdominal wall as possible to avoid injury to intra-abdominal or retroperitoneal structures. The Veress needle inside the expandable sheath is then removed and the trocar cannula inserted through the plastic sheath, expanding the trocar and fixing it to the abdominal wall by its snug fit. Suture fixation is sometimes necessary in smaller infants with a thin abdominal wall. A 30° 4-mm scope is advanced through the umbilical trocar after a pneumoperitoneum has been instilled. This scope is then used for surveillance during the placement of the other four trocars. The second trocar is placed in the right upper quadrant. This trocar should be positioned at the inferior margin of the liver border in the right anterior axillary line. The articulated retractor is passed toward the left upper quadrant and tightened to form its working position. It is then secured to the frame of the operating table by way of a retractor holder. The third, fourth, and fifth trocars are then placed under laparoscopic surveillance. All but the umbilical trocars are reusable 3-mm or 4-mm trocars, except in patients over 20 kg, when a larger liver retractor is passed through a 5-mm trocar. Trocar site 3 is used for the endoscope and is also the prospective gastrostomy site if a gastrostomy button is placed. Trocar sites 2 and 4 are the working ports for the operating surgeon. Trocar site 5 is the initial entry point and is also used for surveillance during placement of the other four ports. When the endoscope is moved to port 3 to perform the operation, port 5 is used for intraperitoneal access by the surgeon's assistant

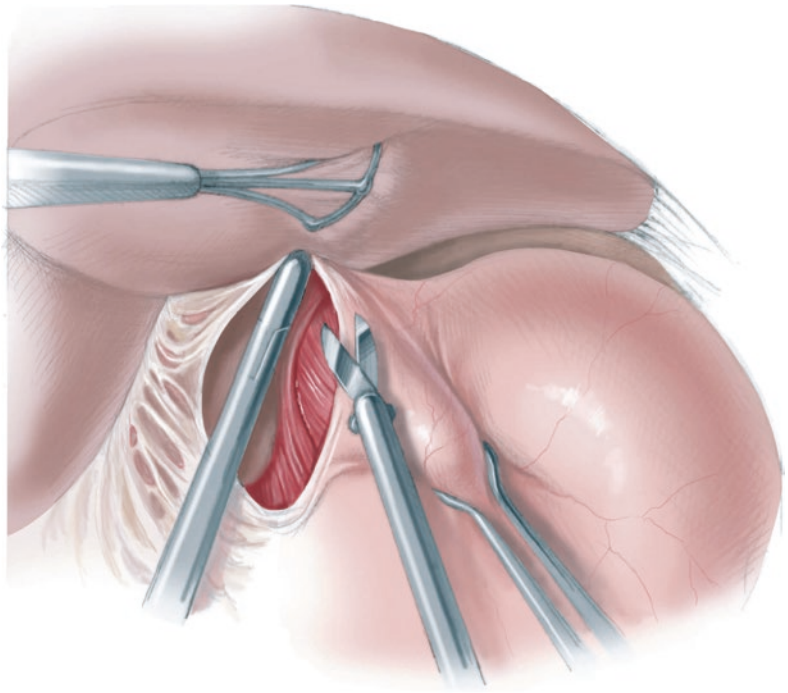

Fig. 7.3 The intraperitoneal dissection is begun by opening the upper part of the hepatogastric ligament. The dissection is performed sharply. The hepatic branches of the vagus nerve are divided. The small vessels to the liver are also divided using electrocautery. Care is taken to avoid transecting the left hepatic artery, which can be found in this ligament in a very small number of patients. The dissection is carried up to the hiatus in an avascular plane. The phreno-oesophageal ligament is opened between the oesophagus and the right crus. The peri-oesophageal plane should be entered cleanly to avoid excessive bleeding and to facilitate a faster operation. The dissection is continued in this plane bluntly and sharply over the top of the oesophagus and down the left side. The anterior vagus nerve is usually tightly adherent to the muscle of the oesophageal wall, but occasionally the nerve falls away and is only loosely associated with the oesophageal wall. Linear structures along the anterior oesophageal wall should be carefully evaluated before dividing them. The cleavage between the oesophagus and the left crus should be carried posteriorly until the fundus of the stomach is encountered

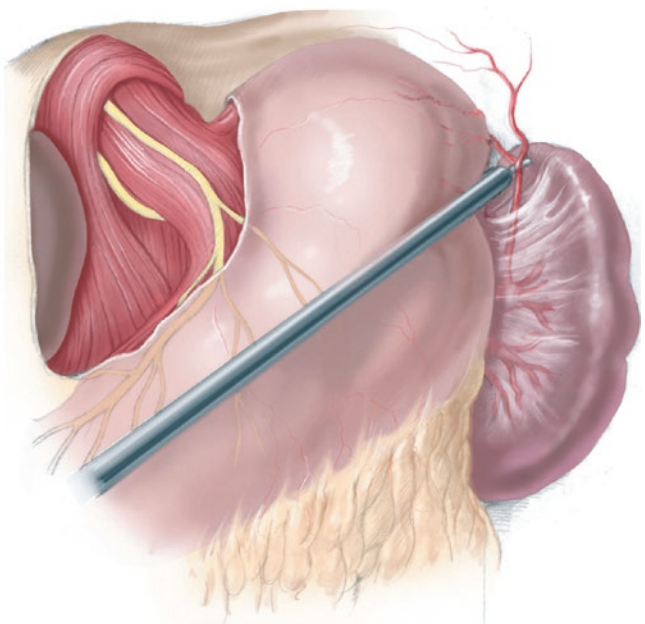

Fig. 7.4 The short gastric vessels are divided routinely. Dividing these vessels allows much better visualization of the left crus and also contributes to a better geometry of the fundoplication wrap. In most patients, the vessels are divided with a hook electrocautery. In large or obese patients, an ultrasonic scalpel is useful in dividing the short gastric vessels. The gastrosplenic ligament is opened at the mid portion of the spleen. The dissection is carried cephalad from this point. Most patients have both an anterior and posterior leaflet of the gastrosplenic ligament. Vessels run in both leaflets

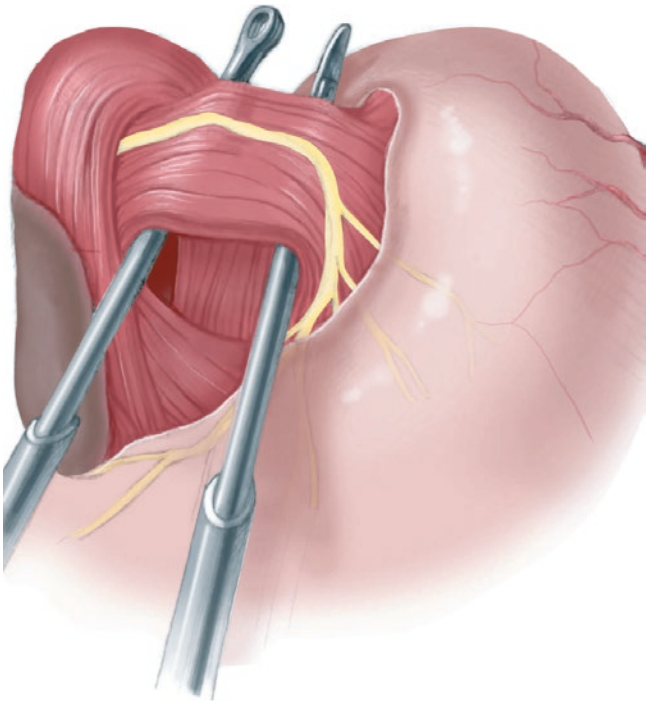

Fig. 7.5 The left crus should be followed as it courses toward the right side behind the oesophagus. Dissection is performed from both sides. The fundus is pulled down using a grasper through the umbilical port site, which allows excellent visualization of the left crus. For visualization of the right crus, the grasper is pulled downward and toward the splenic flexure of the colon. By dissecting both sides alternately using the left crus as a guide, a window is safely formed behind the oesophagus. The tissues tethering the oesophagus to the crura are further divided circumferentially to lengthen the intra-abdominal portion of the oesophagus. The posterior vagus is vulnerable and should be identified and preserved. An instrument passed through the umbilical trocar is used to retract downward on the gastro-oesophageal junction while lengthening the intra-abdominal oesophagus. Circumferential dissection around the oesophagus is continued as far into the mediastinum as necessary to provide at least 2.5 cm of oesophagus in the abdomen with no downward tension on the oesophagus. If less than 2.5 cm of oesophagus remains in the abdomen after releasing the downward tension on the oesophagus, consideration should be given to lengthening the abdominal oesophagus by tubularizing the upper stomach. As much as possible, the fascia covering the crura should be left intact. Care should also be taken to avoid entry into the pleural cavity on either side. If a hole is made in the pleura, it should be enlarged to avoid the development of a tension pneumothorax caused by a one-way ball-valve effect. The pneumothorax can be evacuated by needle thoracentesis at the end of the operation

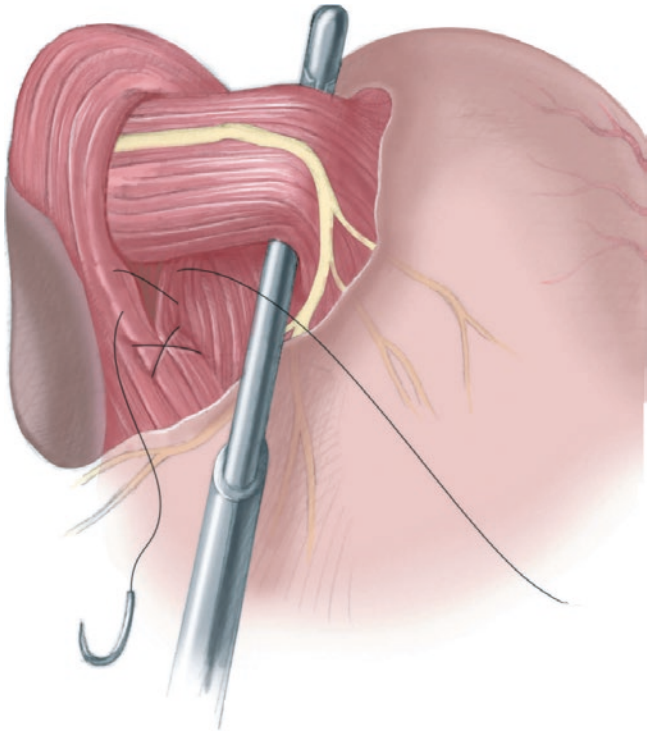

Fig. 7.6 The crura are closed in every case by approximating them behind the oesophagus with nonabsorbable sutures. Generous bites of crus are taken on both the left and right sides and are tied snugly. The aorta is located behind the posterior aspect of the left crus and should not be incorporated in the suture closing the crura. The author prefers to close the crura with the dilator withdrawn into the oesophagus. Great care should be taken to avoid closing the hiatus too tightly. If a large dilator is left in the intra-abdominal oesophagus, suture placement is more difficult and the hiatus is often left larger than it should be. With large hiatal defects, the hiatus may require both posterior and anterior closure. Once the hiatus is closed around the oesophagus, at least 2 cm or more of oesophagus is fixed in the abdomen, utilizing three or four collar sutures. These sutures are usually placed at the 11, 7, and 3 o'clock positions on the oesophagus, incorporating a portion of the oesophageal wall and coapting it to the associated crus. For large hiatus hernias, four or more collar sutures may be needed to adequately close the oesophageal hiatus

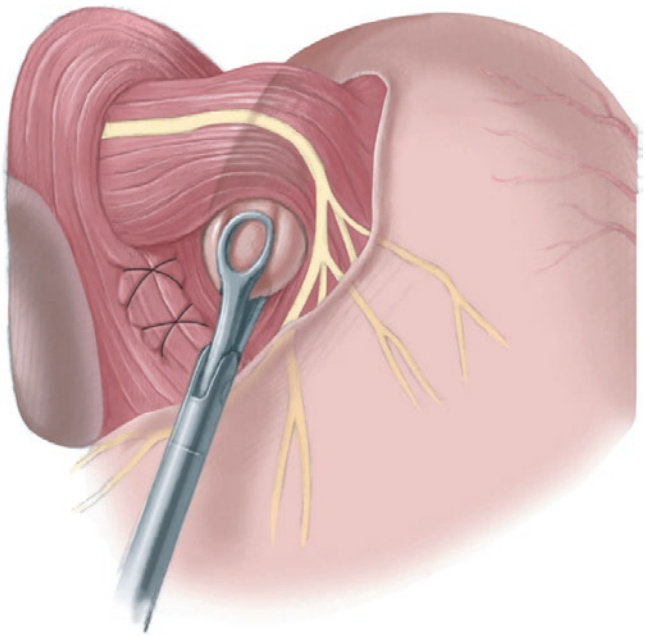

Fig. 7.7 The mobilized fundus is pushed up beside the left side of the oesophagus. A grasper via the umbilical port is used to lift the oesophagus, exposing the fundus behind the oesophagus. The fundus is grasped and pulled through the retro-oesophageal window. The fundus is fluffed until a geometric symmetry is achieved. A "shoeshine" manoeuvre is used to confirm the fundal wrap and to avoid attaching the fundus to the mid portion of the stomach

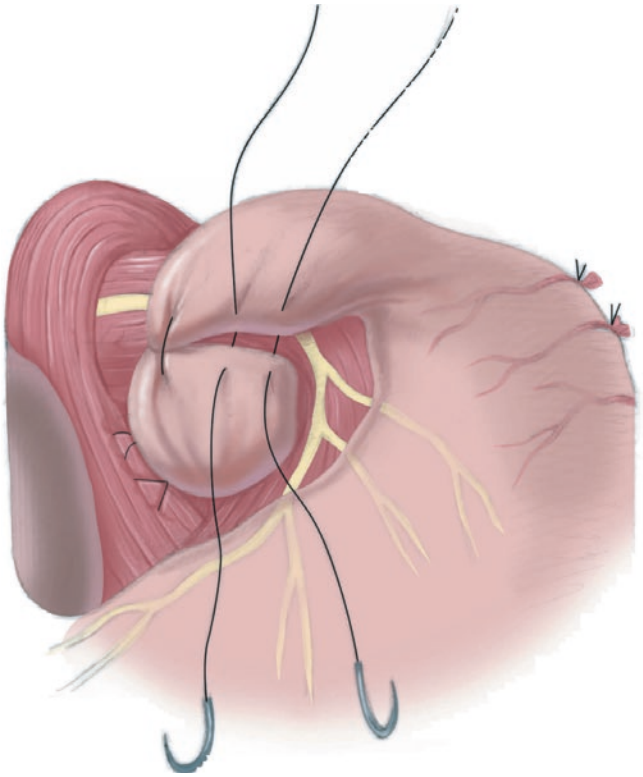

Fig. 7.8 The dilator should be repositioned into the stomach at this time. The left side of the fundus is then basted to the right side of the fundus with a single stitch that does not incorporate oesophageal tissue. The wrap should be loose and should encircle the oesophagus. Two or three nonabsorbable sutures are placed above and/or below the first stitch, incorporating the left and right sides of the fundal wrap and securing them to the oesophagus. The wrap should be no more than 1.5–2 cm in length and should lie loosely around the oesophagus. A figure-of-eight suture is then placed near the bottom of the wrap as a second layer to secure the fundoplication and prevent wrap breakdown

7.2.1 Operative Procedure: Gastrostomy Placement (Figs. 7.9–7.11)

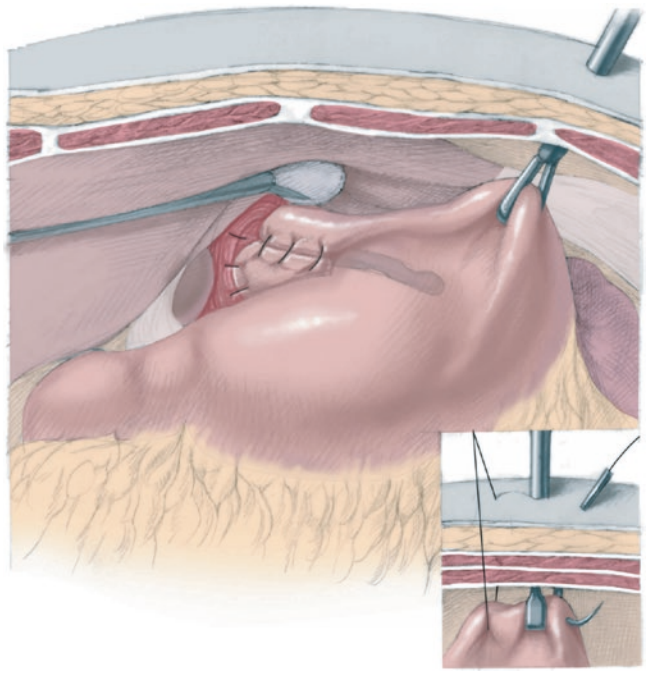

Fig. 7.9 If a gastrostomy is to be placed, the laparoscope is moved back to the umbilical port. A locking grasper is passed through the medial left upper quadrant trocar site. This trocar is initially positioned with the intention of using this site as the gastrostomy site. The stomach is grasped near the greater curvature at the junction of the body and antrum. If a fundoplication was not performed, the stomach should be grasped close to the lesser curvature. Using a large curved needle with a monofilament suture swaged to the needle, a U-suture is passed through the abdominal wall through the stomach, taking a 0.5–1-cm bite of stomach, and back through the abdominal wall. Passing the suture into the gastric lumen does not seem to lead to complications. A second U-suture is passed parallel to the first, 1.5 cm lateral to the first suture. The grasper is then removed, along with the trocar

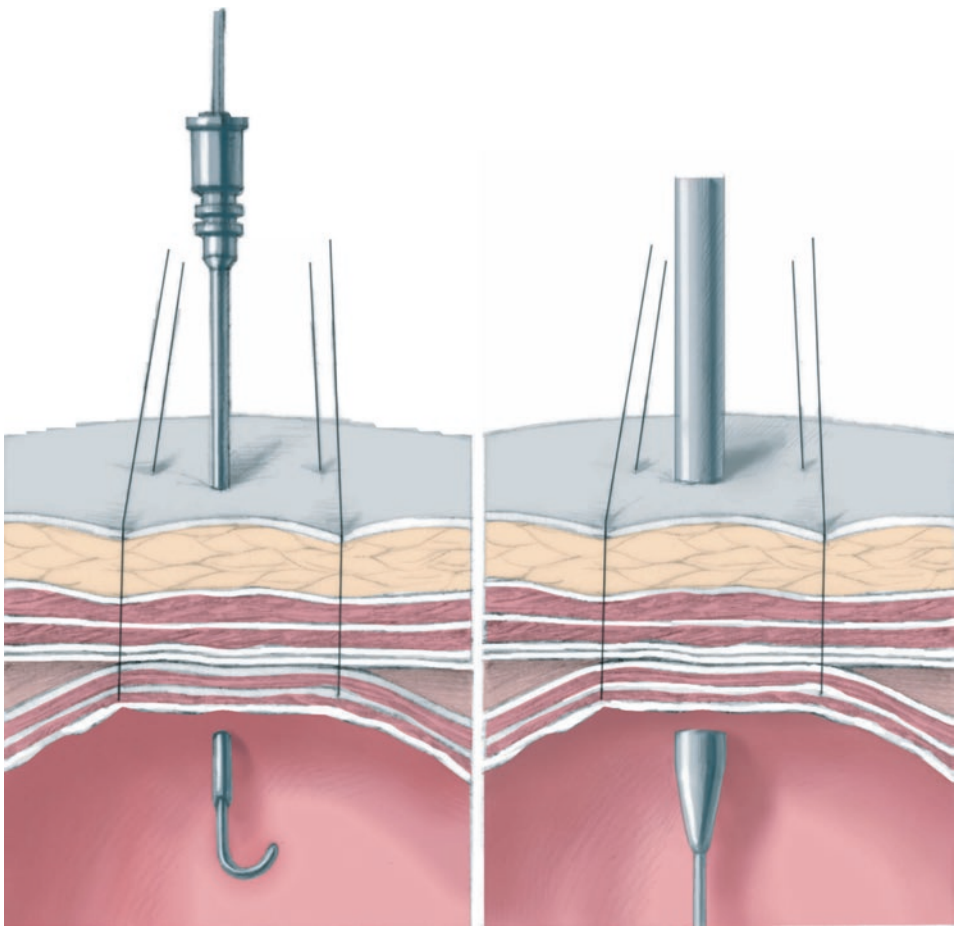

Fig. 7.10 The anaesthetist passes a single-lumen orogastric tube into the stomach and inflates the stomach with 60–120 cc of air. A hollow needle is passed through the medial left upper quadrant trocar site into the inflated stomach. The passage of the needle into the lumen of the stomach should be visualized completely and should not occur on the blind side of the stomach. A J-wire is then passed through the needle into the stomach and the needle is removed over the J-wire. The tract is dilated with vascular dilators from a size 8 French up to a size 20 French. The 20-French dilator should be passed through the abdominal wall only and not into the stomach. The U-suture should be allowed to slacken during passage of the 20-French dilator, to avoid passing the dilator into the stomach

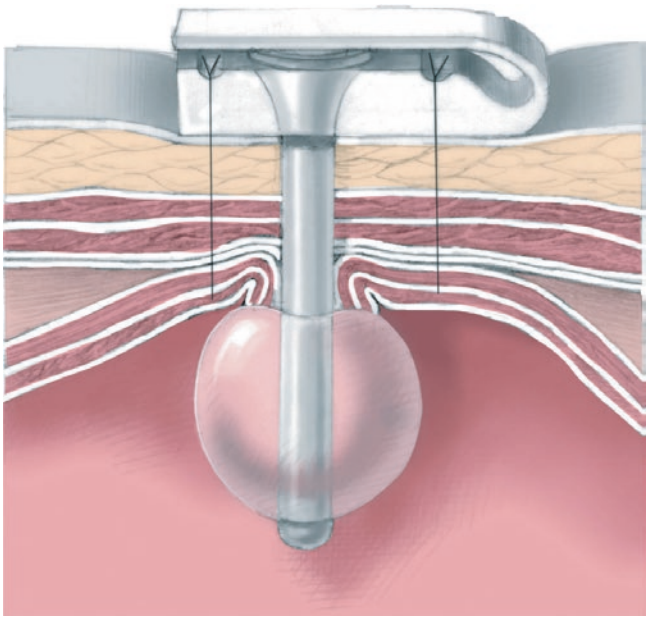

Fig. 7.11 The gastrostomy button is stiffened by passing the 8-French dilator through it. The dilator and balloon button are passed over the guide wire. Gentle twisting of the balloon button while holding counter-traction on the U-suture allows the balloon button to slip into the stomach under laparoscopic surveillance. The balloon should be inflated under direct visualization. The U-sutures are slackened at this point to make certain that the stomach is independently held against the abdominal wall by the inflated balloon button. The U-sutures are then tied over the wings of the balloon button. The laparoscope is passed through the lateral left upper quadrant trocar site to look at the gastrostomy button from a different angle to ensure that it is properly positioned and remains inflated

7.2.2 Operative Procedure: Closure

The liver retractor is removed using laparoscopic surveillance. The umbilical trocar is the first trocar to be removed after the pneumoperitoneum is evacuated. The fascia of the umbilicus is closed using a groove director to protect the underlying bowel and omentum. A simple or figure-of-eight suture is used to close the umbilical fascia. Once closure of the umbilical fascia has been achieved, the pneumoperitoneum is reinstated and the umbilical closure is visualized from a lateral port site to confirm that the omentum has not been incorporated in the umbilical closure. The other trocars are then removed. The fascia in these other trocar sites usually does not require closure. The skin is closed with subcuticular sutures and skin strips. The umbilical skin should be closed carefully with rapidly absorbable braided suture. Careless closure of the umbilicus can result in postoperative granuloma formation.

7.3 Postoperative Care

The patient is fed clear liquids on the day of surgery. Pureed foods are useful for 3–4 weeks to avoid the dysphagia associated with oedema of the fundoplication wrap. Discharge is 1–3 days after surgery. Postoperative pain is controlled with intravenous ketorolac, scheduled acetaminophen, and ibuprofen. Narcotic agents are used only when necessary. The U-sutures in a gastrostomy are removed on the second postoperative day.

7.4 Complications

The mortality rate after fundoplication is nearly zero, but the postoperative morbidity is about 5–20% with open or laparoscopic surgery. The most common complication is recurrent reflux; a special risk factor is mental disability accompanied by frequent choking, belching, or rumination. A further risk factor is preoperative dystrophy, so these patients should undergo preoperative nutritional therapy in order to bring them into an anabolic state.

Frequent causes of recurrences are inadequate closure of the diaphragmatic crura, short oesophagus, and inadequate mobilization of the oesophagus. Postoperative oesophageal dilatation after a too-tight Nissen fundoplication should be avoided, as it is associated with the inability to vomit, breakdown of the fundoplication wrap, and/or herniation of the stomach into the chest.

Common late complications are gas-bloat syndrome (mostly as a consequence of the smaller gastric volume), dysphagia, postoperative pain, and diarrhea. Most of these problems resolve spontaneously after a few months.

Conclusion

Fundoplication is a highly reliable therapy in children with persistent or life-threatening GER. Laparoscopic fundoplication is superior to open fundoplication and should be in the repertoire of all paediatric surgeons who operate on children with GER.

Suggested Reading

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Achalasia

8

Paul K. H. Tam and Patrick H. Y. Chung

8.1 Introduction

Achalasia (a Greek term meaning "does not relax") is a rare motility disorder characterized by an absence of normal oesophageal peristalsis, an increased basal resting pressure, and failure of complete relaxation of the lower oesophageal sphincter (LOS). Less than 5% of all cases present before the age of 15 years, giving an estimated incidence of 0.1 per 100,000 children. Males and females are equally affected.

The condition was first described in 1674 by Willis, who successfully treated a patient by repeated oesophageal dilatation using a sponge-tipped whale bone rod. In the early 1900s, based on observations in 100 reported cases, von Mikulicz suggested cardiospasm as the aetiology. In 1914, Heller described cardiomyotomy, a procedure that carries his name and forms the basis of all surgical approaches to this problem up to this date. The original operation consisted of two myotomies anteriorly and posteriorly on the lower oesophagus, performed through a laparotomy. A single anterior cardiomyotomy was subsequently found to be adequate for symptomatic relief. The operation has been performed through a thoracotomy, as well as thoracoscopically and laparoscopically with or without an additional antireflux procedure.

8.2 Pathogenesis

The pathogenesis of primary achalasia is not well understood. The most consistent histologic finding is a decrease or loss of myenteric ganglion cells, which is more pronounced in advanced cases. The degenerative process especially involves neurones producing neuropeptides and nitric oxide, the latter being identified as inhibitory neurotransmitters. Loss of inhibitory innervation causes increased tonic contraction and interference with normal relaxation of the LOS, as well as aperistalsis of the oesophageal body. There are no specific histologic changes in the oesophageal muscles. The cause of the neuronal damage remains unknown. Various mechanisms—autoimmune, infectious, genetic, toxic, and primary—have been proposed. The finding of myenteric inflammation, which is predominantly lymphocytic, the presence of serum autoantibodies to myenteric plexus, and the increased frequency of class II histocompatibility antigens in patients with achalasia supports an autoimmune aetiology, whereas the similarity between achalasia and Chagas' disease caused by *Trypanosoma cruzi* suggests that a neurotropic infectious agent may be responsible. Rarely, familial cases and association with microcephaly and other congenital anomalies have been observed.

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8.3 Diagnosis

Patients usually present one or more of the following symptoms: vomiting or regurgitation of undigested food, progressive dysphagia, weight loss or failure to thrive, choking, retrosternal discomfort, and pulmonary problems such as recurrent coughing or chest infections. Vomiting and dysphagia are the most common initial symptoms. Vomiting occurs more frequently in infants and young children, whereas dysphagia is more common in older children.

Chest X-ray may show an air-fluid level in the oesophagus, and there may be a soft tissue shadow in the mediastinum on the left hemithorax, corresponding to a dilated lower oesophagus. Sometimes pneumonic changes are seen. The characteristic radiologic features of achalasia in a contrast swallow are proximal dilation of the oesophagus with a smooth tapering of the gastro-oesophageal junction (bird's-beak sign or rat-tail deformity). There is an absence of coordinated peristaltic waves in the proximal oesophagus, and a persistent failure of relaxation of the LOS on swallowing.

Endoscopy confirms a dilated oesophagus, which funnels smoothly towards a narrowed LOS. Retained food or yeast oesophagitis may be noted in the oesophagus. Although the LOS is closed, it provides little resistance to the advancing endoscope.

Oesophageal manometry is the "gold standard" for the diagnosis of achalasia. Diagnostic features include a failure of relaxation of the LOS on swallowing and absence of peristalsis in the body of the oesophagus. Features that are characteristic but not required for the diagnosis include elevated resting LOS pressure (>45 mm Hg), and resting pressure in the oesophageal body exceeding that in the stomach. Recently, the use of high-resolution manometry (HRM) has allowed a more detailed assessment of oesophageal pressure and better prediction of patient outcome.

8.4 Treatment

Symptomatic relief can be achieved by lowering LOS pressure with oral nitrates or calcium channel blocker (nifedipine) medication or with intrasphincteric injection of botulinum toxin. The oral medication can result in a 50% decrease in LOS but is commonly associated with side effects such as headache. Experience with intrasphincteric injection of botulinum toxin is limited in children. Recent studies suggest a mean duration of effect of 4 months, and more than half of patients are expected to require a repeat injection within 6 months. The need for life-long medication (with its associated side effects) or repeated injections limits the role of medical therapy to those children who are not suitable for treatment by dilatation or surgery.

Definitive treatment of achalasia consists of dilatation or oesophago-cardiomyotomy. Dilatation should be guided by endoscopy/fluoroscopy and can be achieved using either rigid or balloon dilators, the latter being the preferred choice in children. Endoscopic balloon dilatation can be used for primary treatment or as a secondary procedure when symptoms recur after surgery. Our experience suggests that dilatation is less effective than surgery for long-term symptomatic relief.

8.4.1 Operative Procedure: Heller's Oesophago-Cardiomyotomy

Heller's oesophago-cardiomyotomy remains the mainstay of treatment for achalasia and can be performed via the abdomen or thorax, either as an open procedure or by the minimally invasive approach, and with or without a concomitant fundoplication (Figs. 8.1, 8.2, 8.3, 8.4 and 8.5). Preoperatively, any yeast oesophagitis should have been eradicated with antifungal medication. The patient is kept on clear fluids a day prior to surgery to minimize the risk of aspiration. Any retained food must be washed-out under oesophagoscopic control before anaesthesia induction, which may be time consuming. Finally, preoperative endoscopy ensures complete emptying of dilated oesophagus and a large feeding tube or a balloon catheter is introduced into the stomach.

After the surgery, a nasogastric tube is left overnight. Fluid diet is commenced after a contrast study confirms the absence of a leak and when gastric stasis has resolved. Return to normal feeds is usually faster for laparoscopic procedures.

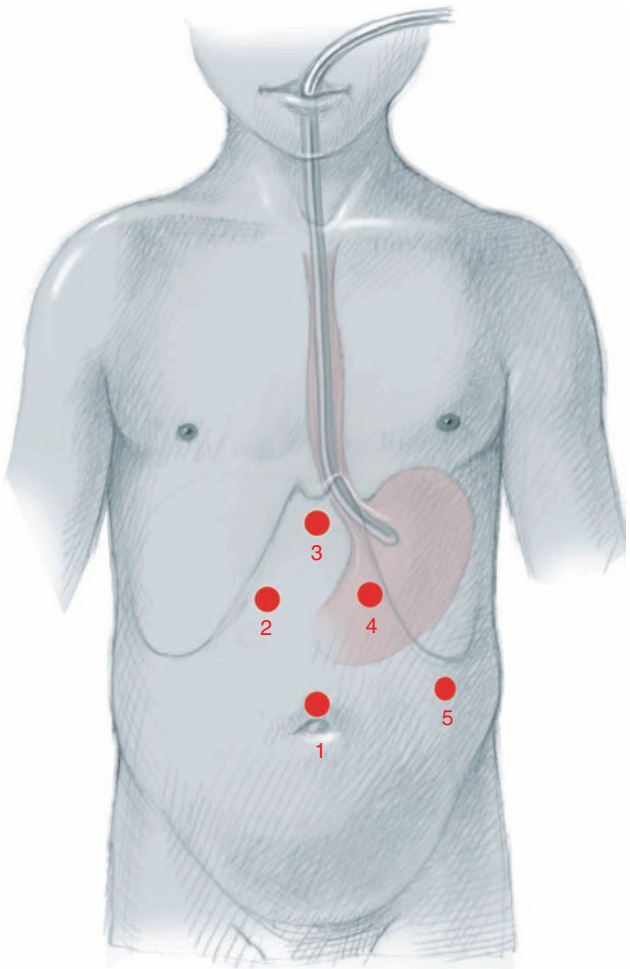

Fig. 8.1 Depending on the surgeon's preference, surgical access can be achieved via the abdomen or left thorax. The abdominal approach is more popular, and allows concomitant fundoplication to be performed more easily. With the patient supine, an upper mid-line is made for laparotomy. For laparoscopic access, the patient is placed in a lithotomy position with the surgeon at the end of the operating table; four to five ports are placed as shown. The telescope is placed in the supraumbilical port (*1*). To expose the oesophagus in the open procedure, the left lobe of the liver is retracted superiorly and medially; the left triangular ligament may be divided to enhance the exposure. For the laparoscopic approach, a cotton-tipped rod is inserted into the epigastric port (*3*) to retract the caudate lobe of the liver cephalad. Instrumentation is carried out via the remaining ports (*2*, *4*, *5*)

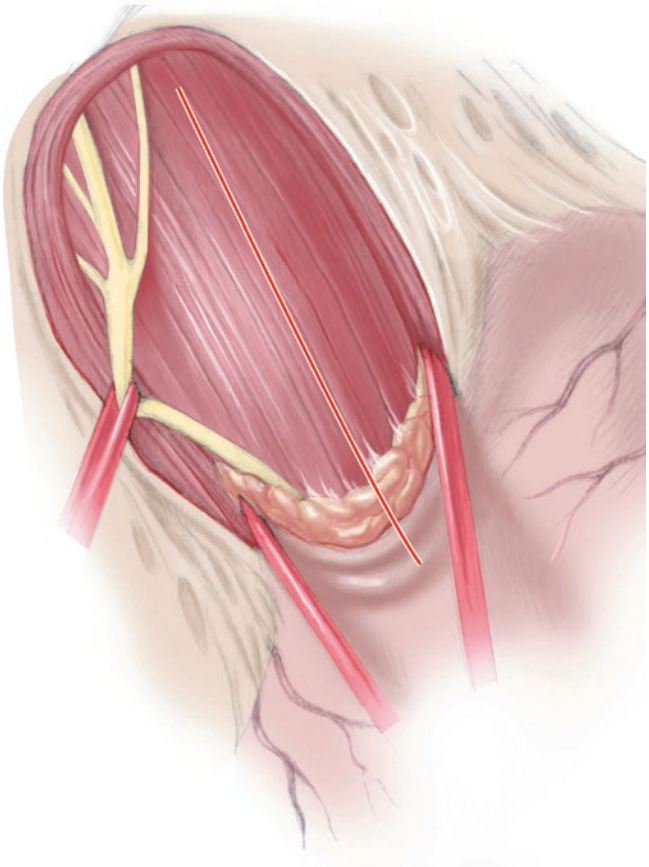

Fig. 8.2 The phreno-oesophageal ligament is incised. The anterior vagus is seen on the anterior wall of the oesophagus and should be preserved. The hiatal window is identified adjacent to the caudate lobe and the tissues between the oesophagus and the crura are divided. The abdominal oesophagus is further freed by blunt dissection into the posterior mediastinum, taking care not to penetrate the pleura proximally. The posterior vagus is preserved. The site of myotomy is marked by electrocautery to the left of the anterior vagus. The myotomy extends for 4–6 cm above the gastro-oesophageal junction and 0.5–1 cm below it. A superficial incision is made with the diathermy tip. The thickened oesophageal muscle is then divided and separated by blunt dissection with grasping/preparation forceps until the submucosal plane is reached. Great care is taken to avoid mucosal perforation

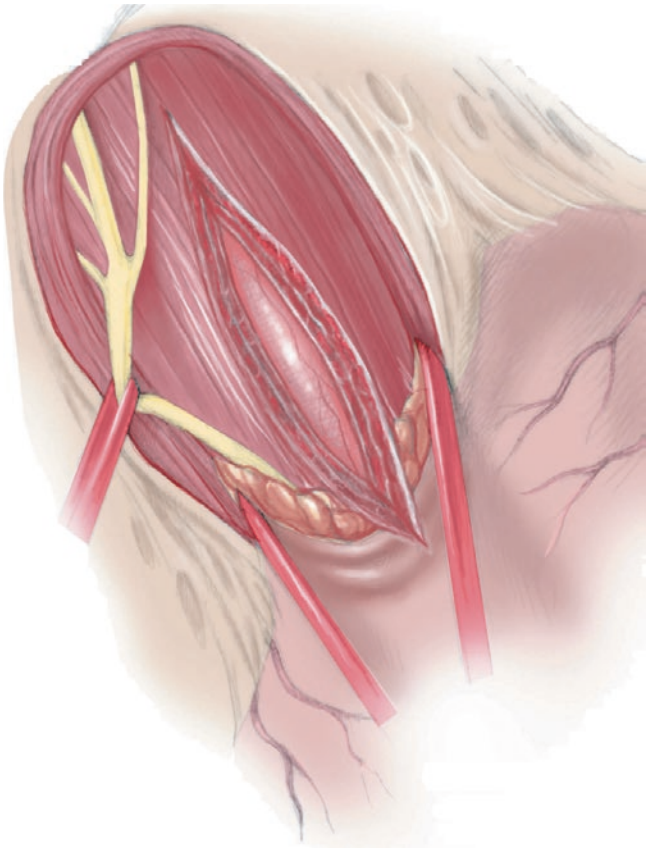

Fig. 8.3 Myotomy is continued proximally and distally with the diathermy hook and blunt dissection until all constricting muscles have been separated and the mucosa is seen bulging outwards; the muscular edges should be undermined for 50% of the oesophageal circumference. The gastro-oesophageal junction is recognized by the "collar-like" configuration of the circular muscles. The gastric muscles are usually more adherent to the mucosa. Mucosal perforation is tested by insufflation of the oesophagus; if perforation is present, it should be repaired by fine suture. A widened hiatus should be narrowed by one or two nonabsorbable, deep sutures placed through the crura. The wound is closed in the usual manner

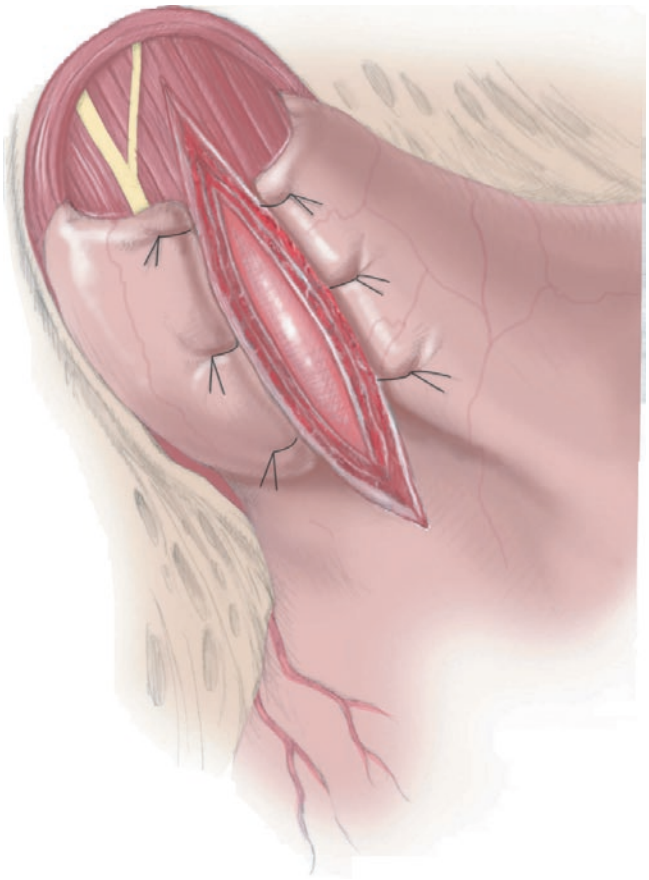

Fig. 8.4 To avoid the long-term complication of gastro-oesophageal reflux after myotomy, many surgeons recommend a concomitant fundoplication. Details of the procedure are separately described (*see* Chap. 6). The fundoplication should be loose, to avoid dysphagia. A posterior 180° (Toupet) fundoplication can be performed over the distal 1–1.5 cm of the oesophagus. The fundus is sutured separately to the cut edge of the oesophageal muscle on either side, using three nonabsorbable, interrupted sutures. In addition to providing an antireflux mechanism, this procedure holds the myotomy edges apart

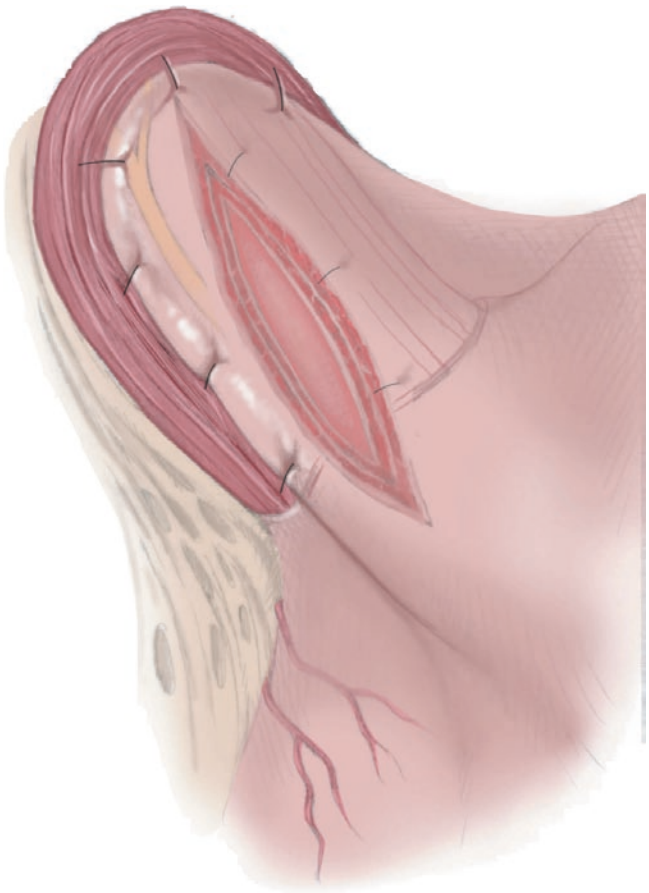

Fig. 8.5 Alternatively, an anterior 180° (Dor or Thal) fundoplication is performed. The anterior fundus is "draped" over the anterior oesophagus, covering the myotomy. This procedure may be more appropriate for patients with mega-oesophagus, as the posterior fundoplication is more likely to result in outflow obstruction. An anterior fundoplication also may provide additional cover after repair of a mucosal perforation

8.4.2 Operative Procedure: Balloon Dilatation

Endoscopic balloon dilatation (Fig. 8.6) has been reported to be effective in up to 90% of patients, but most authors (including ourselves) have been unimpressed with its efficacy as primary treatment for children with achalasia. Multiple dilatations are often required, with 20–75% of patients needing repeated dilatations. Serious complications include oesophageal perforation (0–16%) and symptomatic gastro-oeosophageal reflux (15–35%).

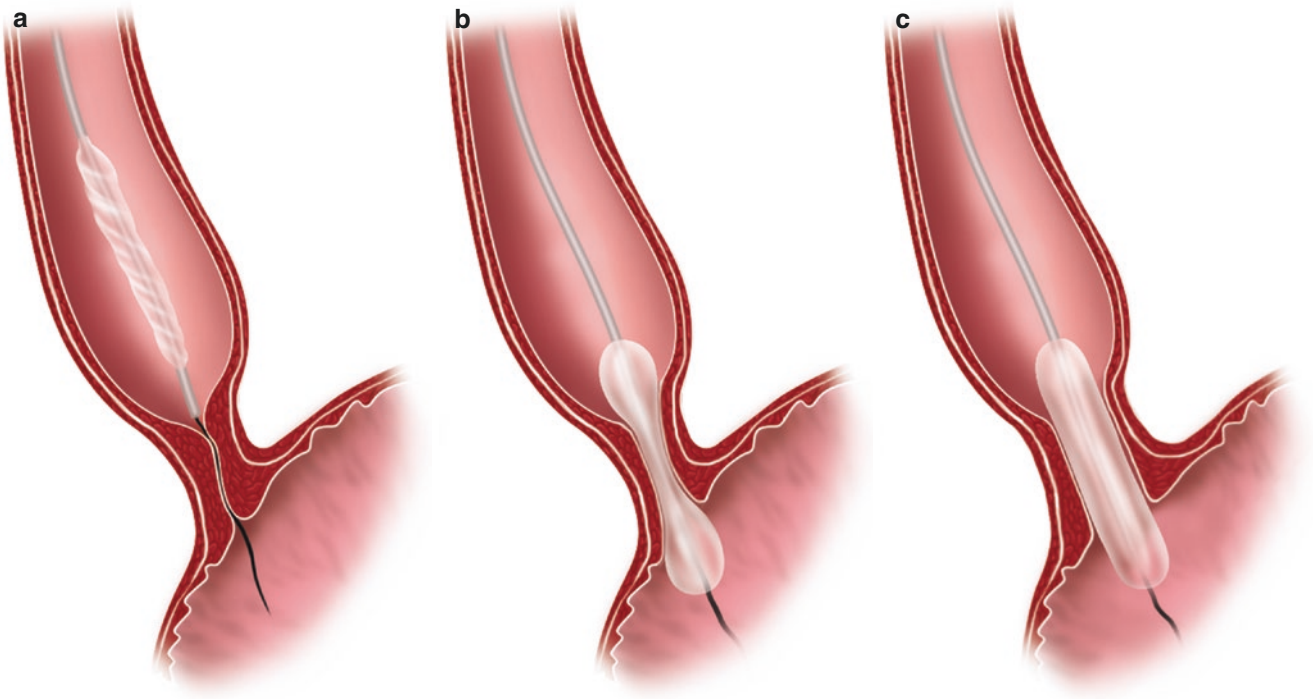

Fig. 8.6 Balloon dilatation can be guided by endoscopy/fluoroscopy or a combined approach. A guidewire is inserted through the gastro-oesophageal junction (**a**) and the expandable balloon is passed over the guidewire into the gastro-oesophageal junction (**b**). When the position is confirmed, the achalasic segment can be dilated by inflating the balloon (**c**). The dilatation process should not be prolonged, to avoid pressure-induced necrosis and subsequent stricture formation

8.4.3 Operative Procedure: Trans-Oesophageal Myotomy

Most recently, the successful development of natural orifice endoscopic surgery (NOTES) in human clinical use has inspired surgeons to perform trans-oesophageal endoscopic myotomy as a treatment for achalasia (Fig. 8.7). Similar to endoscopic submucosal dissection for early oesophageal cancer, myotomy can be achieved via the division of the inner muscle layer through a submucosal tunnel after a small mucosal opening in the oesophagus; the mucosal opening is closed with endoscopic clips. Symptom remission in 89% of patients at 6 months and 82% at 12 months has been reported in a recent prospective, multicenter study on adult patients, and short-term success has also been reported in a small number of children. Long-term outcome remains undetermined, however, especially for the paediatric population.

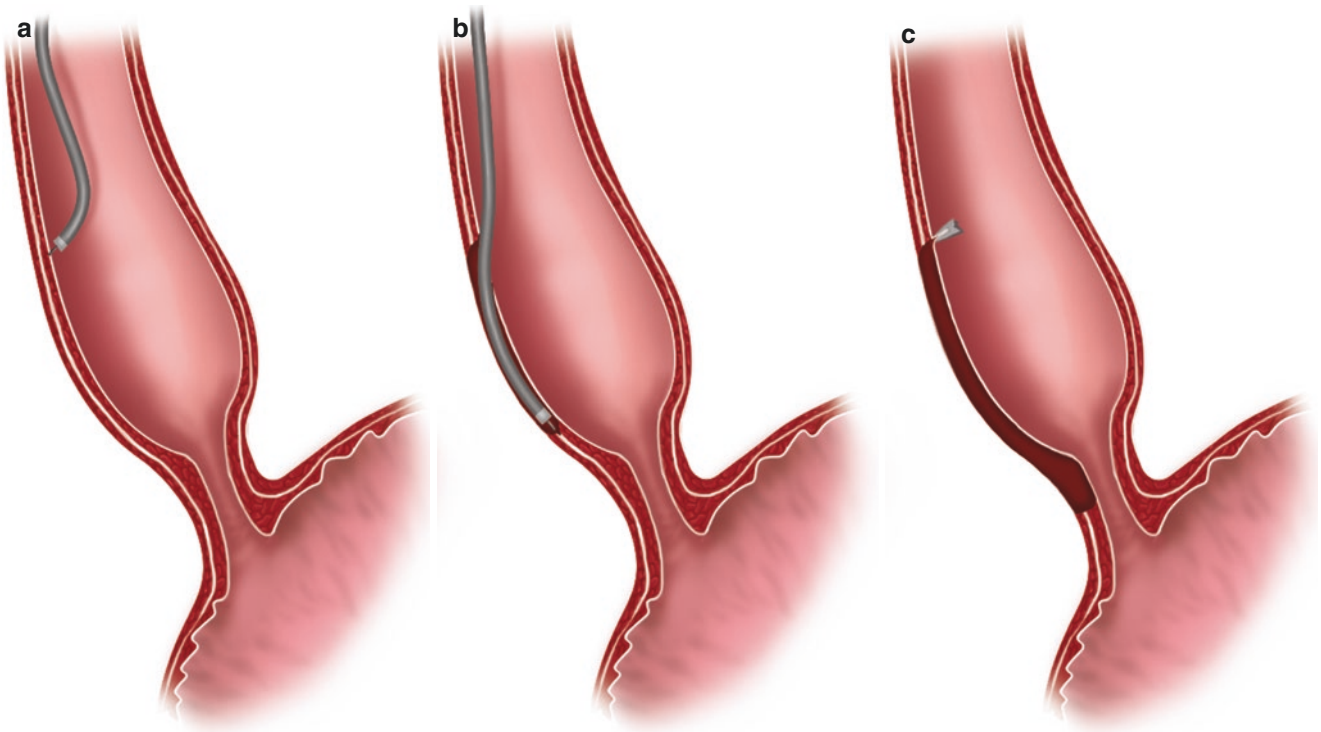

Fig. 8.7 Trans-oesophageal myotomy is performed under endoscopic guidance. A submucosal space is created at the lower oesophagus via a mucosal window (**a**). The inner circular muscle layer and the gastric muscle bundles are incised under direct vision (**b**). The incision should start 5 cm above the gastro-oesophageal junction and extend to 2 cm below the junction. A metal clip can be applied to close the mucosal window after the procedure (**c**)

8.5 Results

Heller's oesophago-cardiomyotomy is the method of choice for the treatment of achalasia in children. In recent years, laparoscopic surgery has become the standard operative approach in many centres over the world, with a reported mean success rate of 89% (76–100%) at a median follow-up of 35 months. The overall rate of complications, including oesophageal perforation, atelectasis, dysphagia, and gastro-oesophageal reflux, is 6.3% (0–35%). Most series report near-zero mortality. A recent meta-analysis comparing laparoscopic Heller myotomy versus endoscopic balloon dilatation has demonstrated superior short-term and long-term efficacy favouring myotomy as the first-line treatment for achalasia. Conversion to open myotomy is required in 10% of laparoscopic procedures, usually as a result of intraoperative oesophageal perforation, but with increasing experience, even oesophageal perforations can be repaired laparoscopically. A poor result following oesophago-cardiomyotomy can be due to mega-oesophagus, incomplete myotomy, or gastro-oesophageal reflux. Incomplete myotomy usually responds to secondary pneumatic dilatation. Gastro-oesophageal reflux is preventable by a concomitant fundoplication during myotomy; performing a fundoplication as a second operation after an initial myotomy without fundoplication is technically more difficult.

With the development of robotic technology in paediatric populations, robotic-assisted Heller's myotomy has been reported. Results from early studies showed better short-term and long-term outcomes than conventional surgery and appeared to be equivalent to laparoscopic surgery. When the three procedures were compared, the minimally invasive approaches resulted in less morbidity (9.08% for open surgery, 5.19% for laparoscopic surgery, 4.02% for robotic surgery) and lower rates of ICU admission.

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Colonic Replacement of the Oesophagus

9

Devendra K. Gupta and Shilpa Sharma

The colon is the most time-tested and frequently used organ for oesophageal replacement. The advantages of colonic replacement include good blood supply, easy access, and availability of adequate length with a vascular pedicle while leaving enough colon behind for appropriate function. Most surgeons adopt this procedure because of the fast learning curve and ease of performing the procedure, minimal early complications, and good long-term results.

9.1 Indications

The most common indications for oesophageal replacement in children are wide-gap or long-gap oesophageal atresia (with diversion done with cervical oesophagostomy and a gastrostomy) or an oesophageal stricture developing after corrosive ingestion. Other rare indications include prolonged oesophageal candidiasis leading to severe dysmotility and recurrent chest infections, epidermolysis bullosa and strictures following the ingestion of foreign bodies, perforation injuries, and injection sclerotherapy for oesophageal varices related to portal hypertension.

9.2 Preoperative Planning

The child should have adequate nutrition and weight gain to tolerate the major procedure and have good postoperative healing. This surgery is best performed in children more than 1 year of age, who can sit and eat, with a weight of about 10 kg and a haemoglobin level of at least 10 g/dL. It is not unusual to perform oesophageal replacement in much younger and smaller children, however, including newborns if necessary.

The colon should be prepared with polyethylene glycol solution taken orally at the rate of 25 mL/kg per hour for 4 h, followed by clear water. The same may be repeated in an older child if the bowel preparation has not been clear. Alternatively, normal saline may be used at the same rate for 2 h with a repeat dose if required. The serum electrolytes should be monitored by a morning sample on the day of surgery.

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9.3 Operative Procedure

In replacing the oesophagus with the colon, attention should be given to the route, the surgical approach, the incision, and the length of the segment required. Generally, a combined approach is needed, with neck and abdominal incisions. A mediastinal route, unless severely fibrosed, is preferred to a substernal route, even though it is more than 1 cm longer. It is wise to mobilize the oesophagus with the neck dissection first, before opening the abdomen.

It should be kept in mind that the colon segment will shrink by about 10% after it has been segregated from the main bowel on its vascular pedicle. On the other hand, extra length will lead to redundancy and dilatation, with later stasis of food and recurrent vomiting.

Figures 9.1, 9.2, 9.3, 9.4, 9.5, 9.6, 9.7, 9.8 and 9.9 illustrate the steps in colonic replacement of the oesophagus.

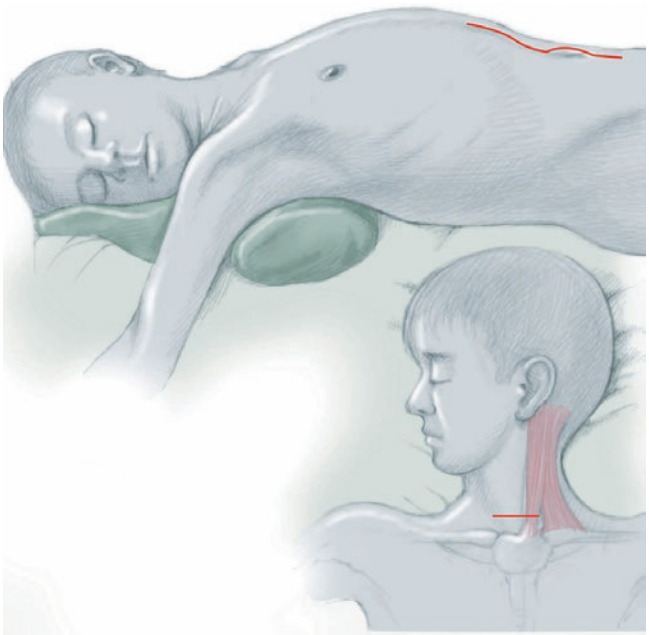

Fig. 9.1 The patient is placed in a supine position with a sand bag under the shoulder and the neck turned to the opposite side from the site of the cervical oesophagostomy. In cases of caustic ingestion, the authors prefer the neck to be turned towards the left. A soft red rubber catheter is placed into the oesophagus through the mouth to allow identification of the oesophagus during neck dissection. A right transverse supraclavicular incision is made for about 3–4 cm. If an oesophagostomy is present, it is mobilized with an elliptical incision, after applying stay sutures around it

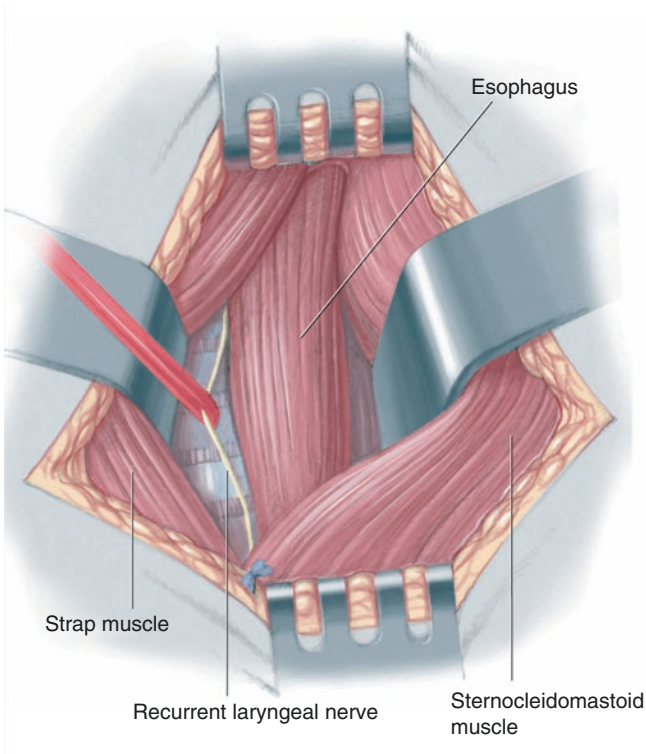

Fig. 9.2 After incising the skin, the subcutaneous tissue, and the platysma in the same plane, the cervical fascia is opened along the anterior border of the sternomastoid. As the deeper dissection continues, the strap muscles are either divided or retracted in the line of incision. The carotid sheath, with the internal jugular vein and common carotid artery, is gently retracted laterally. The recurrent laryngeal nerve is identified and retracted medially. The oesophagus is identified and dissected all around. The oesophagus is encircled with a tape and mobilized proximal to the stricture segment. If an oesophagostomy has been done earlier, it is mobilized for about 2–3 cm to prevent injury to the blood supply. In cases of caustic ingestion, dissection around the proximal and the distal oesophagus is mostly done bluntly, using the finger through the posterior mediastinum, combined with the use of scissors under vision to divide the fibrous tissue. The mobilized oesophagus is kept wrapped in a warm gauze and the attention is diverted to the abdomen

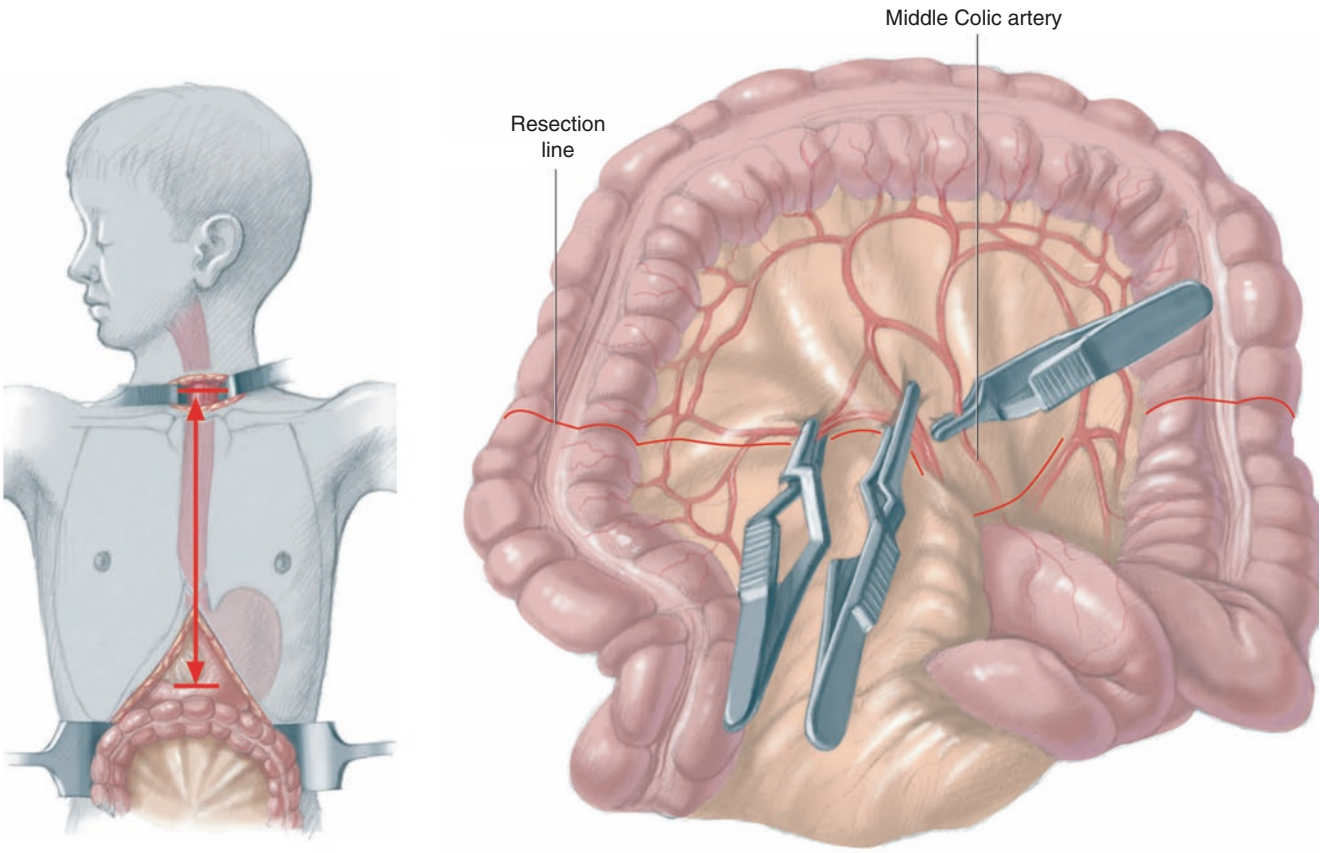

Fig. 9.3 A midline incision is made over the abdomen from the xiphisternum to the umbilicus and extended down around the umbilicus if need be. The transverse colon is mobilized from the ascending to the descending colon and exteriorized to assess the vascular supply. The gastrostomy, when present, is assessed for the site and the need for closure. The colonic graft is chosen based on the blood supply. Usually, it is based on the territory supplied by the upper left colic artery, more prominent compared with the right side in over 80% of cases. Division of the middle colic vessels will be required. Before dividing the major vessels, it is mandatory to use a bulldog clamp to verify adequate circulation to the proposed segment of colon. If there are any vascular anomalies, the graft can be based on the right or the middle colic arteries. The length of the colon segment should be equal to the distance between the antrum and the site of anastomosis in the neck

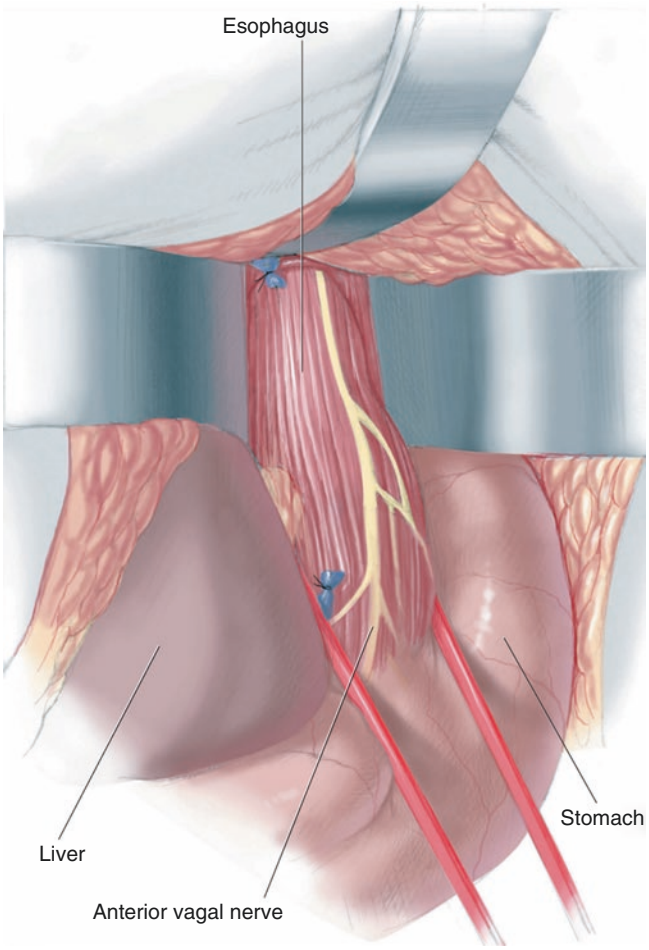

Fig. 9.4 The next step is to mobilize the lower part of the oesophagus in cases with caustic ingestion. The left triangular ligament of the liver is incised with diathermy, over a gauge sponge placed between the left lobe of the liver and the diaphragm. The oesophagus is then dissected all around at the hiatus after the division of the phreno-oesophageal ligament. The right and the left vagi are evident at this stage, and both are divided. Sometimes the posterior vagus can be saved with meticulous intrathoracic dissection. The oesophagus is encircled with a tape to facilitate mobilization. The hiatus is explored, utilizing two malleable retractors. Under direct vision, all oesophageal vessels are coagulated. Traction is obtained with the help of the tape, and the blunt dissection (usually with the finger) is kept very close to the oesophageal wall to avoid injury to surrounding structures. With blunt and sharp dissection, the oesophagus is freed as high as possible. Care is taken to avoid entering the pleural cavity. In case of inadvertent pleural injury, an intercostal tube drain is inserted. The blunt dissection is continued from above and below until the oesophagus is freed completely. It is essential to avoid aggressive dissection in the region of the aortic arch and to stay close to the oesophageal wall. If the oesophagus is badly adherent and impossible to mobilize safely, it should be left in situ without causing unnecessary injury to neighbouring structures. In that case, one can switch over to the substernal (retrosternal) route for the replacement

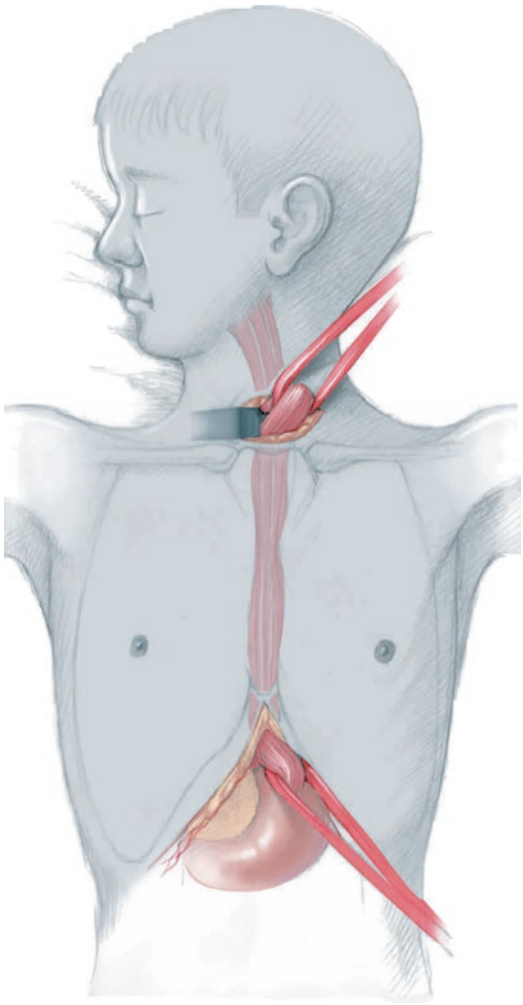

Fig. 9.5 Once the oesophagus has been freed from all its attachments, oesophagectomy is done by dividing the oesophagus in the neck and pulling it down to the abdomen. The gastric end is closed in two layers. A red rubber catheter with the silk suture attached is left in the mediastinum, to be used for guidance for the later passage of the colon through the hiatus

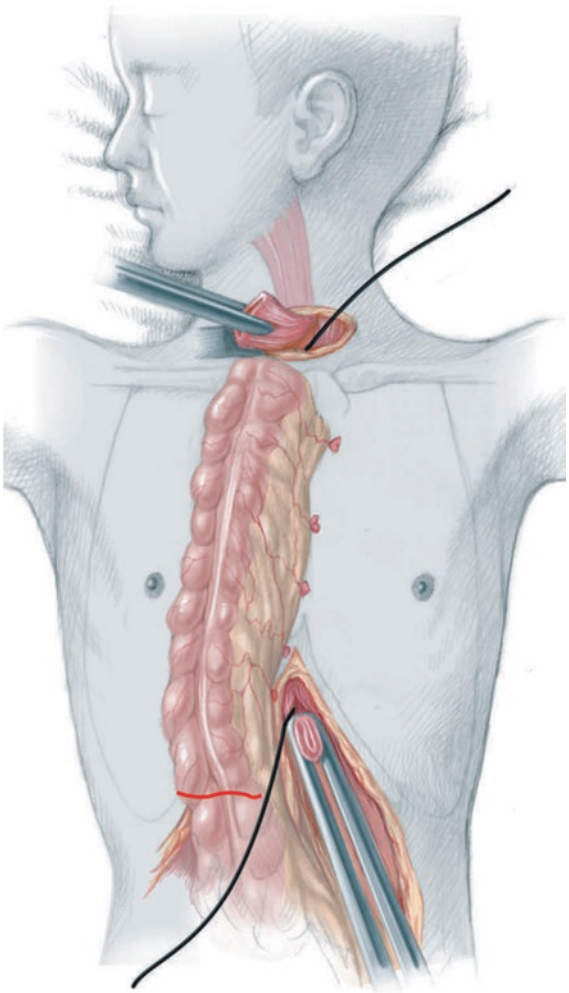

Fig. 9.6 The colon is reevaluated and the pulsation of the marginal artery is examined carefully. The exact measurement of the colon is reassessed after the oesophageal resection and the colon is divided between noncrushing intestinal clamps. The graft is washed with saline and diluted povidone iodine solution. After the wash, the colon graft is kept open on both sides without using any clamps

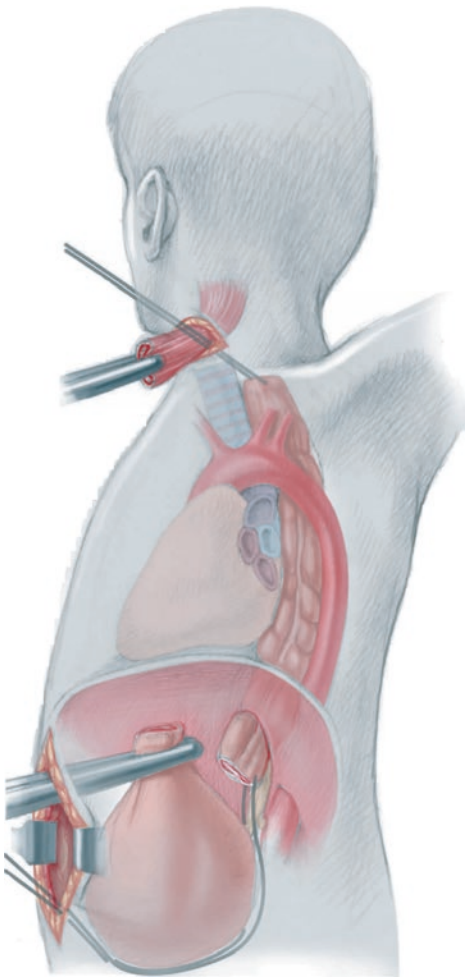

Fig. 9.7 A passage is created behind the stomach, through which the colon is taken up in an isoperistaltic manner. To facilitate passage through the chest, the silk suture previously present is sutured to the proximal end of the colon and pulled through the cervical incision until the colon is placed in the posterior mediastinum. Care should be taken to avoid causing any torsion of the pedicle or traction of the vessels. Viability of the graft is confirmed by noting the bleeding from its cervical end. If there is any redundancy, the redundant part is resected at the cervical and the gastric end, avoiding injury to the pedicle. If the oesophagus has not been resected and a colon bypass procedure is planned, then a retrosternal tunnel is made by blunt dissection, dividing the endothoracic fascia at the upper end from the neck incision after detaching the strap muscles at the suprasternal notch and at the lower end by separating the diaphragm and the rectus abdominis muscle slips from the xiphoid cartilage and the lower sternum. The tunnel is enlarged using fingers, remaining close to the under surface of the sternum and taking care not to injure the pleura. A red rubber catheter with a long silk suture is passed through the tunnel

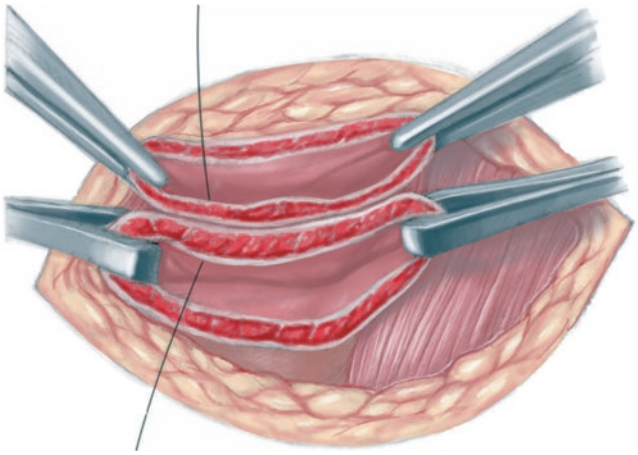

Fig. 9.8 The proximal oesophagus is examined to rule out any proximal stricture in the oesophagus or the pharynx. If there is not much disparity in size, an end-to-end, single-layer anastomosis is made using 4/0 absorbable (PDS or Vicryl) sutures. If the colonic end is slightly bigger, the oesophagus is spatulated posteriorly to accommodate the large size of the colon for the anastomosis. If the oesophagus is markedly smaller in diameter than the colon, an end-to-side oesophagocolic anastomosis is made, with closure of the colonic stump. Fixation of the colon to the neck muscles is done to avoid traction on the neoanastomosis. Suturing of the strap muscles is important to support the neck during swallowing. The wound is closed in layers, leaving a drain in place. In cases of caustic pharyngeal strictures, an end-to-side pharyngocolic anastomosis to the wall of the pharynx is created. First, a hockey-stick type of incision should extend up from the lower neck to the angle of the mandible. Then the dissection should go deep to the wall of the pharynx, which is opened on stay sutures posterior to the thyroid cartilage. A healthy mucous membrane should be available for the pharyngolic anastomosis. The colonic graft should be long enough to reach the pharynx. A wide, single-layer, end-to-side anastomosis is made without any tension. A nasogastric tube is left as a splint for 7–10 days

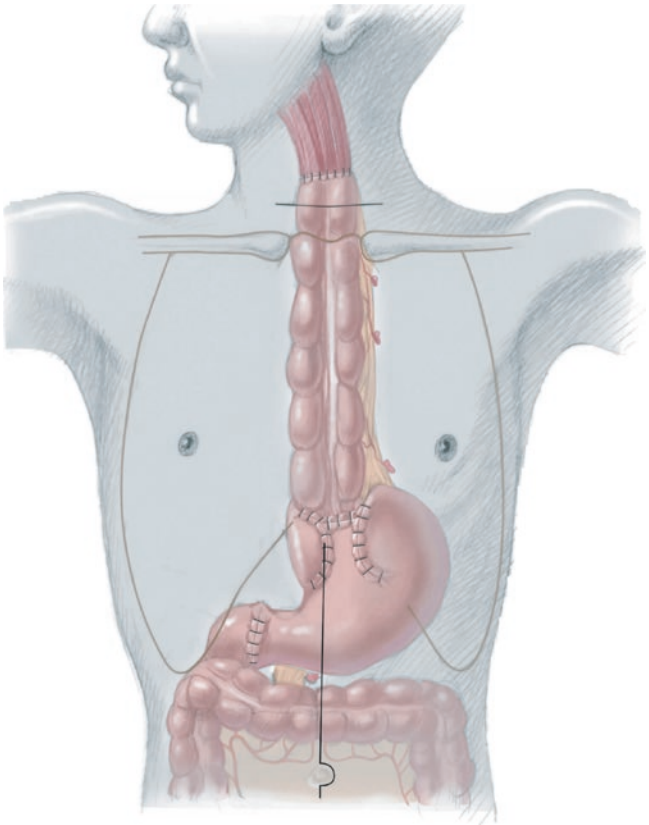

Fig. 9.9 In the abdomen, the gastrocolic anastomosis is performed in two layers, usually near the cardia or to the anterior wall of the stomach proximal to the antrum. The pedicle of the colon should always be placed behind the stomach, never in front of it, to avoid a mechanical gastric outlet obstruction. The colon should be positioned correctly to avoid hinging by the liver edge. The colon should be fixed to the edges of the diaphragm at the lower end of the tunnel in cases of retrosternal colon and to the edge of the hiatus if the posterior mediastinal route is used for the colonic replacement. Pyloroplasty is done in all cases if the oesophagectomy has been done, as both the vagi are usually sacrificed in the mobilization process. It is performed as a Heineke-Mikulicz type with a single-layer anastomosis. Finally, the colocolic anastomosis is performed to establish bowel continuity. Care should also be taken to close the window in the mesentery after the colonic resection. Gastrostomy or a feeding jejunostomy is optional and is considered only if a prolonged ileus is anticipated or the anastomosis has been precarious. The abdomen is closed in layers with a mediastinal drain

9.4 Postoperative Care

The drains are removed after 4–5 days, once the output has stopped. Patients can be kept on IV fluids or total parenteral nutrition, or can be fed by the gastrostomy or jejunostomy for 7–10 days. A contrast study is performed, and if there is no leakage, feeding is started. Any leaking usually stops within a week or so. Gastrostomy, if performed, is removed about 3 months after surgery. In cases with proximal anastomotic strictures, dilation may be done; if that is unsuccessful, surgical revision of the colo-oesophageal anastomosis is planned.

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Gastric Transposition for Oesophageal Replacement

10

Lewis Spitz and Arnold Coran

10.1 Introduction

Every attempt should be made to retain the child's own oesophagus, but this aim cannot be achieved in some circumstances:

- *Oesophageal atresia*, particularly very-long-gap pure atresia, where delayed primary anastomosis has failed, or complicated oesophageal atresia, when a primary repair has disrupted the oesophagus and a cervical oesophagostomy has been established
- *Caustic oesophageal damage* that fails to respond to dilatation
- *Prolonged foreign body impaction* causing injury to the oesophagus
- *Tumours of the oesophagus* such as diffuse leiomatosis or inflammatory pseudotumour
- *Motility disorders*

There are four recognized methods of oesophageal substitution:

- Colon interposition
- Gastric tube oesophagoplasty
- Jejunal interposition
- Gastric transposition

Gastric transposition has been our procedure of choice for oesophageal replacement for over 20 years. It has the following advantages:

- The stomach has an excellent blood supply.
- Adequate length to reach the cervical region can usually be achieved.
- The procedure involves a single anastomosis.
- The leak and stricture rates are relatively low.
- The procedure itself is simple to perform.

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10.2 Operative Procedure

The surgeon should be capable of performing the various alternative methods of oesophageal replacement. Bowel preparation is recommended, to ensure an empty colon in the event that the stomach is unavailable for the transposition procedure.

Figures 10.1, 10.2, 10.3, 10.4, 10.5, 10.6, 10.7 and 10.8 illustrate the technique of gastric transposition for oesophageal replacement.

A feeding jejunostomy is highly recommended for infants with oesophageal atresia who have not previously been established on oral feeding. In addition to the usual postoperative management following any major procedure, it has been our practice to electively paralyse and mechanically ventilate our patients for a minimum of 48–72 h after the surgery.

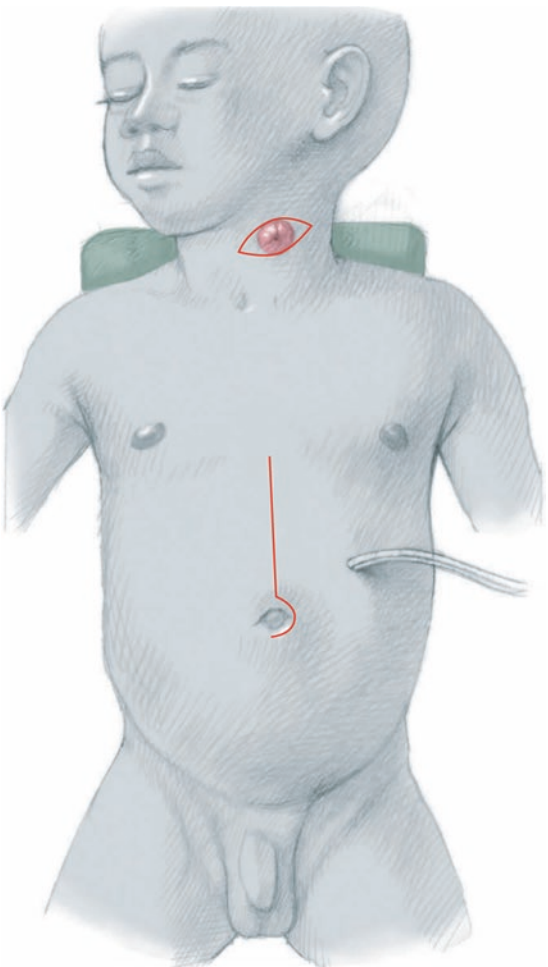

Fig. 10.1 A midline upper abdominal incision or a left subcostal incision is made. An elliptical incision around the cervical oesophagostomy or, alternatively, a right or left low transverse cervical incision is made to expose the cervical oesophagus. A lateral thoracotomy may be required if the surgeon encounters any difficulty in mobilizing the thoracic oesophagus, which may have been damaged by caustic oesophagitis or repeated attempts to retain the child's own oesophagus

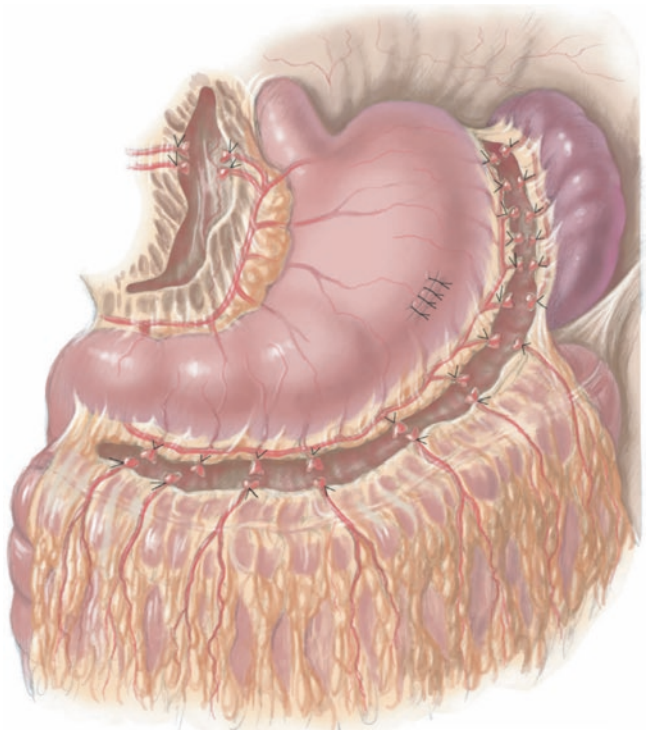

Fig. 10.2 The stomach is exposed, the gastrostomy site is taken down, and the defect in the stomach is closed. The greater and lesser curvatures of the stomach are mobilized, preserving the integrity of the right gastroepiploic and right gastric arcades. The mobilization of the stomach continues proximally by dividing the short gastric vessels between the fundus of the stomach and the spleen and by ligating and dividing the left gastric artery and vein

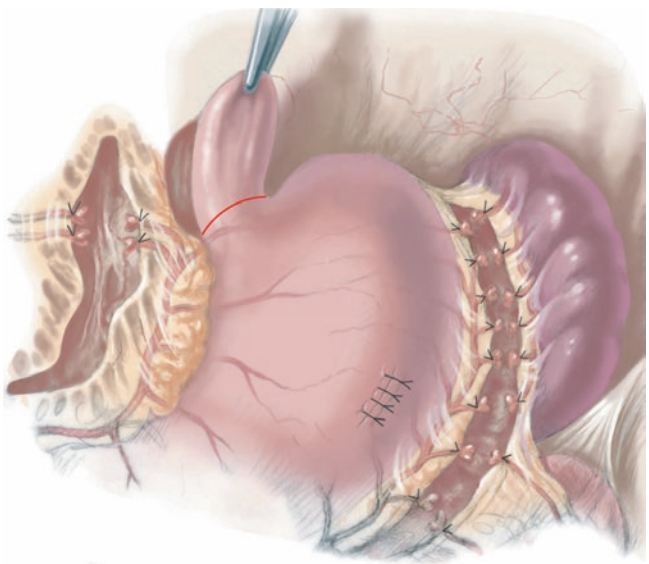

Fig. 10.3 The stump of the distal oesophagus (in the case of long-gap atresia) is mobilized from the posterior mediastinum by dividing the phreno-oesophageal membrane and dissecting out the oesophagus. The anterior and posterior vagal nerves are divided. The oesophagus is divided at the oesophago-gastric junction (*red line*) and the defect in the stomach is repaired

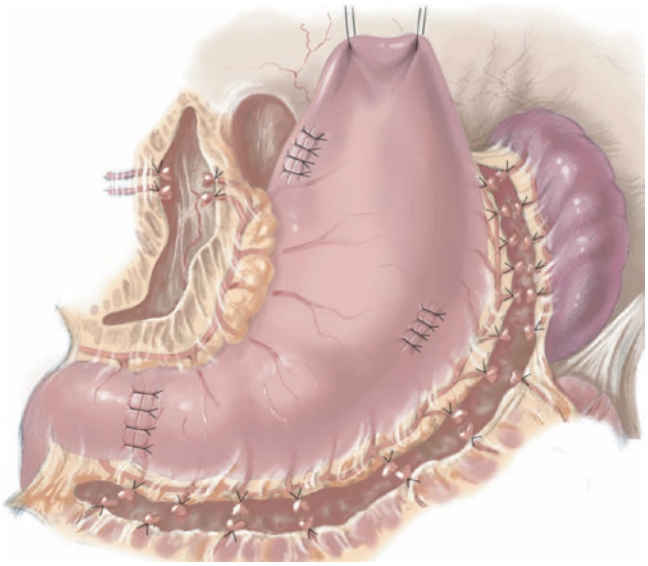

Fig. 10.4 A pyloroplasty or pyloromyotomy is performed. The sutured gastrostomy site and the closed-off gastro-oesophageal junction are shown. The highest point on the stomach and the place for the oesophago-gastric anastomosis is the top of the fundus. Two sutures of different materials are placed in the fundus. The orientation of these sutures is used to ensure that rotation of the stomach does not occur while it is being pulled up into the neck. Alternatively, a 32 Fr stiff chest tube can be sewn to the fundus with two sutures. It is used to identify the orientation of the stomach and to draw the stomach fundus up into the neck

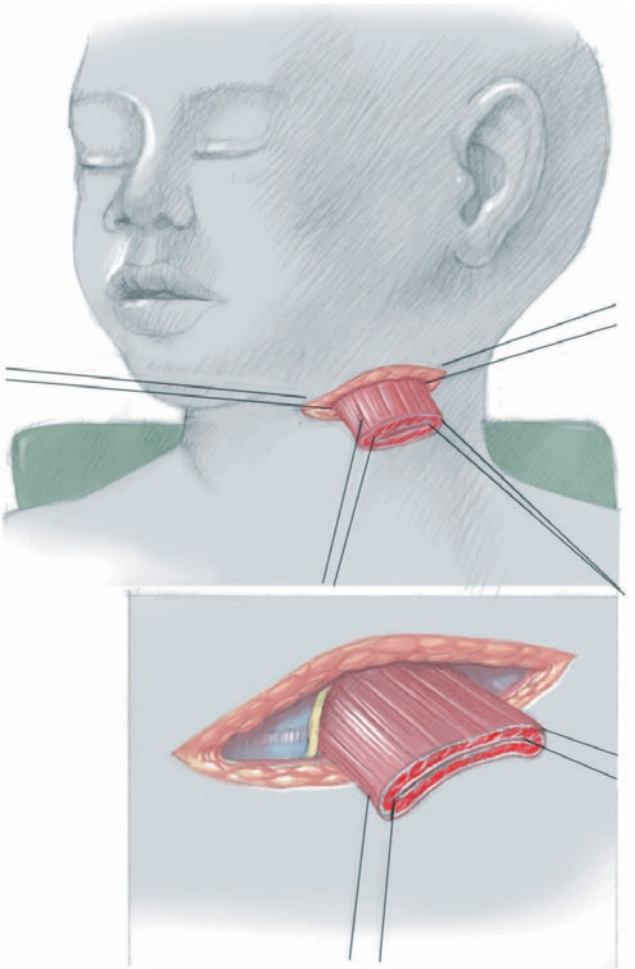

Fig. 10.5 Via the cervical incision, the full thickness of the oesophagus is mobilized. It is easy to enter into the submucosal plane during the dissection, but doing so should be avoided, as the vascularity of the oesophagus will be impaired. The recurrent laryngeal nerves must be preserved during the mobilization procedure

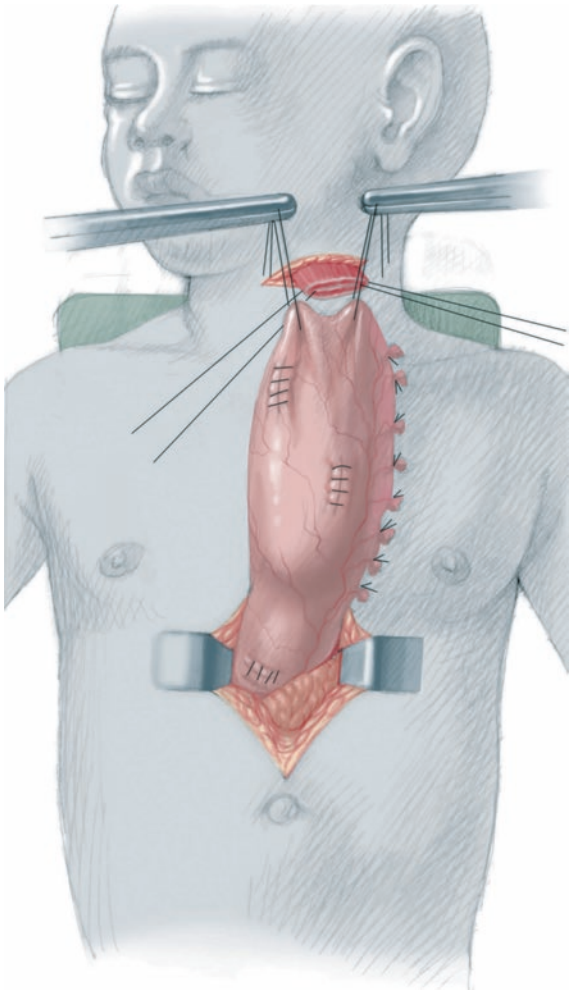

Fig. 10.6 Using the "stay-sutures" as guides, the stomach is pulled up through the hiatus in the diaphragm and through the posterior mediastinal tunnel until the fundus appears at the cervical incision. The transposition should be smooth and under no tension, and the stay-sutures should be correctly oriented to avoid twisting of the stomach in the posterior mediastinum

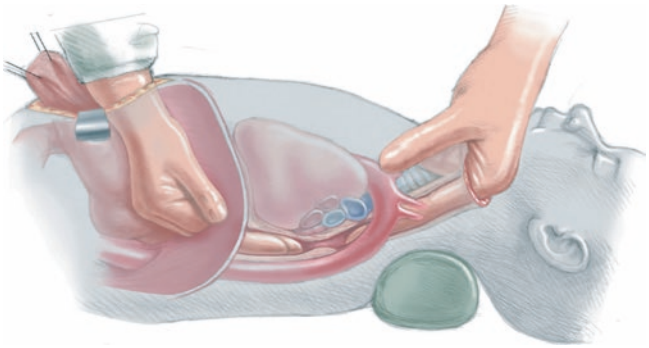

Fig. 10.7 The plane of dissection for the mediastinal tunnel is directly anterior to the prevertebral fascia. From above the dissection, proceed immediately posterior to the trachea and caudally into the posterior mediastinum. From below, through a widened hiatus, dissection is carried out under vision in the prevertebral space behind the heart. The tunnel is completed from above and below by gentle digital dissection in the posterior mediastinum. If any problems are encountered in creating the posterior mediastinum tunnel by blunt finger dissection, it is advisable to perform a lateral transpleural thoracotomy and complete the dissection under direct view. This approach is also essential to remove a scarred oesophagus or a tumour of the oesophagus

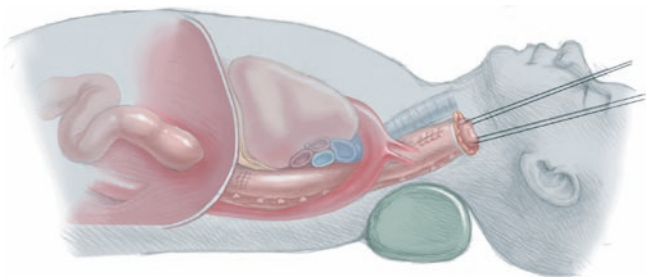

Fig. 10.8 The anastomosis between the end of the cervical oesophagus and the top of the fundus of the stomach is fashioned using a single layer of 5/0 or 6/0 sutures, taking the full thickness of the walls of the oesophagus and the stomach. Before completing the anterior wall of the anastomosis, a size 10F–12F nasogastric tube is passed, with the tip in the intrathoracic stomach. The wounds are closed, with a soft rubber drain at the cervical incision

10.3 Complications

In more than 200 cases by Lewis Spitz and over 190 cases by Arnold Coran, the mortality of this procedure was about 5%. The morbidity has been significant, including these problems:

- Anastomotic leak (12%–20%)
- Anastomotic stricture (20%–30%)
- Swallowing problems (30%)
- Delayed gastric emptying (5%–9%)
- Complications with the jejunal feeding tube (4%)
- Dumping syndrome (1%–3%)

Most of the children prefer to take small, frequent meals, although a normal eating pattern is generally established in older children. Many of the patients (particularly children who are born with oesophageal atresia) grow at a slower rate than normal and are in the lower half of the growth charts for both weight and height.

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Thoracoscopy

11

Bethany J. Slater and Steven S. Rothenberg

11.1 Introduction

Thoracoscopy is a technique that has been in use since the early 1900s but has undergone an exponential increase in popularity and growth over the past two decades. Until the late 1980s, most thoracoscopies were purely diagnostic. It wasn't until the early 1990s, with the dramatic revolution in technology associated with laparoscopic surgery in adults, that more advanced diagnostic and therapeutic procedures have been performed in children. The development of high definition cameras, superior optics, and smaller instrumentation has enabled pediatric surgeons to perform even the most complicated thoracic procedure thoracoscopically. Currently, most of the operations that have been classically performed through a formal thoracotomy can now be performed in a video-assisted fashion using a number of small incisions. The term *VATS* is often used and stands for video-assisted thoracoscopic surgery. This technique provides excellent visualization of the relevant anatomy and pathology, and drastically reduces the pain, recovery, and long-term morbidity associated with the procedures.

There are a wide variety of indications for thoracoscopic procedures in children, and the number continues to expand with advances and refinements in technology and technique. Currently, thoracoscopy is being used extensively for lung biopsy and wedge resection in cases of interstitial lung disease (ILD) and metastatic lesions. At many centers, more extensive pulmonary resections, including segmentectomy and lobectomy, are now being routinely performed for cavitary lesions, bullous disease, intralobar and extralobar sequestrations, congenital lobar emphysema (CLE), and cystic pulmonary adenomatoid malformations (CPAM). Thoracoscopy is also extremely useful in the evaluation and treatment of mediastinal masses. It provides excellent access for biopsy and resection of mediastinal structures such as lymph nodes, thymic lesions, cystic hygromas, foregut duplications, ganglioneuromas, and neuroblastomas. Other advanced intrathoracic procedures that have been described in children include decortication for empyema, closure of patent ductus arteriosus, division of vascular rings, repair of hiatal hernia and congenital diaphragmatic defects, esophageal myotomy for achalasia, thoracic sympathectomy for hyperhidrosis, anterior spinal fusion for severe scoliosis, and primary repair of esophageal atresia. Thoracoscopy has also gained popularity in the treatment of childhood and adolescent cancer.

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11.2 Anesthetic Considerations

A good working space is of paramount importance for thoracoscopy. In children, the working space is often very limited, so techniques have been developed to provide as much room as possible, primarily by collapsing the lung. One-lung ventilation by selective intubation of the main bronchus with a cuffed tube or by the use of a double-lumen tube is a good option but is applicable only in larger children (over 10 years of age or 30 kg) because the smallest available double-lumen tube is a 28 Fr. Another option is a bronchial blocker. This device contains an occluding balloon attached to a stylet on the side of the endotracheal tube. After intubation, this stylet is advanced into the bronchus to be occluded and the balloon is inflated (Fig. 11.1). Unfortunately, size is again a limiting factor, as the smallest blocker currently available is a 6.0 tube. In smaller patients, however, a Fogarty balloon catheter can be placed either adjacent to a standard endotracheal tube or through the lumen to achieve occlusion of the desired bronchus. For most infants and small children, a selective mainstem intubation of the contralateral bronchus with a standard uncuffed endotracheal tube is effective. This can usually be done blindly, without the use of a bronchoscope, simply by turning the head to the contralateral side. It is also important to use an endotracheal tube one half to one size smaller than the anesthesiologist would choose for a standard intubation, or the tube may not pass into the mainstem bronchus, especially on the left side.

At times, this technique will not lead to total collapse of the lung, as there may be some overflow ventilation if the endotracheal tube is not totally occlusive. This problem is overcome by the routine use of a low-flow (1 L/min), low-pressure (4 mm Hg) CO_2 infusion during the procedure to help keep the lung compressed. If adequate visualization is still not achieved, then the pressure and flow can be gradually turned up until adequate lung collapse is achieved. Pressures of 10–12 mm Hg usually can be tolerated without significant respiratory or hemodynamic consequences. This technique requires the use of a valved trocar rather than a nonvalved port. By using small tidal volumes, lower peak pressures, and a higher respiratory rate, enough lung collapse can be achieved to allow for adequate visualization. In neonates with tracheoesophageal fistula (TEF) or other congenital malformations, CO_2 alone can be used to deflate the lung. Once the lung is collapsed, it will stay that way until the anesthesiologist makes a conscious effort to re-expand it. The surface tension of the collapsed alveoli in the newborn keeps the lung collapsed without the use of excessive pressures. Close collaboration between surgeon and anesthesiologist is imperative to prevent problems with hypoxia and excessive hypercapnia, and to ensure the best chance at a successful procedure. It is very important for the surgeon to be patient enough to allow the body to seek a new equilibrium.

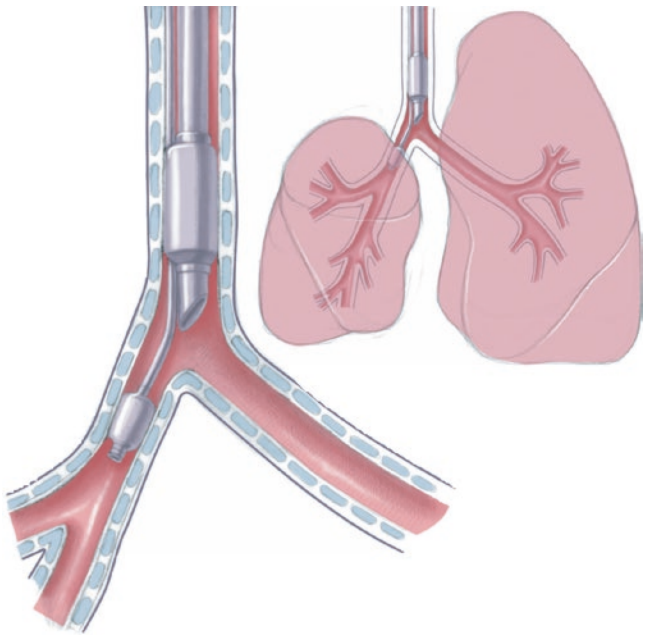

Fig. 11.1 One-lung ventilation with a bronchial blocker, collapsing one lung, can be used to increase the amount of working space for thoracoscopy in children

11.3 Positioning

The surgeon, the operative target area, and the screen should be in line. This means that the surgeon stands behind the back of the patient for anterior mediastinal surgery, and in front of the patient for posterior mediastinal surgery. In most cases, it is better to have the assistant on the same side of the table as the surgeon, to prevent working in a paradox (against the camera); when right-handed, the assistant is usually to the left of the surgeon. The scrub nurse usually stands on the opposite side (Fig. 11.2). For most thoracoscopic procedures it is advantageous to have two monitors, one on either side of the table. The monitors should be placed between the patient's shoulders and hips, depending on the site of the lesion.

Positioning of the patient is determined by the lesion and the type of procedure. Thoracoscopic procedures should be performed with the patient in a position that allows for the greatest access to the areas of interest and uses gravity to aid in keeping the uninvolved lung or other tissue out of the field of view. For routine lung biopsies or lung resections, the patient is placed in a standard lateral decubitus position, which provides for excellent visualization and access to all surfaces of the lung. This position is also the most beneficial set-up for decortications, pleurodesis, and other procedures in which the surgeon may need access to the entire pleural or lung surface. For anterior mediastinal surgery such as thymectomy, aortopexy, or biopsy, or for resection of anterior tumors or lymph nodes, a three-quarters posterolateral decubitus position should be chosen (Fig. 11.3). For posterior mediastinal surgery, including foregut duplications, esophageal atresia, and procedures involving the esophageal hiatus, a three-quarters anterolateral decubitus position should be used (Fig. 11.4). In addition, the patient can then be placed in Trendelenburg or reverse Trendelenburg as needed to help keep the lung out of the field of view. When all of the above measures are taken, usually no retractors are needed. If they are needed, they should be used with care, as they can easily damage the lung or other tissue that must be retracted.

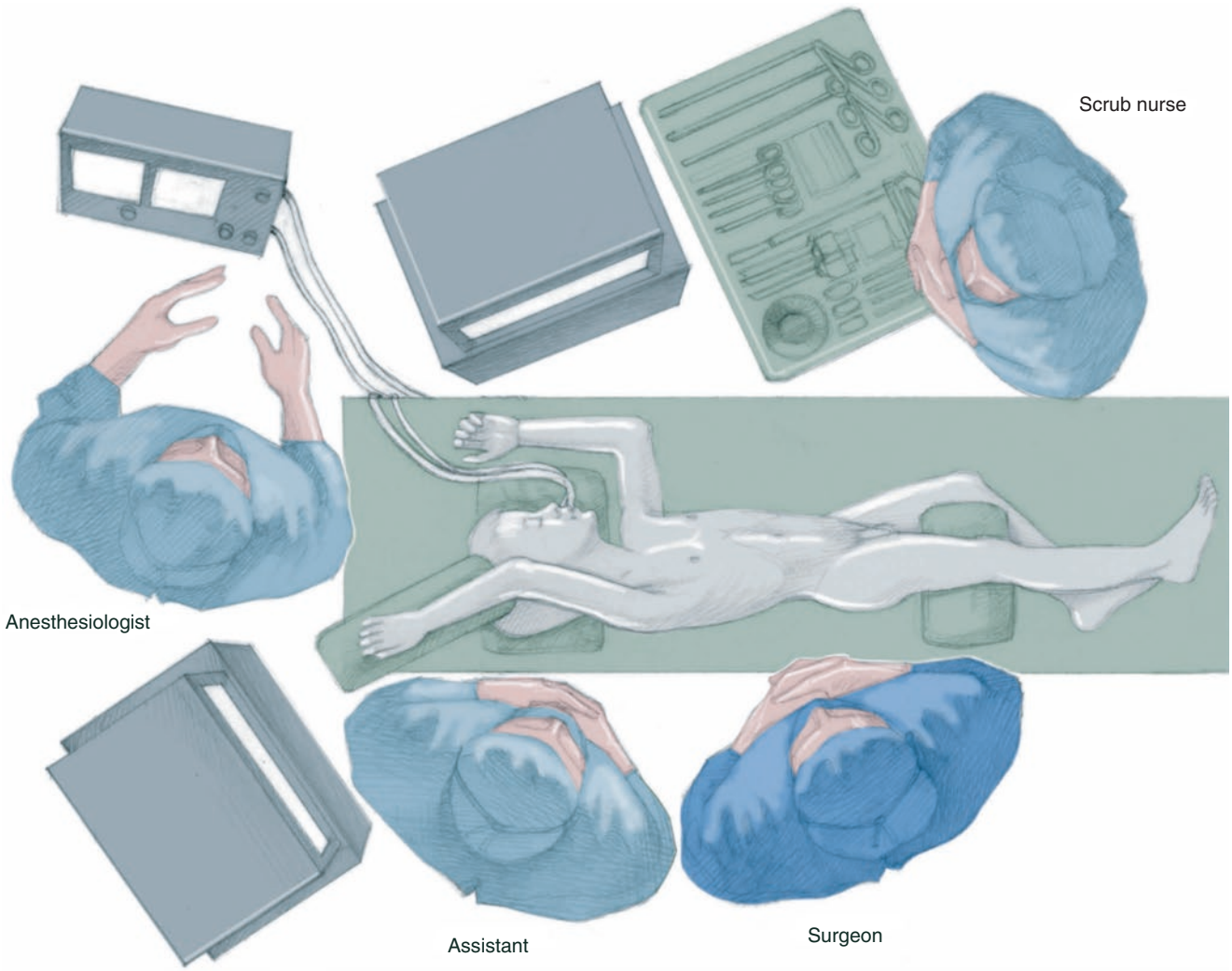

Fig. 11.2 For thoracoscopy, the surgeon, the operative target area, and the screen should be in line (*i.e.*, behind the back of the patient for anterior mediastinal surgery). The assistant usually stands on the same side of the table as the surgeon, with the scrub nurse on the opposite side. Two monitors are used

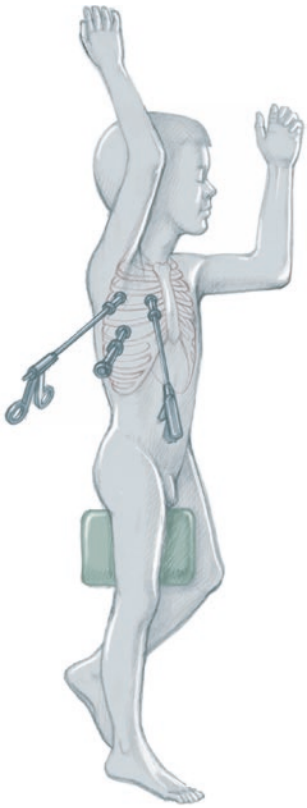

Fig. 11.3 Three-quarters posterolateral decubitus position for anterior mediastinal surgery

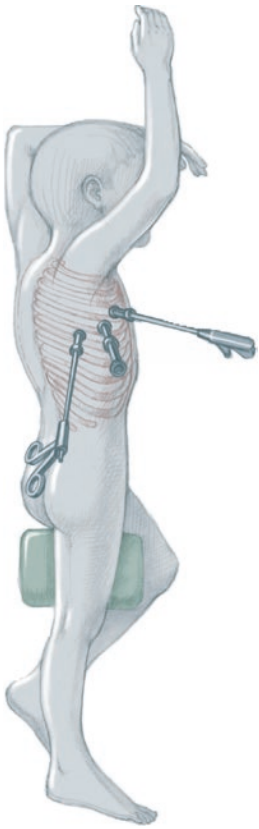

Fig. 11.4 Three-quarters anterolateral decubitus position for posterior mediastinal surgery

11.4 Trocar Placement

The cannulae can be inserted in a closed or open manner. When the closed technique is chosen in larger patients, a radially expandable cannula is usually used, to protect the intercostal neurovascular bundle from injury. A Veress needle with radially expandable sheath is punctured through the intercostal space at the desired place. Air is allowed to enter the chest through the needle so that the pleurae detach. The Veress needle is then removed and the sheath is left behind for dilatation with the cannula and blunt trocar. In the open technique, a small incision is made through the skin. Next, the wound is deepened just over the upper border of the rib until the pleural cavity is opened and air is sucked into the chest. A cannula with blunt trocar is then inserted. The hole in the thorax wall for the cannula should be as small as possible, so that the tissues fit snugly around the cannula in order to avoid CO_2 leakage. All secondary cannulae are inserted in the same method but under direct vision. Especially in small children, who have a rather thin body wall, cannulae have a tendency to slide further into the body cavity, thereby further limiting the working space, or to slide out. Using radially expandable cannulae may lessen this tendency. The best way to prevent this gliding in and out is to suture the trocar to the skin. In infants, standard 3 mm reusable trocars are used. A small piece of Silastic catheter can be placed around them, which is then sutured to the skin to avoid trocar migration.

Positioning of the trocars varies significantly depending on the procedure being performed and the site of the lesion. Thoughtful positioning of the trocars is more important than with laparoscopic surgery, because the chest wall is rigid, so the mobility of the instruments will be more restricted than in the abdomen. The most ergonomic position of the cannulae is triangular or V-shaped. The telescope cannula is inserted at the tip of the V, and the cannulae for the working instruments are positioned at the top of the limbs of the V. Ideally, the angle of the V should be about 90°. In general, the camera port should be placed slightly above and between the working ports to allow the surgeon to look down on the field of view. This will also minimize instrument dueling, which can be a significant problem in smaller infants. The trocar placement can be tentatively planned based on preoperative imaging studies and then modified once the initial trocar is placed.

11.5 Instrumentation

The equipment used for thoracoscopy is basically the same as that for laparoscopy. In general, 5-mm and 3-mm instrumentation is of adequate size, so 5-mm and smaller trocars can be used. The smaller the diameter of the telescope, the poorer the quality of the picture and the less light that can be transmitted. Telescopes with a diameter of 5 mm or 4 mm are of sufficient quality to be used for most endoscopic operations in children. The most commonly used scopes have an angle of 30°. In contrast to 0° scopes, angled scopes allow one to look around structures, which has great advantages. Most telescopes have a length of 33 cm. For procedures in smaller children and infants, it is also helpful to have smaller lenses, such as a short 30° scope that is 16–18 cm long and 3 or 4 mm in diameter, as well as specifically designed shorter instruments (18–20 cm). These tools enable the surgeon to perform much finer movements and dissection, allowing advanced procedures to be performed in infants as small as 1 kg. For most endoscopic surgical operations in children, instruments of 3–5 mm are appropriate. A good-quality digital camera and light source are also extremely important to allow for adequate visualization, especially when using smaller telescopes.

Disposable instrumentation that should be available includes hemostatic clips, endoloops (pretied ligatures), and an endoscopic linear stapler. Ligating loops can be used to seal leaking lung or to take a lung biopsy (Fig. 11.5). The linear stapler is an endoscopic version of the GIA used in

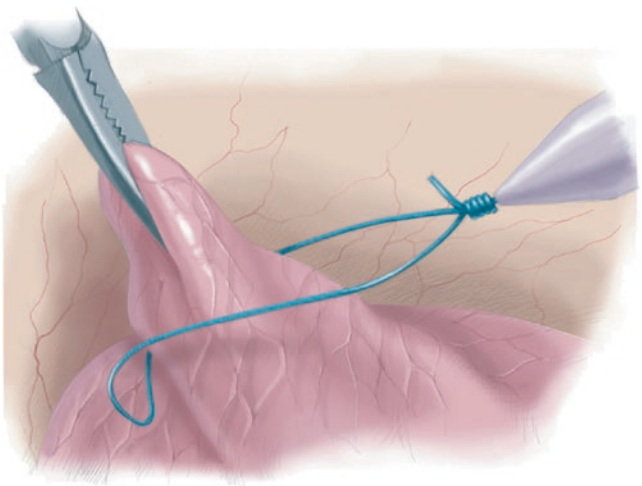

Fig. 11.5 A ligating loop can be used to seal leaking lung or to take a lung biopsy

open bowel surgery. It lays down four to eight rows of staples and divides the tissue between them, providing an airtight and watertight seal. This is an excellent tool for performing wedge resections of the lung, but until recently its size required placement of a 12-mm trocar and it could not be used in patients much under 10 kg because of the limited size of their thoracic cavity. New 5-mm staplers have recently been developed for use in smaller infants and children. There are also a number of energy sources available that provide hemostasis and divide tissue. These include monopolar and bipolar cautery, ultrasonic coagulating shears, and the LigaSure™ (Covidien), all of which can be helpful in difficult dissections. The LigaSure™ is a bipolar, high-frequency electrocautery instrument that can seal vessels with a diameter up to 7 mm, but its working jaw length is too large for infants. A 3-mm sealer device, which can seal vessels up to 5 mm in diameter, has recently been developed and is available for use in smaller children. It is also helpful to have one of the various tissue glues available for sealing lung and pleural surfaces.

A major problem in endoscopic suturing is the introduction of the needle. Most needles will not fit through a 3-mm trocar. In small children, the needle can be put directly through the body wall. Once the suturing has been finished, the needle must be directed back through the wall. Another possibility is to straighten the curved needle so that it will fit (together with the needle holder) through the cannula. The tying of the knot can be done extracorporeally with a knot pusher or intracorporeally.

Conclusion

Thoracoscopy has revolutionized surgery not only in adults but also in infants and children. Almost all operations that were classically performed through a thoracotomy can now be performed using VATS. Thoracoscopic surgery has clearly shown significant benefits over standard open thoracotomy in many cases. With continued improvement and miniaturization of the equipment, the procedures that can be performed and the advantages to the patient should continue to increase.

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Repair of Pectus Excavatum

12

Robert C. Shamberger

Pectus excavatum is a congenital deformity of the anterior chest wall. It consists of two primary elements. The first component is posterior depression of the body of the sternum, generally beginning at the level of the insertion of the second or third costal cartilages. The second component is posterior depression of the attached costal cartilages. This depression generally involves ribs 3 through 7 and sometimes will extend to the level of the second costal cartilage. In older teenagers, the posterior depression of the ribs will involve part of the osseous component as well as the cartilage component. This congenital deformity will be apparent within the first year of life in more than 90% of affected children. It occurs most often in families with a history of chest wall deformity, and has been estimated to have an incidence of 1 in 300 to 1 in 400 births.

The physiologic implications of pectus excavatum have been evaluated for the past four decades. It has been demonstrated that a "restrictive" pulmonary defect occurs in individuals with pectus excavatum. The total lung capacity and the vital capacity are below normative values. The values for an individual often do not fall out of the "normal range," but taken as a group, individuals with pectus excavatum do have decreased pulmonary volume compared with normals. The extent of this impairment varies depending upon the severity of the depression and the depth of the chest. Recent results from a multicenter study of patients with pectus excavatum demonstrated a relatively small decrease in their lung function preoperatively; the improvement after surgical correction was approximately 6–10% [1]. The second physiologic impairment that has been demonstrated is a decrease in the filling capacity of the heart (especially the right ventricle) produced by anterior compression from the depressed sternum. Studies dating back to those of Beiser et al. [2] have shown a decreased stroke volume, particularly in the upright position, associated with significant chest wall deformity. Though subsequent studies have shown variable results when using radioisotope techniques, this impairment is clearly one of the components of decreased cardiopulmonary function in patients with severe pectus excavatum. Workload studies have demonstrated that individuals with pectus excavatum develop symptoms of fatigue earlier than normal in gaited exercise protocols. Studies by Cahill in 1984 [3] and Peterson in 1985 [4] demonstrated the level of exercise tolerance increased after repair of the chest wall deformity.

Several considerations determine the patient's appropriateness for repair. These include the degree of psychologic distress created by the deformity, the extent to which cardiopulmonary symptoms impair physical activity, and the results of pulmonary function and physiologic exercise studies.

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12.1 Techniques for Repair

Techniques for repair of pectus excavatum have evolved significantly since it was first repaired in 1911. Modern approaches date to 1949, when Ravitch [5] first reported a technique that involved excision of all deformed costal cartilages with the perichondrium, and division of the xiphoid and the intercostal bundles from the sternum. A sternal osteotomy was created and the sternum was secured anteriorly with Kirschner wire fixation. This approach was modified in the 1950s by Baronofsky [6] and Welch [7], who stressed the need to preserve the perichondrial sheaths to allow optimal cartilage regeneration for durability of the repair. At about the same time, fixation with metallic struts anterior to the sternum was developed by Rehbein and Wernicke [8]. Retrosternal strut fixation was described by Adkins and Blades in 1961 [9]. Recent innovations for strut fixation have included the use of such materials as bioabsorbable struts, Marlex mesh, or Dacron vascular graft, but no evidence demonstrates that these are better than traditional metallic struts.

In 1998, Donald Nuss et al. [10] first described a method for repair of pectus excavatum utilizing a heavy metal strut to displace anteriorly the sternum and depressed costal cartilages without resection or remodeling of any of the costal cartilages. This technique is also known as the minimally invasive repair of pectus excavatum (MIRPE). The Nuss technique has been shown to be quite durable and results in excellent correction of the posterior depression of the sternum and costal cartilages. In 2002, Croitorou et al. [11] reported the use of this method in a larger and older cohort of 303 patients. The primary complication encountered was late bar displacement, requiring bar repositioning in 8.6% of cases, including 50% of those in whom a stabilizer was not used. Allergic reactions to the metal strut were also recognized, often with rash and erythema overlying the bar or with pleural effusions. This study also identified the importance of placing the bars and stabilizers in a subcutaneous position, not submuscularly, to avoid extra osseous bone formation around the strut. The occurrence of "over-correction" of the deformity was seen infrequently, primarily in children with connective tissue disorders (Marfan's and Ehler-Danlos syndromes). Kelly et al. [12] recently summarized a large experience with the MIRPE technique in 1215 patients and reviewed the changes made to the procedure. One bar was placed in 69% of their patients, and two bars in 30%. Good or excellent surgical outcome at the time of bar removal was seen in 95.8% of the patients. Complications with bar displacement decreased from 12% in the first decade to 1% in the second. Allergy to nickel was identified in 2.8% of patients, the vast majority prior to surgery. Wound infection occurred in 1.4% (17 patients), four of whom required surgical drainage. During the interval of this study, the median age at surgery went from 6 to 14 years. Modifications of the technique have included the routine use of unilateral or bilateral thoracoscopy; techniques to minimize the risk of dissection between the sternum and heart in patients with severe depressions, including elevation of the sternum manually through an infraxiphoid incision or first placing a more superior, transmediastinal tunnel and leaving the introducer in place to elevate the sternum while dissecting the lower tunnel; the use of titanium bars when a metal allergy is identified; using a bar 1 in. shorter than the measurement from right to left midaxillary line; more frequent use of two bars in patients with severe depressions or in older patients; and use of a metal stabilizer to decrease the risk of rotation of the bar.

12.2 Surgical Technique

This chapter presents both the current open technique with its modifications (which I use) and the innovative Nuss technique (MIRPE).

12.2.1 Open Repair

Figures 12.1, 12.2, 12.3, 12.4, 12.5, 12.6, 12.7 and 12.8 illustrate the operative procedure for the current open technique.

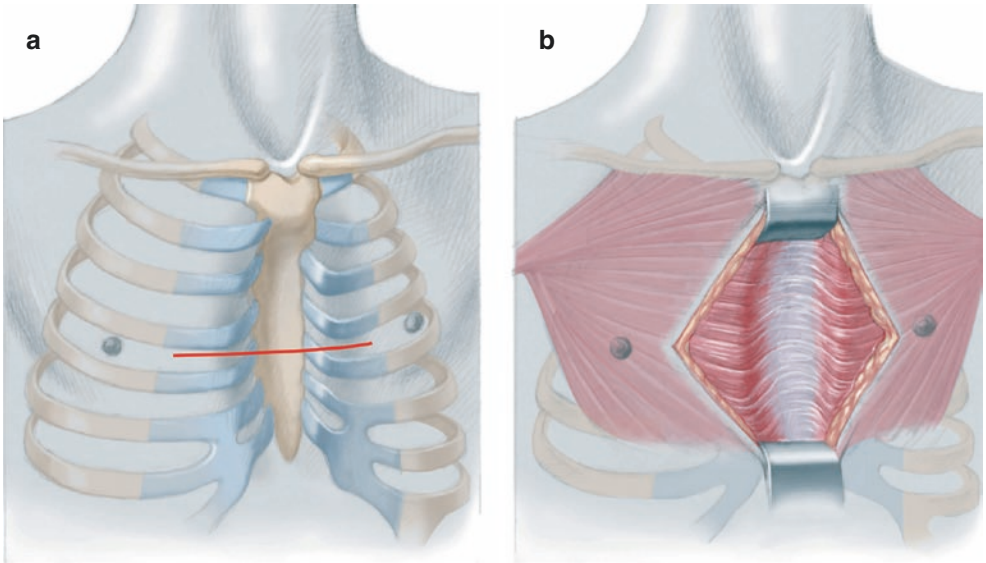

Fig. 12.1 A transverse skin crease incision is placed below and within the nipple lines. In females, it is of particular importance to see that this is placed in the future inframammary crease to avoid unsightly tethering of a scar between the two breasts. The skin flaps are then elevated superiorly to the level of the apex of the deformity and inferiorly to the tip of the xiphoid. The flaps are developed just anterior to the pectoral fascia to keep them well vascularized. The pectoral muscles are then elevated off the sternum, being cautious to keep intact all of the muscle and overlying fascia

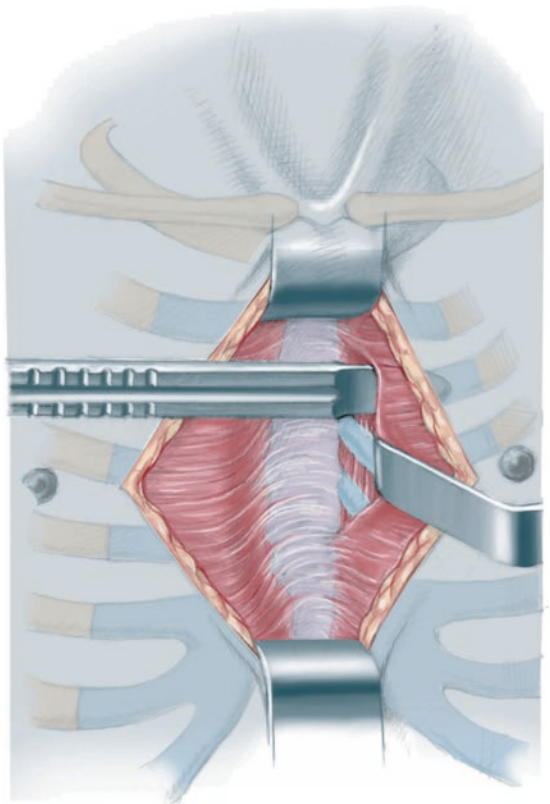

Fig. 12.2 To facilitate identification of the appropriate plane of dissection, the muscle is first elevated just anterior to one of the costal cartilages. When this plane is defined, an empty knife handle is then inserted anterior to the costal cartilage and passed laterally. It is then replaced with a right angle retractor to elevate the muscle anteriorly. This step is then repeated anterior to the next costal cartilage just above or below the first rib defined. Elevation of the muscle flap between the two right angle retractors facilitates identification of the correct plane of dissection. The origin of the salmon-colored pectoral muscles is divided with electrocautery, making certain to stay out of the intercostal bundles, which are covered with a glistening white fascia. Injury of the intercostal bundles can result in significant bleeding. The muscle flaps are mobilized laterally to the costochondral junction or to the lateral extent of the deformity. Generally, cartilages 3 through 7 are involved, but sometimes the second cartilage is also included

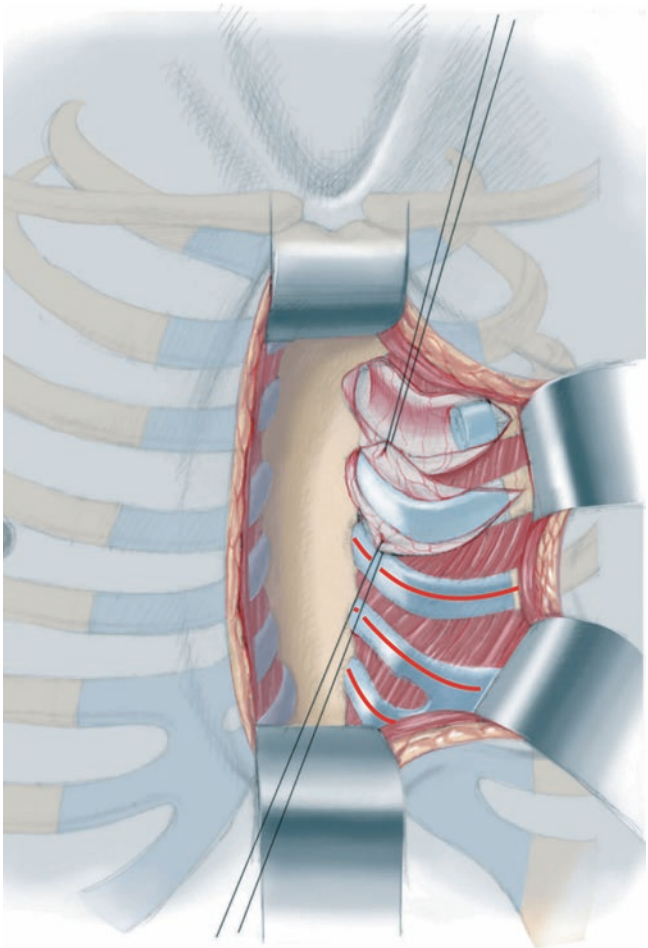

Fig. 12.3 Incisions are then placed through the perichondrial sheaths parallel with the axis of the cartilage. It is helpful to keep the incision on the flat anterior aspect of the rib. The perichondrial sheaths are dissected off the costal cartilage utilizing Welch perichondrial elevators (Codman and Shurtleff; Raynham, MA). Freeing the edge of the perichondrium from the medial aspect of the rib facilitates this process by providing better visualization of the posterior aspect of the cartilage. The cross-sectional shape of the ribs must be remembered. Ribs 2 and 3 are fairly flat, ribs 4 and 5 are round, and ribs 6 and 7 have a narrow width and greater depth

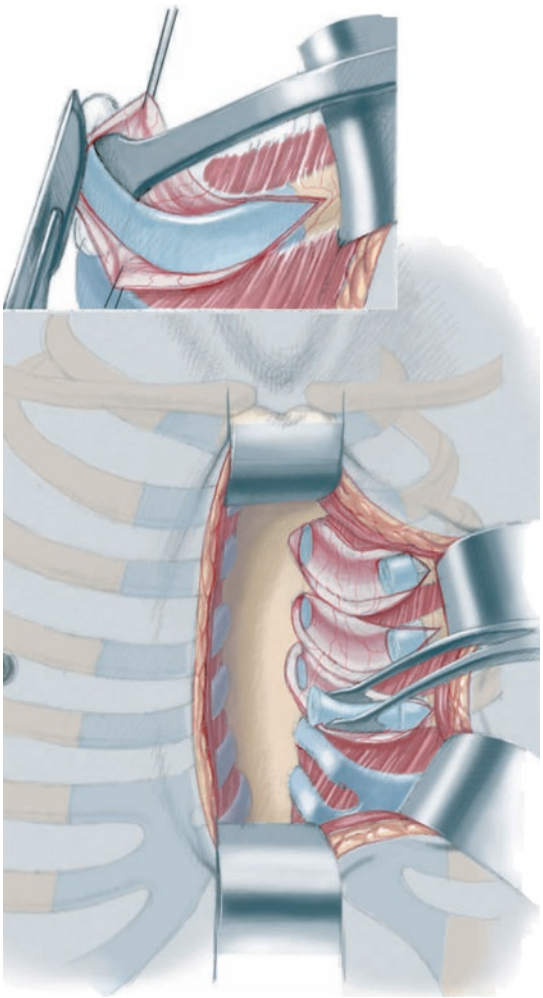

Fig. 12.4 The medial aspect of the cartilage is then incised from the sternum (*Inset*), with the posterior aspect protected by the Welch perichondrial elevator. Incising the cartilage directly adjacent to the sternum will also minimize the risk of injury to the internal mammary vessels, which are generally 1–1.5 cm lateral to the margin of the sternum. To minimize any impairment of subsequent growth of the ribs, 1–1.5 cm of the costal cartilage is preserved with the costochondral junction

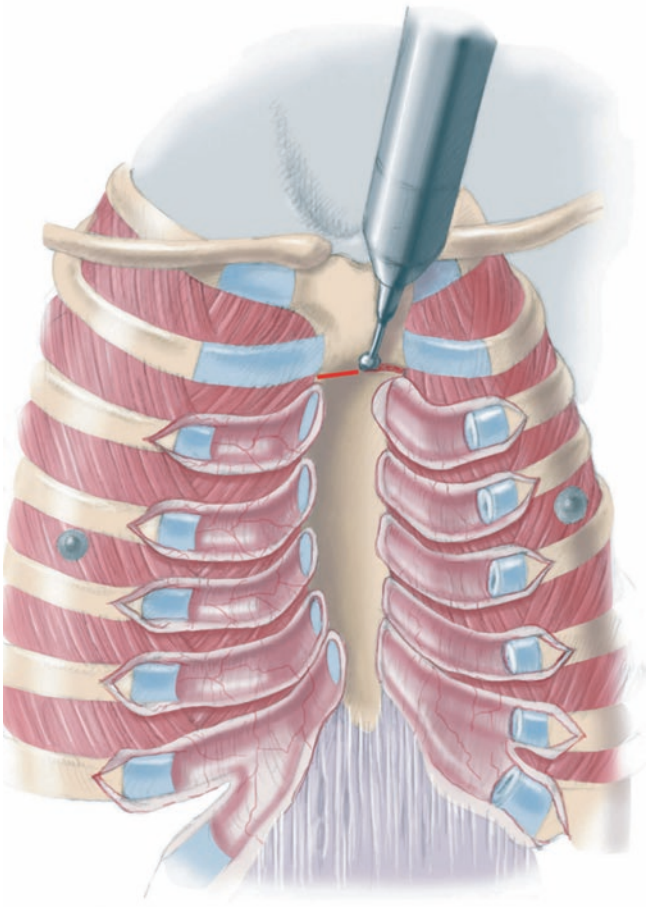

Fig. 12.5 The wedge osteotomy is then created on the anterior surface of the sternum at the apex of the deformity. I use a Hall air drill (Zimmer, Inc.; Warsaw, IN). The segment of bone is then mobilized using one of the wings of the perichondrial elevators, but without entirely dislodging it from the sternum. Leaving it partially in place will facilitate rapid healing of the fracture

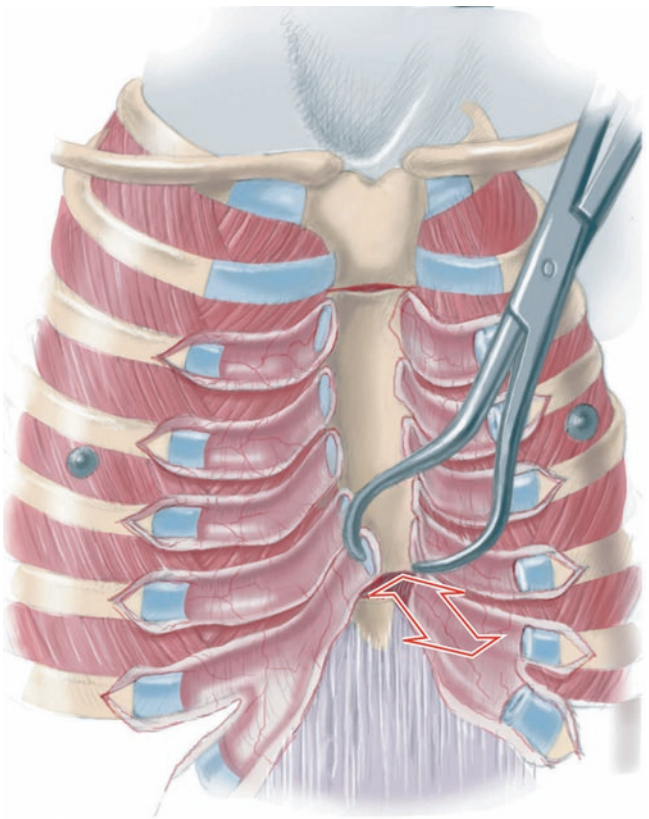

Fig. 12.6 The sternum is then elevated with a towel clip, and posterior pressure is applied to the upper portion of the sternum to fracture the posterior sternal plate. In the past, the xiphoid was divided along with the rectus muscle from the tip of the sternum, but I currently avoid this step to minimize the occurrence of an unsightly depression at the base of the sternum. Using a posterior sternal strut, it is also unnecessary to divide the lower perichondrial sheaths, as was done in the past. (This division of the lower perichondrial sheaths also contributed to the depression below the sternum.) If the xiphoid produces an unsightly protrusion when the sternum is in its corrected position, it can be divided from the sternum using a lateral approach with cautery. This technique avoids taking down the rectus attachment

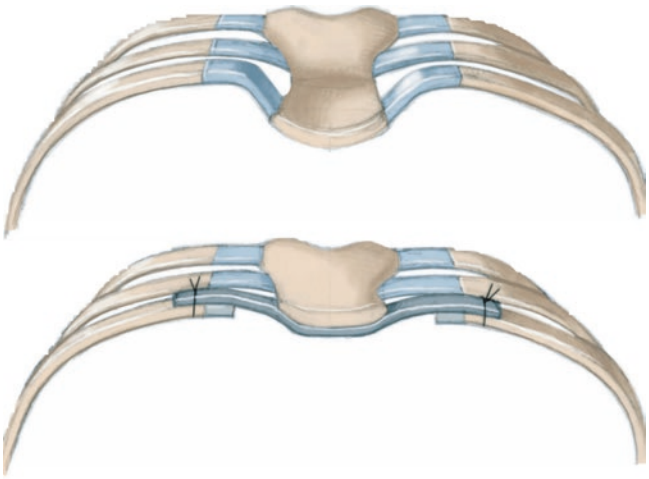

Fig. 12.7 This figure depicts the retrosternal strut, which is tunneled posterior to the sternum. The retrosternal tunnel is made by partially dividing one of the perichondrial sheaths directly adjacent to the sternum. A tunnel is then created posterior to the sternum with a Schnidt clamp, which is brought out directly adjacent to the sternum on the contralateral side to avoid injury to the internal mammary vessels. Before passing the strut behind the sternum, it is preformed so that there is a slight depression in which the sternum will sit, and the strut is curved somewhat posteriorly on each end to allow it to conform to the shape of the ribs and avoid any unsightly protrusions into the skin and the muscle. The Schnidt clamp is then used to draw the strut behind the sternum with the concave portion of the strut anterior. Once it is behind the sternum and in an appropriate position just anterior to the ribs on each side, it is rotated 180 degrees. It is important in this step to make certain that the strut is deep to the pectoral muscle flap to provide adequate soft tissue coverage over the strut. The strut is then secured to the anterior periosteum of the rib laterally with two heavy #0 absorbable sutures, which will secure the strut in position

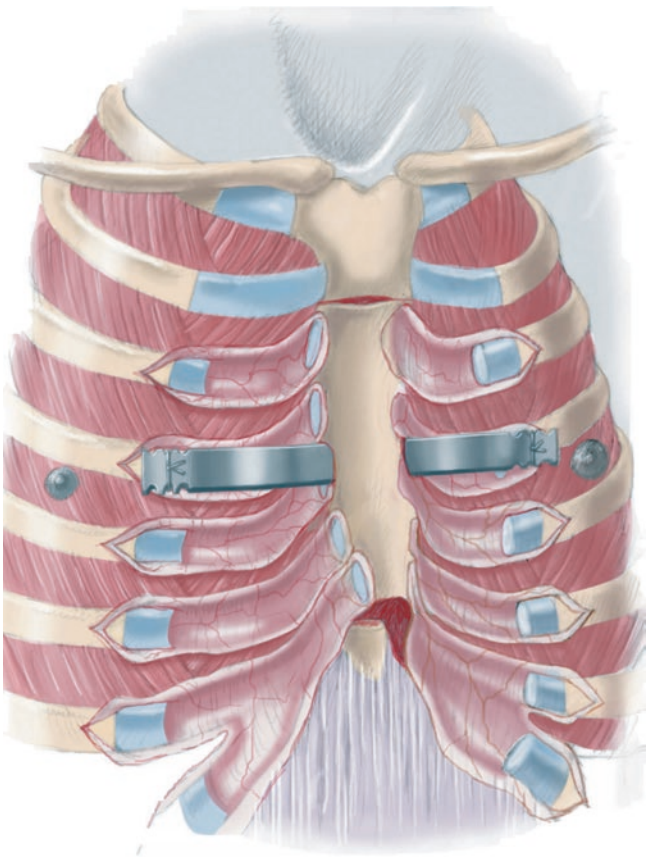

Fig. 12.8 This figure depicts the position of the retrosternal strut from an anterior perspective, with it secured to the ribs on each side. The pectoralis major muscle flaps are then approximated over the sternum. The flaps are advanced inferiorly to compensate for the fairly bare lower portion of the sternum, so that it is covered with soft tissue. At the inferior aspect, the flap is attached to the rectus muscle with interrupted absorbable sutures

12.2.2 The Nuss Minimally Invasive Repair of Pectus Excavatum

The innovative Nuss (MIRPE) technique is shown in Figs. 12.9, 12.10, 12.11, 12.12, 12.13 and 12.14.

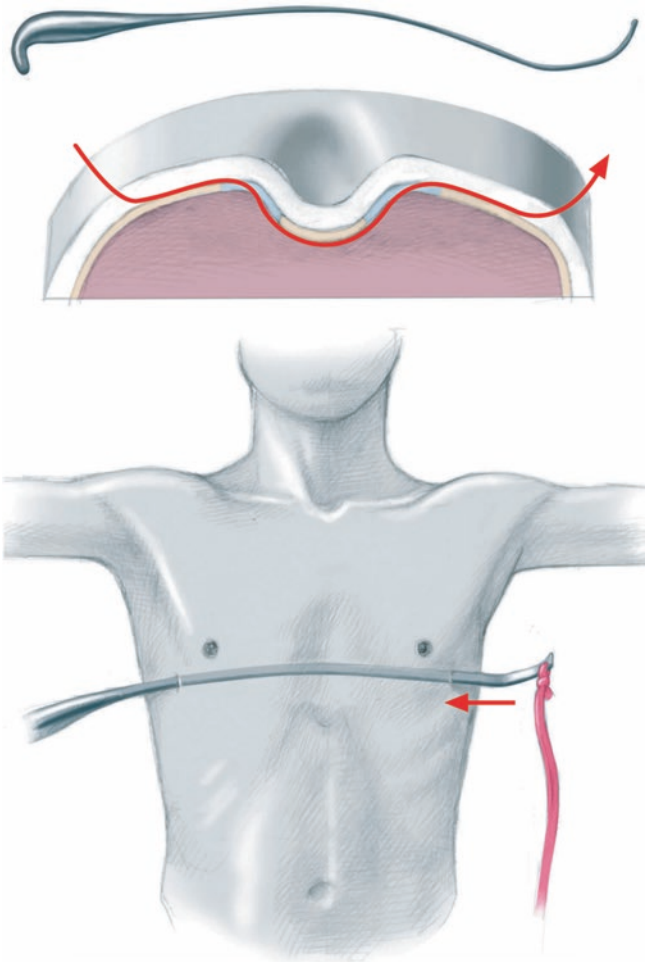

Fig. 12.9 Two incisions are made at the anterior axillary line at the level of maximal sternal depression. A pectus tunneler (Biomet Microfixation; Jacksonville, FL) or long clamp is then passed through one lateral incision along the chest wall and enters into the pleural cavity at the inner aspect of the pectus ridge. It is tunneled behind the sternum and anterior to the pericardium, and is brought out the contralateral side. The point of exit from the thorax is also aimed at the inner aspect of the pectus ridge. Thereafter, it is passed along the outside of the chest wall and out through the skin at the anterior axillary line. An umbilical tape is then grasped by the clamp or pectus tunneler and brought through the tunnel. Two tapes are often used, in case one breaks. Several adaptations have been utilized to minimize the risk of cardiac injury from this maneuver. The first adaptation, now widely used, involves a thoracoscope to monitor the passage of the tunneler behind the sternum. A second adaptation, less frequently used, is to make a small incision at the tip of the sternum, through which a bone hook can be inserted. The sternum is elevated anteriorly to broaden the retrosternal space as the clamp is passed across the chest

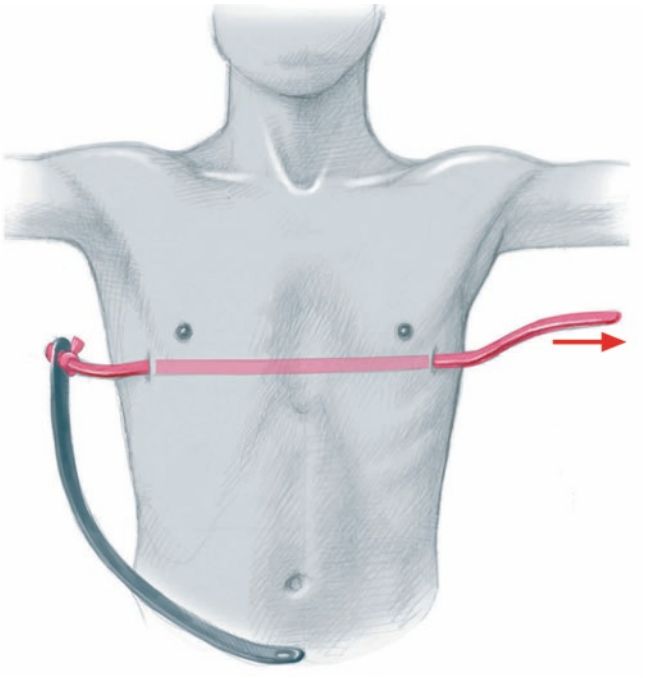

Fig. 12.10 The preformed strut (Biomet Microfixation; Jacksonville, FL), which has been pre-measured and bent to ensure that it fits the breadth of the patient's chest, is then brought through the chest and passed so that the concave surface is anterior

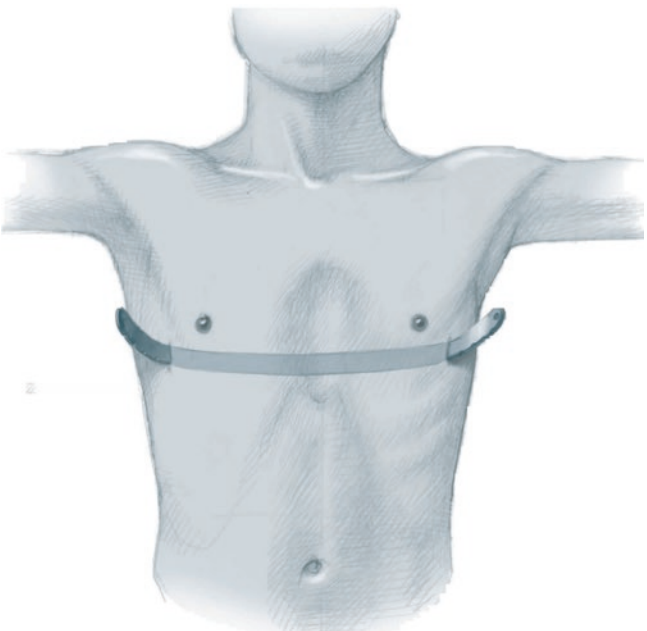

Fig. 12.11 Once the bar is in position, it is rotated 180 degrees with a special "Pectus Flipper" (Biomet Microfixation; Jacksonville, FL) to elevate the sternum and costal cartilages. During this maneuver, the skin and muscle flaps are elevated over the end of the bar so that the bar sits directly along the chest wall

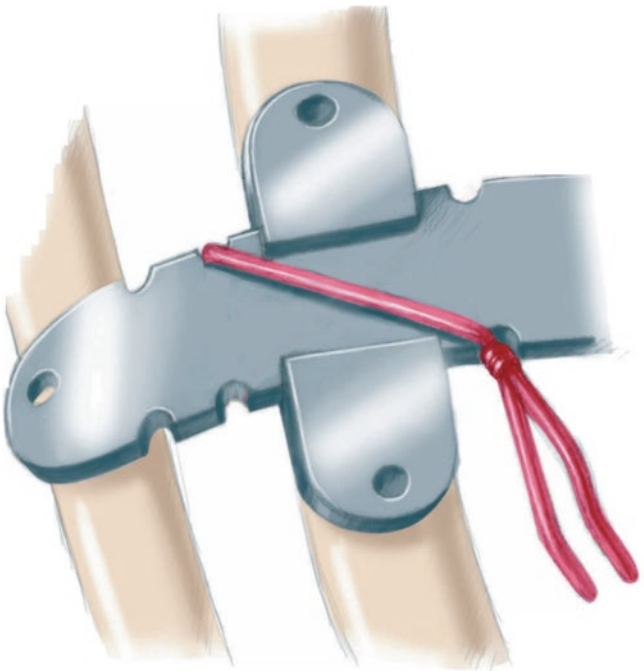

Fig. 12.12 When this procedure was initially performed, the most frequent complication was rotation of the pectus strut. To reduce this risk, a "stabilizer" may be attached to both sides of the strut with heavy #3 wire or suture. Once attached to the strut, it is then sutured to the soft tissues of the chest to provide secure fixation and prevent rotation of the bar and loss of correction of the deformity

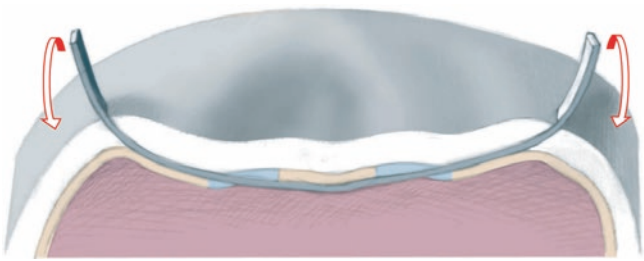

Fig. 12.13 The pectus strut in position prior to rotation

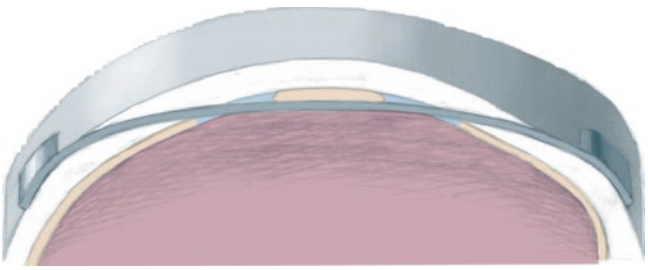

Fig. 12.14 The bar in the final position, displacing the sternum and costal cartilages anteriorly to correct the pectus excavatum deformity. The bar is electively removed in 2 to 3 years

12.3 Results

The overall results of repair of pectus excavatum should be excellent. The perioperative risks must be limited. The most significant complication is a major recurrence, which has been described in large series as occurring in 5–10% of patients. A limited pneumothorax requiring aspiration is infrequent and rarely is a pneumothorax of such magnitude to require a thoracostomy tube. Wound infection should be rare with the use of perioperative antibiotic coverage and protective coverage of the skin during the operative procedure to minimize any contamination by skin flora.

Long-term outcome of the Nuss procedure in teenagers is well documented. The most frequent complication described in early use of the minimally invasive procedure was rotation of the strut. Lateral stabilizers have significantly decreased the incidence of this complication. Other complications described include pneumothorax, pericarditis, and hemothorax. Complications unique to the minimal access procedure that have not occurred with the standard open technique include thoracic outlet syndrome and the rare occurrence of a carinate deformity after repair. An allergic reaction to the metal Lorenz struts has occurred in 1% of patients, who present with rashes along the area of the bar requiring replacement with bars composed of other alloys. Older patients seem to encounter significant pain equivalent to that of the open repair [1].

Conclusions

Both techniques appear to achieve excellent correction of the deformity [13, 14]. Complication rates of each technique were equivalent in a multi-institution study. Repair of pectus excavatum is important for children who are either psychologically distressed or physiologically impaired by their deformity.

Suggested Reading

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Pulmonary Malformations

13

Henry L. Chang and Keith T. Oldham

Though generally uncommon, congenital pulmonary malformations comprise a cluster of anomalies that frequently require surgical evaluation. Symptomatic lesions typically present in the prenatal period as fetal distress, in the neonate with respiratory embarrassment, or during childhood with recurrent pneumonia. An increasing number of pulmonary malformations are being diagnosed prenatally, leading to earlier involvement of the surgeon, with a wider array of therapeutic options available. Though varied in their pathology, the definitive treatment of these lesions is generally resection, and although the technology by which resection is done continues to change, the anatomy and fundamental surgical principles of resection have not. A brief review of the specifics of the more common malformations follows, with a description of a lobectomy highlighting the general principles of lung resection that follow.

13.1 Common Pulmonary Malformations

Congenital lobar overinflation (CLO), or congenital lobar emphysema, is characterized by air trapping and overdistension of one or more anatomically normal lobes. This distension causes compression of the adjacent parenchyma and can result in mediastinal shift with cardiorespiratory compromise. CLO is believed to result most commonly from structural deficiency or absence of supportive cartilage in the affected lobar bronchus, leading to expiratory collapse of the airway and impedance to expiratory flow. CLO is most often seen in the Caucasian population, with a male preponderance. It is most common in the left upper lobe (40–50%), with other sites affected less frequently: right middle lobe, 30–40%; right upper lobe, 20%; and lower lobes, 1%.

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Congenital cystic adenomatoid malformations (CCAM) are a group of cystic lobar hamartomatous lesions pathologically characterized by proliferation of terminal respiratory structures in an adenomatoid pattern. Though several classification systems have been proposed in the past, the current schema of macrocystic (cysts >5 mm) or microcystic (solid, or cysts <5 mm) is of particular clinical import, as described below. CCAMs show no racial preponderance or predilection for laterality, but they are slightly more common in males and in the lower lobes.

Prenatally diagnosed lesions may demonstrate spontaneous regression in up to 15% of fetal CCAMs with serial ultrasonographic examinations. Those that do not regress and threaten in utero hydrops or demise warrant fetal intervention with either maternal betamethasone or thoracoamniotic shunting for macrocystic lesions. Open fetal surgery and ex utero intrapartum treatment (EXIT procedures) for these lesions should be performed at highly specialized centers in cases of impending fetal demise.

Pulmonary sequestrations make up 10–30% of cystic bronchopulmonary foregut malformations. They can be intralobar (within the visceral pleura) or extralobar (invested by its own visceral pleura). In both types, however, there is no bronchial communication between the sequestrum and the tracheobronchial tree.

Intralobar sequestrations make up about 50–70% of the pulmonary sequestrations and most commonly involve the posterior and basal segments of the left lower lobe. The arterial supply is usually derived from aberrant branches of the descending thoracic aorta, although aberrant intercostal, brachiocephalic, or abdominal aortic vessels may be encountered. Venous drainage is usually via the associated pulmonary vein. Extralobar sequestrations are most commonly found in the left lower chest but may occur anywhere, with subdiaphragmatic locations having been reported. These sequestrations also derive arterial blood supply from the descending aorta, with up to 20% having an aberrant vessel traversing the diaphragm.

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https://doi.org/10.1007/978-3-662-56282-6_13

Bronchogenic cysts arise from the trachea, bronchus, or other conducting airways but have usually lost their connection with the parent structure. They are usually simple and contain mucus, but air-fluid levels and infection may be seen if there is continuity with the tracheobronchial tree. In contrast to sequestrations, bronchogenic cysts have a normal bronchial blood supply. Although bronchogenic cysts may reside anywhere in the respiratory tract, including paravertebral, para-oesophageal, subcarinal, and cervical presentations, the majority are found in the lung parenchyma or mediastinum.

13.2 Diagnosis and Surgical Planning

With improving technology and the increasing use of obstetric ultrasounds, many pulmonary malformations are detected during the prenatal period. Asymptomatic neonates are now being born with known pulmonary malformations that may not be visible on postnatal chest radiographs. Surgical planning usually requires further localization with either a chest CT scan with contrast or MRI, both of which provide excellent anatomic detail and demonstrate relationships to neighbouring structures.

Nonoperative observation of asymptomatic lesions may be done in some circumstances, but definitive treatment of congenital pulmonary malformations is resection of the affected lobe. The technique by which resection is accomplished is evolving. Experience with thoracoscopic lobectomy for congenital lesions is rapidly maturing, and most reports indicate advantages in terms of shorter hospital stays, diminished intensive care requirements, possibly better long-term outcomes in terms of chest wall development and fewer chest tube days, at the expense of longer operative times. More recently, there have been reports that "lung-sparing" resections utilizing anatomic resections at the segmental level are feasible and safe, with the theoretical advantage of preserving more native lung parenchyma. Long-term data on these limited resections are needed, however. Despite these advancements, it should be noted that lobectomy, whether via open thoracotomy or thoracoscopy, is very well tolerated in infants and remains the gold standard to which other techniques are compared.

13.3 Operative Procedures

Lung surgery in children is generally similar to that in adults except that the diminutive size, the associated lesions, and the unique pathologic entities require certain special considerations. Lobectomy can be performed by conventional thoracotomy or video-assisted thoracoscopy, as noted. As already noted, lobectomy is generally the current procedure of choice for the treatment of congenital lobar emphysema, CCAM, intralobar sequestrations, and some parenchymal lung cysts. Left upper lobectomy is described and illustrated here, but the principles are the same for any lobe resection.

13.3.1 Open Thoracotomy

For an open thoracotomy, the patient is positioned in the lateral decubitus position, with the upper arm extended and placed over the head (Fig. 13.1). Rolled towels and other positioning devices may be placed in order to optimize stabilization and exposure of the operative field. Optimal exposure is gained by transverse or oblique incision over the fourth or fifth intercostal space, below and lateral to the nipple, to avoid cosmetic and functional damage to the breast tissue. There should be some space between the tip of the scapula and the posterior extent of the incision. This space becomes important during closure of the muscle layers, especially if the incision must be extended posterolaterally. Underlying muscle and subcutaneous tissue is divided along the line of incision by electrocautery (Fig. 13.2). To limit postoperative morbidity, it is desirable and usually possible to employ a muscle-sparing approach, affording adequate exposure yet limiting or avoiding division of the serratus anterior and latissimus dorsi. The scapula is elevated off the chest wall by retractor to gain exposure, and palpation is used to count the ribs to the correct interspace. In most situations in infants, the highest palpable rib is the second. Generally, the fourth interspace is used for a lobectomy, although the fifth also can be used effectively.

The incision is then continued with electrocautery just superior to the lower rib of the selected intercostal space, to avoid damage to the neurovascular bundle that runs along the inferior border of each rib (Fig. 13.3). Care must be taken when entering the pleura to avoid injury to the lung parenchyma beneath. A rib spreader is then placed to facilitate retraction (Fig. 13.4). The incision may then be continued anteriorly or posteriorly from inside the chest if further exposure is needed.

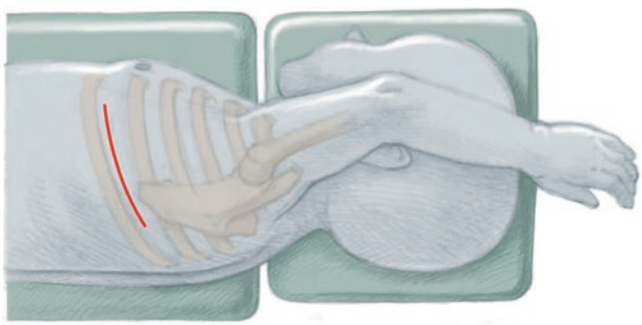

Fig. 13.1 Positioning for left upper lobectomy by open thoracotomy

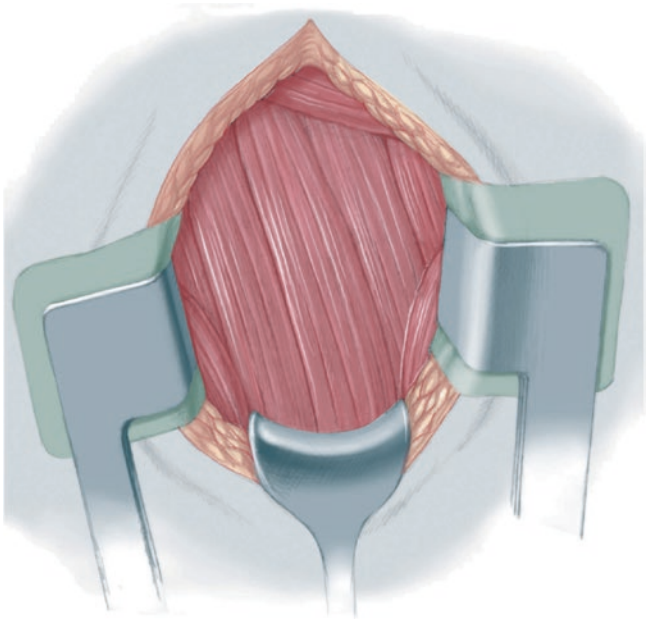

Fig. 13.2 Division of underlying muscle and subcutaneous tissue

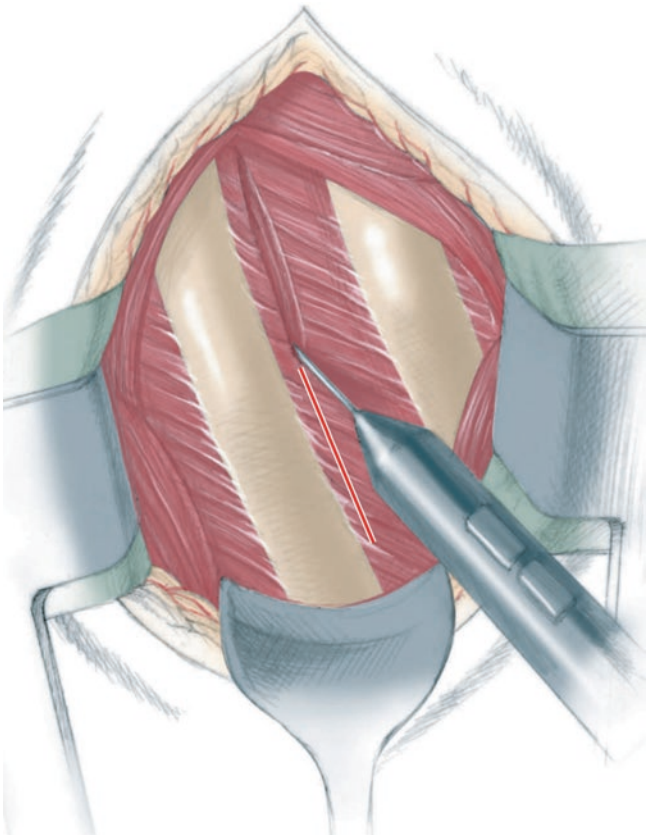

Fig. 13.3 Incision by electrocautery in the intercostal space

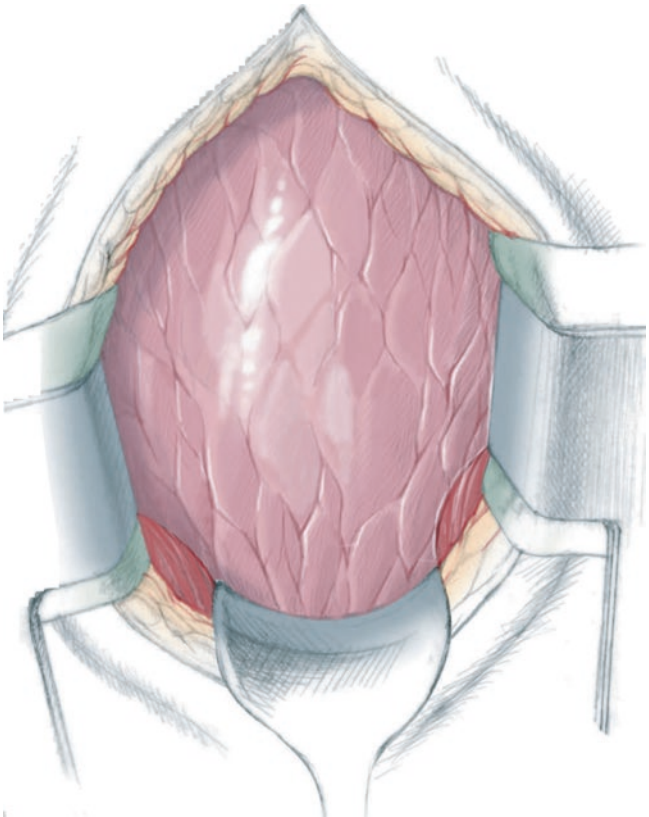

Fig. 13.4 Retraction exposing the lung parenchyma

Gentle lateral and inferior traction on the lobe exposes the hilum. The visceral pleura is carefully incised circumferentially, exposing the hilar structures (Fig. 13.5). Meticulous dissection reveals the left main pulmonary artery as it courses under the aortic arch and crosses the left upper lobe bronchus. Nearby structures to be noted are the left phrenic nerve anteromedially along the mediastinum, and the recurrent laryngeal nerve branching from the vagus under the aortic arch. A review of segmental anatomy of the lung describes four main arterial branches supplying the left upper lobe (Fig. 13.6), but this anatomy can vary. These branches are individually encircled, ligated, and divided, typically using heavy silk and double proximal ligatures. The bronchial blood supply travelling with the left upper lobe bronchus is likewise identified and ligated. Attention is then directed to the left upper lobe venous drainage (Fig. 13.7). Again, individual branches are circumferentially dissected and ligated, using the same approach as for the arterial circulation.

The bronchus is then clamped and divided. Closure of the bronchial stump with commercial surgical stapling devices is appropriate in older children, but size and other technical limitations make this undesirable in infants, where a simple sewn closure is best (Fig. 13.8). Air leaks may be identified for suture repair by filling the chest with warm saline coincident with inflation of the residual lobe by the anaesthesiologist. The inferior pulmonary ligament should be divided at this time to facilitate expansion of the left lower lobe, or it may be done early in the dissection to facilitate exposure. The superior and inferior pulmonary vein sometimes have a common stem outside the pericardium; if unrecognized and divided, this may necessitate

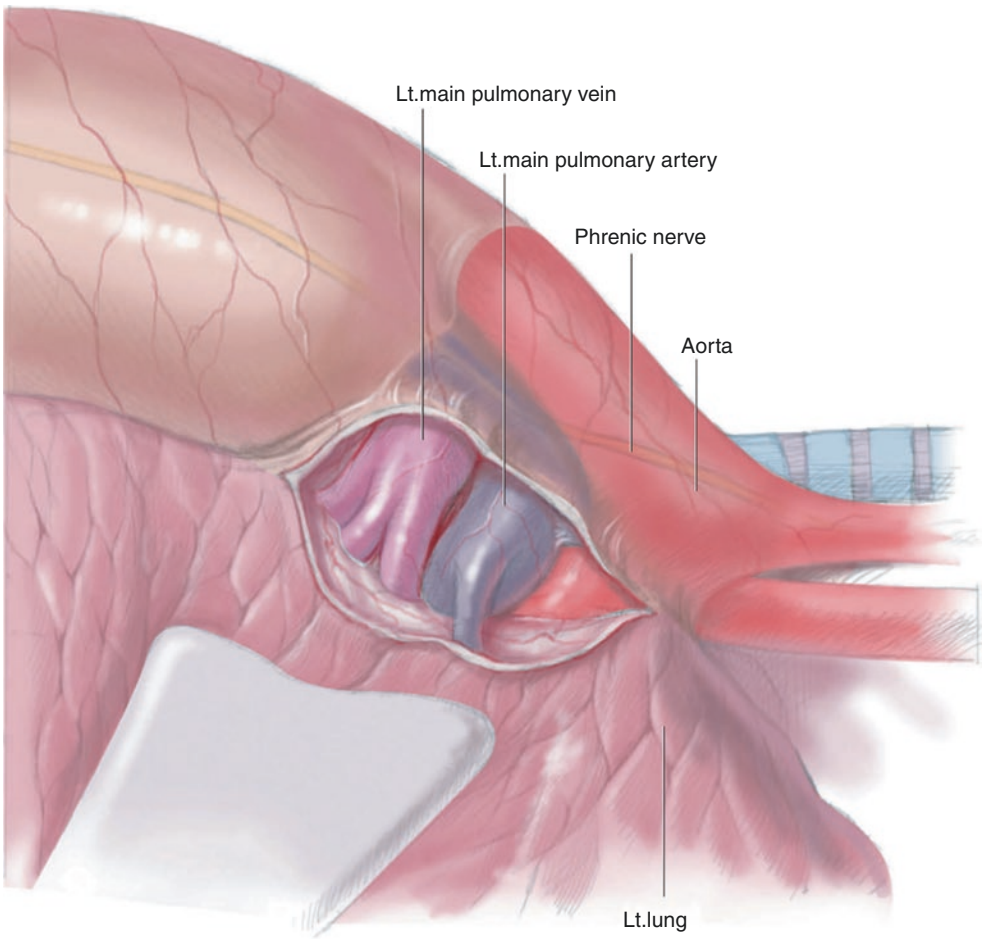

Fig. 13.5 Exposure of the hilum

total pneumonectomy. A chest tube is placed within the pleura for drainage, and the wound is closed in anatomical layers using absorbable suture. Postoperatively, drains can be removed early, provided that no air leak is demonstrable.

13.3.2 Thoracoscopy

The thoracoscopic approach is fundamentally similar to an open thoracotomy in identifying the hilar structures, but surgical clips or other techniques are often deployed to control vascular structures. Port placement depends upon the spe-

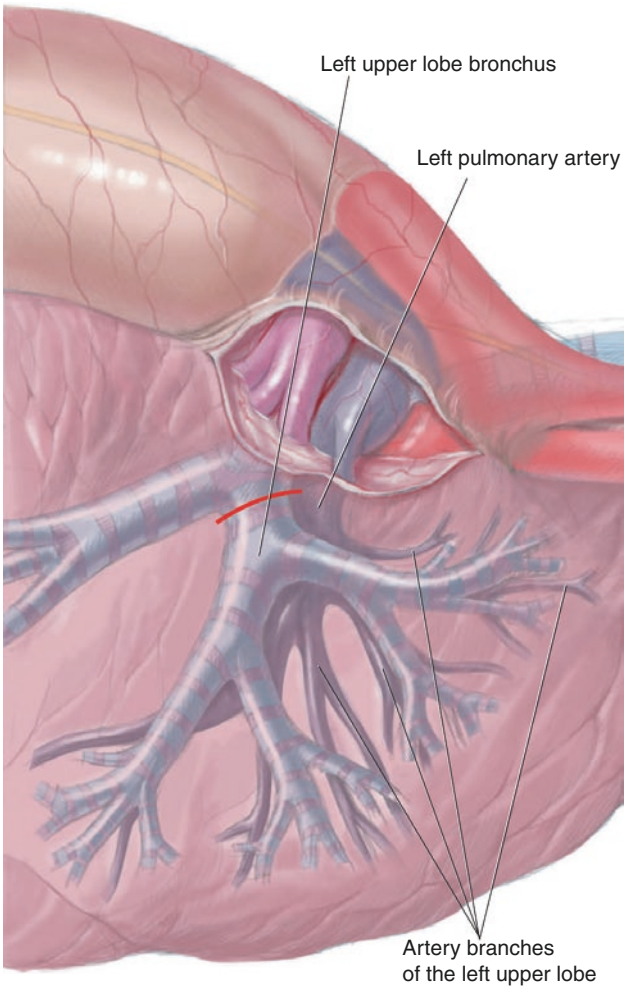

Fig. 13.6 Arterial circulation

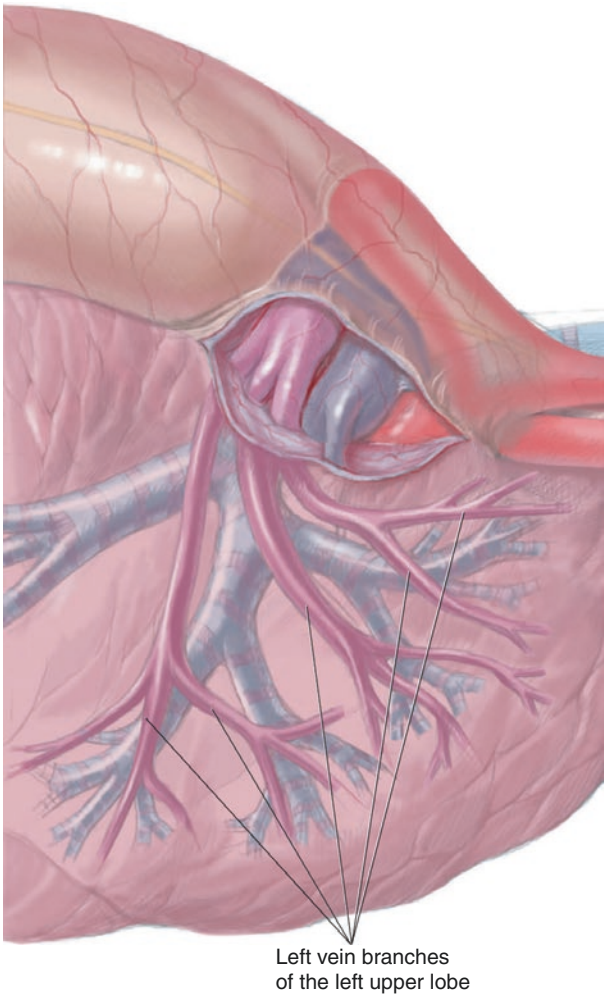

Fig. 13.7 Venous drainage

Fig. 13.8 Sewn closure of the bronchial stump

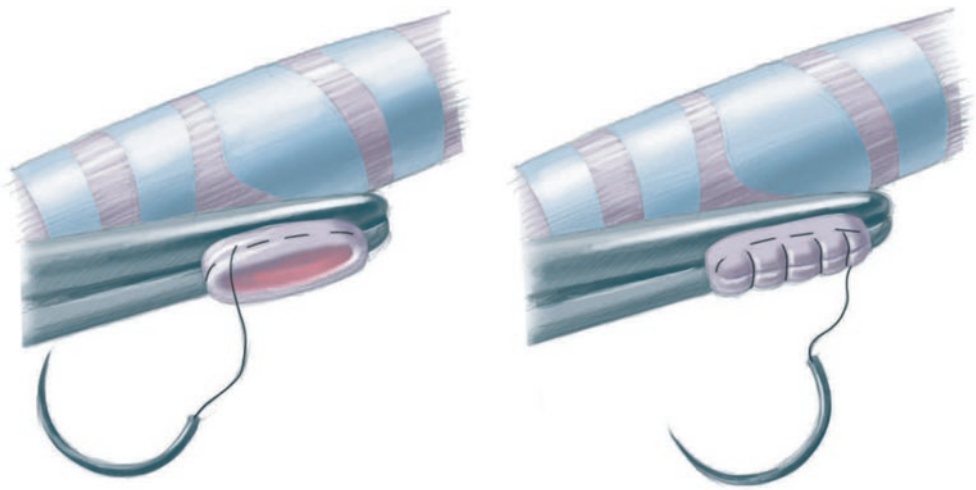

Fig. 13.9 Positioning for left upper lobectomy by video-assisted thoracoscopy

cific procedure to be done. Figure 13.9 (see Chap. 11). depicts the placement scheme for a left upper lobectomy. The fundamental endoscopic principles of triangulation and adequate space between ports applies to thoracoscopy, as it does in laparoscopy.

Conclusion

Lung surgery in neonates and infants is generally similar to that in adults except that the diminutive size, the associated lesions, and the unique pathologic entities require certain special considerations. Of course, the smaller the child, the more care must be taken in order to avoid technical injury. As with all lung surgery, technical problems may result in serious and irreversible consequences. Collaboration with paediatric anaesthesiologists familiar with the unique circumstances of paediatric chest surgery is essential.

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Congenital Diaphragmatic Hernia and Eventration

14

Prem Puri

14.1 Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernia (CDH) is a malformation characterized by a defect in the posterolateral diaphragm, the foramen of Bochdalek, through which the abdominal viscera migrate into the chest during fetal life. The reported incidence of CDH varies from 1 in 2200 to 1 in 5000 births. Polyhydramnios is present in 20% of pregnancies involving an infant with CDH and in 50% of pregnancies associated with infants with CDH who are stillborn. In most series, 80% of posterolateral diaphragmatic hernias have been reported to occur on the left side and 20% on the right side. Bilateral CDH is rare. The size of the defect varies from small (2 or 3 cm) to very large, involving most of the hemidiaphragm. A rim of muscle is usually present around the defect, which is often covered posteromedially with peritoneum. A hernial sac, composed of pleura and peritoneum, has been reported in about 20% of patients.

14.1.1 Antenatal Diagnosis

Widespread use of obstetric sonography has led to an increase in the frequency of antenatal diagnosis of CDH, which is established by demonstration of the abdominal viscera in the chest. Three easily detectable features—polyhydramnios, mediastinal shift, and the absence of an intra-abdominal stomach bubble—should prompt a more careful search for herniated abdominal organs in the chest. Polyhydramnios is present in about 80% of the pregnancies with fetuses who have CDH and has also been associated with poor outcome. Routine antenatal ultrasound detects approximately 50–85% of cases of CDH. Lung-to-head ratio (LHR), the ratio of contralateral lung area to head circumference, measured by antenatal ultrasound, is used to predict the severity and outcome in CDH and to predict patients with poor outcome for antenatal intervention.

Antenatal tracheal occlusion (TO) has been suggested as a potential inducer of increased lung growth. Antenatal fetal endoscopic tracheal occlusion (FETO) has been reported to improve survival in selected severe cases of CDH. With the advancement in fetoscopic techniques, the FETO has now evolved into a percutaneous, minimally invasive procedure using a detachable, endoluminal, inflatable balloon that can be inserted and removed prenatally by endoscopy. However, there have been only a few randomized, controlled trials reporting the outcome of TO.

14.1.2 Postnatal Presentation and Diagnosis

Postnatally, the most severely affected babies present with respiratory distress (cyanosis, tachypnoea, and sternal recession) at birth. Other infants develop cyanosis, tachypnoea, and grunting respirations within minutes or hours after birth. Physical examination reveals a scaphoid abdomen, an increased anteroposterior diameter of the thorax, and mediastinal shift. Breath sounds are absent on the affected side. Associated congenital anomalies may also be seen or may be revealed on further examination. CDH presents beyond the first hours of life in 10–20% of cases.

Diagnosis of CDH is made postnatally by plain radiography of the chest and abdomen, which demonstrates air-filled loops of the bowel in the chest and a paucity of gas in the abdomen. The diaphragmatic margin is absent, there is a mediastinal shift to the opposite side, and only a small portion of the lung may be seen on the ipsilateral side.

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14.1.3 Management Strategies

The mortality rate of infants born with CDH remains high, despite optimal perinatal care. The high mortality rate in CDH has been attributed to pulmonary hypoplasia and associated persistent pulmonary hypertension. In recent years, newer management strategies have emerged in the care of high-risk CDH patients, such as permissive hypercapnia, high frequency ventilation, extracorporeal membrane oxygenation, and delayed surgical repair. These strategies offer some hope of improving overall survival.

14.1.4 Surgical Management

Surgical management of CDH can be via a conventional open abdominal approach (Figs. 14.1, 14.2, 14.3, 14.4, 14.5, and 14.6) or a minimally invasive surgery approach via thoracoscopy (see Chap. 11). There are some concerns in relation to thoracoscopy for CDH repair, particularly the systemic effects of hypercapnia. Carbon dioxide absorbed during insufflation into the chest can lead to significant metabolic and physiologic changes. The impaired respiratory capacity imposed by lung collapse has implications for oxygenation and CO_2 excretion and an increase in arterial alveolar CO_2 gradient. Coupled with impaired ventilation, the end-tidal CO_2 ($ETCO_2$) can markedly increase in infants undergoing thoracoscopy. During thoracoscopic repair of CDH, blood gases and $ETCO_2$ should be closely monitored. Many studies have also reported a higher recurrence rate after thoracoscopic repair of CDH, which has been attributed to more frequent use of patch repair and technical difficulties in achieving a complete closure of the diaphragmatic defect.

After closure of the defect with an open abdominal approach, the abdomen is closed in layers. If the abdominal cavity is small, gentle stretching of the abdominal wall will enable safe closure in most patients. A chest drain should be avoided to prevent barotraumas, as the drain increases the transpulmonary pressure gradient.

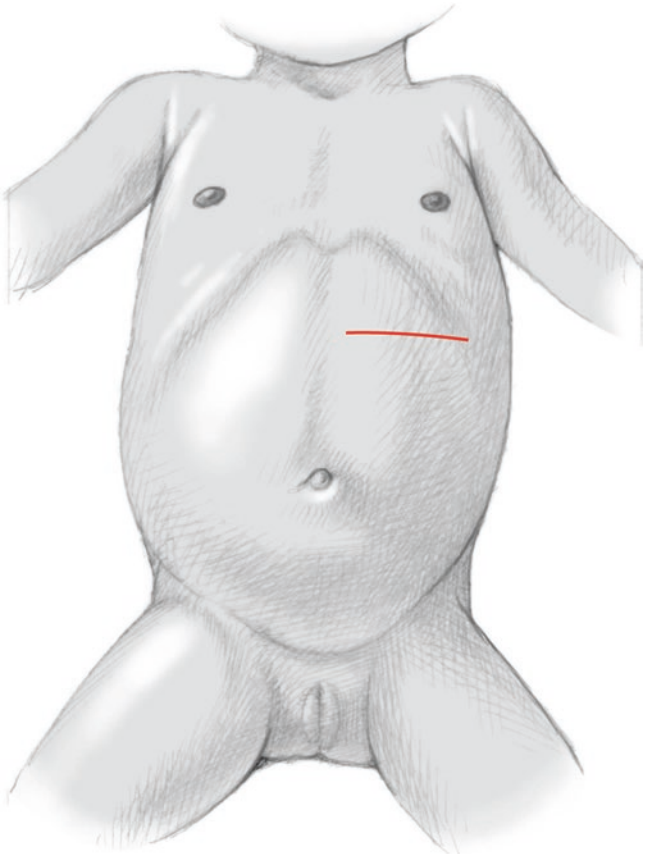

Fig. 14.1 A subcostal transverse muscle-cutting incision on the side of the hernia. General anaesthesia with muscle relaxation is used. The baby is positioned supine on a warm blanket. The most commonly preferred approach is abdominal. This offers good exposure, easy reduction of the abdominal viscera, and recognition and correction of associated gastrointestinal anomalies

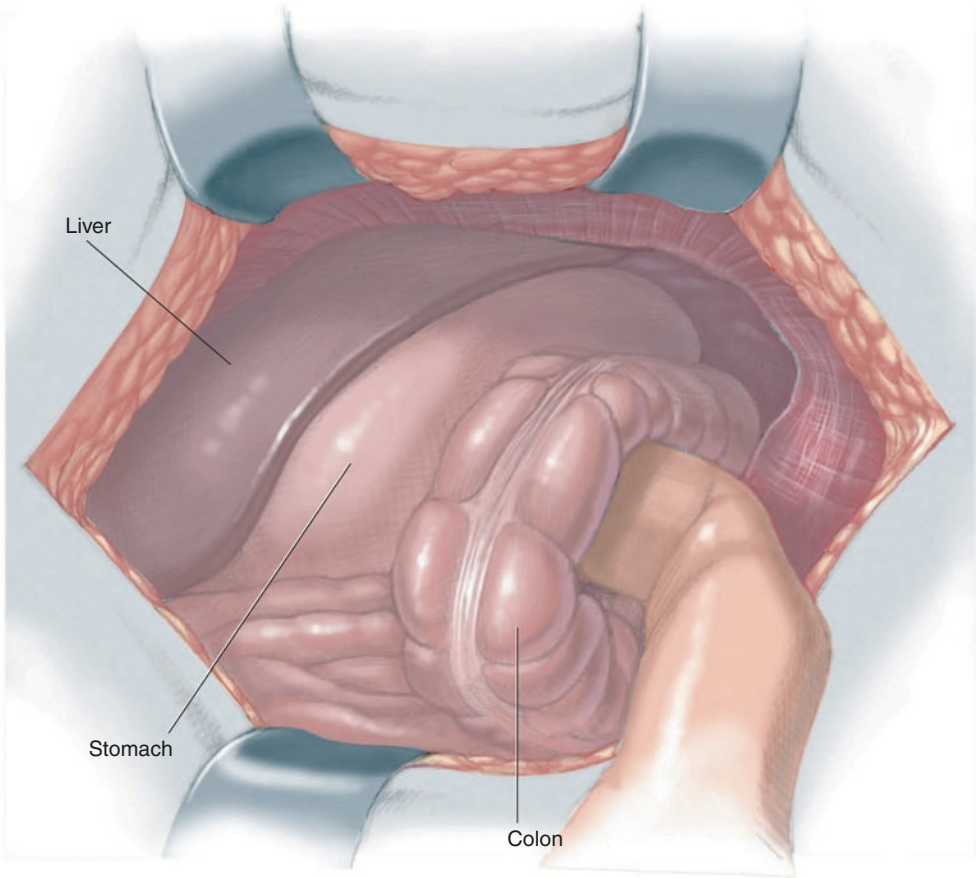

Fig. 14.2 Gently reducing the contents of the left sided hernia in the abdomen. On the right side, the small intestine and colon are first reduced and the liver is withdrawn last. After the hernia is reduced, an attempt is made to visualize the ipsilateral lung. This is usually done by retracting the anterior rim of the diaphragm. Often, a hypoplastic lung can be observed at the apex. A hernial sac, composed of pleura and peritoneum, is present in about 20% of patients. The sac, if present, is excised to avoid leaving a loculated, space-occupying lesion in the chest

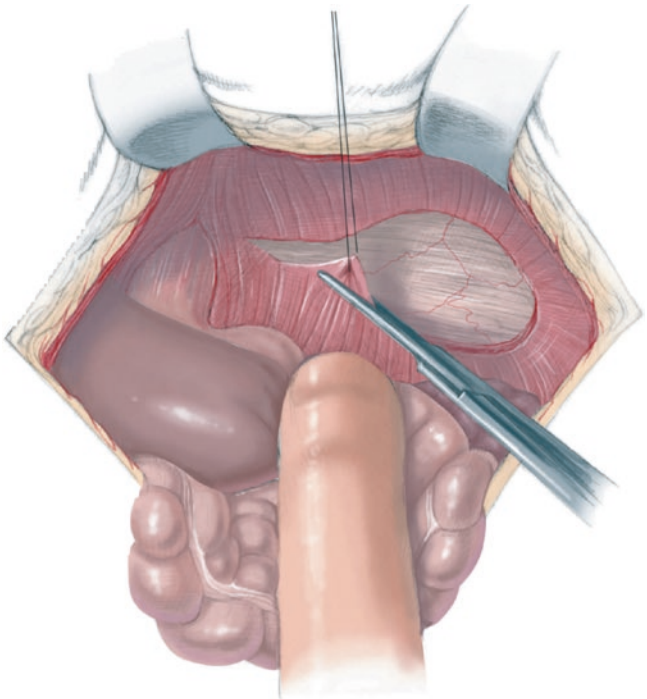

Fig. 14.3 Suturing the diaphragmatic defect. Most defects can be sutured by direct sutures of the edges of the defect. Usually the anterior rim of the diaphragm is quite evident, but the posterior rim may not be immediately apparent and may require dissection for delineation. The posterior rim of the diaphragm is mobilized by incising the overlying peritoneum

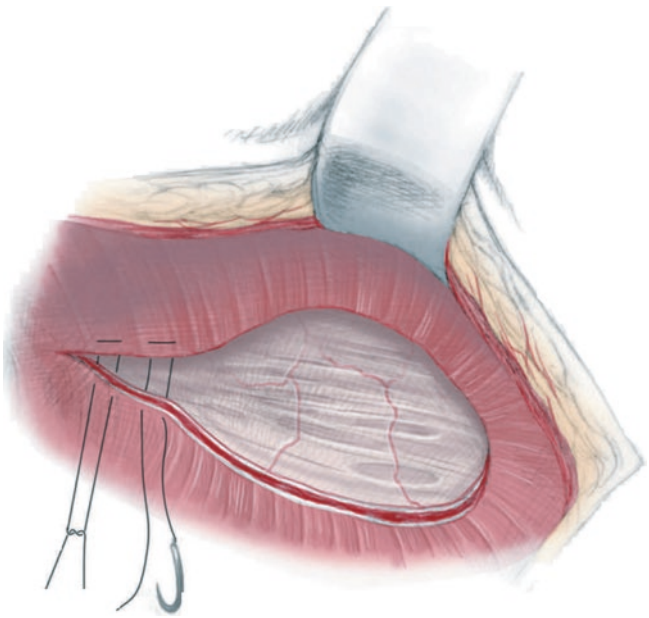

Fig. 14.4 Beginning closure of the defect using interrupted, nonabsorbable sutures

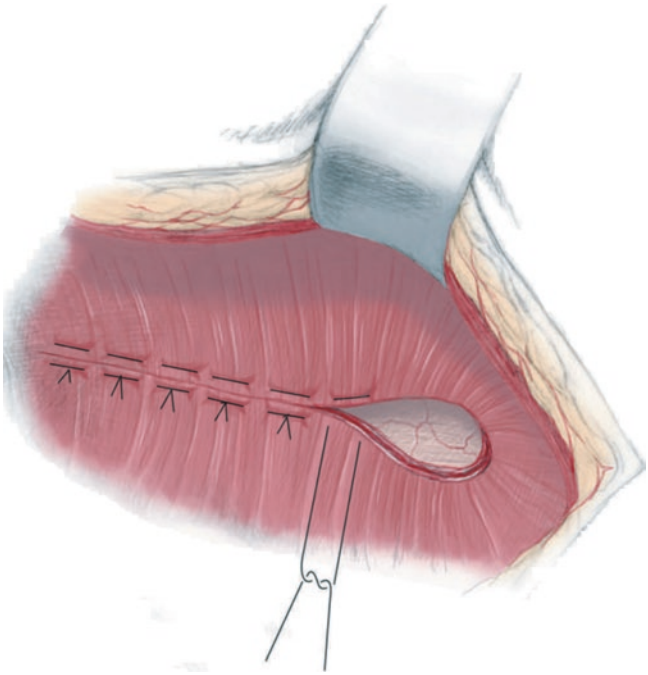

Fig. 14.5 Closure of the defect. Occasionally, the posterior rim is absent altogether, in which case the anterior rim of the diaphragm is sutured to the lower ribs with either periosteal or pericostal sutures

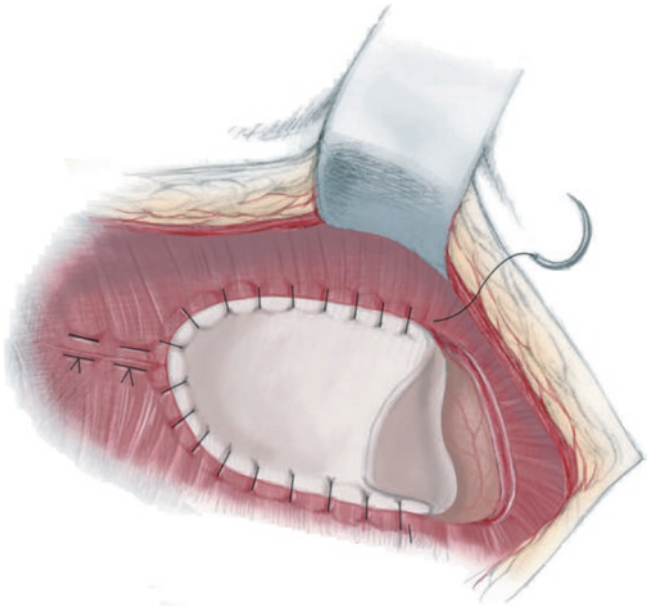

Fig. 14.6 Using a prosthetic patch to close a defect that is too large to repair by direct suture. Various techniques have been described to close these large defects, including the use of prerenal fascia, rib structures, the latissimus dorsi muscle, rotational muscle flaps from the thoracoabdominal wall, and prosthetic patches. The operations involving muscle flaps are too long and complex for critically ill patients and can lead to unsightly chest deformities. Prosthetic materials, including Marlex mesh, reinforced silicone elastomer, preserved pericardial heterografts, preserved dura, and the polytetrafluoroethylene patch (PTFE), have been advocated. The most commonly used prosthetic material presently is Surgisis® Soft Tissue Graft (Cook Medical; Bloomington, IN, USA), which is incorporated into adjacent tissue; this tends to lessen the risk of extension or displacement. The risk of infection is also decreased

14.2 Congenital Eventration of the Diaphragm

Eventration of the diaphragm has been described as an abnormally high or deviated position of all or part of the hemidiaphragm. Eventration may be congenital or may be acquired as a result of phrenic nerve palsy. Congenital eventration is a developmental abnormality that results in muscular aplasia of the diaphragm. In acquired eventration, the diaphragm, which initially had fully developed musculature, becomes atrophic secondary to phrenic nerve damage and disuse. This chapter deals with congenital eventration, but the clinical features and principles of management are similar for both the congenital and acquired forms.

14.2.1 Presentation and Diagnosis

Clinical features range from being asymptomatic to severe respiratory distress. Patients may present later in infancy with repeated attacks of pneumonia, bronchitis, or bronchiectasis. Occasionally, patients present later in childhood with gastrointestinal symptoms of vomiting or epigastric discomfort. In patients with phrenic nerve palsy, there may be a history of difficult delivery. They may present with tachypnoea, respiratory distress, or cyanosis. Physical examination reveals decreased breath sounds on the affected side, mediastinal shift during inspiration, and a scaphoid abdomen.

The diagnosis of eventration is usually made on a chest x-ray. Frontal and lateral chest x-rays will show an elevated diaphragm with a smooth, unbroken outline. Fluoroscopy is a useful investigation for differentiating a complete eventration from a hernia. Paradoxical movement of the diaphragm is seen if complete eventration is present. Ultrasonography is the most useful study in the diagnosis of eventration of the diaphragm and for identification of abdominal organs underneath the eventration. Other investigational modalities include pneumoperitonography, contrast peritonography, radioisotope scanning, and CT scans, but these are rarely required.

14.2.2 Management

Symptomatic patients, especially those with respiratory distress, need prompt supportive care with endotracheal intubation and ventilation with humidified oxygen to minimize the diaphragmatic excursions. A nasogastric tube is passed to decompress the stomach and intravenous fluids are commenced. Surgery is undertaken once the patient's condition is stabilized.

Plication of the diaphragm (Figs. 14.7 and 14.8) has been used for many years to treat eventration. Plication increases both tidal volume and maximal breathing capacity and has been successful in many clinical series. An abdominal approach through a subcostal incision is preferred for left-sided eventration, but a thoracic approach through a posterolateral incision via the sixth space may be used for right-sided lesions. The transabdominal approach allows good visualization of the entire diaphragm from front to back and easier mobilization of abdominal contents. Plication is carried out using nonabsorbable sutures, taking care to avoid injury to the phrenic nerve (Fig. 14.9). In cases of complete eventration, the diaphragm may be strengthened by a muscle flap or prosthetic patch.

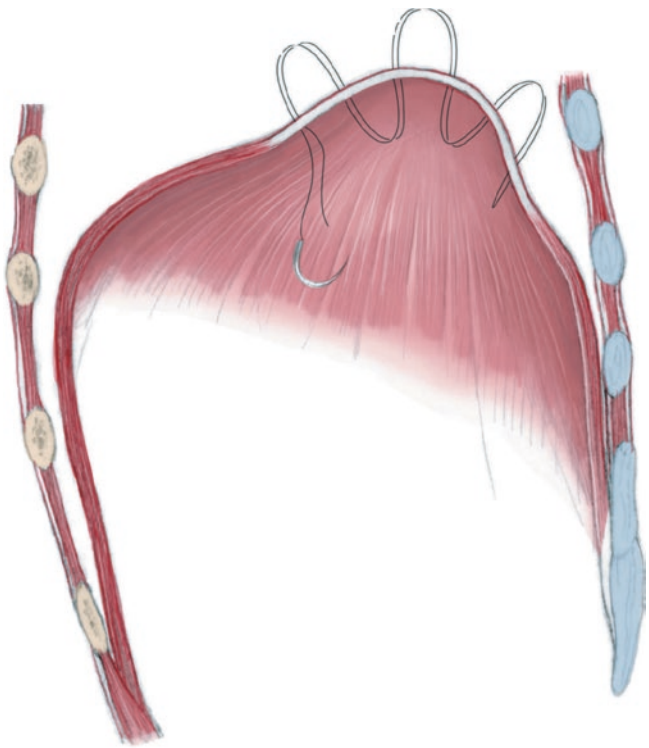

Fig. 14.7 Placing sutures for plication of the diaphragm to treat eventration

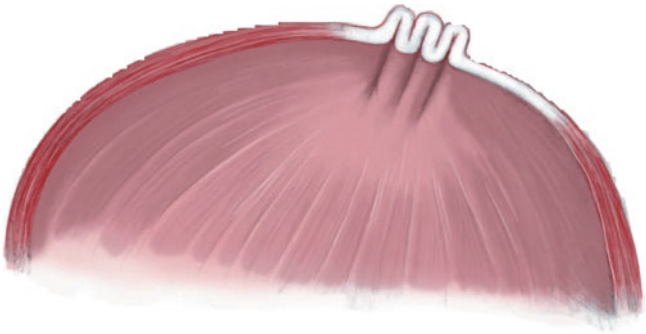

Fig. 14.8 Completed plication

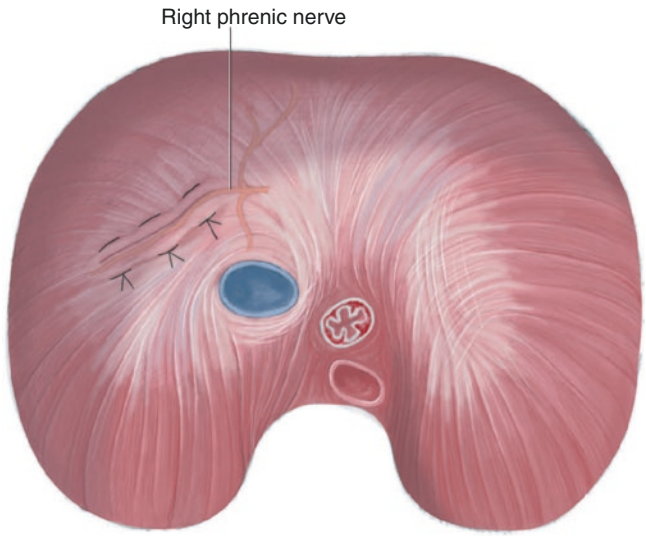

Fig. 14.9 Plication of the diaphragm, avoiding injury to the phrenic nerve

14.3 Postoperative Care

After transfer to the intensive care unit, the infant is kept warm and given maintenance requirements of intravenous fluids. Vital signs are monitored closely, with regular blood gas analyses and monitoring of preductal and postductal oxygenation. Ventilatory support is continued postoperatively with the aim of maintaining preductal PO_2 around 80–100 mmHg, PCO_2 up to 60 mmHg, and pH greater than 7.25 with hyperventilation (rates up to 150 per min) and the lowest possible pressures and low tidal volumes. The intrathoracic air pocket will usually reabsorb, but evidence of increasing air and fluid with mediastinal shift requires insertion of a chest drain. Weaning from ventilation should be meticulous and slow, as small variations in pH, PO_2 and PCO_2 will lead to persistent pulmonary hypertension. Weaning should commence with lowering of FiO_2, then peak pressures, and finally respiratory rate.

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Empyema

15

Michael Singh and Dakshesh Parikh

Paediatric empyema is usually secondary to an underlying pneumonia. *Streptococcus pneumoniae* continues to be the main causative organism. Other organisms include Group A *Streptococcus, Streptococcus viridans*, *Streptococcus anginosus, Staphylococcus aureus*, *Haemophilus influenzae*, *Streptococcus milleri*, and the anaerobic *Peptostreptococcus*. The incidence of paediatric thoracic empyema is increasing globally, and is estimated to be 3.3 per 100,000 children. The highest reported incidence occurs in children under the age of 5 years (53%). The resulting untreated effusion progresses through three stages: exudative, fibropurulent, and organization. Prompt medical management may halt the progression.

15.1 Diagnosis and Treatment Options

The initial investigations include blood cultures, radiograph, and chest ultrasound. Computed tomography (CT) scans of the chest are reserved for complicated patients. The mainstay of treatment includes intravenous antibiotics for sepsis control, and drainage of the effusion to improve lung expansion. If initial medical management fails, then surgical intervention is necessary. The use of intrapleural fibrinolysis (urokinase, tissue plasminogen activator) via a chest drain can produce good results. The surgical options include thoracoscopic debridement, mini thoracotomy and debridement, and thoracotomy with decortication. Thoracotomy and decortication is reserved for advanced and chronic cases.

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15.2 Operative Procedures

15.2.1 Thoracoscopic Debridement

General anaesthesia with central endotracheal intubation is used. Single-lung ventilation is unnecessary. The instruments used include a 5-mm, 0-degree thoracoscope; straight and curved graspers; a suction irrigation set; three 5-mm ports (blunt trocars); and a 16 Fr chest drain.

The patient is positioned laterally on an axillary roll with the affected side up. The surgeon stands towards the patient's back with the stacking system and monitor opposite. The first 5-mm port is inserted bluntly into the intercostal space anterior to the inferior angle of the scapula (Fig. 15.1). Local anaesthetic is infiltrated into the skin and muscles prior to port insertion.

A 5-mm suction device is inserted next, to aspirate as much of the effusion as possible in order to create some working space (Fig. 15.2). A pneumothorax of 6 mm Hg with flows of 1.5 L/min is established.

The second 5-mm working port is inserted under direct vision, in a location so as to give good access to the fibrin peel and septations (Fig. 15.3). The fibrin septations are broken down bluntly and are removed via the working port. The fibrin peel should be removed from the surface of the lung and diaphragm (Fig. 15.4). The result should be good lung expansion.

Any residual blood and effusion should be aspirated, and a 16 Fr chest drain is inserted via one of the ports. The ports are removed; it is not necessary to close the muscle at the port site. The skin can be closed with either sutures or skin glue.

Postoperatively, the chest drain is left on free drainage and a chest x-ray is done the following day. The chest drain is removed when the output is minimal and a chest x-ray shows no reaccumulation of the effusion. Oral antibiotics (co-amoxiclav) are continued for 4 weeks postoperatively.

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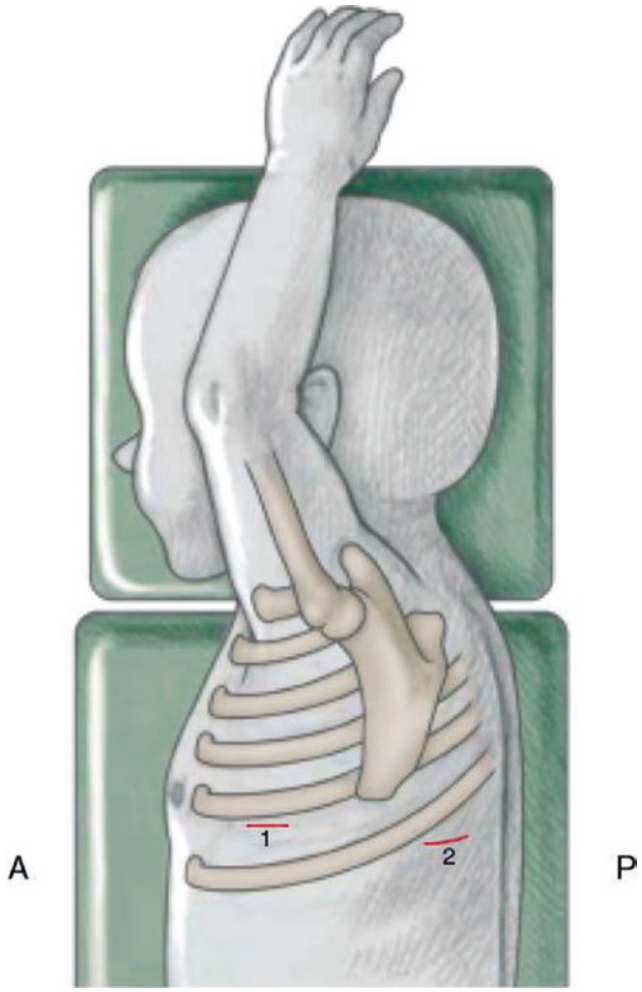

Fig. 15.1 Patient positioning and port placement for thoracoscopic debridement

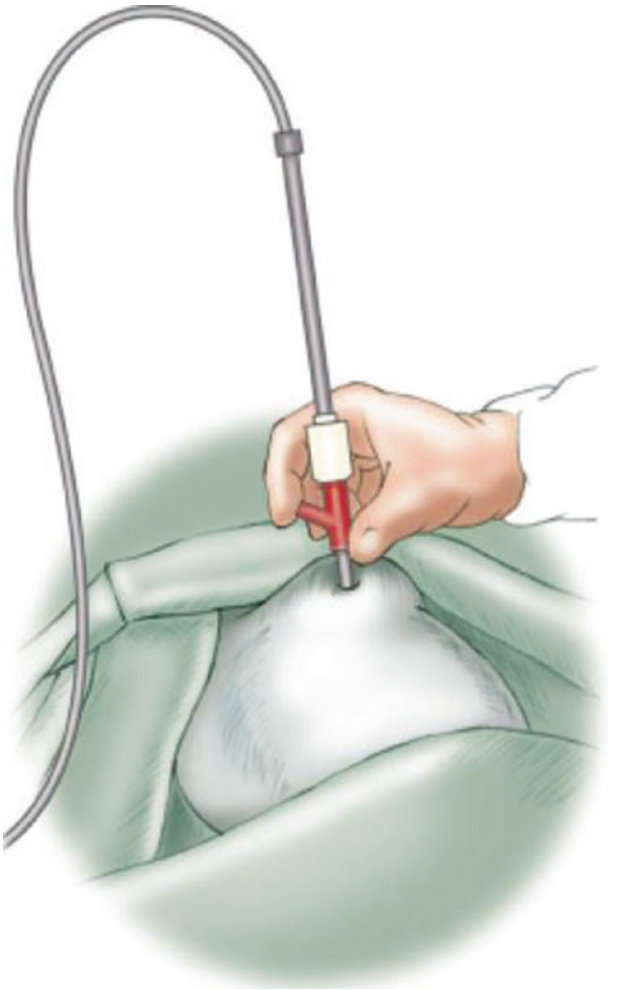

Fig. 15.2 Suction device inserted to aspirate the effusion

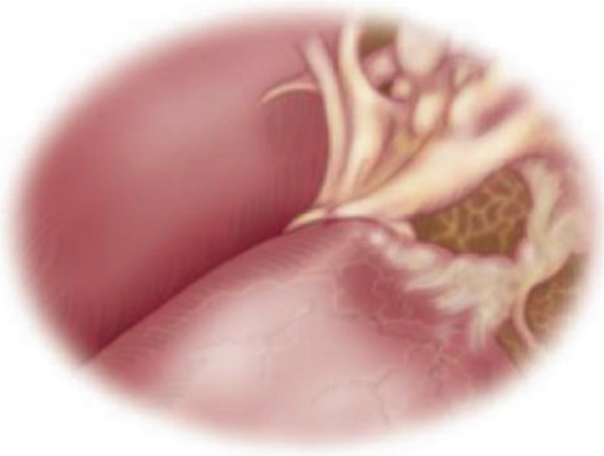

Fig. 15.3 The fibrin peel and septations

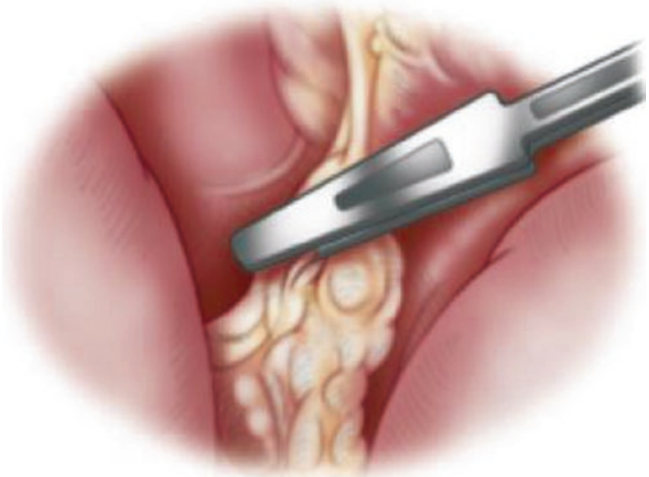

Fig. 15.4 Removal of the fibrin peel

15.2.2 Mini Thoracotomy

Mini thoracotomy is indicated in early empyema when fibrinolysis or thoracoscopic debridement is not available. The patient is positioned as for a thoracoscopic debridement. A 3- to 5-cm transverse incision is made between the anterior and posterior axillary lines over the 5th or 6th intercostal space (Fig. 15.5).

The anterior border of the latissimus dorsi muscle is retracted posteriorly, and the serratus anterior muscle is split (Fig. 15.6). The intercostal muscles are divided with monopolar diathermy and the pleura is opened. The effusion is aspirated and the fibrous peel is removed from the surface of the lung.

Once the lung is re-expanded, a 16 Fr chest drain is inserted and the chest muscles and skin are closed.

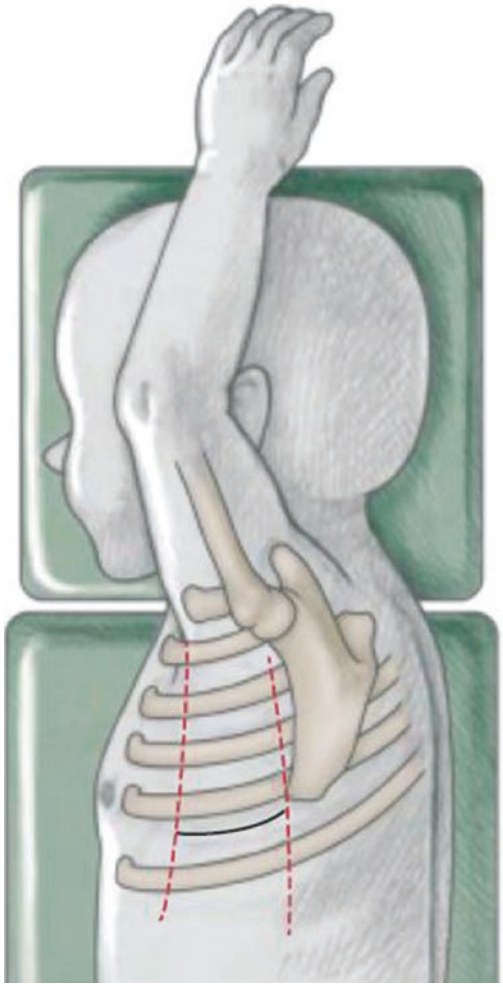

Fig. 15.5 Location of the transverse incision for a mini thoracotomy

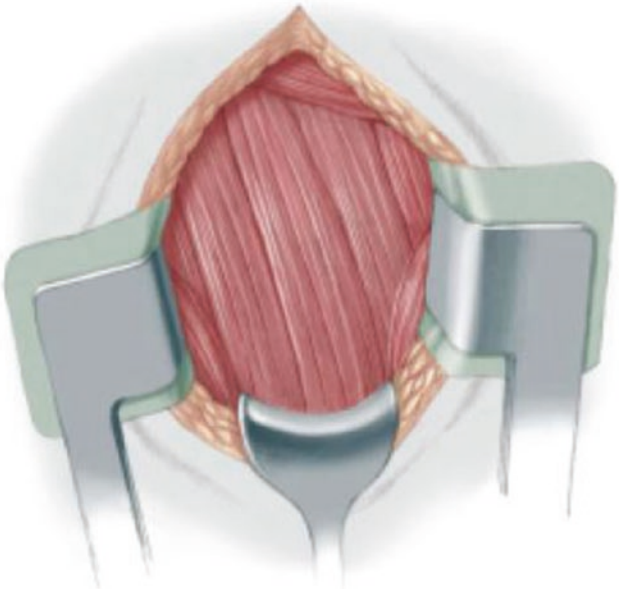

Fig. 15.6 Splitting of serratus anterior muscle

15.2.3 Thoracotomy and Decortication

Thoracotomy and decortication procedure is indicated in patients with advanced empyema, failed fibrinolysis, and bronchopleural fistula.

The patient is positioned as for a thoracoscopic debridement. A 5-cm transverse incision is made between the anterior and posterior axillary lines over the fifth or sixth intercostal space.

The latissimus dorsi and serratus anterior muscles are divided with monopolar diathermy.

The fourth or fifth rib is excised subperiosteally and the pleura is entered (Fig.15.7). The effusion is aspirated.

The loculations are broken down and the thick fibrous peel is removed from the surface of the lung. It is important to debride the interlobar fissures and release the lung from the diaphragm.

Once the lung re-expands, one or two 16 Fr chest drains are inserted. The inner layer of the periosteum is closed with an absorbable suture and the muscles are closed in layers.

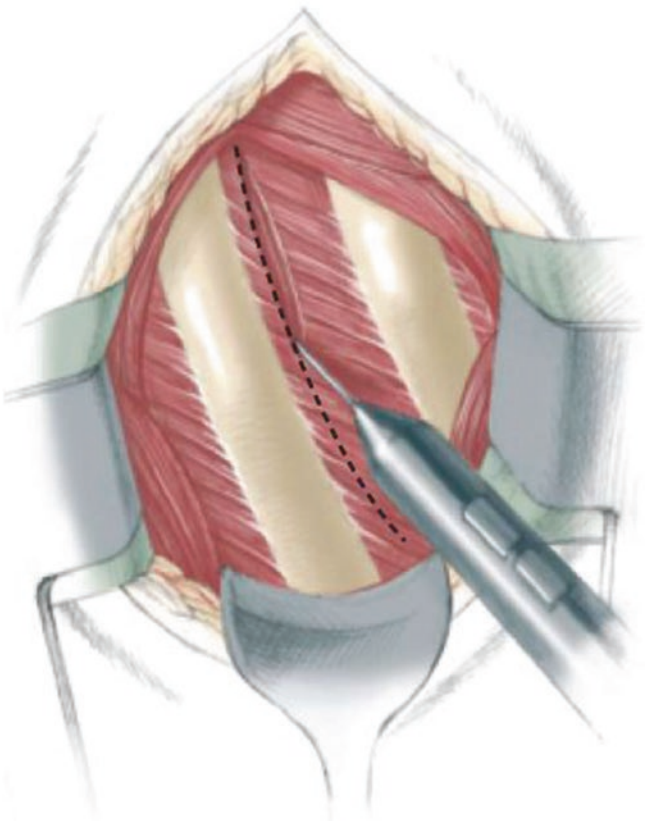

Fig. 15.7 Entry into the pleura

15.2.4 Serratus Anterior Muscle Flap

The serratus anterior muscle can be used to seal a bronchopleural fistula. After the latissimus dorsi is divided, a digitation of the serratus anterior muscle is detached from its anterior insertion and mobilised on its posterior blood supply (Fig. 15.8). After decortication of the lung, the muscle flap is inserted via an adjacent intercostal space and loosely sutured to the open bronchopleural fistula (Fig. 15.9). Chest drains are inserted and the chest closed as above.

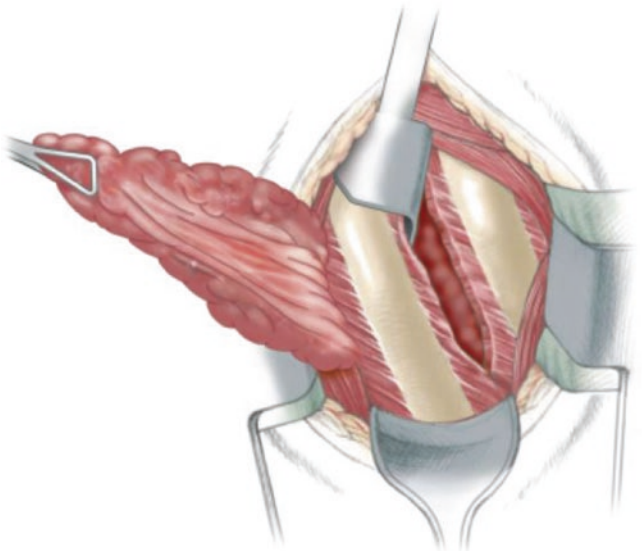

Fig. 15.8 Mobilising a flap of the serratus anterior muscle

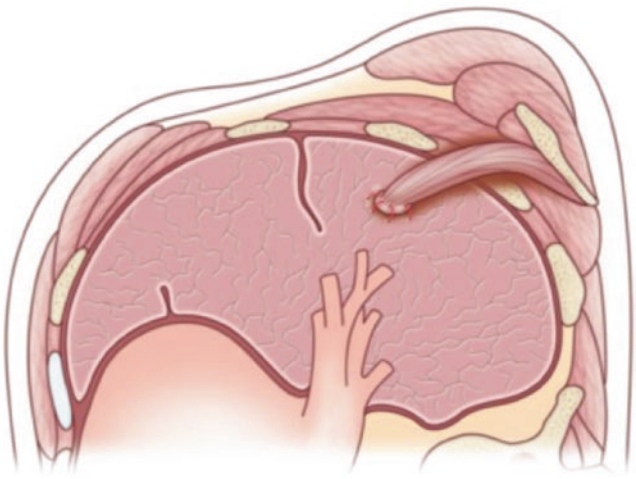

Fig. 15.9 Suturing of the muscle flap to the bronchopleural fistula

Conclusions

Early diagnosis and treatment of the postpneumonic effusion will result in a better outcome. If the effusion is not improving with intravenous antibiotics, then early referral to a paediatric surgical centre is recommended. The early use of drainage and either intrapleural fibrinolysis or thoracoscopic debridement of the empyema can result in resolution in most patients. Thoracotomy and decortication is the procedure of choice if empyema is advanced or complicated.

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Gynecomastia

16

Laura A. Monson and Mary L. Brandt

Gynecomastia, or breast development in males, develops in the presence of elevated estrogen or decreased androgen levels. Gynecomastia typically occurs at three time points: infancy, puberty, and in men over the age of 50. Pubertal gynecomastia is first seen in early to mid puberty, with a peak incidence at 14 years of age; it resolves in most boys within 3 years.

16.1 Evaluation

A thorough history and physical examination will almost always elucidate the cause of gynecomastia. How long the breast enlargement has been present and the velocity of growth should be established. Rapid growth is not characteristic of physiologic gynecomastia and should prompt further evaluation. Patients may report swelling, tenderness, and pain in the breasts. The patient should be asked about testicular swelling or pain, and about any previous testicular surgery. All patients should also be asked about medications, herbal remedies and teas, and the use of recreational drugs. Gynecomastia can lead to significant issues with reduced self-esteem, and questions to elicit symptoms of depression also should be asked. A strong family history of breast cancer may warrant evaluation for a BRAC mutation in the family, as it could affect recommendations for surveillance or treatment.

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In more than 75% of adolescents, gynecomastia will resolve spontaneously. However, the time to resolution is variable. Although many patients experience resolution within 1–2 years, resolution may not be complete until the end of puberty, at age 19 or 20. In cases caused by medications or recreational drugs, treatment involves cessation of the offending substance and watchful waiting. For cases caused by an underlying condition such as kidney failure or hypogonadism, treatment is aimed at correction of the underlying condition.

16.2 Classification

Gynecomastia is most often classified using the grading system (Fig. 16.1) developed by Simon:

- Grade I: Small breast enlargement with localized button of tissue around the areola
- Grade II: Moderate breast enlargement exceeding areola boundaries with edges that are indistinct from the chest
- Grade III: Moderate breast enlargement exceeding areola boundaries with edges that are distinct from the chest with skin redundancy
- Grade IV: Marked breast enlargement with skin redundancy and feminization of the breast

The breast tissue present in gynecomastia is most often tender, firm, and subareolar. One third of boys will present with unilateral gynecomastia. A mass in the breast that is not subareolar and is asymmetric should raise concern for other conditions. A testicular exam is a key component of the physical exam to rule out hypogonadism, suggestive of Klinefelter's syndrome, a varicocele, or possible estrogen-secreting tumors of the testes, such as germ cell tumors, Sertoli cell tumors, or Leydig cell tumors.

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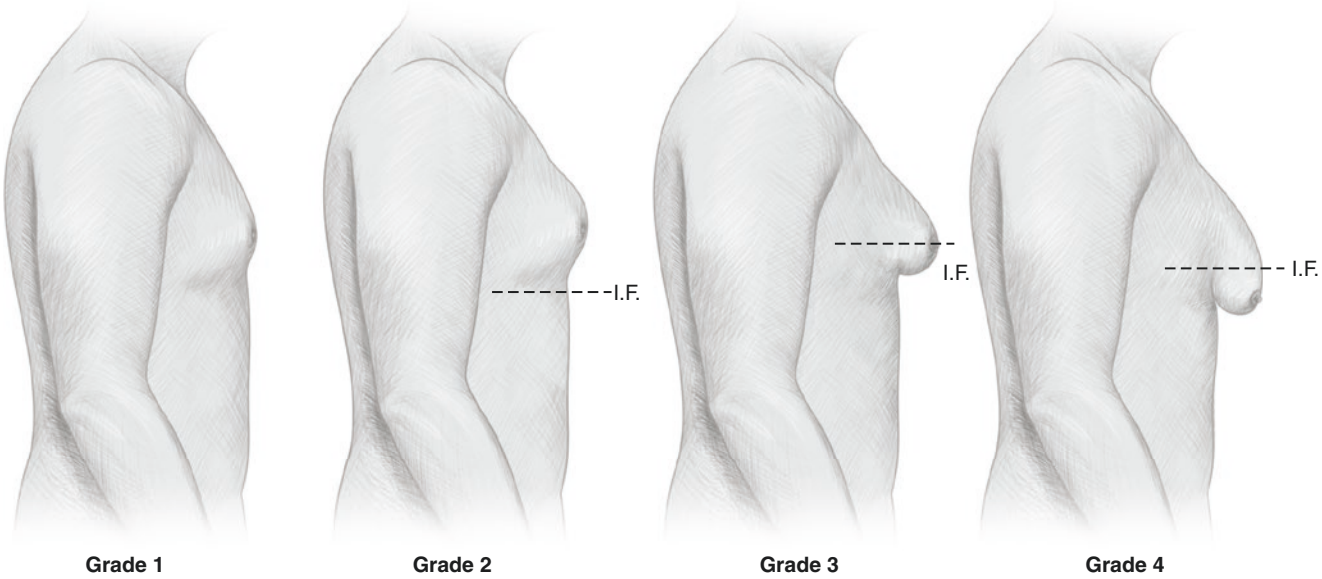

Fig. 16.1 Classification of gynecomastia. IF—inframammary fold

16.3 Surgical Treatment

16.3.1 Subtotal Mastectomy Through a Subareolar Incision

The choice of surgical treatment depends upon the amount of breast tissue and the degree of ptosis. Typical glandular gynecomastia (Grade I or II) is most commonly approached by a subtotal mastectomy via an inferior, semicircular periareolar incision (Fig. 16.2). The subareolar incision is the incision of choice for direct excision because it hides well at the junction of the areola and skin, allowing the patient to remove his shirt in public without noticeable scars. It is important when removing the breast tissue to keep a "button" of tissue approximately 1 cm in depth attached to the areolar complex to maintain a cosmetically normal appearance of the chest wall after surgery.

For boys with large areolas or significant breast enlargement, reduction of the areola and/or redundant skin may be indicated (Fig. 16.3). The average normal male nipple-areolar complex is 2.8 cm in diameter. Several different procedures can be used to reduce the size of the nipple complex for boys who exceed this diameter. Excision of excess areola and/or skin begins with de-epithelializing what will become a dermal flap. Following removal of the excess epithelium, the flap is raised, again with a significant "button" of tissue, allowing the mastectomy to be performed without difficulty. The nipple complex remains vascularized by the remaining subdermal pedicle flap. Closure of the wound is accomplished using the round-block technique.

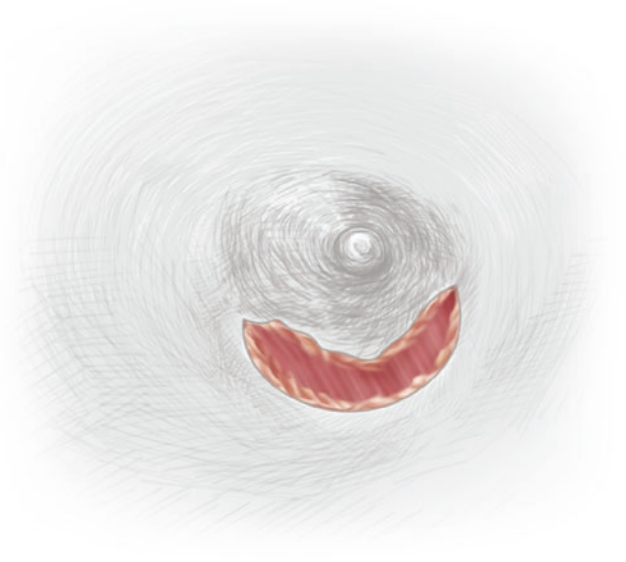

Fig. 16.2 Subtotal mastectomy through a subareolar incision

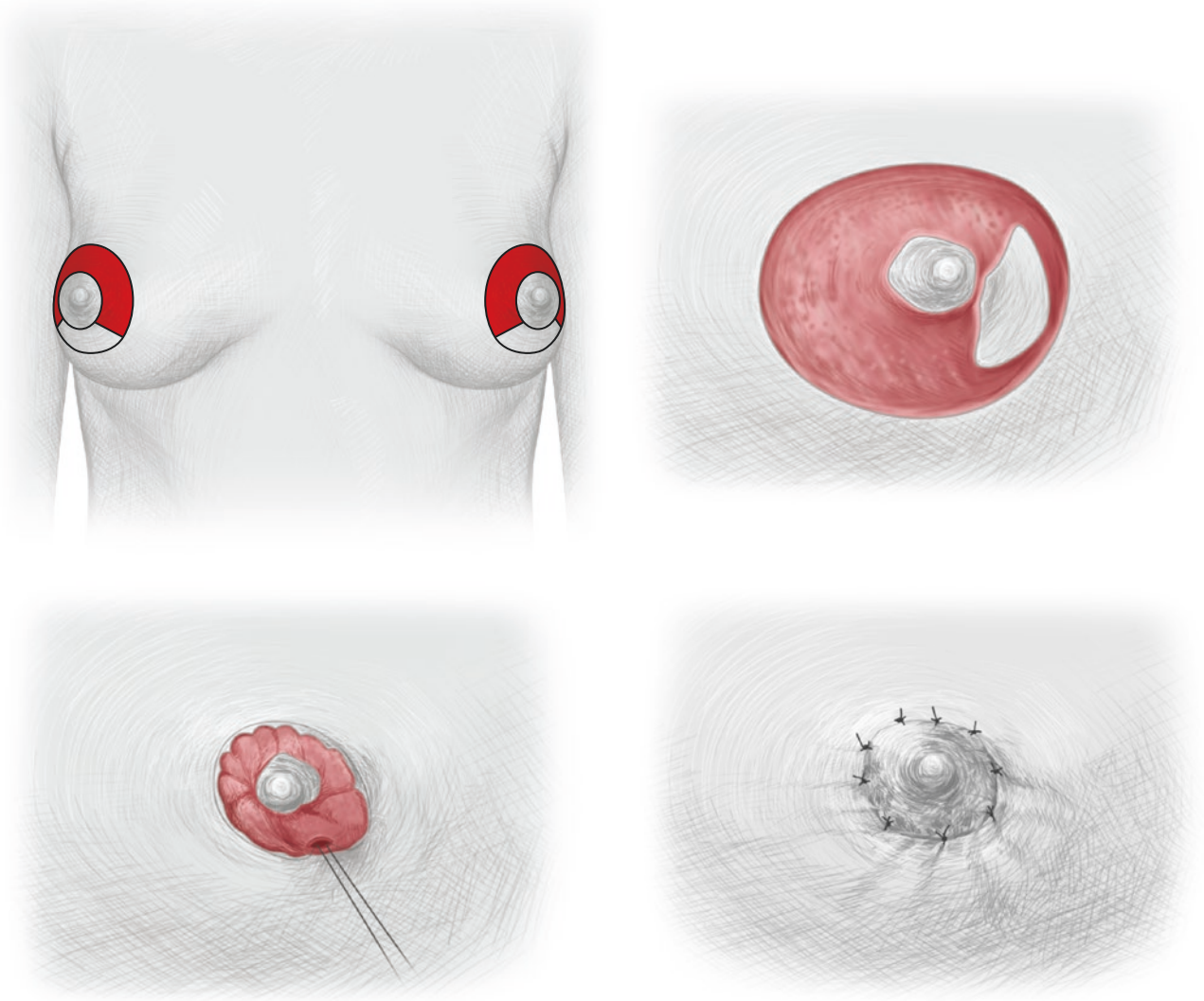

Fig. 16.3 Removal of excessive skin and/or areola for cosmetic reconstruction at the time of subtotal mastectomy. *Top left*, The areola is drawn to a diameter between 2.5 and 3 cm. *Top right*, The area of the pedicle (*stippled*) is de-epithelialized. *Bottom left*, The inferior redundant skin and/or areola is removed. *Bottom right*, The "round block" technique is used to close the defect. Fine sutures are used to complete the skin closure

16.3.2 Ultrasound Liposuction

Ultrasound-assisted liposuction (Fig. 16.4) has changed the approach to gynecomastia in recent years and is particularly effective for young men with good skin elasticity who do not have excess skin. Complications of ultrasound-assisted liposuction include wound infection, wound dehiscence, hematoma, and seroma. Other complications that can occur include undercorrection, overcorrection, contour irregularities, and scarring or keloid formation. For young men with an excessive amount of tissue, poor skin quality, or severe ptosis, liposuction alone may not achieve an adequate result. The patient will be left with loose, hanging skin and a "deflated balloon" appearance. These patients require resection of skin and of breast tissue with reconstruction, as described above.

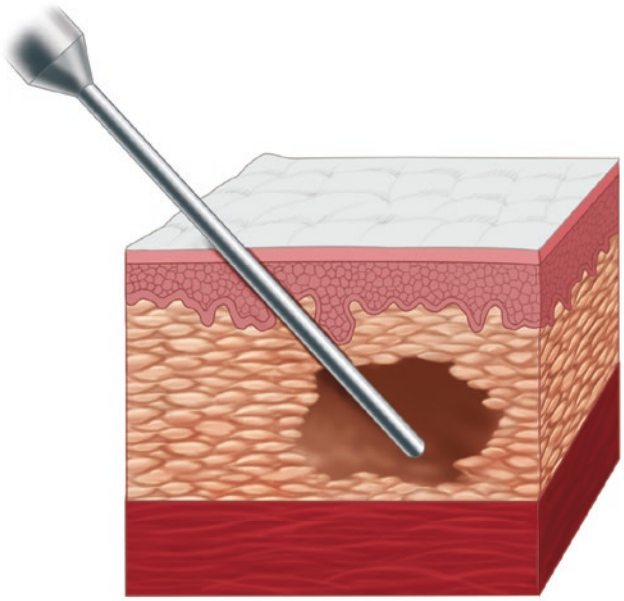

Fig. 16.4 Ultrasound-assisted liposuction

16.4 Postoperative Care and Complications

Regardless of the technique used, supportive compressive garments are essential to assist with seroma resolution and contour irregularities. Drains are commonly placed, although in a retrospective review of 138 adult patients, there was no difference in outcome with or without drains. Weightlifting and other strenuous activities should be avoided for 2–3 weeks to minimize fluid collection.

Complications of surgery are infrequent. They include hematoma, seroma, overcorrection (with indentation of the center of the breast), undercorrection, infection, and scarring. Small seromas can be treated with prolonged use of compression garments and watchful waiting.

Conclusions

Gynecomastia is common in adolescent boys and is usually self-limited. Although most young men will have physiologic gynecomastia, a careful history and physical is essential to diagnose other potential underlying causes of gynecomastia. Surgery is indicated for persistent gynecomastia or for gynecomastia with physical or psychological symptoms. Simple resection through an infraareolar incision is the procedure of choice for Grade I and II gynecomastia. For young men with larger breasts, ultrasonic liposuction, coupled with subcutaneous mastectomy through a periareolar incision when necessary, is the preferred surgical treatment. For adolescents with large areolar complexes or significant redundant skin, more extensive reconstruction may be indicated.

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Extracorporeal Membrane Oxygenation

17

Jason S. Frischer, Charles J. H. Stolar, and Ronald B. Hirschl

Extracorporeal membrane oxygenation (ECMO) is a life-saving technology that affords partial heart/lung bypass for extended periods. ECMO is a supportive rather than a therapeutic modality, as it provides sufficient gas exchange and perfusion in patients with acute, reversible cardiac or respiratory failure. It provides a finite period to "rest" the cardiopulmonary systems, during which time they are spared insults from traumatic mechanical ventilation and perfusion impairment. ECMO was first implemented in newborns in 1974. Since then, the Extracorporeal Life Support Organization (ELSO) has recorded 50,903 neonatal and paediatric patients treated with ECMO for a wide range of cardiorespiratory disorders. In the neonatal period, the most common indications treated with ECMO are meconium aspiration syndrome (MAS), congenital diaphragmatic hernia (CDH), persistent pulmonary hypertension of the neonate (PPHN), sepsis, respiratory distress syndrome (RDS), and cardiac support. For the paediatric population, viral and bacterial pneumonia, acute respiratory failure (non-ARDS), acute respiratory distress syndrome (ARDS), and cardiac disease are the most common pathophysiologic processes requiring ECMO intervention.

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17.1 Indications

Candidates for ECMO are expected to have a reversible cardiopulmonary disease process with a predicted mortality greater than 80–90%, and exhaustion of ventilatory and other therapies. Obviously, these criteria are subjective and vary between institutions. Criteria for mortality risk in neonatal respiratory failure have been suggested to identify infants with greater than 80% mortality:

- Oxygenation index (OI), calculated as FiO_2 × mean airway pressure × 100, divided by PaO_2 (OI > 25 is predictive of a 50% mortality rate and is a relative indication for ECMO; OI > 40 equates with 80% mortality and mandates implementation of ECMO)
- Alveolar-arterial oxygen gradient (A-aDO_2) >625 mmHg for more than 4 h, or an A-aDO_2 > 600 mmHg for more than 12 h

Criteria for high mortality risk are less well defined for older infants and children. The combination of a ventilation index (respiratory rate × $PaCO_2$ × peak inspiratory pressure/1000) >40 and an OI > 40 correlates with a 77% mortality, whereas 81% mortality is associated with an A-aDO_2 > 580 mmHg and a peak inspiratory pressure of 40 cm H_2O or higher. In general, ECMO is indicated in paediatric patients with respiratory failure when the A-aDO2 is >600 mm Hg on FiO_2 1.0 despite optimal treatment. Indications for support in patients with cardiac pathology are based on clinical signs of cardiovascular failure, such as hypotension despite the administration of inotropes or volume resuscitation, metabolic acidosis, oliguria (urine output <0.5 mL/kg/h), and decreased peripheral perfusion.

In addition, the gestational age should be at least 34–35 weeks, because of increased risk for intracranial haemorrhage (ICH), and the birth weight should be at least 2 kg, owing to cannula size limitations. The length of mechanical ventilation prior to ECMO, with its associated

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toxicity from prolonged exposure to high concentrations of oxygen and elevated positive pressure ventilation contributing to the development of bronchopulmonary dysplasia, should be no longer than 10–14 days. Babies with lethal congenital anomalies should not be considered for ECMO support. Treatable conditions such as total anomalous pulmonary venous return and transposition of the great vessels, which may masquerade initially as pulmonary failure, can be corrected with surgery but may require ECMO resuscitation initially. Therefore, echocardiogram should be rapidly obtained to determine cardiac anatomy. There should be no evidence of significant neurologic injury, such as seizures. Patients with suggestion of a small ICH (grades I–II) should be considered candidates for ECMO on an individual basis and should be monitored closely for worsening of the hemorrhage. In fact, any gross active bleeding or major coagulopathy should be corrected in all patients prior to initiating ECMO.

17.2 ECMO Procedure

The goal of ECMO support is to provide gas exchange and oxygen delivery. Figure 17.1 illustrates the various ECMO configurations that are available, and Fig. 17.2 shows the components of a typical ECMO circuit. Figures 17.3, 17.4, 17.5, 17.6, and 17.7 demonstrate the steps in establishing a venoarterial (VA) ECMO circuit.

For venovenous (VV) and double-lumen single-cannula venovenous (DLVV) bypass, the procedure is exactly the same, including dissection of the artery, which is marked with a vessel loop so that a future switch from VV to VA ECMO can be accomplished, if necessary. The catheter tip should be in the mid-right atrium (6 cm in the neonate), with the arterial portion of the catheter pointed toward the ear to direct the oxygenated blood flow towards the tricuspid valve.

Cannula position is confirmed by chest radiograph and by transthoracic echocardiogram when necessary. The venous catheter should be located in the inferior aspect of the right atrium, and the arterial catheter at the ostium of the innominate artery and the aorta. With a double-lumen venous catheter, the tip should be in the mid-right atrium with reinfusion of oxygenated blood flow towards the tricuspid valve.

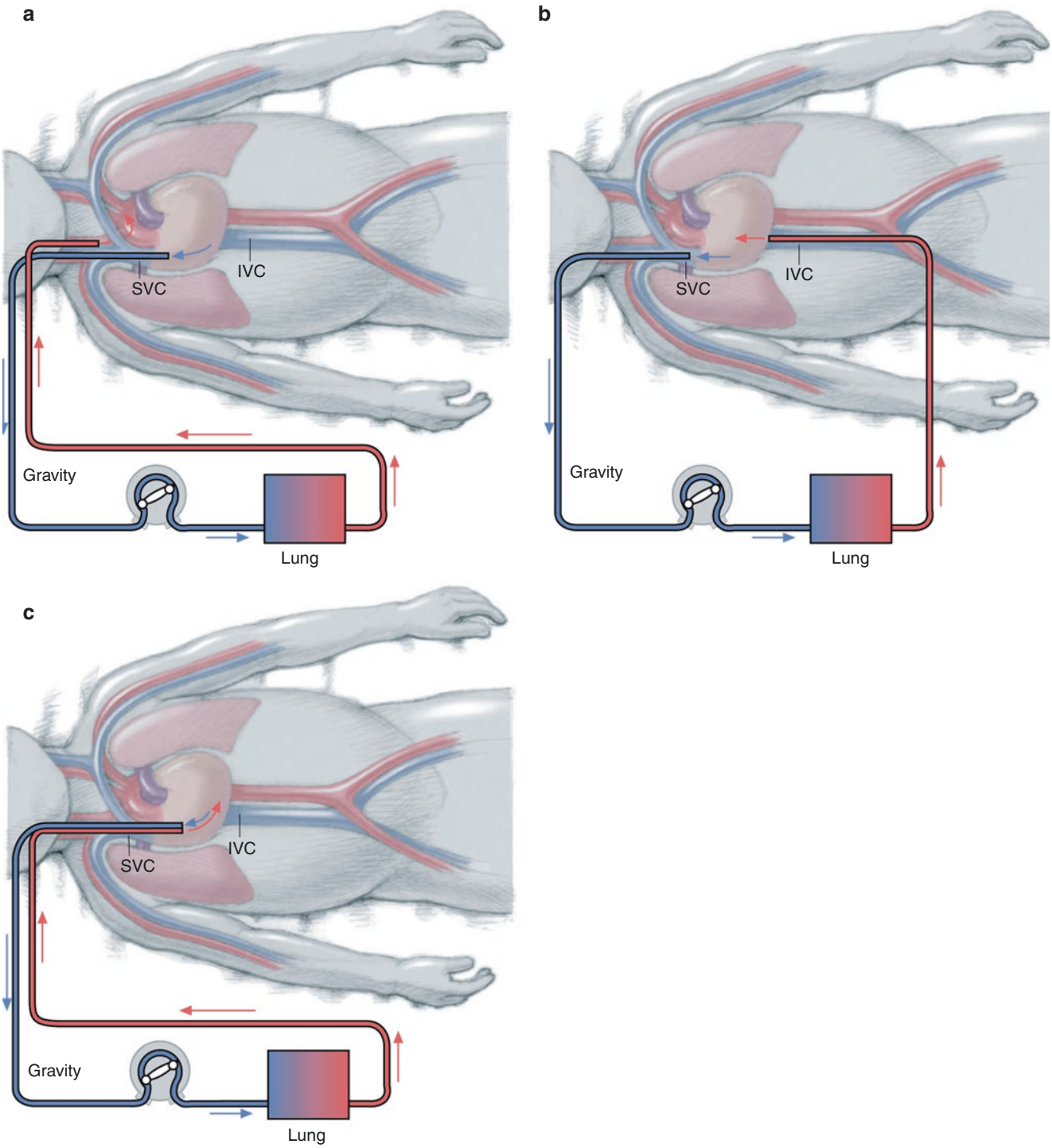

Fig. 17.1 (**a**–**c**) Three different extracorporeal bypass configurations are available: venoarterial (VA), venovenous (VV), and double-lumen single cannula venovenous (DLVV). VA bypass allows for support of both the pulmonary and cardiac systems. Venous blood is drained from the right atrium (RA) through the internal jugular (IJ) vein, and oxygenated blood is returned via the carotid artery (CA) to the aorta. Potential disadvantages of this arrangement include the sacrifice of a major artery; risk of gaseous or particulate emboli into the systemic circulation; reduced pulmonary perfusion; increased afterload, which may reduce cardiac output; nonpulsatile flow; and perfusion of the coronaries by relatively hypoxic left ventricular blood. VV and DLVV avoid these disadvantages and provide pulmonary support but do not provide hemodynamic/cardiac support. VV bypass is accomplished with drainage from the RA via the IJ with reinfusion into the femoral vein or drainage from the inferior vena cava (IVC) or femoral vein with reinfusion into the IJ/RA. DLVV is carried out by means of the IJ. A major disadvantage of VV and DLVV ECMO is that a fraction of freshly infused blood recirculates back into the circuit, requiring about a 20% increase in flow rate. In summary, it is recommended that patients who require only respiratory support use VV or DLVV bypass, and those that require cardiac support use VA ECMO. If necessary, one can convert VV or DLVV to VA support

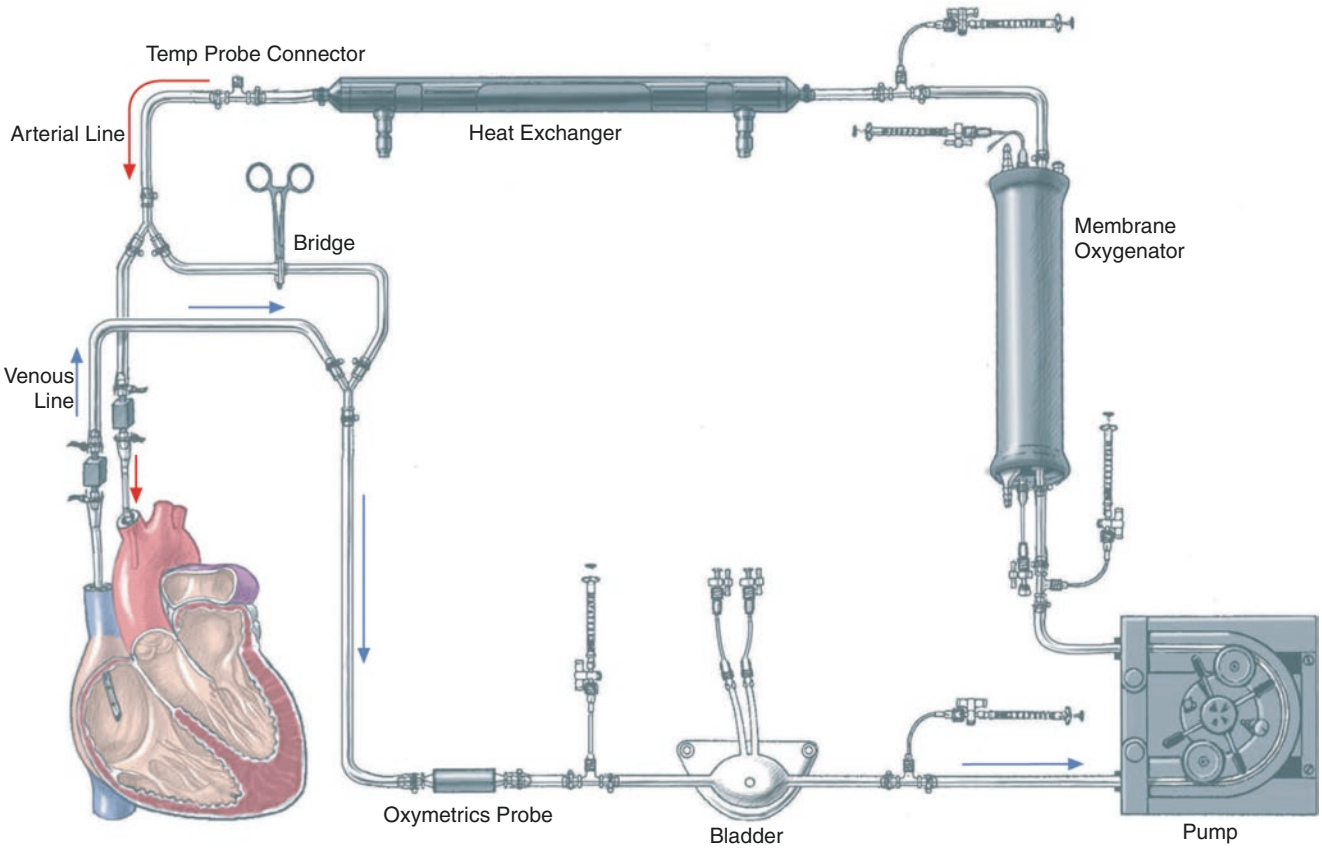

Fig. 17.2 The circuit comprises three main components: a roller pump, a membrane oxygenator, and a heat exchanger. Right atrial blood is drained by gravity siphon into a venous servo-mechanism, which acts to ensure that venous return to the circuit is adequate for the current pump flow. To do so, the servo detects diminished venous return, slows or shuts off the pump, and sounds an alarm, hence stopping blood flow to eliminate the risk of introducing air into the circuit, cavitation, and injury to the right atrium. Next, a roller pump, with continuous servoregulation and pressure monitoring, perfuses the blood through the membrane oxygenator. The oxygenator is a two-compartment chamber composed of a spiral wound silicone membrane and a polycarbonate core, with blood flow in one direction and oxygen flow in the opposite direction. The size of the oxygenator is chosen based on the patient's size. The oxygenated blood then flows through a heat exchanger and is returned to the patient. A bridge is constructed to connect the venous line shortly after exiting the patient and the arterial line just prior to entering the patient, so that during weaning the patient and the circuit can easily form two separate circuits. Many centers now use circuits that incorporate centrifugal pumps and low-resistance, blood-on-the-outside/gas-on-the-inside multiporous hollow-fiber lungs. These systems are much simpler, cannot overpressurize, and are at lower risk for negative-pressure events that bring gas out of solution; they are therefore much safer and require less maintenance and supervision

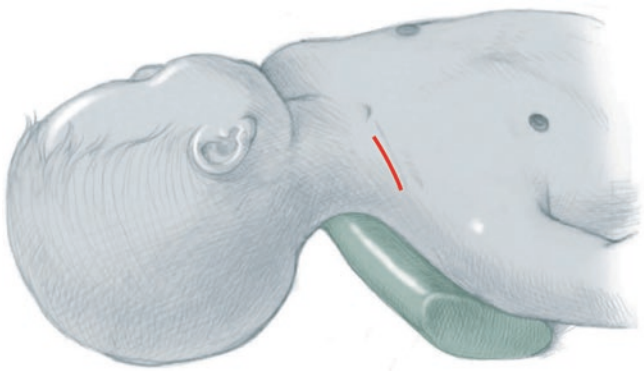

Fig. 17.3 Cannulation can be performed in the neonatal or paediatric intensive care units under adequate sedation, with proper monitoring. The patient is positioned with the head at the foot of the bed, supine, and the head and neck hyperextended over a shoulder roll and turned to the left. Local anaesthesia is administered over the proposed incision site. A transverse cervical incision is made over the sternomastoid muscle, one finger's breadth above the right clavicle

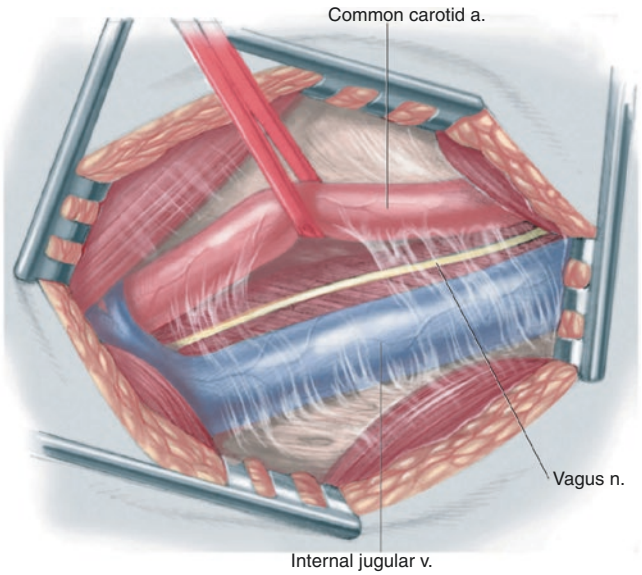

Fig. 17.4 The platysma muscle is divided with electrocautery. Self-retaining retractors are placed and dissection is carried out with the sternomastoid muscle retracted to expose the carotid sheath. Using sharp dissection and meticulous haemostasis, the sheath is opened and the internal jugular vein, common carotid artery, and vagus nerve are identified. All vessels must be handled with extreme care, to avoid spasm. The vein is dissected free first and mobilized over proximal and distal ligatures. Occasionally it is necessary to ligate the inferior thyroid vein. The common carotid artery lies medial and posterior, contains no branches, and is mobilized in a similar fashion. The vagus nerve should be identified and protected from injury

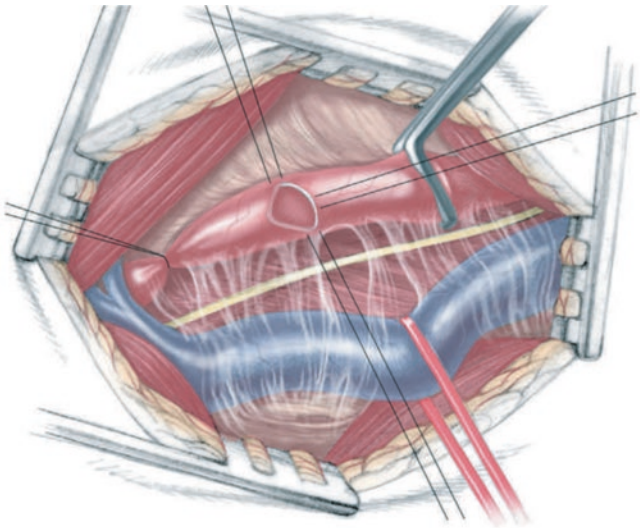

Fig. 17.5 The patient is then systemically heparinized with 50–100 U/kg heparin, which is allowed to circulate for 3 min. The arterial cannula (usually 10F for newborns) is measured so that the tip will lie at the junction of the brachiocephalic artery and the aorta (2.5 cm, one third the distance between the sternal notch and the xiphoid). The venous cannula (12–14 F for neonates) is measured to have its tip in the distal right atrium (6 cm, one half the distance between the suprasternal notch and the xiphoid process). For venoarterial (VA) bypass, the carotid artery is ligated cranially. Proximal control is obtained with an angled clamp, and a transverse arteriotomy is made near the ligature. Stay sutures, using 5/0 or 6/0 Prolene, are placed through the full thickness of the medial and lateral proximal edges of the arteriotomy to help prevent subintimal dissection. The sutures are gently retracted and the clamp is slowly released as the arterial catheter is inserted into the carotid artery to its proper position. The cannula is then fastened into place with two silk ligatures (2/0), with a small piece of vessel loop, on the anterior aspect, inside the ligatures to protect the vessel from injury during decannulation

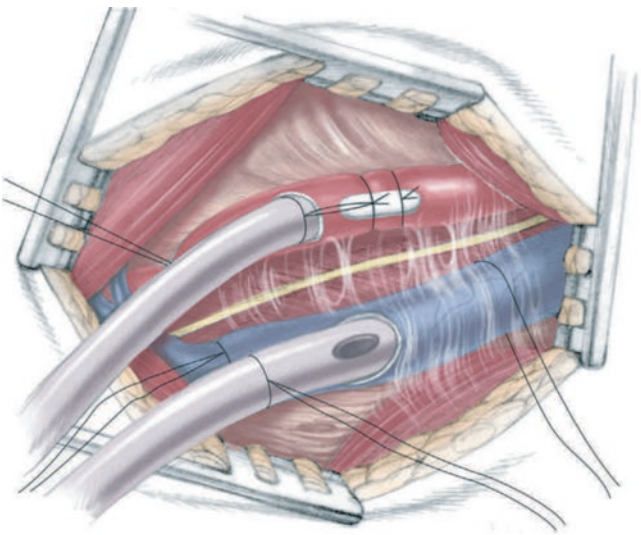

Fig. 17.6 In preparation for the venous cannulation, the patient is given succinylcholine to prevent spontaneous respiration. The vein is then ligated cranially. Gentle traction is placed on the lower ligature to help decrease back bleeding, and a venotomy is made close to the proximal ligation. The drainage catheter is passed to the level of the right atrium and secured in a manner similar to that used for the arterial catheter. The cannulas are then debubbled with back bleeding and heparinized saline. Then they are connected to the ECMO circuit and bypass is initiated

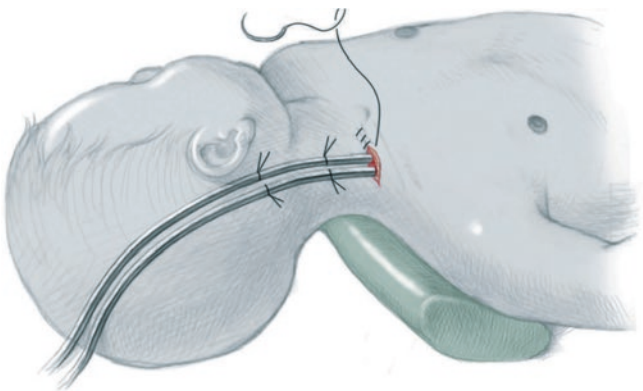

Fig. 17.7 Both cannulas are then secured to the mastoid process using 2–0 suture. The wound is irrigated, meticulous haemostasis is obtained, and the skin is closed in with a running nylon. The site is covered with a sterile dressing and the circuit tubing is fixed securely to the bed

17.3 Patient Management in ECMO

Once the cannulas are connected to the circuit, bypass is initiated and flow is slowly increased to 100–150 mL/kg/min so that the patient is stabilized. Continuous inline monitoring of the venous (prepump) SvO_2 and arterial (postpump) PaO_2, as well as pulse oximetry, is vital. The goal of VA ECMO is to maintain a mixed venous PO_2 (SvO_2) of 37–40 mmHg and saturation of 65–70%. VV ECMO is more difficult to monitor because of variation in the degree of recirculation, which may produce a falsely elevated SvO_2 assessment. Inadequate oxygenation and perfusion are indicated by metabolic acidosis, oliguria, and hypotension. Arterial blood gasses should be monitored hourly, with PaO_2 and $PaCO_2$ maintained as close to normal level as possible. As soon as these parameters are met, all vasoactive drugs are weaned, and ventilator levels are adjusted to "rest" settings. Gastrointestinal prophylaxis is initiated and mild sedation and analgesia is provided, usually with fentanyl and midazolam; the use of a paralyzing agent is avoided. A cephalosporin is often administered for prophylaxis. Routine blood, urine, and tracheal cultures should be taken.

Heparin is administered (typically 30–60 mg/kg/h) throughout the ECMO course, to keep the circuit free of thrombus. The activated clotting time (ACT) should be monitored hourly and maintained at 180–220 s. A complete blood count should be obtained every 6 h, and coagulation profiles daily. To prevent hemorrhage, platelets are transfused to maintain a platelet count above 100,000 mm^{-3}, and some authors sustain fibrinogen levels above 150 mg/dL. To optimize oxygen delivery, red blood cell transfusions can be used to keep the haematocrit above 40%.

Volume management of patients on ECMO is extremely important. It is imperative that all inputs and outputs be diligently recorded and electrolytes monitored every 6 h. All fluid losses should be repleted, and electrolyte abnormalities corrected. All patients should receive maintenance fluids as well as adequate nutrition using hyperalimentation. The first 48–72 h of ECMO typically involves fluid extravasation into the soft tissues. The patient becomes oedematous and may require volume replacement (crystalloid, colloid, or blood products) in order to maintain adequate intravascular and bypass flows, haemodynamics, and urine output greater than 1 mL/kg/h. By the third day of bypass, diuresis of the excess extracellular fluid begins and can be facilitated with the use of furosemide, if necessary.

Surgical procedures, such as CDH repair, may be performed while the child remains on bypass. Haemorrhagic complications are a frequent morbidity associated with this situation, increasing mortality. To avoid these complications, prior to the procedure the platelet count should be greater than 100,000 mm^{-3}, the fibrinogen level should be above 150 mg/dL, the ACT is reduced to 180–200 s, and the ECMO flow is increased to full support. It is imperative that meticulous haemostasis be obtained throughout the surgery. A useful adjunct is fibrinolysis inhibitor aminocaproic acid (100 mg/kg) just prior to incision, followed by a continuous infusion (30 mg/kg/h) until all evidence of bleeding ceases.

As the patient improves, the flow of the circuit may be weaned at a rate of 10–20 mL/h as long as the patient maintains good oxygenation and perfusion. Flows should be decreased to 30–50 mL/kg/min and the ACT should be at a higher level (200–220 s) to prevent thrombosis. Moderate conventional ventilator settings are used, but higher settings can be used if the patient needs to be weaned from ECMO urgently. If the child tolerates the low flow, all medications and fluids should be switched to vascular access on the patient, and the cannulas may be clamped, with the circuit bypassing the patient via the bridge. The patient is then observed for 2–4 h; if the bypass of the circuit is tolerated, decannulation should be performed, done under sterile conditions with muscle relaxant on board to prevent air aspiration into the vein. The catheters are removed and the vessels are ligated. The wound should be irrigated and closed over a small drain, which is removed 24 h later.

17.4 Complications

Extracranial bleeding—either at the site of cannulation or at other sites—is a common complication of the heparinized ECMO patient and is noted in 21% of neonatal cases, 44% of paediatric respiratory cases, and 40% of all cardiac cases. Bleeding at the site of cannulation can often be treated with local pressure or the placement of topical haemostatic agents such as Gelfoam, Surgicel, or topical thrombin. For all sites of bleeding, the platelet count should be increased to >100,000 mm^3 and the ACT lowered to 180–200 s. Sometimes the temporary discontinuation of heparin and normalization of the coagulation status is warranted to help stop the haemorrhage, with availability of a second circuit should acute clotting of the circuit occur. Aggressive surgical intervention is warranted if bleeding persists.

Neurologic sequelae, including learning disorders, motor dysfunction, and cerebral palsy, are a serious morbidity of the ECMO population. These outcomes appear to be due as much to hypoxia and acidosis prior to the ECMO course as to the time on ECMO itself. ICH is the most devastating complication, occurring in 7.4% of newborn patients, with an associated 57% mortality among newborns who have ICH on ECMO. Frequent comprehensive neurologic exams should be performed and cranial ultrasounds obtained daily for the first days of ECMO, with later intervals based on local protocols.

Blood pressure should be carefully monitored and maintained within normal parameters to help decrease the risk of ICH. If necessary, electroencephalograms may be helpful in the neurologic evaluation.

Acute tubular necrosis (ATN), marked by oliguria and increasing blood urea nitrogen and creatinine levels, is often seen in the ECMO patient during the initial 48 h, at which time renal function is expected to improve. If improvement does not occur, poor tissue perfusion must be considered, which may be due to low cardiac output, insufficient intravascular volume, or inadequate pump flow; all of these should be corrected. In the event of continued renal failure, haemofiltration or haemodialysis can be performed to maintain proper fluid balance and electrolyte levels. These are reported to be required in 14% of cases.

Conclusions

As of January 2015, 27,728 neonates (74% survival) and 6569 paediatric patients (57% survival) have been treated with ECMO for respiratory failure, and 13,124 neonatal and pediatric patients have been treated for cardiac failure. In the neonatal population, MAS is the most common indication for ECMO and carries with it a survival rate of 94%. Other frequent diagnoses (with survival rates in parentheses) include PPHN (77%), sepsis (73%), and CDH (51%). Viral pneumonia is the most common indication requiring ECMO amongst the paediatric population, and has 65% survival. Aspiration carries the greatest survival at 68%, whereas non-ARDS respiratory failure has a 54% survival, ARDS 56%, and bacterial pneumonia 59%. Cardiac patients have an overall survival of 46%. Specifically, congenital defects have 49% survival, cardiomyopathy 61%, and myocarditis the highest survival rate, 67%.

Recent medical advances such as permissive hypercapnia, inhaled nitric oxide, and the use of oscillatory ventilation have spared numerous babies from ECMO, yet many children still benefit from this modality.

In summary, any patient with reversible cardiopulmonary disease who meets the criteria should be considered an ECMO candidate. ECMO provides an excellent opportunity to provide "rest" to the cardiopulmonary systems, thus avoiding the additional lung or cardiac injury which otherwise would be necessary to maintain life support.

Suggested Reading

Campbell BT, Braun TM, Schumacher RE, Bartlett RH, Hirschl RB. Impact of ECMO on neonatal mortality in Michigan (1980–1999). J Pediatr Surg. 2003;38:290–5.

Extracorporeal Life Support Organization. International registry report of the extracorporeal life support organization. Ann Arbor, MI: University of Michigan Medical Center; 2015.

Hirschl RB, Bartlett RH. Extracorporeal life support in cardiopulmonary failure. In: Coran AG, Adzick NS, Krummerl T, Laberge JM, Shamberger R, Caldamone A, editors. Pediatric surgery. 7th ed. New York, NY: Mosby; 2012. p. 123–32.

Kim ES, Stolar CJ. ECMO in the newborn. Am J Perinatol. 2000;17:345–56.

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Pediatric Laparoscopic Surgery

18

Amulya K. Saxena

The practice of pediatric laparoscopic surgery approaches patients ranging in size from neonates through infants, toddlers, children, and teenagers, to obese adolescents. Unlike laparoscopic surgery in adults, these size variations in patients are associated with conditions that are rather specific to the age groups: neonatal procedures are most often focused on the management of congenital malformations and pylorus hypertrophy, whereas procedures in infants, toddlers, children, and teenagers deal with a variety of pathological conditions frequently specific to each age group. With the rapid rise in pediatric obesity, laparoscopic surgery in this subgroup of adolescents may include not only the array of bariatric procedures aimed at achieving weight loss, but also procedures to manage common conditions such as appendicitis and gallstones in these patients, thus involving pediatric surgeons who generally do not perform bariatric procedures.

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18.1 Indications

In the past two decades, laparoscopic surgery by enthusiastic pediatric surgeons has grown rapidly from simple diagnostics to complex surgical corrections, winning over skeptics who were initially having doubts or reservations. The present mindset among pediatric surgeons has shifted from whether laparoscopic procedures should be performed in the pediatric population to which conditions are best suited for laparoscopic surgery. Although the principles of laparoscopic surgery in the pediatric population remain the same as in the adult population, miniaturization of instruments and better understanding of the responses of the body to laparoscopic procedures have been instrumental in its safer implementation and greater success.

The following procedures are common indications for laparoscopic surgery by pediatric surgeons:

Appendectomy.
Adrenalectomy.
Antegrade continence enema (ACE) procedure.
Bile duct exploration.
Biliary pancreatic diversion.
Bowel resection.
Cholecystectomy.
Diagnostic laparoscopy.
Drainage of pancreatic cysts.
Exploration for undescended testicle.
Exploration and treatment for inguinal hernia.
Enteric adhesions separation.
Fundoplication.
Gastrostomy.
Heller myotomy (achalasia).
Hepaticojejunostomy (Kasai procedure).
Jejunostomy.
Liver biopsy.
Lymphadenectomy.
Nephrectomy.
Oophorectomy and cystectomy.

P. Puri, M. E. Höllwarth (eds.), *Pediatric Surgery*, Springer Surgery Atlas Series,
https://doi.org/10.1007/978-3-662-56282-6_18

Orchidopexy.
Paraesophageal hernia repairs.
Pelvic exploration (tubo-ovarian pathology).
Pull through for Hirschsprung's disease.
Pull through for imperforate anus.
Pyeloplasty.
Pyloromyotomy.
Rectopexy (rectal prolapse).
Reduction of intussusception.
Repair of diaphragmatic hernia.
Resection of Meckel's diverticulum.
Resection of omental and mesenteric cyst.
Splenectomy.
Treatment of vesico-ureteral reflux.
Tumor biopsy and/or staging.
Varicocelectomy.

18.2 Ergonomics

Ergonomics plays an important role in the outcomes of laparoscopic procedures. The three most important ergonomic considerations are the operating room setup, the posture of the surgical team, and the positioning of the ports.

18.2.1 Operating Room Setup

The setup of the operating room is dictated by the equipment to be used, and can be broadly divided among among three options: the single-monitor option, the dual-monitor option, and the operating suite. In the single-monitor option, a monitor is fixed on a laparoscopic equipment trolley and is positioned in such a way that the members of the surgical team can view the procedure on the screen from their respective positions (Fig. 18.1a). In the dual-monitor option, the laparoscopic equipment trolley has a monitor placed on the top of it, and a second monitor is attached on a swivel arm for flexibility of movement. This option offers better team placement around the operating table and improves visualization of the procedures by the team assisting the surgeon (Fig. 18.1b). Operating suites for laparoscopic surgery offer an equipment tower that is suspended from the roof of the operating room, as well as multiple independent monitors that can be optimally placed to meet the requirements of each individual member of the team (Fig. 18.1c).

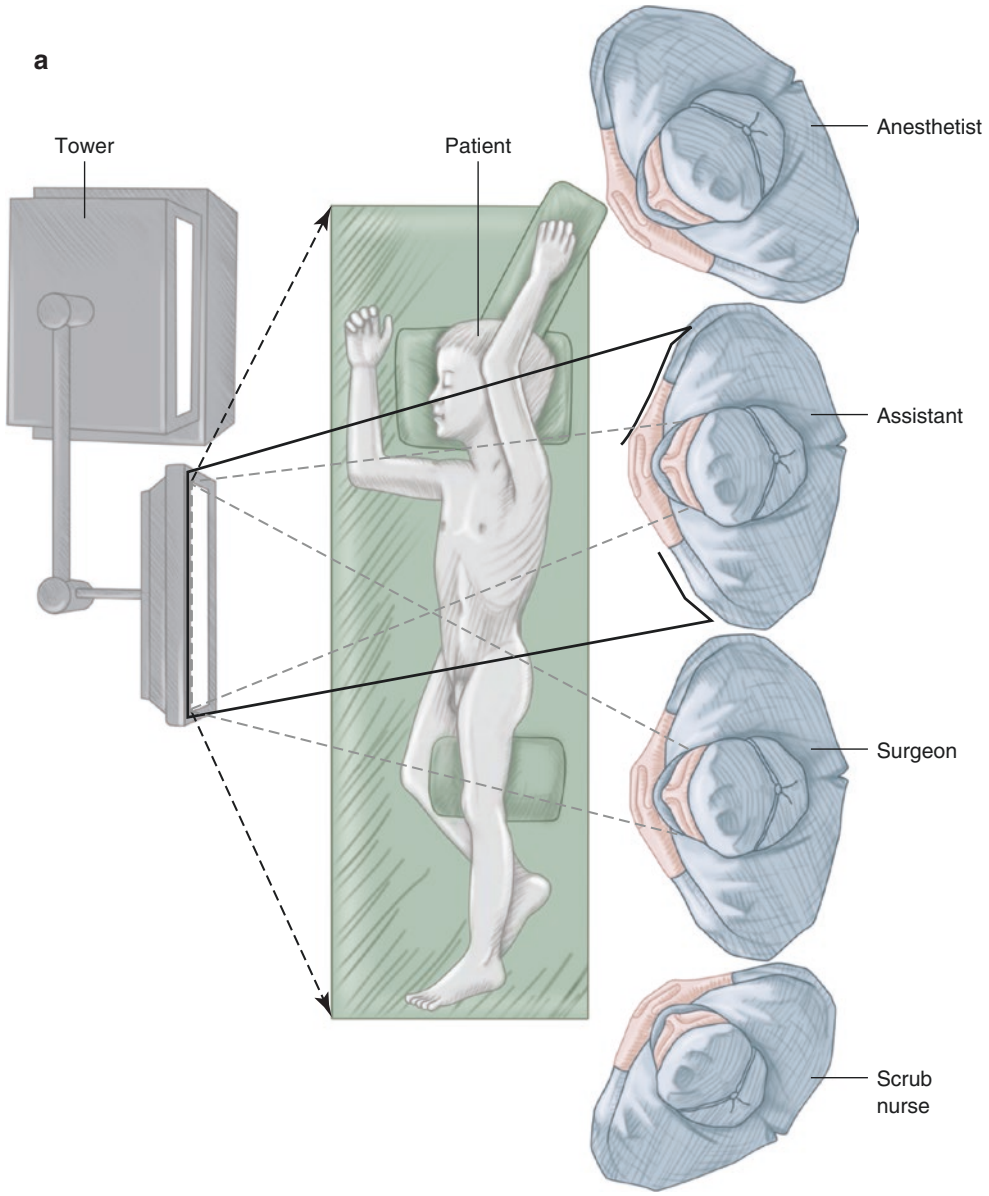

Fig. 18.1 Setup of the operating room: (**a**) Single-monitor option. (**b**) Dual-monitor option and (**c**) Operating room suite

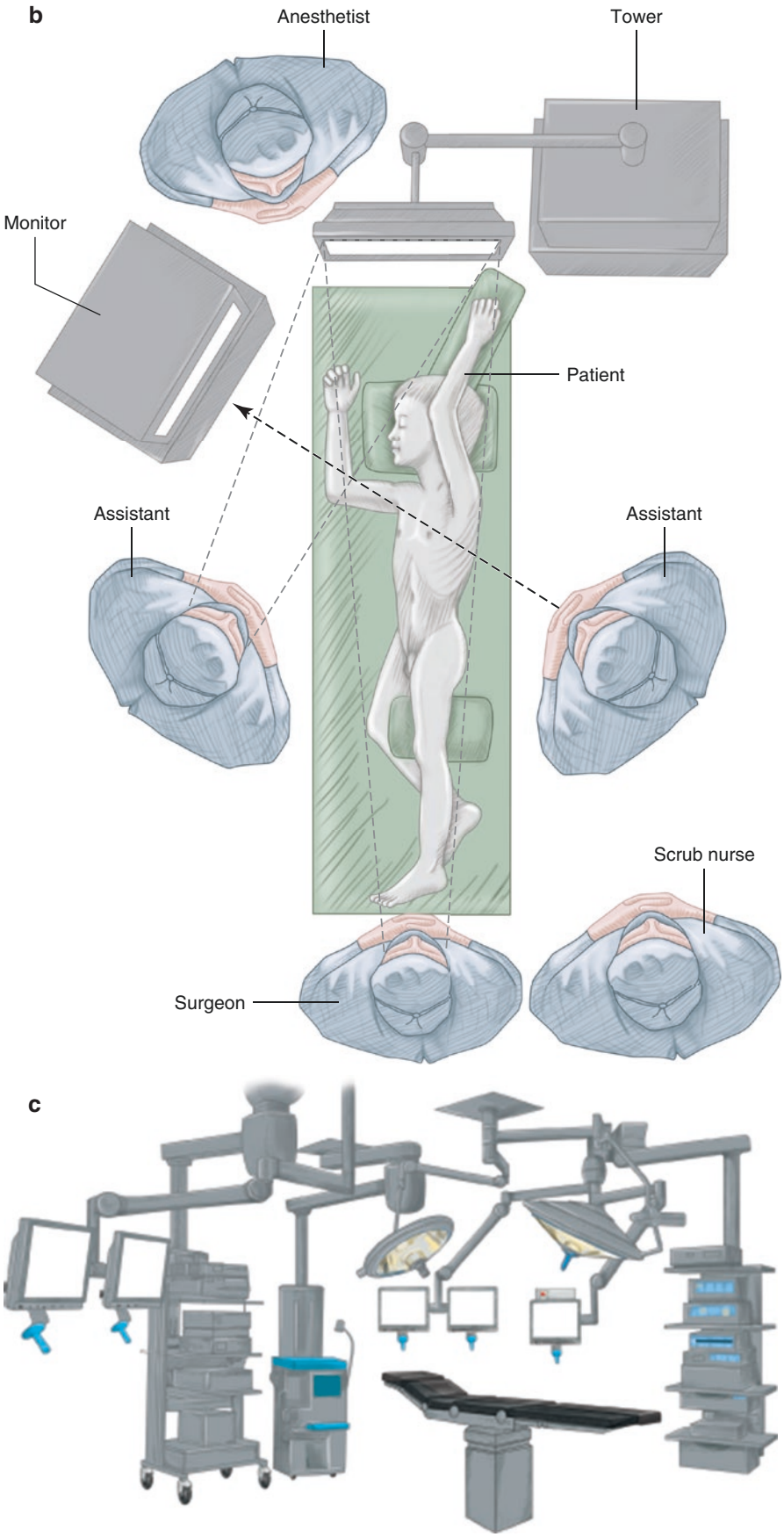

Fig. 18.1 (continued)

18.2.2 Posture of the Surgical Team

The posture of the surgical team with regard to patient positioning is another important factor that should be considered in the ergonomics of laparoscopic surgery. Because single-monitor setups with fixed trolleys set the trend for laparoscopic surgery, schools of laparoscopic surgery advocated straight posture of the surgeon with 90° flexion of the elbow and the operating table positioned at an appropriate height. A straight line of sight was also advocated for the surgeon, but with flat screens, a tilt of 10°–20° could be offered (Fig. 18.2a). This posture cannot be considered to be ergonomically optimal, however, numerous reports on posture-related fatigue and difficulties in completing procedures were published by groups that evaluated ergonomics. For complex procedures or procedures that require a longer time to complete, the monitor is placed so that the line of sight does not require extension of the neck (Fig. 18.2b). Also, the position of the elbows is most comfortable when the hands are 110°–120° from the shoulders. To enable this posture, the patient table is positioned slightly lower. The position of the wrist in relation to the instrument handle also plays an important role in performing procedures, as strength of the hand is affected when there is an unlar or radial wrist deviation (Fig. 18.3). Attention also must be paid to the ergonomics of the camera operator, with options such as having the camera operator seated during the entire procedure, or offering sterile gauze pads on which to rest the elbows during a long procedure.

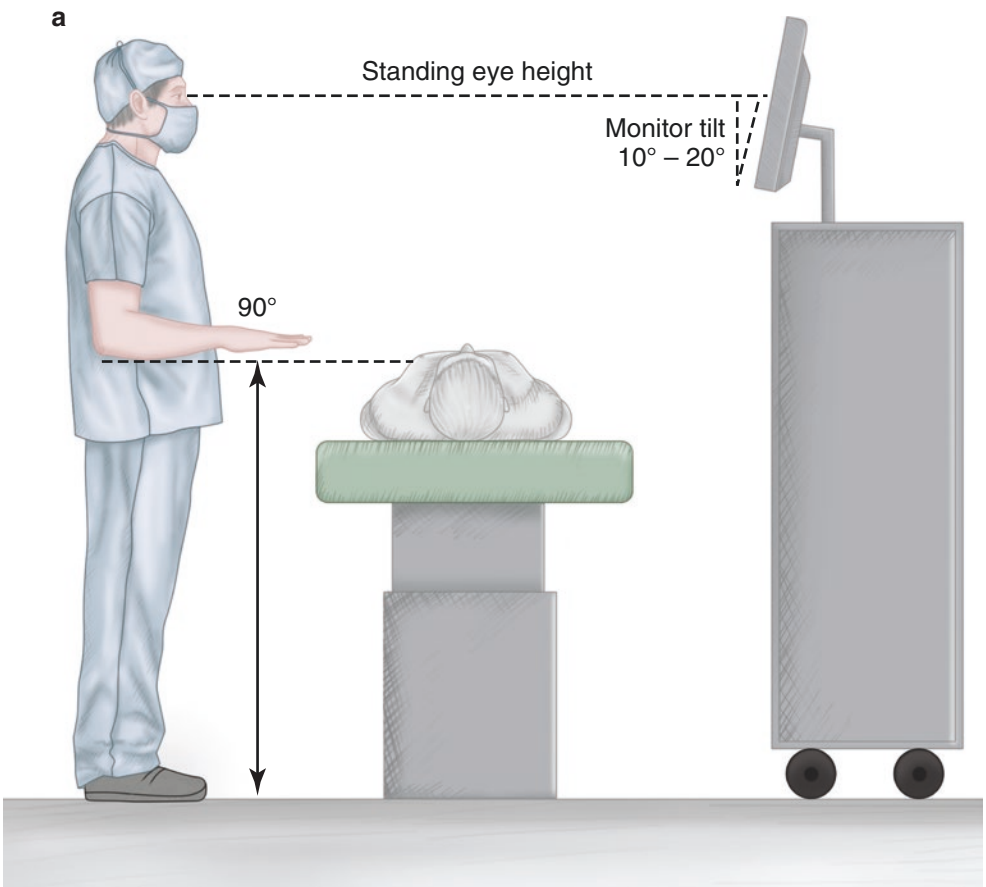

Fig. 18.2 Posture of the surgeon. (**a**) Traditional straight posture. (**b**) Improved ergonomic posture

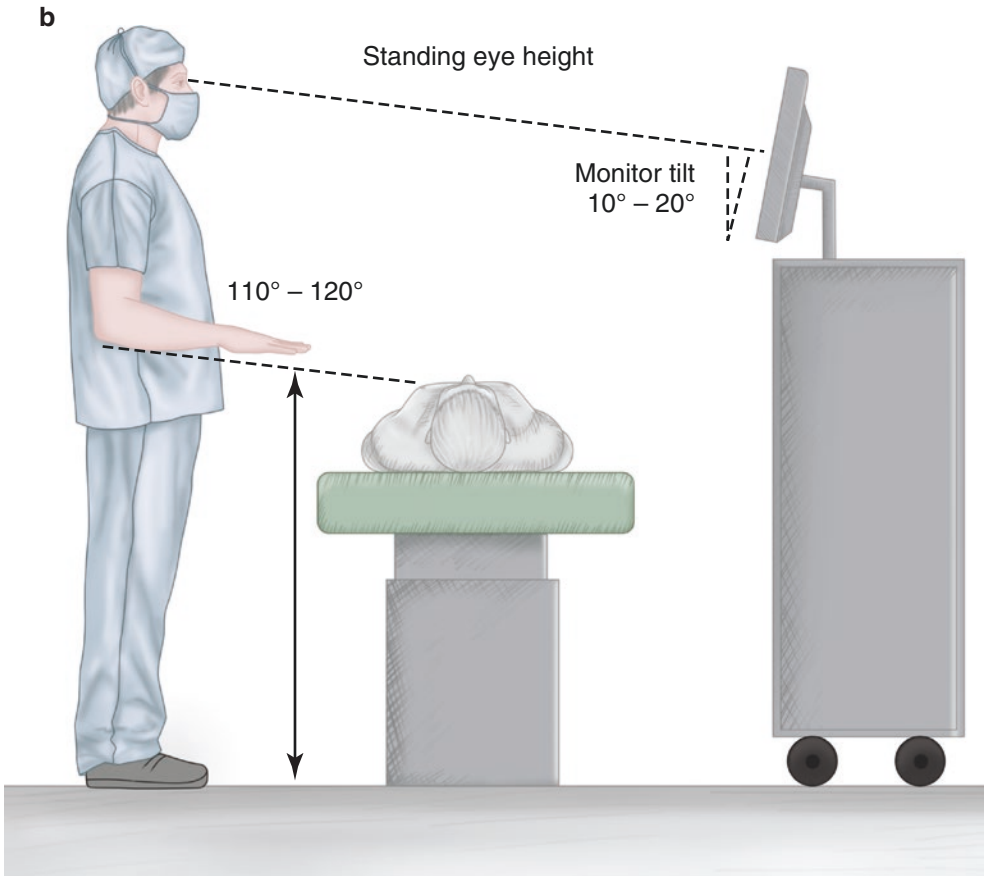

Fig. 18. 2 (continued)

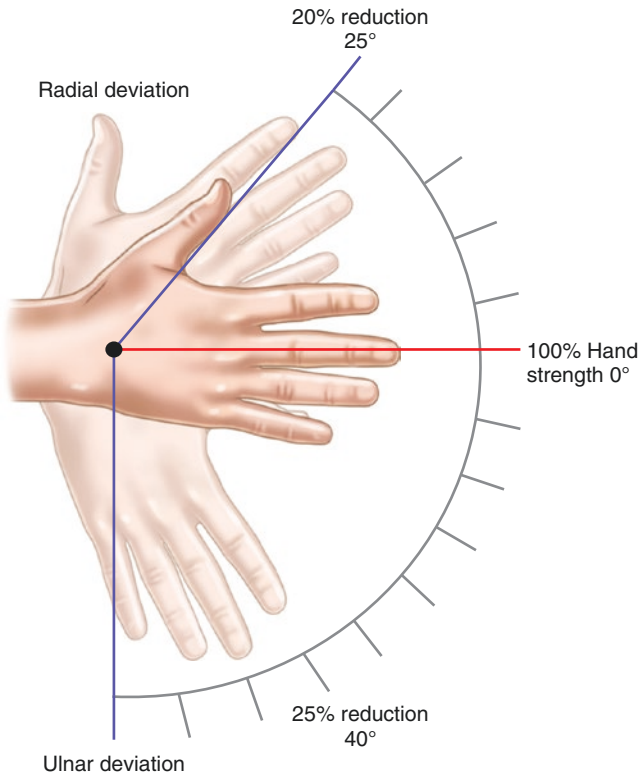

Fig. 18.3 Variations in strength of the hand during laparoscopic procedures when maintained at different angles to the wrist

18.2.3 Placement of Ports

Placement of ports is the third important aspect when considering the ergonomics of laparoscopic surgery. The degrees of movement of the instrument handles outside the abdomen are disproportionate to the degrees of movement of the instrument tips at the site of the procedure, a fact that should be considered during the placement of the ports (Fig. 18.4a). For procedures that involve suturing and knot tying, triangulated placement of the ports is necessary. The camera port in placed in line with the target organ, and the work ports are placed on either side of the camera port but positioned closer to the target organ, considering the degrees of movement and the working distance (Fig. 18.4b). Ideal working distance for manipulation, suturing, and knot tying is 10 cm between the work ports, which enables a 4-cm distance at the target organ to comfortably complete the procedural steps (Fig. 18.4c). In laparoscopic procedures performed on neonates and infants, often it is not possible to attain these optimal distances between the work ports and the optic port. In such cases, the optic port is commonly repositioned from the umbilicus to an alternative site to gain triangulation. Cluttering of instruments should be avoided, as multiple instruments make it more difficult to complete the procedure (Fig. 18.4d).

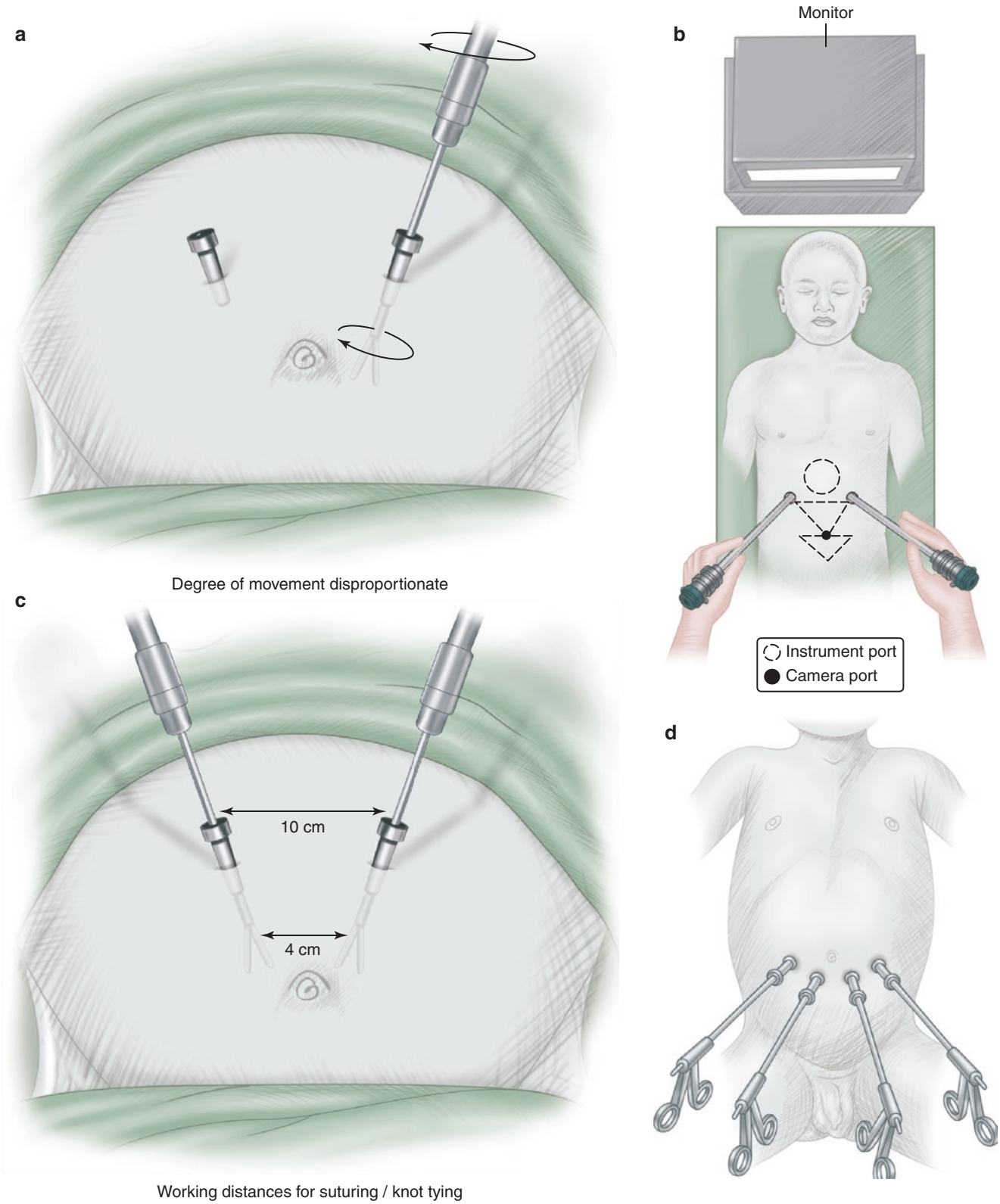

Fig. 18.4 Ideal placement of work ports: (**a**) Consider disproportionate movement of instruments inside and outside the abdomen, (**b**) Desired positioning of the camera and work ports to achieve triangulation, (**c**) Preferred working distances between work ports for suturing and knot tying and (**d**) Avoid instrument cluttering

18.3 Access

Access to the abdominal cavity can be gained using the open access method (Hasson's technique) or the closed access method (Veress needle). In the open access method, entry is gained into the cavities using open surgical methods with placements of ports under complete vision, whereas in the closed method, entry is gained by the insertion of a Veress needle through the immediate subumbilical area in a previously unoperated abdomen. Abdominal access through the umbilicus can be achieved through a superior or inferior umbilical crease incision or a lateral umbilical fold incision, or it can be directly transumbilical. Umbilical access offers several advantages: the area is the thinnest part of abdominal wall; as a central point of the abdomen, it offers an optimal view; it is devoid of major blood vessels; and it gives good esthetic results in scar concealment.

The open-access technique is safe and has found widespread acceptance in pediatric laparoscopic surgery. For this access method, the umbilicus is meticulously cleaned using betadine, and the umbilical folds are held between the forceps. The author's preference is for the left umbilical fold, as this area is completely devoid of embryonic structures that are attached to the umbilicus. Figure 18.5 illustrates the steps in this technique.

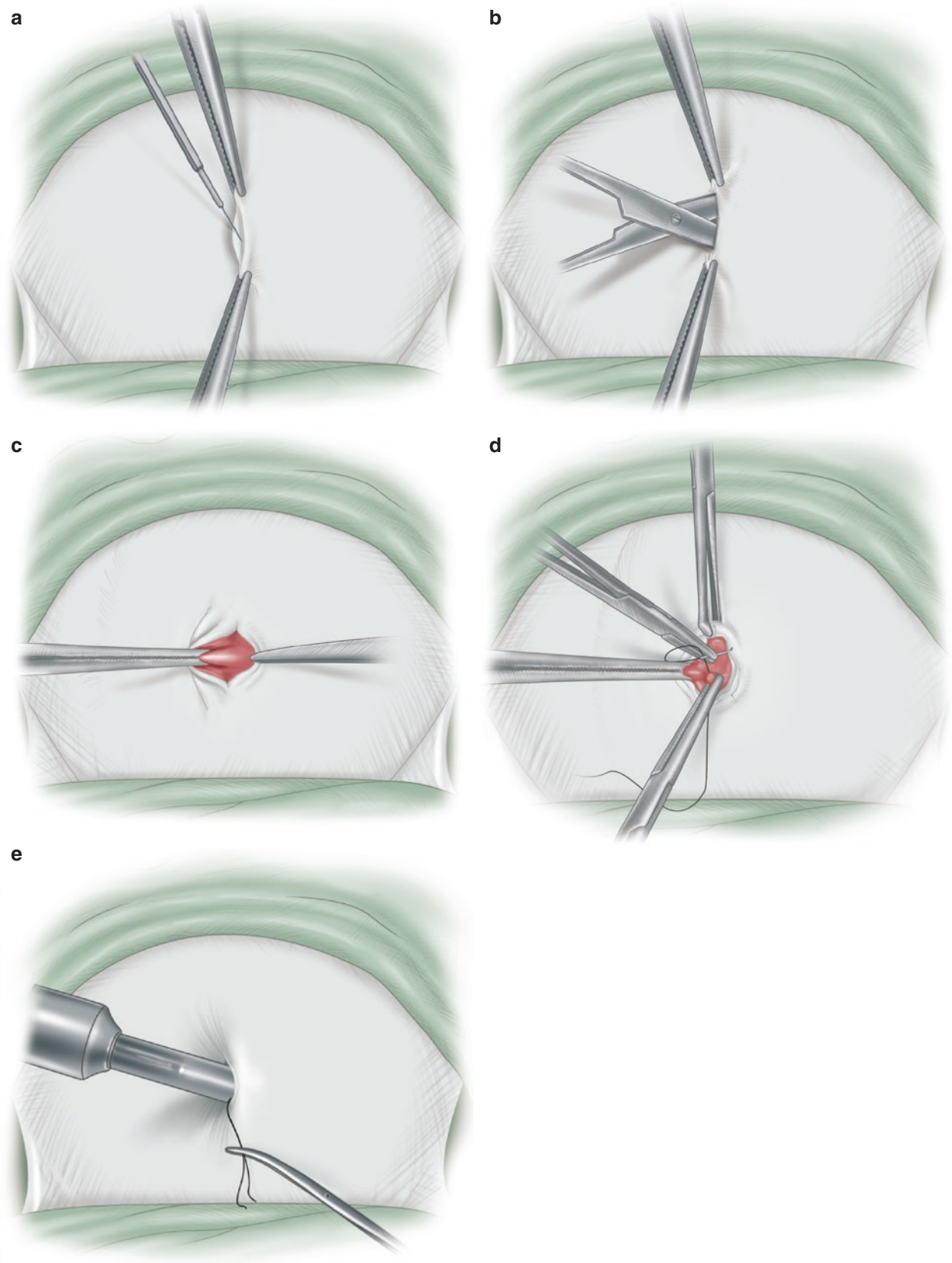

Fig. 18.5 Open access technique. (**a**) The forceps are held in such a way as to offer tension on the umbilical fold to aid in the incision using a scalpel. (**b**) Then, using a scissor, blunt dissection of the subcutaneous tissue is performed to expose the fascia. (**c**) The fascia is incised to expose the peritoneum, which is then breached at this point. (**d**) A purse-string suture is placed to secure the edges of the peritoneum and fascia. (**e**) The purse-string suture is further used to secure the port and to minimize gas leaks. The suture can be tied to close the peritoneum and fascia after completion of the procedure

18.4 Port Systems

Port systems are available with variations in three components: the trocar, the port valve, and the port tip.

18.4.1 Trocars

Trocars may have a blunt, conical, or pyramidal tip (Fig. 18.6a). Blunt-tip trocars are used in the open access technique to aid in the insertion of the port and to exchange ports or replace a displaced port. Conical trocars cause less trauma and less gapping of the abdominal wall wound, but they require more force for their insertion, which consequently increases the risk of visceral injury. The author prefers to incise the skin at the site of the work port and create a tunnel with a scissor until the peritoneum is reached; the port can then be inserted with ease through this tunnel, using a blunt trocar with little or no force, thereby diminishing the possibility of visceral injury. A pyramidal trocar has a pointed cutting tip that requires less force for tissue penetration, thus reducing the risk of visceral injury but increasing the chance of injuring abdominal wall vessels.

18.4.2 Port Valves

Four valve types are used in ports to prevent the escape of insufflated carbon dioxide: flap, ball, trumpet, or diaphragm (Fig. 18.6b). Most ports in contemporary use have a flap valve, with most of those having an external lever. These valves tend to malfunction if tissue gets trapped in the valve mechanism. Also, introducing of sutures through these valves without a suture introducer may cause the needle and suture to be trapped in the valve, a problem that can often be avoided by keeping the valve completely open during introduction of the suture. Likewise, during the removal of instruments, the instruments can become jammed in the valve and cause unintentional removal of the port. This can be prevented by securing the port with a suture, holding the port at this time with the hand, or keeping the valve completely open during instrument removal. The ball valve (fixed or magnet) is an easy system to use, but it has not been popular in laparoscopic surgery. This valve also requires a suture introducer, as there is no lever. The wall of the trumpet valve seals well but manipulation is required with every instrument exchange. This valve can damage the telescope or instruments and is now rarely used. The soft Silastic diaphragm valve can accept different instrument sizes without the need of a reducer.

18.4.3 Port Tips

The port tips have been modified to enable better anchoring and securing of the port during instrument exchanges (Fig. 18.6c). If the port has a smooth sleeve tip (**a**), an anchoring suture may be tied to the valve nozzle to secure the port, but it will not prevent the port from dislocating into the abdomen. With these ports, the author's preference is to use a Steristrip or Silastic tube around the port sleeve, to which the anchoring suture can be secured, thus preventing dislocation in either direction. Likewise, a balloon or winged Malécot arrangement with an outside flange (**b**) can prevent bidirectional dislocation. Another option is a spiral retaining collar (**c**), through which the cannula can be passed. This type of port tip has not been popular. A port with a spiral tip sleeve (**d**) also offers a good alternative option for secure port fixation.

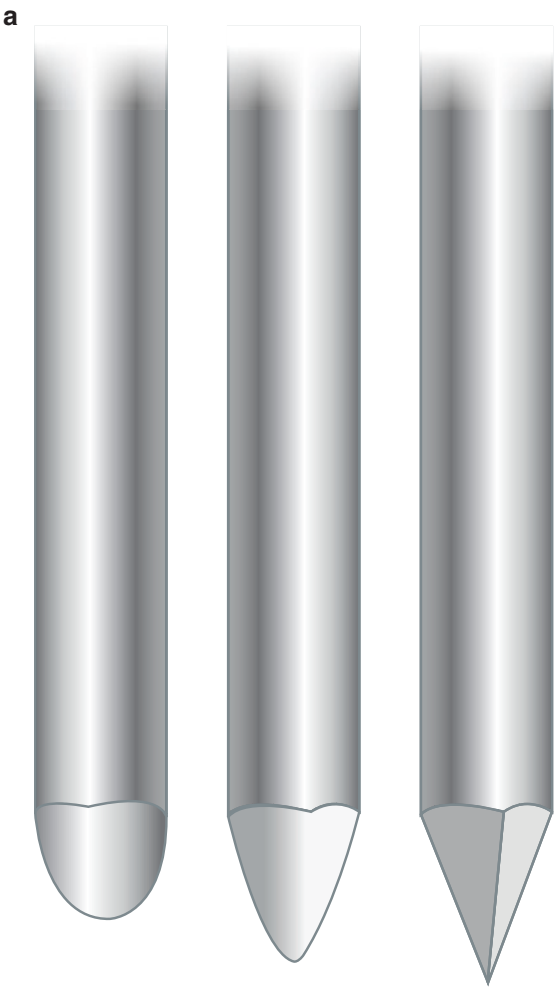

Fig. 18.6 Port systems: (**a**) Options in trocar tips, (**b**) Variations in port valves and (**c**) Port tip alternatives

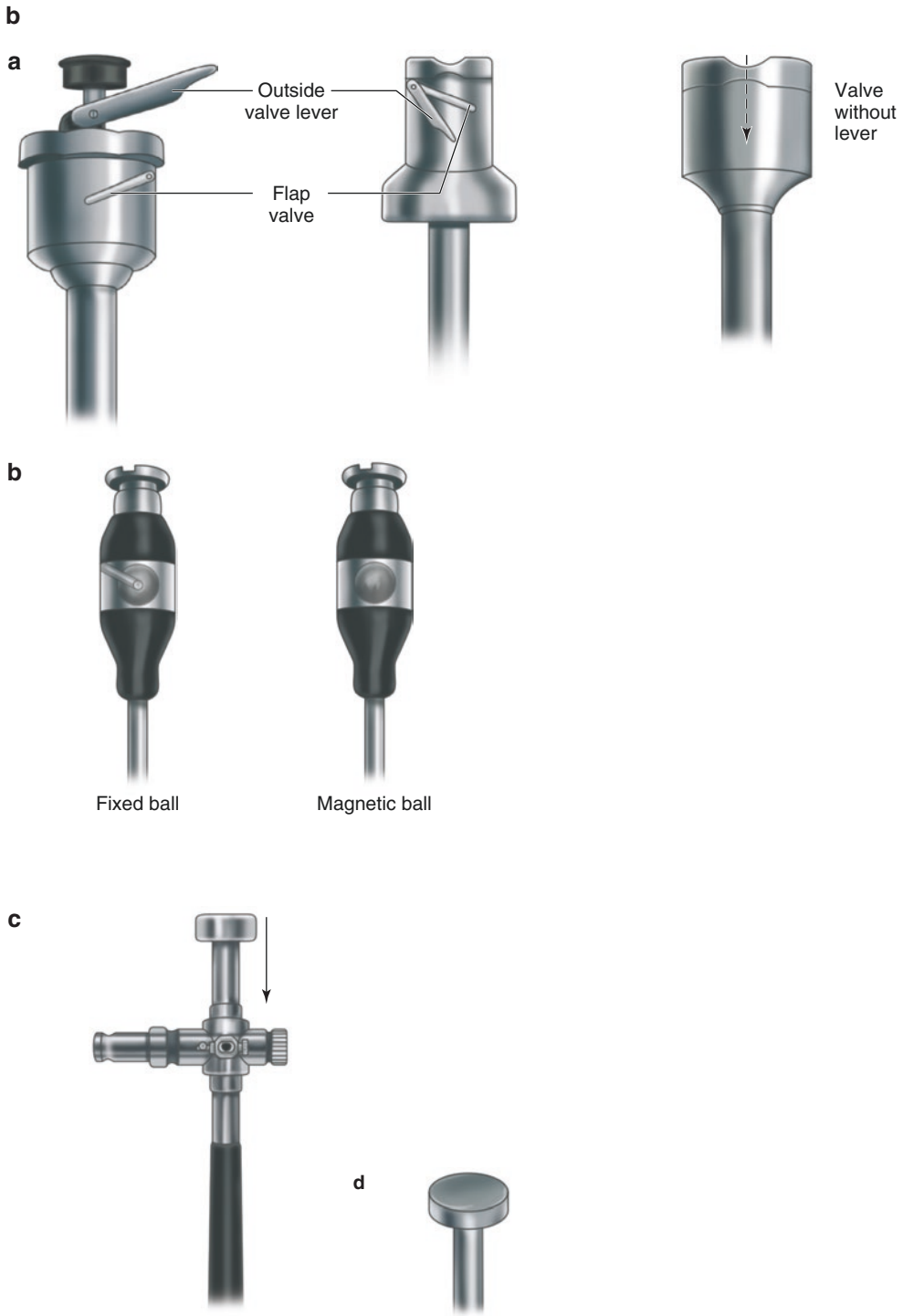

Fig. 18.6 (continued)

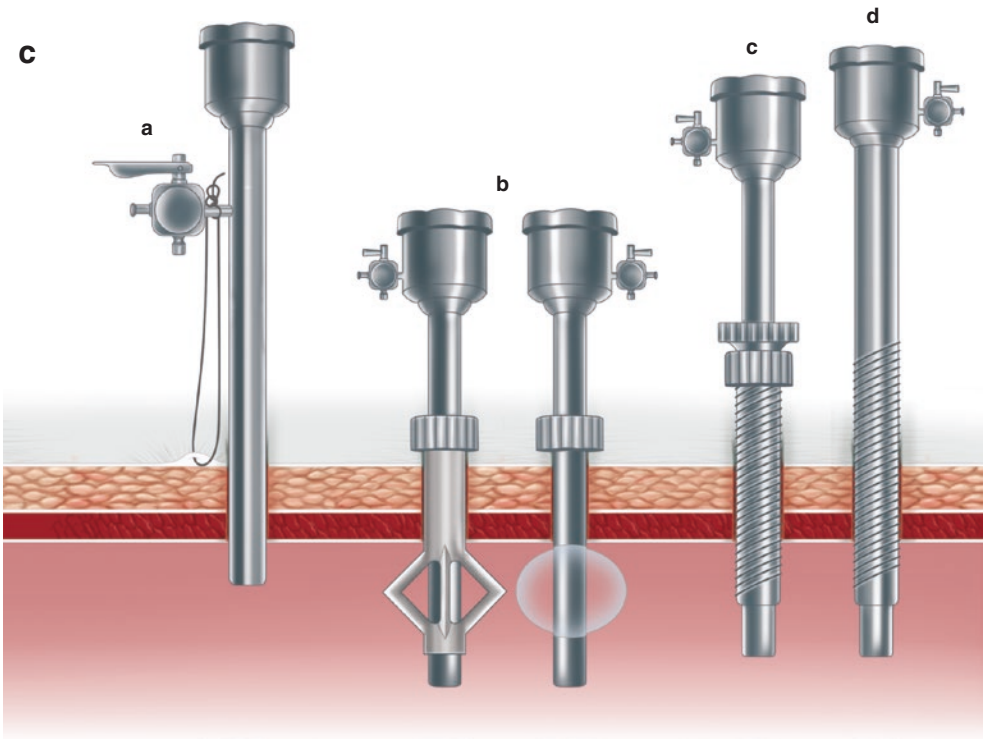

Fig. 18.6 (continued)

18.5 Pneumoperitoneum

Pneumoperitoneum is achieved to provide space in the abdominal cavity to perform laparoscopic procedures after the placement of the optic port (first port) and before the placement of the work ports. Flow should be started at an initial rate of 0.5 L/min to rule out any obstruction. Once the pneumoperitoneum has been established, the insufflation rate can be changed to a higher setting of approximately 0.5–2 L/min to speed up the process. When the limit is reached, the rate of flow can be decreased to 0.1–0.2 L/min.

The surgeon decides upon the pressure desired for the pneumoperitoneum. Although no absolute values can be recommended, pressures in the following ranges are considered to be safe:

- 6–8 mmHg for infants
- 8–12 mmHg for small children
- 12–15 mmHg for older children and adolescents

Figure 18.7 can be used as a guideline for safe insufflation pressures in pediatric patients, including those of normal weight as well as obese adolescents.

Rarely, complications could arise from CO_2 insufflation for pneumoperitoneum during laparoscopy. These include gas embolism, cardiovascular compromise, and hypercapnia. The risks are minimized by the use of low-pressure CO_2 insufflation in children. Slight increases in end tidal CO_2 and peak airway pressures that may be detectable intraoperatively can usually be compensated for by slight hyperventilation. A low complication rate (1–2%) of laparoscopic surgery in children has been observed in high-volume centers with regards to pneumoperitoneum.

Once pneumoperitoneum is established, it is compulsory to explore the entire abdominal cavity before insertion of the work ports. The inspection is commenced in the left upper quadrant and proceeds with 360° rotation of the scope in a clockwise direction. After assessing the presence of additional pathology, the work ports are placed into the abdomen. The procedures are then begun based on the abdominal pathology and the plan of management.

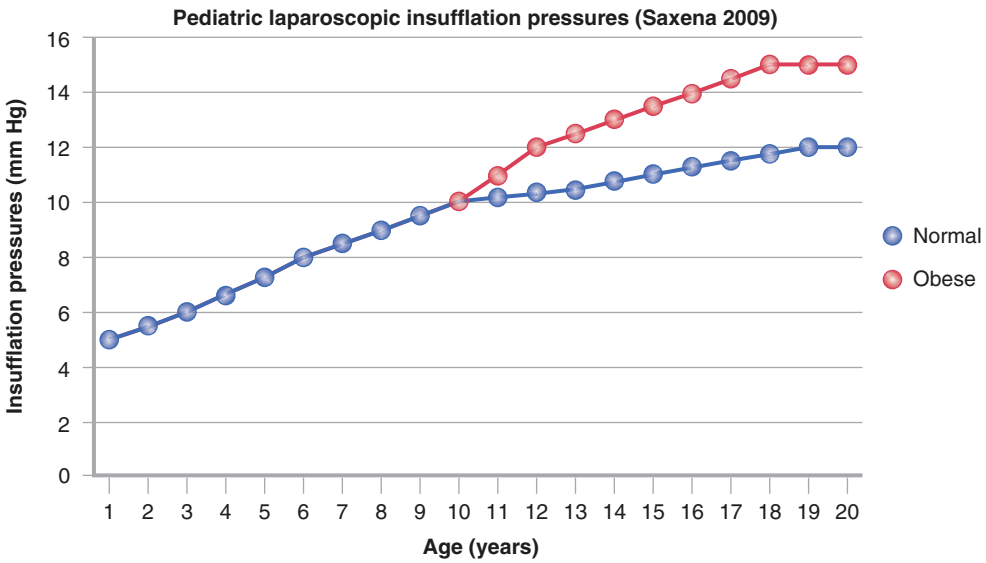

Fig. 18.7 Pediatric laparoscopic insufflation pressures (*Adapted from* Saxena AK. Effects of insufflation. In: Saxena AK, Höllwarth ME, editors. Essentials of pediatric endoscopic surgery. Springer-Verlag: Berlin; 2009. pp. 59–62)

18.6 Avoiding Complications of Electrosurgery

Minimal bleeding, which is a minor obstacle in open surgery, may completely obscure the view in endoscopic surgery and prevent safe tissue preparation. Therefore, in laparoscopic procedures, the surgeon is more reliant on the energy source for hemostasis and cutting than in open surgery. Several problems are inherent in the use of energy sources during laparoscopy. Hazards of electrosurgery in laparoscopic surgery include insulation failure, capacitive coupling, and direct coupling.

Hazards in insulation failure occur when exposed metal in the handle of the instrument may cause burns to the surgeon‘s hands. Further signs of insulation failure within the port by lower-frequency electrical currents must be recognized, as they can cause neuromuscular stimulation and jerking of the abdominal wall or diaphragm. Most dangerous, however, are the defects in the electrode shaft, which cause an injury outside the view of the surgeon (Fig. 18.8a).

Capacitive coupling occurs when electric current is transferred from one conductor (the active electrode), through intact insulation, into adjacent conductive materials (abdominal organs) without direct contact (Fig. 18.8b). The magnitude of the coupled charged is directly proportional to the voltage and the port diameter, hence capatcitive coupling is increased with use of higher voltages. Longer length of instruments, thinner insulation and narrow ports increase the risk of this type of injury. It is important to bear in mind that some degree of capacitive coupling occurs in all monopolar electrosurgical instruments. Capacitive coupling can be minimized by activating the active electrode when it is in contact with the intended tissue and by limiting the time of high-voltage peaks.

In direct coupling, the activated electrode touches other metal instruments, creating an environment for energy to be transferred to tissue outside the laparoscopic field of view (Fig. 18.8c). Ports housing conductive instruments should be made of metal to enable dissipation of stray energy over a large area and to reduce the heat production from the stray current.

A number of guidelines for use of monopolar devices in laparoscopic surgery should be followed:

- All insulations should be inspected for defects.
- Lower power settings should be used for both cutting and coagulation.
- Apply a low-voltage waveform whenever possible.
- A brief activation is better than a prolonged one.
- Metal-to-metal sparking (direct coupling) should be avoided.
- The electrode should be activated only when it is touching the target tissue.

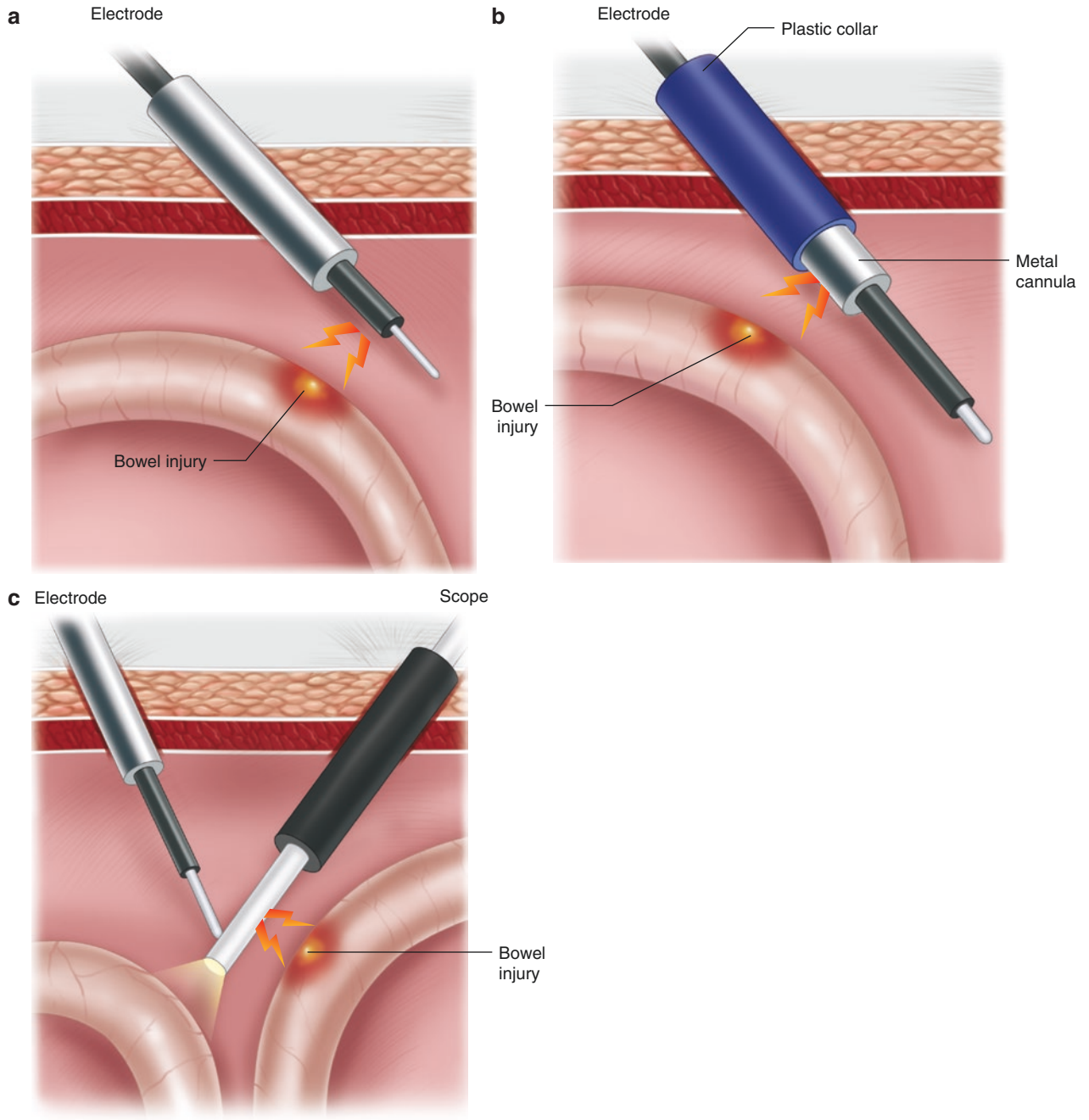

Fig. 18.8 Hazards of electrosurgery. (**a**) Injury caused by insulation failure in the electrode shaft. (**b**) Injury from capacitative coupling. (**c**) Injury from direct coupling

18.7 Removal of Instruments

After completion of a procedure, it is important to remove the work ports under vision. When work ports are no larger than 5 mm, the fascia is not closed and the port insertion site is closed with a 4–0 Vicryl subcutaneous stitch. Umbilical optic ports as well as work ports sites >5 mm require fascia closure, to avoid the risk of port site hernia, if left open. The umbilical optic port site incision is closed with the purse-string suture that was placed at the time of port insertion. If the suture was not placed initially or was detached during the course of the procedure, a new purse-string suture is placed to close the fascia. Care should be taken to avoid trapping of omentum at the time of knot-tying. Also, if any bleeding is observed from the port sites, careful exploration of the bleeding vessel should be performed and managed appropriately. Owing to the increase in abdominal pressure from insufflation, these bleeding vessels may be compromised and begin to bleed again once the abdomen is deflated. Care must be taken to identify these bleeding vessels at the time of completion of the procedure, as they bear the risk of either bleeding inside the abdomen or outside, necessitating later exploration under general anesthesia. Fascia closure of port sites >5 mm in obsese children and adolescents may be challenging and the application of special devices may be benificial as they offer the advantage of not enlarging the skin incision to reach the fascia and also secure closure of the fascia under vision (Fig. 18.9).

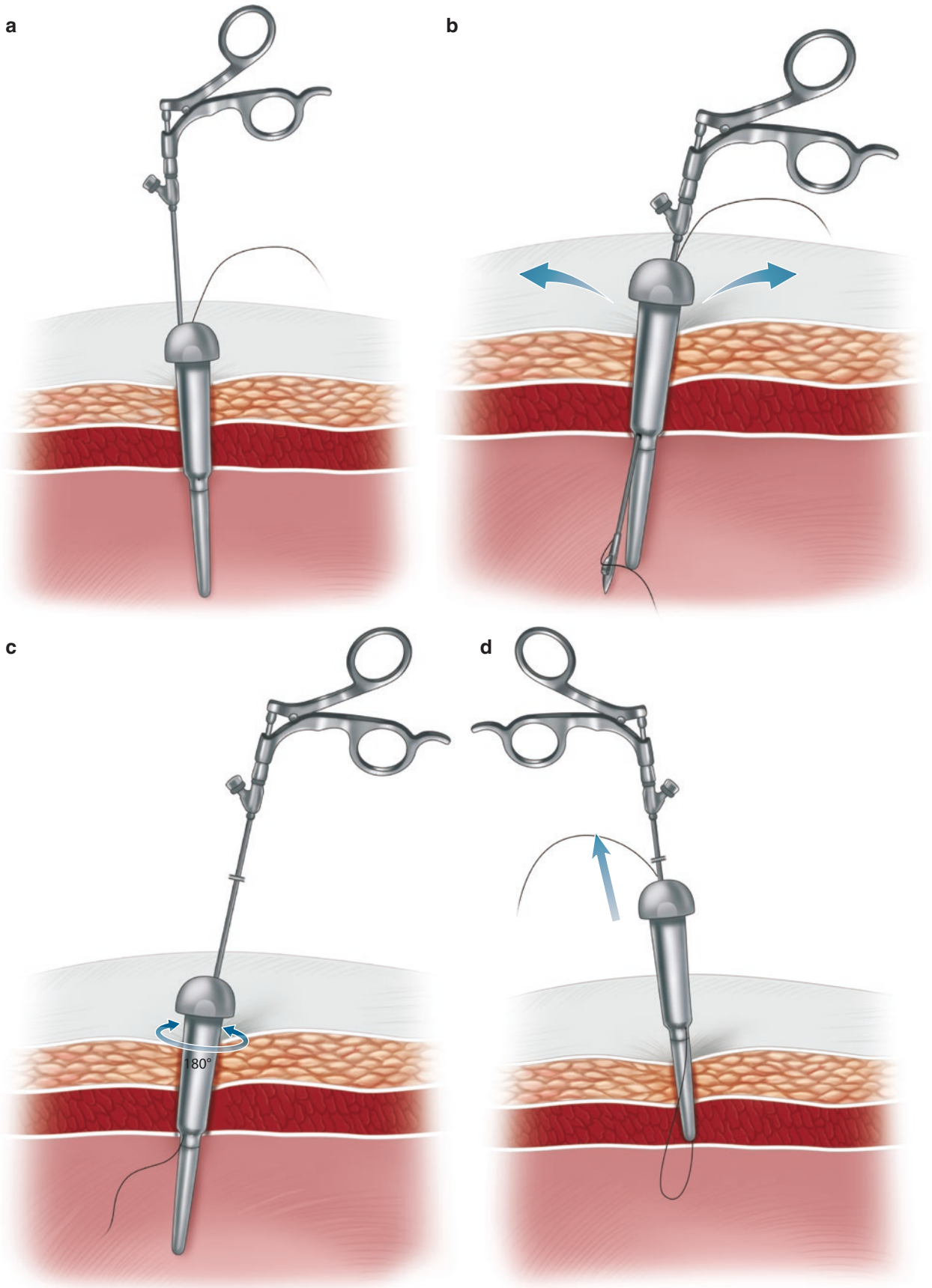

Fig. 18.9 (**a**–**d**) Busche device for secure closure of fascia under vision in obese adolescent children

Conclusions

Laparoscopic procedures, unlike open procedures, require the knowledge and application of high-technology equipment and new instruments with multiple functions. All laparoscopic procedures require a systematic approach to perform the operations safely. The checklist below should be used as a guide to perform "SECURE" procedures in laparoscopic surgery:

***S**copes:* Scope size and the angle of view (0°–70°) should be chosen in advance.

***E**quipment check:* Run an equipment check before every procedure.

***C**O_2:* Check gas level in the cylinder and set insufflator values (pressure/flow).

***U**nrestricted movement:* Have an adequate length of sterile cable/tube on the table.

***R**ecording:* Remember to turn the recording device "ON" when the procedure begins.

***E**mergency:* Have a back-up procedure plan and on-service help for equipment.

Suggested Reading

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Hernias: Inguinal, Femoral, Umbilical, Epigastric, and Hydrocele

19

Juan A. Tovar and Leopoldo Martinez

19.1 Introduction

Inguinal hernia is one of the most common surgical conditions in infancy, with a peak incidence during the first 3 months of life. The diagnosis of inguinal hernia is made with increasing frequency in newborns, and this period carries a particularly high risk of incarceration. The incidence of hernia also is much higher in premature infants, who now survive in growing numbers with sophisticated intensive care management. Direct hernia is exceedingly rare at this age; nearly all congenital indirect inguinal hernias develop because the processus vaginalis remains patent after birth. The most common presentation of inguinal hernia in a child is a groin bulge, extending towards the top of the scrotum. The treatment of inguinal hernia is always surgical. In infants and toddlers, herniotomy can be performed through the external inguinal orifice without any attempt at parietal reinforcement. In older children, however, the length of the canal makes it advisable to open the external oblique aponeurosis in order to achieve a high ligation of the sac. Although more than 80% of major surgical pediatric centers still prefer open technique, there are many surgeons who repair the hernia by laparoscopy, with either single or multiple ports or with transcutaneous or intracorporeal sutures. Recent evidence-based literature suggest that these advantages should be carefully weighed. Undoubtedly, it seems clear that this approach may be useful for recurrent hernias in which inguinal canal scarring makes open dissection difficult.

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The incidence of congenital indirect inguinal hernia in full-term neonates is 3.5–5%. The incidence of inguinal hernia in preterm infants, 9–11%, is considerably higher. The incidence approaches 60% as birth weight decreases to 750 g. Inguinal hernia is more common in males than in females. Most series report a male preponderance over females ranging from 5:1 to 10:1. Of all inguinal hernias, 60% occur on the right side, 25–30% on the left, and 10–15% are bilateral.

The anatomy of the inguinal canal varies slightly with age. In adults and children, the internal and external inguinal orifices are widely separated, whereas in young infants they practically overlap. In girls, the anatomy is similar except for the absence of spermatic elements, which are replaced by the round ligament.

A hydrocele of the tunica vaginalis usually presents as a soft, nontender, fluid-filled sac that may transilluminate. Most hydroceles usually involute spontaneously during the first 12 months of life. Those that persist beyond 1 year of age are associated with a patent processus vaginalis and require operative intervention, the same as for an inguinal hernia.

Femoral hernias are rare in children. The diagnosis is based on the observation of a groin swelling located underneath the external inguinal orifice, although this location is easily missed because, unless the bulge is visible upon examination, relatives and doctors will first interpret its appearance as the expression of an inguinal hernia. This explains why 50% of these patients are mistakenly operated upon for inguinal hernia; only when the sac is not found does exploration of the femoral area allow diagnosis and repair. The femoral orifice, located below the inguinal ligament, allows passage of the femoral vein, artery, and nerve from the pelvis to the thigh. The hernial orifice is always medial and the sac is therefore in close contact with the femoral vein.

Umbilical hernia is as a result of failure of closure of the umbilical ring. The hernial sac protrudes through the defect. Most umbilical hernias have a tendency to resolve spontane-

P. Puri, M. E. Höllwarth (eds.), *Pediatric Surgery*, Springer Surgery Atlas Series,
https://doi.org/10.1007/978-3-662-56282-6_19

ously. In view of the favourable natural history of umbilical hernias, surgical indications are limited to those hernias located above the umbilicus, those that persist beyond the age of 4 years, and those occurring in children with connective tissue disorders.

Epigastric hernia (fatty hernia of the linea alba) usually occurs in the midline of the anterior abdominal wall. It is typically a small defect through which preperitoneal fat protrudes and may cause pain.

19.2 Operative Procedures

19.2.1 Inguinal Hernia Repair

Figures 19.1, 19.2, 19.3, 19.4, 19.5, 19.6, 19.7, 19.8, 19.9, and 19.10 illustrate the open and laparoscopic repair of inguinal hernias.

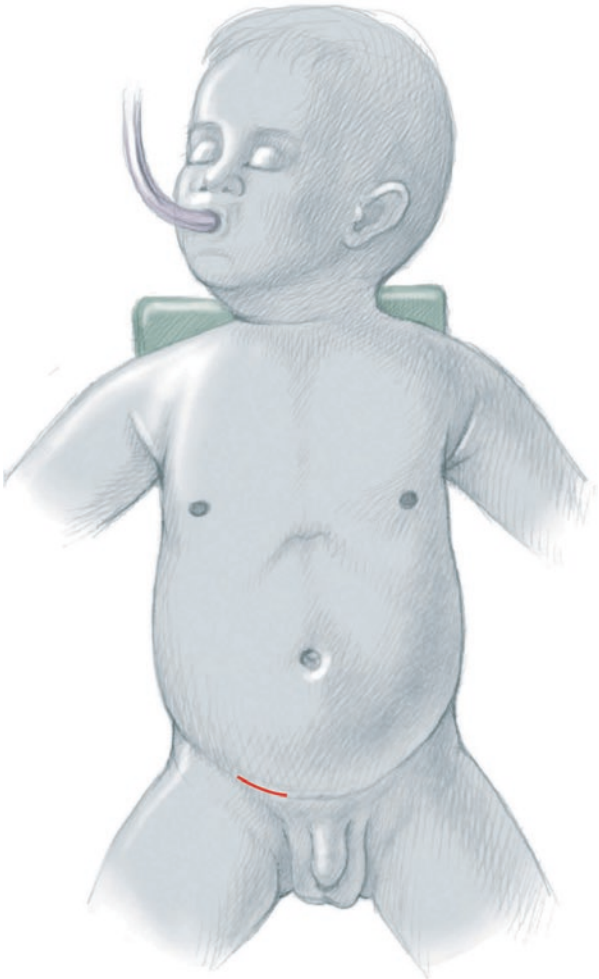


Fig. 19.1 General anaesthesia and spinal anesthesia with or without endotracheal intubation or laryngeal mask is preferred. Premature infants undergoing surgery have an increased risk of life-threatening postoperative apnea. The use of spinal anaesthesia in low-birth-weight infants undergoing inguinal hernia repair is associated with a lower incidence of postoperative apnea. The infant is placed in the supine position on a heating blanket. A 1.5-cm transverse inguinal skin crease incision is placed above and lateral to the pubic tubercle

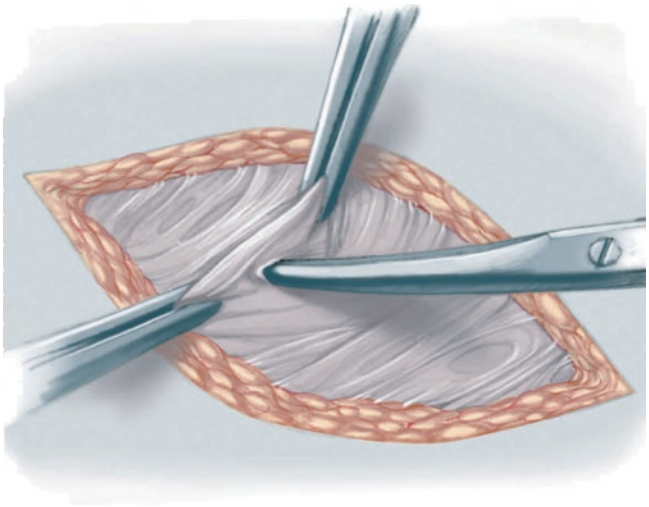


Fig. 19.2 The subcutaneous fat and the fascia of Scarpa (which is surprisingly dense in infants) are opened. Small-toothed Adson forceps are used to grasp them

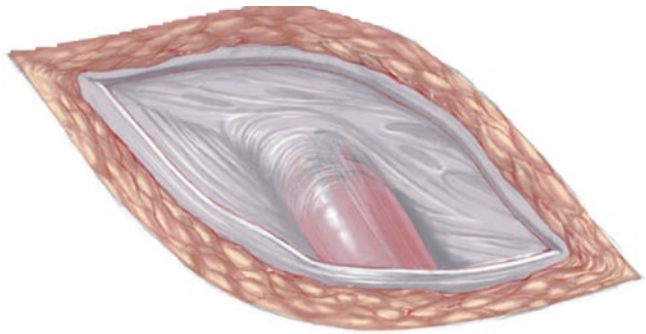


Fig. 19.3 Using blunt scissors or cautery, the external oblique aponeurosis and external ring are exposed. The external inguinal ring is not opened except in older children and adolescents

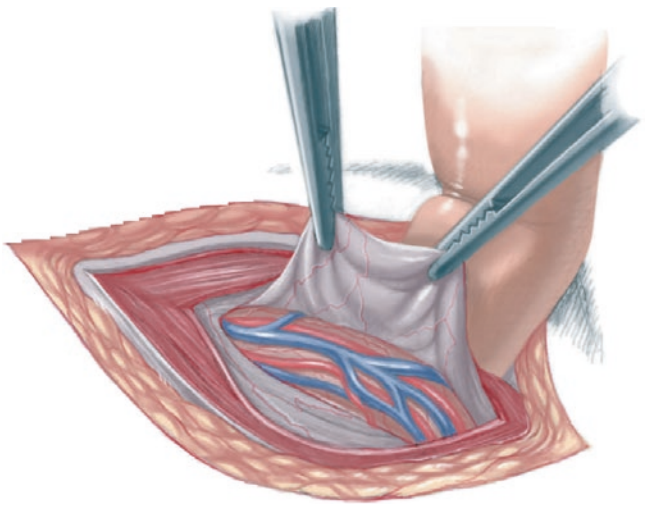


Fig. 19.4 The external spermatic fascia and cremaster are separated along the length of the cord by blunt dissection. The hernial sac is seen and is gently separated from the vas and vessels. A haemostat is placed on the fundus of the sac

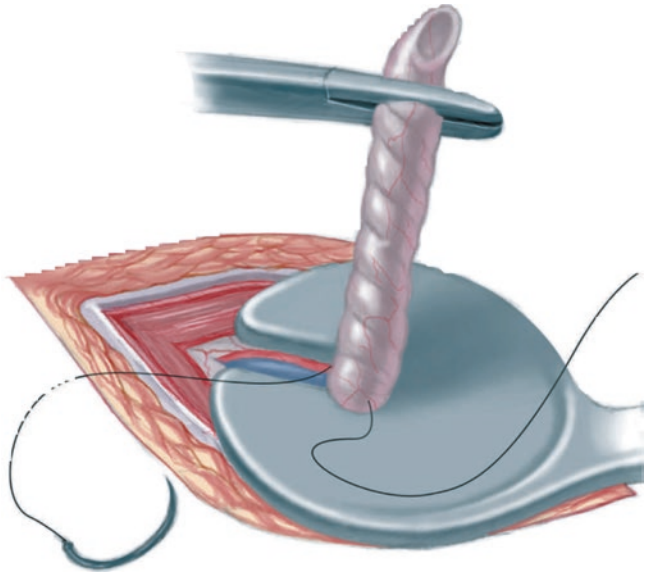


Fig. 19.5 The sac is divided between the clamps and twisted so as to reduce its content into the abdominal cavity. The spoon can be used to keep vas and vessels away from the neck of the sac. The sac is transfixed with a 4/0 stitch at the level of internal ring, which is marked by an extraperitoneal pad of fat. The part of the sac beyond the stitch is usually excised. In the case of hydrocele, the distal part of the sac is widely slit to allow adequate drainage of fluid. In girls, the operation is even more straightforward because there is no risk for the vas or the vessels, and the external orifice can be closed after excising the sac

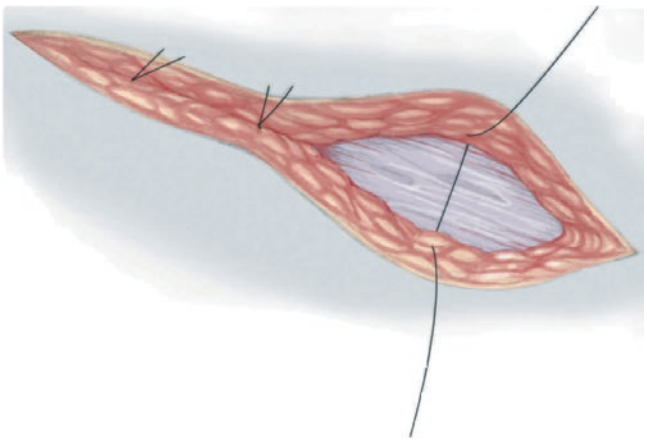


Fig. 19.6 Subcutaneous tissues are approximated using two or three 4/0 absorbable interrupted stitches

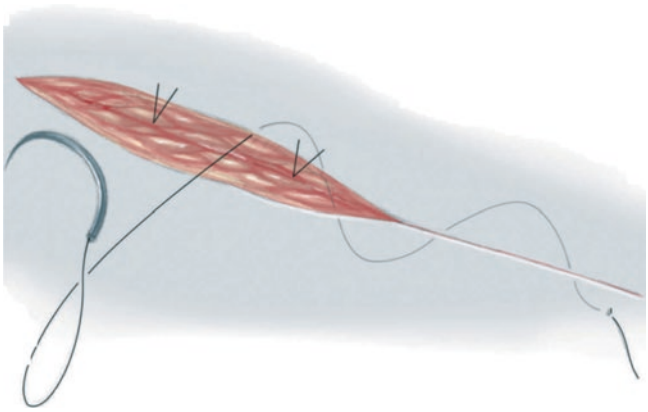


Fig. 19.7 The skin is closed with a 5/0 absorbable continuous subcuticular suture. A small dressing can be applied over the wound if necessary. At the end of the operation, the testis, always tractioned upwards during operative manoeuvres, must be routinely pulled back into the scrotum to avoid iatrogenic ascent

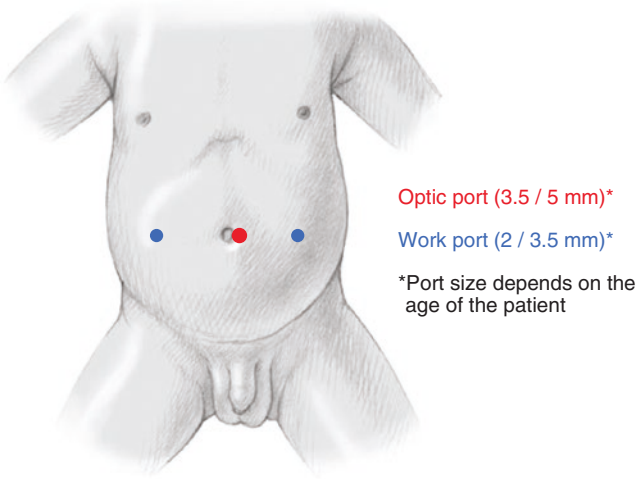
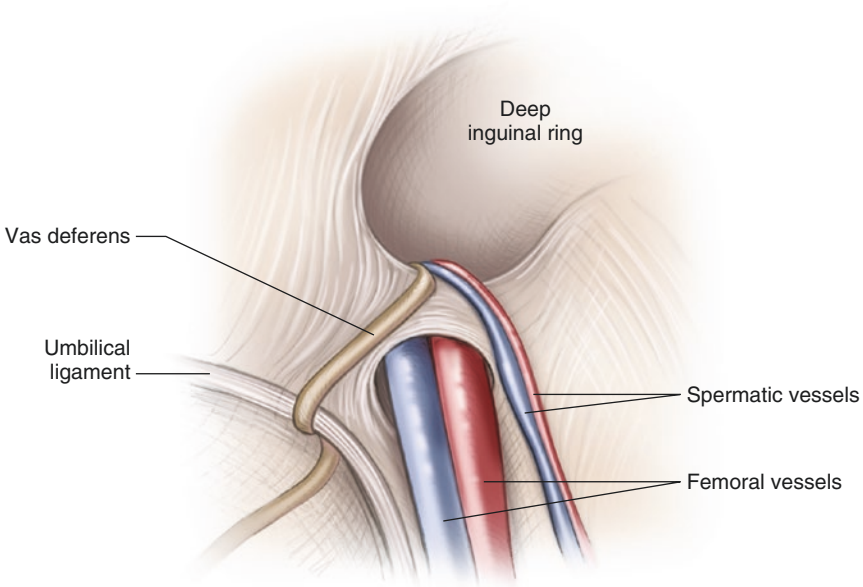


Fig. 19.8 Laparoscopic approach for inguinal hernia. An umbilical access is usually preferred for locating the camera. Two other 2-mm accesses in both flanks are created later

Fig. 19.9 Laparoscopic view of a right inguinal hernia. The deep inguinal ring is clearly open, and both the vas deferens and spermatic vessels run through it. Femoral vessels, clearly visible below the inguinal ring, are always medial to the spermatic vessels



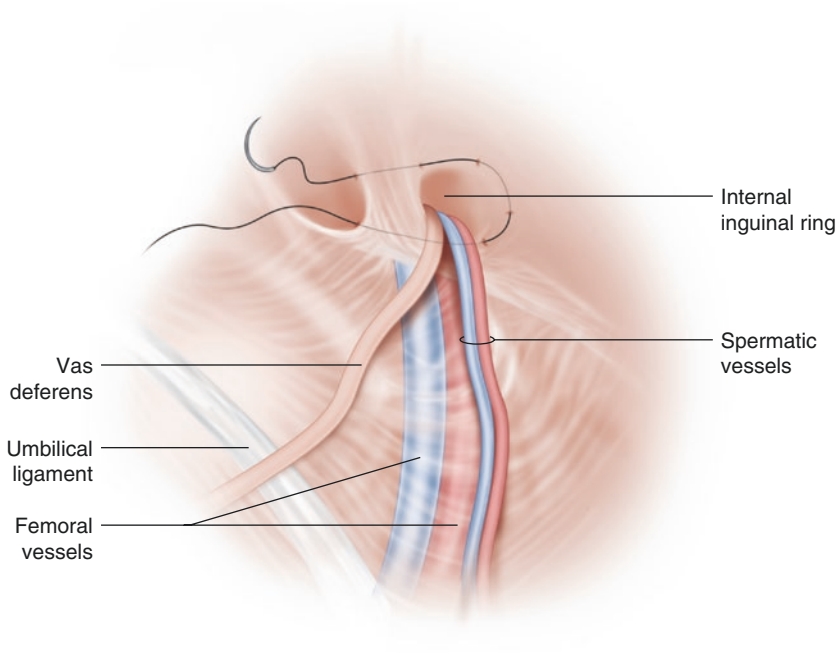


Fig. 19.10 Laparoscopic repair of a right inguinal hernia. Most surgeons who use the laparoscopic technique use an intracorporeal suture to surround the inguinal ring and close it. The suture must be placed carefully to avoid damage to the vas deferens or vascular structures

19.2.2 Femoral Hernia Repair

Figures 19.11, 19.12, and 19.13 show the repair of a femoral hernia.

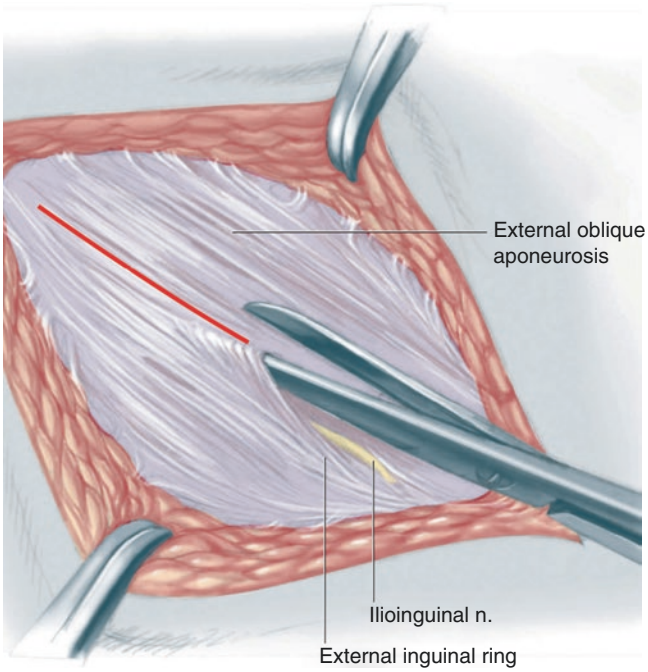


Fig. 19.11 Approach to a femoral hernia. The operative approach for femoral hernia is initially identical to the more commonly used approach for inguinal hernia. An inguinal skin crease incision is made and the subcutaneous layers and Scarpa's fascia are opened in order to expose the external oblique aponeurosis at the level of the external inguinal ring. The aponeurosis is incised longitudinally, taking care to preserve the ilioinguinal nerve. The inguinal canal is opened dorsally, sectioning with cautery the conjoined tendon and the fascia transversalis

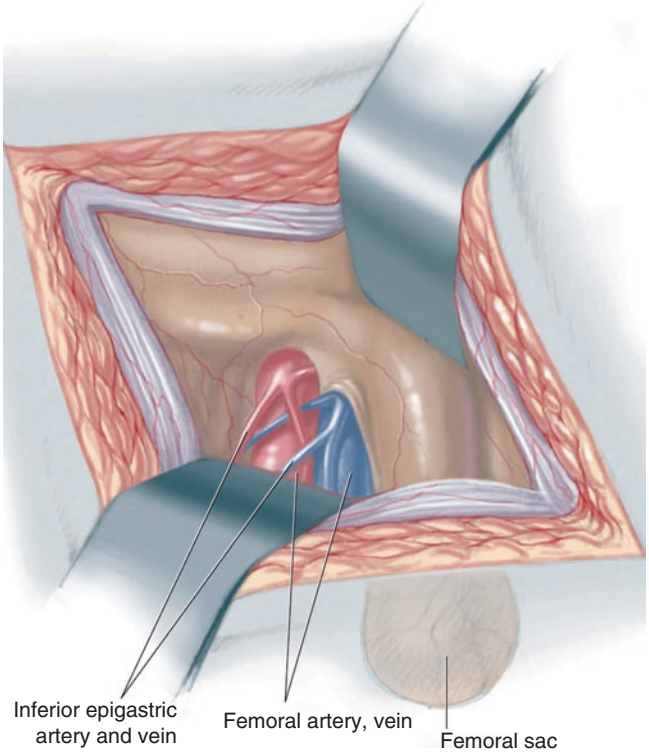


Fig. 19.12 Exposure of a femoral hernia. The spermatic cord is retracted in order to obtain access to the femoral region. The sac is identified and delivered into the wound, avoiding damage to the femoral vein, which is in close contact with the sac laterally. It may be convenient to ligate and divide the inferior epigastric vessels in order to better expose the femoral area from behind

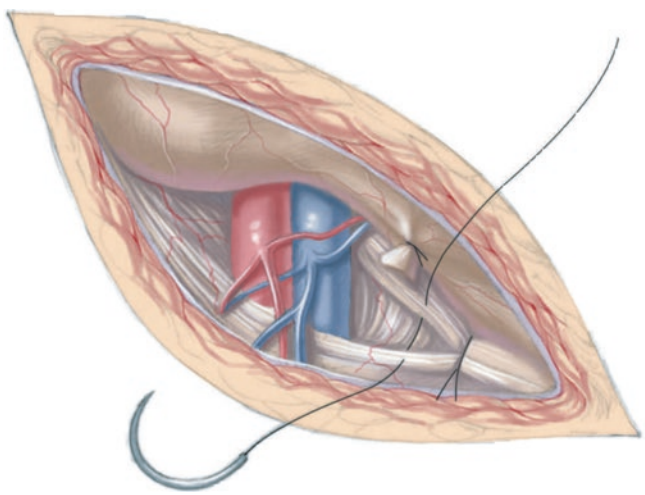


Fig. 19.13 Repair of a femoral hernia. The sac is then opened to ensure that it has no contents; it is subsequently suture-ligated with a fine stitch flush with the peritoneum. The femoral defect is then narrowed by approximating the internal insertions of the Cooper ligament and the inguinal ligament with two or three fine, nonabsorbable stitches, taking care not to compress the femoral vessels. The inguinal canal is reconstructed and the superficial layers and the skin are closed as for an inguinal hernia. Femoral hernia repair can also be accomplished by an infrainguinal approach

19.2.3 Umbilical Hernia Repair

Figures 19.14, 19.15, 19.16, and 19.17 show the repair of an umbilical hernia.

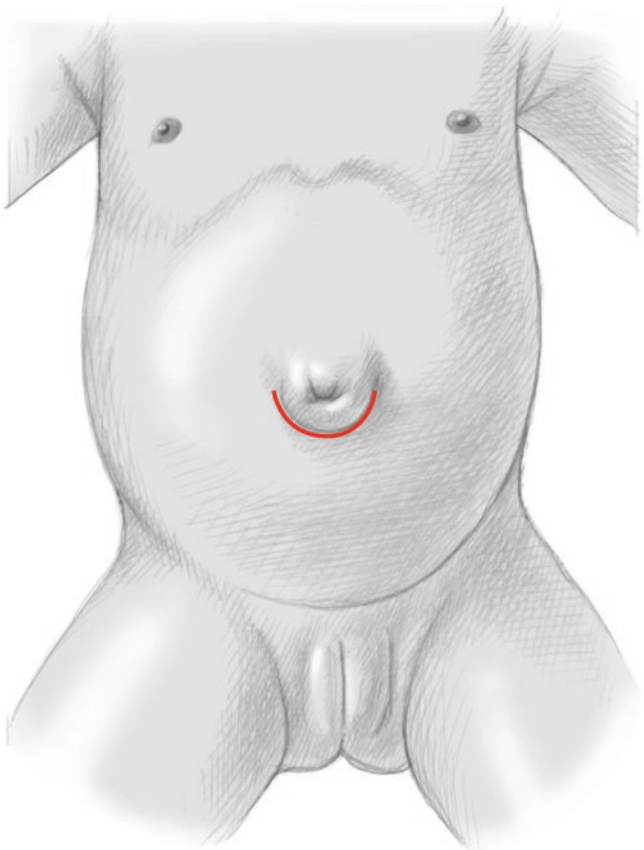


Fig. 19.14 Approach to an umbililical hernia. Umbilical hernia repair is carried out under general anaesthesia. A semicircular incision is made in the skin crease immediately below the umbilicus

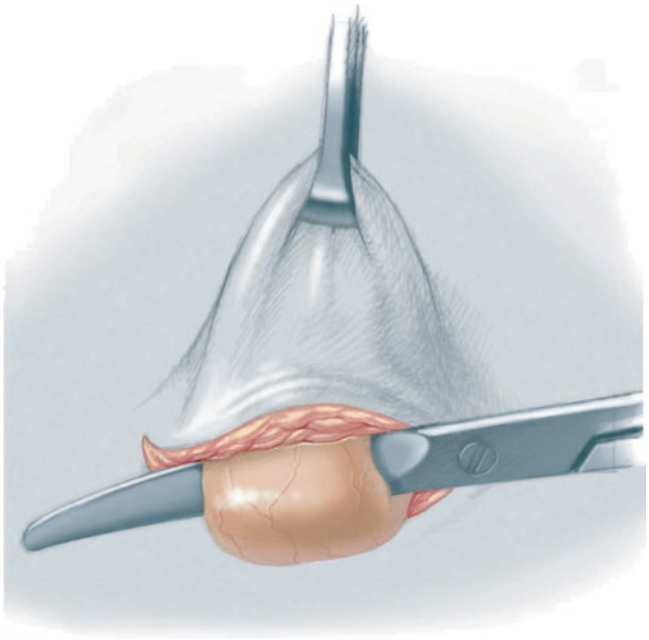


Fig. 19.15 Exposure of an umbilical hernia. The subcutaneous layers are dissected in order to expose the hernial sac. By blunt dissection with a mosquito clamp, a plane is developed on both sides of the sac and the sac is encircled with a haemostat and is divided

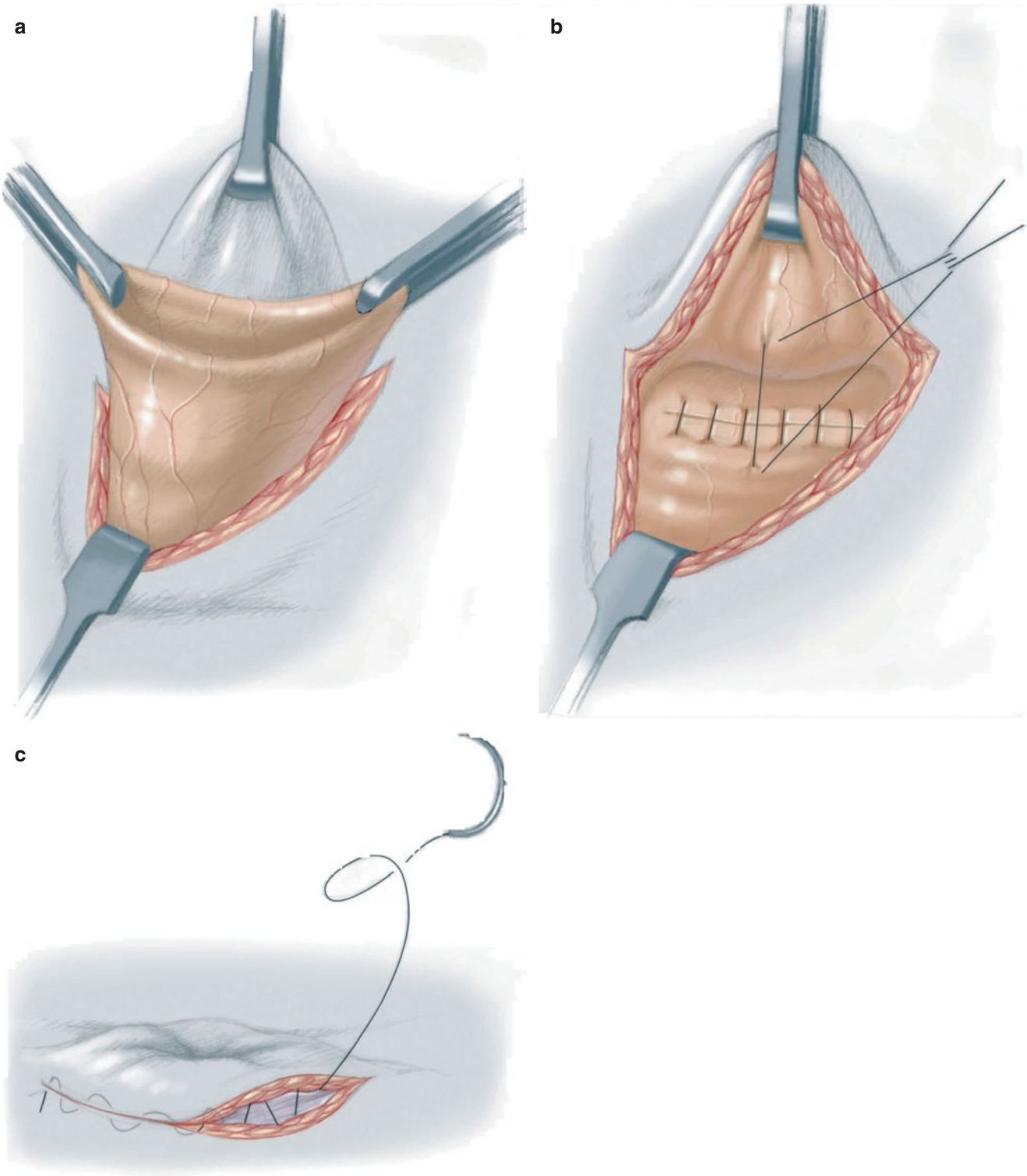


Fig. 19.16 (**a**, **b**) Repair of an umbilical hernia. A clamp is placed on either side of the umbilical defect for traction. The defect is closed by interrupted 2/0 absorbable sutures. A stitch is used to invaginate the umbilical scar, tractioning it downwards and fixing it to the subcutaneous layer in the midline. (**c**) Closure of umbilical hernia repair. The wound is closed with several interrupted sutures placed in the subcuticular plane. A slightly compressive dressing is maintained for 24 h

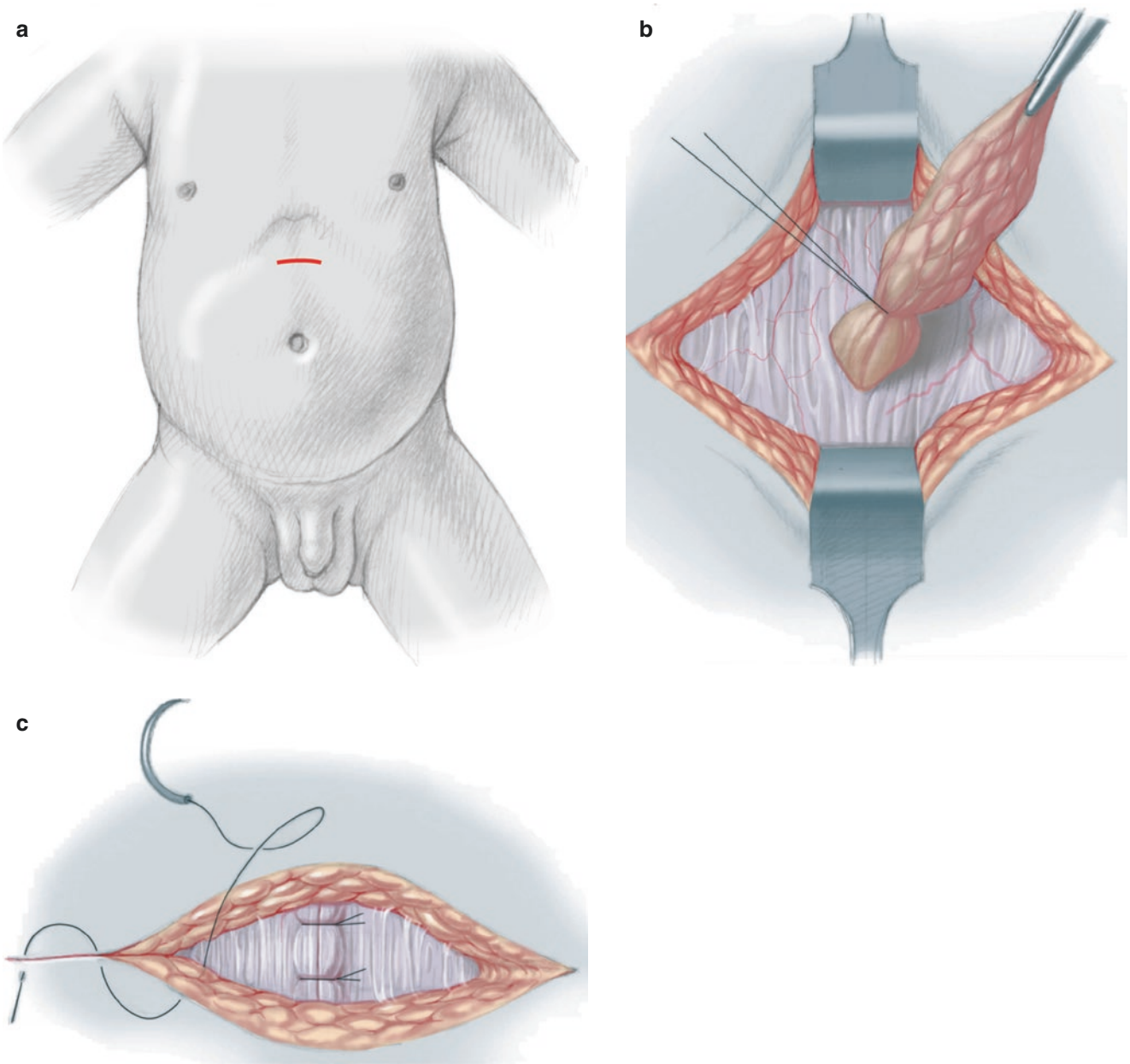


Fig. 19.17 (**a**–**c**) Approach to an epigastric hernia. It is important to mark the location of the defect before anaesthesia, because in the recumbent position they are often impossible to palpate along the widened linea alba. A transverse incision is made directly over the previously marked location of the hernia. Repair of an epigastric hernia. The fatty mass protruding through the linea alba defect is excised after a transfixation stitch. Closure of epigastric hernia repair. The defect in the linea alba is closed with interrupted 3/0 absorbable sutures. The skin is approximated using subcuticular sutures

19.2.4 Epigastric Hernia Repair

Epigastric hernias are repaired when they are prominent or when they are symptomatic.

19.3 Complications After Hernia Repair

The overall complication rates after elective hernia repair are low (about 2%) but they rise to 8–33% for incarcerated hernias requiring emergency operations. A number of complications are possible after inguinal hernia repair:

- Haematoma. This can be avoided with meticulous attention to haemostasis. It is rarely necessary to evacuate a wound, cord, or scrotal hematoma.
- Wound infection. The risk is low and should not exceed 1%.
- Gonadal complications. These are due to compression of the vessels by incarcerated viscera. Though large numbers of testes look nonviable in patients with incarcerated hernia, the actual incidence of testicular atrophy is low; the testis should not be removed unless it is frankly necrotic.
- Intestinal resection. This is necessary in about 3–7% of patients in whom the hernia is not reduced. It may cause some additional morbidity corresponding to the resection itself and contamination of the field.
- Iatrogenic ascent of the testes. This event is relatively rare; slightly more than 1% of patients operated upon for inguinal hernia during infancy required subsequently orchidopexy. This complication is probably due to entrapment of the testis in the scar tissue or failure to pull it down into the scrotum at the end of the operation and to maintain it there.
- Contralateral exploration: Only 5.2% of patients older than 1 year with an initial unilateral hernia will develop a metachronous contralateral hernia, so contralateral groin exploration can be avoided. Up to 14 contralateral explorations should be needed to prevent one metachronous hernia.
- Recurrence. The acceptable recurrence rate for inguinal hernia repair is less than 1%, but when operation is performed in the neonatal period, this complication can occur in up to 8% of patients. The factors that predispose to recurrence are ventriculoperitoneal shunts, sliding hernia, incarceration, and connective tissue disorders. Recurrence may be indirect or direct. Indirect recurrence is due to either failure to ligate the sac at a high level, tearing of a friable sac, a slipped ligature at the neck of the sac, missed sac, or wound infection. Direct hernia may be due to inherent muscle weakness or injury to the posterior wall of the inguinal canal.
- Mortality. The mortality rate of inguinal hernia operations at this time should be zero.

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Omphalocele

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Stig Sømme and Jacob C. Langer

Exomphalos (also known as omphalocele) is a condition seen in newborn infants. It is thought to result from failure of the intestines to return to the abdomen after the migration into the umbilical cord that occurs between the sixth and tenth week after conception. The incidence of exomphalos has not changed over the past several decades. There is no known environmental, racial, or geographic predilection, although in rare cases there may be a familial predisposition. Exomphalos is also associated with a lower than normal birth weight and gestational age.

Exomphalos is characterized by a central defect at the umbilical ring; a membrane composed of visceral peritoneum, Wharton's jelly, and amnion covers the eviscerated abdominal contents. The umbilical cord inserts onto the exomphalos sac. The sac usually contains loops of small and large intestine and stomach; about 50% contain liver. Cases in which the liver is out and the size of the defect is very large are often referred to as "giant omphalocele", although there is no consensus on a precise definition of this term. The abdominal muscles are normally developed. Rupture of the sac is reported in 10–18% of cases. This rupture can happen in utero, at time of delivery, or after delivery.

Exomphalos is frequently associated with other anomalies, the most common of which are cardiac and gastrointestinal tract abnormalities. Chromosomal abnormalities are often seen, particularly in children with small defects that do not contain liver. Exomphalos is also associated with Beckwith-Wiedemann syndrome, cloacal exstrophy, and pentalogy of Cantrell. Many children with large omphaloceles also suffer from pulmonary hypoplasia, although this may be difficult to diagnose prenatally.

20.1 Prenatal Diagnosis and Management

Exomphalos is often suspected because of an elevated level of maternal serum α-fetoprotein. Prenatal diagnosis can be accurately made using prenatal sonography. Exomphalos can be differentiated from gastroschisis because of the location of the defect and the presence of a sac, although this differentiation may be more difficult if the sac has ruptured. If exomphalos is detected or suspected, it is important to search for other abnormalities. In addition to a thorough ultrasound examination, amniocentesis for karyotype analysis should be recommended, and fetal echocardiography should be done to look for major cardiac abnormalities. The mother should be transferred to a perinatal centre with experienced neonatal and surgical support. Although there is no clear evidence to support routine caesarean section, most practitioners will recommend caesarean delivery for fetuses with a large exomphalos, to avoid liver injury and rupture of the sac.

20.2 Postnatal Management

A number of measures should be taken immediately after birth:

- Nasogastric tube placement to decompress the stomach
- Intubation, if the child is in respiratory distress
- Coverage of the sac with moist gauze and plastic foil
- Intravenous fluids
- Routine neonatal bloodwork
- Temperature control with a heating lamp
- Vitamin K administration
- Antibiotics, if the sac is ruptured

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In addition, a thorough assessment for other abnormalities must be performed, which will directly affect decisions related to the care of the child. A detailed physical examination, radiologic studies, echocardiography, and abdominal ultrasound are important to identify any associated anomalies. Because some large defects are associated with pulmonary hypoplasia, oxygenation and ventilation should be carefully assessed and respiratory support using intubation and mechanical ventilation should be instituted if necessary. The exomphalos itself should be evaluated to determine its size, contents, and integrity.

Newborns with abdominal wall defects require more intravenous fluids in the first few days of life than a normal infant, owing to evaporative loss and third spacing. The daily intravenous fluid requirement must be adjusted based on the hourly urine output and other parameters for end-organ perfusion. Infants treated with the use of a silo are at a particularly high risk of fluid, protein, and temperature loss.

20.3 Surgical Management

Based on the clinical status of the patient and the characteristics of the exomphalos, there are three broad categories of options for the surgical management of this condition: primary closure, various types of staged closure, and nonoperative management with late closure.

20.3.1 Primary Closure

Small to moderate defects, particularly when the liver is not in the sac, may be closed primarily (Figs. 20.1, 20.2, and 20.3). For very small defects, the umbilical cord can be left behind to give better cosmetic results.

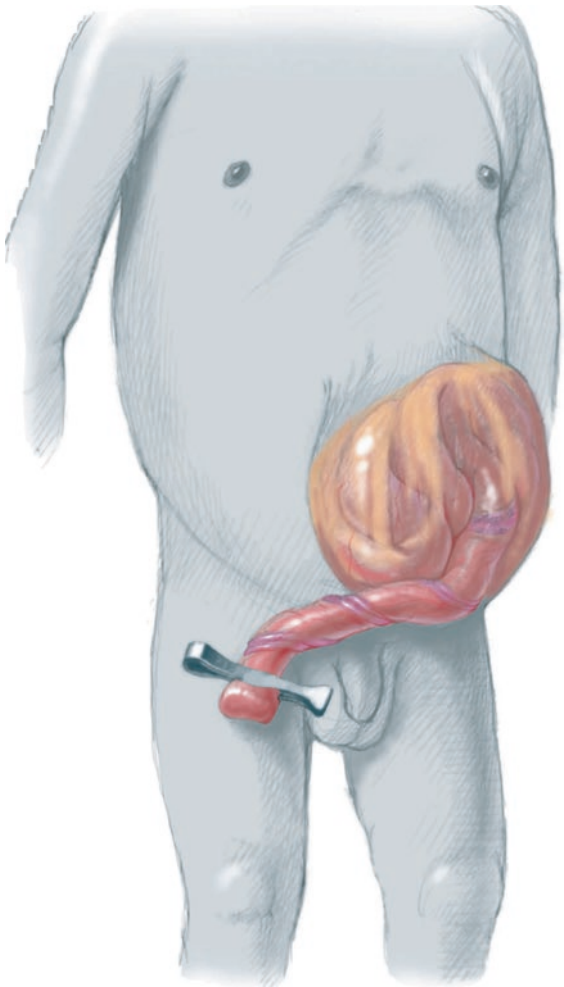

Fig. 20.1 The umbilical vessels are ligated and divided

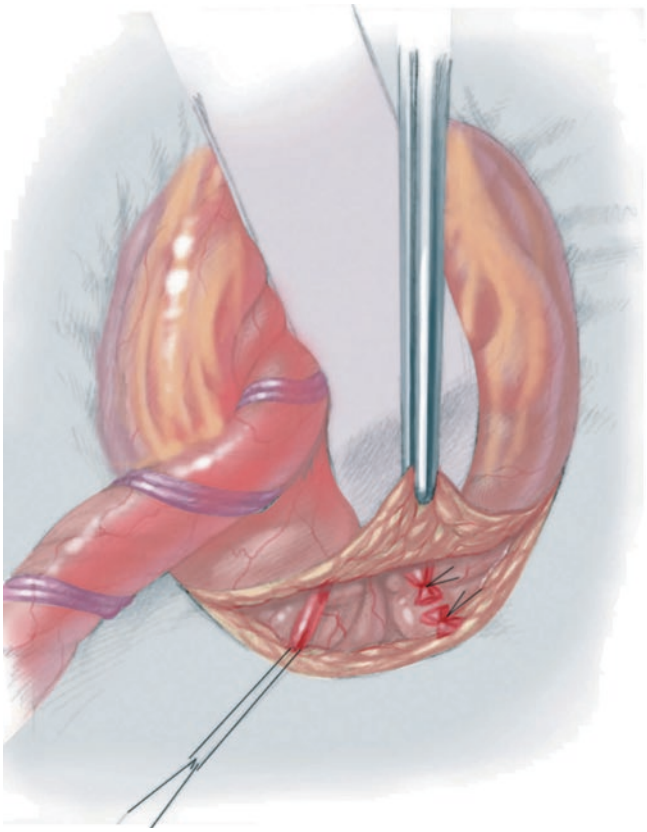

Fig. 20.2 The sac is removed

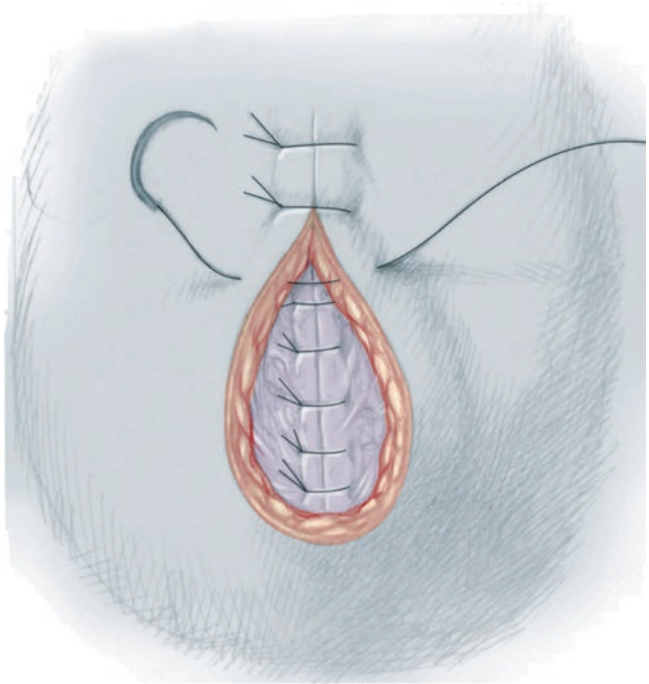

Fig. 20.3 The skin is undermined enough that a secure fascial closure can be accomplished. Absorbable or nonabsorbable sutures may be used. The skin is then closed

20.3.2 Staged Closure

For larger defects, it may not be possible to close the fascia, but there may be enough skin to achieve skin closure over the viscera. In most cases of skin closure alone, the patient is left with a ventral hernia that must be closed at a later time. This technique (Figs. 20.4 and 20.5) was originally described by Gross in 1948. Some surgeons opt to insert a nonabsorbable or bioabsorbable patch into the fascia and then close the skin over the patch (Fig. 20.6). The advantage of a nonabsorbable patch is a lower incidence of subsequent hernia formation, but there is a risk of infection or erosion of the patch into underlying bowel. Bioabsorbable patches have a much lower risk of these complications, but they may be associated with a higher incidence of ventral hernia.

In 1967, Schuster was the first to describe a staged closure using a "silo" made from a sheet of Silastic reinforced with Dacron to gradually reduce the viscera over several days to a week, after which the fascia and skin are definitively closed (Figs. 20.7, 20.8, and 20.9). This technique is useful for children with a large or ruptured exomphalos.

A similar technique, sequential sac ligation, uses the exomphalos sac as a silo (Fig. 20.10). It requires a sac which is relatively strong, and it is more difficult if the liver is adherent to a large part of the sac, but it can be performed at the bedside in the nursery, with only minimal sedation. The technique involves gently kneading the sac to release minor adhesions between the sac and the intestine or liver. Traction is applied to the sac to slowly reduce the contents, and the sac is then twisted and ligated. Once the viscera are reduced as much as possible, the child is taken to the operating room for definitive closure.

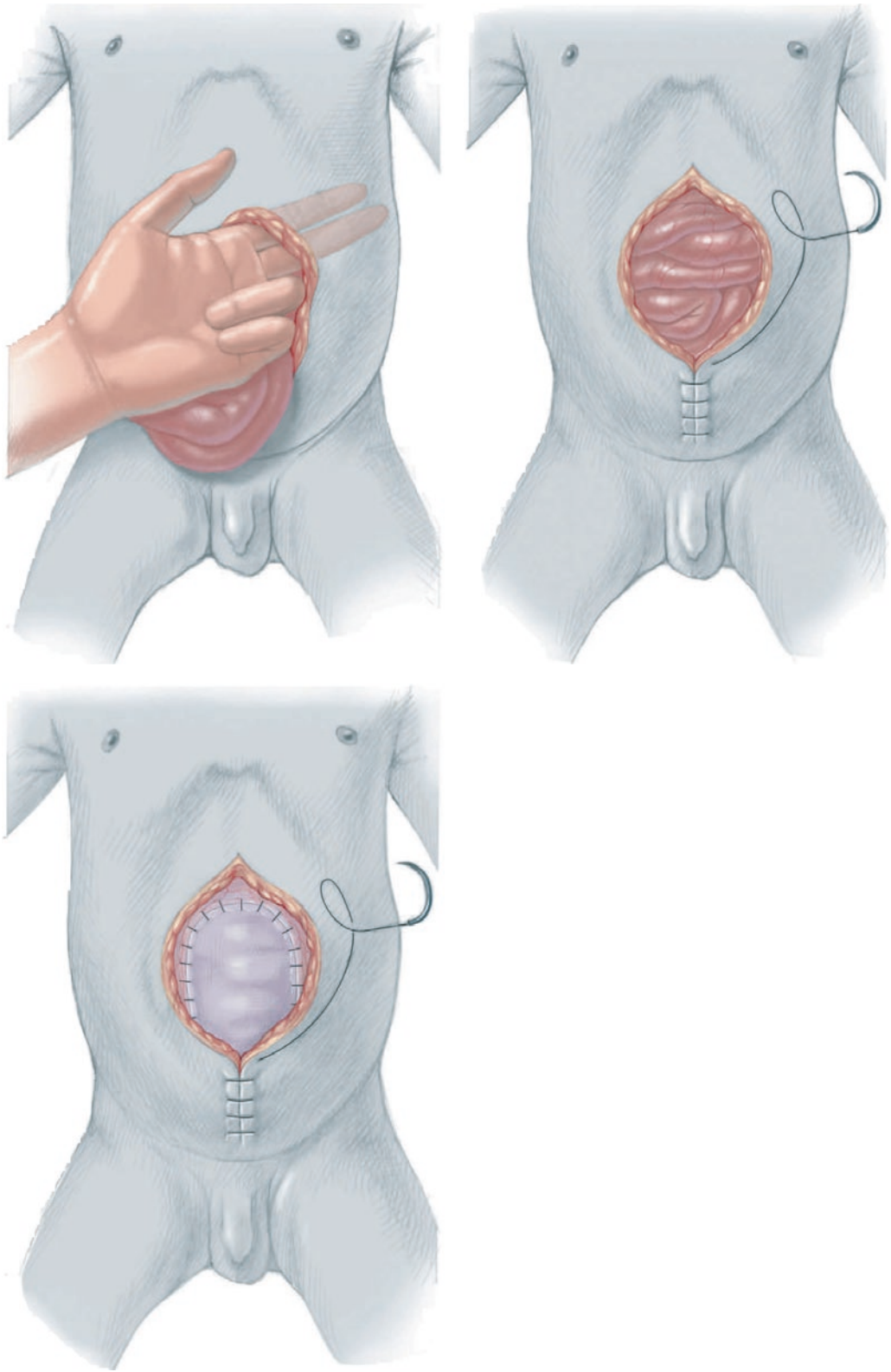

Fig. 20.4–20.6 For larger defects, it may not be possible to close the fascia, but there may be enough skin to achieve skin closure over the viscera. This technique was originally described by Gross in 1948. The sac is usually removed, although some surgeons prefer to leave the sac intact and dissect between the edge of the sac and the skin to the level of the abdominal wall muscle. The skin is undermined as far out as possible, to permit skin closure with minimal tension. At this point, some surgeons opt to insert a patch into the fascia. The skin is then closed over the patch. In most cases of skin closure, the patient is left with a ventral hernia, which must be closed at a later time

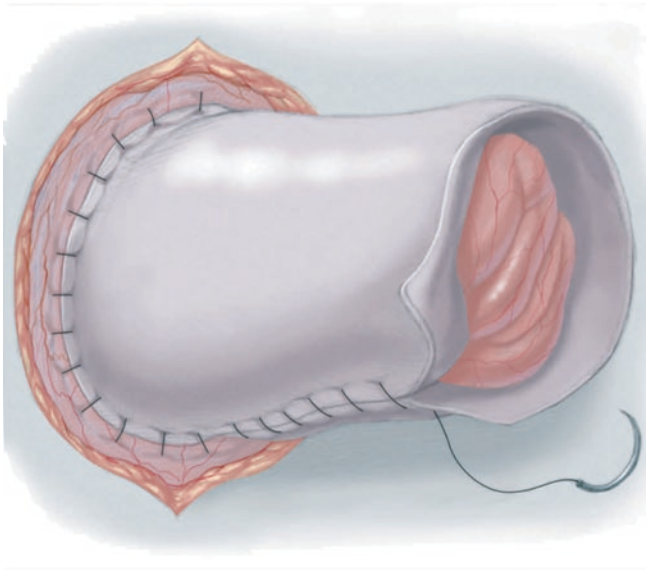

Fig. 20.7 The use of a silo was first described by Schuster in 1967. The concept of a silo is to use a sheet of silastic reinforced with Dacron to gradually reduce the viscera over several days to a week, and then to definitively close the fascia and skin. This technique is useful for children with a large or ruptured exomphalus. The silastic sheeting is sutured to the edges of the musculae fascial layer, after as much of the intestine and liver as possible have been returned to the abdomen. Some surgeons also include the skin in these sutures. Although most surgeons remove the sac, some prefer to leave it intact and dissect between the edge of the sac and the skin to the level of the abdominal wall muscle. In some infants, the neck of the sac at the abdominal wall is relatively small, and the fascial opening must be enlarged to allow gradual reduction of the viscera. Monofilament nonabsorbable sutures are then placed around the edges to avoid any gaps through which intestine can herniate. The silo is then closed over the top, by suturing it to itself. It should be perpendicular and suspended or supported to avoid any kinks in the intestine or the hepatic vessels. Care must also be taken not to injure the hepatic veins, which may course very close to the superior fascial edge

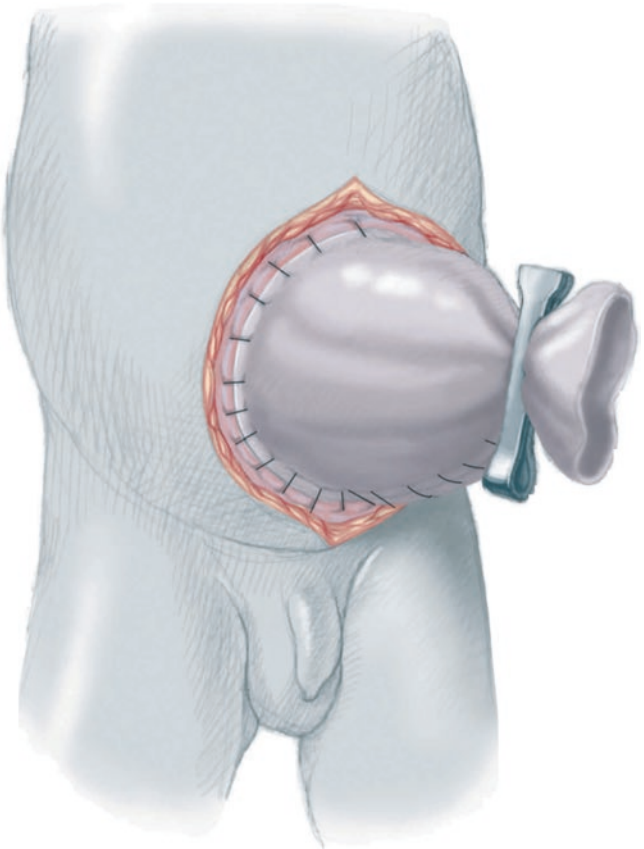

Fig. 20.8 The sac is then gradually reduced at least once daily until all of the viscera have returned to the abdominal cavity. Various techniques have been used to close the top of the silo, including sutures, umbilical cord clamps, umbilical tapes, and roller devices. Once the viscera are completely reduced, the child is brought back to the operating room and the fascia and skin are closed

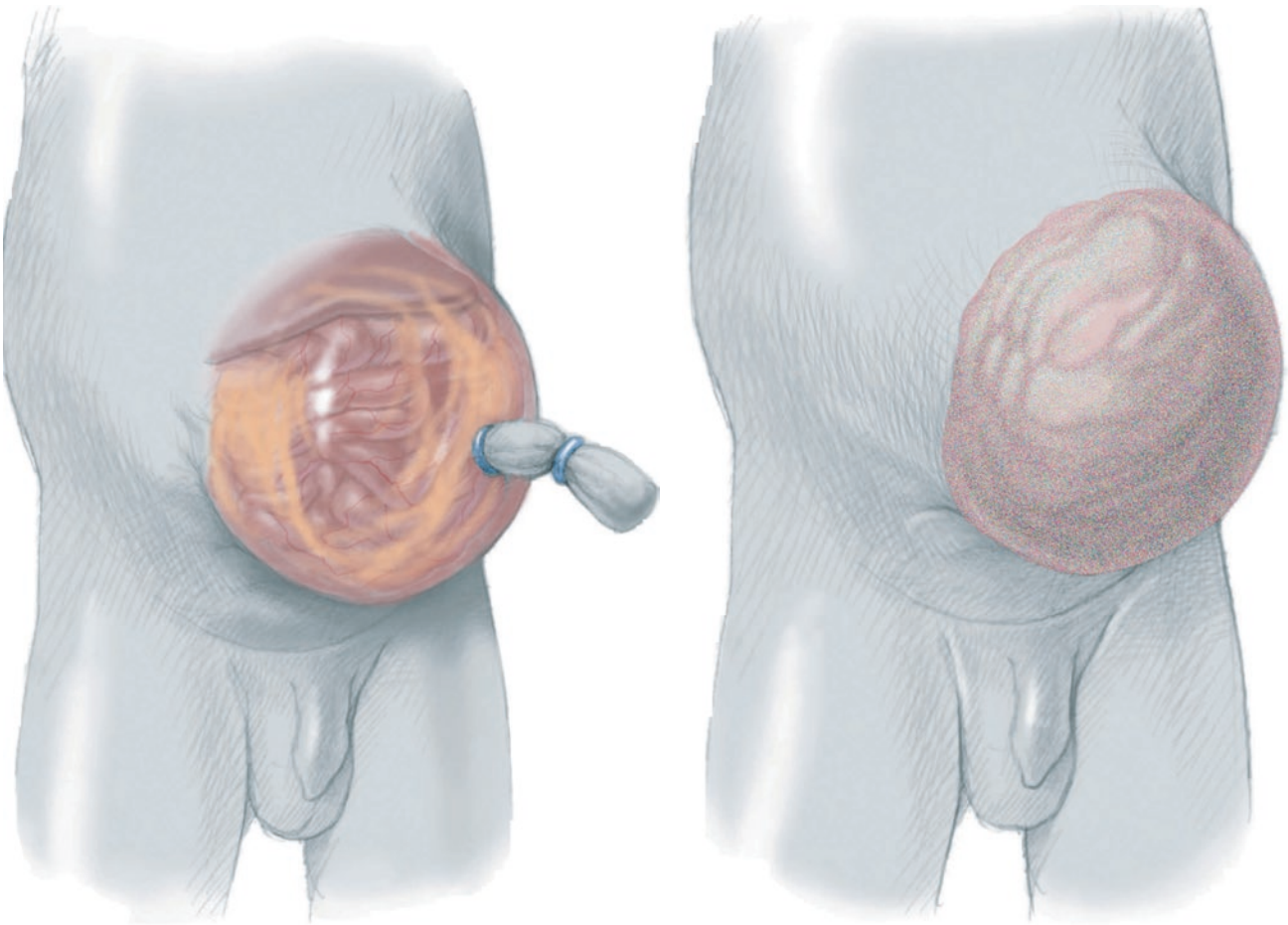

Fig. 20.9, 20.10 This recently described technique uses the exomphalus sac as a silo. It requires a sac which is relatively strong, and it is relatively difficult if the liver is adherent to a large part of the sac. However, it can be performed at the bedside in the nursery, with only minimal sedation. The technique involves gently kneading the sac to release minor adhesions between the sac and the intestine or liver. Traction is applied to the sac to slowly reduce the contents, and the sac is then twisted and ligated with umbilical ties. Once the viscera are reduced as much as possible, the child is taken to the operating room for definitive closure. Some infants with exomphalus are very poor candidates for any kind of surgical intervention. This includes premature infants, those with chromosomal abnormalities, and those with significant congenital heart disease or pulmonary hypophasia. For these children it is best to cover the sac with a material which allows it to form granulation tissue and eventually epithelialize. Early on mercurochrome or iodine solution were used for this purpose, but there were problems with toxicity; this resulted in the abandonment of this practice. The use of plastic sheeting ("Op-site") has been described. We currently recommend silver sulfadiazine, which prevents infection and results in a good bed of granulation tissue. It takes several months for this to occur, and another several months for the granulation tissue to epithelialize. The resulting huge ventral hernia can be repaired electively whenever the child's underlying cardiac, pulmonary, or other conditions have improved. This may take several years in some cases

20.3.3 Nonoperative Management with Late Closure

For infants with large defects that cannot be closed primarily, and infants who are very poor candidates for any kind of surgical intervention, it is best not to perform an operation in the neonatal period. This group includes premature infants, those with chromosomal abnormalities, and those with significant congenital heart disease or pulmonary hypoplasia, in addition to the healthy newborn with "giant omphalocele". In these children, it is best to cover the sac with a material that promotes formation of granulation tissue and eventual epithelialization (escharotic therapy). Early on, mercurochrome or iodine solution were used for this purpose, but problems with toxicity led to abandonment of this practice. The use of plastic sheeting ("Op-site") has been described. We currently use silver sulfadiazine or antibiotic ointment, which prevents infection and results in a good bed of granulation tissue. It takes several months for this to occur, and another several months for the granulation tissue to epithelialize. The resulting large ventral hernia can be repaired electively when the child is healthy and the abdominal wall can be closed with minimal tension, typically any time after 8–12 months of life. In some cases, however, it may take several years and may require several stages and/or the use of a patch, particularly for the superior segment of the defect. Some authors have advocated the use of a tissue expander to increase the intraabdominal domain and accelerate the process. Others have reported success using a component separation technique, either in the neonatal period or after epithelialization of the sac using escharotic therapy. In a healthy infant, "giant omphalocele" can often be closed without much tension and without use of a patch, if the defect has been gently compressed with a self-adherent elastic wrap (e.g., Coban™) or similar external compression for a few months leading up to surgery.

20.4 Postoperative Management

Care of the infant following definitive closure requires a neonatal or pediatric intensive care unit (ICU) for all but the smallest defects. Infants that have undergone repair of larger defects usually require postoperative mechanical ventilation for days to weeks, depending on their pulmonary status. It is important to carefully observe the child for signs of abdominal compartment syndrome, such as oliguria, acidosis, intestinal ischemia, and liver dysfunction. Infants with other congenital malformations require continued investigation and management as needed.

Intra-abdominal pressure monitoring using intragastric or intravesical catheters during abdominal wall closure (either primary or secondary) is an important adjunct to prevent abdominal compartment syndrome, which may result in high airway pressures, oliguria, and intestinal ischemia due to decreased organ perfusion. Monitoring can also be continued in the ICU during the postoperative period. Intra-abdominal pressures above 15–20 mmHg or an increase in central venous pressure of more than 4 mmHg are associated with visceral ischaemia in both animal and human studies.

After closure of the abdomen, infants with exomphalos often develop an ileus, although intestinal function usually returns more quickly than in infants with gastroschisis. A nasogastric tube is therefore necessary. Total parenteral nutrition should be initiated early. Many surgeons place a central venous catheter at the time of the operation for this purpose.

20.5 Outcomes

The outcome for infants with exomphalos is dependent on gestational age, the presence of associated chromosomal and structural anomalies, the presence or absence of pulmonary hypoplasia, and the size of the defect. Long-term problems that are commonly seen in these infants include gastro-oesophageal reflux, feeding disorders, and adhesive bowel obstruction, which may require additional surgical procedures such as fundoplication, ventral hernia repair, feeding tube insertion, and lysis of adhesions. Most of these issues can be corrected, however, or will improve on their own with time. Most infants with exomphalos who do not have severe additional anomalies or pulmonary hypoplasia do very well on long-term followup.

Suggested Reading

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Gastroschisis

21

Marshall Z. Schwartz

Gastroschisis (Fig. 21.1) is one of several congenital abdominal wall defects that evolves in the first four weeks after conception. It is generally accepted that this congenital abdominal wall defect is embryologically different from omphalocele. The anomaly is thought to be the result of a defect at the site where the second umbilical vein involutes. Nonrotation of the bowel always accompanies this anomaly, and there is an increase in intestinal abnormalities, including atresia (mostly involving the small intestine), perforation, and infarction resulting from in utero midgut volvulus or vascular thrombosis. Unlike omphalocele, however, gastroschisis is not accompanied by any increase in anomalies of other organs. The incidence of gastroschisis is approximately 1 in 4000–6000 live births. Infants with gastroschisis typically are slightly premature (35–37 weeks of gestation) and frequently have growth retardation, with birth weights from approximately 2000–2500 g.

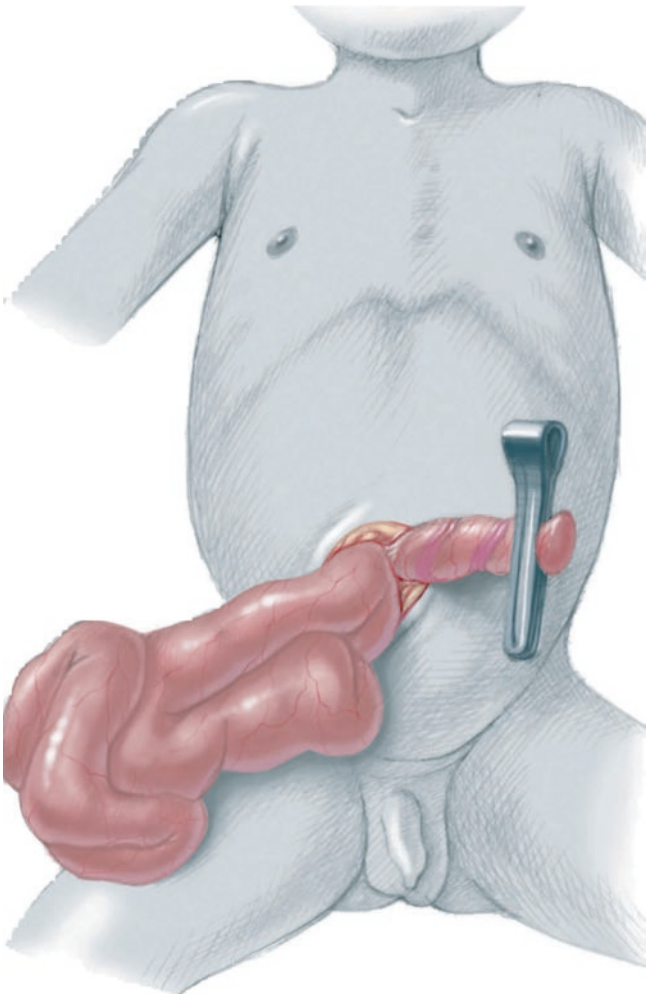

Fig. 21.1 The specific features of gastroschisis include an abdominal wall defect measuring 2–3 cm in diameter, which is almost always to the right of a normal umbilical cord. There is no sac covering the herniated contents, which typically include the entire intestinal midgut. There is shortening of the mesentery and thickening of the bowel wall. The bowel surface may be covered with a fibrin "peel". Depending on the size of the abdominal wall defect, it is possible that the stomach, the urinary bladder, and/or the fallopian tubes and ovaries in a female may be herniated through the abdominal wall defect

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21.1 Prenatal Diagnosis and Management

Most abdominal wall defects can be diagnosed in utero after 14 weeks gestation, when the fetal midgut has returned to the peritoneal cavity. If gastroschisis is noted on fetal ultrasonography, it is strongly recommended that serial examinations be performed to look for changes in the size and thickness of the bowel and the diameter of the abdominal wall defect. Significant bowel wall thickening and bowel dilatation, especially associated with a decrease in the diameter of the abdominal wall defect, may be indications for earlier delivery to avoid bowel infarction. It is important to provide an opportunity for the family to meet with a fetal management team including perinatology, paediatric surgery, and neonatology to review the abdominal wall abnormalities and the likely course following delivery. The recommended mode of delivery has been a subject of controversy over the past several decades. It is generally believed that caesarean section is not necessary unless for obstetric reasons. Elective premature delivery is also unnecessary unless there is evidence of intestinal vascular compromise.

21.2 Postnatal Management

Management immediately following delivery and prior to surgical correction requires prompt attention and is critical to the outcome. Two goals are most important: (1) providing a mechanism to maintain normal thermogenesis, and (2) establishing intravenous access to provide appropriate fluid resuscitation. Infants with gastroschisis are usually hypovolemic and require at least 125–150% of maintenance intravenous fluid to establish and maintain adequate hydration. Establishing intravenous access initially can be done through a peripheral intravenous site, but infants with gastroschisis require central venous access. Once intravenous access is established, it is optimal to institute broad spectrum antibiotic coverage and appropriate intravenous nutrition. To avoid having the bowel get more distended, a nasogastric tube should be inserted and placed on suction.

21.3 Surgical Management for Primary Abdominal Wall Closure

A significant percentage of infants with gastroschsis (reported to be from 60% to nearly 100%) can undergo reduction of the herniated intestinal contents and primary abdomial wall closure. General anaesthesia including muscle relaxation is required for the appropriate intra-operative management of gastroschisis. The bowel and anterior abdominal wall should be prepped. It is my preference to use a warm, dilute 50/50 mixture of povidone iodine and saline. As shown in Fig. 21.2, the umbilical cord should be clamped and tied 2–3 cm above the abdominal wall; the excess umbilical cord is then removed. At this point, appropriate draping is indicated. Because the abdominal wall defect in gastroschisis is relatively small (2–3 cm), it may be difficult to reduce the herniated midgut through this opening. Thus, after prepping and draping the infant's abdomen and bowel and having the surgeons gowned and gloved (NOTE: drapes are not shown in the figures to simplify the drawings), it may be necessary to enlarge the abdominal wall opening, as in Fig. 21.2.

After enlarging the abdominal wall defect (Fig. 21.3), the bowel can be reduced into the peritoneal cavity. The degree of thickening and fibrin peel determines how malleable the herniated bowel is and how easily it is to place it within the abdominal cavity. If the initial assessment suggests that primary closure may not be obtainable, two techniques have been described to facilitate a primary abdominal wall closure. The first approach is to attempt to empty the intestinal contents either retrograde into the stomach (which then can be aspirated through the nasogastric tube) or antegrade into the colon and out the rectum. However, if the bowel has an extensive peel and and is clumped as suggesterd in Fig. 21.1, these manipulations may result in further damage to the bowel or injuy to the blood supply. A second technique is manual stretching of the anterior abdominal wall to increase the size of the peritoneal cavity. Although gentle stretching is potentially advantageous, vigorous stretching can result in haemorrhage and swelling of the rectus muscles in the rectus sheath, resulting in a rigid and painful abdominal wall. My personal preference is to not perform either of the described maneuvers and thus, avoid creating a worse situation.

If it is possible to reduce all of the herniated intestinal contents into the peritoneal cavity, primary closure should be undertaken (Fig. 21.4). It is important to identify good fascial edges for the closure. The placement of sutures at the level of the umbilicus is especially important, to avoid a later umbilical defect.

Before closing the skin, any compromised or ischaemic skin should be trimmed. Steps to relieve tension at the skin edges are important (Fig. 21.5).

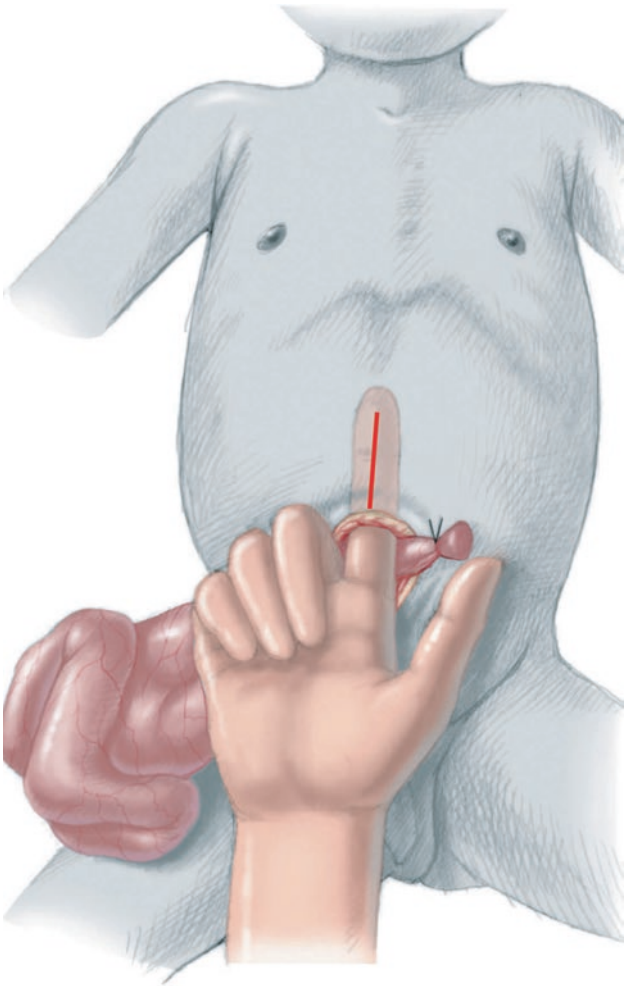

Fig. 21.2 The optimum way to extend the opening is by extending the gastroschisis defect superiorly by incising the fascia along the midline, with a finger placed below the fascia to avoid an injury to the bowel. Extending the defect superiorly is safer than a caudal incision because the bladder is very close to the inferior aspect of the abdominal wall defect, limiting the ability to extend the opening inferiorly

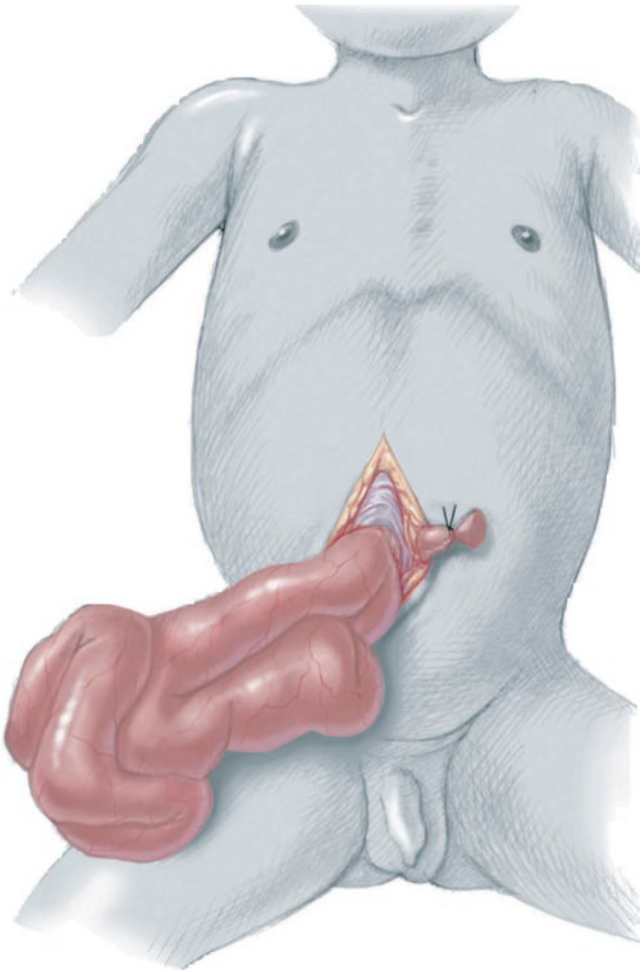

Fig. 21.3 After enlarging the abdominal wall defect, the bowel can be reduced into the peritoneal cavity

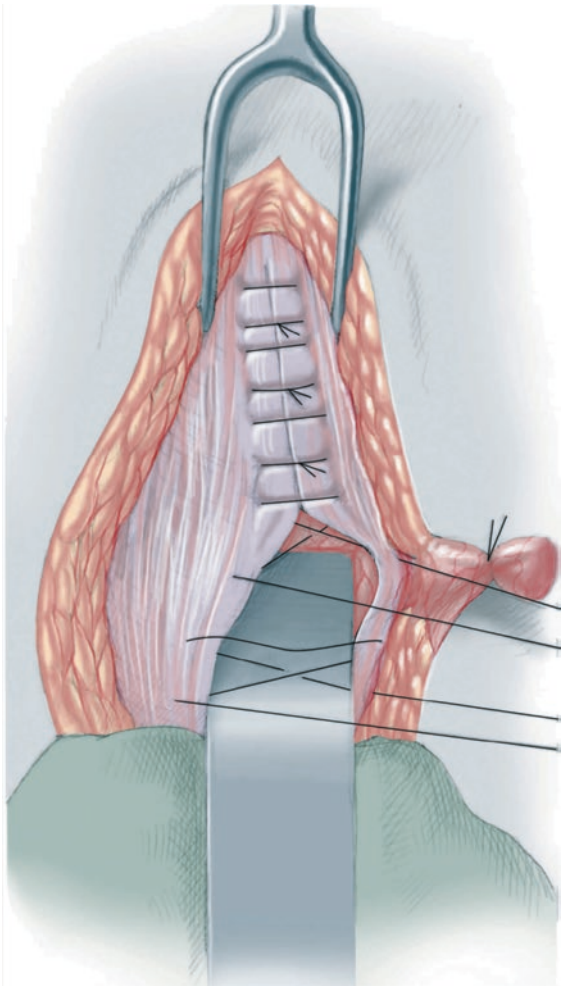

Fig. 21.4 If all of the herniated contents have been reduced into the peritoneal cavity and abdominal wall closure can be accomplished without excessive abdominal wall tension, primary closure should be undertaken. It is important to identify good fascial edges for the closure. The choice of suture material and the technique for placement of sutures is up to the surgeon. My preference (as shown in Fig, 21.5) is interrupted, figure-of-eight, or running suture. It has been my approach to use 3/0 or 2/0 absorbable braided suture if there is mild to moderate tension, and 3/0 or 2/0 monofilament sutures if there is moderate to significant tension. These sutures are placed in a figure-of-eight fashion. It is preferable to place all of the sutures prior to tying them. An important point in patients with gastroschisis is the placement of sutures at the level of the umbilicus. The incidence of an umbilical defect following gastroschisis closure is high. To avoid this, the fascia lateral to the umbilical ring should be clearly identified and used for placement of the suture. If the sutures are placed medial to the umbilical ring, an umbilical defect requiring subsequent repair is highly likely. In tying the sutures in sequence, a thin ribbon retractor placed in the peritoneal cavity underneath the fascia is advantageous, to avoid trapping the bowel during tying of the sutures

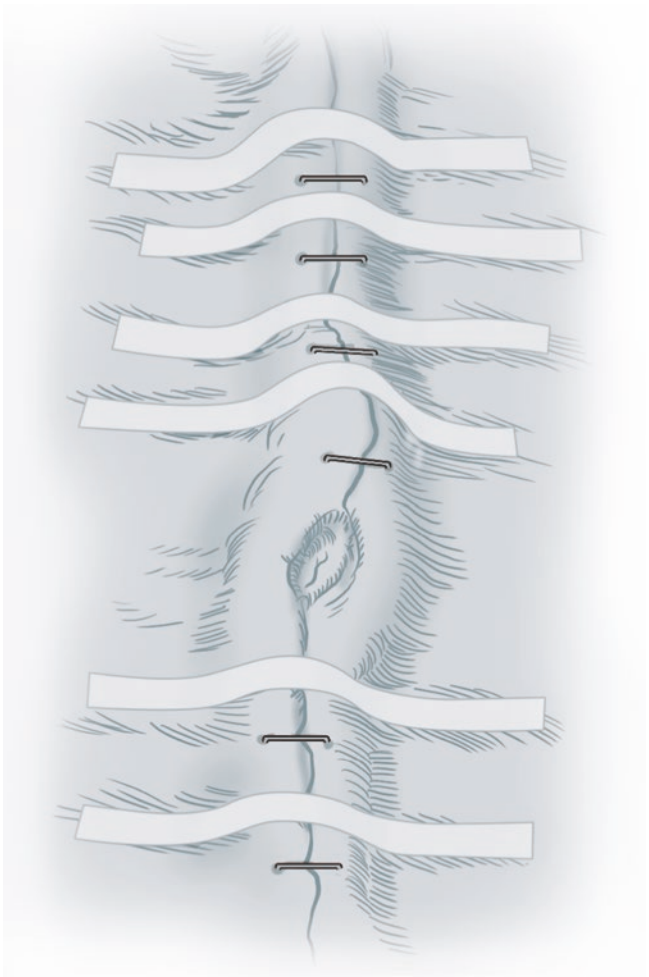

Fig. 21.5 The degree of tension on the skin sutures can dictate the type of closure. It is advantageous to place long skin strips between the stitches or staples to distribute the tension. Relieving the tension at the skin edges will help to diminish the risk of skin disruption

21.3.1 Staged Reduction Using a Silo

If it is determined at the time of the birth or at the initial operative procedure that primary closure is not possible, then an abdominal wall "silo" can be created. This technique, initially described by Schuster and colleagues, has undergone several modifications since its initial description in 1967, but the concept remains the same. A Silastic sheet is sewn to the abdominal fascia circumferentially (Fig. 21.6) and then is used to create a sac around the herniated contents (Fig. 21.7). As much bowel as will be tolerated and without excessive tension is reduced into the peritoneal cavity and then a running suture line is placed across the top of the sac (Fig. 21.8). On successive days (once or twice per day, as tolerated by the infant), the sac is squeezed as much as possible to reduce the herniated contents, and a row of running suture in the silo is placed to maintain the reduction. This staged reduction of the sac produces progressive stretching of the abdominal cavity with simultaneous reduction of the swelling and rigidity of the bowel. Once the bowel has been reduced into the peritoneal cavity and the fascial edges approximate enough to allow removal of the silo, primary fascial and skin closure is performed in the operating theatre. The goal is to try to reduce the bowel into the peritoneal cavity in 7 to 10 days and close the fascial defect and skin in the operating room. After 14 days, the sutures in the fascia holding the silo in place begin to pull away, and the technique may fail (Fig. 21.9).

Recently, the use of a preformed, spring-loaded silo bag frequently, placed at the bedside in the NICU, (Fig. 21.10) in infants with gastroschisis has been shown to be associated with improved fascial closure rates, fewer ventilator days, more rapid return of bowel function, and fewer complications. After the bowel is placed in the preformed silo and the ring is positioned (Fig. 21.11), the reduction process is similar to the previously described technique except that successive ties around the bag as the bowel is reduced instead of placing a suture line. Using this technique, reduction of the herniated bowel has been successfully performed with or without anaesthesia and without enlarging the abdominal wall defect.

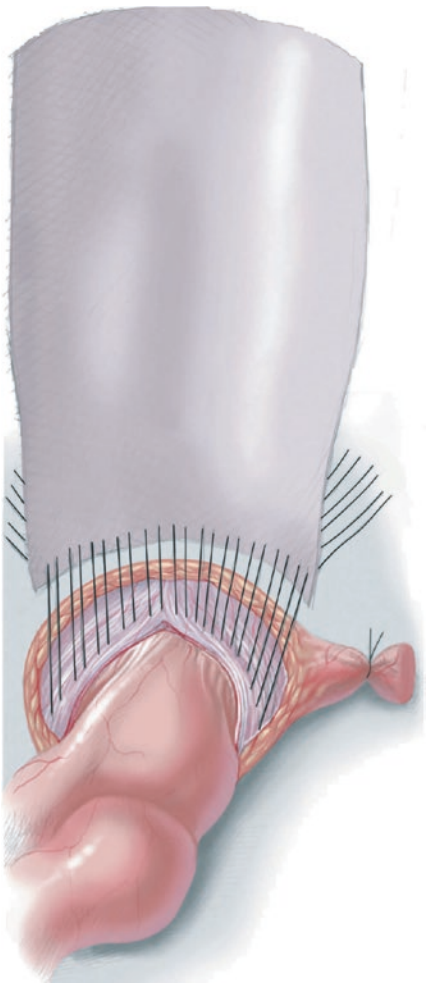

Fig. 21.6 To create an abdominal wall "silo", reinforced Silastic sheeting is sutured to the fascial edges with horizontal mattress sutures of interrupted 3/0 silk suture

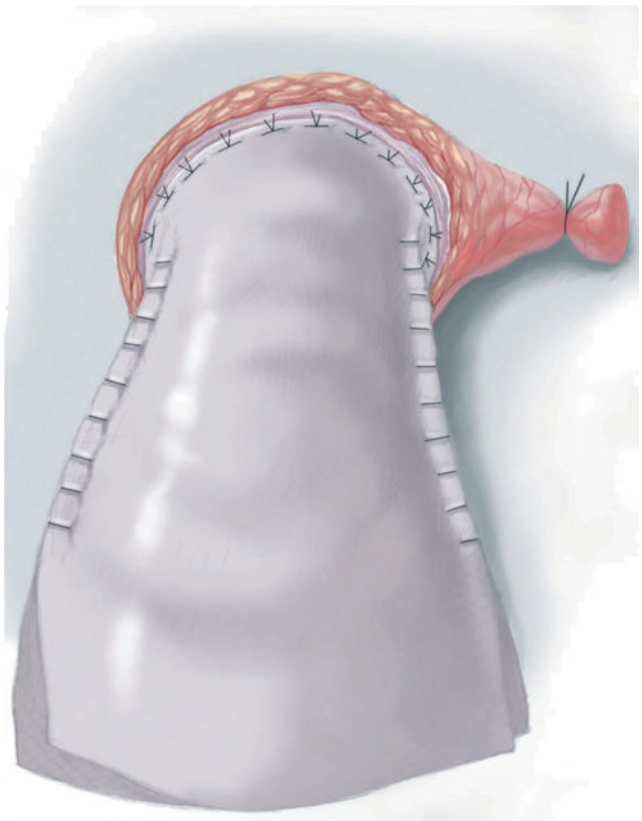

Fig. 21.7 After the Silastic sheets are attached to the fascia on either side of the defect, they are then sewn around the herniated contents with a running suture

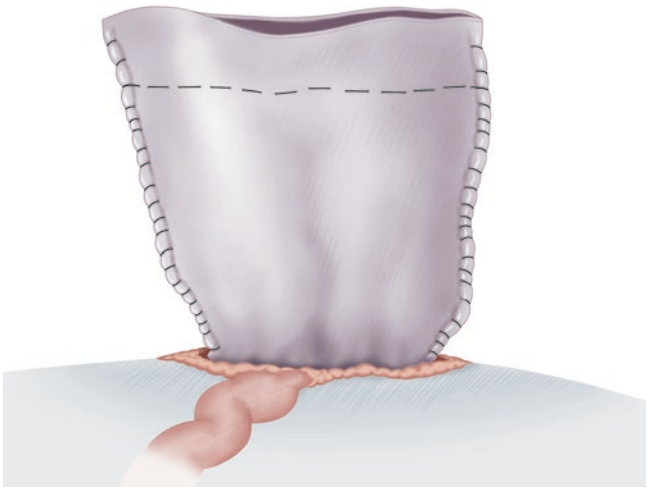

Fig. 21.8 A row of running suture is placed accross the top of the Solo just above the bowel after some bowel has been reduced into the peritoneal cavity. On successive days or as rapidly or slowlly as tolerated, the bowel is gradually completely reduced into the peritoneal cavity

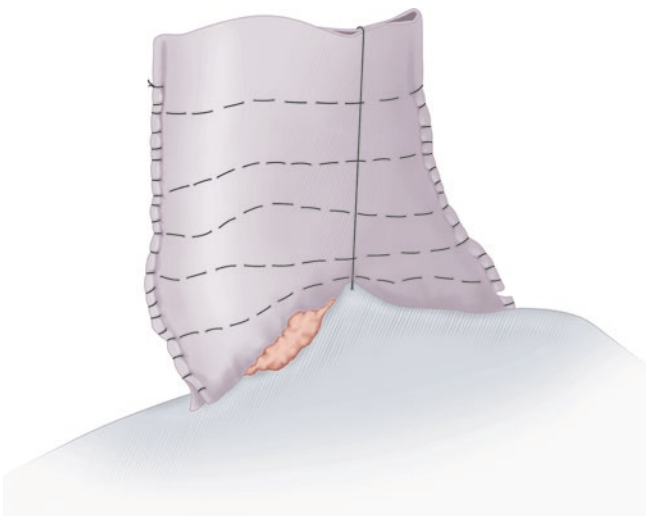

Fig. 21.9 This figure demonstrates complete reduction of the intestinal contents. If done daily, based on the number of rows of suture, the whole procedure took 5 days. However if it took 14 or more days, the sutures in the fascia holding the silo in place may begin to pull away, and the technique may fail as suggested by the loop of bowel pooking through

Fig. 21.10 A preformed, spring-loaded silo bag for the treatment of infants with gastroschisis

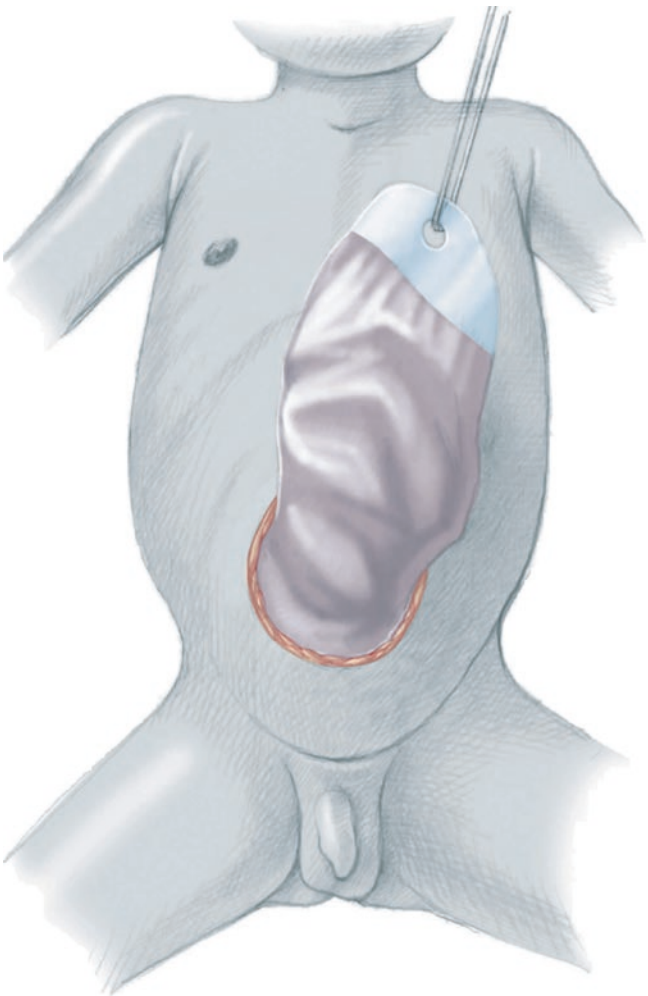

Fig. 21.11 The bowel is placed in the preformed silo and the ring is placed into the peritoneal cavity and positioned just deep to the abdominal wall fascia. This illustration shows a rather large bag and the abdominal wall defect has been enlarged

21.3.2 Sutureless ("Plastic") Gastroschisis Closure

A more recent approach, referred to as "sutureless" or "plastic" gastroschisis closure, has been described. In this technique, the bowel is reduced in the usual fashion but without extending the defect, either primarily or after placement in a silo, but instead of placing sutures to approximate the fascia, the defect is covered with the umbilical stump or a nonadherent dressing. An occlusive dressing is then placed over the site and the wound is allowed to granulate. Once granulation tissue covers the wound bed, the area is covered with dry dressings. Proposed advantages of this technique can include less significant changes in intra-abdominal pressure and decreased narcotic and sedation requirements. Many infants have an umbilical hernia defect following this method of repair, but many of these hernias resolve spontaneously, similar to isolated umbilical hernias.

21.4 Postoperative Management and Outcomes

The outcome for patients with gastroschisis has dramatically improved. Whereas the mortality was 80–90% four or five decades ago, now survival is more than 90%. The improvement in outcome is related to the availability of intravenous nutrition and the use of staged closure when indicated. Late complications and mortality are related to sepsis from either an intra-abdominal or wound complication or from sepsis from a central venous catheter placed for parenteral nutrition. Another late complication, usually after feedings have begun and several (3–4) weeks after abdominal wall closure, is necrotizing enterocolitis. This can be very virulent and with rapid progression. The etiology is unknown.

Once there is evidence of bowel function, it is appropriate to begin enteral feeding. An elemental formula may be better tolerated. When the infant has reached adequate caloric intake enterally, then discharge is appropriate.

In the absence of complications during the recovery from surgery and resolution of the bowel oedema, infants with gastroschisis usually reach goal feedings within 3–4 weeks. Long-term complications after discharge from the hospital are unusual.

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Hypertrophic Pyloric Stenosis

22

Takao Fujimoto

Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical condition encountered in early infancy, occurring in 2–4 per 1000 live births. It is characterized by hypertrophy of the circular muscle, causing pyloric narrowing and elongation. The incidence of disease varies widely with geographic location, season, and ethnic origin. Boys are affected four times more often than girls.

There is evidence of a genetic predisposition to the development of this condition. Siblings of patients with IHPS are 15 times more likely to suffer the condition than children who have no family history of IHPS. The cause of hypertrophic circular muscle of the pylorus is still obscure. Various hypotheses have been advocated, including abnormal peptidergic innervation, abnormality of nitrergic innervation, abnormalities of extracellular matrix proteins, abnormalities of smooth-muscle cells, and abnormalities of intestinal hormones.

22.1 Presentation

The typical clinical presentation of infants with IHPS is nonbilious vomiting, usually occurring at 2–8 weeks of age in full-term infants. The development of IHPS in preterm infants has been described previously as a rare entity, but a recent large population study in the United States reported a significant increase in IHPS rates among premature infants, and the preterm infants typically presented at a later chronological age than full-term infants. Initially there is only regurgitation of feeds, but over several days, vomiting progresses to be characteristically projectile. It occasionally contains altered blood in emesis (appearing as brownish discolouration or coffee grounds) as a result of gastritis and/or oesophagitis.

22.2 Diagnosis

The diagnosis is usually based on the clinical history and physical examination of a "palpable pyloric tumour". Ultrasonographic scanning of the abdomen reveals a typical hypoechoic ring with an echogenic centre of increased muscle thickness. In difficult and/or complicated presentations, a contrast meal may be required, which shows a characteristic narrowed, elongated pyloric canal.

22.3 Treatment

Persistent nonbilious vomiting in these patients results in chloride depletion, metabolic alkalosis, and dehydration. Haematological and biochemical analysis should be undertaken. Any fluid, electrolyte, and acid-base imbalance should be corrected prior to surgery. Oral feeding should be discontinued and a nasogastric tube should be inserted prior to surgery to keep the stomach empty. The operation for pyloric stenosis is not an emergency and should never be undertaken until serum electrolytes have returned to normal.

Ramstedt's pyloromyotomy is the universally accepted operation for pyloric stenosis. A 3-cm transverse right upper quadrant, muscle-splitting incision provides excellent exposure and direct access to the pylorus with minimal retraction. Another incision that is commonly used is a supra-umbilical fold incision. Although a supra-umbilical skin-fold incision has a better cosmetic result, it has been argued that delivery of the pyloric tumour can be difficult and time-consuming and may damage the serosa of the stomach or duodenum by tearing. Some surgeons have used transumbilical intracavitary pyloromyotomy without delivering the pyloric tumor outside. In recent years, more and more centres are employing laparoscopic pyloromyotomy as the surgical approach. The main advantage of the laparoscopic pyloromyotomy is the superior cosmetic result.

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A nasogastric tube must be placed before the induction of anaesthesia, if the tube was not placed preoperatively. If a barium meal study has been carried out prior to surgery, it may be necessary to remove the residual barium meal by gastric aspiration and irrigation.

22.3.1 Operative Procedure: Ramstedt's Pyloromyotomy

Figures 22.1, 22.2, 22.3, 22.4 and 22.5 illustrate an open procedure.

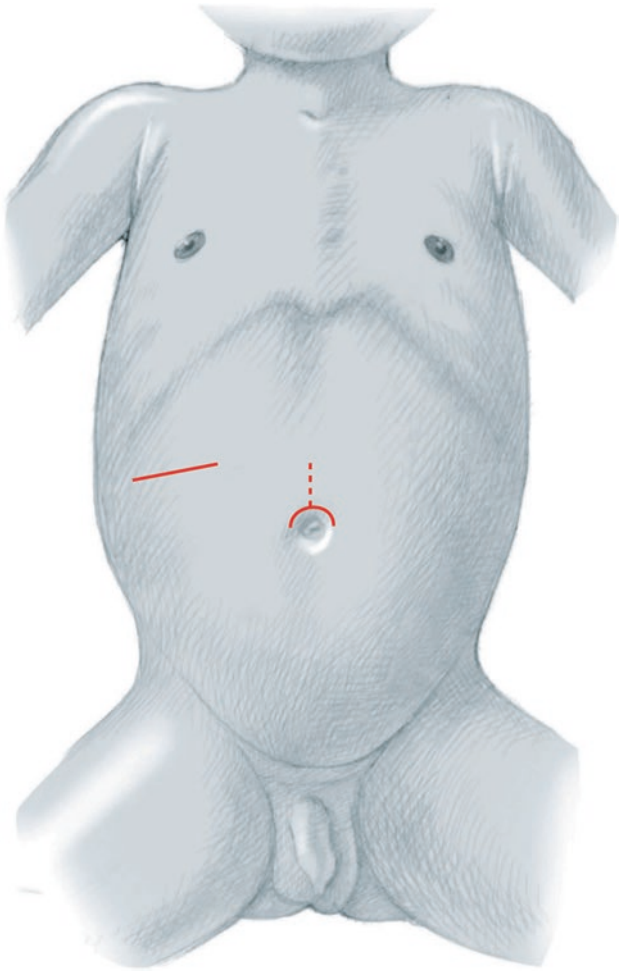

Fig. 22.1 The patient is placed in the supine position. After the induction of anaesthesia and endotracheal intubation, careful abdominal palpation will usually identify the site of the pyloric tumour. A transverse incision 2.5–3 cm in length is made lateral to the lateral border of the rectus muscle. The incision is deepened through the subcutaneous tissue; the underlying external oblique, internal oblique, and transverse muscles are split. The peritoneum is opened transversely in the line of the incision.When a supraumbilical skin fold incision is employed, a circumumbilical incision is made through about two thirds of the circumference of the umbilicus. The skin is undermined in a cephalad direction above the umbilical ring and the linea alba is exposed. The linea alba is divided longitudinally in the midline from the umbilical ring to as far cephalad as necessary to allow easy delivery of the pyloric tumour

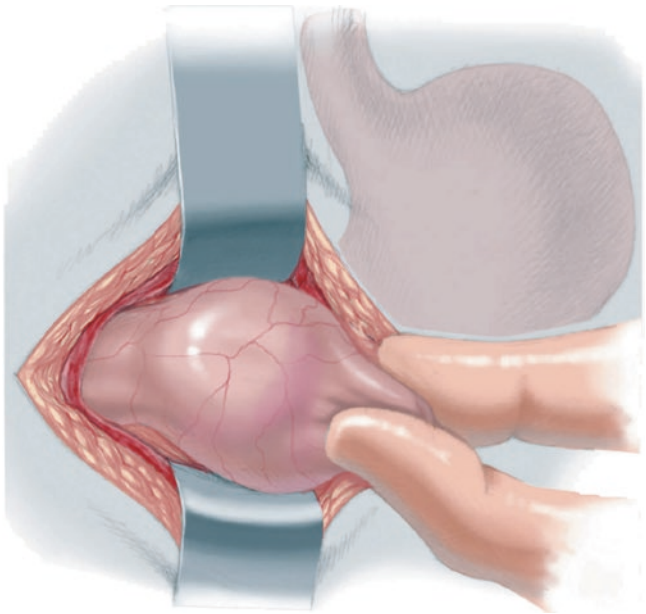

Fig. 22.2 The stomach is identified, is grasped proximal to the pylorus with a noncrushing clamp, and is brought through the wound. The greater curvature of the stomach then can be held in a moist gauze swab, and the pylorus can be delivered through the wound with traction inferiorly and laterally. Grasping the duodenum or pyloric tumour directly by forceps should be avoided, as it often results in serosal laceration, bleeding, or perforation

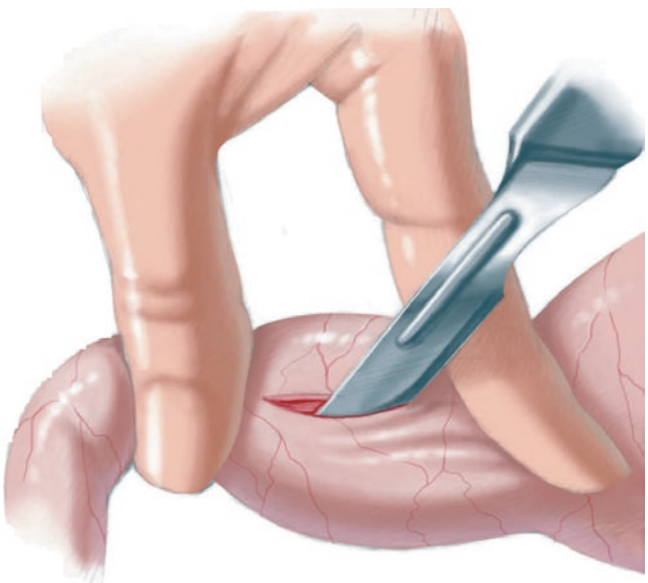

Fig. 22.3 The pylorus is held with surgeon's thumb and forefinger to stabilize and assess the extent of hypertrophied muscle. A seromuscular incision is made over the avascular area of the pylorus with a scalpel, commencing 1 or 2 mm proximal to the prepyloric vein along the gastric antrum. The incision should go far enough onto the gastric antrum (at least 0.5–1.0 cm from the antropyloric junction, where the muscle is thin)

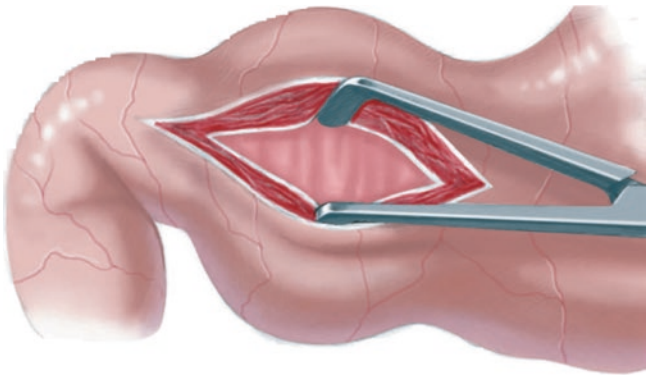

Fig. 22.4 The scalpel handle is used to further split the hypertrophied muscle down to the submucosal layer. Then the pyloric muscle is spread widely. The spreader is placed at the midpoint of the incision line and the muscle is spread perpendicularly; spreading must be continued proximally and distally. Gentle spreading is required to obtain a complete myotomy. Mucosal tears are most common at the pyloroduodenal junction because of the attempt to split all remaining muscle fibres. To reduce the risk of mucosal tears, care should be taken when spreading the pyloric muscle fibres at the duodenal end

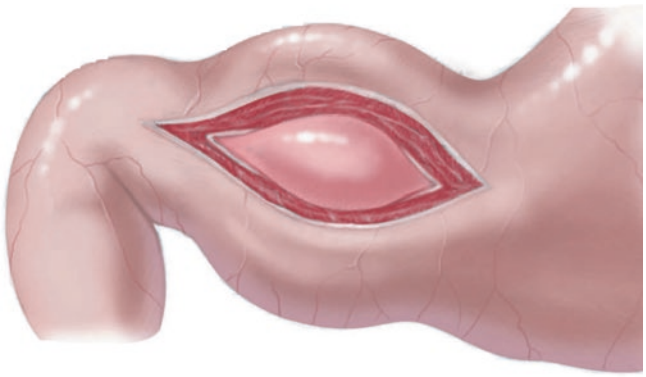

Fig. 22.5 Loose prolapsing of intact mucosa is evidence of a satisfactory myotomy. To test the mucosal injury, the stomach is inflated through the nasogastric tube, and passage of air through the pylorus to the duodenum is confirmed. Then the pylorus is dropped back into the abdomen. Bleeding from the myotomy edge or submucosal surface is frequently seen, but it is generally venous and always stops after returning the pylorus to the abdominal cavity. The posterior rectus fascia and peritoneum are approximated with a running 4/0 absorbable suture material, and the anterior fascia is closed with 5/0 absorbable suture material

22.3.2 Operative Procedure: Laparoscopic Pyloromyotomy

Figures 22.6, 22.7, and 22.8 show the procedure for laparoscopic pyloromyotomy (LP).

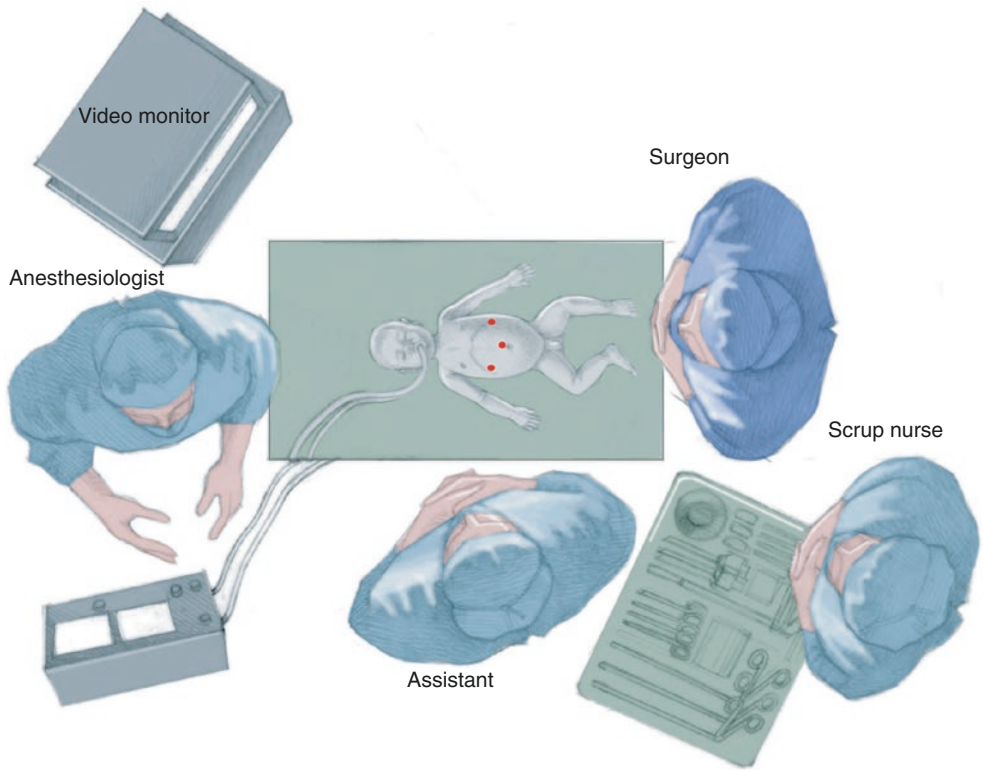

Fig. 22.6 For the laparoscopic procedure, the patient is placed in the supine position at the end of the operating table (or 90° to the anaesthesiologist). The video monitor is placed at the head of the table, and the surgeon stands at the end of the table with the assistant to the patient's right. The abdomen is scrubbed and draped in a sterile fashion. Attention must be paid to ensure the appropriate preparation of the umbilicus. The access sites are injected with local anaesthetic (0.25% bupivacaine) with epinephrine, which is used to reduce the postoperative pain and reduce the risk of bleeding from the stab wound. The author prefers an open procedure for insertion of the primary port. A curvilinear supraumbilical incision (4.0–5.0 mm) is made and carried down to the peritoneal cavity. At the level of the umbilical fascia, 4/0 absorbable suture material is placed circumferentially to anchor the port; it will also be used for closure of the peritoneal cavity after the laparoscopic pyloromyotomy is completed. Intra-abdominal pressure is maintained at 8 mm Hg, and the insufflation rate is set at 0.5 L/min. In the right midclavicular line just below the costal margin (just above the liver edge), a no. 11 scalpel blade is used to make a 2- to 3-mm stab incision under direct vision. Also using the no. 11 scalpel blade, a second stab incision is made under direct vision, just below the costal margin in the left midclavicular line. An atraumatic grasper is placed directly through the right upper quadrant stab wound and is used to retract the inferior border of the liver superiorly and expose the hypertrophic pylorus. A retractable myotomy knife (retractable arthrotomy knife or Endotome) is inserted directly through the left stab wound. Working ports are usually not necessary and instruments are directly introduced through these stab wounds

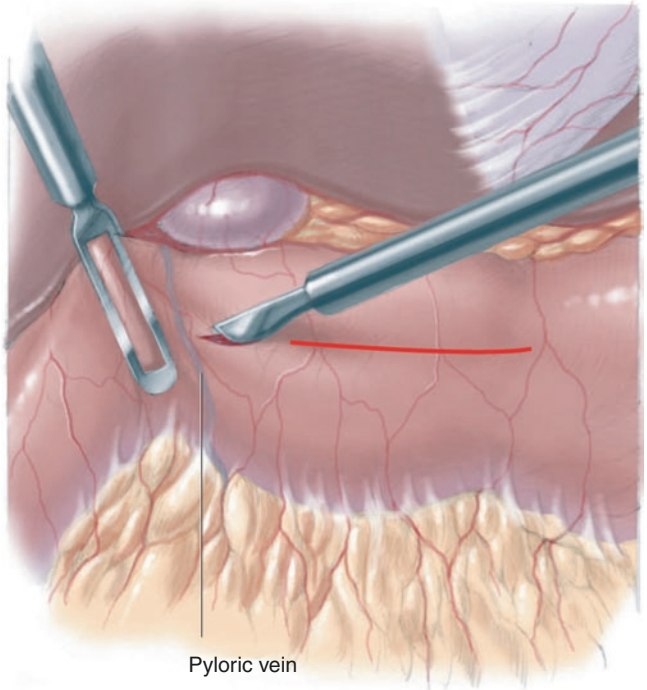

Fig. 22.7 The working instruments (the retractable myotomy knife and atraumatic laparoscopic grasper) are used to assess the extent of the hypertrophied pylorus by palpating the margins of the pylorus as one would do with thumb and forefinger in the open procedure. The duodenum is then grasped just distal to the pyloric vein (pyloroduodenal junction) and retracted using the atraumatic grasper to expose the avascular surface of the hypertrophic pylorus. In positioning the pylorus for myotomy, lateral and slightly anterocephalad retraction of the distal pylorus achieves excellent exposure of the avascular surface of the hypertrophic pylorus. This manoeuvre also exposes the proximal margin of hypertrophied muscle that is seen as a deep fold in the wall of the stomach. A seromuscular incision is made over the hypertrophic pylorus with the retractable myotomy knife, commencing 1 to 2 mm proximal to the pyloroduodenal junction and extending to the gastric antrum. The incision should go far enough onto the antrum (at least 0.5–1.0 cm proximal to antropyloric junction). Care must be taken at this stage that this incision is deep enough to allow the insertion of the pyloric spreader blades; it must penetrate the pyloric muscle somewhat deeper than is usual with the conventional open procedure

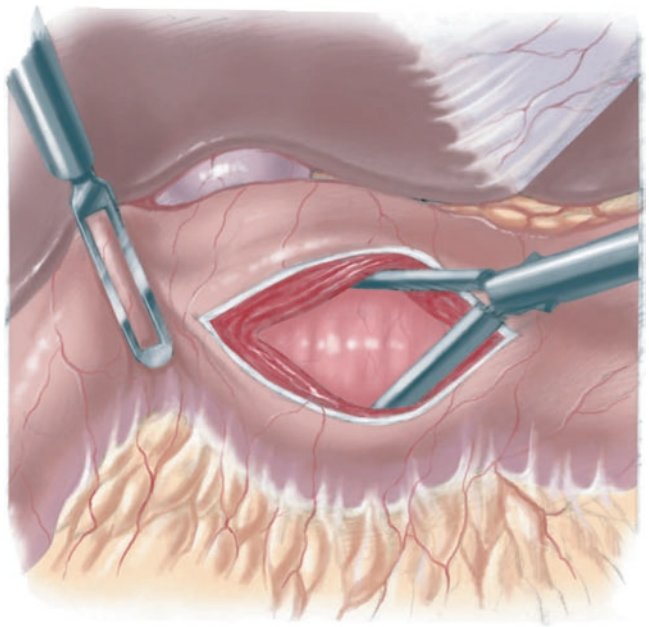

Fig. 22.8 After the muscle is incised, the blade is then retracted and the sheath of the knife is used to further split the hypertrophied muscle fibres, as the scalpel handle is used in the open procedure, until mucosa is visualized. The retractable myotomy knife is removed and a laparoscopic pyloromyotomy spreader is introduced into the abdominal cavity directly through the left stab wound to complete the pyloromyotomy. The spreader is placed in the midpoint of the seromuscular incision line and the muscle is spread perpendicularly. Once the initial spread reaches the mucosa, spreading must be continued proximally and distally. Pushing the spreader towards the mucosa or rapid spreading can result in mucosal tear. To avoid mucosal tears, the spreader should not be placed at the proximal and distal edges of the incisional (myotomy) line. To test for mucosal injury, the stomach is inflated through the nasogastric tube (160–180 mL), as is usually done in open techniques. Bulging of the mucosal layer with no evidence of defect should be confirmed. Greenish or yellowish fluid at the myotomy area is a sign of mucosal tear. After the successful myotomy, the instruments are withdrawn under direct vision and the pneumoperitoneum is evacuated. The nasogastric tube is also removed after completing the surgery. The umbilical fascia is reapproximated with the 4/0 absorbable suture material, which is already in place, and the skin of all the wounds is reapproximated with skin adhesive tapes

22.4 Complications

Mortality associated with pyloromyotomy is rare today. Early diagnosis and proper perioperative management reduces complications. In spite of these advances, there remains about an 8–10% incidence of associated perioperative morbidity such as perforation, wound infection, and wound dehiscence.

In an open procedure, essentially right upper quadrant incision and circumumbilical incision, manipulation of and tension on the pylorus to deliver it through the wound can induce oedema in the muscle layer, mucosal swelling, and occasionally serosal laceration. A laparoscopic pyloromyotomy (LP) is a less traumatic operation. The tolerance of an early feeding regimen after LP confirms the lack of trauma to the pylorus during the procedure, which we feel is the most important benefit of LP. The use of 3.0-mm instruments improves cosmesis.

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Gastrostomy

23

Michael W. L. Gauderer

A gastrostomy (Gr. *gaster* stomach and *stomoun* opening or mouth) is a conduit between the skin surface and the gastric mucosa. Long-term gastric access commonly is maintained by means of a short skin-level tubular device. Its simplicity notwithstanding there are few interventions for which there is a greater palette of options.

23.1 Indications

In infants and children, gastrostomies are indicated primarily for long-term enteral feedings and less frequently for decompression. The addition of a transpyloric tube to a gastrostomy device provides both. In the past four decades, advances in perioperative management have led to a more selective use of gastrostomies in patients with various typical pediatric surgical conditions such as congenital anomalies of the gastrointestinal tract and the abdominal wall. On the other hand, the use of gastrostomies in infants and children without surgical pathology has increased markedly. The main indication for direct gastric access in these patients is an inability to swallow, usually secondary to central nervous system impairment. Additional indications for a gastrostomy are the need to provide feeding supplementation in children unable or unwilling to consume adequate calories orally, long-term administration of unpalatable diet or medications, and access for esophageal bougienage.

When feeding is the main indication, two important questions must be addressed:

First, *nasogastric tube or gastrostomy?* Nasogastric tubes should be preferred if the expected duration of enteral access is relatively short, such as 1 or 2 months, because the newer, small feeding tubes are highly biocompatible and lubricous, remaining smooth and soft for prolonged periods of time. Gastrostomies (or gastrotomies plus jejunal extension) should be considered when gastric (or jejunal) access is expected to last more than several months.

Second, *gastrostomy only or gastrostomy plus antireflux operation?* Neurologically impaired children, the main candidates for gastrostomy, frequently have foregut dysmotility and associated gastroesophageal reflux. Additionally, anatomical abnormalities of the epigastrium, shunts or catheters, and scoliosis are not uncommon. Because gastrostomies may unmask reflux, these children should be evaluated prior to placing a stoma, usually with an upper gastrointestinal contrast series and a combined, multichannel intraluminal impedance and pH study. Endoscopy with biopsy, manometry, and gastric emptying studies may be added, if deemed necessary. Unfortunately, these studies are not particularly helpful in predicting post-gastrostomy reflux. For this reason, a helpful and simple approach is a trial of nasogastric tube feedings for 1–2 weeks. If these are tolerated, only a gastrostomy is placed. If they are not, a gastrostomy plus a temporizing transgastrostomy feeding jejunostomy tube is placed. If the management of gastroesophageal reflux remains a problem, a "protective" antireflux operation can be added, usually without taking down the gastrostomy. It is worth noting that careful dietary manipulation and the addition of contemporary medications have markedly reduced the need for antireflux procedures in children.

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23.2 Choice of Technique

There are four basic approaches for establishing a gastrostomy:

- **Gastrostomy *with* laparotomy**. This technique consists of the formation of a short, serosa-lined channel from the anterior gastric wall around a catheter. This catheter is placed in the stomach and made to exit either parallel or vertically to the serosa. The anterior gastric serosa is apposed to the peritoneal surface of the anterior abdominal wall with sutures. The Stamm technique (Figs. 23.1, 23.2, 23.3, 23.4, 23.5 and 23.6) is the most widely employed

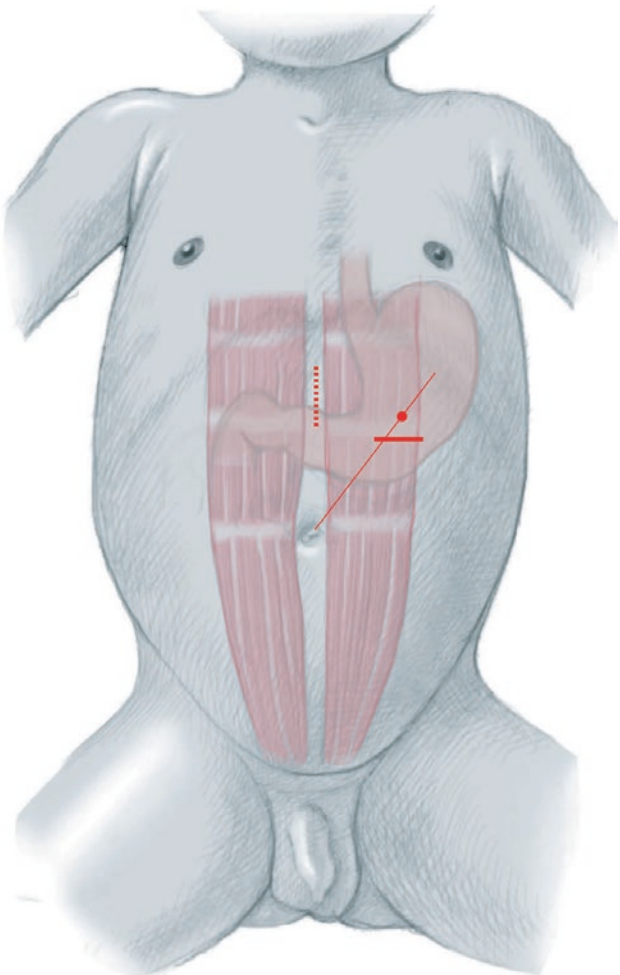

Fig. 23.1 The stomach is approached through a short, transverse supraumbilical incision. Fascial layers are incised transversely and the muscle is retracted or transected. The catheter exit site should be approximately at the junction of the lower two thirds and the upper one third of a line drawn from the umbilicus to the mid portion of the left rib cage, over the mid rectus muscle. It can be useful to mark the site prior to making the main incision. A vertical incision is used in children with a high-lying stomach or a very narrow costal angle. Generally, catheters should not be brought out through the incision, because doing so may predispose to wound complications and leakage

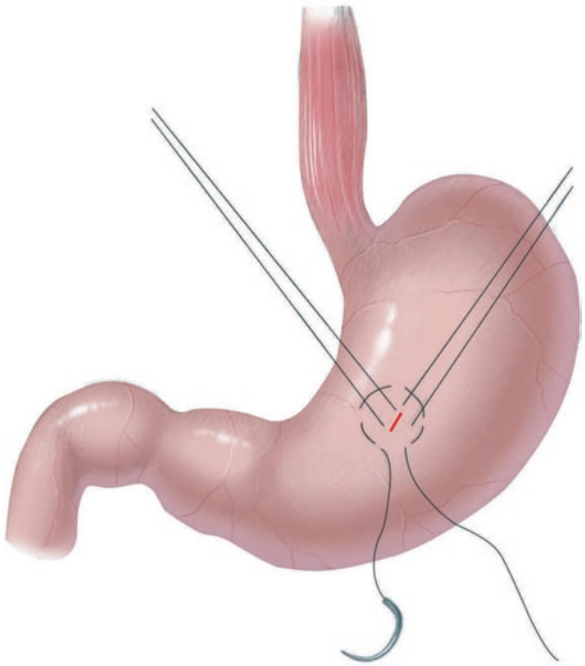

Fig. 23.2 A site on the mid body of the stomach is selected for the catheter insertion, taking into consideration the previously described caveats. Two traction guy sutures are used to lift the gastrostomy site, and a purse-string suture of age-appropriate synthetic absorbable material is placed. The diameter of the purse-string should be adequate for invagination of the gastric wall after insertion of the catheter, but not excessive, to avoid narrowing of the stomach, particularly in small children. A lower guy suture (*not depicted*) may be added to pull the stomach caudally, enhancing the exposure and allowing better gastric access

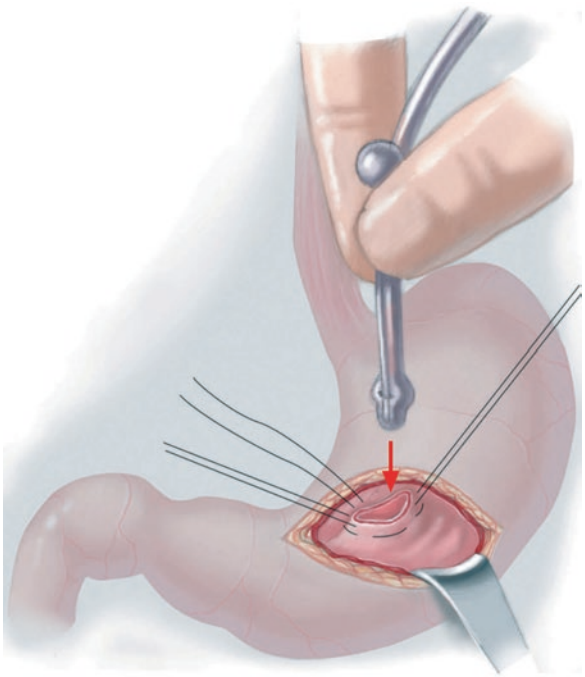

Fig. 23.3 The gastrotomy is performed with fine scissors or cautery while the guy sutures are lifted to prevent injury to the posterior gastric wall. The de Pezzer catheter (or another suitable catheter) is introduced using a simple stylet while the sutures are elevated

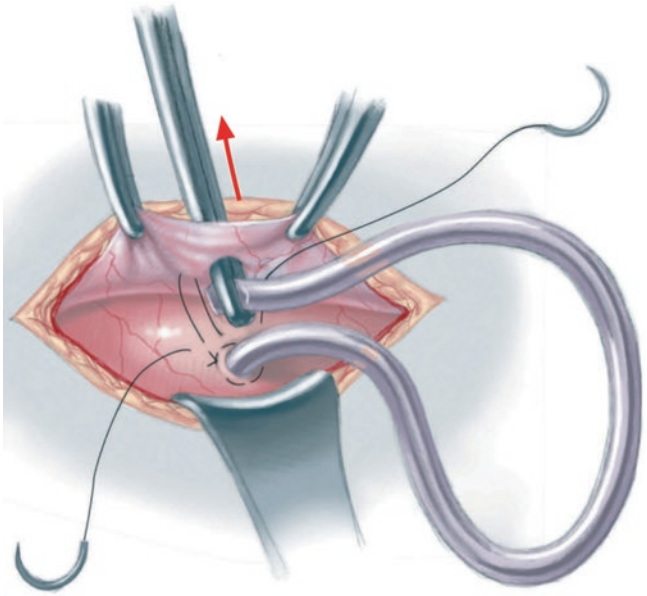

Fig. 23.4 A stab counter-incision for the catheter exit is made, usually at a slightly cranial site. A curved hemostat is introduced into it (initially without piercing fascia, muscle, and peritoneum), pushing the anterior gastric wall inward. A continuous synthetic, absorbable monofilament suture (preferably with a needle at both ends) is used to anchor the stomach to the anterior gastric wall. After the posterior 180 degrees of the "anastomosis" are completed, the peritoneum and fascia are incised and the tip of the clamp is pushed through. The catheter end is grasped and the tube is brought out through the counter-incision

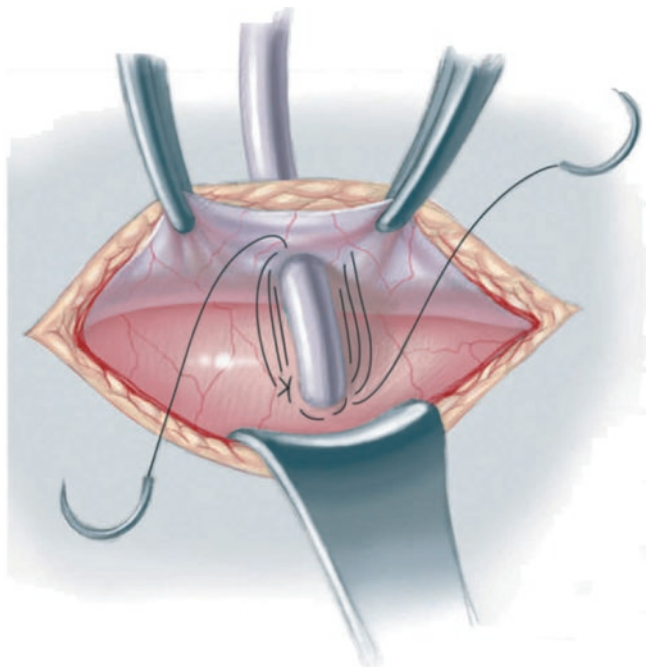

Fig. 23.5 Placement of the continuous monofilament suture then completes the apposing "anastomosis." When tied, this suture provides a 360-degree fixation of the stomach to the abdominal wall and a watertight seal. It also obviates the need for a second purse-string suture

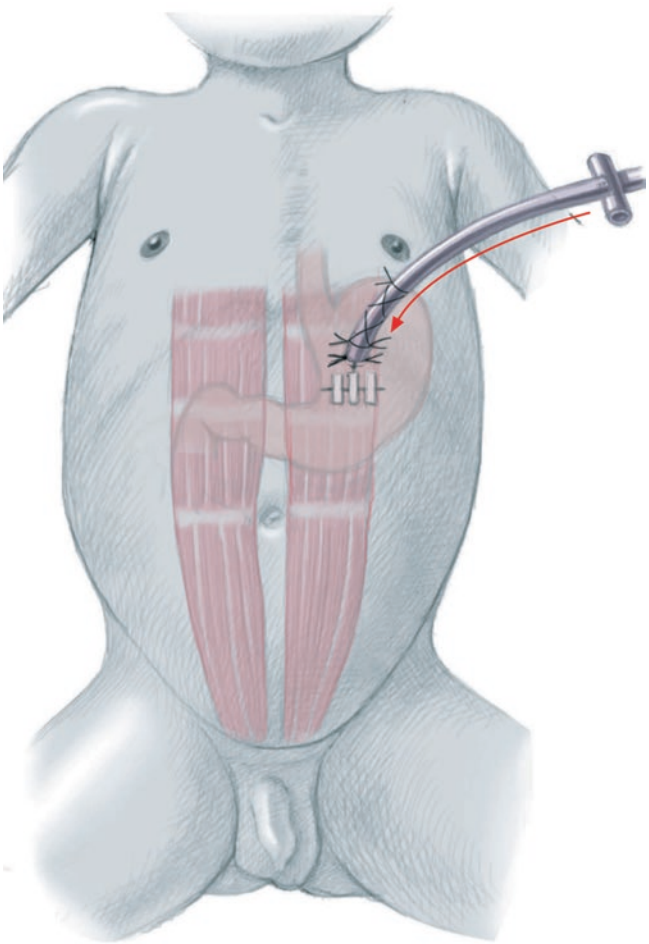

Fig. 23.6 The abdominal wall layers are closed with synthetic absorbable sutures, and the skin is approximated with subcuticular stitches and adhesive strips. The catheter is secured with synthetic monofilament sutures, which are removed 1–2 weeks after the operation. If the tube is to remain long-term, a small, immobilizing cross-bar is added to prevent distal catheter migration. Care must be taken to avoid excessive pressure on this cross-bar, to prevent pressure necrosis

gastrostomy in this group. It is suitable for children of any size and even the smallest stomach.

- **Gastrostomy *with* laparotomy *without* catheter**. In this procedure, a tube is created from a full-thickness gastric flap leading to the skin surface, where it is anchored with sutures. A catheter is introduced only intermittently for feeding. The Janeway procedure is an example. Despite this advantage, this technique is not well suited for children.
- **Gastrostomy *without* laparotomy**. This group encompasses various techniques in which a catheter or skin-level device is placed with the aid of flexible endoscopy or radiological assistance. In the percutaneous endoscopic gastrostomy (PEG) techniques, the catheter can be introduced from inside the stomach to the skin surface (Pull and Push methods) or in the opposite direction (Introducer

method). The latter, as well as the radiological techniques, require temporary fixation of the stomach to the abdominal wall (such as with T fasteners) for catheter placement. The original Gauderer pull PEG is illustrated. Laparoscopic control may be added for safety, particularly if anatomical abnormalities such as scoliosis, organomegaly, or malrotation are present.
- **Gastrostomy *without formal* laparotomy**. With this approach, which has gained wide acceptance among pediatric surgeons, a gastrostomy device is placed with the assistance of laparoscopic visualization and minimally invasive instrumentation. Although this category encompasses multiple options, all techniques are based on previously established operations. A couple of laparoscopically aided procedures are illustrated.

The selection of gastrostomy technique depends on multiple factors, including its main purpose; the age and size of the child; the underlying pathology; the experience of the operator with open, endoscopic, or laparoscopic procedures; and the availability of one or more of the various skin-level devices. Gastrostomies placed by interventional radiology techniques follow the same basic principles, but are not described here. Gastrostomies with laparotomy are usually employed as an adjunct to a major intra-abdominal intervention. Percutaneous endoscopic procedures are simple and expedient. The deployment of "button"-type, nonballoon skin-level devices at the time of the PEG procedure is possible and obviates the need to exchange the long tube for a skin-level device. Laparoscopically aided gastrostomies have a good safety record but are slightly more complex. One of their main advantages is that they allow the primary insertion of various balloon-type and non–balloon-type skin-level devices. What all gastrostomies must have in common is a reliable approximation of the anterior gastric wall serosa to the peritoneum of the anterior abdominal wall.

23.3 General Guidelines

Choosing the Tube. The choice of gastrostomy tube should take into consideration the purpose of the stoma, the patient's size, and the technique employed, as well as device availability. At this time there are several options:

- "Traditional" long tubes, retained intragastrically, either by means of a balloon (Foley type) or without a balloon (de Pezzer or mushroom type, Malecot or wing type, or flat disk type)
- Skin-level devices or buttons, retained intragastrically, either balloon type or without a balloon (mushroom type)
- Hybrid tubes that can be converted from long shafts to skin-level devices, usually with intragastric mushroom-type or disc-type retaining domes
- Other tube types. Most contemporary access devices are made of either silicone rubber or polyurethane. Progress continues in developing longer-lasting, more biocompatible and bacteriostatic implements that are easier to use and resistant to accidental removal

The length and the diameter of the gastrostomy tube (notably a skin-level device or "button") vary with the distance between the gastric mucosa and the skin surface, as well as the type of nutrient or medication to be administered. Skin-level devices should be long enough to allow for slight "play" (thus avoiding tissue injury through compression) and to account for future weight gain. The diameters of tubes commonly employed in children range from 12F (12Ch) to 20F. It is important to remember that, when selecting a PEG catheter for the "pull" technique, the intragastric, retaining dome should be collapsible enough to slide smoothly through the oropharynx, esophagus, and esophagogastric junction.

Placement of the Stoma Site. The stoma site on the abdominal wall should be placed away from the rib cage because, with the child's growth, the distance between the two tends to become shorter. A gastric access device that is too close to the ribs will cause discomfort and interfere with care. Additionally, pivoting motion resulting from breathing and moving can lead to stoma enlargement and leakage. In small children, the linea alba tends to be broad and very thin and also should be avoided as an exit site.

The stoma site on the stomach should be in the midbody—away from the greater curvature, because the proximity to the transverse colon can lead to a gastrocolic fistula; away from the fundus, to allow for a possible future fundoplication; and away from the presumed site of the gastric pacemaker, near the splenic hilum. It is critical to avoid the antrum to prevent pyloric obstruction by the catheter tip and interference with gastric emptying. A stoma in this position is also more likely to leak. Placements that exert downward traction on the stomach, particularly along the greater curvature, can interfere with the angle of His at the esophagogastric junction and therefore should be avoided.

23.4 Operative Procedures

A single-dose prophylactic intravenous antibiotic is advisable for all procedures, especially the PEG. Local wound infiltration with a long-lasting local anesthetic is recommended for all incisions.

23.4.1 Stamm Gastrostomy

The Stamm gastrostomy operation is performed using general endotracheal anesthesia. A nasogastric tube is inserted to evacuate contents and help identify the stomach in children with abnormal upper abdominal anatomy. The patient is positioned with a small roll behind the back to elevate the epigastrium, then prepared and draped. For infants, a thin plastic, small-aperture drape is used to help with temperature maintenance. We prefer to use silicone rubber de Pezzer-type catheters ranging in size from 12F or 14F (neonates) to 20F (adolescents), or PEG-type catheters in which the "dome" has been modified to allow insertion with a stylet. For preterm infants or neonates with a very small stomach (*e.g.*, esophageal atresia without fistula), a 10F T tube (as used in biliary surgery) can be employed.

Figures 23.1–23.6 illustrate the Stamm gastrostomy operation.

This technique can be modified to allow for the insertion of a skin-level device instead of a long tube. Either a nonballoon or a balloon-type device may be used. An initially placed long catheter can also be converted to a skin-level device, without removing the initially placed catheter, using a skin-level conversion kit.

If the placement of a continuous suture between the gastric serosa and the peritoneum is technically difficult, interrupted sutures may be employed.

For immediate postoperative venting or decompression, the catheter can be connected directly to a sputum specimen trap to avoid additional lengthy tubing.

23.4.2 Percutaneous Endoscopic Gastrostomy (PEG)

PEG procedures are best performed in the operating room. In older children and those able to tolerate upper endoscopy without compromising the airway, the procedure may be undertaken using local anaesthesia with sedation as needed. Younger children require general endotracheal anesthesia, primarily because of anticipated difficulties with airway management. Two operators are required—one for the endoscopy and one for the insertion of the guidewire and pulling/guiding the catheter into place. For the endoscopy, the smallest available flexible pediatric gastroscope is used. The catheter, with its retaining internal "dome" or disk, must be soft and collapsible enough to glide atraumatically through the oropharynx and esophagus (Fig. 23.7).

Contraindications to PEG are inability to perform upper tract endoscopy safely or to identify transabdominal illumination and clearly recognize an anterior gastric wall indentation. Anatomical abnormalities such as marked scoliosis, intestinal malrotation, and organomegaly, as well as the presence of intra-abdominal shunts, require caution; the procedure is best performed by adding laparoscopic control. Ascites, coagulopathy, and intra-abdominal infections render the procedure inadvisable.

Figures 23.8, 23.9, 23.10, 23.11, 23.12, 23.13 and 23.14 illustrate a PEG procedure.

The long gastrostomy tube can be converted to a skin-level device by using a changeable port-valve of the skin-level conversion kit type.

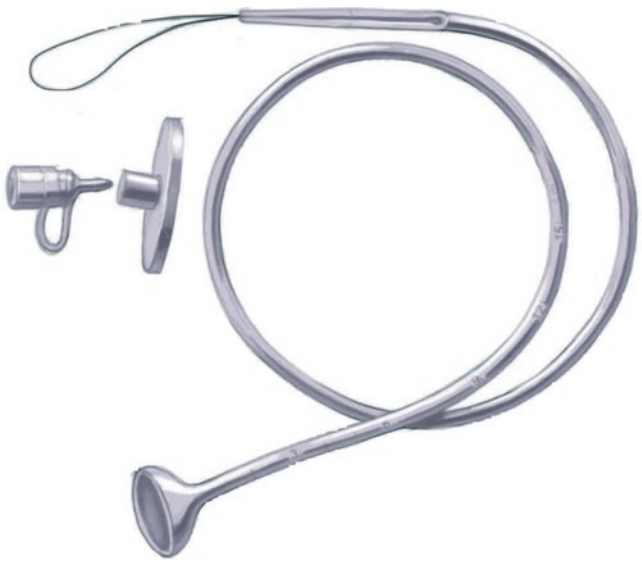

Fig. 23.7 A size 14F–16F silicone rubber catheter is well suited for younger children, and a 20F tube is used for older children and adolescents

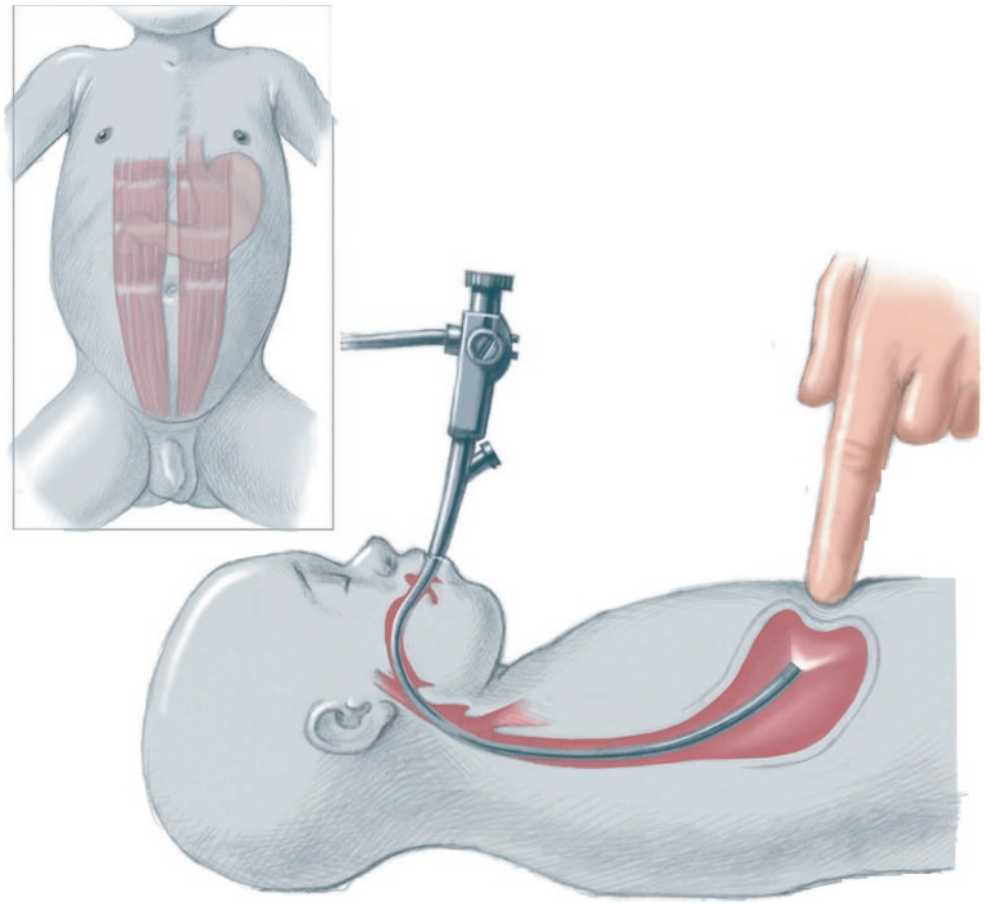

Fig. 23.8 The abdomen is prepared and draped. The catheter exit site is tentatively selected. The gastroscope is inserted, but the stomach is not immediately insufflated. The snare is advanced into the operating channel of the scope. Once all necessary equipment is available and ready for use, the room lights are dimmed and the stomach is insufflated. Under-insufflation or, more importantly, over-insufflation should be avoided to minimize the possibility of accidentally piercing the colon. Excessive insufflation of the small intestine tends to push the transverse colon in front of the stomach and into harm's way. It also distorts the stomach, which may interfere with correct catheter placement. Once the stomach is appropriately distended, digital pressure is applied to the proposed gastrostomy site, which usually corresponds to the area where transillumination is brightest. Transillumination and clear visualization of an anterior gastric wall indentation are key points. Without these, laparoscopic control or an "open" gastrostomy should be employed. Long-lasting local anesthetic is drawn into a syringe, and the proposed PEG site is injected. The needle is advanced further, and continuous aspirating pressure is applied to the plunger. Air bubbles should be visible in the remaining fluid when the tip of the needle is seen by the endoscopist. If air bubbles are noticed before the needle tip is in the stomach, the colon or other intestinal loop may be interposed between the stomach and the abdominal wall. Another safety tip is to estimate the distance between the gastric mucosa and the skin, and use the needle length as a guide. Excessive length warrants caution. In patients with an abnormal epigastric anatomy, the addition of laparoscopy is recommended

Alternatives aimed at avoiding conversion of the long-tube to a skin-level device, such as a catheter-button combination are commercially available. Such products allow the initial placement of a non-balloon button. It is important, however, to ensure that this type of skin-level device is neither too tight nor too loose. In the latter case, special supporting/adjusting rings are included in the kit.

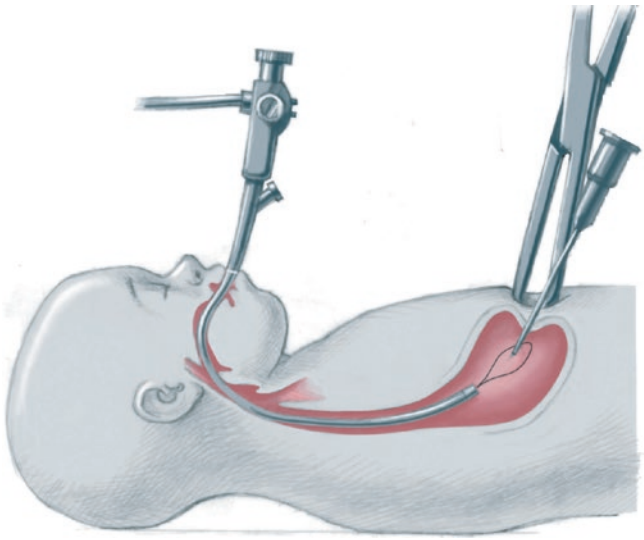

Fig. 23.9 A skin incision 8–10 mm in length is made. A fine-tipped, curved hemostat is applied to maintain the intragastric indentation. The gastroscope is moved gently in small increments. The endoscopist then deploys the polypectomy snare around (or over) this "mound"

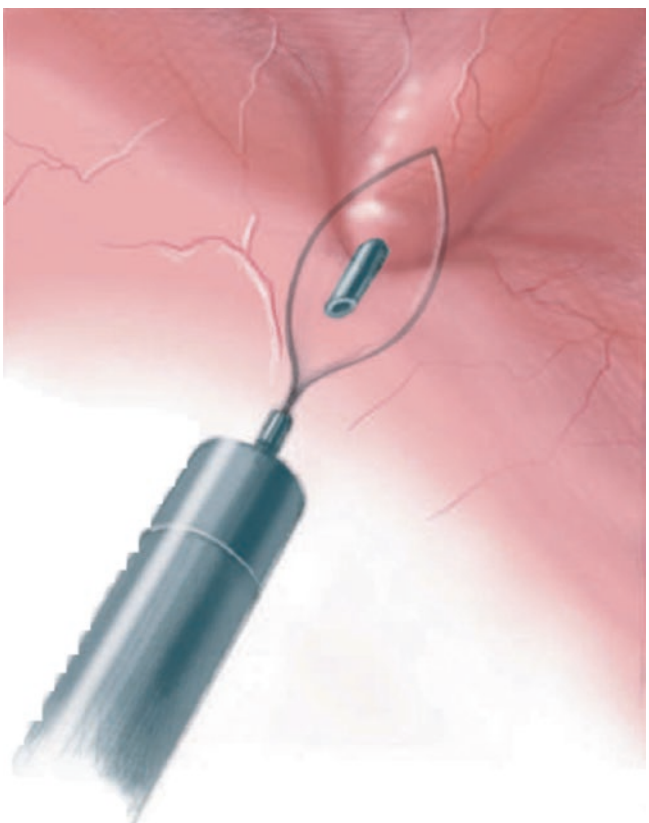

Fig. 23.10 The intravenous needle-cannula, which will allow the placement of the guidewire, is placed in the incision between the slightly spread prongs of the hemostat. The needle-cannula is then firmly thrust through the abdominal and gastric walls, exiting through the tip of the mound into the loop of the polypectomy snare. The snare is partially closed, but is not tightened around the cannula

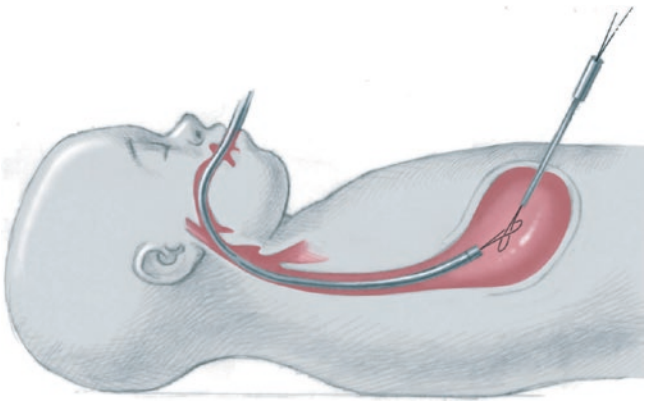

Fig. 23.11 The needle is removed and the plastic-coated, looped steel guidewire is inserted through the cannula. The polypectomy snare is allowed to slide away from the cannula and is tightened around the guidewire. (An alternative method is to retrieve the wire with an endoscopic alligator or biopsy forceps)

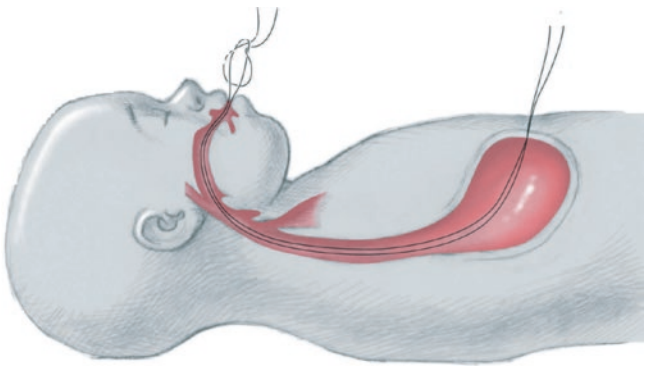

Fig. 23.12 The guidewire is then pulled back, along with the endoscope, from the stomach, through the esophagus, exiting from the patient's mouth. The guiding tract is thus established

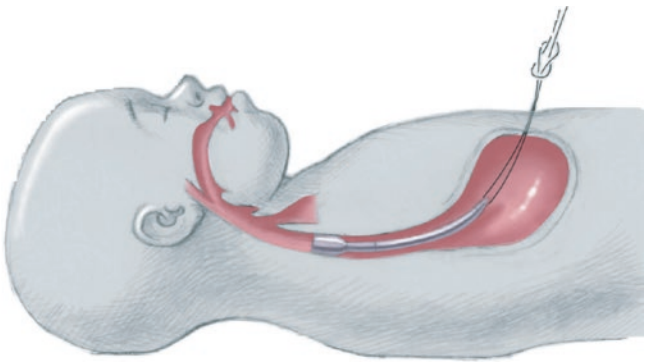

Fig. 23.13 The catheter is attached to the guidewire by interlocking the two steel wire loops and is lubricated. Traction is applied to the abdominal end of the guidewire, guiding the catheter through the oesophagus and stomach and across the gastric and abdominal walls. With an age-appropriate catheter, the intragastric retainer (dome) collapses enough to slide through the oesophagus without injury. For purposes of this diagram, a shortened catheter is shown. The commercially available catheters are long enough to permit the tapered end of the tube to exit through the abdominal wall before the gastric retainer enters the patient's mouth, allowing complete control of the catheter during placement

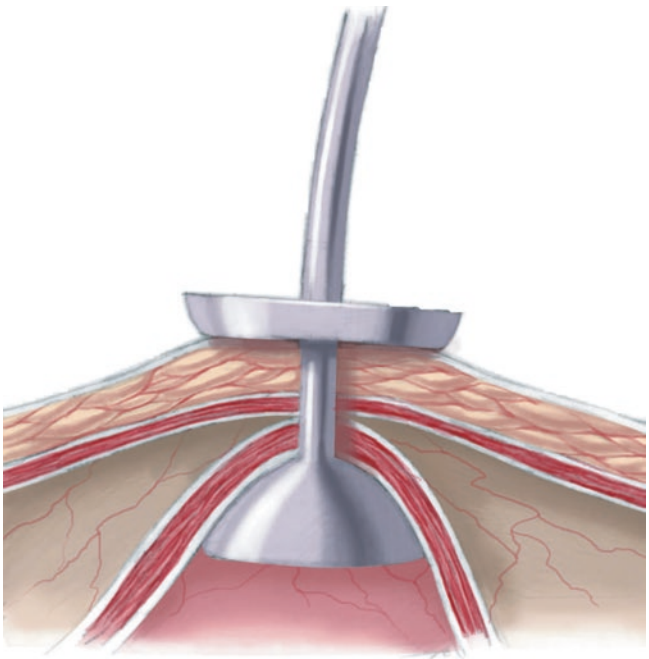

Fig. 23.14 Traction is continued until the gastric and abdominal walls are in loose contact. The markings on the catheter shaft help in judging the correct distance from the mucosa to the skin. The retaining external cross-bar or disk is advanced only enough to produce a good approximation of the gastric serosa to the abdominal wall peritoneum. Excessive approximation can lead to ischemia with tissue necrosis and embedding of the retainers. The catheter is cut to the desired length, and the feeding adapter is attached. No sutures are used, and the catheter is connected to a small, clear plastic sputum specimen trap. A dry gauze pad and tape are applied without kinking the tube

23.4.3 Laparoscopically Aided Gastrostomies

Laparoscopically aided gastrostomies are performed under general endotracheal anesthesia. Several approaches (and variations) for establishing the stoma have been developed. In addition to the videoscopically controlled "pull" PEG, commonly employed methods are based on adaptations of the "push" PEG using the Seldinger technique and U-stitches, and a "minimally open" Stamm technique.

23.4.3.1 Seldinger Method Plus U-Stitches

The most suitable gastrostomy site is selected in the left upper quadrant, following the previously described guidelines, and is marked. A nasogastric tube is introduced. A small transverse incision is made in the lower umbilical fold. Pneumoperitoneum is established in the usual, age-appropriate manner, and a 5-mm laparoscope is introduced. The left epigastrium is inspected. The needle used for the infiltration of the local anesthetic is then pushed through the abdominal wall and the appropriate relation between the anterior gastric wall and the future stoma is established. A 6- to 8-mm skin incision is made, and a 5-mm trocar is inserted. A bowel grasper is introduced, and the stoma site on the anterior gastric wall is lifted toward the parietal peritoneum. (Alternatively, instead of placing a trocar through the gastrostomy site, a small right-lower-quadrant incision is made under direct visualization and a 3-mm bowel grasper is inserted. The instrument grasps the gastric wall at the stoma site and lifts it against the parietal peritoneum). A U-stitch of a 2-0 monofilament suture is passed through the abdominal wall, through the anterior gastric wall, and back out through the abdominal wall. A second U-stitch is passed parallel to the first one, 1.5–2 cm apart, forming a square. Special care is taken to avoid including the posterior gastric wall. The sutures are lifted, keeping the stomach in contact with the abdominal wall. The grasper and the trocar are removed.

Figures 23.15 and 23.16 depict the use of the Seldinger-type guidewire and U-stitches to place a balloon-type "button."

To avoid tissue necrosis, the guy sutures should not be tied too tightly. They can be removed after 7–10 days. Although earlier removal is possible, it is probably not safe because the balloon may inadvertently deflate or rupture, in which case the stomach would separate from the abdominal wall.

Because the U-stitches must be removed, an alternative method (not depicted), is to place subcutaneous absorbable sutures. This technique requires two small stab incisions in addition to the umbilical site for the scope and the gastrostomy-site incision. Three or four absorbable sutures hold the stomach up, instead of the large U-stitches. Atraumatic needles and sutures are introduced into the peritoneal cavity through the gastrostomy site (after the incision has been made). The small needles are grasped by a laparoscopic needle holder. The stitches are placed through the anterior gastric wall around the future gastrotomy site. The needle then exits through the same gastrostomy skin incision. When lifted, these sutures bring the anterior gastric wall against the peritoneum. The Seldinger-guided balloon gastrostomy "button" is placed following the above-described method. After it is placed and inflated, the sutures are tied, resting above the fascia, next to the tube shaft, below the skin. The umbilical opening is closed with sutures, and the other two stab wounds, with adhesive strips.

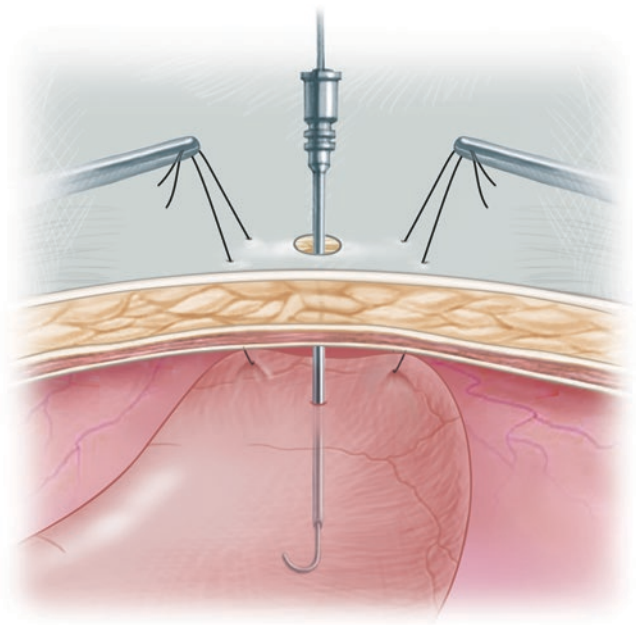

Fig. 23.15 The stomach is insufflated with air through the nasogastric tube, and a needle is inserted through the trocar site into the gastric lumen, between the two U-stitches. A Seldinger-type guidewire is passed through the needle into the stomach. The tract is sequentially dilated over the guidewire with tapered dilators (provided with the insertion kit) to the size required to insert either a Foley-type catheter or a balloon-type skin-level device. These are placed over the same guidewire. Stiffening of the catheter shaft with a thin dilator can be helpful during the insertion

23.4.3.2 Minimally Open Stamm Technique

This "hybrid" procedure is simple and does not require advanced laparoscopic skills. However, it may not be suited for older children with a thicker abdominal wall.

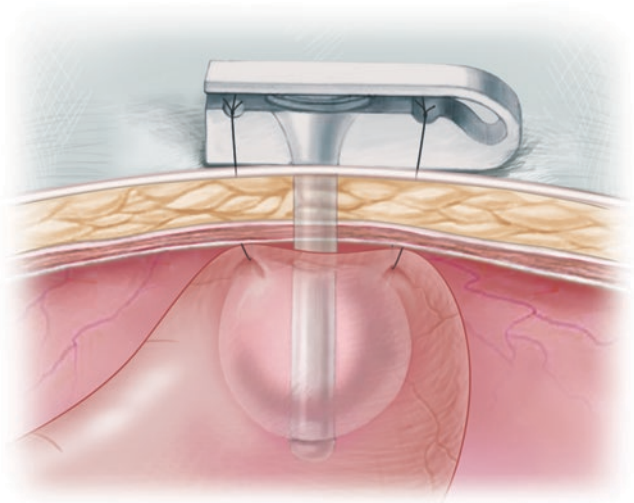

Fig. 23.16 A balloon-type "button" has been introduced and inflated under direct visualization to ensure adequate placement. The dilator and Seldinger wire are removed. The previously placed guy U-stitches are tied over the wings of the "button," securing the gastric wall to the parietal peritoneum and the device to the abdominal wall. Pneumoperitoneum is discontinued and the instruments are removed. The trocar site at the umbilicus and, if employed, the auxiliary incision, are closed with absorbable sutures

23.5 Postoperative Care

Enteral feedings are begun following an open gastrostomy once the ileus has resolved, and on the day after the operation for the minimally invasive procedures. The dressing is removed after 24 h, the wound is examined, and, if external immobilizers have been used, their tension is adjusted in order to avoid excessive pressure that could lead to tissue damage. Thereafter the stoma is left uncovered. We avoid harsh antiseptic solutions; after a few days, we simply use soap and water for cleaning. Granulation tissue tends to form after a couple of weeks and is controlled with gentle applications of silver nitrate. If granulation tissue becomes excessive, it leads to leakage and must be excised. The application of a steroid plus antifungal combination cream may help prevent recurrence. Once the epithelialization of the tract begins, no medication should be used.

23.6 Complications

Although generally considered a basic procedure, gastrostomy is associated with a long list of complications related to technique, care, and catheter use. Serious technique-related problems include separation of the stomach from the abdominal wall (leading to peritonitis), wound separation, hemorrhage, infection, injury to the posterior gastric wall or other organs, and placement of the tube in an inappropriate gastric position. Separation of the stomach from the abdominal wall is usually due to inadvertent, premature dislodgement of the tube, particularly with balloon-type devices, or a disruption during a catheter change. It requires immediate attention. It is generally managed with a laparotomy, although in select cases a laparoscopic correction is possible. Most complications can be avoided by carefully choosing the procedure and stoma device, considering it a major intervention and using meticulous technique, reliably approximating the stomach to the abdominal wall, exiting the catheter through a counter incision (in "open" procedures), and avoiding tubes in the midline or too close to the rib cage.

Among the most serious long-term problems are the so-called buried bumper syndrome (or external catheter migration) and severe gastrostomy leakage. The first complication becomes apparent when there is difficulty with the administration of feedings, which may also be associated with pain. This mishap can be avoided by always allowing (and checking for) sufficient "play" between the skin-level device or external bumper and the skin. Severe leakage is initially managed using conservative measures. If these fail, the stoma should be relocated.

Should a long-standing gastrostomy no longer be needed, the gastric access device is simply removed. Spontaneous closure usually follows. If the tract is well established, lined with skin and mucosa, this gastrocutaneous fistula may continue to drain. Simple extraperitoneal excision of the tract, with a few sutures in the fascia, subcutaneous layer, and skin, should suffice to close the communication.

23.7 Follow-Up

All children with gastrostomies should have careful follow-up to prevent and manage long-term catheter-related complications and should be monitored for manifestations of foregut dysmotility, particularly gastroesophageal reflux.

Suggested Reading

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Malrotation

24

Augusto Zani and Agostino Pierro

Malrotation is a congenital abnormal positioning of the midgut. Intestinal development is traditionally described as a process of elongation, rotation, and fixation. The process begins in the fifth week of gestation. Elongation of the bowel exceeds abdominal cavity expansion and the bowel herniates from the abdomen. As the bowel returns to the abdomen, it rotates 270° anticlockwise around the superior mesenteric artery (SMA). Rotation is completed by week 10 of gestation, with the SMA contained within a broad mesenteric base attachment. The distal duodenum comes to lie across the midline towards the left upper quadrant, attached to the posterior abdominal wall by the ligament of Treitz at the duodeno-jejunal (D-J) flexure. The caecum passes to the right and downwards and becomes fixed to the posterior abdominal wall. This latter process may be incomplete at birth, giving rise to a "high" caecum, a variant of normal in the neonate.

Three features of malrotation are the most common:

- The D-J flexure lies right of midline.
- The dorsal mesenteric attachment is narrow.
- Peritoneal folds (Ladd's bands) cross from the colon and caecum to the duodenum, liver, and gallbladder, possibly obstructing the duodenum.

Whether Ladd's bands are substantial enough to cause mechanical obstruction is debatable. The narrowed mesenteric base can lead to midgut volvulus, bowel obstruction, and mesenteric vessel occlusion. Antenatal volvulus can result in bowel atresia.

24.1 Presentation and Diagnosis

Malrotation is estimated from autopsy studies to occur in 0.5–1% of the population, although only 1 in 6000 live births will present with clinical symptoms. Incidence is slightly higher in males than females. Symptoms are seen in the first month of life in 50–75% of patients, and 90% will present before 1 year of age, but presentation can occur at any age. Malrotation is present in patients with gastroschisis, exomphalos, and congenital diaphragmatic hernia. Coexistent congenital anomalies (cardiac anomalies, bowel atresia, duodenal web, anorectal anomalies, orthopaedic anomalies) are common and affect 50% of children with malrotation. Malrotation is also associated with situs inversus, asplenia, and polysplenic syndromes.

Acute bowel obstruction due to Ladd's bands or intermittent midgut volvulus can present with vomiting, typically bilious, as the commonest presenting feature, accompanied by colicky abdominal pain and abdominal distention. Abdominal tenderness and blood per rectum in an infant is suggestive of bowel ischaemia, which could be due to midgut volvulus. Older children without acute volvulus more often present with chronic episodic obstructive symptoms, failure to thrive, malabsorption, diarrhoea, and nonspecific colicky abdominal pain. Up to 10% of diagnoses of malrotation are made as an incidental finding.

Plain abdominal radiograph is often normal; features suggestive of malrotation with or without midgut volvulus are a distended stomach and proximal duodenum with a paucity of gas distally, either throughout or unilaterally. An upper gastrointestinal contrast study is the investigation of choice for any child presenting with bilious vomiting. The study should be performed urgently to look for findings characteristic of malrotation:

- The D-J flexure is seen right of the left vertebral pedicle and/or inferior to the pylorus.
- The duodenum passes caudally and anteriorly.

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- Contrast tapering or a "corkscrew" appearance suggests obstruction and/or volvulus.

Caecal position is highly variable and may be normal in up to 15% of cases of malrotation, so contrast enema is not always helpful.

Abdominal ultrasound may show reversal in the relationship of the SMA to the superior mesenteric vein (SMV). In a normal situation, the SMV is located to the right of the SMA; an SMV to the left of the artery is suggestive of malrotation. Computer tomography is not indicated in infants and children.

24.2 Surgical Management

All symptomatic patients with positive investigative findings should undergo urgent laparotomy. Management of the asymptomatic patient is more controversial. The risk of bowel ischaemia due to midgut volvulus is invariably present, and most surgeons would proceed to prompt operation.

Whether to look for and/or correct rotation anomalies is controversial in:

- Patients undergoing repair of abdominal wall defects or diaphragmatic hernias, who invariably have some degree of malrotation but rarely have subsequent volvulus
- Asymptomatic patients with malrotation in whom the diagnosis is made incidentally during evaluation for non-specific complaints, prior to fundoplication for gastro-oesophageal reflux, and in those with heterotaxy syndromes
- Monozygotic twins with proven malrotation in the sibling.

24.2.1 Laparotomy

The principles of the procedure have remained almost unchanged since it was originally described by Ladd in 1936. After a right upper quadrant transverse incision is made (Fig. 24.1), the umbilical vein is divided and ligated. The peritoneal fluid is examined. Frequently it is clear; blood-stained fluid implies bowel ischaemia and volvulus; faecal staining indicates bowel perforation.

The midgut is delivered from the wound and examined. Any volvulus should be derotated, noting the number of turns (Fig. 24.2). The bowel is examined for viability and any ischaemic bowel should be wrapped in a damp swab and re-examined after 5–10 min. Nonviable bowel is resected and a primary anastomosis is formed. If extensive ischaemic bowel of doubtful viability is present, a second-look laparotomy is performed after 24 h, with the aim of minimizing the extent of bowel resection required.

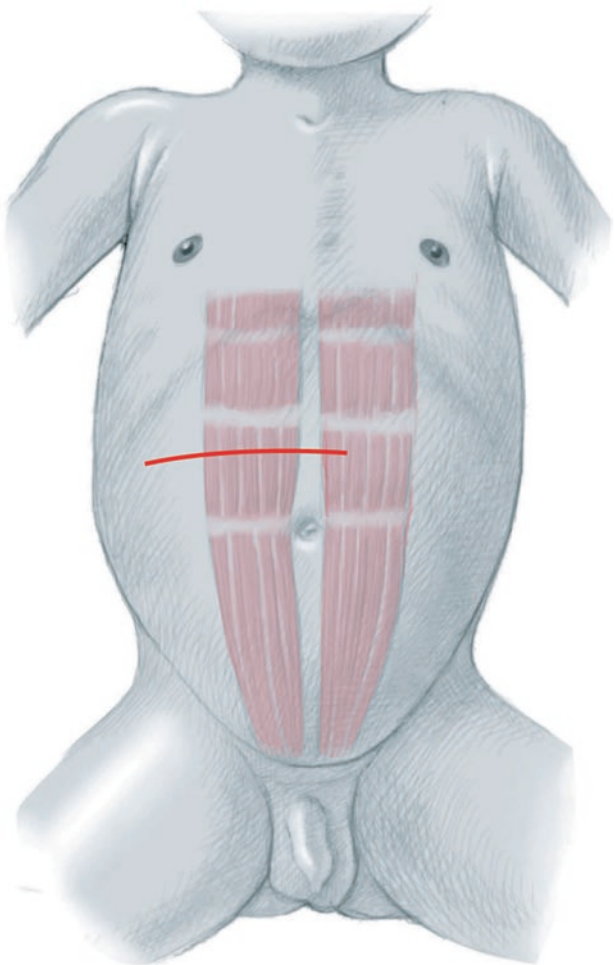

Fig. 24.1 The patient is positioned supine, legs extended. A right upper quadrant transverse incision is made

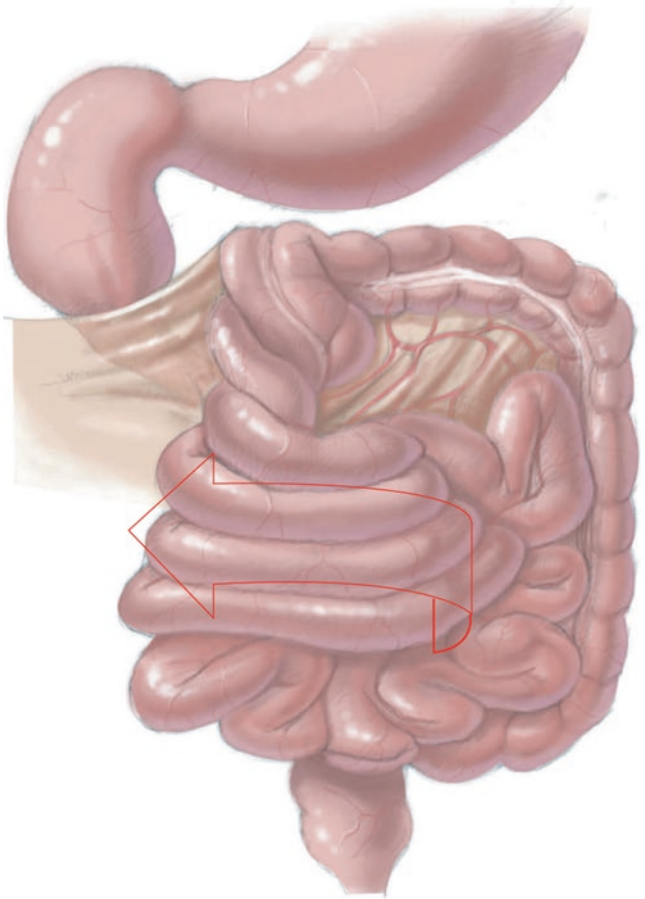

Fig. 24.2 The midgut is delivered from the wound and the base examined. Any volvulus should be derotated anticlockwise (*arrow*), noting the number of turns

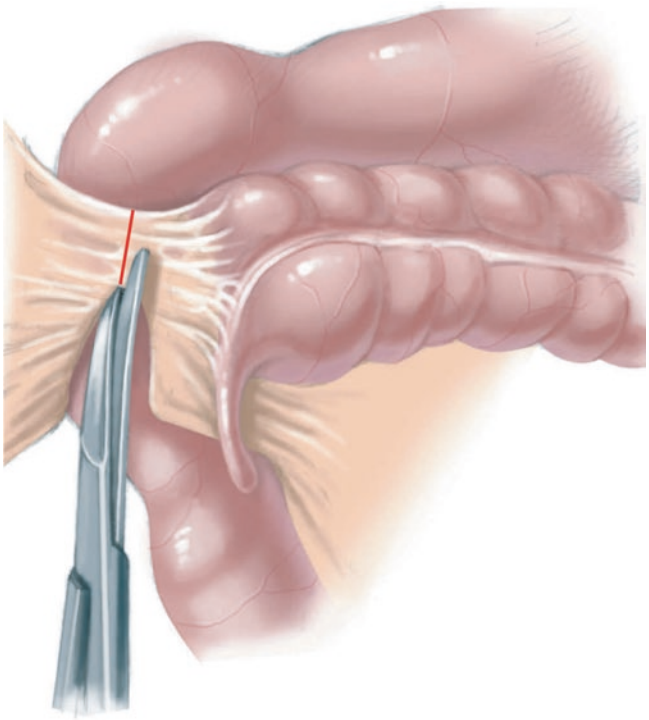

Fig. 24.3 Ladd's bands, if present, are divided

A novel technique to deal with mesenteric thrombosis was recently described. This technique includes digital massage of the superior mesenteric vessels after derotation to restore intestinal perfusion, followed by postoperative systemic infusion of tissue-type plasminogen activator.

Ladd's bands, if present, are divided (Fig. 24.3). The SMA is identified and the mesenteric base is broadened as much as possible by division of the peritoneal folds (Fig. 24.4). At the end of the procedure, the small bowel is placed in the right hemi-abdomen and the large bowel in the left hemi-abdomen. There is no need to apply any fixation sutures, as adhesions and the broad base to the mesentery developed by the Ladd's procedure usually stabilize the bowel. The abdomen is closed per routine.

The abnormal position of the appendix may cause diagnostic problems in future, so removal has been advocated. However, whether an appendectomy should be performed in neonates is subject to debate. Some surgeons opt for an inversion of the appendix; others leave the appendix untouched to prevent potential additional morbidity.

The nasogastric (NG) tube is aspirated hourly for the first 24 h. Intravenous fluids are continued postoperatively and NG tube fluid loss is replaced, millilitre for millilitre, with normal saline and potassium chloride (20 mmol/L saline). Enteral feeds are restarted when aspirates are clear and reducing in volume, usually after 24 h.

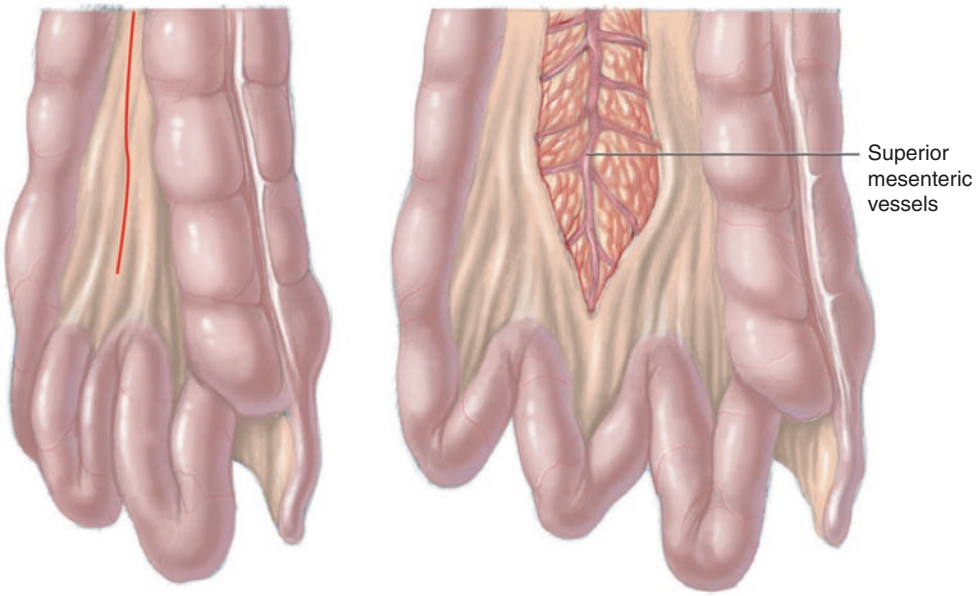

Fig. 24.4 The superior mesenteric artery is identified and the mesenteric base is broadened as much as possible by division of the peritoneal folds, taking care not to injure the superior mesenteric vessels

24.2.2 Laparoscopy

Laparoscopy can be used in nonacute cases of malrotation without volvulus (incidentally diagnosed malrotation). Three ports are placed into the abdominal cavity (Fig. 24.5), for the laparoscope and for nontraumatic grasping forceps that will be used to manipulate the bowel. The anatomy is defined and Ladd's bands identified. The duodenum is exposed and Ladd's bands are divided (Fig. 24.6). After division, the bowel is examined along its length for any further causes of obstruction. The root of the mesentery is broadened by dividing the peritoneal folds. Care must be taken not to injure the major vessels. Appendectomy can be carried out, either by using an endoloop for intracorporeal ligation or by delivering the appendix through a trocar site and excising it extra-abdominally in smaller patients. The trocar sites are then closed.

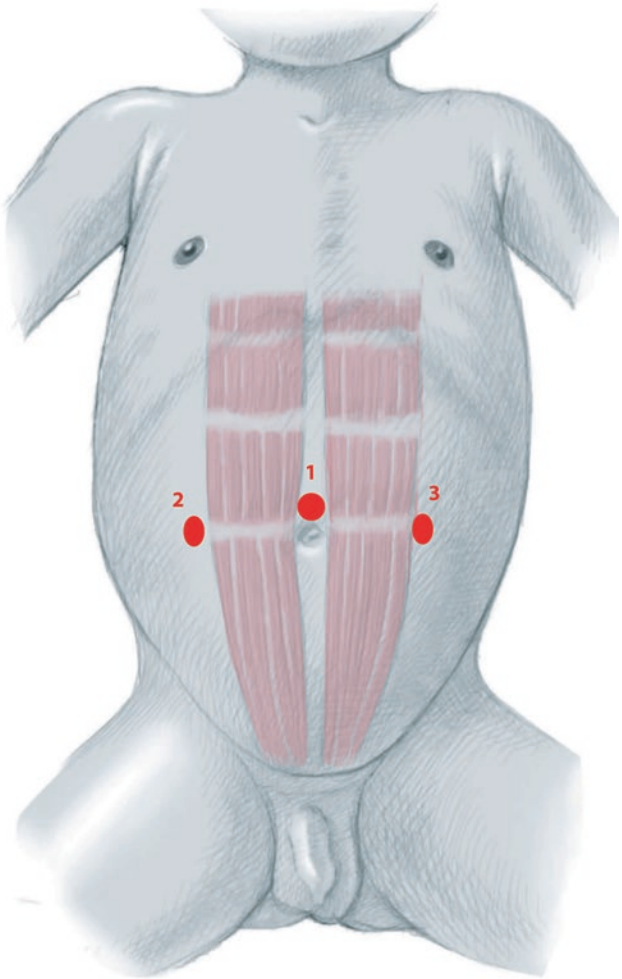

Fig. 24.5 For laparoscopic repair of malrotation, the patient is positioned supine. A 5- or 10-mm port is placed into the abdominal cavity under direct vision. Carbon dioxide is insufflated via the port. The final intra-abdominal pressure should be 8–10 mmHg in an infant, or 10–12 mmHg in an older child. The flow rate of carbon dioxide is set between 0.5 and 1.5 L/min. The laparoscope is inserted into this port. Two further 5-mm ports are placed under direct camera vision in the left lower quadrant and right lower quadrant. Nontraumatic grasping forceps are inserted into these ports to manipulate the bowel

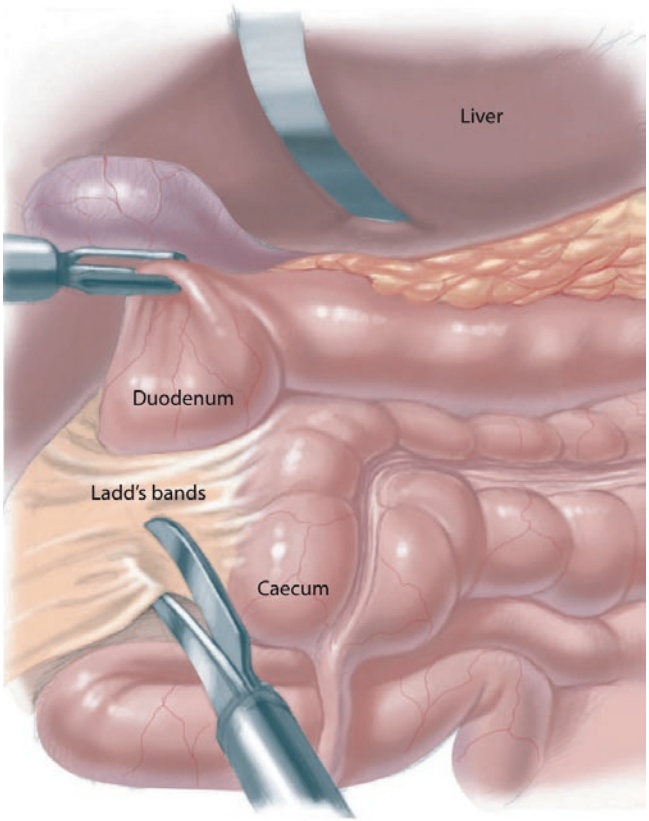

Fig. 24.6 The anatomy is defined and Ladd's bands identified. Care must be taken to correctly identify landmarks such as the duodenum and ascending colon. To gain access to the duodenum, it is useful to raise the head of the operating table and elevate the right flank. The ascending colon falls towards the left side of the abdomen. The duodenum is exposed and Ladd's bands are divided using a combination of sharp dissection and electrocautery

24.3 Outcomes

The outcome of patients undergoing Ladd's procedure for isolated malrotation is very good; most make a full recovery. The commonest postoperative complication is adhesional obstruction (3–5%). Midgut volvulus occurs in 45–65% of children with malrotation and still carries a mortality rate of 7–15%, and necrosis of more than 75% of the midgut is associated with short bowel syndrome. Up to 18% of children with short bowel syndrome on long-term total parenteral nutrition have an original diagnosis of midgut volvulus.

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Duodenal Obstruction

25

Yechiel Sweed and Arcady Vachyan

Congenital duodenal obstruction is the result of several embryologic defects in foregut development, canalization, or rotation. In addition, abnormal embryologic relationships between the duodenum and other structures in close anatomic proximity, such as the pancreas and portal vein, may also lead to congenital duodenal obstruction. Ladd classified these lesions as either intrinsic or extrinsic. Intrinsic lesions include duodenal atresia, duodenal stenosis, or duodenal web, whereas annular pancreas, malrotation, peritoneal bands, and anterior portal vein are classified as extrinsic.

The incidence of duodenal obstruction is reported to be 1 in 5000–10,000 births. In patients with intrinsic duodenal obstruction, there is a high incidence of associated anomalies, especially Down syndrome, which occurs in about 30% of these patients. Other associated anomalies include congenital heart disease, malrotation, annular pancreas, oesophageal atresia, urinary tract malformations, anorectal anomalies, and other bowel atresias.

25.1 Diagnosis

The diagnosis of duodenal obstruction may be suspected prior to the child's birth from prenatal ultrasonography, which may identify maternal polyhydramnios and demonstrate distension of the stomach and the first portion of the duodenum with swallowed amniotic fluid. Although prenatal ultrasonography is an accurate predictor of duodenal obstruction and allows preparation of parents, physicians, and institutions for the anticipated arrival of the patient needing prompt care at birth, it has neither influenced the incidence of associated life-threatening anomalies nor changed the survival rate.

The clinical presentation of duodenal obstruction is usually characterized by feeding intolerance and by onset of vomiting in the first 24–48 h of life. Because 80% of obstructions are located in the postampullary region of the duodenum, the vomitus in most cases is bile-stained. A careful physical evaluation for associated anomalies is performed. Cardiac and renal ultrasonographic examinations are also indicated because of the high incidence of associated malformations in other organ systems.

Diagnosis is achieved in most cases by plain abdominal radiographs, which demonstrate dilated stomach and duodenum, giving the characteristic appearance of a "double-bubble" sign. No gas is observed beyond the second bubble in instances of complete obstruction. In this setting, the plain film is sufficiently diagnostic, and no further imaging of the gastrointestinal tract is necessary. In partial duodenal obstruction, a plain film of the abdomen also will show a "double-bubble" appearance, but there is usually some air in the more distal intestine. Early upper gastrointestinal contrast radiography is indicated in these patients in order to establish the cause of incomplete duodenal obstruction.

Incomplete duodenal obstruction may lead to delayed onset of symptoms, and the diagnosis of duodenal diaphragm with a central aperture is sometimes delayed for months or years.

25.2 Preoperative Management

Although duodenal atresia is a relative emergency, the infant should not be rushed to the operating room until his or her haemodynamic and fluid and electrolyte status is stable. If the clinical history and findings on physical examination indicate that the baby is in no distress, and the radiograph is consistent with the usual presentation of duodenal atresia with no air beyond the second bubble, surgery should be per-

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formed within the first 2 days of life. Immediate surgical exploration should be performed, however, in patients with duodenal obstruction caused by malrotation resulting in extrinsic compression related to Ladd's bands across the duodenum or acute volvulus of the midgut.

Preoperative management consists of nasogastric decompression and fluid and electrolyte replacement. Care is taken to preserve body heat and avoid hypoglycaemia, as most of these newborn patients are premature or small for date. Preoperative systemic antibiotics are administered at least 30 min before the start of the operation.

25.3 Surgical Management

For most causes of congenital duodenal obstruction, duodenoduodenostomy via an open approach is the traditional surgical procedure. Over the past decade, however, the application of minimally invasive surgical (MIS) techniques and the advent of smaller laparoscopic instruments have expanded the potential of laparoscopy for repair of this obstruction.

25.3.1 Open Approach to Duodenal Obstruction

The baby is placed supine on the table, on a warming blanket, with a small roll under the upper abdomen. Endotracheal anaesthesia is used. A nasogastric tube is passed to decompress the stomach. An intravenous infusion is set up. The abdominal skin is prepared by cleaning with prewarmed povidone iodine.

A transverse supraumbilical abdominal incision is made 2 cm above the umbilicus, starting in the midline and extending laterally into the right upper quadrant (Fig. 25.1). A small incision is made in the posterior fascia and peritoneum after these are drawn up with forceps. To enlarge this initial incision, two fingers are inserted and the fascia and peritoneum are cut along the length of the wound. The underlying structures are retracted.

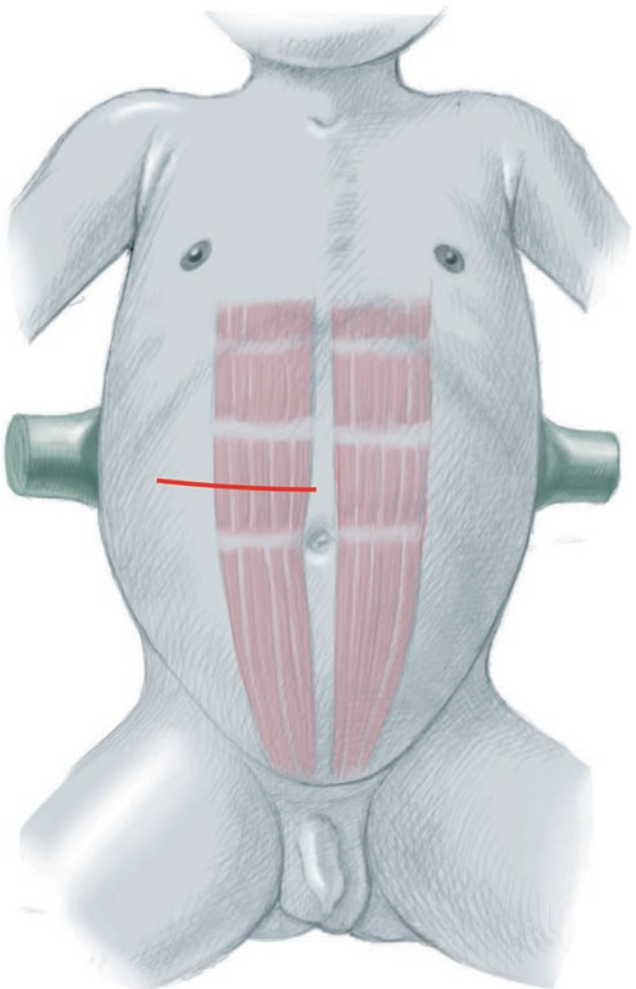

Fig. 25.1 The baby is placed supine on the table, on a warming blanket, with a small roll under the upper abdomen. A transverse supraumbilical abdominal incision is made 2 cm above the umbilicus, starting in the midline and extending laterally into the right upper quadrant

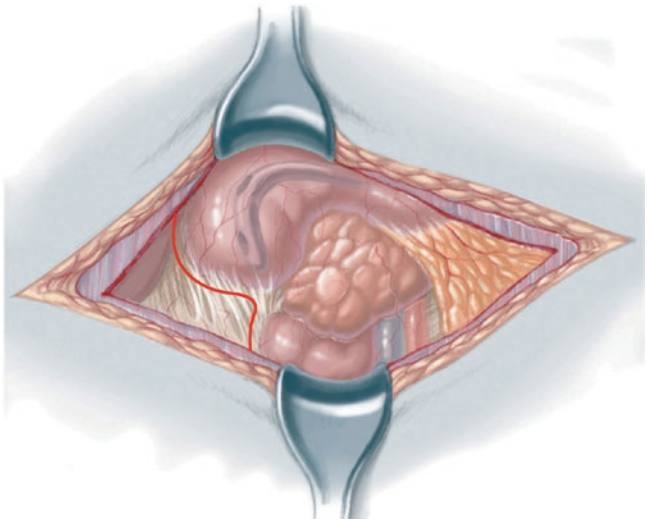

Fig. 25.2 After exposing the peritoneal cavity, the surgeon inspects the entire bowel for the presence of other anomalies. The ascending colon and the hepatic flexure of the colon are mobilized medially and downwards to expose the dilated duodenum. The duodenum is then adequately mobilized and freed from its retroperitoneal attachments (Kocher manoeuvre) (*red line*). Great care must be exercised not to dissect or manipulate either segment of the duodenum medially, to avoid injury to the ampulla of Vater or the common bile duct. The duodenum distal to the site of obstruction is small and decompressed. The requirements for distal mobilization vary according to the location of the atresia and to the gap between the two segments. If necessary, the ligament of Treitz is divided, and mobilization and displacement of the distal duodenum is performed behind the superior mesenteric vessels, thus allowing a satisfactory anastomosis to be performed without any tension

After exposing the peritoneal cavity (Fig. 25.2), the surgeon inspects the entire bowel for the presence of other anomalies. There may be an associated annular pancreas or malrotation in one third of the patients. If the colon is in a normal position, malrotation is probably not a coexisting factor.

The stomach and first portion of the duodenum are usually thickened and dilated. The liver is carefully retracted superiorly. The ascending colon and the hepatic flexure of the colon are mobilized medially and downwards to expose the dilated duodenum.

The duodenum is then adequately mobilized and freed from its retroperitoneal attachments (Kocher manoeuvre). Great care must be exercised not to dissect or manipulate either segment of the duodenum medially, to avoid injury to the ampulla of Vater or the common bile duct. The tube in the stomach is then passed distally into the dilated duodenum and helps to locate the point of obstruction and determine if a "windsock" abnormality is present.

The type of atresia as well as any pancreatic abnormality (annular pancreas) or the presence of a rare preduodenal portal vein are noted. In patients with an annular pancreas, the pancreatic tissue should never be divided and should be bypassed. The duodenum distal to the site of obstruction is small and decompressed. The requirements for distal mobilization vary according to the location of the atresia and to the gap between the two segments. If necessary, the ligament of Treitz is divided, and mobilization and displacement of the distal duodenum is performed behind the superior mesenteric vessels, thus allowing a satisfactory anastomosis to be performed without any tension.

Duodenoduodenostomy is the procedure of choice for patients with duodenal atresia, stenosis, and annular pancreas. Two surgical techniques—side-to-side duodenoduodenostomy or proximal transverse–to–distal longitudinal ("diamond-shaped") anastomosis—may be performed.

25.3.2 "Diamond-Shaped" Anastomosis

Diamond-shaped duodenoduodenostomy has been reported to allow earlier feeding, earlier discharge, and good long-term results. With two traction sutures, the redundant wall of the proximal duodenum is pulled downward to overlie the proximal portion of the distal duodenal segment (Fig. 25.3). A transverse incision in the distal end of the proximal duodenum and a longitudinal incision in the smaller limb of the duodenum distal to the occlusion are made so as to allow good approximation of the openings without tension (Fig. 25.4) and distal atresias are ruled out. The papilla of Vater is located by observing bile flow from gentle compression of the gall bladder.

A single-layer anastomosis is created using Vicryl sutures (Fig. 25.5). Before completion of the anastomosis, a transanastomotic feeding tube may be passed down into the upper jejunum for an early postoperative enteral feeding.

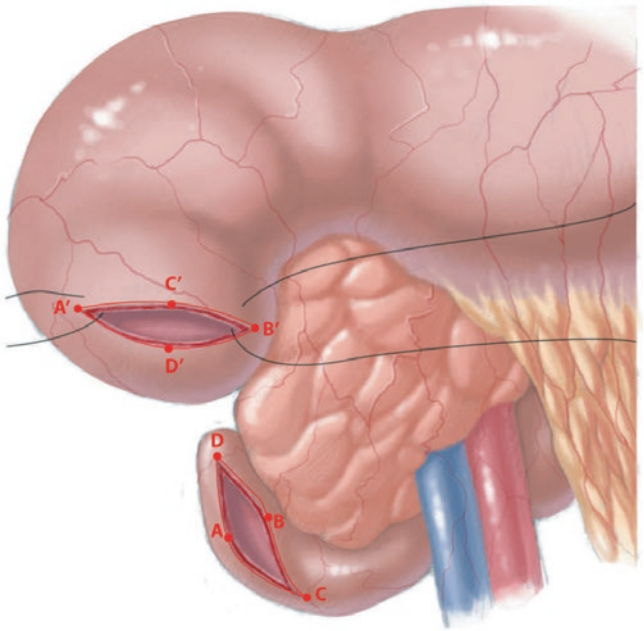

Fig. 25.3 To perform a diamond-shaped anastomosis, two traction sutures in the redundant wall of the proximal duodenum are used to pull it downward to overlie the proximal portion of the distal duodenal segment. A transverse incision is made in the distal end of the proximal duodenum and a longitudinal incision is made in the smaller limb of the duodenum distal to the occlusion. These are made in such a position as to allow good approximation of the openings without tension

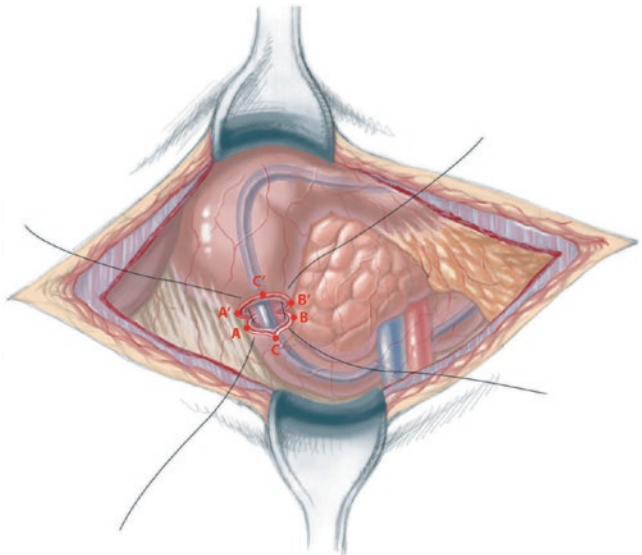

Fig. 25.4 The orientation of the sutures in the diamond-shaped anastomosis and the overlapping between the incisions. At this stage, a small Nélaton catheter is passed distally through the opening made in the distal segment, and 20–30 mL of warm saline is injected to rule out atresias distally. The catheter is then removed

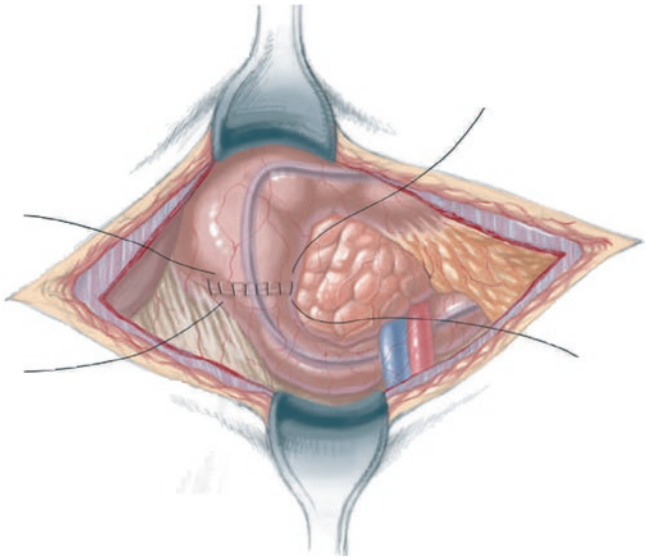

Fig. 25.5 A single-layer anastomosis using interrupted 5/0 or 6/0 Vicryl sutures with knots tied inside the posterior wall of the anastomosis and interrupted sutures with anterior knots tied outside the anterior wall. Before completion of the anterior part of the anastomosis, a transanastomotic feeding tube (5F silicone) may be passed down into the upper jejunum for an early postoperative enteral feeding

25.3.3 Repair of Duodenal Web

When abdominal exploration yields a diagnosis of duodenal web (identified by the advancement of the gastric tube into the proximal dilated duodenum), the dilated duodenal wall is incised (Fig. 25.6) and the duodenum is opened. The windsock duodenal web must be clearly identified because the visible transition from the distended proximal duodenum to the small downstream duodenum may be several centimetres distal to the base of the web. Traction applied at the apex of the web deforms the duodenum at its point of attachment and allows excision at the base (Fig. 25.7).

The duodenal membrane is usually localized in the second part of the duodenum, and occasionally in the third portion. It can be complete or may have a central hole. Anatomically, the ampulla of Vater may open directly into the medial portion of the web itself—anteriorly, posteriorly, or with dual openings into the membrane—or it may open close to it.

This close relationship mandates the identification of the papilla of Vater before excision of the web. The web is opened along the lateral side of the membrane and excision from the duodenal wall takes place (Fig. 25.8). An intermittent bile flow is usually seen via the papilla of Vater,

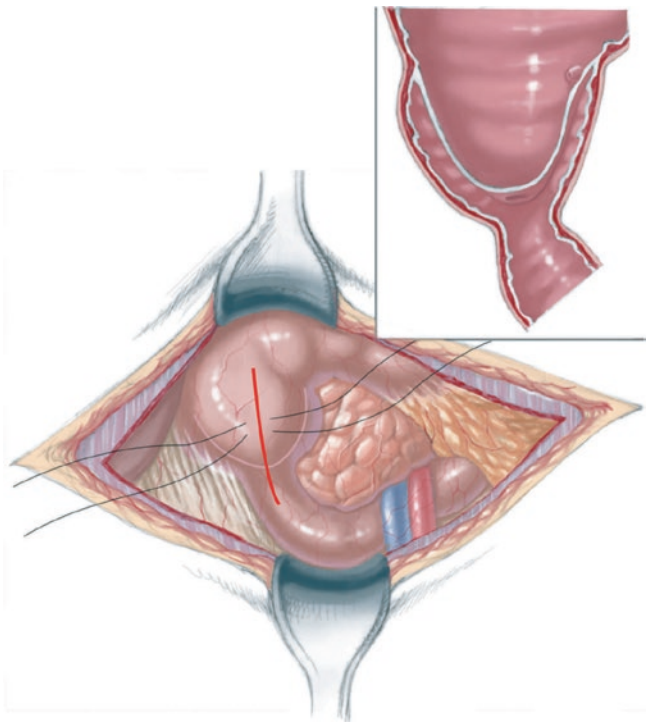

Fig. 25.6 To repair a duodenal web (identified by the advancement of the gastric tube into the proximal dilated duodenum), two stay sutures are placed at the anterior dilated duodenal wall. A longitudinal incision of 2.5–3 cm (*red line*) is performed above the "transitional zone" between the wide and the narrow segments of the duodenum, and the duodenum is opened

indicating to the surgeon the exact line of excision. The resection line is then oversewn and the duodenum is closed transversely (Fig. 25.9). Because of the pitfalls in cases of lax membrane that may bulge downwards distally into the distended duodenum (the so-called windsock phenomenon), and in order to avoid missing the anomaly, the patency of the distal duodenum must be identified by inserting a catheter through the duodenotomy before its closure.

Following completion of the web resection and closure of the duodenum, the abdominal cavity is irrigated with 50 mL of sterile warm saline. The wound is closed in layers: the peritoneum and the posterior fascia and the anterior fascia by two layers using continuous 4/0 Dexon or Vicryl sutures. The skin is closed with a running intracuticular suture using 5/0 Vicryl or Dexon suture.

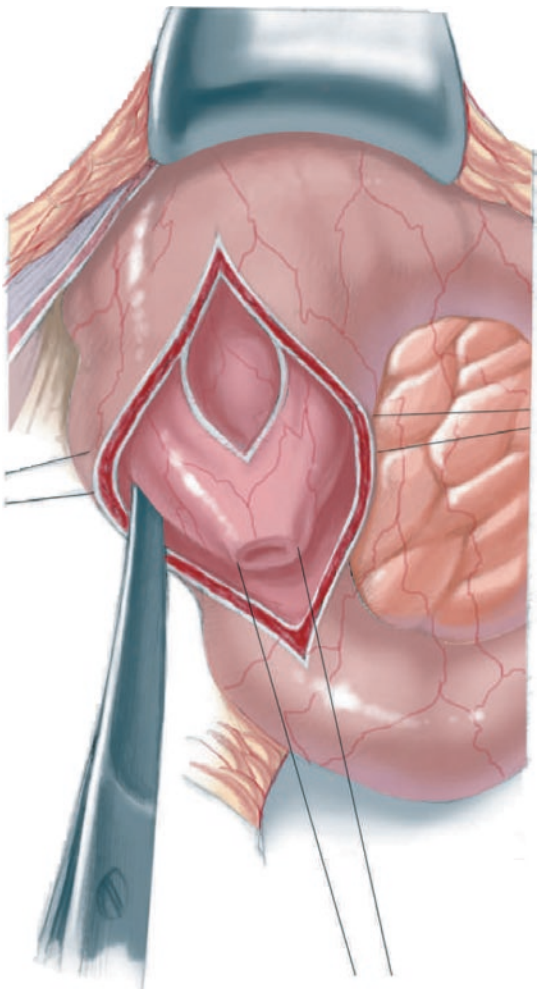

Fig. 25.7 Two other stay sutures are placed at the margins of the duodenal incision. Traction applied at the apex of the web deforms the duodenum at its point of attachment and allows excision at the base. A single 4/0 Vicryl stay suture is placed at the centre of the membrane. The web is opened along the lateral side of the membrane and excision from the duodenal wall takes place, leaving a rim of tissue of 2–3 mm

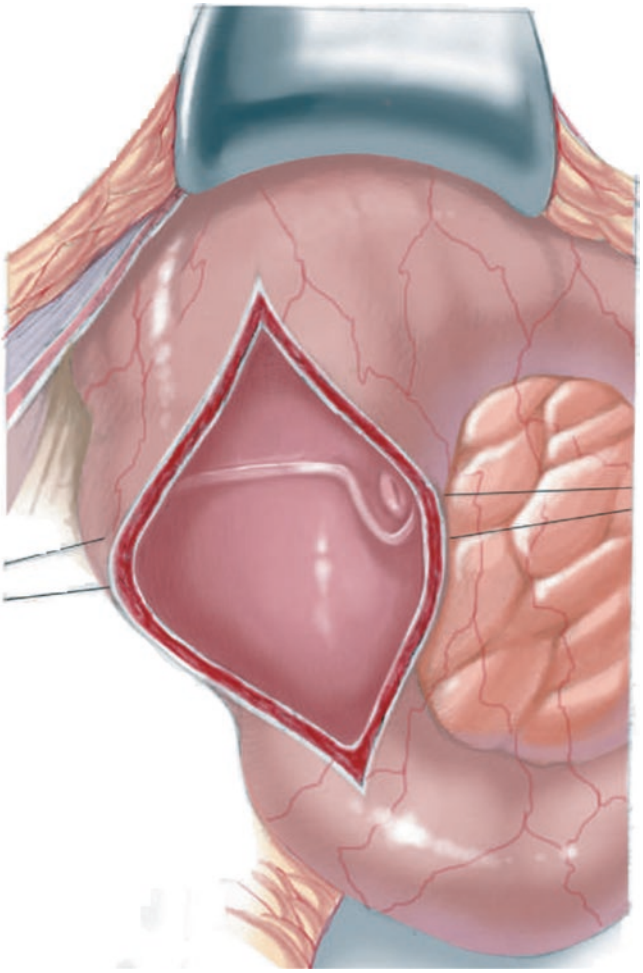

Fig. 25.8 The medial portion of the membrane should remain intact, to avoid damage to the ampulla of Vater

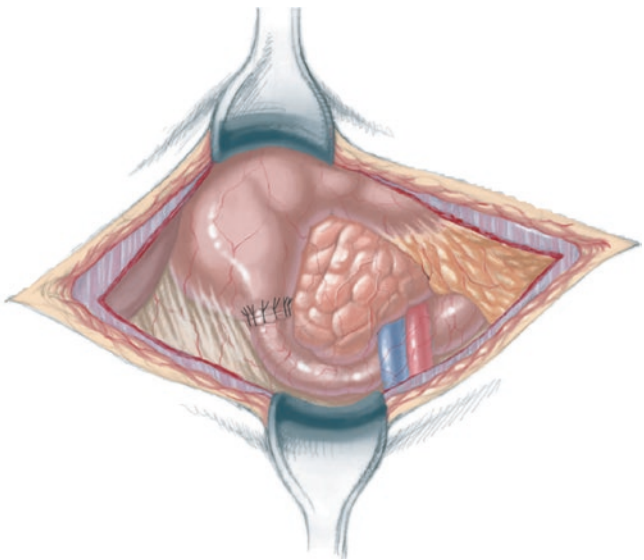

Fig. 25.9 The resection line is oversewn using interrupted 5–0 absorbable sutures. The duodenum is then closed transversely with interrupted sutures

A nasogastric tube is left in place for postoperative gastric drainage. A gastrostomy may be performed if the need is anticipated. Intravenous therapy and antibiotics are continued postoperatively. The patient is kept without oral intake until stool is passed and limited clear or pale-green gastric drainage is noted (<1 mL/kg per h). The commencement of oral feeding may be delayed for several days and occasionally for 2 weeks or more. Postoperatively, patients may have a prolonged period of bile-stained aspirate, mainly because of the inability of the markedly dilated duodenum to produce effective peristalsis. Many surgeons therefore use transanastomotic tubes for feeding in the early postoperative period.

25.3.4 Laparoscopic Management of Duodenal Obstruction

For the laparoscopic approach, neonatal (3-mm) laparoscopic instruments and trocars are used. The patient is positioned supine at the end of the operating table and the surgeon stands at the patient's feet. The abdomen is insufflated and then two additional trocars are inserted (Fig 25.10). A 3-mm grasping forceps for lifting the liver can be also introduced in the left upper quadrant without a trocar. A better view of the dilated duodenum can be also achieved by using a suture to lift up the falciform ligament. The suture is inserted through the abdominal wall in the right upper quadrant, lifts the ligament, and then is passed back through the abdominal wall and tied.

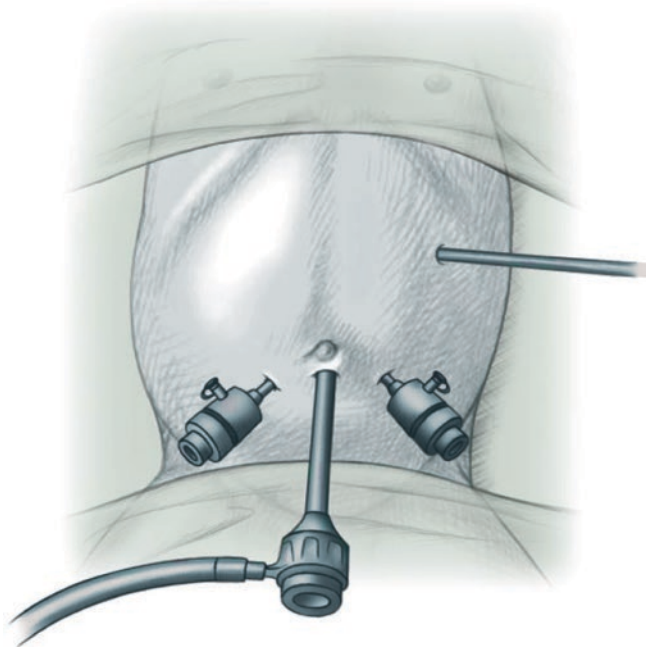

Fig. 25.10 For the laparoscopic approach, the abdomen is insufflated with CO_2 (6 mmHg, 2 L/min) through a 5-mm umbilical port for a 30° laparoscope. Then two additional 3-mm trocars are inserted under direct vision in the right lower quadrant and left middle abdomen. A 3-mm grasping forceps for lifting the liver can be also introduced in the left upper quadrant without a trocar

The first surgical step is to mobilize the colon and the duodenum. A stay suture is inserted through the abdominal wall to move the bulky part of the bulbus duodeni out of the way, allowing a view of the distal duodenum and a more convenient approach to the anastomosis. A transverse incision is made in the distal wall of the dilated duodenum (Fig. 25.11), followed by a longitudinal incision in the distal collapsed duodenum for the "diamond-shaped" anastomosis of Kimura. As with the open repair, stay sutures are placed at each corner to facilitate the anastomosis (Fig. 25.12).

Before suturing the anterior wall of the anastomosis, the anesthesiologist advances a Nélaton catheter, which is grabbed laparoscopically and inserted into the distal collapsed segment. Patency of the proximal small bowel loops is cleared by washing saline solution via the Nélaton catheter. Once the anastomosis is completed, the ports are removed and the sites are closed with absorbable sutures.

When a duodenal membrane is suspected, a longitudinal incision is made on the anterior wall of the duodenum, crossing from the distended duodenum to the distal collapsed duodenum (Fig. 25.13). A urinary catheter is inserted through the abdominal wall directly into the distal duodenal segment, the balloon is filled, and the catheter is gradually pulled back. A membrane with an aperture will stretch itself on top of the balloon. The membrane is incised carefully in its lateral aspect and the longitudinal incision is closed.

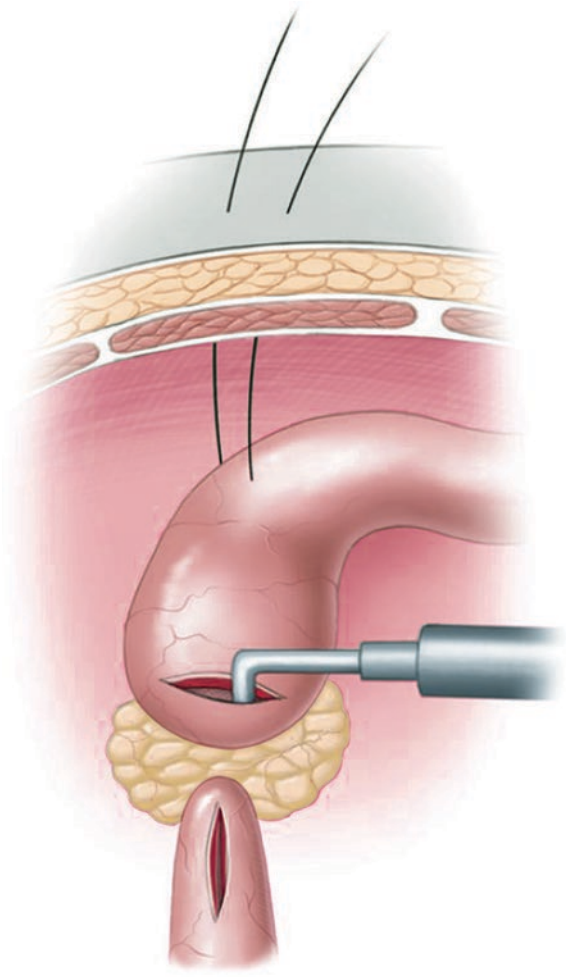

Fig. 25.11 The colon is mobilized to the left, and the duodenum is then adequately mobilized and freed from its retroperitoneal attachments (Kocher maneuver). A stay suture with a large needle is then inserted through the abdominal wall close to and below the right costal margin. The stay suture is introduced into the bulbus duodeni to move the bulky part of the bulbus out of the way, allowing a view of the distal duodenum and a more convenient approach to the anastomosis. A transverse incision using the hook is made in the distal wall of the dilated duodenum, followed by a longitudinal incision in the distal collapsed duodenum for the diamond-shaped anastomosis of Kimura

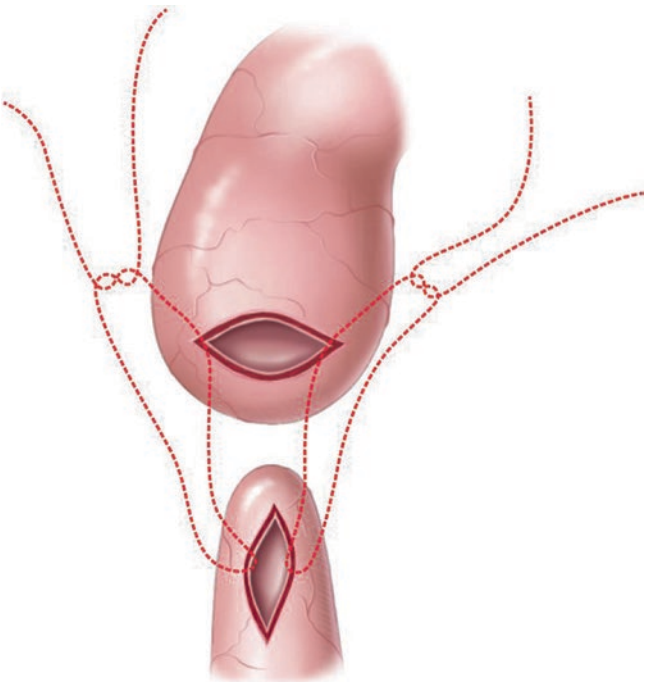

Fig. 25.12 Stay sutures are placed at each corner to facilitate the diamond-shaped anastomosis. The anastomosis is created with either a separate running suture for the posterior and then anterior walls, or single interrupted stitches of Vicryl. Intracorporeal knot tying is used

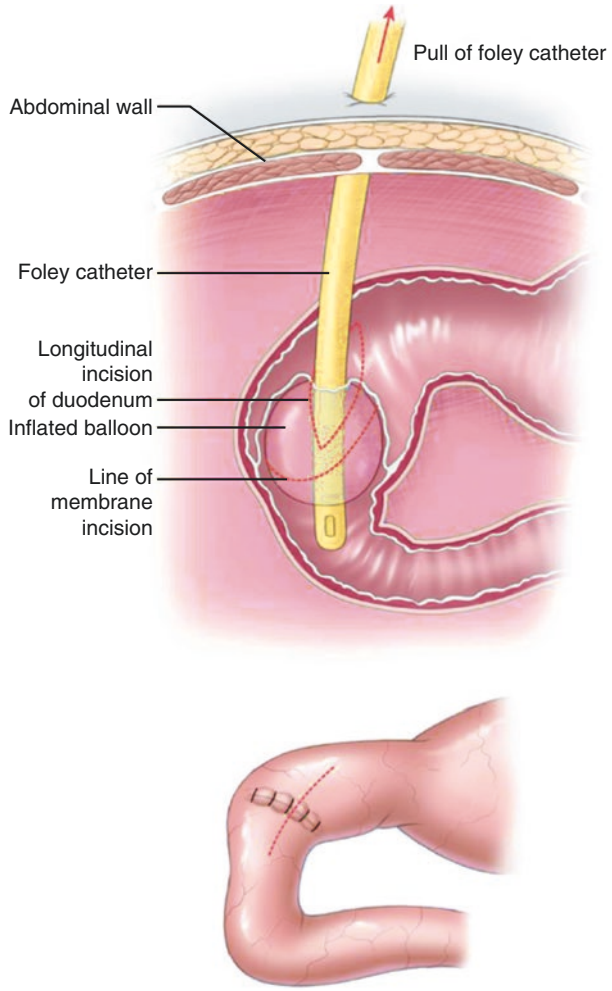

Fig. 25.13 When a duodenal membrane is suspected, a longitudinal incision is made on the anterior wall of the duodenum, crossing from the distended duodenum to the distal collapsed duodenum A urinary catheter is inserted through the abdominal wall directly into the distal duodenal segment without a trocar. The balloon is filled and the catheter is gradually pulled back. If a membrane with an aperture is present, it will stretch itself on top of the balloon. The membrane is incised carefully in its lateral aspect, leaving the medial part of the membrane intact, to prevent injury to the papilla Vater. The longitudinal incision is closed transversely with a running suture or several interrupted sutures

25.4 Outcome and Complications

Long-term outcome after repair of congenital duodenal obstruction is excellent, with contemporary operative survival exceeding 95% and with most patients reported to be asymptomatic, with normal growth.

The experience with laparoscopic duodenoduodenostomy demonstrates that it can be performed safely and successfully in the neonate with excellent short-term outcomes. The last retrospective studies comparing the surgical outcome of laparoscopic repair versus open repair found that laparoscopy is a safe and effective technique. The results, including operative time, length of stay, time to full feeding, and complication rate, were similar in both groups.

The main factors contributing to the mortality in patients with duodenal obstruction are a high incidence of associated anomalies, prematurity, and low birth weight. The associated complex cardiac defects continue to be the leading cause of death, particularly in infants with trisomy 21, but recent reviews document that advances in both paediatric cardiology and cardiac surgery in neonates and infants have reduced this mortality significantly.

Early postoperative complications include anastomotic obstruction, continuing obstruction due to missed anomalies, leakage at the anastomosis, and prolonged adynamic ileus. Upper intestinal radiographic studies are necessary to reveal the source of the problem.

The late gastrointestinal complications include megaduodenum, duodenogastric reflux, gastritis, peptic ulcer, and gastro-oesophageal reflux. Megaduodenum is a particularly troublesome problem, which may result either from anastomotic obstruction or from an inherent motility disorder of the proximal duodenum.

Suggested Reading

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Jejunoileal Atresia

26

Alastair J. W. Millar and Alp Numanoglu

Defects in the continuity of the small bowel, one of the most common causes of neonatal intestinal obstruction, can morphologically be divided into either stenosis or atresia. Jejunoileal atresia has a prevalence rate of approximately 1:1000 live births, with a third of infants either born prematurely or small for dates. The anomaly is rarely genetically determined; fewer than 1% of babies will have chromosomal or other associated anomalies.

Most jejunoileal atresias or stenoses result from a localized intrauterine vascular insult to the developing bowel with ischaemic necrosis and subsequent reabsorption of the affected segment(s). Additional pathology in the form of intrauterine fetal intussusception, malrotation and midgut volvulus, thromboembolic occlusions, transmesenteric internal hernias and incarceration, or snaring of fetal bowel in a gastroschisis or exomphalos further supports the ischaemic hypothesis.

The ischaemic insult also adversely influences the structure and subsequent function of the remaining bowel. Histological and histochemical abnormalities can be observed up to 20 cm cephalad from the end of the atretic proximal segment. The distal bowel is unused and potentially normal in function.

26.1 Diagnosis

Prompt recognition of intestinal atresia is essential for the early institution of management. A prenatal history of polyhydramnios and ultrasonography showing bowel dilatation with or without echogenic bowel and obstructed fetal intestine are strong indicators of congenital intestinal atresia. A positive family history will help to identify hereditary forms. Hereditary forms are usually autosomal recessive conditions and may be associated with immune deficiency.

In the newborn baby, intestinal atresia or stenosis may present initially with large intragastric volumes at birth (>20 mL gastric aspirate), followed by persistent bile-stained vomiting, but symptoms may be delayed for more than 24 h in 20% of children. Abdominal distension is frequently present from birth, and the more distal the obstruction, the more generalized the abdominal distension. Proximal jejunal atresia often presents with gastric distension and one or two loops of visible bowel in the upper abdomen relieved by nasogastric tube aspiration. With delay in presentation, increasing intraluminal pressure and/or secondary torsion of the distended bowel proximal to the atresia may lead to ischaemia, perforation, and peritonitis.

The differential diagnosis includes midgut volvulus, intestinal stenosis, meconium ileus, duplication cyst, internal hernia, strangulated inguinal hernia, Hirschsprung's disease, and ileus due to sepsis, birth trauma, maternal medications, prematurity, or hypothyroidism.

The diagnosis of jejunoileal atresia can be established in most cases by an abdominal roentgenogram with air as contrast medium. Erect and supine abdominal radiographs show distended air- and fluid-filled loops of bowel. The more distal the obstruction, the greater the number of distended loops of bowel and fluid levels that will be observed. Occasionally, intraperitoneal calcification may be seen on the plain radiographs, signifying intrauterine bowel perforation, meconium spill, and dystrophic calcification. In the presence of complete obstruction, a contrast enema is usually performed to exclude concomitant colonic atresia, to document the calibre of the colon, and to locate the position of the caecum as an indication of malrotation. With incomplete upper small bowel obstruction, an upper gastrointestinal contrast study may be indicated to demonstrate the site and nature of the obstruction and to exclude midgut volvulus.

The clinical and radiologic presentation of jejunoileal stenosis will be determined by the level and degree of stenosis. The diagnosis is often delayed because of subclinical symptoms and findings.

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26.2 Classification

The morphologic classification of jejunoileal atresia into types I–IV has significant prognostic and therapeutic implications:

- Stenosis (12%) is characterised by a short, localized narrowing of the bowel without discontinuity or a mesenteric defect. The bowel is of normal length.
- Atresia type I (23%) is represented by a transluminal membrane or short atretic segment causing complete intestinal obstruction. The bowel remains in continuity, has no mesenteric defect, and is of normal length (Fig. 26.1).
- Atresia type II (10%) has the blind-ending proximal bowel attached to the collapsed and underdeveloped distal bowel by a fibrous cord along the edge of the mesentery. The proximal bowel is distended and hypertrophied for several centimetres. There is no mesenteric defect and the bowel length is not foreshortened (Fig. 26.2).
- Atresia type IIIa (16%) is similar to type II except that the fibrous connecting cord is absent and there is a V-shaped mesenteric defect. The bowel length may be foreshortened (Fig. 26.2).
- Atresia type IIIb (apple peel) (19%) consists of a proximal jejunal atresia often with associated malrotation, absence of most of the superior mesenteric artery, and a large mesenteric defect. The distal bowel is coiled in a helical configuration around a single perfusing artery arising from the right colic arcades. There is always a significant reduction in intestinal length. These infants are usually of low birthweight and may have associated anomalies (Fig. 26.3).
- Atresia type IV (20%) represents multiple segment atresias like a string of sausages or a combination of types I–III. Bowel length is always reduced (Fig. 26.3).

The most proximal atresia determines whether it is classified as jejunal or ileal. Although single atresias are most commonly encountered, 6–12% of infants have multiple atretic segments, and up to 5% may have a second colonic atresia. The appearance of the atretic segment is determined by the type of occlusion, but in all cases the maximum dilatation of the proximal bowel occurs at the site of the obstruction. When presentation is delayed, it is often hypoperistaltic and of questionable viability.

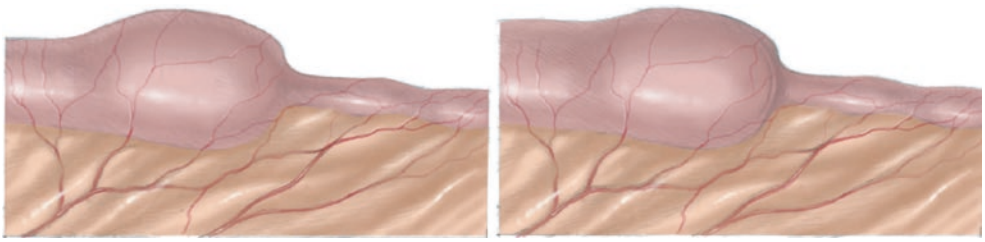

Fig. 26.1 Intestinal stenosis and type I atresia

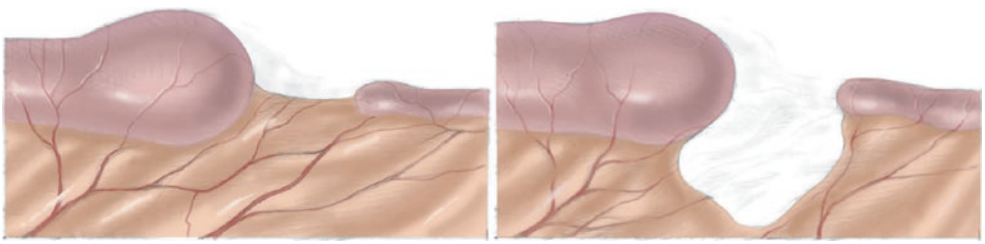

Fig. 26.2 Type II and IIIa atresias

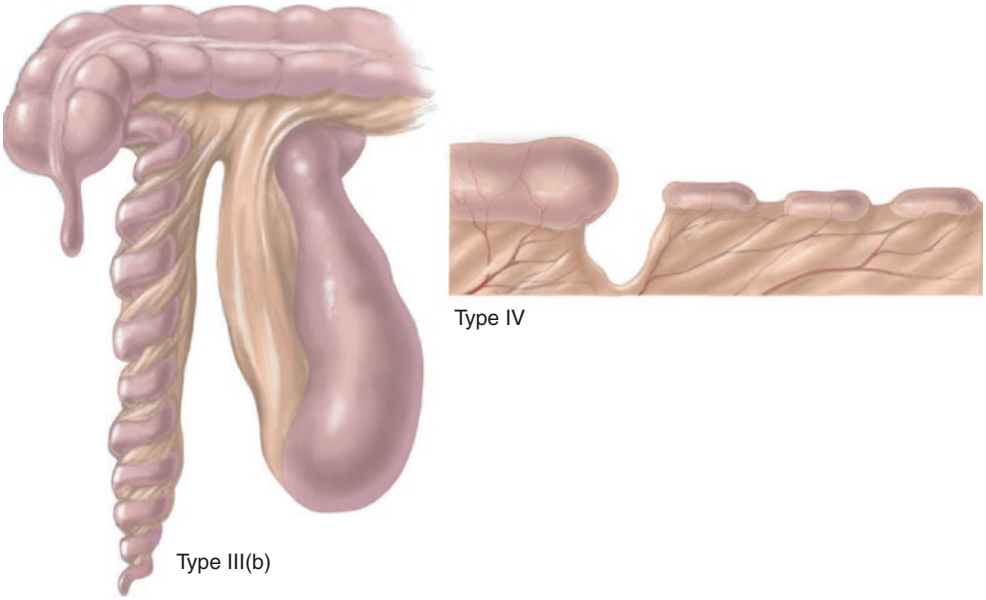

Fig. 26.3 Type IIIb (apple peel) and Type IV multiple atresias

26.3 Surgical Management

The infant is placed supine on a warming blanket, and the exposed abdomen is cleaned and sealed with plastic adherent drapes. Access to the abdominal cavity is obtained through an adequate transverse supraumbilical incision transecting the rectus abdominis muscles and dividing the ligamentum teres between ligatures. Alternatively, a minimally invasive approach via the umbilicus as intra-abdominal access (with or without prior single-port laparoscopy) may be preferred. When using a transumbilical approach, the umbilical cicatrix is dilated and retracted using a Lone Star retracting system. The umbilical vessels are ligated and the abdominal cavity is entered.

The small intestine can easily be exteriorised from the abdominal cavity by gentle pressure on the wound edges and manual delivery of the intestine. The operative procedure will then be determined by the anatomic-pathologic findings (Fig. 26.4).

The operative procedures include the following steps:

1. Identification of atresia type and possible aetiology.
2. Confirmation of patency of distal small and large bowel (if patency of the colon has not been demonstrated by contrast enema prior to surgery) with saline injection.
3. Resection of the proximal bulbous atretic segment.
4. Careful untwisting of volvulated bowel, especially in type IIIb atresia, in which vascular supply to the surviving distal bowel supplied by a single vessel may become compromised.
5. Limited distal bowel resection.
6. Accurate measurement of residual bowel length proximal and distal to the anastomosis.
7. Single-layer end-to-end or end-to-back anastomosis. End-to-end anastomosis is preferred and can be achieved in most cases with a difference in lumen size between proximal and distal bowel of up to 8 to 1, using an extramucosal, interrupted suturing technique.
8. Bowel lengthening procedures have no place during the initial surgery.
9. Gastric decompression postoperatively is best achieved with a Replogle tube on low, continuous suctioning. Neither decompression Stamm gastrostomy nor transanastomotic feeding tubes are recommended. If parenteral nutrition is not available, a transanastomotic feeding tube may be useful to provide enteral nutrition until functional intestinal continuity and prograde peristalsis have been established.
10. The fashioning of a proximal or distal stoma is indicated only in the presence of established peritonitis or compromised vascularity of the remaining intestine.
11. Additional steps may include derotation of a proximal jejunal atresia, back resection to the distal second part of the duodenum, and tapering of the duodenum if very dilated. If total bowel length is significantly reduced (types III and IV), the bulbous dilated segment proximal to the atresia is preserved. As prograde peristalsis of this bowel is deficient, the lumen calibre should be reduced. Maximum mucosal conservation is achieved by inversion plication prior to anastomosis to the distal bowel, but experience has shown that inversion plication often leads to later recurrence of dilatation. A better strategy may be to perform an end-to-end anastomosis and maintain the infant on partial parenteral nutritional support until a plateau of adaptation has been obtained, when a bowel-lengthening procedure such as serial transverse enteroplasty (STEP) or longitudinal intestinal lengthening and tailoring (LILT) can be done, if indicated by the ongoing requirement for parenteral nutrition.

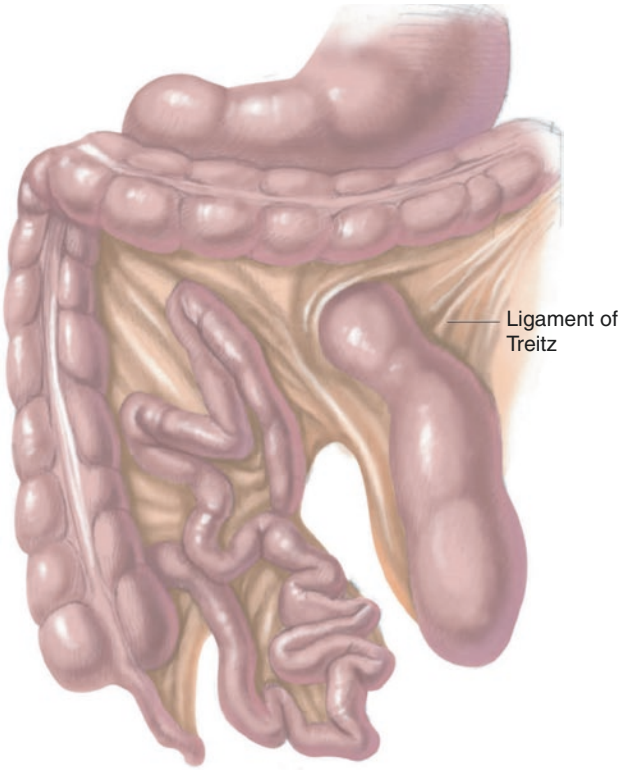

Fig. 26.4 Surgical exploration

Detection of distal atretic areas: It is imperative to exclude distal atresias, which have a prevalence rate of 6–21% (Fig. 26.5). This aim is best achieved with a preoperative contrast enema to exclude an associated colonic atresia, and by injecting saline into the distal collapsed small bowel and following the fluid column distally until it reaches the caecum.

Bowel length measurement: The total length of the small bowel is measured along the antimesenteric border. Once bowel resection has been completed, residual bowel length is of prognostic significance and may determine the method of reconstruction, especially in types III and IV atresias. The normal bowel length is approximately 250 cm in the full-term infant and 115–170 cm in the premature infant.

The surgical procedure is similar for all three types of abdominal access. The atretic and adjacent distended proximal and collapsed distal bowel is isolated with sterile moist swabs. The intestinal content is milked backwards into the stomach, from where it is aspirated; a pus swab sent for culture and sensitivity. Alternatively, proximal bowel contents are milked into the bulbous blind end if it is to be resected. An atraumatic bowel clamp is then placed across the bowel a few centimetres proximal to the elected site for transection. If the total bowel length is deemed to be adequate (>70 cm + ileocaecal valve), the bulbous, hypertrophied proximal bowel is resected (5–15 cm) alongside the mesenteric bowel border in order to preserve maximal mesentery for later use, until bowel of near-normal diameter has been reached. The bowel should then be divided at right angles, leaving an opening approximately 0.5–1.5 cm in width. The blood supply should be adequate to ensure a safe anastomosis. A very limited distal small bowel resection (2–3 cm in length) should then follow (Fig. 26.6). The resection line should be slightly oblique towards the antimesenteric border to ensure that the openings of the proximal and distal bowels are of approximately equal size, to facilitate easy axial end-to-end or end-to-back (Dennis-Browne) single-layer anastomosis. The discrepancy in luminal width of the proximal and distal bowel may vary by 2–5 cm, however, depending on the distance from the stomach.

In type IIIb or high jejunal atresia, the proximal bowel should be derotated and resection of the bulbous portion may be extended into the third or second part of the duodenum without jeopardizing the ampulla of Vater. The distal 'apple peel' component of type IIIb atresia may require release of restricting bands along the free edge of the distally coiled and narrow mesentery to avoid kinking and interference with the blood supply. The large mesenteric defect is usually left open, but with proximal bowel resection, the residual mesentery can be used to obliterate the defect. Furthermore, to prevent kinking of the marginal artery after completion of the anastomosis, the bowel must be replaced in the peritoneal cavity very carefully, in a position of nonrotation.

Although isolated type I atresia is best dealt with by primary resection and anastomosis, multiple diaphragms have been successfully perforated with transluminal bougies passed along the entire length of the affected small bowel.

Multiple type IV atresias (present in 18% of infants) are often localized, necessitating en bloc resection and a single anastomosis rather than multiple anastomoses. It is important, however, to maintain maximum bowel length to avoid the short bowel syndrome.

Similar techniques are used for intestinal stenosis and type I atresias. Procedures such as simple transverse enteroplasties, excision of membranes, bypassing techniques, or side-to-side anastomosis are no longer utilized. They fail to remove the abnormal, dysfunctional segments of intestine and thus increase the risk of the blind loop syndrome.

Anastomosis: The anastomosis is either end-to-end or end-to-back (Dennis Browne method), using 5/0 or 6/0 polypropylene or polydiaxanone stitches (Figs. 26.7 and 26.8). The mesenteric border of the divided ends is united with a stay suture, and a matching stitch is placed at corresponding points of the anti-mesenteric borders of the divided ends. The "anterior" edges of the bowel are then united with interrupted through-and-through extramucosal or inverting

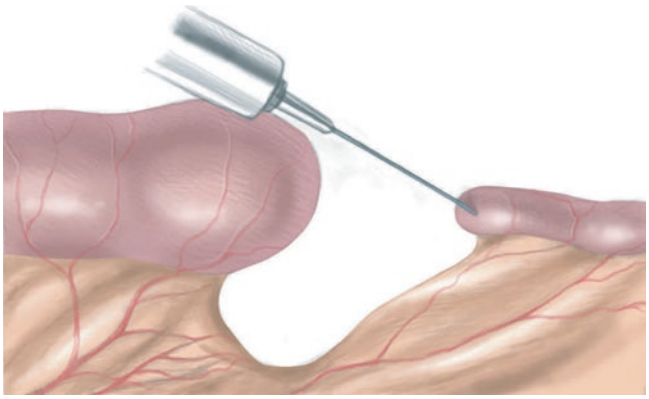

Fig. 26.5 Confirmation of patency of distal bowel

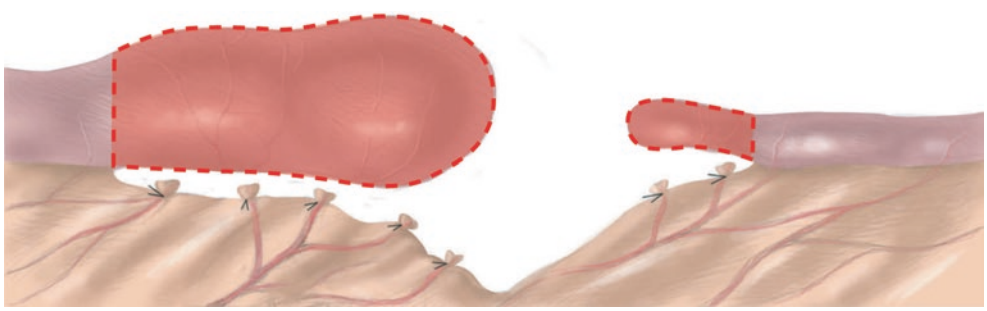

Fig. 26.6 Proximal and distal resection

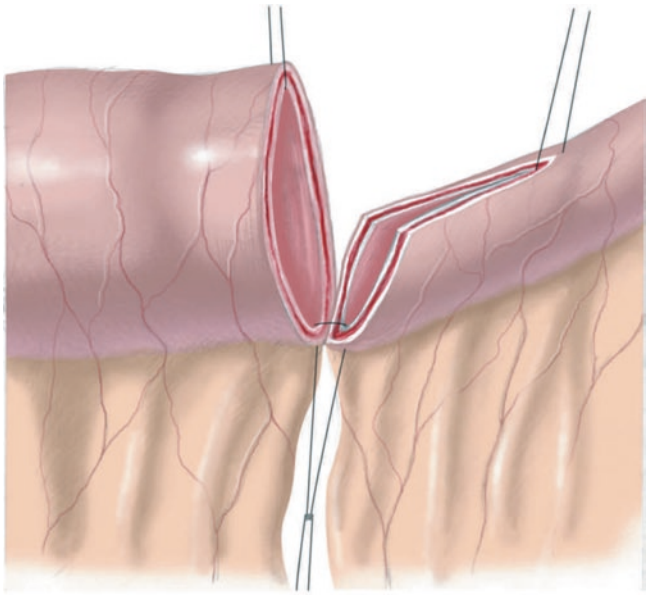

Fig. 26.7 Anastomosis start set up

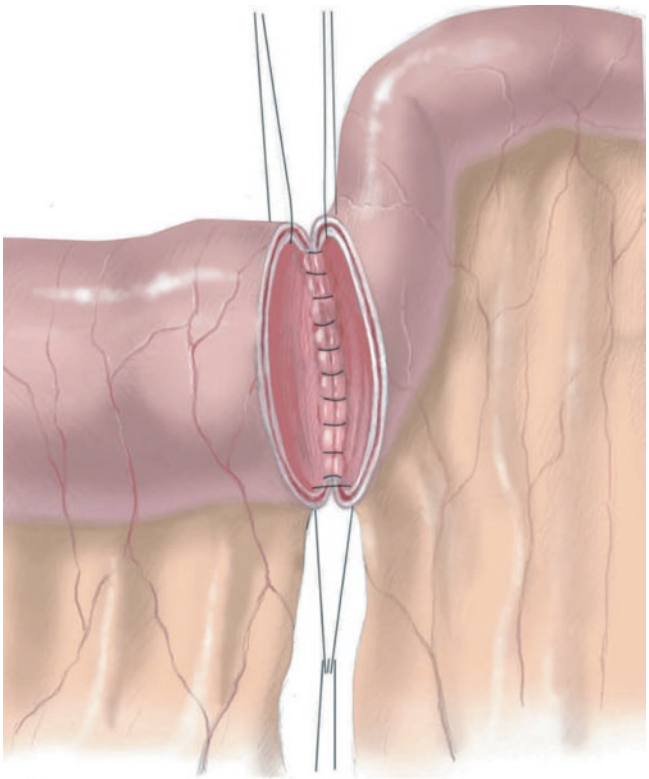

Fig. 26.8 180° rotation of bowel to expose posterior wall

Connell stitches, which are tied on the serosal surface. After completion of one half of the anastomosis, the bowel is rotated through 180° and the "posterior" anastomosis is completed (Fig. 26.9). Alternatively, the posterior edge of the bowel is anastomosed with the stitches tied on the mucosal surface, followed by anastomosis of the "anterior" edges with interrupted stitches tied on the serosal surface. The suture lines are inspected for anastomotic integrity or tested with saline injection on completion.

Mesenteric closure: The defect in the mesentery is repaired by approximating or overlapping the divided edges with interrupted sutures, taking great care not to incorporate blood vessels or to kink the anastomosis (Fig. 26.10). Closure of the large mesenteric defect can be facilitated by using the preserved mesentery of the resected proximal bowel.

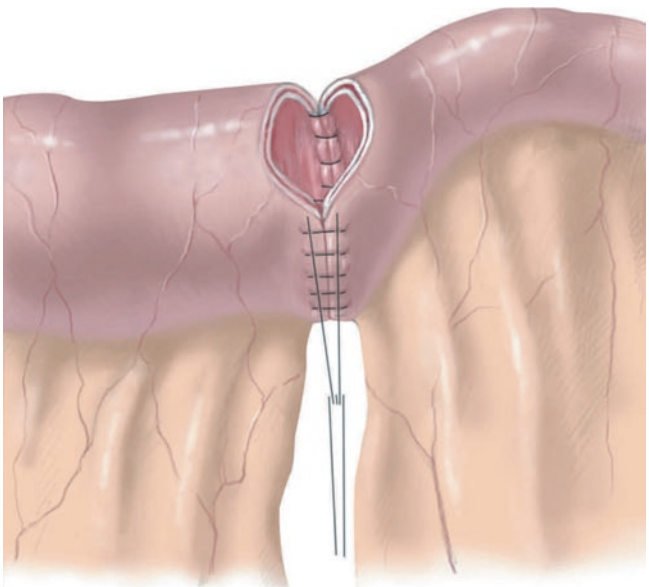

Fig. 26.9 Suturing of posterior wall

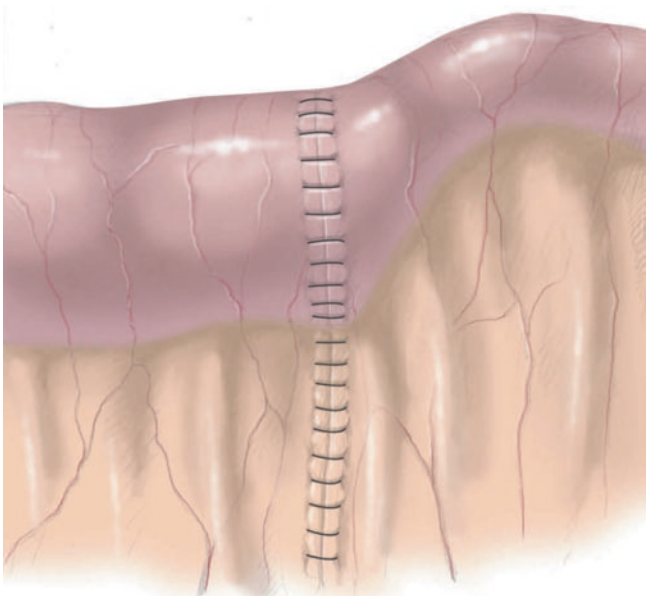

Fig. 26.10 Closure of mesentery

Wound closure: The peritoneal cavity is thoroughly irrigated with warm saline to remove all macroscopic debris, and the bowel then returned to the abdominal cavity. Care is taken not to kink the anastomosis.

The abdomen is closed by approximating en masse all the layers of the abdominal wall (excluding Scarpa's fascia) with a single, continuous 4/0 monofilament absorbable suture, followed by subcutaneous and subcuticular absorbable stitches. If a transumbilical approach has been used, the umbilical defect is closed with a purse-string suture. In general, transanastomotic tubes are not recommended, but they may be used in more proximal atresias to provide enteral nutrition in the early postoperative period. Transanastomotic tubes exiting through the distal bowel have been used to provide temporary decompression of the dilated bowel proximal to the atresia if it was not resected prior to performing the anastomosis.

26.4 Alternative Surgical Techniques

Alternative surgical techniques may be required if the ischaemic insult has resulted in an atresia with markedly reduced intestinal length, if large resections of abnormal or multiple atretic segments are required, or if the measured length of the residual small intestine is less than 70 cm.

26.4.1 Tapering Duodenojejunoplasty

Tapering duodenojejunoplasty has several indications:

- As part of bowel length preservation where the proximal atretric segment is grossly dilated and hypertrophied over an extended distance (typical in type IIIb atresia or jejunal atresia).
- To equalize disparity in anastomotic lumen size.
- For the correction of a failed inversion plication procedure.
- To improve function in a persistently dilated, nonfunctioning megaduodenum following surgery for high jejunal atresia.

The atretic jejunoduodenum is derotated and the antimesenteric segment of the dilated proximal segment is resected over a 22–24 French catheter. The resection may extend cephalad to the second part of the duodenum. An intestinal autostapling device may be used to facilitate the resection and anastomosis. The longitudinal anastomotic line is reinforced with an absorbable 5/0 or 6/0 Lambert suture (Figs. 26.11 and 26.12). Tapering can be done safely over a length of 20–35 cm. The tapered bowel is then primarily anastomosed to the distal bowel (± equal bowel diameters) and placed on the right side as for corrected malrotation, with the caecum in the left hypochondrium.

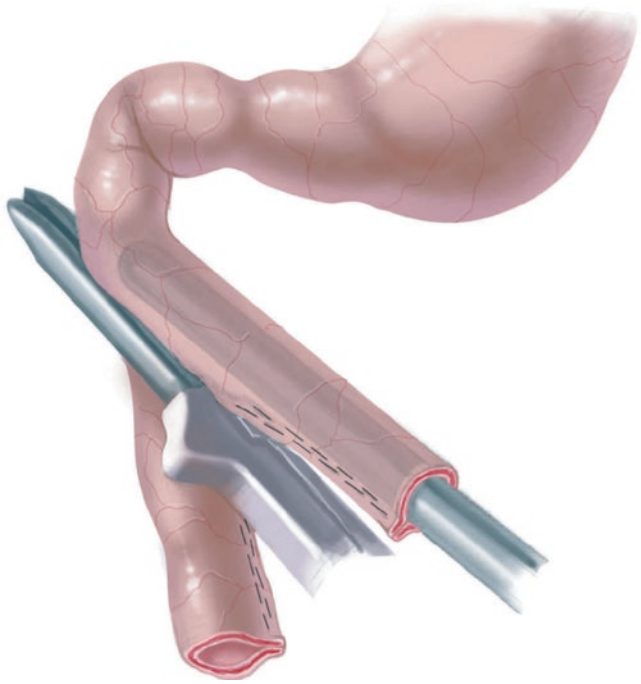

Fig. 26.11 Staple tapering of proximal bowel

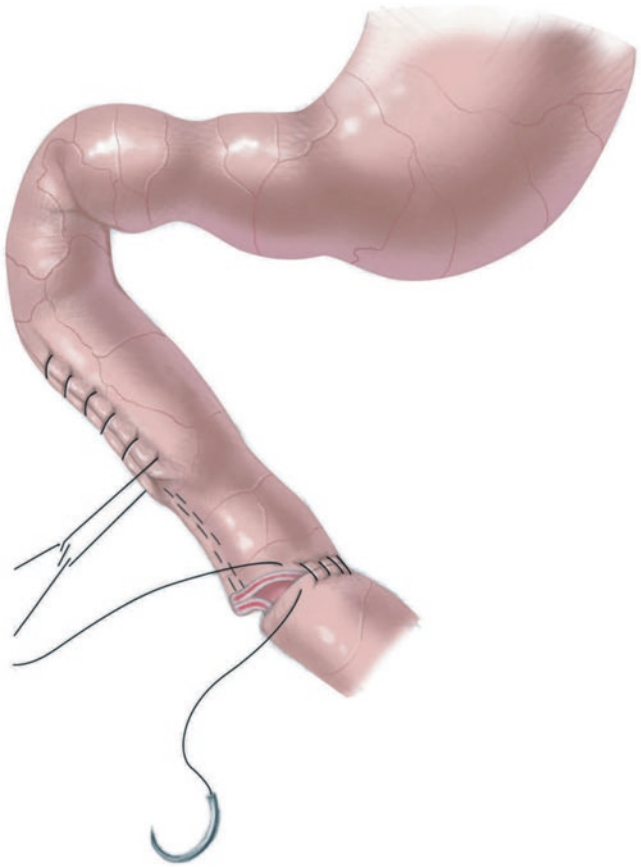

Fig. 26.12 Reinforcement of staple line

26.4.2 Plication

The derotation and back resection methods are used as described for tapering procedures. The plication method has the advantages of both reducing the risk of leaking from the antimesenteric suture line and conserving mucosal surface area; it may even facilitate the return of bowel peristalsis. More than half of the antimesenteric bowel circumference may be infolded into the lumen over an extended length without causing an obstruction, with care being taken not to narrow the lumen excessively (Figs. 26.13 and 26.14). The 'keel' must be trimmed and closed with interrupted sutures. The main drawback of this method is unravelling of the suture line within a few months, necessitating revision. The bowel is left in a position of derotation, with the duodenojejunum placed on the right side of the abdominal cavity, with the mesentery broad-based and the caecum in the left hypochondrium, as in a Ladd's procedure.

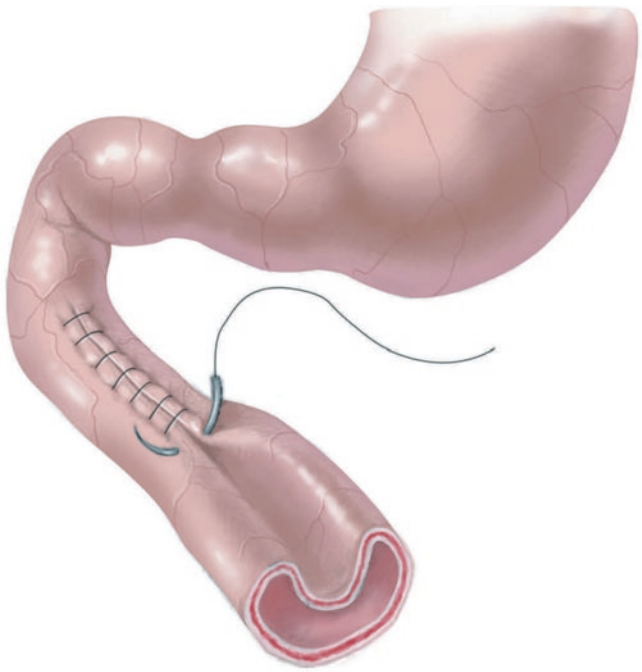

Fig. 26.13 Plication

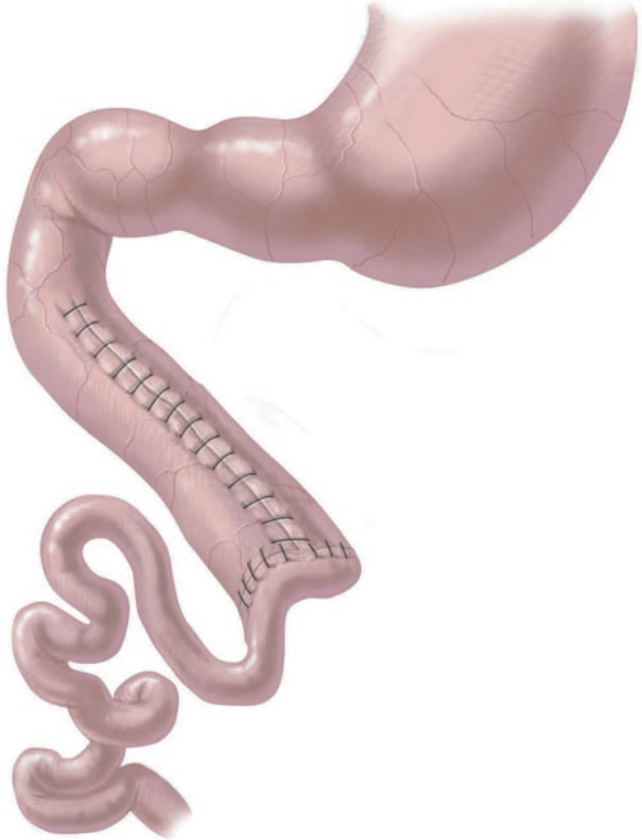

Fig. 26.14 Completion of anastomosis

26.4.3 Antimesenteric Seromuscular Stripping and Inversion Plication

This technique prevents unravelling of the plication method and preserves maximal mucosal surface for absorption. Two antimesenteric converging seromuscular strips about 2 mm in width are resected (easily done by stabilising the proposed line of resection with straight, nontoothed forceps). A seromuscular strip is then excised, taking care not to damage the mucosa. The two denuded mucosal strips are then approximated with a running monofilament suture. The 'keel' of inverted bowel should be trimmed and the bowel edges approximated with interrupted sutures prior to anastomosis to the distal bowel (Figs. 26.15, 26.16 and 26.17). Proximal and distal luminal size can be approximated to facilitate the anastomosis.

Fig. 26.15 Antimesenteric seromuscular stripping

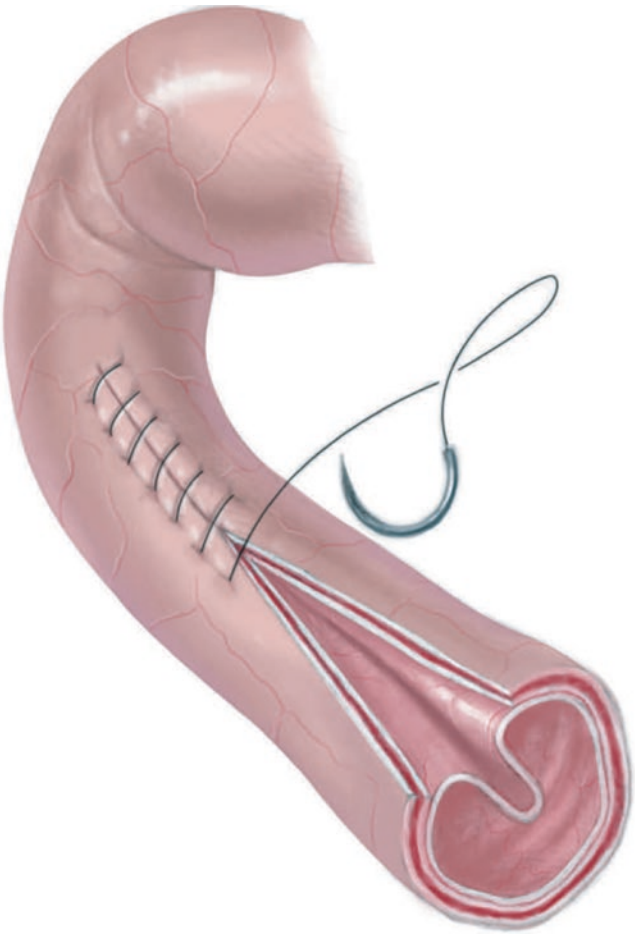

Fig. 26.16 Plication of bowel wall following seromuscular stripping

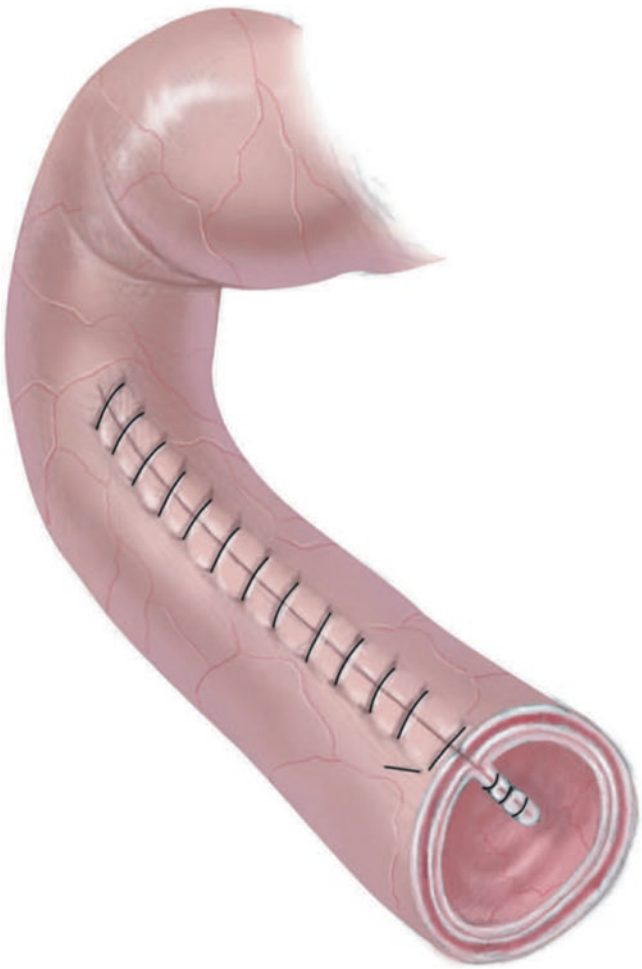

Fig. 26.17 Completion of seromuscular stripping and inversion plication

26.5 Results and Complications

The overall survival amongst newborn babies with intestinal atresia has increased from a dismal 10% in 1952 to over 90% at present, primarily owing to a change in the surgical procedure from primary anastomosis without resection to liberal resection of the blind proximal and distal ends followed by end-to-end anastomosis. Despite improvements in surgery, anaesthesia, and postoperative care, type IIIb atresia still carries a mortality of 19%, however, predominantly due to gangrene of the proximal end of the distal segment (7%), anastomotic leak (15%), and stricture formation (15%). The prognosis of intestinal atresia is further determined by genetic factors, prematurity (30%), delay in presentation, associated diseases such as cystic fibrosis, malrotation (45%), exomphalos, gastroschisis, Hirschsprung's disease, other gastrointestinal atresias, infarction of the proximal atresia with peritonitis, sepsis, pneumonia, and the complications of prolonged parenteral nutrition.

Understanding the pathogenesis of atresia and adapting surgical procedures to minimize loss and conserve bowel length ensured that most infants will have sufficient bowel length for normal alimentary tract function and overall growth and development.

The incidence of postsurgical complications such as anastomotic leaks, stricture formation, transient intestinal dysfunction, blind loop syndrome, and the short bowel syndrome can be minimized by careful attention to the presenting anatomic detail, meticulous surgical technique, and maximal bowel length conservation methods. Because of the high incidence of unravelling, the plication technique is rarely used.

The short bowel syndrome is a major factor influencing outcome. It may be due to predisposing factors such as extensive intrauterine bowel loss, operative factors such as overzealous bowel resection and ischaemic injury to the bowel, or postoperative complications. Under ideal circumstances, a survival rate of more than 70% can be expected in most infants with less than 25 cm of jejunoileum, especially if the ileocaecal valve is intact.

Several surgical procedures have been identified to improve the outcome of the short bowel syndrome, including serial transverse enteroplasty (STEP) and Bianchi's longitudinal intestinal tailoring and tapering (LILT), as well as reversal of segments of bowel, interposition of colonic segments, and methods to increase mucosal surface area for absorption purposes. Most are of experimental value only, except for bowel-lengthening procedures; these procedures should not be performed initially, but rather only after the failure of conservative methods to stimulate and allow maximum bowel adaptation. Full bowel adaptation may require 6–18 months.

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Meconium Ileus

27

Guido Ciprandi and Massimo Rivosecchi

Meconium ileus is the earliest clinical manifestation of cystic fibrosis (CF) and occurs in 8–10% of patients with CF at birth. CF is an autosomal recessive disorder caused by the presence of mutations in both copies of the gene for the protein cystic fibrosis transmembrane conductance regulator (CFTR). CFTR is involved in the production of sweat, digestive fluids, and mucus. When CFTR is not functional, secretions that are usually thin instead become thick. CF is diagnosed by a sweat test and genetic testing. The sweat test has been used since the early 1950s to detect anomalies in chloride metabolism. The reference interval for sweat chloride levels among infants 5–6 weeks of age has recently been established to be <30 mmol/L.

The clinical features are due mainly to the presence of abnormal, inspissated and viscid mucus of intestinal origin. In neonates affected by this condition, the impacted meconium produces an intraluminal obstruction in the midileum, leading to a progressive distension. In about 40% of patients, meconium ileus is complicated by intestinal volvolus, atresia, gangrene and necrosis, perforation, peritonitis with abdominal calcifications, and meconial pseudocyst. Meconium ileus accounts for 9–33% of all neonatal intestinal obstructions, with an incidence of 1:2500 newborns. It is the third most common cause of neonatal small bowel obstruction, after ileal and duodenojejunal atresia and malrotation.

Relief of the intestinal obstruction has been obtained by irrigating the intestine with various solutions such as normal saline, 1% *N*-acetylcysteine, hyperosmolar ionic radiopaque diatrizoate, surfactant, or DNase (an endonuclease able to hydrolyse phosphodiester bonds). With surgical and medical efforts, the survival rate at 1 year has increased from 10% to 90%, and the operative mortality is drastically decreased to 15–23% of treated newborns in the past 10 years.

27.1 Symptoms and Diagnosis

Polyhydramnios is the most frequent feature observed in prenatal diagnosis of complicated forms of meconium ileus. The presence of fetal hyperechogenic bowel on the ultrasound, associated with dilated bowel and/or ascites could be indicative of an intestinal obstruction. A family history of CF is clearly evident in almost 25% of these patients. Meconium ileus is uncommon in premature infants (5–12%), and associated congenital anomalies are rare.

In meconium ileus, the proximal, the middle, and the distal ileum show different aspects. In the first portion, nearly normal evidence is present, with a progressive dilatation at the mid-portion borderline. In the *proximal ileum*, the content has a semiliquid consistency and is not yet viscous. Marked and severe dilatation of the *middle ileum* is always seen, and the intestine contains thick, dark green, putty-like meconium that is firmly adherent to the walls. The intestinal obstruction causing a hyperperistalsis is responsible for the congestion and hypertrophy of the walls. The *distal ileum* is full of concretions called "rabbit pellets"—grey-stained and with a typical beaded appearance. This small bowel condition is responsible for a narrow, empty, and small colon, never used, which is called *microcolon*.

The main symptoms include abdominal distension (96%), bilious vomiting (50%), and delayed passage of meconium (36%). From a clinical point of view, it is possible to recognize two different conditions: a simple, uncomplicated, nonsurgical type, and a complicated, severe type, which has a

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mortality of at least 25% of all cases. In the first type (58%), signs and symptoms of a distal ileal obstruction are seen not later than 48 h after birth:

- Generalized abdominal distension with dilated and visible as well as palpable loops of bowel
- Bilious vomiting
- No stools
- Narrowing of the anus and rectum, with only a dense and rubber-like grey meconium sticking to the anal wall.

In the second type (42%), the neonate represents a surgical emergency and must be treated within 24 h of birth, when the signs of hypovolemic shock or sepsis are not well established. A fetus with complex meconium ileus is at increased risk of postnatal bowel obstruction and perforation. The usual X-ray image of a fine, granular, "soap-bubble" appearance ("Singleton's sign") or a "ground-glass" appearance ("Neuhauser's sign") is due to dense meconium mixed with air, typical of the distal ileum; this picture is usually located in the mid-abdomen or in the right iliac fossa. When the meconium ileus is complicated, the abdominal radiograph may show calcification as a result of meconium peritonitis due to a fetal perforation of the intestine. A "double-bubble" image or air-fluid levels can be seen when a secondary ileal atresia (single or double) is the final bowel remodelling after a complete volvulus associated with severe ischaemic damage. If the intestinal perforation occurs early in the antenatal period, the X-ray appearance of a round rim of calcification underlines a meconium pseudocyst.

27.2 Treatment

The first steps in treatment include nasogastric tube decompression, antibiotic prophylaxis, and correction of dehydration, electrolytes and hypothermia.

27.2.1 Contrast Enema

For uncomplicated cases (Fig. 27.1), a contrast enema with water-soluble and hyperosmolar or iso-osmolar contrast is the medical treatment of choice and is safe for the mucosa (Fig. 27.2). A recent study that administered various enema solutions in a mouse model showed that surfactant and diatrizoic acid (Gastrografin®) were more efficacious for the in vivo relief of constipation than perflubron, polysorbate 80 (Tween® 80), GoLYTELY, DNase, *N*-acetylcysteine, and Viokase. Intestinal mucosal damage was absent, and viscosity was significantly reduced in vitro.

The enema evacuation should be obtained under fluoroscopic control, with a gentle and progressive increasing of the intraluminal pressure, thus avoiding unexpected fractures of the colon. A correct procedure prevents leakage of the contrast medium and catheter dislocation by taping the buttocks. If the contrast medium fails to progress into the dilated small bowel loops, the presence of an acquired atresia is definite and the radiologist must stop the examination because of a high risk of perforation. Of neonates undergoing this procedure, 50% benefit from the enema alone over the next 48 h, without any additional treatment; in some cases, a second enema may be used to achieve complete evacuation of the meconium filling the ileal loops. Acetylcysteine adminis-

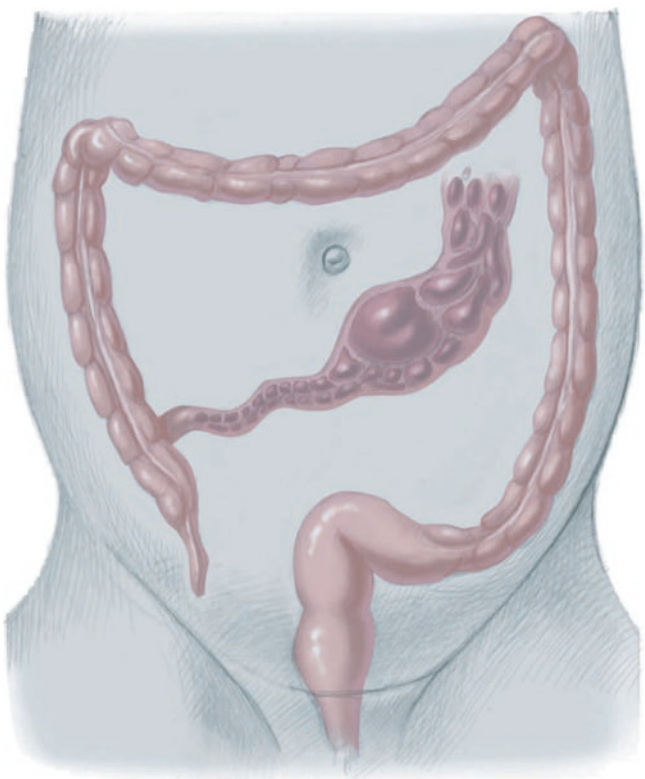

Fig. 27.1 In uncomplicated meconium ileus, dilated small bowel loops are filled with meconium

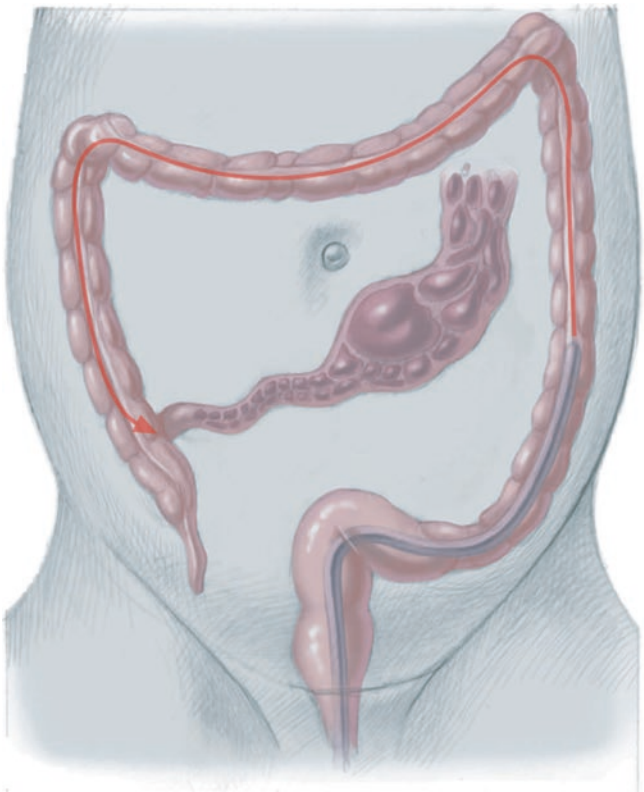

Fig. 27.2 Enema under fluoroscopic control may achieve complete evacuation of the meconium filling the ileal loops

tered by mouth is useful and helps to relieve the obstruction. Radiographs are taken at 3, 6, 12, 24, and 48 h, with the aim of evaluating progression and possible complications. At this time feeding is begun. Hypovolemic shock and early perforation are possible complications, but they can be avoided with an appropriate and meticulous procedure.

27.2.2 Enterotomy Irrigation

Nonoperative management using a water-soluble contrast enema fails in only 6–10% of uncomplicated cases. If no significant difference in intestinal diameters is seen and no microcolon is present, a limited enterotomy and repeated warm saline irrigations through a smooth catheter provide the best result (Fig. 27.3). Meconium discharge may be manually supported, using an enterotomy placed in the dilated hypertrophic ileum. The two-way catheter is directed with care, clearing the small and the large bowel. After exclusion of an atretic intestinal segment, the enterostomy is closed by single, interrupted seromuscular stitches.

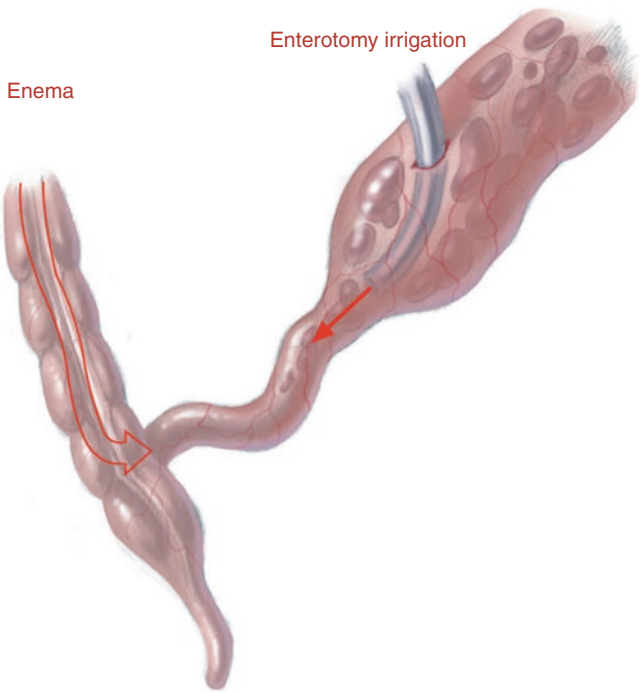

Fig. 27.3 An enterotomy in the dilated, hypertrophic ileum allows repeated warm saline irrigations through a smooth catheter to allow meconium evacuation

27.2.3 Double-Tube Ileostomy

Approximately half of neonates with meconium ileus cannot be treated adequately with irrigations alone, or they have additional intestinal obstruction complicated by neonatal intestinal perforation or ileal atresia secondary to intrauterine perforation. These patients always require a surgical procedure such as resection of the dilated meconium-filled ileum and ileal anastomosis (as shown in Chap. 26). Additionally, complicated cases of meconium ileus are seen in newborns with an extreme difference in the diameters of the proximal and distal ileum and a significant microcolon. In those cases, the double-tube ileostomy technique according to Rehbein has proved to be an effective treatment that avoids the secondary laparotomy for stoma closure.

A horizontal, predominantly right-sided laparotomy is performed approximately 2 cm below the umbilicus. A small transverse incision into the enlarged ileum is performed, approximately 5–7 cm in front of the narrow part filled with stool pellets. Four stay sutures are inserted into the margins of the incision. If an atresia exists, the atretic part is resected and the thickened meconium from the proximal part and the grey stool pellets from the distal part are evacuated by numerous irrigations with warm saline through a 5- to 8-Ch feeding tube; the manoeuvre is supported by gentle manual forward and backward manipulation. Once all intestinal contents are evacuated, a 10-Ch feeding tube with larger cut openings is inserted through the enterotomy in the proximal nondilated ileum, and a second 5-Ch tube is inserted into the distal narrow ileum or microcolon in a T-tube fashion (Fig. 27.4).

The enterotomy is closed around the tubes and is tightly fixed to the appropriate part of the ventral abdominal wall, in a similar fashion as in a gastrostomy. Both tubes are carefully fixed with nonabsorbable sutures to the skin (Fig. 27.5). If a bowel segment was resected, the stoma for the tubes is best situated approximately 5 cm in front of the anastomosis, and the small tube is passing the anastomosis far into the narrow intestine. Postoperatively, the large tube serves for suctioning and evacuation of the intestinal contents; the small distal tube serves for constant irrigation with increasing amounts of fluid (first saline; later, predigested milk), thereby promoting rapid enlargement of the ileum and the microcolon. As soon as the evacuations through the large tube become less and normal bowel movements occur, it is indicative that most of the intestinal contents are passing by distally. The double tubes can then be simply removed. We have used this double-tube method successfully since the 1980s and no longer perform cutaneous enterostomy.

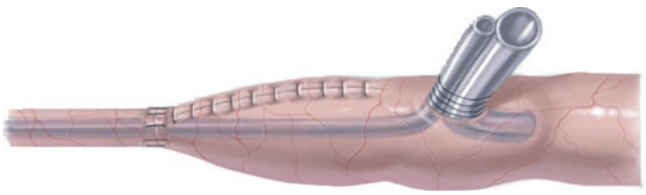

Fig. 27.4 A 10-Ch feeding tube with larger cut openings is inserted through the enterotomy in the proximal nondilated ileum, and a second 5-Ch tube is inserted into the distal narrow ileum or microcolon in a T-tube fashion. If a bowel segment was resected, the stoma for the tubes is best situated approximately 5 cm in front of the anastomosis, and the small tube passes the anastomosis far into the narrow intestine

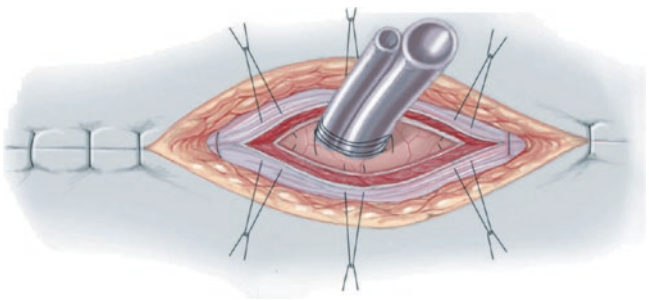

Fig. 27.5 The enterotomy is closed around the tubes and is tightly fixed to the appropriate part of the ventral abdominal wall, in a similar fashion as in a gastrostomy. Both tubes are carefully fixed to the skin with nonabsorbable sutures

27.2.4 Enterostomy

Surgical techniques developed in the past have consisted of a resection of the enlarged bowel segment and temporary decompression by means of a distal or proximal enterostomy. The simplest form is a double-barrelled ileostomy according to Mikulicz, with the two loops brought out side-to-side (Fig. 27.6). This solution is quick and avoids an intra-abdominal anastomosis. More technical alternatives described later have included a distal ileostomy with end-to-side ileal anastomosis (Bishop-Koop), known as a "distal chimney enterostomy" (Fig. 27.7). This procedure consists of a Roux-en-Y anastomosis between the end of the proximal segment and the side of the distal segment, at least 3–5 cm from the open end. The open limb of the distal segment is used as an ileostomy. A variation of this technique has been described that uses an angulating proximal segment, which is obliquely anastomosed with the distal stump (Fig. 27.8). Proximal chimney enterostomy, the so-called Santulli procedure, consists of a proximal ileostomy with end-to-side ileal anastomosis (Fig. 27.9). The end of the distal limb is anastomosed to the side of the proximal limb, the end of which is used as the enterostomy. This technique should facilitate irrigation as well as decompression of the proximal small bowel.

All of these enterostomies can be closed by an end-to-end anastomosis when uninhibited passage of intestinal contents is established, usually in 7–12 days.

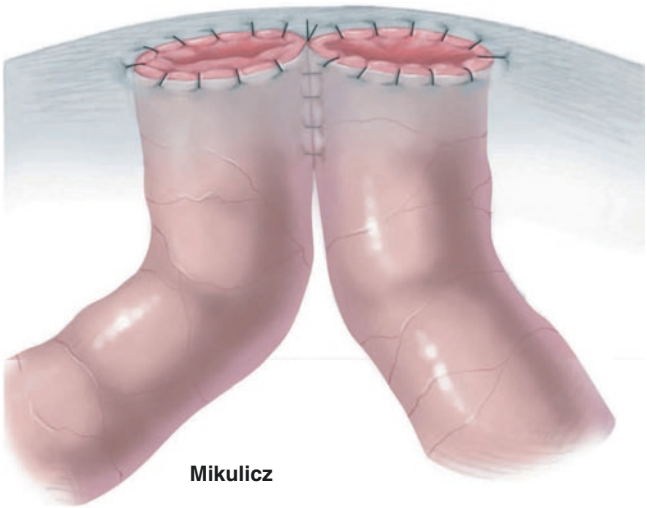

Fig. 27.6 A double-barrelled ileostomy according to Mikulicz, with the two loops brought out side-by-side

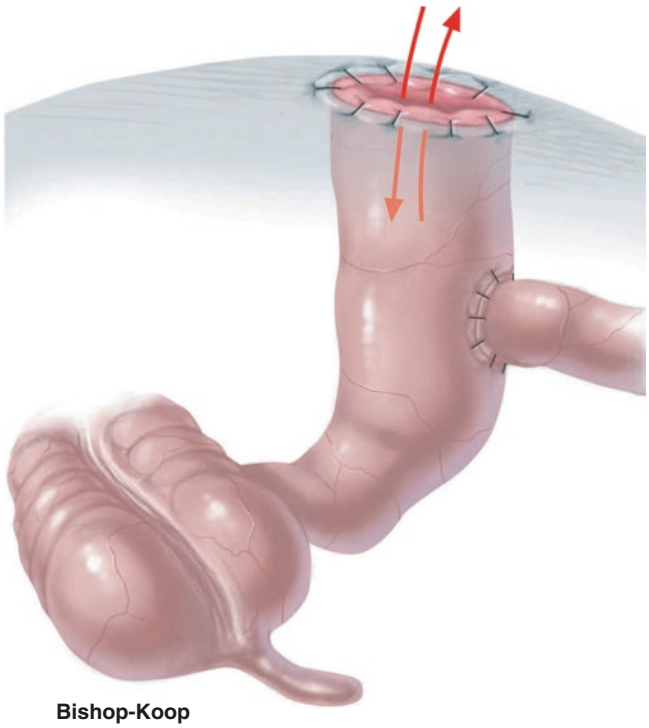

Fig. 27.7 A distal ileostomy with end-to-side ileal anastomosis (Bishop-Koop), known as a "distal chimney enterostomy". This procedure consists of a Roux-en-Y anastomosis between the end of the proximal segment and the side of the distal segment, at least 3–5 cm from the open end. The open limb of the distal segment is used as an ileostomy

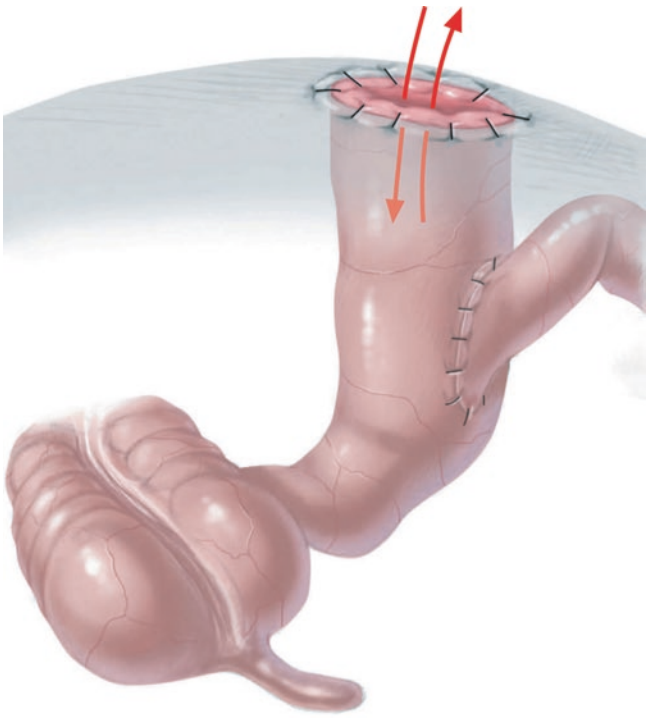

Fig. 27.8 A variation of the Bishop-Koop technique uses an angulating proximal segment, which is obliquely anastomosed with the distal stump

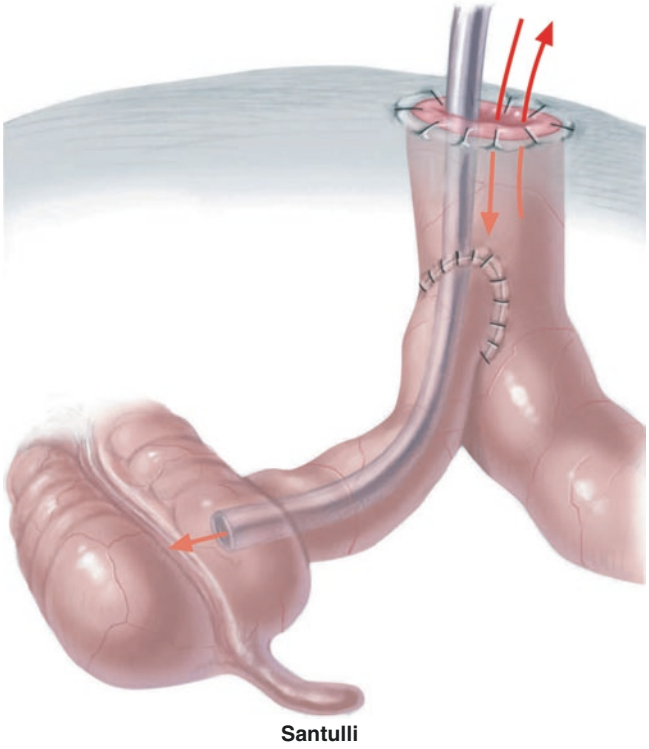

Fig. 27.9 Proximal chimney enterostomy, the so-called Santulli procedure, consists of a proximal ileostomy with end-to-side ileal anastomosis. The end of the distal limb is anastomosed to the side of the proximal limb, the end of which is used as the enterostomy. This technique should facilitate irrigation as well as decompression of the proximal small bowel

27.2.5 Terminal Ileostomy

When a terminal ileostomy or colostomy is desired in neonates or children, the preferred method is the nipple-valve system formation (Fig. 27.10). This is an easy technical procedure that avoids any kind of infiltration, stricture, retraction of the neostoma. The intestinal segment used is 2–3 cm long and has a serosal surface that is free of fat and has a good vascular supply. The seromuscular wall is sutured to the fascia in four quadrants. The stitch is then taken through the skin seromuscular wall and the edge of the stoma, thereby creating the nipple. Four to six sutures are normally needed, depending upon the size of the stoma.

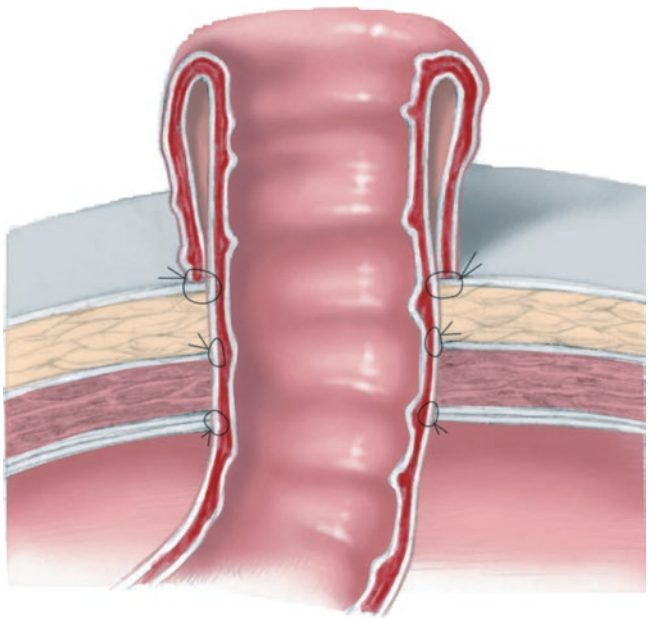

Fig. 27.10 Nipple-valve system for creating a neostoma. The intestinal segment used is 2–3 cm long and has a serosal surface that is free of fat and has a good vascular supply. The seromuscular wall is sutured to the fascia in four quadrants. The stitch is then taken through the skin seromuscular wall and the edge of the stoma, thereby creating the nipple. Four to six sutures are normally needed, depending upon the size of the stoma

27.3 Outcome and Complications

Generally, the less the extent of bowel resection, the earlier the recovery of peristalsis. Actually, bowel resection with primary anastomosis has been proved to be as effective and safe as stoma formation, and it is associated with a reduced length of initial hospital stay. The most important complication is pulmonary infection, which occurs in at least 8–10% of these patients. Anastomotic leakage may occur for various reasons, including a technical mistake, an insufficient blood supply, or an unrecognized distal obstruction. Delayed recovery of peristalsis is another frequently observed complication and is due to abnormal stretching of the intestinal walls during fetal life. Total parenteral nutrition is the support of choice and a central venous catheter is mandatory in these situations.

Meconium ileus may be an early indication of a severe phenotype of CF, as suggested by the significantly lower pulmonary function found in children with a history of meconium ileus compared with age- and sex-matched children who did not have meconium ileus. In this view, patients without CF demonstrated better growth and functional status, and had a lower incidence of lung diseases and gastrointestinal problems. It is also possible that patients affected by meconium ileus have a more severe form of CF that, from a pathological point of view, consists of more severe compromise of mucous glands.

The complicated forms of meconium ileus are susceptible to additional long-term surgical complications, including especially small bowel obstructions (of adhesive origin) and blind loop syndromes (30% of the cases). These long-term problems are more commonly ascribed to a higher incidence of peritonitis and small bowel ischaemia. Very few patients undergoing enterotomy and irrigation manifest long-term surgical morbidity. There is general agreement that resection and stomas could be avoided with more meticulous attention to care and clearing the more dense concretions from the intestinal walls.

Long-term complications are rarely seen in neonates affected by uncomplicated meconium ileus who were not operatively treated, and only mild and transient complications have been observed in newborns treated with minor surgical procedures, such as enterotomies and irrigations. Meconium ileus does not directly affect long-term functional or social status in children with CF. Their quality of life is mainly correlated with the extension of lung lesions, which are the most limiting factor with respect to school, social life, and physical activity.

Survival of neonates with meconium ileus has improved over the past two decades because of neonatal intensive care and improved surgical technique and medical treatment. In general, an overall immediate survival of 90% is achieved using the modern protocols; nearly all deaths occur in adolescents. Few children die because of liver or septic complications. Deaths are mainly due to sepsis (primary or secondary to pulmonary interstitial emphysema) or aspiration pneumonia. In a large series reported and analysed by Fuchs, only one child died because of the meconium ileus itself.

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Gastrointestinal Duplications

28

Mark D. Stringer

Alimentary tract duplications, sometimes known as enteric cysts, enterogenous cysts, or reduplication cysts, are rare congenital malformations that may occur anywhere from mouth to anus. They are usually single, vary widely in size, and are more often cystic than tubular. Unlike tubular duplications, most cystic lesions do not communicate with the gut lumen. Intestinal duplications are typically located on the mesenteric aspect of the intestine, share a smooth muscle wall and common blood supply with the adjacent bowel, and are lined by alimentary tract mucosa. Occasionally, they are located separate from the alimentary tract, such as in the retroperitoneum. Heterotopic mucosa, most often gastric but occasionally respiratory or pancreatic, is present in 20–30% of all alimentary tract duplications but in a higher proportion of tubular duplications.

28.1 Pathogenesis

There is no satisfactory single explanation to account for the development of gastrointestinal duplications. The pathogenesis of those with associated vertebral anomalies can be explained by abnormal adherence of the endoderm of the primitive gut to the notochord (the split notochord theory). Some hindgut duplications are an expression of caudal twinning, and some intestinal lesions may result from an intrauterine mesenteric vascular accident or defective recanalization of the developing gut tube.

28.2 Presentation

Enteric duplications frequently cause symptoms in infancy or early childhood, although some remain asymptomatic until adult life. An increasing proportion are detected incidentally during imaging for unrelated symptoms or conditions, including routine prenatal ultrasonography. Presenting symptoms and signs depend on the location and size of the duplication, and whether it contains heterotopic gastric mucosa. Potential complications include obstruction of the airway (coughing, wheezing, and/or pneumonia), oesophagus (dysphagia), or intestine (abdominal pain and vomiting). Cysts lined by heterotopic gastric mucosa are prone to ulceration, haemorrhage, or perforation. Intussusception and segmental volvulus are other possible complications of small bowel duplications. Occasionally, duplications are complicated by infection. Malignant degeneration has been reported as a late complication of duplication cysts in adults, but there have been no such reports in children under 16 years of age.

About half of all enteric duplications are related to the midgut, with the ileum being the single commonest site (Fig. 28.1); just over one third are foregut in origin. Vertebral anomalies such as bifid or hemi-vertebrae or vertebral fusion may also be present; they are most often associated with thoracic and hindgut duplications. Rarely, in such cases, there may be associated intraspinal pathology, when the duplication is often referred to as a neurenteric cyst. Other recorded associated congenital malformations include congenital cardiac disease, oesophageal atresia, congenital diaphragmatic hernia, congenital pulmonary malformations, and myelomeningocele with foregut duplications; intestinal malrotation or (less commonly) intestinal atresia with midgut duplications; and genitourinary duplication, bladder exstrophy, and imperforate anus with hindgut duplications.

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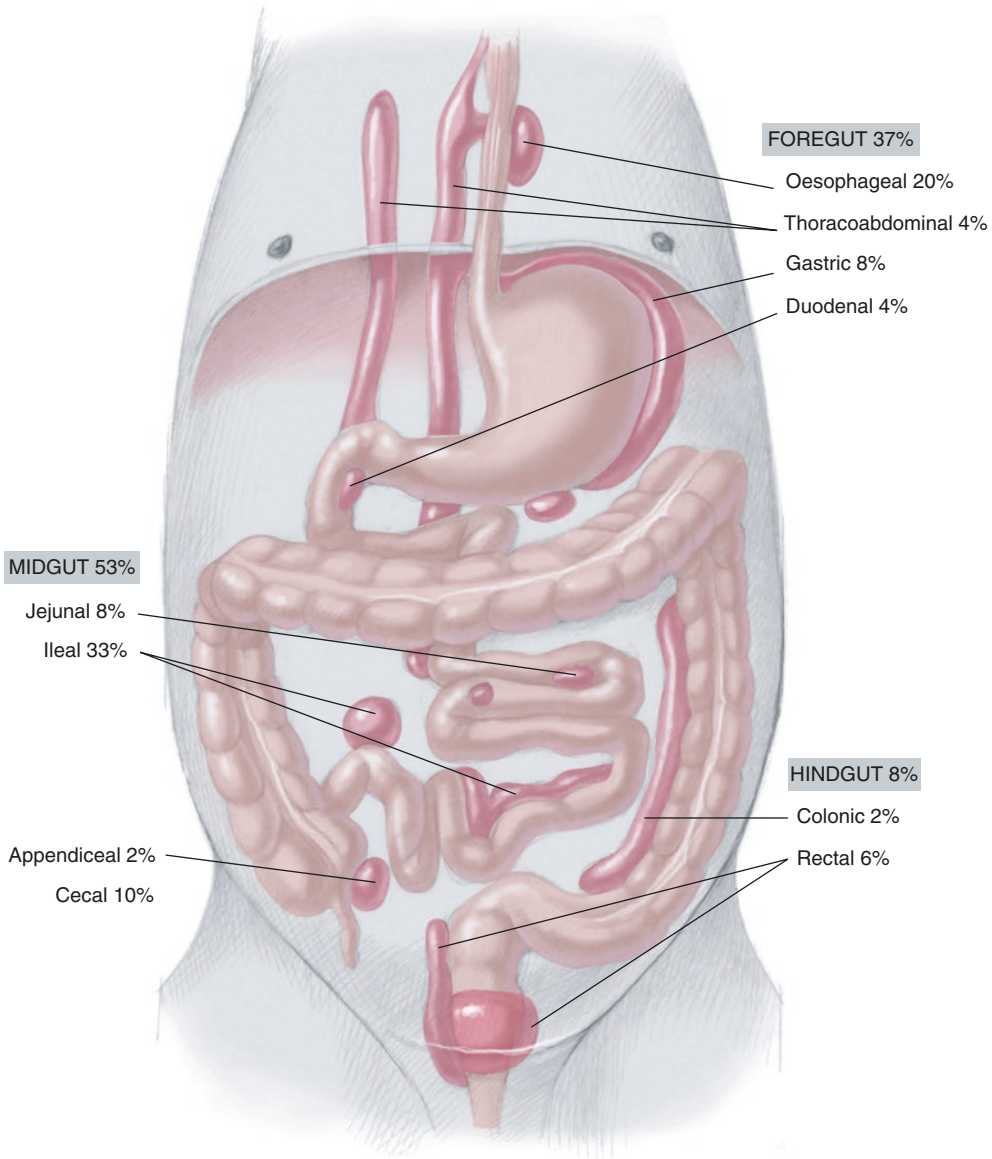

Fig. 28.1 Types of enteric duplications and their distribution

28.3 Preoperative Imaging

Imaging is a critical aspect of preoperative preparation. The choice of imaging is dictated by the site of the duplication, clinical urgency, and the potential for associated anomalies. Isolated small bowel duplications often require no preoperative investigations other than abdominal sonography, a plain abdominal radiograph and routine blood tests. Occasionally, a technetium-99m pertechnetate radionuclide scan is helpful in detecting heterotopic gastric mucosa. In contrast, thoracoabdominal lesions demand detailed radiologic imaging of mediastinal, abdominal, and spinal components. MRI and/or CT scans enable evaluation of the cranial and caudal extent of the cyst. Upper gastrointestinal contrast studies and endoscopy may be necessary in selected cases. The possibility of an additional intestinal duplication must be considered with foregut duplications.

Magnetic resonance cholangiography or, in older children, endoscopic retrograde cholangiopancreatography (ERCP) are valuable in the assessment of pancreatic and selected duodenal duplications. Magnetic resonance angiography may assist in planning surgery for large retroperitoneal lesions. Pelvic duplications are best imaged by CT or MRI in conjunction with a contrast enema, fistulogram, endoscopy, and urinary tract sonography.

28.4 Surgical Treatment

Alimentary tract duplications are best treated by *early complete excision*. The surgeon must be familiar with the regional anatomy and the range of available operative techniques. Rarely, complete resection is unduly hazardous, so alternative techniques such as mucosal stripping or fenestration are necessary. Incomplete excision may lead to late, potentially fatal, complications such as cyst recurrence, infection, meningitis (neurenteric cysts), gastrointestinal bleeding, and perforation. At operation, a careful search is made for additional duplications and associated malformations. Asymptomatic simple appendiceal duplications found in association with cloacal or bladder exstrophy should be preserved for use in subsequent reconstructive surgery. Patients should receive prophylactic intravenous broad spectrum antibiotics at the induction of anaesthesia.

28.4.1 Oesophageal Duplications

These are usually intramural cystic lesions, more often related to the right side of the oesophagus. They may or may not communicate with the oesophageal lumen. A nasogastric tube in the oesophagus may assist with identification.

Cervical oesophageal duplications can be removed via a supraclavicular approach. The neck is extended with a sandbag between the shoulders. Care must be taken to avoid injury to structures such as the recurrent laryngeal nerve. The cyst is fully mobilized and excised by dissecting close to its wall.

Intrathoracic oesophageal duplication cysts can be approached via a transpleural posterolateral thoracotomy (typically right-sided). Excision of the cyst is achieved by mobilizing the cyst and either excising it intact or transecting it close to the wall of the oesophagus (Fig. 28.2) and removing any residual cyst mucosa (Fig. 28.3). Injury to the vagus and phrenic nerves and the thoracic duct must be avoided. Any communication with the oesophageal lumen should be closed with sutures. The residual muscular defect in the oesophageal wall is repaired and can be buttressed using the muscular fringe from the duplication (Fig. 28.4). Complete mucosal excision is essential to avoid recurrence; marsupialization and laser ablation of residual cyst mucosa is inadequate. Closing the muscular defect in the oesophageal wall is necessary to avoid the potential development of an oesophageal diverticulum. The integrity of the oesophageal mucosa can be checked before closing the muscle layer by insufflating air through a nasogastric tube or a narrow-calibre, flexible, fiberoptic endoscope. Occasionally, the presence of an adjacent oesophageal stricture or ulcer will require a segmental oesophageal resection. The chest is closed and pleural drainage is not usually necessary.

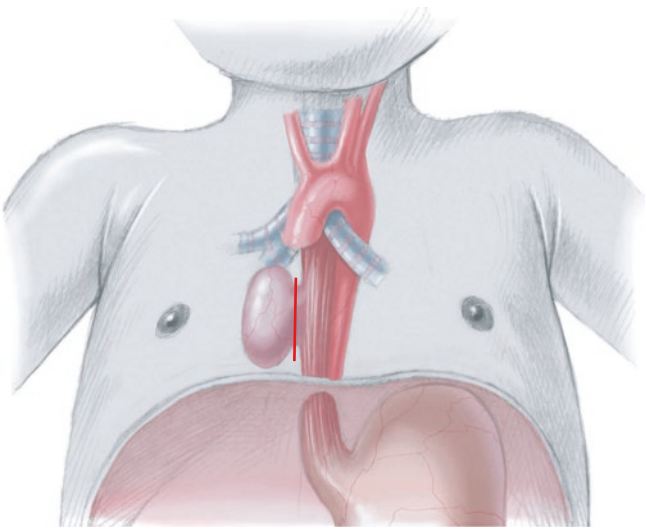

Fig. 28.2 Excision of an oesophageal duplication cyst

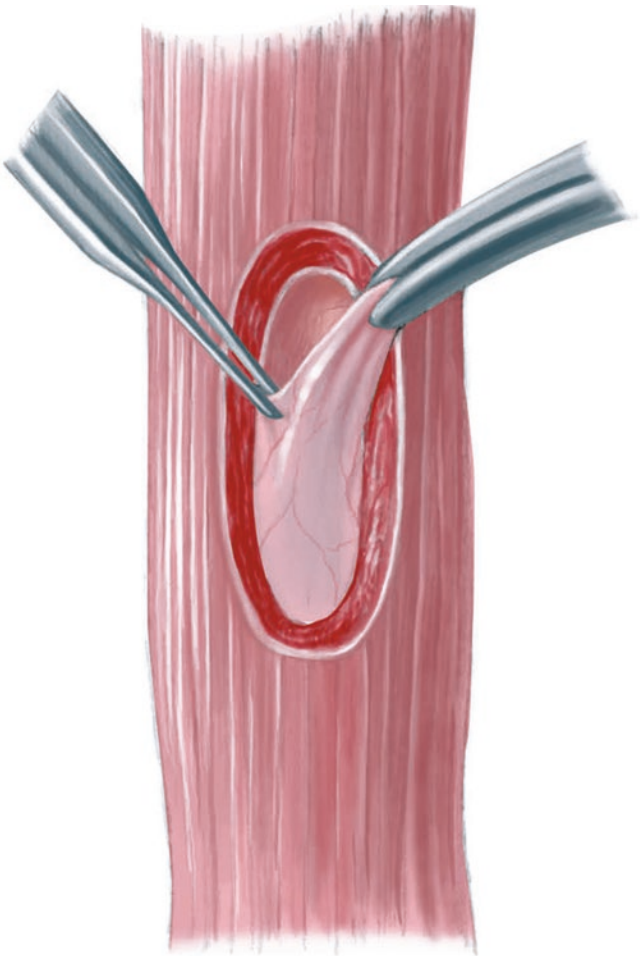

Fig. 28.3 Removal of the cyst's residual mucosal lining adherent to the wall of the oesophagus

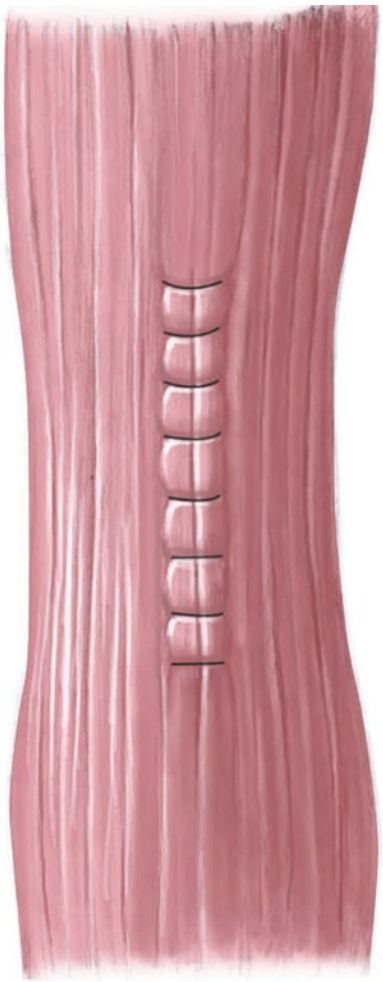

Fig. 28.4 Closure of the oesophagus

Video-assisted thoracoscopic resection of a thoracic oesophageal duplication is now the preferred technique if feasible, as it enables a faster recovery with reduced postoperative discomfort and improved cosmesis. Single lung ventilation or use of a bronchial blocker allows collapse of the ipsilateral lung to facilitate surgical exposure. In the left lateral decubitus position, a 5- or 10-mm 30° telescope is introduced under direct vision just inferior and anterior to the angle of the scapula using gentle blunt dissection. An insufflation pressure of about 4 mm Hg is usually safe and effective. Two or more 5-mm instrument ports are subsequently sited on either side of the camera port to allow for triangulation of dissecting instruments (graspers, dissectors, hook electrocautery, etc.) (Fig. 28.5). Decompression of the cyst may be necessary to improve working space. After dissection, if the cyst remains intact, it is aspirated to enable removal through one of the port sites. Before removing the camera, the lung is re-expanded and checked for air leaks. It is not necessary to leave a chest drain unless there are concerns about an air leak or the integrity of an oesophageal repair.

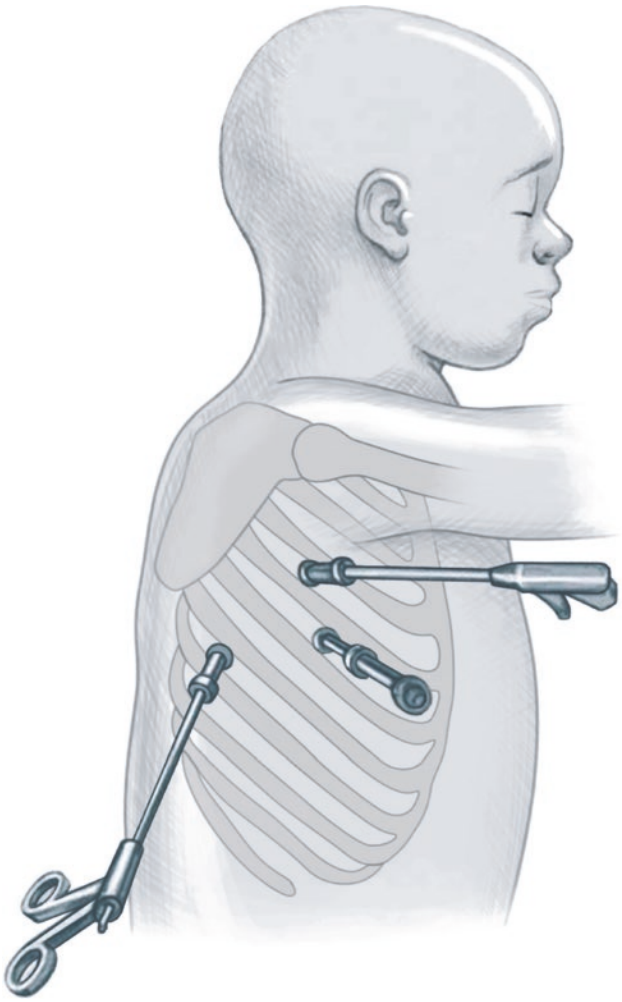

Fig. 28.5 Location of ports for thoracoscopic resection of a thoracic oesophageal duplication

28.4.2 Thoracoabdominal Duplications

A thoracoabdominal duplication usually descends to the right of the oesophagus (but slightly separate from it), in the posterior mediastinum. It communicates through the diaphragm with the stomach, duodenum, pancreas, jejunum, or ileum. A detailed preoperative assessment of the mediastinal, abdominal, and potential spinal components of the duplication is essential. These lesions are best approached by separate thoracic (posterolateral thoracotomy) and abdominal incisions (Fig. 28.6). Occasionally, a laminectomy is also necessary to deal with an intraspinal component of the duplication. This should be anticipated from careful preoperative imaging; multidisciplinary planning involving neurosurgical expertise is advisable in such cases. A staged approach, excising the thoracic and abdominal component of the thoracoabdominal duplication sequentially, is best avoided, because an undrained segment of the duplication may cause serious complications.

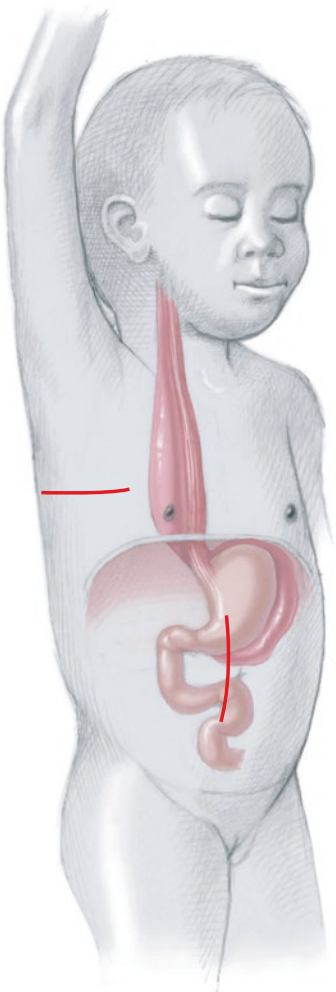

Fig. 28.6 Location of incisions for excision of a thoracoabdominal duplication

Complete excision of the duplication may involve dissecting its upper extremity free from a bony vertebral attachment. This may require a gouge or chisel. Some thoracoabdominal duplications are complicated by peptic ulceration, which can erode into the lung parenchyma and cause haemoptysis. In such cases, excision may require oversewing the fistula or (rarely) a pulmonary lobectomy. The duplication is traced distally and usually passes behind the diaphragm, where it may become tenuous. The duplication is then pulled up into the chest and divided between ligatures before closing the chest with pleural drainage and proceeding to laparotomy. Alternatively, the thoracic component of the cyst is posted through the diaphragmatic defect into the abdomen, where it is subsequently retrieved. The abdominal portion most often appears as a tubular lesion communicating with the jejunum, but it may end blindly along the greater curve of the stomach or communicate with the stomach, pancreas, or duodenum. It usually can be excised without difficulty.

28.4.3 Gastric Duplications

Cystic duplications of the stomach do not usually communicate with the gastric lumen; very rarely, they may communicate with the pancreas. Most greater curve or pyloric duplication cysts can be completely excised by dissecting the cyst off the gastric submucosa and repairing the residual seromuscular defect in the stomach with absorbable sutures. Gastric mucosal integrity can be checked by insufflating air via a nasogastric tube.

Small gastric duplications are sometimes more simply excised by a wedge resection of the cyst and a margin of stomach followed by a two-layered gastric closure (Fig. 28.7). Extensive tubular duplications of the greater curve of the stomach are best treated by partial resection, stripping off any residual mucosal lining attached to the stomach, and repair. A less optimum approach is to divide the septum separating the tubular duplication from the gastric lumen with linear staplers introduced via proximal and distal gastrotomies (Fig. 28.8). This division should be as complete as possible to avoid leaving a diverticulum. Since the mucosal lining of the duplication is not removed, a risk of long-term complications remains.

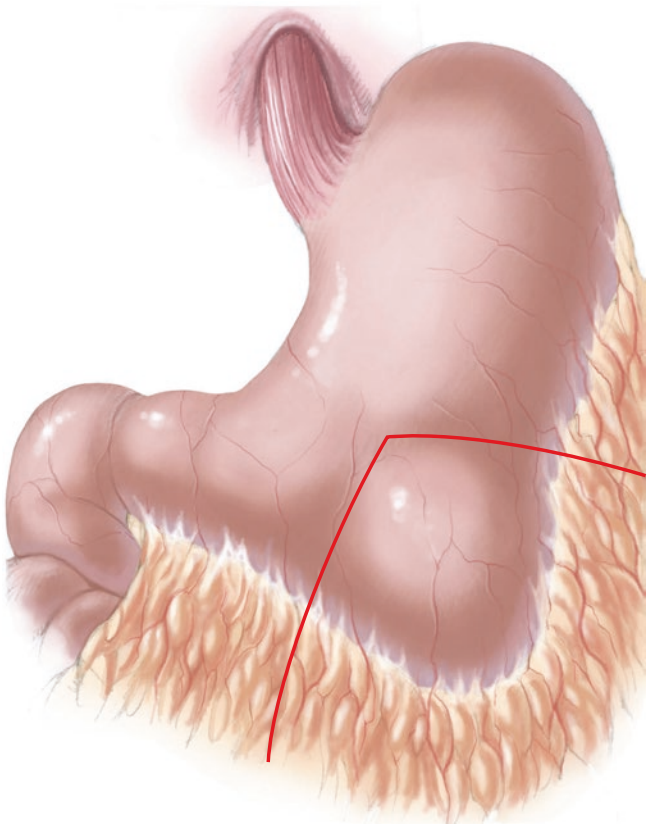

Fig. 28.7 Wedge resection of a gastric duplication cyst

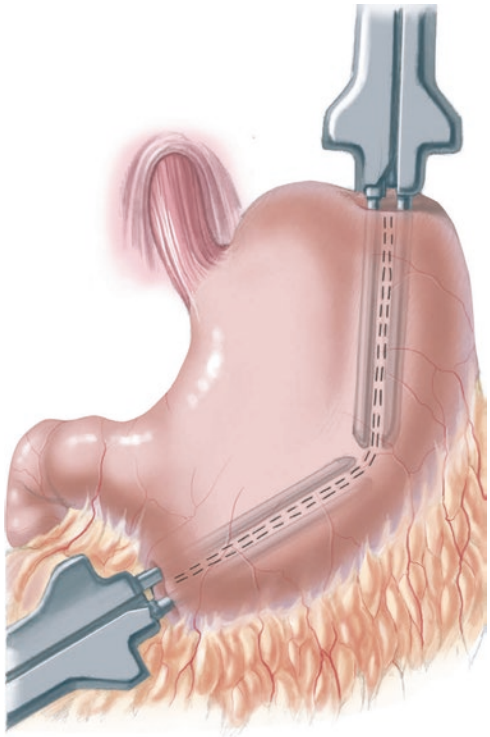

Fig. 28.8 Using linear staplers to divide the septum separating a greater curve gastric tubular duplication from the gastric lumen

Laparoscopic excision of smaller gastric duplication cysts is an alternative technique, and can be achieved more easily with a linear stapling device.

28.4.4 Duodenal Duplications

Some duodenal duplications are simple cystic lesions that can be excised easily without risk of injury to pancreatic or bile ducts. Others, on the medial aspect of the second or third parts of the duodenum, are potentially complex and may communicate with the pancreatic and/or common bile duct. Previous pancreatitis or peptic ulceration secondary to heterotopic gastric mucosa can compound the surgical difficulties. Facilities for intraoperative cholangiopancreatography should be available.

In open surgery, the duodenum is approached via a right upper quadrant incision and "Kocherised" to elevate it into the wound. The adjacent peritoneal cavity is packed off with swabs soaked in dilute aqueous povidone iodine, and an oblique incision is made in the lateral duodenal wall to expose the medially situated duplication cyst and major duodenal papilla. If needle aspiration of the cyst yields bile, an on-table cholangiogram should be performed to delineate the bile duct connection. It may be useful to perform a cholecystectomy and pass a fine probe or catheter distally through the cystic duct down into the duodenum.

Surgical management is dictated by the anatomy of the duplication. Ideally, the cyst should be completely excised, with division and ligation of any ductal communication (Fig. 28.9). Occasionally, partial excision with mucosectomy of the remaining part of the cyst is appropriate. Wide fenestration into the duodenal lumen is another option, but only if heterotopic gastric mucosa is excluded by intraoperative biopsy and the cyst window is made sufficiently large to allow free dependent drainage without the tendency to form a cul-de-sac. The edges of the fenestration must be carefully oversewn to achieve haemostasis, avoiding injury to adjacent ducts.

Selected duodenal duplications are amenable to minimally invasive techniques, but the potential complexity of cysts in the head of the pancreas or around the major duodenal papilla should prompt caution. Endoscopic marsupialisation of duodenal duplication cysts has been reported, but the paucity of long-term results and the potential later risk of malignant degeneration in adults is a concern.

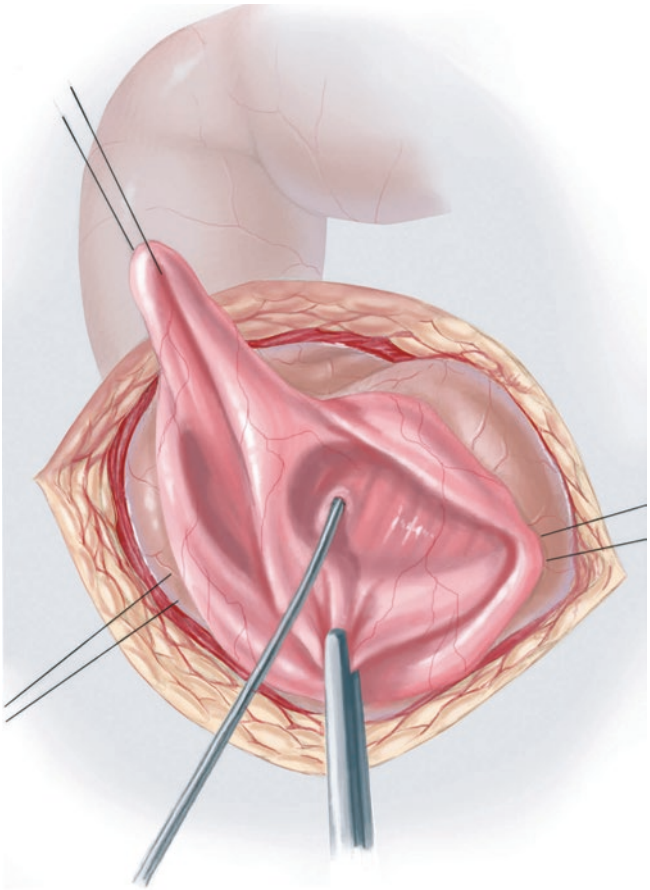

Fig. 28.9 Excision of duodenal duplication. Stay sutures have been placed on the collapsed duplication cyst prior to its resection and a probe inserted into the major duodenal papilla to identify the bile and pancreatic ducts

28.4.5 Pancreatic Duplications

Rarely, a duplication cyst is located exclusively within the head of the pancreas and can cause recurrent pancreatitis. Surgical approaches include *complete* local excision with Roux loop drainage of the residual cavity or a Whipple-type pancreatoduodenectomy. Roux loop drainage of the cyst alone is sometimes sufficient but recurrent pancreatitis is a risk if the mucosal lining of the cyst has not been destroyed by inflammation. If the duplication lies in the tail of the pancreas, enucleation may be possible (with care to oversew any ductal communication) or it can be excised by a distal pancreatectomy with splenic preservation (laparoscopic or open).

28.4.6 Small Bowel Duplications

Cystic lesions of the ileum or jejunum are the most common duplications, and complete excision is relatively straightforward using open or laparoscopic-assisted techniques. The continuity of the muscle coat of the normal bowel and duplication cyst usually make excision of the cyst alone unnecessarily difficult compared with simple segmental resection of the bowel (Fig. 28.10). For localised lesions, the duplication should be resected with the adjacent intestine, after ligation/coagulation and division of relevant mesenteric vessels. If the bowel is obstructed, gentle atraumatic clamps placed across the intestine (but not the mesentery) will help prevent contamination. Before dividing the bowel with needle cautery, the surgical field should be protected by surrounding gauze swabs soaked in dilute aqueous povidone iodine. End-to-end anastomosis is performed using a single layer of interrupted, extramucosal absorbable sutures (Figs. 28.11 and 28.12). Laparoscopic assisted resection can be achieved by preliminary aspiration of the cyst and exteriorisation of the affected segment of bowel.

Short tubular duplications (Fig. 28.13) may be excised in continuity with the parallel segment of small bowel. Care is needed to achieve complete excision of the duplication at the proximal and distal margins of the tubular duplication; the

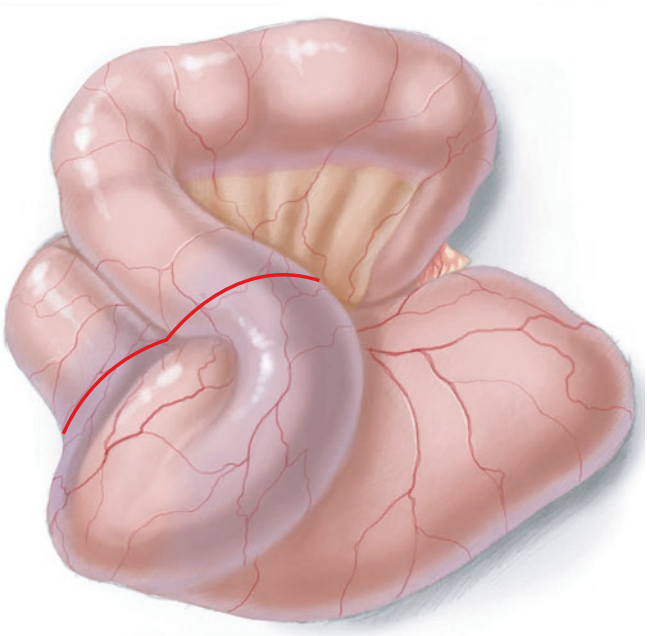

Fig. 28.10 Segmental resection of a small bowel duplication cyst

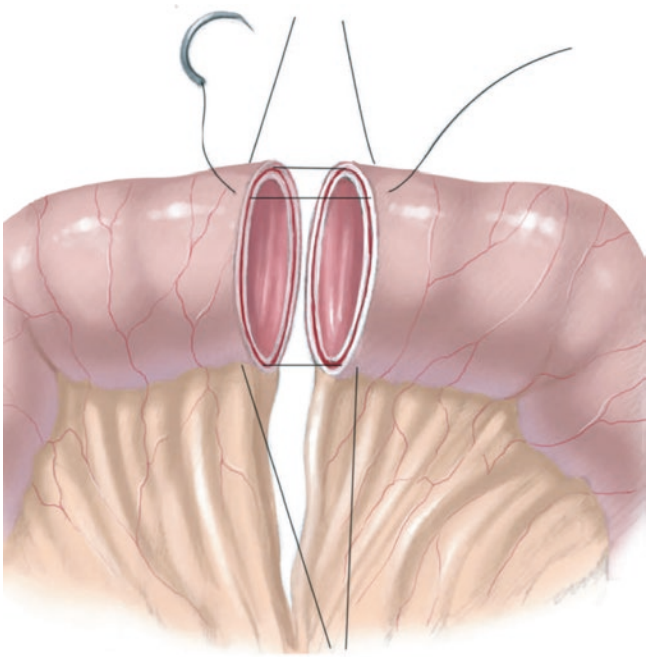

Fig. 28.11 End-to-end anastomosis with interrupted extramucosal sutures after a localised small bowel resection for a duplication cyst

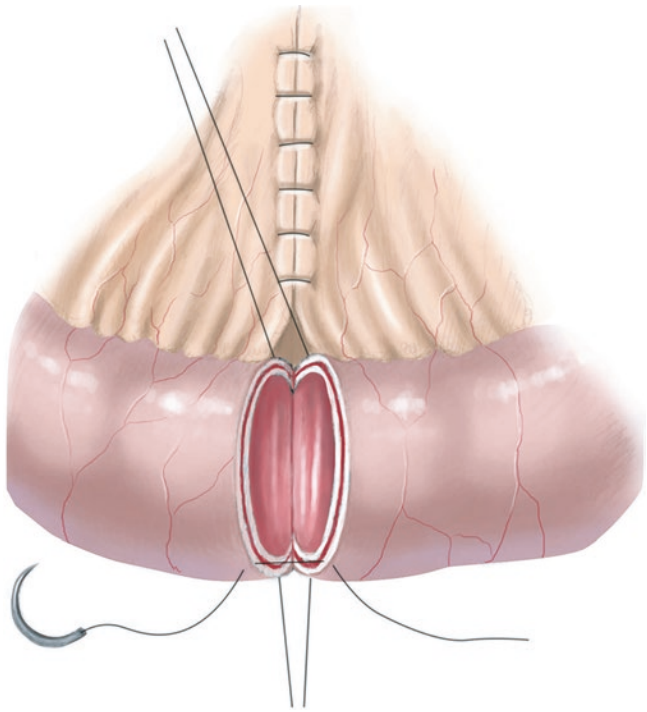

Fig. 28.12 Completion of the end-to-end anastomosis after bowel resection

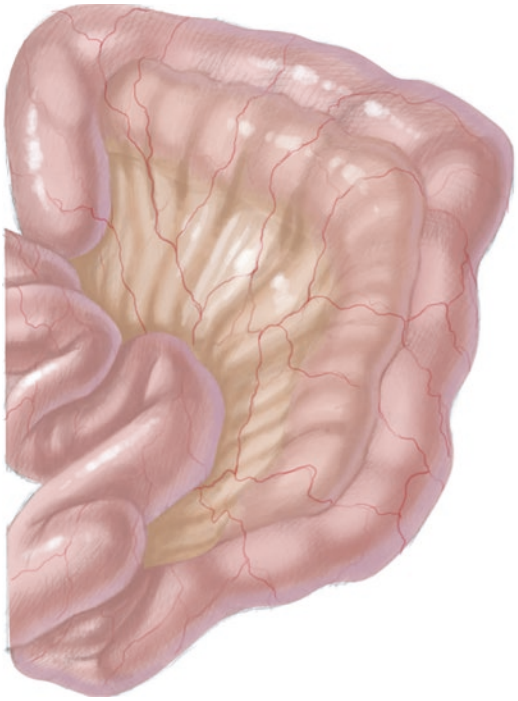

Fig. 28.13 A short tubular small bowel duplication

exact point of termination of the duplicated bowel can occasionally be difficult to determine.

Long tubular duplications, where the length of the unaffected small intestine is a potential concern, are more challenging. The tubular duplication runs parallel to the native bowel within the adjacent mesentery. Complete resection of the duplication alone risks rendering the native small bowel ischaemic. In these cases, submucosal resection of the duplication is an alternative but difficult option. The mucosal lining of the duplication is stripped out using a series of longitudinal seromuscular incisions (Fig. 28.14a). Bipolar diathermy and gentle, blunt pledget dissection are useful. The residual seromuscular sleeve of the tubular duplication may be safely left in situ provided haemostasis has been achieved. For tubular duplications within the mesentery but separate from the native intestine, careful separation of the two leaves of the mesentery and division of vessels in one or other leaf only may enable excision of the duplication without jeopardising the blood supply of the native bowel. Whichever technique is used, it is essential to check the viability of the remaining intestine and to resect the entire junction of duplicated and normal bowel (Fig. 28.14b) because heterotopic gastric mucosa is frequently present in tubular intestinal duplications. If there is an associated intestinal malrotation, a Ladd's procedure will be required.

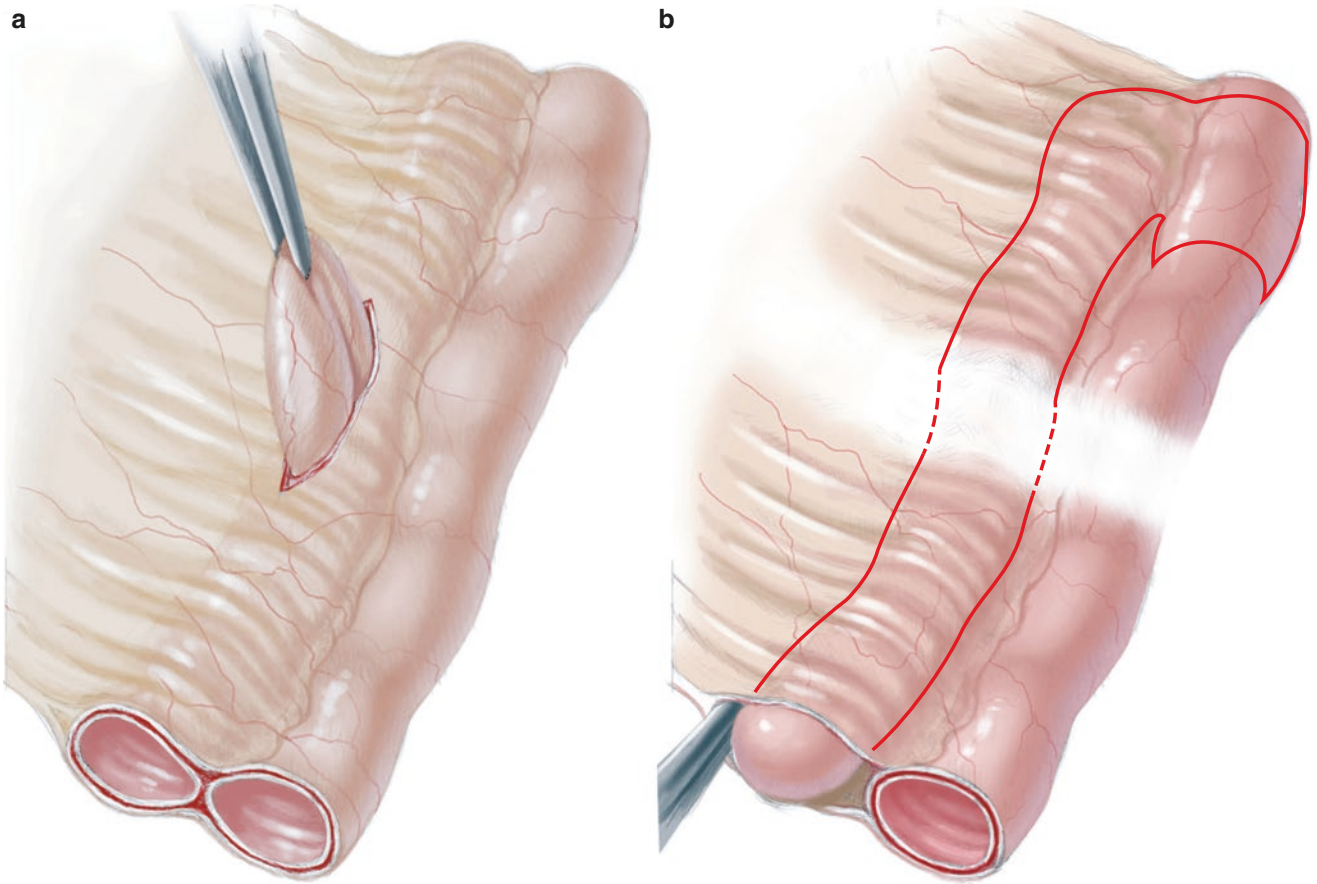

Fig. 28.14 (**a**) Stripping out the mucosal lining of a long tubular small bowel duplication. (**b**) The entire junction of normal and duplicated bowel must be resected

28.4.7 Colonic (and Appendiceal) Duplications

All cystic and most tubular duplications of the colon can be excised by segmental colonic resection (Fig. 28.15). The bowel is reanastomosed with an end-to-end, single-layer, extramucosal suture technique. Total colonic duplications are rare; the duplicated bowel may lie lateral or medial to the normal colon and often has a proximal connection (Fig. 28.16a). Heterotopic gastric mucosa is rarely found, so distal fenestration is possible, which can be performed with a linear stapling device introduced through an enterotomy near the distal margin of the duplication (Fig. 28.16b). The distal end of the septum must be divided completely to avoid leaving a problematic spur. If there is complete hindgut duplication with two perineal openings, then a preliminary double defunctioning colostomy is recommended. The duplicated bowel can later be transected at the level of the rectum and the proximal part anastomosed to the 'normal' rectum, with excision of the mucosa of the redundant distal rectal segment.

Asymptomatic duplications of the appendix found in association with cloacal or bladder exstrophy can be retained for later use in reconstructive surgery.

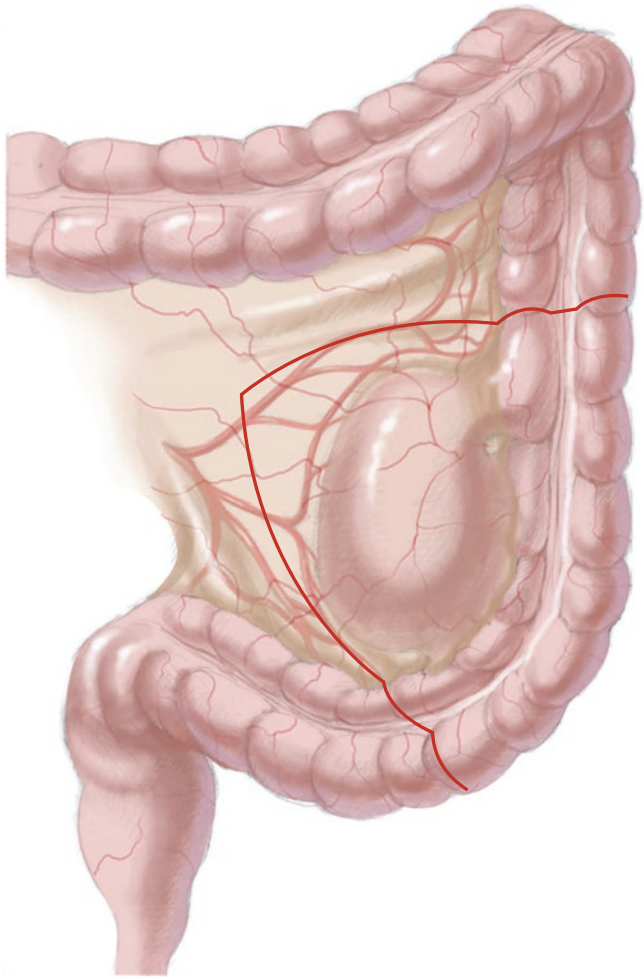

Fig. 28.15 Segmental resection for cystic duplication of the colon

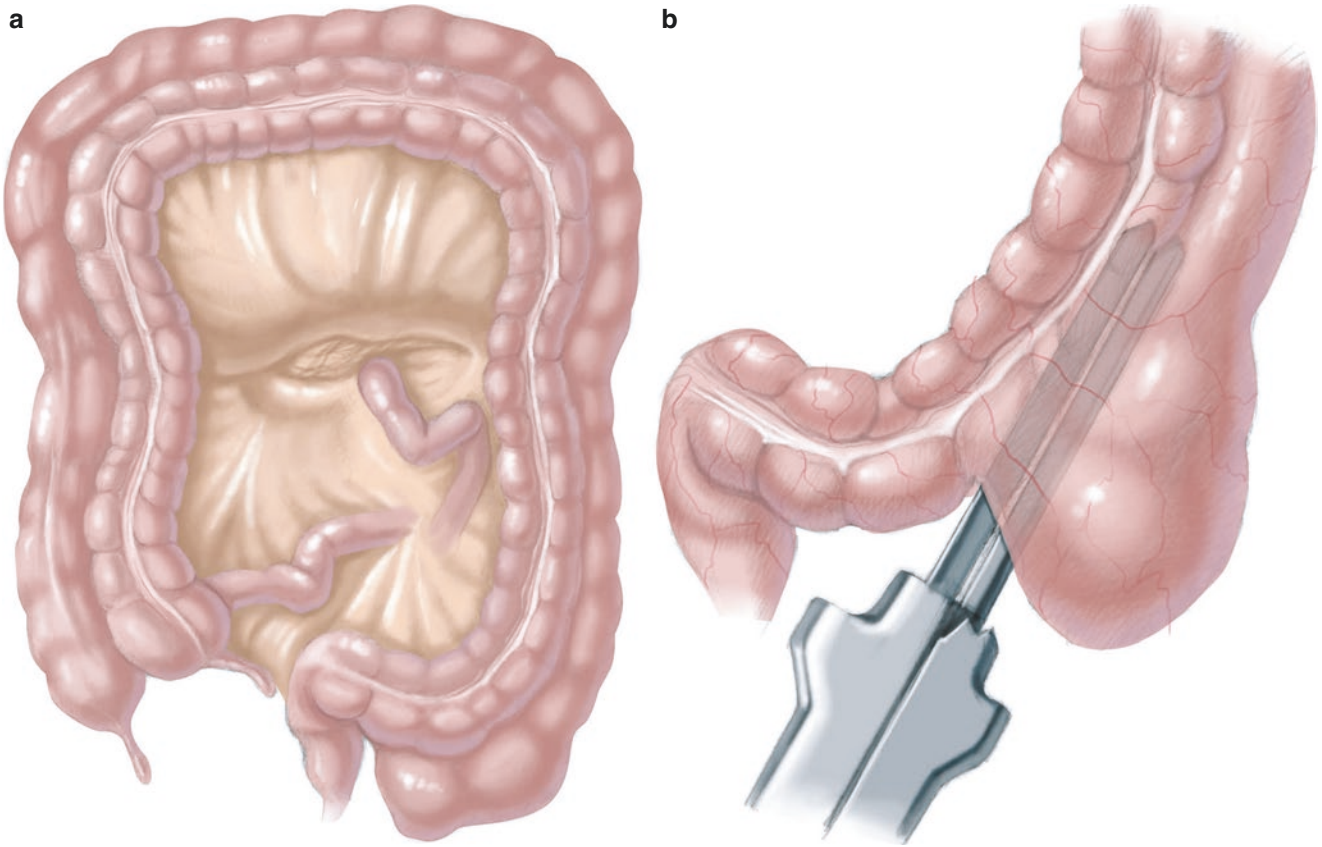

Fig. 28.16 (**a**) Total duplication of the colon and appendix. (**b**) Distal fenestration of total colonic duplication

28.4.8 Rectal Duplications

Rectal duplications often present in the neonatal period as a perineal mucosal swelling (Fig. 28.17) and/or a fistula extending to the perianal skin or anorectum. They may be confused with perianal sepsis. Some are cystic lesions which may or may not have an external or internal communication (Fig. 28.18). These are usually found in the retrorectal space and do not communicate with the urinary tract. Cystic rectal duplications must be distinguished from cystic sacrococcygeal teratoma, tailgut cyst, and an anterior meningocele. When associated with an anorectal malformation and sacral anomalies, the cyst may be part of the Currarino syndrome. Rarely, a rectal duplication cyst presents as rectal prolapse in an infant. Rectal duplications may be excised by one of several approaches.

Small submucosal cysts can be excised endorectally. As the anus is dilated with an anal retractor, the cyst bulges forward. The rectal mucosa over the cyst is incised and, working within the submucosal plane of the rectum, the cyst is gradually dissected free (Fig. 28.19). Local injection of 1:200,000 adrenaline and bipolar diathermy may assist the dissection. The cyst is best left intact until nearing complete excision, as the wall tension can facilitate the dissection. The incision in the rectal mucosa is repaired with interrupted absorbable sutures.

The posterior sagittal approach provides excellent exposure of the retrorectal space for excision of tubular and larger cystic duplications (Fig. 28.20). With attention to bowel preparation and antibiotic prophylaxis, a covering colostomy can be avoided. Infected rectal duplication cysts are best treated by preliminary perineal drainage followed by resection once the inflammation has settled. Rectal duplications should be completely excised because of a risk of late malignant degeneration.

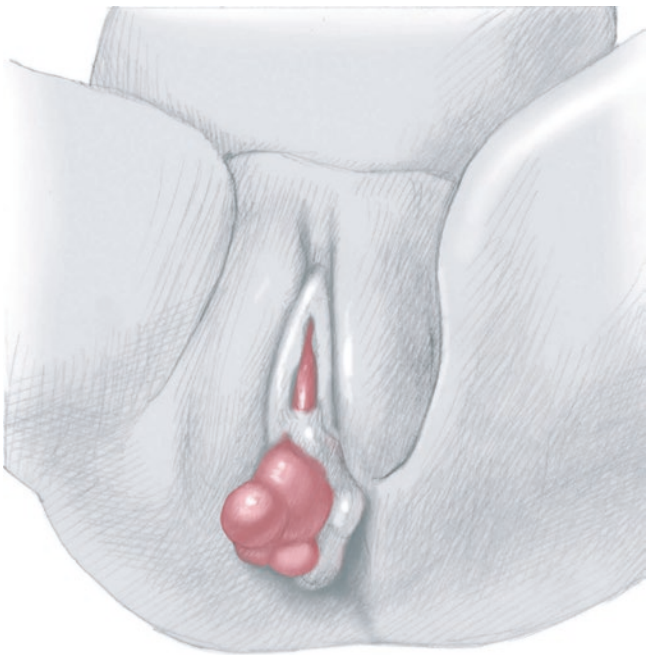

Fig. 28.17 Rectal duplication

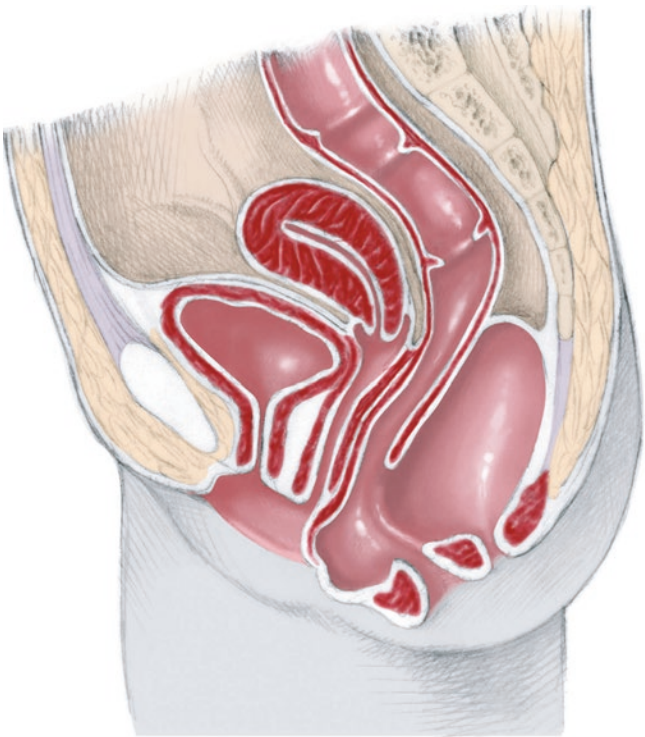

Fig. 28.18 Cystic retrorectal duplication cyst with internal communication with the native rectum and a postanal fistula

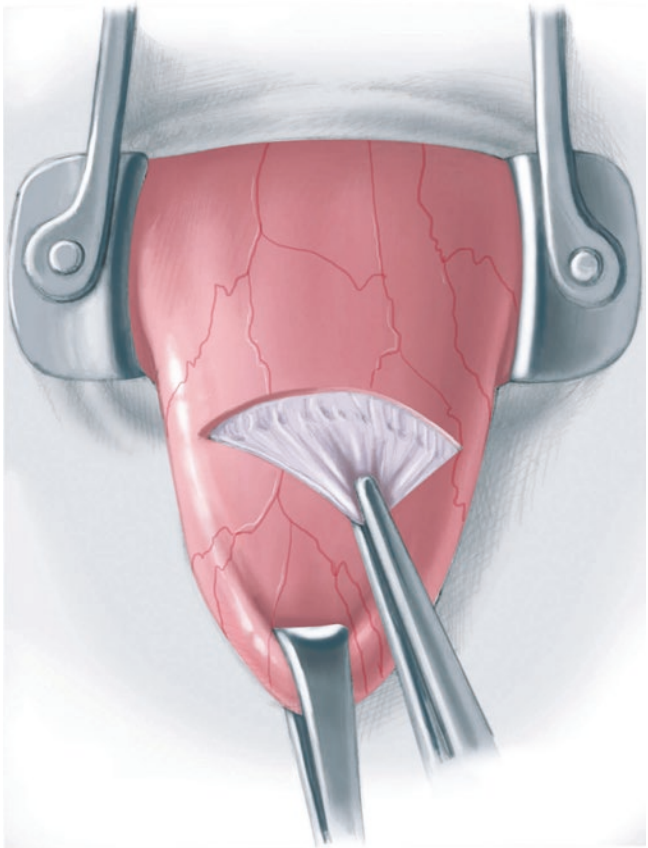

Fig. 28.19 Endorectal dissection of a submucosal cystic duplication of the rectum

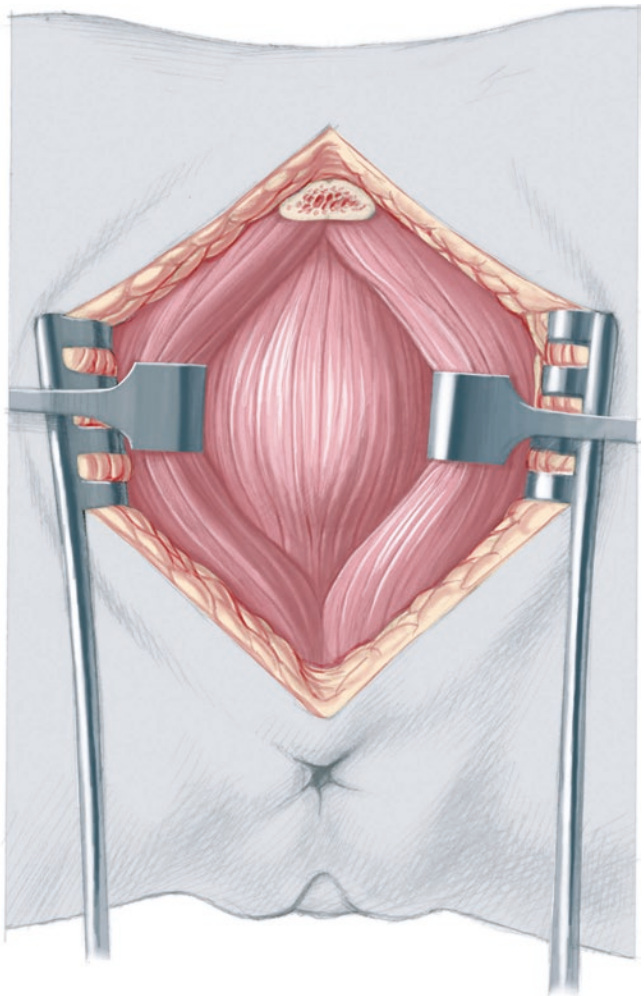

Fig. 28.20 Posterior sagittal approach to a retrorectal duplication cyst

28.4.9 Other Duplications

Lingual duplication cysts are most often located in the anterior two thirds of the tongue and are best treated by complete surgical excision, usually via an intraoral, sublingual approach. *Retroperitoneal* duplication cysts may be very large and are frequently adherent to retroperitoneal structures, including the pancreas. Care is required during surgery to avoid injury to mesenteric and renal vessels and nearby viscera. *Intradural* or *extradural* duplication cysts in the vertebral canal are best managed in conjunction with a neurosurgeon.

28.5 Postoperative Care and Complications

All excised duplications should be sent for pathologic examination to assess the completeness of the excision, to document the presence of heterotopic mucosa, and to exclude neoplasia. Specific aspects of postoperative clinical management are determined by the site of the duplication cyst and the complexity of the surgical procedure.

Surgical complications are related to the location and size of the cyst, the presence of a communication with the gastrointestinal tract or spinal canal, the existence of heterotopic gastric mucosa, and potential injury to nearby structures.

Conclusions

Several steps are key in the successful surgical management of gastrointestinal duplications:

- Thorough understanding of the variable pathology of these lesions
- Careful preoperative imaging of the duplication
- Awareness of possible associated pathology
- Operative planning
- Complete excision when feasible
- An understanding of alternative techniques if excision is deemed too hazardous

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Short Bowel Syndrome

29

Michael E. Höllwarth

The term "short bowel" was defined by Rickham in 1967 as a small intestinal remnant of 75 cm or less in the newborn, which equals 30% of normal small bowel length in that age group. A more functional description, preferred by most authors, defines a "short bowel syndrome" (SBS) as a state of significant maldigestion and malabsorption due to an extensive loss of functional absorptive intestinal surface area.

The prevalence of SBS has been increasing over the past two decades because of the enormous progress in intensive care of babies with either severe acquired intestinal diseases (e.g., necrotizing enterocolitis and volvulus) or congenital malformations leading to the SBS (e.g., multiple intestinal atresia). Rarely, SBS is caused by a genetically determined disease such as a congenital short bowel or a total intestinal aganglionosis.

Following extensive loss of small bowel, the symptoms of an individual baby depend on the absorptive capacities of the intestinal remnants. Resection of the jejunum is well tolerated owing to the enormous adaptive capacity of the ileum, the intact enterohepatic circulation of bile salts, and the preserved absorption of fat-soluble vitamins and vitamin B12. In contrast, if the entire ileum is lacking, absorption of nutrients is significantly more difficult because of the limited intestinal adaptation capacities of the jejunum. Nonabsorbed intestinal contents, including bile acids, spill over into the colon and may cause significant diarrhea. Furthermore, loss of the ileum leads to a reduction of the bile salt pool, malabsorption of fat and fat-soluble vitamins, and a vitamin B12 deficiency.

The pathophysiological process that follows an extensive loss of small bowel is called *intestinal adaptation*. It includes (1) morphological changes leading to an increase of absorptive surface area, (2) functional changes resulting in augmentation of the absorptive capacity of the remaining enterocytes, and (3) increased intestinal diameter with a concomitant reduction of motility, thereby slowing down the intestinal transit time of chyme.

The presence of intraluminal food is the most important driving force for intestinal adaptation. Enteral nutrients stimulate gastrointestinal secretions and hormones that are known to exert trophic effects on the mucosa. Recent evidence suggests that glucagon-like peptide 2, human growth hormone, epidermal growth factor, and insulin-like growth factor I may play an important role in the process of intestinal adaptation.

Surgery is indicated only for selected patients with any one of three conditions: (1) *when the absorptive surface area is definitely too small* to allow enteral feeding; (2) when *severe dysmotility in grossly dilated loops* entails stagnation of chyme; and (3) when *intestinal transit time is too fast* to allow sufficient absorption of nutrients. In the first group of patients, intestinal transplantation is the mainstay of surgical therapy. In the second group, peristalsis can be improved by intestinal tapering or tapering and lengthening. In the third group, antiperistaltic segments, colonic interposition, intestinal valves, and/or artificial invagination have been used in selected patients. These surgical techniques are illustrated and described in detail on the following pages (Figs. 29.1, 29.2, 29.3, 29.4, 29.5, 29.6, 29.7, 29.8, 29.9, 29.10, 29.11, and 29.12)

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https://doi.org/10.1007/978-3-662-56282-6_29

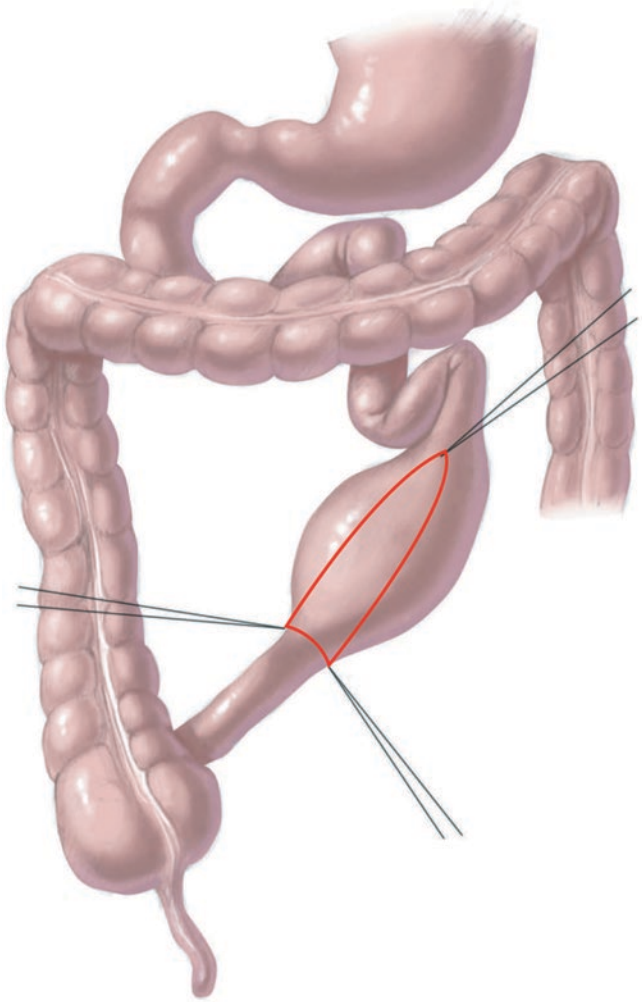

Fig. 29.1 Tapering. In patients with enough absorptive surface area—at least 50 cm with ileocecal valve—the tapering can be performed by resection of a long triangular or elliptical antimesenteric segment. The bowel segment is isolated from surrounding adhesions and the chosen length for the tapering is marked by 5/0 stay sutures, which indicate the lateral margins of the planned triangular resection. One stay suture is located exactly at the antimesenteric line and indicates the end of resection, which is at the tip of the triangle. The resection can be performed by means of a GIA stapler in very large, dilated loops (*see* Chap. 22)

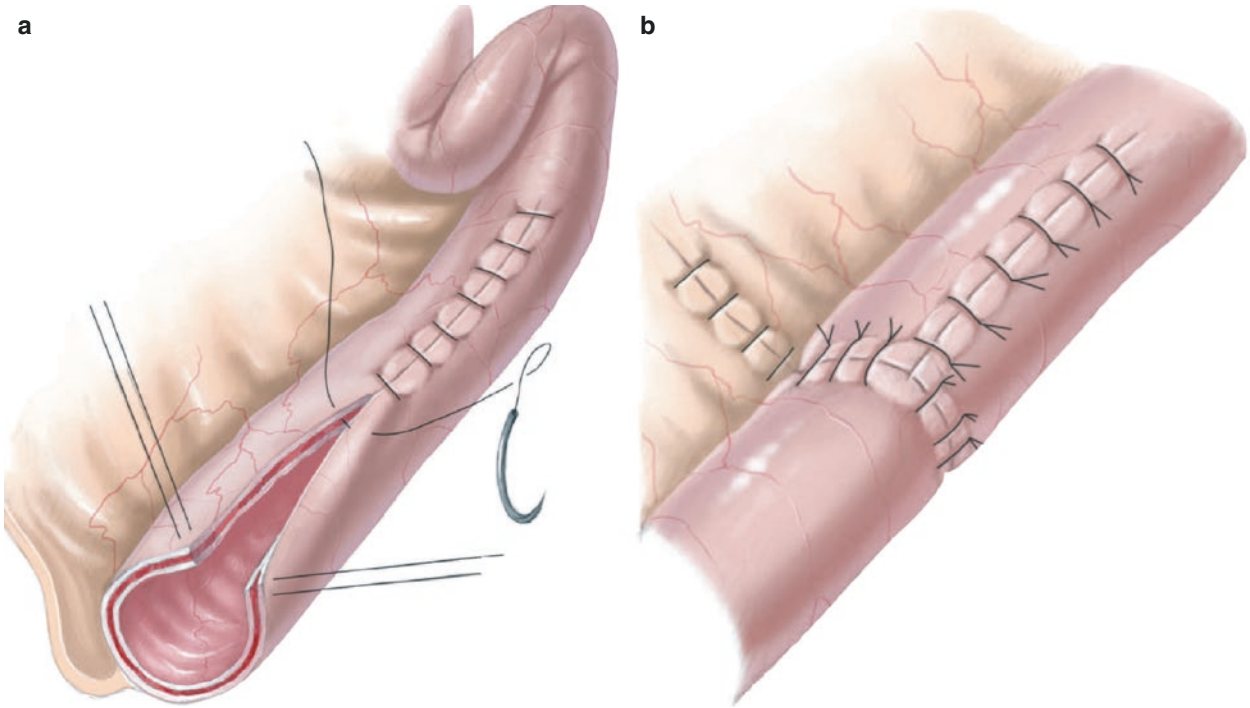

Fig. 29.2 (**a**, **b**) In small babies, we prefer to resect the antimesenteric redundant part with sharp scissors, thus allowing some bleeding from the resected margins, and we try to avoid carefully any disturbance of the local circulation, which inevitably would result if we use cautery. The antimesenteric anastomosis can be accomplished either by a continuous running 6/0 absorbable suture or by 6/0 interrupted sutures

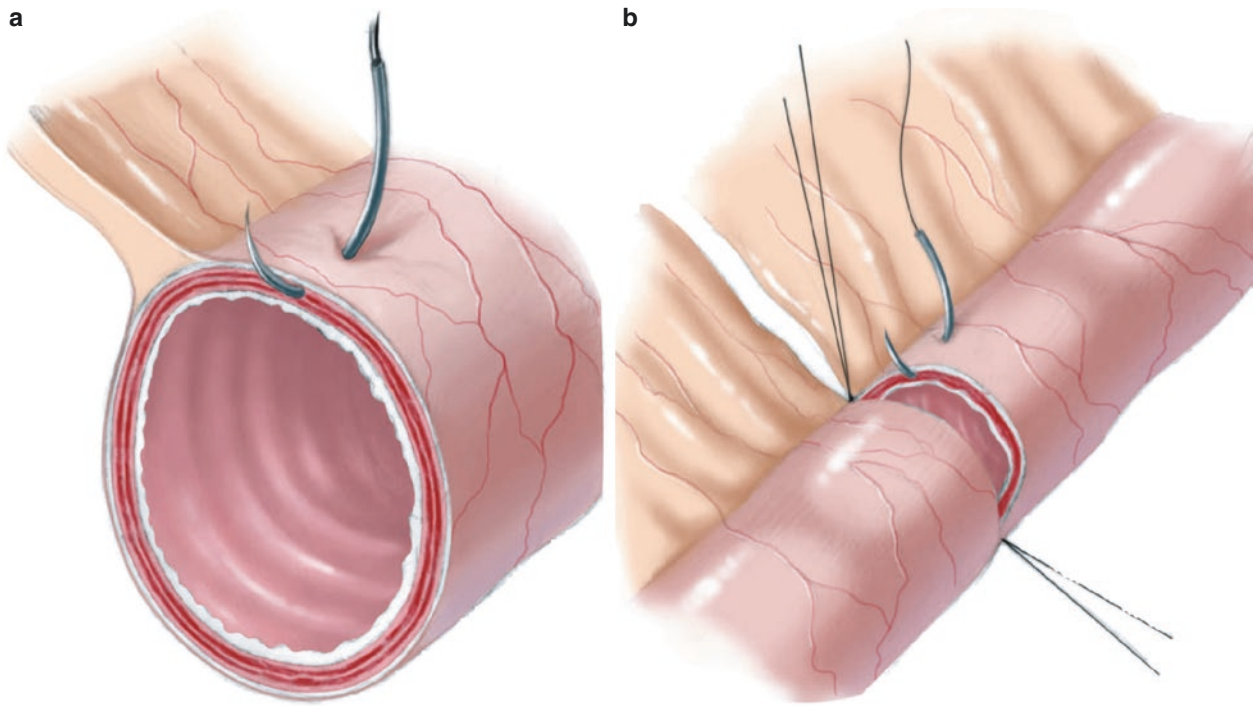

Fig. 29.3 (**a**–**d**) It is important in performing any kind of intestinal anastomosis that the stitches should include only the seromuscular layer, leaving out the mucosal layer. If the stitches take equal amounts of tissue on either side, both segments of the bowel are perfectly adapted and the mucosal edges will lie side by side. Another important part of this technique is avoiding too much tension when tying knots, thus not compromising circulation. This technique was originally described by Halsted in 1912 and has the advantage of preventing mucosal necrosis just under the stitch, thereby supporting the rapid and perfect healing of the anastomosis. If, additionally, an anastomosis to the distal or proximal part of the intestinal tract must be performed, the same surgical technique is useful. First, the two intestinal ends are brought together with two to four 5/0 stay sutures. Thereafter, the anastomosis with interrupted 6/0 stitches begins at the anterior wall. The needle takes a good part of the seromuscular layer on either side. The suture is tied carefully, avoiding any strangulation. The single interrupted sutures are continued in the same way until the whole anterior wall is anastomosed. The posterior wall is sutured in a similar way, after turning around

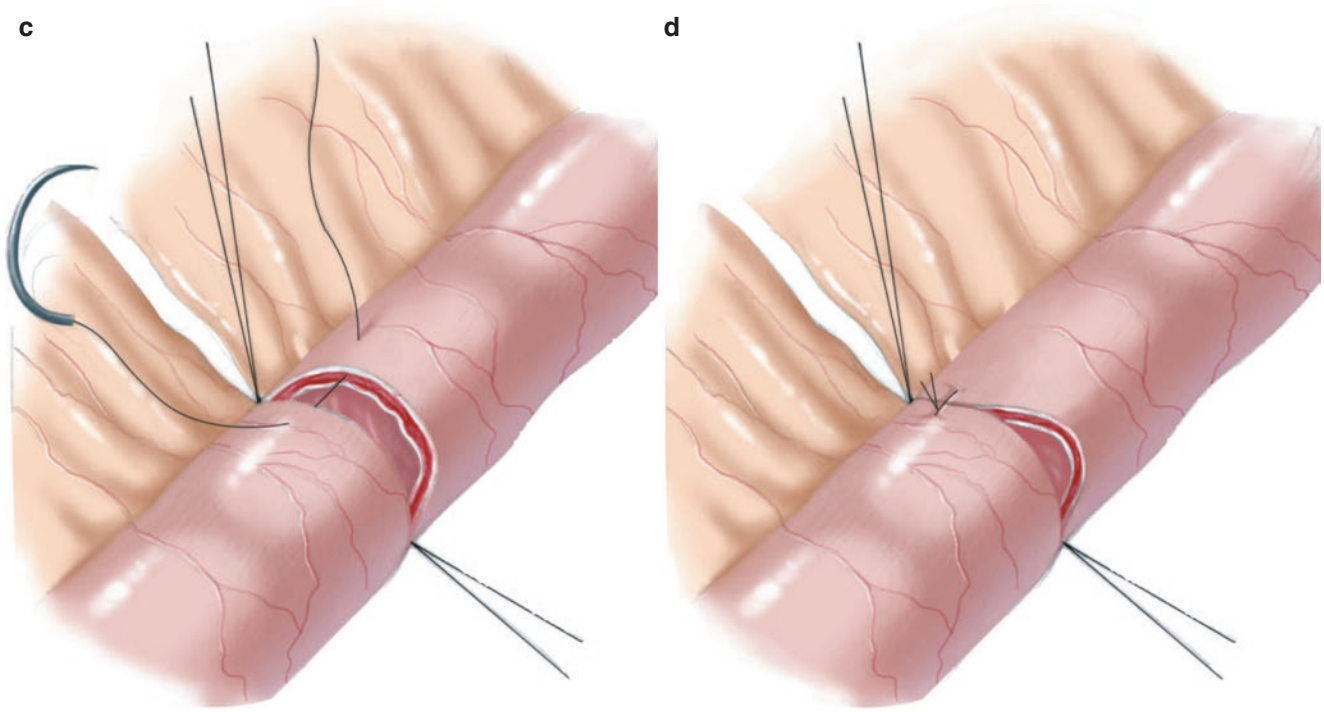

Fig. 29.3 (continued)

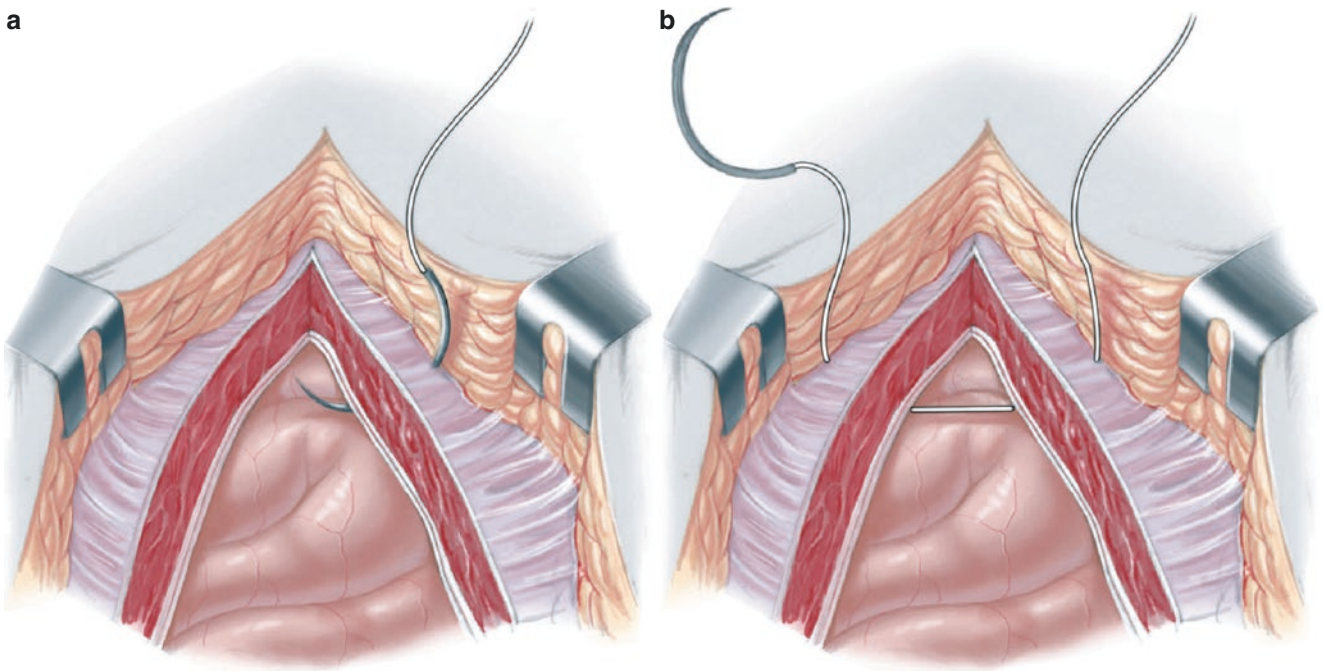

Fig. 29.4 (**a**–**d**) The abdominal wall is closed using 3/0 or 4/0 interrupted single-layer figure-of-eight absorbable sutures. The knot must be tied rather loosely, without strangulating the tissue

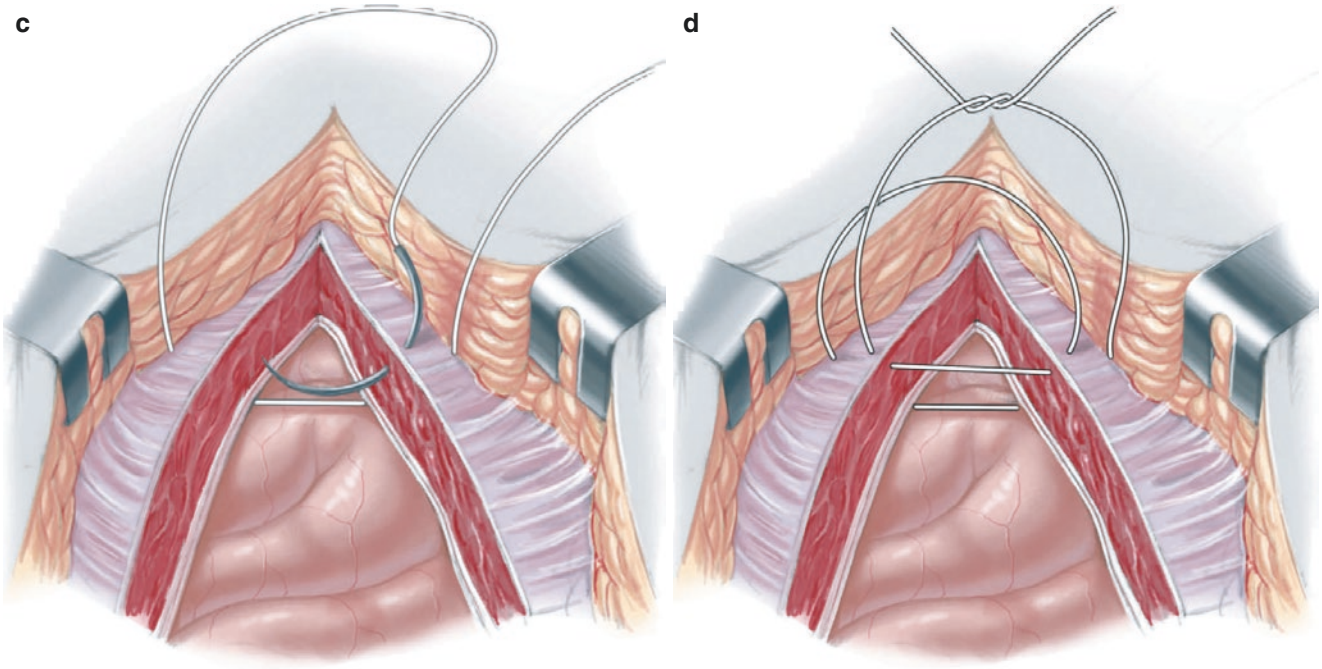

Fig. 29.4 (continued)

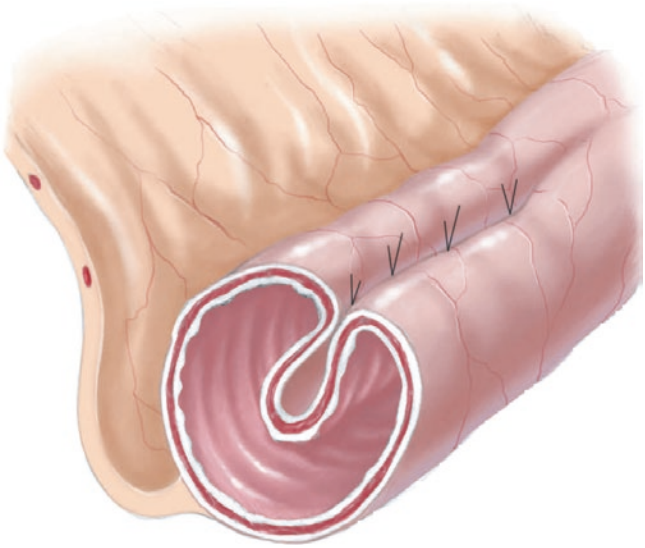

Fig. 29.5 Infolding. This method has the same effect as tapering but saves all existing absorptive surface area. Therefore, it is indicated for patients with a rather short intestinal tract, when it seems advisable not to sacrifice any mucosal surface. The intestinal tract can remain closed in most patients. The intestinal loops that are selected for the infolding method are marked with 5/0 stay sutures on the lateral side and on both ends. The lateral margins are approximated with 5/0 or 4/0 nonabsorbable seromuscular stitches, enfolding the tissue between the margins. It is reported that the plicated segments are prone to breakdown with time, and some authors suggest resecting a serosal strip or a triangular segment on each side in order to support the development of dense adhesions (*see* Chap. 26)

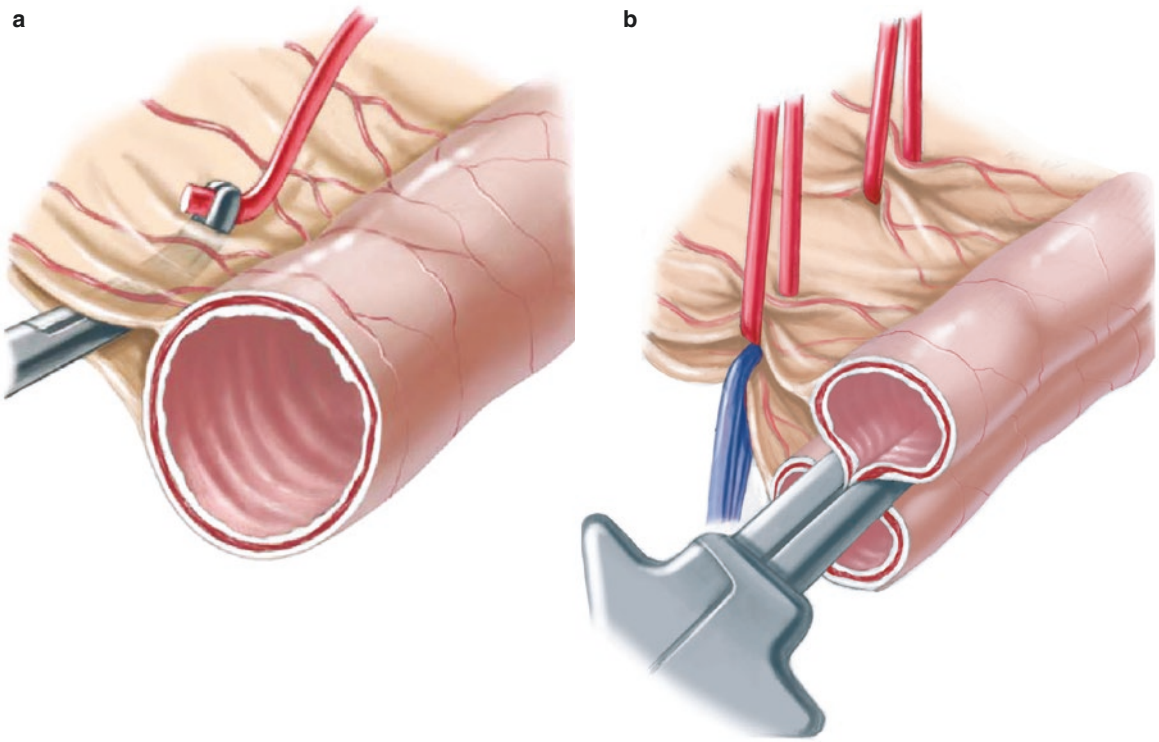

Fig. 29.6 (**a**, **b**) Tapering and lengthening. Bianchi first reported an experimental procedure combining the method of tapering with the use of the redundant tissue for lengthening the bowel. Indications for this method are patients with very short bowel segments but largely dilated loops characterized by inefficient to-and-fro peristalsis, stasis of chyme, and bacterial overgrowth. The technique is based on the fact that the vessels coming from the mesentery are divided extramurally in branches supplying either side of the bowel separately. Therefore, careful longitudinal division of a dilated intestinal segment between the branching vessels results in two intestinal halves with an intact blood supply for each side. The first step of the procedure consists of careful separation of the vessel branching in the right and left group and supplying either part of the selected intestinal tract. The space between the vessel layers can be opened by introducing a haemostat and spreading the two layers. The small vessel groups on each side are secured by fine vessel loops of different colours for the right and left parts. If the space between the branches is large enough that a GIA stapler can be introduced, the intestinal loop can be divided in the midline and anastomosed longitudinally, step by step

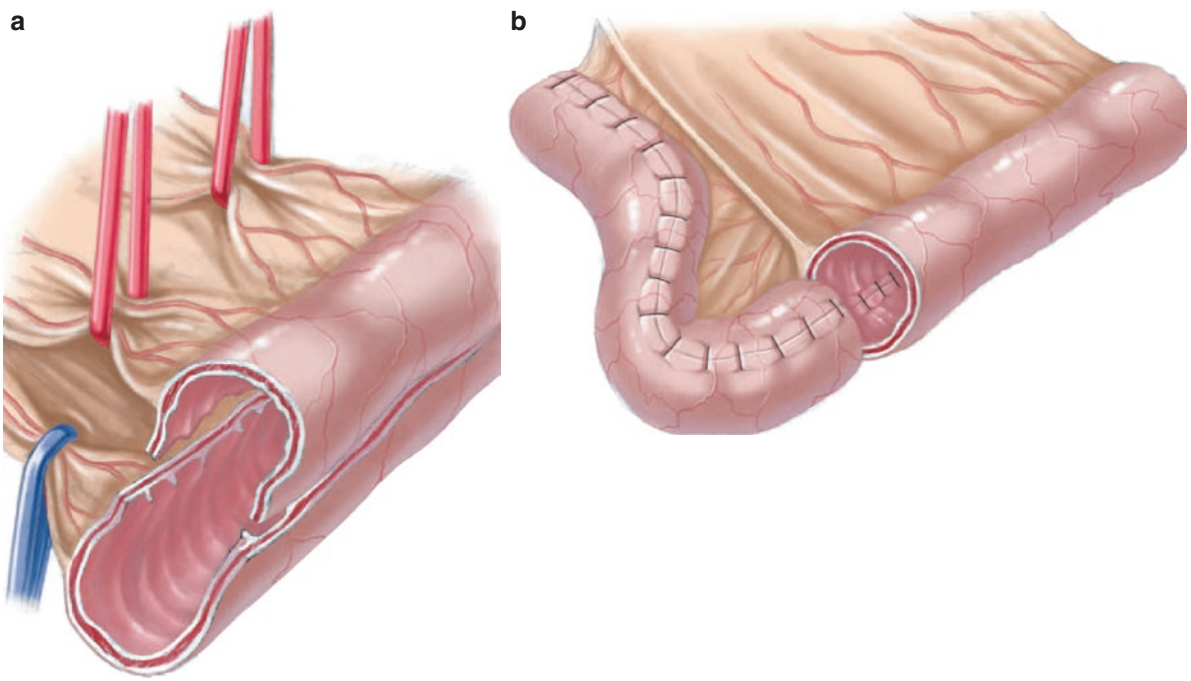

Fig. 29.7 (**a**, **b**) If the space is too small to insert a stapler—which is the case when the bowel is not extremely dilated—then a sharp longitudinal division is our preferred method for tapering and lengthening. We try to avoid any cautery or use it very cautiously. Seromuscular 6/0 absorbable sutures are used for the longitudinal anastomosis of the divided segments of the bowel

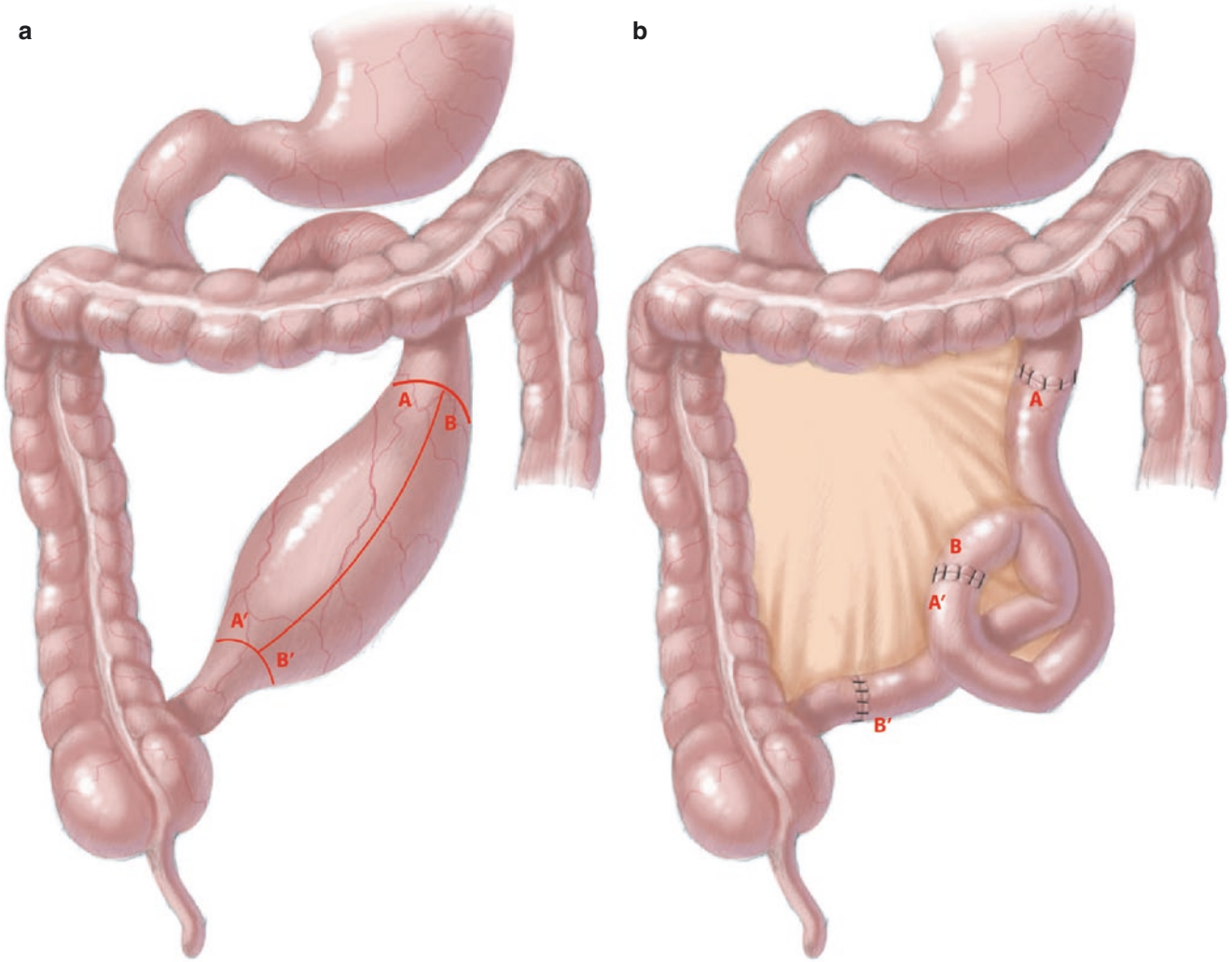

Fig. 29.8 (**a**, **b**) Because both sections of the bowel hang on the same mesenteric segment, a helix-like isoperistaltic anastomosis is easier to perform than an anastomosis with the two segments sliding one on the other. The helix technique avoids traction on the nutrient vessels, which is critical; necrosis of the divided segments has been reported

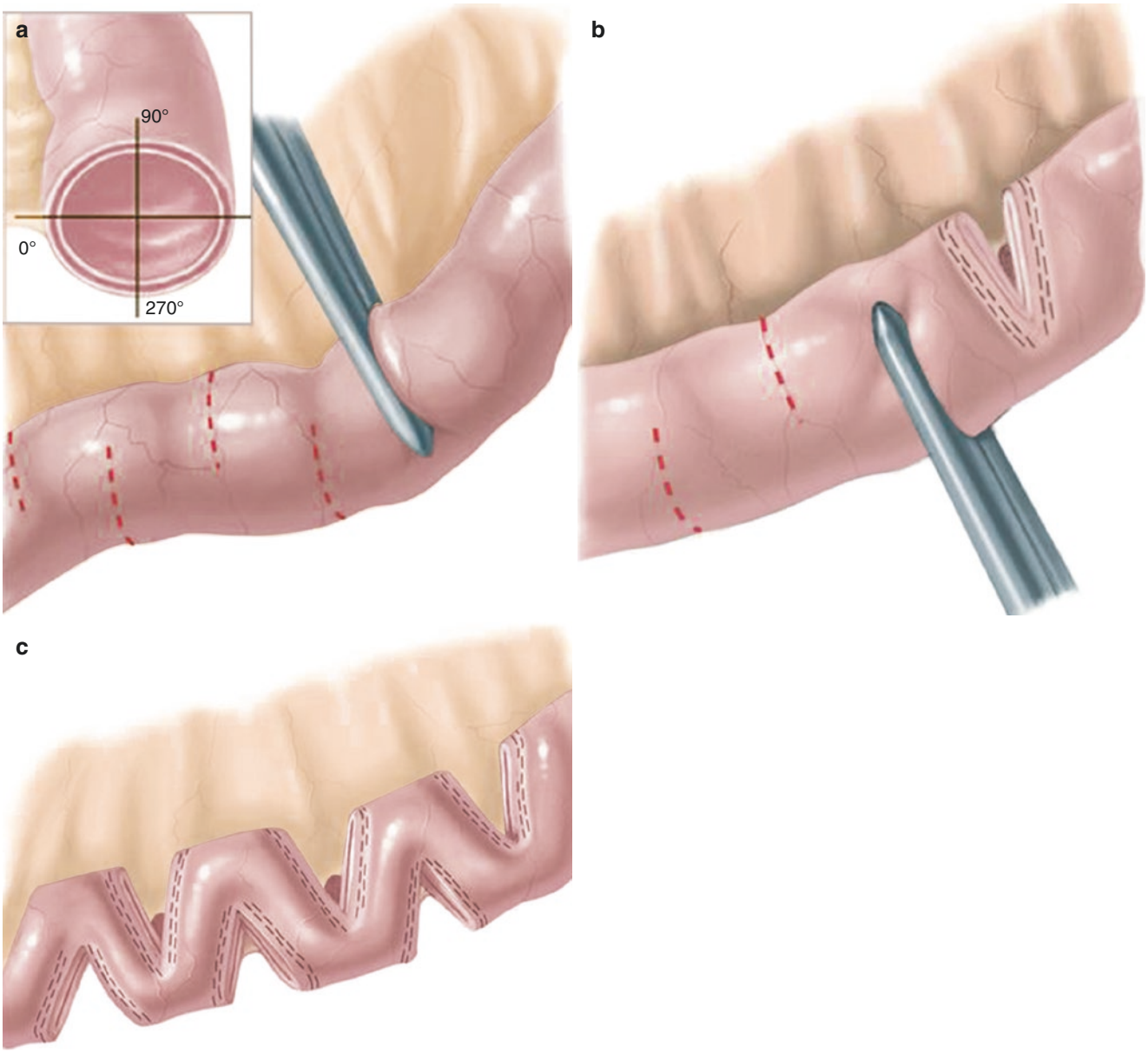

Fig. 29.9 (**a**–**c**) Serial transverse enteroplasty (STEP) is a new procedure to refashion dilated intestinal loops, thereby improving peristalsis and motility. It is technically much easier than the Bianchi method. The dilated bowel segment is decompressed and laid out flat. Mechanical staplers are used one after the other in turn on the mesenteric and antimesenteric site to reduce the lumen and to prolong the passage time. The diameter of the remaining lumen should not be smaller than 2.5 cm. The central corner is most critical for bleeding, so an extra stitch is recommended. In contrast to Bianchi's method, the procedure has no limits on length. Redilatation may occur and re-STEP procedures have been published

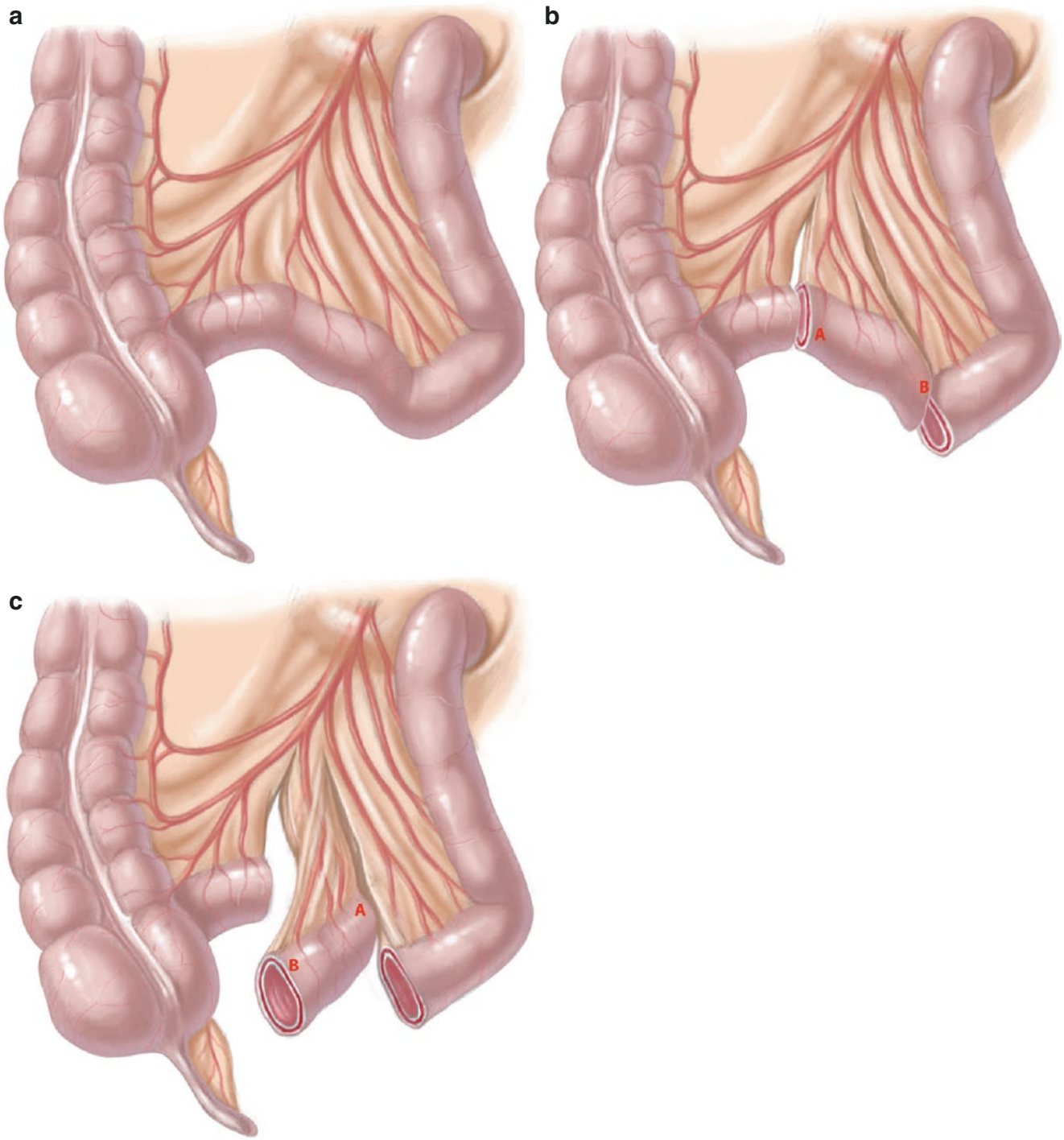

Fig. 29.10 (**a**–**c**) The use of antiperistaltic segments is indicated only in patients with good propulsion of the luminal chyme; it is contraindicated in those with disturbed motility. A number of different intestinal interpositions have been used in the past, both experimentally and (in some selected cases) clinically. The method for placing an antiperistaltic small or large bowel segment, as well as the interposition of an isoperistaltic colonic segment, are briefly demonstrated. Reversal of distal small bowel loops has been studied experimentally for years. The ideal length of the reversed segment appears to be 10 cm in adults and 3 cm in infants. The antiperistaltic segment acts as a physiological valve either by causing retrograde peristalsis or by functioning as an effective brake for the passage of chyme. The distal ileum (if available) is best used as an antiperistaltic segment shortly before the ileocecal valve. In an infant, a 3-cm segment of small bowel is first identified in regard to its blood supply, which should come from one major branch of the mesenteric vessels. Care must be taken that neither the blood supply to the remainder of the small intestine nor the supply to the colon, if present, is disturbed. The segment is isolated and the appropriate mesenteric base is isolated in a way that the segment can be reversed by 180° without impairing the blood flow. Finally, the proximal and distal intestinal anastomoses are performed as previously described. In the same way, a 3- to 5-cm antiperistaltic segment of the colon can be used. If no ileocecal valve is present, the best way is to reverse the first part of the colon immediately after the small intestine, but if the ileocecal valve has been preserved, the antiperistaltic colonic segment can be interposed between the distal small bowel and the valve. The method has rarely been used in humans and long-term results are not available

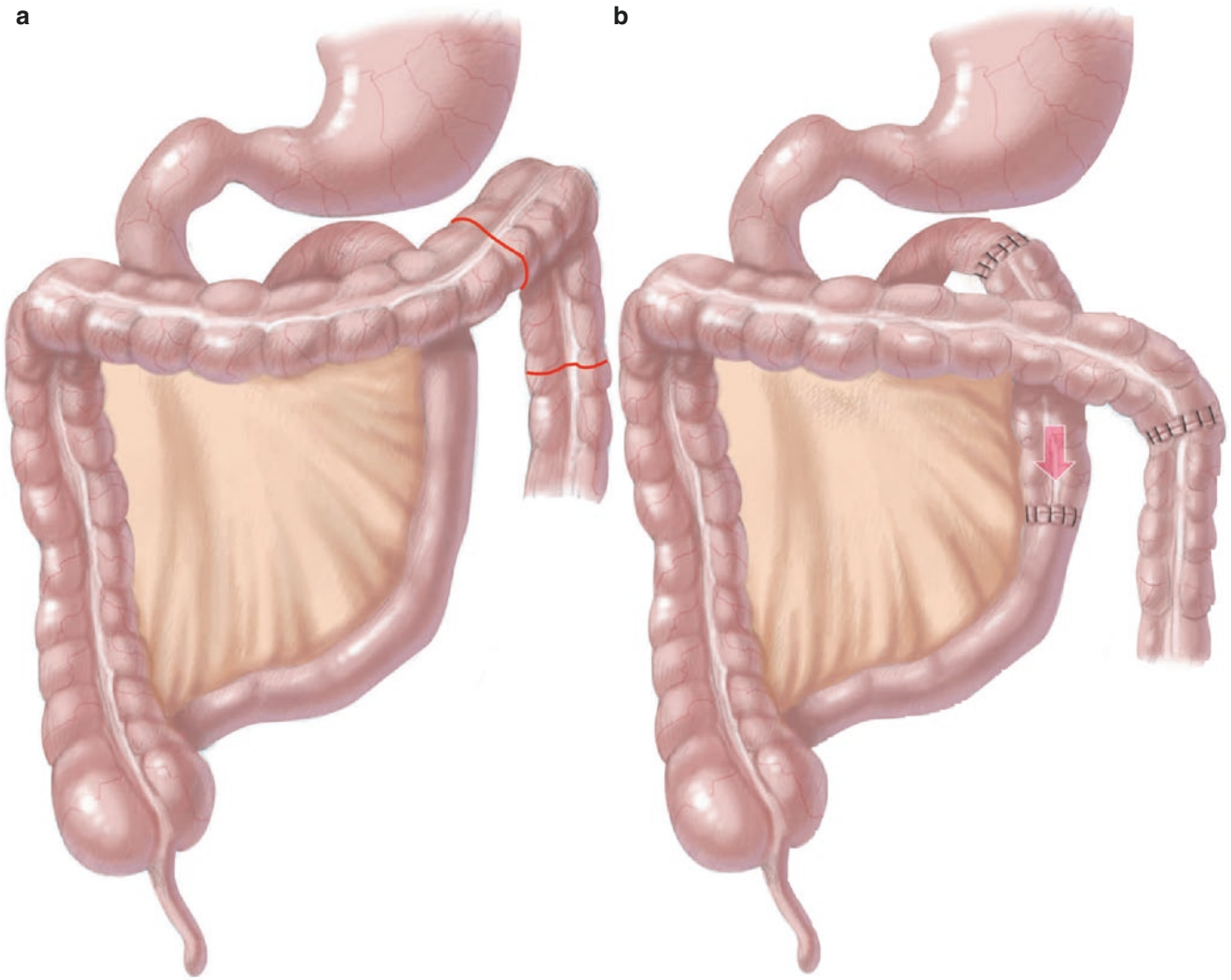

Fig. 29.11 (**a**, **b**) Isoperistaltic interposition of colon has the advantage of using none of the small bowel remnants. Isoperistaltic colonic interposition is best done with a 10- to 15-cm segment into the proximal part of the small intestine. Experimental evidence exists that the isoperistaltic colon prolongs transit time and gains some absorptive qualities. Favorable but highly variable results have been reported from the use of this method in children, with some groups reporting improved nutrient absorption and weaning from parenteral nutrition

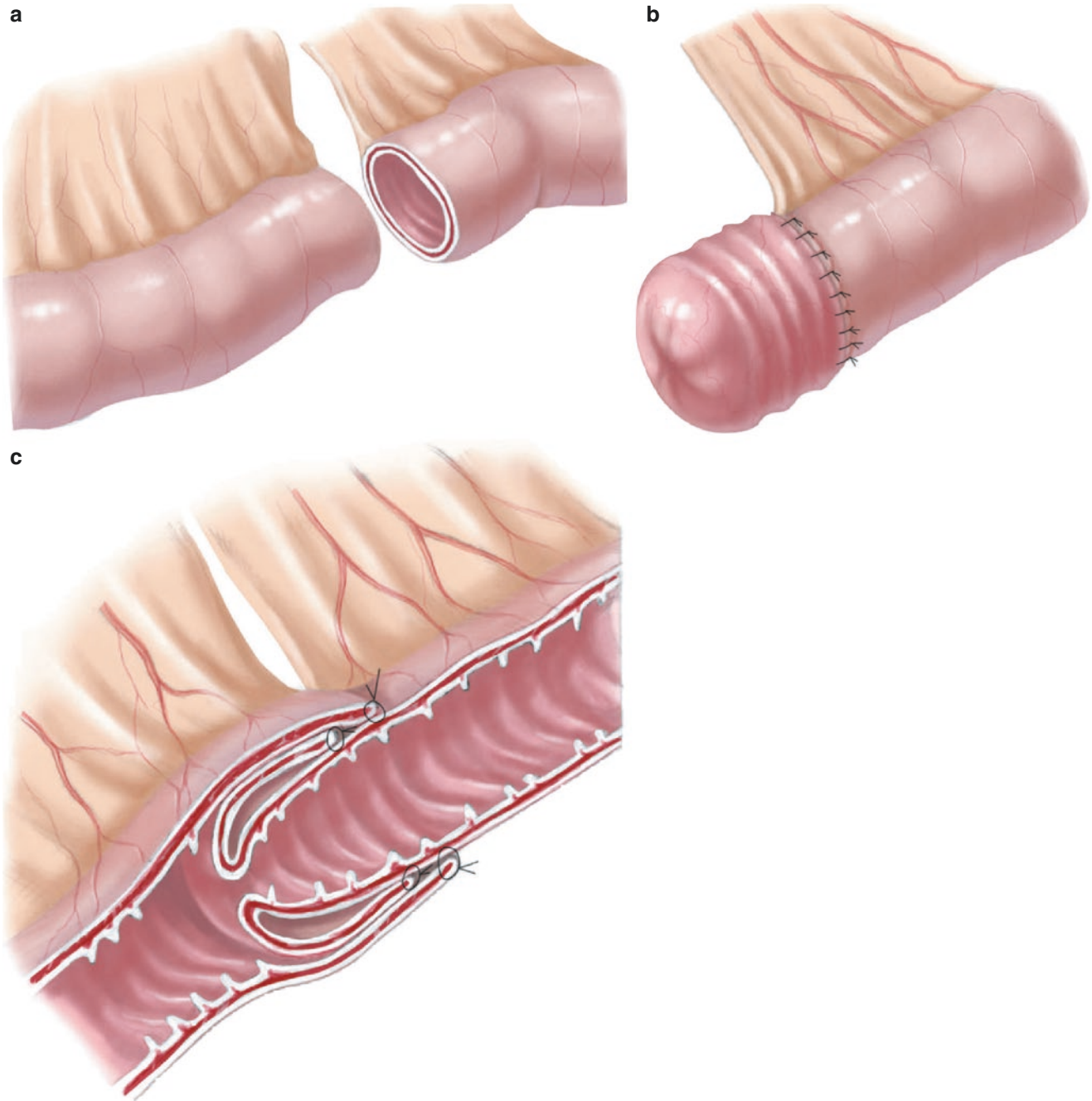

Fig. 29.12 (**a**–**c**) The benefits of the ileocecal junction on long-term outcome of babies with short bowel syndrome (SBS) has been questioned, but a large body of evidence indicates that it has a powerful impact on intestinal transit time, slowing the passage of intraluminal nutrients into the colon. Therefore, a variety of experimental surgical procedures have been devised to slow the intestinal transit time by creating artificial valves. The valve must be placed at the distal end of the small bowel. The intestine is transected at an appropriate level, and the last 2–4 cm of the end of the proximal bowel are everted and firmly fixed by 4/0 or 5/0 seromuscular interrupted sutures onto the underlying seromuscular bowel wall. The distal intestinal segment is then pulled over the everted bowel and finally anastomosed to the everted segment and to the proximal intestine by seromuscular interrupted stitches. The result corresponds to a typical prograde intussusception and may act as a valve similar to the ileocecal valve. A valve less than 3 cm can also be constructed in a reversed manner by everting the distal end of the small bowel, thereby creating a retrograde intussusception. This method may be more efficient, but definite clinical experience is very scarce

29.1 Results and Conclusions

The mainstay of the treatment of a newborn or child with SBS consists of a sophisticated enteral stimulation with nutritional equilibrium individually balanced among carbohydrates, proteins, and fatty acids. Enteral nutrition is the best stimulus for intestinal adaptation. The use of additional hormonal therapies has not yet proved sufficiently effective by controlled studies. Weaning from parenteral nutrition should be possible in more than 80% of the patients. Crucial for successful weaning is the presence of good, propulsive intestinal motility—with or without adjunct surgical measures. Therefore, the methods described in this chapter are indicated only in some individuals as a helpful adjunct therapy to the enteral nutritional program and must be planned very carefully.

Tapering is indicated in patients with sufficient intestinal length but severely dilated small bowel with impaired propulsive peristalsis. The intestinal content in the enlarged intestinal loops causes bacterial overgrowth, inflammation, and bacterial translocation with recurrent sepsis. Grossly dilated loops can be primarily due to an intentionally limited resection of small bowel in patients with intestinal atresia, or they can be the result of the process of intestinal adaptation with subsequent dilatation of the intestinal tract.

The method of intestinal tapering and lengthening (Bianchi 1999) attracted great attention in the past and has been used even in newborns with atresia and very short bowel remnants. However, many of these children (predominantly those with severe motility disorders) died later with total parenteral nutrition (TPN)–associated liver failure. Therefore, this method is now recommended only for carefully selected patients, particularly children with SBS who are about 1 year old and are not suffering from life-threatening TPN-associated liver failure. Furthermore, the method is indicated only in patients whose small intestine is no longer than 30–40 cm. Today, serial transverse enteroplasty (STEP) is the preferred method in many centers for secondary dilated intestinal loops, because it is technically easier and can be performed on longer segments. Initial long-term results are very promising.

Basically, all surgical measures intended to prolong intestinal transit time must be located at the distal end of the small bowel in order to allow as much digestion and absorption as possible in the proximal band. A difficulty with all these surgical methods is the need to find an ideal balance that prolongs the transit time without producing an ileus. One exception is the isoperistaltic interposition of a colonic segment, which must be placed in the proximal part of the small bowel because it not only prolongs the passage time but also seems to take on some absorptive capacities, thereby increasing the absorptive surface area. None of these techniques have been used extensively in patients (mostly because of the small number of patients who may profit from each particular procedure), so prospective studies are lacking.

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Hirschsprung's Disease

30

Prem Puri and David Coyle

Hirschsprung's disease (HD) is characterised by an absence of ganglion cells in the distal bowel extending proximally for varying distances. The absence of ganglion cells has been attributed to failure of migration, proliferation, and survival of neural crest cells. The earlier the arrest of migration, the longer the aganglionic segment. The pathophysiology of Hirschsprung's disease is not fully understood. There is no clear explanation for the occurrence of a spastic or tonically contracted aganglionic segment of bowel.

The aganglionosis is confined to the rectosigmoid in 80–85% of patients and 15–20% of patients have long-segment aganglionosis or total colonic aganglionosis. The incidence of HD is estimated to be 1 in 5000 live births. The disease is more common in boys, with a male-to-female ratio of 4:1. The male preponderance is less evident in long-segment HD, where the male-to-female ratio is 1.5–2 to 1.

30.1 Symptoms and Diagnosis

Of all cases of HD, 80–90% produce clinical symptoms and are diagnosed during the neonatal period. Delayed passage of meconium is the cardinal symptom in neonates with HD. Over 90% of affected patients fail to pass meconium in the first 24 h of life. The usual presentation of HD in the neonatal period includes constipation, abdominal distension, and vomiting during the first few days of life. About one third of the babies with HD present with diarrhoea. Diarrhoea in HD is always a symptom of enterocolitis, which remains the commonest cause of death.

In recent years, nearly all cases of HD have been diagnosed in the neonatal period. The diagnosis of HD is usually based on clinical history, radiologic studies, and anorectal manometry, as well as histologic examination of rectal wall biopsy specimens, the gold standard for diagnosis. Barium enema performed by an experienced radiologist using careful technique should achieve a high degree of reliability in diagnosing HD in the newborn. It is important that the infant should not have rectal washouts or even digital examinations prior to the barium enema, as such interference may distort the appearance of the transitional zone and give a false-negative diagnosis. A typical case of HD will demonstrate flow of barium from the undilated rectum through a cone-shaped transitional zone into dilated colon. In the presence of enterocolitis complicating HD, barium enema may demonstrate spasm, mucosal edema, and ulceration.

The diagnosis of HD is confirmed on examination of rectal biopsy specimens. The introduction of histochemical staining technique for the detection of acetylcholinesterase activity in suction rectal biopsy has resulted in a reliable and simple method for the diagnosis of HD. Immunohistochemical examination of paraffin-embedded specimens for calretinin has further improved the accuracy of diagnosis, especially in the case of smaller specimens. Full-thickness rectal biopsy is rarely indicated for the diagnosis of HD. Once the diagnosis of HD has been confirmed by rectal biopsy examination, the

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infant should be prepared for surgery. Intraoperative biopsies for frozen sections are taken to determine the extent of aganglionosis and the level of the transition zone.

30.2 Surgical Procedures

Many centres are now performing one-stage pull-through operations in the newborn with minimal morbidity rates and encouraging results. The advantages of operating on the newborn are that the colonic dilatation can be quickly controlled by washouts, and the calibre of the pull-through bowel at operation is near normal, allowing for an accurate anastomosis that minimizes leakage and cuff infection. A number of different operations have been described for the treatment of HD. Four operations are most commonly used:

- Rectosigmoidectomy, developed by Swenson and Bill
- Retrorectal approach, developed by Duhamel
- Endorectal procedure, developed by Soave
- Deep anterior colorectal anastomosis, developed by Rehbein

The basic principle in all these procedures is to bring the ganglionic bowel down to the anus. The long-term results of any of these operations are very satisfactory if they are performed correctly.

Since the 1990s, a variety of one-stage pull-through procedures in the newborn using minimally invasive laparoscopic techniques have been described. The major purported benefits of this approach are reduced adhesion formation, excellent visualisation of the pelvic structures, and shorter postoperative recovery time.

30.2.1 Staged Treatment with Colostomy

Staged treatment for HD with up-front colostomy formation is now generally reserved for those with enterocolitis or those too unwell to tolerate primary pull-through, or in patients where colonic decompression is not possible with washouts and rectal stimulation alone. The abdomen is opened via the Pfannenstiel incision (Fig. 30.1). Full-thickness rectal biopcies are performed for frozen section to confirmed the extent of aganglionosis (Fig. 30.2).

Many surgeons prefer right transverse colostomy; others advocate performing colostomy just above the transition to ganglionic bowel. Ileostomy is indicated in patients who have total colonic aganglionosis. A right transverse colostomy is convenient in usual cases. We perform a loop colostomy over a skin bridge. A V-shaped incision is made in the

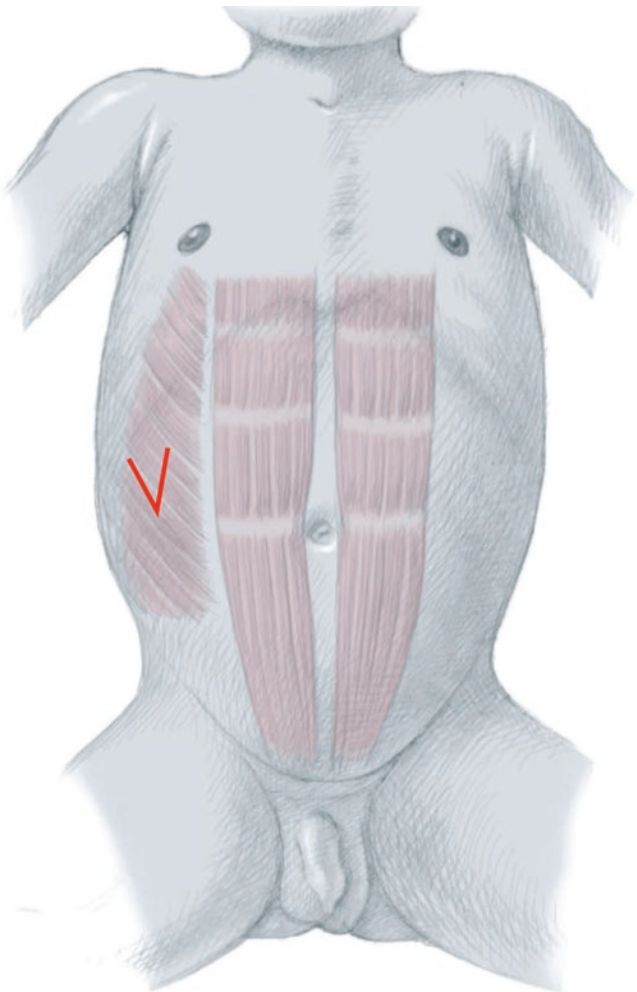

Fig. 30.1 The abdomen is opened via the Pfannenstiel incision

right upper quadrant (Fig. 30.1), and the V-shaped skin flap is reflected upwards. The external oblique is split and the internal oblique and transverse abdominis muscles are divided with diathermy. The peritoneum is opened.

An opening is made in the mesocolon of the selected segment of transverse colon. The skin flap is pulled through the opening in the mesocolon (Fig. 30.3) and sutured to the opposite skin margin. A few interrupted absorbable sutures of 4/0 or 5/0 are placed between the peritoneum, the muscle layers of abdominal wall, and the seromuscular layer of colon. The colon is opened longitudinally along the antimesenteric border using diathermy (Fig. 30.4). The bowel is sutured to the skin using interrupted 4/0 absorbable sutures (Fig. 30.5).

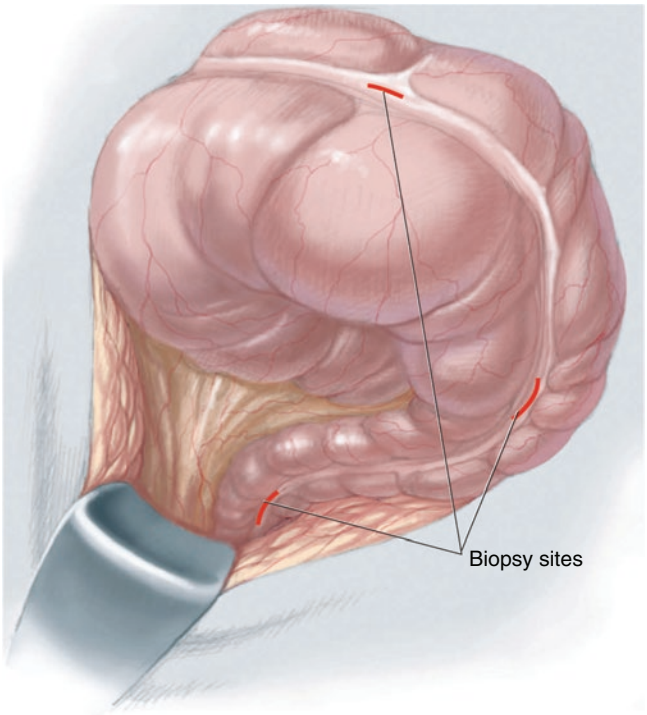

Fig. 30.2 The biopsy site is selected by observing the apparent transitional zone. In the usual case of rectosigmoid aganglionosis, three seromuscular biopsies are taken along the antimesenteric surface without entering the lumen. One biopsy is taken from the narrowed segment of bowel, a second biopsy from the transition zone and a third biopsy from the dilated portion above the transition zone. Biopsies are assessed intraoperatively by frozen section, to determine the level of ganglionic bowel

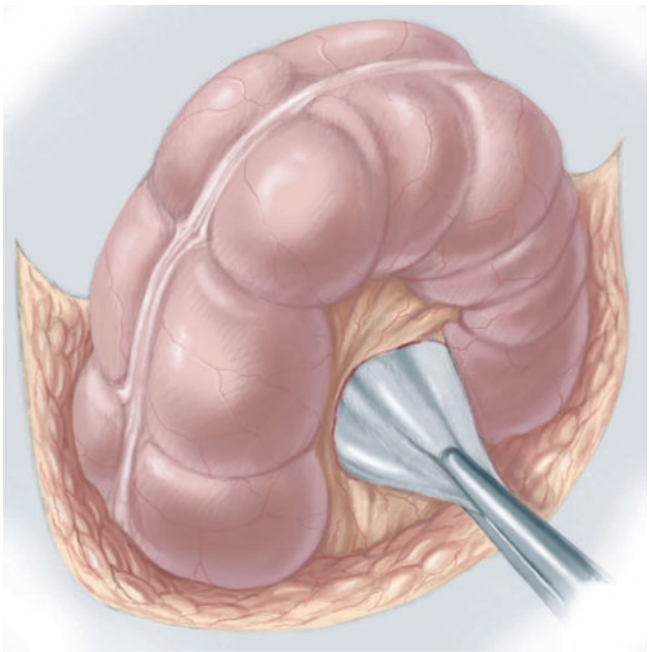

Fig. 30.3 An opening is made in the mesocolon of the selected segment of transverse colon. The skin flap is pulled through the opening in the mesocolon and sutured to the opposite skin margin

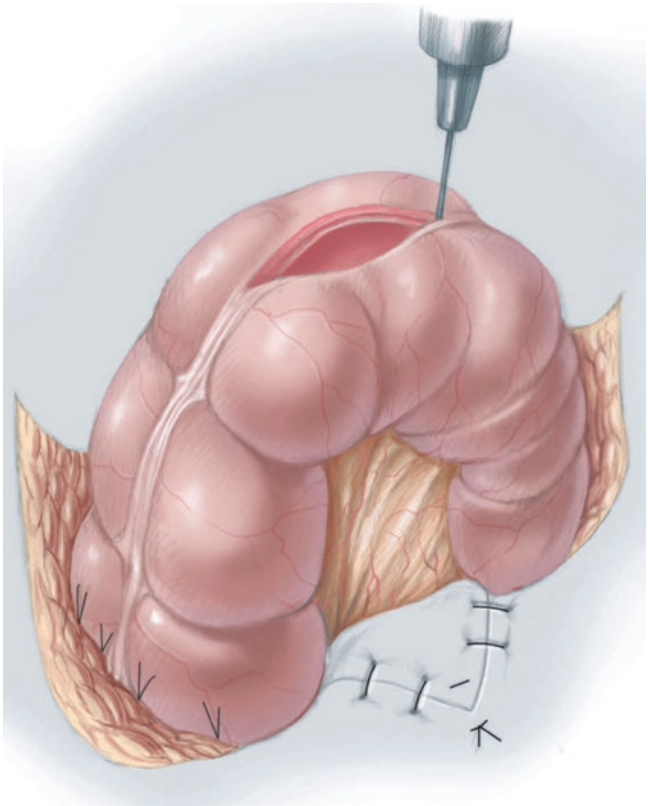

Fig. 30.4 A few interrupted absorbable sutures of 4/0 or 5/0 are placed between the peritoneum, the muscle layers of abdominal wall, and the seromuscular layer of colon. The colon is opened longitudinally along the antimesenteric border using diathermy

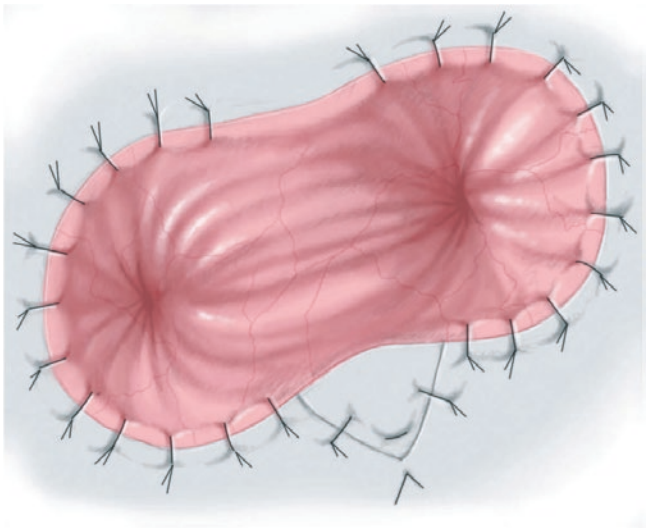

Fig. 30.5 The bowel is sutured to the skin using interrupted 4/0 absorbable sutures

30.2.2 One-Stage Transanal Endorectal Pull-Through Operation

Of patients with HD, 80–85% have rectosigmoid aganglionosis. A one-stage pull-through operation can be successfully performed in these patients using a transanal endorectal approach without opening the abdomen. This procedure is associated with excellent clinical results and permits early postoperative feeding, early hospital discharge, and no visible scars. Once the diagnosis of HD is confirmed, rectal irrigations are carried out twice a day for 3 days before surgery. Intravenous gentamicin and metronidazole are started on the morning of operation.

The authors prefer the one-stage, transanal endorectal pull-through operation for classic rectosigmoid HD and Swenson's pull-through operation for long-segment HD because of their simplicity and lack of complications. We have not used diversionary colostomy for usual cases.

The patient is positioned on the operating table in the lithotomy position. The legs are strapped over sandbags. A Foley catheter is inserted into the bladder. In infants and children, the anus can be everted using a self-retaining Lone Star retractor, which usually provides excellent exposure of the dentate line. The hooks of the retractor can then be placed inside the dentate line to protect it during mucosal dissection. In neonates, the dentate line can be exposed with the placement of sutures. The rectal mucosa is circumferentially incised using cautery, approximately 5–10 mm above the dentate line, and the submucosal plane is developed (Fig. 30.6). The proximal cut edge of the mucosal cuff is held with multiple 4/0 silk sutures, which are used for traction. The endorectal dissection is then carried proximally, staying in the submucosal plane; some surgeons carry out this procedure in the Swenson plane.

When the submucosal dissection has extended proximally to a point above the peritoneal reflection, the rectal muscle is divided circumferentially, and the full thickness of the rectum and sigmoid colon is mobilized out through the anus (Fig. 30.7). This requires division of rectal and sigmoid vessels, which can be done under direct vision using cautery or ligatures. Once the peritoneal reflection is divided, placement of an anterior serosal stay suture will prevent unwanted twisting of the colon as it is pulled through (Fig. 30.8).

When the transition zone is encountered, full-thickness biopsy sections are taken and frozen section confirmation of ganglion cells is obtained. Following confirmation of the level of the proximal extent of the transition zone, the rectal muscular cuff is split longitudinally, either anteriorly or posteriorly, and the colon is then divided several centimetres above the most proximal normal biopsy site (Fig. 30.9).

A standard Soave-Boley anastomosis is performed (Fig. 30.10). No drains are placed. The patient is started on oral feeds after 24 h and discharged home on the third postoperative day. Digital rectal examination is performed 2 weeks after the operation. Routine rectal dilation is not performed unless there is evidence of a stricture.

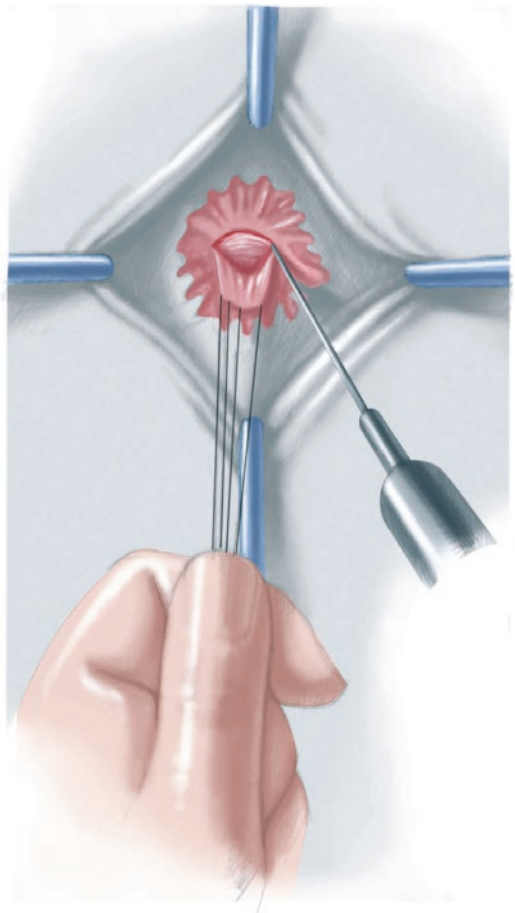

Fig. 30.6 The rectal mucosa is circumferentially incised using cautery, approximately 5–10 mm above the dentate line, and the submucosal plane is developed

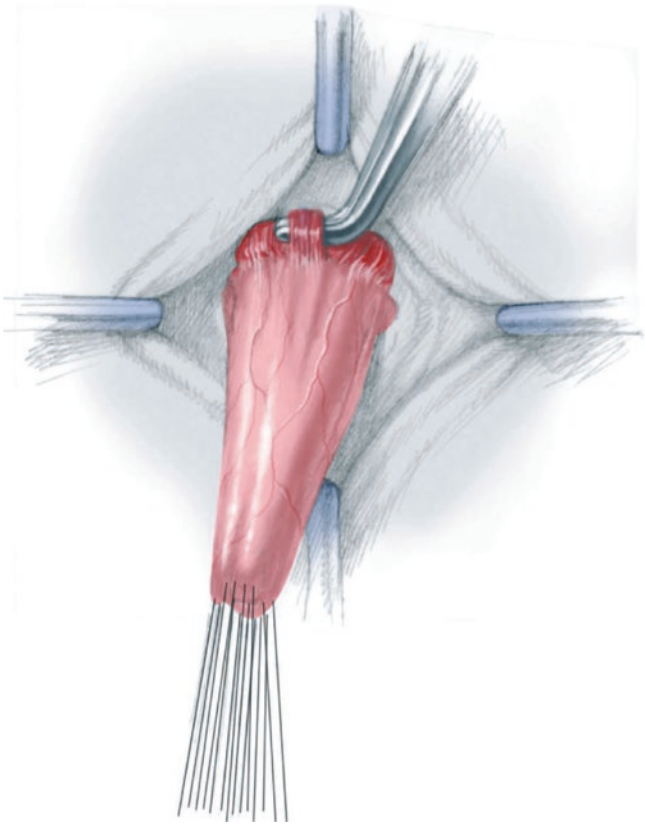

Fig. 30.7 When the submucosal dissection has extended proximally to a point above the peritoneal reflection, the rectal muscle is divided circumferentially, and the full thickness of the rectum and sigmoid colon is mobilized out through the anus. This requires division of rectal and sigmoid vessels, which can be done under direct vision using cautery or ligatures

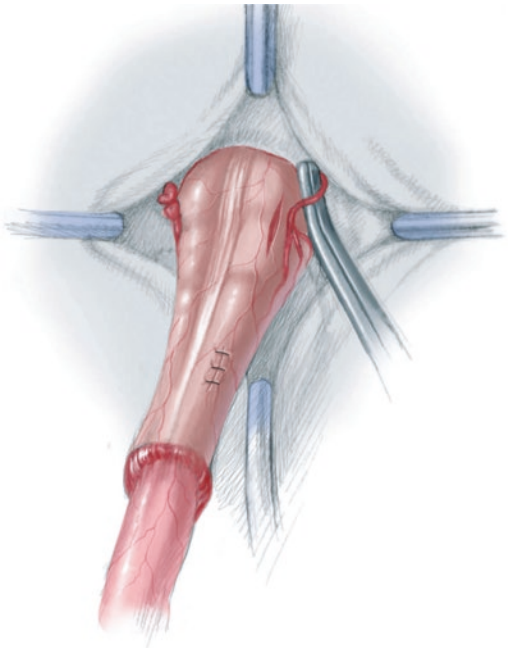

Fig. 30.8 When the transition zone is encountered, full-thickness biopsy sections are taken and frozen section confirmation of ganglion cells is obtained

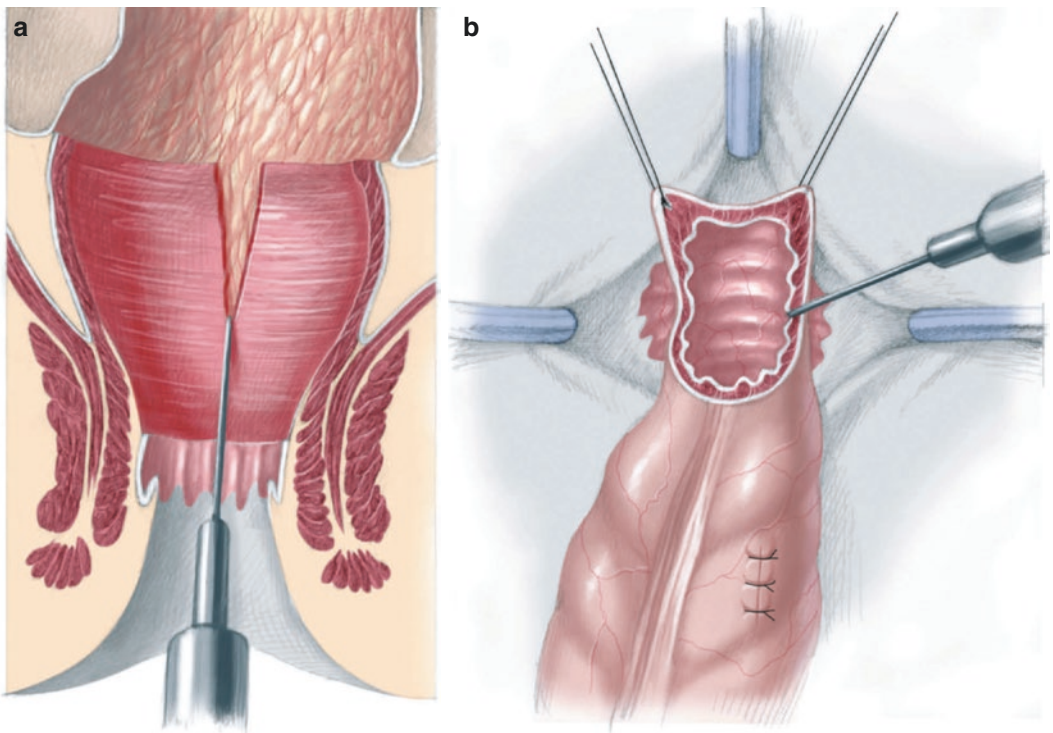

Fig. 30.9 (**a**) Following confirmation of the level of the proximal extent of the transition zone, the rectal muscular cuff is split longitudinally, either anteriorly or posteriorly. (**b**) The colon is then divided several centimetres above the most proximal normal biopsy site

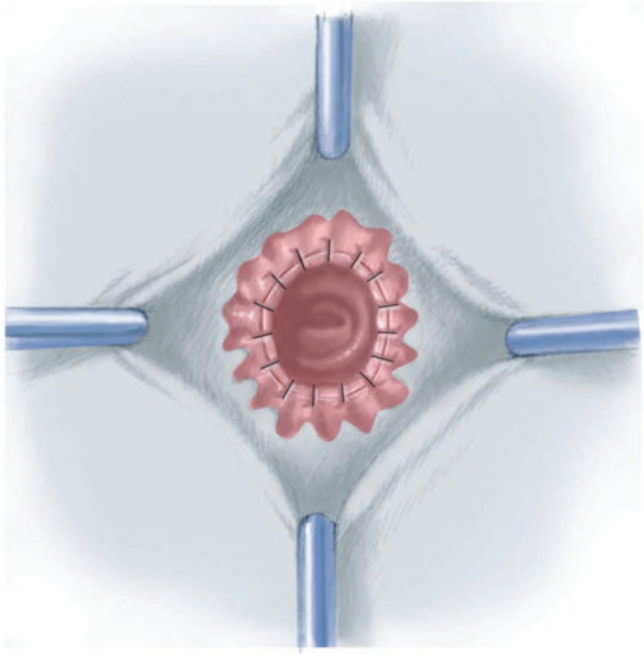

Fig. 30.10 A standard Soave-Boley anastomosis is performed. No drains are placed. The patient is started on oral feeds after 24 h and discharged home on the third postoperative day. Digital rectal examination is performed 2 weeks after the operation. Routine rectal dilation is not performed unless there is evidence of a stricture

30.2.3 Swenson's Pull-Through Operation

Swenson's pull-through operation is preferred for long-segment HD. For Swenson's pull-through operation, the patient is positioned on the operating table to provide simultaneous exposure of the perineum and abdomen. The pelvis is allowed to drop back over the lower end of the table and the legs are strapped over sandbags. A Foley catheter is inserted into the bladder. The abdomen is opened via a paramedian incision. (Some surgeons prefer a Pfannenstiel incision when performing a Swenson's pull-through operation in the neonate.) Extramucosal biopsies are taken at intervals along the antimesenteric border and assessed by frozen section to determine the level of ganglionated bowel. The sigmoid colon is mobilized by dividing the sigmoid vessels and retaining the marginal vessels. It may be necessary to mobilize the splenic flexure to obtain adequate length. The proximal level of resection above the ganglionated level that was determined by frozen section is selected and the bowel is divided between intestinal clamps or staples.

The peritoneum is divided around its lateral and anterior reflection from the rectum, exposing the muscle coat of the rectum. At this point, the bowel is divided at the rectosigmoid junction and removed. Dissection extends around the rectum, keeping very close to the bowel wall. It is essential to maintain the dissection close to the muscular wall in order to prevent damage to the pelvic splanchnic innervation. All vessels are electrocoagulated under direct vision. Sufficient tension-free length is obtained by dividing the inferior mesenteric pedicle, carefully preserving the marginal vessels. Dissection is carried down to the level of the external sphincter posteriorly and laterally, but does not extend as deeply anteriorly, leaving about 1.5 cm of intact rectal wall abutting the vagina or urethra.

The mobilized rectum is intussuscepted through the anus by passing a curved clamp or a Babcock forceps through the anal canal; an assistant places the closed rectal stump within the jaws of the clamp. When the dissection has been completed, it should be possible to evert the anal canal completely when traction is applied to the rectum. An incision is made anteriorly through the rectal wall about 1 cm from the dentate line, extending halfway through the rectal circumference. A clamp is inserted through this incision to grasp multiple sutures placed through the cut end of the proximal colon (Fig. 30.11). An outer layer of interrupted 4-0 absorbable sutures is placed through the cut muscular edge of the rectum and the muscular wall of the pull-through colon. When the outer layer has been completed, the proximal bowel is opened and an inner layer of interrupted 4-0 absorbable sutures is placed. When anastomosis is completed, the sutures are cut, allowing the anastomosis to retract within the anus.

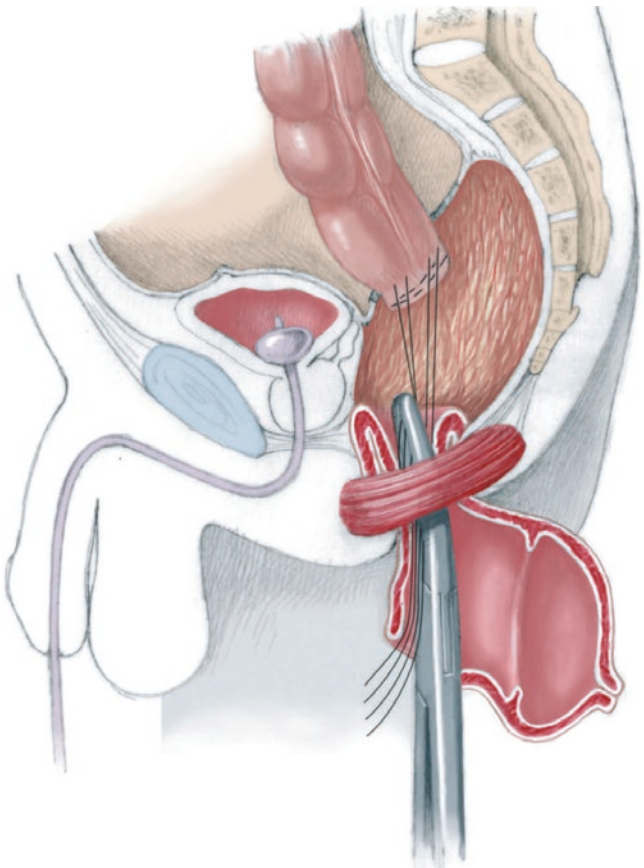

Fig. 30.11 In a Swenson's pull-through operation, a clamp is inserted through the incision in the rectal wall to grasp multiple sutures placed through the cut end of the proximal colon

30.2.4 Duhamel Pull-Through Operation

The advantage of the Duhamel pull-through is that very little manipulation of the rectum is performed anteriorly, thus avoiding injury to the genitourinary innervation. The rectum is divided and closed just above the peritoneal reflection. The redundant aganglionic bowel is resected. The retrorectal space is created by blunt dissection down to the pelvic floor. The posterior rectal wall is incised 1.5–2 cm above the dentate line, sponge-holding forceps is inserted into the retrorectal space, and ganglionic bowel is pulled through (Fig. 30.12). The anterior half of the pulled-through ganglionic bowel is anastomosed to the posterior wall of the aganglionic rectum, and the remainder of the colorectal anastomosis is completed by approximating the aganglionic rectum to the posterior wall of the pulled-through ganglionic bowel. Finally, an extra-long automatic stapling device is used to complete the side-to-side anastomosis between the aganglionic rectum and the ganglionic pulled-through bowel. Some surgeons complete the side-to-side anastomosis before closing the rectal stump, thereby preventing any residual septum.

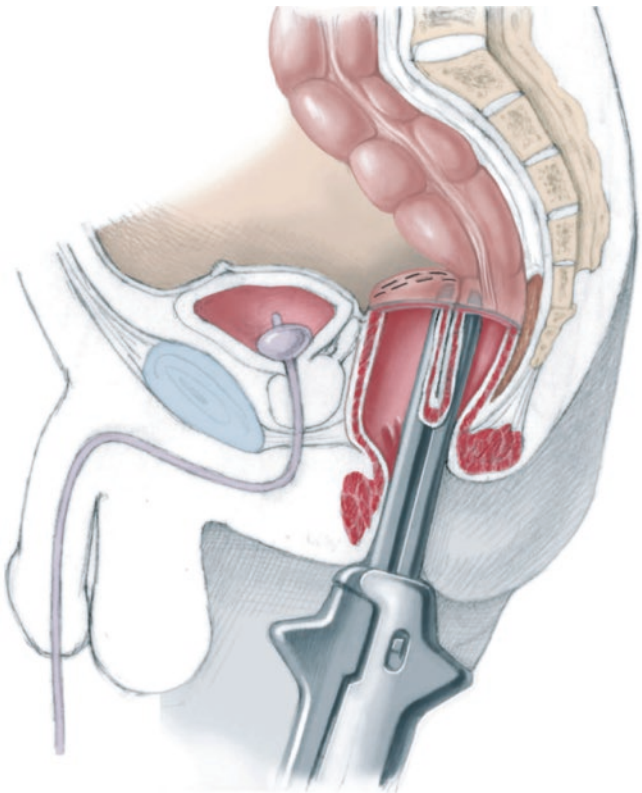

Fig. 30.12 In Duhamel operation, rectum is divided and closed just above the peritoneal reflection

30.2.5 Soave (Endorectal) Pull-Through Operation

In Soave or endorectal pull-through, the first steps of the operation are similar to those described for a Swenson's or Duhamel operation. The colon is mobilized and resected about 4 cm above the peritoneal reflection. The endorectal dissection begins 2 cm below the peritoneal reflection. The seromuscular layer is incised circumferentially and the mucosal-submucosal tube is freed distally. The mucosal dissection is continued distally to the level of the dentate line. The mucosa is incised circumferentially 1 cm above the dentate line. A Kelly clamp is inserted from below and the ganglionic bowel is pulled through (Fig. 30.13). The coloanal anastomosis is completed using 4/0 absorbable sutures.

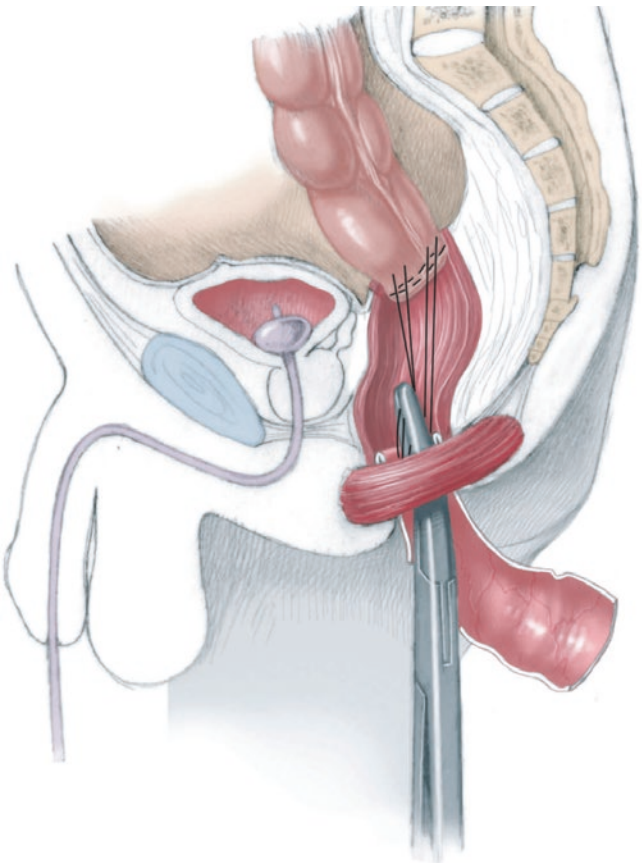

Fig. 30.13 A Kelly clamp is used to pull through the ganglionic bowel in the Soave technique

30.2.6 Rehbein's Technique

Rehbein's technique differs from the Swenson's procedure in that the anastomosis is a low, anterior colorectal anastomosis (Fig. 30.14). In this procedure, 3–5 cm of the terminal aganglionic rectum is left behind, which is anastomosed to the ganglionic bowel.

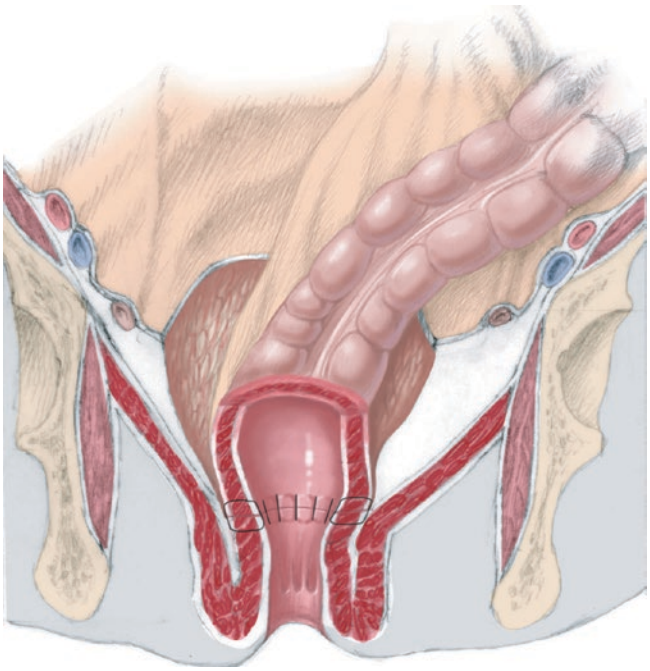

Fig. 30.14 Rehbein's technique uses a low, anterior colorectal anastomosis

30.2.7 Minimally Invasive Techniques

Minimally invasive modifications of the Soave, Duhamel, and Swenson techniques have evolved over the past two decades. In the laparoscopic-assisted pull-through, preoperative preparation is the same as for open surgery. Infants and smaller children are positioned transversely across the operating table, and older children are positioned supine with legs in stirrups. Use of a whole-body antiseptic skin preparation below the level of the nipples allows the surgeon to access both the abdomen and the perineum as the need arises.

Usually three to four trocars are placed: a camera port placed either supraumbilically or in the right subcostal area, one trocar high in the right iliac fossa, and one in the left lower quadrant (Fig. 30.15). An optional fourth trocar may be placed in the left upper quadrant to allow for traction on the colon in cases of long-segment aganglionosis. A 30° laparoscope is used to provide optimal visualisation.

The first step in the laparoscopic procedure is establishment of the level of the transition zone by taking seromuscular biopsies with a sharp-tipped Metzenbaum scissors for frozen section examination. Intracorporeal suture closure of the biopsy site can be performed while awaiting histology. As the presence of aganglionosis proximal to the splenic flexure may alter the operative approach greatly, it is advisable to wait until the level of the most proximal extent of the transition zone is known before proceeding with further dissection.

The next step is dissection of the mesocolon, beginning with the superior rectal vessels. This can be accomplished with hook diathermy, an ultrasonic Harmonic scalpel, or a LigaSure® device. Preservation of the pedicle, including the marginal artery, may be necessary in those with aganglionosis extending beyond the rectosigmoid region. Division of the lateral peritoneal attachments of the descending colon follows, if necessary. The most distal extent of dissection depends on which technique is being used. In the laparoscopic-assisted Duhamel procedure, the distal rectum can be transected using a laparoscopic stapling device, and a laparoscopic grasper can be used to pull the proximal transected colon through an incision in the posterior rectum.

Perineal dissection should proceed as in the open or endorectal pull-through. It is critical to use the laparoscope to ensure that the pulled-through colon is not twisted, and in the case of the laparoscopic-assisted Duhamel technique, to ensure that the mesentery of the pulled-through colon is posterior, in order to avoid transection of the mesentery when the spur between the native rectum and pulled-through colon is being divided transanally.

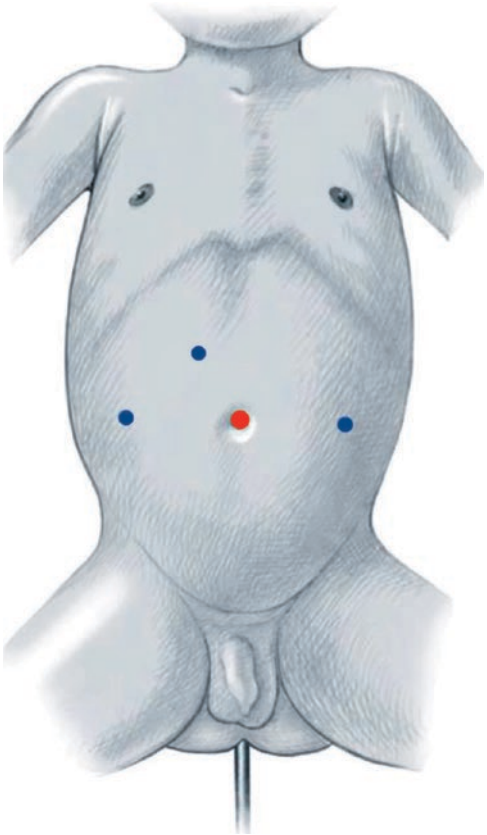

Fig. 30.15 Placement of trocars for laparoscopic-assisted pull-through procedures

30.3 Outcome

The vast majority of patients treated with any one of the standard pull-through procedures achieve satisfactory continence and function with time. The attainment of normal continence depends on the intensity of bowel training and the patient's social background and intelligence. Mental handicap, including Down syndrome, is invariably associated with long-term incontinence.

Suggested Reading

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Anorectal Anomalies

31

Alberto Peña, Andrea Bischoff, and Marc A. Levitt

Anorectal malformations represent a wide spectrum of defects. Surgical techniques to repair the most common types of anorectal malformations seen by a general pediatric surgeon are presented in order of complexity, from the simplest to the most complex.

Malformations considered "low" traditionally have been approached perineally, without a protective colostomy, whereas malformations that are considered "high" are treated with a colostomy in the newborn period, with subsequent definitive repair of the malformation and eventual closure of the colostomy. Surgical management has been evolving towards fewer operations and minimally invasive procedures. Anorectal malformations have been included in this trend.

31.1 Colostomy

Colostomy is a vital procedure for children born with high and complex malformations. The circumstances in which this procedure is performed vary from one institution to another and from one country to another. Some surgeons feel confident approaching newborns without a protective colostomy. This is feasible and safe if the surgeon has experience in the management of these defects. Some surgeons prefer a safer path in which they perform a protective colostomy, particularly when the baby is premature or has severe associated anomalies.

The colostomy that is best in the management of anorectal malformations is a descending colostomy. The surgeon must understand that all colostomies performed in a mobile portion of the colon tend to prolapse. In the type of colostomy described here, the mobile portion is the distal stoma (mucous fistula), so the surgeon must either fix this distal sigmoid colon to the anterior abdominal wall or make the mucous fistula very small (4 mm in diameter), as it will be used only for colonic irrigations or a distal colostogram. The incision is created in the left lower quadrant, and the surgeon must be sure that both the stomas are sufficiently separated as to be able to accommodate the stoma bag over the proximal stoma. The mucosa fistula should not be included under the stoma bag. The colostomy can be created using a laparoscopic technique, which eliminates a skin incision between the two stomas.

When creating a colostomy in a newborn, the surgeon should look for the descending colon and select the first portion of mobile sigmoid to open the colostomy (Fig. 31.1). That part of the colon is usually very distended and full of meconium. The surgeon can place a purse-string suture, make an orifice in the centre, and pass a catheter to irrigate the sigmoid until all the meconium has been removed. This simple maneuver facilitates the manipulation of the colon and helps to create a better colostomy. It is also vital because removing the distal meconium reduces the risk of urinary

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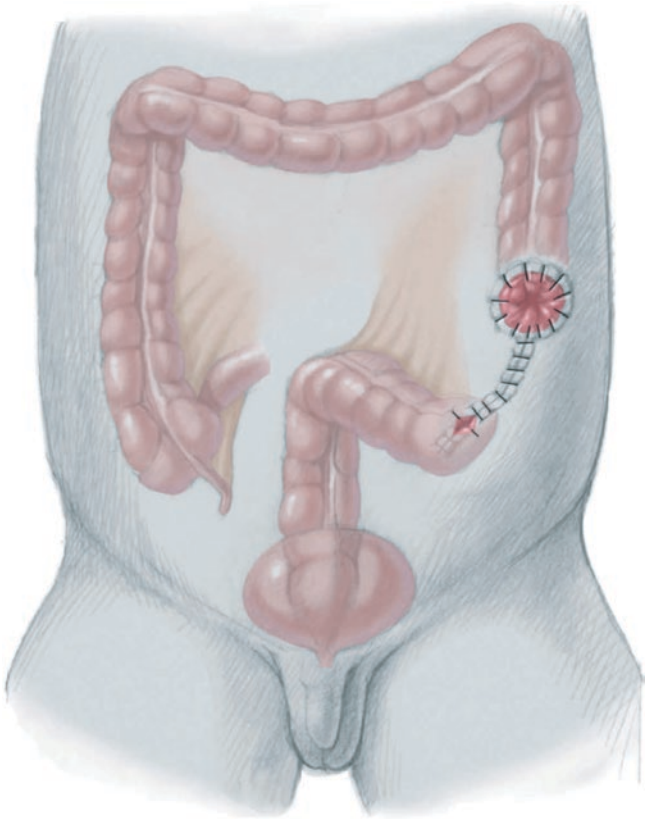

Fig. 31.1 Colostomy in a newborn

tract contamination and facilitates the future distal colostogram.

Loop colostomies are contraindicated in children with anorectal malformations. They tend to prolapse, and stool can pass into the distal stoma, which may provoke faecal impaction in the distal rectum and also contaminate the urinary tract with faeces.

31.2 Male Defects

31.2.1 Perineal Fistula

This malformation represents the simplest on the spectrum. In this defect, the rectum opens immediately anterior to the centre of the sphincter, yet the anterior rectal wall is intimately attached to the posterior urethra (Fig. 31.2). The anal orifice is frequently narrowed. These patients will have bowel control with or without an operation, but without surgery, they may have problems with control later in life if the stool is loose or during athletic activities. We prefer to operate on these babies to give the child an anal opening that is centered in the sphincter and appropriately sized, and to achieve a better cosmetic effect. We also believe that the operation should be done soon after the diagnosis is made. We typically perform these operations without a colostomy in the newborn baby. The operation can be delayed but should be performed within the first few months of life.

The baby is placed in prone position with the pelvis elevated. It is *mandatory* to place a catheter in the patient's urethra to help prevent the most common intraoperative complication, which is a urethral injury. The incision extends mid-sagittally approximately 2 cm posterior to the anal orifice, dividing the sphincter mechanism posterior to the intended anal opening. Multiple fine, nonabsorbable sutures are placed in the fistula orifice to exert traction to facilitate dissection of the rectum. The posterior incision divides the

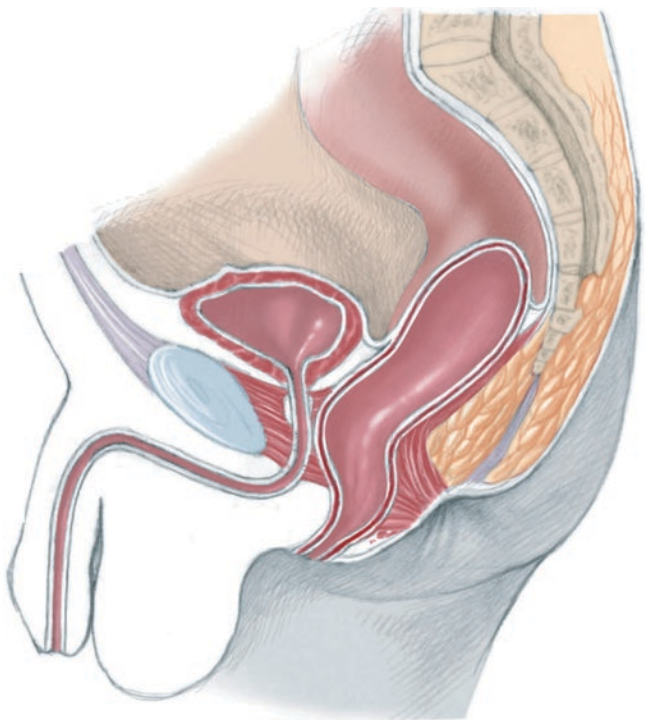

Fig. 31.2 Perineal fistula

sphincter until the posterior rectal wall (with its characteristic whitish appearance) is identified. The surgeon continues dissecting in this plane, first the lateral walls of the rectum and eventually the anterior rectal wall. While dissecting the anterior rectal wall, the surgeon must put special emphasis on trying to avoid urethral injury, as there is no plane of separation between the rectum and urethra. Once the rectum has been mobilized enough as to be moved back to be placed within the limits of the sphincter, the limits of the sphincter are determined with an electrical stimulator. The perineal body is then reconstructed with long-term, fine absorbable sutures, and the rectum is anchored to the posterior edge of the muscle complex. An anoplasty is performed within the limits of the sphincter, using 16 circumferential 6/0 long-term absorbable sutures. The baby is kept NPO for 5–7 days after surgery to ensure good healing. Intravenous antibiotics are given for 24 h.

Dilatations are performed twice per day beginning 2 weeks after surgery. The parents learn to advance one size (1 mm) every week until they reach the size adequate for the patient's age: no. 12 for a newborn baby, no. 13 for a 4-month-old, no. 14 for an 8-month-old, no. 15 for a 1-year-old, and no. 16 for older patients.

31.2.2 Rectourethral Fistula

This group of patients includes two specific subtypes: rectourethral bulbar fistula (Fig. 31.3) and rectoprostatic fistula (Fig. 31.4). These two variants represent the majority of male patients with anorectal malformations. It is vitally important to recognize the difference between these types, which have different prognostic and therapeutic implications. In our experience, patients with rectourethral bulbar fistula have an 80% chance of having bowel control by the age of 3, whereas that number is 60% for those with rectoprostatic fistula. Patients with a rectoprostatic fistula have a 60% incidence of associated defects, compared with 30% for patients with rectourethral bulbar fistula. The rectoprostatic patients require a more demanding perirectal dissection to mobilize the rectum, which is located higher in the pelvis. These operations can be performed within the first few months of life. If the neonatal approach with no colostomy is to be attempted, perhaps the best patient for this management would be the one who has a rectal pouch located below the coccyx, so that the surgeon will know for sure that posterior sagittal opening will find the rectum. The dissection of this distal rectum must be meticulous, as it is intimately attached to the urethra.

If the surgeon does not have a clear and reliable image that shows the rectum located below the coccyx, he or she should *never* approach a patient posterior sagittally without a colostomy and without a distal colostogram. The distal

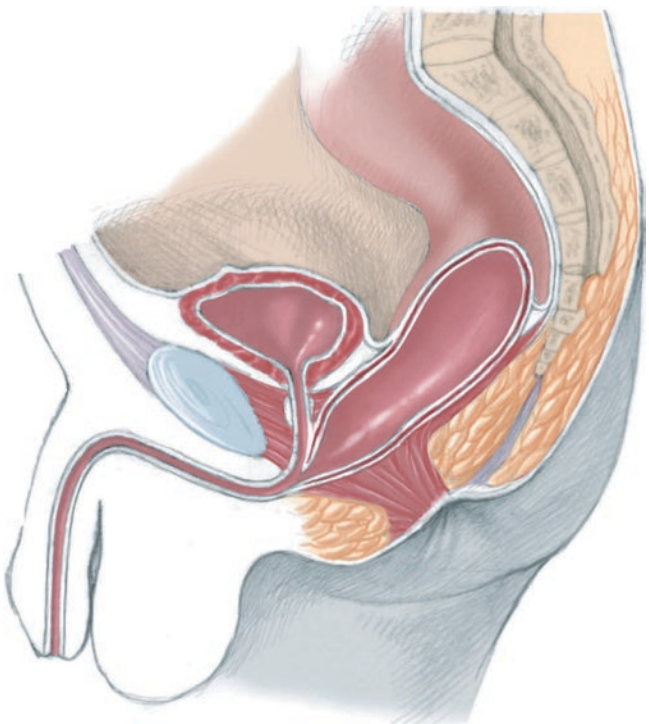

Fig. 31.3 Rectourethral bulbar fistula

colostogram, which is by far the most valuable study in defining the anorectal anatomy, can be done in patients with anorectal malformations only when the patient already has a colostomy. We have seen catastrophic complications during the performance of posterior sagittal operations in male patients who did not have a distal colostogram.

A Foley catheter is inserted and the patient is placed in prone position with the pelvis elevated. The incision is a posterior sagittal one, between the buttocks, running from the lower portion of the sacrum just above the coccyx down to the anal dimple. The limits of the sphincter are electrically determined. The incision goes through skin, subcutaneous tissue, parasagittal fibres, muscle complex, and levator muscle (Fig. 31.5). When the surgeon is dealing with a rectourethral bulbar fistula, he or she can expect to see a bulging rectum as soon as the levator muscle is split. In cases of rectoprostatic fistula, the rectum is much smaller and it may not bulge through the incision; the surgeon expects to find it immediately below the coccyx. The surgeon should not look for the rectum in the lower part of the incision in patients with rectoprostatic fistula. Looking for a rectum without preoperative evidence that the rectum is there is the main source of complications in this approach. The surgeon will instead find the urethra, vas deferens, prostate, or seminal vesicles and can damage the nerves important for urinary control and sexual potency.

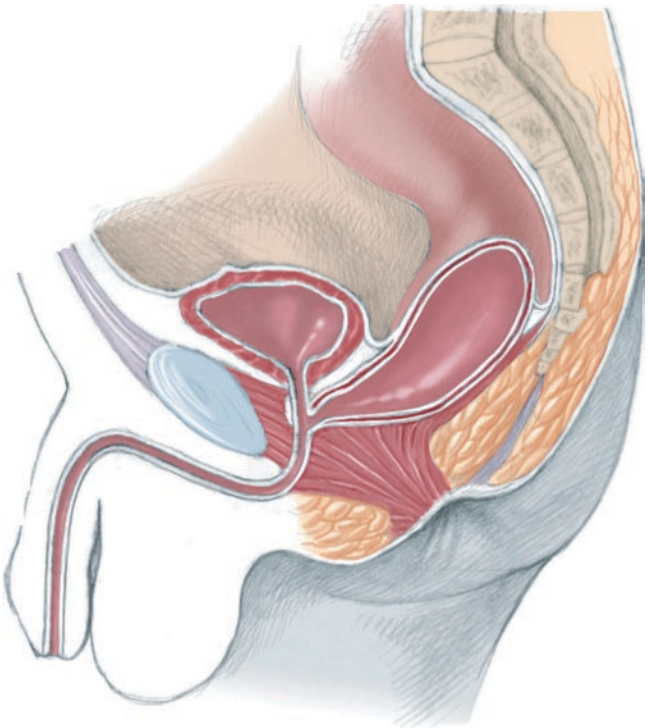

Fig. 31.4 Rectoprostatic fistula

The posterior rectal wall is easily identified by its characteristic whitish appearance. The surgeon must keep in mind that there is a fascia covering the rectum posterior and laterally, which must be removed. The dissection of the rectum must be performed as close as possible to the rectal wall without injuring the rectal wall itself. The posterior rectal wall is opened in the midline, between two 5/0 silk stitches (Fig. 31.6). The incision is continued distally, staying in the midline while placing stitches in the edges of the rectum. The traction on these stitches will allow the surgeon to see the lumen of the rectum (Fig. 31.7). When the incision continues, extending distally and anteriorly in the midline, it ends directly into the rectourethral fistula, which usually is identified as a 1- to 2-mm orifice.

Multiple 6/0 silk sutures are placed in a semicircumference above and lateral to the fistula site. These multiple sutures serve the purpose of exerting uniform traction on the rectal mucosa to facilitate the separation of the rectum from the urinary tract. The fascia that covers the rectum is then

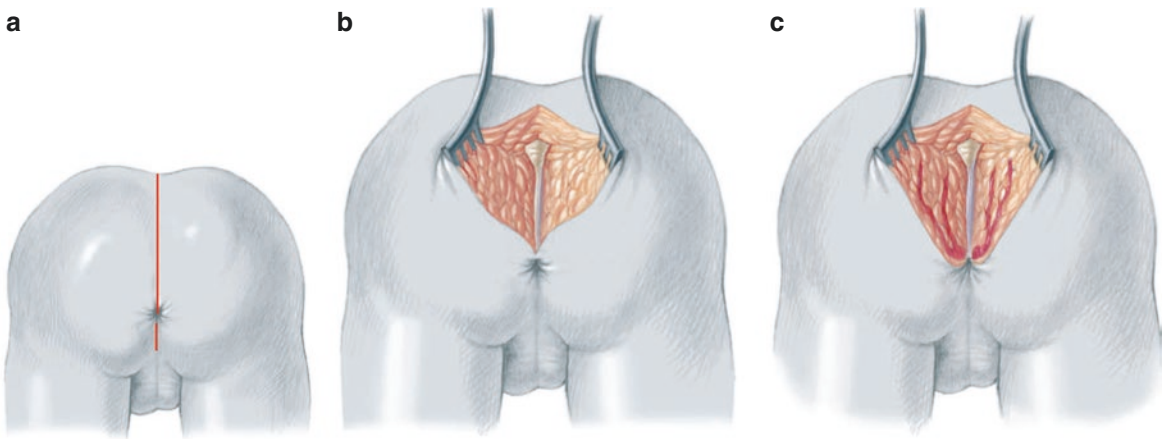

Fig. 31.5 Posterior sagittal incision to treat rectourethral fistula

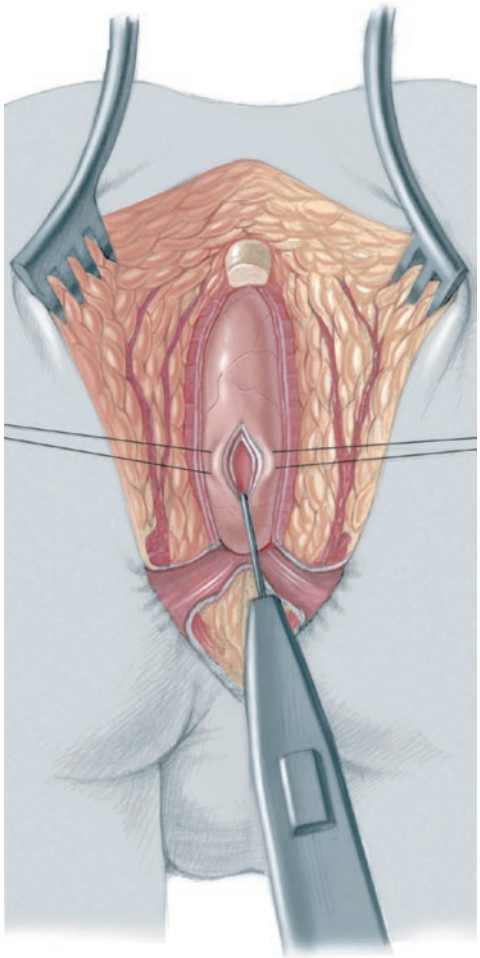

Fig. 31.6 Opening of the posterior rectal wall in the midline, between two 5/0 silk stitches

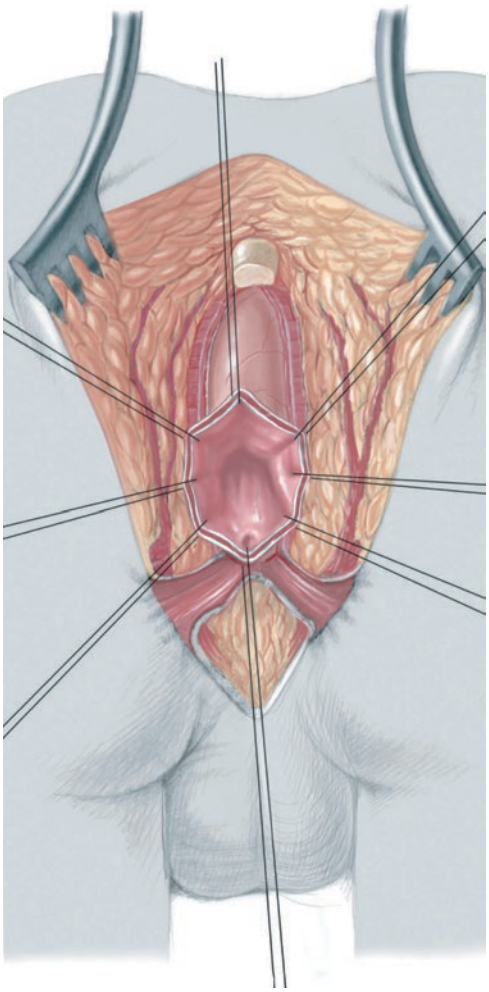

Fig. 31.7 Traction on stitches placed in the edges of the rectum to allow the surgeon to see the lumen of the rectum

removed, creating the lateral planes of the rectum. The mucosa of the anterior rectal wall distal to the multiple 6/0 silk sutures is divided at a depth of 1 mm (Fig. 31.8).

The dissection continues between the rectum and urinary tract in a submucosal plane for approximately 5–10 mm and then gradually becomes a full-thickness dissection, using as a guide the lateral planes, until the rectum is completely separated from the urinary tract (Fig. 31.9). The separation of the urethra from the rectum is the most delicate part of the operation, during which most serious complications occur. The fistula site is closed with three or four 6/0 long-term absorbable sutures (Fig. 31.10).

The surgeon should then evaluate the size of the rectum and decide if it is necessary to taper the rectum to fit within the limits of the sphincters. In the past, many of these tapering procedures were required, probably because patients came for surgery later in life, after having had inadequate colostomies and therefore developing a megarectum. Babies now are referred earlier and better, totally diverting colostomies are being performed, so megarectums are less common and tapering is rarely necessary.

When a tapering procedure is required, we recommend removing an adequate portion of the posterior rectal wall (Fig. 31.11), and then closing this wall in two layers of interrupted, long-term absorbable sutures. Tapering on the anterior wall is contraindicated, as it would leave a rectal suture line against the urethral fistula repair, and a recurrent fistula may develop.

The limits of the sphincter are then electrically determined and marked with temporary silk sutures (Fig. 31.12). The perineal body is reconstructed.

The posterior edge of the levator muscle is electrically determined, and the rectum is placed in front of the levator. The posterior edges of the levator muscle are sutured together with interrupted 5/0 long-term absorbable sutures. The distal continuation of the levator muscle is called the muscle complex. The posterior edge of this muscle structure is sutured together in the midline with interrupted 5/0 long-term

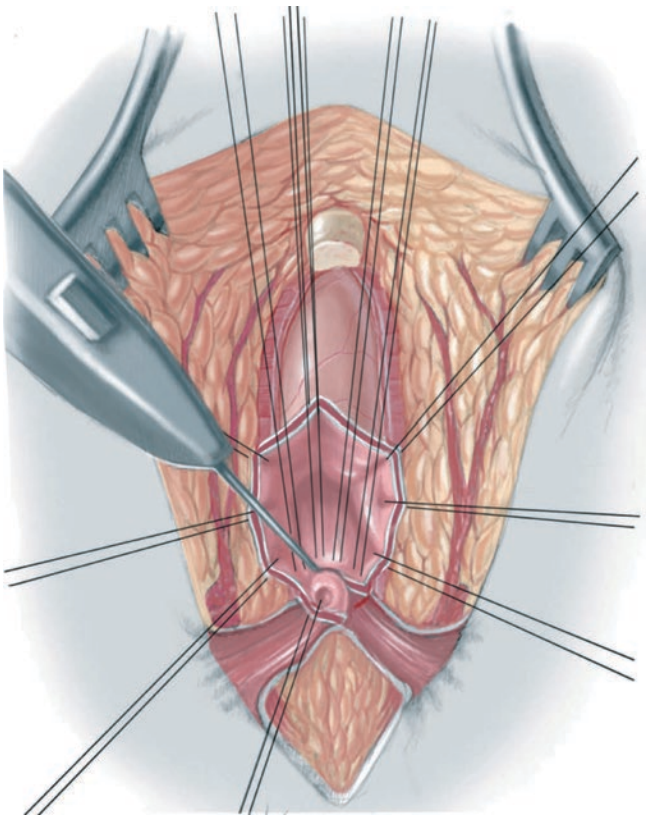

Fig. 31.8 Multiple 6/0 silk sutures are placed above and lateral to the fistula site, and the mucosa of the anterior rectal wall distal to these sutures is divided at a 1 mm depth

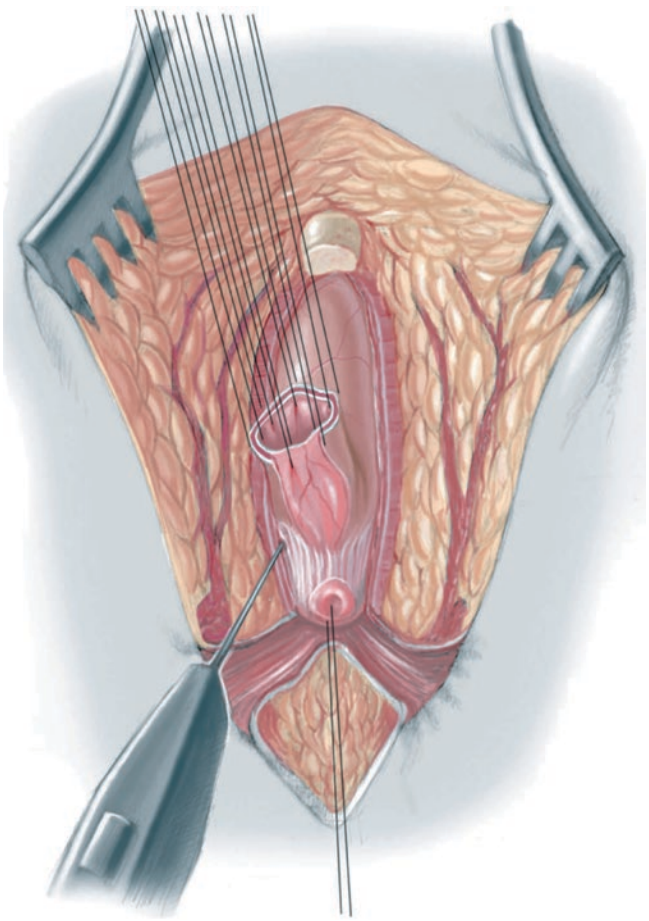

Fig. 31.9 Dissection between the rectum and urinary tract

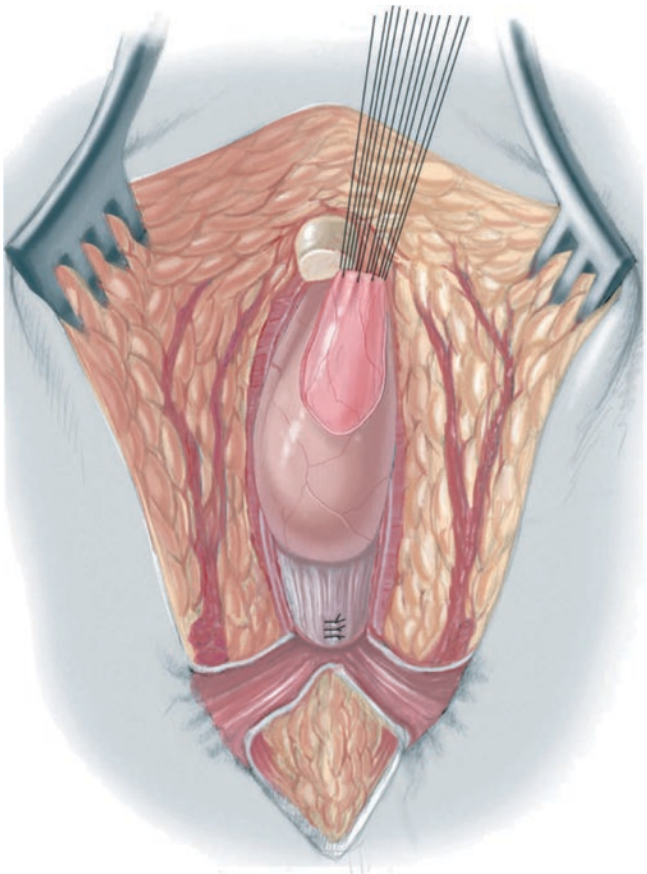

Fig. 31.10 Closure of the fistula site

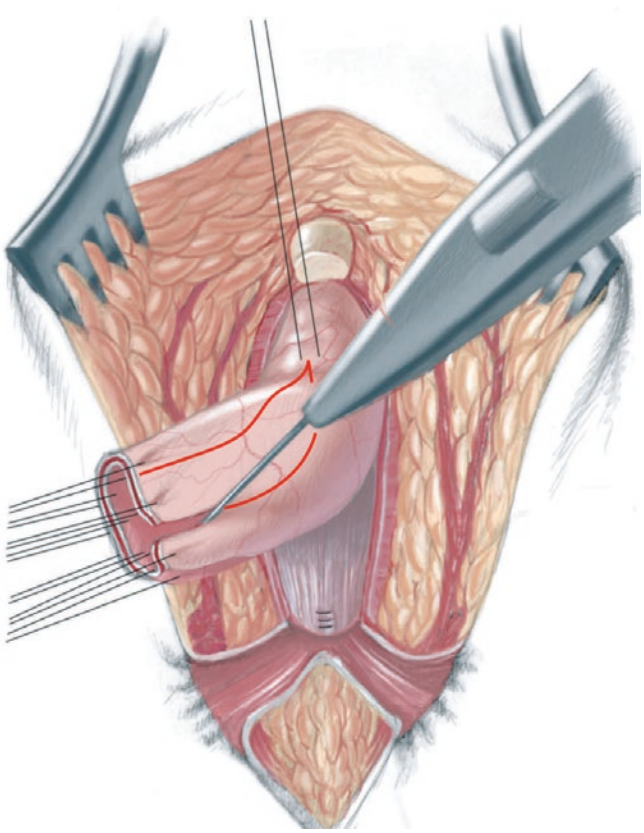

Fig. 31.11 Removal of a portion of the posterior rectal wall to taper the rectum

absorbable sutures (Fig. 31.13). These sutures also take a bite of the posterior rectal wall in order to anchor the rectum in a good position to avoid retraction, prolapse, or both.

The rest of the incision is closed meticulously, reapproximating all the layers of the wound (Fig. 31.14). An anoplasty is performed with 16 circumferential stitches of 6/0 long-term absorbable sutures (Fig. 31.15). Any excess part of the rectum that has been damaged or does not have adequate blood supply is trimmed off.

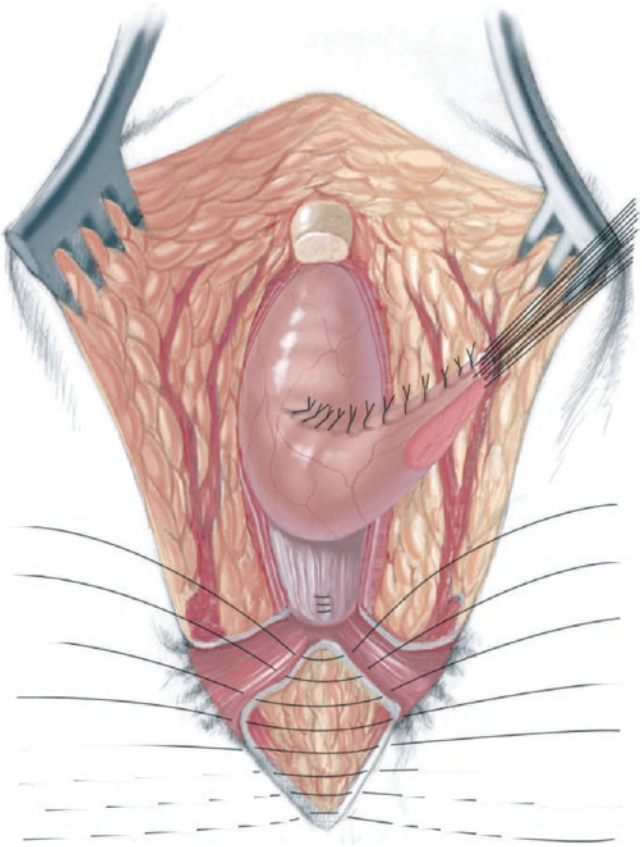

Fig. 31.12 The limits of the sphincter are marked with temporary silk sutures

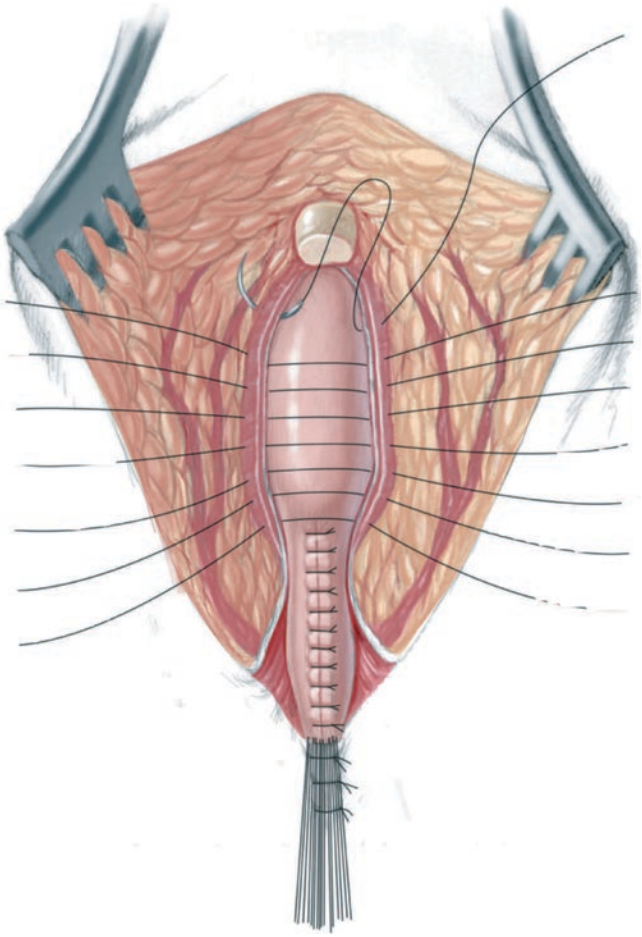

Fig. 31.13 Suturing of the posterior edges of the levator muscle and the posterior edge of the muscle complex

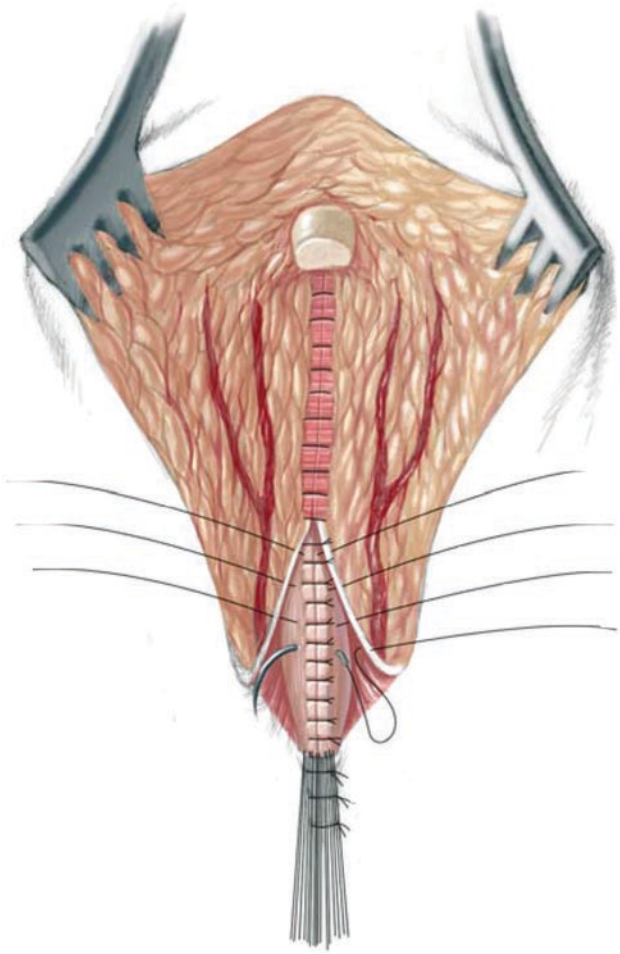

Fig. 31.14 Closure of the rest of the incision, with all layers of the wound reapproximated

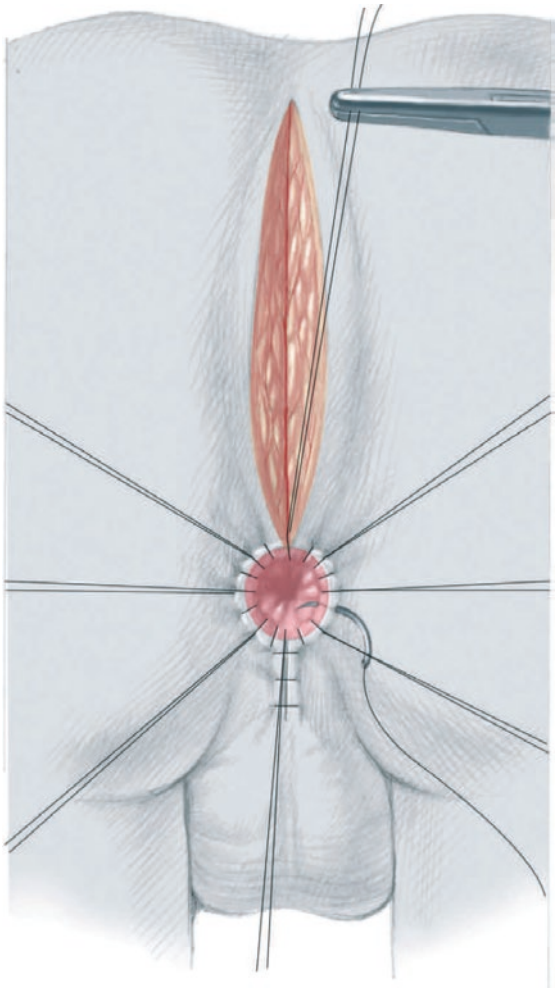

Fig. 31.15 Anoplasty, performed with 16 circumferential stitches of 6/0 long-term absorbable sutures

31.2.3 Recto–Bladder Neck Fistula

This malformation is the highest of all defects seen in male patients. It occurs in 10% of patients, and 90% of them have associated defects. Usually, the sacrum is hypodeveloped. This particular group of malformations is the only one that requires, in order to be repaired, not only a posterior sagittal approach but also an abdominal one, either by laparotomy or laparoscopy.

Minimally invasive procedures have been extended to anorectal malformations, and we believe that they have specific indications in patients who formerly required a laparotomy. In a case of recto–bladder neck fistula, the rectum can be separated from the urinary tract laparoscopically, avoiding a laparotomy. Unfortunately, these patients do not have a good functional prognosis. In our experience, only 15% of them have voluntary bowel movements by the age of 3. These patients require a posterior sagittal approach to create the space through which the rectum will be pulled down. During the laparotomy or laparoscopy, the surgeon must separate the rectum from the urinary tract. Fortunately, in these very high malformations, the common wall between the rectum and the urinary tract is very short. In other words, the rectum connects to the bladder neck in a "T" fashion (Fig. 31.16).

When we operate on these patients, we perform a total body preparation, including the entire lower half of the body, in order to have access to the perineum and to the abdomen, when necessary. The operation is started via laparoscopy. The distal rectum is dissected and then ligated where it narrows down and connects to the bladder neck. To avoid damage to the ureters and vas deferens, the surgeon must keep in mind that they run very close to the distal rectum as they approach the trigone of the bladder. Adequate length is gained on the rectum. The legs are then lifted up and a midsagittal incision is performed. Using the concavity of the sacrum as a guide, a mosquito clamp is safely passed into the pelvis posterior to the ligated bladder neck fistula, and the distal rectum is grasped (Fig. 31.17). The anoplasty is performed as previously described (Fig. 31.18), and the posterior sagittal incision is closed.

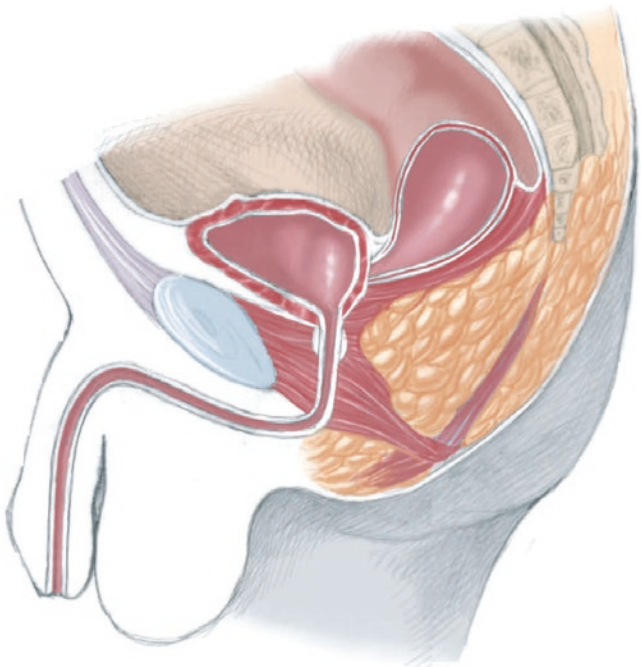

Fig. 31.16 Recto–bladder neck fistula

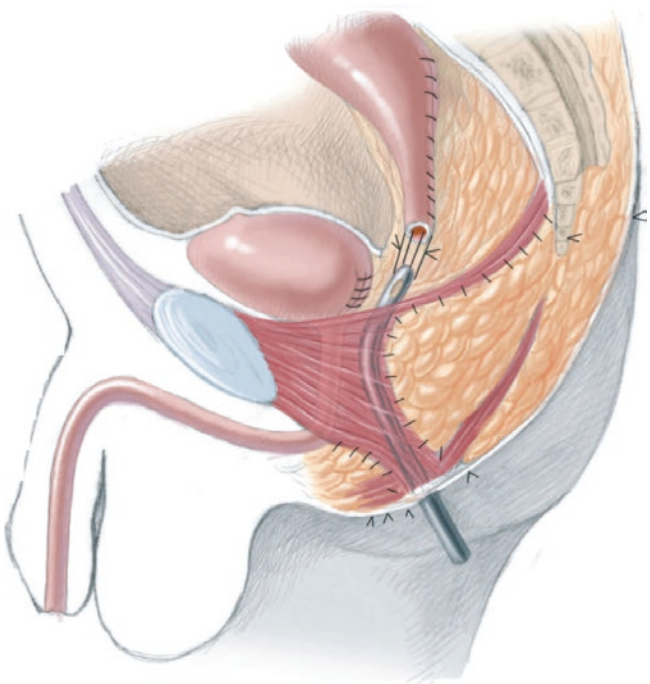

Fig. 31.17 A mosquito clamp is used to grasp the distal rectum posterior to the ligated bladder neck fistula

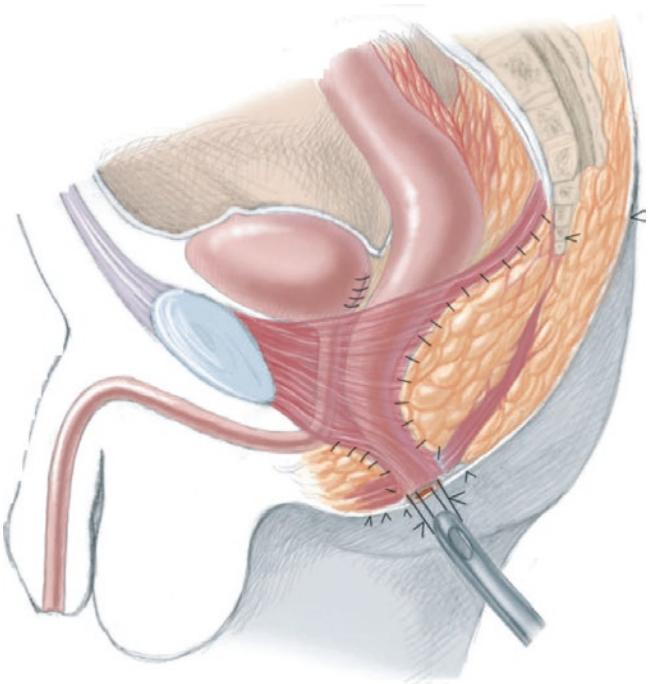

Fig. 31.18 The rectum is pulled down and anoplasty is performed

31.2.4 Imperforate Anus Without Fistula

This particular malformation (Fig. 31.19) is unique. The height of the defect is located approximately 1–2 cm above the perineal skin, at the level of the bulbar urethra. This malformation is found in only in 5% of cases, half of whom have Down syndrome. The patients with these defects have good prognosis, with a good sacrum and good sphincters. Of our patients with this defect without Down syndrome, 90% have bowel control, and 80% of our Down syndrome patients have bowel control. The technique to repair this malformation is not necessarily simpler than the one for rectourethral bulbar fistula, because the rectum is intimately attached to the posterior urethra. The surgeon must open the posterior rectal wall and create a plane of dissection between the anterior rectal wall and the urethra, a manoeuvre that requires meticulous dissection.

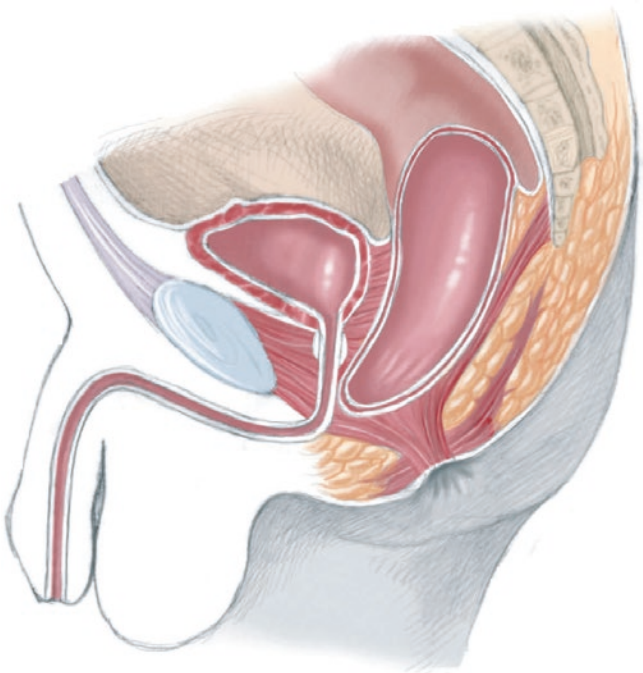

Fig. 31.19 Imperforate anus without fistula

31.3 Female Defects

There are also a spectrum of defects in females. We have formed the view that many patients described as having had an operation to repair a "rectovaginal fistula" actually never had that particular defect. Rather, most of them suffered originally from a cloaca that was misdiagnosed; the surgeon repaired the rectal component of the defect and left the patient with a persistent urogenital sinus. Or the baby may have been born with a rectovestibular fistula and the surgeons misdiagnosed it as a rectovaginal fistula and performed an abdominoperineal procedure for a malformation that could have been repaired posterior sagittally with good results; sadly, the patient was left incontinent for faeces.

31.3.1 Rectoperineal Fistula

This defect (Fig. 31.20) is equivalent to the rectoperineal fistula already described in males. Bowel control exists in 100% of our patients, and less than 10% have associated defects. The patients are faecally continent with and without an operation, but without repair, they can have incontinence when stools are loose or during athletic activities. Constipation is a constant sequela and should be treated aggressively. (This is true also for male patients with perineal fistulas.) We have learned that the lower the defect, the greater the chance of constipation. We have also learned that constipation is a self-perpetuating and self-aggravating condition that eventually produces severe megacolon, chronic faecal impaction, and overflow pseudo-incontinence; it must be vigilantly avoided.

At our institution, the operation to repair this defect is performed at birth. We offer this operation to our patients to provide an anus of adequate size, perfectly positioned in the sphincter mechanism, and one that creates a normal-length perineal body. We perform this operation during the newborn period, before the baby leaves the hospital. Delayed surgery is acceptable but should be performed in the first few months of life.

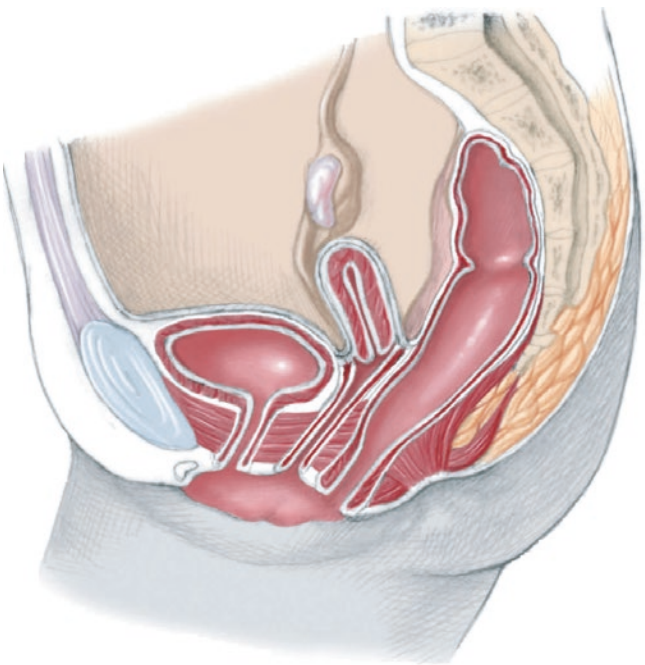

Fig. 31.20 Rectoperineal fistula in a female

The patient is placed in prone position with the pelvis elevated. Multiple 6/0 silk stitches are placed around the fistula site. The incision is about 1.5–2 cm long and divides the sphincter mechanism in the posterior midline. We dissect the rectum as previously described for perineal fistulas in male patients, but in this case it is dissected off of the posterior wall of the vagina. The perineal body is reconstructed as shown and the rectum is anchored to the posterior edge of the muscle complex. An anoplasty is performed.

These patients are kept NPO for 7 days to ensure adequate healing. Antibiotics are given for 24 h.

31.3.2 Rectovestibular Fistula

This defect (Fig. 31.21) is perhaps the most important anorectal malformation in females. It is by far the most common defect seen in females, and it has an excellent functional prognosis when managed correctly. Unfortunately, girls with these defects often suffer from more complications after a failed attempt at repair. For many years, it has been very controversial whether this malformation should be treated with a protective colostomy or should be repaired primarily at birth. Again, we believe that this depends very much on the experience of the surgeon. When a baby is born with this malformation at our institution, we repair this malformation during the newborn period if the baby is otherwise healthy. If the baby is premature or has associated defects, it is always safer to open a protective colostomy or dilate the fistula until definitive repair can be performed. The surgeon must keep in mind that dehiscence and infection in patients with anorectal malformations not only represent a few more days in the hospital and an ugly scar but also represent the possibility of worsening the prognosis for bowel control.

Patients with a vestibular fistula have an excellent prognosis. Bowel control exists in 95% of our patients; 70% have constipation that is manageable but needs to be treated aggressively.

The most important anatomic feature that the surgeon should understand is that the rectum and vagina share a long common wall that must be separated, creating a plane of dissection where it does not exist, in order to mobilize the

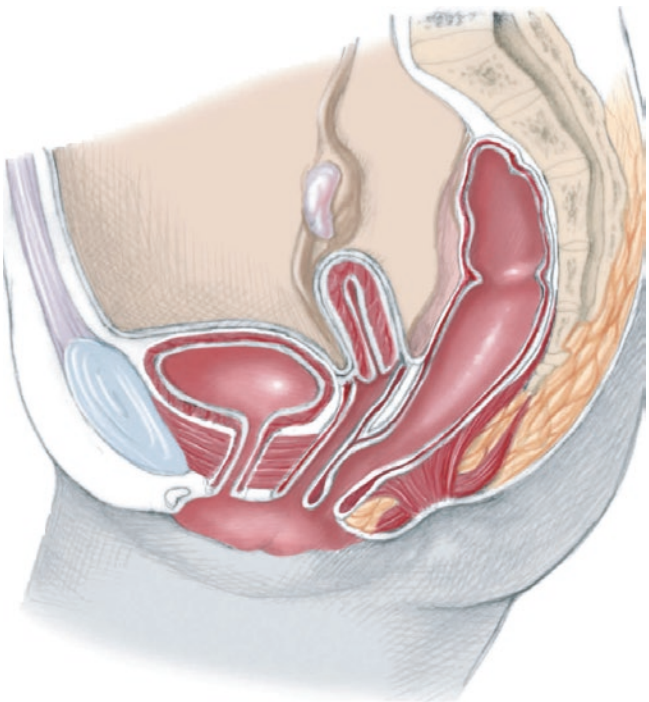

Fig. 31.21 Rectovestibular fistula in a female

rectum and put it in the right place. We believe that most of the complications that we have seen from treatment of this malformation originate from the lack of adequate separation of these two structures, which leaves tension on the anoplasty and leads to dehiscence.

For surgical repair, these patients are placed in prone position with the pelvis elevated. The incision usually runs from the coccyx down to the fistula site. Multiple 6/0 sutures are placed circumferentially in the fistula opening. Traction always facilitates the dissection of these delicate structures. The sphincter mechanism is divided posteriorly until the posterior rectal wall is identified, and then the plane of dissection is established, removing the fascia that covers the rectum. The dissection then continues laterally. The surgeon must put special emphasis on a very meticulous separation of the rectum from the vagina. The purpose of this dissection is to make two walls out of one. Holes in the rectal wall or the vaginal wall should be avoided. Once the rectum has been completely separated from the vagina, the surgeon determines the limits of the sphincter electrically and reconstructs the perineal body, bringing together the anterior limits of the sphincter.

The rectum is then placed within the limits of the sphincter as well as the muscle complex. Only the lower part of the levator is visible because the incision is rather limited in this operation. The rectum is anchored to the posterior edge of the muscle complex (Figs. 31.22 and 31.23) and the anoplasty is performed as in the previously discussed cases. When the patient is a newborn, we keep the baby 5–7 days with nothing by mouth postoperatively, giving intravenous antibiotics for 24 h. Occasionally, we see patients that come later in life without a colostomy; in those cases, we clean the bowel meticulously the day before surgery with a balanced electrolyte solution and keep the patient 7 days with nothing by mouth, receiving concentrated glucose solution. Following this routine, we reduce the incidence of perianal dehiscence.

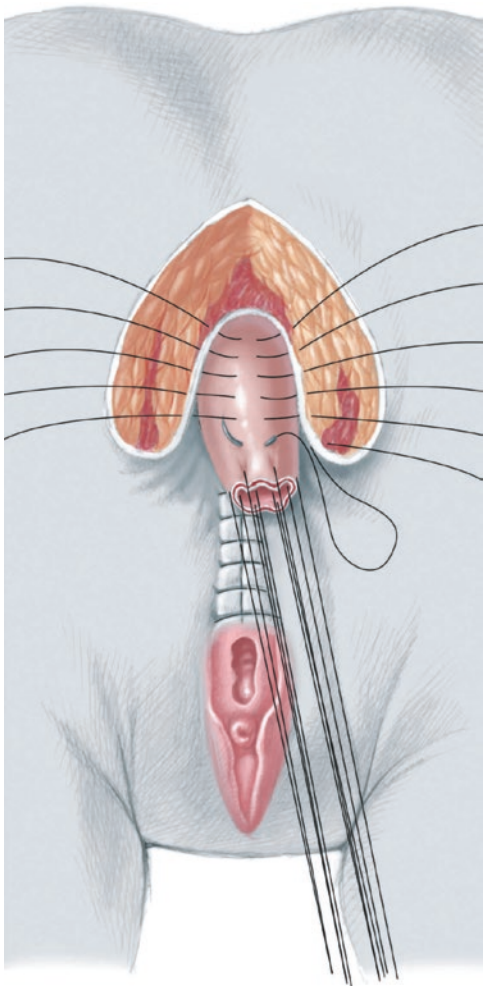

Fig. 31.22 In repairing a rectovestibular fistula, the rectum is anchored to the posterior edge of the muscle complex

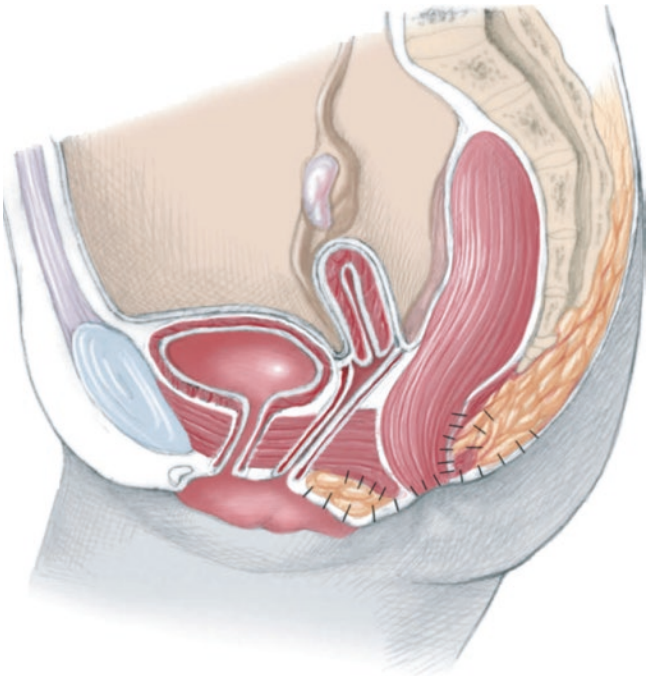

Fig. 31.23 Completed repair of a rectovestibular fistula

31.3.3 Cloaca

A cloaca is defined as a malformation in which the rectum, vagina, and urethra are congenitally fused, forming a common channel and opening in a single perineal orifice at the same location where the normal female urethra is located (Fig. 31.24). These three structures share common walls that are very difficult to separate, and the length of the common channel has important therapeutic and prognostic implications. Cloacas represent a spectrum of malformations, and we have been learning many important lessons about their treatment.

It was 1982 when the first patient with a cloaca was operated on using the posterior sagittal approach. Since then, we have operated on over 500 patients with cloaca. Based on our experience, we now believe that there are two distinct groups of patients with cloacas—those with a common channel shorter than 3 cm and those with a common channel longer than 3 cm—and it is important for the pediatric surgeon and pediatric urologist to recognize this separation.

Cloacas with a short common channel (<3 cm) represent the majority of all cloaca patients, so we believe that there may be enough cloacas of this type in the world for general pediatric surgeons to learn to repair them successfully. Cloacas with a short common channel can be repaired by the posterior sagittal route, without opening the abdomen.

On the other hand, cloacas with a common channel longer than 3 cm, because of their complexity and rarity, should be repaired by individuals with special training that includes a special dedication to pediatric urology. Patients suffering from cloaca with a short common channel have a relatively low incidence of associated urologic, spinal, and vertebral defects, whereas patients with long cloacas have a very high frequency of associated complex pelvic anomalies (mainly urologic, but also vertebral), which make the repair of this group very challenging.

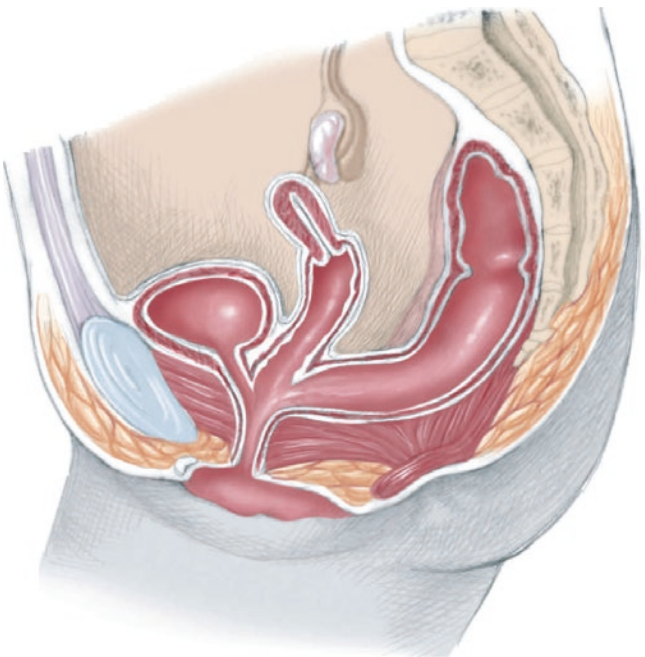

Fig. 31.24 A cloaca

When a baby is born with cloaca, the surgeon must keep in mind that approximately 50% of these patients suffer from a very giant vagina full of fluid ("hydrocolpos"). The hydrocolpos may compress the trigone, interfering with the drainage of the ureters and therefore provoke bilateral megaureters and hydronephrosis.

All babies with a cloaca should have a complete urologic evaluation at birth, including an ultrasound of the kidneys and ultrasound of the pelvis. The baby should not be taken to the operating room without this evaluation. If the baby suffers from hydrocolpos, it is mandatory that the surgeon drain the hydrocolpos at the same time that he or she opens a colostomy. Not draining a hydrocolpos may produce persistent hydronephrosis and induce an inexperienced pediatric urologist to perform ureterostomies or nephrostomies when they are not indicated. The drainage of the vagina usually takes care of the problem of hydronephrosis. Failure to drain a tense hydrocolpos also may produce infection of the vagina (pyocolpos), perforation, and sepsis.

The colostomies must be completely diverting (separated stomas) to avoid contamination of the urinary tract. The definitive cloaca repair can take place during the first few months of the baby's life, provided that she is growing and developing normally.

About 30% of the patients also have duplicated Mullerian structures. In other words, they have different degrees of septation of the hemi-vaginas and two hemi-uterii. These have significant potential obstetric implications.

Before the planned operation, the patient is placed in lithotomy position and a cystoscopy is performed. If the surgeon finds that the patient has a cloaca with a common channel shorter than 3 cm, it may be assumed that the malformation can be repaired posterior sagittally. Surgeons may predict that they do not have to open the abdomen and that the final functional prognosis, both for urinary and faecal function, is going to be reasonably good. On the other hand, if the patient has a longer common channel, she must be operated on by a highly experienced and skilled surgeon or team of surgeons.

In cases of a short common channel, the patient is placed in a prone position with the pelvis elevated. The incision runs from the middle portion of the sacrum all the way down to the single perineal orifice. The entire sphincter mechanism is divided posteriorly and then the most posterior structure (usually the rectum) is exposed.

A total urogenital mobilization is performed. The rectum is separated from the vagina in the same way as in the cases of rectovestibular fistula. Once we expose the malformation,

multiple 6/0 silk stitches are placed, taking the edges of the common channel and the edges of the vaginal walls. We use those stitches to exert a uniform traction (Fig. 31.25). Then urethra and vagina together are mobilized as a total urogenital mobilization. Another set of sutures is placed in a transverse fashion, taking the mucosa of the common channel 5 mm proximal to the clitoris, and then the entire common channel is divided, full thickness, creating a plane of dissection between the pubis and the common channel. This is a natural plane of dissection and therefore it is very easy to dissect. We reach the upper part of the pubis, and there we can identify avascular fibrous structures, the suspensory ligaments of the urethra, which fix the genitourinary structures to the pelvis. These are divided, and that allows an immediate gain of length of the urogenital structures of approximately 2–4 cm. With some additional dissection of the lateral vaginal wall, this gain is enough to repair a malformation with a short common channel.

The goal of this mobilization is to move what used to be a urethral meatus all the way down, to be placed immediately behind the clitoris, so it is visible and accessible for future urethral catheterizations when indicated. The vagina comes down together with the urethra, and the edges of the vagina are sutured to the skin of the perineum, forming the new labia. What used to be the common channel is divided in the midline, creating two flaps that are preserved and sutured to the new labia. All this reconstruction is performed with interrupted 6/0 long-term, absorbable sutures (Fig. 31.26).

With these kinds of techniques, 80% of patients with a good sacrum have bowel control, and approximately 80% remain dry of urine and can empty the bladder voluntarily without intermittent catheterization; 20% require intermittent catheterization to empty the bladder. Fortunately, they have a good bladder neck and they remain dry between catheterizations. When the sacrum is very abnormal or there is an associated tethered cord, the results are not as good, and the need for intermittent catheterization is higher.

The repair of cloacas with longer common channels (>3 cm) represents a real technical challenge and requires a great deal of experience in the management of these cases. For that kind of patient, the pediatric surgeon and/or pediatric urologist should have experience in the management of the urinary structures, including bladder reconstructions, bladder neck reconstructions, ureteral reimplantations, bladder augmentation, and Mitrofanoff procedures, as well as vaginal replacements using rectum, colon, or small bowel.

The repair of cloacas with long common channels should begin with total body preparation for access to both the perineum and the abdomen. If the common channel is longer than 5 cm, we recommend opening the abdomen directly in the midline, because the rectum and vagina are not accessible posterior sagittally and are more easily accessed through the abdomen. If the common channel is between 3 and 5 cm, the surgeon can open posterior sagittally, find the vagina or vaginas, and can try to repair them by performing the total

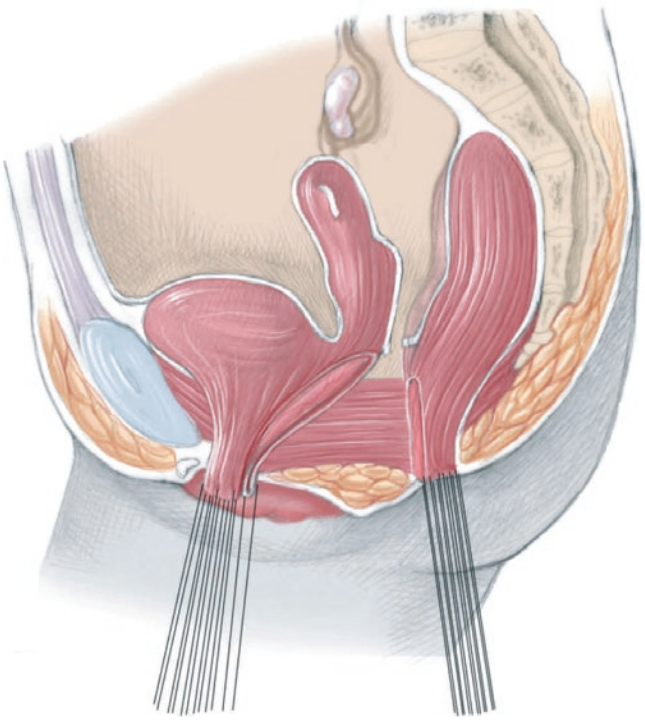

Fig. 31.25 Stitches placed to provide traction for repair of a cloaca with a short common channel

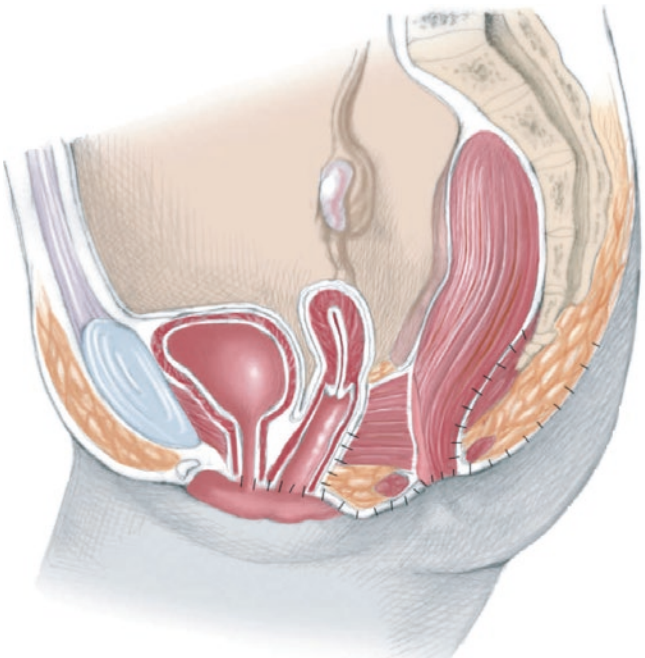

Fig. 31.26 Completed repair of a cloaca

urogenital mobilization. If the total urogenital mobilization proves not to be enough to repair the malformation, then the operation must be completed through a laparotomy. The separation of the rectum from the vagina is not difficult in very high malformations; it is similar to what we described in the separation of the rectum from the bladder neck in male patients. On the other hand, the separation of the vagina from the urinary tract in a case of a cloaca with a long common channel is a very delicate manoeuvre that requires expertise and finesse. Once the vagina has been completely separated (which may take several hours), the surgeon then has to make important decisions about how to repair the vagina. The separation of the vagina from the urinary tract should be performed with the bladder open in the midline and with ureteral catheters. The ureters run through the common wall that separates the vagina from the bladder, so the surgeon should be ready to deal with the fact that the ureters may be right in the plane of separation. Once the vagina has been separated, the surgeon should evaluate the size of the mobilized vaginas and the length that is needed to bring the vagina down. The patient may have a very large vagina that reaches the perineum, or a vaginal replacement using colon or small bowel may be needed.

31.4 Outcome

Clinical results differ for each type of malformation. A recent review of the authors' series showed that 100% of patients with rectal atresia and perineal fistula had voluntary bowel movements, as did 95% of those with vestibular fistula, 90% with bulbar urethral fistula, 85% with imperforate anus without fistula, 80% with cloaca, 75% with rectourethral prostatic fistula, and 25% with bladder neck fistula. Soiling in patients who have voluntary bowel movements usually represents a manifestation of faecal impaction and disappears when constipation is treated properly. Patients who have voluntary bowel movements and never soil are considered "totally continent".

Constipation is a common sequela seen after the repair of an anorectal malformation. Interestingly, patients with lower defects, and therefore with better prognosis for bowel control, suffer a higher incidence of constipation and vice versa. Constipation correlates directly with the degree of rectosigmoid dilation at the time of colostomy closure. Therefore, every effort should be made to try to keep the rectosigmoid empty and decompressed from day 1 in these patients.

Urinary control can be expected in the overwhelming majority of male patients after repair of imperforate anus, provided that a good surgical technique was performed. Urinary incontinence is a concern only in male patients with absent sacrum or spinal anomaly or in some female patients with cloaca. In cloacas, when the common channel is shorter than 3 cm, approximately 20% of patients require intermittent catheterization to empty the bladder. The remaining 80% have urinary control. When the common channel is longer than 3 cm, 80% of the patients require intermittent catheterization to empty the bladder. Fortunately, after the repair of a cloaca, patients have a good bladder neck. When the bladder becomes completely full, the patient starts suffering from overflow urinary incontinence, but intermittent catheterization keeps these patients completely dry.

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Intussusception

32

Holger Till and Erich Sorantin

32.1 Introduction

Intussusception represents the most common abdominal emergency in infancy and early childhood. The overall incidence is about 1–4%, and approximately 95% of all intussusceptions occur within the first 2 years of life. Interestingly, neonates are affected in less than 1% of all cases.

By definition, intussusception means that one portion of the bowel is telescoping into the adjacent distal portion. Such phenomena probably happen quite often with ongoing peristalsis of the small intestine and recover spontaneously, but once the bowel remains telescoped, especially through the ileocoecal valve, its blood supply becomes increasingly impaired. Progressive oedema of the bowel wall finally leads to mucosal bleeding, insufficient arterial blood supply, increased venous stasis, and even bowel necrosis.

This pathophysiology explains the typical clinical symptoms, which in the early stage are predominantly vomiting and intermittent abdominal pain. These colicky attacks typically cease as quickly as they began, and the child is quiet in between. With increasing bowel wall oedema and compression of the mesenteric blood supply, bilious vomiting, abdominal distension, dehydration, tachycardia, fever, and shock will develop. These severely sick patients must be stabilized before being taken to radiology.

Examining the child, the right lower quadrant seems empty, but a tender mass—mostly in the right upper quadrant—can be felt in about 85% of cases. In a later stage of intussusception, mucous clots and occult or even gross blood may be seen on rectal examination.

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Anatomically, the ileocolic form is the most common type, representing 80–90% of all cases, followed by ileo-ileal forms. Usually a "solid" leading point of the intestinal wall serves as the head of the intussusception, being drawn into the distal gut by peristaltic activity. In infants and younger children with gastrointestinal infections by adenoviruses or rotaviruses, the swollen Payer's plaques protrude into the lumen of the bowel, initiating intussusception. In older children, pathological leading points such as Meckel's diverticulum, polyps, appendix, or a duplication cyst may be expected. Rarely, intramural haematoma (as in Schönlein-Henoch disease) or neoplastic lesions like a carcinoid tumour or a Burkitt lymphoma could be present.

In most paediatric centers around the world, children with intussusception are managed conservatively because of the high efficacy of a nonsurgical reduction. Nevertheless, surgery remains important in cases with incomplete reductions, especially when additional pathologies are suspected or complications such as perforations or ischemic bowel occur. According to Sorantin, there are several cornerstones of management:

- Confirmation or exclusion of intussusception
- Confirmation or exclusion of complications
- Performance of nonsurgical reduction
- Identification of other pathologies mimicking intussusception

A variety of imaging modalities are available and highly efficient for such a workup. The following list suggests the order in which the various techniques are employed:

- Ultrasound, including Doppler ultrasound
- X-ray films in two planes
- Diagnostic enema

In most centres, ultrasound represents the first choice to diagnose intussusception, because it is readily available, uses

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no radiation, and provides a diagnostic sensitivity of 98.5% and specificity of 100%. Several findings are characteristic:

- Soft tissue mass, usually having a diameter between 2.5 and 5.0 cm
- "Doughnut sign" (Fig. 32.1): The edematous bowel wall appears as a hypoechoic or anechoic external rim, whereas the hyperechoic center represents the intussuscepted mesenteric fat. The *white arrows* indicate a lymph node within the intussusception.
- "Target sign" (a variant of the "doughnut sign"): Within the central echogenic area there is an additional anechoic area (which probably represents fluid at the apex of the intussusceptum), resulting in a targetlike appearance.
- "Multiple concentric ring sign": If the oedema is less severe, the various layers of the mucosa and submucosa are less stretched and thinned, and form multiple concentric rings
- "Pseudo-kidney sign": In the longitudinal scan, the intussusception resembles a kidney, with a hypoechoic rim and a hyperechoic center

Doppler ultrasound provides information about the vascular supply of the intussusception. Finding of a reduced perfusion is not a contraindication to nonsurgical reduction. Figure 32.2 shows an ultrasound with colour Doppler in the lower right quadrant. A target sign indicates intussusception. The colour Doppler tracings indicate normal perfusion in this case at the time of investigation.

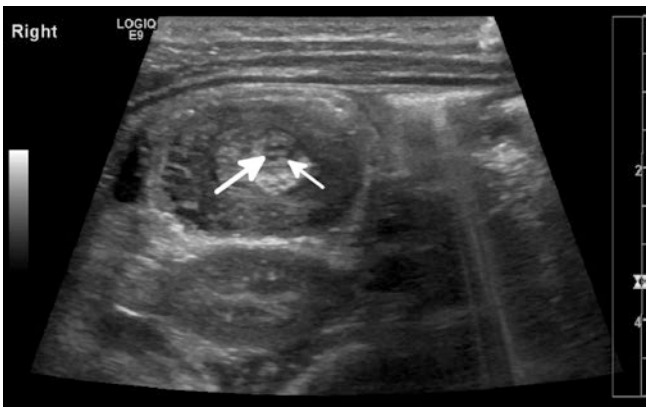

Fig. 32.1 The "doughnut sign" on ultrasound, representing intussusception

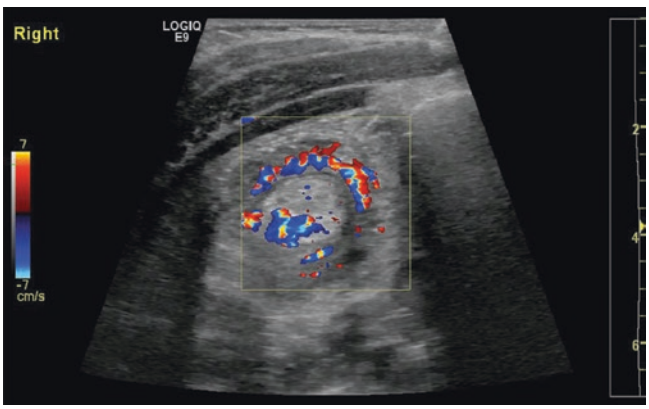

Fig. 32.2 Ultrasound with colour Doppler, showing a target sign representing intussusception, but normal perfusion

32.2 Nonsurgical Treatment

Before starting any conservative treatment the paediatric radiologist should inform the paediatric surgeon and the operating staff in case incomplete reduction or perforation occurs. Basically, there is no contraindication for any conservative trial of reduction with respect to duration of intussusception or position of the apex, but nonsurgical treatment must not be undertaken in a patient with clinical signs of shock, peritonitis, or severe obstruction.

Nonsurgical reduction has a longstanding history; it was first described by McDermott in 1994. In principle, an enema pushes back the intussuscepted bowel. The progress of the reduction can be monitored by fluoroscopy or ultrasound. As contrast media, air or water-soluble iodine solutions can be used for fluoroscopy, and physiologic saline, for ultrasound. A success rate of up to 95% has been reported for nonsurgical reduction, so this procedure should be considered as a first-line treatment.

At the authors' institution, air is insufflated through a rectal tube under fluoroscopy control and the intussusception is outlined as a soft tissue structure, which moves orally due to the air insufflation. The procedure is successful once the air influxes into the small bowel, usually as a blush. The basic guideline is that no more than three attempts should be performed, with a maximum duration of 3 min and a maximum pressure of 100 mm Hg. If complete reduction cannot be achieved, surgery should be considered.

Figure 32.3 uses fluoroscopy (last image hold technique) to depict a nonsurgical reduction. The intussusception appears as soft tissue mass (*white asterisk*), and small and large *white arrows* outline the borders of the intussusception. In Fig. 32.4, there is free inflow of air in the small bowel, confirming the complete reduction. The *white arrows* mark the swollen lips of the ileocoecal valve.

When ultrasound is used to monitor the nonsurgical reduction, the saline enema must be followed from the rectum to the small bowel, requiring the sonographer to be skilled in bowel ultrasound to depict the typical findings and appearance. Figures 32.5 and 32.6 illustrate a nonsurgical reduction by ultrasound guidance and saline enema, in the right lower quadrant, axial plane. In Fig. 32.5, the *asterisk* marks the intussusception within the ileocoecal valve and the *"X"* depicts the saline enema. Figure 32.6 documents the successful reduction of the ileocolic intussusception. The *asterisk* marks the ileocoecal valve lips and the *arrows* show the opening area of the valve with free inflow of the saline enema to the terminal ileum.

Bowel perforation can occur as a complication of the procedure, requiring immediate decision-making. Tension

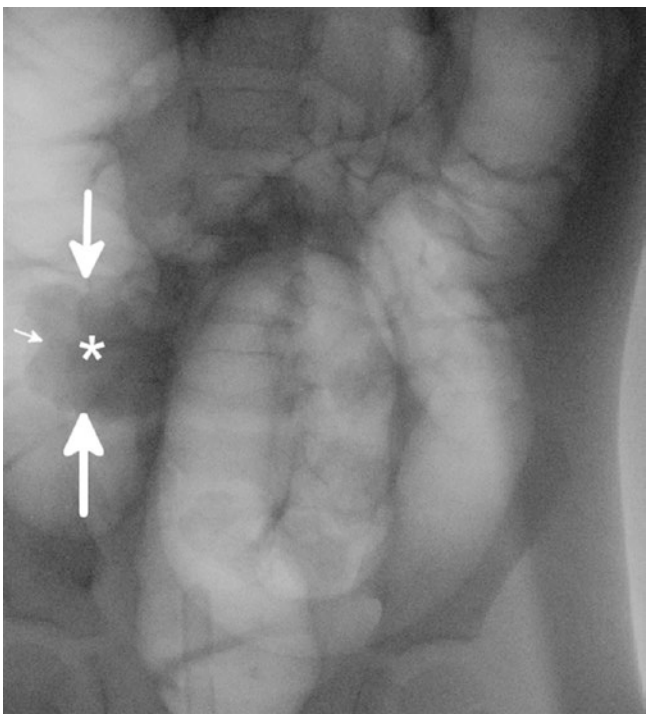

Fig. 32.3 Fluoroscopy depicting nonsurgical reduction of intussusception

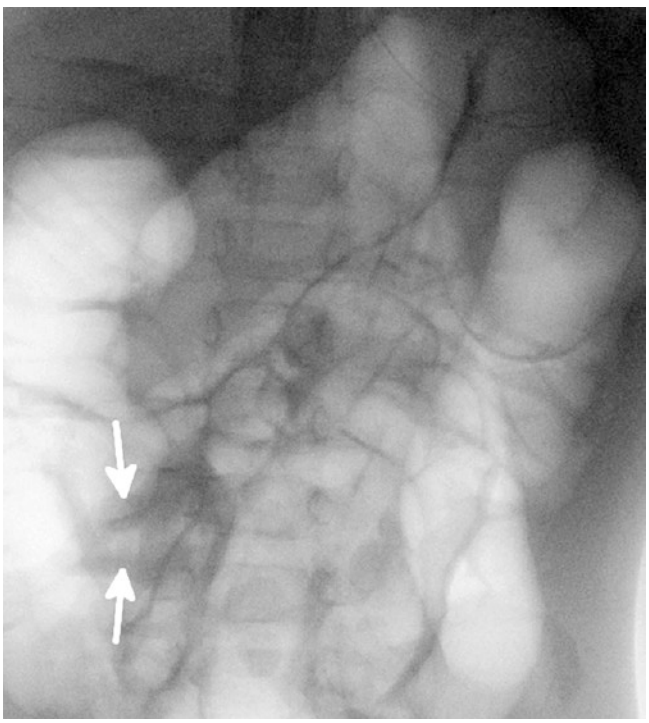

Fig. 32.4 Fluoroscopy showing free inflow of air in the small bowel, confirming the complete reduction of intussusception

pneumoperitoneum due to trapped air (at pressures of up to 100 mm Hg) is an extremely rare but serious complication that may require immediate needle decompression to prevent hemodynamic compromise and cardiovascular shock. All other cases with incomplete nonsurgical reduction or perforation are taken immediately to the operating theatre for surgical treatment.

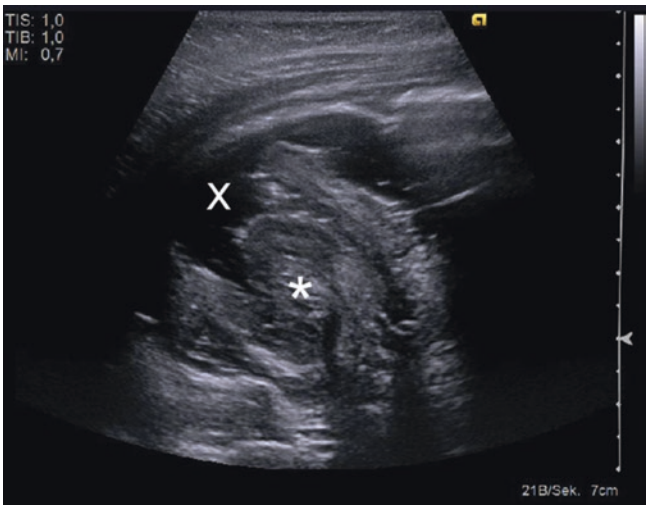

Fig. 32.5 Ultrasound depicting a nonsurgical reduction procedure by saline enema

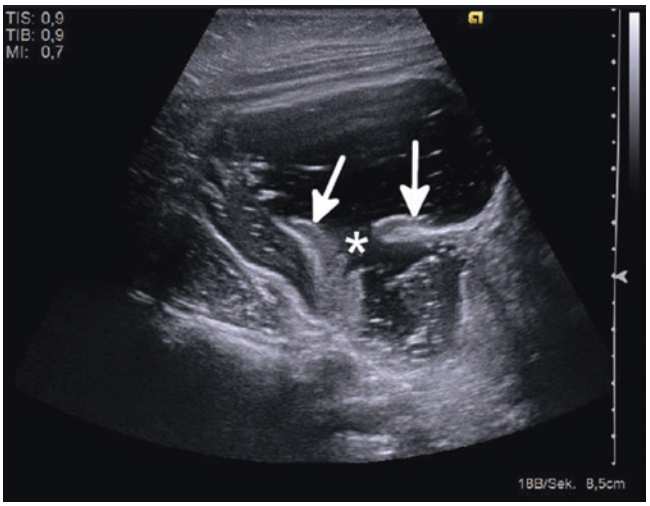

Fig. 32.6 Ultrasound showing successful reduction of an ileocolic intussusception

32.3 Surgical Treatment

The indications for surgery include incomplete radiological reduction, suspected pathological leading point, or cases of perfoation or peritonitis. Preparation of the patient for surgery includes decompression of the gastrointestinal tract by open nasogastric tube, monitoring of vital parameters, and basic laboratory studies. Antibiotics should be started preoperatively, because bacterial translocation can occur during unsuccessful nonsurgical reduction of intussuception.

On the operating table, the child is positioned supine (Fig. 32.7). In cases of incomplete reduction with the remaining apex close to the ileocoecal valve, oblique incision in the right lower quadrant may be sufficient. When the intussusception reaches the transverse colon or progressive disease develops, which may require bowel resection, a transverse abdominal incision may be preferable.

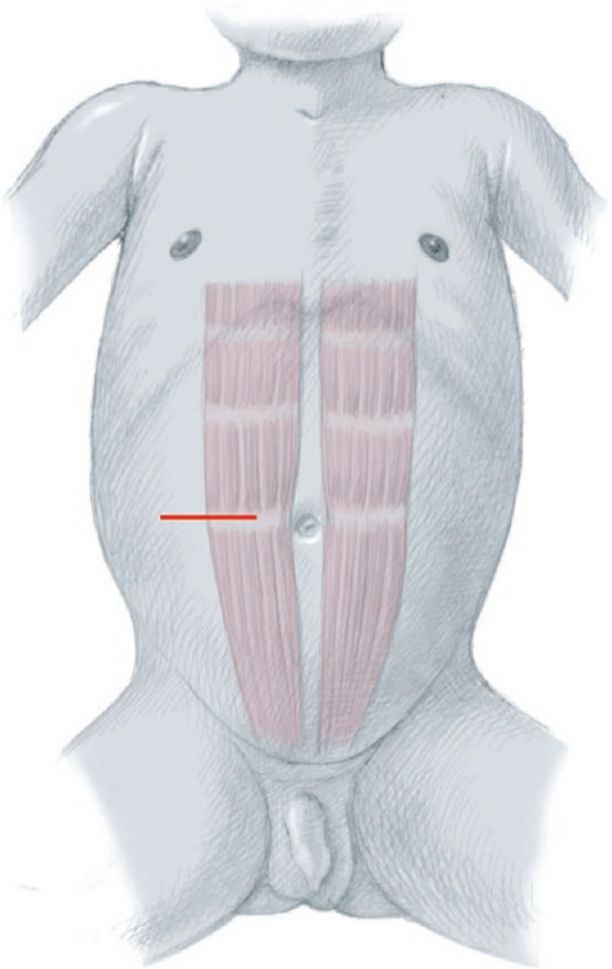

Fig. 32.7 Positioning for surgical treatment of intussusception

By inspection and palpation, the apex of the intussusception is located. In most cases, the cecum is mobile enough to access it adequately for manual reduction. Otherwise, a local dissection of the lateral attachments of the ascending colon may be advisable. Manual reduction must be performed very carefully and slowly. Taking the apex of the intussusceptum between two fingers of one hand and the adjacent part of the distal bowel into the other hand, the intussusceptum is gently squeezed towards the ileocoecal valve (Fig. 32.8). Pulling on the intussuscepted bowel should be avoided, as it causes tears and ruptures. Once the reduction has been successfully performed, the intussusceptum must be inspected. It usually looks quite oedematous or even ischemic, but the color usually improves quickly. Necrotic bowel should be resected.

Furthermore a pathological leading point must be identified by stepwise inspection and palpation of the bowel wall. It is important not to confuse a swollen and thickend Peyer's patch with a pathological leading point. A Meckel's diverticulum must be resected. Most centres recommend a simultaneous appendectomy. If a Burkitt lymphoma is encountered as a leading point, it should be resected along with the intussuception. In advanced stages, sampling of peritoneal fluid and tumour material for cytology, histology, immunophenotyping, and molecular biology will be required (Fig. 32.9).

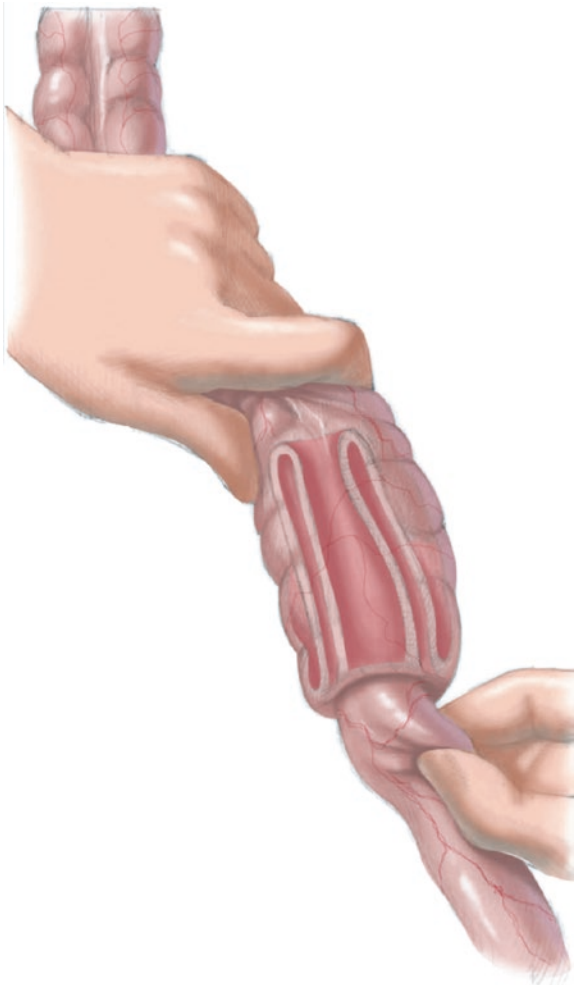

Fig. 32.8 Manual reduction of intussusception

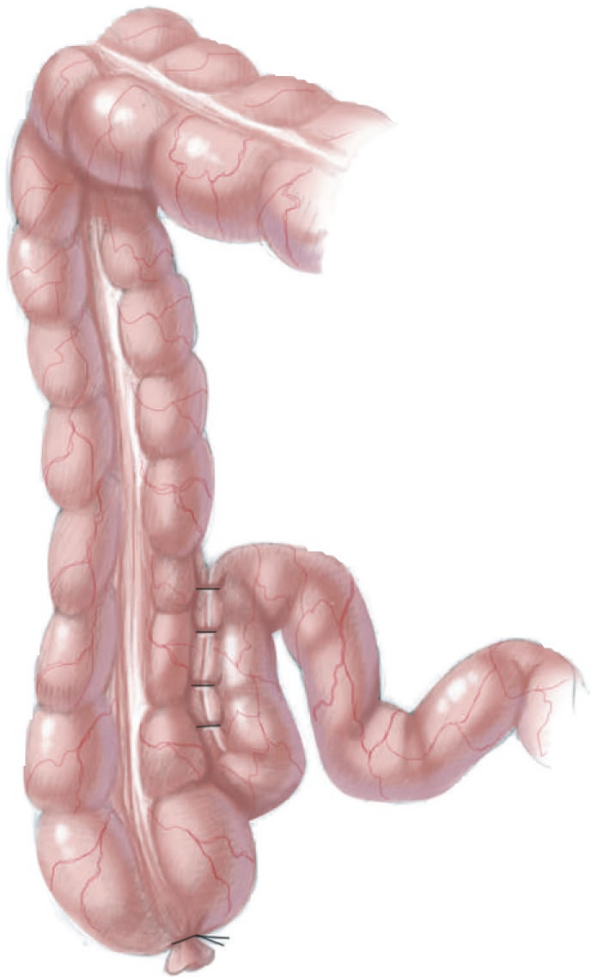

Fig. 32.9 Fixation of the bowel wall

32.4 Postoperative Care

The postoperative management depends on the extent of the disease, surgical procedures performed, and the overall condition of the child. In uncomplicated cases with nonsurgical reduction, enteral feeding should be started the following day. The more complex cases may require a short-term nasogastric tube and intravenuous fluid. Any child who again becomes symptomatic must be evaluated by ultrasound for recurrence. Children who are asymptomatic can be discharged as soon as enteral feeding has been established.

32.5 Recurrent Intussusception

Recurrent intussusception is seen after nonsurgical reduction in up to 13% of all cases, and 30% of all recurrences occur within the first postoperative day. Irritability and discomfort are the first signs of an early recurrence. Nonsurgical reduction may be repeated, especially in infants and younger children with gastrointestinal infections. Hsu et al. analysed the recurrence rate in 686 children and found 15.7% recurrence after the first barium enema reduction, 37.7% after the second, 68.4% after the third, and 100.0% after the fourth reduction. In our institution, we recommend surgery after three recurrences following successful nonsurgical reductions.

Chronic intussusception is a rare condition. It is defined as a nonstrangulating intussusception lasting a minimum of 14 days, mostly due to pathological leading points. The clinical features include chronic diarrhoea and colicky pain. The diagnosis can be easily achieved by ultrasound, contrast studies, or even laparoscopy.

32.6 The Role of Laparoscopy

The role of laparoscopy for cases with intussusceptions remains a matter of discussions. Some authors recommend it as the first line of surgical treatment. It must be remembered, however, that the small intestine in acute intussusception is usually dilated, allowing only small laparoscopic working spaces. Thus the risk of iatrogenic perforations must be considered. Wei et al. found that the operating time and rate of conversion to open surgery were significantly higher in children with the intussusceptum in the transverse and descending colon. The biggest disadvantage of laparoscopy is that the bowel cannot be palpated for pathological lesions as well as in open surgery.

The technique requires three ports. A 5-mm scope with a 30° lens is placed in the umbilicus, preferably with an open Hasson technique because of the dilated bowel. Insufflation pressure should be limited to a maximum of 5–7 mm Hg and the CO_2 flow to 0.5–1.0 L/min. Two more working ports 3–5 mm in diameter are then introduced under direct vision in the left abdomen, aiming for an ergonomic triangle with the tip towards the cecum. Taking two atraumatic graspers, the cecum with the intussuscepted ileum is identified and assessed for the extent of the disease and localization of the apex. In cases of perforation, most experts would immediately convert to open surgery. In uncomplicated cases, the apex is pushed towards the ileocoecal valve with the atraumatic grasper. It must be mentioned again that any pulling, as tempting as it may be, can easily lead to seromuscular tears. Once the laparoscopic reduction has been successfully performed, the surgical tasks remain the same as in open surgery, comprising a careful inspection of the telescoped bowel for viability, perforations, and leading points. Recently, Apelt et al. published a systematic review of the safety and efficacy of the use of minimally invasive surgery in paediatric intussusception. In 10 studies treating 273 cases, the laparoscopic success rate was 71.0%, with a shorter length of hospital stay than the open group (mean 4.0 *vs* 7.1 days; $p < 0.01$) and only one case of laparoscopic intestinal perforation (0.4%). These authors concluded that laparoscopy is safe and effective in the treatment of pediatric intussusception.

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Appendectomy

33

Girolamo Mattioli

Acute appendicitis is the most common surgical emergency in childhood. Appendicitis may present at any age, although it is uncommon in preschool children. Approximately one third of children with acute appendicitis have perforation by the time of operation. Despite advances in improved fluid resuscitation and better antibiotics, appendicitis in children, especially preschool children, is still associated with significant morbidity.

33.1 Diagnosis

The diagnosis of acute appendicitis in childhood can sometimes be difficult. Definite diagnosis is made in only 50–70% of patients at the time of initial assessment. The rate of negative pediatric appendectomy is in the range 3–28% in various reports. The patient's history and clinical examination are the most important tools for the diagnosis of appendicitis. Periumbilical pain is often the first symptom, followed by vomiting and fever. When the inflammation progresses, the pain localizes to the right lower quadrant, and right lower quadrant tenderness develops. Laboratory investigations and plain radiographs are neither sensitive nor specific in the diagnosis of appendicitis. In recent years, ultrasonography of the right lower quadrant has been shown to be a useful tool in the evaluation of patients with clinical findings that are suggestive but not diagnostic of appendicitis. According to the literature, a non-compressible appendix with a diameter of more than 6 mm is considered pathologic. If the appendix is not visible in the ultrasound, indirect signs of inflammation are inflamed fat and/or echo poor formations near the cecal pole with pain or tenderness during compression or free fluid near the cecum. Ultrasound investigation of the appendix has sensitivity of 80–94%, specificity of 90%, and an overall accuracy of 90%. CT imaging may be helpful in selected cases but is rarely needed. Additionally, recent data have shown that repeated CT scans impose a significant radiation dose to the child with the risk of late cancer development.

In patients with an uncertain diagnosis of acute abdominal pain, a policy of active observation in hospital is usually practised. A repeated structured clinical examination is simple and noninvasive.

Children with perforated appendicitis must be treated preoperatively to prevent dehydration and generalized sepsis. Antibiotics against aerobic and anaerobic bacteria are essential to reduce complications and to prepare the patient for the surgical procedure.

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33.2 Surgical Treatment

33.2.1 Open Technique

A transverse incision (Fig. 33.1) (Rockey-Davis incision or Lanz incision) or a slightly oblique right lower quadrant skin crease incision (McBurney incision) across McBurney's point is recommended. The two incisions are similar, but the former gives a better cosmetic result and the latter provides optimal exposure of the appendix.

The muscular layers are split in the direction of their fibres (Fig. 33.2). Transversalis fascia and preperitoneal fat are dissected and the peritoneum is grasped. Peritoneum is opened with a scalpel and free fluid is sent for culture.

The mesoappendix is divided (Fig. 33.3) and the appendiceal base is clamped and ligated (Fig. 33.4). Stump inversion is optional. Several studies have reported no difference as regards wound infection and postoperative fever between one group in which the appendix was ligated and doubly invaginated and another group in which it was simply ligated. If pus is present, the abdomen should be irrigated with saline. Drains are not necessary. The abdominal wall is closed in layers. The skin is usually closed by subcuticular absorbable sutures, even in the case of perforation. Primary wound closure after perforated appendicitis is safe, economical, and advantageous in pediatric practice.

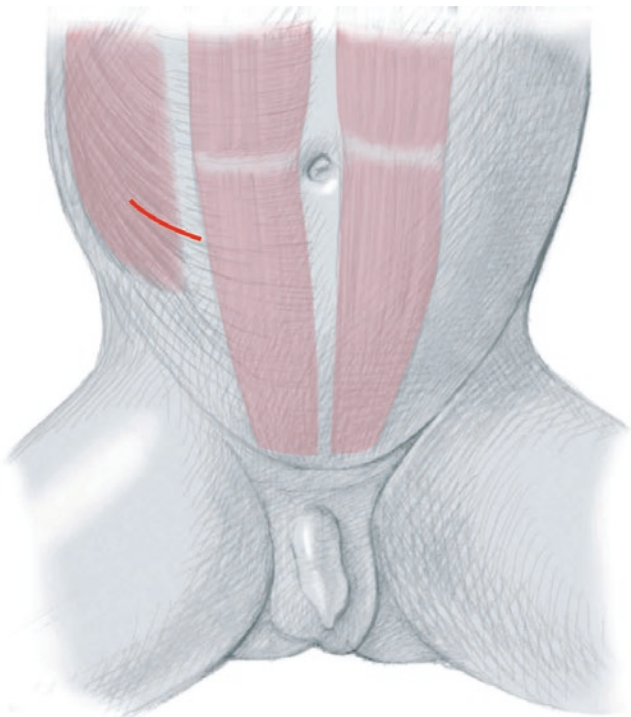

Fig. 33.1 A transverse incision for open appendectomy

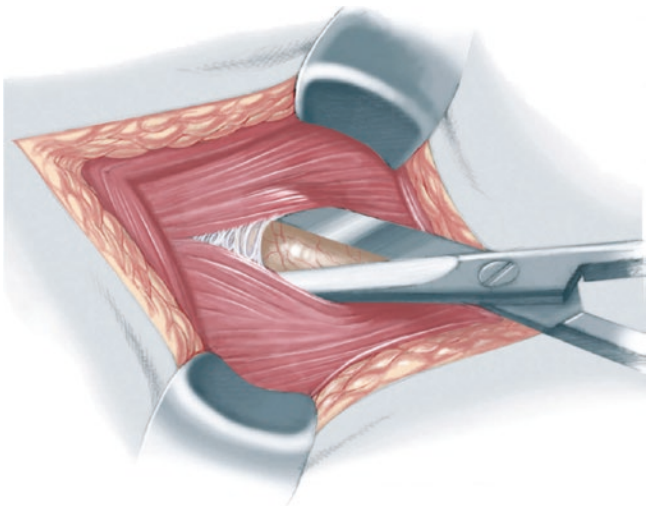

Fig. 33.2 Splitting of the muscle layers and dissection of the transversalis fascia

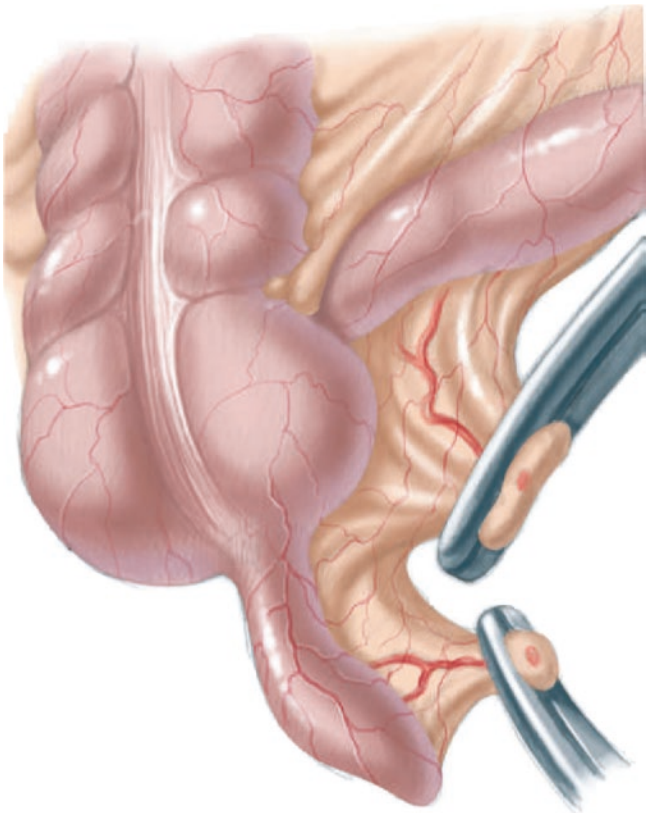

Fig. 33.3 Division of the mesoappendix

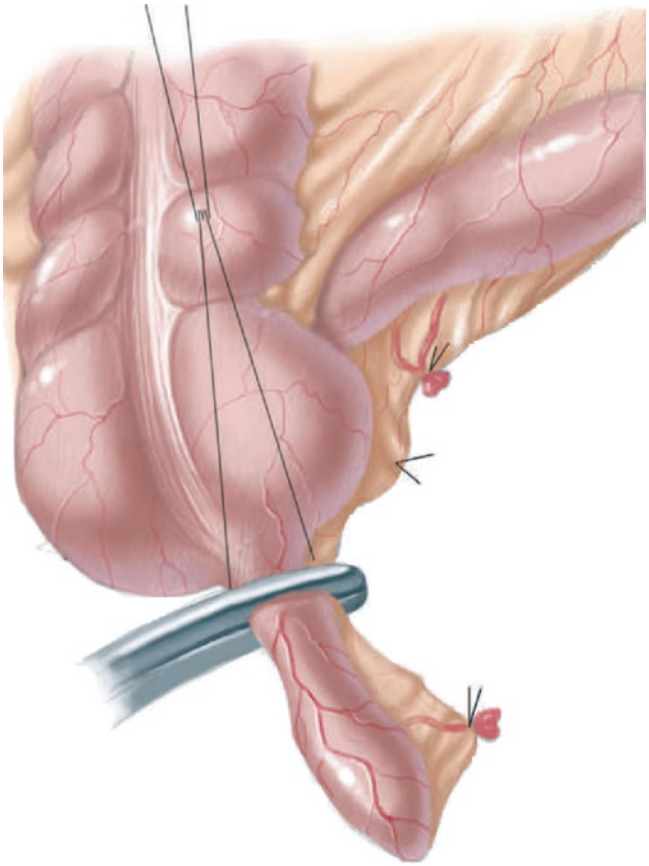

Fig. 33.4 Clamping and ligation of the appendiceal base

33.2.2 Laparoscopy-Assisted Appendectomy

Laparoscopy-assisted appendectomy can be performed using three different methods: the complete laparoscopic approach, the transumbilical laparoscopic appendectomy (TU-LAP), and the single-incision laparoscopic appendectomy (SILA). The complete laparoscopic procedure is performed using three ports (Fig. 33.5). The first is inserted through the navel, initially for the telescope and afterwards for the operating instruments and the stapler or the loops, as well as for the extraction of the appendix. The second port is positioned in the left iliac region for the telescope and for the operating instruments. The third port is positioned in the right iliac region to grab the appendix. A direct transparietal suture is inserted in the right iliac flank to keep the appendix in tension during the dissection. Before resecting the appendix, an exhaustive evaluation of the entire abdominal cavity and pelvic region in female patients must be done.

Skeletonizing of the appendix is performed using a bipolar instrument or a monopolar hook (Fig. 33.6); large vessels are ligated using clips or regular sutures. Generally, clips or sutures are not required for inflamed tissues and small-sized vessels. The base of the appendix is closed and cut using staplers, or it is ligated with two preformed loops and cut between them with scissors. There are no differences between two techniques in terms of postoperative complications.

TULAP is performed using a 10-mm telescope with an operating channel through the umbilicus. The appendix is grabbed and pulled through the umbilicus. The procedure is thereafter completed from outside using a conventional approach through the umbilicus. The advantage of TULAP is the good cosmetic result from the limited dimension of the scars.

SILA is performed using a single, small umbilical incision through which multiple laparoscopic instruments are placed, either through a single port device with multiple conduits or through multiple, closely spaced ports. The benefit of SILA is mostly cosmetic.

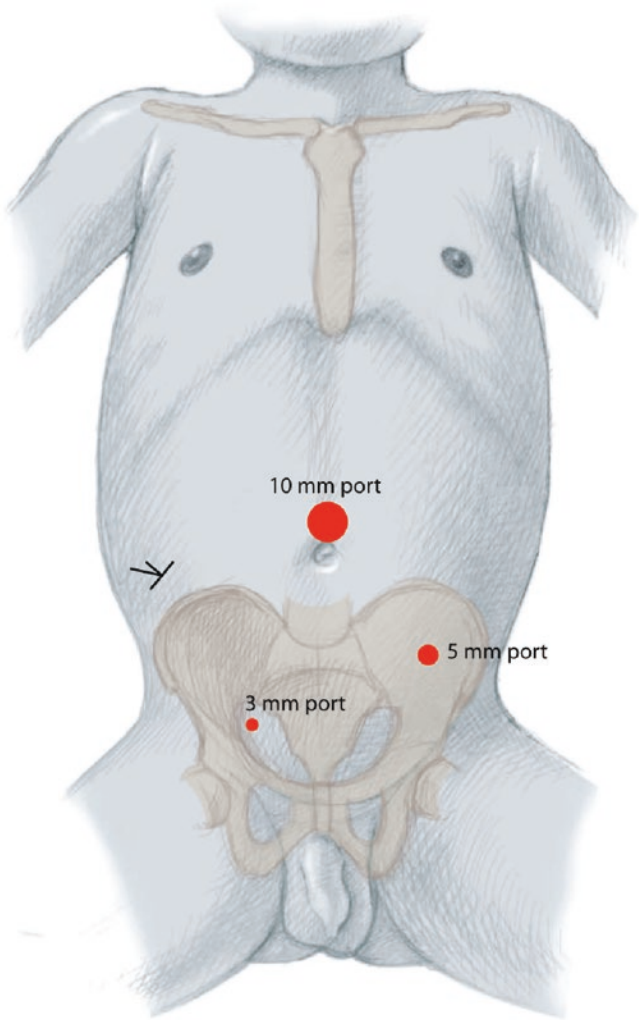

Fig. 33.5 Ports for laparoscopy-assisted appendectomy

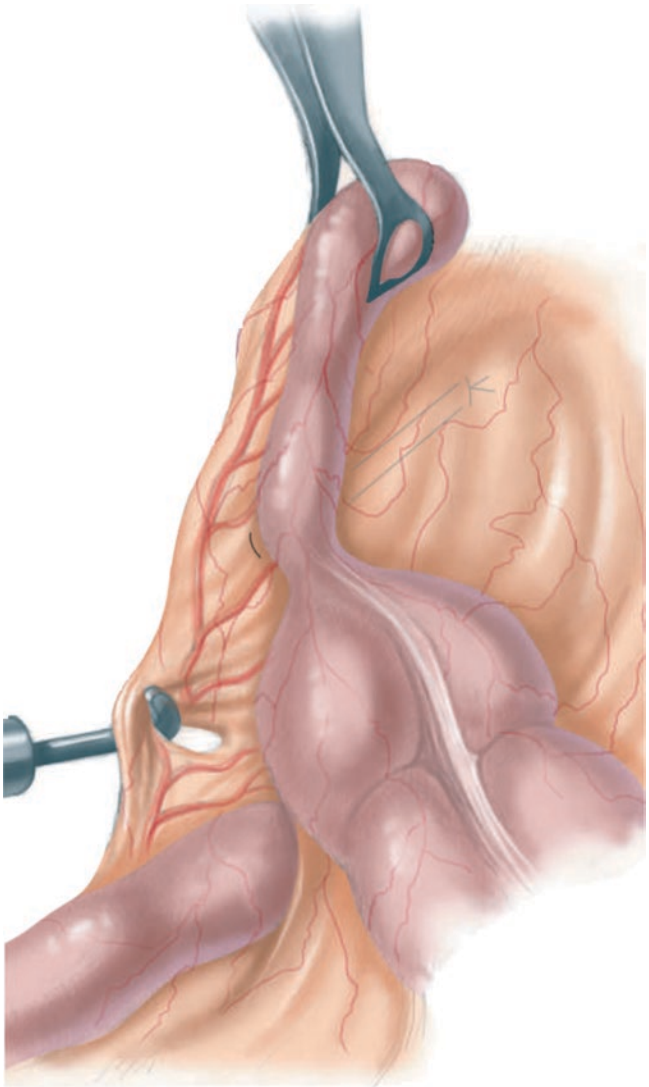

Fig. 33.6 Skeletonizing of the appendix

33.3 Outcomes

Advances in perioperative care, current surgical techniques, and antibiotics recommendations have resulted in a nearly zero mortality rate and low morbidity in children with appendicitis, whether simple or perforated. The long-term outcome of the vast majority of patients who undergo appendectomy in childhood is very good, with a late risk of adhesive intestinal obstruction less than 1%.

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Omphalomesenteric Duct Remnants

34

Kenneth K. Y. Wong and Paul K. H. Tam

In the human embryo, the omphalomesenteric duct exists as a long, narrow tube that joins the yolk sac to the digestive tube. During the seventh week of gestation, the duct usually undergoes complete obliteration. Incomplete obliteration results in omphalomesenteric remnants, which may be completely asymptomatic throughout life. Some types of omphalomesenteric remnants can be apparent in the newborn infant, and others may cause various problems. For example, some remnants have the potential to develop into adenocarcinoma owing to scattered glandular epithelial nests.

34.1 Embryology

At about the 20th day in embryo, the precursors of the enteric nervous system migrate and colonize to form the original gut, involving the foregut (oesophagus, stomach, and duodenum), the midgut (small intestine, caecum, ascending colon, and proximal transverse colon), and the hindgut (distal transverse colon, descending colon, sigmoid, and rectum) (Fig. 34.1).

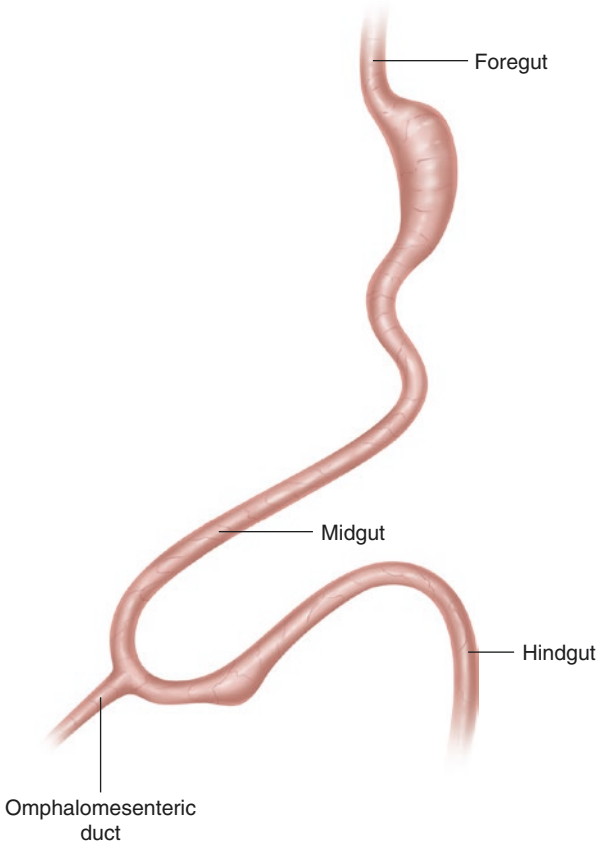

Fig. 34.1 Schematic diagram showing the omphalomesenteric duct located between the midgut and umbilicus during embryogenesis

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Development of the gut in the embryo is characterized by the elongation starting at the end of the fourth week of gestation. At the apex of the tube, the intestinal loop remains connected and open to the yolk sac via the omphalomesenteric (vitelline) duct. At the end of the fifth week of gestation, the midgut starts to be isolated from the yolk sac through the obliteration and resorption of the omphalomesenteric duct. This process should be completed by the end of the seventh week of gestation (Fig. 34.2b, c).

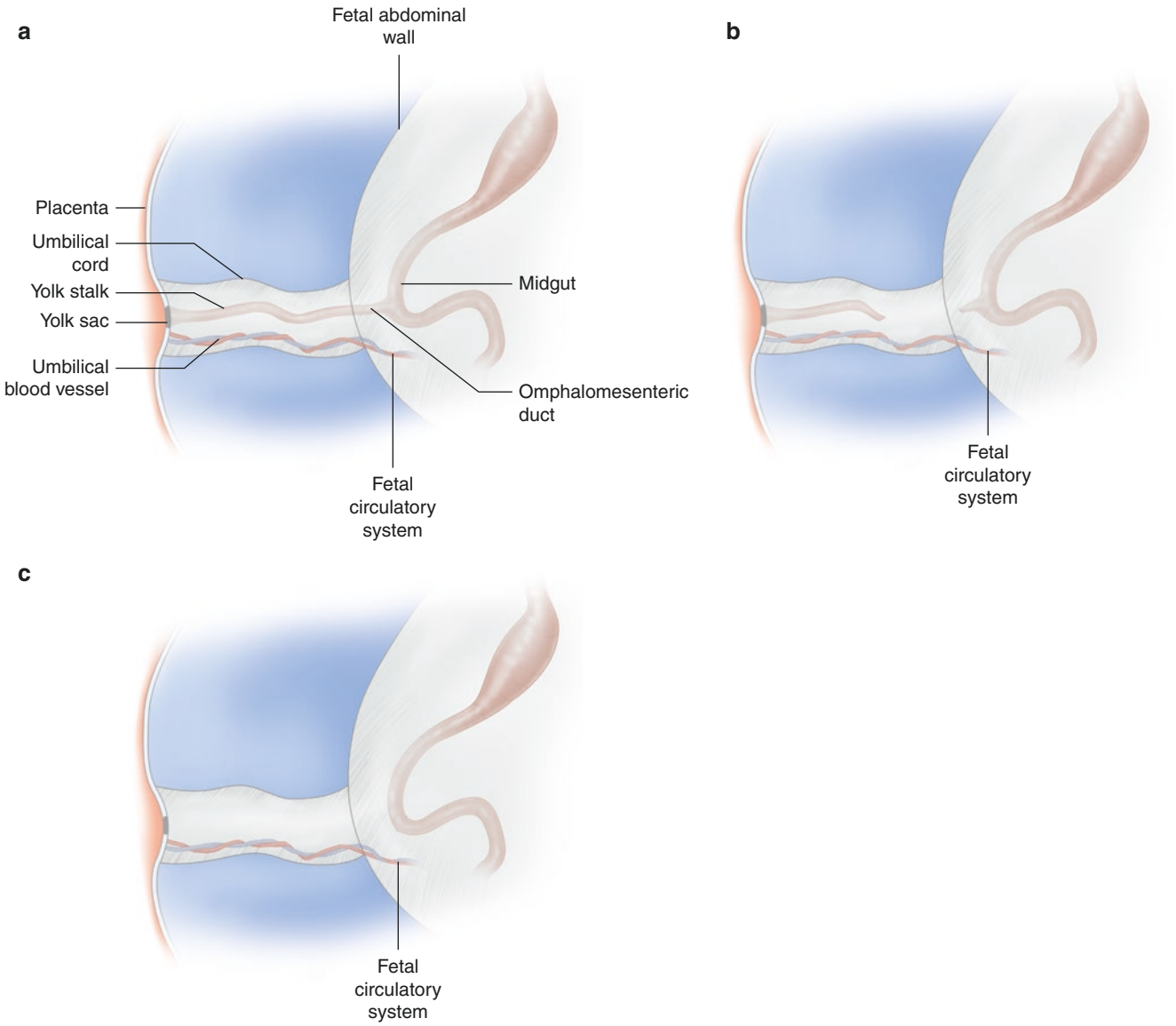

Fig. 34.2 Schematic diagrams showing the normal development of omphalomesenteric duct and the yolk stalk. (**a**) Connection of open intestinal loop to the yolk sac via the omphalomesenteric (vitelline) duct during early gestation. (**b**) Gradual obliteration and resorption of omphalomesenteric duct starting from the end of the fifth week of gestation. (**c**) Complete resorption of the omphalomesenteric duct by the end of seventh week of gestation

34.2 Pathology and Clinical Presentation

Because of incomplete obliteration and resorption, omphalomesenteric duct may persist after birth in some people, giving rise to omphalomesenteric remnants with various anatomic anomalies. These can be classified into four types: Meckel's diverticulum, umbilical polyp, omphalomesenteric duct cyst, and persistent omphalomesenteric duct (Fig. 34.3). Urachal remnant and umbilical granuloma, although not of the same embryologic aetiology as omphalomesenteric remnants, must be considered as differential diagnoses.

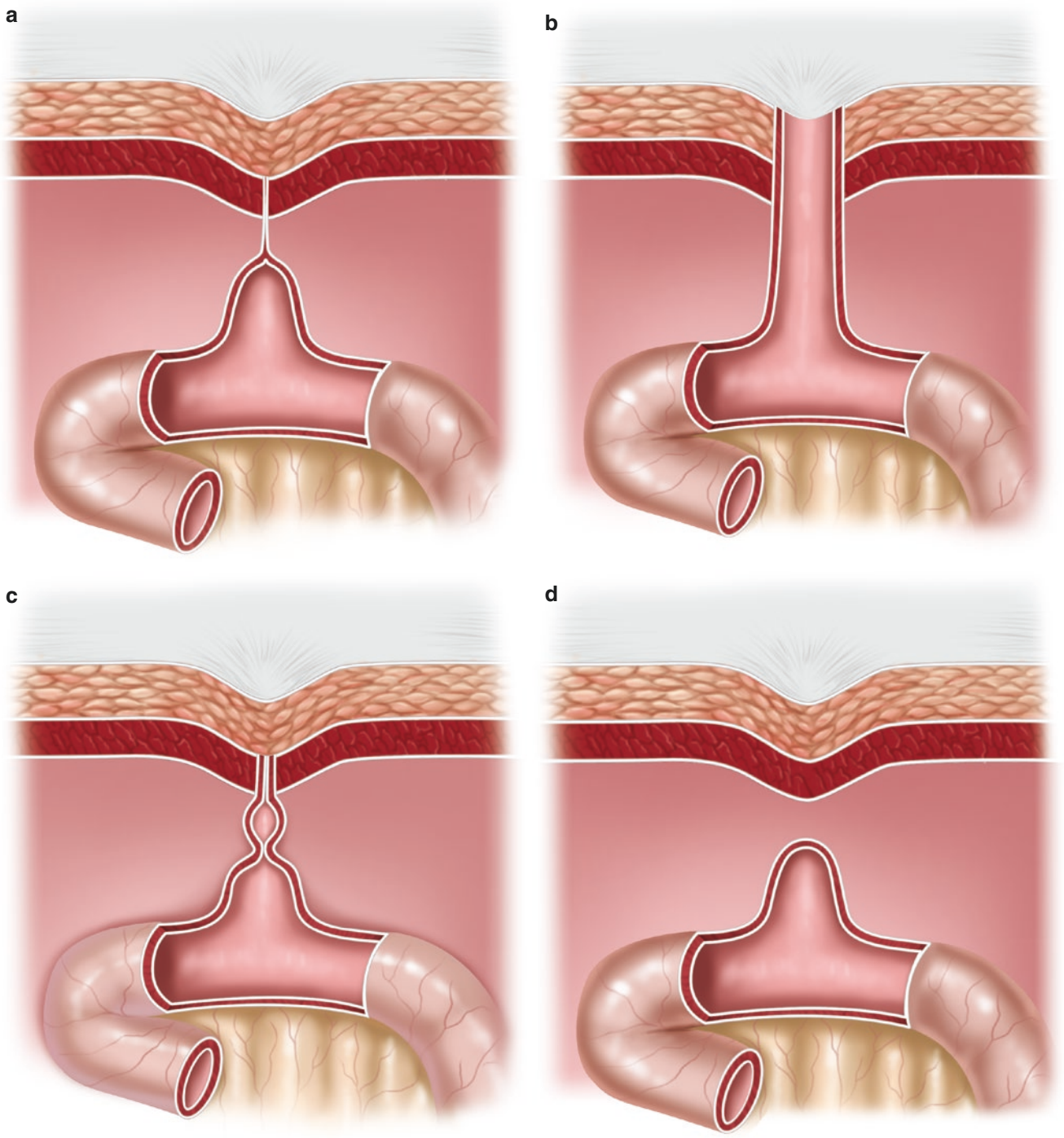

Fig. 34.3 Schematic diagrams showing the different anatomical variations of omphalomesenteric remnants. (**a**) Obliterated omphalomesenteric duct. (**b**) Patent omphalomesenteric duct. (**c**) Omphalomesenteric duct cyst. (**d**) Meckel's diverticulum

34.2.1 Meckel's Diverticulum

The most common omphalomesenteric remnant seen is the Meckel's diverticulum, which is the persistence of the enteral end on the antimesenteric side of the small intestine. It was first described in 1809 by the German anatomist Johann Friedrich Meckel (1781–1833). This anomaly occurs in about 2% of the population. It is typically located within 2 ft of the ileocoecal valve. Meckel's diverticulum is usually lined by typical ileal mucosa, as in the adjacent small bowel. Ectopic gastric mucosa can be seen, and duodenal, colonic, pancreatic, and hepatobiliary tissues have also been reported. Complications of the diverticulum include bleeding (due to acid secretion from gastric mucosa), diverticulitis, and intussusception. The risk of complications decreases with increasing age, with no predictive factors for the development of complications. Asymptomatic diverticula are usually found incidentally at laparotomies for other conditions. A recent study showed that resection of incidentally detected Meckel's diverticulum had a significantly higher postoperative complication rate than leaving it in situ, and that 758 resections would need to be performed to prevent one death from the condition. As a result, there is no evidence to support the resection of incidentally detected Meckel's diverticulum.

For diagnosis, the presence of ectopic gastric mucosa with the secretion of gastrin can be detected using Tc-99 radioisotope scanning, although a negative result does not exclude the presence of Meckel's diverticulum. The sensitivity of Tc-99 is about 80–85%. In these cases, diagnostic laparoscopy can be a very useful tool.

34.2.2 Umbilical Mucosal Polyp/Umbilical Cyst

An umbilical polyp is a remnant of gastric or intestinal mucosa at the umbilicus. The bright red, polypoid tissue produces a persistent discharge, which may be blood-stained. It is often confused with an umbilical granuloma, which should respond to simple cauterization treatment. The diagnosis of an umbilical polyp may be confirmed by biopsy to look for the presence of intestinal or gastric mucosa, whereas specific multinucleate giant cells can be noted in umbilical granuloma tissue. As umbilical polyp may be associated with Meckel's diverticulum because both are remnants of the omphalomesenteric duct, the presence of the visible polyp may serve as a warning for otherwise obscure intra-abdominal symptoms.

34.2.3 Omphalomesenteric Duct Cyst

The obliterated omphalomesenteric duct may contain one or more cysts. The clinical presentation of this condition is similar to obliterated duct, with a risk of small bowel volvulus. The cysts are usually lined by a columnar, mucin-secreting epithelium. Thus, the cyst can sometimes become infected and the child will present with pain and fever. The infant who has this anomaly usually presents a cystic mass with increasing size in the middle or inferior belly. Occasionally, the intestinal duct dislocates owing to cystic mass compression, and the infant presents obstructive symptoms correlated with adhesive bowel volvulus. Diagnostic ultrasonography usually confirms the potential cystic mass before surgery.

34.2.4 Persistent Omphalomesenteric Duct (Patent or Obliterated)

The omphalomesenteric duct may be fully patent and present as an omphalo-ileal fistula. Nonetheless, this is a rare finding that usually presents in the neonatal period with gastrointestinal drainage. The fistula may contain ectopic gastric, colonic, or pancreatic tissue. Further, an umbilical 'polyp' consisting of intestinal mucosa may also be present, caused mainly by inflammatory tissue proliferation due to persistent chemical stimulation of bowel content. In the newborn, the persistent discharge can often result in periumbilical excoriation. Rarely, the ileum may even prolapse through the omphalo-ileal fistula, giving rise to the so-called steer-horn abnormality. The diagnosis is confirmed clinically by passing a catheter through the fistula into the small intestine and aspirating small bowel content, or by injecting radiographic contrast medium into the fistula.

Incomplete obliteration of the omphalomesenteric duct can result in the presence of a fibrous connective cord attached to the umbilicus. Although mostly asymptomatic, this persistent obliterated omphalomesenteric duct can cause intestinal obstruction or even volvulus.

34.3 Management

Surgery remains the best management option for omphalomesenteric remnants. The approaches for the various anomalies are described below.

34.3.1 Meckel's Diverticulectomy

As mentioned previously, it is not essential to remove the Meckel's diverticulum in otherwise asymptomatic patients, as the risks outweighs the benefits. Meckel's diverticulum should be excised in symptomatic children, however. The procedure is now mostly laparoscopically assisted or is even performed totally laparoscopically in many centers. Laparoscopy is also useful in helping to diagnose symptomatic patients with negative isotope scans. The principles of laparoscopic resection remain the same as for open surgery. Although some would argue for the resection of Meckel's diverticulum alone, it is the authors' preference to resect a short segment of ileum together with the diverticulum to ensure that all abnormal mucosa is removed.

In centres where laparoscopy is not feasible, open surgery is adopted. Here, the access to the peritoneal cavity is via a 2-cm periumbilical incision (Fig. 34.4). With this approach, an excellent postoperative cosmetic result is ensured. After gaining access into the peritoneum, the small bowel is brought out and run through. The Meckel's diverticulum is situated about 2 ft from the ileocaecal valve, on the antimesenteric border of the distal ileum, and may be bound to the adjacent small bowel mesentery by a covering of peritoneum. At the operation, these adhesions are divided to mobilize the diverticulum. The blood vessels on the mesenteric side are ligated and divided. Ileal resection with primary end-to-end anastomosis with single-layer, interrupted absorbable 4-0 sutures is carried out, ensuring the removal of all ectopic tissue. The fascia of the subumbilical incision is closed using 3-0 absorbable sutures, and the skin is approximated with subcuticular 5-0 suture reinforced with adhesive strips.

34.3.2 Excision of the Omphalomesenteric Duct

For a patent omphalomesenteric duct, the umbilical skin surrounding the fistula should be preserved for better cosmesis. Thus, a circumferential incision is made around the orifice of the fistula. A separate skin-crease incision is made below the umbilicus. The superior skin flap, which includes the umbilicus, is elevated and the fistula is brought out through the subumbilical incision. The abdominal wall fascia is incised transversely on either side of the fistula. The umbilical vessels and the urachus are individually ligated and divided as the peritoneal cavity is entered. The fistula is then traced to its origin at the distal ileum.

The blood supply to the fistula should be ligated and divided near its origin on the mesentery. Similar to Meckel's diverticulum, the fistula should be excised with a small margin of ileum, which is then repaired with a single, interrupted layer of absorbable 4-0 sutures. The linea alba is then repaired and the subumbilical incision is closed with subcuticular 5-0 suture. The circular defect in the center of the umbilicus may be left to heal by secondary intention if it is small, or it may be loosely closed with a purse-string suture. The healed wound should resemble the umbilicus.

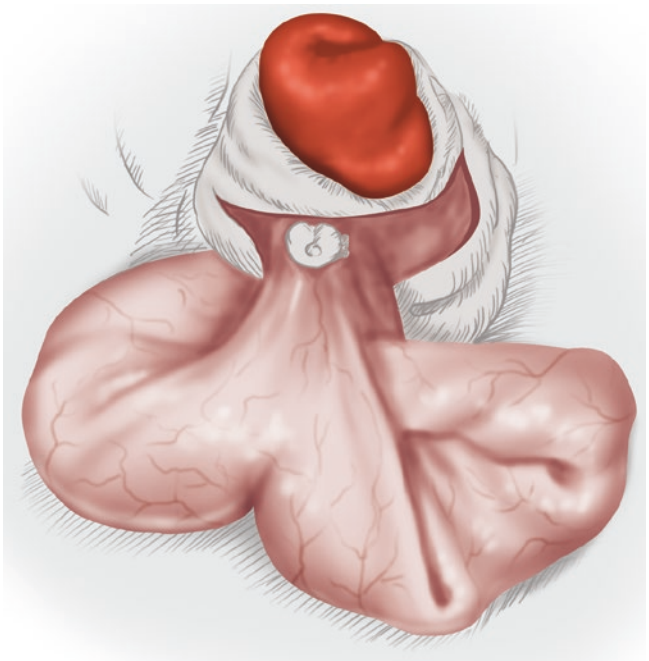

Fig. 34.4 Dissection of a patent omphalomesenteric duct

34.3.3 Excision of an Umbilical Polyp

For the surgical management of umbilical polyp, limited exploration of the peritoneal cavity is advisable, because of the possibility of an underlying connection to the ileum by a remnant of the omphalomesenteric duct. The approach is via a circumferential incision around the polyp, trying to preserve as much of the normal umbilicus as possible. The skin defect is repaired using an absorbable purse-string suture. A subumbilical incision is made as described above. The abdominal wall is opened transversely and the peritoneal cavity is entered. If an omphalomesenteric duct remnant is present, it is resected.

34.4 Complications

For conditions that involve peritoneal access (either open or laparoscopic) and intestinal resection, early postoperative complications include anastomotic leak, adhesions formation, postoperative ileus, and wound infection. These complications are rare (<5%), however. Intestinal obstruction from adhesions may present as a late event.

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Ulcerative Colitis

35

Risto Rintala

Approximately 15–20% of all cases of ulcerative colitis have their onset during childhood. In the West, the incidence of paediatric ulcerative colitis ranges between 1 and 4 per 100,000 per year. The onset of symptoms typically occurs in prepuberty or puberty. A trend towards earlier appearance of symptoms has been noted recently, with some patients already developing symptoms in their early school years.

The aetiology of ulcerative colitis is still unclear, so there is no curative treatment. The medical therapy of ulcerative colitis is based on systemic or local suppression of the immune response of the large bowel. Oral 5-aminosalicylic acid (5-ASA) regimens are usually used as first-line therapy for mild to moderately active disease. In more severe disease, corticosteroids are usually used as induction therapy that is continued with aminosalicylic acid medication. Steroid dependency should not be tolerated, so other options such as immunomodulatory maintenance medication with thiopurines can be used in patients who are not responding to 5-ASA therapy. Biologic medications (infliximab, adalimumab) can be used in induction and maintenance treatment in steroid-dependant disease that cannot be controlled by thiopurines.

Ulcerative colitis is more aggressive in children than in adults. Children present more often with widespread disease and develop pancolitis more often than adults. Therefore, children require more aggressive medical treatment than adults and usually need corticosteroids to control the initial disease. Systemic corticosteroids are a major concern; the adverse effects of high-dose corticosteroid treatment on a growing and developing body are significant and often are an indication for surgical treatment.

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35.1 Surgical Treatment

Between 25% and 40% of children with ulcerative colitis undergo surgical treatment. As most patients today can be stabilised by medical treatment, emergency operations for toxic megacolon, unremitting bleeding, or refractory fulminant colitis are not common. The typical indications for surgery of ulcerative colitis are poor response to optimal medical treatment, dependence on high-dose corticosteroids with significant side effects, delay in growth and maturation, and severe extraintestinal manifestations of the disease. Surgery should not be considered as a primary or early treatment of ulcerative colitis. A significant proportion of patients achieve long-term symptom relief with conservative treatment and may remain in remission with minimal or no medication. Moreover, the functional outcome following restorative proctocolectomy is not comparable to normal bowel function. When patients go through several exacerbation phases of the disease, they gradually learn to accept that their bowel will function from a few times to several times a day. Before proctocolectomy is undertaken, Crohn's disease should be ruled out with every possible measure, as Crohn's disease patients should not undergo restorative proctocolectomy.

35.1.1 Choice of Procedure

The gold standard of surgery for ulcerative colitis has been proctocolectomy and permanent ileostomy. Limited colonic resections, as well as colectomy and ileorectal anastomosis, have been abandoned, as these procedures have been associated with a high incidence of complications and recurrence of the disease. Proctocolectomy and permanent ileostomy gives excellent control of ulcerative colitis and related symptoms, but it is not very well tolerated by children and adolescents because of the significant social restrictions and permanently altered body image that are related to this operation. Since late 1970s, restorative proctocolectomy with

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ileoanal anastomosis has gained overall acceptance as the standard operative procedure for both adult and paediatric ulcerative colitis. Many paediatric surgeons advocate the use of an ileal reservoir; the most popular and easiest to construct is the J-pouch. Some paediatric surgeons still use a straight ileoanal anastomosis without a reservoir.

A two-stage operation (colectomy and pouch formation with ileostomy in the first stage and stoma closure in the second stage) is the most common elective approach for ulcerative colitis. A three-stage operation (first colectomy, then pouch formation, and finally stoma closure) should be considered in patients with high-dose steroid use or severe malnutrition, and when Crohn's disease has not been completely excluded. Restorative proctocolectomy without ileostomy may be considered in patients without any risk factors (steroids, anastomotic tension).

The laparoscopic approach can be used safely in children. The rate of complications may be less than for open surgery, and the cosmetic results are definitely superior.

35.1.2 Preoperative Preparation

Restorative proctocolectomy is a major operation with a significant incidence of postoperative complications. Septic complications are common, as most patients with refractory ulcerative colitis are immunosuppressed because of high-dose corticosteroid treatment. The nutritional status of many patients is often not very good because of long-term diarrhoea and poor nutrient intake. To avoid septic complications, it is imperative that systemic corticosteroids are tapered to the lowest possible level, or preferably changed to locally acting budesonide, which has fewer systemic immunosuppressive effects. The patient's nutritional status should also be improved. It is usually possible to do this by dietary measures. Parenteral nutrition is only rarely required to restore proper nutritional status.

There is no need for preoperative bowel preparation. The site of the covering loop ileostomy should be marked with water-resistant marker pen before the operation. A stoma site in the right lower abdominal quadrant is best determined when the patient is sitting.

Prophylactic antibiotic treatment (cefotaxime and metronidazole) is started at the induction of the anaesthesia. The operation is undertaken under general anaesthesia; the use of nitrous oxide as an anaesthetic is best avoided, as it may distend the bowel. Insertion of an epidural catheter for local anaesthetic infusion to control postoperative pain is advisable. Additional pain control can be achieved by administrating opioids by a patient-controlled analgesia (PCA) system. A bladder catheter is inserted and left in place until pain control with opioids and epidural catheter can be discontinued.

35.1.3 Operative Technique: Open Approach

The patient is positioned in a lithotomy position with a 10–15° Trendelenburg tilt. The abdomen is prepped from lower chest to perineum. A midline incision starting from the midpoint between the xiphoid process and umbilicus and extending through or around the umbilicus down to the suprapubic region is used to get free access to the whole length of the colon (Fig. 35.1). Usually there is no need to use self-retaining wound retractors, which may cause wound edge ischaemia and increase postoperative wound pain. The whole length of the bowel is inspected to rule out Crohn's disease.

Before colectomy is started, it is important for the surgeon to assess whether the terminal ileum will reach down to the perineum. If the rotation of the bowel is normal and the terminal ileum reaches the pubic bone, it is very likely that an ileoanal anastomosis can be performed without undue tension. After mobilisation of the ileocaecal region, the ileum is transected by a GIA stapler flush to the ileocaecal junction (Fig. 35.2).

The peritoneal reflections of the ascending colon and hepatic flexure are mobilised. The splenocolic ligament is severed and splenic flexure mobilised. The greater omentum can be preserved if it does not tear much during its dissection

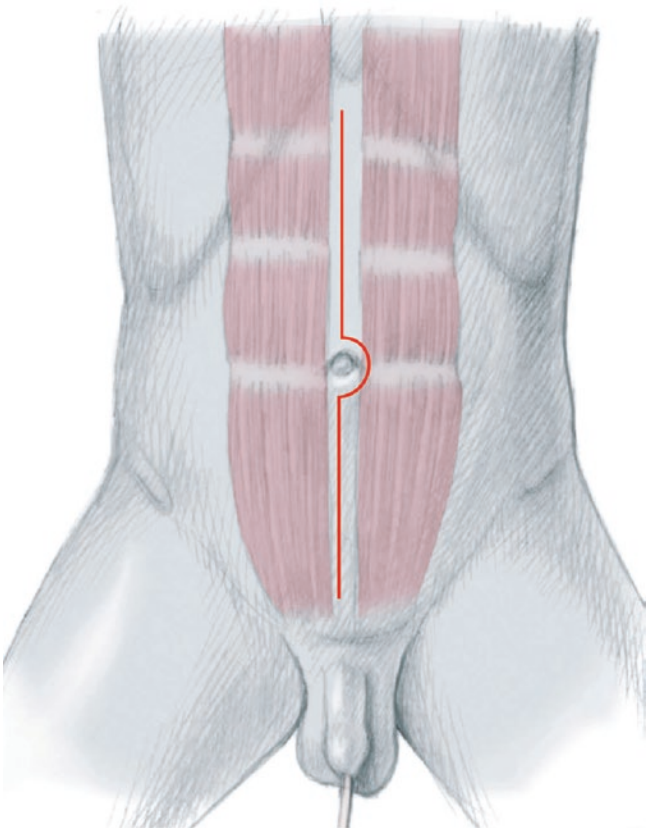

Fig. 35.1 Incision for open approach to restorative proctocolectomy

off from the transverse colon. The dissection of the omentum is best performed by bipolar cautery or scissors; the attachments between the transverse colon and omentum are transected flush to the colonic wall. The lateral peritoneal reflections of the descending and sigmoid are transected. The vessels in the colonic mesenterium are ligated or cauterised near the bowel wall; usually only the main arteries to the colon, right, middle, and left colonic arteries need ligatures. The colon is transected at the junction of the sigmoid and rectum with a GIA stapler (Fig. 35.3). The whole colon can be now removed from the operative field.

Stay sutures or a right-angled large clamp on the proximal rectal stump facilitate the dissection of the rectum (Fig. 35.4). These allow the surgeon to pull and move the bowel on either side freely. The mesentery of the rectum in patients with severe ulcerative colitis is often inflamed and very thick. Dissection within the mesentery is time-consuming and bloody. The easiest way to proceed is to keep the plane of dissection right on the rectal wall. The small vessels entering the bowel are cauterised flush to the bowel wall. Broad and long-bladed retractors and cranial pulling from the rectal stump facilitate the dissection.

The dissection is continued down to the level of the pelvic floor. Rectal finger examination is useful to assess the adequacy of abdominal dissection. If the lowest level of abdominal mobilisation is within 3–4 cm from the anal verge, no problems are expected in the transanal mucosectomy and rectal pull-through (Fig. 35.5).

The next stage in the operation is the mobilisation of the ileum to reach to the anal canal (Fig. 35.6). The ileocolic artery is ligated and transacted. The mesentery of the ileum is mobilised up to the level of the proximal superior mesenteric artery. The root of the mesentery may have to be mobilised from the duodenum and lower rim of the pancreas. The mesenteric arteries to the distal two or three vascular arcades of the terminal ileum are ligated and transected proximally. To ensure a tension-free ileoanal anastomosis, the distal end of the ileum or the tip of the J-pouch should reach in front of the pelvic rim, to the base of the penis in males or the anterior vestibulum in females (Fig. 35.6).

The J-pouch should be between 7 and 10 cm in length. The terminal ileum is folded, and the antimesenteric tip of the future pouch is longitudinally opened with a cautery needle. The opening should be kept short (1.5–2 cm), as it widens significantly when pulled through to the anus. The GIA stapler blades are inserted into each arm of the pouch; the stapler is closed and fired (Fig. 35.7). One firing of a 75-mm stapler or two of a 50-mm stapler is usually sufficient

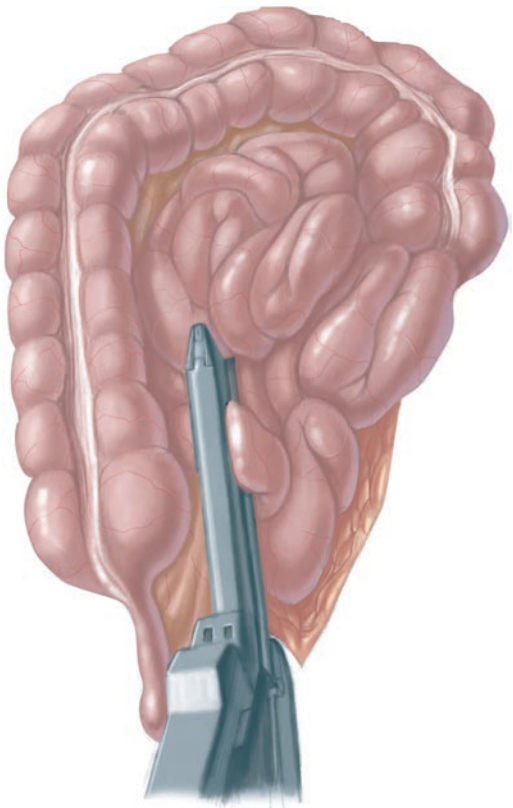

Fig. 35.2 Transection of the ileum

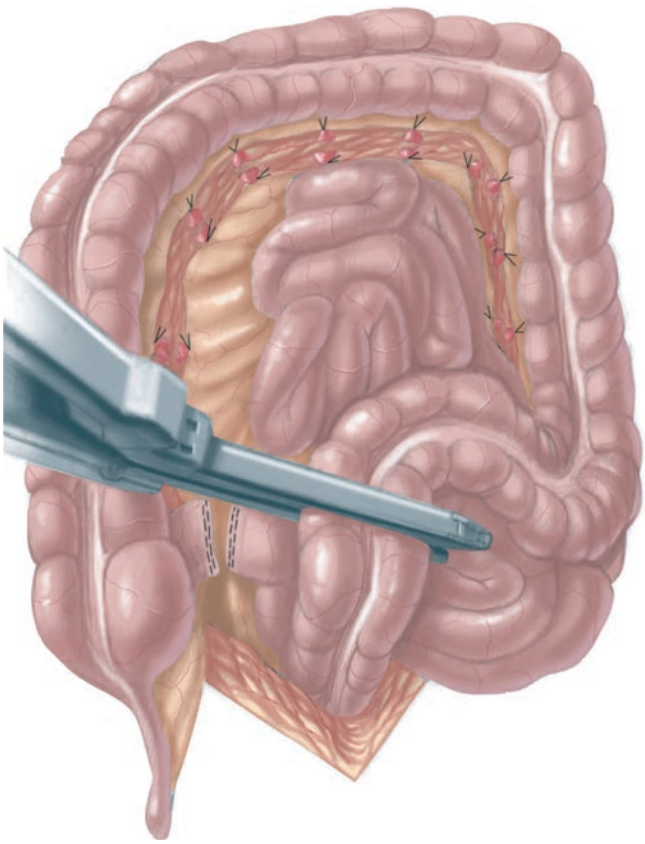

Fig. 35.3 Mobilization and transection of the colon

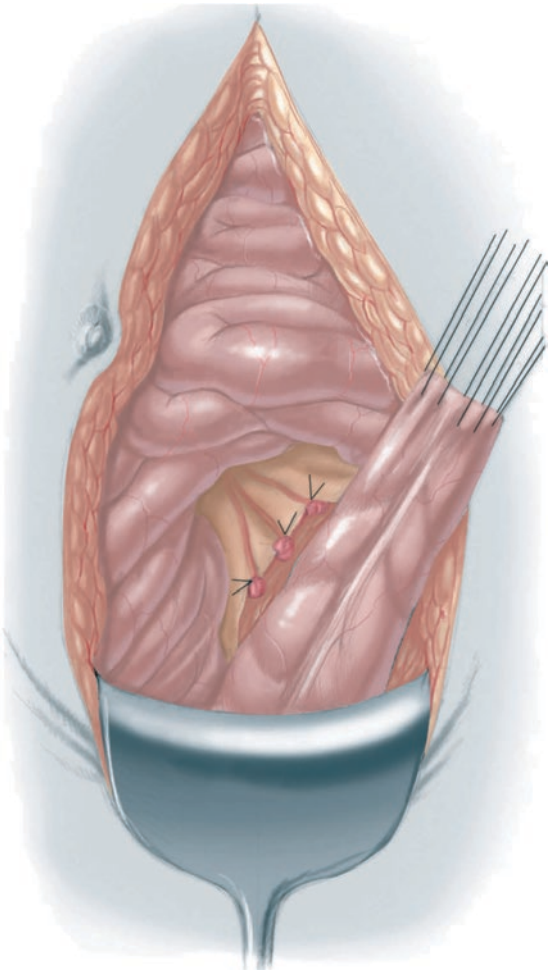

Fig. 35.4 Dissection of the rectum

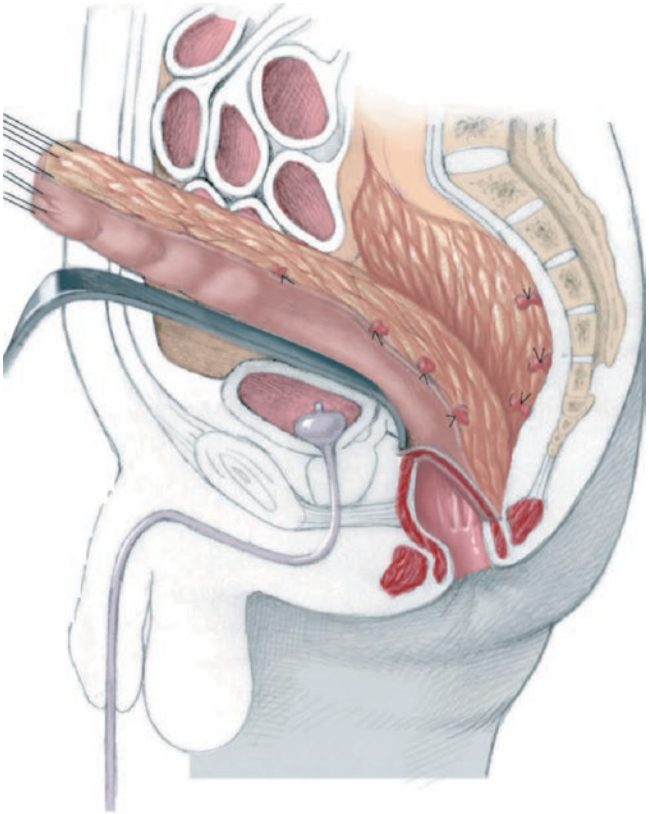

Fig. 35.5 Dista extension of abdominal dissection

to construct a pouch. The stapled suture line can be reinforced with 4-0 or 5-0 absorbable sutures. The pouch and terminal ileum are wrapped in warm and moist swap and returned the abdomen. The abdominal incision is loosely packed with warm and moist swaps.

The perineal phase of the operation starts by insertion of holding stiches between the mucocutaneous junction of the anal canal and a colostomy ring (Fig. 35.8). These keep the anus open and dilated, and give excellent access to the anal canal. Epinephrine in saline (1:100,000) is injected under the mucosa to lift it up and decrease bleeding during the initial phases of transanal mucosectomy.

The transanal mucosectomy is started above the dentate line. A rim (5–10 mm) of anal transitional epithelium should be left *in situ*, otherwise the sensibility of the anal canal is significantly decreased and the anal sampling reflex may be lost. Disease recurrence in the transitional anal epithelium is unlikely. The whole circumference of the anal canal mucosa is incised and the mucosectomy is started. The *dotted line* in Fig. 35.9 depicts the line of dissection between the mucosa and the rectal muscle. Some surgeons prefer to use multiple stay sutures in the mucosa just above the level of the mucosal incision to facilitate mucosectomy. The author uses small, triangular clamps to grasp the edge of the mucosal cuff (Fig. 35.10). The mucosectomy is performed by a combination of sharp and blunt dissection with scissors. In colitis ulcerosa, the mucosectomy is much more difficult than in noninflammatory conditions, and blood loss is often also significant. Preoperative treatment with locally acting cortisone foam or suppositories may decrease blood loss and make the dissection easier. The mucosectomy is continued for 5–8 cm until the level above the pelvic floor is reached.

The cuff, consisting of the muscular lining of the anal canal and distal rectum, can be transected transanally when the pelvic cavity is entered at the upper end of the mucosectomy. Pulling from the mucosal tube tents the proximal end of the muscular cuff inside the more distal cuff; thus the cuff can be safely transected without damaging the urethra and prostate. Another option is to evert the rectum through the anus and sever the muscular cuff outside the anus at the upper end of the mucosectomy. Bleeding from small vessels in the cuff can be controlled by cautery. The mobilised remaining rectum and the mucosal tube are then removed through the anus (Fig. 35.10).

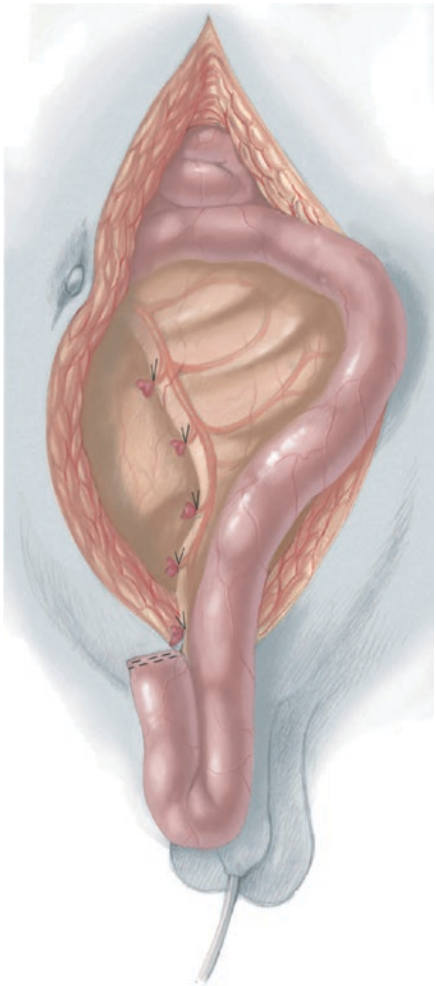

Fig. 35.6 Mobilisation of the ileum to reach to the anal canal

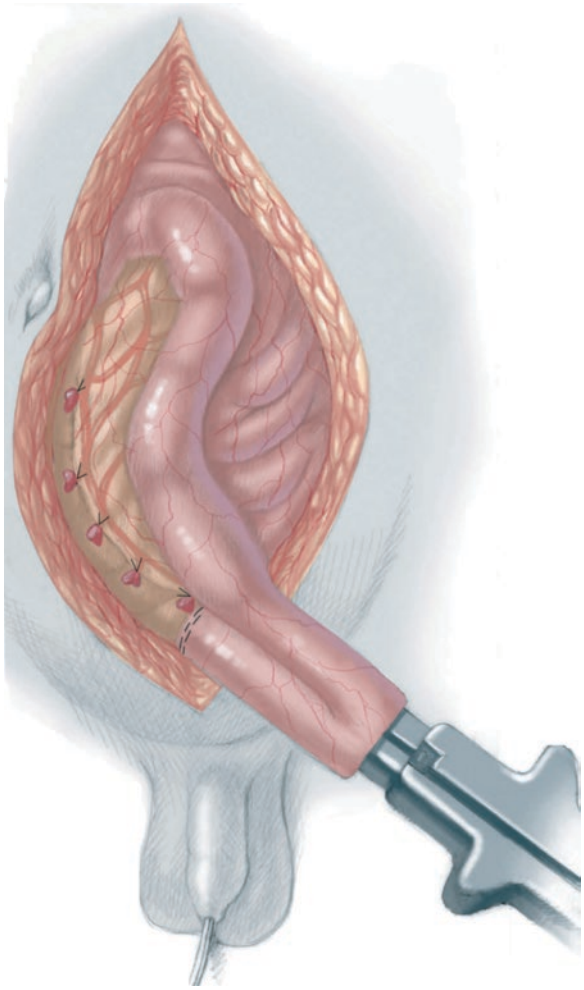

Fig. 35.7 Construction of the J-pouch

A long, soft clamp is inserted through the anal muscular cuff to the pelvis. The J-pouch (or distal ileum in case of straight pull-through) is grasped with the clamp and pulled through to the anus (Fig. 35.11). The assistant confirms through the laparotomy incision that the mesentery of the pulled-through pouch is not twisted. The mesentery of the pulled-through J-pouch (or distal ileum) is the tightest component of the pulled-through segment and requires the shortest route. Therefore it is natural that in the pelvis the mesentery is positioned anteriorly to the bowel; this does not mean that the pull-through segment would be kinked.

As there is often significant tension when the first anastomotic stitches are inserted, it is advisable to use four-quadrant holding stitches to initiate the suturing of the anastomosis. The anastomosis is performed in one layer, with interrupted sutures between the full-thickness ileum and anal canal (Fig. 35.12). A suitable material is absorbable 4-0 sutures. The tension in the suture line is usually relieved when the sutures between the anus and the stoma ring are cut, allowing the suture line to retract within the anal canal.

The space between the pulled-through bowel and the posterior peritoneum is closed with running 4-0 absorbable sutures. The pelvic cavity is inspected for bleeding sites. A round skin disk is cut off the stoma site. A cruciate incision is made to the abdominal muscle fascia at the site of the stoma. The opening in the fascia and abdominal muscles is bluntly enlarged to accommodate two fingers. An opening is made in the anterior peritoneum and a loop of ileum as near as possible to the ileoanal anastomosis is pulled out for stoma formation. The abdominal incision is closed in layers and the stoma is matured over a stoma rod (Fig. 35.13). Generally there is no need to drain the pelvic cavity.

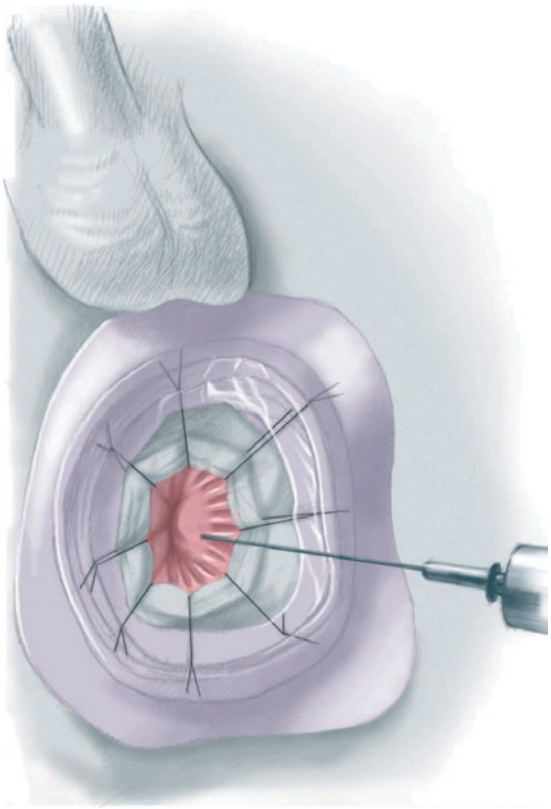

Fig. 35.8 Stitches between the mucocutaneous junction of the anal canal and a colostomy ring

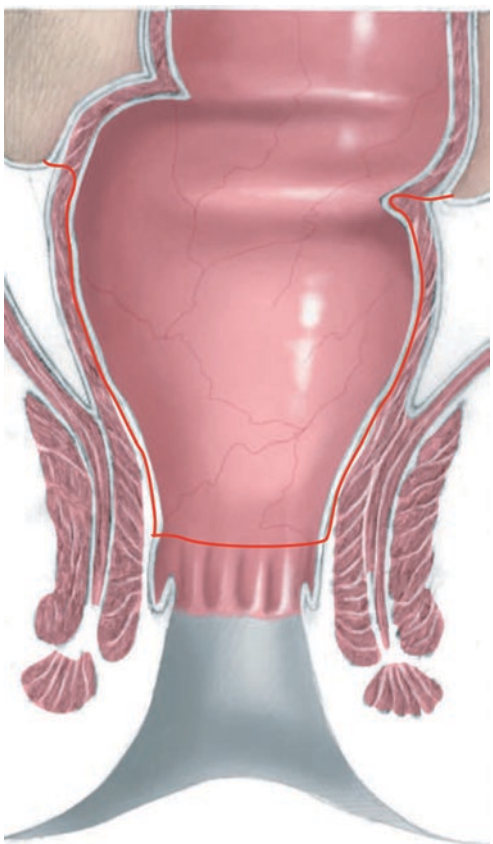

Fig. 35.9 Steps in the transanal mucosectomy

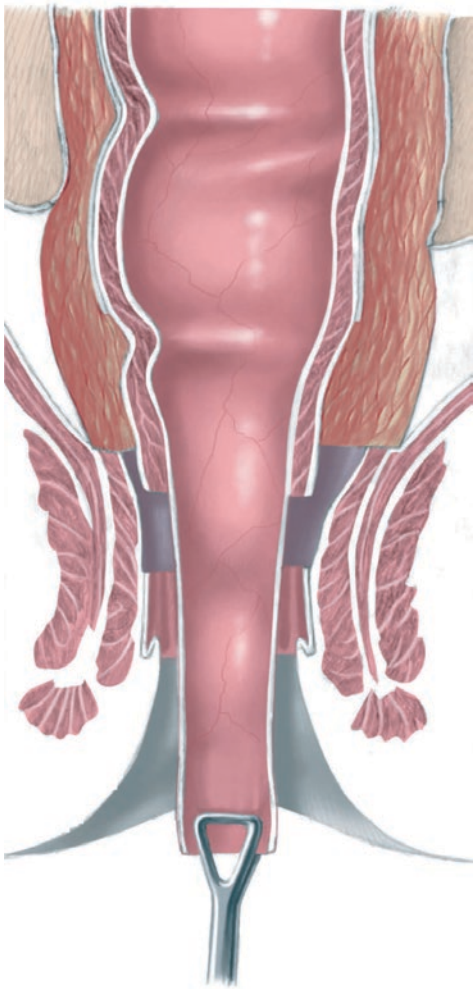

Fig. 35.10 Transanal removal of the remaining rectum and mucosal tube

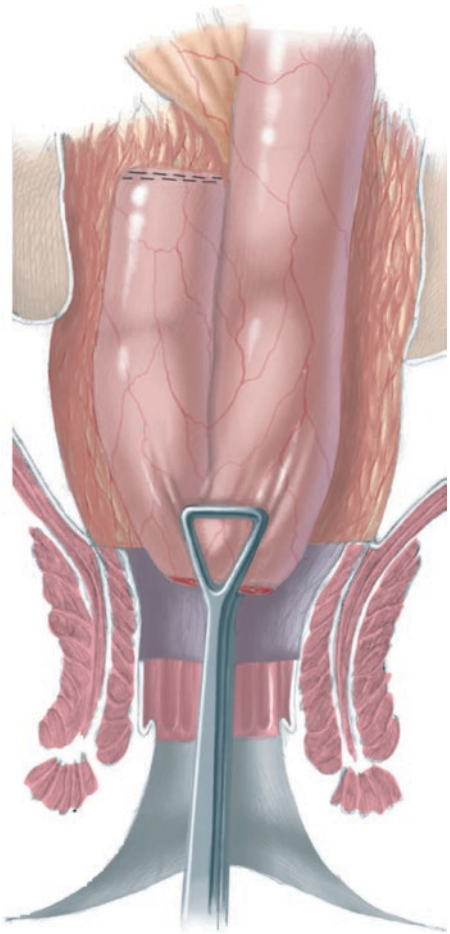

Fig. 35.11 Pulling the J-pouch through to the anus

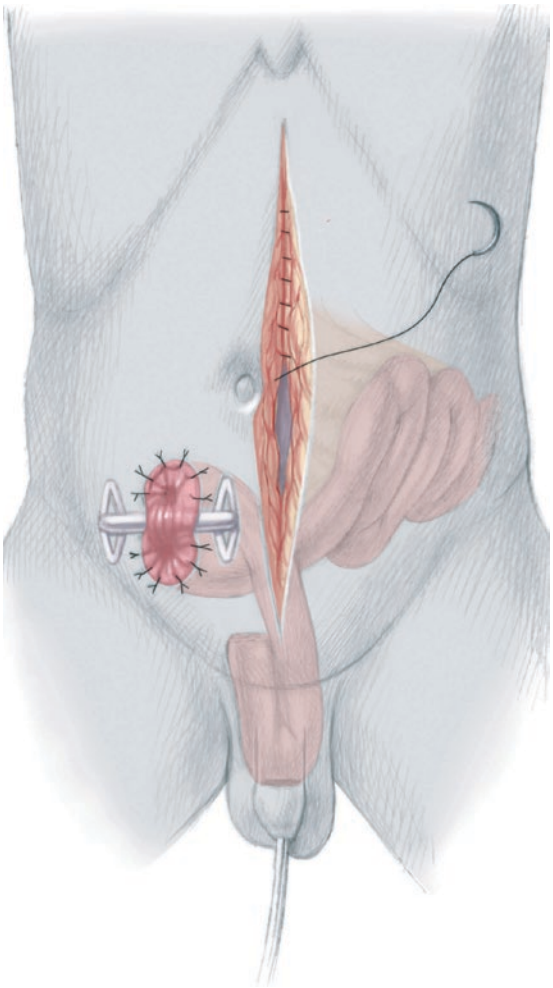

Fig. 35.13 Stoma formation

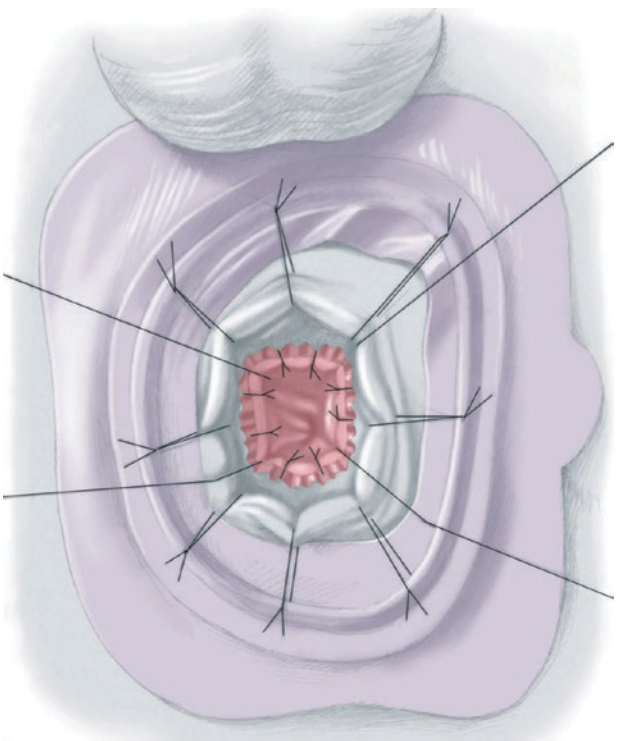

Fig. 35.12 Suturing of the anastomosis

35.1.4 Operative Technique: Laparoscopic Approach

The position of the patient and the prepping of the abdominal area in the laparoscopic operation are similar to those for the open approach. The first trocar in inserted through the umbilicus, either openly or after Veress needle insufflation. The other trocars are placed under direct vision lateral to rectus muscles to avoid injury to epigastric vessels (Fig. 35.14). Four or five trocars are used in all. One trocar is placed to the site selected for the ileostomy.

The small bowel is completely inspected through by using two Babcock clamps to rule out any evidence of Crohn's disease. The dissection of the colon can be initiated on either side; the author prefers to start on the left. The lateral peritoneal attachments are divided by Harmonic scalpel or vessel sealing device (LigaSure®) after identification of the left ureter. The mesentery is divided near the bowel wall by using either Harmonic scalpel or vessel sealing technology. The splenic flexure is mobilised by dividing the splenocolic ligament. The most difficult part of the operation is the mobilisation of the transverse colon (Fig. 35.15). The omentum may be preserved, but doing so is often difficult and not necessary.

After the mobilisation of the transverse colon, the hepatic flexure is taken down (Fig. 35.16) to enable dissection of the right lateral peritoneal attachments. Again, the right ureter must be visualized. After mobilisation and devascularisation of the right colon, the terminal ileum is transected with an endostapler device.

After completion of the colectomy, the patient is positioned into a steep head-down tilt to keep the pelvis free of the small bowel and mobilised large bowel. The rectum is dissected with the same instruments (Harmonic scalpel or vessel sealing device) and diathermy hook, staying at the bowel wall to minimize damage to pelvic nerves (Fig. 35.17). The dissection is continued deep to the pelvic floor in a manner similar to the open technique.

A short Pfannenstiel incision is performed and the rectum is transected with a stapler. The mobilized and devascularized colon and rectum are removed and a J-pouch is fashioned in the same manner as described for the open technique (Fig. 35.18). The perineal phase of the operation and the formation of the loop ileostomy are also similar to the open technique.

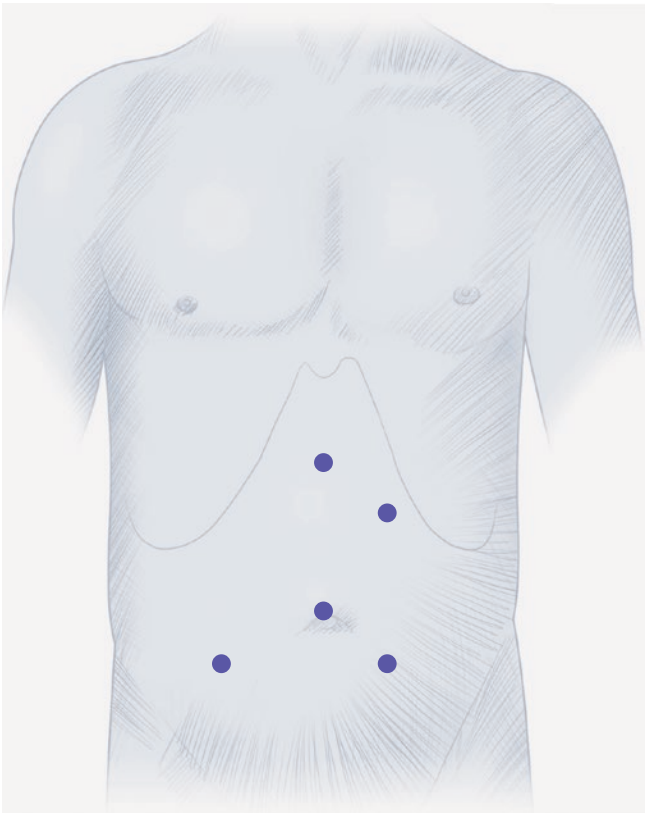

Fig. 35.14 Location of trocars for a laparoscopic approach

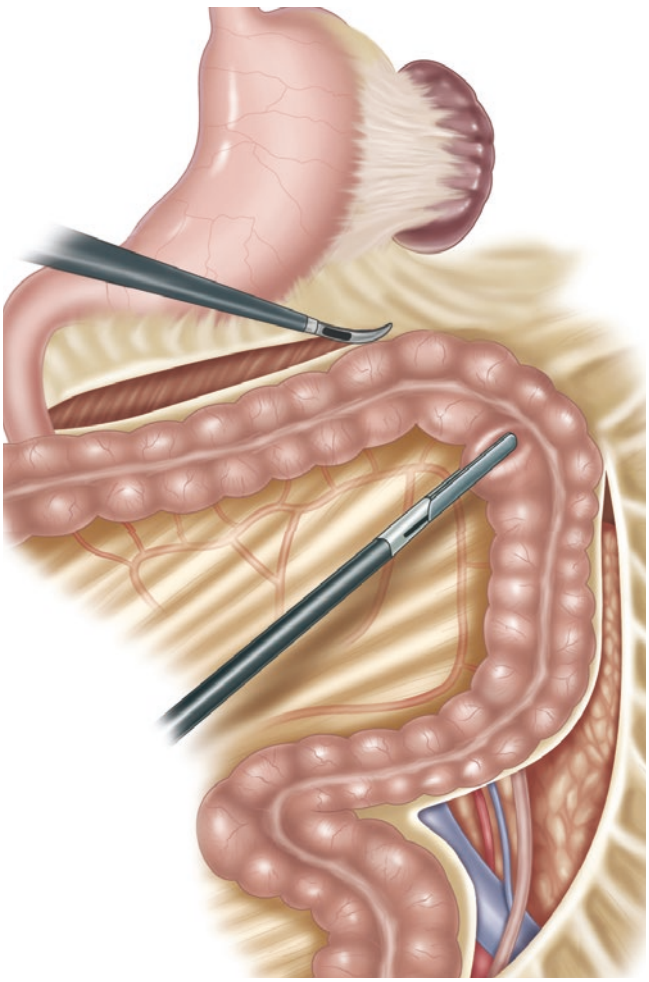

Fig. 35.15 Dissection of the transverse colon

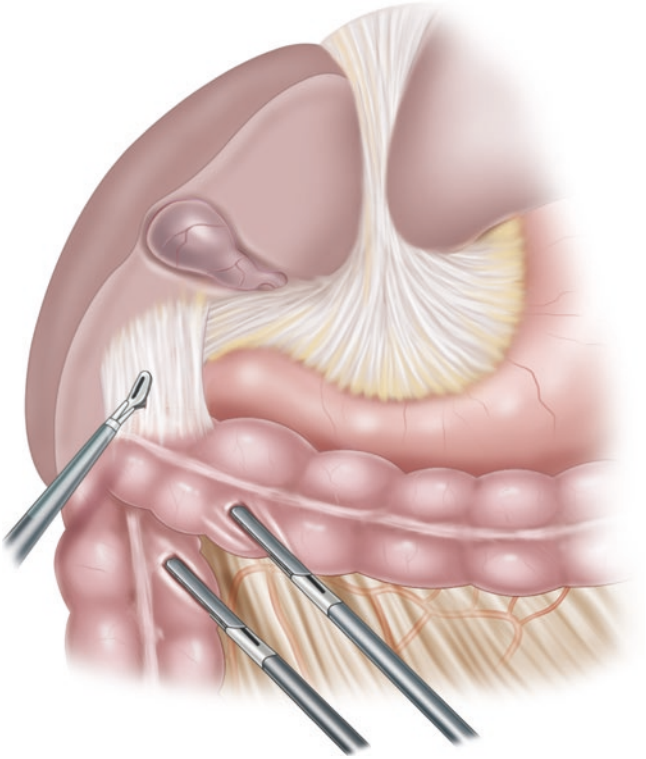

Fig. 35.16 Mobilization of the hepatic flexure

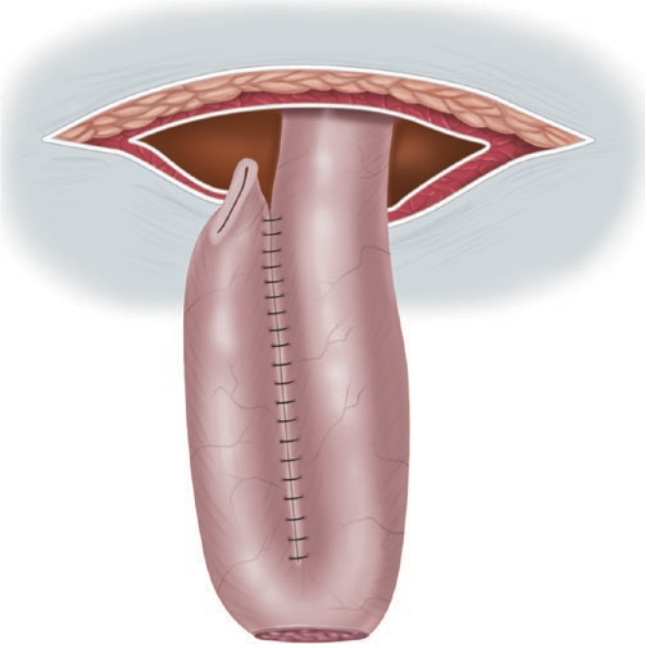

Fig. 35.18 The J-pouch

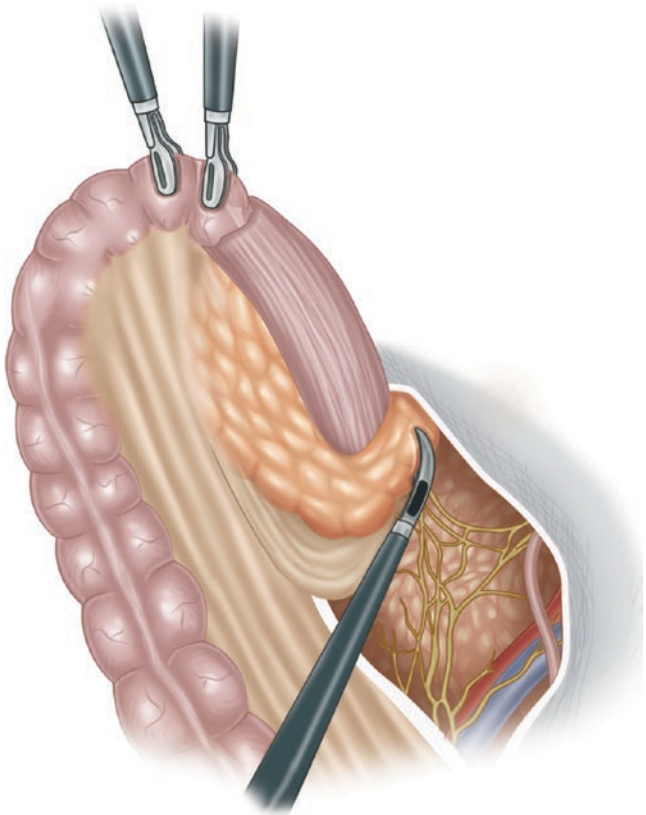

Fig. 35.17 Dissection of the rectum

35.2 Postoperative Care

Postoperative gastric decompression by nasogastric tube is usually not required. The bladder catheter can be removed when epidural anaesthesia is discontinued. Antibiotic prophylaxis is continued for 72 h postoperatively. If the patient was taking high-dose corticosteroids preoperatively, postoperative parenteral corticosteroid therapy is required until oral intake of medication is possible. Postoperative corticosteroids can be discontinued when sufficient function of the patient's own adrenal glands is confirmed by laboratory tests.

Oral intake of liquids is usually possible on the first or second postoperative day. Enteral feeding is encouraged and most patients tolerate full enteral feeding within the first 5 postoperative days. Stoma output is often excessive when the bowel resumes motility, and stoma losses should be replaced according to output and the electrolyte content of the stoma fluid. In most cases, Ringer's lactate is sufficient for replacement. Oral sodium supplementation to decrease stoma output is started as soon as the patient tolerates intake of salt tablets.

Postoperative dietary management consists of a lactose-free, low-residue diet. The amount of sodium supplementation can be monitored by spot urinary sodium measurements. The urinary sodium concentration should be kept higher than 20 mmol/L. Insufficient salt intake leads to increased and watery stoma output.

The protecting stoma can be taken down when healing of the pouch and ileoanal anastomosis is confirmed. A distal loopogram 3–6 weeks after the operation is used to assess the integrity of the ileoanal anastomosis and J-pouch. The immediate postoperative phase is characterised by loose and frequent bowel movements, occurring up to 10–12 times per 24 h. Antipropulsive medication (loperamide) is useful in slowing down gut motility. The bowel frequency gradually decreases to two to seven bowel movements per 24 h within 3–6 months. A low-residue diet and salt supplementation are helpful during the adaptation phase.

35.3 Outcome and Complications

Ileoanal anastomosis has revolutionised the treatment of ulcerative colitis in children as well as in adults. Long-term patient satisfaction following the operation is excellent despite a high incidence of postoperative complications. In children with a pouch ileoanal anastomosis, the functional outcome in terms of faecal continence is good. Most patients resume full daytime faecal continence within 6 months after closure of the stoma. A few patients suffer from slight nighttime staining that may require protective pads. In the absence of major postoperative complications, gross faecal soiling is practically nonexistent. The frequency of bowel movements 6–12 months postoperatively is between 4 and 10 per 24 h.

Early and late complications occur in 20–60% of patients. The most common are bowel obstructions or wound infections in patients who have been using high-dose corticosteroids prior to the operation. Pelvic septic complications or separation of the ileoanal anastomosis occur in less than 10% of the cases. Acute or chronic inflammation in the pouch (pouchitis) is an innate problem related to ileoanal pull-through for ulcerative colitis. Its incidence varies between 50% and 70% of the patients. Most acute bouts of pouchitis respond rapidly to a short course of oral antibiotics such as metronidazole. Chronic pouchitis is much less common, occurring in less than 10% of the patients. The treatment of chronic pouchitis consists of courses of low-dose antibiotics, with oral corticosteroids (preferably budesonide) used in recalcitrant cases. Probiotics may be helpful in maintaining remission. Chronic pouchitis may be a presentation of Crohn's disease; eventually 5–15% of patients who have undergone ileoanal anastomosis for ulcerative colitis are discovered to have Crohn's disease. Another symptom that should raise suspicion of Crohn's disease is pouch fistulisation, especially recurrent fistulas.

Despite the many potential postoperative problems, a great majority of patients who have had restorative proctocolectomy for ulcerative colitis resume a highly satisfactory lifestyle with complete faecal continence and acceptable frequency of daily bowel movements.

Suggested Reading

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Crohn's Disease

36

Risto Rintala

The incidence of Crohn's disease in children has clearly been increasing since the 1970s. The age at onset of the disease is similar to the age for ulcerative colitis. The diagnostic delay in Crohn's disease—usually more than a year from the onset of symptoms—is usually significantly longer, however. The symptoms are usually relatively vague in the early stages of the disease. The most common symptom is nonspecific abdominal pain. General symptoms are more common in patients with Crohn's disease. Growth failure and delay of sexual maturation are significantly more common in patients with Crohn's disease than in those with ulcerative colitis. The presentation of Crohn's disease in children is usually less localised than in adults. Crohn's disease in younger children typically affects the colon, and pancolitis is not uncommon. Moreover, the disease is significantly more aggressive in children than in adults.

As in ulcerative colitis, the aetiology of Crohn's disease is unclear. It is likely that a genetic predisposition exists for Crohn's disease, as a family history of the disease is more common in Crohn's patients than in those with ulcerative colitis.

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36.1 Treatment

There is no definitive curative treatment for Crohn's disease, which is an affliction of the whole gastrointestinal tract. Surgery does not offer any permanent cure. Unlike in ulcerative colitis, surgical treatment is palliative and is aimed at treating the complications of the disease.

36.1.1 Medical Treatment

The medical treatment of Crohn' disease has evolved significantly during the past decade. Generally speaking, the more widespread and more distal in the bowel the disease activity is, the more aggressive medical management must be. Exclusive enteral nutrition with liquid formula feeds is recommended as safe first-line therapy to induce remission. In moderate to severe disease, an alternative induction medication is corticosteroids. As with ulcerative colitis, steroids should not be used in maintenance therapy. Antibiotics are useful as primary or adjuvant therapy for penetrating perianal Crohn's disease. Exacerbations, relapses, or refractory Crohn's disease require more effective medication; biologic drugs including TNF-α antibodies (infliximab, adalimumab) may be more effective than steroids to induce remission. Thiopurines or methotrexate may be helpful in maintaining remission, but many patients also require biologic drugs to stay in remission. Biologic therapy is effective in the management of fistulizing Crohn's disease. Internal and severe rectoperineal fistulas respond well to infliximab therapy with adjuvant antibiotics, and most patients resume a longer period of remission.

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36.1.2 Surgery

It is evident that the development of drug therapies has reduced the need for surgical therapy for Crohn's disease. The indications for surgery are limited to cases that are refractory to medical therapy or in which medical therapy is poorly tolerated. Acute indications include medically unmanageable toxic megacolon or acute bleeding, both of which are rare. Subacute or chronic conditions that may require surgery include refractory strictures, internal or external fistula, and intra-abdominal abscesses.

The main principle in the surgical treatment of Crohn's disease is to save bowel length. Radical resections are not indicated; resection should be limited to the segment of bowel that is causing symptoms. Isolated skip lesions are left alone if they do not cause obstruction. Most bowel resections in Crohn's disease can be performed with laparoscopic techniques. In most cases, the actual bowel anastomosis is made outside the abdominal cavity. A typical laparoscopic resection in Crohn's disease is ileocecal resection.

In adults, strictureplasty has been shown to be an effective bowel-saving surgical method for multiple fibrotic stenoses. The long-term outcome in terms of disease activity, risk of recurrence, and quality of life has been very similar to the outcome following resectional surgery. There are only a few reports of strictureplasty for Crohn's disease in children, but the preliminary results are similar to those in adults.

Major bowel resections are sometimes required in patients with severe and widespread colonic disease. In these cases, rectum should be spared. Ileoanal anastomosis with or without a pouch is generally not indicated for Crohn's disease, as the long-term outcome is very unpredictable and often poor, and pouch complications are very common.

Perianal manifestations of Crohn's disease are common in children. These include skin tags, fissures, and fistulas. In most cases, the perianal manifestations cause mild symptoms or are asymptomatic. Surgical treatment should be considered only for severely symptomatic rectoperineal or rectovaginal fistulas. If surgery is required for perianal fistulas, a conservative approach is best tolerated. This includes drainage of possible abscesses and application of a silicon, noncutting seton thread to the fistula tract. The seton can be left in place for months. An acceptable cure rate can be achieved if this conservative approach is combined with biologic drugs. In most recalcitrant perianal disease, the resection of the most diseased (usually left) colonic segment and temporary bowel diversion may increase the success rate of the fistula repair. In very severe perianal disease, especially if it is associated with severe rectal manifestations, proctectomy may be the only possibility to guarantee a reasonable quality of life.

Figures 36.1, 36.2, 36.3, 36.4, 36.5, 36.6, 36.7 and 36.8 illustrate techniques that may be used in children with Crohn's disease who require surgical treatment, including a laparoscopic ileocecal resection (Figs. 36.4–36.7).

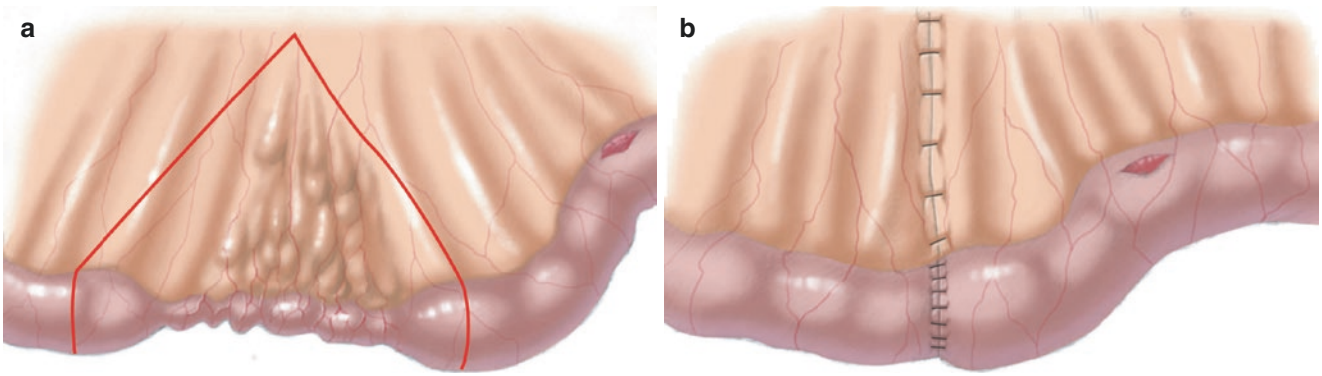

Fig. 36.1 (**a**, **b**) Resection of a localised stricture. The resection margin to healthy bowel does not need to be longer than a few centimetres. Anastomosis is performed with an absorbable suture material with one-layer interrupted stitches

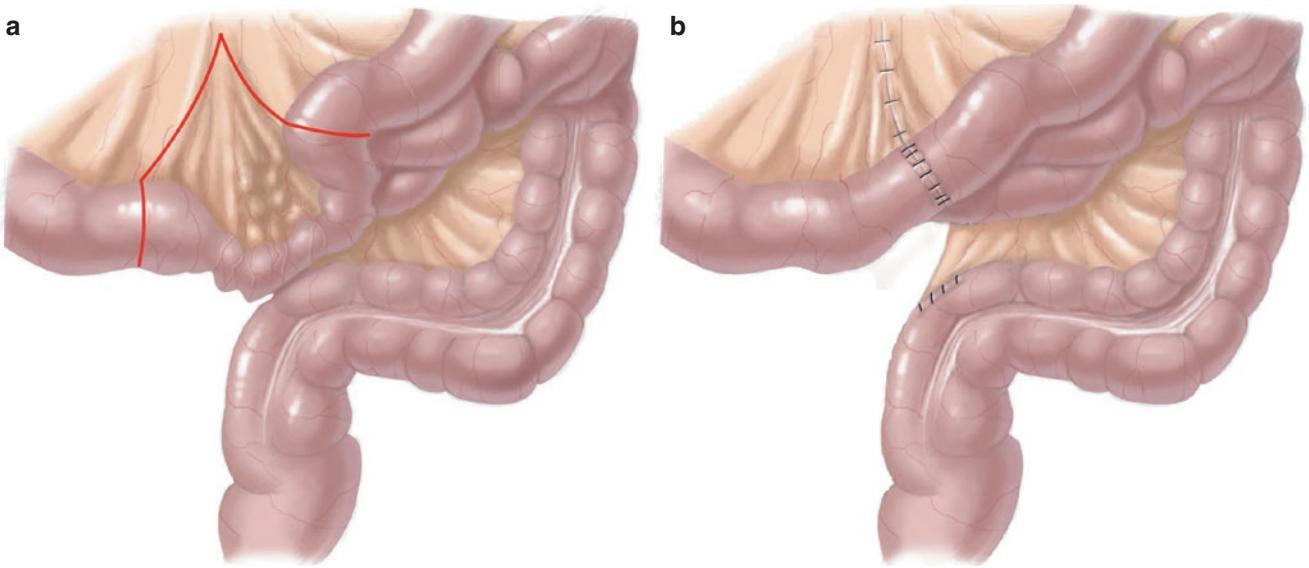

Fig. 36.2 (**a**, **b**) Repair of an internal fistula. In children, internal fistula is often between the ileum and the sigmoid bowel. The ileum is usually affected by the disease and the sigmoid is healthy except for the fistula site. The proximal disease segment is resected with primary anastomosis and the distal fistula opening is excised and closed with interrupted sutures

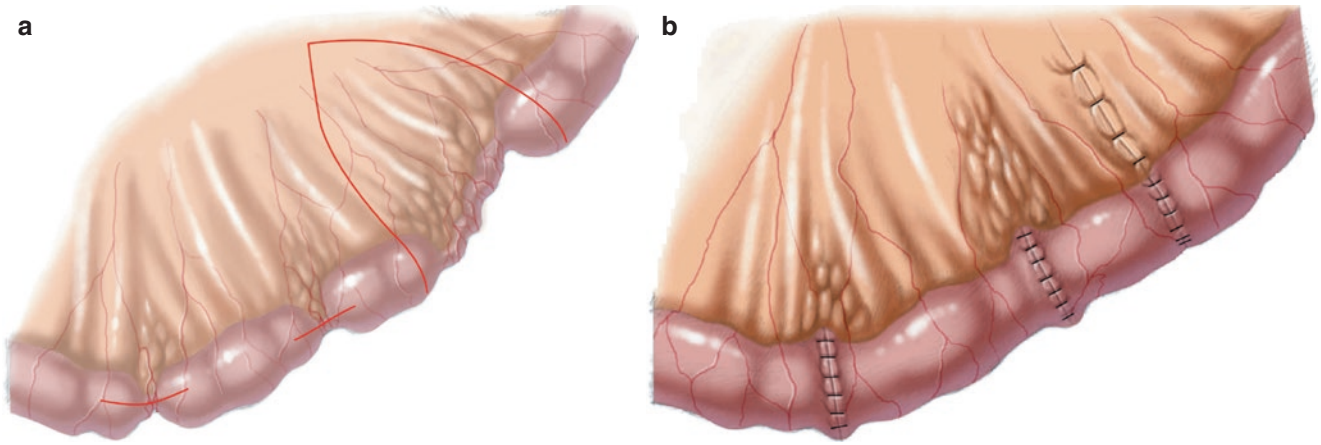

Fig. 36.3 (**a**, **b**) Strictureplasty for multiple short jejunoileal strictures. Strictureplasty can be combined with resection of longer stricture segments. The stricture site is opened longitudinally so that the incision extends to the macroscopically healthy bowel. The longitudinal opening is closed transversely with interrupted sutures

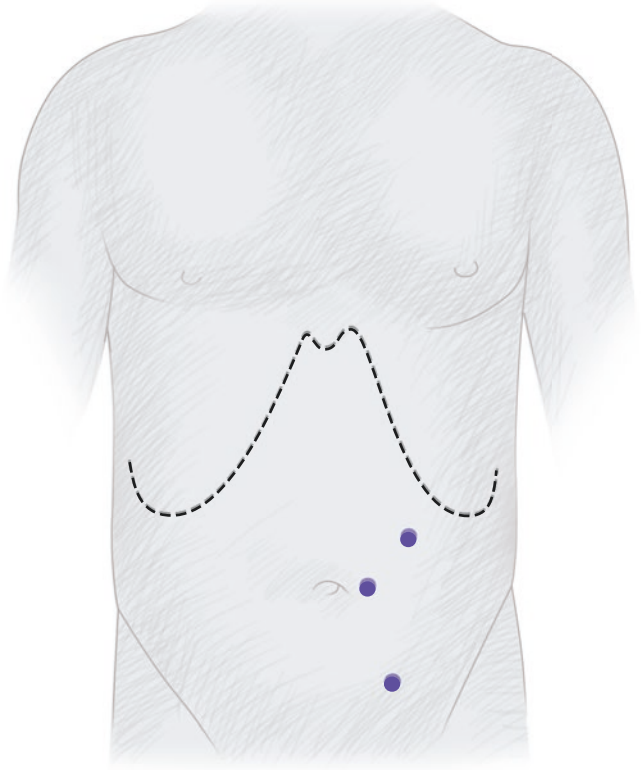

Fig. 36.4 Laparoscopic ileocecal resection. Port sites for laparoscopic ileocecal resection. The first trocar in inserted through umbilicus either openly or after Veress needle insufflation. The other trocars are placed under direct vision lateral to rectus muscles to avoid injury to epigastric vessels

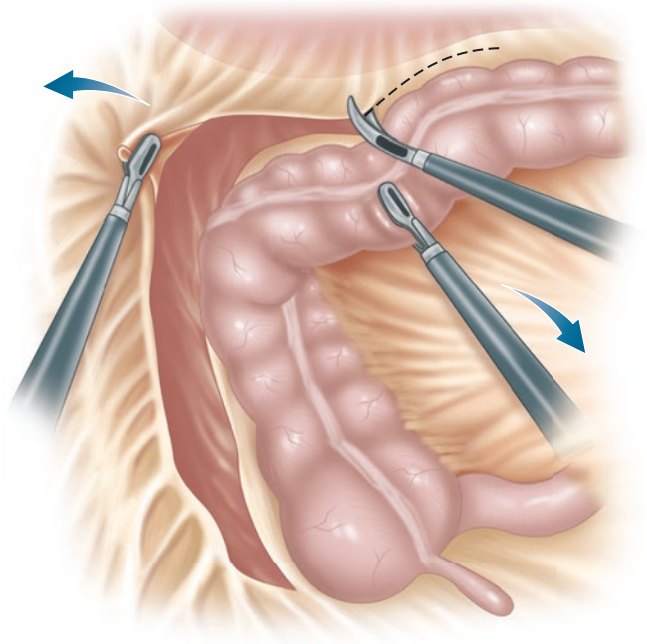

Fig. 36.5 The lateral attachments of the right colon are mobilized with a Harmonic scalpel or vessel sealing device (LigaSure®). The right ureter should be visualised before any vessel coagulations. The right colon and cecum are fully mobilised, along with the terminal ileum. The ileocecal area can be devascularized intracorporeally or extracorporeally after exteriorization of the bowel segment

Fig. 36.6 A short incision is made in the right iliac fossa, and the mobilised (and devascularized) ileocecal segment is brought through the incision. The diseased segment is resected

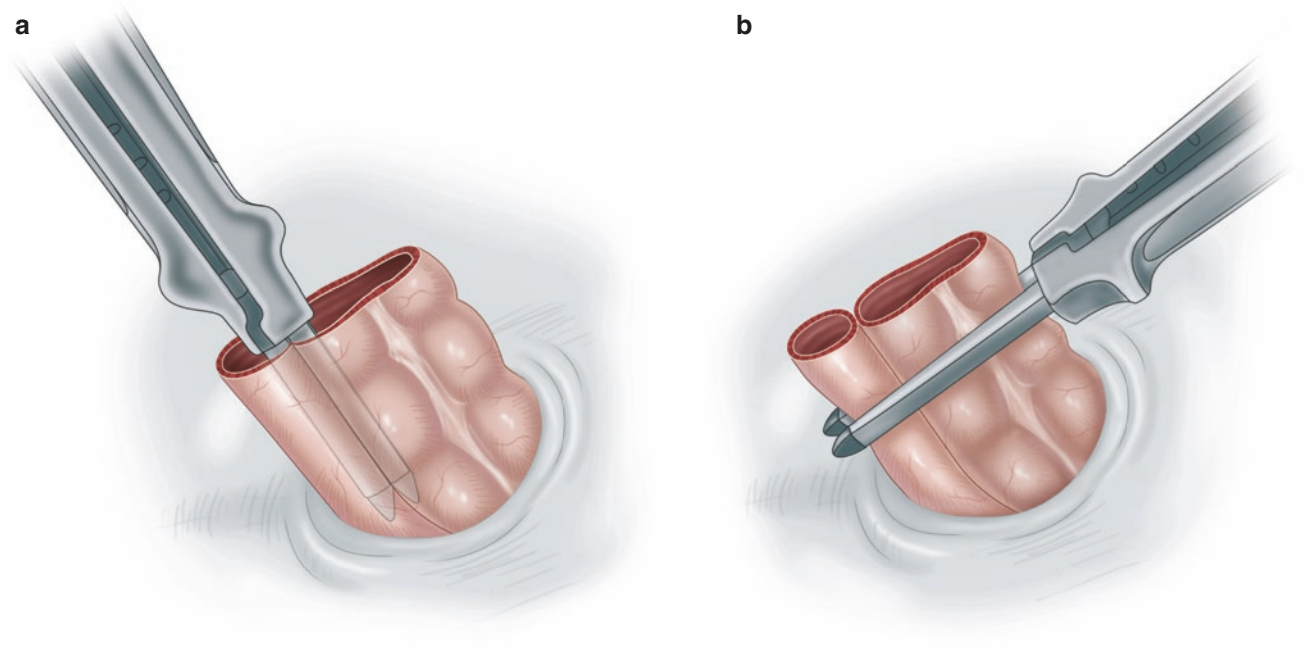

Fig. 36.7 An ileocolonic "functional end-to-end anastomosis" is performed by two firings of a linear stapler

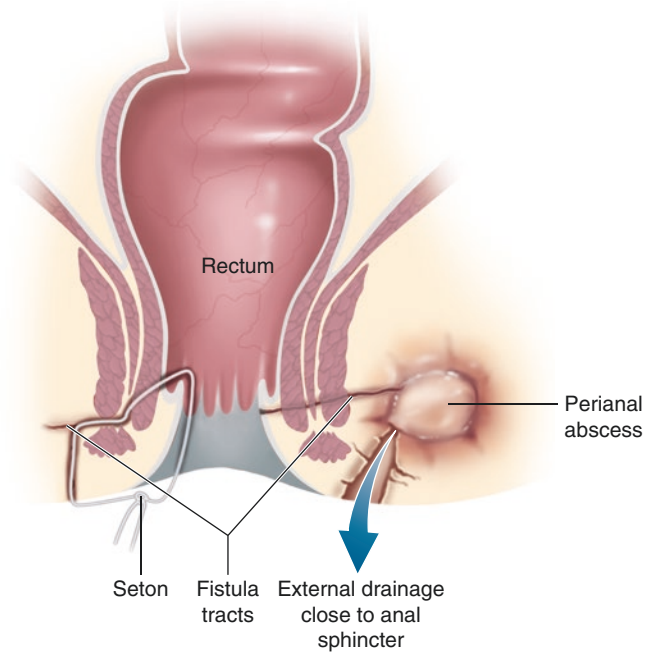

Fig. 36.8 Drainage of a perianal abscess. After drainage, a noncutting silicone seton is applied through the fistula tract (left side)

36.2 Outcome

Operative treatment of Crohn's disease is aimed at management of complications of the disease that are refractory to medical treatment. Although permanent cure cannot be expected following surgery, many patients resume longer periods of full remission with a normal lifestyle. Proper surgical treatment in selected patients can also decrease or even abolish the need for immunosuppressive or other potentially harmful medication for months or years.

Suggested Reading

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Stomas for Large and Small Bowel

37

Andrea Bischoff and Alberto Peña

The creation of an intestinal stoma is an operation frequently performed in pediatric surgery, especially for cases of anorectal malformations or necrotizing enterocolitis, and occasionally for Hirschsprung's disease. Its purpose is to divert the fecal stream, for various reasons. The diversion can be total (separated stomas) or partial (loop colostomies).

Colostomy in pediatrics is a technically demanding operation with a significant associated morbidity, which is the reason behind the general tendency to perform more primary colorectal procedures without a protective colostomy in children. It is understandable and justified to look for treatment modalities that avoid the performance of a colostomy, but there are many clinical conditions in which a stoma is mandatory. The morbidity related with the opening and closure of stomas is considered preventable, provided that the surgeon pays attention to important technical details.

37.1 Types of Colostomies

Two types of colostomies may be performed:

- *Loop colostomy*, in which both stomas (proximal and distal) are created together. These colostomies are easy and quick to create and to close, but they have several disadvantages: the potential for frequent passing of stool from proximal to distal bowel; fecal distal impaction and urinary tract fecal contamination in cases of anorectal malformation.
- *Separated stomas*, in which both stomas are completely separated. This type guarantees that stool will not pass into the distal bowel, but it is a time-consuming and technically demanding operation.

The location of the colostomy also has important implications:

- *Sigmoid colon*. Colostomies of the sigmoid colon make it easy to clean the distal colon and to perform a distal colostogram. They can interfere with the final reconstruction of an anorectal malformation, however, because of the short distal colon.
- *Transverse colon*. Colostomies in this location leave a long distal segment, which may represent an advantage for further anorectal and/or vaginal reconstruction, but they also present several disadvantages:
  - Inability to clean the distal colon, leaving meconium or stool that may be a source of infection during future reconstruction
  - Inability to perform a successful diagnostic distal colostogram
  - Megarectosigmoid as a result of continuous accumulation of mucous and mucosal desquamation. As a result, tapering will be needed during rectal repair, and more serious constipation is likely
  - Potential hyperchloremic acidosis as a result of urine absorption, in cases of anorectal malformation
  - Urinary tract infection

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P. Puri, M. E. Höllwarth (eds.), *Pediatric Surgery*, Springer Surgery Atlas Series,
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37.2 Goals of a Colostomy

In the management of anorectal malformations, the objectives of a colostomy are very specific:

- Because more than 85% of patients with an anorectal malformation have a distal communication with the urogenital tract, the colostomy must be totally diverting, to avoid fecal contamination of that tract.
- Both stomas must be completely separated.
- The stoma bag must cover only the proximal stoma.
- The recommended location of the stomas is the left abdomen (Fig. 37.1).
- The proximal stoma must include a fixed portion of the descending colon and must be located in the center of a triangle represented by the iliac bone, the umbilicus, and the lowest rib.
- Special attention must be paid to leave enough length of distal colon to allow a successful future pull-through.
- The distal colon must be tapered enough to create a 3-mm, nonprolapsed mucous fistula, which will be used only for irrigations (Figs. 37.1 and 37.2) and for injection of contrast material with diagnostic purposes.
- Avoid fecal contamination.
- Fix both the proximal and distal limb to the peritoneum and aponeurosis.
- The proximal stoma must be wide open and have good blood supply.

An important step during the creation of a colostomy is to irrigate the distal portion of the bowel with saline solution until it is completely clean of meconium. Leaving meconium distally can result in fecalomas and contamination of the urinary tract when there is a fistula between the rectum and the urinary tract.

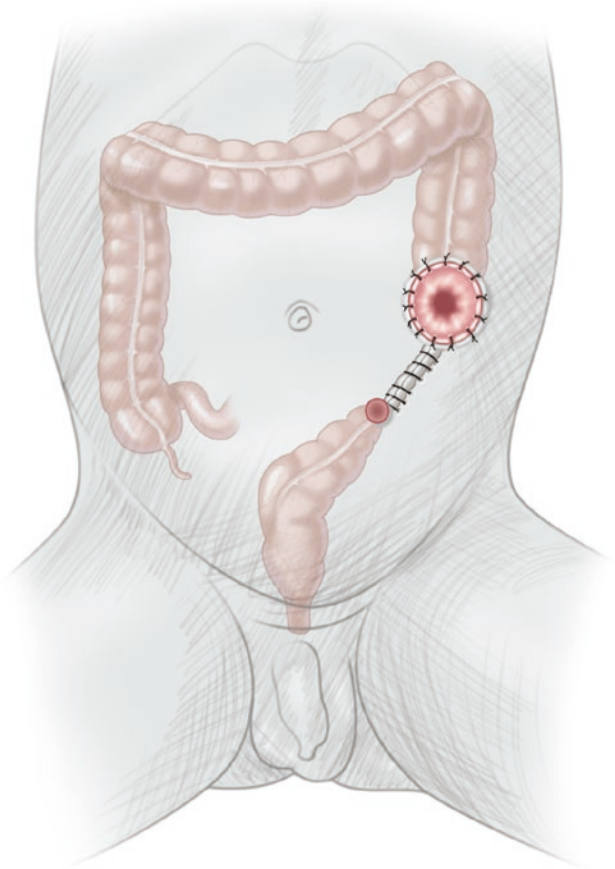

Fig. 37.1 Descending colostomy, with mucous fistula

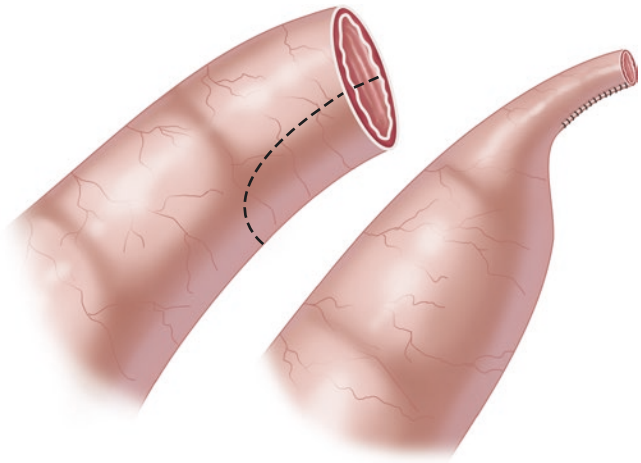

Fig. 37.2 Distal stoma made tiny and flat to help avoid prolapse

37.3 Most Common Error: Stoma Mislocation

There are several types of stoma mislocation. The diagnosis of a mislocated colostomy is best determined by a distal colostogram, which is a routine study done prior to the main repair of an anorectal malformation.

37.3.1 Proximal and Distal Stomas Too Close to Each Other

The colostomy bag covers both of these stomas and allows passage of stool to the distal stoma. As a consequence, some patients suffer from recurrent urinary tract infections. Also, the stoma bag may be difficult to adapt to the skin. This problem can be managed by performing the main repair earlier than anticipated, or by revising the stomas to separate them. If proceeding with the main repair in a patient with this kind of colostomy, the patient may pass stool through a reconstructed area, which may increase the risk of infection. Alternatives include cleaning the entire gastrointestinal tract preoperatively and then keeping the patient fasting for a period of time (about a week), receiving parenteral nutrition; or using a heavy purse-string of an absorbable suture in the distal stoma to occlude its lumen temporarily and avoid the postoperative passage of fecal material.

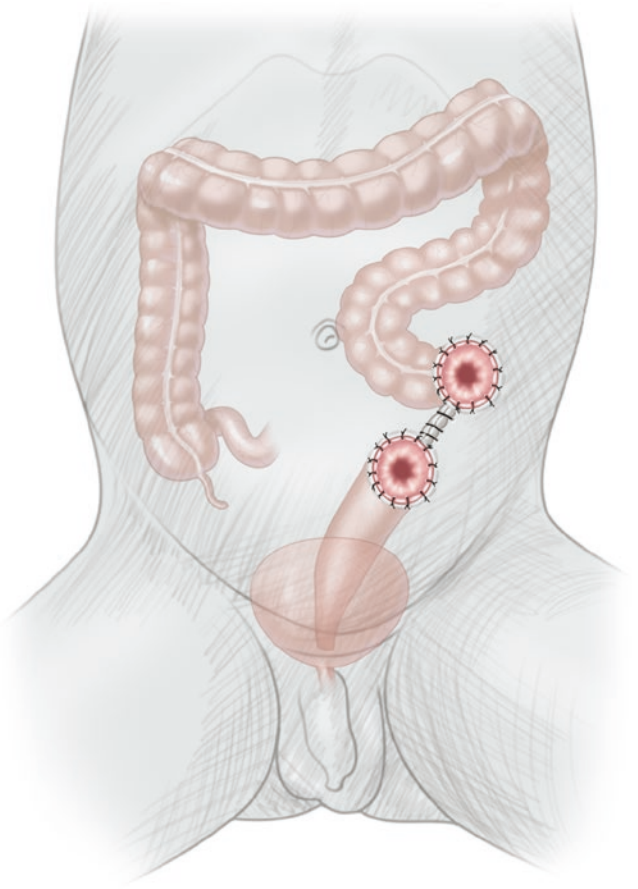

Fig. 37.3 Colostomy placed too distal, with not enough distal bowel for the pull-through

37.3.2 Colostomy Created Too Distal in the Sigmoid

When the colostomy is too distal in the sigmoid, there is insufficient distal length for the rectal pull-through (Fig. 37.3). In this case, the surgeon has several options. The first is to perform the pull-through, taking down the distal stoma (mucous fistula) and closing it as a Hartman pouch. Sometimes the piece of bowel is so short that it will make a very difficult future colostomy closure because the anastomosis will have to be done deep down in the pelvis, behind the bladder. The second option is to close the colostomy at the time of the main repair and then pull down the rectum, leaving the patient without the benefit of a protective colostomy. For that option, we recommend cleaning the entire gastrointestinal tract preoperatively and leaving the patient to fast for 7–10 days, receiving parenteral nutrition. The third option is to close the colostomy, pull the rectum down, and create a proximal new colostomy to divert the stool and protect the perineum.

It is vital to try to save the distal rectum, and to avoid discarding it and pulling through the proximal stoma. Saving the distal rectum is particularly important in patients with anorectal malformations, in whom the resection of the rectum will result in constant passing of stool and a greater chance of fecal incontinence.

37.3.3 Inverted Stomas

The functional (proximal) stoma is inadvertently placed in an inadequate location, making it very difficult to adapt the stoma bag (Fig. 37.4). An additional problem occurs when the surgeon mistakenly tapers the bowel, assuming that it is the mucous fistula, which may provoke an obstruction. This complication may require a re-operation. Another problem with an inverted stoma is that the distal bowel may be under tension, requiring mobilization of the distal stoma in order to have adequate length for the pull-through.

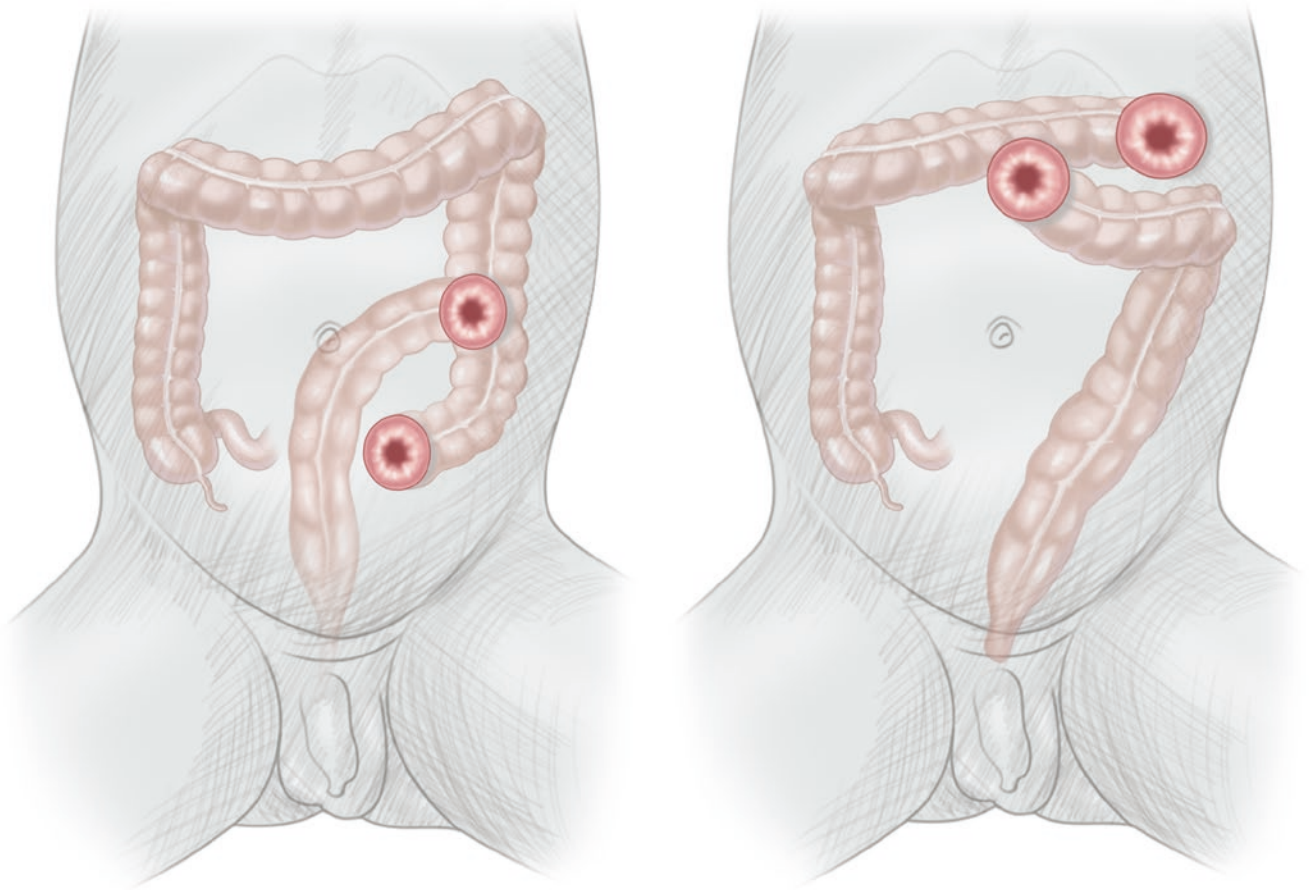

Fig. 37.4 Inverted stomas

37.3.4 Sigmoid Colostomy in the Upper Abdomen

In this case depicted in Fig. 37.5, the surgeon planned to do a transverse colostomy, made an incision in the upper abdomen, found a piece of colon, and brought it out as a colostomy, believing it was the transverse colon. Instead, a sigmoidostomy was inadvertently opened in the upper abdomen, which then can interfere with the pull-through. When opening a transverse colostomy, the surgeon must remember that newborns have a very dilated and mobile sigmoid colon, which can reach the diaphragm. When this error is detected before the pull-through, the colostomy must be moved to the lower abdomen.

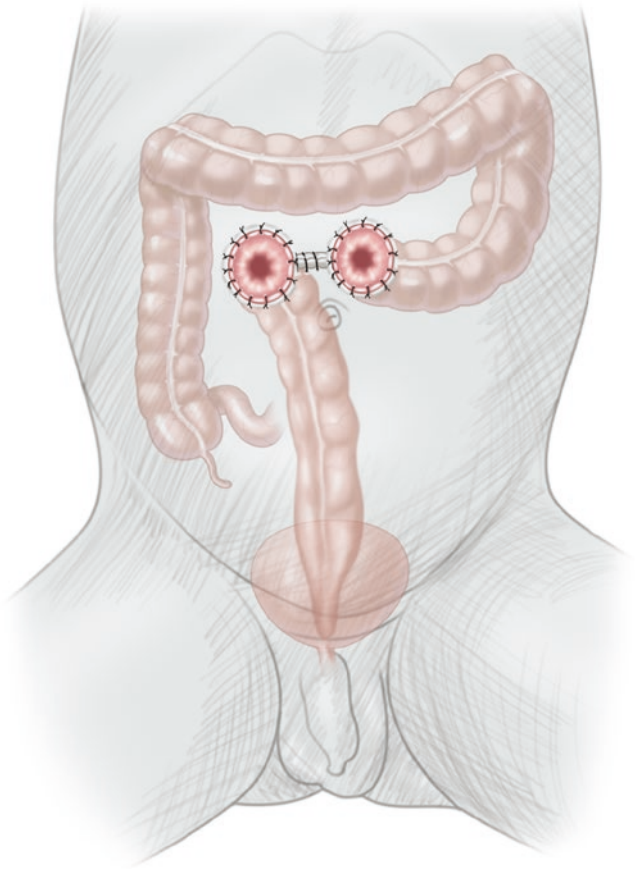

Fig. 37.5 Right upper sigmoidostomy instead of a right transverse colostomy

37.4 Most Common Complications of Colostomies

37.4.1 Prolapse

Prolapse can be a serious complication that can result in intestinal loss due to ischemia. Most prolapses can be avoided by creating the colostomy adjacent to a fixed portion of the colon. If the colostomy must be created in a mobile portion of the bowel, the colon must be fixed to the anterior abdominal wall, approximately 6–7 cm proximal to the stoma. Figure 37.6 identifies the mobile and fixed portions of a normally rotated colon and points out the segments that are prone to prolapse and will need fixation to the abdominal wall.

If a patient presents with significant prolapse, surgical repair may be required. A simple technique is to insert a large amount of packing gauze soaked in povidone-iodine in the prolapsed bowel, gently reducing the prolapse. Then the abdomen is palpated, feeling for the mass that corresponds to the packing gauze inside the bowel, naturally oriented inside the abdomen. An incision is then made on top of the palpated mass, usually about 5 cm away from the stoma. The incision goes through skin, subcutaneous tissue, muscle, aponeurosis, and peritoneum. The bowel full of gauze is easily identified. The peritoneum and aponeurosis are closed with interrupted Vicryl stitches, including in each stitch a bite of the bowel wall (without taking the packing gauze) and securing it to the abdominal wall, to avoid future prolapse. With this technique, the stoma will not be touched.

37.4.2 Stenosis

When stenosis occurs, the patient may suffer from obstructive symptoms. When opening a stoma, it is important to create an adequate space for the functional bowel to pass through without being compressed by the fascia. Avoid creating stomas through a simple stab wound. A circle of skin must be resected, as well as a circle of aponeurosis, muscle, and peritoneum. Most cases of stoma stenosis do not respond to dilations and need reoperation.

37.4.3 Retraction

Retraction is a technical mistake and therefore a preventable problem. An acute, early retraction can be a surgical emergency. A late retraction may represent a serious difficulty in managing the stoma, because it will be hard to adapt the stoma bag. The reoperation includes the mobilization of the bowel higher above the skin surface, in order to have enough colon to achieve a successful “mature” (everted) stoma.

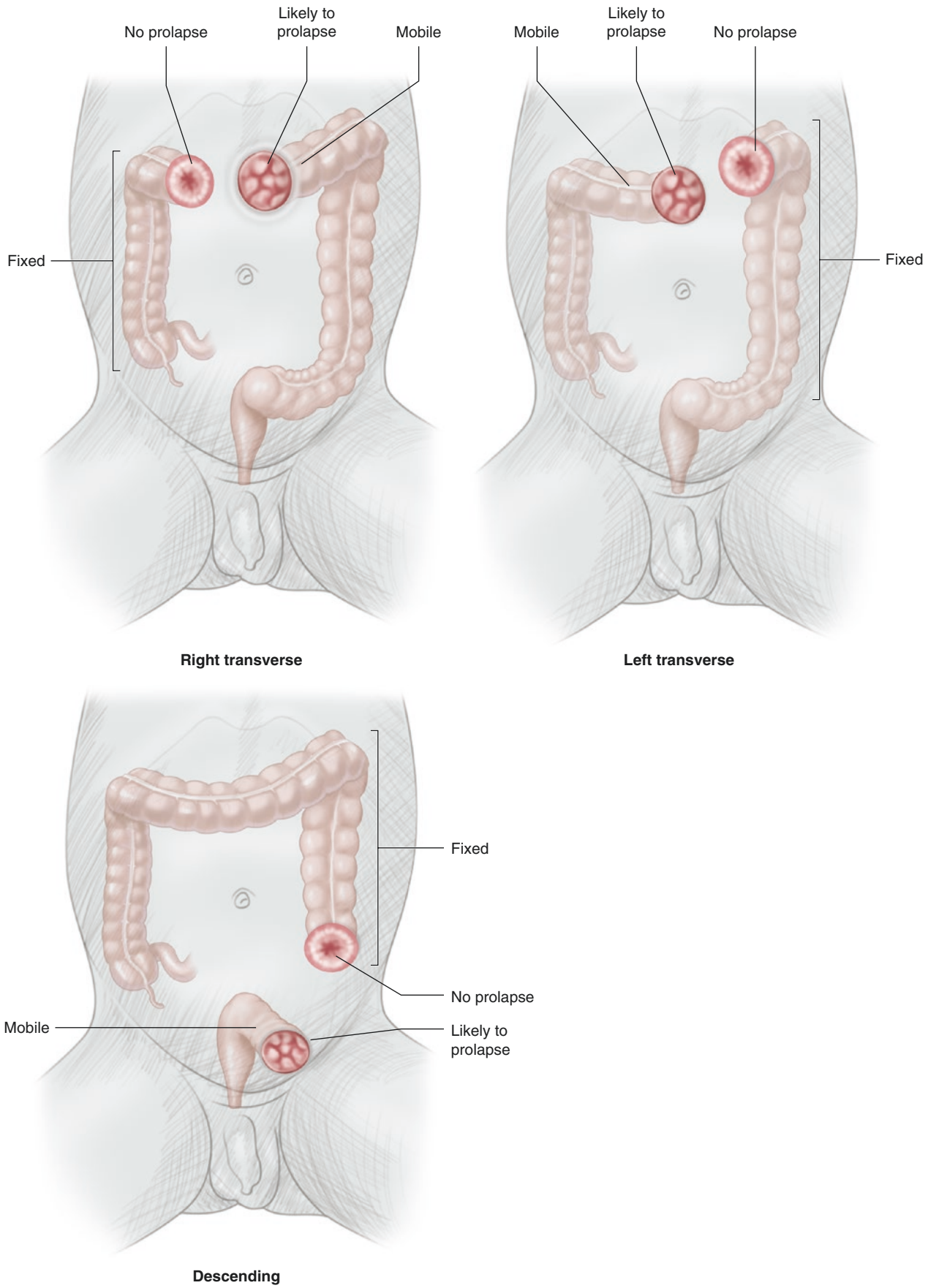

Fig. 37.6 Right transverse colostomy (distal stoma likely to prolapse); left transverse colostomy (proximal stoma likely to prolapse); descending colostomy (distal stoma likely to prolapse)

37.5 Stomas in Cloacal Exstrophy

A special comment should be made about stomas in patients born with cloacal exstrophy. A common misconception is that these patients have a short, and therefore useless, colon. As a consequence, there is a tendency to perform an ileostomy at birth and use the colon for urogenital reconstruction or simply discard it. This procedure must be avoided because the colon is extremely valuable for these patients. It allows them to form solid stool, making them future candidates for a colonic pull-through. It is very important to incorporate every single piece of colon into the fecal stream, regardless of how small it is. Over time, those small pieces of bowel grow considerably. Another advantage of incorporating colon in the fecal stream is that those patients become easier to manage than a patient having an ileostomy. Patients who have undergone an ileostomy leaving the distal colon attached to the bladder are often observed to suffer from hyperchloremic acidosis, which may interfere with growth and development. Such patients benefit from a "rescue operation" taking down the ileostomy, separating the colon from the urinary tract, and opening a real end colostomy.

37.6 Ileostomies

Ileostomies are commonly performed in patients with total colonic aganglionosis and necrotizing enterocolitis. Because by definition the ileostomy will always be made in a mobile portion of the bowel, the risk of prolapse is very high, so it is indicated to fix the bowel to the anterior abdominal wall about 6 or 7 cm proximal to the stoma. All the other complications described for colostomy also apply to ileostomies.

The other particular characteristic of ileostomy is that electrolyte disturbances may occur. Patients need to maintain good hydration, and parents need to be keen observers of stoma effluent. These patients tend to have significant sodium losses and may need oral sodium supplementation.

37.7 Colostomy Closure

Colostomy closure in the pediatric population is also associated with a high morbidity rate due to potential avoidable complications such as anastomotic dehiscence, stricture, wound infection, bleeding, and death.

Following are the steps of the operative and perioperative protocol for colostomy closure:

- Admission on the day before surgery
- Clear liquids by mouth
- Repeated proximal stoma irrigations with saline solution, 24 h prior to the operation
- Administration of IV antibiotics during anesthesia induction, continued for 48 h
- Meticulous surgical technique, including packing of the proximal stoma, plastic drape to immobilize the surgical field, and multiple silk sutures in the mucocutaneous junction of the stomas to provide uniform traction that allows the surgeon to identify the correct dissection plane, remaining as close as possible to the bowel wall (Fig. 37.7)
- Careful hemostasis with emphasis on avoiding contamination and cleaning the edges of the stomas to allow a precise anastomosis (Fig. 37.8)
- A two-layer, end-to-end anastomosis with separated, long-term 6-0 absorbable sutures (Fig. 37.9)
- Generous irrigation of the peritoneal cavity and subsequent layers with saline solution, closure by layers to avoid dead space, avoidance of hematomas, and wound coverage with Dermabond® (Fig. 37.10)
- Postoperatively, no nasogastric tubes are used, and the patients receive clear fluids on the first postoperative day, if they are not distended or nauseated. Most patients can be discharged home on the second or third day following the operation.

If the size discrepancy between the proximal and distal stomas is greater than 5:1 and the diameter of the distal stoma is less than 1 cm, the colostomy closure can present a serious challenge. In such cases, it is recommended to perform an end-to-side anastomosis, plus a window-type stoma created about 5–10 cm proximal to the anastomosis (Fig. 37.11). During the first few postoperative days, the fecal output through the window may seem large, but eventually that output decreases and the amount of stool passing through the downstream bowel increases, until the window closes up, the anastomosis is efficient, and the microcolon grows.

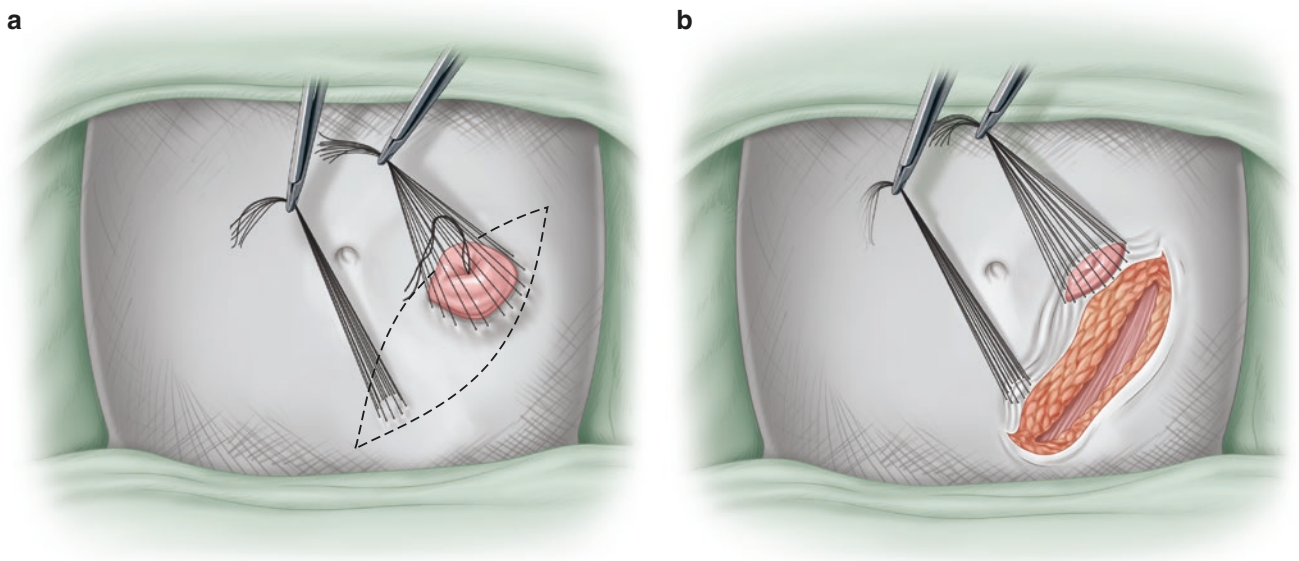

Fig. 37.7 Multiple silk sutures in the mucocutaneous junction, to allow for uniform traction. The *dotted line* shows the elliptical incision. The opening is performed layer by layer

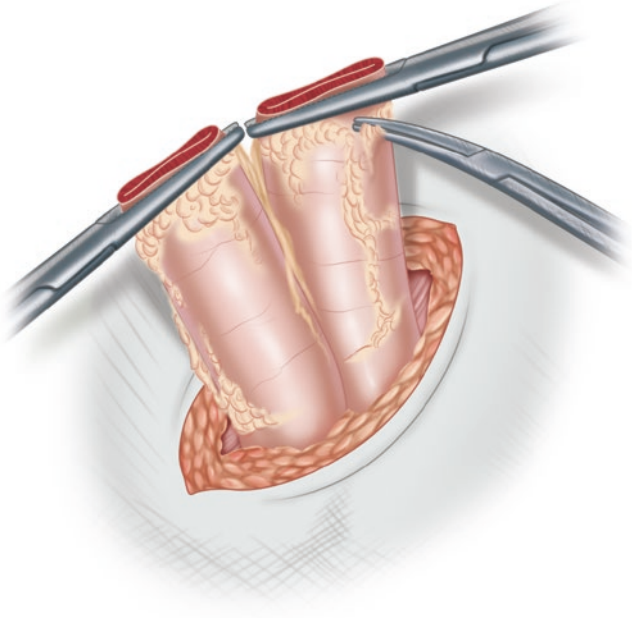

Fig. 37.8 Cleaning the edges of the stomas, preparing for the anastomosis

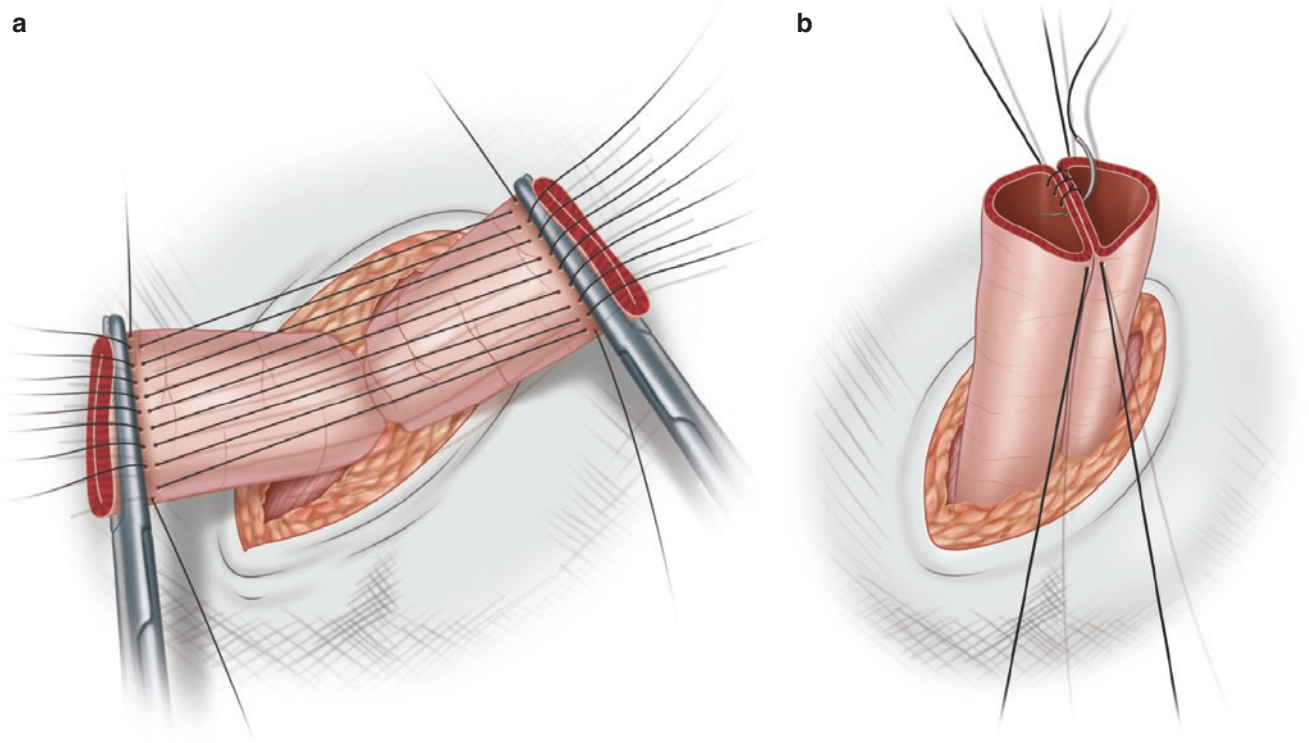

Fig. 37.9 Two-layer anastomosis: (**a**) External layer of posterior wall. (**b**) Internal layer of posterior wall. (**c**) Internal layer of anterior wall. (**d**) External layer of anterior wall

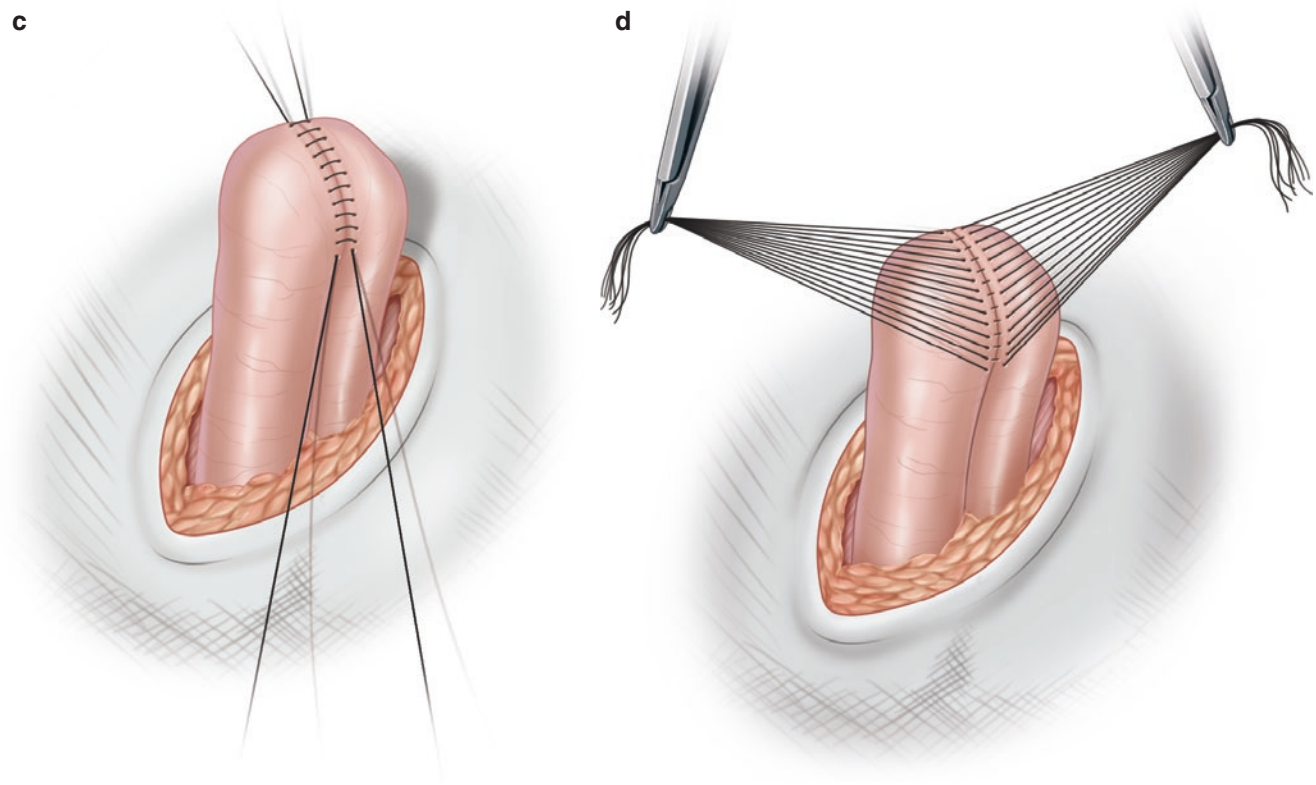

Fig. 37.9 (continued)

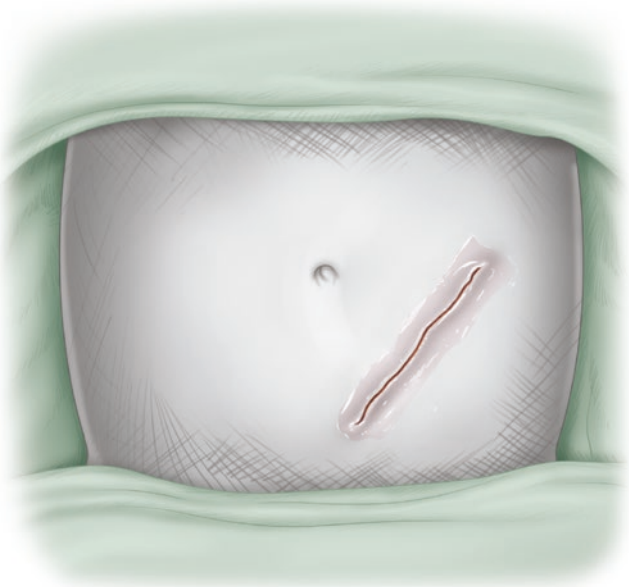

Fig. 37.10 Closed wound, covered with Dermabond®

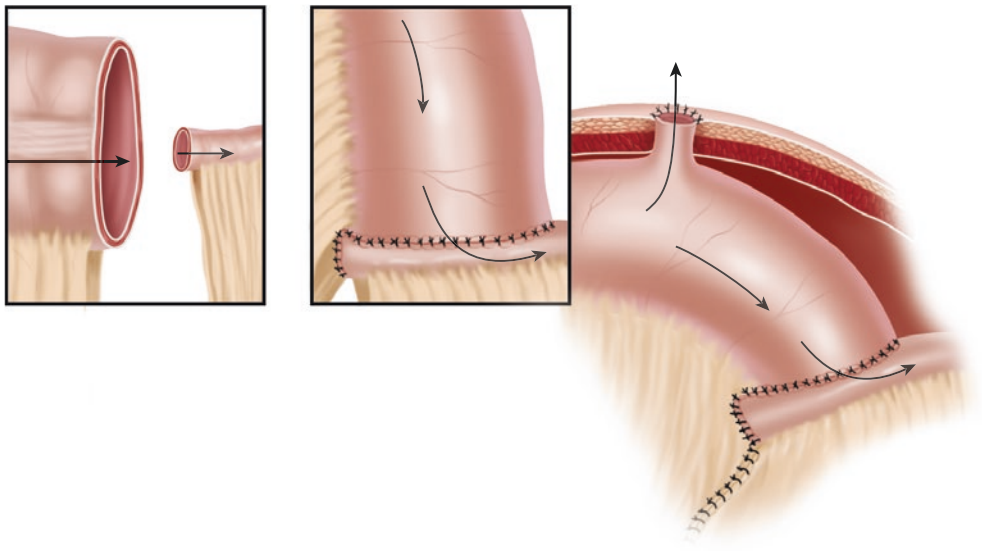

Fig. 37.11 End-to-side anastomosis for size discrepancy greater than 5:1. A window-type stoma is also created about 5–10 cm proximal to the anastomosis

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Rectal Biopsy

38

Augusto Zani

Rectal biopsy is performed in the paediatric age group almost invariably to rule out Hirschsprung's disease, for which it represents the gold standard for diagnosis. This concept was introduced in 1948 by Swenson and Bill, who first described the method of full-thickness transrectal biopsy to accurately identify aganglionosis. The seminal discovery that the level of aganglionosis is identical in the submucosal and the myenteric plexus paved the way for a less invasive technique for rectal biopsy, using a suction capsule. The advantage of this technique is that it can be safely performed at the bedside or in the clinic, it does not require general anaesthesia or hospitalization, and it is relatively painless.

38.1 Instruments and Techniques

Of all the various submucosal biopsy devices described over the years, the most popular one has been the Noblett forceps. This instrument consists of a blunt-ended tube with a side hole, an attachment for a suction tube and syringe, and a knife blade that is triggered once the suction is applied to the rectal mucosa. Nowadays, the rbi2 (Aus Systems, Allenby Gardens, South Australia), a novel forceps device that uses the same principle of the Noblett forceps, has become the most widely used device on the market. According to Hall et al., this forceps device offers superior efficacy over the Noblett forceps, and its use is likely to be more cost-effective.

Regardless of the technique and the instrument used, classically the absence of ganglion cells (on frozen sections or on hematoxylin/eosin stains) and the presence of thick (>40 μm) nerve trunks (acetylcholinesterase expression) define the diagnosis of Hirschsprung's disease. Whilst a full-thickness biopsy provides the pathologist with larger samples containing both submucosa and muscularis propria, a suction biopsy is more superficial and impedes comparison between the smaller and sparse submucosal ganglion cells and the more compact myenteric ones. To facilitate the diagnosis of aganglionosis from submucosal biopsies, a number of alternative diagnostic neuronal markers have been reported in the literature over the years, and some have been introduced into clinical practice, such as calretinin, nicotinamide adenine dinucleotide phosphate diaphorase (NADPH-d), and S-100.

38.2 Preoperative Care

Before performing a rectal biopsy, several steps are essential:

- Ensure that the patient has no signs of acute enterocolitis or other clinical illness
- Use washouts for decompression
- Obtain informed consent from the parents
- Ensure that a pathologist is available to receive the specimen

It is the author's practice to administer a single prophylactic dose of amoxicillin/clavulanic acid to cover the procedure.

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38.3 Rectal Biopsy Procedures

38.3.1 Rectal Suction Biopsy

The rectal suction biopsy device (Fig. 38.1) is assembled according to manufacturer instructions. If necessary, a pressure manometer is attached to the instrument in order to exert adequate negative suction pressure to the rectal mucosa, making it easier to obtain an adequate tissue sample and reducing the risk of unsuccessful biopsy.

Infants are held in the lithotomy position or with legs up (Fig. 38.2), and children are placed in the left lateral knees-bent position. Although this procedure is also described in adults, in the author's practice rectal suction biopsy is reserved for infants and toddlers, as older children are less compliant.

The instrument is adequately lubricated and inserted into the anus. It is important that the side hole of the capsule is positioned facing the posterior or lateral wall of the rectum, in order to avoid perforation into the rectovesical or rectovaginal pouch of the peritoneal cavity in case of an accidental full-thickness biopsy. Negative suction pressure should be gently applied to the rectal wall, so that the mucosa is drawn inside the side hole of the capsule and is then cut by a moveable blade, once the blade is triggered. If a pressure manometer is used, a pressure of −150 cm H_2O should be applied for 1 to 5 s to enable adequate tissue suction. Pressing the capsule too forcefully onto the mucosal surface will result in unsuccessful biopsy.

Specimens are taken at 3, 3.5, and 4 cm from the anal verge to avoid the anatomic hypoganglionic zone and diagnostic confusion. At each time, the specimen is retrieved from the capsule, placed on wet gauze for frozen sections, and adequately labelled.

Fig. 38.1 The rectal suction biopsy device

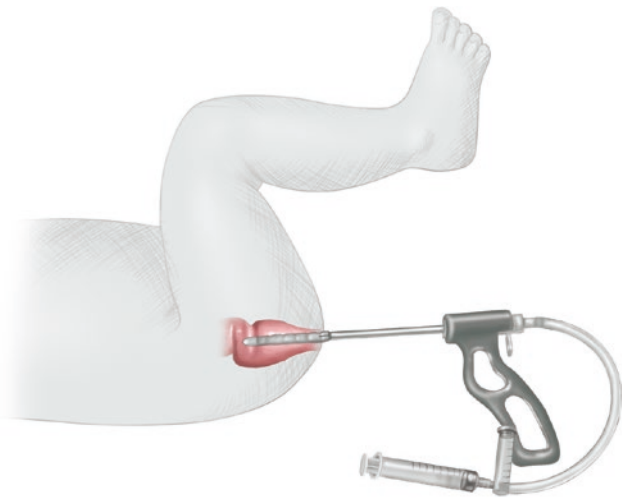

Fig. 38.2 Position for rectal suction biopsy

38.3.2 Open Rectal Biopsy

An open rectal biopsy is performed under general anaesthesia in selected cases, such as previous inadequate tissue sampling with suction technique (usually following two unsuccessful attempts), in older children who would be incompliant with suctioning technique, or as an additional part of another procedure in a patient already under general anaesthesia.

The patient is held in the lithotomy position. A self-retaining retractor, such as a nasal speculum or a Park's retractor, is introduced in the anus and held in position by an assistant.

A first stay stitch is placed on the midline in the posterior rectal wall at least 2 cm above the dentate line, the ends are tied, and the needle is cut off. Applying traction on this stay stitch, a second stay stitch is placed 2–3 cm higher and its ends are tied, but the needle is left attached (Fig. 38.3).

The tissue between the two stay sutures is retracted with forceps and an adequate specimen is excised using sharp-pointed scissors (Fig. 38.4). Generally, only one specimen of adequate size is required. The needle of the second stay stitch is used to repair the defect with running locking suture, which is finally tied with the end of the first stay stitch. If the running locking suture has not achieved satisfactory haemostasis, bipolar diathermy can be employed.

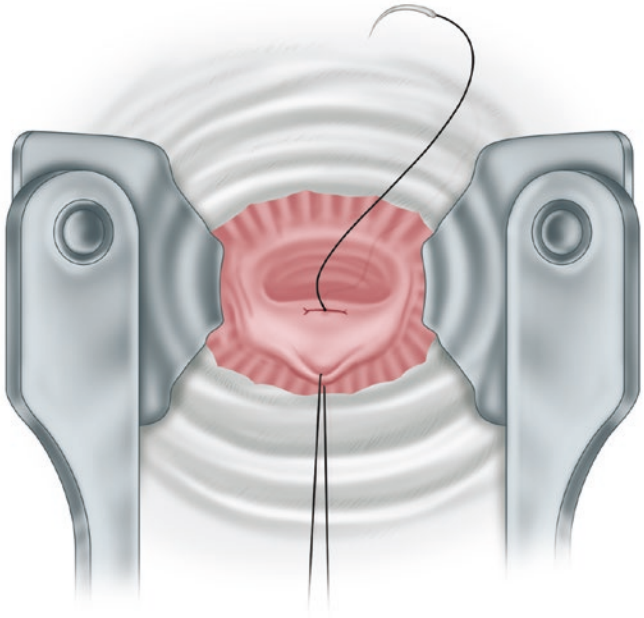

Fig. 38.3 Placement of stay stitches in open rectal biopsy

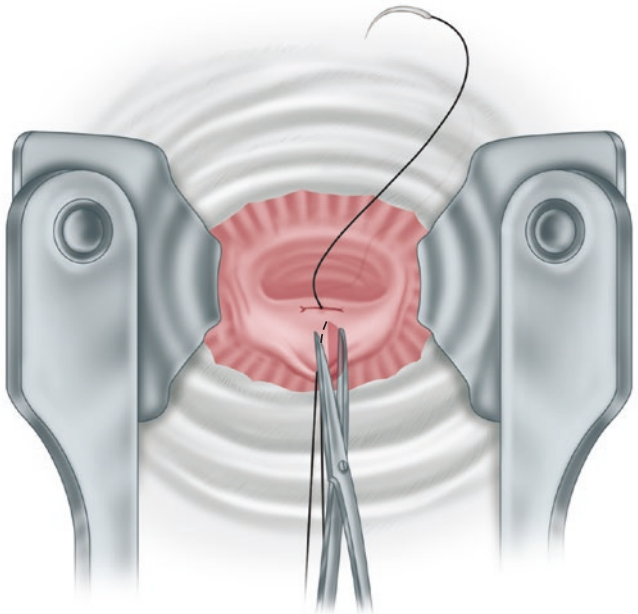

Fig. 38.4 Excision of the specimen in open rectal biopsy

38.4 Postoperative Care

After a rectal biopsy, it is essential to ascertain that the patient is in no acute distress and vitals are stable and that the specimens are adequately labelled and delivered to the histopathology department in a timely fashion. No *per rectum* medications, suppositories, or enemas should be administered for at least 24 h.

Outpatients who underwent a rectal suction biopsy can be discharged home after at least 2 h of postbiopsy observation. Children who had an open rectal biopsy are usually admitted on a day-surgery ward and observed for at least 4 h after anaesthesia, before being discharged home.

Common analgesics are advised for all patients. Parents must be told that the child may pass a small amount of blood from their anus, particularly when they pass stools. If active bleeding does not stop spontaneously or if the patient develops abdominal distension and signs of peritonism, parents must seek medical advice as soon as possible.

38.5 Complications

Several types of complications are possible:

- *Infection*. Cases of pelvic sepsis have been reported in the literature, especially as a result of unrecognised rectal perforation, justifying the use of preprocedural prophylactic antibiotics.
- *Bleeding:* Bleeding that stops spontaneously is a common event. Rarely, bleeding requires blood transfusion or diathermy/stitching.
- *Insufficient sample.* This is the commonest complication of the rectal suction technique, especially if the biopsy is performed by an inexperienced operator. The retrieval of more than one specimen is thus justified.
- *Perforation:* This is a rare complication (<1%) that can usually be managed nonoperatively with antibiotics, bowel rest, and nasogastric suction; sometimes surgery is required.

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Rectal Prolapse in Children

39

Paolo De Coppi

Prolapse of the rectum is a relatively common and self-limited problem in children, with a peak incidence between 1 and 3 years of age. The prolapse can be either partial (only rectal mucosa) or complete (full thickness).

Rectal prolapse is often idiopathic in children and the vast majority of these patients have no predisposing factors, except that the sacrum in this age group is not curved as in older age groups but rather straight towards the anus allowing to prolaps the rectum in cases of difficult defacation or constipation. However, it can be caused by factors that can cause undue straining, such as diarrhea and constipation. It is also associated with autism, neuromuscular problems such as meningomyelocele, or exstrophy of the bladder, and there is an increased incidence of rectal prolapse in children with cystic fibrosis associated with tenacious stool, chronic cough, and loss of perirectal fat. More than one fifth of patients with cystic fibrosis will develop rectal prolapse, and cystic fibrosis will be diagnosed later in 20% of children seen with rectal prolapse between 6 months and 3 years of age. Rectal prolapse is also rarely associated to Shigellosis in neonates, scleroderma, lymphoid hyperplasia of the distal colon (which also acts as a leading point for rectal intussusception), parasites and other infectious agents, and HIV infection.

39.1 Diagnosis

The assessment of children presenting with rectal prolapse should include a general history and physical examination to exclude associated etiological factors such as rectal polyps, in which the polyp acts as a leading point for the intussusception. Initial investigations should include a sweat test or gene probe to exclude cystic fibrosis. Stool analysis is also usually performed.

Clinical examination is important, but it may not reveal the prolapse. Modern technology allows documentation which is often diagnosed by the parents themselves. Most commonly, the prolapse is incomplete, limited to 2–3 cm of mucosa protruding from the anus and classically displaying radial folds. Complete, full-thickness rectal prolapse is more unusual; in this case, the mucosal folds are circumferential.

The prolapse may reduce spontaneously or may require manual reduction. When the prolapse has been present for some hours, the mucosa becomes edematous, smooth, and featureless: only the size and palpable thickness of the wall will differentiate the two types. Rarely, children with rectal prolapse present to the emergency room with mucosal bleeding.

39.2 Initial Management

The steps of the initial management of rectal prolapse are nonoperative and aim to facilitate normal stooling without excessive straining:

1. prescribing a laxative (e.g., lactulose)
2. encouraging a high-fiber diet
3. encouraging regular, prompt defecation from a sitting, not squatting, position

The initial management is designed to break the cycle that led to the a mechanical prolapse in the first place. Laxatives used to soften the stool and hence eliminate the urge to defecate may allow the pelvic floor muscles to recover and retain their tone. It is also advisable to prevent prolonged defecation periods. Over 90% of children who experience rectal prolapse during the first 3 years of life respond to conservative treatment.

In the past, external support to the perianal region during defecation was recommended, but there is no evidence that it

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prevents recurrence, and the authors do not support this practice.

39.3 Surgical Management

Surgical management is required in refractory cases. Among the surgical procedures described are Thiersch's operation, posterior sagittal rectopexy (PSRP), and more recently, laparoscopic rectopexy (LSRP) with or without the use of a synthetic mesh material.

Under general anesthesia, an initial rectal examination and proctoscopy are performed to exclude rectal polyps. If the rectum is loaded with hard stool, this should be evacuated. The specific goals of surgical management of full-thickness rectal prolapse are to eradicate the external prolapse of the rectum, improve continence, improve bowel function, and reduce the incidence of recurrence.

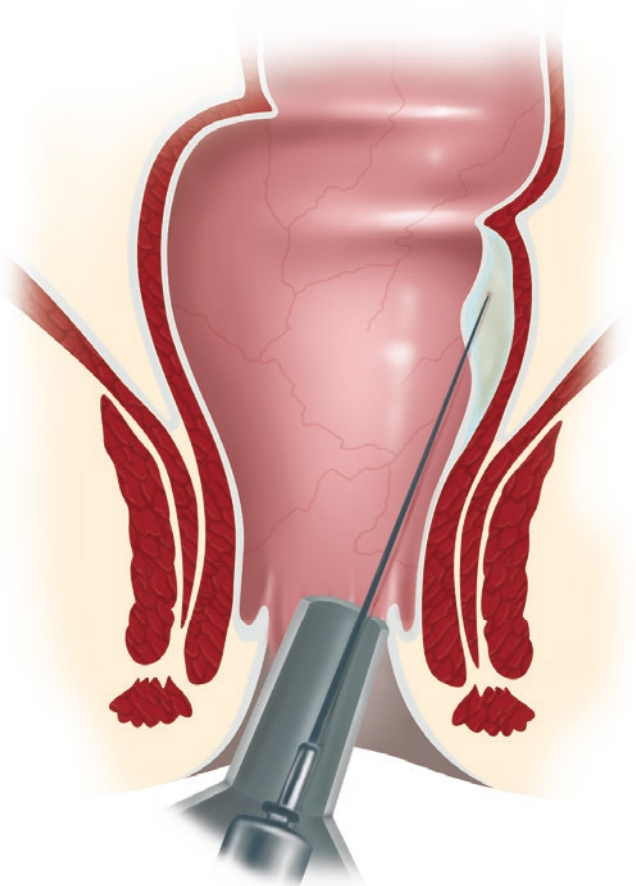

Fig. 39.1 Injection of mucosal prolapse

39.3.1 Thiersch's Operation

A proctoscope of appropriate size is gently introduced into the lower anorectal region (Fig. 39.1). A long, 23-gauge needle is placed under vision into the submucosal plane of the lower rectum approximately 4 cm from the anal verge. Then 1–2 mL of 5% phenol, 30% saline solution, or 5% ethanolamine oleate is injected into the submucosal space in each of the four quadrants. A bulge at the injection site or blanching of the mucosa will indicate that sufficient sclerosant has been injected.

The child is placed in the lithotomy position and the perianal area is prepared and draped (Fig. 39.2). Two small incisions are made 2 cm from the anal verge at 12 o'clock and 6 o'clock. A length of absorbable suture material (e.g., 0 caliber polydioxanone [PDS]) is threaded from the posterior incision to the anterior incision around the anus, just deep to the external sphincter muscle. The suture is continued from anterior to posterior so that eventually a ring is placed around the anus.

With an assistant's finger or a Hegar's dilator held inside the anal canal, the suture is pulled and tied inside the posterior incision. Absorbable sutures are used to close the two incisions. Thiersch's procedure acts by narrowing the anal orifice and thereby mechanically preventing the prolapse.

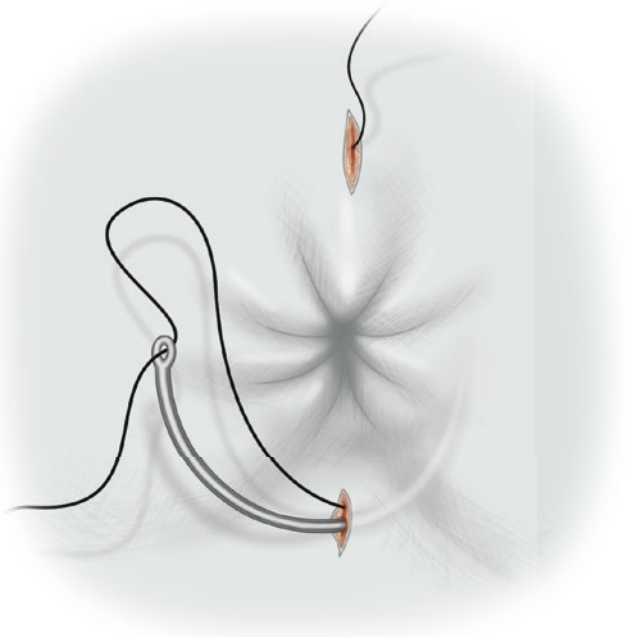

Fig. 39.2 Thiersch's operation (modified)

39.3.2 Eckehorn Procedure

The patient is placed in prone jackknife position on the table. The anus is spread wide open by the assistant with two long Langenbecks holders. The largest available 180° curved needle with one end of 0-PDS thread is introduced as far as possible into the rectum and pulled out through the seromuscular layers close to one side of the sacrum. The PDS is held there by the assistant. The needle is pulled back and the other end of the PDS suture is threaded into the needle, which is then again introduced as far as possible into the rectum and pulled out at the other side of the sacrum. The two ends of the PDS thread are now tied firmly together over a swab. The dorsal rectum wall is fixed to the sacrum and the local inflammation results in a permanent fixation of the rectum to the sacrum. Complications such as abscess formation are unusual. The PDS is removed after 8–10 days. Clinical experience shows satisfying long-term results in most cases, and a low rate of recurrence.

39.3.3 Posterior Sagittal Rectopexy

The patient is placed in the jackknife position (Fig. 39.3). A midline skin incision is made from the coccyx and extended halfway to the anus. This incision is deepened towards the coccyx. If the distance from the coccyx to the anus is short and the operating field is limited, the coccyx may be excised. The parasagittal fibers and levator muscle are divided exactly in the midline using cautery, taking care not to incise the muscle complex. The rectum is then dissected free for two thirds of the circumference and up to 10–15 cm vertically. Three or four permanent seromuscular sutures (3/0 or 4/0 polypropylene [Prolene]) are then placed in a longitudinal, U-shaped, mattress pattern. When these sutures are pulled together, the redundant rectum is drawn together. A further set of sutures may then be passed through the last segment of the sacrum and tied on its surface. The muscle layers are then approximated and the wound is closed.

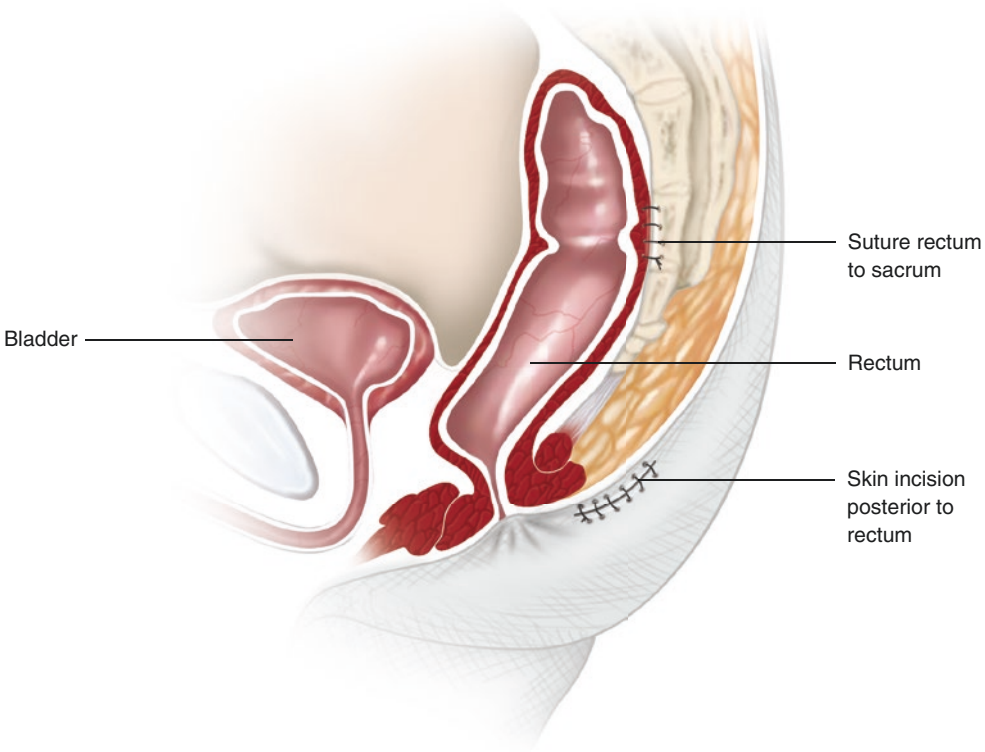

Fig. 39.3 Posterior sagittal rectopexy (PSRP)

39.3.4 Transanal Mucosal Sleeve Resection

The patient is placed prone in the jackknife position (Fig. 39.4). The prolapse is gently drawn out and four quadrant traction sutures are placed through the submucosa at the apex. Epinephrine solution (1:200,000) may be injected to separate the mucosal and submucosal layers from the muscular layers, defining the plane of dissection (**a**). A circumferential incision is made through the mucosal and submucosal layers approximately 1 cm proximal to the pectinate line, and blunt dissection is used to strip this layer from the underlying muscle (**b**). The denuded muscle layer is gradually reduced into the pelvis while the traction sutures are used to pull the mucosal sleeve in the opposite direction. When the submucosal layer has been separated from the entire length of the prolapse, it is divided longitudinally into two halves (**c**). As the sleeve is incised circumferentially, single absorbable sutures are placed to approximate the edges of the proximal and distal mucosal cuffs. Traction is maintained on the sleeve until the resection is complete; the sutures are then cut and the anastomosis retracts into the pelvis (**d**).

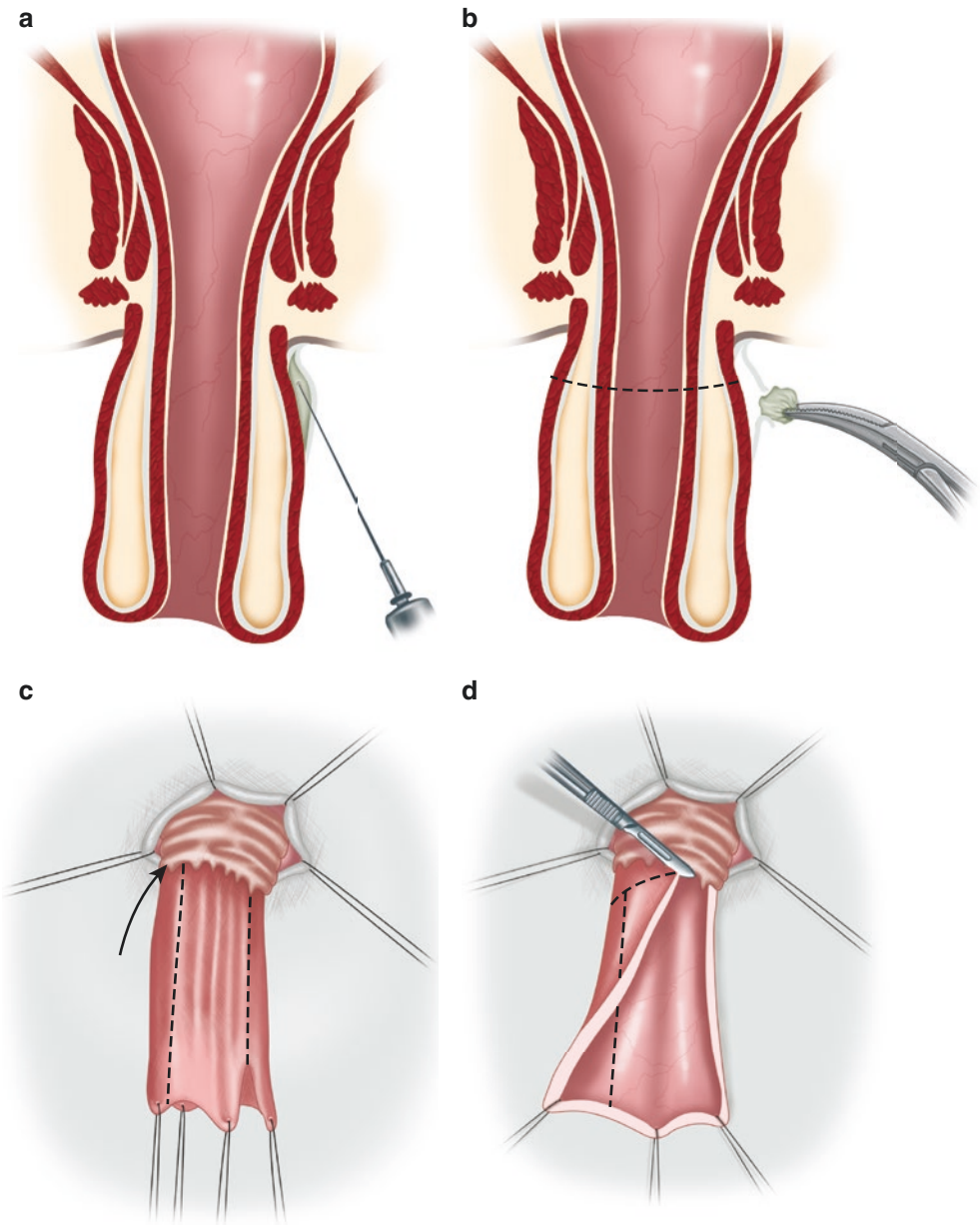

Fig. 39.4 Transanal mucosal sleeve resection (**a**–**d**)

39.3.5 Laparoscopic Abdominal Rectopexy

Many open abdominal procedures previously advocated for severe recurrent rectal prolapse are now performed using laparoscopic techniques such as the suture rectopexy and the modified sling rectopexy using a polypropylene mesh to secure the rectum; in the opinion of the authors, this is the preferable technique because it gives the best view and surgical control of the procedure. Indeed, a growing body of evidence in the literature supports the concept that laparoscopic surgical techniques can safely provide the benefits of low recurrence rates, improved functional outcome, less postoperative pain, short hospital stay, and early return of bowel function for patients with full-thickness rectal prolapse.

The patient is placed in the lithotomy position. A nasogastric tube and urethral catheter are placed. A pneumoperitoneum is established under direct vision by placing a Hasson cannula. A 5-mm, 30° laparoscope is passed to inspect the intra-abdominal contents. Three further 5-mm trocars are then inserted under direct vision, two in the right paraumbilical region and one in the left paraumbilical region (Fig. 39.5).

Laparoscopic technique includes a retrorectal dissection, starting from the peritoneal reflection on the right side of the rectum and extending from the sacral promontory to the pelvic muscular floor in the rectosacral bloodless plane. Great care is given to the identification of the iliac vessels and ureter. In the absence of pelvic floor laxity, suture rectopexy to the sacral promontory and suture sigmoidopexy to the left lateral peritoneum are done without mesh. In cases with laxity and weakness of the pelvic floor and in patients with neuropathic conditions (spina bifida and meningomyelocele), additional retrorectal mesh is recommended. The mesh is tailored to fit the retrorectal space, where it is fixed to the rectum by two to four nonabsorbable stitches. Then the mesh and the rectum are dragged up and fixed to the sacral promontory, with closure of the peritoneal defect.

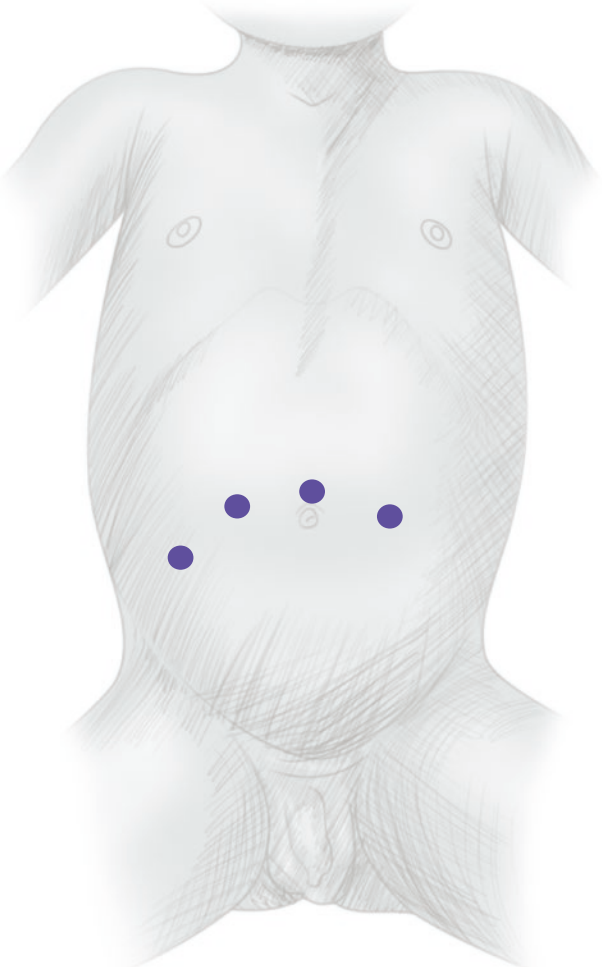

Fig. 39.5 Laparoscopic abdominal rectopexy: trocar positions

39.3.6 Postoperative Care and Complications

As in nonoperative management, regular bowel habits are encouraged. Stool softeners may be advocated for 3–6 months, with advice to avoid sitting on the toilet for long periods.

All of the procedures may be associated with infection and the formation of perianal abscess. Usually these complications resolve fully with conservative treatment, including antibiotic therapy, but occasionally incision and drainage may be required.

Rarely, serious scarring and stricture formation may result, causing deformity of the rectum and leakage of mucus or fistula formation. The Thiersch suture may cause stool retention and fecal impaction if tied too tightly, in which case suture removal should be performed. Disruption of the skin wounds and exposure of the knots may occur if they have not been buried sufficiently.

Most patients presenting with a simple mucosal rectal prolapse respond to conservative, nonoperative management.

Following injection of sclerosant, recurrence occurs in 10–20% of cases. In these cases, injection therapy may be repeated 4–6 weeks later. Many different treatments have been suggested for persistent or severe cases that are resistant to injection therapy. Encircling procedures, abdominal rectopexies, and abdominal perineal bowel resections and have a recurrence risk of approximately 25%. Posterior sagittal and transanal procedures have a higher success rate, between 80% and 100%.

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Bariatric Surgery

40

Brian Dalton and Thomas H. Inge

Pediatric obesity has reached epidemic proportions in the United States over the past three decades. At least 18% of children are estimated to be obese (BMI >95th percentile for age) and 4–7% of children are severely obese (BMI >99th percentile for age). This increased prevalence of childhood obesity has been accompanied by an alarming increase in obesity-related comorbidities at a younger age (e.g., diabetes mellitus, hypertension, dyslipidemia, obstructive sleep apnea, nonalcoholic steatohepatitis [NASH]).

40.1 Selection Criteria

Nonsurgical weight loss plans have proven ineffective in obese and severely obese children and adolescents. A large number of children cannot complete a medical weight loss program, and those that do typically do not lose sufficient weight to reverse comorbid conditions (average 3–5 kg), and weight loss that is achieved is not typically sustainable. Given the unsuccessful nature of medical weight loss programs in adolescents, surgical weight loss procedures increasingly have been used as an effective alternative.

Selection criteria for bariatric surgery in adolescence currently closely follow the adult criteria. All patients with a BMI ≥35 kg/m^2 with significant comorbidities such as moderate to severe obstructive sleep apnea (apnea-hypoapnea index >15), type 2 diabetes mellitus, severe NASH, or pseudotumor cerebri can be considered for bariatric surgery. Also, individuals with a BMI ≥40 kg/m^2 and less severe comorbidities (e.g., milder obstructive sleep apnea, prediabetes, dyslipidemia, hypertension, weight-related quality of life impairment, weight-related renal injury) may be evaluated for bariatric surgery. Prior to any surgical intervention, all patients should be evaluated by a weight loss team (e.g., surgeon, medical specialist, registered dietician, mental health specialist, social worker) to assess the patient's suitability for surgery.

40.2 Bariatric Surgical Procedures

The three most common bariatric procedures performed in adolescents are Roux-en-Y gastric bypass (RYGB), vertical sleeve gastrectomy (VSG), and adjustable gastric banding (AGB). A recent trend that has been observed is the increase in VSG and concomitant decrease in the use of AGB for weight-loss surgery. The vast majority of these procedures are performed laparoscopically in adolescents. These surgical approaches were shown to be as safe in adolescents as in adults, with perioperative complication rates (for major and minor events within 30 days of surgery) of 26% for RYGB and 16% for VSG. AGB is not approved by the U.S. Food and Drug Administration (FDA) for patients under the age of 18. Outcome data collected in FDA-approved studies of the device in adolescents have shown mixed results, with varying complications and weight-loss performance. Data from a more definitive and larger study sponsored by the manufacturer has not been published.

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40.2.1 Roux-en-Y Gastric Bypass

The patient is placed in supine position on the operating table. After induction of general anesthesia, access to the peritoneal cavity is obtained using a bladeless, 12-mm trocar placed supraumbilically. This trocar is carefully inserted through each body wall layer identified using simultaneous guidance with the 0° laparoscope that is positioned in the center of this viewing trocar. Two other 12-mm trocars and two 5-mm trocars (one subxiphoid for liver retraction) are placed (Fig. 40.1).

The omentum is retracted cephalad and the transverse mesocolon is elevated anteriorly. The jejunum is divided approximately 50 cm from the ligament of Treitz. Most surgeons typically create a 100- to 150-cm Roux limb for most patients. The biliopancreatic limb is approximated to the distal end of the Roux limb with a single stitch at the antimesenteric border. The Harmonic scalpel is used to make opposing enterotomies, and a side-to-side jejunostomy is created with an endoscopic GIA stapler. The enterotomy used for the stapler is then closed by running suture or another stapler application (Fig. 40.2).

Antecolic and retrocolic Roux limb construction each has its theoretical risks and benefits. The antecolic is technically more straightforward and avoids the risk of bleeding from the vasculature in the often-thick transverse colon mesentery. This method is preferred in many high-volume bariatric centers. In the antecolic technique, the omentum is replaced inferior, and when it is bulky, it is divided from the free edge to the transverse colon. This technique minimizes the mass of tissue the Roux limb must traverse and therefore reduces the tension on the gastrojejunostomy when the patient stands upright. The antecolic technique does result in a rather large Petersen's defect, however, with potential for future internal herniation. Some surgeons routinely close the large defect and some do not. The exact risk of internal herniation is not known, nor is it completely eliminated by closure, because postoperative loss of fat mass (particularly abdominal fat mass) may well compromise the closure that was performed.

Creation of the lesser curve gastric pouch is then started by retraction of the left lateral section of the liver anteriorly to expose the gastroesophageal junction. Using Harmonic and blunt dissection anteriorly, a plane is opened between the stomach wall and the diaphragm at the angle of His. A small gastrotomy is then created in the anterior body of the stomach for introduction of the EEA stapler anvil. The anvil with spike attached is passed through the previously widened left upper quadrant (LUQ) incision and guided into the stomach through the gastrotomy, with the assistance of a suture and articulating grasper. The point of the articulating grasper, now within the lumen of the stomach (4–5 cm from the gastroesophageal junction on the lesser curvature), is advanced cephalad then articulated to point anteriorly. With assistance of the Harmonic scalpel, the point of this instrument penetrates the gastric wall from inside to out. The anvil spike is then pulled from the lumen of the stomach, leaving the wider anvil inside, which will form the gastrojejunal anastomosis. The gastrotomy on the greater curve is then typically closed using a linear stapler (Fig. 40.3).

The gastric pouch dissection next begins 6–10 cm inferior to the gastroesophageal (GE) junction on the lesser curve, and the stomach is thus partitioned with the previously placed anvil left within the newly formed pouch. Using ultrasonic energy, the dissection is carried posterior, staying close to the gastric wall until the lesser sac is encountered posterior to the stomach. The lesser sac is then entered from the greater curve attachments, and posterior attachments of the stomach are transected to achieve continuity with the lesser curve dissection. An Endo GIA stapler with 1.5-mm (closed staple height) staples is used to create the pouch. The first firing is transverse across the lesser curve, flush with the anvil of the EEA stapler. Once this is completed, any remaining lesser sac attachments are transected. The vertical portion of the staple line is advanced with multiple firings of the Endo GIA stapler toward the angle of His. Once the angle of His is reached, the pouch construction is complete (Fig. 40.4).

Laparoscopic gastrojejunal anastomosis can be performed by numerous techniques. Our preferred method is an end-to-side EEA-stapled anastomosis. The previously created Roux limb is brought to the gastric pouch in an antecolic fashion. The antimesenteric border of the Roux limb is then opened for 4–6 cm. The 25-mm EEA stapler is inserted through the LUQ port and into the Roux limb enterotomy while providing counter-traction to ensure that the stapler head is entirely in the lumen. The spike is extended through the small bowel wall. The anvil grasper is used to mate the anvil spike to the stapler spike. After ensuring that no redundant tissue is caught between stapler and anvil, the stapler is fired and then removed from the abdomen and replaced by a 15-mm trocar (Fig. 40.5). The specimen is inspected to ensure that two complete tissue "donuts" are present, indicating an intact anastomosis. The enterotomy is then resected using an Endo GIA stapler. The integrity of the anastomosis can be tested under saline with insufflation of air via an orogastric tube.

A drain is left near the gastrojejunal anastomosis, exiting through the right upper quadrant (RUQ) incision. The 15-mm trocar site is closed percutaneously with laparoscopic guidance, utilizing the Carter-Thomason (Cooper Surgical Inc.; Trumbull, CT) closure system. Skin incisions are closed in the standard fashion.

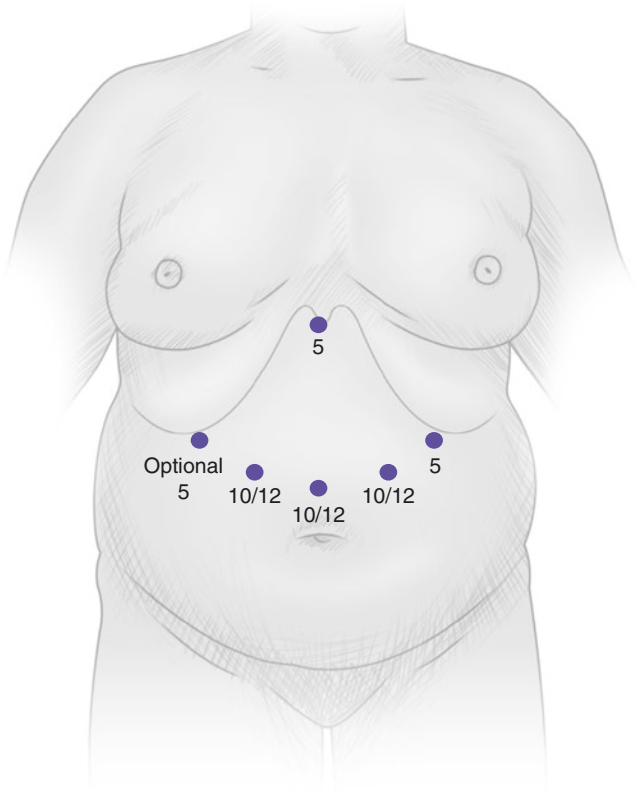

Fig. 40.1 Trocar placement for Roux-en-Y gastric bypass (RYGB)

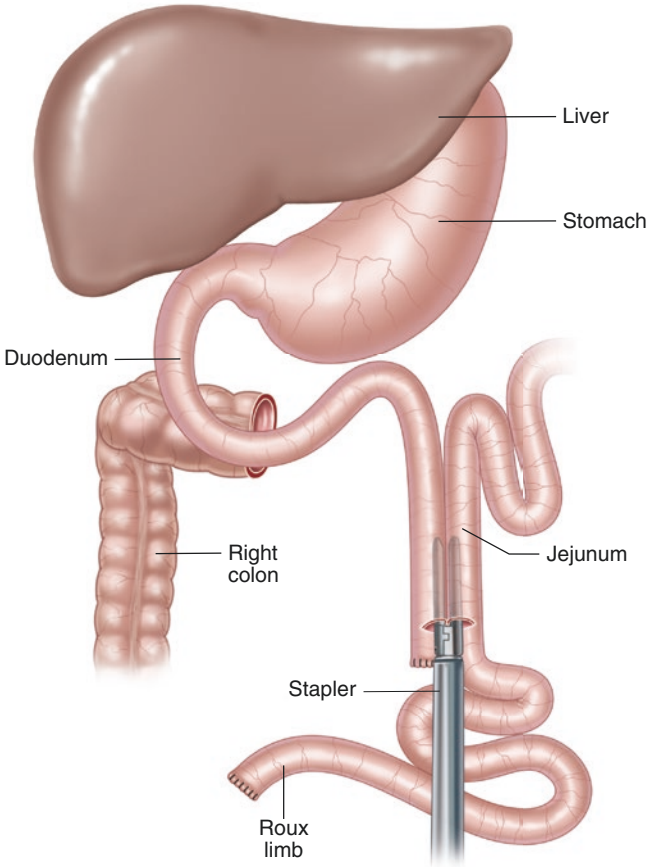

Fig. 40.2 Creation of a side-to-side jejunostomy for RYGB

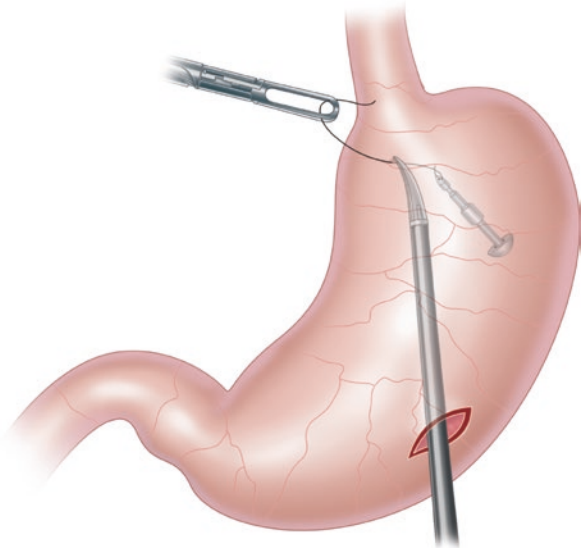

Fig. 40.3 The gastrotomy on the greater curve is then typically closed using a linear stapler

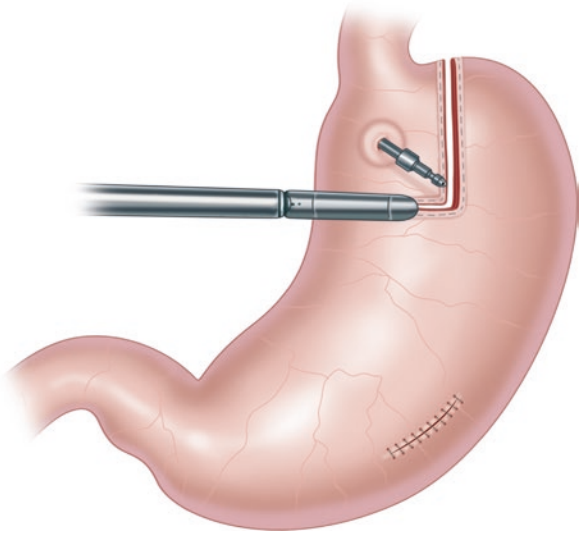

Fig. 40.4 Gastric pouch dissection

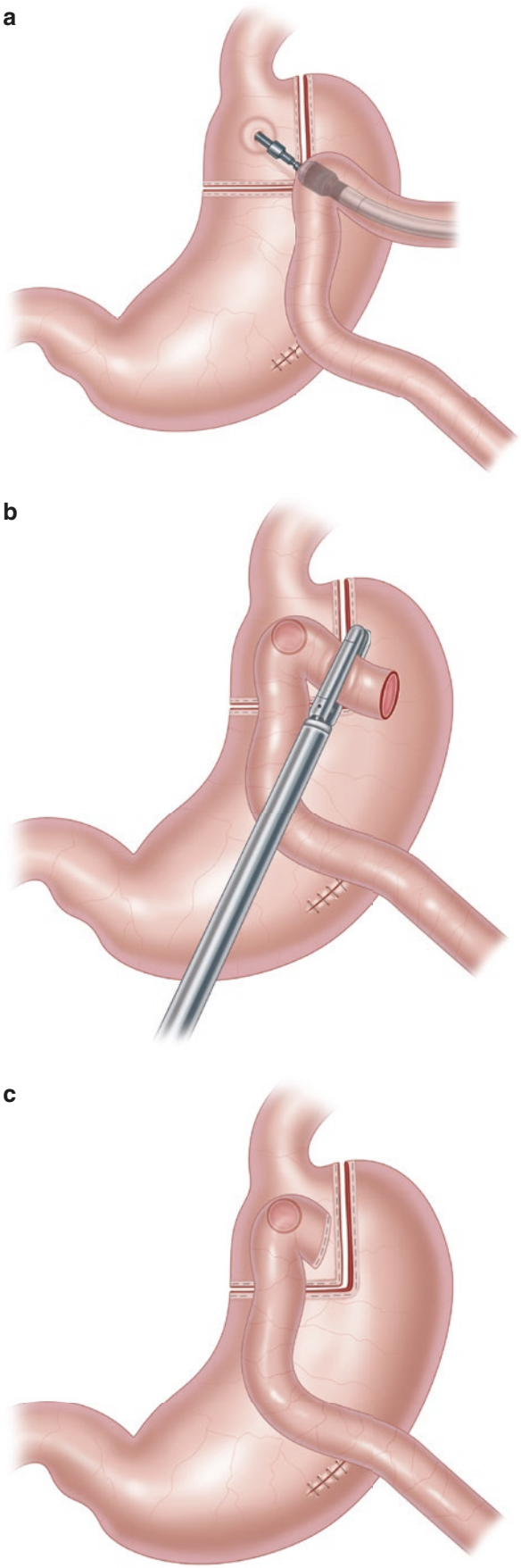

Fig. 40.5 (**a**–**c**) Creation of the gastrojejunal anastomosis

40.2.2 Vertical Sleeve Gastrectomy

For VSG, the patient position and port placement are similar to the RYGB procedure, with three 5-mm ports and two 12-mm ports used (Fig. 40.6). A liver retractor is placed and a 34Fr orogastric tube is inserted by anesthesia to assist in the dissection.

The greater curve dissection is started 6 cm proximal to the duodenum, measured by umbilical tape. The greater omentum is dissected away from the stomach starting at this point, and this dissection extends to the left crus of the diaphragm. All attachments to the greater curve and fundus must be transected (Fig. 40.7).

Gastric resection is initiated 6 cm from the pylorus, using a 60-mm Endo GIA stapler with staple line reinforcement (Gore-Tex material is typically used). The first firing is across the antrum abutting the orogastric tube previously inserted (Fig. 40.8a). Subsequent firings are aimed toward the left crus next to a 34F orogastric tube (Fig. 40.8b). When near the GE junction, the stapler is pointed slightly lateral to avoid esophageal injury. The result is small triangular portion lateral to the esophagus. Many experts advise that this superior-most portion of the staple line should be reinforced with a figure-of-eight suture to prevent leak. In addition, some advise that this end of the staple line should be suspended from the left crus to recreate the angle of His. The resected portion of the stomach can usually be removed through a 12-mm port site with a Kelly clamp after widening the fascial defect.

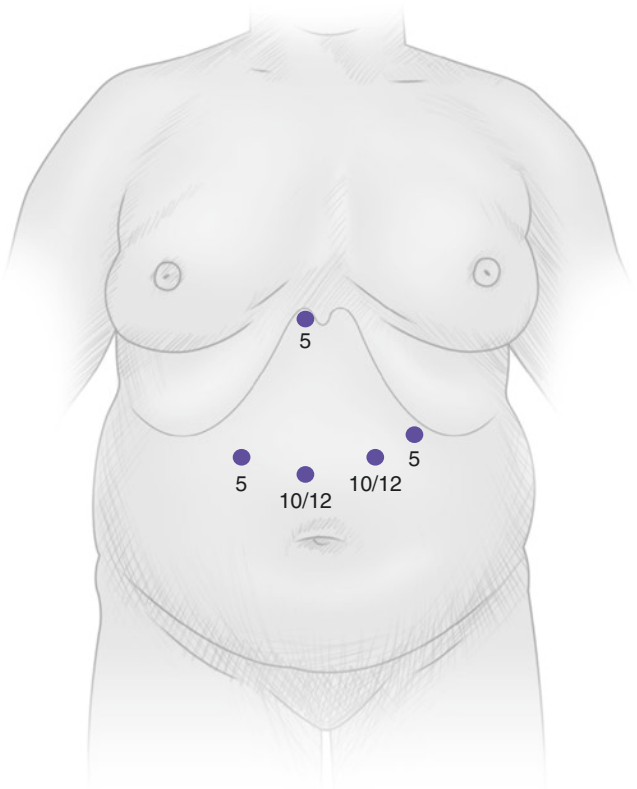

Fig. 40.6 Trocar placement for vertical sleeve gastrectomy (VSG)

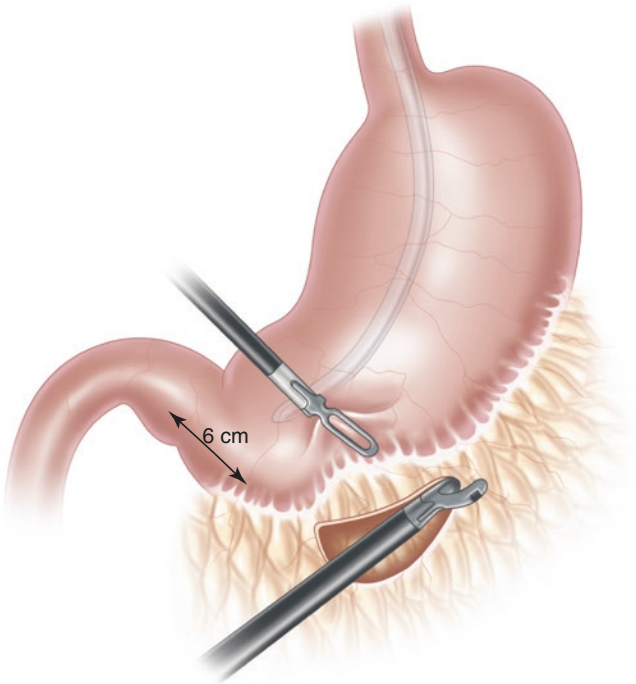

Fig. 40.7 Greater curve dissection for VSG

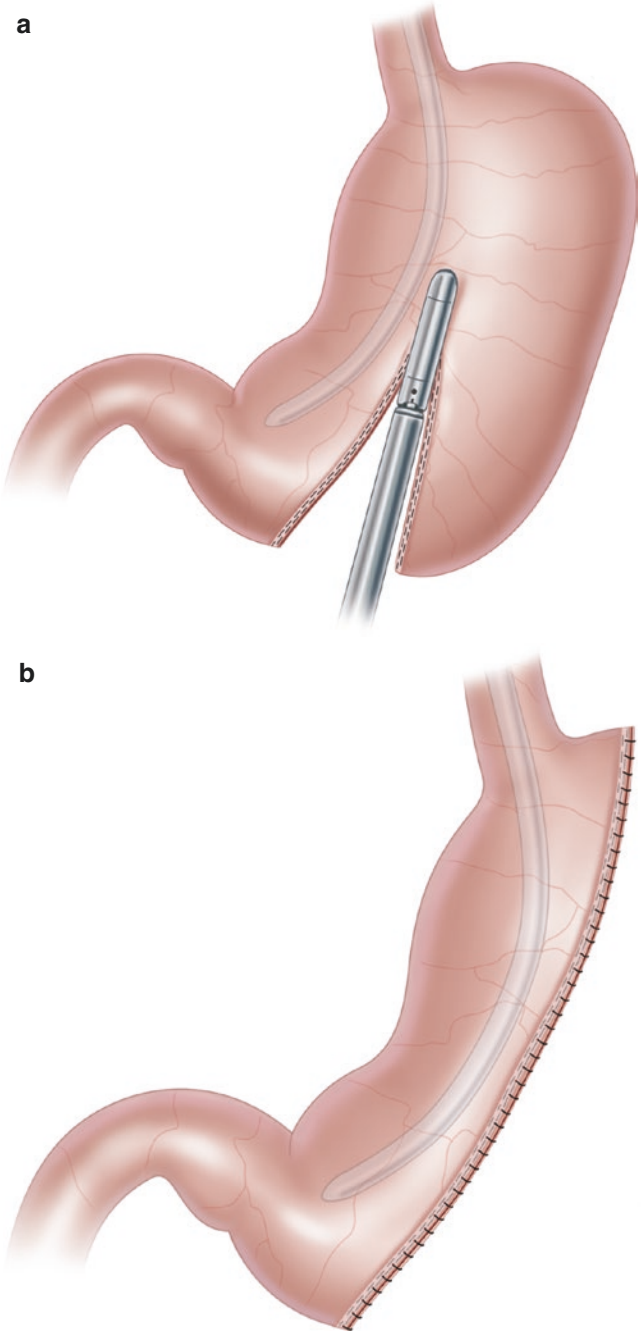

Fig. 40.8 Gastric resection in VSG

40.2.3 Adjustable Gastric Band

As of 2014, no weight loss devices, including the AGB, have been approved by the U.S. FDA for use in patients less than 18 years of age, but some surgeons have used the device in pediatric age groups, and short-term (1–2 year) outcome data have been published, which show benefits and risks of the device. To place the band, one 15-mm port, one 12-mm port and two 5-mm ports are used, in a similar arrangement to RYGB and VSG. A liver retractor is again used to retract the left lateral section anteriorly. An energy device is used to dissect the angle of His to expose the left crus. The pars flaccida is then dissected using an articulating 5-mm instrument to expose the right crus, creating a connecting plane to the left-sided dissection near the GE junction. The hiatus should be inspected for hiatal hernia before placement of the band. Transection of the short gastric vessels is usually unnecessary and may lead to band migration. The suture end of the lock-end flap is then attached to the blunt dissector and the band is passed from left to right in the retrogastric plane created during dissection (Fig. 40.9). The band is then locked in place and rotated so the band lies on the lesser curve. The body of the stomach is then plicated over the band to prevent band slippage (Fig. 40.10).

The band tubing is then brought out through a separate incision closer to the xiphoid to aid in visualization of the fascia. The tubing is then attached to the injector port and the port is fixed to the fascia with suture. The band can be primed with a small amount of fluid or left deflated per manufacturer's recommendations at this time. The 15-mm port is again closed with laparoscopic assistance. Drain placement is typically not necessary.

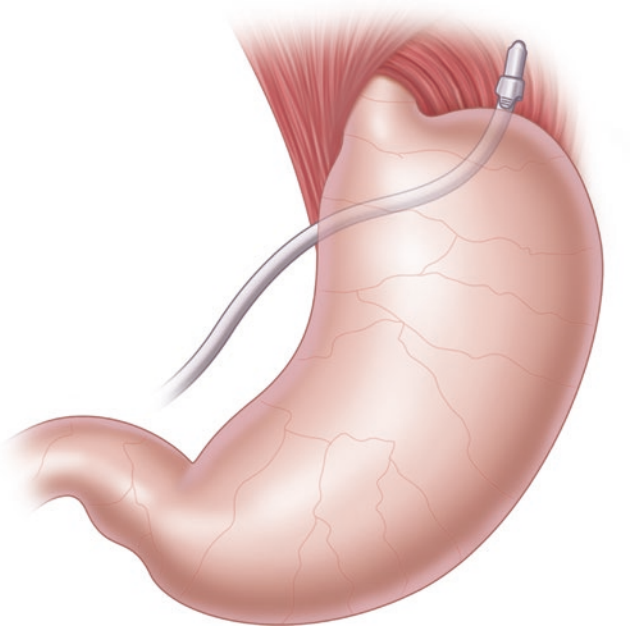

Fig. 40.9 Passing the adjustable gastric band (AGB) in the retrogastric plane

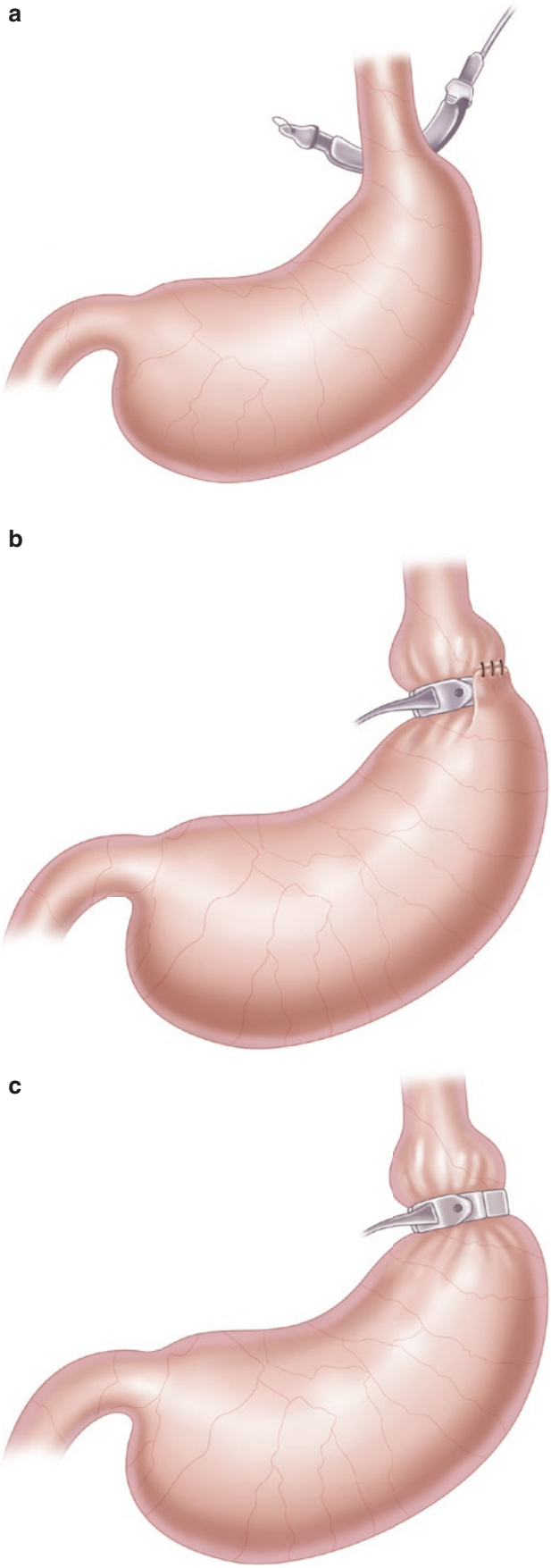

Fig. 40.10 Putting the AGB in place, with plication of the body of the stomach over the band

40.3 Postoperative Care

Postoperatively, all patients can usually be monitored in a non-intensive care unit setting. Some surgeons obtain an upper gastrointestinal (UGI) contrast study the morning of postoperative day (POD) 1. Others selectively obtain a study only if there are signs or symptoms of a leak or obstruction. Clear liquid is initiated early following operation. Normally RYGB and VSG patients stay in the hospital for two or three nights after surgery and are sent home on a clear liquid diet, which is then advanced to a high-protein liquid diet starting after discharge and continuing for 1 month postoperatively. AGB patients are normally started on a liquid diet on POD 0 and sent home on POD 1. AGB patients can be advanced to a small-portion solid diet over 2–4 weeks. These patients consume small portions and delay drinking liquids for 30 min after solid intake.

Follow-up after RYGB and VSG involves appointments at 2 and 6 weeks; 3, 6, 9, 12, 18, and 24 months; and then yearly. Monitoring labs, including serum chemistry, CBC, prothrombin time, and vitamin B1 and B12 levels are obtained at annual intervals. Vitamin supplementation is usually needed because of the restrictive diet (low-calorie, low-carbohydrate) and micronutrient malabsorption with RYGB and VSG. Vitamin and mineral supplementation normally consists of a pediatric multivitamin, vitamin B1, vitamin B12, calcium, and iron supplementation for menstruating females. RYGB patients should avoid nonsteroidal anti-inflammatory medicines because of the increased risk of ulcer formation at the gastrojejunal anastomosis. Birth control is strongly recommended during rapid weight loss (until 18–24 months postoperatively) for women of childbearing age.

Five lifestyle rules are emphasized to each patient:

- Eat protein first
- Drink 1.9–2.8 L of water or other sugar-free liquids per day
- Do not snack between meals
- Exercise at least 30 min daily
- Take vitamin and mineral supplements

40.4 Outcomes

Weight loss with all three surgical procedures has been shown to be significant in teenagers. A recent study from Sweden showed an average BMI reduction from 45 to 30 kg/m^2 over a 2-year period for adolescents that had undergone RYGB. A significant reduction in comorbidities was also reported. At Cincinnati Children's Hospital Medical Center, a body composition analysis showed a loss of both lean body mass and fat mass of 20% at 3 months following RYGB. From 3 to 12 months postoperatively, however, fat mass was reduced by a further 40% while lean body mass remained stable. VSG resulted in a very similar 30–35% reduction in BMI at 1 year in adolescents. Likewise, AGB has been shown to reduce excess body weight and BMI significantly (about 28% reduction in BMI, compared with 3–5% reduction with lifestyle modifications), but the weight loss rate is slower than with VSG and RYGB. AGB may also require revisional procedures more often. The National Institutes of Health sponsored the Teen Longitudinal Assessment of Bariatric Surgery (Teen-LABS) Study, which has reported the perioperative (30 day) safety outcomes of all three surgical procedures, but found that RYGB and VSG were performed much more commonly in the adolescent population. Three-year outcomes from this prospective study are now also showing significant and durable weight loss and comorbidity resolution for VSG and RYGB cohorts. Further publications from this study group will help to elucidate long-term outcomes of adolescent patients who have undergone weight-loss surgery.

Suggested Reading

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Biliary Atresia

41

Masaki Nio, Hideyuki Sasaki, Hiromu Tanaka, and Ryoji Ohi

Biliary atresia remains one of the most intractable gastrointestinal surgical diseases in infancy. The introduction of liver transplantation (LTx) has revolutionized the protocols for the treatment of this condition, hepatic portoenterostomy (the Kasai procedure) is still the first-line surgical treatment.

The main clinical manifestations of biliary atresia are persistent jaundice, clay-colored feces, and hepatomegaly. Although symptoms do not vary greatly, anatomic findings of the biliary tract vary from case to case. Only about 10–15% of patients with biliary atresia have an extrahepatic bile duct large enough to perform a mucosa-to-mucosa anastomosis to the intestine. These correctable cases can undergo hepaticoenterostomy. The remaining 85–90% of patients do not have a viable bile duct amenable to a conventional anastomosis. For these noncorrectable-type cases, hepatic portoenterostomy should be performed.

41.1 Surgical Treatment: Hepatic Portoenterostomy

Hepatic portoenterostomy was first devised in 1957 as corrective surgery for patients with the noncorrectable type of biliary atresia. The basis of this procedure is that intrahepatic bile ducts are patent in early infancy, and minute intrahepatic bile ducts are present in the cone-shaped fibrous tissue, replacing extrahepatic biliary radicles. In hepatic portojejunostomy, extrahepatic bile ducts, including fibrous remnants at the porta hepatis, are completely removed, and bile drainage is established by the anastomosis of an intestinal conduit to the transected surface at the porta hepatis. Microscopic biliary structures at the liver hilum drain bile into the intestinal conduit, and, in time, an autoapproximation occurs between the intestinal and ductal epithelial elements.

Successful hepatic portoenterostomy depends on early diagnosis and surgery (preferably in the first 30 days after birth), adequate operative technique, prevention of postoperative cholangitis, and precise postoperative management.

41.1.1 Preoperative Care

In addition to routine preoperative care for neonates and young infants undergoing surgery, vitamin K (1–2 mg/kg per day) is usually given for several days before surgery. Packed red blood cells are cross-matched, and preoperative broad-spectrum antibiotics are administered.

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https://doi.org/10.1007/978-3-662-56282-6_41

41.1.2 Operative Technique

Surgery is performed under general anesthesia with tracheal intubation. The patient is placed in the supine position. A pillow is placed under the back of the patient for better exposure. Laparotomy is performed through a right subcostal or upper abdominal transverse incision (Fig. 41.1). Some pediatric surgeons recommend mobilizing and completely exposing the liver to enable adequate visualization of the porta hepatis. The authors do not use this technique because of its potential invasiveness; we believe that enough exposure is obtained for Kasai portoenterostomy without this technique.

Liver biopsy is obtained. Then, a catheter is inserted into the small gallbladder to perform cholangiography. After atresia of the extrahepatic bile duct is confirmed by cholangiography, the gallbladder is freed from the liver (Fig. 41.2), and dissection is advanced along with the cystic duct toward the common hepatic duct. Dissection is performed with the help of surgical loupes at 2.5× magnification.

The superficial peritoneum on the hepatoduodenal ligament is opened, and the anatomy of the involved bile duct and hepatic arteries is assessed. Dilated lymphatic channels around the hepatoduodenal ligament should be carefully ligated and divided to avoid excessive lymphatic fluid loss, which occasionally causes intractable postoperative ascites. The common bile duct remnant is carefully dissected, because it is often adherent to the surrounding tissues.

The common bile duct is clamped and divided adjacent to the duodenum. After the common bile duct is severed, it is raised, and the hepatic duct remnant is freed from underlying hepatic arteries and the portal vein (Fig. 41.3). The hepatic duct usually transforms into a cone-shaped fibrous tissue, which is situated cranial to the bifurcation of the portal vein. It continues adjacent to the portal tracts in the liver. Even if a cyst-like structure is present, it should be removed and should not be used for the anastomosis to the intestine.

The separation of fibrous remnants from right and left portal veins is carefully advanced posteriorly. The bifurcation of right and left portal veins must be retracted to obtain proper exposure of the porta hepatis. Several small branches bridging from the portal vein to fibrous remnants are identified and divided between ligatures, facilitating downward displacement of the portal vein (Fig. 41.4). The posterior aspect of fibrous remnants is exposed deep and wide enough behind bilateral main branches of the portal vein. The caudate lobe also should be well visualized behind the bifurcation of the portal vein and caudal to fibrous remnants.

Dissection between the anterior aspect of fibrous remnants and the quadrate lobe of the liver is also sufficiently advanced. The fibrous remnant at the porta hepatis is vertically divided in the middle (Fig. 41.5), taking care not to injure the liver capsule between the quadrate and caudate lobes.

The divided remnants on both sides are independently and completely removed in order without injuring the liver capsule (Fig. 41.6). The transection of fibrous remnants is carefully performed using small, round scissors or a sharp scalpel at the level of the posterior surface of the portal vein. The transected surfaces of the fibrous remnants are located close to the bilateral main branches of the portal vein, and the surrounding liver capsule should be at the same level.

Although some surgeons confirm the presence of microscopically patent ducts at the level of the anastomosis using frozen sections perioperatively, we do not use frozen-section guidance because we always transect the portal bile duct remnants at the same level. Hemorrhage from the superficial incision of the porta hepatis is occasionally considerable. Irrigation with warm saline stops the bleeding, usually within 10 min. Oozing at the porta hepatis is usually controlled by the anastomosis of the posterior row of the portoenterostomy. Ligation or cautery should not be applied because of the possibility of accidental obliteration of small bile ducts that may be opening on the transected surface.

The next step is the construction of a Roux-en-Y loop of the jejunum (Fig. 41.7). A Roux-en-Y anastomosis with the ascending limb is constructed, with an approximate length of 50 cm (or a weight of 10 cm/kg). The end of the gastric limb of the intestine is anastomosed not to the antimesenteric surface but to the lateral surface of the hepatic limb of the intestine. Currently, most surgeons avoid using a cutaneous stoma because of the frequent bleeding that occurs when the patient develops portal hypertension. Also, technical difficulties will arise if subsequent LTx becomes necessary, and no reduction has been observed in the incidence of cholangitis.

A spur valve is created, which may prevent cholangitis. Two centimeters of half of the seromuscular layer facing the gastric limb are removed from the biliary limb proximal to the anastomosis (Fig. 41.8). The gastric and biliary limbs are then coapted over the denuded mucosa with sutures along the edges of the removed seromuscular layer (Fig. 41.9).

After the completion of valve creation, the hepatic limb is brought up retrocolically. The end of the intestine is anastomosed around the transected end of the fibrous remnants at the porta hepatis with full-thickness interrupted sutures using 5/0 monofilament absorbable sutures (Fig. 41.10). Sutures must not be placed on the transected surface of fibrous remnants, in which minute bile ducts are present. After all inter-

rupted sutures of the posterior row are placed in position, they are tied.

The anterior row is sutured in a manner similar to the posterior row of the anastomosis (Figs. 41.11). Again, it should be emphasized that sutures must not be placed on the transected surface of fibrous remnants but rather on the liver tissue around the transected area.

Several interrupted seromuscular sutures are added using 5/0 braided absorbable sutures between the jejunum and the quadrate lobe of the liver and/or the hepatoduodenal ligament, if reinforcement of the anastomosis is believed to be required (Fig. 41.12). Irrigation of the abdominal cavity with saline should be performed sufficiently. The intestine, particularly the jejunum including the biliary limb, is carefully placed in order within the abdominal cavity to prevent ileus. A Penrose drain is placed in the foramen of Winslow. After the viscera of the upper abdomen, including the small intestine and the liver, is covered by Seprafilm®, the abdominal cavity is closed in layers. A central venous catheter is placed for postoperative management, and surgery is concluded.

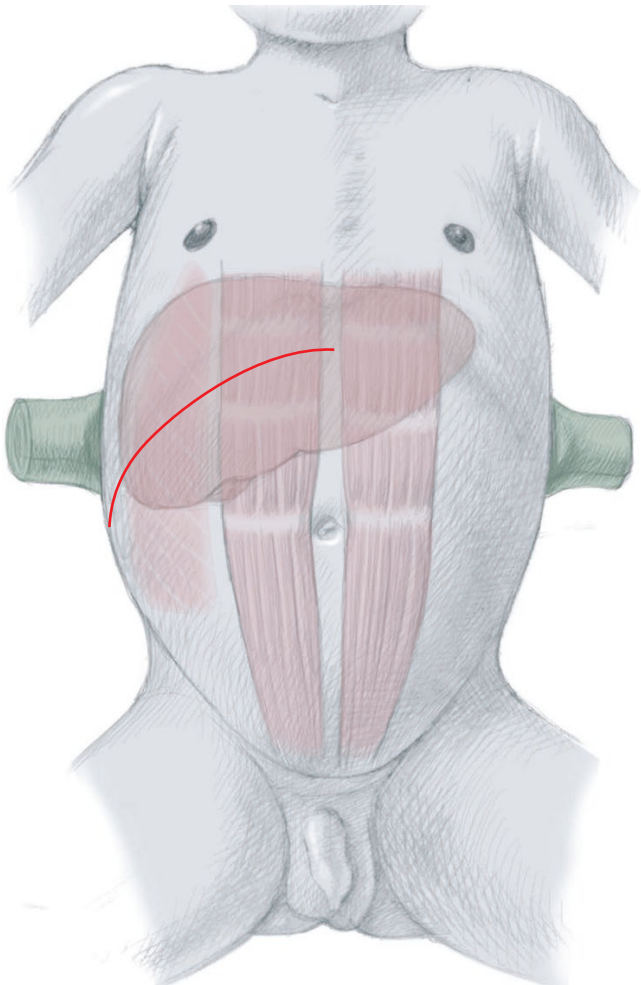

Fig. 41.1 Incision for Kasai portoenterostomy

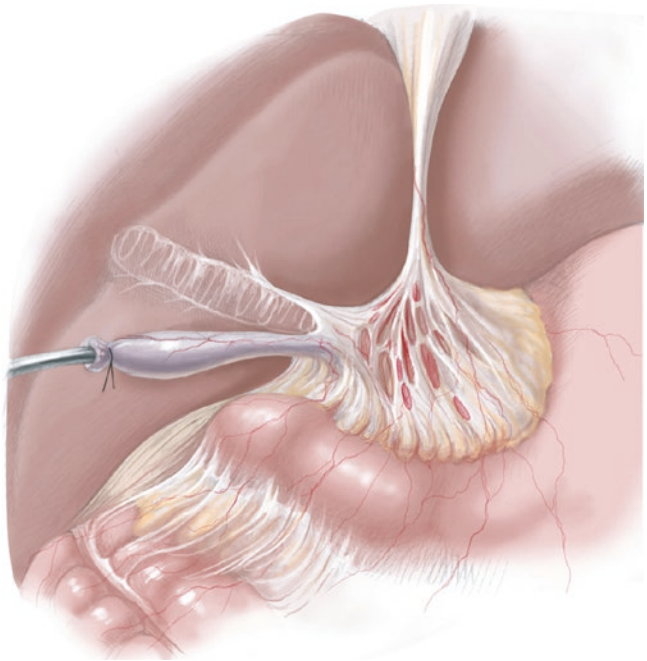

Fig. 41.2 Freeing the gallbladder from the liver

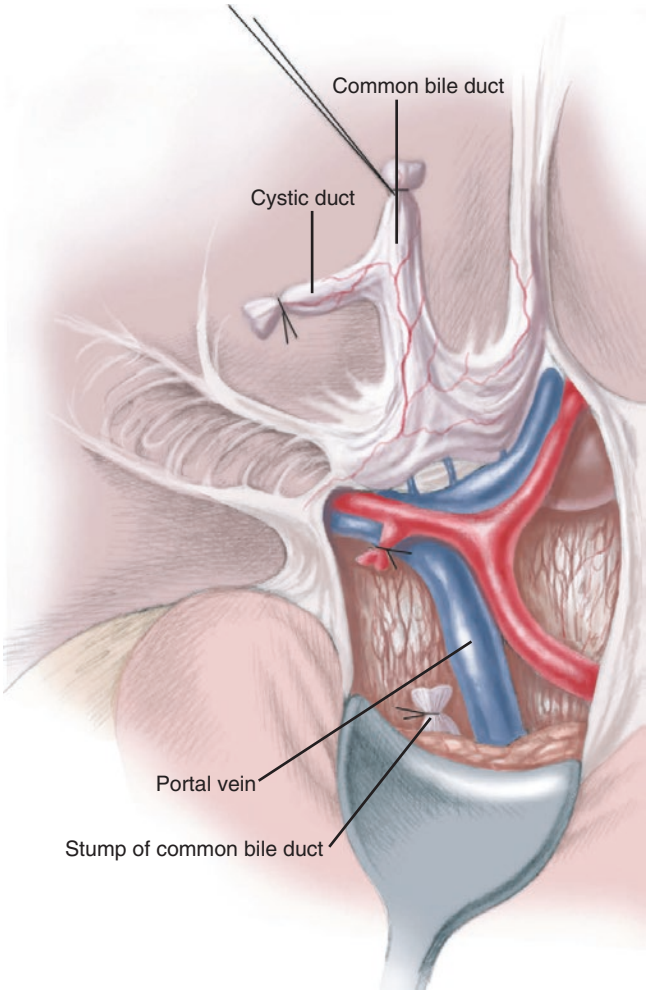

Fig. 41.3 Severed common bile duct

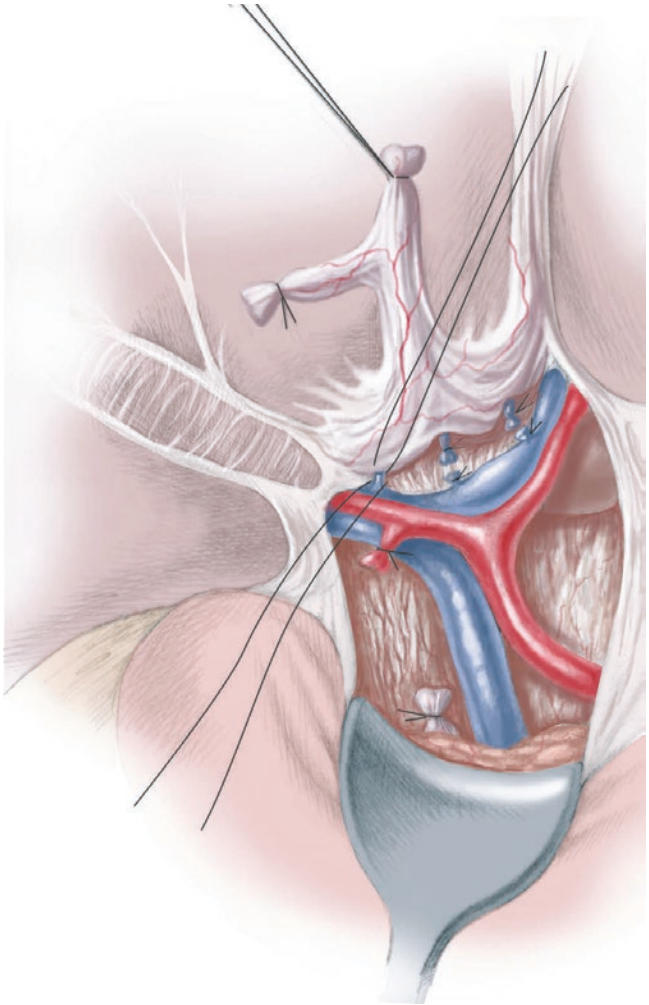

Fig. 41.4 Downward displacement of the portal vein to expose the porta hepatis

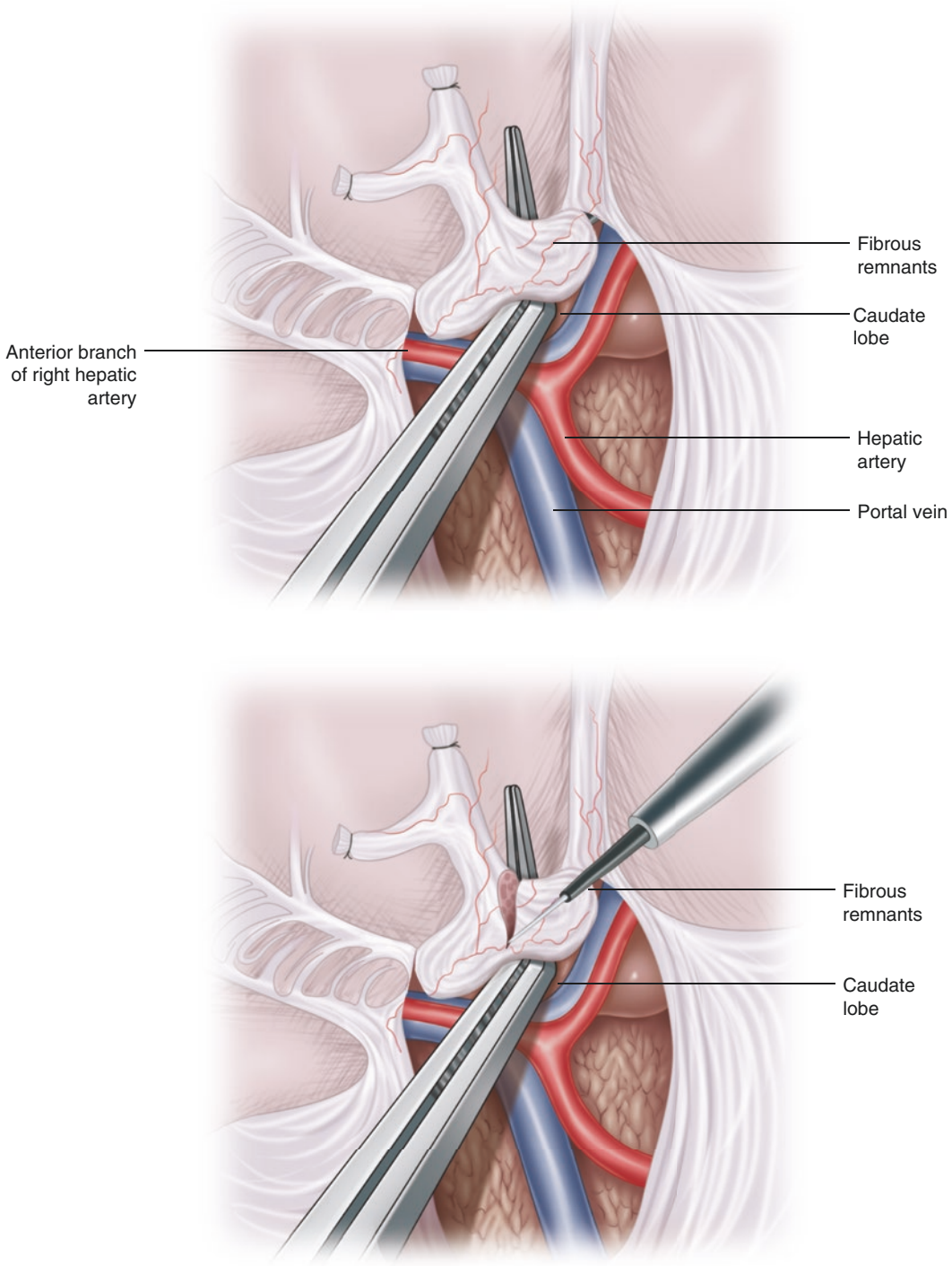

Fig. 41.5 Dissection of the fibrous remnant at the porta hepatis

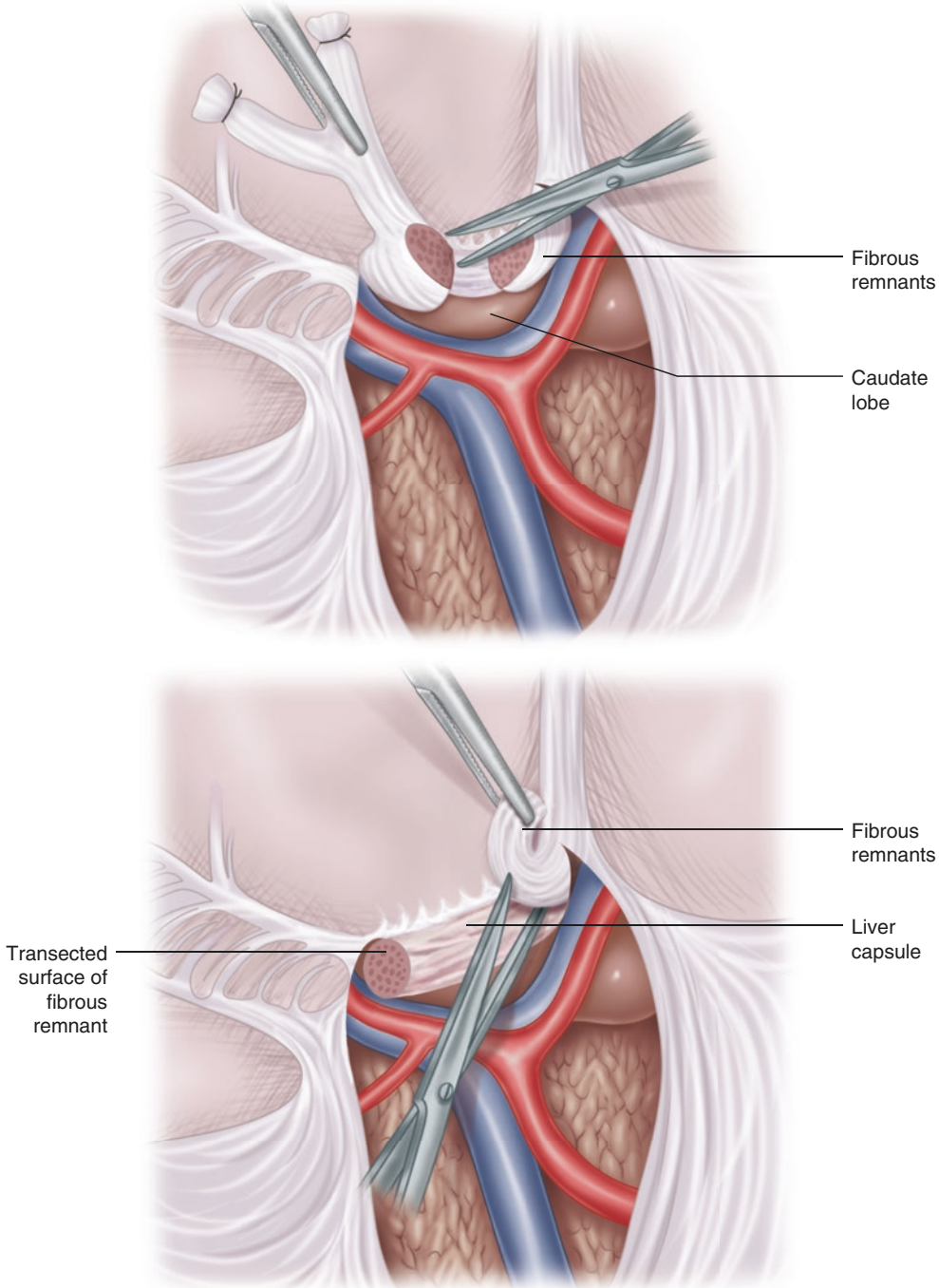

Fig. 41.6 Removal of the fibrous remnants

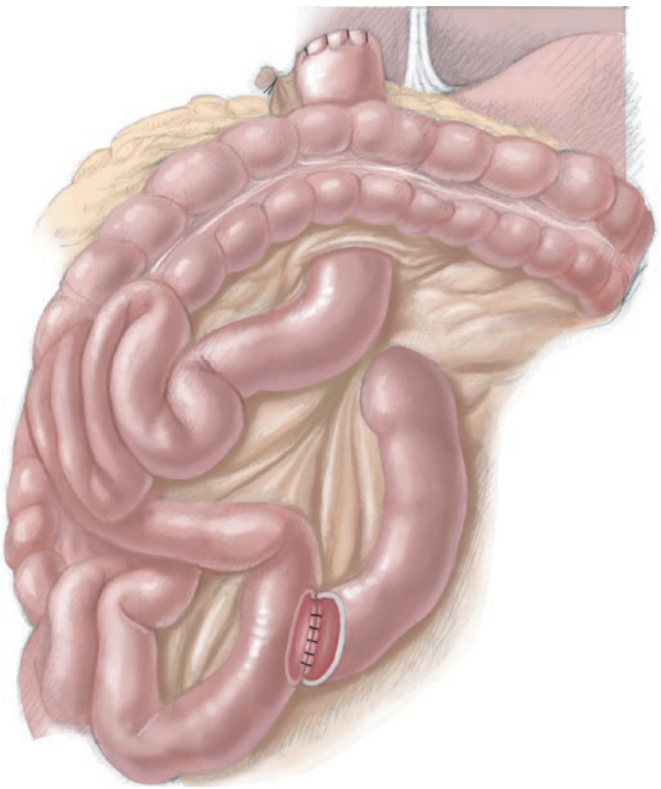

Fig. 41.7 Construction of a Roux-en-Y loop of the jejunum

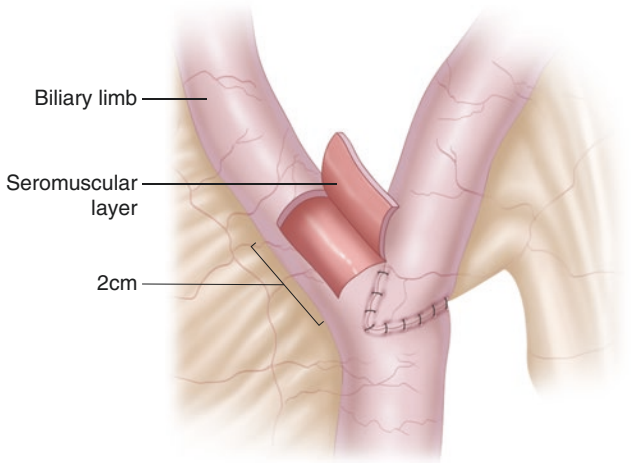

Fig. 41.8 Removal of a portion of seromuscular layer to create a spur valve

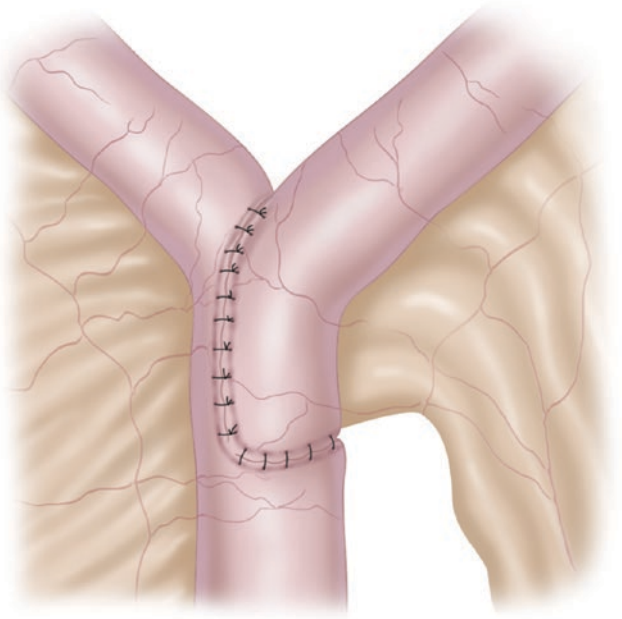

Fig. 41.9 Coaptation of the gastric and biliary limbs

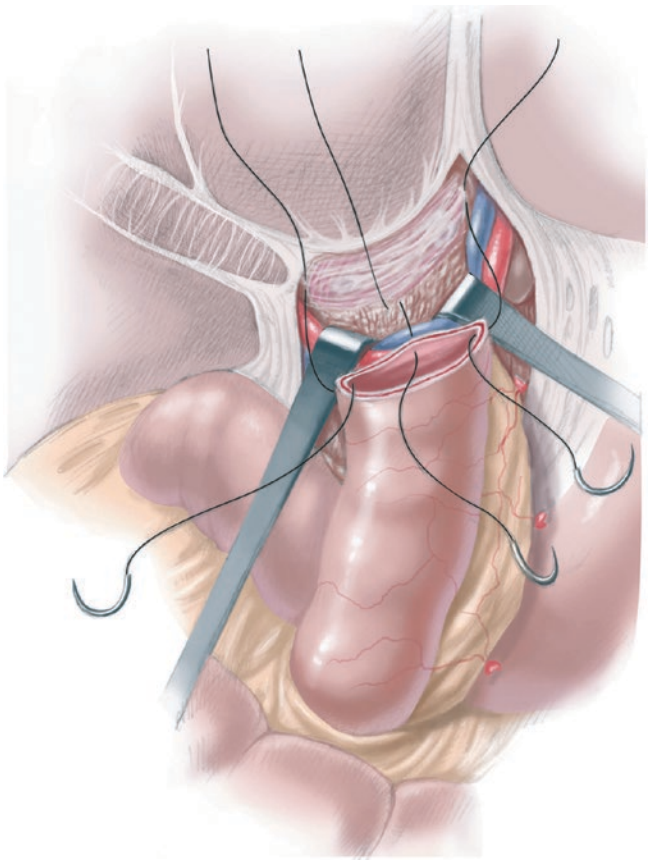

Fig. 41.10 Creation of the anastomosis: suturing of the posterior row

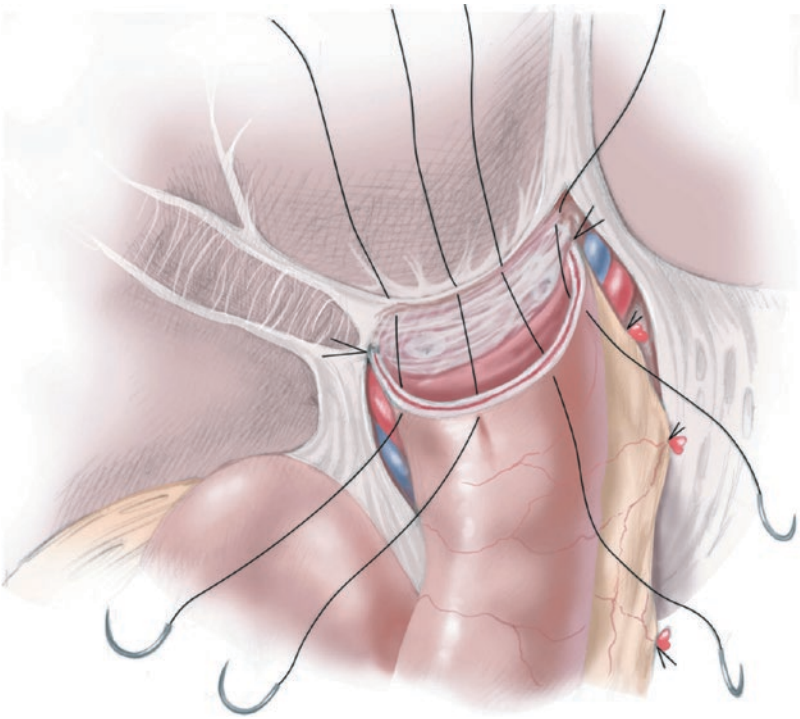

Fig. 41.11 Suturing of the anterior row

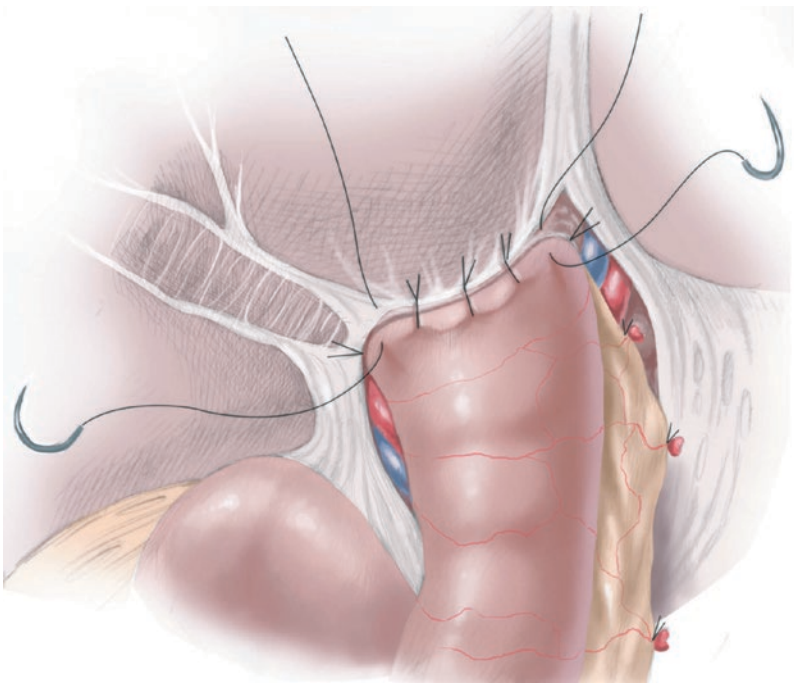

Fig. 41.12 Placement of sutures between the jejunum and the quadrate lobe of the liver

41.1.3 Postoperative Care

Patients are placed on oxygen via nasal cannula, and intravenous fluid is given. Decompression of the gastrointestinal tract should be performed by nasogastric aspiration and enema. Oral feeding is usually initiated on the fifth or sixth postoperative day, when bowel activity resumes. Prevention of postoperative cholangitis is the main goal of postoperative management. For this purpose, we routinely use antibiotics, choleretics, and steroids as postoperative medication. For antibiotics, amikacin sulfate at a dose of 8 mg/kg per day, q8h, is administered for 7 days postoperatively. Then cephalosporin at a dose of 50–80 mg/kg per day is given intravenously for a few months, until the serum bilirubin level is below 2 mg/dL. For choleretics, ursodeoxycholic acid at a dose of 20 mg/kg per day is given orally as soon as oral feeding is initiated. Prednisolone, 4 mg/kg per day, is initiated after the seventh postoperative day. Steroid therapy is continued intravenously for 4 days and is then switched to oral administration of prednisolone at 4 mg/kg bid every other day until the serum bilirubin level is below 2 mg/dL, or for a maximum of 3 months following hepatic portoenterostomy.

41.2 Complications

The most frequent and serious postoperative complication is cholangitis, which can result in fatal septicemia or reobliteration of the hepatic portoenterostomy. Although there are various modified reconstruction procedures to prevent cholangitis, the authors now perform hepatic portojejunostomy using a long Roux-en-Y limb with a length of 50 cm or a weight of 10 cm/kg equipped with a spur valve, as shown in this chapter. The incidence of cholangitis during the first year following Kasai portoenterostomy, however, remains at approximately 30%. Cirrhosis and portal hypertension and/or hypersplenism also have been documented as late complications. It has been shown that both late surgery and the complication of cholangitis aggravate hepatic fibrosis and induce portal hypertension.

41.3 Outcomes

Between 1953 and 2013, a total of 350 patients with biliary atresia underwent surgery in our hospital. Of these, 35 patients underwent hepatic portoenterostomy with a spur valve during the most recent 13 years. Bile drainage postoperatively was achieved in 100% of cases and 85.7% (30 patients) became jaundice-free. The incidence of postoperative cholangitis in the first year following hepatic portoenterostomy was 25.7%. Of the patients treated using this procedure, 26 (74.3%) are still alive with their native liver and without jaundice, eight underwent subsequent LTx, and one is waiting for LTx due to recurrent jaundice. No patient died.

Conclusions

The combination of hepatic portoenterostomy with subsequent LTx is the treatment of choice for patients with biliary atresia. It is important, however, to attempt preservation of the patient's native organ by continuing efforts to achieve the best possible results with hepatic portoenterostomy.

Our current strategy for surgical treatment of patients with this disease includes (1) early diagnosis, including prenatal diagnosis; (2) hepatic portoenterostomy without stoma formation; (3) careful postoperative care, particularly for the prevention of postoperative cholangitis; (4) revision of hepatic portoenterostomy only in selected patients who showed good bile drainage after the first procedure; (5) early LTx in patients with absolutely failed hepatic portoenterostomy; (6) avoidance of laparotomy for the treatment of esophageal varices and hypersplenism (endoscopic injection sclerotherapy and partial splenic artery embolization); and (7) consideration of primary LTx for patients with advanced liver damage at the time of referral.

Suggested Reading

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Choledochal Cyst

42

Atsuyuki Yamataka, Joel Cazares, and Hiroyuki Koga

The most common types of choledochal cyst are cystic or fusiform. Among the rare types are diverticulum of the common bile duct, choledochocele, and Caroli's disease. Choledochal cyst is nearly always associated with pancreatico-biliary malunion (PBMU) and is often associated with dilatation of the intrahepatic bile ducts (IHBD). Before cyst excision, detailed information must be obtained about IHBD abnormalities such as ductal stenosis and dilatation, presence of debris or stones, and intrapancreatic bile duct anomalies such as PBMU or dilatation of the pancreatic duct and presence of debris or protein plugs in the common channel. Today, magnetic resonance cholangio-pancreatography (MRCP) and endoscopic retrograde cholangio-pancreatography (ERCP) allow preoperative visualization of the precise anatomy of the hepato-biliary-pancreatic ductal system in most cases of choledochal cyst. Although MRCP is highly accurate, it may not clearly visualize PBMU if the patient is an infant or a young child, and ERCP is generally contraindicated if pancreatitis is present. If information obtained from MRCP and/or ERCP is insufficient, intraoperative cholangiography is indicated.

42.1 Classification

Choledochal cyst is commonly associated with PBMU involving concurrent abnormalities of the common channel, pancreatic duct, and intrahepatic ducts. Figure 42.1 illustrates the classification of choledochal cysts:

With PBMU:

(a) Cystic dilatation
(b) Fusiform dilatation
(c) Without biliary dilatation

Without PBMU:

(d) Cystic diverticulum of the bile duct
(e) Diverticulum of the distal bile duct (choledochocele)
(f) Intrahepatic bile duct dilatation alone (Caroli's disease)

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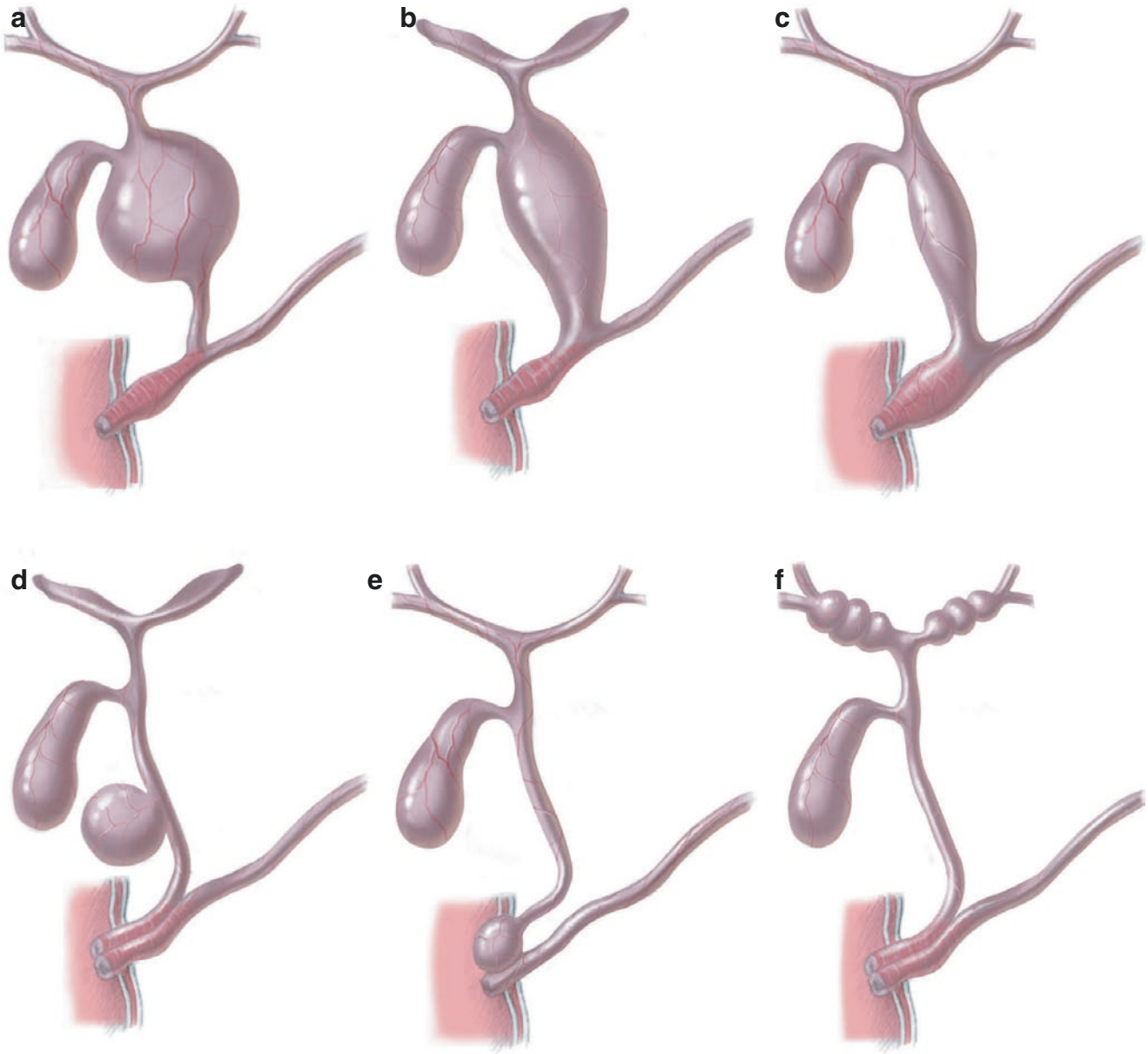

Fig. 42.1 Classification of choledochal cysts

42.2 Surgical Management

The principles of surgical management of choledochal cyst include the following steps:

1. Preoperative imaging (MRCP or ERCP) or intraoperative cholangiography to demonstrate the entire hepato-biliary-pancreatic tract
2. Excision of the extrahepatic bile duct
3. Intraoperative endoscopy (IE)
4. Dissection and excision of the distal choledochus
5. Adequate excision of the common hepatic duct at the correct level
6. Hepaticojejunostomy (HJ) anastomosis (preferably end-to-end)
7. Roux-en-Y biliary reconstruction

The authors now perform IE routinely during cyst excision to determine the level of resection of the distal common bile duct, to irrigate the common channel and wash out any debris or protein plugs present, to examine the ampulla of Vater for stenosis, and to examine the proximal IHBD. IE was introduced in 1986 and was initially performed using a pediatric or neonatal cystoscope.

Surgical problems with cystic choledochal cyst are most often encountered on the proximal side of the pathology, occurring as a result of anatomic variants of the common hepatic duct, uncertainty in relation to the excision level of the common hepatic duct, dilated IHBD, and debris and/or stenosis in the IHBD. In contrast, surgical problems with fusiform choledochal cyst most often arise on the distal side of the malformation and are due to uncertainty in relation to the excision level of the distal choledochus, debris in the common channel, and complicated PBMU.

42.2.1 Open Choledochal Cyst Excision

The patient is placed supine on an operating table with facilities for intraoperative cholangiography. We prefer an extended right subcostal incision. After careful macroscopic examination of the choledochal cyst and liver, the gallbladder is mobilized from its bed together with the choledochal cyst. In comparison with fusiform choledochal cysts, there are usually more adhesions between a cystic choledochal cyst and surrounding vital structures such as the portal vein and hepatic artery, especially in older children. In adolescents and adults, the adhesions are often very dense, and great care is required during cyst excision.

Prior to commencing specific dissection of a cyst, its anterior wall is always opened first by incising the anterior wall of the cyst transversely (Fig. 42.2). Because anatomical variants of the common hepatic duct are often found in cystic choledochal cyst, this incision should be made below the centre of the cyst. By opening the anterior wall of the cyst, the posterior wall of the cyst is visible directly from the inside, and the choledochal cyst can be freed from surrounding tissues including the portal vein and hepatic artery more easily than by dissecting the cyst free without incising the anterior wall (Fig. 42.3).

If a choledochal cyst is extremely inflamed and adhesions are very dense, mucosectomy of the cyst should be performed rather than full-thickness dissection to minimize the degree of surgical stress to which the patient is exposed. In order to prevent postoperative pancreatitis and/or stone formation due to persistence of residual cyst, the distal common bile duct should be resected as close as possible to the pancreatico-biliary junction (Fig. 42.4). In cystic choledochal cyst, the distal common hepatic duct is narrow—sometimes so narrow that it looks blind-ended and cannot be identified specifically. Thus, if mucosectomy is completed up to the pancreatico-biliary junction, it is unlikely that a residual cyst will develop within the pancreas (Fig. 42.5). In contrast, in fusiform choledochal cyst, excision of the distal common hepatic duct is more difficult because the distal common bile duct is still wide at the pancreatico-biliary junction; if it is not excised properly, the likelihood that the distal common bile duct will be left within the pancreas is high.

Following mucosectomy, the distal end of the cyst is transfixed twice using 3/0 or 4/0 absorbable sutures. The distal stump is either left or is buried in the muscle wall of the surrounding cyst (Fig. 42.6).

Because there are fewer adhesions associated with fusiform choledochal cyst, dissection of the proximal common bile duct is relatively easy (Fig. 42.7), and the common hepatic bile duct can then be prepared for the hepaticojejunostomy (HJ) anastomosis (Fig. 42.8).

Fusiform choledochal cyst is usually associated with complicated PBMU as well as debris and/or protein plugs in the common channel. Pancreatic duct anomalies are also often present. IE should be performed up to the distal common bile duct in fusiform cases to confirm that dissection can be performed safely.

If the distal common bile duct is resected along the *red line* (Figs. 42.9), over time a cyst will re-form around the distal duct left within the pancreas, leading to recurrent pancreatitis, stone formation in the residual cyst, or malignant changes in the residual cyst. In contrast, if the distal duct is resected along the *blue line* (Figs. 42.9)—that is, just above the pancreatico-biliary junction—cyst re-formation due to residual duct within the pancreas is unlikely (Fig. 42.9, *bottom right*).

Before the introduction of IE, it was difficult to excise the pancreatic portion of a fusiform choledochal cyst completely and safely for fear of injuring the pancreatic duct. IE prevents injuries to the pancreatic duct and reduces the risk for postoperative complications due to incomplete resection, such as recurrent pancreatitis, stone formation, and carcinoma.

Cystic choledochal cyst is often associated with stenosis and dilatation of the intrahepatic bile ducts (IHBD). If stenosis is present and left untreated at the time of cyst excision, stones may form in the IHBD and cholangitis may arise later in life. The incidence of postoperative complications such as recurrent cholangitis, stone formation, and anastomotic stricture is increased in patients with dilated IHBD. Surgical correction is required for congenital IHBD stenosis if it is severe and the IHBD distal to the stenosis is extremely dilated (Fig. 42.10). When stenosis exists at the hepatic hilum or at the first branch of the IHBD, surgical correction such as dilatation or ductal plasty can be performed, but if the stenosis is located distal to the first branch of the IHBD, treatment is difficult. Large, diffusely dilated IHBD in both lobes cannot be corrected, but liver resection such as segmentectomy or lobectomy may be recommended at a later stage if the dilatation is localized to a single lobe.

The ideal length of common hepatic duct required for successful anastomosis is up to approximately 10 mm (Fig. 42.11); a longer common hepatic duct may become kinked, leading to bile stasis in the IHBD (**a**). However, the lumen of the common hepatic duct should be inspected before trimming it to make it shorter because there may be ductal anomalies such as a luminal stenosis (**b**), separate opening (**c**), or septum (**d**). Variations in the anatomy of the common hepatic duct have been described in the literature and have been experienced firsthand by the authors. Such variations can affect the success of hepaticoenterostomy after transection of the cyst, but since IE became a routine part of cyst excision, anatomic variations have not posed any problems.

Cyst excision and Roux-en-Y HJ is the treatment of choice for choledochal cyst in children. End-to-end anastomosis during HJ is recommended to prevent elongation of the blind pouch if the ratio between the diameters of the common hepatic duct and the proximal Roux-en-Y jejunum at the proposed site of anastomosis is less than or equal to 1:2.5 (Fig. 42.12a). If end-to-side anastomosis is unavoidable, the common hepatic duct should be anastomosed as close as possible to the closed end of the blind pouch so there will be no blind pouch at the HJ anastomosis site (Fig. 42.12b). If an end-to-side anastomosis is performed far from the closed end of the blind pouch, the blind pouch will grow as the child grows, and an elongated blind pouch can contribute to bile stasis in the pouch and IHBD (especially if they are dilated), leading to stone formation.

Some surgeons predetermine the length of the Roux-en-Y jejunal limb (e.g., 30, 40, 50, or 60 cm) without considering the size of the child, which tends to make the Roux-en-Y jejunal limb unnecessarily long, especially in infants and younger children. The length of the Roux-en-Y limb should be customized so that the Roux-en-Y jejunojejunostomy fits naturally into the splenic flexure after the jejunojejunostomy is completed and returned to the peritoneal cavity. This is done by placing the jejunojejunostomy at the umbilicus and bringing the distal end of the limb up to the xiphoid process. By doing this, there will be no redundancy of the Roux-en-Y limb.

When a jejunojejunostomy and Roux-en-Y limb are used, the native jejunum and the Roux-en-Y jejunal limb should be approximated for up to 8 cm cranially from the jejunojejunostomy, both to ensure bile in the Roux-en-Y limb and to ensure that contents of the native jejunum flow smoothly down into the jejunum distal to the jejunojejunostomy. Otherwise, the jejunojejunostomy tends to become T-shaped, and there may be reflux of jejunal contents into the Roux-en-Y limb, leading to dilatation of the jejunal limb and biliary stasis in the Roux-en-Y limb.

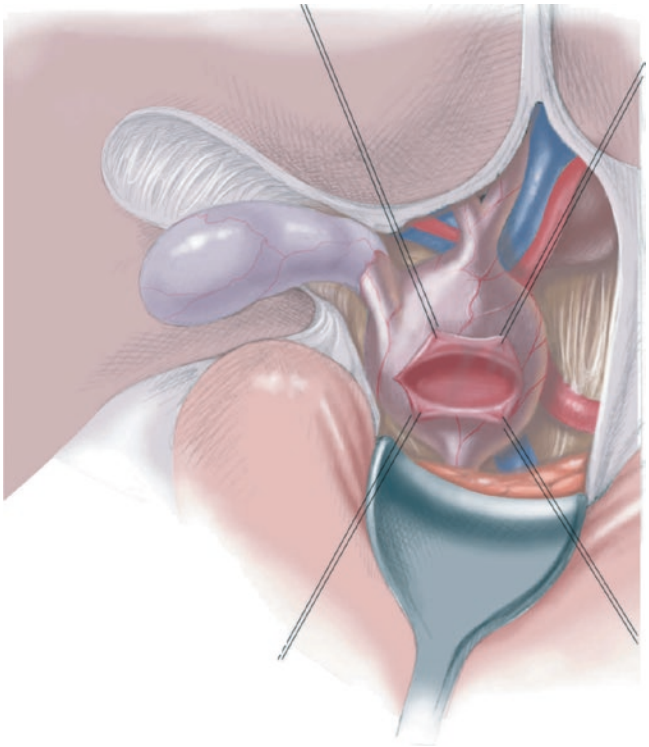

Fig. 42.2 Opening the anterior wall of a cyst

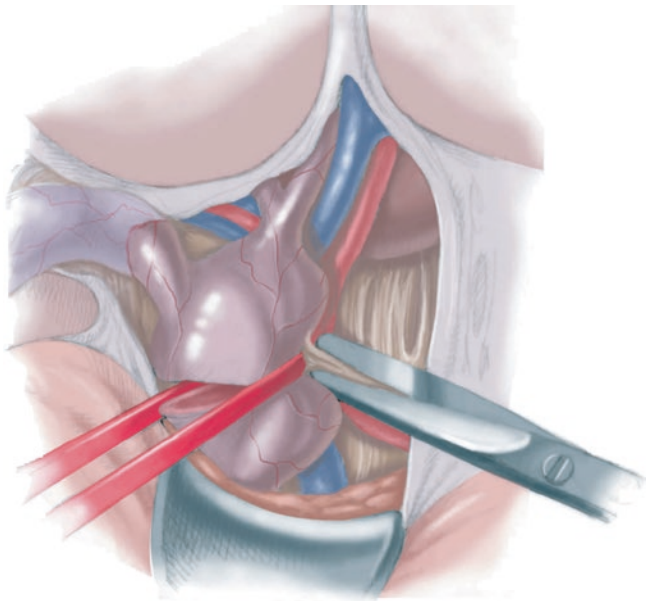

Fig. 42.3 Freeing the cyst from surrounding tissues

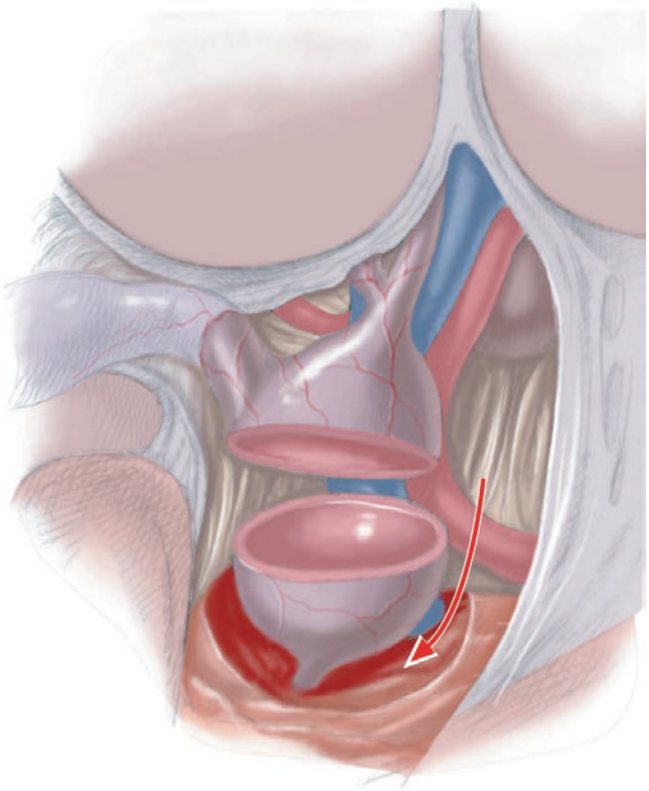

Fig. 42.4 Preparing for mucosectomy of an inflamed cyst

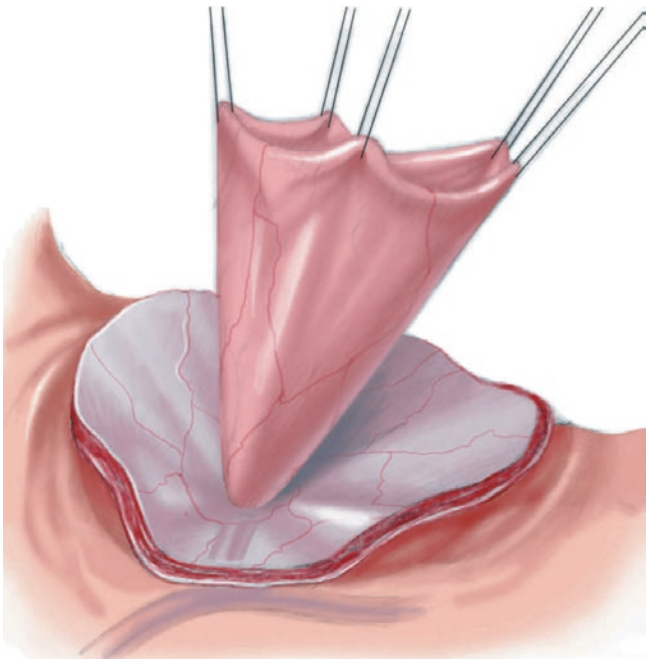

Fig. 42.5 Performing mucosectomy

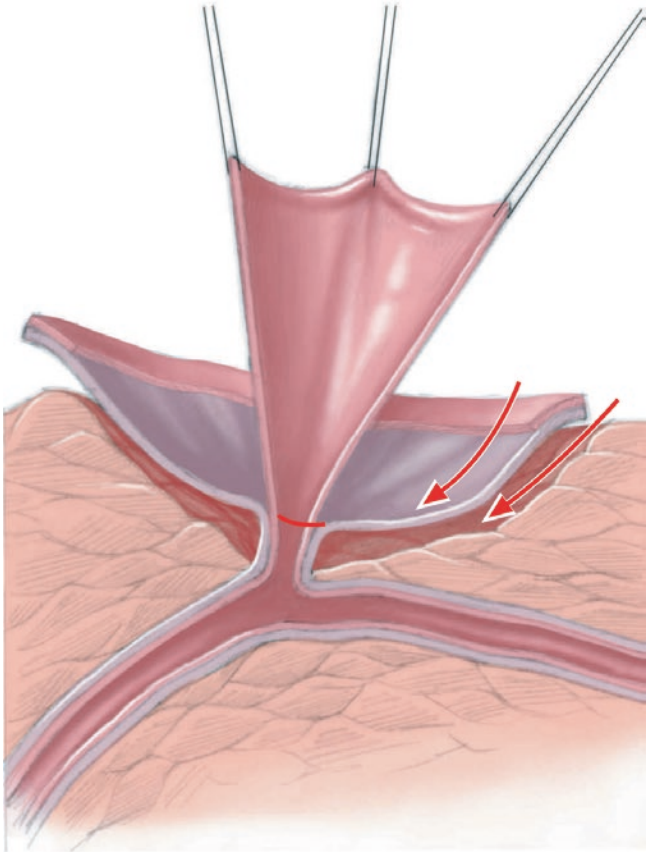

Fig. 42.6 After mucosectomy

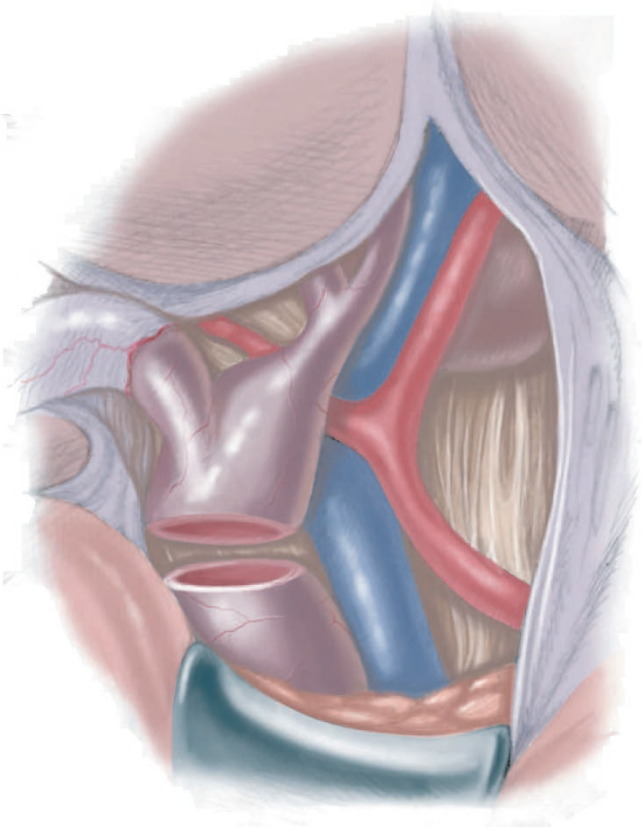

Fig. 42.8 Common hepatic bile duct prepared for hepaticojejunostomy (HJ) anastomosis

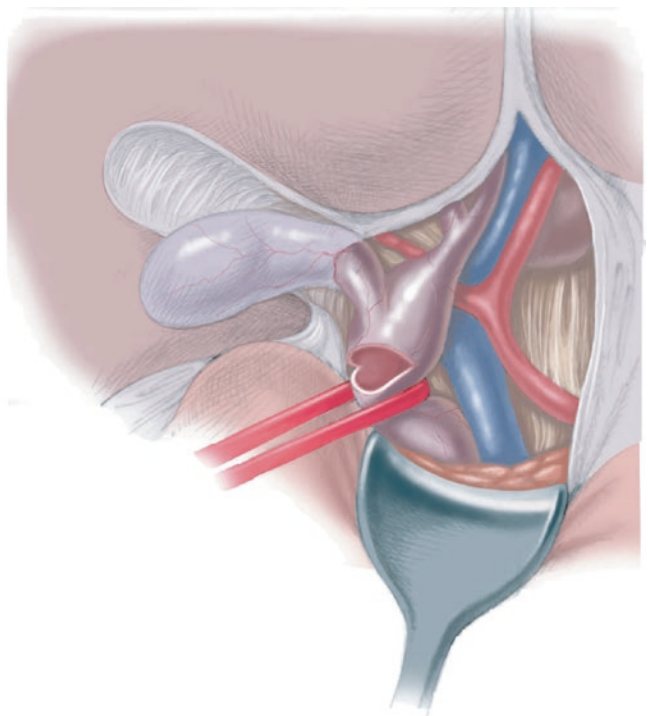

Fig. 42.7 Dissection of the proximal common bile duct

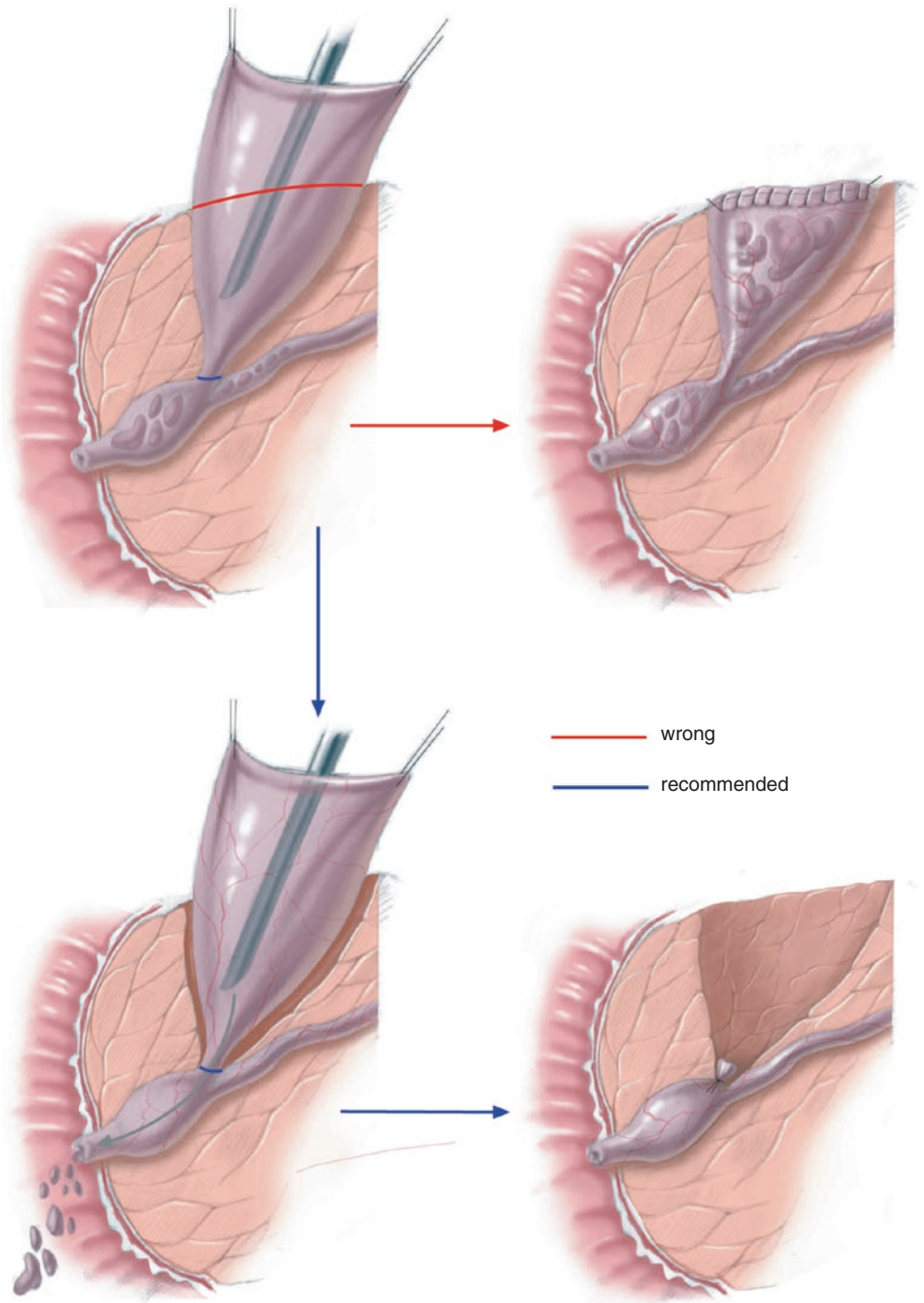

Fig. 42.9 *Top left*, Fusiform choledochal cyst: choosing location for resection of the distal common bile duct. *Top right*, Wrong resection, with subsequent stone formation in residual duct. *Bottom left*, Recommended resection. *Bottom right*, Result of recommended resection eliminating residual duct

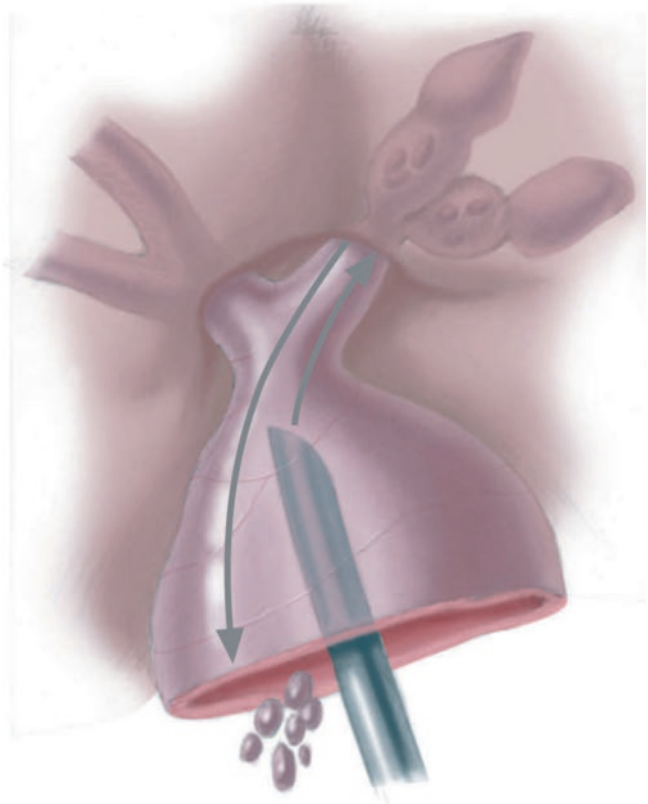

Fig. 42.10 Surgical correction of severe intrahepatic bile duct (IHBD) stenosis when the IHBD distal to the stenosis is extremely dilated

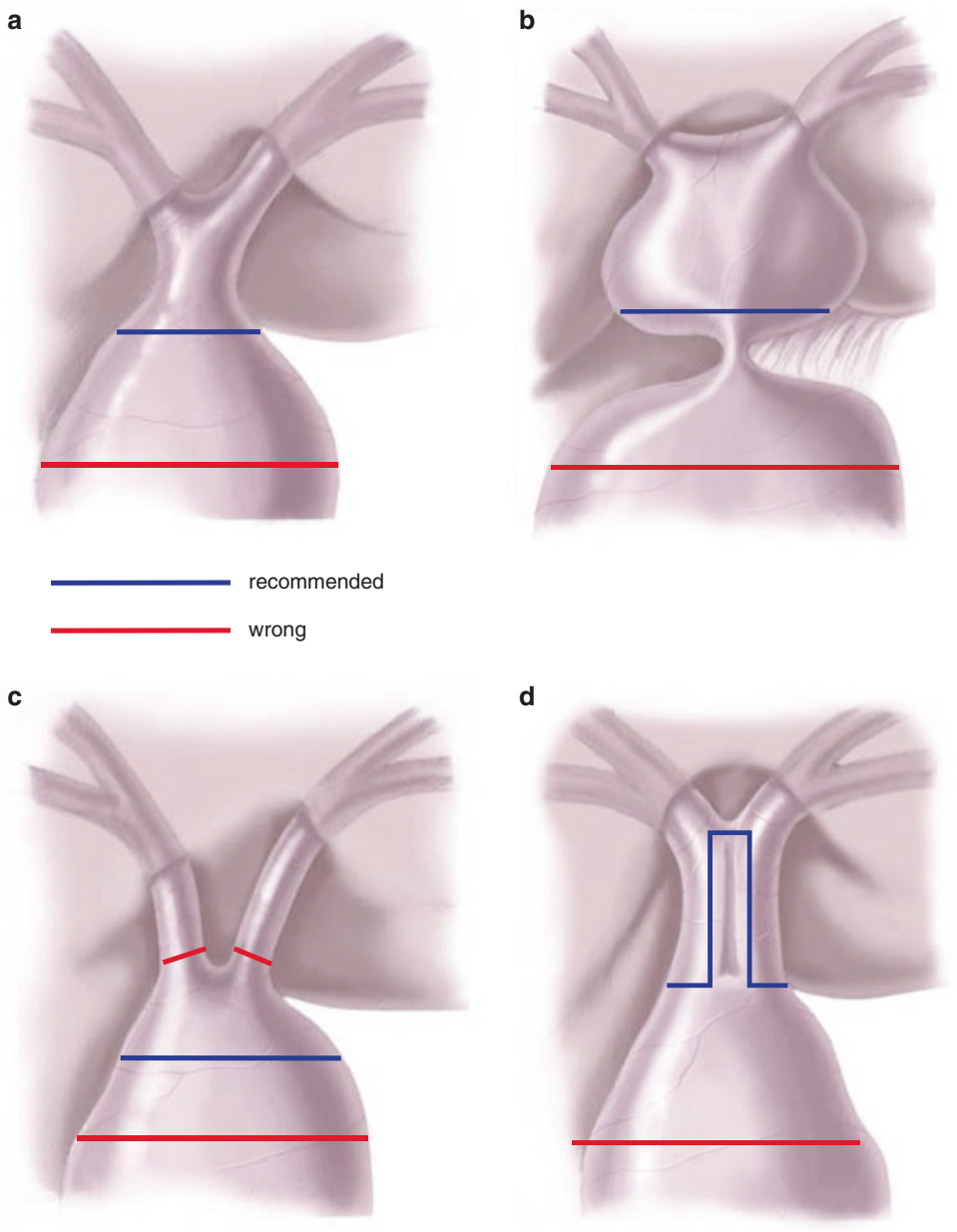

Fig. 42.11 (**a**) The recommended length of common hepatic duct required for successful anastomosis. (**b**–**d**) Possible ductal anomalies

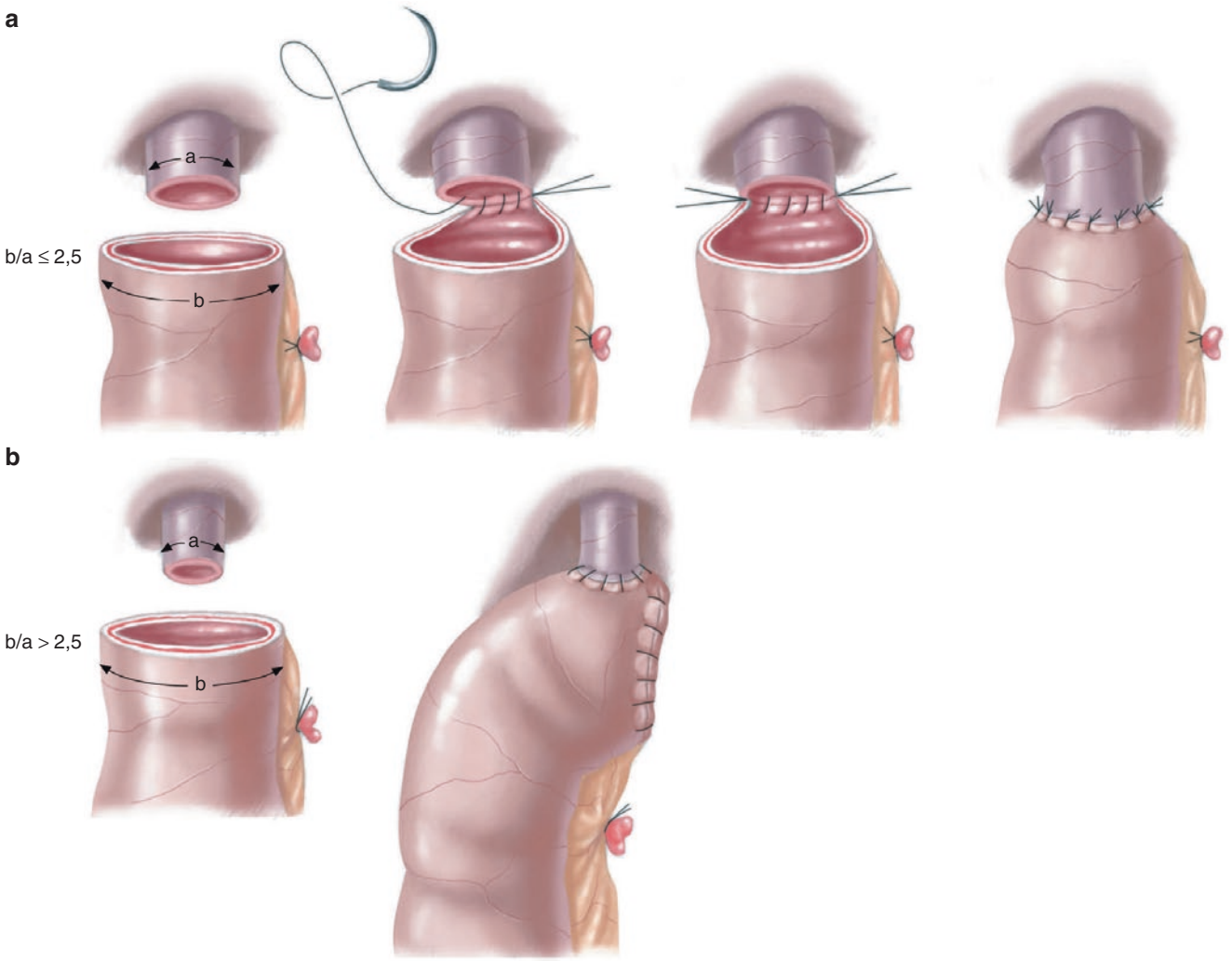

Fig. 42.12 End-to-end anastomosis during HJ (**a**). End-to side anastomosis (**b**)

42.2.2 Laparoscopic Choledochal Cyst Excision

The authors perform laparoscopic excision of a choledochal cyst according to the open complete excision technique presented earlier with routine IE. For laparoscopic excision, IE is retermed intralaparoscopic endoscopy.

The patient is positioned at the foot of the table and the table is tilted 15° to be in a reverse Trendelenburg position. The surgical team is initially positioned with the surgeon at the patient's feet, an assistant with a camera on the surgeon's left, and another assistant on the right with the monitor placed towards the patient's head (Fig. 42.13). Later, for the HJ anastomosis, the surgeon will move to the patient's right side in order to face the monitor for the whole procedure.

An open Hasson technique through a supraumbilical incision is used for inserting either a 5- or 10-mm, 30° or 45° laparoscope; CO_2 pneumoperitoneum is established at a pressure of 10–12 mmHg. Three additional 5-mm trocars are inserted in the right upper quadrant, left paraumbilical area, and left upper quadrant (Fig. 42.14). After initial inspection of the abdomen, intraoperative cholangiography is performed through the gallbladder if MRCP or ERCP have not precisely delineated the exact anatomy of the hepato-bilary-pancreatic ductal system.

Adequate exposure of the porta hepatis is achieved by introducing a percutaneous stay suture just below the xiphoid process to snare the falciform ligament and retract the liver (Fig. 42.15). Babcock forceps to further expose the porta hepatis are inserted through the left subcostal port in the anterior axillary line to grasp and elevate the gallbladder to facilitate dissection of the choledochal cyst from surrounding structures, such as the portal vein and hepatic artery. The cystic artery is identified, clamped and divided (Fig. 42.16). Dissection of the choledochal cyst is commenced along a plane created by peeling away the peritoneum from the cyst using monopolar electrocautery and a Maryland dissector starting from the anterior wall, moving laterally and medially along the lower one third of the cyst (Fig. 42.17). As mentioned in the description of open cyst excision, the anterior wall of the cyst is opened first in order to expose the posterior wall of the cyst.

At this point, IE is performed before transecting the posterior wall (Fig. 42.18). An additional 3.9-mm trocar is placed in the left epigastrium for the introduction of a fine pediatric ureteroscope (see Fig. 42.14). The tip of the ureteroscope is inserted into the common channel through the distal choledochal cyst to remove protein plugs, as shown in Fig. 42.9, but under laparoscopic control. Subsequently, the ureteroscope can also be inserted through the left paraumbilical trocar for intralaparoscopic hepatic bile duct endoscopy to remove debris in the IHBD, if present (Fig. 42.19).

After the cyst is freed, the distal part is divided as close as possible to the pancreaticobiliary junction and the stump is ligated with an Endoloop ligature. The exact level of transection of the distal common bile duct can also be determined, if the orifice of the pancreatic duct in the common channel is identified (see Fig. 42.9). The proximal cyst is divided just below the hepatic duct bifurcation into right and left hepatic ducts, aproximately 10 mm below the porta hepatis (Fig. 42.20).

A segment of proximal jejunum distal to the ligament of Treitz is exteriorized by extending the umbilical port incision, and bowel reconstruction is performed extracorporeally according to the same principles as described earlier for open cyst excision: (1) customizing the length of the Roux-en-Y limb; (2) approximating the Roux-en-Y limb to the native jejunum for 8 cm cranially; (3) using a scalpel, not monopolar diathermy, to create a 5-mm antimesenteric enterotomy for the HJ anastomosis; (4) returning the exteriorized jejunum to the abdominal cavity with the closed end of the jejunal limb being brought up via a retrocolic window to the porta hepatis.

Technically, the enterotomy should be created near the closed end of the Roux-en-Y limb to allow the common hepatic duct to be anastomosed as close as possible to the closed end of the blind pouch (Fig. 42.21). The edges of the enterotomy are temporarily apposed with two 7/0 PDS sutures to prevent spillage of bowel contents when it is brought through the retrocolic window. The sutures are cut at the time of HJ anastomosis. Avoidance of monopolar diathermy to create the 5-mm enterotomy prevents lateral thermal injury to tissues around the enterotomy, which could be implicated in postoperative anastomosis leakage.

Two additional 3- to 5-mm ports are inserted for the HJ anastomosis: one lateral right subcostal port, and one between the right subcostal and right upper quadrant ports (see Fig. 42.14). To perform the HJ anastomosis in an end-to-side fashion, interrupted 5/0 or 6/0 absorbable sutures are placed using the right upper quadrant port for the needle holder in the right hand, the 5-mm port for the scope, and the 3-mm subcostal port for the needle receiver in the left hand. Both the right and left edge sutures are exteriorized and used as traction sutures during anastomosis of the anterior wall to enhance accuracy, especially when the HJ anastomosis diameter is less than 9 mm (Figs. 42.22 and 42.23).

A tube drain is inserted in the pouch of Winslow. The resected cyst and gallbladder are extracted through the umbilical port. The trocars are removed and the wounds are closed.

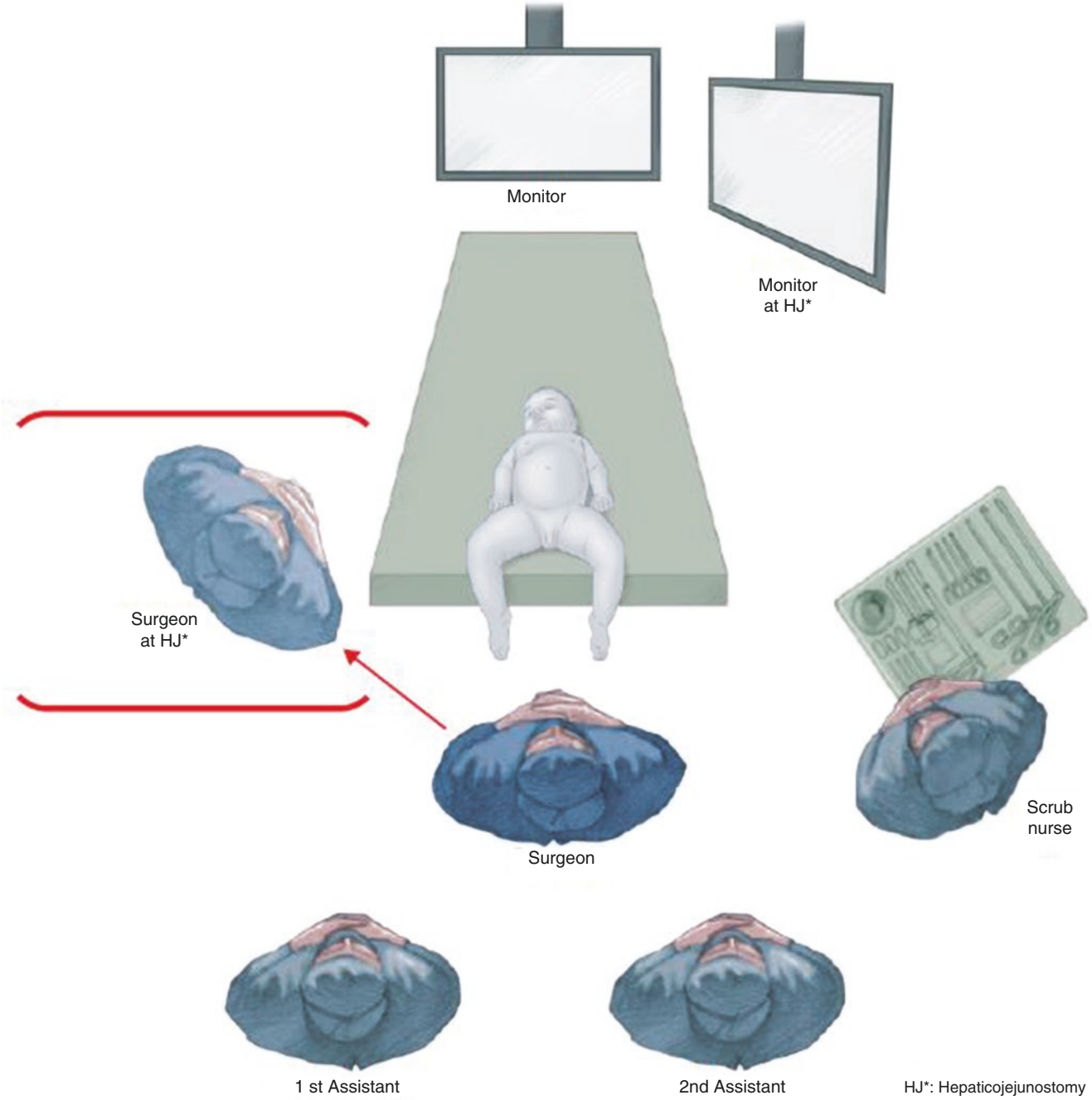

Fig. 42.13 Room setup for laparoscopic excision of a choledochal cyst

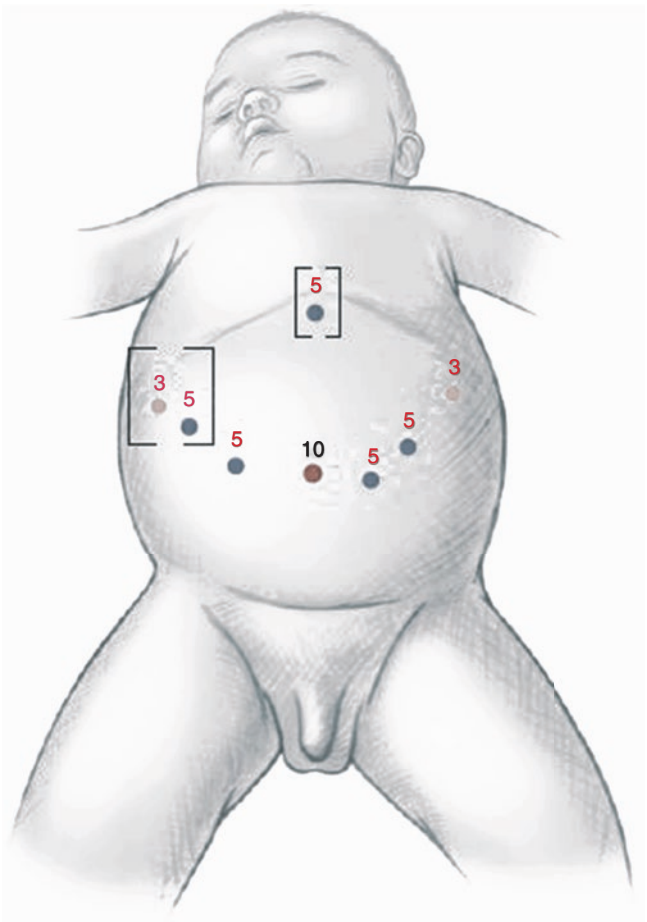

Fig. 42.14 Port locations

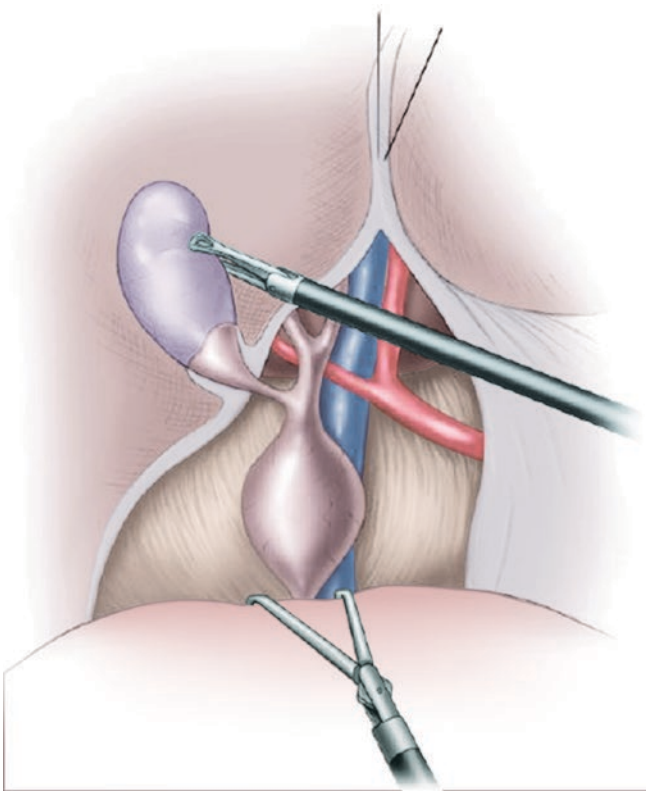

Fig. 42.15 Exposure of the porta hepatis

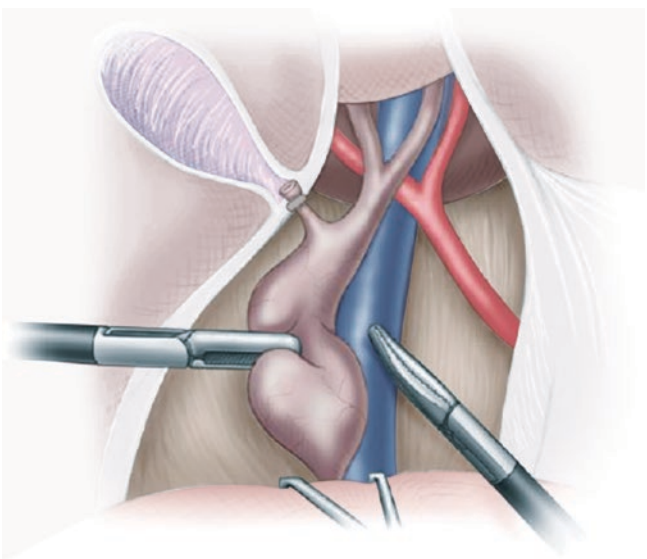

Fig. 42.16 Identification and division of the cystic artery

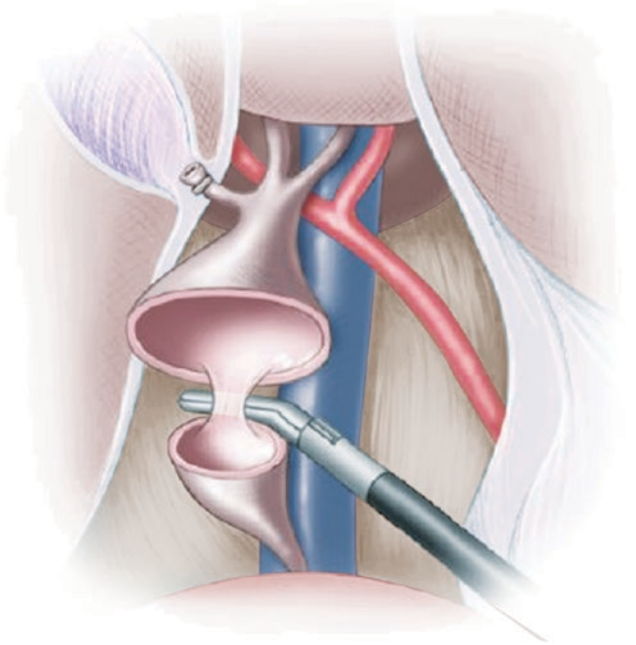

Fig. 42.17 Commencing the dissection of the cyst

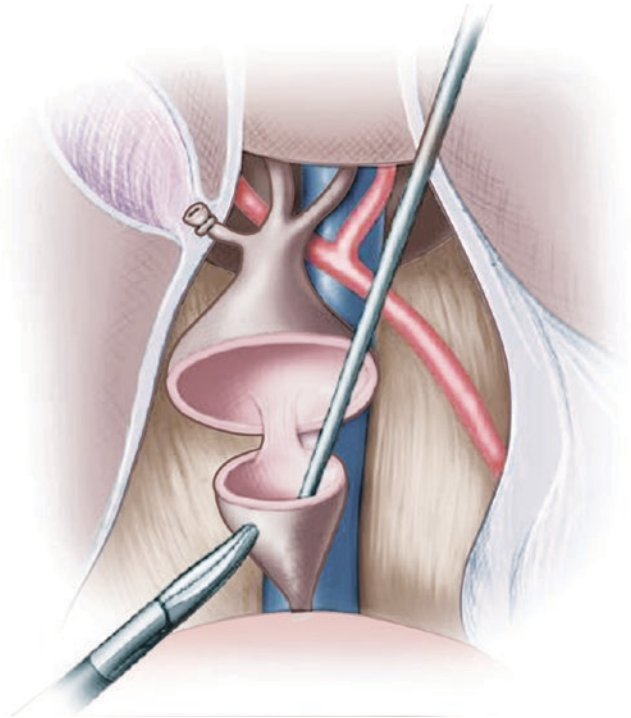

Fig. 42.18 Intralaparoscopic endoscopy

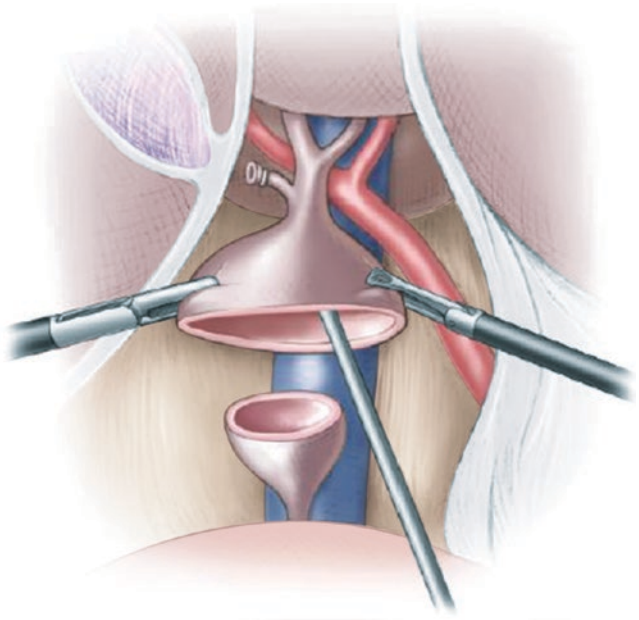

Fig. 42.19 Intralaparoscopic hepatic bile duct endoscopy to remove debris

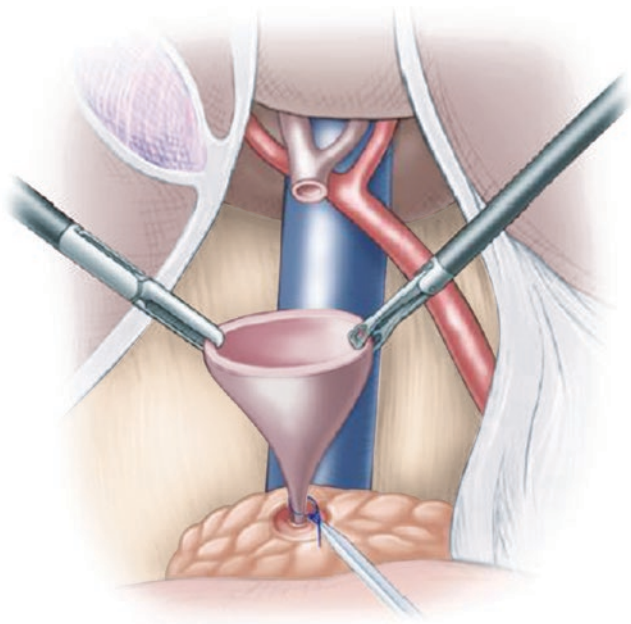

Fig. 42.20 Division of the proximal cyst

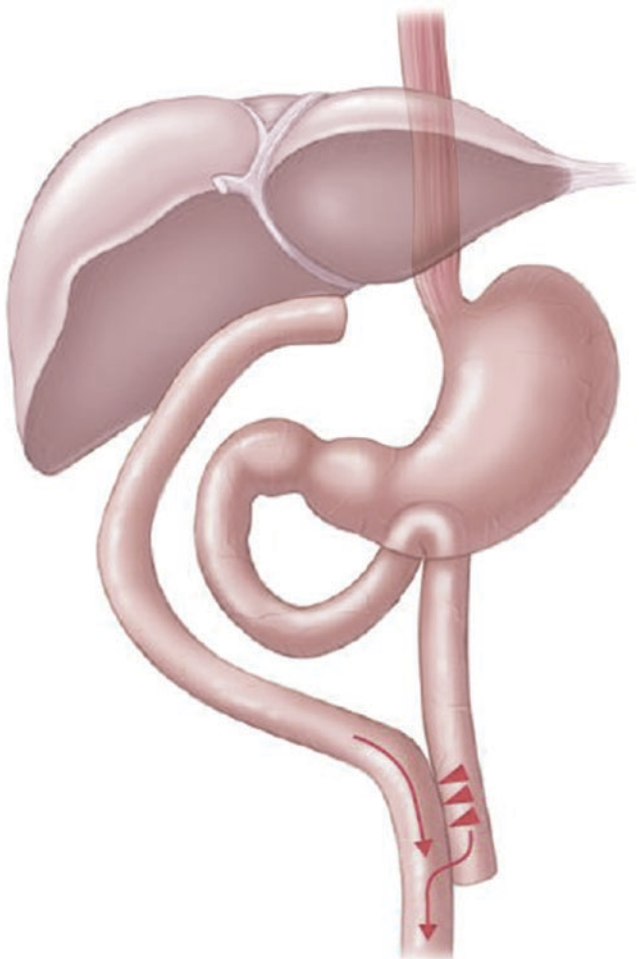

Fig. 42.21 Creation of Roux-en-Y

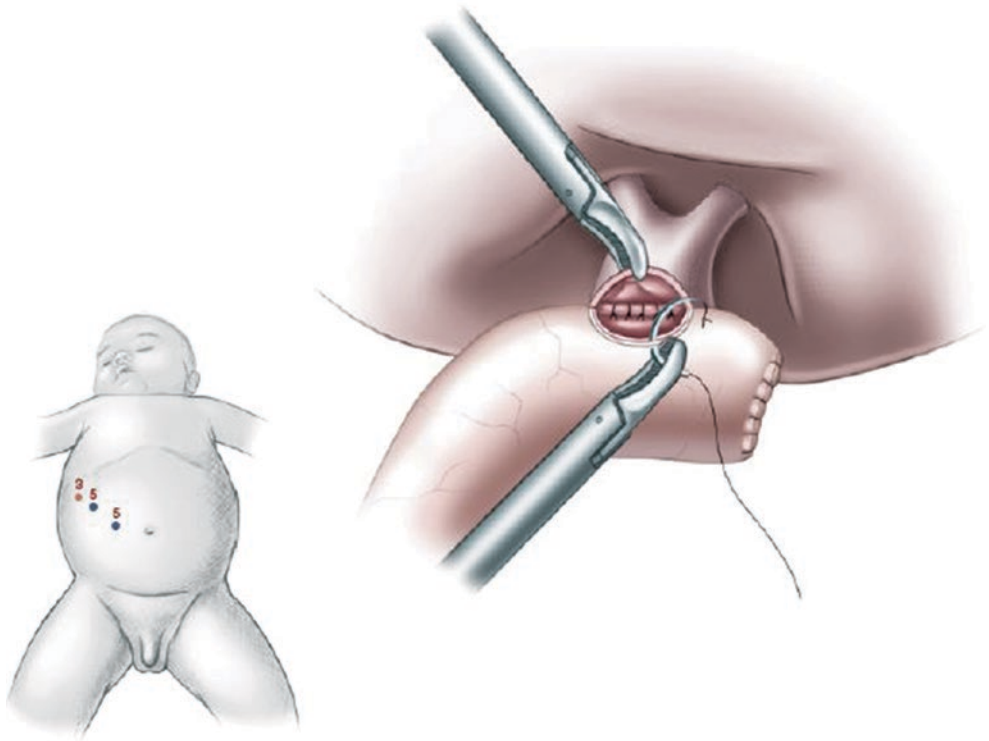

Fig. 42.22 The anterior wall of HJ anastomosis

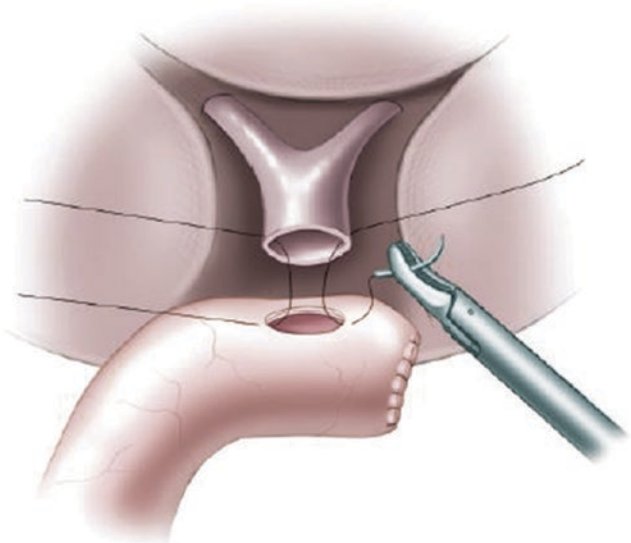

Fig. 42.23 Right and left edge suture exteriorization during anastomosis of the anterior wall to facilitate accuracy

42.3 Outcome

In a report comparing laparoscopic cyst excision with open cyst excision in children, operative time was found to be longer and overall costs higher when laparoscopy was used, but there was significantly less blood loss and the duration of hospitalization was shorter. There were no significant differences in the incidence of bile leakage or wound infection rates. Although technically challenging and time-consuming, laparoscopic excision imparts less surgical stress on patients than open excision, and parents are generally more satisfied with the smaller scars.

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Cholecystectomy

43

Alan Mortell and Farhan Tareen

Cholecystitis and cholelithiasis are increasing in frequency in infancy, childhood, and adolescence; the incidence is reported to be between 0.15% and 0.22%. There is no sex predilection before puberty, but after puberty the frequency of gallbladder stones increases significantly in females (4:1).

43.1 Pathogenesis

The origin of gallstones remains unknown in approximately 80% of pediatric patients. The commonest cause of cholelithiasis is haematologic disease (such as sickle cell disease, hereditary spherocytosis, and thalassaemia), so gallstones are more often seen in older children and adolescents. In the absence of an underlying haematologic disorder, most gallstones are associated with obesity, adolescent pregnancy, a positive family history for cholelithiasis, the use of oral contraceptives, the presence of a choledochal cyst, cystic fibrosis, previous cardiac transplantation, and the use of extracorporeal membrane oxygenation (ECMO). Total parenteral nutrition often results in cholestasis and stone formation in neonates and infants.

43.2 Diagnosis

Patients usually present with pain, jaundice, or both. Less often than in adults, patients present with fever, right upper quadrant tenderness, and leukocytosis, suggesting the presence of cholecystitis. Real-time ultrasonography, the first-line investigation to confirm the diagnosis of cholecystitis and cholilithiasis, has an accuracy of 96%. Nuclear cholescintigraphy may be useful if the diagnosis remains uncertain after ultrasonography. Magnetic resonance cholangiopancreatography (MRCP) is a noninvasive modality that may be indicated to delineate stones if there is dilatation of the common bile duct (CBD) on ultrasound or if the patient presents with obstructive jaundice. These patients are generally placed on antibiotics and oral intake is withheld until it is clear that their acute inflammation is resolving.

43.3 Surgical Treatment

Patients with asymptomatic cholelithiasis are not scheduled for surgery unless they develop symptoms or complications. When possible, we prefer to perform cholecystectomy as an elective procedure after the inflammation has resolved. When pain or cholecystitis persists, however, cholecystectomy is performed without delay. If CBD stones are present, it is important to consider preoperative endoscopic retrograde cholangiopancreatography (ERCP) with sphincterotomy for removal of bile duct stones followed by laparoscopic cholecystectomy.

Today, laparoscopy is recognized as the "gold standard" procedure for cholecystectomy because of better cosmesis, less postoperative pain, and shorter hospital stay than with open cholecystectomy. The principles of the open and laparoscopic procedures are essentially the same, except for the incision. These procedures are routinely performed under general anaesthesia with endotracheal intubation. A nasogastric tube is placed for gastric decompression, and a bladder catheter may be used to empty the bladder.

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43.3.1 Laparoscopic Cholecystectomy

The patient is placed supine on the operating table. To facilitate access to the patient, we place the patient's arms at the patient's side rather than at right angles to the patient. The patient is placed on a table suitable for fluoroscopy, under the assumption that an operative cholangiogram may be necessary. The abdomen is prepped and draped as is customary, so that the entire abdomen from xiphoid to pubis and from posterior axillary line to posterior axillary line is accessible. Four cannulas generally are used (Fig. 43.1), although it is possible to use fewer ports. In some cases, a single port can be used (SILS: Single Incision Laparoscopic Surgery). The position of the ports depends somewhat on the patient's size.

We begin the procedure with the insertion of an umbilical cannula of 10 mm diameter (Fig. 43.2). Although many are comfortable with the use of a Veress needle, we believe that the safest approach is an open Hasson approach to cannula insertion. A 10-mm incision first is made in the umbilical ring, either cephalad or caudad to the umbilicus. We make this incision with a number 15 blade and carry the incision through the skin to the fascia.

Two haemostat clamps are then used to grasp the fascia so that it can be incised. This incision is carried down to the peritoneum, which is opened sufficiently so that the cannula can be inserted. A right-angled retractor can be used to elevate the abdominal wall to facilitate cannula insertion. The cannula is fixed to the abdominal wall by placing a suture through the skin to a small ring of rubber catheter that was cut and placed around the outside of the cannula for this purpose. Alternatively, a balloon port can be used to anchor the umbilical access point. Proper cannula position is checked using a 5-mm telescope. The abdomen then is insufflated with CO_2 gas to 15 torr (12 mmHg).

Three other cannulas are inserted. Their position depends somewhat on the patient's size. In general, one needs a cannula for the gallbladder and liver retractor, one for a grasper to hold and manipulate the neck of the gallbladder, and another for dissection, duct and vessel ligation, and for the telescope when it is time for removal of the gallbladder. The gallbladder/liver retractor is placed in the mid to anterior axillary line between the level of the umbilicus and the level of the superior iliac crest (for smaller patients). A cannula is inserted at about the mid-clavicular line between the umbilicus and the costal margin (usually closer to the costal margin). The final cannula is inserted at about the midline and the mid-clavicular line (in smaller patients). All these cannulas can be inserted directly under the watchful eye of the surgeon's telescope/camera.

The gallbladder is grasped at the fundus and the gallbladder and liver are retracted superiorly and anteriorly (Fig. 43.3). A self-retaining retractor can be used to secure this cannula in place for most of the procedure. The right subcostal cannula is used for the retractor while we begin the dissection with a Maryland dissector. It is helpful to position the patient in a reverse Trendelenburg position with the right side up.

Dissection is begun by stripping away the peritoneum to expose the cystic duct and artery. Once the duct and artery are exposed, we apply clips to these structures and then divide them (Fig. 43.4). We advise against using electrocautery here so as to avoid any injury.

If there is any doubt as to the anatomy, if the duct is larger than it should be, or if the preoperative studies suggest the possibility of CBD stones, an intraoperative cholangiogram is performed. We clip the cystic duct as it joins the gallbladder and make a small nick in its wall, large enough to insert the cannula for the study (Fig. 43.5). We then use an 8- to 12-gauge plastic catheter as an "access port" for the cholangiogram catheter, and insert this directly through the abdominal wall. To minimize the risk of injury, we prefer a balloon-tipped catheter, which is inserted directly into the duct through a nick in the duct midway between the gallbladder and the CBD. Inflation of the balloon holds it in place. The cholangiogram can then be done under fluoroscopic control while the duct is observed to ensure that there is no leak of contrast.

If the cholangiogram is negative for retained stones, another clip is applied after the catheter is removed. If the cholangiogram shows that stones are present, there are three options: (1) the patient can be opened for exploration; (2) the operation can be completed and the patient referred for postoperative ERCP; or (3) a flexible endoscope can be used to examine and free the duct of stones (in larger patients). The third option in smaller patients consists of introducing a pediatric cystoscope into the duct for passage of a stone basket.

After the cystic artery and duct are divided, a right-angled hook electrocautery device or endoscopic shears connected to cautery are used to free the gallbladder from its bed (Fig. 43.6). After this is accomplished, the gallbladder bed is inspected for haemorrhage, which is controlled if found.

We then move the telescope from its umbilical site to the midline or left subcostal cannula to visualize removal of the gallbladder from the 10-mm umbilical cannula site. When stones are present, we open the gallbladder (after it has been partially exteriorised) and use a stone forceps to remove the stones until the gallbladder can be extracted. The abdomen then is inspected for haemorrhage and lavaged if bile has been spilled during the procedure.

We then close the fascia for all wounds of 5 mm or greater (in small children), or close the umbilical wound to prevent later herniation. All wounds are infiltrated with a long-acting local anaesthetic for postoperative analgesia. Patients are awakened from anaesthesia and discharged from the hospital when they can ambulate and tolerate liquids, usually on the day of surgery or after overnight hydration in the case of patients with sickle cell disease.

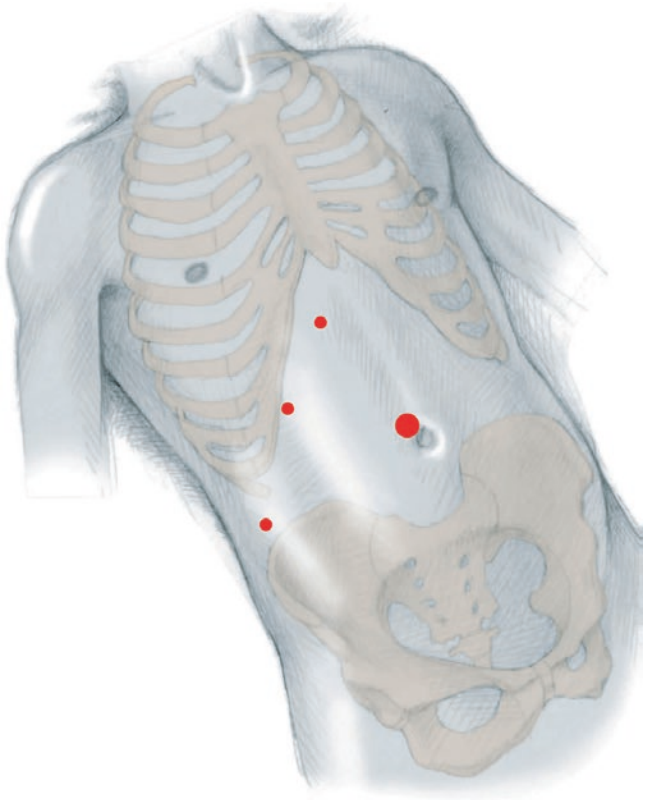

Fig. 43.1 Port positions for laparoscopic cholecystectomy

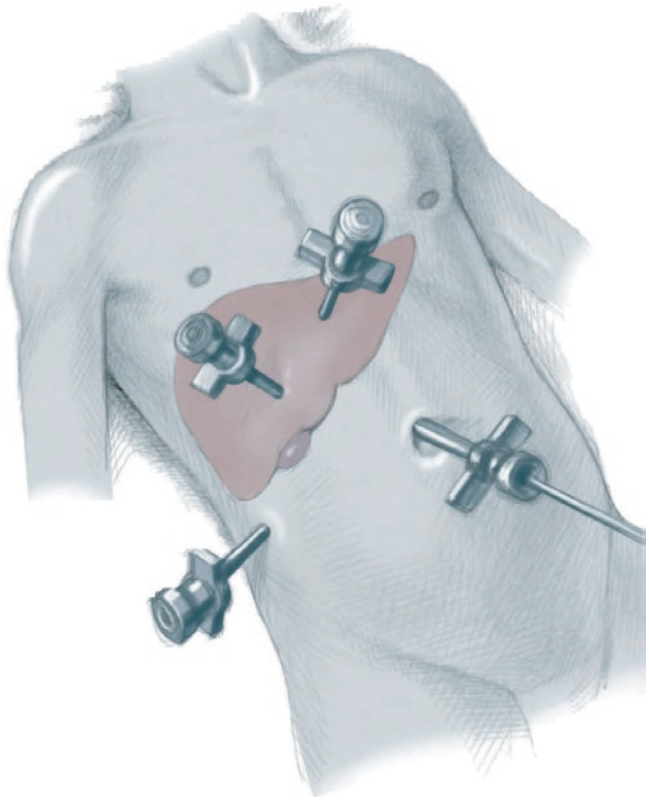

Fig. 43.2 Placement of the four cannulas that are generally used

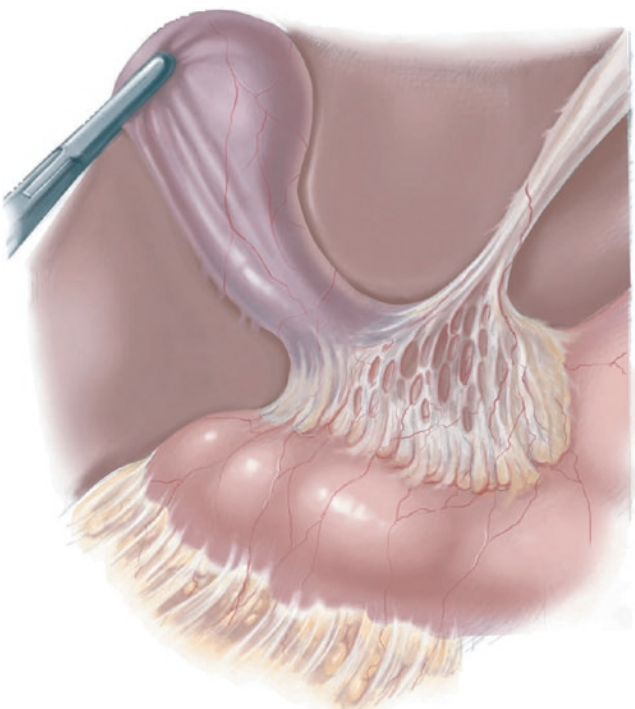

Fig. 43.3 Grasping the gallbladder and retracting it

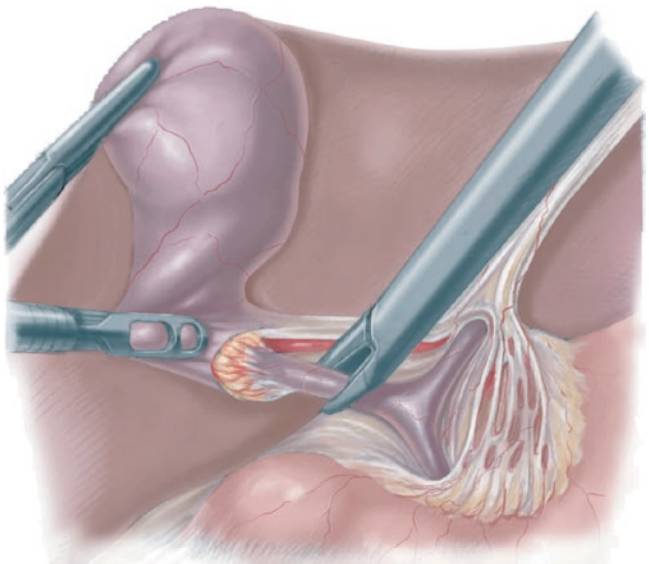

Fig. 43.4 Exposing and dividing the cystic duct and artery

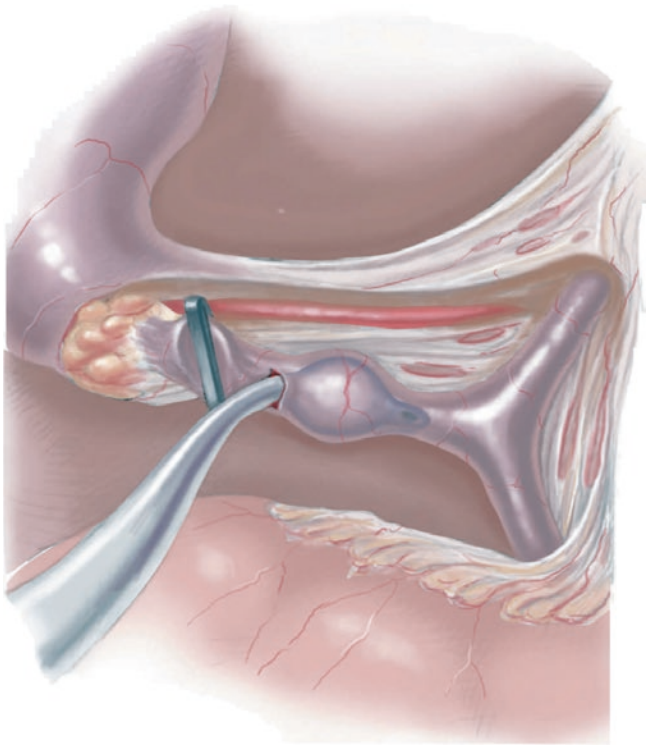

Fig. 43.5 Insertion of an intraoperative cholangiogram catheter

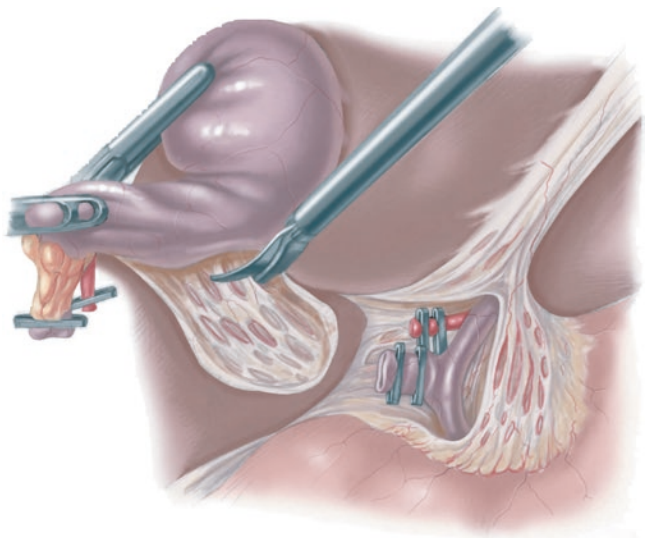

Fig. 43.6 Freeing the gallbladder from its bed

43.3.2 Alternative Procedures

Laparoscopic cholecystectomy has replaced open cholecystectomy for the vast majority of cases. Rarely, however, access cannot be achieved because of previous surgery, or the patient cannot tolerate the abdominal insufflation. In those cases, a subcostal or midline abdominal incision is made, the intestines and liver are retracted out of the way, and essentially the same procedure is carried out.

More recently, some surgeons have elected to perform the cholecystectomy from the fundus toward the porta hepatis. This technique may be helpful in cases of severe inflammation or marked oedema.

43.4 Complications

The most serious complication of this procedure is an unrecognized injury to the bile ducts, which can be prevented by clear visualization of the cystic artery and duct prior to division (the key point in cholecystectomy). This injury can occur when the anatomy is unclear, so the advice is to perform a cholangiogram when there is any doubt. Such injury can also occur from an electrical burn from using electrocautery near the portal area, so the recommendation is to avoid this practice.

When the gallbladder is accidentally opened during its dissection, bile and stones can spill into the peritoneal cavity. We make a modest effort to lavage and evacuate this spilled material, but rarely have we had postoperative problems occur from this event.

Conclusion

Laparoscopic cholecystectomy is one of the more gratifying procedures we do. It is quick, simple, and effective, and we see few problems afterwards.

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Congenital Hyperinsulinism of Infancy

44

Agostino Pierro, Augusto Zani, and Lewis Spitz

Hyperinsulinism as a cause of persistent hypoglycaemia in infancy and childhood is very uncommon (1:50,000). The importance of preventing hypoglycaemia (and reducing the likelihood of neurologic damage) by the administration of adequate quantities of carbohydrate cannot be overstressed.

44.1 Diagnosis

The diagnosis of congenital hyperinsulinism of infancy (CHI) is based on several criteria:

- Inappropriately raised plasma insulin levels for blood glucose concentration
- Glucose infusion rate greater than 10 mg/kg/min to maintain a blood glucose level above 2.6 mmol/L
- Low free fatty acid and blood ketone bodies during hypoglycaemia
- Glycaemic response to glucagons despite hypoglycaemia

In essence, the diagnosis is established by measuring insulin in a blood sample taken during hypoglycaemia (blood glucose <2.6 mmol/L). Among the lesions causing hyperinsulinism, 40–50% are focal. It is important to identify these lesions in order to avoid unnecessarily performing a near-total pancreatectomy.

There are two main histological variants of CHI: focal and diffuse.

- *Focal forms* are typically sporadic in inheritance and are restricted to a small area of the pancreas (2.5–7.5 mm). Within the focal lesion, several abnormal lobules are interspersed with acinar foci and a second endocrine-cell population can be detected. These features are representative of an abnormal developmental process rather than a tumorigenic process. Around the main focal lesion, there may be satellite areas of abnormal pancreatic tissue, so clear resection margins are crucial to avoid recurrence.
- *Diffuse forms* are typically inherited in an autosomal recessive or dominant manner and involve every β-cell throughout the whole pancreas, with variable involvement of the islets. The islet pattern is preserved but contains active β-cells with abundant cytoplasm and abnormal nuclei.

To distinguish between focal and diffuse subtypes, a genetic analysis is performed. Typically, children with diffuse CHI have homozygous recessive or compound heterozygote mutations in the *ABCC8* and *KCNJ11* genes (which encode the SUR1 and KIR6.2 proteins of the pancreatic β-cell). Focal CHI is associated with loss of heterozygosity for paternally inherited mutations in the KATP genes. Infants without characteristic genetic mutations of diffuse CHI undergo an 18F-fluoro-L-dihydroxyphenylalanine (^{18}F-DOPA) positron emission tomography (PET) scan combined with a CT scan. This PET-CT scan distinguishes between diffuse and focal disease: diffuse CHI exhibits a diffuse uptake on the PET-CT scan. In focal disease, the scan assists in localizing the site of the focal lesion, allowing a more directed pancreatic resection.

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44.2 Treatment

It is essential to insert a central venous catheter to monitor blood glucose levels and to provide a reliable route for intravenous glucose administration. Drug treatment includes diazoxide with chlorothiazide, nifedipine, octreotide, and new medicines such as long-acting octreotide.

The indications for surgery are failure to respond to intensive medical treatment for diffuse lesions, or the presence of a focal lesion. There are two surgical approaches to diffuse or focal CHI: open and laparoscopic. Diffuse forms of CHI that are not responding to medical treatment require a near-total pancreatectomy, whereas focal forms of CHI benefit from a partial pancreatectomy that includes a complete excision of the focal lesion.

44.3 Operative Techniques

44.3.1 Open Near-Total Pancreatectomy for Diffuse CHI

The pancreas is exposed by dividing the vessels in the gastrocolic omentum to expose the head, body. and tail of the pancreas (Fig. 44.1).

Any suspicious nodules (particularly if the preoperative studies have indicated focal disease) should be excised and sent for frozen-section histopathologic examination (Fig. 44.2).

Near-total pancreatectomy for diffuse disease starts with mobilizing the tail and body of the pancreas (Fig. 44.3). Bipolar coagulation of short pancreatic vessels is carried out, following which the vessels are divided. Starting at the tail of the pancreas in the hilum of the spleen, the dissection proceeds towards the neck of the pancreas. The distal part of the tail of the pancreas is divided and the end is sent for frozen-section histopathology to confirm the diffuse nature of the condition (i.e., diffuse or focal form of the disease).

In order to perform the resection of the uncinate process, the superior mesenteric vein is retracted to the left and the uncinate process is carefully dissected out from behind the vein until it is completely free from any attachments (Fig. 44.4).

The common bile duct is defined above the first part of the duodenum and a soft rubber sling is passed around the duct at this point (Fig. 44.5). From within the C-loop of the duodenum, a passage is created behind the first part of the duodenum, and the rubber sling is transposed behind the duodenum to appear within the C-loop. The aim of this manoeuvre is to identify the distal course of the common bile duct in or posterior to the head of the pancreas and to preserve its integrity during resection of the head of the pancreas.

The resection includes the head, body, and tail of the pancreas, including the uncinate process, leaving a small sliver of pancreatic tissue within the C-loop of the duodenum and that part of the pancreas that lies around the common bile duct and between the duct and the duodenum (Fig. 44.6). Before closing the abdominal incision, it is important to check the integrity of the common bile duct by applying gentle pressure on the gallbladder and observing for any bile leak. A suction drain is left in the pancreatic bed for 24–48 h postoperatively.

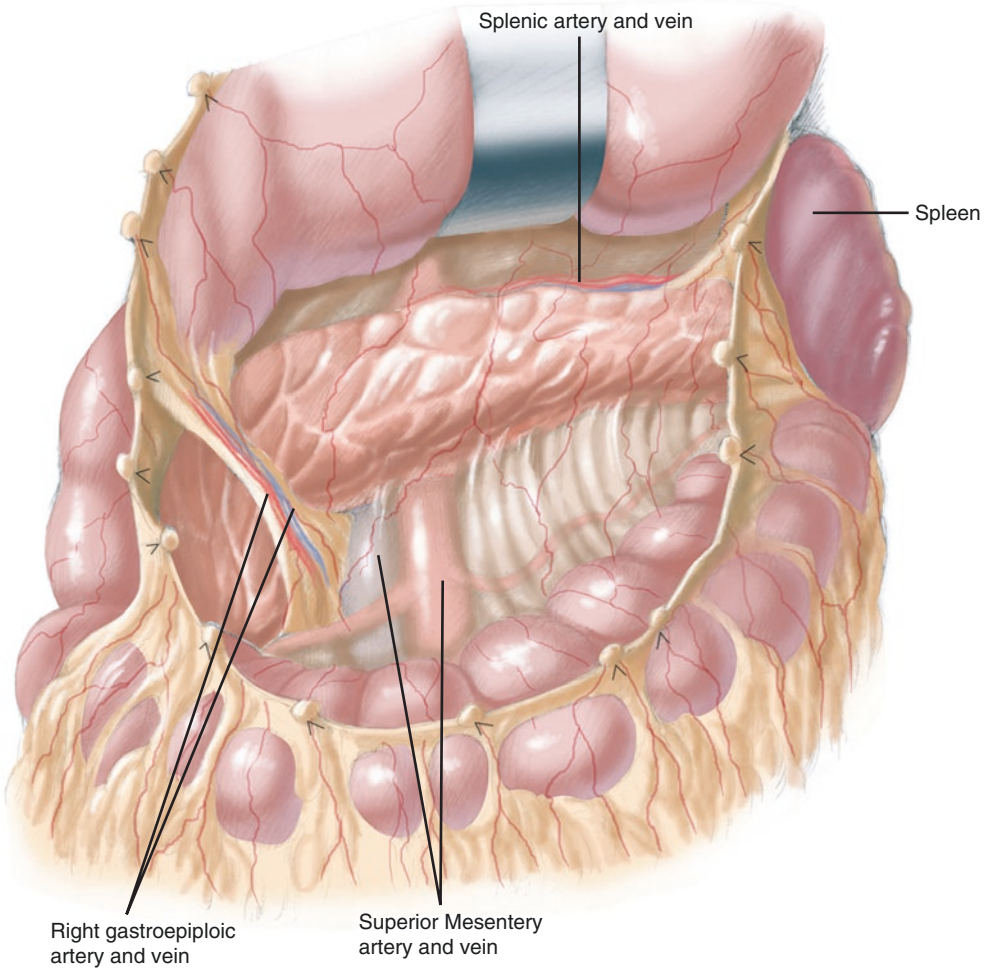


Fig. 44.1 Exposure of the pancreas

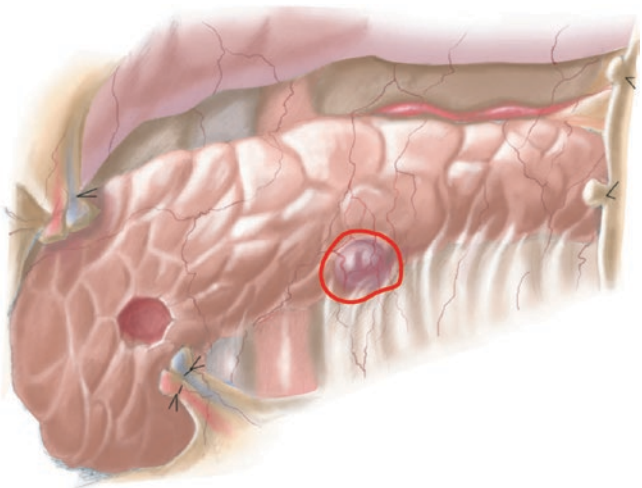


Fig. 44.2 Excision of suspicious nodules

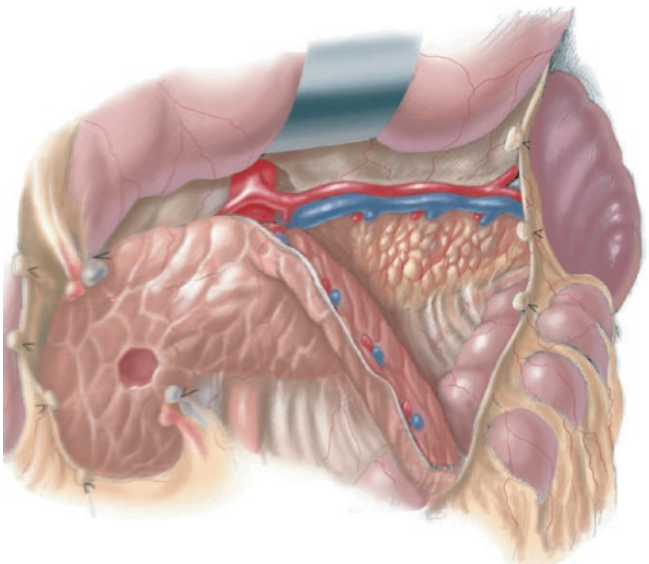


Fig. 44.3 Mobilizing the tail and body of the pancreas

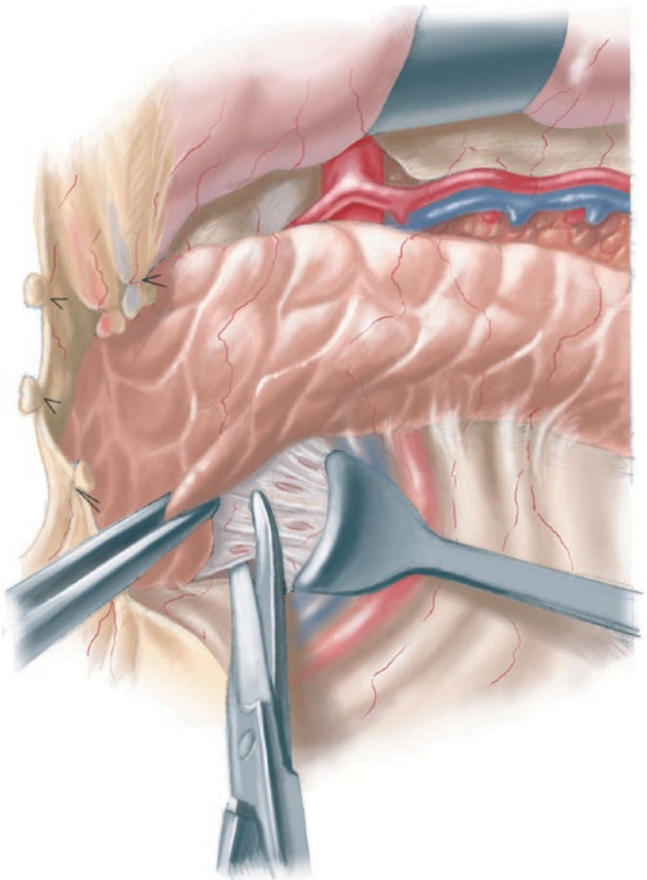


Fig. 44.4 Resection of the uncinated process

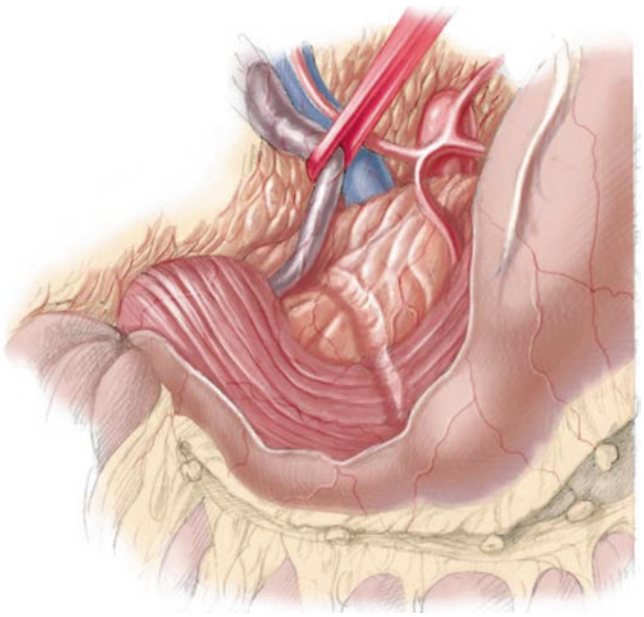


Fig. 44.5 Using a rubber sling to identify the distal course of the common bile duct

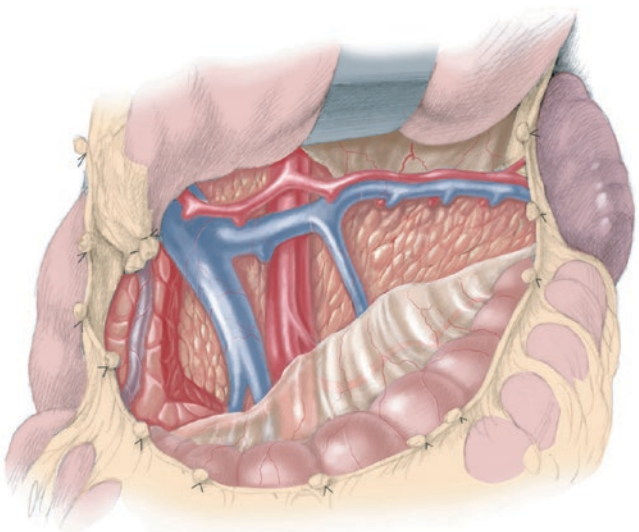


Fig. 44.6 The completed near-total resection

44.3.2 Laparoscopic Near-Total Pancreatectomy for Diffuse CHI

An umbilical 10-mm Hasson port is inserted by an open technique, and a 5-mm 30° camera is introduced. Three additional ports are placed: a 5-mm port in the left lower quadrant, a Nathanson retractor at the epigastrium, and a further working port (3 or 5 mm) in the right flank (Fig. 44.7). The gastrocolic omentum is divided, the lesser sac is entered, and the Nathanson retractor is used to retract the stomach. The head of the patient is elevated.

A stay suture is inserted into the tail of the pancreas, which is used to retract the pancreas superiorly. Dissection of the pancreas proceeds towards the head (Fig. 44.8). The short pancreatic vessels passing from the splenic vessels into the pancreas are divided using a 3-mm hook diathermy at very high coagulation settings. These vessels are the most common source of intraoperative hemorrhage, which may be controlled by applying gentle pressure using an atraumatic bowel grasper. The pancreatic tail is transected using the Harmonic Scalpel (Ethicon Endosurgery, Cincinnati, OH). The pancreatic tail is removed via the umbilical 10-mm port and sent for frozen section analysis to confirm the diagnosis of diffuse CHI. No further resection takes place until this confirmation has been obtained. Subsequent dissection is facilitated by the insertion of a stay suture at the cut surface of the remaining pancreas, which is resected in segments of approximately 2 cm. As dissection nears the head of the pancreas, stay sutures are placed in the uncinate process and the head of pancreas, which are retracted superiorly. A near-total pancreatectomy is achieved by leaving an adequate amount of pancreatic tissue along the medial border of the duodenum.

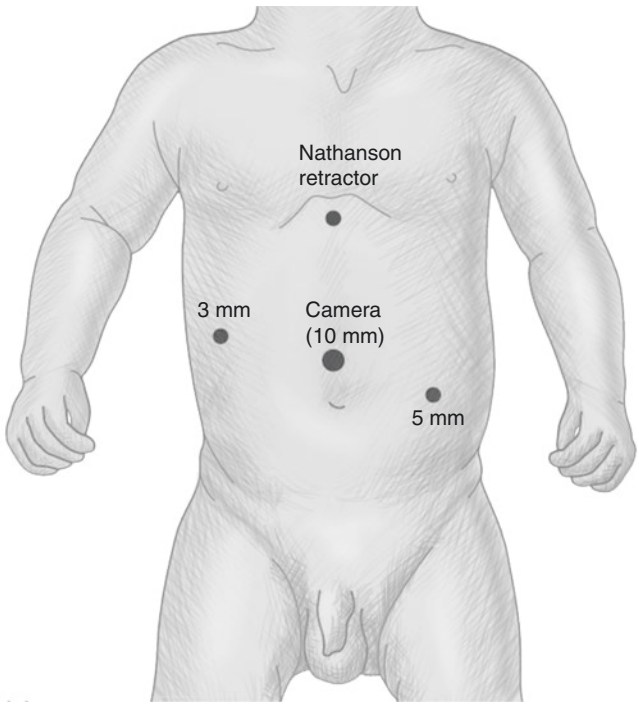
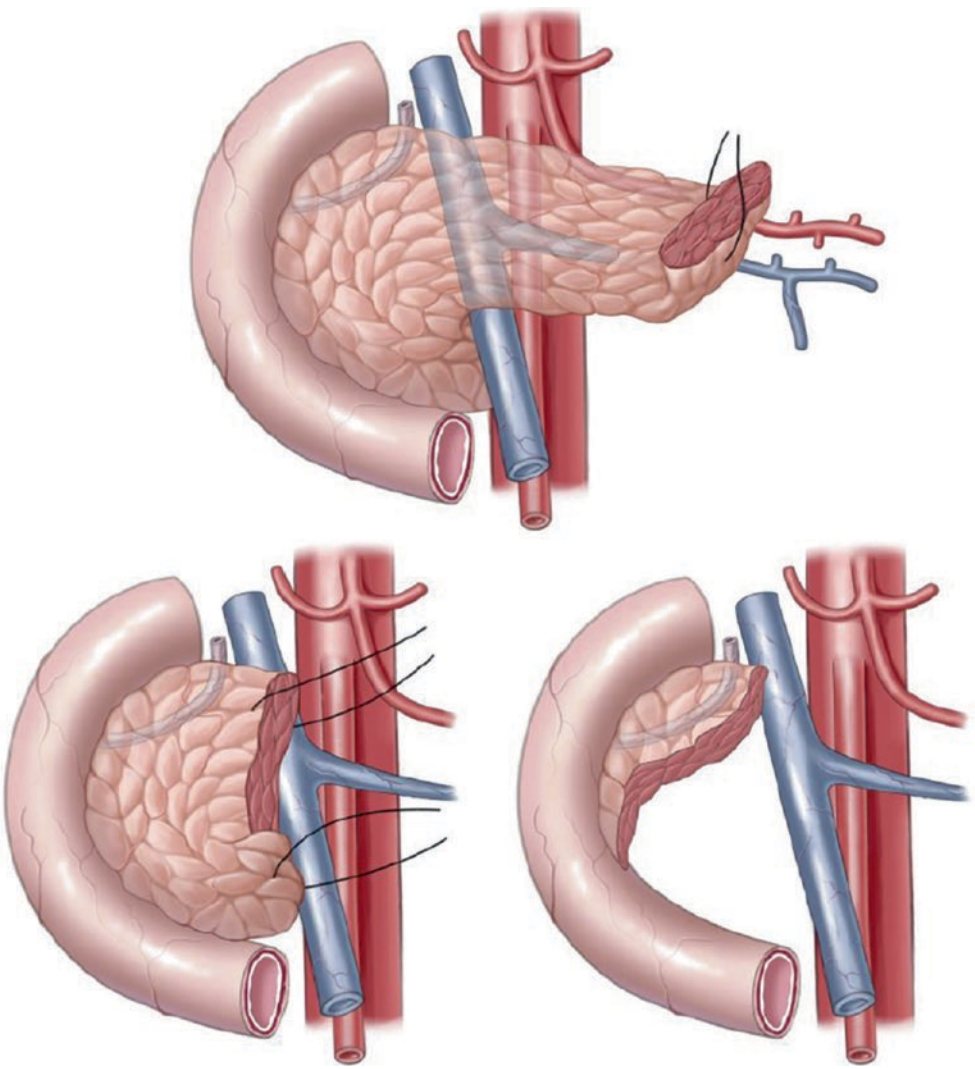


Fig. 44.7 Port placement for laparoscopic near-total pancreatectomy

Fig. 44.8 The steps in a laparoscopic near-total pancreatectomy



44.3.3 Partial Pancreatectomy for Focal CHI

In patients with focal CHI, a localized resection of the focal lesion is curative. The diagnosis of focal CHI and its localization is made using the ^{18}F-DOPA-PET CT scan. This imaging technique allows localization of the lesions, which are usually deep in the parenchyma and hardly visible macroscopically. The ^{18}F-DOPA-PET CT scan is beneficial for the preoperative surgical planning: in focal lesions of the head or neck of the pancreas, open or laparoscopic resection of the pancreatic head with a small rim of surrounding normal pancreatic tissue is carried out and pancreato-jejunostomy is performed to allow drainage of the distal pancreas. In distal focal lesions, a distal pancreatectomy is performed.

Focal lesion in head or neck of pancreas: The operation can be performed open or laparoscopically using the same approach described above. The pancreas is inspected for a nodular area, which indicates the site of the focal lesion. In focal CHI, the lesion is often deep within the parenchyma of the pancreas, however, and may not be visually evident. It is very rare to encounter superficial lesions that can be enucleated without major pancreatic resection. The pancreas is transected distally to the location of the focal lesion indicated by the ^{18}F-DOPA-PET CT scan. A stay suture is inserted to elevate the portion of the pancreas to resect and to facilitate the division of the short pancreatic vessels passing from the splenic vessels into the pancreas. These vessels can be divided using bipolar diathermy (open technique) or 3 mm hook diathermy (laparoscopic technique). When portion of the pancreas that should contain the focal lesion on the basis of the ^{18}F-DOPA-PET CT scan has been excised, frozen section histologic examination is performed to confirm the presence of the focal lesion and its complete resection (normal pancreas at the resection margin). Further pancreatic excision is carried out if the focal lesion is not seen on histology or it is not completely removed.

The head of the pancreas is dissected in the direction of the duodenum, carefully avoiding injuries to splenic vein and artery, portal vein and common bile duct. The pancreas is excised to include the area where the focal lesion is suspected to be present (Fig. 44.9). Histologic confirmation of complete excision of the focal lesion is required before performing a pancreato-jejunostomy. This anastomosis is done to allow drainage of the distal pancreas and can be performed via open laparotomy or via laparoscopy.

Focal lesion in body or tail of pancreas: In patients with more distal lesions, the entire procedure is best performed laparoscopically, but some surgeons continue to prefer the open approach. The operation starts by mobilizing the tail of the pancreas as described above for the near-total pancreatectomy. The portion of the tail/body of the pancreas including the focal lesion is excised (Fig. 44.10, *grey area*). The pancreas is transected using the Harmonic Scalpel, which in my experience avoids the need to suture the distal end of the pancreas. Histologic confirmation of complete excision of the focal disease is essential. Drains are not needed.

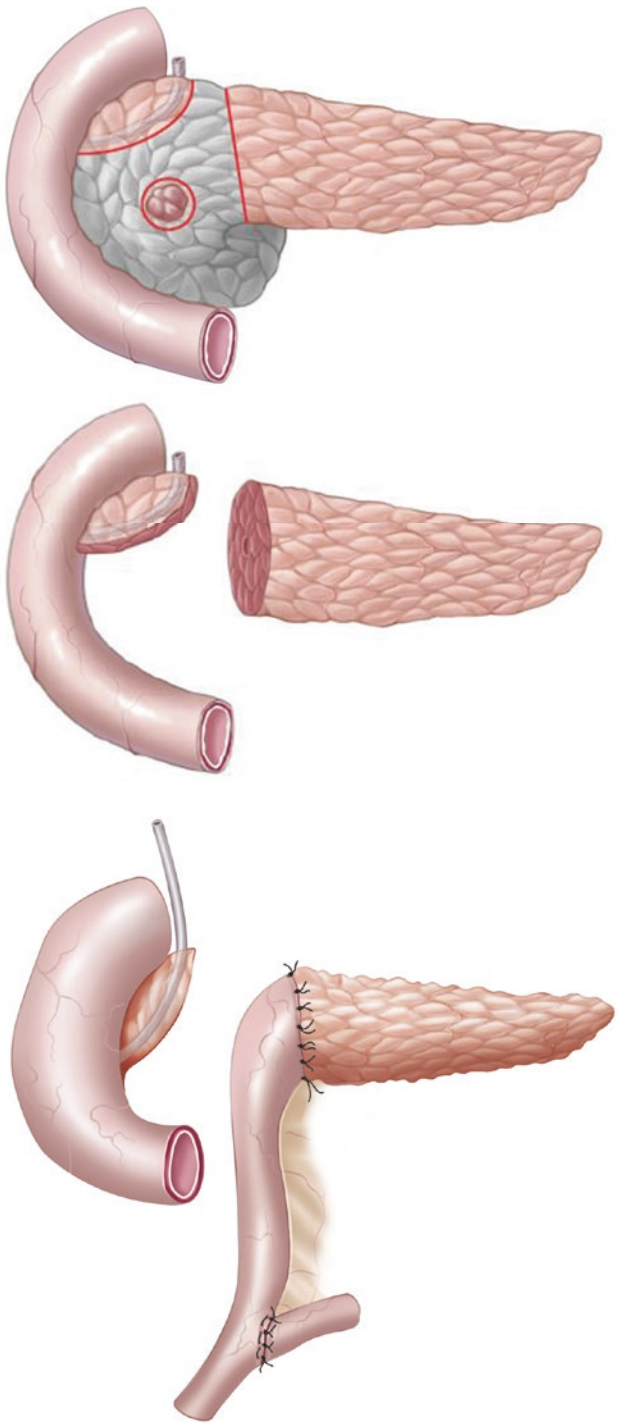


Fig. 44.9 Excision of the area of a focal lesion in the head or neck of the pancreas, followed by creation of a pancreato-jejunostomy

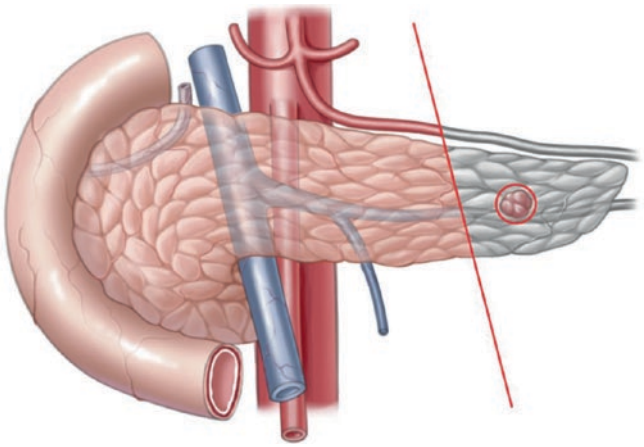


Fig. 44.10 Excision of the area of a more distal focal lesion

44.4 Postoperative Management

Enteral feeding is recommenced as early as the first postoperative day and is gradually advanced, reducing the intravenous infusion of dextrose. A period of gastroparesis can occur, particularly after excision of the head of the pancreas for focal lesions or near-total pancreatectomy for diffuse CHI. The management of postoperative hypoglycemia may require medical treatment, but it is usually transient. Further resection of the pancreas is needed if bolus enteral feeding and cessation of continuous intravenous glucose are not achieved.

44.5 Complications

Intraoperative haemorrhage should not occur. The most frequently encountered problem is trauma to the bile duct (2–10%). If the duct injury is detected intraoperatively, immediate repair can be carried out by either direct suture or choledochoduodenostomy. Late stricture from ischaemia can occur weeks to months postoperatively, and this too requires drainage by choledochoenterostomy. Other complications include wound sepsis, adhesion intestinal obstruction, and prolonged ileus. The long-term requirements for insulin therapy and exocrine pancreatic replacement need to be carefully assessed.

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Splenectomy

45

Peter Borzi

The spleen provides important phagocytic clearance of senescent red cells and platelets, as well as entrapment and destruction of encapsulated bacteria such as *Streptococcus pneumoniae* and *Neisseria meningitidis*. This normal clearing mechanism becomes pathological when target red cells or platelets are seen as abnormal by the spleen or when increasing splenic mass sequesters or destroys otherwise normal cells. In childhood, most commonly, congenital spherocytosis, sickle cell disease, or autoimmune haemolytic anaemia can precipitate acute haemolytic crises, profound anaemia, and jaundice, as well as a chronic compensated anaemia from splenic hyperfunction.

45.1 Presentation of Splenic Disorders and Splenomegaly

Idiopathic thrombocytopenic purpura (ITP) secondary to splenic platelet destruction manifests as petechiae, bruising, and, rarely, nonactive overt bleeding (e.g., mucous membrane bleeding). Giant splenomegaly can produce combinations of thrombocytopenia, anaemia, and possibly leucopenia. Infiltrative processes such as lipid deposition disorders (e.g., Gaucher's disease) and malignancy, such as Hodgkin disease or juvenile chronic myeloid leukaemia and portal hypertension, can create enormous splenic enlargement, hyperfunction, and risk of rupture from minor trauma.

45.2 Consequences of Splenectomy

Splenectomy will reverse some of the above adverse consequences, but at the cost of thrombocytosis and increased senescent red cells (Howell-Jolly bodies on blood smear). With loss of efficient clearance of encapsulated bacteria comes an increased risk of overwhelming postsplenectomy infection (OPSI). In general, OPSI is a small risk (4.5% lifelong) when compared with the pathological effects of the spleen. OPSI is seen with greater frequency in the presence of a dysfunctional reticuloendothelial system (e.g., immunosuppression), malignancy, or radiation therapy, or in the very young (<4 years). Caution is recommended when considering splenectomy in the presence of these clinical states.

Preoperative immunization is obligatory in elective splenectomy. Commercial vaccinations with *extended serotype coverage* are available for pneumococcus, meningococcus, and *Haemophilus influenza*; ideally, they should be administered at least 3–4 weeks before surgery. Postoperative vaccination may be required in the acute setting, and either way, seroconversion can never be assumed. The use of appropriate antibiotics in the setting of infection with asplenia is to be stressed to the patient and parents or other caregivers. Splenic preservation should be the surgical objective when possible, such as when exploration is needed for uncontrolled splenic bleeding secondary to abdominal trauma. A referral to a national spleen register may be an appropriate for longterm follow up and updates for vaccinations and information e.g., Spleen Australia.

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45.3 Operative Techniques

Splenectomy traditionally was carried out as an open procedure. Laparoscopic splenectomy has become the standard for normal-size or moderately enlarged spleens and when concomitant cholecystectomy is indicated. With experience, giant spleens can be removed as a completely laparoscopic procedure, or as laparoscopic-assisted or hand-assisted laparoscopic splenectomy (HALS). Laparoscopic partial splenectomy is an increasingly popular technique for splenic preservation in the clinical settings of some benign vascular tumours, splenic cysts, splenic infarcts, trauma, and even hereditary spherocytosis. Single-incision laparoscopic splenectomy (SILS) remains an emerging alternative approach that has not proven to be more advantageous than laparoscopic splenectomy, except for its cosmetic advantage.

45.3.1 Positioning and Incisions

Splenectomy can be performed in the supine, semilateral, or lateral positions. Except in the presence of enormous splenomegaly, when there is a need to access the splenic hilum early in dissection, it is preferable to utilize gravity and positioning to enhance exposure by encouraging the stomach, liver, colon, and small bowel to fall away from the operative site. The semilateral and lateral positions with the patient supported and strapped to the operating table allow reasonable intraperitoneal positioning, including reverse Trendelenburg during laparoscopic splenectomy (Fig. 45.1). Intraoperative orogastric decompression is imperative to enhance exposure, especially at the upper short gastric vessels.

For elective splenectomy, a transverse subcostal incision with possible extension across the midline allows good exposure and the ability to retrieve the spleen after mobilization and devascularization. Port sites for laparoscopy are somewhat variable; Fig. 45.1 shows a commonly used strategy. The dorsal and epigastric ports may change depending on the size of the spleen. The need for other procedures such as cholecystectomy requires additional port placements. The epigastric or right-sided port may be extended to allow the insertion of a hand-assisted device for HALS.

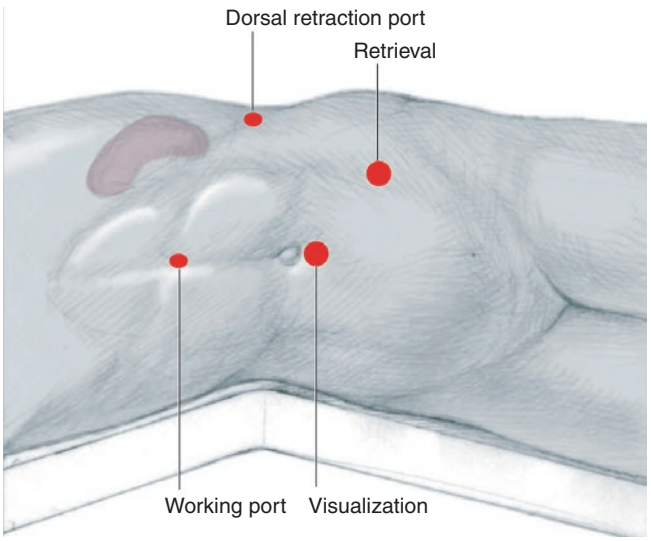

Fig. 45.1 Positioning for splenectomy, with port sites commonly used for laparoscopy

45.3.2 Exposure and Mobilization

Knowledge of the anatomical attachments and vascular anatomy is essential for completing an uncomplicated splenectomy (Fig. 45.2). In particular, the splenocolic, splenorenal, and phrenosplenic ligaments maintain splenic attachment to the posterior abdominal wall and diaphragm. The gastrosplenic ligament above and the gastrocolic ligament below cover the omental bursa, the distal pancreas, and splenic vessels.

Initial release of the phrenocolic and splenocolic ligaments allow the splenic flexure and adjacent transverse colon to fall away from the inferior splenic pole. Extending this dissection superomedially, one may encounter the left gastroepiploic artery before entering the lesser sac. Division of the lower short gastric vessels creates a window onto the distal pancreas and allows isolation of the splenic artery and vein in its more accessible and tortuous distal segment before entering the splenic hilum. These vessels can be tied or clipped in continuity more proximally if hilar dissection seems difficult, particularly when dealing with an enormous spleen. Continued release of the upper short gastrics completes the anterior dissection.

Medial retraction of the spleen gives good exposure to the splenorenal ligament and the embedded tail of the pancreas. This posterior mobilization is continued with release of the phrenosplenic ligament. In this stage of mobilization, the spleen can be retrieved from the splenic bed onto the abdominal wall for closer inspection of the pancreatic tail and vascular pedicle.

With laparoscopic splenectomy, splenic mobilization usually progresses with a similar anatomical dissection. Following complete release of the short gastric vessels, the dissection returns to detachment of the splenorenal ligament inferiorly only, allowing access to the retropancreatic and hilar plane passing superiorly above the pedicle through the posterior peritoneum of the upper lesser sac. The splenic pedicle is then isolated on all sides. Leaving the upper splenorenal and phrenosplenic ligaments intact gives better stability and controlled exposure to the pedicle than a floppy and totally mobilized spleen. Early ligation of the splenic vessels above the pancreas is preferable if hilar exposure difficulties are expected, but it is generally not mandatory.

Animal studies have found that about 25% of viable spleen is required for adequate splenic function. Partial splenectomy will then involve more careful consideration of the recognized vascular pedicles if splenic polar preservation is to be considered. Depending on the position of the lesion to be excised within the spleen, selective dissection

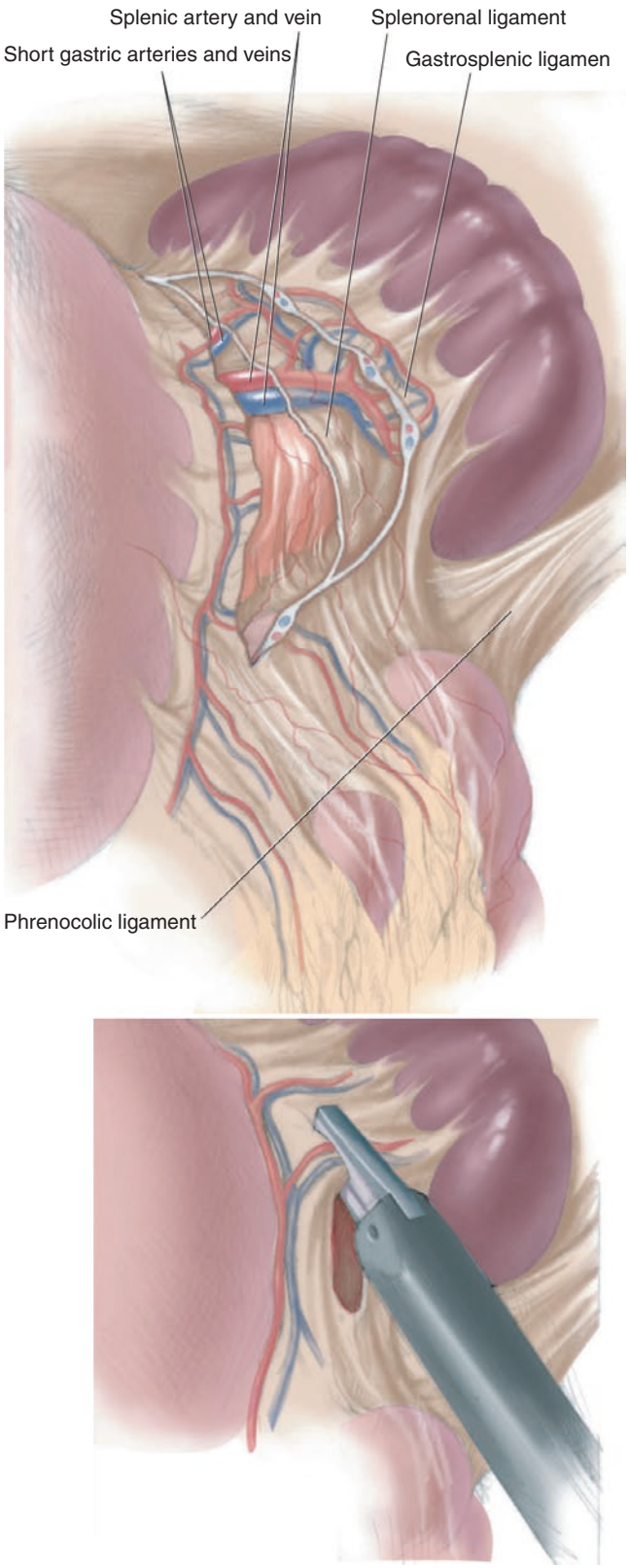

Fig. 45.2 Anatomical attachments and vascular anatomy. *Inset* shows division of the lower short gastric vessels

near the hilum of the relevant branches of the splenic artery and then the splenic vein is needed. Transection of the devascularised parenchyma is completed with the use of an Ultracision® harmonic scalpel or electrosurgical bipolar vessel sealing device (LigaSure™). To prevent postoperative haemorrhage, leaving a cuff of devascularised rim of spleen or the use of topical haemostatic agents may be considered.

With the spleen (or part of it) devascularised and completely freed from all attachments, retrieval via an Endobag™ allows controlled morcellation with a mechanical device or sponge-holding forceps, preventing intraperitoneal spillage. Sometimes it may be necessary to keep the spleen architecture intact for pathological analysis. A low Pfannenstiel incision or extension of the retrieval port site to contain a retrieval bag may be necessary.

45.3.3 Accessory Spleens

The presence of accessory spleens at the time of splenectomy has been variably reported to occur in 10–31% of patients. More than 90% occur in the supracolic compartment in and around the splenic hilum (particularly along the line of the distal splenic artery and adjacent hilum), the gastrosplenic and splenorenal ligaments, and adjacent to the greater omentum (Fig. 45.3). After entering the abdominal cavity, these sites of accessory spleniculi must be sought out early. At splenic mobilization, the surgeon needs to be observant and remove any accessory splenic tissue encountered. Doing so will avoid possible recurrent haemolysis or thrombocytopenia resulting from hypertrophy of the spleniculi. In most patients (63.3%), only one accessory spleen is present, but 17% have three or more.

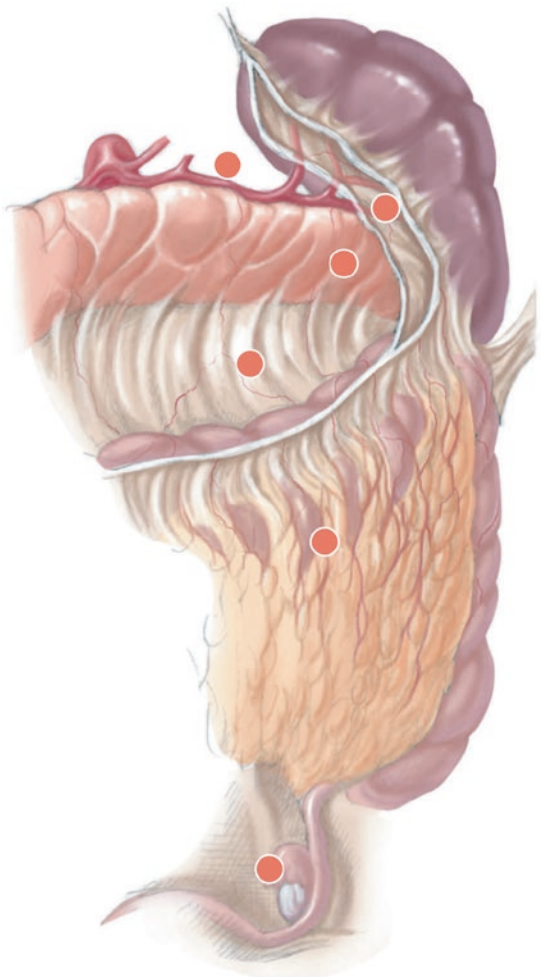

Fig. 45.3 Common locations of accessory spleens (*orange circles*)

45.3.4 Traumatic Splenic Injury

In the presence of a traumatic splenic injury with continuing uncontrolled haemorrhage, rapid mobilization and isolation of the pedicle is the priority, to reduce continued blood loss and maximize the chance of splenic preservation. The incision may need to be modified depending the possibility of other visceral injuries. After rapid entry into the peritoneal cavity, large clots and free blood are removed and air is introduced into the left subphrenic space, from which often the spleen becomes more mobile, descending closer to the wound. The splenic mass and fossa is packed with surgical sponges. With medial retraction of this splenic mass, lateral incision of the splenorenal and upper phrenosplenic ligaments superiorly and medially (Fig. 45.4) allows greater mobility. Delivery of the inferior splenic pole into the wound can achieve progressive retrieval of the spleen onto the anterior abdominal wall, taking care not to avulse the upper short gastric vessels or damage the splenic flexure. In this position, the potential for splenic salvage can be assessed. Polar devascularization may be required, or direct suture with haemostatic gauze and/or omental plug may be necessary to preserve the integrity of functioning splenic tissue.

45.3.5 Completing Splenic Retrieval

Early splenic devascularization by splenic vessel ligation through the lesser sac is generally considered a safe practice for elective splenectomy and giant splenomegaly, or where dense diaphragmatic and gastric adherence to the spleen is expected. Alternatively, initial anterior and posterior ligamentous splenic release and subsequent pedicle ligation can be applied also in the setting of elective splenectomy (Fig. 45.5); it is definitely indicated in the presence of splenic trauma. Laparoscopic splenectomy can be performed by either of the above methods, with the polar vessels divided individually with clips or suture ligation or with a linear stapling device after initial mobilization of the tail of the pancreas. Care must be taken to avoid splenosis during splenic retrieval, particularly in the presence of haemolytic disorders. Laparoscopically, the mobilized and devascularized spleen is placed in a sturdy, impermeable bag with the open end delivered through the retrieval port site. The wound edges are protected while the specimen is fragmented and removed. Devices such as Endocatch II (Tyco Healthcare, Mansfield, MA; Covidien, 555 Long Wharf Drive New Haven, CT, USA) allow somewhat easier insertion of the spleen into the retrieval pouch.

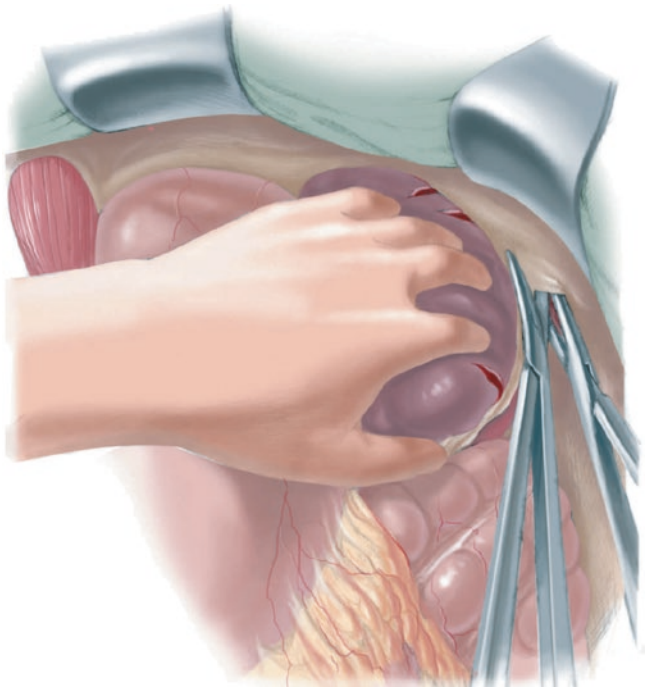

Fig. 45.4 Incision of the splenorenal and upper phrenosplenic ligaments for mobilization in cases of traumatic splenic injury

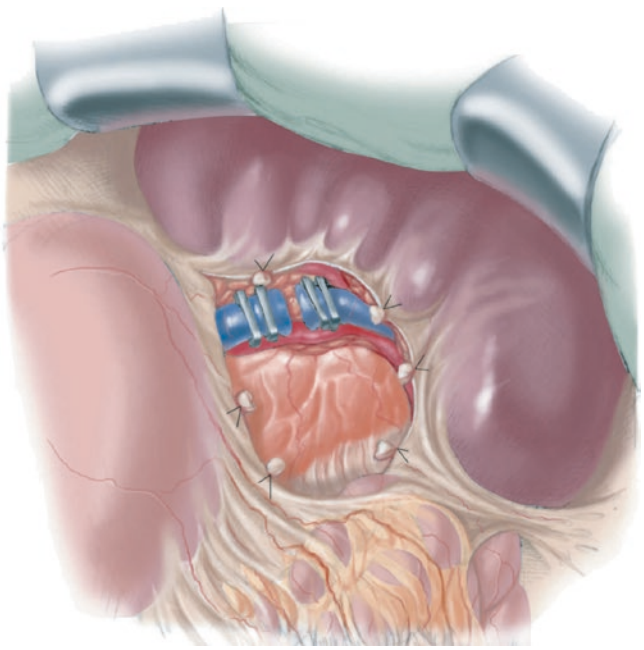

Fig. 45.5 Completed splenectomy, showing devascularization and ligamentous splenic release

45.4 Results and Complications

Postoperative paralytic ileus from the intraperitoneal and retroperitoneal dissection may be expected, depending on the size of the spleen, the circumstances at the time of removal, and the need for concomitant cholecystectomy. Nasogastric decompression during the perioperative period may need to be extended for 24–48 h after surgery. Persistent ileus and continuing vomiting raises the suspicion of a possible pancreatic injury. Slow mobilization and inadequate diaphragmatic movement suggests left lower lobe pulmonary collapse. Subphrenic collections can similarly attract a sympathetic pleural effusion and pulmonary infection. Finally, injury to the stomach, although very uncommon, can evolve where extensive adhesions between the greater curve of the stomach and an inflammatory spleen are encountered.

OPSI should be minimized by preoperative immunization and intraoperative and continued postoperative antibiotic prophylaxis for a minimum of 6–12 months. In the long term, any clinical infection or procedures that increase the risk of potential sepsis, such as dental, gastrointestinal, or genitourinary surgery, require antibiotic prophylaxis for the duration of the infection or operation and the immediate convalescence.

Rebound thrombocytosis is generally not of clinical concern in children with preoperative ITP. In more than 90% of cases, complete reversal of symptomatic thrombocytopenia would be expected, with return of the platelet count to the normal range or above. In the presence of more nonspecific haemolytic disorders, 60–70% will recover, but most of the rest will have some transient recovery of platelet count but a bleeding tendency that remains subclinical.

Clinically significant recurrent thrombocytopenia needs further evaluation for missed accessory spleens. Contrast CT scans or red-cell-labelled isotope scans are suggested, with consideration being given to a second-look laparotomy or laparoscopy.

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Portal Hypertension

46

Mark Davenport

Portal hypertension (PHT) has no formal definition in children but can arbitrarily be considered as a portomesenteric pressure greater than 10 mmHg. Sustained PHT leads to the development of varices at the sites of the portosystemic watershed, together with splenomegaly. The principal site for the development of varices is the oesophagogastric watershed, but haemorrhoids and rectal varices may become apparent even in children and adolescents. Occasionally, ectopic varices develop around surgical anastomoses such as a Roux loop and can be a cause of obscure gastrointestinal bleeding. The third site of venous overlap is the retroperitoneum and anterior abdominal wall. Spontaneous shunts may develop in the retroperitoneum; they seldom cause clinical symptoms, but they may be problematic at surgery. The most obvious sign of a significant shunt in the anterior abdominal wall is the well-known *caput medusae* sign, comprising dilated periumbilical veins radiating from the umbilicus. These develop along the obliterated umbilical vein and arise most commonly in cases of long-standing cirrhosis.

46.1 Causes of Portal Hypertension in Children

There are three main causes of significant PHT in children, listed here in descending order of frequency:

- Portal vein obstruction (PVO) (usually synonymous with thrombosis)
- Liver cirrhosis
- Liver fibrosis

Figure 46.1 expands this list and provides a structure by dividing these causes into three approximate levels of venous obstruction: the prehepatic, hepatic, and posthepatic levels. The hepatic level is further divided into cirrhotic and fibrotic, depending largely on the preservation of hepatocyte functionality. The posthepatic level is divided into obstruction at the level of the hepatic veins (Budd-Chiari syndrome) or at the level of the hepatic venules (veno-occlusive disease). Rare causes of PHT such as arterioportal hypertension will not be considered further.

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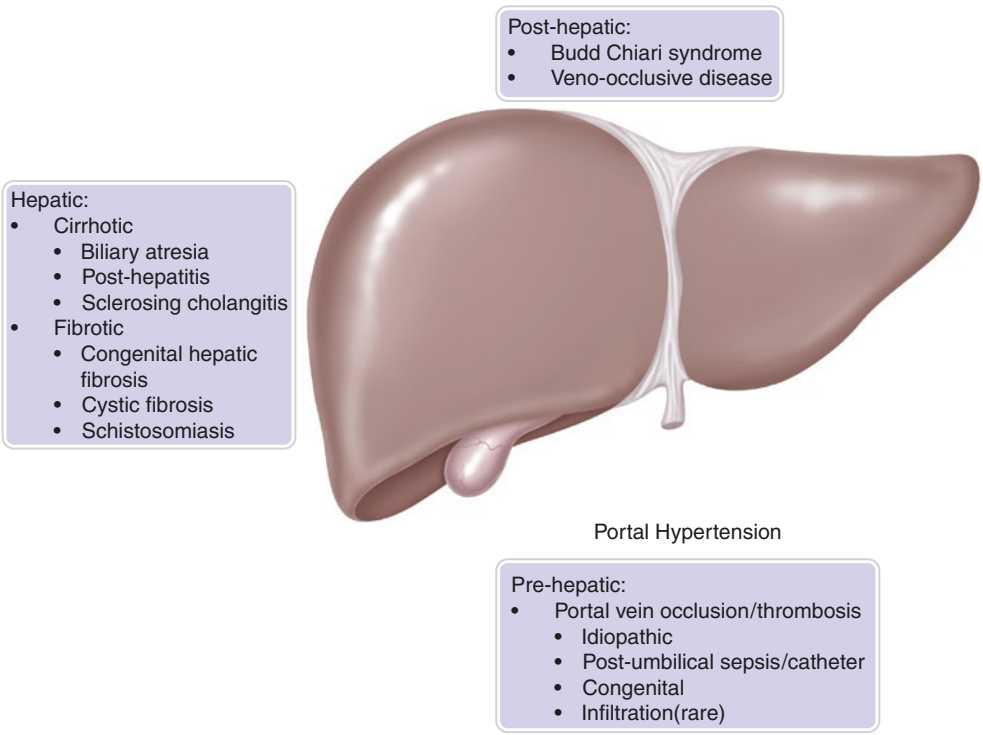

Fig. 46.1 Classification of portal hypertension in children

46.1.1 Portal Vein Thrombosis

Portal vein thrombosis is the commonest single cause of significant PHT in children and is easily distinguished clinically from cirrhotic liver disease, the next most common cause. It is relatively common in the general population (perhaps surprisingly so), with some post-mortem studies suggesting that PVO is present in about 1%. More recent studies based on screening ultrasonography suggest that the true incidence may be even higher, at about 5%.

The cause of PVO is not clear in most cases (idiopathic), but the presence of other clinical features may suggest some aetiologic factors. About 20% will have other extra-abdominal abnormalities such as skeletal defects or cardiac and craniofacial anomalies, suggesting that the PVO is also congenital in origin. A history of umbilical vein catheterisation may be seen in 10–20% of cases, but to put this into context, fewer than 1% of episodes of umbilical vein catheterisation cause PVO. Other neonatal events such as sepsis and dehydration also appear more frequently than expected.

Intra-abdominal sepsis in older children (due to appendicitis, for instance) may cause showers of septic emboli to end up in the portal vein (portal pyaemia) and cause thrombosis (<10% of most series). Less common is direct portal vein damage—possibly iatrogenic or traumatic—or infiltration by tumour (such as hepatoblastoma). PVO also may be seen as a complication after splenectomy, owing to clot propagation along the splenic vein. This risk now appears to be more common following laparoscopic splenectomy than it was when splenectomy was performed as an open technique.

A number of inherited defects of thrombolysis also predispose to PVO, including Factor V Leiden mutation, deficiencies of protein C and possibly protein S, and anti-thrombin III deficiency.

The thrombotic segment can be discrete and limited or much more extensive, involving the intrahepatic portal venous system or the entire porto-mesenteric venous system. This element needs to be defined by MR or CT venography prior to consideration of shunt surgery. The cavernoma or cavernomatous transformation commonly seen radiologically represents collateralisation and can restore hepatopetal venous flow to some extent.

46.1.2 Hepatic Fibrosis and Cirrhosis

Fibrosis is not a normal finding in the liver, and its formation may be prompted by a number of disease processes. Congenital hepatic fibrosis refers to a genetic condition characterised by early fibrosis, typically in association with fibrocystic disease of the kidneys. PHT is therefore almost invariable in these patients, although they usually do not present until later in childhood.

Extensive bridging fibrosis, together with attempts at hepatocyte regeneration, may be further defined as cirrhosis, which is seen in many conditions of childhood. The commonest patients are those born with biliary atresia and other survivors of the Kasai operation. Other causes include

postinfective hepatitis, α-1-antitrypsin deficiency, cystic fibrosis, and in older children, Wilson's disease.

46.1.3 Budd-Chiari Syndrome

This syndrome was named for a nineteenth century British physician (George Budd, 1808–1882), who worked at Kings College Hospital, and an Austrian pathologist (Hans Chiari, 1851–1916). The name is used to describe the clinical features surrounding hepatic vein and outlet occlusion. It is rare in children, with some cases due to a congenital web but most occurring as a result of abnormal thrombogenesis, typically related to malignancy. Veno-occlusive disease, a condition affecting the smaller hepatic venules, arises secondarily to certain drugs used in anticancer therapy and is particularly common following stem cell transplantation.

In addition to the features of PHT, these patients have hepatomegaly due to venous congestion ultimately leading to fibrosis. There is also selective preservation and usually hypertrophy of the caudate lobe, which has venous drainage separate from the rest of the liver.

46.2 Features of Portal Hypertension in Children

46.2.1 Oesophageal Varices

Bleeding from oesophageal varices, often without warning, may be the commonest mode of presentation of PHT in a child. Typically, the underlying aetiologic factors of PVO, such as neonatal intensive care and umbilical vein insertion, have been forgotten or disregarded. In other conditions, such as older children with biliary atresia, the varices may have been detected with an endoscopic surveillance programme. Bleeding from a ruptured varix manifests itself as potentially exsanguinating haematemesis and melaena. Varices seldom present surreptitiously or silently with chronic gastrointestinal bleeding. Indeed, if the child is recognised to be anaemic, then this will be due to hypersplenism and should be accompanied by a low white cell count and thrombocytopaenia.

46.2.2 Splenomegaly

Sustained PHT with a duration of months to years will cause an increase in the size of the spleen. Initially, it may be just palpable in the left hypochondrium, but with progression its direction shifts to cross the midline towards the right lower quadrant. About 20% of patients with PVO will present with palpable splenomegaly and may have to endure a complex series of investigations in haematology departments to exclude haematological malignancies before PVO is considered seriously. The blood film features of hypersplenism should always be present if the size of the spleen has increased sufficiently to be palpable.

46.2.3 Ascites and Other Features

Obvious ascites usually implies a degree of liver involvement and cirrhosis and is a much more complex pathophysiological phenomenon than simply an elevated portal venous pressure. Therefore most children with clinically obvious ascites have cirrhosis or a Budd-Chiari syndrome and hepatomegaly rather than hepatic fibrosis or PVO.

There may be evidence of growth failure in children with cirrhosis, and this may also be an observation in some cases of PVO. In cirrhosis, growth failure is due to major changes in protein synthesis, abnormalities in hormonal control of metabolism and catabolism, and a diminution in caloric intake. In PVO, it may occur as a consequence of the smaller volume liver that is invariably seen, and possibly can be due to a degree of growth hormone resistance.

Hyperammoniaemia and hepatic encephalopathy are uncommon in children with cirrhosis unless there is decompensation (perhaps seen following a large gastrointestinal bleed). Hyperdynamic circulation and the formation of spider naevi again are features of cirrhosis rather than PVO.

46.3 Diagnosis of Portal Hypertension

The diagnostic strategy aims to uncover the cause and, where possible, discriminate between the two principal causes, PVO and cirrhosis. Diagnosis may involve MR and CT scanning supported by expert interpretation of colour Doppler ultrasonography. Liver biopsy is usually regarded as essential if cirrhosis is thought to be the cause, but it also may be warranted in PVO, particularly if shunt surgery is to be considered.

In idiopathic PVO, a search should be made for evidence of abnormal coagulation and a tendency to increased thrombogenesis as possible causes. Slight prolongation of the INR (International Normalised Ratio) is typical of those with PVO and need not have any prognostic significance. Levels of anti-thrombin III, protein C, and protein S should be assessed.

The main consequence of PVO is the development of varices, and full upper gastrointestinal endoscopy to the level of the third part of the duodenum should show the presence and grade of varices in the stomach and oesophagus, together with any stigmata suggesting imminent risk of bleeding. The rectum and distal sigmoid should also be endoscopically assessed for the presence of rectal varices and haemorrhoids. Hypersplenism may be defined as thrombocytopaenia ($<100 \times 10^9$/L), leucopaenia, and anaemia.

46.4 Treatment

The correct management of chronic PVO and PHT is controversial, and there is considerable overlap and variability in opinion. Essentially, management can be divided into medical management and surgical management. The indication as to when to proceed to surgical decompression of PHT can be difficult and many factors can have a bearing.

46.4.1 Treatment of Oesophagogastric Varices

The emergency management of bleeding varices—or much more usually, recently bled varices—involves the restoration of circulating blood volume and correction of any apparent coagulopathy and thrombocytopaenia (if $<50 \times 10^9$/L). A number of pharmacological measures can be started to try to reduce the underlying PHT, such as octreotide and terlipressin (Glypressin©). Torrential bleeding may warrant the use of a Sengstaken-Blakemore tube to tamponade the bleeding varix. This device should always be placed under general anaesthesia with a protected airway (Fig. 46.2). Ideally, the child should proceed to endoscopy and definitive variceal treatment such as banding or sclerotherapy.

There are two principle endoscopic methods of achieving definitive control of oesophageal varices: injection sclerotherapy (Fig. 46.3) and banding (Fig. 46.4). Both cause thrombosis and obliteration of variceal columns in the distal oseophagus.

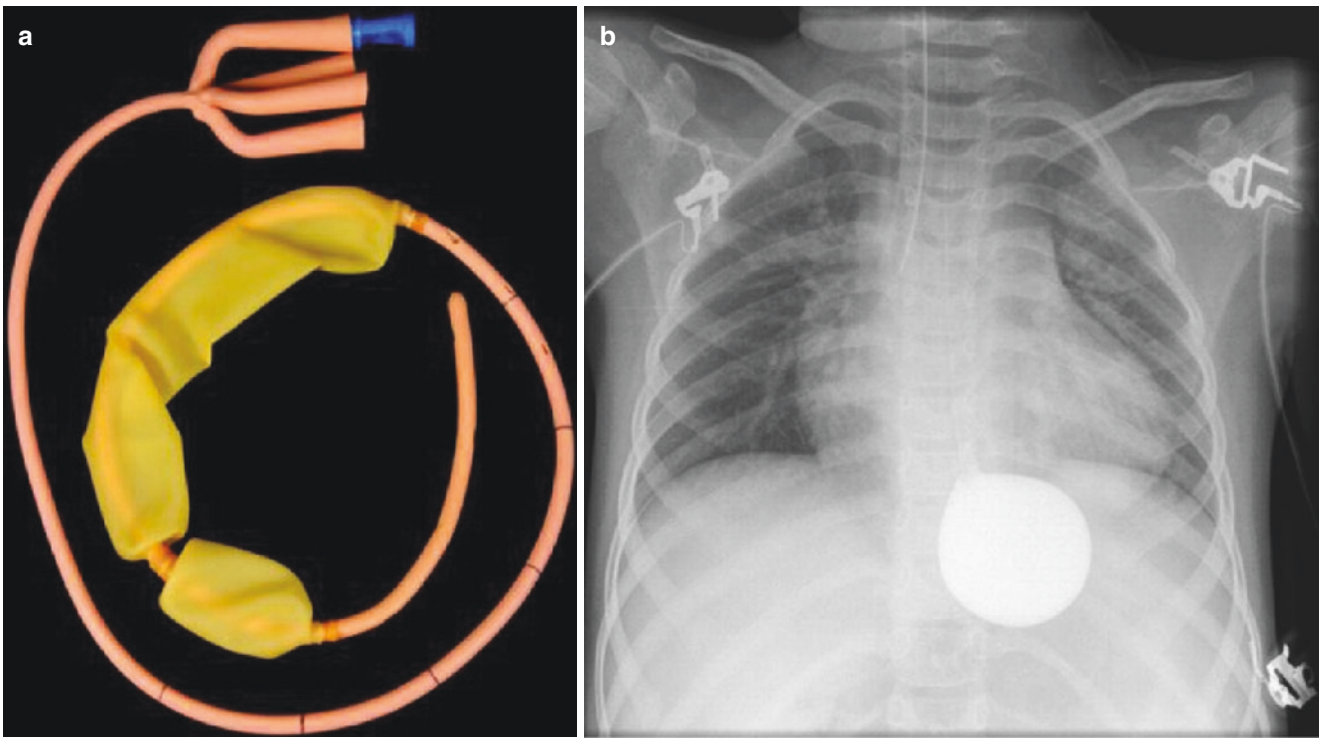

Fig. 46.2 Sengstaken-Blakemore tube (SBT). This device (**a**) has inflatable gastric and oesophageal balloons, together with two lumens, one proximal for aspiration of saliva in the pharynx and proximal oesophagus, and the other for gastric aspiration. The tube is inserted through the mouth until it is clearly in the stomach. The gastric balloon is inflated with diluted contrast (to check position) and is pulled back to impact the gastro-oesophageal junction (GOJ), thus exerting variceal compression (**b**). In children, it is usually not necessary to then inflate the oesophageal balloon. Pressure is usually maintained by bringing the tube to the side of the mouth and taping it to the cheek. If bleeding stops, then traction is maintained for about 24 h to allow correction of any coagulopathy, with relaxation for 5 min every 4–5 h to prevent mucosal ischaemia. (Reproduced from Jayakumar et al. [2015], with permission of Elsevier)

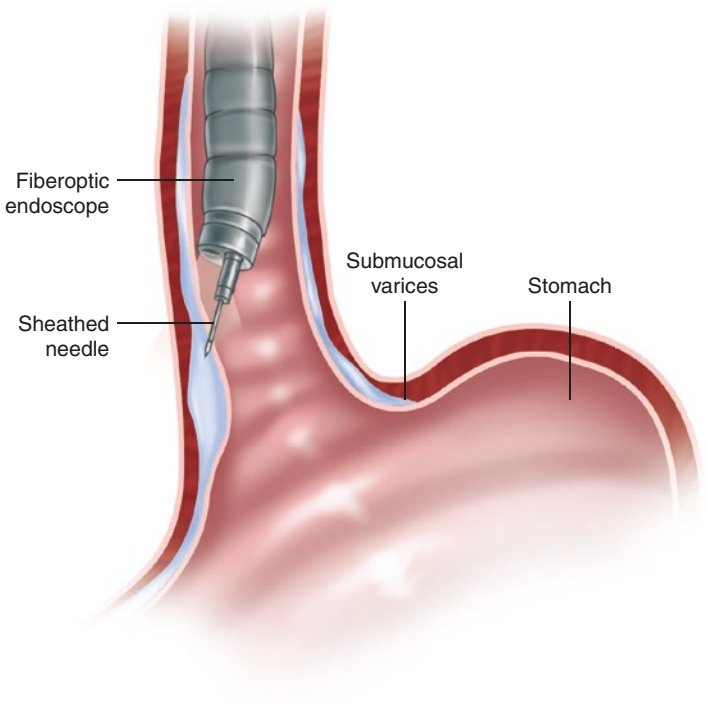

Fig. 46.3 Injection sclerotherapy.The column is visualised with an end-viewing endoscope of appropriate size. The primed injection needle is advanced into the lumen of the vein and 1 to 2 mL of sclerosant (e.g., ethanolamine oleate [2–5%]) is injected under vision. Ideally, the injection is intravariceal,butparavariceal injection will have the same effect.Continue the injection as the needle is withdrawn to reduce bleed back.Compression using the end of the endoscope should be performed for about 3 to 5 minutes, and then the lumen can be washed to repeat the injection elsewhere

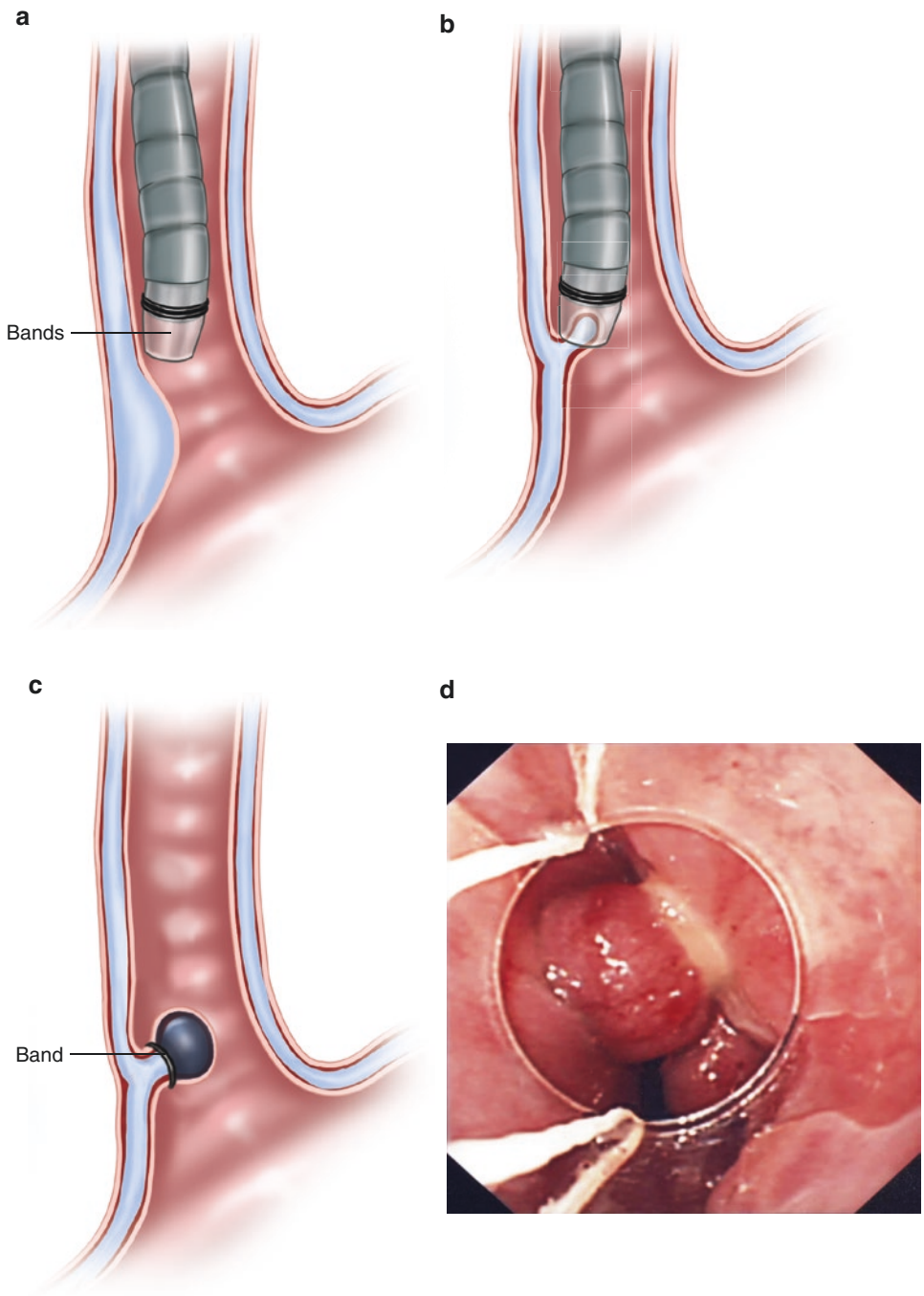

Fig. 46.4 Variceal banding. The bander consists of a transparent chamber with multipleprestretched bands and a stringlike system passing back through the working channel to allow the bands to be pulled off the end. Starting at the base of the most prominent varix (usually left-sided), the end is angled into the column (**a**) and suction is applied (**b**). The suction must be sufficiently vigorous to suck in the column and fill the chamber completely ("red-out"). The bander is then triggeredto throw the band tightly around the neck. Suction is stopped and the scope is withdrawn slightly to view the results. The appearance of the banded varix should resemble a mushroom (**c**, **d**). The next target is then visualised, a little further back and usually on the opposite wall. Avoid circumferential, sequential banding, which may cause so much mucosal injury that it results in a stenosis. Ultimately, there will be spontaneous necrosis, with passing of the band in about 5 to 7 days. There is always a small mucosal ulcer thatmay bleed, but bleeding is unusual

46.4.2 Surgical Treatment of Portal Hypertension: Meso-Rex Bypass

The advent of Meso-Rex Bypass (MRB) into the surgical repertoire during the 1990s reignited the debate as to the role of surgery early in the course of PVO. (It clearly has no role in cirrhosis.) The indication for MRB varies according to the institution. Some offer it to children with varices that have never bled, but this seems rather a low threshold for a complex operation that has the potential for complications and that usually leaves an obvious scar in the neck. We tend to reserve MRB for patients with recurrent bleeding episodes following a period of endoscopic treatment.

The aim of MRB is to circumvent the thrombosed portal vein by a venous conduit between the remnants of the left portal vein in the Rex fossa and typically the superior mesenteric vein (Fig. 46.5). Preoperative preparation is crucial in being able to identify a possible vein in the liver. The best investigation is the invasive retrograde hepatic venogram showing contrast reflux through the liver parenchyma. Current MR or CT imaging is not sensitive enough to be reliable. A history of an umbilical vein catheter is also a contraindication.

The success of the MRB procedure is probably related to the degree of atrophy or patency of its run-off into the intrahepatic portal venous system. The long-term success rate was only 60% in a French series of 43 children who underwent an attempted MRB.

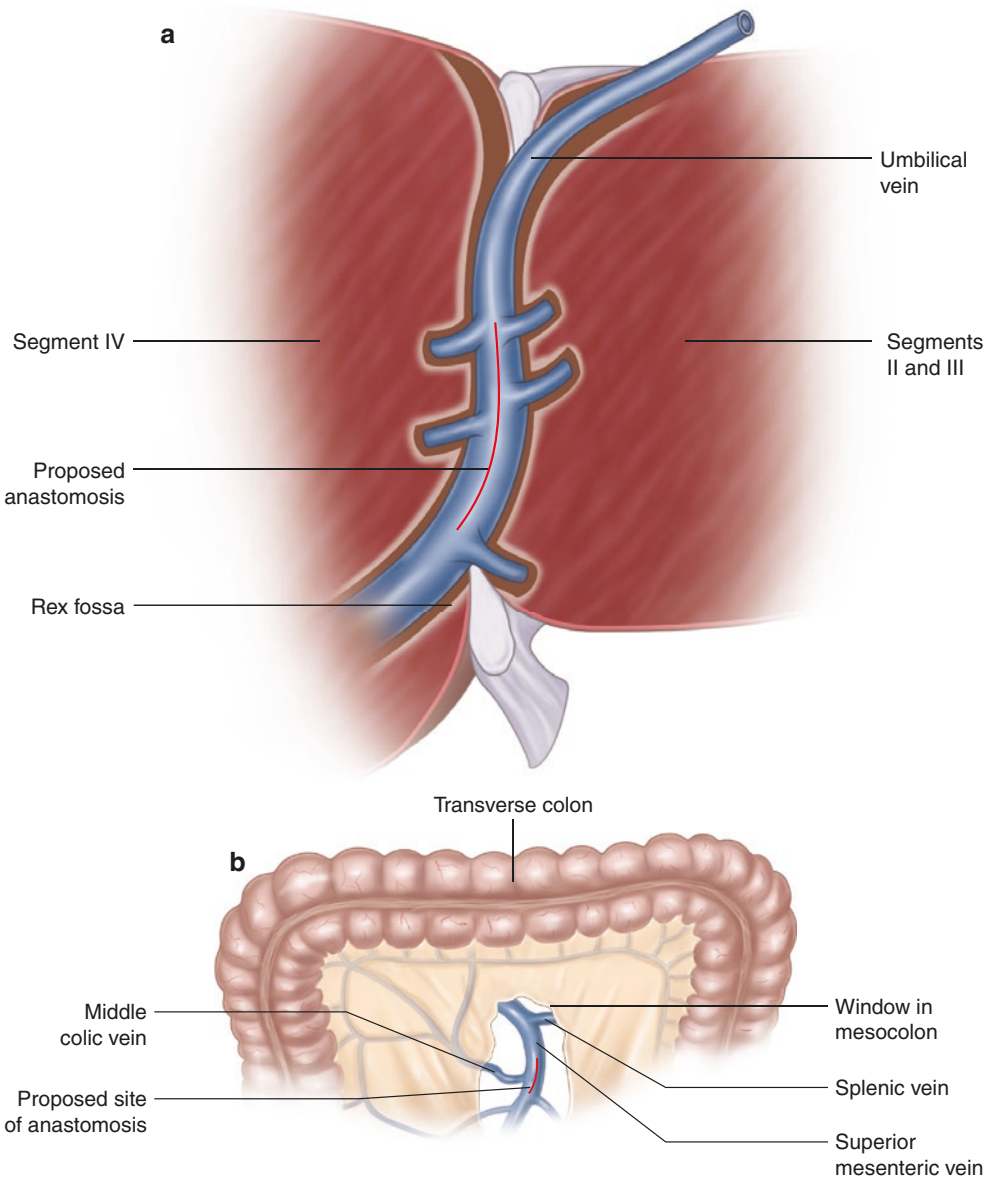

Fig. 46.5 Meso-Rex bypass. (**a**) The abdomen is explored through a transverse incision. The umbilical vein is dissected back to the liver. Sometimes the Rex fossa is open; otherwise the isthmus of liver tissue must be opened widely to expose the junction with the left portal vein (LPV). Typically there are paired venous tributaries into adjacent segments arising from the LPV. These are slung and controlled. The main vein is exposed and confirmed to have an adequate lumen and run-off. Sometimes direct on-table angiography can be performed at this point. (**b**) Attention is then shifted to the infracolic compartment. Follow the middle colic vein into the mesentery to its junction with the superior mesenteric vein (SMV). Trace this vein until the splenic vein is visualised joining from the left. Sling and control an appropriate segment of vein as the takeoff for the conduit. Measure the length required using a retrogastric route superficial to the pancreas. The left internal jugular vein is the commonest choice for the venous conduit. Its patency and that of its fellow on the other side should have been confirmed by preoperative ultrasound. Sacrifice does not usually cause any measureable ill effects on venous drainage, but sometimes transient hemifacial oedema can be seen. The vein lies behind the sternomastoid, which can be preserved and retracted through a longitudinal cervical incision. Beware of cranial nerve XI (accessory nerve) coming from the posterior aspect of this muscle and crossing the posterior triangle. Mobilise the vein from behind the clavicle to the mastoid, typically dividing the common facial vein at the midway point. Preserve the vagus nerve (X) at the back of the carotid sheath and beware of the thoracic duct joining distally. Oversew either end carefully, leaving a long suture end to avoid vein retraction and bleeding. The proximal vein, washed in heparinised saline, is anastomosed to the preselected SMV using (typically) 6/0 Prolene, using running or interrupted sutures or a combination on the anterior and posterior walls. If using running sutures, avoid narrowing and leave a "growth factor". The conduit is placed behind the stomach and in front of the pancreas in as direct a route as possible. The distal vein is sutured to the LPV in similar fashion. Avoid air emboli and twisting of the graft. Using Doppler ultrasound intraoperatively to confirm and measure flow is important. Direct measurement of mesenteric pressure should also demonstrate a significant fall (although perhaps not to the same extent as with other shunts). The abdomen is closed with drains into the various recesses. These drains should be removed only when the child is on a normal diet, as there is a risk of chylous ascites from transection of abdominal lymphatics

46.4.3 Surgical Treatment: Conventional Portosystemic Shunts

Conventional portosystemic shunts still have a role, particularly when patients have limited access to specialist medical care and cannot rely upon rapid endoscopy and resuscitation. For these children, secure long-term lowering of portal and mesenteric pressure is essential. Encephalopathy is possible, particularly during the adult years, although the true risk remains entirely unknown. Some authorities, such as Frédéric Gauthier from Bicêtre Hospital, Paris, would say the risk is entirely negligible.

If MRB appears inappropriate, alternatives must be found for those with PVO that is unresponsive to endoscopic or medical therapy. A mesocaval shunt (Fig. 46.6) is an excellent option, as the cava is always uninvolved in the thrombotic process, but it requires a conduit (usually the internal jugular vein). In our institution, we prefer the mesocaval shunt, but some other units prefer to use the left renal vein and splenic vein in various ways (Fig. 46.7). The advantage is that with mobilisation, no conduit is required. The disadvantage is that these may have a higher thrombosis rate.

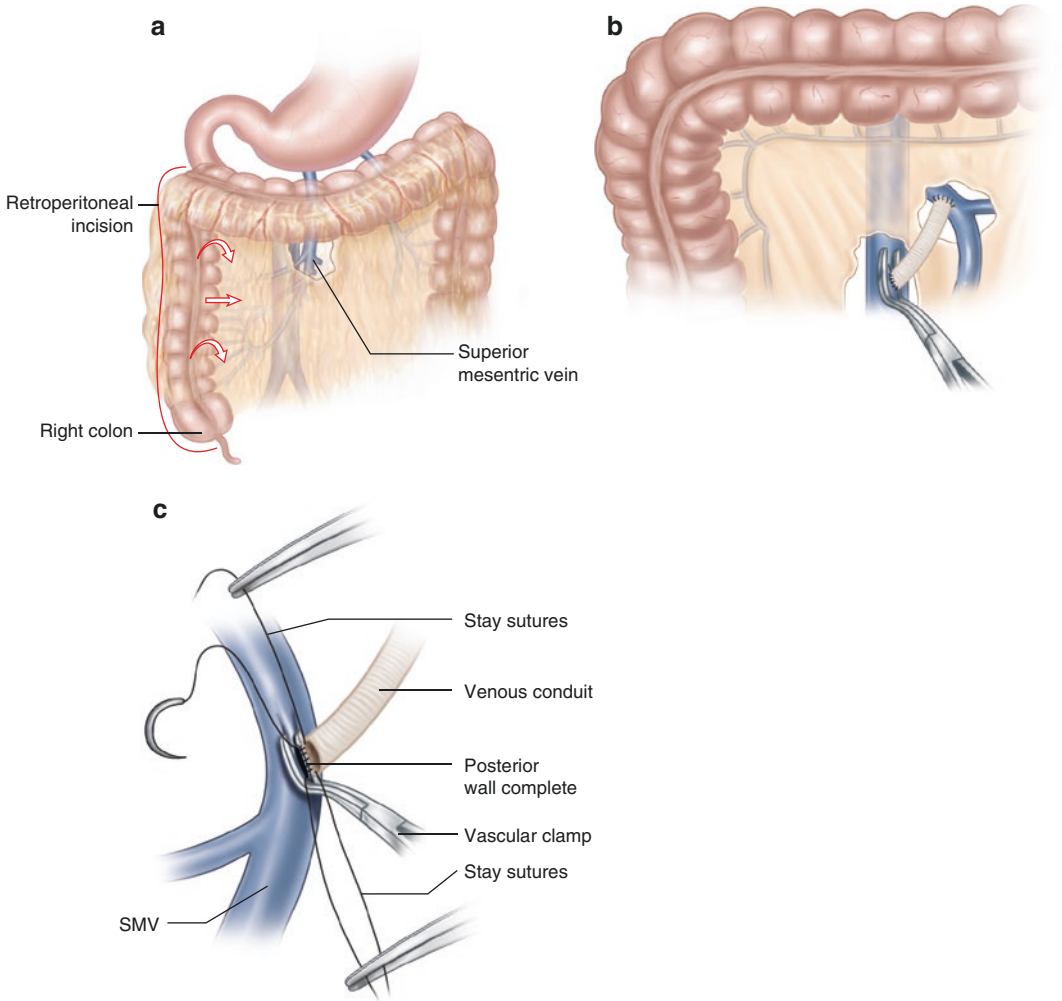

Fig. 46.6 Mesocaval shunt. (**a**) The abdomen is explored through a transverse incision. As before, the middle colic vein is the key to identify the SMV lying within the mesentery. Sling and control an appropriate segment of vein as the takeoff for the shunt. (**b**) The vena cava is approached from the right (**a**). The hepatic flexure and colon are dissected off the right kidney. Sling and control an appropriate segment of the infrarenal cava. Make a tunnel from the back of the SMV through the mesentery directly to the cava, avoiding the third part of the duodenum. Measure the length of conduit required, which should be shorter than a comparable MRB. (**c**) Harvest the left internal jugular vein as before. The appropriate anterior part of the inferior vena cava (IVC) is controlled with a side-biting Cooley vascular clamp (**b**). The distal part of the vein conduit, flushed in heparinised saline, is then anastomosed using a 6/0 Prolene suture. Typically an ellipse of vein wall is excised to open this anastomosis. The side of the preselected SMV is anastomosed with the proximal end of the vein conduit using (typically) a 6/0 Prolene. Again, whether this is a running or interrupted suture is irrelevant. Direct measurement of mesenteric pressure with and without shunt occlusion should show a significant fall. The abdomen is closed with drains into the various recesses

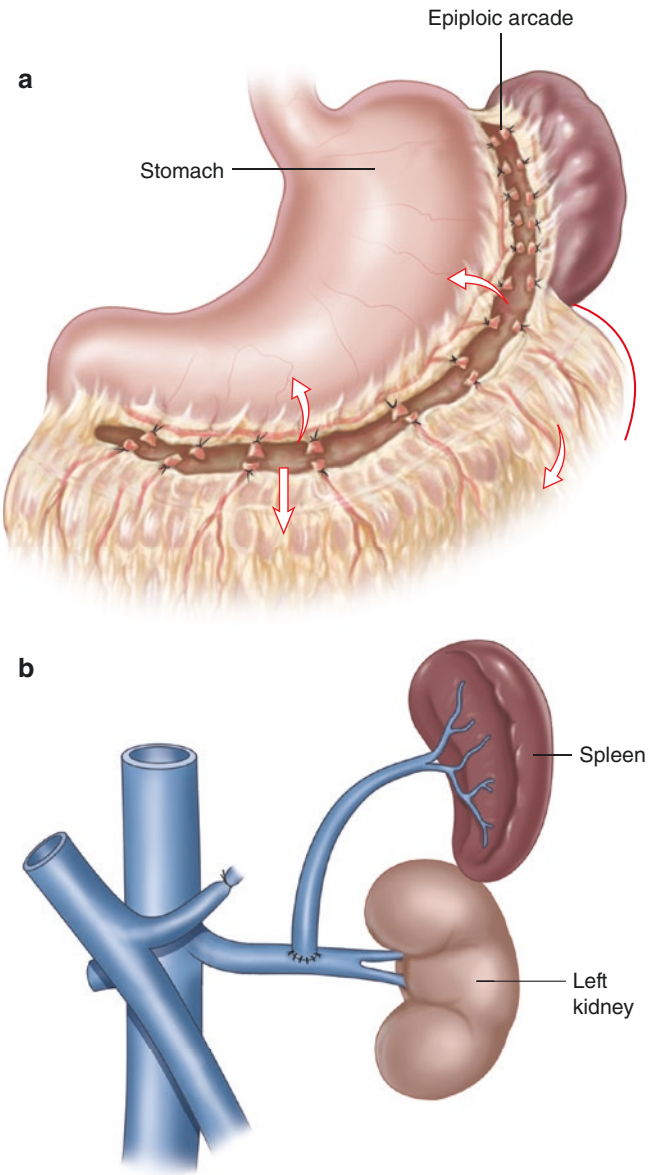

Fig. 46.7 Alternative shunts. The relevant veins can be accessed through a paragastric window (**a**) into the lesser sac or through a dissection on the left side of the mesocolic mesentery. Either approach allows visualisation of the pancreas and the intimate splenic vein with the retroperitoneal renal vein nearby. Mobilisation of the left renal vein (usually with ligation of the adrenal vein) and separation of the splenic vein from its pancreatic bed can allow enough length to perform a side-to-side anastomosis with splenic preservation. Alternatively, the spleen can be removed and the now-redundant splenic vein can be anastomosed to the left renal vein. Finally, the splenic vein can be ligated near to its junction with the SMV and the now-distal part is anastomosed to the left renal vein (**b**). When combined with a portal-azygous venous disconnection, this latter technique was the basis of the Warren shunt, though it was little used in children

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Liver Transplantation

47

Alastair J. W. Millar

Until relatively recently, treatment options for the debilitating effects of liver disease in children were limited to supportive therapy and immunosuppressive agents. With surgical conditions such as biliary atresia, some success had been achieved with timely diagnosis and early surgical intervention, but in many cases there was inevitable progression of liver disease even with expert management. Liver transplantation offered the only chance of a cure for these unfortunate children. Early experimental studies were conducted in the 1950s and early 1960s in Boston and Denver. The first attempt at human transplantation was performed in 1963, but it was not until 5 years later that long-term success was achieved. In 1983, the National Institutes of Health Consensus Development Conference in the USA accepted liver transplantation in children and adults as deserving a wider clinical application. Advances in organ procurement and preservation, surgical technique (particularly the use of size reduction of the graft, which to some extent alleviated the graft shortage for infants and children), anaesthetic management, and preoperative and postoperative care, as well as refinements in immunosuppression over the past three decades have resulted in a much improved outcome with an ever-increasing list of indications being identified (Table 47.1). The current expected 5-year survival is now greater than 85%, and excellent quality of life is the rule rather than the exception. The longest survivor is well more than 45 years after transplantation. Anxieties remain over organ donor scarcity, long-term side effects of the immunosuppressive therapy, financial implications, and some ethical issues. The focus of attention has now shifted from an initial target of early post-transplant survival to quality of life in the long term. The transformation of a miserable, jaundiced invalid into an active, healthy child remains a powerful stimulant to paediatricians and transplant surgeons alike to actively pursue liver transplantation for these children.

Shortage of donor organs has been tackled in an imaginative way, initially with size reduction of the donor liver for transplantation, and then with splitting the liver into two functioning units for two recipients, as well as using segments of liver procured from living donors and, more recently, the use of donors after cardiac death.

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Table 47.1 Indications for which liver transplantation has been performed in children

Category	Specific indications
Metabolic (inborn errors of metabolism)	Alpha-1 antitrypsin
	Tyrosinaemia
	Glycogen storage disease type III and IV
	Wilson's disease
	Perinatal haemochromatosis
	Hypercholesterolaemia
	Cystic fibrosis
	Hyperoxaluria (+ renal transplant)
	Haemophilia A + B
	Protein C deficiency
	Crigler-Najjar (type 1) syndrome
	Urea cycle disorders
Acute and chronic hepatitis	Fulminant hepatic failure (viral, toxin- or drug-induced)
	Chronic hepatitis (B, C, etc.; toxin, autoimmune, idiopathic)
Intrahepatic cholestasis	Neonatal hepatitis
	Alagille syndrome
	Biliary hypoplasia
	Familial cholestasis
Obstructive biliary tract disease	Biliary atresia
	Choledochal cyst with cirrhosis
	Alagille syndrome
	Sclerosing cholangitis
Neoplasia	Hepatoblastoma
	Hepatocellular carcinoma
	Sarcoma
	Haemangioendothelioma
Miscellaneous	Cryptogenic cirrhosis
	Congenital hepatic fibrosis
	Caroli's disease
	Budd-Chiari syndrome
	Cirrhosis from prolonged parenteral nutrition

47.1 Indications

Liver disease has been generally underestimated as a cause of death in children, probably because many liver conditions in children have led to rapid deterioration. Almost all forms of liver disease in children can be complicated by hepatocellular failure. These include acute and subacute liver failure from metabolic, toxic, or viral insults and chronic parenchymal disease of varying causes, the most common of which are biliary atresia, biliary hypoplasia, chronic active hepatitis, post viral hepatitis, and some metabolic diseases. Some metabolic diseases are expressed in hepatocellular disease, whereas others, such as tyrosinaemia, Wilson's disease, hyperglycoproteinaemia, some glycogen storage diseases, and hyperoxaluria, may have more widespread systemic effects.

47.2 Assessment

In general, liver transplantation should be considered as a therapeutic option in all cases of chronic liver disease before the condition of end-stage liver disease is realised. Doing so allows for a thorough assessment of the child and the family (Table 47.2) and gives time for full and frank discussions of treatment options, which can prepare all concerned as much as possible for the transplant procedure and its aftermath. The family's capacity to sustain long-term compliance after transplantation, so crucial to the success of this endeavour, must be assessed. Transplant candidacy inevitably results in enormous emotional stress for parents while waiting for the appropriate donor, as their child's condition often deteriorates from nutritional depletion, bleeding from gastro-oesophageal varices, and episodes of systemic infection. Family life may become disrupted. Those that live far from the transplant centre may feel constrained to relocate, which in itself can cause major domestic distress to families. The value of a sympathetic social worker to provide support cannot be overstated.

There are indeed few reasons for refusal for transplantation. These include uncontrolled systemic bacterial, viral, or fungal infections; malignancy outside the liver; fixed cyanotic pulmonary arteriovenous shunting with pulmonary hypertension; active chronic hepatitis B; HIV/AIDS; or other major cardiorespiratory, neurologic, or renal disease that would be incompatible with quality long-term survival. To a certain extent, these are all relative contraindications. Psychosocial factors also may be a reason for refusal. Parental substance abuse, severe psychiatric problems, and poor preoperative compliance with therapy are factors that must be carefully examined. Compliance is more difficult to predict in children with acute hepatic failure, as the time from presentation to decision to transplantation is much shorter.

Table 47.2 Investigation of the liver transplant candidate

Area of investigation	Specific testing
Full symptom history and physical examination	
Haematology	Complete blood count, clotting profile
	Blood group, tissue typing
	Serum biochemistry, liver function tests
Radiology	Chest and abdominal X-ray
	X-ray of wrists and long bones
Imaging	Ultrasound with Doppler for size and flow in the portal vein and unusual anatomy
	CT with contrast or magnetic resonance angiogram if vascular anomaly is suspected
Renal function	Urine analysis, biochemistry, and creatinine clearance
Cardiopulmonary function	Full evaluation, particularly if hepatopulmonary syndrome is evident
Serology	Hepatitis A,B,C; CMV, EBV, HSV, HIV, measles, varicella
Infection screen	Cultures of urine, sputum, blood, stool, ascites fluid and swabs from nose and throat, dental check
Nutritional evaluation	Micronutrients: iron, zinc, selenium, manganese; vitamin levels A and E
Developmental assessment	
Psychosocial assessment	

CMV—cytomegalovirus; EBV—Epstein-Barr virus; HIV—human immunodeficiency virus; HSV—herpes simplex virus

As the outcome of the operation is so much better in recent years, indications for early transplantation would be evidence of impaired synthetic function of the liver, including prolonged prothrombin time, reduced serum cholesterol levels, and low serum albumin. Clinical indicators include the presence of ascites, bleeding from oesophageal varices not controlled by sclerotherapy or banding, and poor response to nutritional resuscitation. Those with acute hepatic failure who develop encephalopathy, hypoglycaemia, and prothrombin time of greater than 50 seconds should be considered for transplantation, although the data to guide us in avoiding death without transplantation and unnecessary transplantation remain elusive. Although it has not gained widespread application, auxiliary partial orthotopic liver transplantation (APOLT) is in theory an attractive option, performed in the hope that the native liver will recover and eventually immunosuppression can be withdrawn.

All patients require initial confirmation of the diagnosis, intensive medical investigation, and nutritional resuscitation to treat the complications of the liver disease, portal hypertension and nutritional deprivation. Immunization status must be reviewed and supplemented with hepatitis A and B immunization, and *Haemophilus influenzae*, pneumococcal, and meningococcal vaccine in most cases. Blood-group identical or compatible donors are much preferred, as the long-term survival with blood-group incompatible donors is significantly less; the incidence of biliary stricture is increased. In recipients less than 1 year of age, blood-group incompatible donors can be used in urgent situations. Patients are generally listed according to urgency on a scoring system measuring the serum bilirubin (mg/dL), the International Normalised Ratio (INR), and the serum albumin (mg/dL). The Pediatric End-Stage Liver Disease (PELD) score is most widely used in children. The PELD score = $0.480 \times \text{Log}_e$ (bilirubin mg/dL) + $1.857 \times \text{Log}_e$ (INR) − $0.687 \times \text{Log}_e$ (albumin g/dL) + 0.436 if patient is <1 year (scores for patients <1 year listed for liver transplantation; continue to include the value assigned for age of <1 year until the patient is actually aged 2 years) + 0.667 if the patient has growth failure (<−2 standard deviation) × 10 (then round to the nearest whole number) [1].

47.3 Donor Identification, Screening, and Management

The process of deceased organ donation and transplantation requires, in the first instance, donor identification, followed by donor screening to exclude risk of transmission of serious disease (cancer and infection). The donor is then managed to optimise the quality of the grafts at the time of procurement. Consent or authorization must be obtained within the legal framework of the country. Organ retrieval involves removing the organs, preparing them for transportation to the appropriate recipient, and finally, organ allocation and engraftment.

Contraindications to organ donation include evidence of HIV/AIDS or active hepatitis B infection, presence or history of extracranial malignancy and disease of unknown aetiology. Viral screening of donors is essential. This screening should include hepatitis A, B, and C; cytomegalovirus (CMV), Epstein-Barr virus (EBV), and HIV.

Donor organ suitability and function is difficult to predict. Increasingly, marginal heart-beating, brain-dead donors are being used, with a surprisingly low incidence of primary poor function or nonfunction. Age limits for donors are being extended, but the preferred donors are stable patients less than 50 years of age with a short intensive care unit stay (<3 days), little requirement for inotropic support, and normal or near-normal liver function; the expected incidence of impaired function after transplantation from these donors is less than 5%. Liver biopsy is useful if steatosis is suspected. More than 50% fatty infiltration would preclude the use of the liver in most centres. Organs procured after cessation of cardiac output—so-called non-heart beating donors or donation after cardiac death—have been used, but less frequently for children; the incidence of biliary strictures and early poor function or nonfunction is increased.

47.4 Surgical Technique

47.4.1 Donor Organ Retrieval

Surgical techniques used for donor retrieval and recipient liver removal and engraftment have evolved over the past 40 years. Most donor livers are removed as part of a multiorgan procurement procedure in brain-dead, heart-beating donors, including various combinations of kidneys, liver, heart or heart and lungs, small bowel, and pancreas. Cold University of Wisconsin (UW) solution and the histidine-tryptophan-ketoglutarate (HTK) organ preservation solutions are the most widely used preservation solutions.

Two principal techniques are used: a careful, complete dissection and excision technique, or the so-called rapid technique described by Starzl. A midline incision is made from the suprasternal notch to the pubis. The sternum is opened and spread with a sternal retractor. The subcostal diaphragm is incised on each side to obtain better exposure. The abdominal part of the operation includes mobilization of the caecum and right colon to expose the aortic bifurcation, and control of the aorta above the coeliac axis by incision of the right crus of the diaphragm, being careful to avoid entering the oesophagus. The aorta at this level is encircled with tape to enable cross clamping prior to infusion of cold preservation solution. The inferior vena cava (IVC), infrarenal aorta, and bifurcation are identified, dissected, and encircled with tapes below the renal vessels. The inferior mesenteric artery may be ligated in continuity to prevent loss of preservation fluid into the bowel if the intestine is not being procured. A careful search is conducted for any vascular abnormalities, which most frequently would be a left hepatic artery branch from the left gastric artery and an accessory or replaced right hepatic artery from the superior mesenteric artery (SMA), which may run through the head of the pancreas and behind the

portal vein and bile duct. The liver is then mobilised by division of the falciform, right triangular, and left coronary ligaments. An appropriate-sized cannula is placed in the inferior mesenteric or superior mesenteric vein so that the tip lies at the junction with the splenic vein and is secured with a tape tie. A large-bore cannula is placed in the aorta with the tip approximately opposite the renal arteries, the distal common iliac vessels are tied off, and the donor is given a heparin (3 g/kg). A large-bore cannula may be placed in the IVC, which is connected to an away suction to drain off venous blood and perfusate. The gallbladder and biliary tree are thoroughly irrigated with ice cold saline. The cardiac procurement can proceed now if the heart is suitable.

Procurement commences with infusion of preservation solution through both the portal vein and aorta after cross clamping the aorta at the level of the diaphragm and incising the suprahepatic vena cava within the pericardium. The timing depends on whether the heart is being procured. Topical cold saline and slush ice is lavaged into the peritoneal cavity to assist in cooling both the kidneys and liver. Normally, 2 L of UW preservation solution or 5–8 L of HTK solution are infused through each cannula, according to the size of the donor. If the SMA is not ligated, the bowel will blanche; this will allow portal perfusion as well as arterial perfusion, particularly if the inferior mesenteric vein is used for access. The porta hepatis is divided distal to the gastroduodenal artery, which is ligated, and the portal vein is divided at the junction with the splenic vein. The proximal part of the SMA is defined and is dissected down to the aorta on its left side, preserving 2–3 cm if accessory or replaced right hepatic arteries are present. In the 'rapid' technique, the liver is removed with a patch of aorta including the base of the SMA and the coeliac axis, particularly if abnormal arterial supply has been identified. The liver is then removed from above downwards. The resected liver includes a cuff of diaphragm around the bare area, along with the retrohepatic cava and part of the right adrenal gland, which is cut through. The infrahepatic IVC is divided above the renal veins.

Once the organs have been removed, they are placed in a plastic bag and the liver is further perfused with 500 mL of preservation solution via the portal vein, the hepatic artery, and the bile ducts. The liver then is placed in two plastic bags, surrounded by an additional litre of preservation solution and packed in ice for transportation. Once the kidneys are removed, the iliac arteries and veins are retrieved and stored in preservation solution in case they are needed for subsequent vascular reconstructions.

Normal liver volume is estimated to be between 2% and 4% of body weight. Graft size should not be less than 0.8% of recipient body weight, to avoid small-for-size syndrome.

47.4.2 Back Table Preparation

Back table preparation of the donor liver will depend on whether a whole liver, split liver, or reduced-size liver graft is to be used. The liver is placed with the inferior surface uppermost within the inner plastic bag and is surrounded by packed sterile ice. Upper and lower cava are defined and redundant diaphragmatic muscle is excised. Phrenic veins are ligated and sutured closed, and the arterial and venous anatomy is clearly dissected. Arterial reconstruction on the back table may be required, using microsurgical techniques and the available arteries to fashion one primary inflow to be sutured to the recipient common hepatic artery. For instance, in whole live transplants with an accessory or replaced right hepatic artery arising from the SMA, a short segment is preserved and anastomosed to the coeliac artery trunk; if already divided, it can be anastomosed to the stump of the splenic artery.

The plane of transection when size reduction is planned is usually just to the right of the falciform ligament in left lat-

eral segment transplants and in the median plane for left liver transplants taking into account the segmental anatomy of the liver (Figs. 47.1, 47.2, 47.3, 47.4, 47.5, 47.6, 47.7, 47.8 and 47.9). Even more careful dissection is required where split liver transplantation is being used, in which the liver is divided into two functional units for two recipients (Figs. 47.10, 47.11, 47.12 and 47.13). Decisions must be made as to which porta hepatis structures will go with which graft. To a certain extent, this depends on local preference, but many centres opt to export the right-sided graft for an adult recipient with the full complement of structures of bile duct, portal vein, and hepatic artery, keeping the left lateral segment for the local graft and paediatric recipient in much the same way as a living donor transplant. A back-table cholangiogram is very useful in deciding exactly how to split the graft and will alert one to the need for possible separate segment 2 and 3 hepatic duct jejunostomy in the recipient. If a reduced-size liver is used, the caudate lobe is always excised. The left portal vein is carefully dissected from the caudate lobe with ligation of all tributaries and is preserved to achieve maximum length with division at its junction with the right branch of the portal vein. The site of division of the right portal vein is sutured closed transversely for the right lobe graft. In most cases of paediatric transplants in which a reduced-size or split liver graft is used, the donor IVC is discarded or left with the right split and a cuff of left/middle hepatic vein is preserved on the paediatric graft to be sutured to a preserved vena cava and opened hepatic vein confluence of the recipient. The division of liver tissue may be performed using the standard forceps clamp technique and bipolar diathermy with suture, Ligaclip, and ligation of vascular and biliary structures. The ultrasonic dissector, LigaSure, or Harmonic Scalpel also may be used. The portal vein, hepatic artery, and bile ducts are again perfused to identify any leaks, which are sutured closed. The cut edge of the liver is then sprayed with two layers of tissue glue. The graft remains in preservation solution surrounded by ice until the recipient is ready for engraftment. The graft may be further reduced in size by excision of more graft tissue (monosegment graft) to achieve donor-to-recipient weight ratios of up to 15 to 20:1. The decision as to how this is done depends on the shape of the left lateral segment (Fig. 47.16). With a 'flat fish' left lateral segment graft, much of segment 2 is removed, whereas with a graft having a larger anterior/posterior volume, a triangular slice of liver including both the more lateral parts of segments 2 and 3 is removed. Contraindications to splitting are anatomical variants that include absent portal vein bifurcation, a segment 3 hepatic vein draining directly into a middle hepatic vein distant from the liver surface, a small atrophic left liver, and an accessory right hepatic artery arising from the left hepatic artery. Splitting of the liver takes 2.5–3.5 hours, and this time must be taken into account when preparing the recipient. Although rarely performed, an auxiliary partial orthotopic liver transplant (APOLT) may be attempted in a situation of acute liver failure, where the native liver is expected to recover, after which immunosuppression can be withdrawn. An APOLT or heterotopic graft may also be attempted to replace metabolic defects of the native liver but these techniques have not been widely performed.

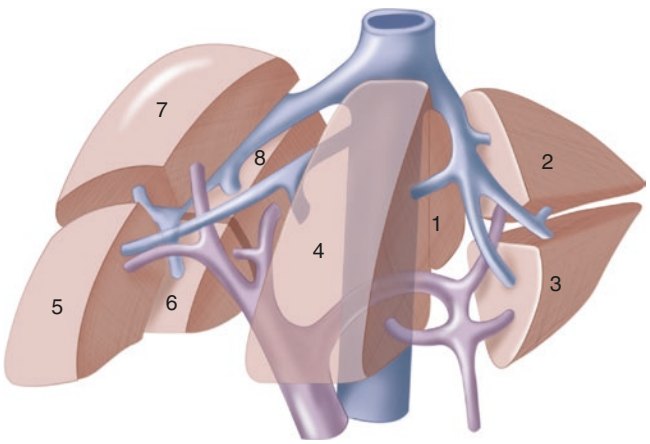

Fig. 47.1 The portal and hepatic vein segmental distribution in the liver showing the numbered Couinaud classification of the segmental anatomy of the liver

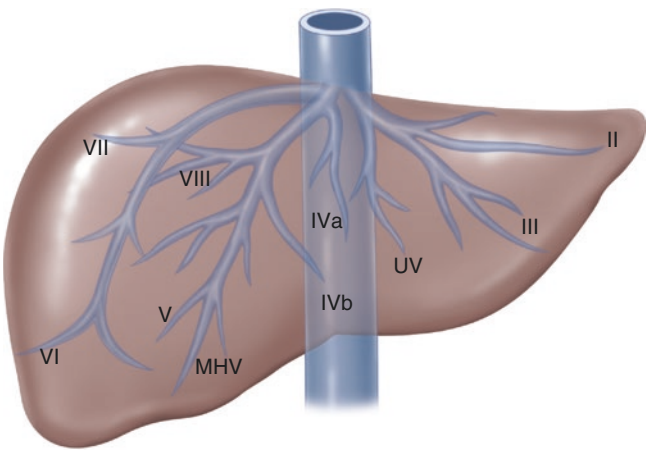

Fig. 47.2 Hepatic vein anatomy of the liver (MHV - middle hepatic vein, UV - site of umbilical vein)

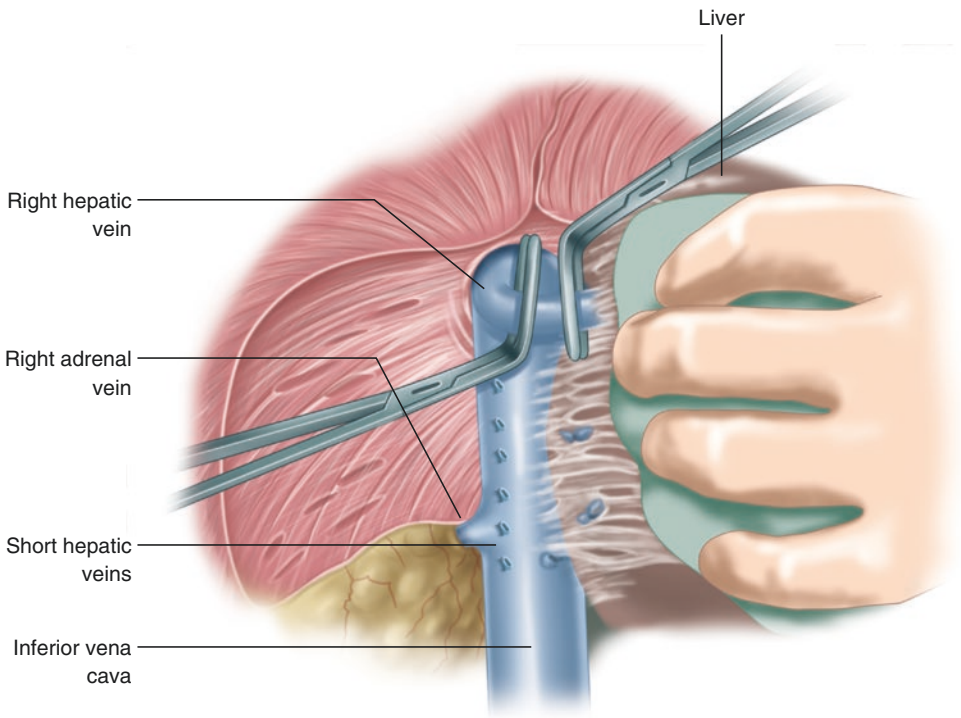

Fig. 47.3 Mobilization of the right lobe of the liver off the inferior vena cava (IVC)—an important step in explantation of the liver when the IVC is to be preserved, as in most paediatric transplants

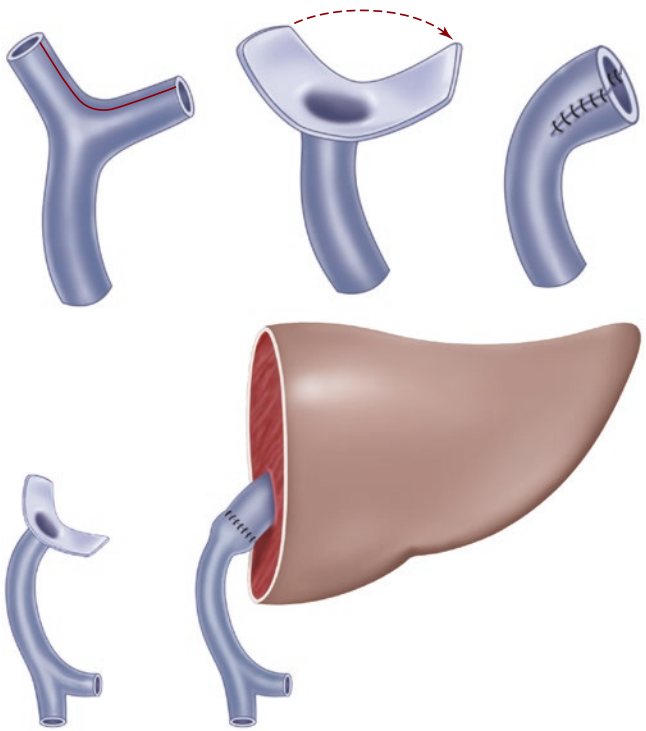

Fig. 47.4 Recipient portal vein bifurcation is opened to provide a more appropriate size match for anastomosis to an adult-size graft portal vein. The opened bifurcation may be sutured directly to the adult size vein or a funnel may be fashioned by suturing the flaps on each side as depicted

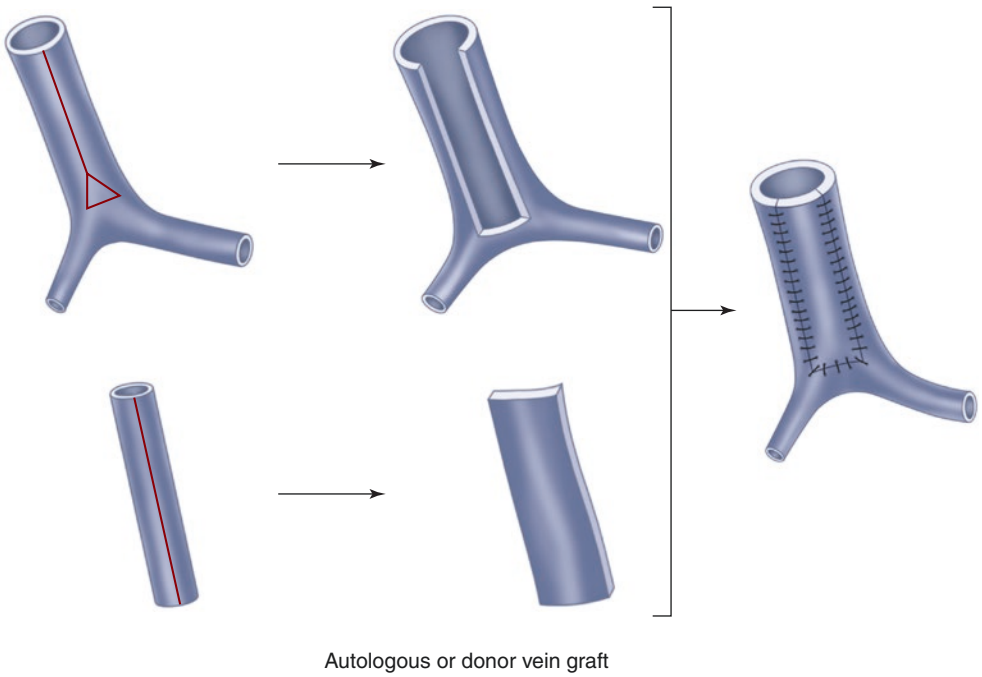

Fig. 47.5 Anterior portal vein patch venoplasty for a hypoplastic recipient portal vein

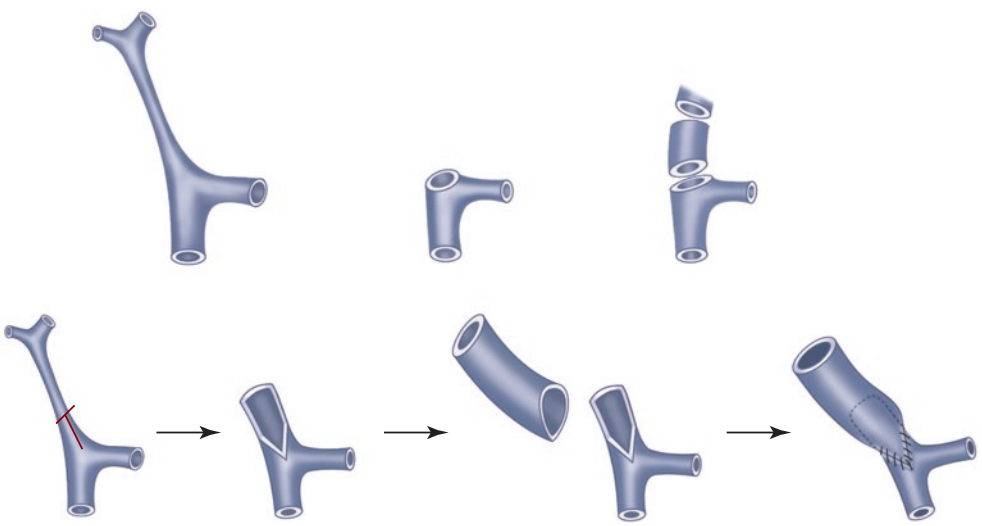

Fig. 47.6 Other techniques of dealing with a hypoplastic recipient portal vein: (1) Interposition of an iliac vein graft onto the superior mesenteric vein and splenic vein confluence. (2) Excision of very narrow sclerotic portal vein, incising anteriorly, and then suturing a vein graft to the expanded portal vein segment

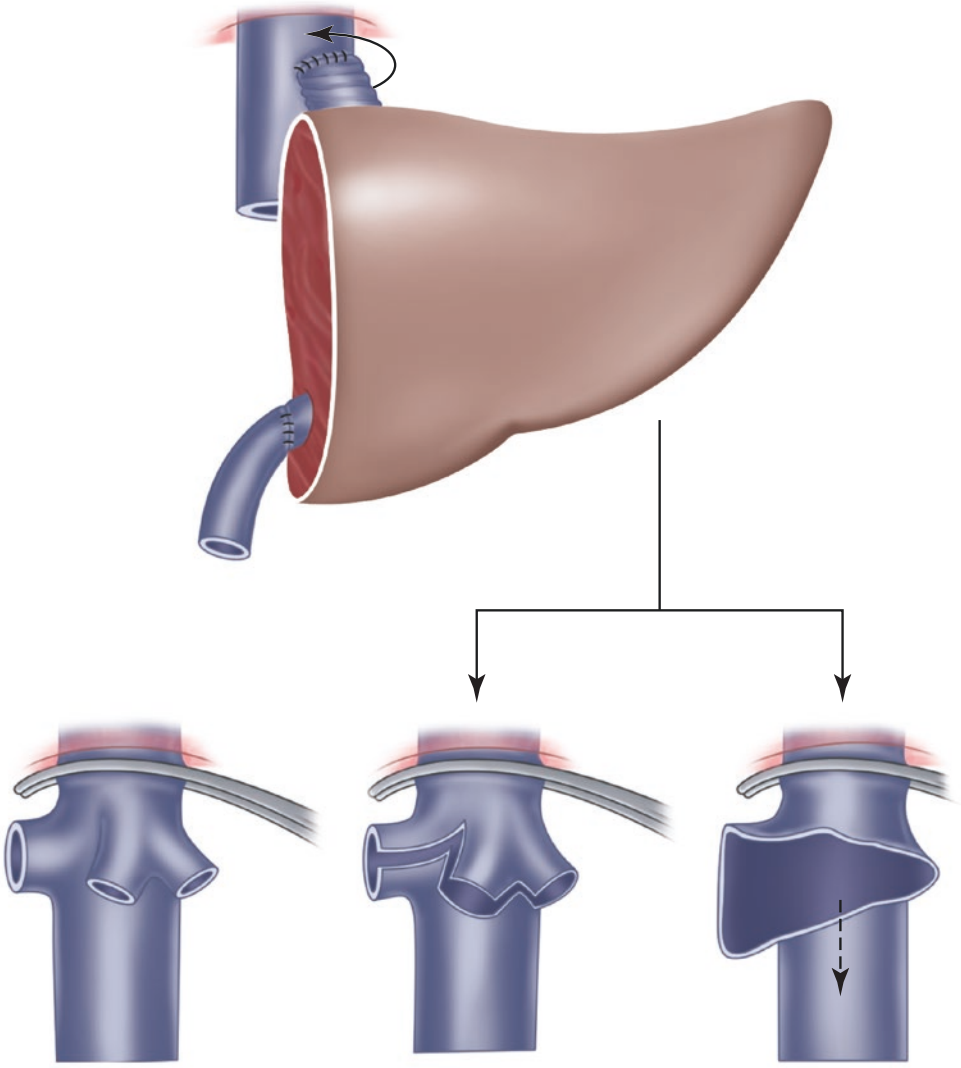

Fig. 47.7 To avoid rotation and kinking at the anastomosis of the hepatic vein and IVC as depicted in this figure, the three hepatic veins are opened to make a large triangular orifice, which may be enlarged further by caudal incision on the anterior aspect of the IVC (see arrow)

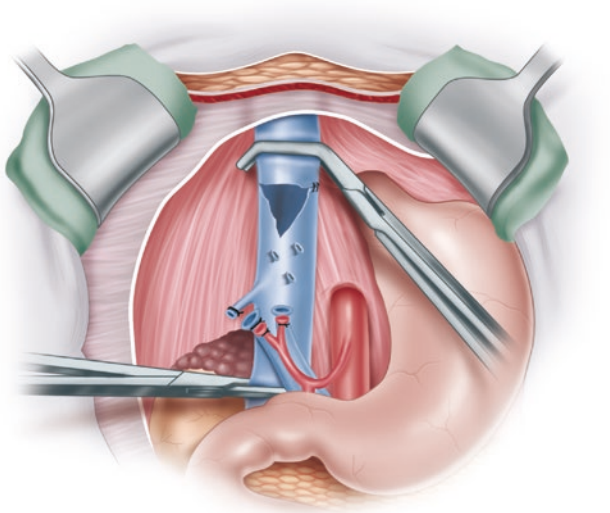

Fig. 47.8 The preserved IVC after recipient liver explantation. The orifice of the left hepatic vein has been sutured closed, and a large triangular orifice is evident in preparation for graft hepatic vein anastomosis

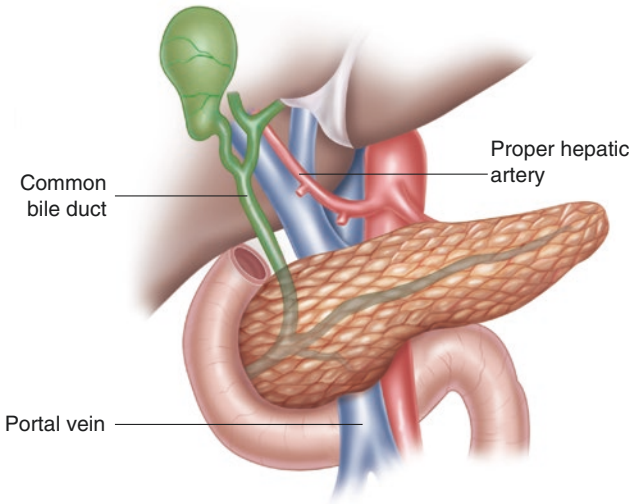

Fig. 47.9 The usual vascular and biliary anatomy at the porta hepatis

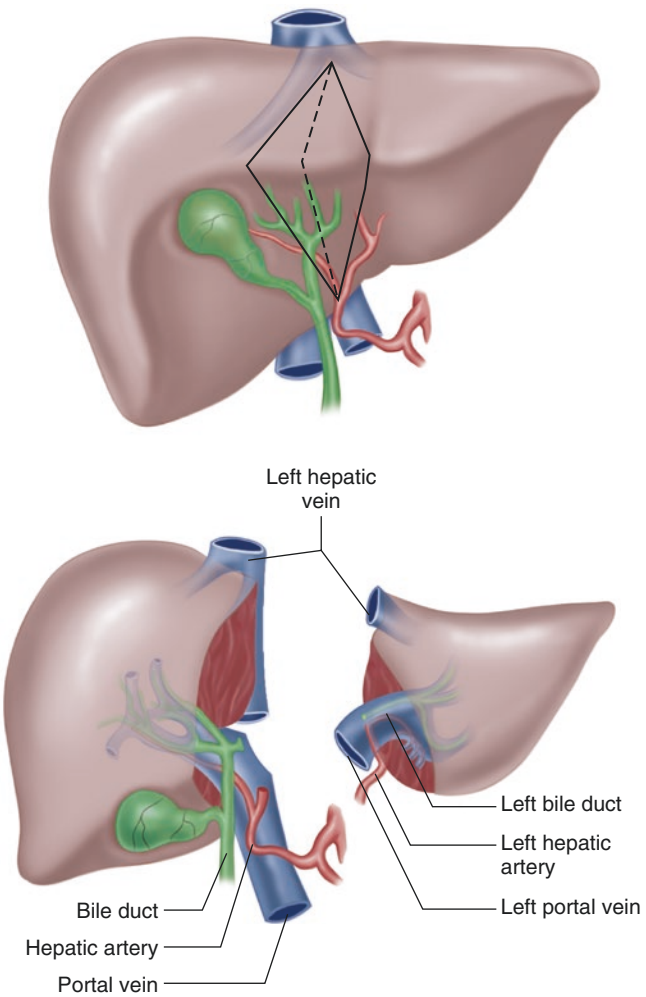

Fig. 47.10 Vascular and biliary division after splitting the liver

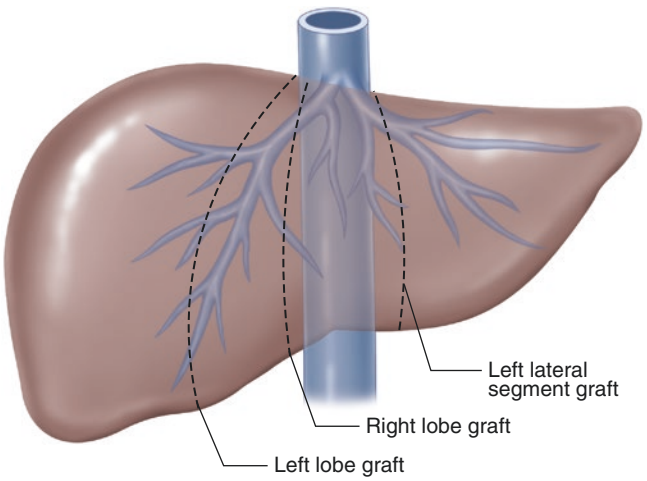

Fig. 47.11 The three planes of division for different types of split liver grafts

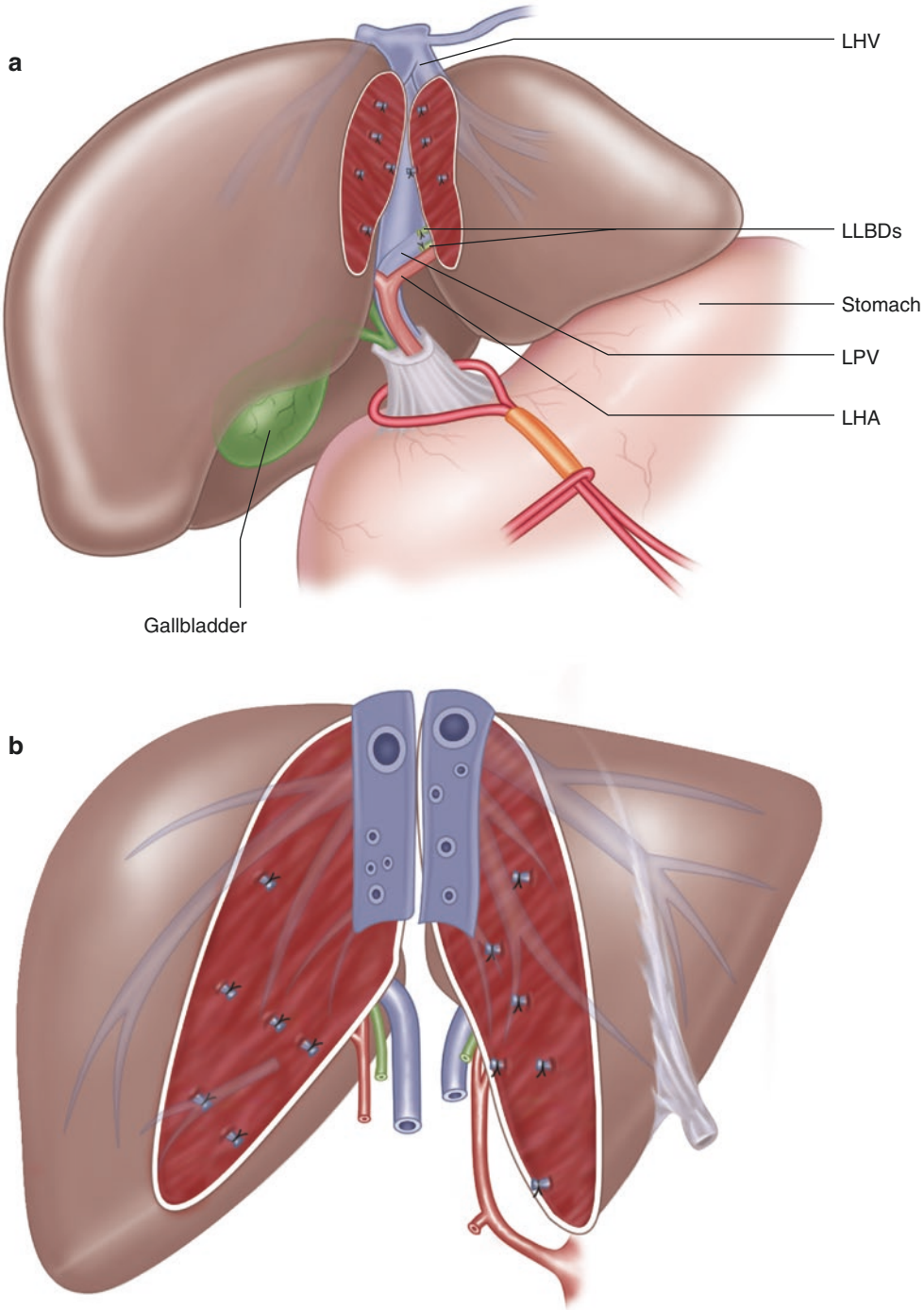

Fig. 47.12 (**a**) Classic split liver graft, with the segment 2 and 3 graft for a paediatric recipient and the right liver for an adult recipient. (**b**) Full median right/left split grafts for two paediatric recipients or a child and a small adult. LHA—left hepatic artery; LHV—left hepatic vein; LLBDs—left lobe bile ducts; LPV—left portal vein

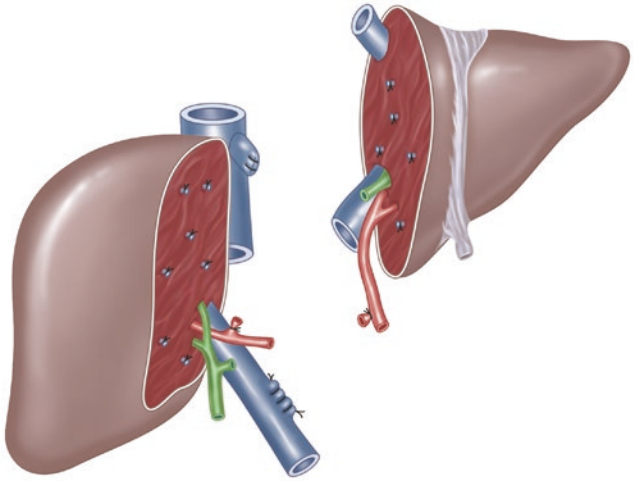

Fig. 47.13 Split liver transplantation with the main trunk of the hepatic artery retained with the left lateral segment graft

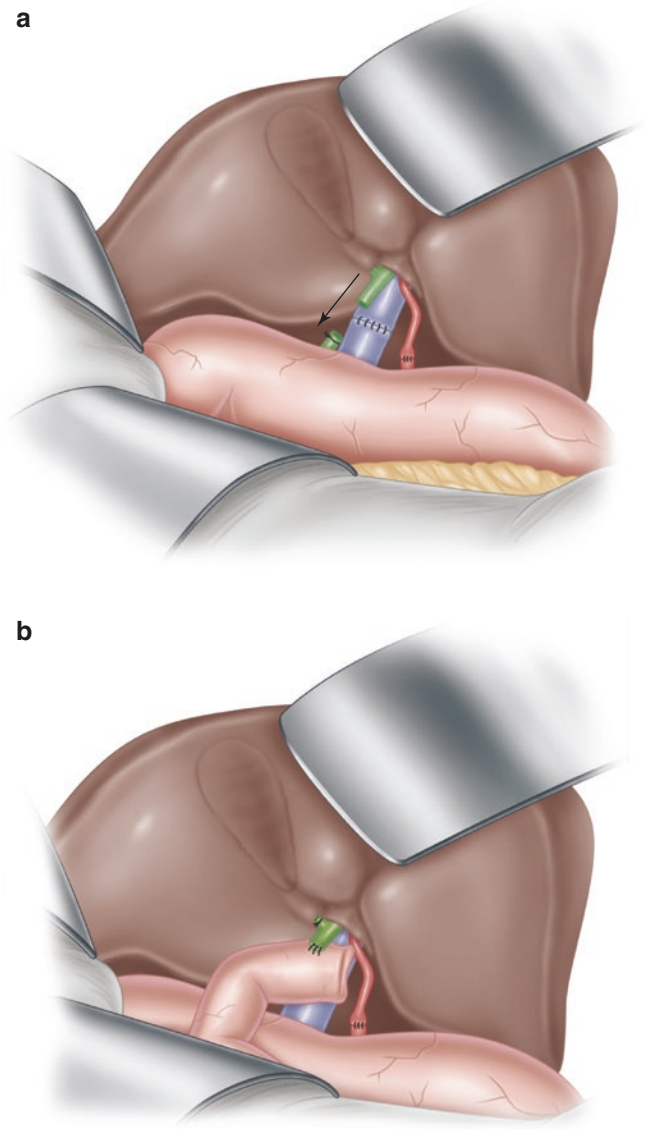

Fig. 47.14 Whole liver graft with gall bladder removed, portal vein and arterial anastomoses completed, and duct-to-duct anastomosis in progress (**a**) and with a choledocho-jejunostomy completed (**b**)

47.4.3 The Recipient Operation

The recipient operation is timed so that the estimated hepatic graft ischaemic time is less than 12 hrs. Much longer preservation times have been recorded, but the incidence of cholestasis and graft dysfunction is increased. The recipient operation commences with a laparotomy incision, which may be subcostal, chevron-shaped, and extended in the midline to the xiphoid process for extra exposure. If a Kasai procedure has been done, the previous scar may be used as guide. Prior to incision, subcutaneous injection of 1:200,000 adrenaline in saline may reduce bleeding from large subcutaneous veins traversing the incised tissue. Self-retaining subcostal retractors are an essential aid in obtaining the necessary exposure. Careful suture ligation of large, thin-walled veins in the abdominal wall, falciform ligament, adhesions, and omentum are required to keep blood loss to a minimum. Once the infrahepatic and subcostal areas are freed from the surrounding structures and liver using cautery and sharp dissection, the flaps of abdominal wall are sewn back over the costal margins to the recipient skin or drapes and a subcostal self-retaining retractor (e.g., the Thomson system) is inserted with cranial and lateral traction to provide maximum exposure. The left lobe is further separated from the diaphragm and spleen by division of the left triangular ligament. The porta hepatis is dissected next; in children with biliary atresia, this requires the portoenterostomy to be taken down, with preservation of the Roux-en-Y loop for later hepatic duct anastomosis.

It is useful at this stage to dissect out the hepatic artery to proximal to the gastroduodenal in preparation for revascularisation. This step avoids unnecessary delay at the engraftment stage of the procedure. The portal vein is dissected back to the level of the pancreas and isolated. The rest of the liver is carefully mobilised, with division of the gastrohepatic ligament and the falciform and triangular ligaments together with the right retroperitoneal reflexion. Mobilisation of the liver off the IVC, which is frequently preserved to facilitate reduced-size transplantation or piggyback engraftment, should be done by careful dissection and ligation/suture of multiple small hepatic veins draining from the caudate lobe. This step is assisted by carefully dividing the vena cava ligament and ligating the right adrenal vein and sometimes the right hepatic vein (Fig. 47.3). The suprahepatic and infrahepatic vena cava are dissected and encircled with tape.

The portal vein must be assessed as to its suitability for use. If it is sclerotic and narrow, it is best to dissect back to the splenic vein confluence or prepare a segment of iliac vein, which is sutured to the recipient portal vein prior to

engraftment (see below). A trial of cross clamping of the portal vein and IVC is done to ensure that cardiac output in the recipient is maintained. This is particularly important in cases where the recipient liver is not cirrhotic, such as recipients with oxalosis (in which cardiovascular reserve may be compromised) or Alagille syndrome without cirrhosis with very good portal vein flow and a relatively hypoplastic artery. Occasionally in larger children (>25 kg), a porta-systemic bypass pump may be required if cross clamping is not tolerated and one is unable to maintain venous flow through a preserved IVC. The IVC is clamped above and below the liver, right and left portal vein branches are encircled and ligated or clamped as close to the liver as possible to preserve the bifurcation, and the hepatic artery is ligated and divided. Once the recipient liver has been removed, meticulous haemostasis of the retroperitoneum must be ensured with a combination of suture ligation and cautery. If the recipient IVC is to be preserved, liver removal is most easily done by carefully incising the diseased liver clear of the cava and when the liver has been removed, individually suturing all small areas of leakage from divided direct caudate lobe hepatic veins. The IVC is prepared for the donor liver by dividing the bridges between the separate hepatic veins. This step creates a wide orifice for the hepatic vein-to-cava anastomosis, which is enlarged further by making an incision on its anterior surface caudally for approximately 1–2 cm to make a wide triangular orifice for the 'piggyback' graft (Figs. 47.7 and 47.8). The graft hepatic vein is matched in size to the recipient hepatic vein defect. The graft of a reduced-size liver is sewn in rotated 40–60° in an anticlockwise direction so that the graft cut surface faces posterolaterally. A living donor or split liver graft may also be slightly rotated, but should remain in an anatomical position. Engraftment should begin with the upper caval anastomosis, which is usually performed with continuous left and right posterior polypropylene or polydioxanone 4/0 or 5/0 sutures, with interrupted anterior sutures if conventional transplantation is being done. Prior to completion of the anastomosis, the portal vein is briefly opened, allowing a blood flush to ensure that a thrombus has not developed during clamping; the liver is flushed clear of preservation solution via the portal vein with normal saline, blood, or a colloid solution through an incomplete anterior hepatic vein caval anastomosis if a piggyback technique has been used. The lower cava anastomosis is then performed if required, being sure not to cause any stricture or kinking. A 'growth factor' of about one third of the diameter of the vessel is usually sufficient to prevent this from occurring. The recipient portal vein is usually used for the anastomosis; in reduced-size transplants where the donor liver is of large diameter, the bifurcation of the recipient portal vein is opened to create a trumpeted end for anastomosis (Fig. 47.4). If the portal vein is hypoplastic, then the anastomosis is done at the level of the confluence with the splenic vein, after careful dissection under the head of the pancreas, or a graft is sutured (Fig. 47.5); this is done during the anhepatic phase by suturing a vein graft from the donor iliac vein or recipient inferior mesenteric vein onto either the portal vein/splenic vein confluence or onto the recipient portal vein, which has been partially cut back and incised along its anterior aspect. Figure 47.6 illustrates other techniques. The donor to recipient portal vein anastomosis is carefully performed, avoiding torsion and tension or redundancy, using continuous sutures between triangulated stay sutures on traction to avoid any narrowing or concertina effect. Alternatively, posterior continuous and interrupted anterior suture techniques or the use of a generous 'growth factor' can achieve the same effect.

In reduced-size grafts, plenty of portal vein length should be left to avoid having any tension on the vein, which may result in stretching and compromise flow as the cut surface of the graft lies against the recipient diaphragm at an angle of 40–60° from the median plane and the portal vein has to extend far to the recipient's right side before entering the vein. With split-liver and living donor grafts, the left lateral segments should lie more in an anatomical position because of the relative shortness of the portal vein. The donor hepatic artery is flushed with heparin saline to remove air and blood clots, and an anastomosis is done to the recipient common hepatic artery. End-to-end microvascular techniques are preferred, using 7/0 or 8/0 Prolene or polydioxanone. Vascular grafts using donor external iliac artery to the infrarenal aorta or from the supracoeliac aorta have been used with success, when the recipient hepatic artery is small. This is particularly the case with Alagille syndrome and cases of biliary atresia splenic malformation (BASM) syndrome. The donor liver is usually revascularized with removal of the suprahepatic clamp first, followed by removal of the infrahepatic clamp, a pause to check for any major bleeding, and then opening of the portal vein. The arterial anastomosis is then performed between the donor coeliac trunk, trimmed to the appropriate size, or graft hepatic artery variant and the recipient common hepatic artery. There is some experimental evidence of less reperfusion injury with early arterial revascularisation.

After careful haemostasis of bleeding areas such as the free edge of a reduced-size liver or any of the other major bleeding points, the operation is completed by performing the biliary reconstruction (Figs. 47.14 and 47.17). The bile duct (Fig. 47.9) is trimmed back such that good bleeding from the cut edges is obtained, and it may be incised anteriorly to extend the anastomosis and avoid stricturing. In patients with biliary atresia and those with a reduced-size graft, a 35-cm Roux-en-Y choledochojejunostomy or hepaticojejunostomy is performed with fine interrupted absorbable sutures. If a reduced-size graft is used and the duct-to-jejunum anastomosis is distal to the cystic duct confluence, it is important not to close the cystic duct orifice, as it may develop into a retention mucous-filled cyst and obstruct the bile duct. If the hepatic ducts of segments 2 and 3 are far

apart, two individual anastomoses may be required. If they are close together, they may be approximated with interrupted sutures to make a single orifice for the hepaticojejunostomy (Fig. 47.15).

Occasionally in paediatric cases, a duct-to-duct anastomosis may be performed with a whole liver graft in a recipient with a normal extrahepatic biliary system (Fig. 47.14). Cholecystectomy is performed with careful ligation of the cystic duct. Stents or T tubes are optional and are now rarely used. Haemostasis is obtained, closed tube drains are placed to the suprahepatic and infrahepatic spaces, and the wound is closed with continuous polydiaxonone sutures. If there is any tension at sheath closure because of bowel oedema or large graft size, it is wise to insert a temporary patch of GoreTex or other nonadherent material (Fig. 47.18), as a 'tight' abdominal closure is associated with abdominal compartment syndrome and an increased incidence of vascular thrombosis. It is usually possible to obtain skin closure over the patch without too much tension. A temporary patch can be removed 5–10 days later with sheath approximation without tension. A more permanent patch may also be used such as porcine dermal collagen (Fig. 47.17).

47.4.4 Living Related Donors

Living related donation of the left lateral segment, first successfully performed by Strong, has become widely accepted as a method of acquiring a liver graft in the face of severe donor shortages, particularly in countries with cultural or religious reticence to accept brain death in a ventilated, heart-beating donor. There are clear advantages in the planned nature of the procedure, which is preferably performed before end-stage liver disease in the recipient, with an excellent-quality graft and short ischaemic time.

The use of a living donor also increases the availability of donor organs in general for other patients on the waiting list. The only advantage to the donor is a psychological one, and there is a current morbidity of about 10% (wound sepsis, hernia, bile leak, and adhesive bowel obstruction). There is also a reported mortality of about 0.2%, although more than 1000 of these operations were done in one centre in Japan without donor mortality. Ethical concerns appear justified, as with more widespread transplant activity, increasing mortality

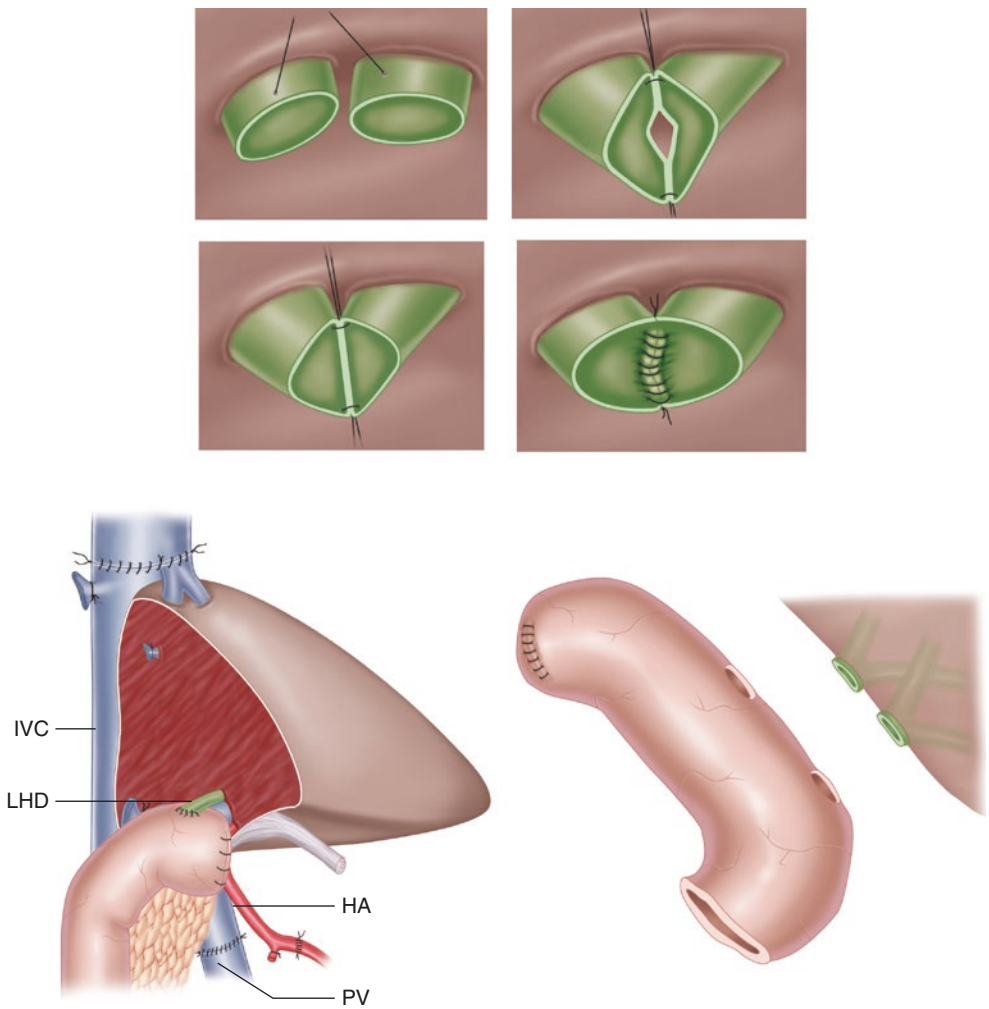

Fig. 47.15 In left lateral segment grafts, if the segment 2 and 3 ducts are separate at the line of resection, they can be approximated and fashioned into one orifice when close enough together; if not, two separate hepaticojejunostomies must be done. CBD—common bile duct; HA—hepatic artery; PV—portal vein

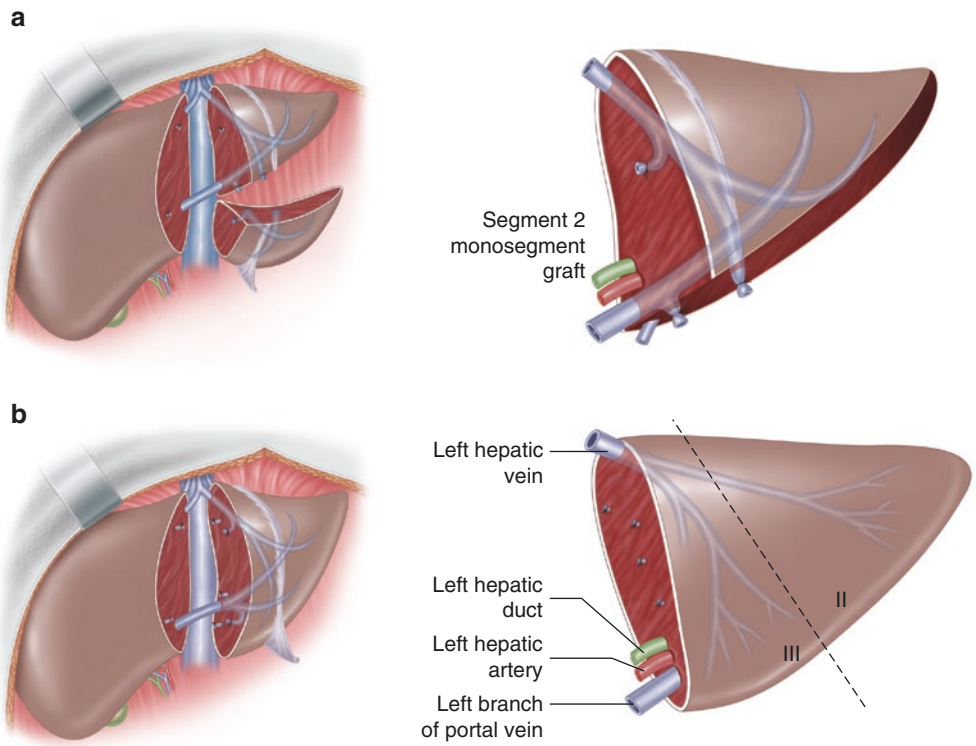

Fig. 47.16 Preparation of a monosegment graft, depending on the shape of the left lateral segment. If the anteroposterior (AP) diameter is large (**a**), much of segment 3 is discarded; if the AP diameter is less ('flat fish' segment), segment 2 can be excised (**b**)

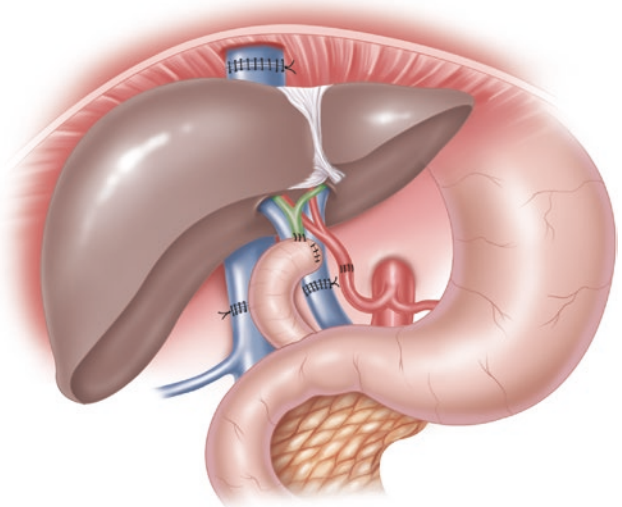

Fig. 47.17 A classic whole liver graft at the completion of surgery, with a choledocho-jejunostomy and standard vascular anastomoses

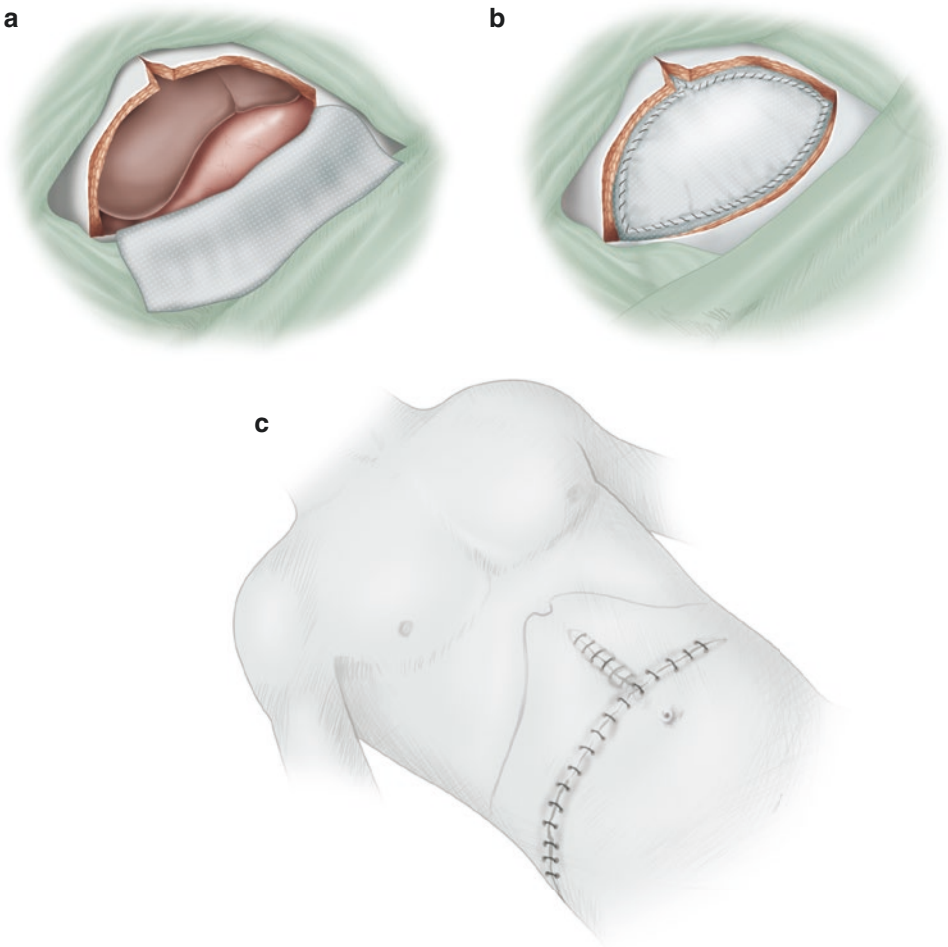

Fig. 47.18 (**a–c**) Large whole graft requiring abdominal patch closure to avoid abdominal compartment syndrome. The patient shown is a 14-year-old boy weighing 24 kg who underwent whole liver/kidney retransplantation from a 30-kg donor

and morbidity have been recorded. The donor should first undergo a thorough screening, both clinical and psychological, without coercion, and should be given an option to withdraw at any time before the transplantation.

47.5 Postoperative Care

Patients are monitored with all aspects of modern postoperative intensive care and usually require ventilation for a period of 24–48 h. Immunosuppression protocols have become standardized using tacrolimus, azathioprine, or mycophenolate mofetil and steroids; some centres use an anti-CD 25 monoclonal antibody (basiliximab). mTOR inhibitors rapamycin or everolimus can be added later. Liver ultrasound with colour flow Doppler is performed frequently to confirm vascular patency and the absence of biliary dilatation. Liver biopsies are performed if indicated by increasing serum liver enzyme activity or bilirubin levels, using a Menghini needle (diameter 1.4 mm), unless biliary dilatation is observed on ultrasonography. Biopsy specimens are routinely assayed for viral and bacterial activity.

47.6 Surgical Complications

Surgical complications (Table 47.3) may present early and late, but they may be reduced to an absolute minimum with meticulous technique. Biliary complications continue to be a significant problem, with an overall incidence of 10–20%, particularly in living related left lateral segment and split-liver grafts. These complications include bile leak, anastomotic strictures, and non-anastomotic intrahepatic strictures of the donor bile duct with sludge formation.

It is imperative with all suspected biliary complications to ensure that the hepatic artery is patent, using Doppler ultrasound or angiography (CT or MR), as hepatic artery thrombosis may cause ischaemia and necrosis of the biliary tree.

Simple bile leaks are diagnosed in the early postoperative period by the presence of bile in drainage fluid or in percutaneous aspirate of fluid collections around the liver. Early biliary complications are best treated by immediate surgery and re-anastomosis if required. Late stricture formation may be satisfactorily dealt with by endoscopic or percutaneous balloon dilation or stenting, and finally by retransplantation. Graft ischaemia either from hepatic artery thrombosis or stenosis can be a devastating complication. The incidence (about 5%) is much less with the use of reduced-size liver transplants and microsurgical techniques for living donor transplantation. Nevertheless, when it occurs it may lead to graft necrosis, intrahepatic abscess, biliary necrosis, and bile leakage. A massive rise in enzyme activity, particularly in the first few days after transplantation, may be the first sign. Immediate intervention with thrombectomy and re-anastomosis is only occasionally successful.

Late presentations of hepatic artery thrombosis or stenosis may be less acute and typically present with gram-negative sepsis, liver abscess, or biliary complications, mainly stricture. An early hepatic artery thrombosis usually means that the patient will require an urgent liver re-transplant. Late thrombosis may be asymptomatic, and if so, can be ignored. Although technical factors usually account for most cases, it is advisable to maintain the haematocrit at

Table 47.3 Summary of common postoperative surgical problems

Type of problem	Specific complication
Biliary tract	Anastomotic leak—often associated with hepatic artery thrombosis
	Stenosis or stricture
	Infection (cholangitis—usually associated with anastomotic stricture)
Vascular (thrombosis, stenosis)	Hepatic artery
	Portal vein
	Inferior vena cava (supra and infrahepatic)
	Hepatic vein (left lateral segment grafts), Budd-Chiari recurrence
Miscellaneous	Bowel perforation (steroids, diathermy)
	Diaphragm paresis/paralysis
	Gastrointestinal haemorrhage (peptic ulceration, varices)
	Infected collections
	Abdominal compartment syndrome (large-for-size graft)
	Wound-related (infection, dehiscence, hernia)
	Primary nonfunction

about 30% to improve microvascular flow. Low-dose heparin infusion is used for "at risk" arteries, and aspirin/persantin prophylaxis is given routinely for the first 3 months.

Portal vein thrombosis (about 2% incidence) usually presents with a degree of liver dysfunction, prolonged INR, ascites, and portal hypertension, which may be heralded by an oesophageal variceal bleed. Immediate thrombectomy may be successful. Late thrombosis or stenosis may be amenable to percutaneous balloon angioplasty. Where portal vein thrombosis is established, a meso-portal (Rex) shunt, with a vein graft taken from the internal jugular vein and interposed between the superior mesenteric vein and the left branch of the portal vein, may be curative.

IVC thrombosis or venous outflow obstruction are now rare, particularly since the triangular technique of hepatic vein to caval anastomosis has been practiced (Fig. 47.7, 47.8). Thrombosis in the IVC may develop either in the immediate postoperative period (presenting with ascites and lower body oedema) or later, when it is due to regeneration of the graft and twisting at the anastomosis of the hepatic vein to the cava. Thrombolytic therapy may be successful in late thromboses but should be avoided in early thromboses, as uncontrollable bleeding may occur from raw surfaces, particularly if a reduced-size liver was transplanted.

Hepatic venous outflow obstruction is suspected if there is persistence of ascites and is confirmed on Doppler ultrasound and transjugular venography. Liver biopsy findings of congestion and red cell extravasation around central veins may also be evident. An expandable stent placed via the transjugular route may be curative.

47.7 Retransplantation

Graft failure may occur at some time in 10–20% of patients, who then need retransplantation. Early indications may be primary nonfunction, early hepatic arterial thrombosis, severe drug-resistant acute rejection, and established chronic rejection. Early retransplantation is technically a much less traumatic procedure than the original transplant, although the patient may be in a poorer condition. Outcome depends largely on the indication for retransplantation; it is quite good for technical causes but less satisfactory for rejection or infection. An increasingly poorer outcome can be expected after third and fourth retransplants, and the efficacy and ethics of these interventions are in question.

47.8 Long-Term Survival and Quality of Life

One-year survival higher than 95% is being achieved in the best centres, with predicted 10-year survival of at least 75%. Patients receiving grafts for acute liver failure have done less well, with a higher early death rate usually associated with cerebral complications and multiorgan failure. Excellent quality of life can be achieved, and most children are fully rehabilitated.

It is increasingly evident, however, that prolonged cholestatic jaundice and malnutrition in infancy may have late effects. Despite good physical rehabilitation, evidence of significant cognitive deficits is common; these present during early schooling as learning difficulties and attention deficit disorder. As in any immunosuppressed patient, there are toxic side effects, particularly renal function impairment and greatly increased lifetime incidence of neoplasia.

Conclusion

Careful planning, extensive preparation of personnel, a broad base of skills, and good teamwork between health professionals are required for the development of a successful paediatric transplant programme. Surgical technique, anaesthetic skills, and medical care of the highest order are essential.

A patient with a liver transplant is a patient for life and requires complete commitment from the transplant medical and surgical team. This commitment cannot be abrogated after the patient is discharged from hospital.

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Spina Bifida

48

Jonathan R. Ellenbogen and Conor L. Mallucci

Neural tube defects (NTDs) occur in 1 in every 1000 births, arising as a result of aberrations in the embryologic development of the neural tube. They encompass a variety of congenital anomalies ranging from spina bifida occulta to anencephaly. Geographic variations exist in the incidence of spina bifida and other NTDs. Lower socioeconomic groups have a higher incidence of developing spina bifida, and the incidence is higher in Caucasians than in black people. NTDs are thought to be multifactorial in origin, with both genetic and environmental factors playing a role in their development.

Whilst spina bifida remains the most common congenital central nervous defect, the overall incidence of NTDs is in decline. Several factors are thought to account for this change: increased antenatal diagnosis, periconceptual folic acid, changing social attitudes, and improved standards of living and nutrition.

48.1 Types of Neural Tube Defects

The term *spina bifida* encompasses a range of malformations now also grouped under the general term *spinal dysraphism*; various other terms have been used, including *spina bifida occulta* and *spina bifida cystica* (including meningocele and myelomeningocele).

The term *spina bifida occulta* refers to the form of 'closed' spinal dysraphism (with intact skin cover) not accompanied by the extrusion of the contents of the vertebral column. There may be congenital loss of the spinous process and variable amounts of lamina without exposure of neural tissue and/or skin changes heralding underlying conus and filum anomalies such as lipomyelomeningocele.

A meningocele is an epithelium-lined sac filled with cerebrospinal fluid (CSF), which communicates with the spinal subarachnoid space. There is a defect in the vertebral arches with extension of the meninges through the defect, but no abnormality of neural tissue. It accounts for about 5% of all spina bifida cystica cases. Meningocele, which is most frequently observed in the lumbar region, is usually not associated with neurologic deficit or hydrocephalus.

Myelomeningocele is one of the most common congenital malformations and the most common form of NTD. There is a congenital defect in the vertebral arches and the overlying vertebral fascia is absent over a variable number of vertebral segments. There is cystic dilatation of the meninges with underlying structural and functional abnormality of the spinal cord. A neural plaque is centrally placed, around which is a cystic lesion with attenuated meninges and skin. CSF usually leaks from the exposed neural plaque. Although the pathological changes are obvious at the site of the lesion, additional changes involve the whole of the nervous system and other systems, especially the genitourinary and skeletal systems. Most patients have some degree of sensory loss and paralysis of the lower limbs, with muscle power imbalance resulting in flexion deformity of the hips and hyperextension of the knees. Bilateral hip dislocation and clubfoot deformity are common associated problems. Innervation to the bladder and bowel is abnormal, resulting in problems with continence and more typically a neuropathic bladder. The resultant morbidity is significant and affects normal ambulation, preservation of renal function, and social continence. Nearly all infants who are born with myelomeningocele have the Arnold–Chiari type II malformation, which includes a constellation of anomalies that include hindbrain herniation (downward displacement of the medulla, fourth ventricle, and cerebellum into the spinal canal), brainstem abnormalities, low-lying venous sinuses, and a small posterior fossa. This malformation is also associated with hydrocephalus (requiring a ventriculoperitoneal shunt in approximately 60–90% of children) and developmental brain abnormalities.

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48.2 The Role of Foetal Surgery

There is a potential role for foetal surgery as neural tube defects are diagnosed prenatally. Techniques using endoscopy, percutaneous foetoscopic patch coverage, and hysterotomy have been described. The Management of Myelomeningocele Study (MOMS) was a three-centre, randomized prospective trial in the United States that compared prenatal surgery (before 26 weeks of gestation) versus standard postnatal repair of myelomeningocele. The study found that prenatal surgery for myelomeningocele reduced the need for CSF shunting (40% in the prenatal-surgery group and 82% in the postnatal-surgery group) and improved the composite score for mental development and motor function at 30 months. Prenatal surgery was associated with increased foetal risks, however, and an increased risk of preterm delivery and uterine dehiscence at delivery. Foetal surgery is a very complex procedure performed in a limited number of centres. This chapter therefore discusses only the accepted postnatal surgical approach.

48.3 Postnatal Surgery

48.3.1 Preoperative Care

Before closure of the defect, informed consent is obtained from the child's parent or legal guardian. The initial management is aimed at stabilizing the infant, with the aim that all actively treated spina bifida patients should undergo closure of the defect within 24–48 h after delivery. Early defect closure prevents infection and further damage to exposed neural tissue; the aim is to preserve existing function. A minority of children have such severe deformity that primary closure may not be indicated. These patients are usually managed by supportive nursing care, and closure is performed later if the child survives.

The infant should be kept euvolemic and normothermic, with latex precautions in place because 73% of these infants are allergic to proteins present in latex. The myelomeningocele should be covered in sterile, saline-moistened gauze and wrapped in plastic cling-film to minimize dehydration and prevent contamination. The infant is nursed lateral or prone to ensure that no pressure is placed on the neural placode.

48.3.2 Surgical Technique

The principle of surgery is to reconstruct the spinal cord by five-layer closure of the pia, arachnoid, lumbar fascia, subcutaneous tissue, and skin. The vascular supply to the neuroplaque is maintained and unnecessary neural injury is avoided.

After general anaesthesia and insertion of an endotracheal tube, the patient is turned prone and cotton (Gamgee) supports are placed under the pelvis and chest (Fig. 48.1). The head is turned to the right through 90°. It is important to

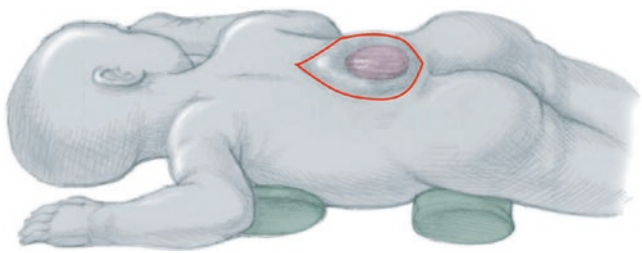

Fig. 48.1 Patient positioning

place additional supports beneath each foot. It is not necessary to have blood available for this procedure, but most surgeons would have a group and save order with the laboratory. A neonatal diathermy pad is usually applied to the abdomen or chest. A warming bear-hugger is routinely used. Bipolar diathermy should be used throughout. The table is in the Trendelenburg position to minimize CSF drainage from the spinal defect. Swabs are taken from the lesion for microbial examination and culture.

An antiseptic soak is placed over the anus. The lesion is covered with a warm swab, and the surrounding skin is prepped with chlorhexidine and draped with generous margins to facilitate additional skin dissection if required. An incision is made at the lowermost portion of the sac using a sharp scissors or scalpel. The skin is incised at the junction of the neural placode arachnoid membrane and the dystrophic epidermis. This incision is carried cranially, staying away from the neural plaque, which should not be handled where possible. Incision into the sac results in a release of CSF and some bleeding, which is easily dealt with using bipolar diathermy. Nerve roots and blood vessels are now seen traversing the sac and disappearing anteriorly through the dural layer; they should be preserved. The dural layer is clearly visible as a whitish, fibrous layer. The sac is elevated, taking care not to traumatize the neural plaque. The membrane between the edge of the skin defect and the neural plaque is removed carefully to avoid inclusion cysts (dermoid/lipoma) (Fig. 48.2). Bleeding from the neural edge is dealt with using bipolar diathermy and with minimal handling of the plaque itself. At intervals, chlorhexidine soaks may be applied to the area.

The plaque will now be seen to lie on an easily recognizable dural layer. The dura is freed laterally and then superiorly and inferiorly to normal intact dura, and the layer is carefully dissected from the underlying fascia. This dissection is carried medially until the nerve roots come into view (Fig. 48.3). Epidural veins may prove difficult at this point, but these can be controlled by bipolar diathermy.

The mobilized dural layer is now tubularised over the exposed neural plaque, using a continuous monofilament 6/0 absorbable suture on an 11-mm round-bodied needle (Fig. 48.4). A watertight closure is the aim. Occasionally it is not possible to complete the dural tube completely, and a small portion of vertebral fascia is used to achieve closure. This is preferable to compromising the plaque. Postoperative CSF leakage may herald the development of hydrocephalus,

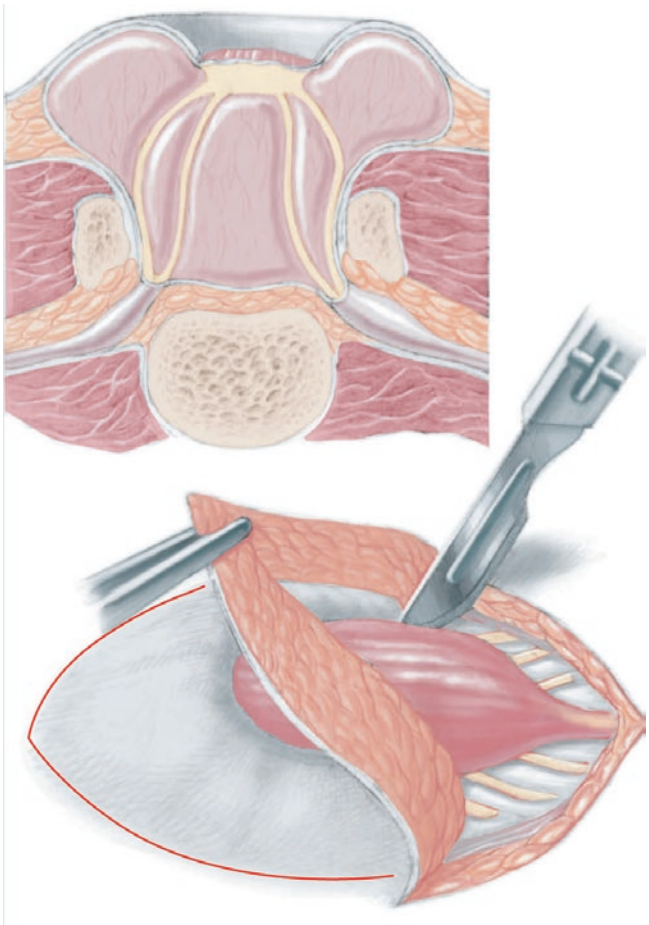

Fig. 48.2 Removal of the membrane between the edge of the skin defect and the neural plaque (*bottom*)

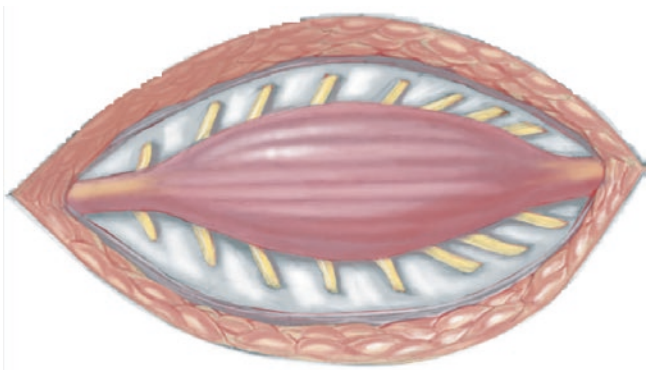

Fig. 48.3 Freeing the dural layer exposes the nerve roots

which usually responds to the insertion of a ventriculoperitoneal shunt. Where possible, the dural tube is reinforced using an additional lumbodorsal fascial covering obtained by mobilizing the fascia from the underlying muscle. It is usually impossible to cover the dural tube completely with fascia, especially at the lower end.

The subcutaneous tissue is closed with 3/0 absorbable interrupted sutures, and then the skin is closed with interrupted nylon 4/0 stitches (Fig. 48.5). Occasionally skin closure requires considerable skin undermining and rarely the use of lateral releasing skin incisions or complex flap repairs involving plastic surgery. The skin closure is supported with wound strips and a dressing is applied.

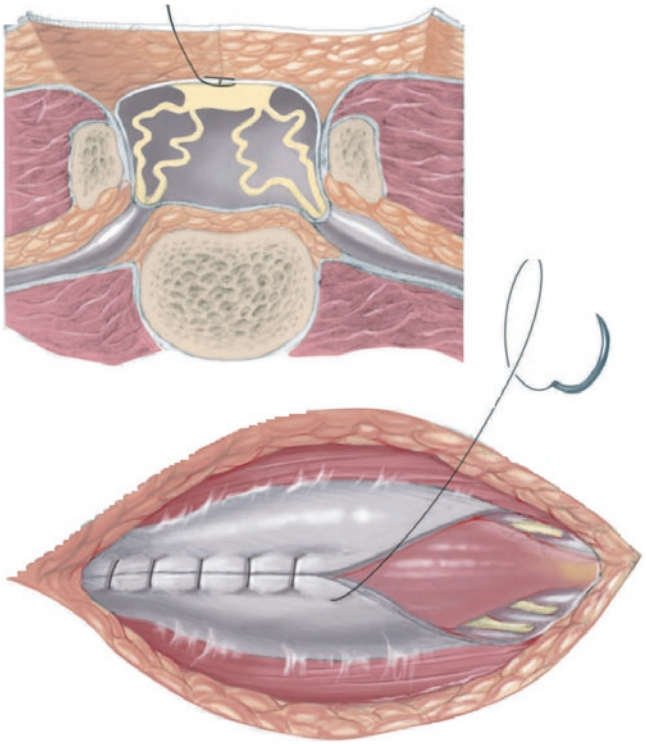

Fig. 48.4 Suturing of the dural layer to create a dural tube

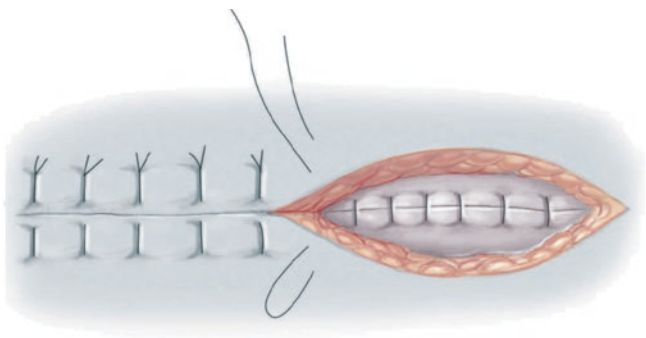

Fig. 48.5 Closure of the subcutaneous tissue and the skin

48.3.3 Postoperative Care

The infant is nursed in a prone or a lateral position. A plastic drape is positioned between the wound and the anus to prevent contamination of the wound. Careful attention is paid to keeping the area clean. Feeding is commenced once the bowel starts working. The wound is periodically inspected.

Regular head circumference measurements are performed and can be supplemented by cranial ultrasound to monitor progress of the associated hydrocephalus. A ventriculoperitoneal shunt is inserted if the head circumference rises precipitously or the ventricular diameter increases beyond 50–60% of the diameter of the skull and progressive ventriculomegaly is confirmed on serial scans.

48.4 Other Surgical Considerations

Associated vertebral kyphosis may occasionally require an osteotomy to facilitate closure in the primary setting. This is more likely when lesions are initially treated conservatively and back closure is performed as a secondary event.

Meningocele is closed as an elective procedure. Because there is no exposure of neural elements, closure is straightforward, with sac excision and closure over a drain.

48.5 Management Strategy

The most appropriate management strategy for these patients is a continued source of medical, ethical, and legal debate. A multidisciplinary team approach incorporating neurosurgeons, paediatricians, neurologists, endocrinologists, urologists, orthopaedic surgeons, physiotherapists, social workers, psychologists, and nursing staff must be utilized, as diagnosis of a neural tube defect may have devastating consequences for both the patient and the family. The aim is to provide the patient and family with the least disability possible and therefore the best quality of life achievable.

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Hydrocephalus

49

Kai Arnell and Tomas Wester

Hydrocephalus is defined as an excessive amount of cerebrospinal fluid (CSF) under increased pressure in the ventricular system. The incidence is 0.4–2.5 per 1000 live births. CSF is mainly produced by the choroid plexus of the ventricular system. It is circulating through the foramen of Monro to the third ventricle and then through the aqueduct of Sylvius to the fourth ventricle. The CSF enters the subarachnoid space through the foramina Luschka and Magendie. It is absorbed into the blood stream by the arachnoid villi on the surface of the brain. The CSF production is age-dependent. Neonates produce approximately 25 mL/24 h, and adults produce approximately 700 mL/24 h. The intraventricular pressure at rest varies between 0 and 10 cm H_2O.

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49.1 Causes

The cause of hydrocephalus can be obstruction to the flow of CSF (noncommunicating hydrocephalus), overproduction of CSF, or failure of its re-absorption (communicating hydrocephalus). Pathological processes that cause hydrocephalus are congenital malformations, neoplasms, bacterial meningitis, prenatal infections (toxoplasmosis, listeriosis, or cytomegalovirus infection), subarachnoid or intraventricular haemorrhage, and overproduction of CSF by choroid plexus papillomas. The three most important malformations that cause congenital hydrocephalus due to obstruction of the CSF flow are aqueductal stenosis, Arnold-Chiari II malformation, and Dandy-Walker malformation. Aqueductal stenosis accounts for about 15% of cases of hydrocephalus and occurs in several anatomical forms. Myelomeningocele, when combined with Arnold-Chiari II malformation, is associated with hydrocephalus in about 90% of cases. This malformation leads to CSF flow obstruction due to herniation of the medulla oblongata, cerebellar tonsils, and vermis through the enlarged foramen magnum and compression of the fourth ventricle. The Dandy-Walker malformation is a developmental defect of the foramina of Luschka and Magendie, with absence of the cerebellar vermis, a distended fourth ventricle, and an enlarged posterior fossa. Hydrocephalus can be an isolated finding but it can also be part of various malformation syndromes, such as Meckel-Gruber, Walker-Warburg, and X-linked hydrocephalus.

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49.2 Clinical Features

An important clinical feature of hydrocephalus in neonates and infants is enlargement of the head circumference, which can easily be overlooked if the head circumference is not measured repeatedly. The shape of the head is often abnormal. The anterior fontanel is large and bulging. Cranial suture separation and distended scalp veins are common. The" sunset sign" may be present. Vomiting, behavioural changes, drowsiness, and headache are common symptoms in infants and young children with hydrocephalus. Other symptoms are failure to thrive, irritability, delayed motor and psychosocial development, and mental retardation.

49.3 Diagnosis and Monitoring

Ultrasound, CT scans, and MRI distinguish hydrocephalus from other conditions. Hydrocephalus is frequently diagnosed with antenatal ultrasound. Ultrasound is also the method of choice for serial examinations of premature babies to detect intracranial haemorrhages and secondary hydrocephalus. CT and particularly MRI are useful to show the detailed anatomy and reveal the cause of hydrocephalus as well as associated abnormalities or neoplasms. The investigation of the patient with hydrocephalus also includes a TORCH screen and chromosomal analysis if a prenatal infection or a chromosomal abnormality is suspected. The patient should undergo an eye examination, and associated anomalies such as a cardiovascular malformation should be excluded.

Mild hydrocephalus, caused by intracranial haemorrhage, is sometimes monitored by repeated measurement of the skull circumference, ultrasound scans, and clinical signs of progressive hydrocephalus; it may finally resolve without treatment. On the other hand, in cases with progressive ventricular dilatation, rapidly increasing skull circumference, and clinical features of increased intracranial pressure, the condition requires treatment by surgical diversion of CSF.

49.4 Surgical Treatment

Ventriculoperitoneal shunts are the primary option for surgical diversion of CSF in infants and children. Ventriculoatrial shunts are used only under certain circumstances because of the risk of serious sequelae such as thrombosis, pulmonary embolism, and septicaemia. Many different shunt systems are available. Systems with a programmable valve are preferred.

Endoscopic third ventriculotomy has become an option particularly for treatment of aqueductal stenosis. The technique eliminates the need for a shunt and thereby the complications of shunting. There appear to be a correlation between success rate and age, with poorer outcome in infants less than 6 months of age, although conflicting data have been reported.

49.4.1 Preoperative Routines

The preoperative routines are important to minimize the risk of *Staphylococcus epidermidis* shunt infections. The patient should be carefully prepared before the operation. Cloxacillin is administered as prophylaxis at induction of general anaesthesia.

The child is placed in a supine position on the operating table with the head turned to the left if the shunt is to be placed on the right side (Fig. 49.1). The right shoulder and neck are elevated to stretch the skin and avoid creases that make the subcutaneous tunneling of the abdominal catheter difficult. The scalp should be shaved and well prepared. The placement of the incision depends upon whether a frontal or a parieto-occipital approach is selected for ventriculoperitoneal shunting.

Fig. 49.1 Patient positioning for placement of a ventriculoperitoneal shunt

49.4.2 Frontal Approach

A frontal approach has several advantages. Less brain is traversed than with the posterior approach, and the distance to the ventricle is shorter, which makes cannulation of very small ventricles easier. The disadvantage of a frontal approach is that a longer subgaleal tunnel is required and an extra occipital incision is needed for tunneling. The sagittal and coronal sutures are marked, as well as the place of entrance of the ventricular catheter, which should be located 2 cm from the midline and 2 cm anterior to the coronal sutures. In neonates and infants with massive hydrocephalus, the catheter will be placed in the fontanel. In most cases, the ventricular catheter will be inserted at the edge of the fontanel or through the frontal bone (Fig. 49.2). It is important to avoid having any part of the shunt system lying under the wound. Prepare the child and drape it in a sterile fashion. The entire operation field should be covered with a plastic sheet to avoid contact between the shunt system and the skin. The skin is very thin in neonates, particularly in preterm babies, and the blood supply is vulnerable, so infiltration with xylocaine and adrenaline is not used. Bipolar diathermy is preferred for haemostasis. The flap is carefully dissected from the underlying galea and a subcutaneous stay suture is put in the flap. When entering at the edge of the fontanel, the galea is opened and a circular hole is nipped in the bone edge. In older children with bone under the flap, the periosteum is removed. A burr hole is made. The bone is very thin in neonates and infants, and it is important to be careful. The dura is coagulated using a fine-pointed forceps, and a small opening is made. Ventricular catheters may be either straight or prebent, but prebent catheters are recommended for infants and children. From a CT scan, the optimal length of the ventricular catheter can be estimated. The ventricular catheter is introduced through the small opening to get a snug fit and avoid leakage of CSF. The direction of the ventricular catheter is chosen so that the tip will be located in the anterior horn of the right ventricle. The catheter is passed through the brain perpendicular to the skull, aiming at the inner canthus of the ipsilateral eye.

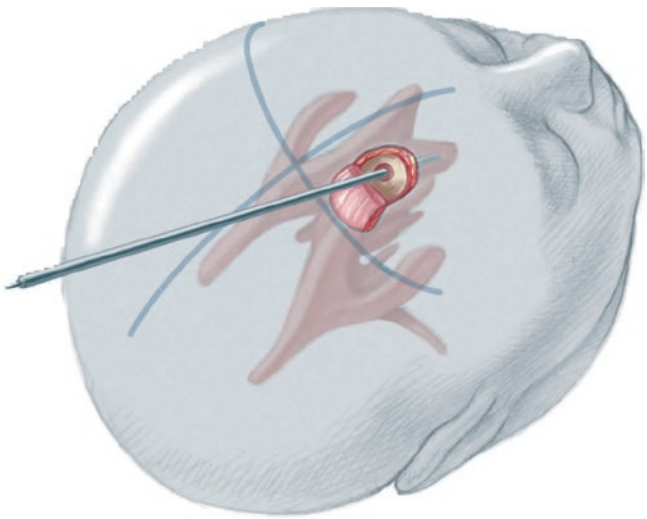

Fig. 49.2 Location of the ventricular catheter using a frontal approach

49.4.3 Parieto-Occipital Approach

In a parieto-occipital approach, the scalp incision is semicircular, lying well behind the posterior parietal eminence. Care must be taken to avoid having any part of the shunt system located under the wound, which would predispose for breakdown of the wound and infection. The burr hole is placed 3 cm above and behind the ear. The catheter is passed parallel to the sagittal suture, aiming at the glabella (Fig. 49.3). The length of the ventricular catheter should allow the tip to extend 1 cm anterior to the coronal suture in order for the catheter to be placed in the anterior horn.

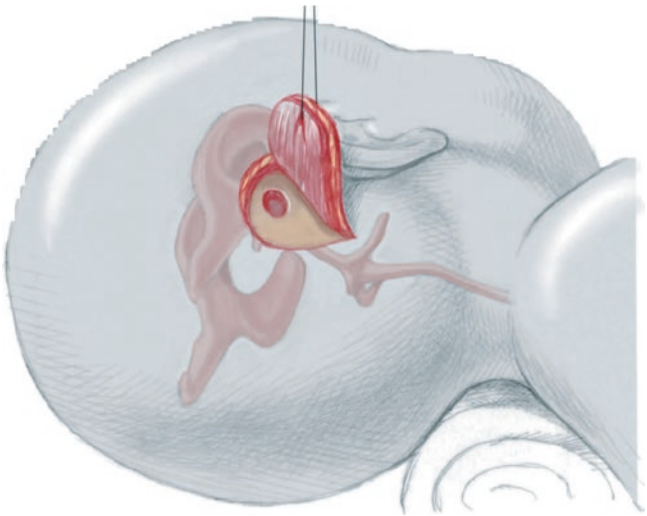

Fig. 49.3 Location of the ventricular catheter using a parieto-occipital approach

49.4.4 Completion of Shunt Placement

The abdominal incision is made 2 cm below the costal margin, over the rectus abdominis muscle (Fig. 49.4). The muscle fibres are split longitudinally and the peritoneal cavity is opened.

The tunneling device is pushed from the scalp incision subcutaneously to the abdominal incision (Fig. 49.5). The passage over the clavicle is controlled to avoid skin contusion or perforation. The stylet is withdrawn and the peritoneal end of the catheter is pushed through the tunneling device.

The ventricular catheter is introduced into the lateral ventricle in the direction of the glabella and the stylet is withdrawn. The valve is pressed to test the function. CSF should be seen dripping distally. The scalp incision is closed with absorbable subcuticular sutures. The system is tested once again before the abdominal catheter is inserted through the small opening into the peritoneal cavity (Fig. 49.6). At least 35–40 cm of the catheter can be placed in the abdomen of a full-term baby. The peritoneum is closed in a water-tight manner, as are the layers of the abdominal wall. Absorbable sutures are used for the skin closure.

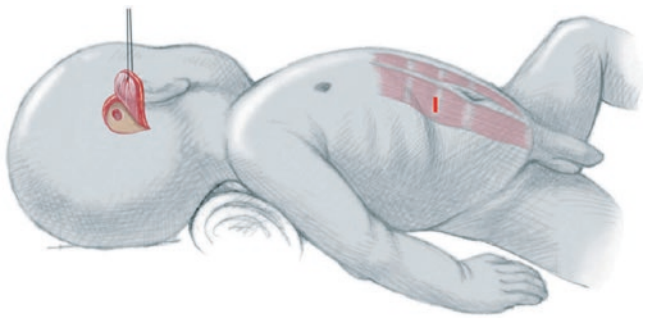

Fig. 49.4 The abdominal incision

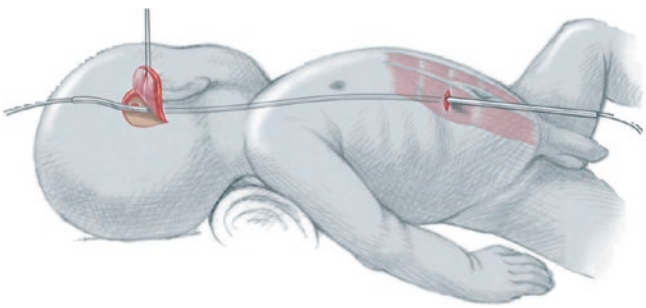

Fig. 49.5 Use of the tunneling device

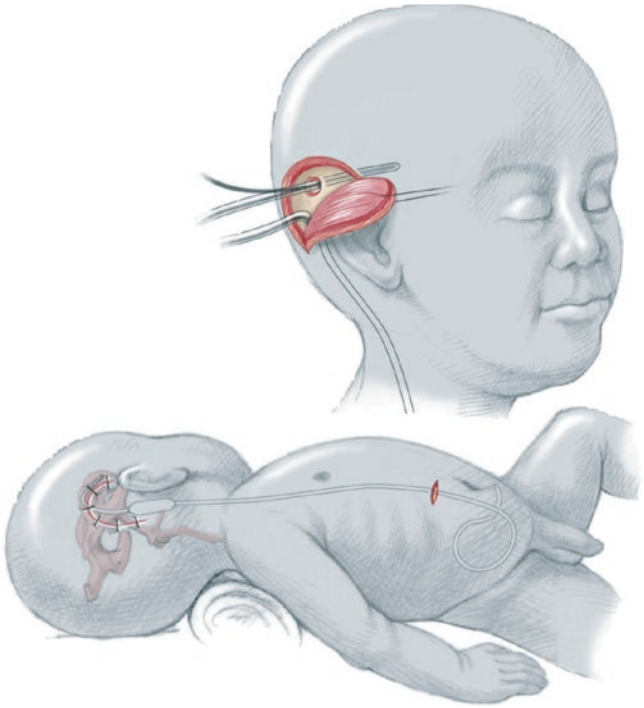

Fig. 49.6 Insertion of the catheter into the peritoneal cavity

49.5 Complications

Shunt complications are relatively frequent, owing to infections, catheter obstruction, improper placement of the catheters, and short or disconnected catheters. Despite careful preparation and antibiotic prophylaxis, there is a risk of perioperative infections with *Staphylococcus epidermidis*, *Staphylococcus aureus*, or *Proprionebacterium* species. Neonates and infants are more sensitive to infections and can also have haematogenous infections with origin in the urinary tract or respiratory tract. The incidence of infections has been reported to range from 10% to 15%. The infections are treated by removal of the infected shunt system, external ventricular drainage, and use of intravenous and intraventricular antibiotics before insertion of a new system.

Technical advances have led to the development of shunt systems that are much more sophisticated than those initially produced 50 years ago. The programmable valves can be noninvasively adjusted to a suitable pressure, giving the brain optimal conditions for development. In spite of the introduction of programmable valves, however, problems due to overdrainage are encountered, resulting in slit ventricles. An antisiphon device can be inserted between the valve and the abdominal catheter to reduce this problem. There are also valves available that include an antisiphon mechanism.

The incidence of reoperations due to shunt malfunction used to be very high. Today, the incidence has decreased because of the awareness of strict preoperative and perioperative routines, antibiotic prophylaxis, and the use of longer catheters.

49.6 Outcome

The long-term outcome of children with hydrocephalus depends on many factors, such as the severity and duration of the ventricular dilatation, the aetiology of the hydrocephalus, associated intracranial malformations, and whether severe infections have occurred. It is important to follow these patients and monitor their neurologic and psychosocial function as well as their growth and onset of puberty.

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Dermal Sinus

50

Jonathan R. Ellenbogen and Conor L. Mallucci

A dermal sinus tract is a form of occult spinal dysraphism. Embryologically, it results from a focal incomplete disjunction of the surface ectoderm and dermal elements from the neuroectoderm. This process likely occurs between the third and eighth weeks of gestation. Later during embryogenesis, the spinal cord ascends relative to the spinal canal and stretches the adhesion cephalad into a long, tubular tract. Dermal sinus tracts are therefore abnormal communications extending from the skin surface to the spinal fascia, dura mater, or neural elements, and are lined with epithelium. Although dermal sinuses can occur anywhere from the upper cervical region to the midsacrum, they are most commonly found in the midline in the lumbar or lumbosacral area. Some dermal sinuses end blindly within the soft tissues superficial to the underlying lamina, but 60–70% reach the subarachnoid space, half attaching to the filum or the conus.

50.1 Diagnosis

A dermal sinus usually manifests as a small dimple or pinpoint ostium, which is often associated with cutaneous findings of hyperpigmentation, hypopigmentation, angiomatous skin changes, hypertrichosis, subcutaneous lipomas, or drainage of debris or fluid from the pit. They occur in the midline or rarely in a paramedian location.

Dermal sinus tracts include both dermal and epidermal elements, and may be associated with CSF leakage, intradural dermoid or epidermoid cysts, spinal cord tethering, or (rarely) teratomas. Diagnosis may be aided by ultrasound (usually up to 6 months of age) or MRI. Ultrasonography readily demonstrates the subcutaneous tract, intraspinal inclusion tumours, and diminished cord pulsations. MRI, including T1-weighted sagittal and axial MRI with gadolinium injection and fat saturation, is the gold-standard diagnostic technique.

Clinical and radiologic features can appear innocuous, leading to delayed diagnosis and failure to appreciate the extent of the abnormality. If the dermal sinus is left untreated, the main complication is infection, with meningitis or intradural abscesses. These infections may lead to permanent neurologic deficits, so dermal sinuses must be diagnosed and operated on early in life. The entire midline area of the skin from the skull to the sacrococcygeal region must be carefully inspected when a child of any age suffers recurrent meningitis or meningitis from an unusual organism. A high index of suspicion must be maintained for all dimples above the intergluteal fold despite a normal examination or neuroradiologic studies. Other common presenting symptoms include those of tethered cord, spinal cord compression secondary to intradural dermoid cyst growth, and hydrocephalus secondary to recurrent meningitis.

Dermal sinuses must be differentiated from benign coccygeal pits, which end blindly and never extend intraspinally, and therefore do not require further imaging evaluation or treatment. Dermal sinus tracts are found above the natal cleft and are usually directed superiorly. By comparison, coccygeal pits are found within the natal cleft, below the top of the intergluteal crease, with a tract extending either straight down or inferiorly. Coccygeal pits occur over the lower sacrum and coccyx and are anatomically located below the level of the subarachnoid space. They are encountered in nearly 5% of newborns, and although they are present from birth, they rarely manifest themselves before adult life. In later years, these small pits or dimples may become pilonidal sinuses or abscesses.

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50.2 Treatment

Dermal sinuses should be electively resected at the time of diagnosis, regardless of patient age. Conservative management of dermal sinuses is not justified because of the potential for complications resulting in meningitis and permanent neurologic dysfunction. General procedure-related morbidity is acknowledged to be very low, with postoperative infection being the most common complication.

50.2.1 Elective Resection

Treatment of these lesions consists of excision of the dimple and the tract, as well as any intradural connections or masses. Even when MRI reveals no intradural anomalies, dermal sinus tracts located above the gluteal crease should be explored to their termination. Surgery in advance of deficits aims to maintain normal neurologic function, allowing these children to develop unimpeded by infection or by motor or bladder paralysis.

After induction of general anaesthesia and placement of a Foley catheter into the bladder, the patient is positioned prone with lateral padded rolls supporting the chest and abdomen. The arms of children less than 2 years of age are best supported alongside the trunk, whereas in older patients, elevation above the shoulder positions the surgeon closer to the patient. Chlorhexidine solution is used to prepare the skin from the intergluteal fold to many spinal levels above the sinus tract. Perioperative administration of intravenous antibiotics is strongly recommended. An elliptical skin incision encircling the sinus opening, and any abnormal skin surrounding it, is made to excise fully the dermal sinus (Fig. 50.1). Purulent material or drainage should be cultured in aerobic and anaerobic medium.

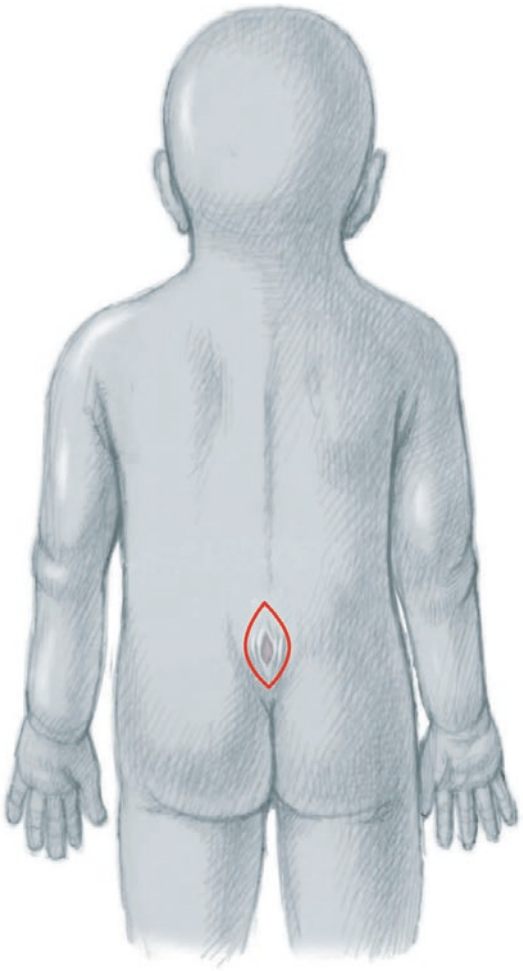

Fig. 50.1 Skin incision encircling the sinus opening

The subcutaneous tissue is divided to expose the fascial defect, and the sinus stalk is circumferentially dissected (Fig. 50.2). Cephalad to the stalk, the paraspinal muscles are elevated with electrocautery in a subperiosteal fashion from the first intact spinous process and lamina. Preparations must be made to continue bone removal across several laminae until the site of attachment to the dura is identified.

If imaging studies or intraoperative observation indicate that the lesion penetrates deeper than the fascia, the dissection should proceed along the tract until its termination is reached.

The dura is opened with an elliptical incision encompassing the tract. Some sinus tracts abruptly end with dural attachment, which is readily apparent after dural opening. In these cases, after confirming normal intradural anatomy, the dura is closed and the wound is closed in layers. When the stalk continues and intradural lesions such as a dermoid or epidermoid cyst or tethered spinal cord are present, then the dissection must proceed into the intradural space (Fig. 50.3). Further dissection is performed with loupe or microscope magnification. Dissection of the stalk from this disordered glial mass may be accomplished using sharp microsurgical technique.

Intra-operative ultrasonography may be useful for identifying syringomyelia or intramedullary dermoid at the site of stalk attachment.

Regardless of the attachment anatomy, a comprehensive inspection should be carried out to look for arachnoid adhesions, dermoid tumours, and a thickened filum terminale (Fig. 50.4). Dermoid inclusion tumours are frequently multiple and can be solidly adherent to the filum and nerve roots within the cauda equina, especially when meningitis has occurred.

Intradural cysts should be completely removed, without opening if this is possible and does not endanger neural elements. Intraspinal and adherent intradural cysts are emptied of their contents and as much as possible is

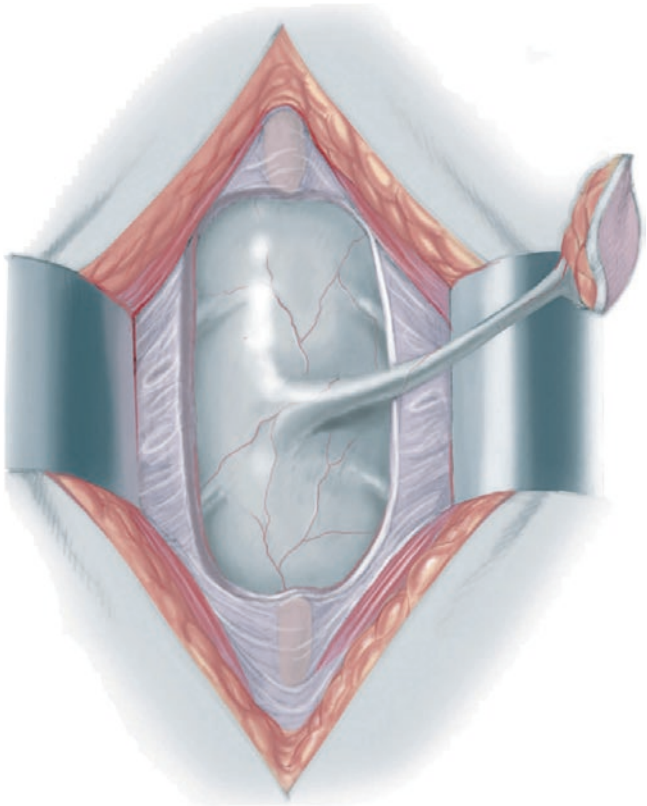

Fig. 50.2 Exposure of the fascial defect and dissection of the sinus stalk

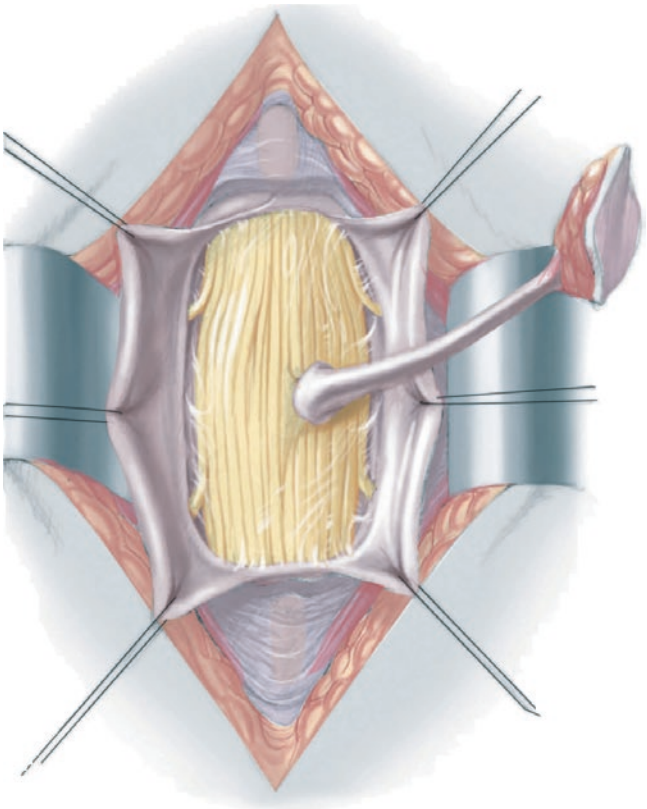

Fig. 50.3 Dissection into the intradural space

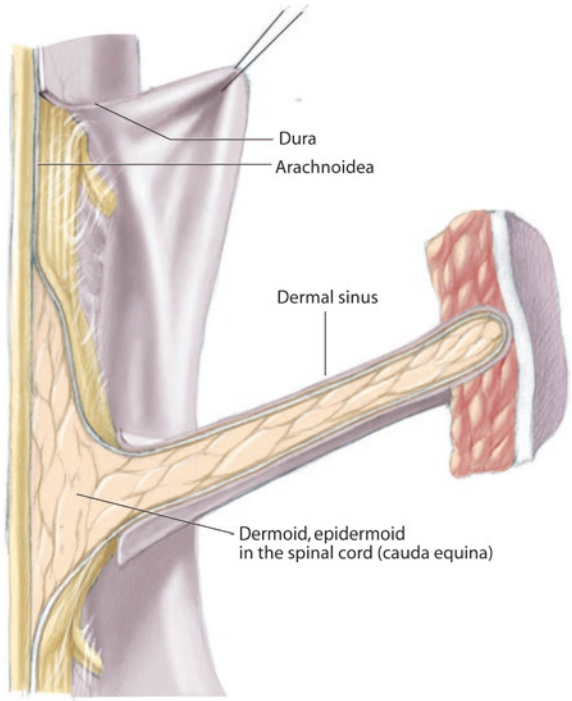

Fig. 50.4 Cross-sectional anatomy

removed, but attempting to remove a hard fibrous capsule densely adherent to neural tissue or a capsule of infected intraspinal cysts is fruitless and may lead to avoidable cord or root injury. Duraplasty is performed if necessary, and the muscles and skin are closely approximated without drainage. Drainage of an extradural abscess after operation may be necessary.

Debulking dermoid cysts from within is the method of choice. The capsule, which may be adherent, can usually be dissected from the surrounding neural elements. Retention of an epithelial surface will result in recurrence of the dermoid cyst and necessitate a repeated operation. An attempt to remove the capsule should be undertaken to prevent further recurrence of the lesion. The initial dissection has the highest likelihood of achieving complete resection. Incompletely resected dermoid tumours may grow slowly over time, and the density of postoperative adhesions and scar preclude total resection at reoperation. Intraoperative neurophysiology monitoring may be useful for complex intradural cases with adherent nerve roots, to avoid any new nerve root injury.

Following complete tract and inclusions tumour resection, the subarachnoid space should be irrigated with a saline solution. Dermoid and epidermoid debris are highly irritative to the spinal fluid, and this manoeuvre may diminish postoperative inflammatory meningitis. If an intramedullary mass has been resected, the pia arachnoid might be sutured and the tubular spinal cord reconstituted.

To minimize postoperative spinal cord tethering, a dural patch graft can be incorporated to ensure a wide contact of cerebrospinal fluid around the lower spinal cord and cauda equina. Fibrin glue may be employed if necessary. The wound is closed in layers, and the paraspinal muscle fascia is reapproximated with running and interrupted sutures in a watertight manner. Skin closure with vertical mattress sutures is preferred, especially if there has been previous infection.

50.2.2 Emergency Surgery

If the lesion is discovered during an episode of meningitis, laminoplasty and intradural exploration should follow after the infection has been controlled by antibiotic therapy. Emergency surgery is required in cases of rapid neurologic deterioration, recurrent infection during antibiotic therapy, or when infection cannot be controlled quickly.

Surgical treatment consists of excision of the dimple and the tract from the skin surface to the deepest projection, including intradural connections or masses. Intradural exploration is warranted, as it may reveal previously unappreciated pathologic findings even when MRI findings are unremarkable.

50.3 Outcomes

The prognosis in patients who already have some neurologic deficit is frequently unfavourable, although some improvement may occur in most patients. Surgery is performed to prevent neurologic deficit in those who do not have it. In patients with incomplete tumour resection, follow-up MRI is recommended at least yearly.

Optimum management of children requires close cooperation among the multidisciplinary team encompassing paediatricians, paediatric surgeons, neurosurgeons, and the multiple specialists involved with congenital disorders and infectious diseases.

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Sacrococcygeal Teratoma

51

Kevin C. Pringle and Hiroaki Kitagawa

Sacrococcygeal teratoma is a rare tumour, occurring in approximately 1 in 40,000 live births. The tumour arises from the caudal end of the spine, usually protruding from the inferior end of the infant's spinal column and displacing the anus forwards. They are much more common in girls, with the female-to-male ratio being at least 3:1. It is generally agreed that sacrococcygeal teratoma is the result of continued multiplication of totipotent cells from Hensen's node, which fail to involute at the end of embryonic life.

These are true neoplasms. Willis (1962) defined the term *teratoma* as follows: "A teratoma is a true tumour or neoplasm composed of multiple tissues of kinds foreign to the part in which it arises". By definition, then, sacrococcygeal teratomata are composed of several types of tissue, usually derived from two or three germ layers. Within any one tumour, the cells can vary from totally benign (even forming well-formed teeth, hair, or other organs) to cells that appear frankly malignant. Although many sacrococcygeal teratomata contain malignant-looking cells (usually described as "immature"), if they are completely excised they do not recur. For this reason, the diagnosis of malignant sacrococcygeal teratoma can be made only if there are distant metastases. The risk of malignancy depends on the site and extent of the tumour and the age at diagnosis. Tumours diagnosed after 2 months of age have a high risk of being malignant. Tumours that are largely "exophytic"—those that protrude from the caudal end of the baby with only a small intra-abdominal component—tend to be benign. Those that have a large intra-abdominal component, however, have a higher risk of behaving in a malignant fashion. In general, as long as a complete resection is obtained, the risk of recurrence is low.

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51.1 Diagnosis

These tumours used to be diagnosed when they presented as a large sacral mass after a difficult delivery, or with an obstructed delivery, but the most common presentation now is an antenatal diagnosis by ultrasound. Series reporting the antenatal diagnosis of sacrococcygeal teratomata have revealed that a significant number of the fetuses diagnosed as having a sacrococcygeal teratoma are likely to die before delivery. Most of the fetuses who have died following antenatal diagnosis had tumours with a mass as great as or greater than the rest of the fetus. It is entirely possible, therefore, that these fetuses die of heart failure as the fetal heart is unable to pump sufficient blood to nourish both the tumour and the rest of the fetus. Certainly, in many of the antenatal series reported, fetal hydrops (nonimmune hydrops) is very common and is associated with an increased risk of fetal demise.

Most cases presenting as neonates to paediatric surgeons will have a large, skin-covered mass protruding from the coccygeal region, pushing the anus and vagina anteriorly. On the surface, large veins may be visible, which usually drain into the surrounding structures. Large tumours may have ruptured (in which case they will bleed profusely) or may have an ulcerated area on the surface. Neonates with a tumour approaching the size of the rest of their body may be delivered prematurely and will often have features of nonimmune hydrops. In all cases, the tumour is firmly attached to (and may be said to arise from) the anterior surface of the coccyx. It may displace the coccyx posteriorly, but the sacrum is almost always normal.

An abdominal ultrasound will determine the size and consistency of any pelvic or abdominal component. It may be necessary to fill the bladder with water to allow it to be used as a sonic window. MRI should clearly distinguish between sacrococcygeal teratoma and anterior meningocele, and may be able to detect the occasional extension of the tumour through the sacral hiatus into the spinal canal.

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51.2 Treatment

These lesions are best resected within the first 24 h after birth, because the gut is usually not colonized in the first 24 h, reducing the risk of infection if the field is contaminated by stool during the resection. Perioperative antibiotics are given immediately before surgery commences and continued for 24–48 h postoperatively. Blood should be cross-matched, and adequate intravenous access is vital. An arterial line may also be useful during the operation. It is useful to obtain blood for α-fetoprotein levels before surgery as a baseline, in order to confirm post-operatively that α-fetoprotein levels continue to fall at a normal rate.

51.2.1 Positioning and Incision

The patient is anaesthetized, intubated and positioned prone with a roll under the hips. The roll is positioned so that the infant's weight is taken on the anterior superior iliac spines (Fig. 51.1). It is vital that the abdomen be left hanging free to ensure that respiration is not inhibited by the baby's weight. For this reason, the baby's shoulders should be supported either by a smaller roll lying transversely across the apex of the chest at the level of the medial ends of the clavicles, or by two rolls running parallel to the spine, each supporting the glenohumeral joints. A catheter should be placed in the bladder to measure urine output throughout the procedure. Many authorities state that the anus should be prepared out of the field. We find that approach both inconvenient and impractical, as access to the anus is often required during the procedure. A Vaseline gauze pack placed through the anus into the rectum (with a suture attached to ease later retrieval) can make it easier to identify the rectum later in the dissection. The cautery pad usually can be placed across the shoulders. A clear plastic drape may conserve body heat and assist in prevention of hypothermia.

A chevron incision is made in the skin over the dorsum of the mass. It is continued down to fascial layers. It is preferable not to dissect beyond the level of the deep fascia at this stage of the dissection. There are often several large veins in the subcutaneous tissue on either side of the midline. These should be divided between ties. The incision should be placed so as to preserve as much normal skin as possible. Excess skin can always be trimmed later if necessary. The apex of the chevron should be over the lower sacrum.

51.2.2 Dissection

In the midline, the dissection should continue directly down to the sacrococcygeal junction, or even down to the fourth or fifth sacral vertebra. The edges of the sacrum are defined, and a clamp is passed across the sacrum at this level, keeping the tips of the forceps against the ventral surface of the bone (or cartilage) to ensure that the forceps pass between the sacrum and the underlying median sacral vessels, which are usually substantial vessels supplying the bulk of the blood supply to the tumour. Once this manoeuvre is complete, the sacrum (which is usually completely, or at least largely, cartilaginous) can be divided with a scalpel at the level of the 5th sacral vertebra and the tumour displaced slightly inferiorly to expose the median sacral vessels (Fig. 51.2). This manoeuvre needs to be carried out with caution, as occasionally the bulk of the venous drainage from the tumour passes through the sacral hiatus and into the epidural plexus. Failure to recognize this possibility can result in a substantial, rapid loss of blood. This bleeding usually can be controlled using simple pressure, taking care to maintain the pressure until the bulk of the arterial inflow has been divided. Even after that has been accomplished, however, blood loss from the epidural plexus may continue, requiring the use of bone wax to control it. It may be necessary to divide some of the attachments of the thinned-out remnants of the levators to the edges of the lower end of

Fig. 51.1 Positioning of the infant for removal of a sacrococcygeal teratoma

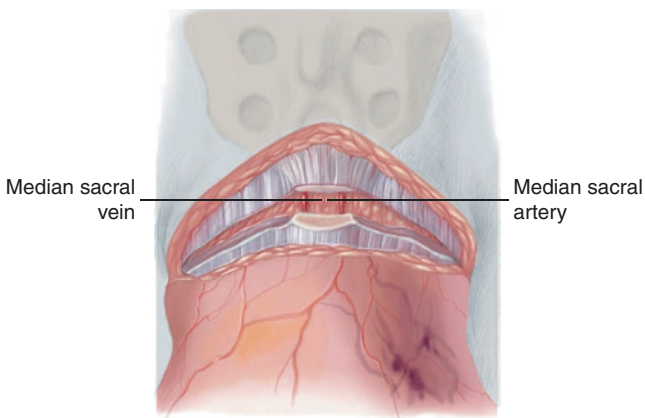

Fig. 51.2 Division of the sacrum to expose the median sacral vessels

the sacrum and coccyx to enable the coccyx to be displaced caudally. The median sacral vessels are then ligated in continuity and divided. This early division of the median sacral vessels is essentially the same as the procedure advocated by Smith and colleagues. This manoeuvre opens a plane of dissection that is outside the tumour capsule, but deep to the thinned-out remnants of the levators and the gluteus maximus. The levators may be so thin as to be almost invisible, but they will contract on stimulation, either with a muscle stimulator or the electrocautery. The dissection should continue laterally in this plane on either side of the midline until the muscles are lost in the fascia of the tumour. At this point, they can be divided along a line parallel to the skin incision, allowing the tumour to be further displaced in a caudal direction.

Attention is then directed to the pelvic extension of the tumour. Using blunt dissection with peanut swabs in the plane anterior to the median sacral vessels, it is usually possible to displace the pelvic component of the tumour anteriorly until its upper extent is reached (Fig. 51.3). This is normally an essentially avascular plane anterior to the sacrum, although some vessels feeding into the tumour from the internal iliac vessels may be encountered laterally. These can usually be controlled with cautery. In most cases, the tumour can be dissected out from the pelvis and rolled inferiorly over the patient's legs.

This manoeuvre exposes the upper end of the rectum, which can be identified by the Vaseline gauze pack or by passing a finger in through the anus. The tumour can be dissected off the rectum with a combination of sharp and blunt dissection (Fig. 51.4), and rolled inferiorly until the plane of dissection moves away from the rectum and the anal canal. At all times during this dissection, it is best to try to maintain the plane of dissection on the capsule of the tumour and to preserve all normal structures no matter how distorted and thinned-out they are. As the tumour is rolled inferiorly, it eventually becomes apparent that the plane of dissection has reached the subcutaneous tissue along the inferior surface of the tumour, posterior to the anus. Once the dissection has reached this point, the dissection can be terminated as long as the inferior skin flap that has been developed is of sufficient length to allow easy closure of the wound. The inferior skin flap can then be divided from the tumour and the tumour delivered from the field. A careful check of the tumour bed is carried out to ensure that meticulous haemostasis has been achieved.

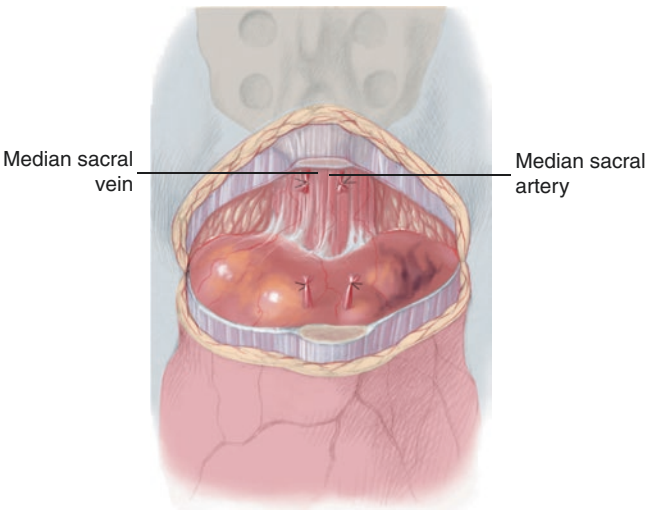

Fig. 51.3 Displacement of the pelvic component of the tumour until its upper extent is reached

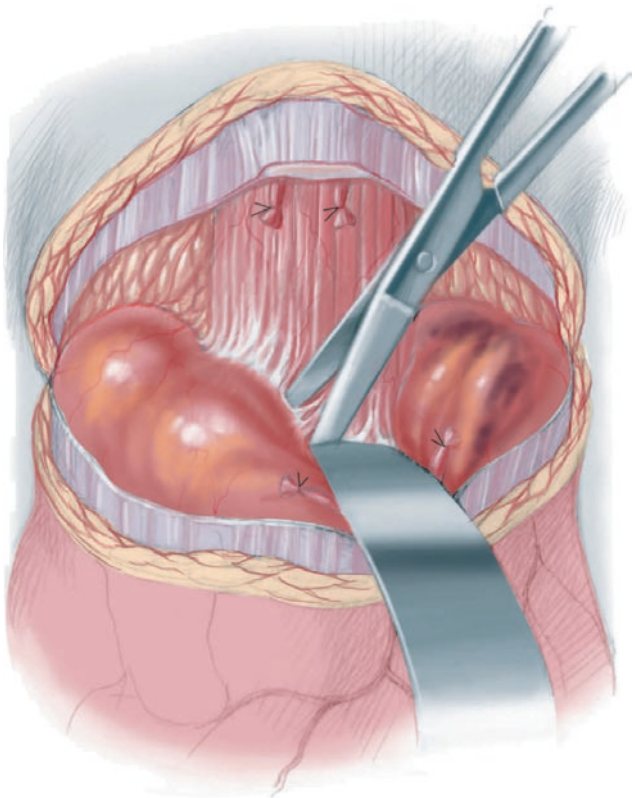

Fig. 51.4 Dissecting the tumour off the rectum

51.2.3 Reconstruction and Closure

Attention is then directed to reconstruction of the pelvic floor and closure of the wound. The remnants of the levator sling are identified and the central portion is sutured to the perichondrium of the anterior surface of the sacrum using a 5/0 monofilament absorbable suture (Fig. 51.5). This same suture is used for all subsequent muscle and fascial reconstruction. These initial fascial sutures, rather than the skin closure, should determine the siting of the anus. This aspect of the reconstruction, therefore, should be carried out with care to ensure both a functional and a cosmetically pleasing result.

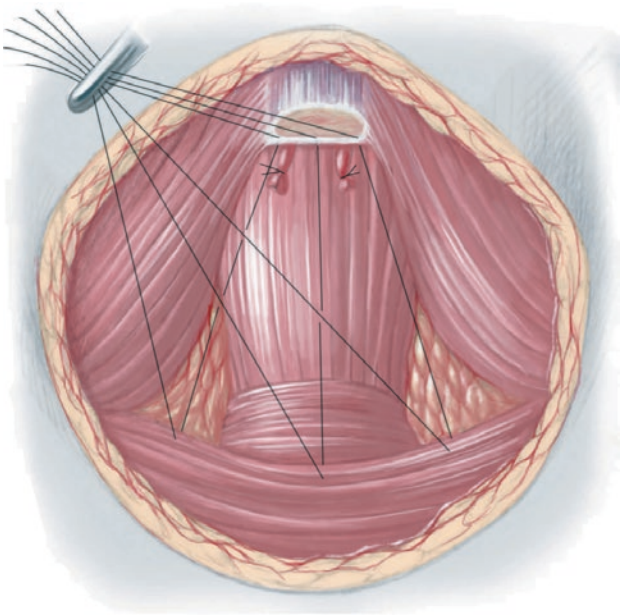

Fig. 51.5 Reconstruction of the pelvic floor

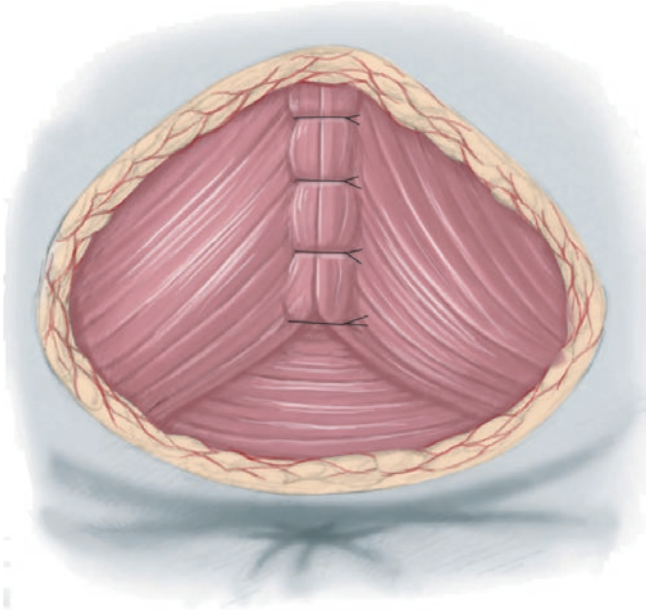

Fig. 51.6 Closure of the medial edges of the gluteus maximus in the midline

If a drain is to be placed, then it is placed at this stage, in the presacral space, led out through the gap in the levators and tunnelled out through the subcutaneous tissue of the buttock. A closed suction drain is preferred.

If remnants of the levators are recognizable lateral to the midline, these are repaired with interrupted 5/0 monofilament absorbable suture. The medial edges of the gluteus maximus are then closed in the midline over the sacrum and the lower part of the levator sling (Fig. 51.6). The skin flaps are then trimmed to length. If possible, the subcutaneous tissues are closed with a running 5/0 monofilament absorbable suture and the skin is closed with a running 5/0 monofilament absorbable subcuticular suture. A Steristrip and collodion dressing is then applied. If it is not possible to close the subcutaneous tissue, then a subcuticular suture may not be adequate for skin closure. In this case, interrupted 5/0 nylon skin sutures are placed (Fig. 51.7). The rectum is packed with Vaseline ribbon gauze at the completion of the procedure in an attempt to obliterate dead space. It is useful to suture a 2/0 silk suture to the end of this pack to aid its retrieval, should the pack become displaced higher up the rectum in the immediate postoperative period.

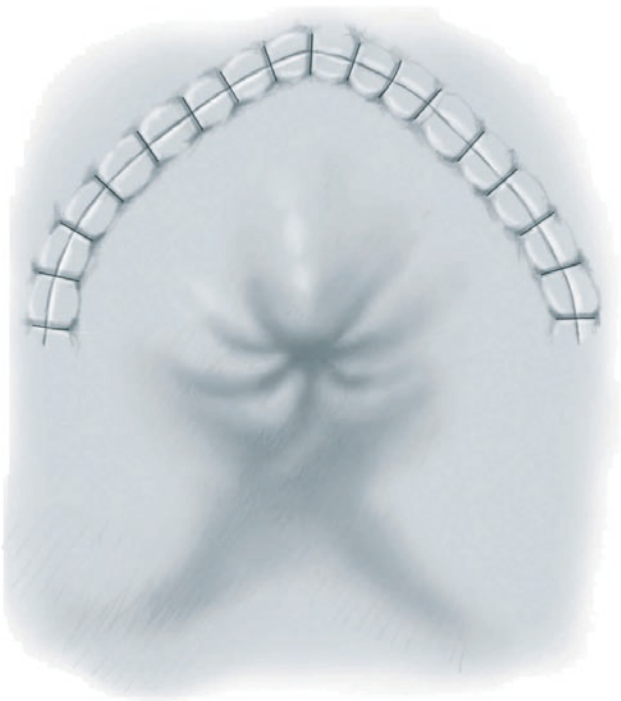

Fig. 51.7 Closure of the skin

51.3 Postoperative Care and Follow-Up

The infant is nursed in a prone position for several days after surgery. The urinary catheter can be removed as soon as the baby's condition is stable, and the infant can be extubated as soon as its respiratory condition allows. The infant can usually be fed as soon as it is extubated. The Vaseline pack is usually removed in the first postoperative day by pulling on the 2/0 silk suture left attached to the distal end. Any drain can usually be removed within the first few days. Levels of α-fetoprotein should be determined immediately after surgery and on discharge. The infant should then be followed up at monthly intervals for 3 months and then every 3 months as discussed below.

Follow-up should continue for at least 5 years. At each visit, a rectal examination will detect any local recurrence and an α-fetoprotein level may detect any distant spread. The α-fetoprotein level is often very high (about 100,000 U or more) and even in normal babies may be over 10,000 U. These high levels usually take more than a year to fall to normal adult levels. It is generally thought that as long as the α-fetoprotein level continues to fall steadily, recurrence is unlikely. In our experience, however, a steadily falling α-fetoprotein level does not rule out the possibility of either a local recurrence or even a malignant recurrence. In our series, all of the local recurrences and a malignant recurrence developed in the context of a steadily falling α-fetoprotein level. For this reason, we have developed a protocol of routine ultrasound examinations and rectal examinations at regular intervals for the first 3 years of life. These are monthly for the first 3 months, then every 3 months until 3 years of age. The ultrasounds are not only an attempt to detect recurrence but also a screening examination to provide early detection of a neurogenic bladder. After the first 3 years, ideally, the patient should be observed every 6 months until school age, and then every 1–2 years until puberty. At any stage, presentation with a urinary tract infection should prompt an early renal ultrasound. The relatively late development of a hostile neurogenic bladder at the age of 3 years in one patient does suggest that normal bladder function cannot be assumed to be present until well into the school years.

51.4 Outcomes

If the excision is complete and no distant metastases are identified at presentation, then life expectancy should be normal, although the appearance of the buttocks usually leaves something to be desired. The prognosis for patients presenting with a malignant sacrococcygeal teratoma must be guarded. Modern chemotherapy has produced some improvement in survival, but the regimens are toxic and the tumor tends to be relatively resistant to therapy. This is a relatively uncommon circumstance, however.

Two recent reports (Derikx et al. 2007; Shalaby et al. 2014) of long-term follow-up of a significant number of patients has revealed that up to 46% report problems with urinary or fecal incontinence. In most cases, the problem is manageable, but some patients require bladder augmentation in order to manage the development of a hostile neurogenic bladder. There have been few reports of problems with pregnancy or delivery in patients who have had a sacrococcygeal teratoma removed in infancy.

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Neuroblastoma

52

Edward Kiely and Michael E. Höllwarth

Neuroblastomas arise in neuroblasts of the sympathetic nervous system. The largest concentration of these cells is in the adrenal medulla and in the sympathetic ganglia chain from the neck to the pelvis. Other collections of sympathetic neuroblasts lie within the preaortic ganglia of Zuckerkandl.

The incidence of neuroblastoma in children is about 1 in 100,000. It accounts for 6–10% of all childhood cancers and 15% of cancer deaths. It is the commonest abdominal malignancy of childhood. About one third of neuroblastomas occur in infancy, and more than half of all patients are younger than 2 years. Fifty percent are detected under the age of 4 years and all but 5% are found before age 10. The male to female ratio (1.2:1) shows a slight preponderance of males. Hereditary factors may play a role, especially in early infancy; neuroblastomas have been described in twins and in a parent and young child.

The tumor occurs in the retroperitoneum in about 75% of patients, in either the adrenal (about 50%) or the paraspinal ganglia (25%). Tumours arising in the posterior mediastinum (20%) or the pelvis/neck (5%) are much less common.

52.1 Clinical Presentation and Staging

Neuroblastomas are characterized by many different clinical presentations. In the abdomen, a hard tumor may be palpable and symptoms include weight loss, failure to thrive, fever, anemia, and hypertension due to renal artery compression the adrenal production of the metabolic active cells. Tumors in the mediastinum may be associated with respiratory distress. In the upper thorax or neck, tumors arising from the stellate ganglion cause a Horner syndrome (ptosis, miosis, and enophthalmos). Paraspinal tumors can extend through the intervertebral foramen to the extramedullary space (so-called dumbbell-shaped lesions), leading to paraplegia. Excessive catecholamine production results in hypertension, sweating, irritability, and occasionally intractable diarrhea. The "dancing eye syndrome" characterized by opsomyoclonus and nystagmus is typical for cerebellar ataxia.

At the time of clinical presentation, about two thirds of patients have locally advanced or metastatic disease. Anemia is often related to bone marrow invasion. The presence of bone and joint pain, a consequence of metastasis, may bring the condition to attention. Metastasis to the retrobulbar bones produces typically unilateral proptosis and ecchymosis.

Various classification and staging systems have evolved over the years in an attempt to predict outcome, compare results, and direct therapy. The International Neuroblastoma Staging System (INSS) is currently the system that is most widely applied. In this system, tumours are staged from 1 to 4S. Stages 1 and 2 are localized and amenable to excision; stage 3 implies extension across the midline with a nonresectable tumour, and stage 4 implies a tumour with distal metastases. Stage 4S is limited to infants with metastases to liver, skin, or mild bone marrow involvement. A more recent staging system is based on the pretreatment imaging and bone marrow morphology, instead of the surgical resection,

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which is dependent upon the approach of the surgeon. Staging is undertaken using cross-sectional imaging (ultrasound examination, CT with three-dimensional reconstruction, and MRI), and isotope scanning together with bone marrow aspirates. Metaiodobenzylguanidine (MIBG) imaging of soft tissue and bone extension of the disease has been shown to be the most sensitive investigation for staging and evaluation of treatment response.

In addition to clinical staging systems, serum markers of disease behaviour are also widely used. These include 24-h catecholamine byproduct secretion (adrenaline, noradrenaline, dopamine, homovanillic acid [HVA], vanillylmandelic acid [VMA]), neurone-specific enolase (NSE), lactate dehydrogenase (LDH), and ferritin. Elevated levels of these markers are associated with a worse prognosis.

Histologically, the appearance of these tumours varies from the more benign ganglion cell and nerve fibre mixture to an undifferentiated, small blue round cell appearance. The classic rosette appearance is sometimes seen. Although histologic grading systems modified by Shimada are in use, they are not universally applied. Stroma-poor tumors have often MYCN amplification indicating aggressive tumor activity. MYCN and Ip deletion are associated with a worse outcome regardless of age and stage; polyploidy is associated with a better outcome. Other markers are being studied.

52.2 Treatment Protocols

Most children with neuroblastoma are treated by pediatric oncologists according to agreed national or international protocols. In general, low-stage tumours with favourable biological profiles are managed by excision alone. Aggressive chemotherapy is used for low-stage tumours with a poor biological profile or for those (the majority) who have locally advanced or metastatic disease. Neuroblastomas are radiosensitive tumors, so radiotherapy is an important part of the treatment regimen. It is used before and after surgery. Surgical resection of the tumor is still a central part of the therapeutic regimen, even if resection is not complete. When planning surgery for any of these children, cross-sectional imaging provides useful information on the site of the tumour and the degree of anatomical distortion. As the tumours are intimately related to major blood vessels, imaging provides some useful information about which vessels will need to be dissected.

52.3 Surgical Procedure

The surgery for neuroblastoma is undertaken under full, intubated general anaesthesia with indwelling vascular monitoring. The procedure described is for resection of a left adrenal primary tumour with nodal disease around the aorta and its branches, but the principle of the operation is the same regardless of the site of the primary tumour. Neuroblastomas do not usually invade the tunica media of major blood vessels. Consequently, a plane of dissection may be developed with a knife between the tunica adventitia and the tunica media. The initial phase of the operation is to display part of the circumference of all the relevant vessels as they traverse the tumour. Each vessel is then cleared in turn, and the tumour may be excised once the vascular anatomy is completely displayed.

The abdomen is opened through an upper transverse abdominal incision. After a full laparotomy, the sigmoid and descending colons are reflected medially, leaving Gerota's fascia intact (Fig. 52.1). The spleen is then mobilized and reflected medially with the pancreas and stomach. All mobile

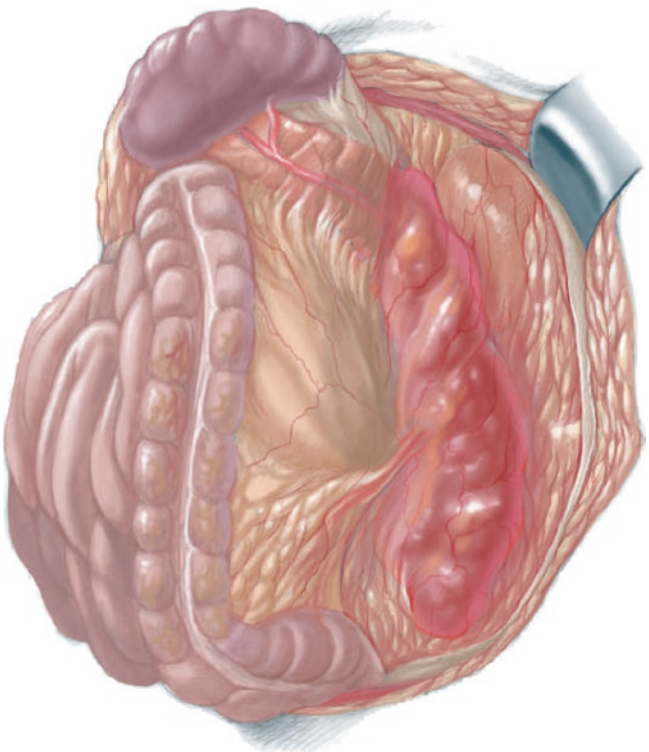

Fig. 52.1 Exposure Retraction of the sigmoid and descending colons

intestine and spleen are placed in an intestinal bag and a table-mounted retractor is used to maintain a clear operative field.

The lower limit of the tumour is then established and the artery is dissected as it emerges from the tumour. This is done by incision of the tunica adventitia longitudinally along the middle of the vessel (Fig. 52.2). If the adventitia is put under tension by the surgeon and the assistant, it opens cleanly once it is incised.

The plane of dissection is now established for the remainder of the operation. The dissection advances proximally in the same fashion (Fig. 52.3), incising tumour and adventitia down to media in 1- to 2-cm steps. It is important for the surgeon and assistant to apply tension to the tissue to be divided, as this eases the dissection. The aortic bifurcation is then reached and the direction of the dissection moves along the middle of the aorta. The origin of the inferior mesenteric artery is encountered shortly thereafter and usually does not present many problems.

When the left renal vein is reached, it is mobilized in a similar fashion and the tumour beneath it is divided and cleared from the anterior wall of the aorta. The origin of the left renal artery is encountered shortly thereafter, and the axis of dissection moves towards the 2 o'clock, rather than the 12 o'clock, position to avoid the origin of the main visceral arteries. The origin of the left renal artery is cleared as before by longitudinal incision of the tumour down to media. The dissection then continues along the aorta until the diaphragm and upper limits of the tumour are resected (Fig. 52.4).

At this time the origin of the coeliac axis is visible, and this artery is exposed as before. Once the coeliac has been cleared, tumour overlying the superior mesenteric artery is managed in similar fashion. If the tumour extends deeply towards the right crus of the diaphragm, then the splenic vein and its junction with superior mesenteric and portal veins are cleared in the same way (Fig. 52.5).

Once all the vessels are in view, they are mobilized circumferentially and the tumour is removed (Fig. 52.6).

After complete excision of the tumour, all vessels can be clearly seen (Figs. 52.7 and 52.8). Any suspicious lymph glands should be removed for histological examination.

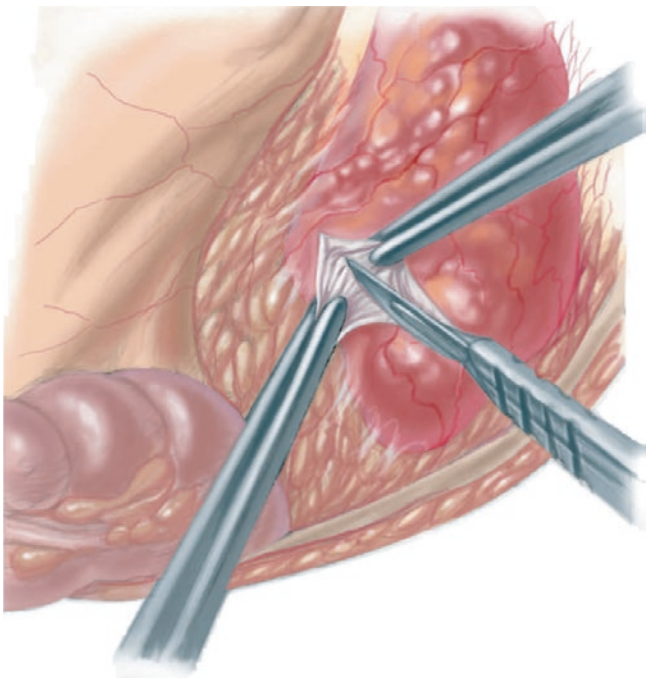

Fig. 52.2 Incision of the tunica adventitia

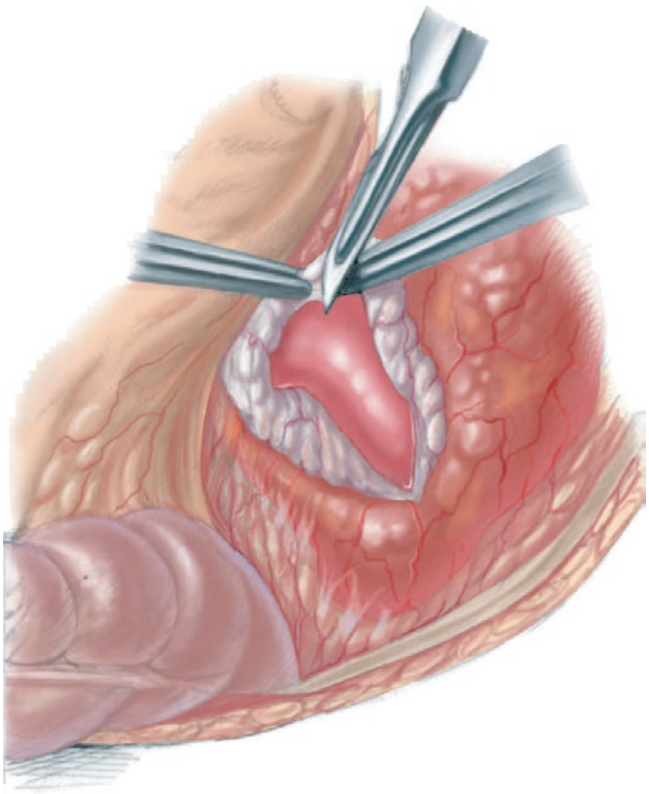

Fig. 52.3 Advancing the dissection

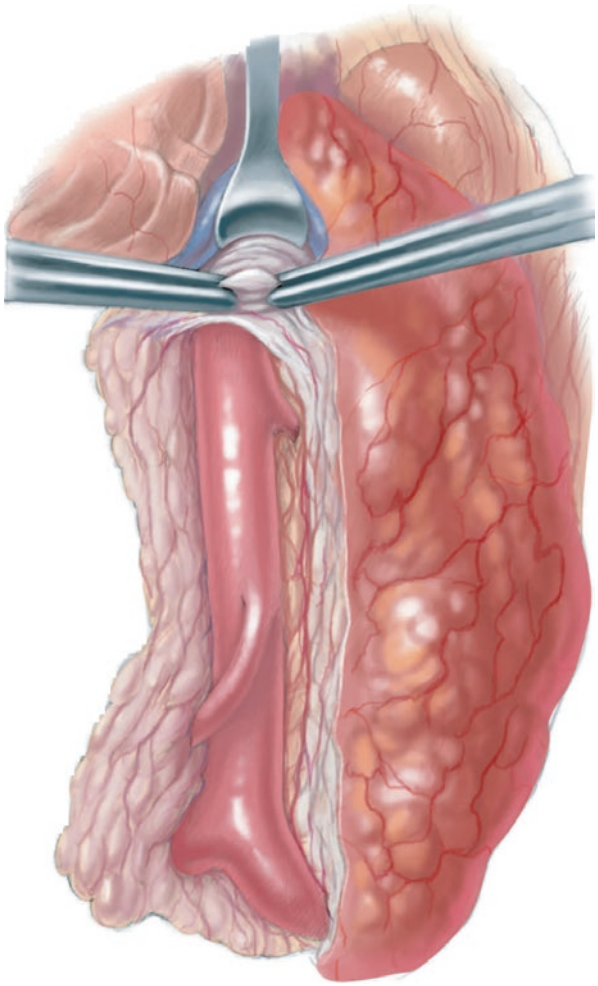

Fig. 52.4 Dissection along the aorta

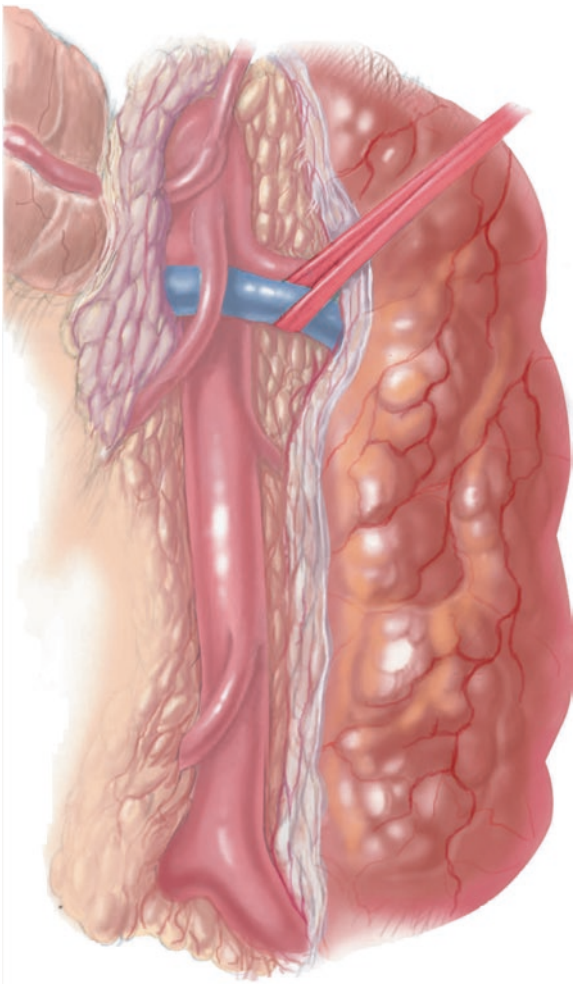

Fig. 52.5 Clearing of tumour from the veins

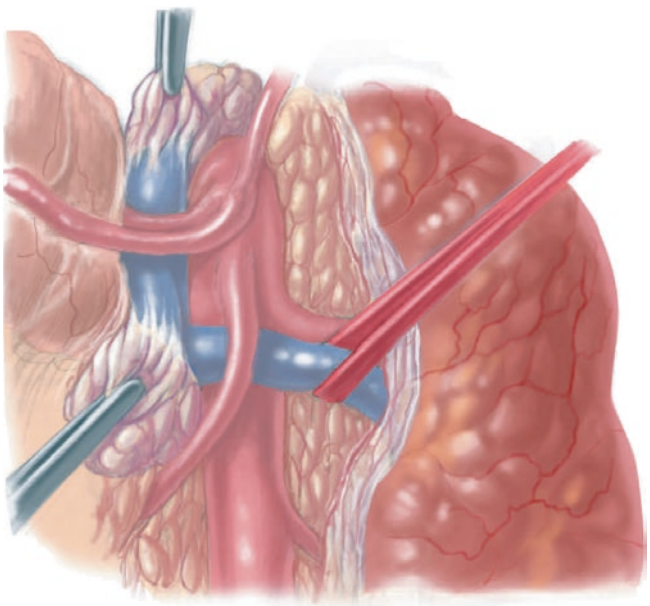

Fig. 52.6 Mobilizing all the vessels

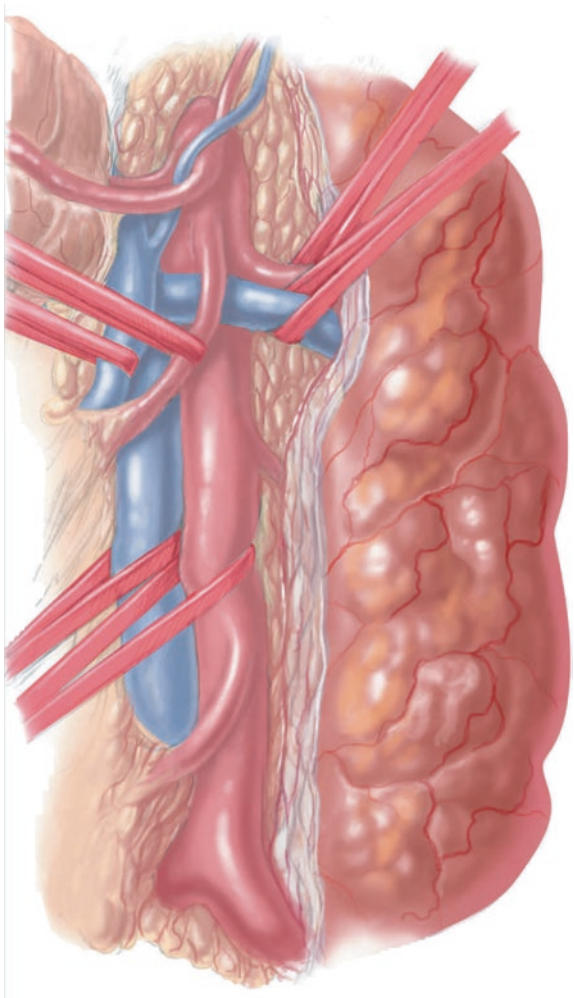

Fig. 52.7 Completing the excision of the tumour

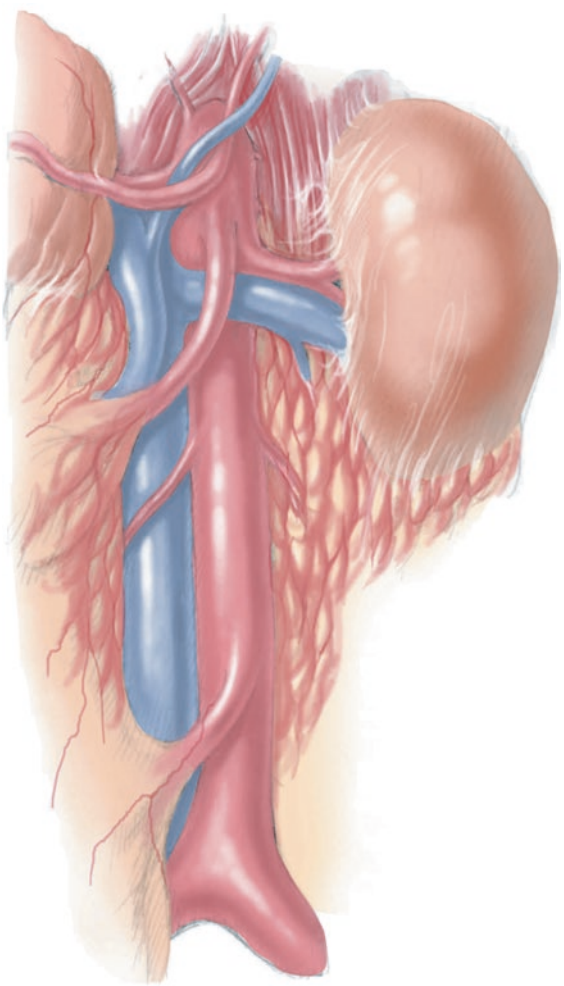

Fig. 52.8 Viewing all vessels after removal of the tumour

52.4 Outcomes

Complete resection is possible in about 90% of patients. Surgical resection can be complete in 85% of patients. The operative mortality is about 1–2%. Laparoscopic resection of adrenal neuroblastoma is feasible in localised tumours and can be performed with results equivalent to those of open surgery. Intraspinal extension can be managed effectively with upfront chemotherapy, and neurologic manifestations of less than 4 weeks' duration are usually reversible. For those undergoing clearance of coeliac and superior mesenteric arteries, diarrhoea has been a particular problem.

Overall 5-year survival in recent statistics is 68% (stage 1, 100%; stage 2, 90%; stage 3, 77%; stage 4, 52%; stage 4S, 80%; high-risk cases, 52%). Gross tumor resection has a clear survival benefit over subtotal resection in stage 3 cases, but the difference is not significant for stage 4 disease. Myeloablative therapy with total-body irradiation followed by autologous bone marrow transplantation significantly increased survival rates in patients with stage 4 disease.

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Wilms Tumour

53

Philip Hammond and Robert Carachi

Renal neoplasms in childhood are usually malignant, the most common being nephroblastoma Wilms tumour (WT). The incidence varies from 10.9 per million in the United States to 2.5 per million in China. A genetic predisposition exists, and nephrogenic cell clusters, (which are premalignant lesions), are found in one third of patients with WT. Some syndromes are associated with WT:

- WAGR (WT, Aniridia, Genitourinary anomalies, mental Retardation)
- Denys-Drash (WT, disorders of sexual development, nephropathy)
- Beckwith-Wiedemann (exomphalos, macroglossia, visceromegaly)
- Hemihypertrophy is associated with an increased incidence of WT

WT1 and *WT2* genes are associated with WT, especially *WT2* with Beckwith-Wiedemann syndrome.

53.1 Presentation

WT presents as a palpable, asymptomatic abdominal mass in a toddler. It is usually discovered when the parents are bathing or dressing the child. Weight loss, malaise, abdominal pain, hypertension, and haematuria may be present. Rarely, the patient may present with a varicocoele, where the left renal vein is occluded by tumour thrombus. On examination, the smooth, rounded tumour occupies most of the abdomen, and in about 10% of patients, the tumour thrombus from the nephroblastoma may invade the inferior vena cava. At times, it may extend to the right atrium, causing cardiac dysfunction or even a pulmonary embolus. Obstruction of the hepatic veins may rarely cause an acute hepatic encephalopathy and produce a Budd-Chiari syndrome. Patients with syndromes such as WAGR, Denys-Drash, Beckwith-Weidemann, or hemihypertrophy should receive regular ultrasound screening because they have an increased risk of developing WT.

53.2 Diagnosis

The mainstay of investigation is imaging. A plain x-ray of the abdomen usually shows a soft tissue mass, and calcification may be seen in about 10% of patients. A plain chest radiograph may show pulmonary metastases. Abdominal ultrasound confirms that the tumour is renal in origin and can demonstrate a normal contralateral kidney. It also evaluates the inferior vena cava for blood flow and for the presence of tumour thrombus. A CT scan will outline the tumour and may show a lesion in the contralateral kidney; it is more sensitive than the plain chest radiograph for the identification of pulmonary metastases. MRI can add a further dimension to renal evaluation, with visualisation of blood vessels. Echocardiography may be necessary to exclude the presence of an intra-atrial extension of the tumour thrombus.

The histology of WT mimics the development of the normal kidney with the proportion of the three components (blastema, tubules, and stroma) varying greatly in different tumours. Histologic differentiation allows good clinical correlation between a "low risk" group of patients who can be cured and a "high risk" group who need more intensive treatment and do less well.

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53.3 Treatment and Staging

The surgical treatment of Wilms tumour involves three stages: (1) making a diagnosis by biopsy; (2) operative excision of the tumour; and (3) staging of the patient.

53.3.1 Staging Systems

The staging system of the National Wilms' Tumor Study (NWTS) uses five stages:

- *Stage I*: Tumour limited to the kidney and completely excised. Surface of the renal capsule intact; no tumour rupture; no residual tumour apparent beyond margin of excision
- *Stage II:* Tumour extends beyond kidney but is completely excised; regional extension of tumour; vessel infiltration; tumour biopsy or local spillage or tumour confined to flank; no residual tumour apparent at or beyond margins of excision.
- *Stage III*: Residual non-hematogenous tumour confined to the abdomen, lymph node involvement of the hilum, periaortic chains or beyond; diffuse peritoneal contamination by tumour spillage; peritoneal implants; tumour extends beyond resection margins, either microscopically or macroscopically; tumour not completely resectable because of local infiltration into vital structures.
- *Stage IV*: Deposits beyond stage III in lung, liver, bone, or brain.
- *Stage V:* Bilateral renal involvement at diagnosis.

The SIOP (International Society of Pediatric Oncology) group subdivides Stage II into IIa, in which the lymph nodes are tumour-free, and IIb, with tumour-positive regional (hilar) lymph nodes.

53.3.2 Preoperative Chemotherapy

A variety of protocols have been devised that show benefit from preoperative chemotherapy once the diagnosis has been established. Thus, on suspicion of a Wilms tumour, preoperative chemotherapy may shrink the tumour, downgrade the staging, and reduce the incidence of intraoperative tumour rupture. Preoperative chemotherapy has been very effective in Europe, where the SIOP trials have demonstrated a reduced incidence of operative tumour rupture in patients who had received chemotherapy. The chemotherapy shrinks the tumour considerably in 80% of cases, and therefore makes the surgery safer (particularly for intravascular tumour thrombus). In the UK, patients currently may have percutaneous biopsies to obtain histologic confirmation of the tumour prior to starting preoperative chemotherapy. (This is an attempt to identify those histologic types that are likely to progress on chemotherapy and may benefit from expedited surgery.) Surgery is normally delayed for several weeks whilst neoadjuvant chemotherapy is administered according to the protocol.

53.3.3 Surgical Procedure

The operation performed for Wilms tumour is a transabdominal nephrectomy. It is essential to have all the necessary prerequisites done before surgery can start, such as a central venous catheter with central venous pressure monitoring, arterial line with continuous arterial pressure monitoring, urinary catheter, etc. Epidural block is part of the routine procedure, and cross-matched blood should be available if needed.

The abdomen and chest are prepared and draped. It is best to have the flank raised with a roll underneath the patient. The operative procedure begins with a large transverse incision (Fig. 53.1). The incision is carried out well into the flank on the involved side and across to the flank on the opposite

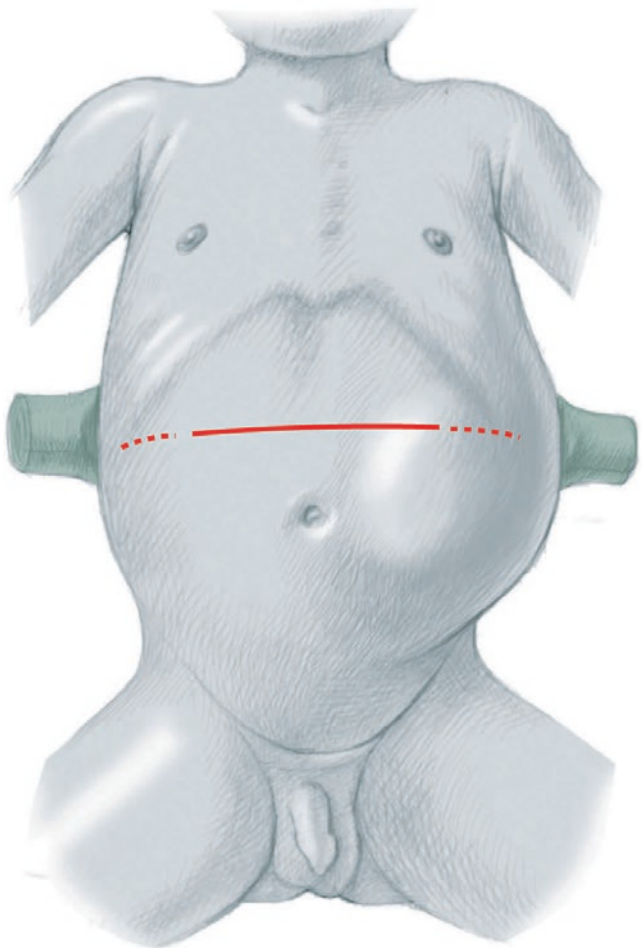


Fig. 53.1 Transverse incision for transabdominal nephrectomy

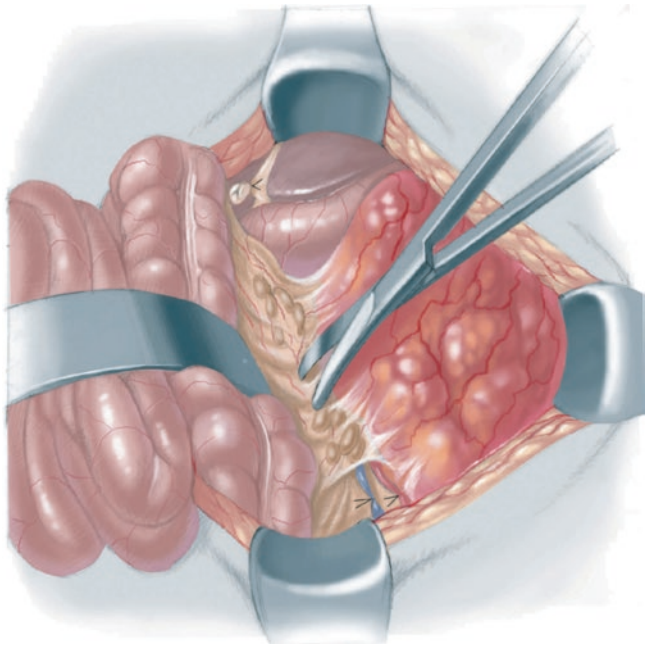


Fig. 53.2 Mobilizing the colon off the tumour

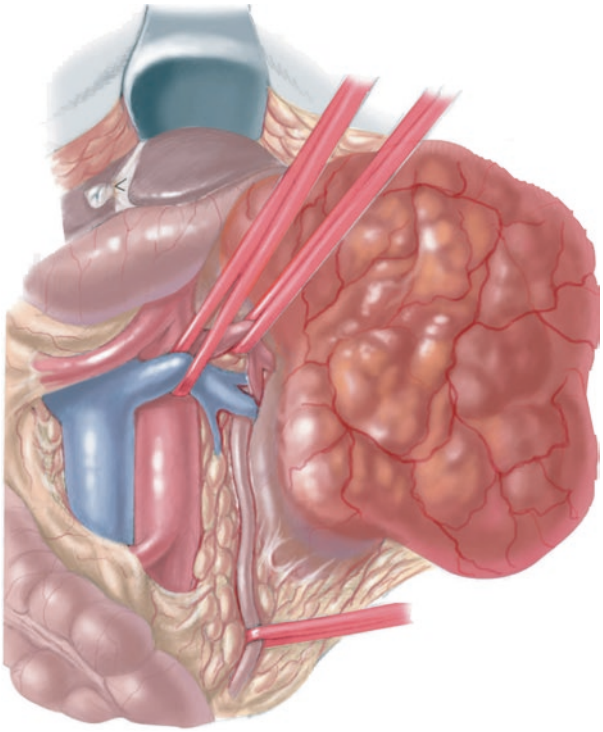


Fig. 53.3 Slings preventing embolization

side. The incision must be large enough to allow large tumours to be mobilized without risking intraoperative rupture. The incision is deepened through the subcutaneous fat and the rectus and oblique muscles.

The peritoneum is then opened and the ligamentum teres is divided between ligatures. Any free fluid (particularly if it is blood-stained) should be sent for cytology, and any peritoneal deposits should be excised and sent for histopathology. The small intestine is then delivered out of the abdomen and protected by warm, moist packs in order to assess the tumour. A full assessment should be made by palpating the tumour, the liver, lymph nodes, and the contralateral kidney (although preoperative imaging may be sufficient).

The retroperitoneal space is opened by an incision made lateral to the reflection of the peritoneum of the ascending colon for a right-sided tumour, or lateral to the descending colon for a left-sided tumour. The colon is mobilized off the tumour and reflected medially (Fig. 53.2). Occasionally the tumour invades the mesentery and the vessels; these need to be ligated and divided, preserving the marginal artery to avoid unnecessary resection of bowel.

A sling is passed around the renal vein or veins, and the contralateral renal vein is visualized. (Tumour may distort the normal anatomy.) A sling around these structures will prevent any risk of embolization when mobilizing the tumour. Sling the renal artery or arteries and the ureter (Fig. 53.3). Dissection of the hilum of the kidney before mobilization is not always possible, especially when the tumour crosses the midline or if large lymph nodes are in the way.

The renal vein and inferior vena cava should be palpated early for an assessment of whether an intravascular thrombus with tumour extension is present. Careful palpation of the contralateral vein is also indicated at this step of the surgical procedure. If a thrombus is detected in the renal vein, or the thrombus is extending into the vena cava, the appropriate vessels can be opened by a transverse incision between slings or after placement of vascular clamps (Fig. 53.4), and the thrombus can be removed with an open-ended suction cannula or a Fogarty balloon catheter. The vein is then closed with a 5/0 monofilament non-absorbable continuous running suture.

In cases without intravascular tumour involvement, the renal artery or arteries are transfixed and the ends are ligated. Double-ligate the renal veins with nonabsorbable suture material (Fig. 53.5). The renal artery (or arteries) should be ligated and divided before the renal vein (veins) to avoid excess congestion of the tumour. Lymph nodes from para-caval, para-aortic, supra-hilar, infra-hilar, mesenteric and bilateral iliac regions should be sampled (and carefully labeled) for staging. A sling is passed around the ureter at the pelvic brim and the gonadal vessels are usually divided (although may be preserved if tumour anatomy allows). The ureter should be isolated off the pelvic brim and transfixed as low down as possible using absorbable suture material.

The kidney is mobilized from the retroperitoneal space (Fig. 53.6). Large veins may need to be divided. Meticulous

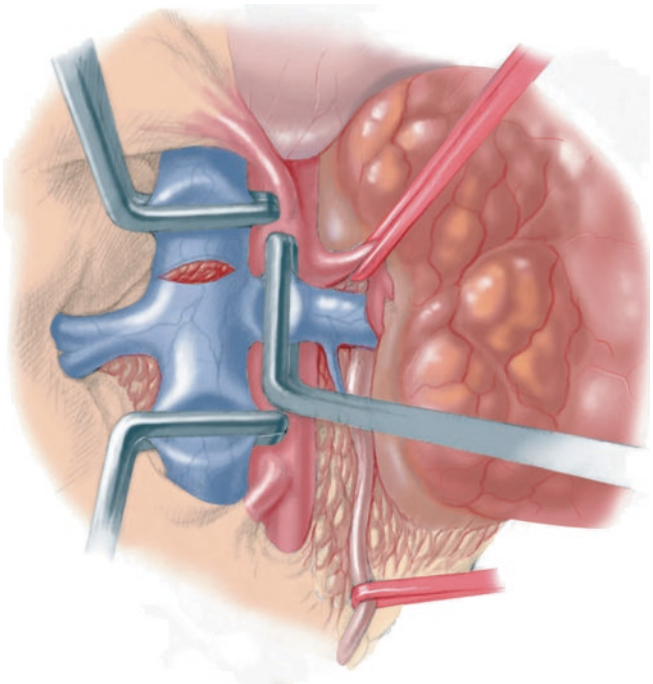


Fig. 53.4 Incision for removal of an intravascular thrombus

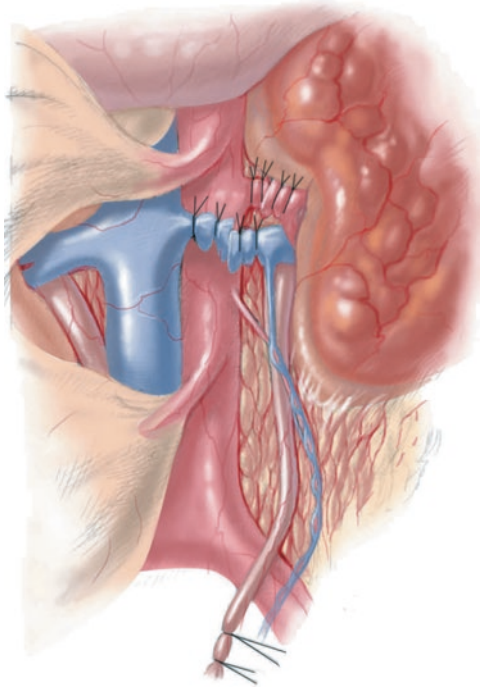
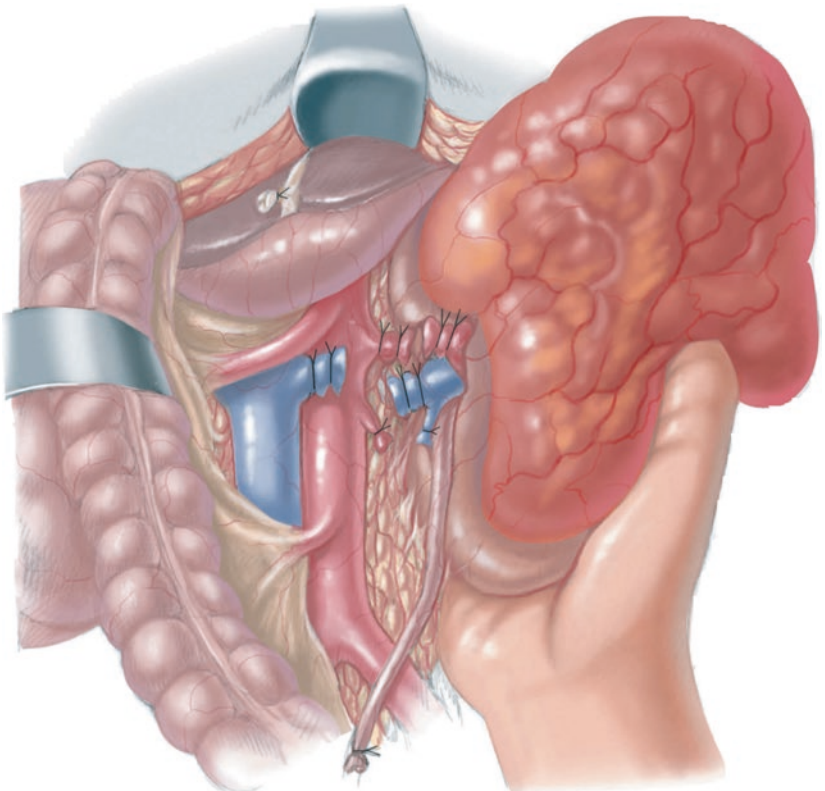


Fig. 53.5 Ligation of renal veins and arteries

Fig. 53.6 Removal of the tumour



dissection is needed. If the tumour is adherent to muscle or diaphragm, this needs to be removed as well. Finger dissection is useful for tissue planes.

The adrenal is removed in most cases (although preservation may be possible in the case of small, lower pole tumours). Any lymph nodes should be included in the mobilization and removed en bloc with the perinephric fat. These should be secured with double sutures and transfixed where appropriate, as in the case of the renal artery. Sometimes large veins in the perinephric fat must be ligated and divided. Direct infiltration of the posterior abdominal wall by the tumour can occur. Similarly, invasion of the diaphragm may require dissection of the muscle together with the tumour. The adjacent liver is usually adherent rather than actually invaded, but invasion of the diaphragm is common and sometimes a portion of the diaphragm must be removed.

It is important to recognize that large tumours may displace the aorta or the vena cava, and the tumour may actually grow behind these large vessels.

Accidental ligation of the aorta or the contralateral renal vein and vena cava as well as injury to the superior mesenteric vessels have been known in the past. To avoid any doubt about which type of blood vessel is being sacrificed, a loop around the vein or artery before ligating is essential. If the adrenal gland is adherent, it may be removed with the tumour. The tumour must be removed en bloc with the hilar lymph nodes and sent fresh to pathology.

The tumour bed should be inspected for haemostasis or residual tumour. Any suspicious area should be removed and sent for biopsy. The contralateral kidney must be carefully inspected for evidence of disease.

Haemostasis needs to be meticulous and the tumour bed should be dry before closure of the wound. Rarely, where there is uncontrolled bleeding, a pack may need to be left in and removed 48 h later. The abdomen is closed in layers, and no drain is left in situ.

53.4 Outcome and Follow-Up

The history of the treatment of patients with WT is one of the most impressive success stories in pediatric surgery. In 1941, W.E. Ladd reported a survival rate of 20%, but today the relapse-free survival is close to 90% in all patients, and even 66% in the histologically high-risk group. Study results of the NWTS (primary surgery) and the SIOP (primary chemotherapy, with or without tumour biopsy) are similar: the rate of complications and tumour rupture is significantly higher with the primary surgery strategy.

Long-term follow-up is important, to allow early detection of a possible metachronous tumour in the contralateral kidney. Synchronous bilateral tumours have an incidence of about 10%; after initial biopsy, chemotherapy should be instituted in these patients, and then partial nephrectomy carried out, as well as lumpectomies. This procedure has been found to be effective in the past. Biopsy only and chemotherapy is indicated in patients with diffuse nephroblastomatosis, but long-term follow-up is also crucial, because late WT occurrence is known.

53.5 Alternative Approaches

Experience with nephron-sparing surgery (NSS) in unilateral WT is increasing. This approach should be considered particularly for children with good response to preoperative chemotherapy who have a single kidney, syndromes predisposing to WT, or other nephrologic conditions. Ideally, for NSS to be feasible, the tumour should be polar, away from renal sinus/vessels and calyces. The aim is to leave two thirds of the kidney in situ. This approach may risk up-staging from stage I to stage III.

Some surgeons have undertaken minimally invasive surgery for WT (particularly for small tumours in the hilum). Careful case selection is vital, and this approach is not currently recommended by SIOP (especially if the alternative would be NSS, which would be preferred). Certainly, if this approach is considered, it is important for the patient to be enrolled in ongoing international studies so that the historical improvements in outcome for this tumour are not compromised by surgical approach.

Suggested Reading

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Liver Tumours

54

Irene Isabel P. Lim and Michael P. La Quaglia

Although primary liver tumours comprise only 5% of pediatric extracranial solid tumours, more than two-thirds are malignant. In Europe and North America, primary malignant liver tumours are the tenth most common pediatric cancer. Malignant tumours can arise from primary hepatocytes, such as in hepatoblastoma and hepatocellular carcinoma, or from the biliary tract (rhabdomyosarcoma). Leiomyosarcoma (arising from hepatic smooth muscle tissue) and angiosarcoma (arising from blood vessels) are rare primary hepatic tumours. Malignant liver tumours require complete resection for cure. Benign hepatic tumours, which account for about one third of all liver tumours in children, also occasionally will require liver resection. Hemangiomas are the most common benign liver tumour, followed by focal nodular hyperplasia. Less common benign tumours include hepatic adenoma and mesenchymal hamartoma.

The liver is also a frequent site of metastatic disease in children. Non-Hodgkin's lymphoma, neuroblastoma, Wilms tumour, desmoplastic small round cell tumour, adrenal cortical carcinoma, osteogenic sarcomas, and a host of other malignancies may metastasize to the liver. Criteria for surgical removal of hepatic metastases include control of the primary site, a solitary or limited number of metastases, good performance status, and a reasonable expectation of prolonged survival or cure.

54.1 Presentation

Together, hepatoblastomas and hepatocellular carcinoma account for over 90% of primary hepatic malignant tumours. Hepatoblastomas are the most common childhood hepatic malignancy, representing nearly 80% of all primary hepatic cancers and 43% to 64% of all hepatic neoplasms. The mean age at diagnosis is 18 months, and only 5% of cases are diagnosed in children over 4 years of age. Children with hepatoblastoma are usually in good health and most commonly present with an asymptomatic abdominal mass. Jaundice is rare, but thrombocytosis is common, and serum α-fetoprotein is well established as both an initial tumour marker and a means of monitoring the therapeutic response. The level is normally less than 20 ng/mL, but it can be elevated more than 20,000 times normal at presentation. Imaging studies often show a large tumour with evidence of central necrosis.

Hepatocellular carcinoma presents in a bimodal fashion, with the first peak occurring at 5 years of age and the second peak at 13–15 years of age. In contrast to hepatoblastoma patients, children and adolescents with hepatocellular carcinoma frequently present with symptomatic and palpable abdominal masses. Symptoms such as pain, anorexia, malaise, nausea, vomiting, and significant weight loss are common. Hepatitis B and C infections are correlated with hepatocellular carcinoma, though the fibrolamellar subtype is not. The α-fetoprotein is elevated in approximately 85% of patients; levels are higher than 1000 ng/mL in most patients with hepatocellular carcinoma but are usually lower than that level in patients with hepatoblastoma. Jaundice is also rare. Multifocal disease is more common in hepatocellular carcinoma, and up to 10% may present with tumour rupture and haemoperitoneum.

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54.2 Treatment Principles

Complete resection of the primary liver tumour is the cornerstone for survival in hepatoblastoma. Multiple studies support the effectiveness of systemic chemotherapy combined with complete surgical resection. Historically, the combination of doxorubicin and cisplatin has been used with great success, but the present-day protocol of cisplatin, 5-fluorouracil, and vincristine is equally effective and less toxic. Very young infants who undergo complete resection may receive a shortened course (three doses) of adjuvant single-agent doxorubicin, which is well tolerated.

The primary treatment of hepatocellular carcinoma is also complete surgical resection, and long-term survival is unlikely without it. Unfortunately, complete resection is often difficult because of the high incidence of multifocal disease within the liver, as well as extrahepatic extension to regional lymph nodes, vascular invasion, and distant metastases. Tumor infiltration resulting in thrombosis of portal and hepatic venous branches is common; even the vena cava may be involved. Historically, the same chemotherapy protocols used for hepatoblastoma were used for pediatric hepatocellular carcinomas. Cisplatin has had activity against hepatocellular carcinoma, but long-term survival still approaches zero, and new therapies are the subject of current research. In particular, sorafenib has been shown to increase median survival by nearly 3 months. Other signaling pathways and molecular targets are currently being investigated as potential targets for pharmacologic therapy.

54.2.1 Preoperative Evaluation

Imaging studies are crucial in both the preoperative evaluation of liver tumours and the planning of major hepatic resections. The PRE-Treatment EXTension (PRETEXT) system is used extensively for operative planning in both hepatoblastoma and hepatocellular carcinoma, and is dependent on the tumour's location with respect to the Couinaud segmental anatomy of the liver (Table 54.1). Doppler ultrasonography can identify whether a mass is cystic or solid, determine the patency of the portal and hepatic veins and vena cava, and identify satellite lesions. At present, MRI, particularly when enhanced with gadolinium-based contrast agents, provides the greatest amount of information regarding hepatic lesions and surrounding veins and bile ducts. Combined with Doppler sonography, MRI of the liver can provide sufficient information with respect to vascular and biliary anatomy, so arteriography is almost never performed. CT arterial portography has evolved to provide an equivalent alternative imaging modality, though the significant radiation exposure limits its use in the pediatric population. If a malignant tumour is suspected, CT scans of the chest and a bone scan are needed to rule out pulmonary and osseous metastases, respectively. Cerebral metastases are rare but have been reported, so patients with neurologic symptoms should undergo cranial MRI. A tissue diagnosis is mandatory, particularly in children, in whom most cases of primary liver malignancy occur in the absence of cirrhosis. Percutaneous needle core or aspiration biopsy is useful for hepatoblastomas but may not be definitive in the case of hepatocellular carcinoma, as the distinction between high-grade dysplasia and malignancy is difficult on biopsies. If hepatocellular carcinoma is strongly suspected and resection is planned, open or laparoscopic biopsy is preferred.

Table 54.1 PRETEXT system for planning of hepatic resections

PRETEXT number	Definition
I	One section involved, three adjoining sections free
II	One or two sections involved, two adjoining sections free
III	Two or three sections involved, one section free
IV	Four sections involved

PRETEXT—PRE-Treatment EXTension

54.2.2 Surgical Anatomy

A thorough understanding of the functional surgical anatomy of the liver is essential. The schema of hepatic anatomy most useful for the surgeon is based on the anatomic classification of Couinaud (Fig. 54.1). The liver is divided into the right and left lobe by the main portal fissure containing the middle hepatic vein. Each lobe is divided into a paramedian and a lateral region by the right and left hepatic veins. The left portal fissure is identified by the falciform ligament externally, but there is no external feature identifying the right portal fissure dividing the right lobe into anteromedial and posterolateral regions. The four regions are subdivided into anterior and posterior segments. Each of the eight Couinaud segments is supplied by a portal triad composed of a branch of the portal vein and hepatic artery, and drained by a tributary of the right or left main hepatic ducts. Segment I (the caudate lobe), situated posteriorly between the vena cava and the ligamentum venosus, receives inflow from both the right and left branches of the hepatic artery and portal vein, and drains directly into the inferior vena cava (IVC) via numerous small veins. The major venous structures comprise approximate boundaries of the four sections generally used in PRETEXT staging and surgical planning: the left lateral section (segments II and III), the left medial section (segment IV), the right anterior section (segments V and VIII), and the right posterior section (segments VI and VII).

Nonanatomic wedge resections with a satisfactory margin may be feasible in the uncommon instance of a small peripheral or pedunculated lesion. More frequently, a major anatomic resection is required. Anatomic resections are associated with less blood loss and greater frequency of clear margin. The extent of the hepatic resection is based on the consideration of several factors, including tumour location and proximity to major vascular or ductal structures. As much as 85% of the liver parenchyma may be removed, with subsequent full and rapid regeneration despite the administration of postoperative chemotherapy. Preoperative portal vein embolization of the segments to be resected has been utilized in adult patients to increase residual hepatic volume and has been met with some success.

The major hepatic resections include left lateral lobectomy (removal of segments II and III), left hepatic lobectomy (II, III, IV), extended left hepatectomy (II, III, IV, V, VIII), right hepatic lobectomy (V, VI, VII, VIII), extended right hepatectomy or right trisegmentectomy (IV, V, VI, VII, VIII), and central hepatic resection (IV, V, VIII). Segment I can also be resected during extended right or left hepatectomy to achieve tumour clearance. Lesions limited to one segment may be resected with the appropriate segmentectomy.

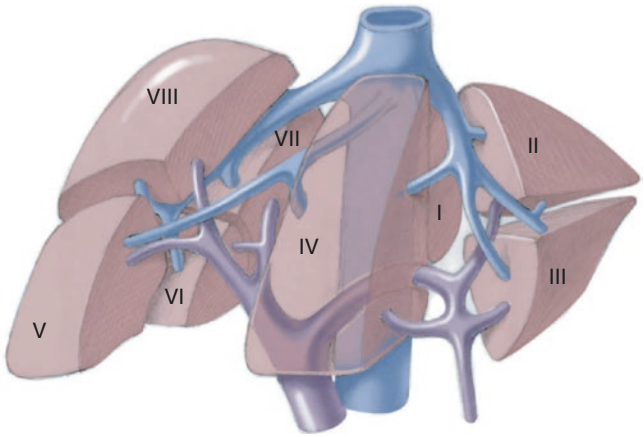

Fig. 54.1 Couinaud's anatomic classification of liver segments, which comprise the caudate lobe (segment I), the left lateral section (segments II and III), the left medial section (segment IV), the right anterior section (segments V and VIII), and the right posterior section (segments VI and VII)

54.2.3 Preoperative Preparation

Preoperative preparations include a comprehensive laboratory evaluation comprising a complete blood count, coagulation profile, and liver function tests. Adequate cross-matched blood should be available. If the patient received doxorubicin as part of the chemotherapy regimen, an echocardiogram evaluating cardiac function should be obtained. Bowel prep is carried out on the day prior to liver resection. A prophylactic antibiotic (usually a first-generation cephalosporin) is administered preoperatively. Gram-negative coverage should also be instituted if the biliary tree has been previously instrumented. An epidural catheter is useful for postoperative pain control unless otherwise contraindicated. A central venous catheter and arterial line are placed to facilitate haemodynamic management.

Preoperative and intraoperative communication with the anaesthesiology team is crucial for any major hepatic resections. The key to successful intraoperative patient care is low central venous pressure (LCVP) anaesthesia in conjunction with hepatic inflow and outflow control. The central venous pressure is kept at or below 3 mm Hg to preclude vena caval distension and facilitate mobilization of the liver and dissection of the retrohepatic vena cava and major hepatic veins. LCVP minimizes hepatic venous bleeding during parenchymal transection and facilitates control of inadvertent venous injury. The blood loss resulting from a vascular injury is directly proportional to the pressure gradient across the vessel wall and the fourth power of the radius of the injury, as illustrated by Poiseuille's Law. In addition to decreasing the pressure component of the equation, LCVP also minimizes the radial component of flow by reducing vessel distension (Fig. 54.2).

Fluid restriction is an important component of this technique and is continued until the resection is complete. Intravascular hypovolemia is counteracted by placing the patient in Trendelenburg position to improve venous return, and small fluid boluses may be given to maintain haemodynamic stability. With this cooperative surgical-anaesthetic technique, major hepatic resections can be performed with minimal blood loss and morbidity.

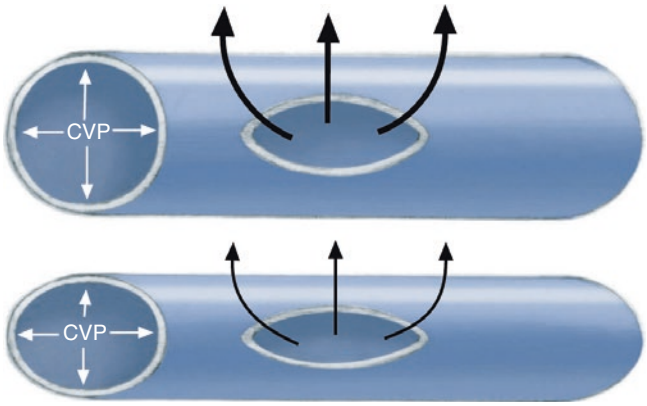

Fig. 54.2 Reduced vessel distension with low central venous pressure (CVP)

54.3 Surgical Technique

The patient is positioned supine with a roll under the upper abdomen to facilitate exposure. Slight right lateral decubitus position with the right side elevated at a 30° angle may be helpful in right hepatectomies. Hepatic resection is usually performed through a chevron, or bilateral subcostal, incision. Frequently, this incision is extended vertically in the midline to allow visualization of the IVC at the confluence of the hepatic veins. Raising a small skin flap in the midline and incising only the midline fascia can sometimes be done (Fig. 54.3a). Doing so avoids the upper skin extension and allows for a more cosmetic closure. A right subcostal incision with vertical midline extension up to the base of xiphoid cartilage ("hockey stick") can also be used for patients with narrow costal margins (Fig. 54.3b). This approach provides similar exposure to a bilateral subcostal incision with vertical midline extension, and results in better healing. A right thoracoabdominal incision (Fig. 54.3c) may be required in patients with large lesions that arise high in the right lobe with or without diaphragmatic invasion. This approach is preferable if caval resection is planned, as it facilitates control of the supradiaphragmatic vena cava and access to the right atrium. A left thoracoabdominal incision is rarely required. The exposure is maintained with a self-retaining retractor attached to the operating table.

The falciform ligament is identified and the ligamentum teres (round ligament) is ligated and divided as the abdomen is entered. The falciform ligament should be divided close to the anterior abdominal wall in order to preserve it for possible future use. The ligamentum teres is used later as a landmark for the umbilical fissure.

The liver is widely mobilized by sharply dividing the right triangular and coronary ligaments to the falciform ligament anteriorly and the IVC posteriorly (Fig. 54.4). The left triangular ligament is also separated from the diaphragm, with care taken not to injure the left phrenic vein. The suprahepatic vena cava and the hepatic veins are visualized. Thorough palpation of the entire liver should be carried out to identify multifocal disease and congenital anomalies. Intraoperative ultrasound, if available, may be performed at this stage to look for satellite lesions and to further delineate vascular and ductal anatomy. Tumour proximity or invasion into a vascular or ductal structure should be identified to better plan resection and reconstruction.

Once resectability is confirmed, the hepatoduodenal ligament is identified and surrounded with a looped vascular tape to expedite a subsequent Pringle manoeuvre (Fig. 54.5). A Rumel tourniquet may also be used. After mobilizing the liver and obtaining control of the hepatoduodenal ligament, the cystic duct and artery should be ligated and divided. The gallbladder infundibulum can be dissected free from the liver and retracted superiorly to expose the hilar plate.

The key principle in all major hepatic resections is control of vascular structures **prior** to parenchymal division. This control can be achieved with extrahepatic vascular control or hepatic pedicle ligation.

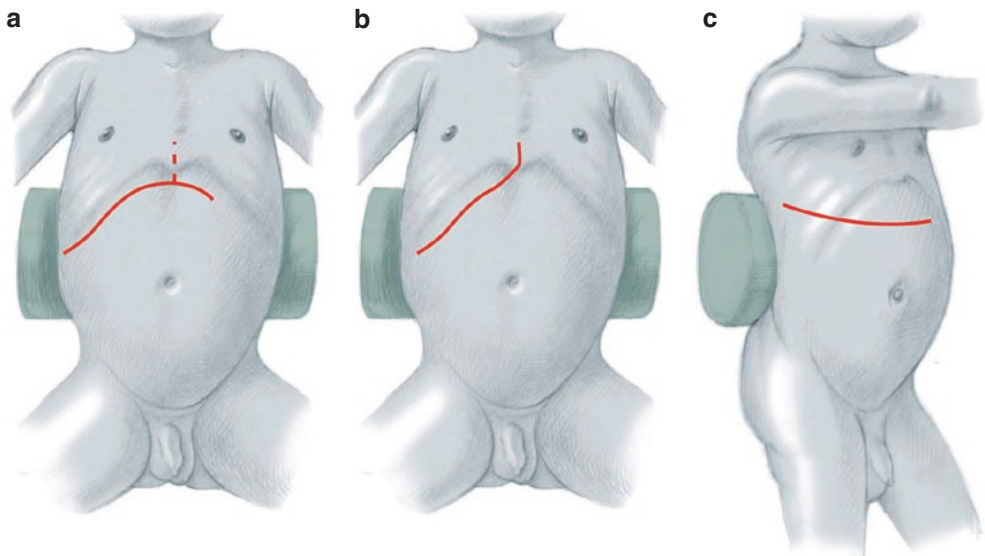

Fig. 54.3 Surgical approaches for hepatic resection: bilateral subcostal incision with midline extension (**a**); right subcostal incision with vertical midline extension to the xiphoid (**b**); right thoracoabdominal incision (**c**)

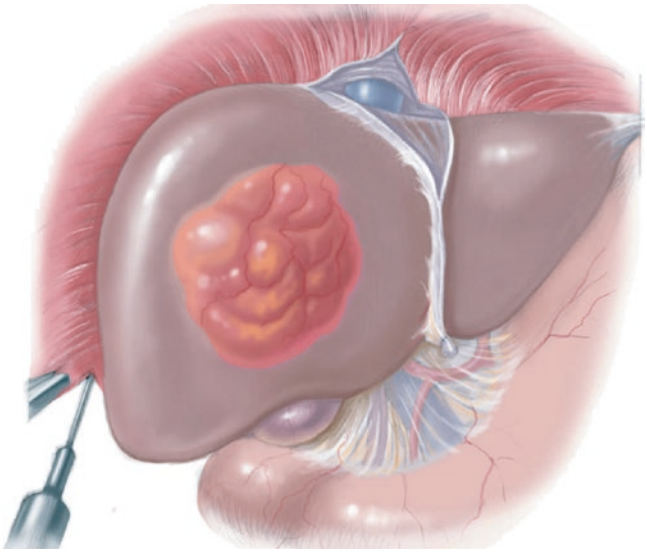

Fig. 54.4 Mobilizing the liver

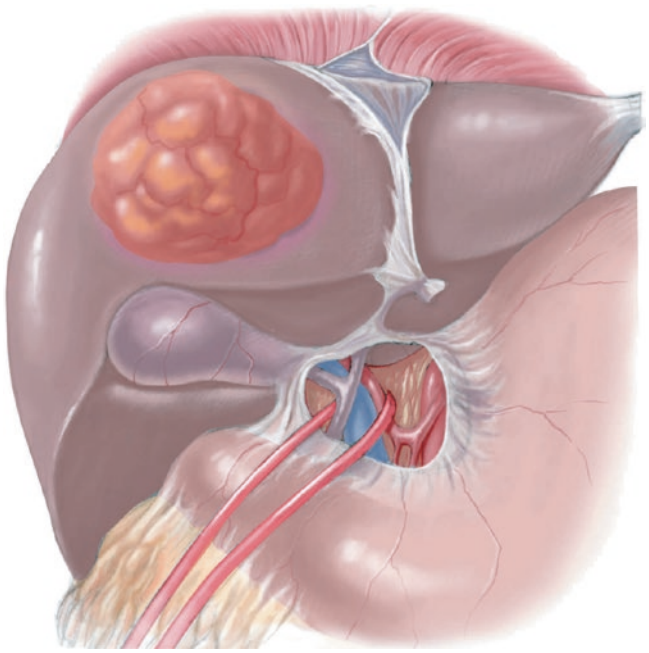

Fig. 54.5 Surrounding the hepatoduodenal ligament with a loop

54.3.1 Right Hepatectomy

Right hepatic resections are the most frequently performed major liver resections (60%). Segments V, VI, VII, and VIII are removed in right hepatic lobectomy. An extended right hepatectomy or right trisegmentectomy also includes removal of segment IV, and segment I may be included as well. The first step in vascular control of segments V through VIII is the exposure of the right hepatic artery and the right branch of the portal vein by lowering the hilar plate. The hilar plate is divided and lowered using sharp dissection to expose the bifurcation of the hepatic artery and portal vein, as well as the confluence of the hepatic ducts.

The right hepatic artery arises from the main hepatic trunk and usually passes posterior to the common bile duct. With the common bile duct gently retracted to the left, the right hepatic artery is isolated and divided. The right branch of the portal vein is then dissected free and divided between vascular clamps or ligated in continuity (Fig. 54.6). The proximal stump is oversewn with fine Prolene sutures. Alternatively, a vascular stapler can be used.

To minimize injury to the biliary tree, the extrahepatic biliary dissection can be withheld and the right hepatic duct controlled and divided intrahepatically during subsequent parenchymal transection. If the right hepatic duct is divided at the hilum, the confluence of the common bile duct and the left hepatic duct must be confirmed to be well preserved and intact.

Alternatively, a right pedicle ligation technique can be employed (Fig. 54.7). Hepatotomies are made at the base of the gallbladder fossa and the caudate lobe. The right pedicle is isolated intraparenchymally through the hepatotomies and divided with a vascular stapler. The advantage of this manoeuvre is that the left pedicle is well protected. If the tumour is close to the hilum, however, extrahepatic inflow control will be necessary to achieve adequate tumour clearance.

Following inflow control of the right lobe, the umbilical fissure must be defined, as the left portal triad enters the liver at the base of the umbilical fissure. The umbilical fissure is exposed and opened by retracting the ligamentum teres superiorly (Fig. 54.8), exposing the left portal vein, left hepatic artery and left hepatic duct. The segmental portal triads to segment IV can be controlled during parenchymal transection to avoid injury to the vascular supply or biliary drainage of the left lateral segment as well as the caudate lobe, if it is to be preserved. If the tumour encroaches upon the umbilical fissure, however, the segmental IV branches should be isolated and divided within the fissure.

Control of the hepatic vein is the most delicate manoeuvre of any hepatic resection. Attempts at early control of the hepatic vein before hilar vascular control can result in hepatic vein injury and haemorrhage. Positioning the patient in Trendelenburg with

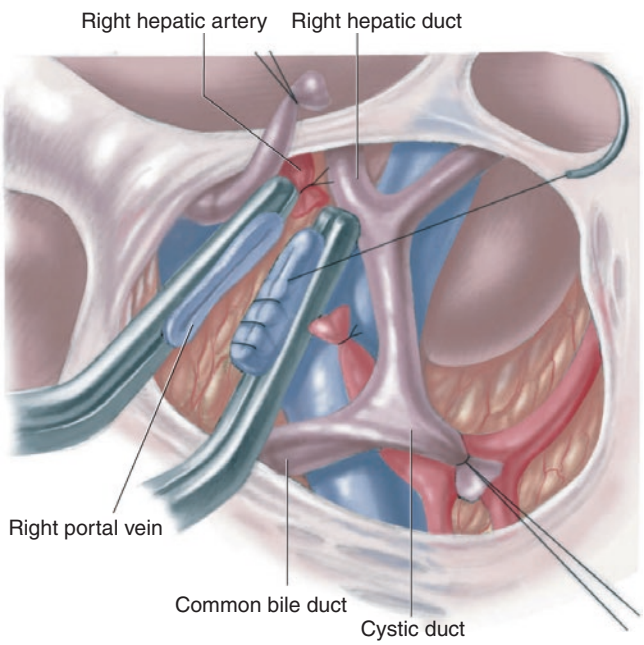

Fig. 54.6 Division of the right portal vein

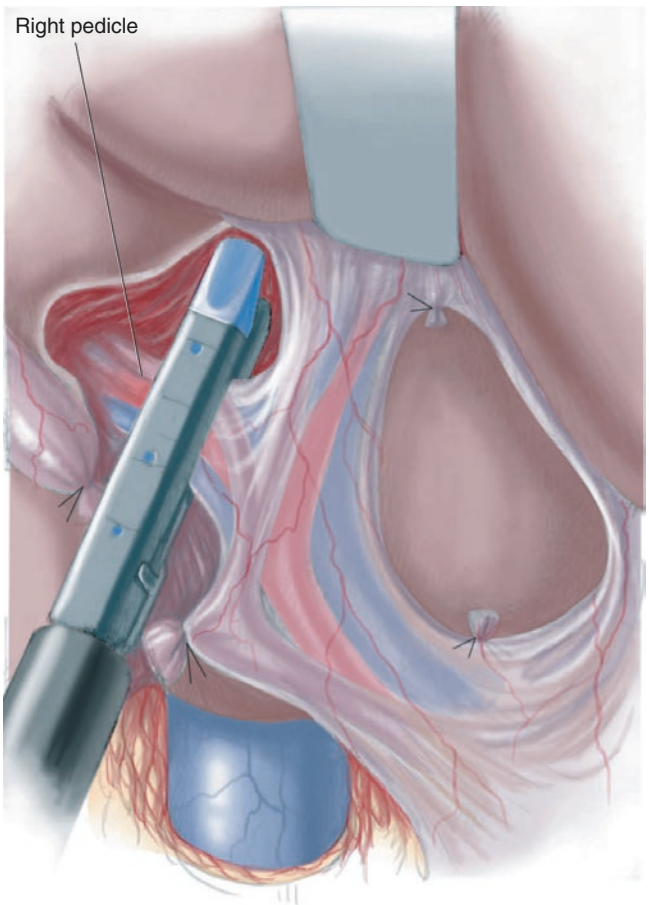

Fig. 54.7 Right pedicle ligation

15° head down reduces pressure in the IVC and hepatic veins, thus minimizing both blood loss and the risk of air embolus.

The division of the right hilar structures will reduce blood flow through the right hepatic vein and facilitate its dissection. The right lobe is rolled toward the midline, exposing the retrohepatic vena cava. The right upper portion of the retrohepatic vena cava is obscured by the IVC ligament. This fibrous band of tissue needs to be sharply divided to expose the right hepatic vein as it meets the retrohepatic vena cava. Next, small, unnamed hepatic veins that extend from the retrohepatic vena cava to the right hepatic lobe and the caudate lobe are ligated or clipped and divided individually in an inferior to superior fashion. For large right-sided tumours, a large unnamed hepatic vein is usually found before the main right hepatic vein. After this vein is divided, the junction of the right hepatic vein and the retrohepatic vena cava is carefully exposed and the right hepatic vein is encircled with a vessel loop. The right hepatic vein is then suture ligated, oversewn, or divided with a vascular stapler (Fig. 54.9).

In an extended right hepatectomy, the middle hepatic vein is also resected. The middle hepatic vein usually joins the left hepatic vein, but on occasion it may enter the vena cava separately. Extrahepatic isolation of the middle hepatic vein can be difficult and may risk injury to the left hepatic vein. Thus, the middle hepatic vein is usually controlled during parenchymal transection.

After vascular isolation of segments IV through VIII is complete, the line of ischaemic demarcation becomes apparent. The liver capsule (Glisson capsule) is scored with electrocautery along the line of devascularization. The hilar vessels are occluded with the Pringle manoeuvre intermittently for no more than 15 min at a time. This manoeuvre ensures minimal blood loss during parenchymal transection. The hepatic substance is divided slowly and carefully after fracturing with a Kelly clamp. Vascular and biliary radicals are meticulously secured with haemoclips or suture ligatures. A Cavitron ultrasonic aspirator, water jet dissector, laser, or Harmonic scalpel may also be used for parenchymal division in difficult cases or based on the surgeon's preference.

If the recurrent branches to segment IV are still intact, parenchymal dissection is carried out anteriorly down to the base of the umbilical fissure and the arterial and portal vessels to segment IV are then identified and divided. This dissection is continued posteriorly toward the vena cava and along the plane just to the right of the falciform ligament. The middle hepatic vein is encountered and controlled by suture ligation or division with the vascular stapler (Fig. 54.10). The left hepatic vein is carefully preserved.

After the specimen is removed and the Pringle manoeuvre is released, the raw cut surface of the liver is observed for haemostasis. The argon beam coagulator is useful in controlling oozing from the cut surface. Major sources of haemorrhage should be suture ligated. A Valsalva manoeuvre will increase the hepatic venous pressure and is helpful in identifying additional bleeding. Topical agents such as thrombin, fibrin glue, or Gelfoam also may be applied.

Occasionally a large tumour in the base of segment IV may encroach upon the left hepatic duct as it enters the umbilical

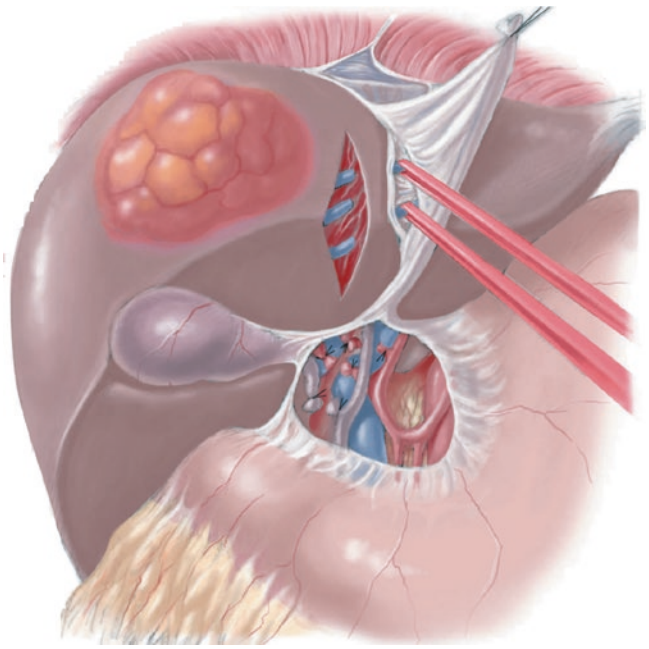

Fig. 54.8 Retracting the ligamentum teres to expose the umbilical fissure

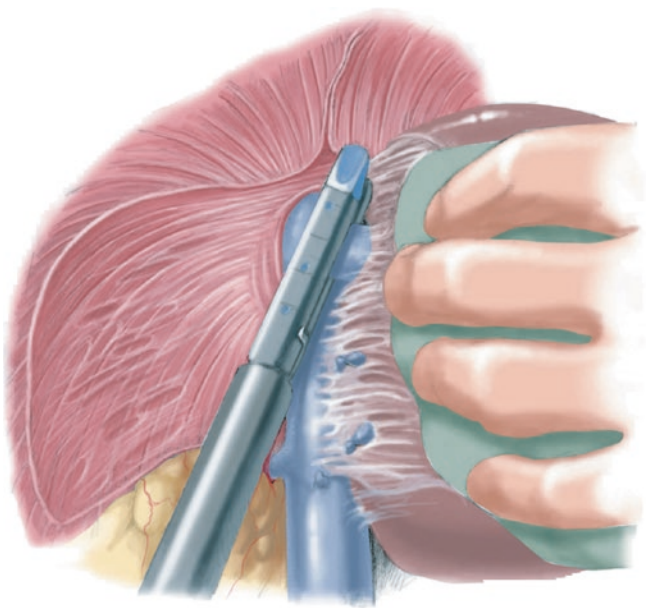

Fig. 54.9 Dividing the right hepatic vein

fissure. In such cases, mobilization of the left hepatic duct at the hilar area can be difficult and a portion of the left hepatic duct may be resected. The biliary drainage can be reconstructed with a Roux-en-Y hepaticojejunostomy (Fig. 54.11).

At the conclusion of an extended right hepatectomy, segments I, II, and III are left intact. The remaining liver parenchyma is then inspected to ensure viability. The operative field is copiously irrigated. Elective and uncomplicated hepatic resections typically do not need drainage.

The midline portion of the incision is closed with interrupted absorbable monofilament sutures. The subcostal incision is then closed by re-approximating the peritoneum and transversus abdominis muscle. The internal oblique and the posterior sheath are closed next, followed by the external oblique and anterior rectus sheath. The subcutaneous space is irrigated again and Scarpa's fascia is approximated to eliminate dead space. Finally, the skin is closed in a subcuticular fashion.

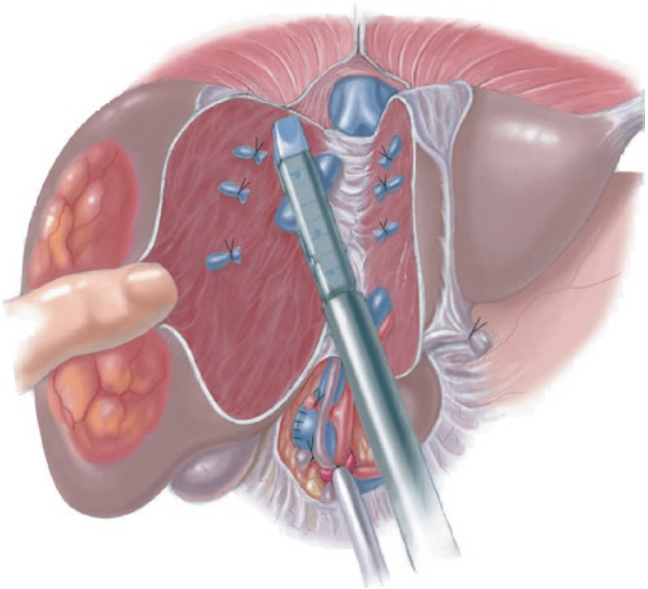

Fig. 54.10 Dividing the middle hepatic vein

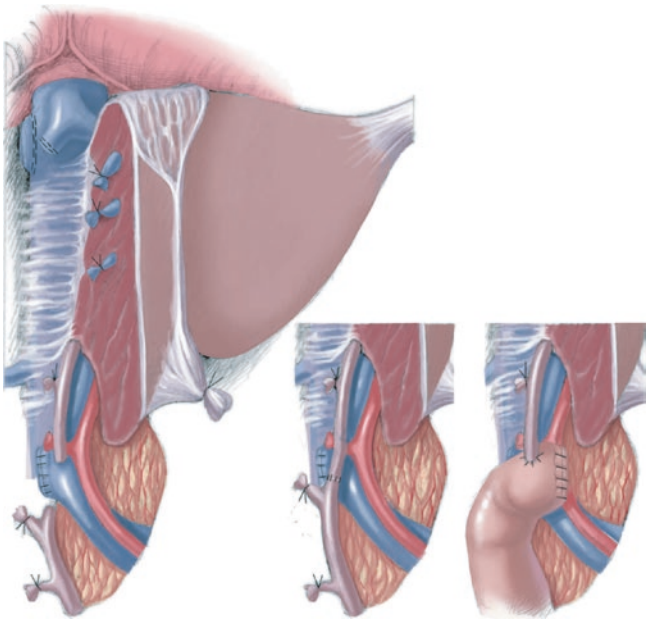

Fig. 54.11 Reconstructing biliary drainage with a Roux-en-Y hepaticojejunostomy

54.3.2 Extended Left Hepatectomy

Extended left hepatectomy is necessary when a large tumour arising from the left lobe of the liver crosses the portal fissure into segments V and VIII. It may also be carried out to clear multifocal disease. The left lobe and the right anterior sector, which consist of segments II, III, IV, V, and VIII, are removed en bloc, and segment I (caudate lobe) may also be included in the resection.

The liver is completely mobilized by dividing both the right and left ligamentous attachments. Inflow control is obtained by the ligation and division of the left hepatic artery at the base of the umbilical fissure (Fig. 54.12). The left portal vein is exposed along the umbilical fissure and the branches to the caudate lobe are identified. The portal vein is divided distal to the take-off of the caudate branches. If the caudate lobe is to be resected, the left hepatic artery and portal vein can be controlled at the hilus. The left hepatic duct is then identified, ligated, and divided.

The suprahepatic vena cava is fully exposed in the bare area of the liver. The middle and left hepatic veins are controlled next. The middle hepatic vein usually joins the left hepatic vein, but occasionally it may enter the vena cava separately. The liver is rotated laterally, exposing the ligamentum venosum, which is then divided. Division of the ligamentum venosum creates a plane in which the middle and left hepatic veins can be encircled and divided (Fig. 54.13). Division of these veins can be accomplished by oversewing the stump or using the vascular stapler. If the caudate lobe is to be removed, it is carefully mobilized from the vena cava by serially ligating the short retrohepatic veins with sutures or clips.

Inflow control to segments V and VIII is accomplished via ligation of the right anterior sectoral pedicle. Hilar dissection is carried out along the right portal vein and right hepatic artery until they branch into anterior and posterior pedicles (Fig. 54.14). Partial parenchymal division may be required for this dissection. The fissure of Gans is a reliable indicator of the course of the posterior pedicle and is used for orientation. Once the anterior pedicle is identified and encircled, it can be suture ligated or divided with the vascular stapler.

Successful vascular control will result in a clear ischaemic demarcation on the surface of the liver and will indicate the plane of parenchymal transection. This plane lies horizontally, lateral to the gallbladder fossa, parallel and just anterior to the right hepatic vein. The parenchymal transection is then carried out along this plane of demarcation as described previously.

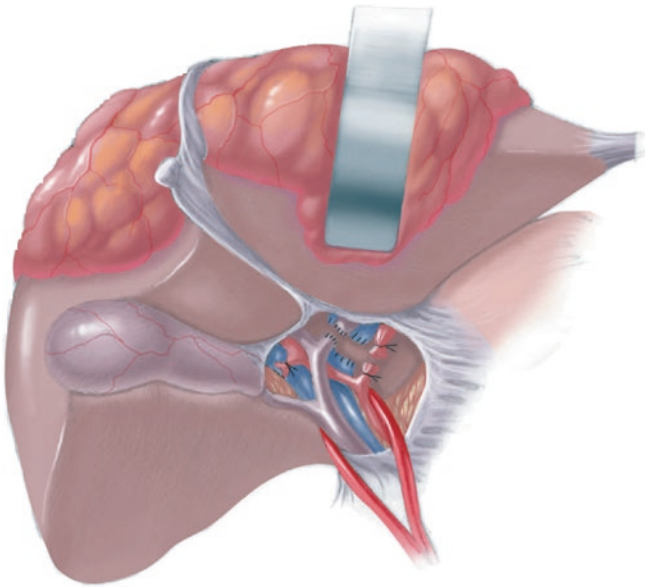

Fig. 54.12 Ligating the left hepatic artery

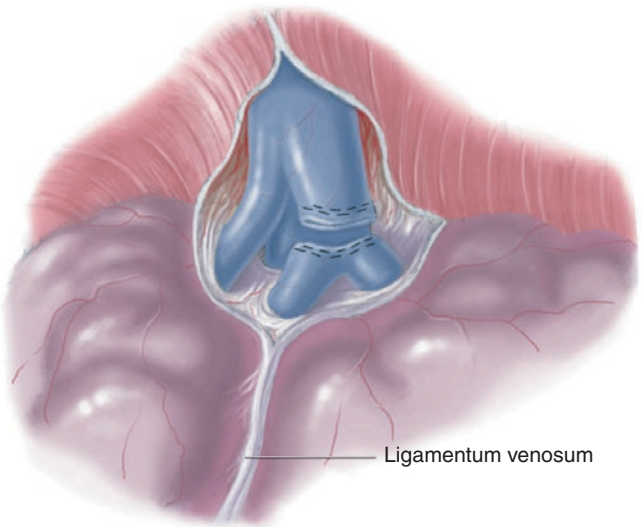

Fig. 54.13 Dividing the middle and left hepatic veins after division of the ligamentum venosum

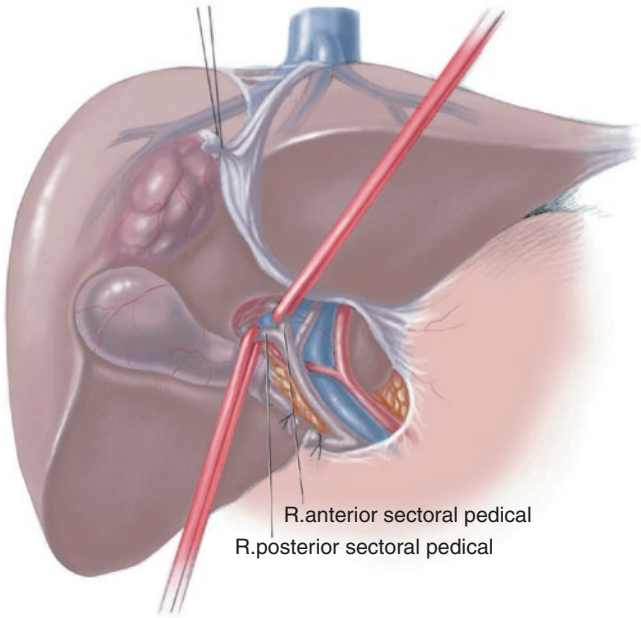

Fig. 54.14 Hilar dissection to control inflow to segments V and VIII

54.3.3 Central Hepatic Resection

Central hepatic resection removes segments IV, V, and VIII. Indicated for centrally located hepatic lesions, central hepatic resection avoids the use of extended hepatectomy. Less extensive resection allows for the preservation of normal hepatic parenchyma and may lower the risk of late complications such as biliary tract stricture.

The liver is mobilized and the porta hepatis is exposed and surrounded with a vascular tape. The hilar plate is lowered and the confluence of the right and left hepatic arteries and portal veins is exposed as described earlier. The recurrent branches of the left portal vein to segment IV are divided in the umbilical fissure, while taking care to preserve the branches to the left lateral segment and the caudate lobe. The middle hepatic vein is ligated as dissection approaches the vena cava (Fig. 54.15). The left hepatic vein should be carefully preserved.

Control of the arterial and portal inflow to segments V and VIII is accomplished by ligation of the right anterior sectoral pedicle (Fig. 54.15). Ligation will lead to a clear demarcation of segments V and VIII anterior to the right hepatic vein, which is used as the plane of parenchymal transection. Segments IV, V, and VIII are now devascularized and isolated from the rest of the liver. A Pringle manoeuvre is performed, and the remaining hepatic substance posteriorly and inferiorly is divided with gentle crushing technique using a Kelly clamp and meticulous haemoclip ligation of vascular and biliary radicals. At the completion of a central hepatic resection, the middle hepatic vein has been ligated and the right and left hepatic veins remain intact.

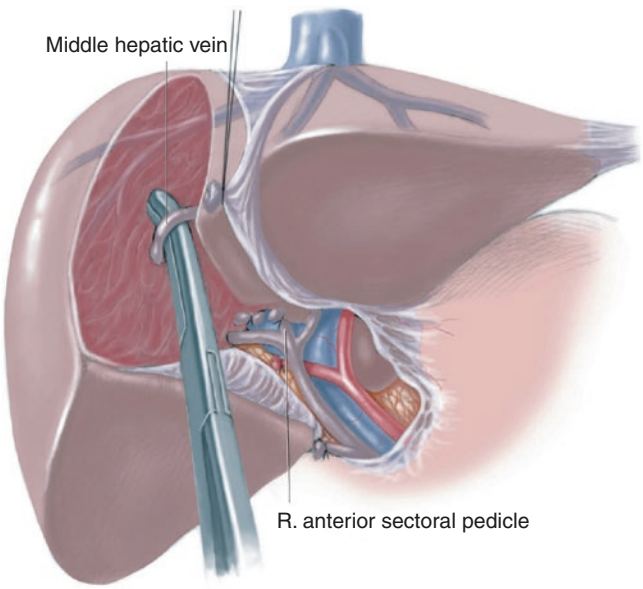

Fig. 54.15 Ligating the middle hepatic vein and the right anterior sectoral pedicle

54.3.4 Segmentectomy

The widespread use of intraoperative ultrasound, combined with superior understanding of Couinaud's classification, has enabled surgeons to perform anatomic segmentectomies successfully. Segmentectomy or multiple simultaneous segementectomies are best reserved for focal lesions in one segment, particularly metastases or smaller tumours. Control of arterial and portal inflow is completed via ligation of the appropriate segment's pedicle. Depending on the location of the lesion, dissection for vascular control may be technically demanding and is aided by advanced preoperative and intraoperative imaging. For example, for a right posterior segmentectomy, the right posterior segmental portal pedicle must be accessed while the right hepatic vein is preserved. With segmentectomy, posterior or central tumors that traditionally would have required right or left hepatectomy may be resected without loss of otherwise normal liver tissue. Functional liver parenchyma is thus preserved, which is of utmost importance for patients with impaired hepatic function. Moreover, in cases of recurrent disease, segmentectomy offers the opportunity to repeat curative resection without compromising functional parenchymal volume.

54.4 Outcomes

Major hepatic resection is safe, even in small infants. The operative mortality is less than 5% and can be reduced to nearly zero with judicious use of invasive monitoring, LCVP, and meticulous surgical techniques.

An overall survival of 85–90% is possible after complete surgical resection of non–stage IV hepatoblastoma. About 50% of patients who present with pulmonary metastases are curable. Removal of the primary hepatic lesions is of utmost importance for survival. If gross disease is left in situ in the primary site, survival rates fall to zero. Some patients with microscopic residual tumor are curable with continued chemotherapy and may benefit from external beam radiotherapy to the primary hepatic site. In a multivariate analysis, factors that have been independent predictors of worse prognosis include a high TNM stage, unresectable tumour, bilobar involvement and multifocal disease, α-fetoprotein <100 or >10^5 ng/mL, distant metastases, embryonal versus fetal histology, and vascular invasion.

In contrast, the overall survival from pediatric hepatocellular carcinoma is only about 20% at 5 years. Curative treatment of hepatocellular carcinoma remains a therapeutic dilemma, and chemotherapy is only partially effective. Complete resection of localized lesions remains the only realistic chance for cure.

54.5 Additional Treatment Options

About 46% of hepatic malignancies are resectable at diagnosis. Often, resection is not feasible if the tumours are large and involve both hepatic lobes. For unresectable tumours, the initial surgical procedure should include a diagnostic biopsy and placement of a vascular access device for chemotherapy. Neoadjuvant chemotherapy will often result in tumour shrinkage and thus allow for subsequent resection. If imaging studies after four cycles of chemotherapy show good response and resectable disease, a second laparotomy is performed. Again, complete resection of the primary tumour is necessary for survival and may require extended hepatic lobectomies or complex biliary reconstructions. The patient should also receive several cycles of chemotherapy after definitive resection.

In some cases, resection of primary disease requires complete hepatectomy and liver transplantation. Transplantation is a potentially curative treatment for chemosensitive hepatoblastomas when partial hepatectomy is not possible because of multifocal disease or suboptimal margins. Tumours with extensive extrahepatic extension or vascular invasion have poorer outcomes with total hepatectomy and hepatic transplantation.

Chemoembolization is a new, adjuvant treatment option in which cisplatin and/or doxorubicin is injected under arteriographic guidance, followed by injection of occluding thrombogenic materials such as Gelfoam into the arteries feeding the hepatic tumour. Data have shown that chemoembolization may result in some tumour reduction but does not improve resectability. In a variation of this approach, some researchers have used super-selective intra-arterial radiometabolic therapy for malignant hepatic tumours. Other approaches include treatment with anti–α-fetoprotein antibodies and the use of viral transfection vectors to attack malignant hepatic cells.

Conclusions

Infants and children with liver malignancies can be treated successfully with complete surgical resection using anatomic resections, with little subsequent morbidity. Patients with hepatoblastoma have excellent prognosis when treated with the combination of complete surgical resection and chemotherapy. Some patients with hepatocellular carcinoma can be cured with liver resection, but the poorer prognosis requires the development of additional treatment options.

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Testicular Tumours

55

Jonathan Ross

Testicular tumours account for 1–2% of pediatric solid tumours. Pediatric testis tumours are pathologically and clinically distinct from their adult counterparts, and these differences demand a different management algorithm. Nearly 90% of adult testis tumours are seminomas, embryonal carcinomas, or non-seminomatous mixed germ cell tumours, but such tumour types are extremely rare in prepubertal children. Conversely, the two most common prepubertal tumours—pure teratoma and pure yolk sac tumour—account for fewer than 1% each of adult tumours. Gonadal stromal tumours are relatively common in children but quite rare in adults. These differences in histologic subtype result in significant differences in natural history: metastases occur in 61% of adult patients but only 9% of children. The critical histologic changes appear to occur at puberty, so tumours occurring in postpubertal adolescent males are best managed under adult algorithms.

A majority of prepubertal testis tumours are benign. Tumours may be of germ cell or stromal origin (the exception being gonadoblastomas, which contain elements of both). Teratomas and epidermoid cysts are the most common tumours and are universally benign in prepubertal patients. Yolk sac tumours are by far the most common malignancy in prepubertal patients. Gonadal stromal tumours have generally behaved in a benign fashion in children, except for some undifferentiated stromal tumours and occasional Sertoli cell tumours in children over 5 years of age.

55.1 Presentation

Most testicular tumours present as a painless, hard mass, though rarely they may present with pain related to an acute bleed. On physical examination, a mass that cannot be separated from the testis is assumed to be a testis tumour until proven otherwise. Ultrasound is very helpful in making this distinction when the physical examination is unclear. Rarely, testis tumours may present with a reactive hydrocele. If a hydrocele is large and firm enough to preclude palpation of the testis, an ultrasound should be obtained.

55.2 Diagnosis and Preoperative Evaluation

Preoperative evaluation of prepubertal patients includes an ultrasound scan and determination of the α-fetoprotein (AFP) level. Ultrasound is helpful in distinguishing intratesticular from extratesticular scrotal masses. The ultrasonographic characteristics of various testis tumours have been described, though the ultrasound by itself cannot determine a histologic diagnosis. Serum AFP is elevated in 90% of children with yolk sac tumours. AFP is very specific for yolk sac tumour, but may be physiologically elevated in normal infants (including those with benign tumours). Generally speaking, a child over 1 year of age with a testicular mass and an elevated AFP can be assumed to have a yolk sac tumour. Infants under 1 year of age with an elevated AFP may have benign tumours, though AFP levels are rarely greater than 100 ng/mL in normal children over 6 months of age. If, based on the AFP level, a tumour is felt to be malignant, then an inguinal orchiectomy is performed. Tumours that are likely to be benign based on AFP level should be managed initially with an excisional biopsy and frozen section analysis. Even tumors that appear on ultrasound to replace the testis may be enucleated, leaving significant residual testicular tissue. If a benign histology is confirmed, then the testis is closed with absorbable suture and returned to the scrotum. If the biopsy reveals a malignant tumour

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(usually yolk sac) or potentially malignant tumour (such as an undifferentiated stromal tumour or a Sertoli cell tumour in an older child), then an inguinal orchiectomy should be performed. If a teratoma is diagnosed and the child is near pubertal age, the surrounding parenchyma should be examined for its pubertal status. If the tubules are immature, then the tumour may be treated as benign, but if the tubules show evidence of maturation, the tumour should be treated as potentially malignant because some adult teratomas behave in a malignant fashion.

Retroperitoneal lymph node dissection (RPLND) plays a very limited role in prepubertal testis tumours. Unlike adults, only a minority of prepubertal patients with metastases have metastases limited to the retroperitoneum, and metastatic disease is very responsive to chemotherapy. Furthermore, the complication rate following RPLND is significantly higher in children than in adults. The only relative indications for retroperitoneal surgery in a prepubertal patient are to biopsy an equivocal node or to excise a persistent retroperitoneal mass following chemotherapy—a rare occurrence.

55.3 Surgical Technique

The patient is placed in the supine position. If possible, a caudal block has been applied. An inguinal skin incision is made, extending laterally from just above the pubic tubercle following Langer's lines. The subcutaneous tissue is divided with cautery. The superficial epigastric vein is often encountered in the lateral corner of the incision and should be controlled with cautery.

The superficial fat may then be separated bluntly with retractors, exposing Scarpa's fascia. Scarpa's fascia is grasped with forceps and opened with scissors (Fig. 55.1). The tissue between Scarpa's fascia and the external oblique aponeurosis is developed by gentle spreading, and Scarpa's fascia is then divided throughout the length of the incision.

A small incision is made in the external oblique aponeurosis near the lateral corner of the incision so that it comes through above the internal ring, where it is unlikely to injure the ilioinguinal nerve. After bluntly pushing the ilioinguinal nerve down from the undersurface of the aponeurosis, the aponeurosis is opened with scissors through the external ring.

The cremaster sheath is opened and, with blunt dissection, the plane behind the cord is developed and a vessel loop is passed behind it. With a combination of blunt and cautery dissection, this plane is developed proximally to the internal ring and the vessels are controlled at the internal ring with a rubber shod clamp or vessel loop. Then the testis is delivered into the operative field and the gubernaculum is divided with cautery (Fig. 55.2).

If the preoperative assessment suggests a malignancy, then the hydrocele sac is left intact and the specimen is ligated with permanent suture and divided at the internal ring without manipulation of the testicle (*not shown*). If a benign tumour is suspected, then the hydrocele sac is opened (Fig. 55.3a) after draping off the testis to isolate it from the field (*drapes not shown*). The tumour is excised or enucleated (Fig. 55.3b) and a frozen section analysis is obtained. If the tumour is benign on frozen section, then the testis is closed with 5/0 absorbable suture (Fig. 55.3c) and returned to the scrotum. If the tumour is malignant, then the cord is doubly suture-ligated with nonabsorbable suture and divided at the internal ring. The entire specimen is sent for histopathologic evaluation. The incision is closed in layers with absorbable suture.

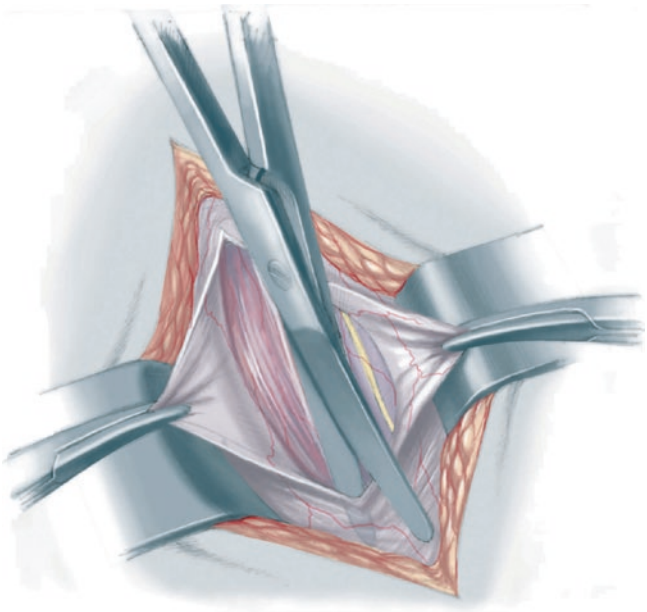

Fig. 55.1 Opening Scarpa's fascia

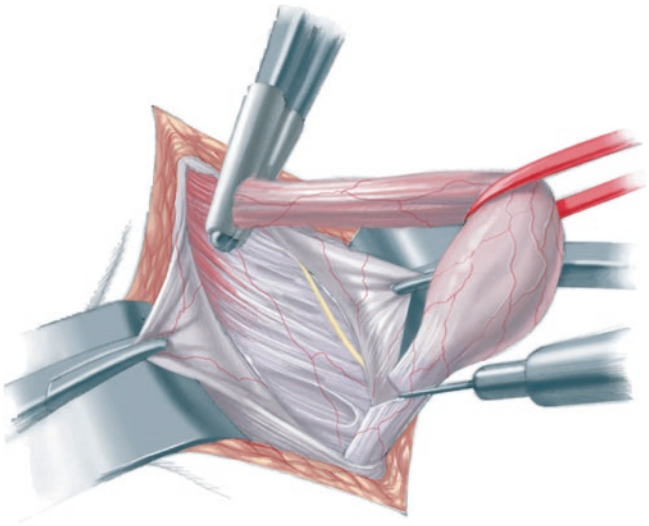

Fig. 55.2 Delivering the testis and dividing the gubernaculum

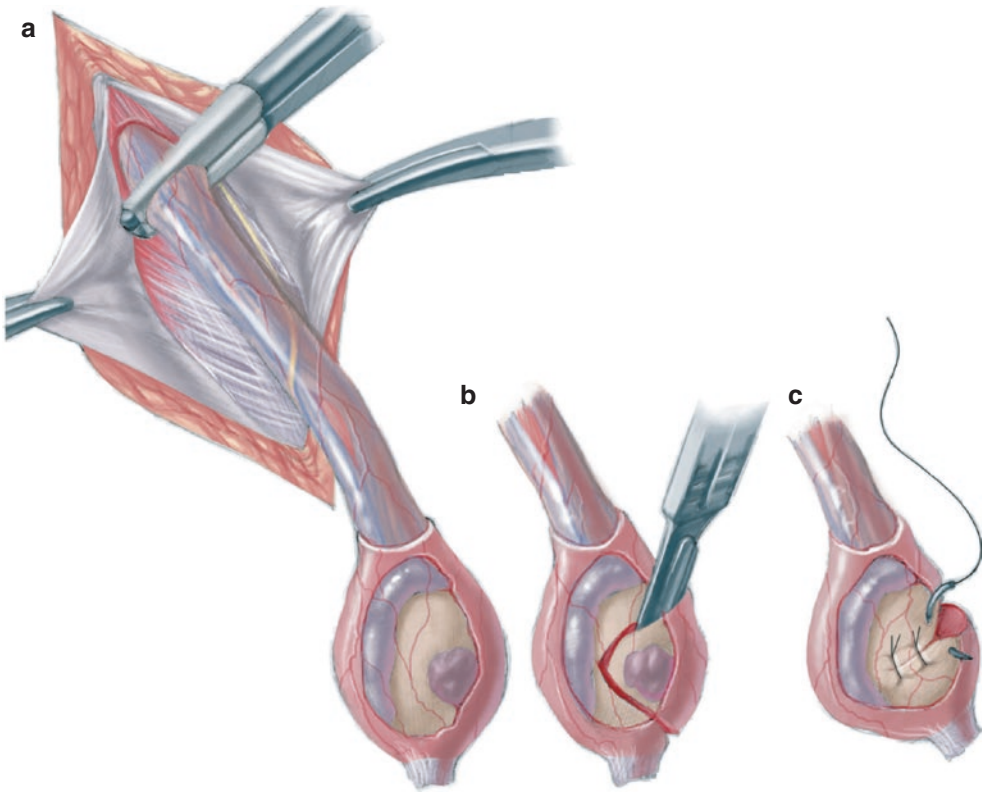

Fig. 55.3 Opening the hydrocele sac (**a**), excising the tumour (**b**), and closing the testis (**c**) in removing a benign tumour

55.4 Complications

Surgical complications are rare following inguinal orchiectomy or tumour excision. Local recurrence following simple excision has not been reported for benign tumours or following inguinal orchiectomy for malignancy. Local recurrence in the scrotum has occurred following transscrotal biopsy of malignant tumours, however, so a scrotal approach for possible malignancies should be avoided. The occasional complications that can occur following inguinal orchiectomy are scrotal infection or haematoma and retroperitoneal haematoma. These complications generally can be avoided with meticulous surgical technique and thorough haemostasis of the raw surface of the inner scrotum.

55.5 Outcome and Follow-Up

The survival of children with testis tumours depends on the histology of the tumour and the presence or absence of metastatic disease. Children with teratomas, epidermoid cysts, and benign stromal tumours do well with excision alone. No long-term follow-up is required.

Patients with yolk sac tumour require a metastatic evaluation consisting of a CT scan of the abdomen and pelvis, a chest x-ray or chest CT scan, and determination of the serum AFP level. The half-life of AFP is 5 days. Approximately 80% of patients will have stage 1 disease (disease limited to the testicle) confirmed by a negative radiographic evaluation and normalization of the AFP level. These patients may be observed closely without adjuvant therapy. Historically, follow-up has included CT scans every 2 months and chest x-rays and AFP levels monthly for 2 years, followed by observation at longer intervals. However, consideration should be given to decreasing the frequency of CT scanning by relying more on AFP levels and MRI, to minimize the long-term risks of radiation exposure from multiple CT scans. The relapse rate for stage 1 yolk sac tumour patients is approximately 20% but virtually all patients can be salvaged with chemotherapy. Patients who present with metastatic disease are treated with adjuvant chemotherapy, and survival is nearly 100% for this group as well. Radiation plays no role in the primary treatment of these tumours. Metastatic stromal tumours, though exceedingly rare, are resistant to treatment. Survival is low for this group.

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Soft Tissue Sarcomas

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Soft tissue sarcomas are a heterogenous group of malignant tumors that arise from primitive mesenchyme, and thus arise in any anatomic location. They are uncommon and account for 7.4% of all cancers in children under 20 years of age. They are classified into rhabdomyosarcoma (RMS) and non-rhabdomyosarcoma soft tissue sarcoma (NRSTS). RMS histologically resembles striated skeletal muscle, and is more common in children under 5 years of age. In contrast, NRSTS is more common in adolescents older than 10 years.

RMS is the third most common extracranial solid tumor in infants and children. Its presentation follows a bimodal age distribution in children under 6 years of age or adolescents aged 10–15 years. RMS is associated with the Li-Fraumeni cancer susceptibility syndrome with germline p53 mutations, Pleuropulmonary Blastoma Family Tumor and Dysplasia syndrome with germline *DICER1* mutations, Costello syndrome with germline *HRAS* mutations, Noonan and Beckwith-Wiedemann syndromes, and neurofibromatosis type 1. Histologically, the cells are characterized by cross-striations similar to those in skeletal muscle or rhabdomyoblasts. Immunohistochemical staining for desmin, myogenin, and MyoD1 is typically positive. The two main histologic subtypes of RMS are alveolar and embryonal. The embryonal subtype is more common in males, presents in younger patients under 8 years of age, and is most commonly found in the head and neck and in genitourinary regions. These tumors often show loss of heterozygosity (LOH) at the 11p15 region with preferential maternal allele loss. Alveolar tumors arise more commonly in the extremities, trunk, and perineal regions, and occur in patients older than 10 years. Most alveolar tumors have translocations between the *FOXO1* gene and the *PAX3* or *PAX7* genes. *PAX3-FOXO1* fusion is more common and is associated with older patient age and worse overall survival, independent of the presence of distant metastases.

NRSTS includes neoplasms of all other mesenchymal tissues besides skeletal muscle. NRSTS is more common in adolescents and adults, with a slight male predominance. In pediatric patients, the most common tumor subtypes are synovial sarcoma (25–30%), and undifferentiated or unclassified sarcoma not otherwise specified (NOS) (20%). Uniquely, in some NRSTS, pathologic grade determines the management but may not correlate with clinical outcome. Infantile fibrosarcoma and hemangiopericytoma appear histologically aggressive but are chemosensitive and may have a relatively benign clinical course following complete surgical resection. Low-grade tumors like desmoid fibromatosis have a high propensity for local recurrence despite extensive resection. Indolent lesions like desmoid tumors can be observed if they are stable, not growing, asymptomatic, or are located in a site where surgery is technically difficult or potentially debilitating. Large, high-grade, infiltrating tumors (>5 cm, T2b, Grade 3) have a tendency for distant metastases and poor patient survival. Many NRSTSs are also characterized by chromosomal translocations, such as *SYT-SSX* fusion in synovial sarcoma and *EWS-WT1* fusion in desmoplastic small round cell tumor. More than two thirds of NRSTSs occur in the extremities or trunk.

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56.1 Diagnosis and Evaluation

56.1.1 Imaging

Initial evaluation of a suspected soft tissue sarcoma begins with imaging studies to strengthen the preoperative diagnosis, guide planning of the diagnostic biopsy, and evaluate for distant disease. Contrast-enhanced CT scans are useful for truncal and intracavitary lesions and to evaluate for pulmonary metastases. Gadolinium-enhanced MRI is useful for extremity lesions, particularly to visualize relationships to neurovascular structures. Positron emission tomography (PET) scans have not been used routinely in pediatric soft tissue sarcomas but may have a role in the evaluation of tumor response to therapy.

56.1.2 Staging

The most commonly used staging system for soft tissue sarcomas involves presurgical and postsurgical evaluations (Tables 56.1 and 56.2). In addition, a TNM staging system is used for NRSTS (Table 56.3). Key prognostic factors for RMS include tumor site and size; the extent of disease as defined by the pretreatment Stage and postresection Group; histologic subtype and biologic factors; and age, with children aged 1–9 having the best prognosis. The prognoses of patients with NRSTS vary greatly with histologic type and patient age; NRSTSs in adolescents behaving more like those in adults. Incomplete surgical resection, intracavitary tumor sites, high mitotic index, and large tumor size—alone (>5 cm) or in proportion to body surface area—are associated with poorer outcomes.

Table 56.1 Children's oncology group soft tissue sarcoma (COG-STS) pretreatment staging system

Stage	Site of primary tumor	T	Size	Regional lymph node involvement	Distant metastasis
1	Favorable	T1 or T2	Any	Any	Absent
2	Unfavorable	T1 or T2	≤5 cm	Absent	Absent
3a	Unfavorable	T1 or T2	≤5 cm	Present	Absent
3b			>5 cm	Any	
4	Any	T1 or T2	Any size	Any	Present

T1—tumor confined to anatomic site of origin (noninvasive); T2—tumor extension and/or fixation to surrounding tissue (invasive)

Table 56.2 COG-STS postresection surgical-pathologic group system

Group	Definition
Group I	Localized, completely resected tumors
Group II	Localized, grossly resected tumors, with any of the following: • Microscopically positive margins • Microscopically negative margins, positive regional lymph nodes • Microscopically positive margins and positive regional lymph nodes
Group III	Gross residual disease after resection or biopsy only
Group IV	Distant metastases present at diagnosis

Table 56.3 TNM staging system for non-rhabdomyosarcoma soft tissue sarcomas (NRSTS)

Stage	T	Regional lymph node involvement	Distant metastasis
1	T1a–T1b	Absent	Absent
2	T2a–T2b	Absent	Absent
3	Any	Present	Absent
4	Any	Any	Present

a—tumor ≤5 cm in greatest dimension; b—tumor >5 cm in greatest dimension; T1—tumor limited to organ or tissue of origin; T2—tumor invades contiguous organs or tissues and/or with malignant effusion; TNM—tumor, nodes, metastasis

56.1.3 Biopsy

Key considerations in planning a diagnostic biopsy are the choice of biopsy technique and the placement of incisions. The least invasive biopsy technique should be selected, to minimize the risk of seeding while ensuring that sufficient tissue is obtained. Core needle biopsies are best for deep-sited tumors. Excisional biopsies should be performed only if the lesion is small and superficial. In most instances, an incisional biopsy is preferred. The incision should be placed in a way that will facilitate complete excision at a potential subsequent wide excision, which should be anticipated in most cases (Fig. 56.1). In extremity tumors, incisions are longitudinal and parallel to the neurovascular bundle, rather than transverse. Biopsy margins are marked to orientate the specimen. Doing so helps to direct the subsequent re-resection if a positive margin is detected on histology. Debulking surgery is typically unhelpful; it confers no survival advantage, does not downstage the disease, and may delay the initiation of adjuvant therapy. It may be useful, however, for intracavitary lesions with obstructive signs and symptoms such as hydronephrosis or bowel or bladder outlet obstruction. If hemorrhage is encountered with vascular lesions, a surgical drain should be placed, because an intralesional hematoma would increase the size and extent of the subsequent resection.

Regional lymph nodes should be biopsied if they are enlarged or clinically suspicious, and they should be actively sought in patients with trunk and extremity tumors, which have an increased risk of nodal metastases, especially T2b grade 3 lesions. RMS of the extremity and perineum and paratesticular RMS in children older than 10 years are associated with a higher incidence of nodal disease that is underdiagnosed on CT scans or MRI, so they require routine regional lymph node dissection to guide subsequent therapy. Sentinel lymph node biopsy and functional imaging with PET scans are not employed routinely to evaluate nodal disease in RMS, but they may help in guiding the need for regional or systemic therapy in select patients.

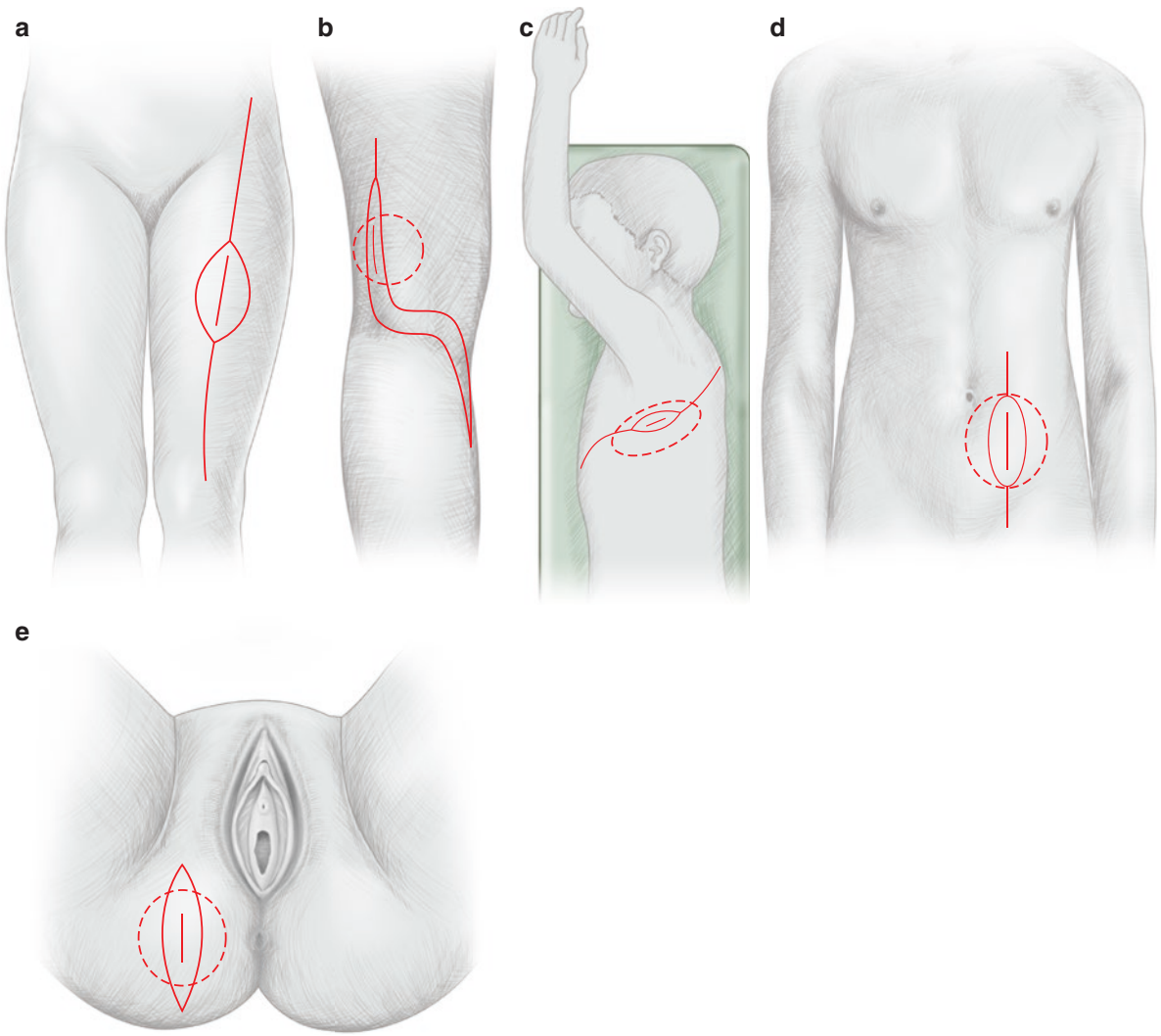

Fig. 56.1 Biopsy for soft tissue sarcomas: principles for planning a biopsy in various anatomical locations. (**a**) Extremity along a limb. (**b**) Extremity over a joint. (**c**) Trunk (chest wall). (**d**) Trunk (abdominal wall). (**e**) Perineum

56.2 Treatment

The management of RMS has transitioned towards a multimodal approach involving multiagent systemic chemotherapy with surgery and/or radiation therapy for local control. In contrast, NRSTS is not chemosensitive in most instances, so complete resection is the mainstay of treatment, with radiation therapy administered for close (<5 mm) margins and high-grade lesions.

The primary tumor should be completely resected with a circumferential margin of at least 5 mm of normal tissue where possible, without compromising form or function. This resection may be performed before chemotherapy if it will not cause significant disfigurement, functional compromise, or organ dysfunction. Marginal or subtotal resections are acceptable in sites that will not allow for a margin of normal tissue—such as RMS of the orbit or genitourinary region, which are sites associated with a good prognosis anyway—or in low-grade NRSTS (Grade 1–2) with less aggressive behavior, such as infantile fibrosarcoma.

If a prior unplanned biopsy or subtotal excision has been performed, and complete wide resection of the residual mass is still feasible, a primary re-excision (PRE) should be done before initiation of adjuvant therapy. This involves making a wider excision of the tumor bed to obtain negative surgical margins (Fig. 56.2). This may reduce the clinical Group, reduce the need for subsequent radiation and chemotherapy, and increase survival in these patients.

If the tumor is initially unresectable, biopsy should be followed by treatment with neoadjuvant multiagent chemotherapy, before employing further surgery and/or radiation therapy for local control. Residual tumor that remains after initial chemotherapy can be removed through a second-look procedure (SLP), also known as delayed primary excision. The finding of viable tumor or positive margins at SLP confers a worse prognosis, but the survival advantage conferred by SLPs is controversial. Radiation therapy is also given for proven lymph node disease, although noting the high morbidity that radiation confers to the adjacent tissues and organs of growing children.

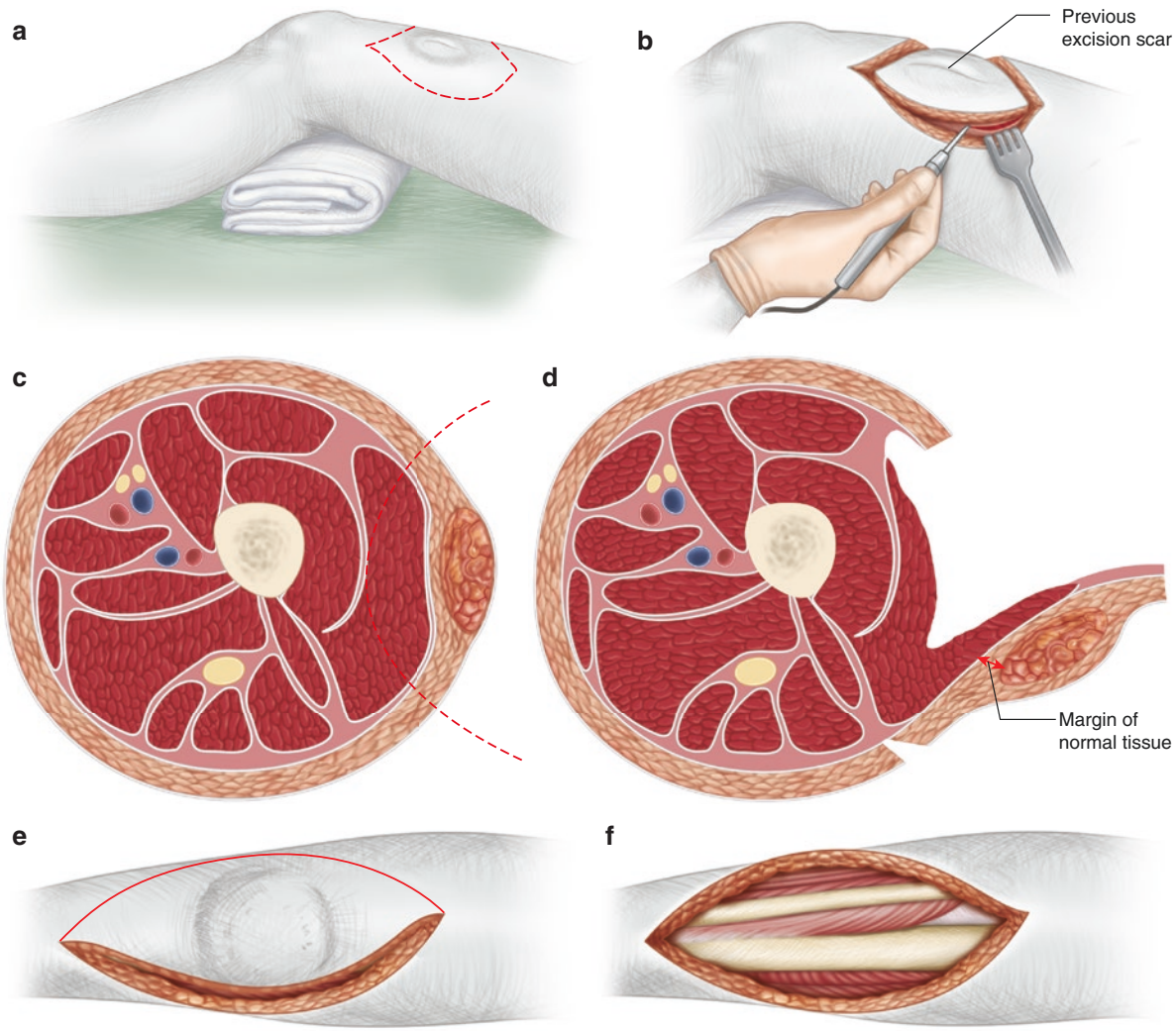

Fig. 56.2 Primary re-excision (PRE) and second-look procedure (SLP). (**a**) Circumferential margin around previous excision scar. (**b**) Circumferential dissection with a margin of normal tissue. (**c**) Deep margin below residual tumor. (**d**) Deep dissection with a margin of normal tissue. (**e**) Complete dissection around residual tumor and scar. (**f**) All residual tumor removed

56.2.1 Resection of Extremity Tumors

The goal of surgery is to perform a wide excision with clear resection margins, while avoiding excessively debilitating surgery. There is no need to resect the entire muscle group. Close margins of several millimeters may be acceptable and practical in children, as there is no evidence to dictate the optimal resection margin for these tumors. Primary tumors of the extremity have a poorer prognosis—particularly primary tumors of the hands or feet, flexor fossae, or groin and axilla, which have high-risk features and often are not amenable to complete resection without amputation. With definitive radiation therapy following complete surgical resection where possible, however, 10-year local control rates of 80–90% are achievable.

A longitudinal incision is made over the lesion, incorporating an ellipse of skin around the existing biopsy scar (Fig. 56.3). The incision is carried directly down to the lesion without raising skin flaps. The dissection continues circumferentially, including a margin of normal tissue with the tumor. The optimal tumor-free resection margin is not well established. In RMS, a margin of 0.5 cm is adequate, but in low-grade and high-grade NRSTS, a margin of more than 1 cm is associated with decreased risk of local recurrence. During the resection, constantly palpating and reorienting the lesion helps to maintain a consistent margin of normal tissue around the lesion.

Adjacent neurovascular bundles are isolated and slung with vessel loops for identification (Fig. 56.4). Maintain meticulous hemostasis while dissecting through the hypervascular tissues surrounding the tumor. If the procedure must be abandoned because of uncontrolled bleeding or significant persistent oozing from the resection site, a surgical drain should be left in place to prevent formation of a hematoma, which may cause vascular embarrassment to the limb or increase the size and extent of the subsequent resection.

During the resection, areas where gross tumor may remain, or areas where surgical margins are close, are noted and marked with surgical clips. The perimeter of the resection bed also should be marked with surgical clips to help in directing subsequent radiation therapy if a resection margin returns as positive or close on histology. Orientate the specimen with marking sutures (Fig. 56.4, *inset*), to help direct subsequent re-resection, should it be required.

If required, afterloading catheters for adjuvant brachytherapy may be placed prior to wound closure. Measure the dimensions of the area to be irradiated to determine the layout and number of catheters required. Leader catheters should be laid parallel to each other 1 cm apart, extending at least 1 cm proximal and distal to the target area as determined by a radiation oncologist. Using a percutaneous insertion device, catheters are inserted away from the wound edges and tunneled in a straight path to directly contact the target area, before exiting on the opposite side (Fig. 56.5a). The catheters should be laid as straight as possible, avoiding kinks and acute angles, and avoiding vital structures. The catheters are trimmed to length and anchored in place with radio-opaque buttons at either end; filaments are left in place within their lumens. The remaining muscle fascia is approximated, and the subcutaneous tissue and skin are closed in layers (Fig. 56.5b). A surgical drain should be placed in the wound bed to prevent formation of a postoperative seroma, which may interfere with wound healing and displace brachytherapy catheters away from the target area.

The limb is wrapped with gauze and a light elastic bandage. It is kept slightly elevated, to control the amount of postoperative wound site edema. Adequate distal circulation is ascertained in the operating room before reversing the patient, and neurovascular monitoring is continued postoperatively. If brachytherapy is to be performed, surgical drains and dressings are left in place until the end of the radiation treatment course.

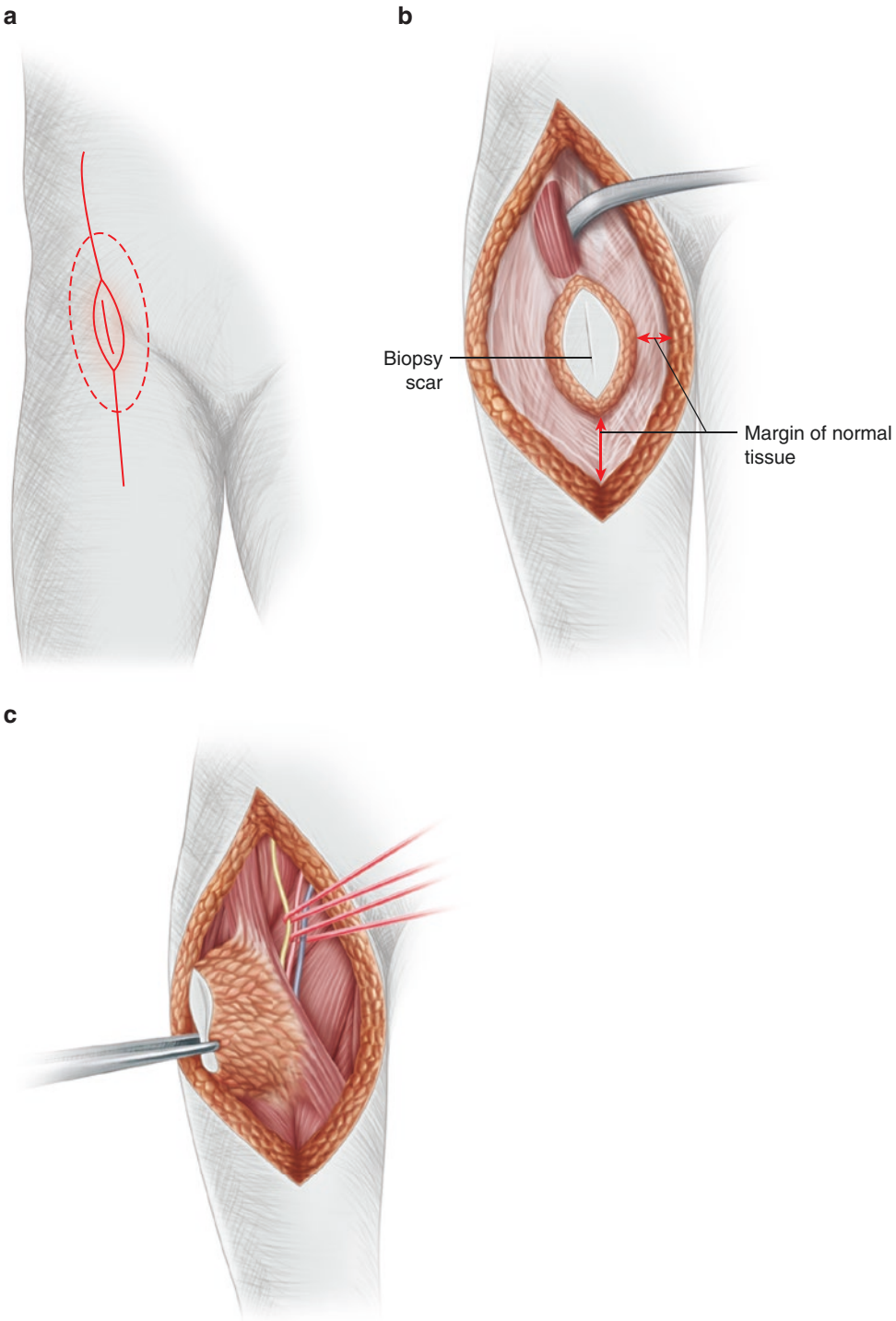

Fig. 56.3 Resection of extremity tumors (beginning the excision). (**a**) Planned excision incorporating previous biopsy scar. (**b**) Circumferential dissection with a margin of normal tissue. (**c**) Deep dissection with a margin of normal tissue

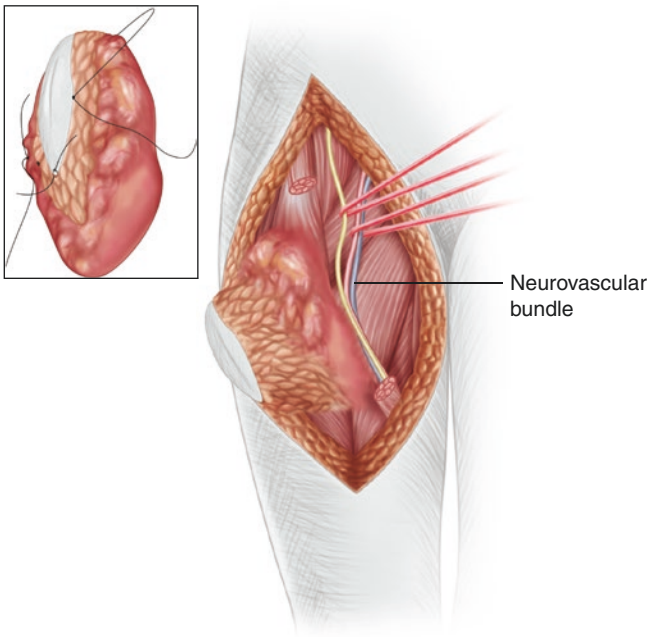

Fig. 56.4 Resection of extremity tumors (neurovascular bundles and marking). Neurovascular structures isolated. *Inset*, Resected specimen orientated with marking sutures

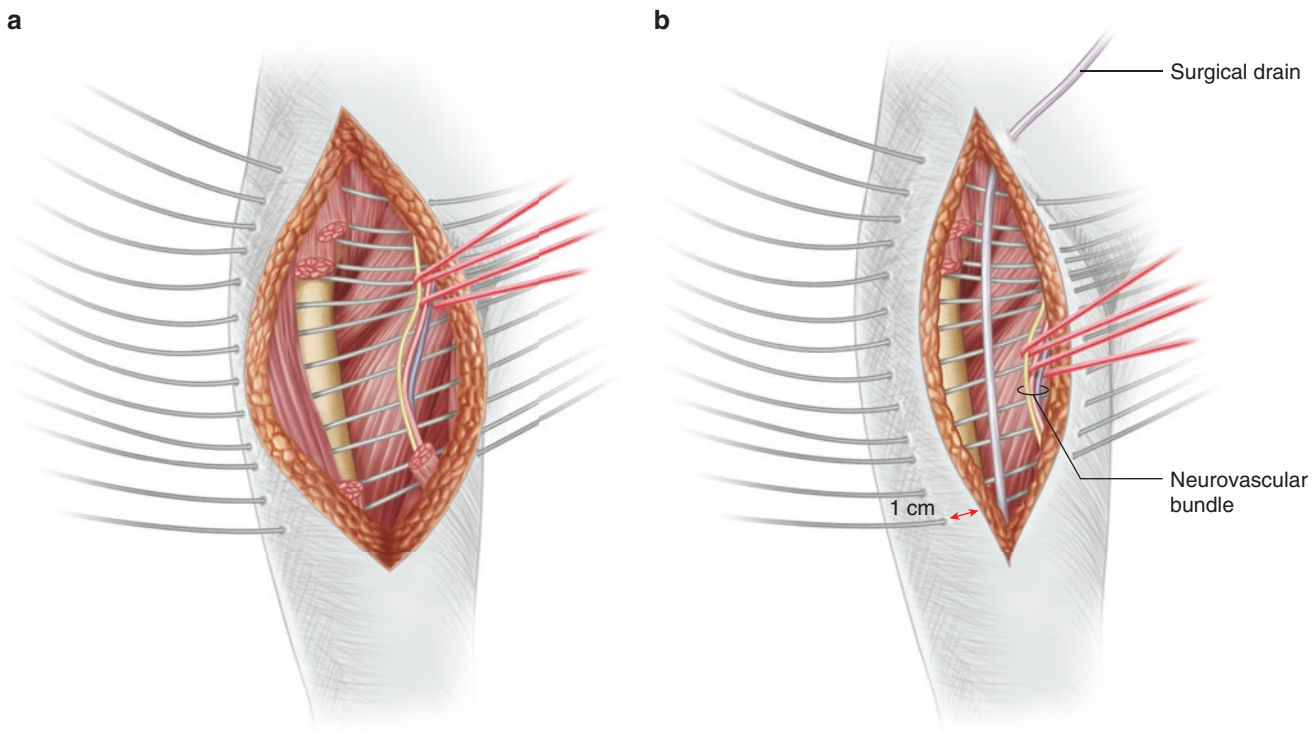

Fig. 56.5 Resection of extremity tumors (closure and brachytherapy). (**a**) Brachytherapy catheters laid against target area. (**b**) Surgical drain placed tumor bed to prevent seroma formation

56.2.2 Resection of Chest Wall Tumors

For truncal lesions of the chest or abdominal wall, complete excision should be the goal of surgical intervention, with prosthetic reconstruction or rotation flaps employed where necessary. Although these tumors may be large on initial presentation, they may become amenable to wide resection with clear margins following preoperative chemotherapy and radiation therapy.

Preoperatively, the craniocaudal and anteroposterior extent of the resection is determined using a recently performed CT or MRI study (Fig. 56.6a). The patient is placed in a lateral position under general anesthesia with indwelling vascular monitoring. A classic posterolateral thoracotomy incision is used, which should provide adequate exposure for resection of most of the lower ribs (Fig. 56.6b). If the upper four ribs are to be included in the resection, the posterior aspect of the incision is curved around the scapula and extended upwards to the neck, the rhomboids and levator scapulae are divided, and the scapula is retracted away from the chest wall (Fig. 56.6c).

At the anterior and posterior limits of the planned resection, the intercostal neurovascular bundles are ligated and divided, and the periosteum around the ribs is stripped using a Doyen periosteal elevator (Fig. 56.7a). The intervening intercostal muscles between the divided ribs and between the anterior and posterior osteotomies are divided (Fig. 56.7b). The muscle fascia of the chest wall and the parietal pleura are left intact on the inner and outer aspects of the tumor. A margin of normal muscle should be retained circumferentially around the tumor.

If the deep aspect of the tumor invades the underlying lung, the dissection should be carried in an extrapleural plane, leaving the parietal pleura attached to the deep component of the tumor and the visceral pleura attached to the involved lung (Fig. 56.7c). Once beyond the limits of the tumor's attachment to the lung, the lung parenchyma is divided using multiple fires of a stapling device, leaving a cuff of normal lung tissue attached to the tumor (Fig. 56.7d). The tumor should thus be resected en bloc with the attached underlying lung tissue.

Limited resections of the chest wall are best reconstructed using a Gore-Tex® soft tissue prosthetic patch. The selected patch should overlap the chest wall defect by approximately 1–2 cm in all directions (Fig. 56.8). Using interrupted nonabsorbable monofilament sutures, the patch is anchored to the ribs above and below, and to the remaining intercostal muscles anteriorly and posteriorly. A second layer of running sutures is placed around the edge of the patch to make an airtight closure. The tension over the patch should be spread evenly, being careful to distribute gathers in the material and to pull the patch taut over the adjacent ribs. Doing so restores the mechanical integrity of the chest wall and avoids a flail segment, which may contribute to paradoxical respiration.

Before placing the final sutures to anchor the prosthetic patch, a chest tube should be inserted through an adjacent rib space, and placed in the appropriate position in the chest cavity. The chest tube should not be inserted under an edge of the prosthetic patch, as it will leave a gap in the prosthetic repair upon its removal. A surgical drain is placed above the prosthetic patch, and the overlying subcutaneous tissue and skin is closed in layers (Fig. 56.9).

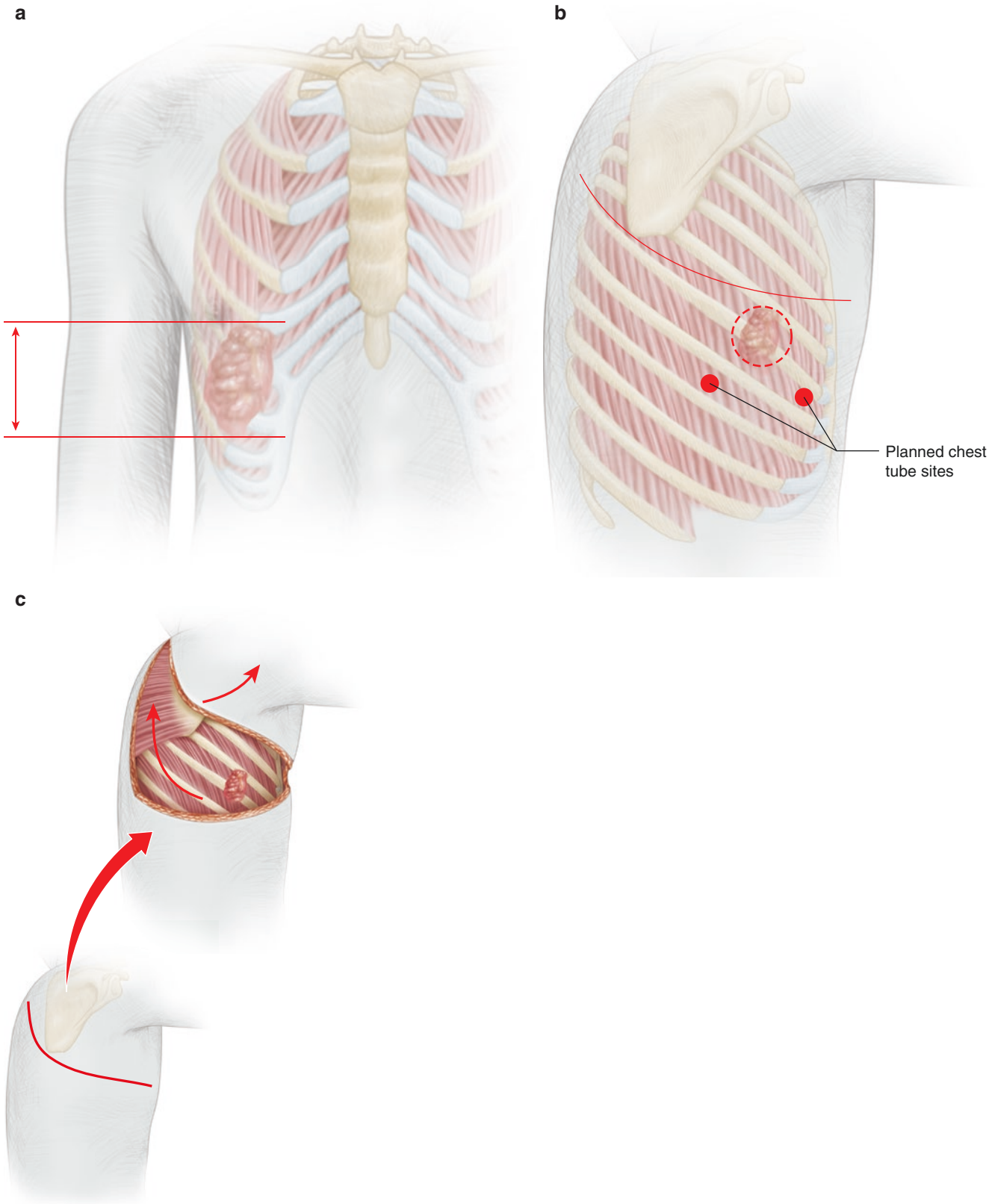

Fig. 56.6 Resection of chest wall tumors (planning, positioning, and incision). (**a**) Preoperative evaluation of craniocaudal extent of mass. (**b**) Thoracotomy incision overlying tumor, and planned chest tube sites. (**c**) Division of rhomboids and levator scapulae for upper thoracic exposure

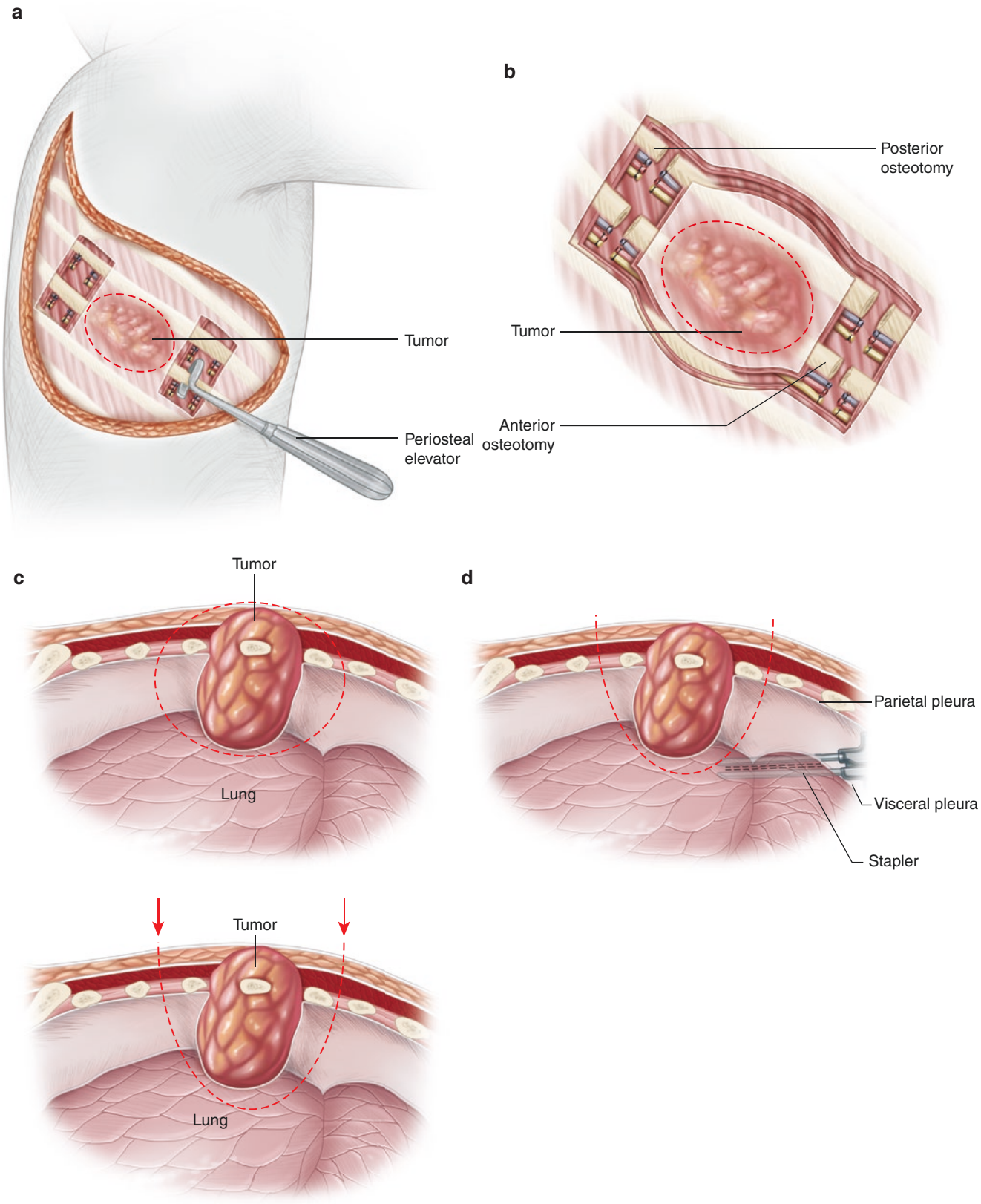

Fig. 56.7 Resection of chest wall tumors (neurovascular bundles and osteotomies). (**a**) Ligation of intercostal neurovascular bundles after stripping periosteum. (**b**) Tumor circumferentially dissected away from surrounding ribs and muscles. (**c**) En-bloc resection of rib tumor with underlying adherent lung. (**d**) Division of underlying lung with stapling device

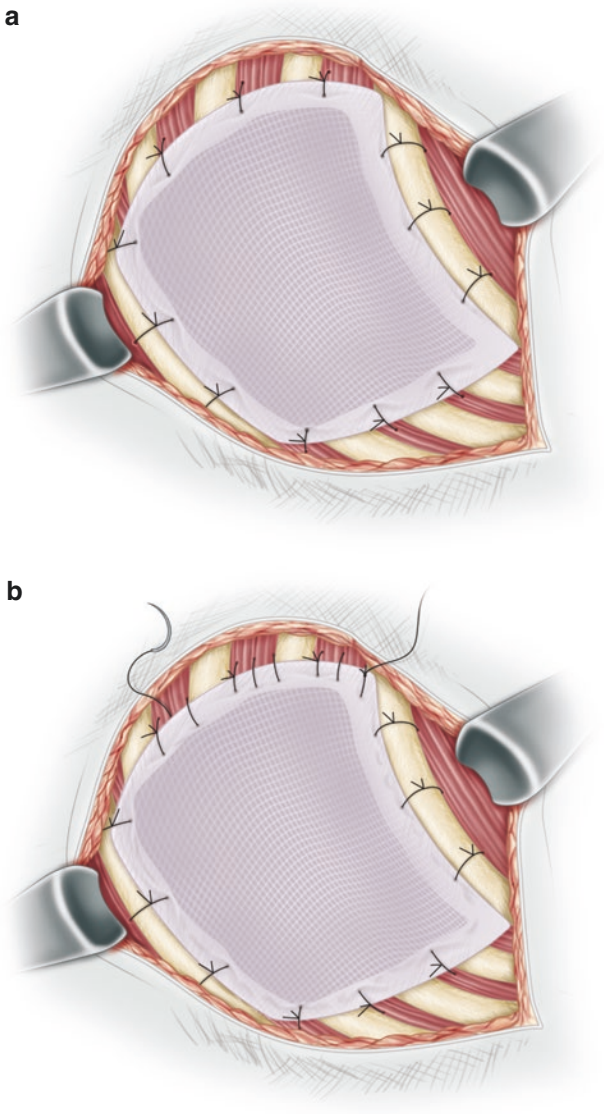

Fig. 56.8 Resection of chest wall tumors (chest wall reconstruction). (**a**) First layer of interrupted mattress sutures to secure patch. (**b**) Second layer of continuous running sutures

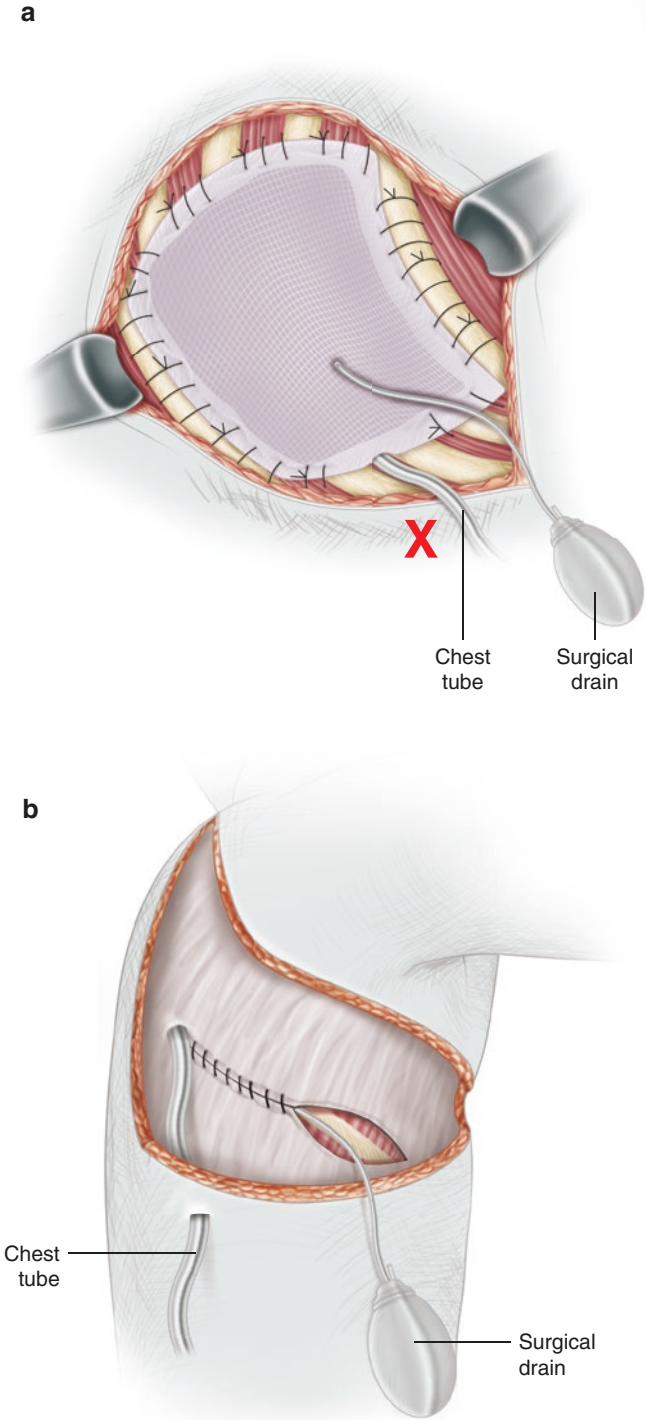

Fig. 56.9 Resection of chest wall tumors (drains and closure). (**a**) Incorrect placement of chest tube under edge of prosthetic patch. (**b**) Closure of wound with surgical drain and chest tube in place

56.3 Follow-Up and Outcomes

In general, primary resection should be performed where technically feasible. In all other instances, adequate biopsy should be followed by neoadjuvant chemotherapy and SLP at week 9–12. If residual disease remains after resection, radiation therapy is added as an adjuvant therapy.

All patients with RMS receive chemotherapy, with regimens usually consisting of a combination of vincristine, actinomycin-D, and cyclophosphamide, tailored according to a risk-based stratification. Radiation therapy is used for local control in patients with microscopic or gross residual disease after biopsy, initial surgical resection, or chemotherapy. Radiation treatment typically follows 1–3 months after chemotherapy and is delivered in fractions over 5–6 weeks. Multimodality treatment protocols have increased survival for nonmetastatic RMS to greater than 70%, and to more than 90% for tumors in genitourinary and orbital sites. However, patients with recurrent or metastatic disease continue to experience survival rates of only 20–30%.

Chemotherapy for NRSTS often consists of regimens incorporating doxorubicin and ifosfamide. Neoadjuvant chemotherapy is the treatment of choice for patients with unresectable disease or when the completeness of initial resection is uncertain, as it may reduce tumor size sufficiently to facilitate subsequent conservative complete resection. The role of adjuvant chemotherapy is uncertain because it has not consistently shown a survival advantage even in children with localized, resectable disease. Radiation is reserved for NRSTS patients with positive surgical resection margins and large (>5 cm) or high-grade tumors. Radiotherapy is important and effective particularly in reducing local recurrences in high-grade tumors with tumor margins less than 1 cm or positive margins of resection. Unfortunately, outcomes for NRSTS have not changed appreciably since the mid-1970s, with 10-year overall survival for children and adolescents stagnant at 50–60%.

Suggested Reading

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Ovarian Tumors

57

Daniel von Allmen and Mary E. Fallat

57.1 Introduction

There is a wide spectrum of ovarian lesions in infants and children; most are benign. Nonneoplastic lesions include functioning cysts. Neoplastic lesions include benign and malignant tumors. Most neoplastic ovarian tumors originate from cell lines that are derived from one of the following lineages:

- Germinal epithelium covering the urogenital ridge
- Underlying stromal elements of the urogenital ridge
- Germ cells that arise from the yolk sac

Cells from each of these lineages may develop into a neoplasm by dedifferentiation, proliferation, and ultimately, malignant transformation. Malignant ovarian tumors likely develop from their benign counterparts as a result of either direct or indirect hormonal stimulation. Epithelial lesions that exhibit histologic and biologic characteristics between benign and malignant forms are designated as tumors with low malignant potential.

The relative frequency of various types of ovarian neoplasms is impacted by age. In adults, most tumors are derived from the epithelial line, and adenocarcinomas predominate. In children, germ cell tumors are most common (60–77% of cases), predominating in all pediatric age groups. Epithelial lesions account for only about 15% of tumors. The peak incidence of sex cord–stromal tumors occurs in the first 4 years of life, whereas epithelial tumors are more commonly seen in older teenagers.

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Management approaches, including surgical approaches, must take into account differences between suspected benign and malignant processes and common pediatric and adult tumor types. In children, tumors typically present at a less advanced stage, and most have a favorable natural history and response to therapy. There are indicators of benign and malignant tumors based on imaging techniques and preoperative biochemical markers. The surgical approach and the need for staging are based on a careful preoperative assessment. Ultimate histologic confirmation supplements the clinical assessment of disease status.

Precise staging of malignancies is based on clinical examination, imaging, surgical exploration, tissue histology, and fluid cytology. The overall goal of surgery for malignant tumors in children is to evaluate the extent of disease, completely resect the tumor, and spare all uninvolved reproductive organs if feasible, as preservation of reproductive potential is a high priority. The uncommon nature of ovarian neoplasms increases the value of evaluation and treatment protocols developed from multi-institutional collaborative studies. This chapter outlines the surgical approach to an ovarian mass, with an emphasis on techniques to spare the ovary when the process is suspected to be benign. Preoperative discussion with the family should always include the potential need for re-operation and a more extensive resection as dictated by the pathology.

The clinical presentation of an ovarian mass is insufficient to distinguish between a benign or malignant tumor. Abdominal pain is the most common symptom, although a number of ovarian lesions are discovered incidentally through imaging performed for other reasons. Abdominal pain can be acute in onset, with a crescendo pattern of severity caused by torsion, rupture, or hemorrhage. A more chronic, insidious pattern of abdominal pain, increasing girth, and marked distention over several weeks to months can also occur. Secondary symptoms include anorexia, nausea, vomiting, constipation, and urinary frequency and urgency due to pressure from the tumor.

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57.1.1 Distinguishing Benign from Malignant Disease Based on Diagnostic Workup

When carrying out the preoperative assessment, obvious malignancy can be predicted by collecting serum tumor markers and performing pelvic ultrasound (US) to determine whether an ovarian mass is complex. Ovarian cysts generally appear as anechoic, thin-walled masses with through transmission. With torsion, fluid debris or septation may be present. Malignant tumors are often larger and appear as complex soft tissue masses with ill-defined, irregular borders and central necrosis, thick septations, or papillary projections. Doppler color-flow imaging and transvaginal US are valuable in postpubertal adolescent patients to determine morphologic characteristics of ovarian lesions. Low color content and/or the presence of normal ovarian tissue adjacent to an ovarian tumor suggests a benign process. Vessels located in central, septal, or papillary projections, together with diffuse vascularity, suggest malignancy.

Preoperative α-fetoprotein (α-fp), β-human chorionic gonadotropin (β-HCG), and cancer antigen 125 (CA-125) are needed for mixed cystic and solid ovarian masses discovered incidentally on imaging studies. Elevated tumor markers and a complex mass on US are indicative of malignancy. CT is helpful in demonstrating tumor extension into adjacent pelvic structures or metastatic disease to liver and lungs. A fluid-filled mass with fat and calcifications is characteristic of benign tumors. Focal solid components arising from the tumor wall are common. Malignant lesions are large and predominantly solid with occasional cystic areas and calcifications. MRI is helpful in distinguishing between ovarian and uterine pathology, as it is not affected by extensive subcutaneous fat and offers excellent soft tissue contrast resolution. MRI enhances characterization of adnexal masses based on benign (fatty components, shading on T2-weighted images) or malignant (vegetations or solid components within cystic masses) criteria.

Positron emission tomography (PET) scanning is a newer modality that may be helpful in distinguishing malignant from borderline ovarian tumors. PET and PET-CT have a role in evaluating patients for recurrent ovarian cancer, particularly those with negative CT or MRI findings and rising tumor marker levels. Fused PET-CT scans are useful in localizing pathologic activity and differentiating this activity from physiologic radiotracer uptake.

If signs of precocius puberty are evident, preoperative testing should include follicle stimulating hormone (FSH), luteinizing hormone (LH), estradiol, and lactate dehydrogenase (LDH) serum levels. An ovary-sparing procedure is the goal if assay results are normal.

57.2 Surgical Guidelines for Ovarian Tumors

Pre-operative preparation includes a type and cross match if the patient has secondary anemia or the tumor is large. Urinary symptoms should be screened with a urinalysis and culture if needed. An enema or more extensive bowel prep may be needed, depending on the planned extent of surgery. Prophylactic antimicrobial therapy, usually with a first-generation cephalosporin, is used to prevent a wound infection; it is not continued postoperatively. Certain types of gynecologic procedures in postpubertal adolescents may benefit from a preliminary bimanual pelvic examination and/or the lithotomy position and insertion of a tenaculum on the uterine fundus to allow manipulation of the uterus during the procedure. Postpubertal or obese patients and other patients at risk should have appropriate intraoperative deep venous thrombosis prophylaxis, such as sequential compression devices. A Foley catheter is generally used to decompress the bladder; it can be removed at the end of the procedure or the next day, depending on the extent of the surgery.

The overall goal of surgery for ovarian tumors in children is to evaluate the extent of disease, completely resect the tumor, and spare all uninvolved reproductive organs. Preservation of reproductive potential is a high priority.

57.2.1 Incisions and Approach

If tumor markers are negative and the mass is likely to be benign, a laparoscopic approach can be considered. Laparoscopic procedures may confer benefits including shorter recovery time and briefer hospital stays. In addition to the umbilical port site, two or three additional port sites may be needed. Upper quadrant port sites will increase the intraperitoneal working distance for instruments, but a lower quadrant port site may also be beneficial to stabilize the tumor. Alternatively, a lower midline or Pfannenstiel incision can be used. The Pfannenstiel incision is a curved incision following the lines of skin cleavage just above the suprapubic skin crease (Fig. 57.1). The incision is carried down to the fascia and an upper supra-fascial flap is developed over the rectus muscle before making a midline fascia incision. Transverse division of the rectus muscles may provide better exposure for larger tumors. For benign unilateral cysts or tumors where the side is known, a small muscle-splitting incision can also be used. For suspected malignancies, a generous midline incision is preferred.

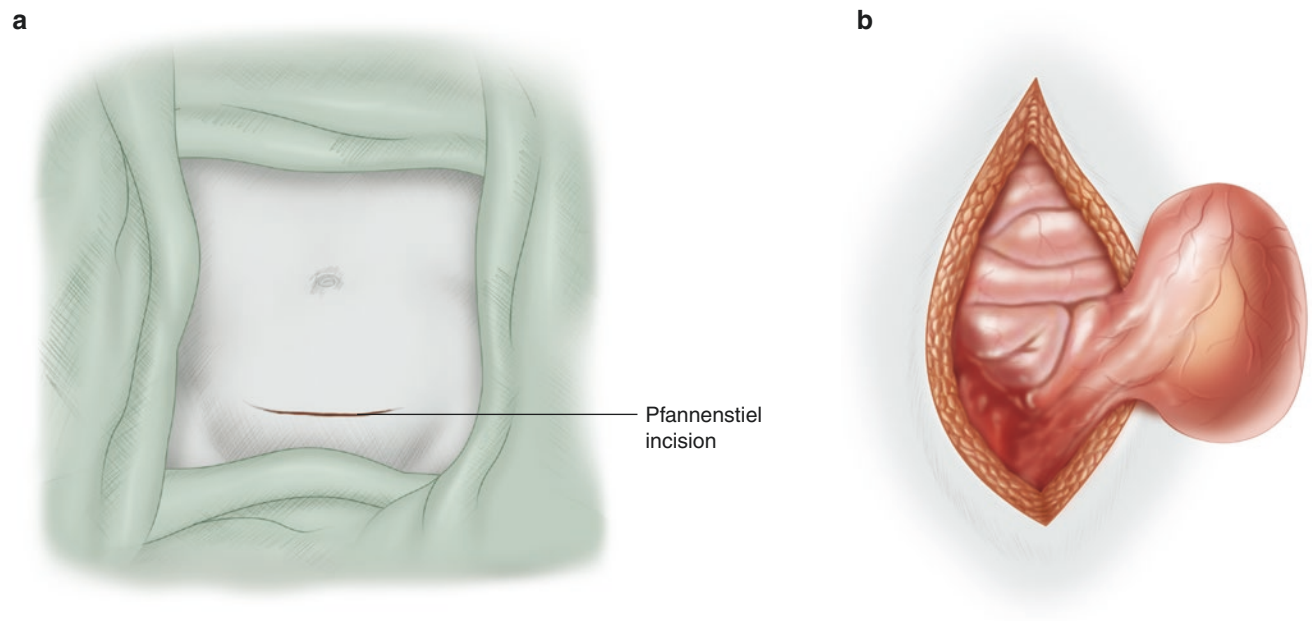

Fig. 57.1 (**a**) Pfannenstiel incision. (**b**) A generous midline incision, preferred for suspected malignancies

57.3 Benign Ovarian Masses

Benign lesions, including benign simple ovarian cyst or benign dermoid tumor, require only tumor resection via ovarian cystectomy. The approach may be open or laparoscopic. For primarily cystic lesions, initial decompression by aspiration may be beneficial to aid in removal of the mass from the peritoneal cavity. For obviously benign cystic masses, a laparoscopic needle decompression can be done through a port site, or the cyst can be manipulated up to the abdominal wall and an angiocatheter or needle can be introduced under direct vision through the abdominal wall and hooked to suction for decompression. The fluid can be sent for cytology in the suction canister. Using an open or laparoscopy-assisted approach, once the cyst is accessible through an incision, an angiocatheter can be placed directly into the cyst through a pursestring suture in the ovarian capsule. A rubber shod can be used to cinch down the suture and minimize leaking (Fig. 57.2). If there is concern about any spillage at all, a plastic bag can be mounted on the cyst first with cyanoacrylate adhesive (Figs. 57.3 and 57.4).

There are several ways to identify the plane between the ovary and the cyst. In the event of torsion, the adnexa must first be detorsed (Fig. 57.5).

The first step in doing an ovary-sparing cystectomy or mass excision is to recognize where the predominant ovarian tissue is located and make a small incision above this area in the ovarian capsule. This can be done with the harmonic scalpel if the procedure is done laparoscopically (Fig. 57.6). A small amount of sterile saline can be injected just beneath the capsule to enhance initial separation of the layers. The incision can be made with the cautery, hot grasper, harmonic scalpel, or a regular scalpel, and the plane developed with the aid of a curved hemostat, moist cotton-tipped swab, or finger dissection if the capsule is thick enough (Fig. 57.7). It is very important to just remove the cyst and preserve as much of the ovarian capsule as possible for future reproductive potential. It is usually possible to remove most of the cyst without removing the ovary unless the ovarian capsule is too adherent. Remember that the cyst is underneath the capsule because the cyst develops in the ovary (Fig. 57.6).

When doing a laparoscopic case, a suction irrigator for aqua dissection will facilitate development of the plane between the ovarian wall and the tumor or cyst while graspers placed on these structures provide traction and countertraction. Then the tumor is placed in an endobag (Fig. 57.8) and removed from the peritoneal cavity by enlarging one of the port sites.

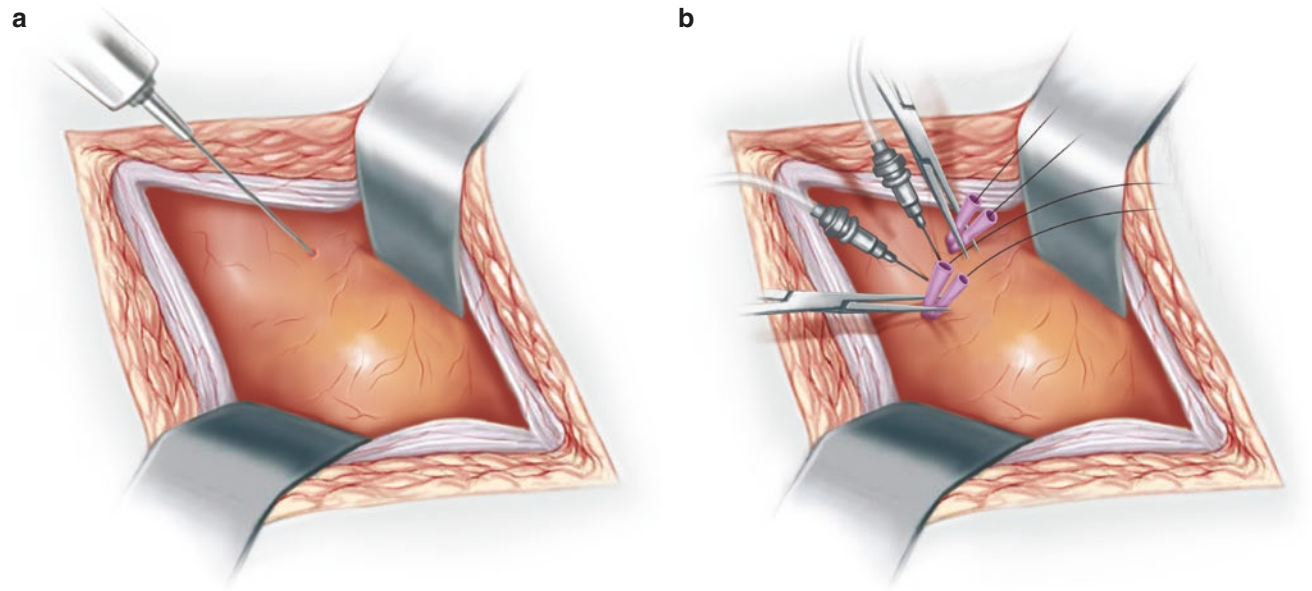

Fig. 57.2 (**a** and **b**) Simple aspiration of a benign cyst with an angiocatheter through a pursestring suture in the ovarian capsule

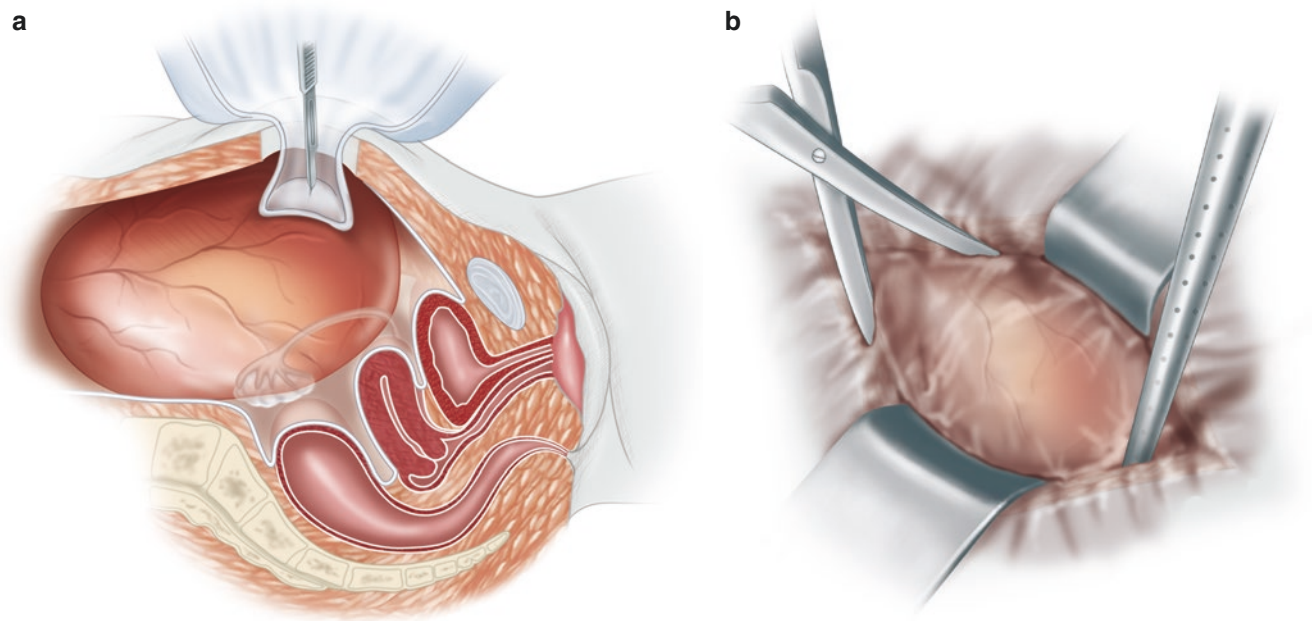

Fig. 57.3 (**a** and **b**) Mounting a plastic bag using cyanoacrylate adhesive for leak-proof puncture of an ovarian cyst

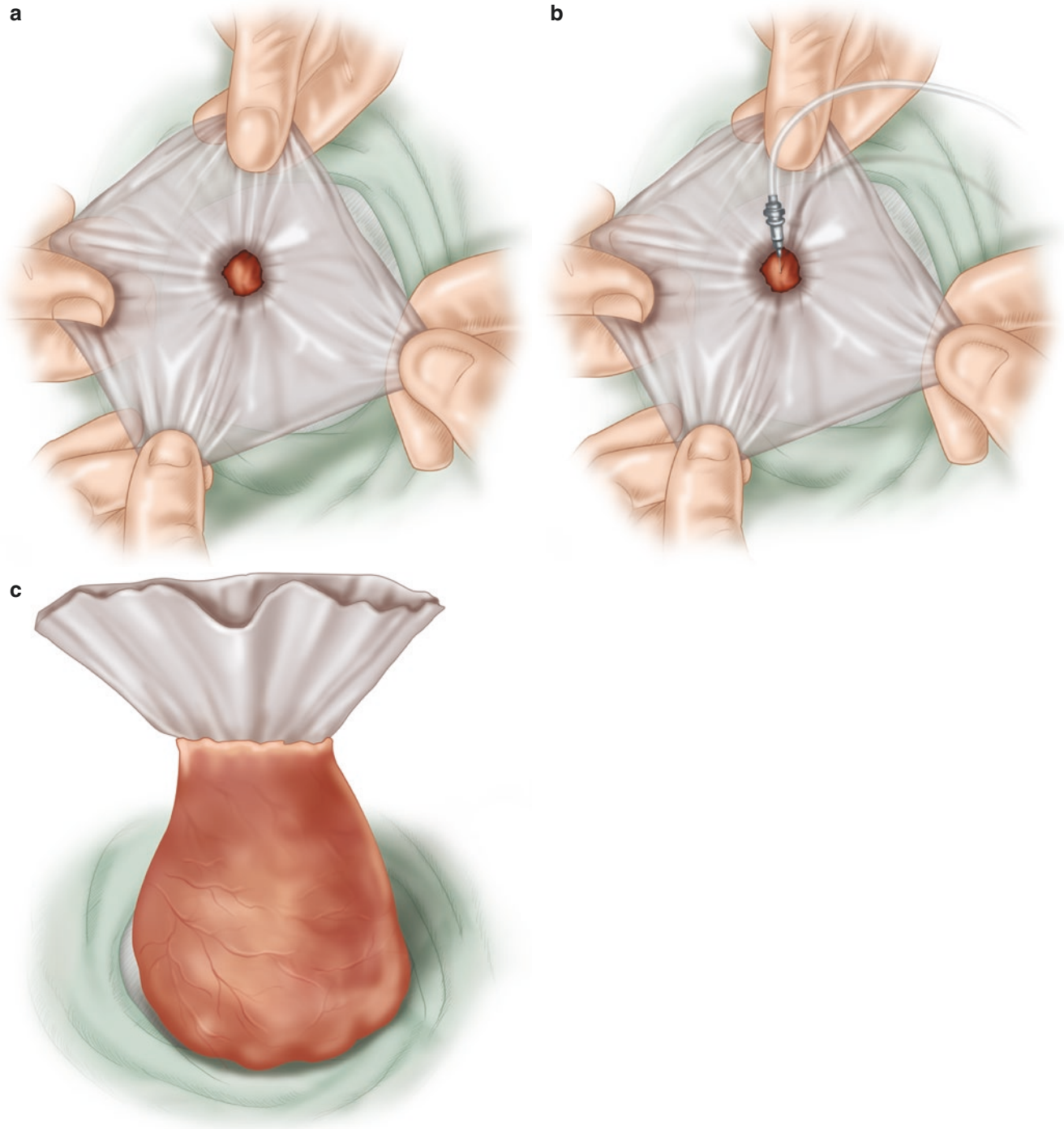

Fig. 57.4 (**a–c**) Needle aspiration through the mounted bag and delivery of the mass outside the abdomen

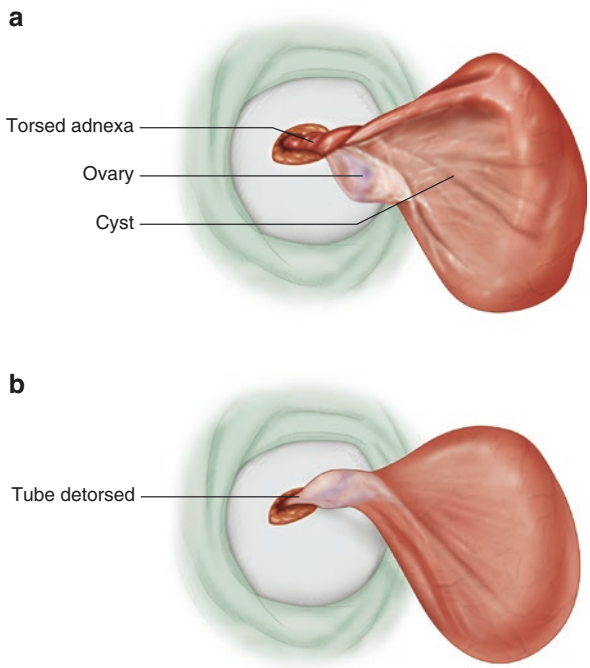

Fig. 57.5 (**a** and **b**) Detorsion of the ovary and tube before cyst removal

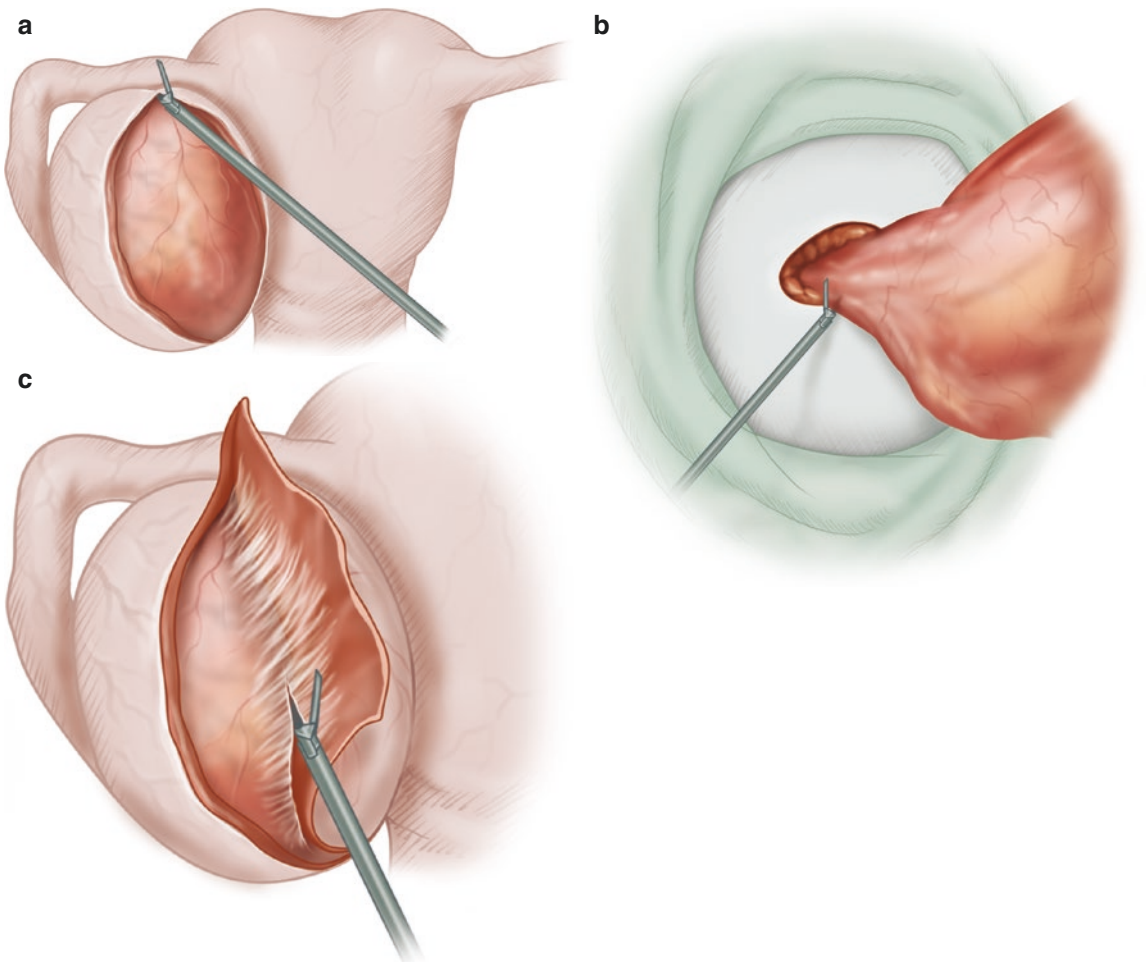

Fig. 57.6 (**a** and **b**) Opening the capsule of the ovary with a harmonic scalpel to get to the underlying cyst. (**c**) The raw surface of the inside of the ovary after the cyst has been removed

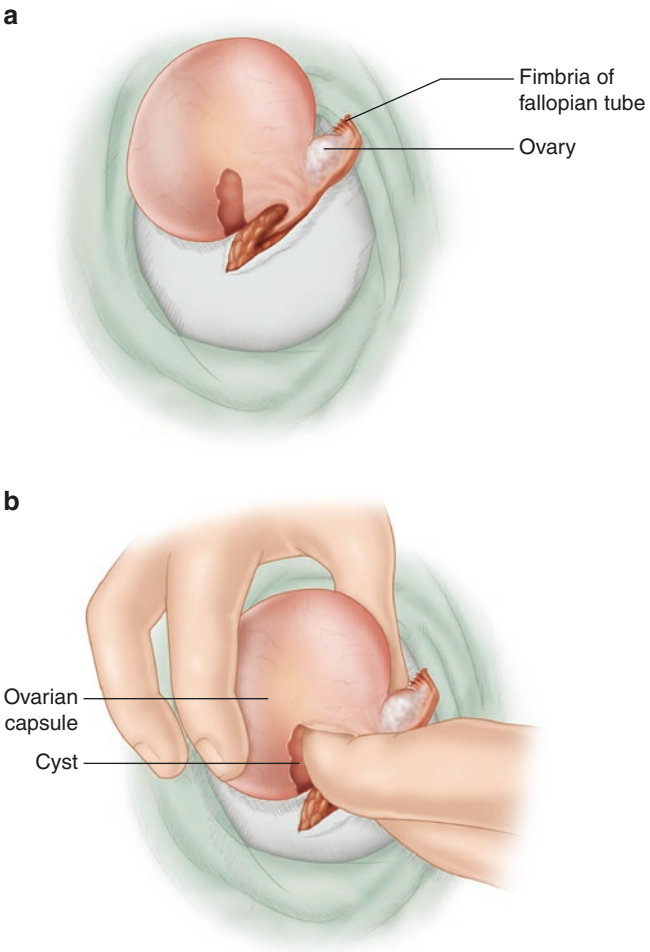

Fig. 57.7 (**a** and **b**) Peeling the capsule off the cyst and preserving the ovary

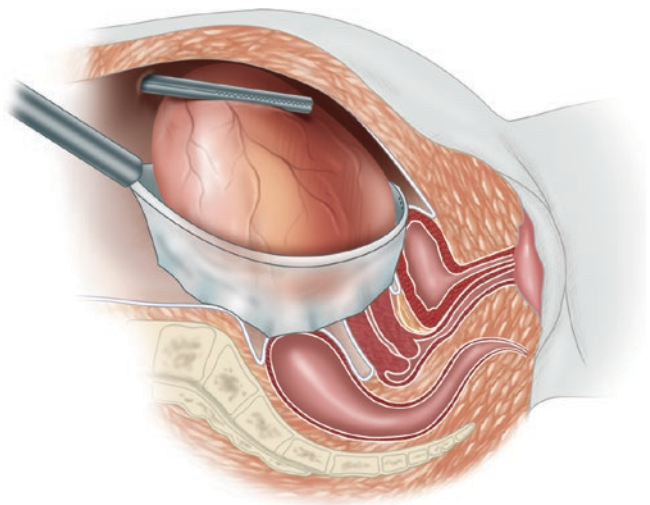

Fig. 57.8 Placing the tumor in an endobag

57.4 Suspicious or Confirmed Ovarian Malignancy

If a suspected ovarian malignancy is detected at the time of laparoscopy, complete surgical staging and resection by conventional laparotomy is recommended. For potentially malignant lesions, an adequate abdominal incision is needed to avoid violation of the tumor capsule. Initial resection should favor a fertility-sparing approach, with anticipatory guidance for the patient and family that a second procedure might be needed after the final pathology is determined. Although uniform surgical guidelines exist, the overall approach to ovarian neoplasms has gradually become more conservative. Staging procedures for malignancies differ for different cell types, which can result in inadequate staging of unsuspected epithelial tumors. Benign tumors, frankly malignant tumors, and those with mixed histologic characteristics often cannot be distinguished based on gross appearance. If in doubt, complete staging is recommended, as required for treatment and prognosis of malignancies.

57.5 Germ Cell Tumors

In the United States, pediatric germ cell tumors are staged according to the system established by the Children's Oncology Group (COG) in 2010. As detailed below, staging guidelines for germ cell tumors proposed by COG include peritoneal fluid aspiration/washings, inspection of the omentum and contralateral ovary, biopsy of suspicious peritoneal and liver nodules (including the subphrenic spaces), biopsy of clinically suspicious lymph nodes, and removal of the primary tumor. Both ovaries are inspected. If an ovarian tumor is found and malignancy suspected, it should be removed by unilateral oophorectomy if the fallopian tube is not involved, or by salpingo-oophorectomy for tube involvement. The contralateral ovary should be inspected and nodules or suspicious areas should be biopsied. Contralateral salpingo-oophorectomy should be avoided in the absence of confirmed malignancy.

Sample/aspirate peritoneal fluid for cytology. If there is fluid in the peritoneal cavity, it can simply be aspirated into a sputum trap or other sterile container directly from the peritoneal cavity. If there is little or no fluid, introduce normal saline into the peritoneal cavity and roll the patient from side to side and in Trendelenburg and reverse Trendelenburg and collect the fluid for cytology.

Resect the tumor. Carefully divide any adhesions between the ovarian mass and the intestine or viscera, to avoid unintentional injury. As illustrated in Fig. 57.9, the infundibulopelvic ligament is divided, either between clamps or with the harmonic scalpel. The ovarian vessels are suture ligated with a large-gauge absorbable suture. The broad ligament is divided. The interstitial portion of the fallopian tube is removed by making an elliptical incision through the thickness of the uterine wall. The corneal artery needs to be controlled with either compression or a suture ligature before separating this area. Mattress sutures are traditional to control bleeding along the ligaments, although the harmonic scalpel will minimize the need for sutures, especially in young patients. Sutures should be tied without tension to avoid crushing the uterine wall. The mesosalpinx is clamped along its entire length. If the mesosalpinx is going to be saved because only an oophorectomy is needed, mattress sutures can be used to avoid the blood supply by staying close to the artery, or the harmonic scalpel can be used right next to the ovary. *Inspect the contralateral ovary*. Wedge biopsy suspicious areas.

Inspect the omentum. An omentectomy (Fig. 57.10) is approached initially by locating the transverse colon and distracting the omentum directly upward. Beginning at the right side adjacent to the posterior taenia coli, a thin and relatively avascular peritoneal layer can be identified and divided. A few small blood vessels may be seen toward the anterior taenia and can be divided using a traditional division of vessels between ties, the cautery, the ENSEAL, the LigaSure, or the harmonic scalpel. The left sided dissection is carried to just below the spleen. Care is taken to avoid injury to the middle colic vessels and the splenic capsule.

Biopsy peritoneal surface lesions. These lesions can be simply enucleated with precise identification of anatomic location for the pathologist.

Biopsy retroperitoneal lymph nodes if suspicious. In conjunction with evaluation of preoperative radiographic imaging, the retroperitoneum is examined and palpated for palpable lymph nodes; these are biopsied with precise identification of location for pathology.

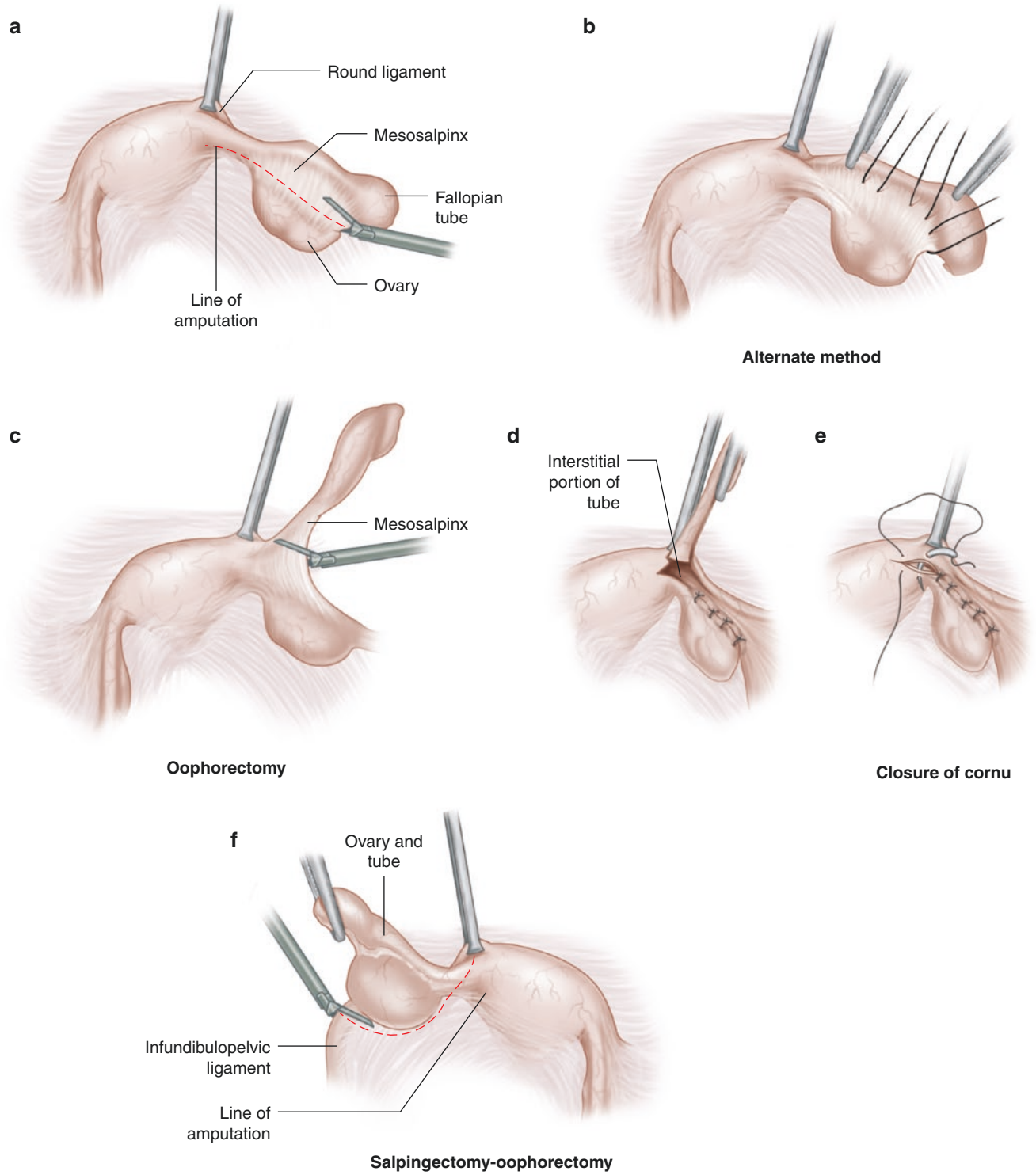

Fig. 57.9 Oophorectomy and salpingo-oophorectomy (**a**–**f**)

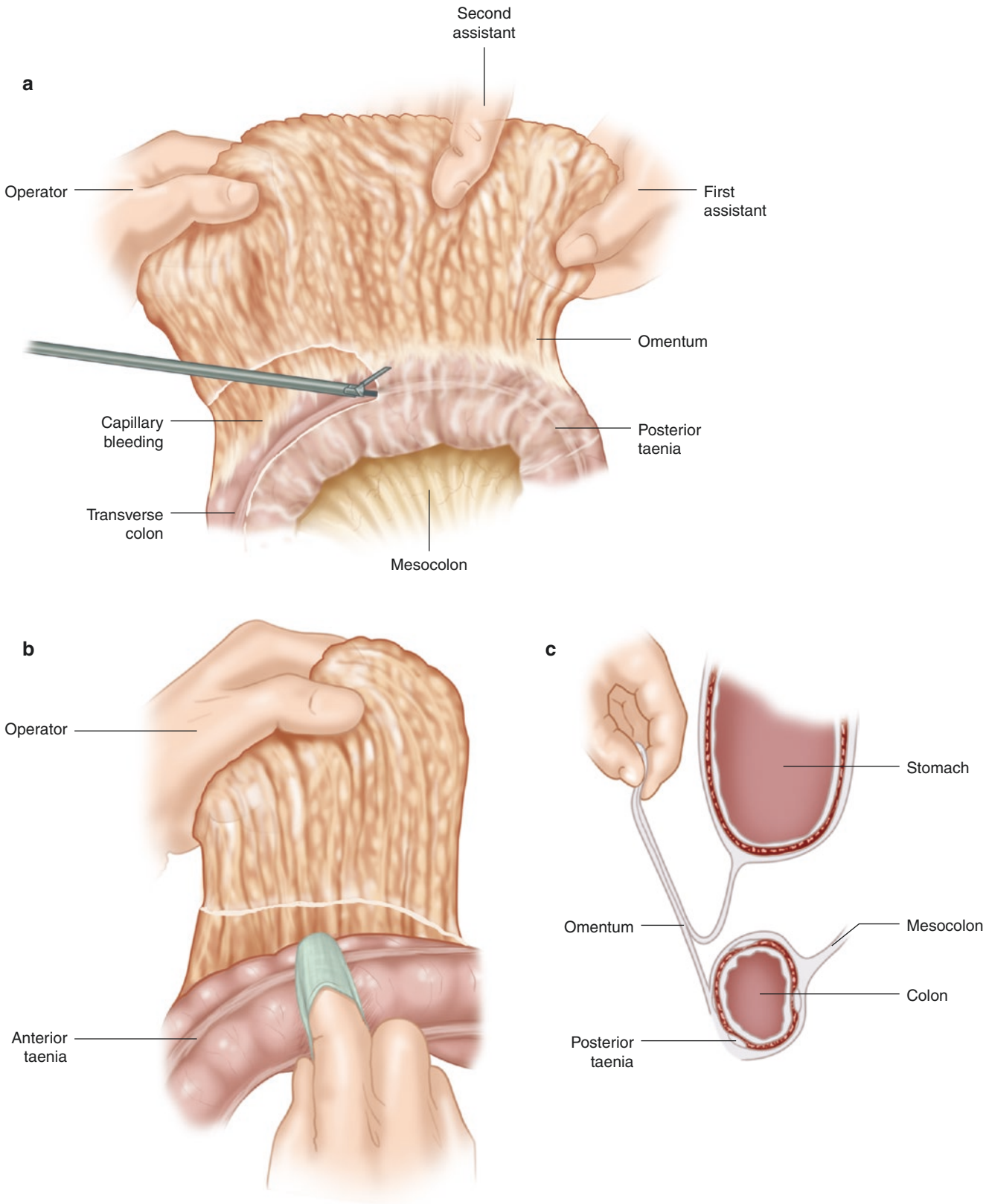

Fig. 57.10 (**a**–**c**) Omentectomy

57.6 Epithelial Cell Tumors

Epithelial cell tumors are usually serous or mucinous. Of serous tumors, 20% are bilateral but few are malignant, contrasted with a malignancy rate of 10% for mucinous tumors, which are generally unilateral. Epithelial tumors are staged using the adult system**,** which requires peritoneal biopsies, peritoneal washings/aspiration, omentectomy, removal of the primary tumor, and an ipsilateral lymph node dissection. Up to 30% of clinically normal lymph nodes can be positive for metastatic disease, so this part of the procedure is not optional. In advanced stages, maximum cytoreduction is associated with improved outcomes. Total abdominal hysterectomy and bilateral salpingo-oophorectomy with omentectomy and resection of as much gross intraperitoneal disease as possible may be needed and is best done with a gynecologic oncologist.

57.7 Tumors with Low Malignant Potential

Initial management of tumors with low malignant potential should include pelvic washings, unilateral ovarian cystectomy, intraoperative frozen section, and meticulous inspection of the contralateral ovary. Pelvic washings should be performed immediately on entry into the abdomen (via either laparoscopy or laparotomy), to minimize possible contamination in the event of intraoperative tumor rupture. If there is no free fluid in the abdomen, lactated Ringer's solution can be used to irrigate the pelvis and paracolic gutters, aspirated, and sent as washings.

57.8 Postoperative Care

Postoperative care of a patient who had removal of a benign or malignant ovarian mass involves early mobilization while controlling pain, removal of the Foley catheter as soon as feasible, and a bowel regimen to prevent constipation. Antimicrobial therapy is not indicated after surgery if no established infection is identified.

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Laparoscopic and Robotic Urology

58

Andrew C. Strine and Paul H. Noh

In the field of pediatric urology, an interest in laparoscopic surgery and robot-assisted laparoscopic surgery (RALS) has been growing since the initial description of diagnostic laparoscopy for the evaluation of nonpalpable testes in 1976. These minimally invasive techniques have been adopted more slowly in children than in adults, but recent improvements in instrumentation and the widespread acceptance of RALS have led to rapid expansion of their application in pediatric urology.

Pediatric urologists have incorporated a wide variety of minimally invasive techniques into their armamentarium. Diagnostic laparoscopy has become the standard of care for the evaluation of nonpalpable testes. Laparoscopic nephrectomy, heminephrectomy, orchidopexy, and varicocelectomy have also proven to be safe and effective in children. Furthermore, RALS has improved the availability of minimally invasive techniques to more pediatric urologists and has facilitated their application to more complex surgery, owing to the technical difficulty and steep learning curve with laparoscopic surgery. RALS allows for greater control and precision than laparoscopic surgery; it offers magnified three-dimensional vision, instruments with seven degrees of freedom and 90° of articulation, motion scaling, reduction of tremor, and improved ergonomics. These features seem to be particularly conducive to reconstructive surgery in pediatric urology, which demands delicate, precise movements in a limited working space. The most significant contribution of RALS has been observed in pyeloplasty, for which an increasing number of pediatric urologists are adopting the RALS approach because of its improved technical ease with intracorporeal suturing. Most recently, RALS has been explored for continent reconstruction (augmentation cystoplasty, Mitrofanoff appendicovesicostomy, anterograde continence enema, and/or bladder neck reconstruction) with encouraging but preliminary results.

The successful application of laparoscopic surgery and RALS to children requires a basic understanding of unique technical considerations and the physiologic effects of pneumoperitoneum.

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58.1 Physiologic Effects of Pneumoperitoneum

The physiologic effects of laparoscopic surgery and RALS are primarily related to pneumoperitoneum. Children generally experience a more pronounced physiologic effect than adults because of the higher compliance of their chest wall, diaphragmatic respiration, and rate-dependent cardiac output.

The ideal gas for pneumoperitoneum should be inexpensive, have minimal physiologic effects, be noncombustible, have minimal peritoneal absorption, be rapidly excreted if absorbed, be highly soluble in blood, and present minimal effects from intravascular embolization. Although an ideal gas does not exist, carbon dioxide (CO_2) is most commonly used for pneumoperitoneum because of its favorable characteristics. Its advantages include its low expense, wide availability, noncombustible nature, high solubility in blood, and rapid excretion by the lungs. It is absorbed across the peritoneum, however, and may cause hypercarbia. Hypercarbia is exacerbated by Trendelenburg position, elevated intra-abdominal pressure (IAP) during insufflation, and prolonged operative times. Certain children with pulmonary disease may also have an increased risk of hypercarbia and metabolic acidosis owing to an inadequate compensatory response.

The central nervous system, pulmonary, cardiovascular, and renal systems are primarily affected by an elevated IAP during insufflation (Table 58.1). An increased intracranial pressure may occur because of the transmission of an elevated IAP via the central venous system and cerebrospinal fluid, although the central nervous system is typically able to maintain its cerebral perfusion. Pneumoperitoneum may also lead to a change in ventilation. An elevated IAP specifically decreases the diaphragmatic excursion, causing a decreased functional residual capacity and compliance as well as an increased peak inspiratory pressure. These pulmonary changes ultimately result in a ventilation-perfusion mismatch and shunting, which may lead to hypoxemia. In the cardiovascular system, the compressive effect of pneumoperitoneum on the inferior vena cava causes decreased venous return followed by an increased peripheral vascular resistance to maintain the cardiac output. The sympathetic nervous system is also stimulated by CO_2, resulting in tachycardia and increased cardiac contractility as well as vasoconstriction via the renin-angiotensin-aldosterone system. These hemodynamic changes may ultimately lead to no change or a mild decrease in cardiac output as well as an increased risk of arrhythmia. Lastly, an increased IAP causes decreased renal blood flow and glomerular filtration rate, both of which result in decreased urine output or even anuria. All of these physiologic effects of pneumoperitoneum are easily reversible by decreasing the IAP.

The generally recommended pressures for insufflation are 15 mm Hg in adolescents (>10 years), 10–12 mm Hg in children (2–10 years), and 8–10 mm Hg in infants (0–2 years); these levels produce minimal physiologic effects. Children typically have a smaller and more compliant anterior abdominal wall than adults, limiting the working space during insufflation. Their peritoneal cavity does not accommodate more than 1–2 L of pneumoperitoneum, compared with 3–5 L in adults. Because of the higher compliance of their anterior abdominal wall, increasing the IAP above these recommended levels does not increase the working space.

Table 58.1 Physiologic effects of Pneumoperitoneum

System	Effect
Central nervous system	
Intracranial pressure	↑
Pulmonary system	
Peak inspiratory pressure	↑
Compliance	↓
Vital capacity	↓
Functional residual capacity	↓
Dead space	←→
Cardiovascular system	
Heart rate	←→ or ↑
Mean arterial pressure	↑
Central venous pressure	↑
Cardiac preload	↓
Systemic vascular resistance	↑
Cardiac output	←→ or ↓
Renal system	
Renal blood flow	↓
Glomerular filtration rate	↓
Urine output	↓

58.2 Surgical Approaches

Various surgical approaches are available to pediatric urologists. The best surgical approach in a particular circumstance is determined by multiple factors, including the experience of the surgeon, the size of the child, the past medical and surgical history, and the location of the targeted organ.

Most pediatric urologists prefer a transperitoneal laparoscopic approach because of the larger working space and familiar anatomic relationships, but a retroperitoneoscopic approach may be preferable in certain circumstances. It is advisable not to violate the peritoneal cavity of children on peritoneal dialysis, to prevent the need for temporary hemodialysis. Similarly, a retroperitoneoscopic approach may be advantageous in children with prior intra-abdominal surgery to avoid any adhesions and the associated risk of bowel injury and open conversion. Other advantages of a retroperitoneoscopic approach include a decreased risk of ileus/bowel obstruction and trocar site–related complications, such as fistula and hernia. Nevertheless, most pediatric urologists have been slow to adopt the retroperitoneoscopic approach because of a lack of familiarity with this approach to the kidney.

The decision between a traditional laparoscopic approach or RAL approach is determined by similar factors in addition to the technical difficulty of the planned surgery. A laparoscopic or retroperitoneoscopic approach is preferred for extirpative surgery (e.g., nephrectomy and heminephrectomy), whereas the RAL approach is increasingly performed for reconstructive surgery such as pyeloplasty, ureteral reimplantation, and complex reconstruction. More recently trained pediatric urologists are also more likely to favor a RAL approach, owing to their greater exposure in residency and fellowship training.

Laparoendoscopic single-site surgery (LESS) is a novel application of laparoscopic surgery that has recently been explored in pediatric urology. LESS is based on the co-axial and fulcrum concepts, whereby the laparoscope and instruments pivot around a single port. LESS offers the same benefits as other minimally invasive techniques in addition to an improved cosmesis because the single port can be concealed around the umbilicus (Fig. 58.1). LESS is more ergonomically challenging, however, owing to the reversal of handedness, limited range of motion, collisions, and fixed trocar site with the laparoscopic instruments pivoting around a single port and crossing in the working space. LESS is typically reserved for extirpative surgery by more advanced laparoscopic surgeons. The future development of a robotic platform for LESS may improve the availability of this minimally invasive technique to more pediatric urologists.

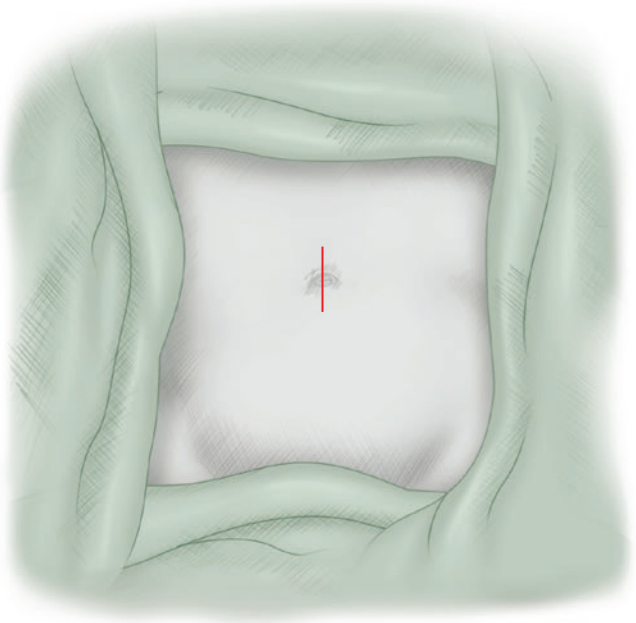

Fig. 58.1 LESS offers the same benefits as other minimally invasive techniques in addition to an improved cosmesis due to an ability to conceal a single port around the umbilicus

58.3 Contraindications

The only contraindications to laparoscopic surgery and RALS are analogous to those for open surgery, including an uncorrected coagulopathy and severe, uncompensated cardiopulmonary disease.

58.4 Positioning

Positioning is a critical step to prevent any iatrogenic injuries and maximize the exposure of the targeted organ, particularly in children, in whom working space is limited. The type of positioning depends on the size of the child, the location of the targeted organ, and the planned surgical approach.

58.4.1 Surgery of the Upper Urinary Tract

Children are generally placed in a lateral decubitus position for a transperitoneal laparoscopic and retroperitoneoscopic approach (Fig. 58.2). All lines and monitors are placed after the induction of anesthesia and carefully padded to prevent any pressure-induced necrosis. A nasogastric or orogastric tube is placed to decompress the stomach, and a Foley catheter to decompress the bladder. The bladder is not a pelvic organ in infants and young children and should be decompressed to prevent its injury and to maximize the working space. The child is transferred into a modified or full lateral decubitus position with the affected side elevated. A full lateral position is preferred for a retroperitoneoscopic approach. The break in the operating table is positioned at the level of the iliac crest and later is flexed to maximize the exposure of the kidney and upper ureter. The ventral or dorsal surface of the child is also positioned on the edge of the operating table to prevent any collision with the instruments for a transperitoneal laparoscopic or retroperitoneoscopic approach, respectively. The head and neck are placed in a neutral position. The upper arm may be tucked alongside the child or placed across the chest with a slightly abducted shoulder, flexed elbow, and pronated hand. The lower leg is flexed at the hip and knee, while the upper leg remains extended. The upper leg is also supported with pillows in a neutral position and parallel to the operating table. Careful padding of all pressure points is important to prevent any neuromuscular injuries, with particular attention to the elbows, hips, knees, and ankles. An axillary roll is also placed in the lower axilla to prevent injury to the brachial plexus. The child is secured to the operating table with padded tape or straps to prevent any movement during rotation of the operating table.

Alternatively, children may be placed in a prone position for a retroperitoneoscopic approach. Two large rolls are placed under the chest and hips of the child, preventing the compression of the abdomen against the operating table. This maneuver allows the intra-abdominal contents to separate from the retroperitoneum and prevents the compression of the retroperitoneum if the peritoneal cavity is inadvertently entered and insufflated. Similar guidelines are otherwise followed for positioning.

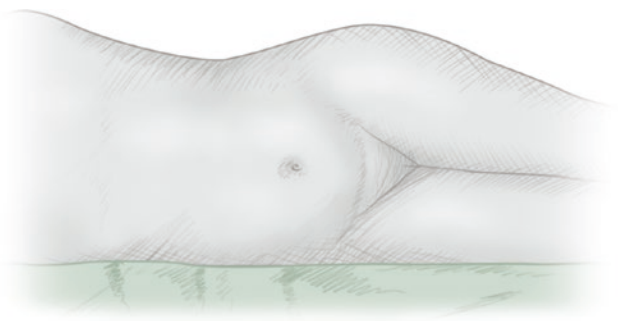

Fig. 58.2 Children are generally placed in a lateral decubitus position for a transperitoneal laparoscopic and retroperitoneoscopic approach

58.4.2 Pelvic Surgery

Children are typically placed in a supine or dorsal lithotomy position for pelvic surgery. For the dorsal lithotomy position, the arms are tucked alongside the child with pronated hands. The legs are also placed in stirrups with the hips, knees, and ankles aligned with the contralateral shoulder. The weight of the legs should reside on the heel rather than on the popliteal fossa (to prevent injury to the popliteal artery) or the lateral surface of the legs (to prevent injury to the common peroneal nerve). The child may be placed in a Trendelenburg position to shift the intra-abdominal contents from the pelvis. Compared with the supine position, the Trendelenburg position may alter the physiologic effects of pneumoperitoneum, including increased intracranial pressure, decreased heart rate, increased mean arterial pressure, increased systemic vascular resistance, and increased cardiac output. Similar guidelines are otherwise followed for positioning.

58.5 Access

There are several unique technical considerations for safely obtaining and maintaining access in children. Children generally have a more compliant anterior abdominal wall, thinner fascia, and a smaller and shallower peritoneal cavity than adults. Their anterior abdominal wall therefore provides less resistance for obtaining access, with close proximity of the underlying visceral organs and vasculature. It is critical to insert the Veress needle and/or trocars with greater care and less force, to prevent any access-related injuries. Transiently increasing the IAP may increase the tension of the child's anterior abdominal wall and facilitate the insertion of trocars.

Maintaining access is also more challenging owing to the dislodgement of trocars during the exchange of instruments and other minor episodes of desufflation, leading to a loss of pneumoperitoneum or preperitoneal insufflation. A Hasson or balloon trocar may be used to prevent its dislodgement. Our preference is to make an incision in the skin and fascia that is only large enough to accommodate a traditional laparoscopic or robotic trocar, which should hold it in place.

58.5.1 Primary Access

Primary access may be obtained using one of three techniques: closed technique with Veress needle, open Hasson technique, and direct trocar insertion. Primary access is typically obtained around or through the umbilicus for a transperitoneal laparoscopic approach. The selection of another site for primary access using an open Hasson technique or even using a retroperitoneoscopic approach should be considered in the setting of prior intra-abdominal surgery to avoid any adhesions.

58.5.1.1 Closed Technique with Veress Needle

The Veress needle has a spring-loaded inner sheath that retracts while being advanced through the anterior abdominal wall and springs forward once the resistance is released from entering the peritoneal cavity. The inner sheath covers the sharp tip of the needle and prevents any injuries to the underlying visceral organs and vasculature. A series of steps are followed to ensure the proper insertion of the Veress needle into the peritoneal cavity. An incision is made at the site for primary access with a scalpel and carried down to the fascia with gentle spreading or electrocautery. For primary access through the umbilicus, it is particularly helpful to evert the umbilicus with sutures or towel clamps on each side. The fascia is secured with sutures or instruments (e.g., Kocher or Allis clamps) for superior retraction. The Veress needle is engaged at a 30° angle toward the pelvis and advanced with proximal control to avoid deep insertion, particularly in children, who have a smaller and shallower peritoneal cavity. Two "clicks" are felt and heard as the Veress needle is advanced through the fascia and parietal peritoneum. The Veress needle is aspirated to confirm its location in the peritoneal cavity. The Veress needle should not be removed if it does not aspirate or if bowel contents, blood, or fluid are aspirated. An alternative site is subsequently obtained for access via a closed technique with a Veress needle or an open Hasson technique, after which the original site is inspected to determine the nature and extent of the injury. If the Veress needle aspirates easily, it is irrigated with 1–2 mL of normal saline and again is aspirated for confirmation. The Veress needle is gently advanced 0.5–1 cm, during which no resistance should be felt. A "drop test" is performed by removing the syringe and observing the remaining normal saline in the Veress needle. A rapid descent of this fluid confirms its location in the peritoneal cavity. Once the insufflation tubing is cleared, the Veress needle is attached to the insufflator. The opening IAP should be less than 10 mm Hg. If the opening IAP is greater than 10 mm Hg, the Veress needle is gently withdrawn to separate its tip from any intra-abdominal contents, such as bowel or omentum. The Veress needle is otherwise removed and replaced if the opening IAP remains greater than 10 mm Hg. Once the opening IAP is less than 10 mm Hg, the peritoneal cavity is insufflated at a high-flow rate. It is not necessary to begin at a low-flow rate, because the size of the Veress needle limits the flow rate to 1.5–2 L per minute. All four abdominal quadrants are monitored for uniform insufflation with visual inspection and percussion. Once the IAP reaches 15 mm Hg, the trocar is advanced into the peritoneal cavity. A variety of different trocars are available, ranging in size from 3 to 12 mm; the choice depends on the size of the child, the purpose of the trocar, and the planned surgical approach. An axial or radial dilating trocar should always be used instead of a cutting trocar, which presents greater risk of access-related injuries and produces larger fascial defects. Another option is

an optical trocar, which has a translucent dilating tip and accommodates a camera, which allows for insertion under direct visualization. Diagnostic laparoscopy is performed to evaluate for any injuries to the underlying visceral organs and vasculature before proceeding with the insertion of additional trocars.

58.5.1.2 Open Hasson Technique

An open Hasson technique is indicated for infants and smaller children as well as in the setting of prior intra-abdominal surgery or failed primary access with the Veress needle. It is the safest technique for obtaining primary access but requires a larger incision. For this technique, an incision is made at the site for primary access with a scalpel and carried down to the fascia with gentle spreading or electrocautery. The fascia and parietal peritoneum are secured with sutures for superior retraction and opened. The trocar is placed into the peritoneal cavity under direct visualization. A traditional laparoscopic or robotic trocar may be used if the incision in the skin and fascia is not excessively large, or a Hasson trocar may be secured to the fascia with sutures. Another option is a balloon trocar, which is secured to the fascia between an inflatable balloon and a moveable foam pad.

58.5.1.3 Direct Trocar Insertion

The direct insertion of an optical trocar without the prior creation of pneumoperitoneum is discouraged because of the risk of access-related injuries.

58.5.1.4 Retroperitoneoscopic Primary Access

Primary access for a retroperitoneoscopic approach is obtained via an open technique. For a child in the full lateral decubitus or prone position, a 1-cm transverse incision is made with a scalpel approximately 1 cm inferior to the tip of the 12th rib and 1 cm lateral to the paraspinous muscles and is carried down to the lumbodorsal fascia with gentle spreading or electrocautery. The retroperitoneal space is entered by bluntly spreading the lumbodorsal fascia and muscle with either a finger or a blunt hemostat. The undersurface of the 12th rib, the psoas muscle, and the lower pole of the kidney are palpated to confirm a retroperitoneal location. Various techniques are used to develop the retroperitoneal space, including blunt dissection with a sponge, a laparoscope, insufflation, and a balloon dilating trocar.

58.5.2 Secondary Access

The insertion of additional trocars is more challenging in children because of their smaller anterior abdominal wall. The closer proximity of trocars may limit the mobility of the laparoscopic and robotic instruments and predispose to collisions. A difference of only a few millimeters may significantly affect the feasibility of the planned surgery, emphasizing the importance of proper placement. Once primary access is obtained, the anterior abdominal wall is inspected in an insufflated state. The planned sites of additional trocars are marked on the skin. Each additional trocar is inserted under direction visualization, using a 30° up camera through the primary access. A variety of different trocars are available, ranging in size from 3 to 12 mm; the choice depends on the size of the child, the purpose of the trocar, and the planned surgical approach. Owing to the higher compliance of the child's anterior abdominal wall, our preference is to advance a scalpel into the peritoneal cavity and gently dilate this track with a hemostat. This maneuver seems to facilitate the insertion of additional trocars with less force. Multiple templates exist for each surgical approach; they should serve as a guideline and be adapted to each individual child. Trocars should be inserted along the midline or lateral to the rectus muscles to avoid the epigastric vessels, and they should be at least 8 cm apart to minimize any collisions. Generally, they are triangulated with a camera trocar being situated between two or more working trocars. This amount of separation may not be feasible in infants and young children. Working trocars may therefore be inserted equidistant from the targeted organ and the camera trocar; this placement seems to allow for an adequate reach and still minimize any collisions. For instance, the camera and two working trocars may be configured along the midline for surgery of the upper urinary tract (Fig. 58.3). Advances in RALS have facilitated the configuration of trocars in infants and young children. Triangulation is not required for trocar placement with RALS. The da Vinci Xi® surgical system (Intuitive Surgical, Sunnyvale, CA) includes a boom-mounted system, less bulky robotic arms with longer reach and improved range of motion, and a laser targeting system.

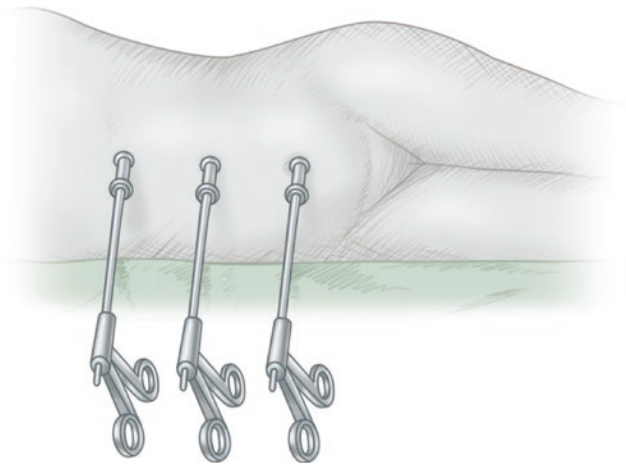

Fig. 58.3 The camera and two working trocars may be configured along the midline for surgery of the upper urinary tract

58.5.3 Laparoendoscopic Single-Site Surgery (LESS)

Access for LESS is obtained via an open technique. An incision is made at the site for access carried down to the fascia. The fascia is incised with electrocautery, after which the parietal peritoneum is opened. Access is typically obtained at the umbilicus for improved cosmesis. It is critical to make an incision in the skin and fascia that is only large enough to accommodate the access port, to prevent its dislodgement. The access port is placed into the peritoneal cavity under direct visualization. A variety of access ports are commercially available. Various combinations of laparoscopes and instruments have also been reported to minimize the limited range of motion and collisions. Curved and articulating instruments have specifically been developed to improve maneuverability. Our preference is to use a deflectable laparoscope and standard 5-mm laparoscopic instruments.

58.6 Closure

Maintaining one's vigilance is important while closing the peritoneal cavity, to prevent any trocar site–related complications such as fistula or hernia. Before closing the peritoneal cavity, the surgical field is inspected for hemostasis at a reduced IAP. An axial or radial dilating trocar typically creates a fascial defect that is approximately half its diameter. Fascial approximation may not be required when 3-mm trocars are used, but omental herniation is still possible even with the smallest trocars.

58.7 Complications

A growing body of evidence supports the safety and efficacy of laparoscopic surgery and RALS in children. Most studies have demonstrated a short-term outcome comparable to that of open surgery, as well as reduced postoperative pain, shorter hospitalization, more rapid convalescence, and improved cosmesis. No surgical approach has proven to be superior to others, with each one having its own advantages and disadvantages.

Few contemporary studies have assessed the complication rate of laparoscopic surgery and RALS in children. In a survey of more than 5400 laparoscopic surgeries performed by 153 pediatric urologists, Peters (1996) reported an overall complication rate of 5.38%. The complication rate decreased to 1.18% after preperitoneal insufflation was excluded, and only 0.39% of complications required a surgical intervention. Laparoscopic experience was the greatest predictor of complications in this survey.

Intraoperative complications related to pneumoperitoneum are easily reversible by decreasing the IAP and/or achieving an adequate compensatory response (e.g., increasing the tidal volume and respiratory rate on the ventilator for hypoxemia). Those related to positioning, access, and closure are largely self-limited and preventable by adhering to proper technique. Life-threatening complications, such as gas embolus and bowel or vascular injuries, are fortunately rare.

Conclusion

A growing body of evidence supports the safety and efficacy of laparoscopic surgery and RALS in children. Most studies have demonstrated a short-term outcome comparable to that of open surgery, as well as reduced postoperative pain, shorter hospitalization, more rapid convalescence, and improved cosmesis. No surgical approach has proven to be superior to others, with each one having its own advantages and disadvantages.

Laparoscopy has evolved from a simple diagnostic procedure to an array of minimally invasive techniques for the treatment of many pediatric urologic conditions requiring both extirpative and reconstructive surgery. Laparoscopic surgery and RALS will only continue to expand, with a focus on more dedicated training, higher-quality research, cost reduction, and continued innovation.

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Pyeloplasty

59

Boris Chertin and Prem Puri

Widespread use of maternal ultrasound has significantly changed the practice of paediatric urology. Pelvi-ureteric junction (PUJ) obstruction is the most common cause of hydronephrosis detected antenatally. Controversy continues on the optimal timing of surgical intervention in children with antenatally detected hydronephrosis, but it is beyond the scope of this chapter to discuss all the aspects of postnatal management of children with prenatal diagnosis of PUJ obstruction. The decision to intervene surgically in these infants has become more complex because spontaneous resolution of antenatal and neonatal upper urinary tract dilatations is being increasingly recognized.

59.1 Diagnosis

The recognition and relief of significant obstruction is important to prevent irreversible damage to the kidneys. Differentiating urinary tract dilatations that are significantly obstructive and require surgery from those that represent more anatomical variants with no implications for renal function is not a simple task, especially in newborns. The important aspect of postnatal investigations is to identify the group of patients who will benefit from early intervention and those who need to be carefully followed up. Currently, surgery is undertaken in infants with deteriorating renal function. Some patients present with urinary tract infection, irritability, vomiting, and failure to thrive. In those patients who had prenatal diagnosis of hydronephrosis, ultrasonography is performed in the first week of life. If hydronephrosis is confirmed, radionuclide studies are undertaken when the child is 6–8 weeks old in order to assess renal function and rule out obstruction. In those patients who present with clinical symptoms, a renal ultrasound is performed, and if it shows hydronephrosis without dilated ureters, the diagnosis is confirmed with radionuclide studies. The most commonly used radionuclides are diethylenetriamine pentaacetate (DTPA) and mercaptoacetyltriglycine (MAG3). Because MAG3 is excreted mostly by the renal tubules and yields better images in infants with compromised renal function and immature kidneys, we and others prefer to use traces with a high extraction rate (such as MAG3) in patients with hydronephrosis.

59.2 Operative Procedures

59.2.1 Choice of Surgical Approach

Various techniques are available to repair a PUJ obstruction. Current methods are classified as dismembered or flap techniques. Dismembered pyeloplasty of the Anderson-Hynes type consists of complete excision of an anatomically or functionally abnormal PUJ, correction of high insertion of the ureters, reduction of the renal pelvis, straightening of lengthy and tortuous proximal ureters, and transposition of the PUJ if obstruction is secondary to an aberrant vessel. Dismembered pyeloplasty enjoys a high success rate and has almost universal applicability. Occasionally, however, a patient has a dependent PUJ with a long, narrow ureteric segment or small intrarenal pelvis; in these cases, dismembered pyeloplasty may leave the surgeon with a short ureteric segment or may require aggressive ureteric and pelvic mobilization, so culp-pleloplasty type is more suitable.

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There are various surgical approaches to pyeloplasty:

- The classic, traditional approach is an extraperitoneal approach via lateral flank incision. All kidneys can be approached by flank incision easily with excellent exposure. The major disadvantage is a painful incision postoperatively.
- The anterior extraperitoneal approach is an excellent approach in younger children with a large renal pelvis, but access is more difficult in obese children.
- The posterior (dorsal) lumbotomy recently has gained wide popularity. The use of muscle splitting rather than muscle cutting makes it almost a minimally invasive procedure. The location of the posterior incision in the crease line has cosmetic advantages and allows direct access to the renal pelvis, but this a bombsight incision, requiring precise localisation of the PUJ. There is also some concern that this approach creates a scar that crosses normal skin folds. A bilateral procedure, if indicated, is possible under the same anaesthesia without position changes, using dorsal lumbotomy. This approach should not be used in older children or those who are significantly obese, however.
- A transperitoneal approach can be used when access to the abdominal contents is required. Recent technological advances and a shift across all the surgical fields to embrace minimally invasive surgery have led to increased utilization of minimally invasive pyeloplasty.
- Laparoscopic Anderson-Hynes dismembered pyeloplasty has yielded almost the same surgical outcome as open surgery and has been accepted as an effective alternative treatment for symptomatic PUJ obstruction in children. The development of robotic instruments designed to perform dismembered pyeloplasty has added a new dimension to laparoscopic suturing and will significantly enhance the ability to perform microanastomosis, which has been the limiting factor preventing many surgeons from performing this technically demanding operation.

59.2.2 Operative Procedure: Dismembered Pyeloplasty

Figures 59.1, 59.2, 59.3, 59.4, 59.5, 59.6 depict the traditional dismembered pyeloplasty.

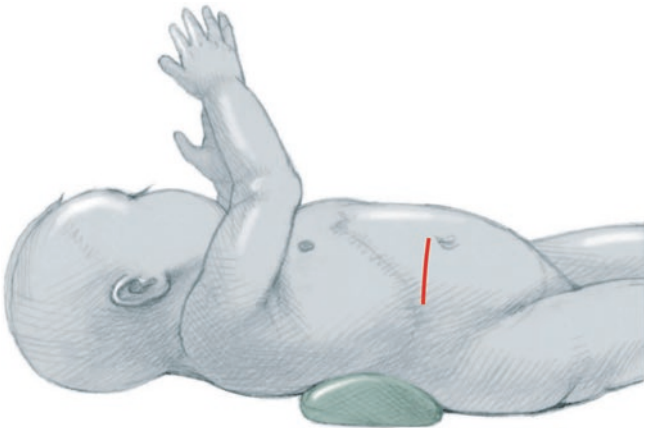

Fig. 59.1 The child is placed on the operating table in a supine position with the affected side elevated on a roll. A horizontal skin incision is made below the 12th rib, extending anteriorly

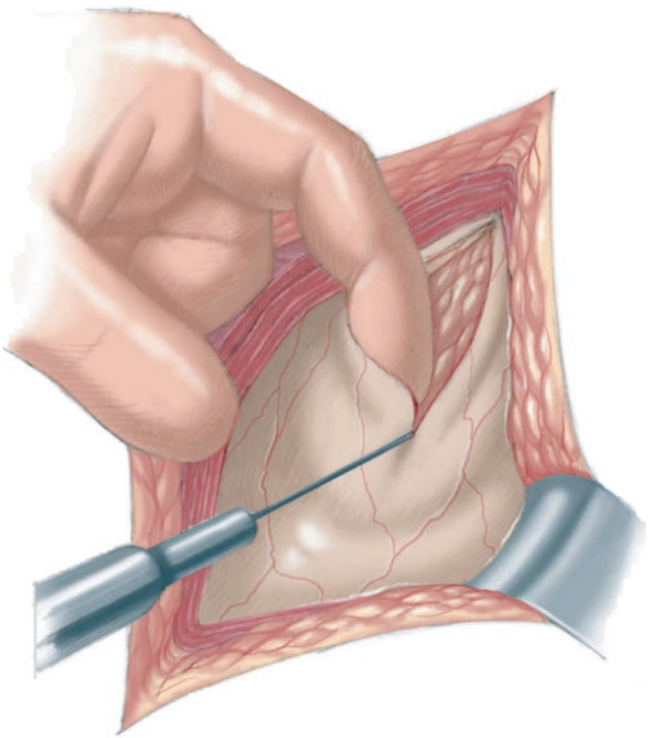

Fig. 59.2 The external and internal oblique muscles are divided by cutting diathermy in order to minimize blood loss. The entry into the retroperitoneum is made at the rib tip and extended posteriorly along the superior edge of the rib. The transversus abdominis is separated by blunt dissection in the lines of its fibres after the lumbodorsal fascia is incised. The peritoneum is retracted medially to allow more working space

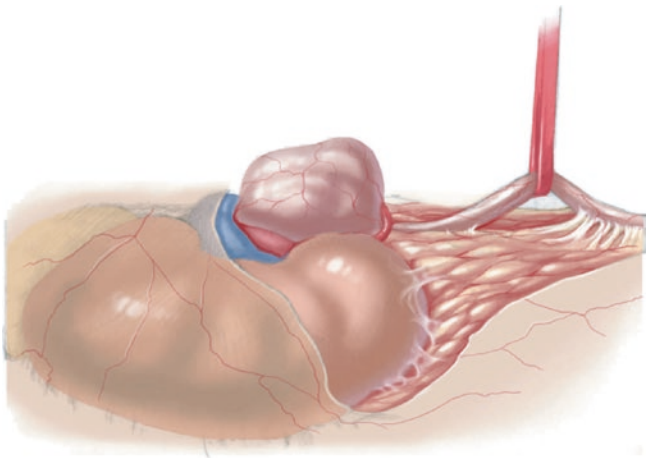

Fig. 59.3 Gerota's fascia is opened longitudinally in the posterior angle of the incision. The lower kidney moiety then comes to view. The lower pole is mobilized using blunt and sharp dissection. In most patients, a PUJ is best approached anteriorly. The main renal vessels and the branches and tributaries around the anterior renal hilum pose added risk for injury using the anterior approach to renal hilum. It is easy to rotate the kidney, retracting the lower pole forwards and upwards to approach the PUJ from behind. The ureter is then identified and mobilized downward

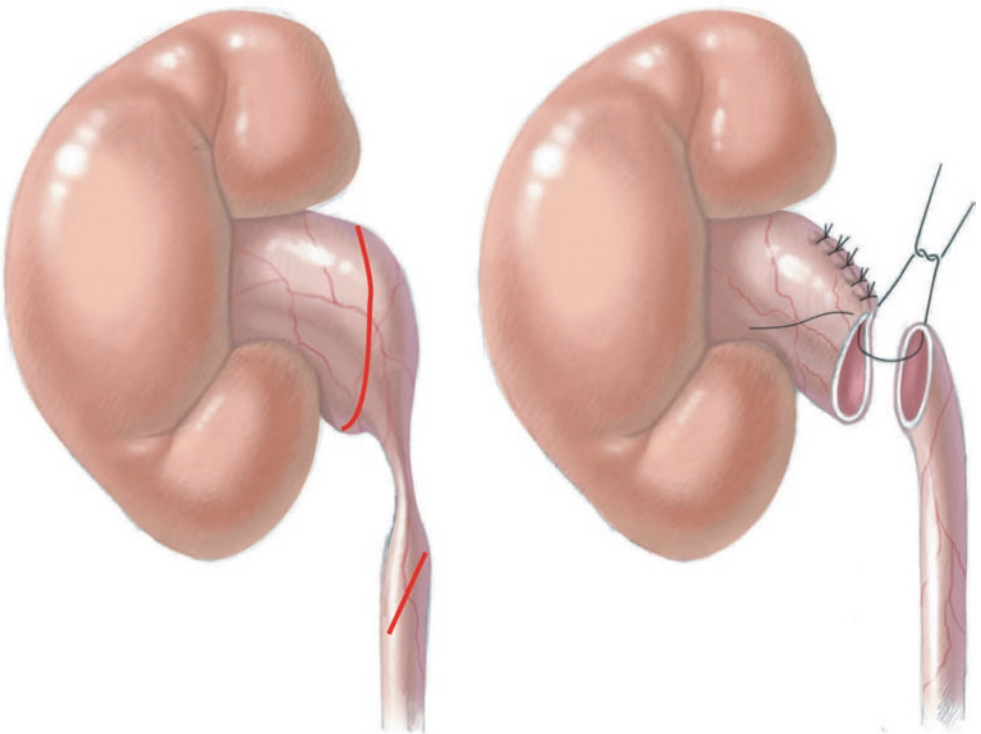

Fig. 59.4 Absorbable 6/0 sutures are placed in three locations: at the superomedial aspect of the pelvis, at the inferolateral aspects of the pelvis, and on the ureter about 5 mm below the PUJ. The ureter is divided obliquely above the ureteric stitch and the redundant pelvis is trimmed. The ureter is spatulated for approximately 2–3 mm by incising its posterolateral margin using fine scissors. At this stage, the decision should be made whether to utilize a nephrostomy tube or ureteric stent. Some surgeons still advocate nephrostomy tube drainage after pyeloplasty, but we believe that this technique is outdated and a double-J stent should be used in most cases. The ureter is then anastomosed to the renal pelvis over the stent. Some surgeons have expressed concern about passing a double-J stent through the tiny ureterovesical junction (UVJ) in small infants, with the risk of later development of UVJ obstruction. Furthermore, the need for a second general anesthesia to remove the stent cannot be ignored. Therefore, some surgeons have used a Pippi-Salle stent nephrostomy tube (Cook, USA) for dismembered pyeloplasty

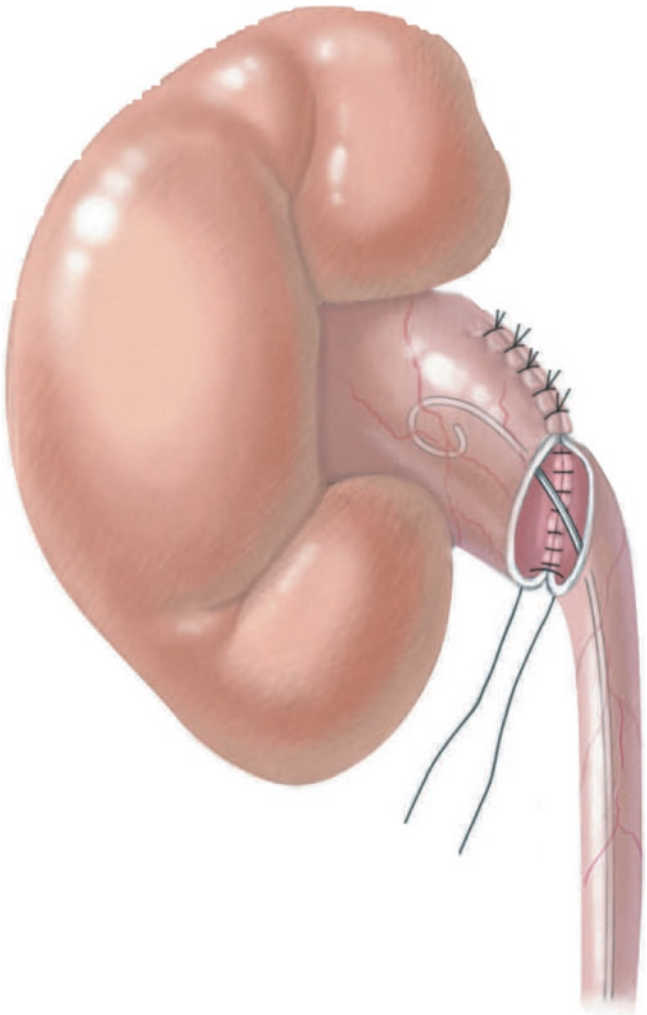

Fig. 59.5 In case of an aberrant renal artery, the anastomosis is performed anterior to the vessel. The first stitch is placed approximating the lower margin of the spatulated ureters and the lower margin of the renal pelvis using absorbable 6/0 sutures. A further interrupted suture is then placed to complete the posterior aspect of the anastomosis. Suture should advance up the posterior wall and then up the anterior wall, keeping the suture knots on the outside of the lumen of the anastomosis. All sutures should include 1 mm of tissue on each side. The sutures should be placed about 1.5–2 mm apart

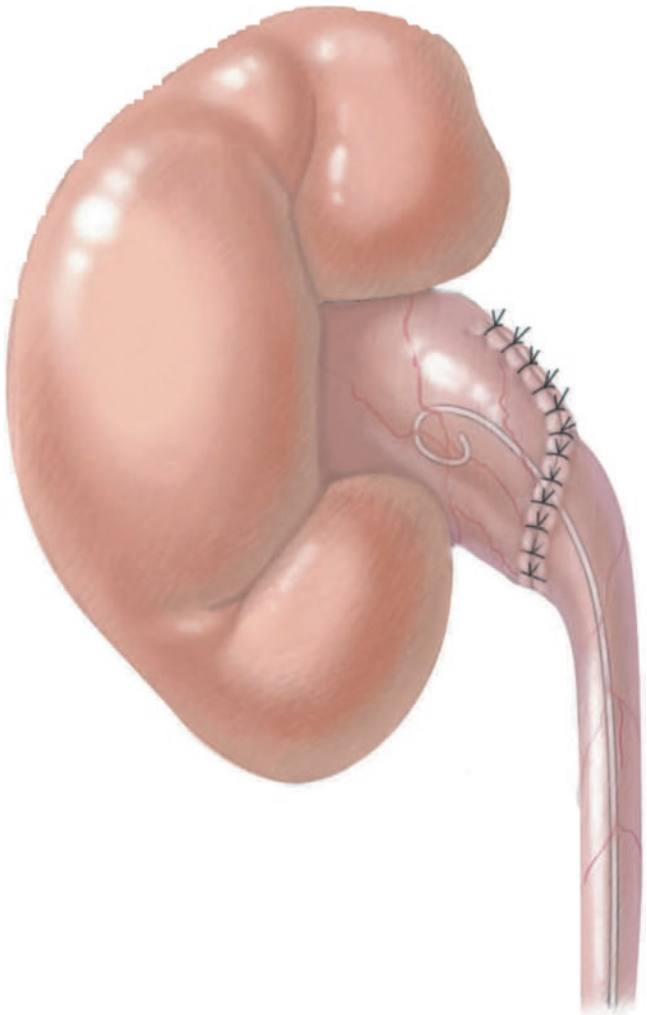

Fig. 59.6 The anterior closure of anastomosis is then performed using either interrupted or running 6/0 suture. The stent, which is passed through the anastomosis, prevents the inadvertent picking up and inclusion of the posterior wall into the stitch line during this stage of anastomosis

59.2.3 Operative Procedure: Flap Pyeloplasty

Figures 59.7, 59.8, 59.9 show the procedure for a spiral flap (culp) pyeloplasty, which is suitable for a long, dependent, stenotic uretero-pelvic obstruction.

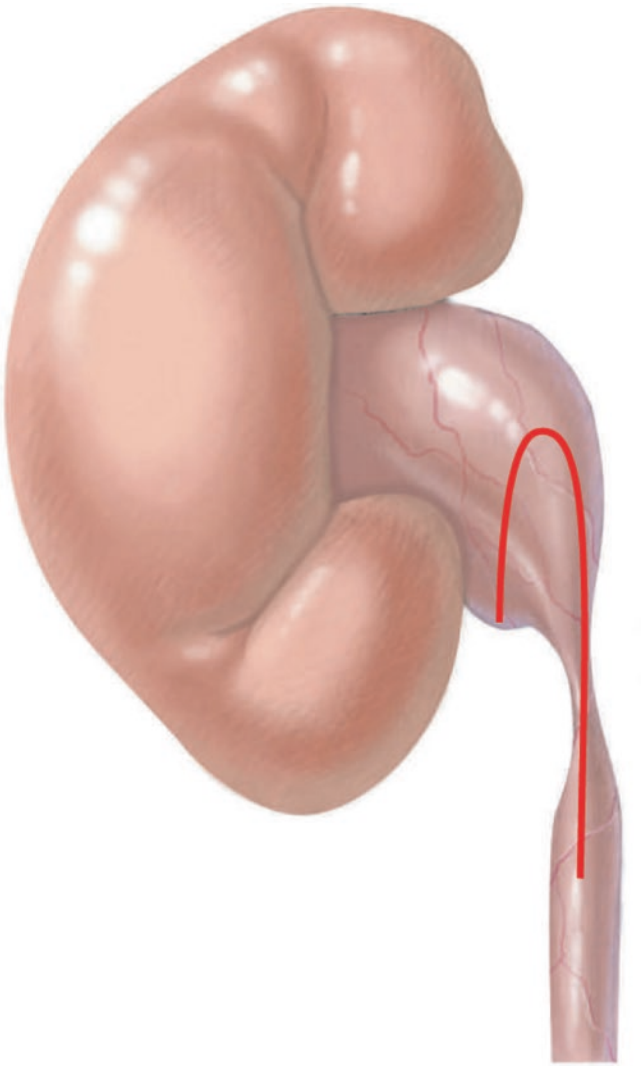

Fig. 59.7 To perform a spiral flap (Culp) pyeloplasty, the incision on the ureters must be adequate, covering the stenotic area

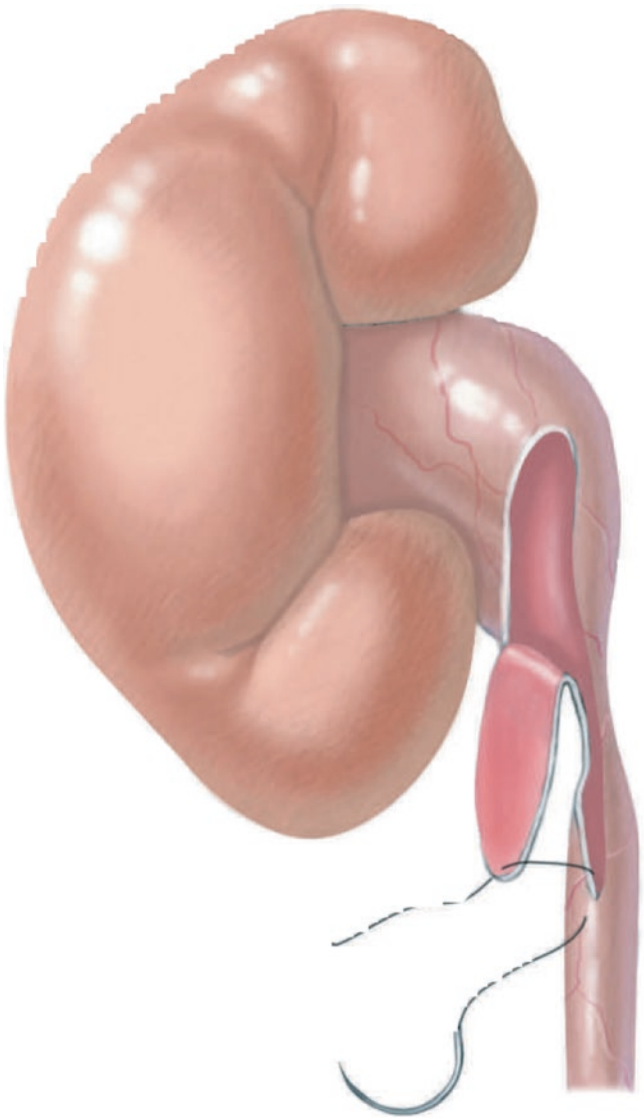

Fig. 59.8 The flap of equal length is based on a broad base

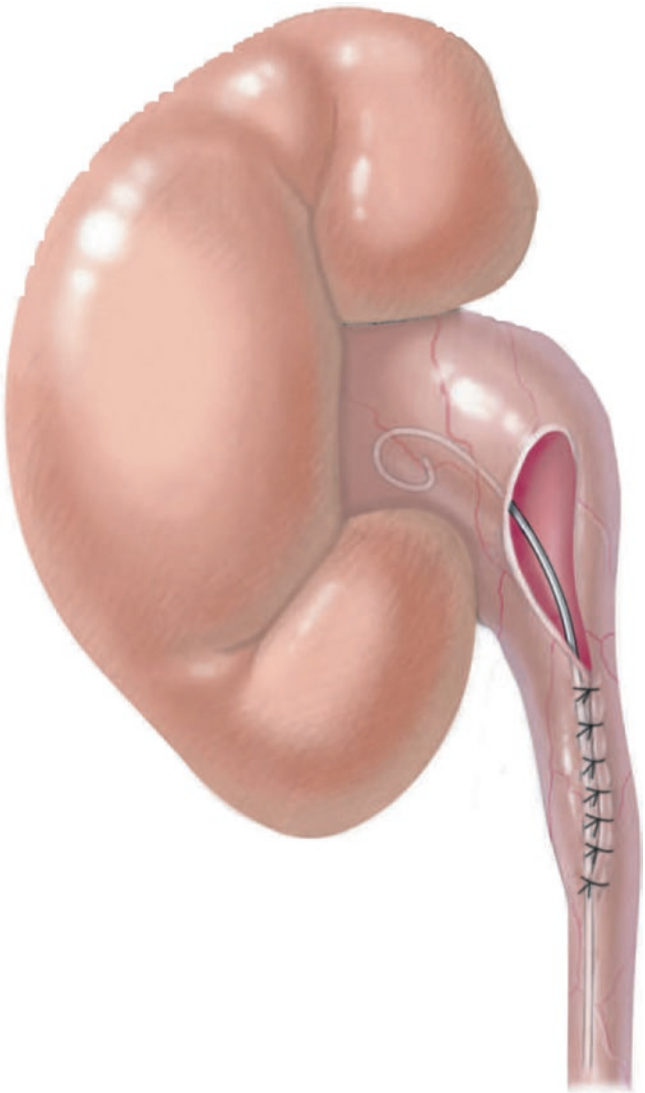

Fig. 59.9 The anterior layer of ureters and flap is sutured using 6/0 sutures

59.2.4 Operative Procedure: Laparoscopic Dismembered Pyeloplasty

Laparoscopic dismembered pyeloplasty can be performed via the transabdominal approach or a retroperitoneal approach. We have adopted a transperitoneal approach because it offers larger operating space, which makes intracorporeal anastomosis easier.

The patients are secured in a flank position and a urinary catheter is inserted. Upon catheter insertion, antibiotic prophylaxis is administered. The bladder is filled with irrigation water stained with indigo carmine. Following the insertion of three 5-mm ports and abdominal insufflation, the upper ureter and the renal pelvis are identified and dissected.

At this stage, a trans-flank holding stitch through the renal pelvis is placed in order to facilitate pelvic dissection and to stabilize the renal pelvis during intracorporeal anastomosis. The ureter is dismembered proximal to the UPJ, at the level of the renal pelvis. The excess tissue of the renal pelvis is used for grasping and manipulating the ureter. The abdomen is desufflated, the lateral port is removed, and the dismembered ureteric end is externalized to skin level, at the port site. The externalized ureter is then spatulated. The JJ ureteral stent is inserted in an antegrade fashion over a hydrophilic guidewire. The presence of the indigo carmine–stained urine indicates the correct position of the distal part of the stent in the urinary bladder**.** The first stitch for future intracorporeal anastomosis is applied on the lower part of the spatulated ureter. Insufflation is resumed and the stented, spatulated ureter is returned to the abdomen. The laparoscopic anastomosis is made in the usual way, using two Vicryl 4/0 or 5/0 running sutures; the excess tissue is then excised and a stent is inserted. The use of excess renal pelvic tissue facilitates manipulation of the ureter and avoids injury to the ureteric tissue in the anastomotic area. Following anastomosis completion, the excess renal pelvic tissue is excised and the renal pelvis is closed using running 4/0 Vicryl suture. A 7-Fr drain is left alongside the anastomosis. The stent is left in place for 4–6 weeks and is then removed under general anesthesia or deep sedation at cystoscopy.

59.3 Results

The objective of pyeloplasty is to achieve a dependent, adequately calibrated, watertight PUJ. Different techniques are available to repair a PUJ obstruction. The dismembered Anderson-Hynes pyeloplasty is suitable for most patients with PUJ obstruction. Use of either a double-J ureteral stent or a Pippi-Salle stent nephrostomy tube has virtually eliminated ureteral leaks and early obstructions.

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Endoscopic Treatment of Vesicoureteral Reflux

60

Prem Puri

Primary vesicoureteral reflux (VUR) is the most common urologic anomaly in children, occurring in 1–2% of the pediatric population and 30–40% of children presenting with a urinary tract infection (UTI). The familial nature of VUR is well-recognised. Several studies have shown that siblings of children with VUR are at much higher risk for reflux than the general population, with a reported prevalence between 26% and 50%. VUR is classified into five grades (Fig. 60.1).

The main goals of treating a child with VUR are preventing recurring febrile urinary tract infections and minimising the risk of renal damage and long-term renal impairment. Renal parenchymal damage can be congenital (hypoplasia or dysplasia) or acquired scarring. Acquired scarring occurs from pyelonephritis-induced renal injury, whereas congenital reflux nephropathy is a result of abnormal embryologic development and subsequent renal dysplasia. Exposure to UTIs in patients with congenital renal dysplasia can lead to progression of renal parenchymal damage. Reflux-associated nephropathy is an important cause of hypertension and end stage renal disease.

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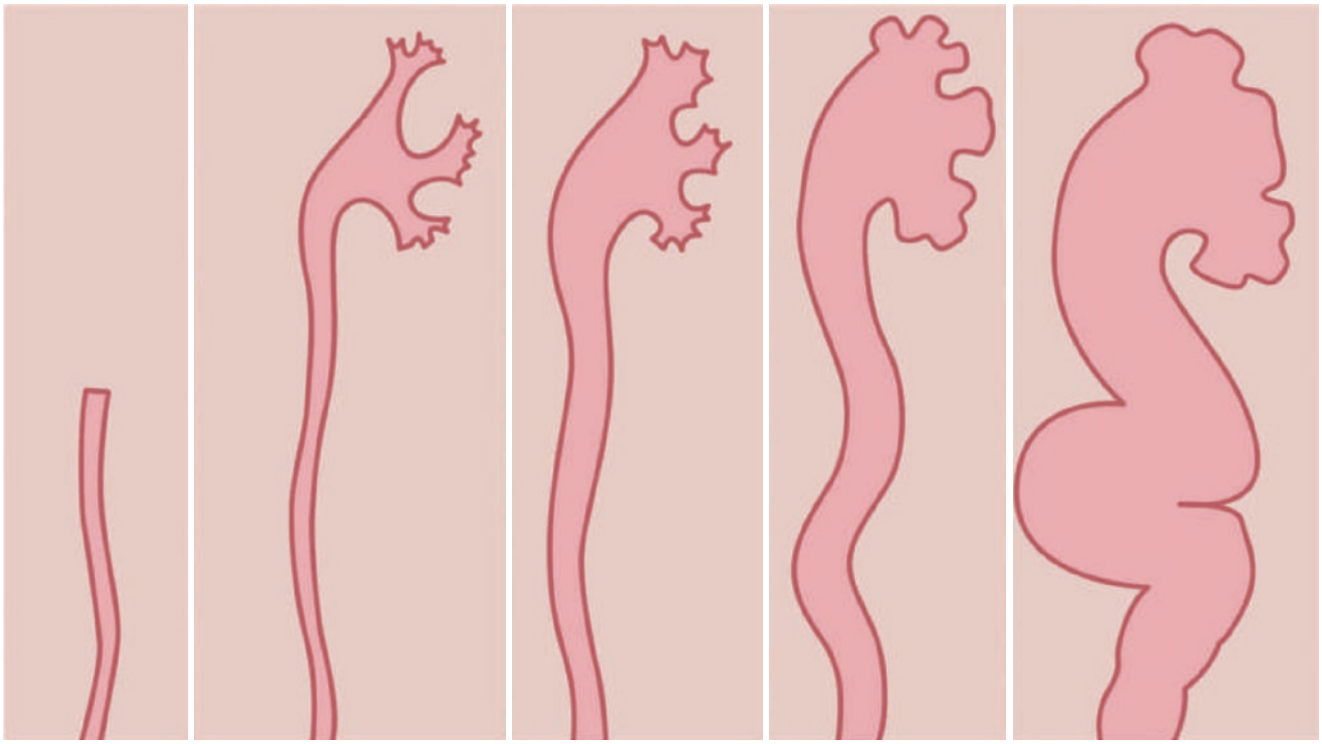

Fig. 60.1 Grades of vesicoureteral reflux (VUR)

60.1 Treatment Options

Options for treatment of VUR include observation (with or without antibiotic prophylaxis) and surgical correction. There has been no consensus regarding when medical or surgical therapy should be used. A number of prospective studies have shown low probability of spontaneous resolution of high-grade VUR during conservative follow-up. Furthermore, all of these studies revealed that observation therapy does carry an ongoing risk of renal scarring. Open surgery is the standard treatment for VUR when indicated. Although ureteral reimplantation is effective, this operation is not free of complications.

Since its introduction in 1984, minimally invasive endoscopic correction of VUR has become an established alternative to long-term antibiotic prophylaxis and open or laparoscopic ureteral reimplantation. The indications for endoscopic therapy for VUR include:

- High-grade primary VUR (grades III–V)
- VUR in duplex renal systems
- VUR secondary to neuropathic bladder and posterior urethral valves
- VUR in failed reimplanted ureters
- VUR into ureteral stumps

A number of tissue-augmenting substances have been used endoscopically for subureteral injection. The most widely used tissue-augmenting substance for injection therapy for VUR is dextranomer microspheres in sodium hyaluronic acid solution (Deflux®; Salix Pharmaceuticals, Raleigh, NC, USA). It has been reported that dextranomer/hyaluronic acid copolymer (DX/HA) is biodegradable, has no immunogenic properties, and has no potential for malignant transformation. The dextranomer microspheres (80–250 μm) are mixed in 1% high-molecular-weight sodium hyaluronic acid solution. Each milliliter of Deflux® contains 0.5 mL sodium hyaluronan and 0.5 mL of microspheres. Since Deflux® was approved by the United States Food and Drug Administration (FDA) in 2001 as an acceptable tissue-augmenting substance, we have used endoscopic DX/HA injection as the first line of treatment in the management of intermediate- and high-grade VUR. We recently reported our experience with endoscopic subureteral DX/HA injection as the first line of treatment in 1551 children with intermediate- and high-grade VUR (2341 ureters). VUR was resolved after the first endoscopic injection of DX/HA in 87.1% of ureters, 7.3% after a second injection, and 1.6% after a third. Febrile urinary tract infection developed in 4.6% of the patients during median follow-up of 5.6 years. None of the patients in this series needed reimplantation of ureters or experienced any significant complications.

60.2 Endoscopic Subureteral Injection (STING) Procedure

The disposable Puri catheter for injection (Storz) is a 4-Fr nylon catheter onto which is swaged a 21-gauge needle with 1 cm of the needle protruding from the catheter (Fig. 60.2). Alternatively, a rigid needle can be used. A 1-mL syringe filled with Deflux® paste is attached to the injection catheter.

All cystoscopes available for infants and children can be used for this procedure. The injection catheter can be introduced through a 9.5F, 11F, or 14F Storz cystoscope; a 9.5F Wolf cystoscope; or a 9.5F or 11.5F angled Wolfe cystoscope (Fig. 60.3).

The patient should be placed in a lithotomy position. The cystoscope is passed and the bladder wall, the trigone, bladder neck, and both ureteric orifices are inspected. The bladder should be almost empty before proceeding with injection, as this helps to keep the ureteric orifice flat rather than away in a lateral part of the field.

The injection of Deflux® paste or any other tissue-augmenting substance should not begin until the operator has a clear view all around the ureteric orifice. Under direct vision through the cystoscope, the needle is introduced under the bladder mucosa 2–3 mm below the affected ureteric orifice at the 6 o'clock position (Fig. 60.4a). In children with grade IV and V reflux with wide ureteral orifices, the needle should be inserted not below but directly into the affected ureteral orifice. It is important to introduce the needle with pinpoint accuracy. Perforation of the mucosa or the ureter may allow the paste to escape and may result in failure.

The needle is advanced about 4–5 mm into the lamina propria in the submucosal portion of the ureter and the injection is started slowly (Fig. 60.4b). As the paste is injected, a bulge appears in the floor of the submucosal ureter. During injection, the needle is slowly withdrawn until a "volcanic" bulge of paste is seen. Most refluxing ureters require 0.5–1.0 mL of Deflux® to correct reflux.

A correctly placed injection creates the appearance of a nipple, on the top of which is a slit-like or inverted crescentic orifice (Fig. 60.4c). If the bulge appears in an incorrect place, such as at the side of the ureter or proximal to it, the needle should not be withdrawn, but should be moved so that the point is in a more favourable position. The noninjected ureteric roof retains its compliance while preventing reflux.

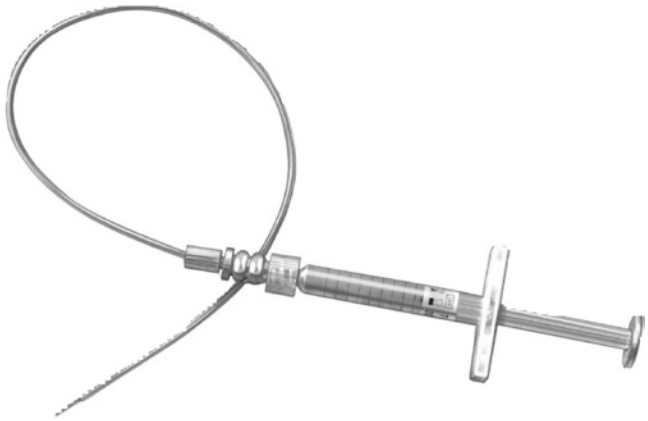

Fig. 60.2 Puri Catheter for endoscopic subureteral injection

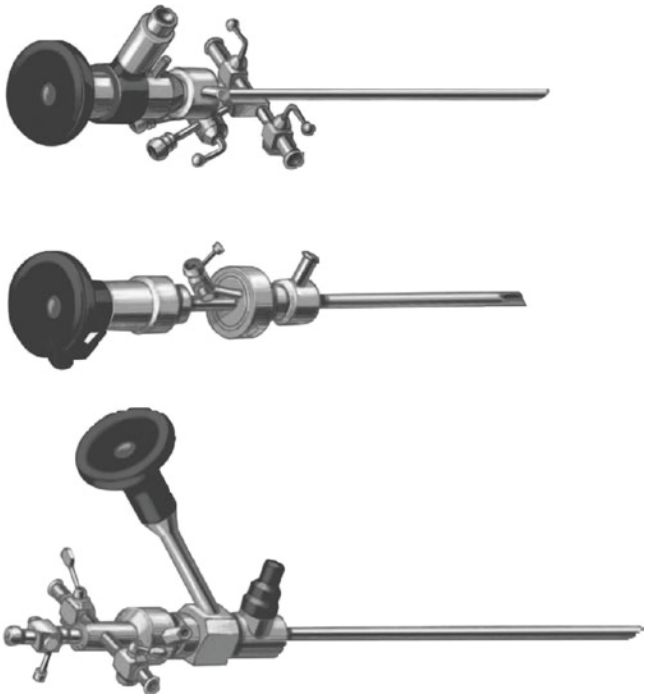

Fig. 60.3 Cystoscopes to introduce the injection catheter

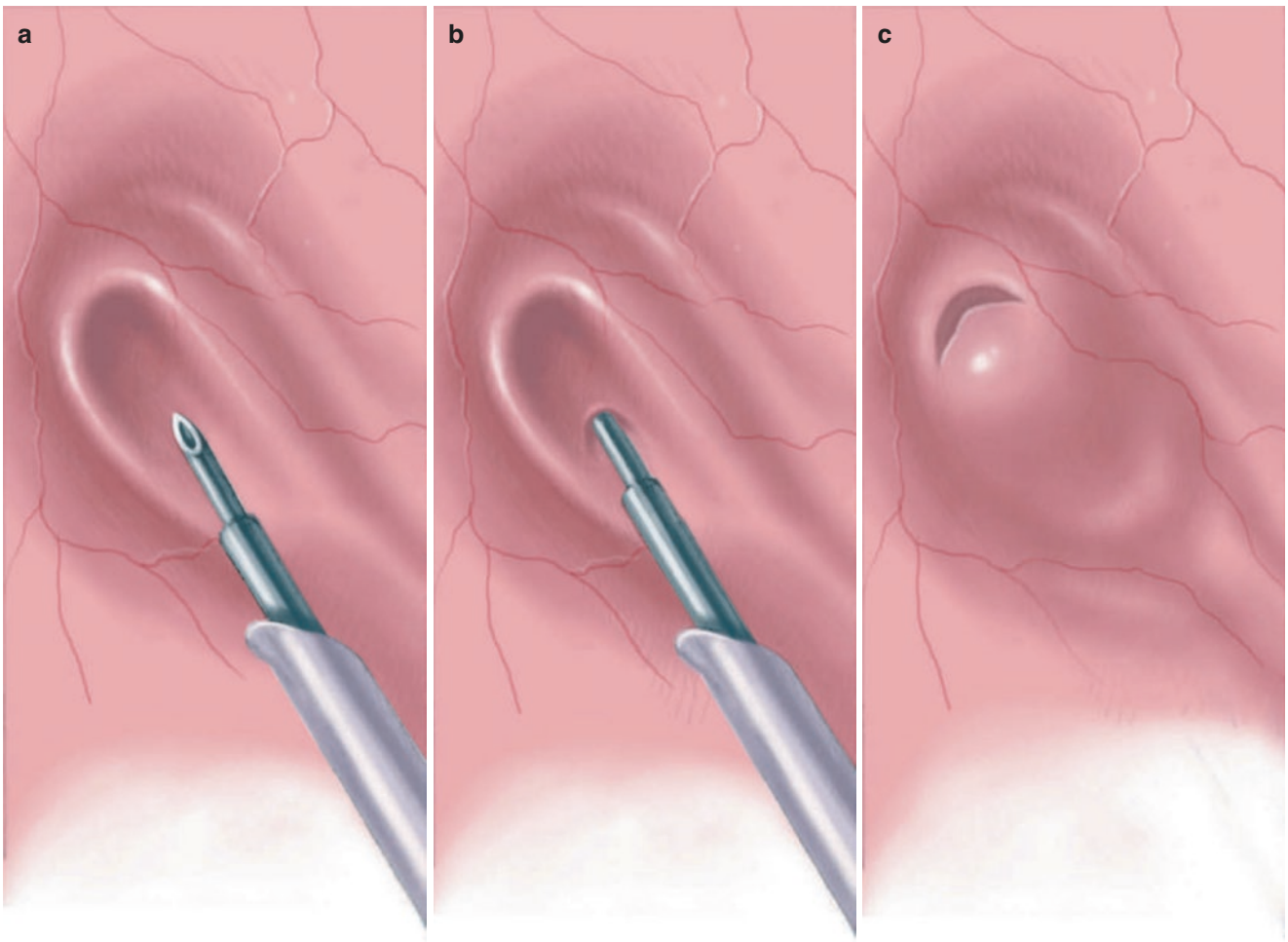

Fig. 60.4 (**a**–**c**) Steps in subureteral injection

60.3 Postoperative Care and Follow-Up

Postoperative urethral catheterization is not necessary. Most patients are treated as day cases. Co-trimoxazole is prescribed in prophylactic doses for 3 months after the procedure. Micturition cystography and renal ultrasonography are performed 3 months after discharge. A follow-up renal and bladder ultrasonographic scan are obtained 12 months after successful endoscopic correction of reflux.

60.4 Complications and Outcome

Endoscopic treatment is a simple, safe, and effective procedure in the management of all grades of reflux. Procedure-related complications are rare. The only significant complication with this procedure has been failure—either initial failure, when the reflux is not abolished by the injection, or recurrence, when initial correction is not maintained. About 15–20% of refluxing ureters require more than one endoscopic injection of paste to correct the condition. Apart from failure to correct reflux, the only other reported rare complication following STING is vesicoureteral junction obstruction.

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Vesicoureteral Reflux: Surgical Treatment

61

Jack S. Elder

Vesicoureteral reflux (VUR) affects approximately 1% of children. VUR predisposes an individual to upper urinary tract infection (UTI) (that is, pyelonephritis). Repeated episodes of pyelonephritis can result in renal scarring (reflux nephropathy), hypertension, impaired somatic growth, renal insufficiency, end-stage renal disease, and complications during pregnancy. VUR can be managed medically or surgically.

Medical management is based on the principles that VUR often diminishes or resolves over time, and maintaining sterile urine minimizes the risk of reflux nephropathy. Medical management includes bladder training (encouraging regular micturition and treating symptoms of bladder/bowel dysfunction) and possibly antibiotic prophylaxis with a daily dose of an antimicrobial such as nitrofurantoin, trimethoprim, or sulfatrim. Many children undergo regular follow-up assessment with a voiding cystourethrogram (VCUG) and renal ultrasonogram (US) every 12–18 months. Medical management is continued until the VUR resolves or improves sufficiently that the VUR no longer seems clinically significant. Many clinicians consider grades I and II VUR to be benign. (*See* Fig. 60.1 for a diagram of grades I–V.)

Surgical management is generally recommended when medical management has failed—that is, the child experiences breakthrough UTI while receiving antimicrobial prophylaxis or has persistent VUR or noncompliance with the prescribed therapy. In addition, children with VUR that is unlikely to resolve, such as grade IV, V, or bilateral grade III VUR, often are managed surgically, as is VUR associated with a complete duplicated collecting system, ureterocele, ectopic ureter, or bladder exstrophy.

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61.1 Types of Surgical Management

Surgical management can be accomplished through an incision ("open surgical treatment"), endoscopically (subureteral injection, covered in Chap. 60), and laparoscopically or robotically with laparoscopic assistance. Because the bladder is an abdominal organ in children, open surgical therapy is easiest when the child is prepubertal. Following puberty, the bladder descends behind the pubic symphysis, and dissection of the ureters is more difficult. The decision whether the child should undergo endoscopic or open surgical management should be a joint decision between the patient's family and the surgeon. This chapter address the options for open and laparoscopic/robotic surgical management of VUR.

The principle of surgical correction of VUR is to create a 4:1–5:1 ratio of submucosal tunnel length to ureteral width. The intramural ureter should be in a fixed portion of the bladder. There are numerous ways to correct VUR; some are intravesical, some are extravesical, and some are combined. The three techniques that are used most commonly are the transtrigonal (Cohen) procedure, the Politano-Leadbetter procedure, and detrusorrhaphy. The first two are intravesical techniques, whereas detrusorrhaphy is extravesical. The advantage of the latter is that there is minimal bladder spasm and haematuria associated with the procedure, because the bladder is not opened widely. In contrast, with an intravesical approach, children typically experience a moderate amount of dysuria, urgency, and haematuria for a week postoperatively.

A modified approach may be recommended if ureteral tailoring is necessary because the ureter is too wide to achieve a 4:1 ratio. With improvements in pediatric anaesthesia and postoperative pain management, children often stay in the hospital for 1–2 days after these procedures. The success rate is 95–98% for grades I through IV VUR and somewhat lower for grade V.

61.2 Operative Principles

During open surgical correction of VUR, several principles apply:

- Optical magnification with loupes is recommended.
- It is helpful to use fine cautery (Pena tip) for the operative procedure.
- Tenotomy scissors are ideal for tissue dissection throughout the entire procedure, because the tips are fine and blunt. Metzenbaum scissors are much wider and do not dissect the tissues as easily.
- The exposed bladder mucosa should not be wiped with a sponge, and suction should not be applied to the bladder mucosa. These manoeuvres will result in significant mucosal oedema, which may make submucosal dissection difficult.
- The submucosal tunnel should be four or five times as long as the width of the ureter.
- Ureteral stents are unnecessary in routine ureteroneocystostomy, but are recommended for VUR in a solitary kidney, reoperative cases, ureteral tailoring, or if there is significant detrusor hypertrophy from posterior urethral valves, neuropathic bladder, or severe bladder/bowel dysfunction.
- If there is a duplicated collecting system, both ureters may be treated as one and re-implanted together in one tunnel. An alternative treatment is to perform a ureteroureterostomy with distal ureterectomy, in which the refluxing ureter is anastomosed to the nonrefluxing ureter, and the distal segment of the refluxing ureter is excised.

61.3 Operative Techniques

61.3.1 Transtrigonal (Cohen) Procedure

The abdomen and genitalia should be prepped with betadine or chlorhexidine, and the urethral meatus should be included in the operative field so that a catheter may be inserted or removed when necessary. Preoperative broad-spectrum antibiotics should be administered.

A Foley catheter should be inserted into the bladder and the bladder is filled manually with sterile water to push the peritoneum superiorly. A Pfannenstiel incision is made one finger-breadth above the pubic symphysis (Fig. 61.1). The limits of the incision should be the lateral borders of the rectus muscles. The incision is carried down to the external oblique fascia and haemostasis is achieved.

Make a transverse incision in the anterior rectus sheath in the line of the incision, exposing the rectus muscles (Fig. 61.2). Using the fine-tip needle electrode (Pena tip) for cautery, develop rectus fascial flaps superiorly, nearly to the umbilicus. It is helpful to grasp the superior rectus fascia with straight mosquito clamps (Fig. 61.3). Using an identical technique, the inferior rectus fascia is mobilized to the pubic symphysis.

Separate the rectus muscles in the midline with a Kelly clamp and use the cautery to incise the linea alba, the midline attachment of the rectus muscles. With tenotomy scissors, incise the transversalis fascia and expose the bladder (Fig. 61.4). The distended bladder is dissected out bluntly. The peritoneum should be swept superiorly to prevent inadvertent peritoneotomy.

The Denis Browne ring retractor is then used to hold the rectus muscles apart. Allis clamps are placed on either side of the midline of the bladder. The detrusor is incised in the midline with the cautery. Ideally, only the muscular layer should be divided first, allowing cauterization of the small arterial vessels in the detrusor. The mucosa then protrudes out and may be cut with tenotomy scissors or the cautery. The bladder is then drained. The bladder is then isolated with 4/0 or 3/0 absorbable traction sutures (placed with one tie) in the four corners of the bladder wall (Fig. 61.5), and a figure-of-eight stitch is placed in the bladder neck to prevent it from spreading open.

The Denis Browne ring retractor is then placed in the bladder. The side blades have two sizes; usually the larger size is necessary. Several moistened gauze sponges are placed in the bladder dome and the malleable blade is inserted and adjusted to retract the dome superiorly. When the malleable blade is inserted, the ureteral orifices should be easily visible. The rake retractor is placed inferiorly. In older children the Denis-Browne retractor may be too small, and instead a child-size Balfour retractor may be necessary, using the bladder blade for retraction superiorly.

The ureteral orifices are identified and cannulated with 8F or 5F pediatric feeding tubes; in infants and very young children it may be necessary to use a 3.5F feeding tube. The catheter should be passed up to the kidney and sutured to the bladder wall with 4/0 absorbable sutures. A clamp is placed on the feeding tube and suture for distal traction, which aids in ureteral dissection.

The ureter is then dissected out (Fig. 61.6). The Pena tip cautery on cutting current or a fresh number 15 scalpel is used to circumscribe the ureter. The mucosa inferomedial to the orifice is grasped with Castroviejo 4″ 0.5-mm tooth forceps, and a deep cut is made in the space between ureter and mucosa. This plane around the ureter is developed by sharp dissection, exposing the underlying detrusor muscle.

The ureter has a pearly white appearance. Dissecting too close to the ureter risks devascularization, and dissecting too far away in the detrusor often results in significant bleeding. Megaureters often have a better intrinsic blood supply, and devascularization during mobilization of the megaureter is uncommon. A small, right angle clamp can be used to develop the plane between the ureter and detrusor, and the clamp may be opened to separate the muscle from the ureter. Muscular attachments to the ureter may be cauterized gently, being careful to keep the tip of the cautery away from the ureter. If there has been a recent urinary tract infection, the ureter tends to be more adherent to the muscle. The ureter is dissected out until the peritoneum is identified and can be swept away.

The ureteral hiatus must then be closed to prevent a diverticulum from forming. Three or four interrupted 3/0 absorbable sutures are placed through the detrusor muscle on each side, starting inferomedially and working superolaterally; the hiatus should not be closed too tightly.

The submucosal tunnel is then made. The mucosa medial to the hiatus should be grasped gently. Using the tenotomy scissors, with the tips pointed anteriorly, the mucosal attachment to the underlying detrusor is incised to establish the submucosal plane. Next, the tenotomy scissors are passed into the plane and spread gently (Fig. 61.7). The scissors should be opened approximately twice as wide as the ureteral diameter. The submucosal tunnel is gradually lengthened. When the tunnel length is four or five times as long as the width, the tips of the scissors should be used to elevate the mucosa. The scissors should be opened slightly and the cautery should be used to open the mucosa. The tips of the scissors are advanced through the mucosa and opened further.

The tip of the feeding tube in the ureter is cut off. A right-angled or curved mosquito clamp is passed backward through the opening in the mucosa toward the ureteral hiatus and the

tip of the feeding tube is grasped. The tip of the feeding tube is then pulled through the submucosal tunnel (Fig. 61.8).

The suture holding the feeding tube is cut and the tip of the ureter is trimmed slightly, being careful to excise any portion of the ureter that seems devascularized. If both ureters are being re-implanted, it is appropriate to place them in the same submucosal tunnel.

The ureter is spatulated slightly. With the feeding tube in place, the ureter is sutured to the bladder mucosa with interrupted 5/0 or 6/0 absorbable sutures; the two distal apical sutures should be placed through the bladder muscle also, to help fix the ureter in place. There should be no tension on the ureter. Small mosquito clamps are placed for traction on the proximal and distal apical sutures to allow easy identification of the new ureteral orifice. The feeding tube should be removed and then reinserted into the ureter; the feeding tube should pass easily through the submucosal tunnel (Fig. 61.9). After the ureter(s) is fixed in place, the bladder mucosa is closed with running 5/0 absorbable sutures. It is unnecessary to leave the ureter stented unless there is significant bladder wall oedema, the ureter is draining a solitary kidney, or the patient is undergoing a secondary procedure.

If a satisfactory submucosal tunnel cannot be made because of mucosal oedema, the mucosa may be incised and peeled back, creating a trough in which to lay the ureter. In fact, the mucosal edges may be sutured to the edge of the ureter, and the epithelium will grow over the ureter, creating a submucosal tunnel.

The bladder is then closed. A two-layer closure is performed. The muscular layer is closed with a running 2/0 polyglycolic acid (PGA) imbricating stitch (Connell); a second layer uses a running 2/0 PGA Lembert stitch. The rectus muscles are approximated with interrupted 3/0 chromic catgut. The rectus fascia is closed with a running 2/0 PGA or PDS (polydioxanone). A Foley catheter is left in place overnight.

If a unilateral transtrigonal ureteroneocystostomy is performed, there is a 10% risk of contralateral reflux, probably secondary to destabilization of the contralateral ureter during mobilization of the refluxing ureter. The risk is 50% if the contralateral ureter refluxed in the past but is no longer refluxing. This complication may be prevented by performing bilateral ureteroneocystostomy or by performing a contralateral Gil-Vernet ureteral reimplant (contralateral ureteral meatal advancement).

After cannulating the ureteral orifice with a feeding tube of appropriate size and suturing it in place, a Y-shaped mucosal incision is made from the medial surface of the ureter medially to the midline of the bladder trigone (Fig. 61.10). The medial wall of the ureter is dissected out, separating it from the underlying detrusor muscle. The medial extension of the mucosal incision is opened also, exposing the detrusor. The ureteral meatus is moved medial, to the midline. The ureteral meatus is fixed to the mucosa and underlying detrusor with several 5/0 absorbable sutures.

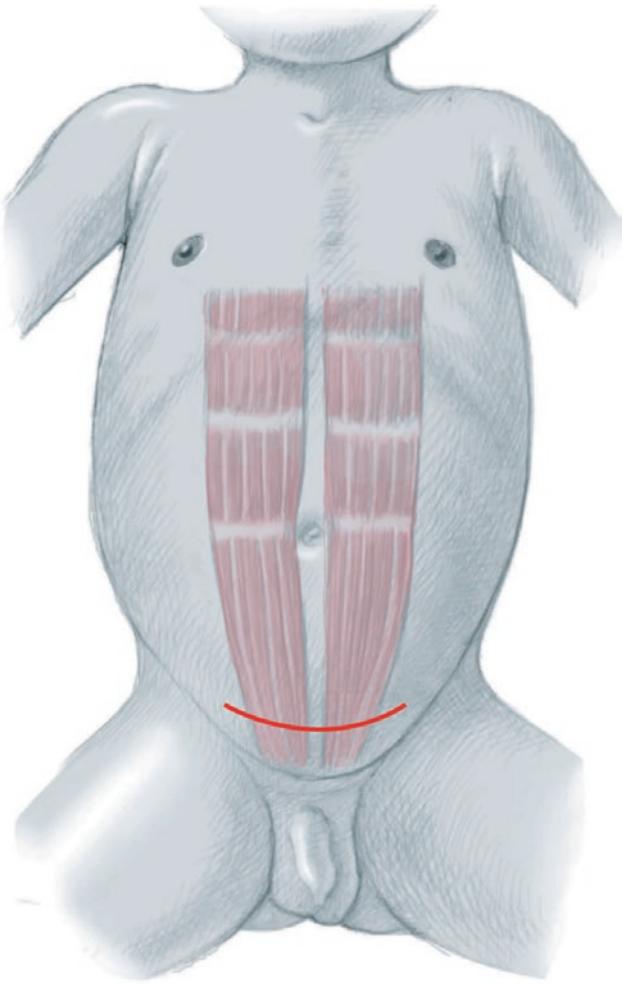

Fig. 61.1 Pfannenstiel incision for transtrigonal procedure

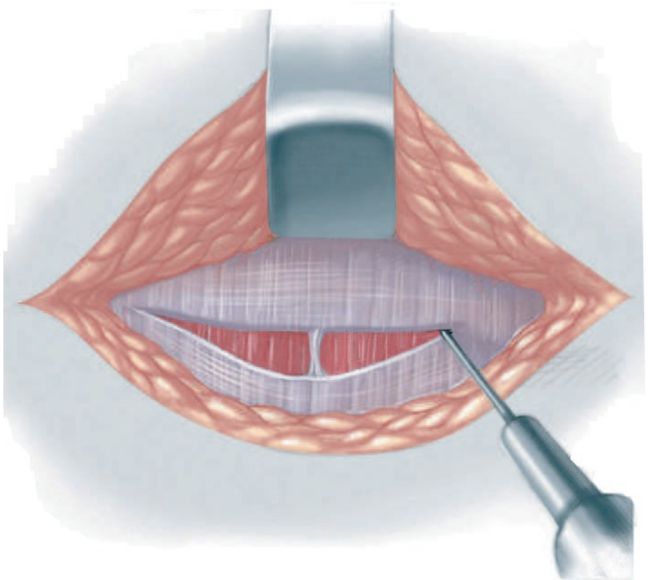

Fig. 61.2 Transverse incision in the anterior rectus sheath

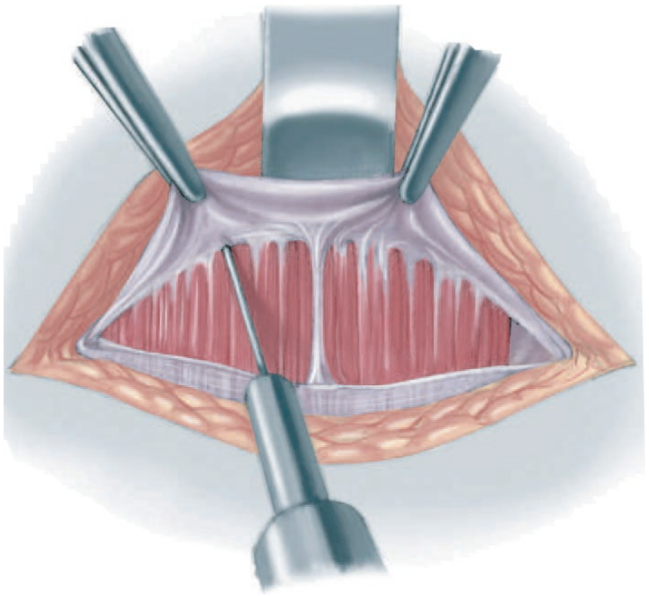

Fig. 61.3 Developing rectus fascial flaps superiorly

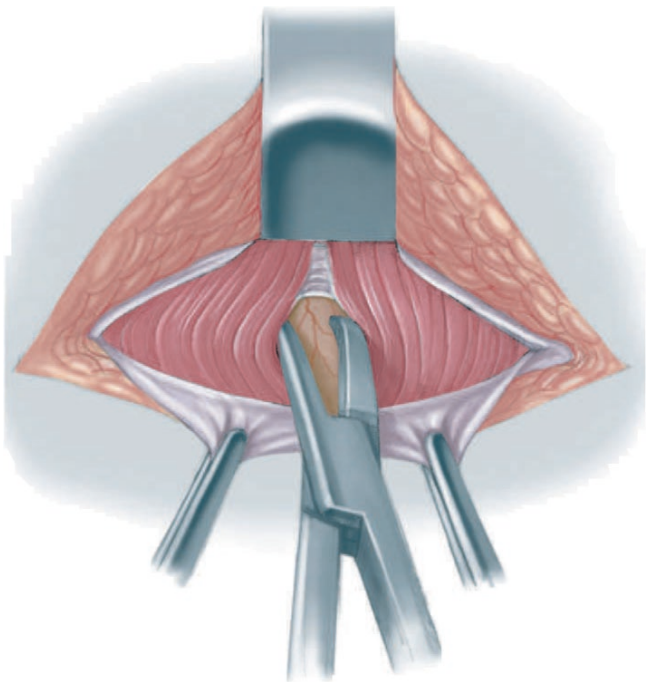

Fig. 61.4 Exposing the bladder

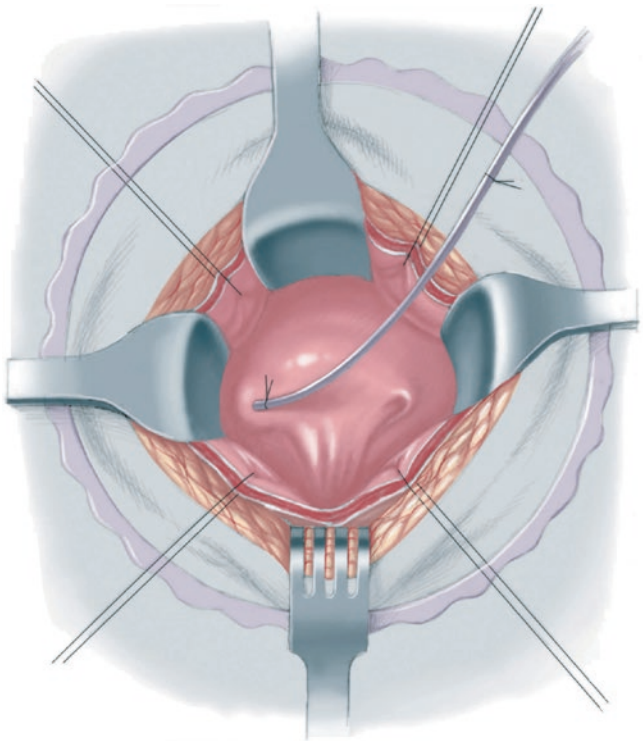

Fig. 61.5 Cannulation of the ureteral orifice, with ring retractor in place

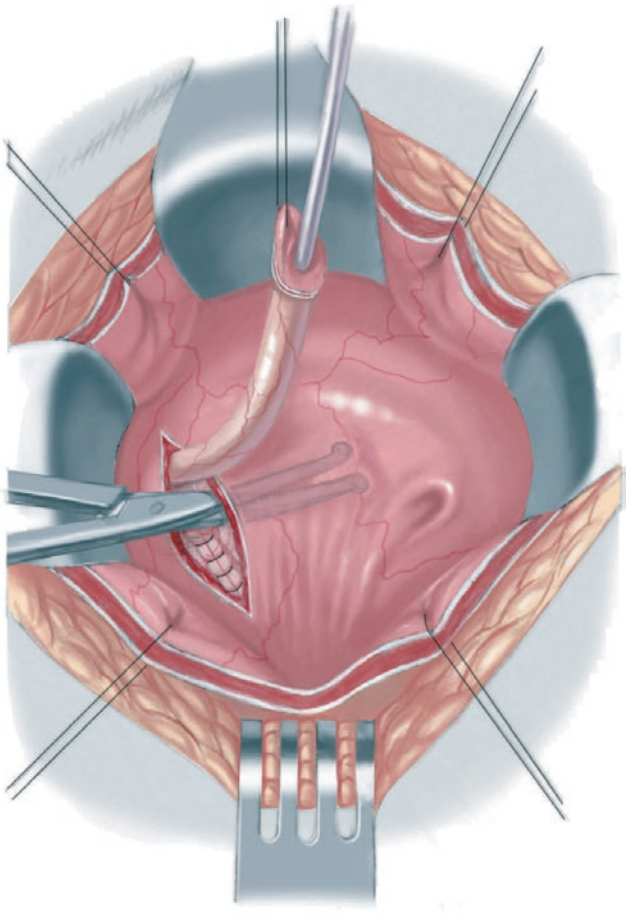

Fig. 61.7 Creating the submucosal tunnel

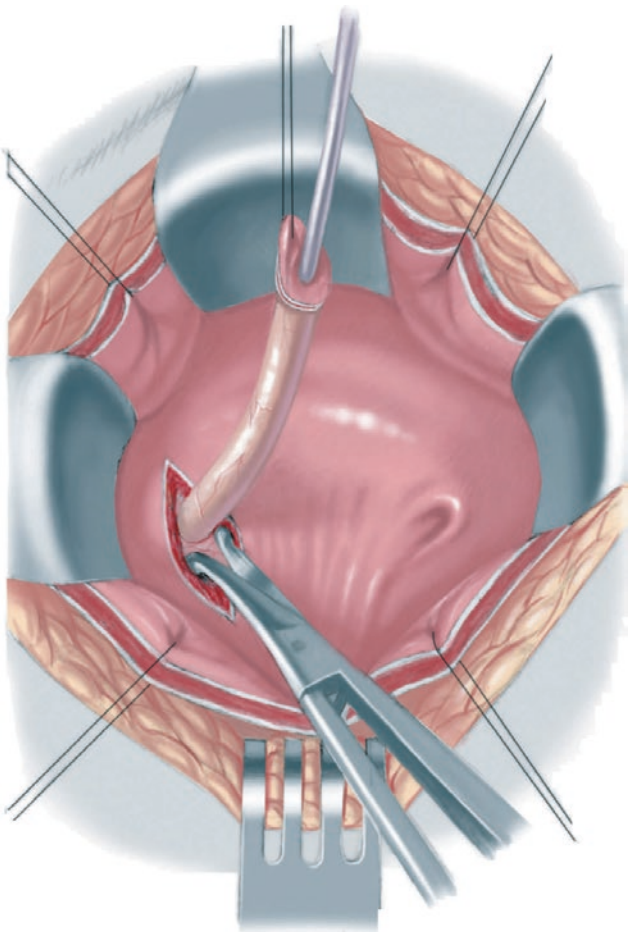

Fig. 61.6 Dissecting out the ureter

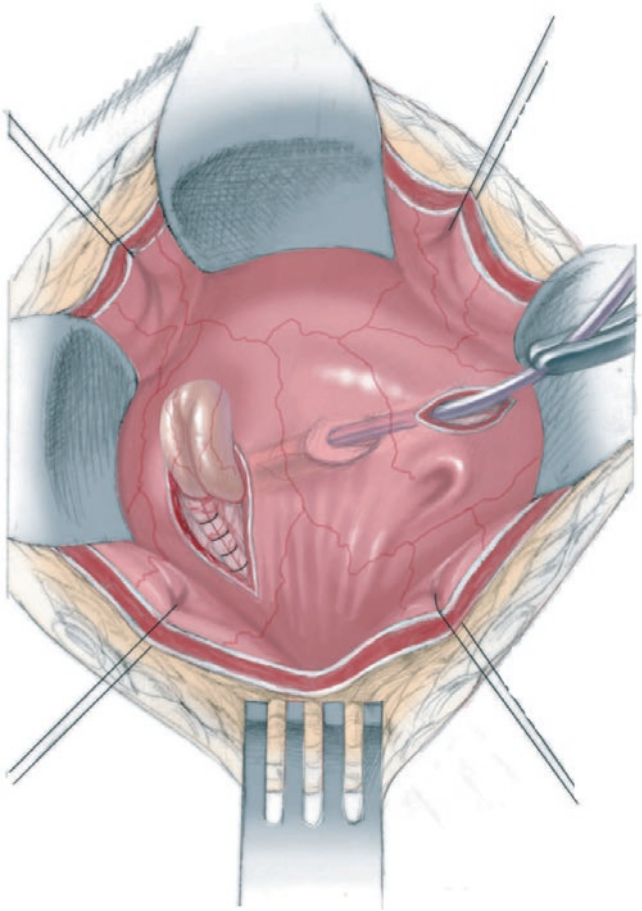

Fig. 61.8 Pulling the feeding tube through the submucosal tunnel

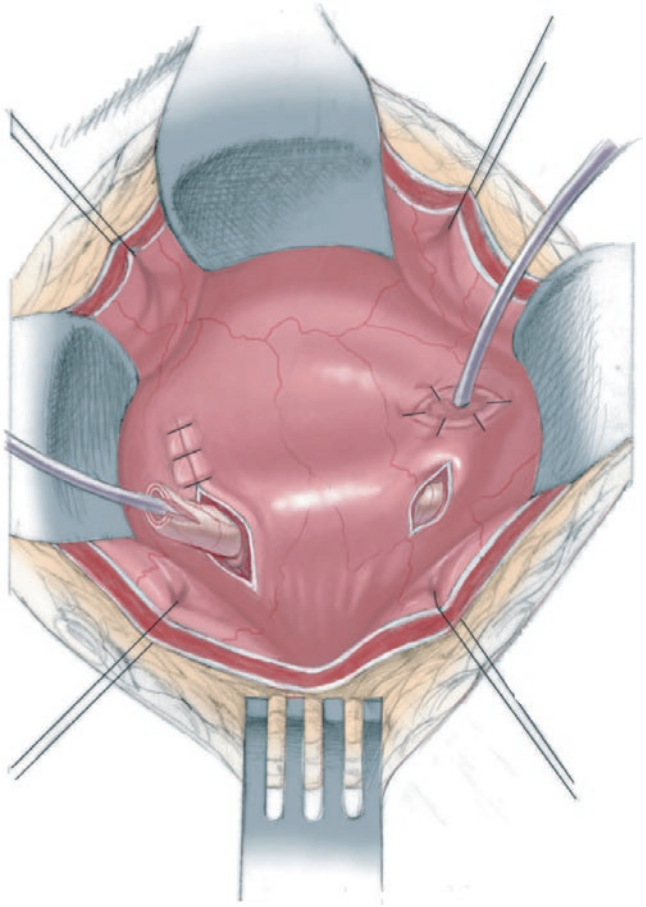

Fig. 61.9 Fixing the slightly spatulated ureter in place

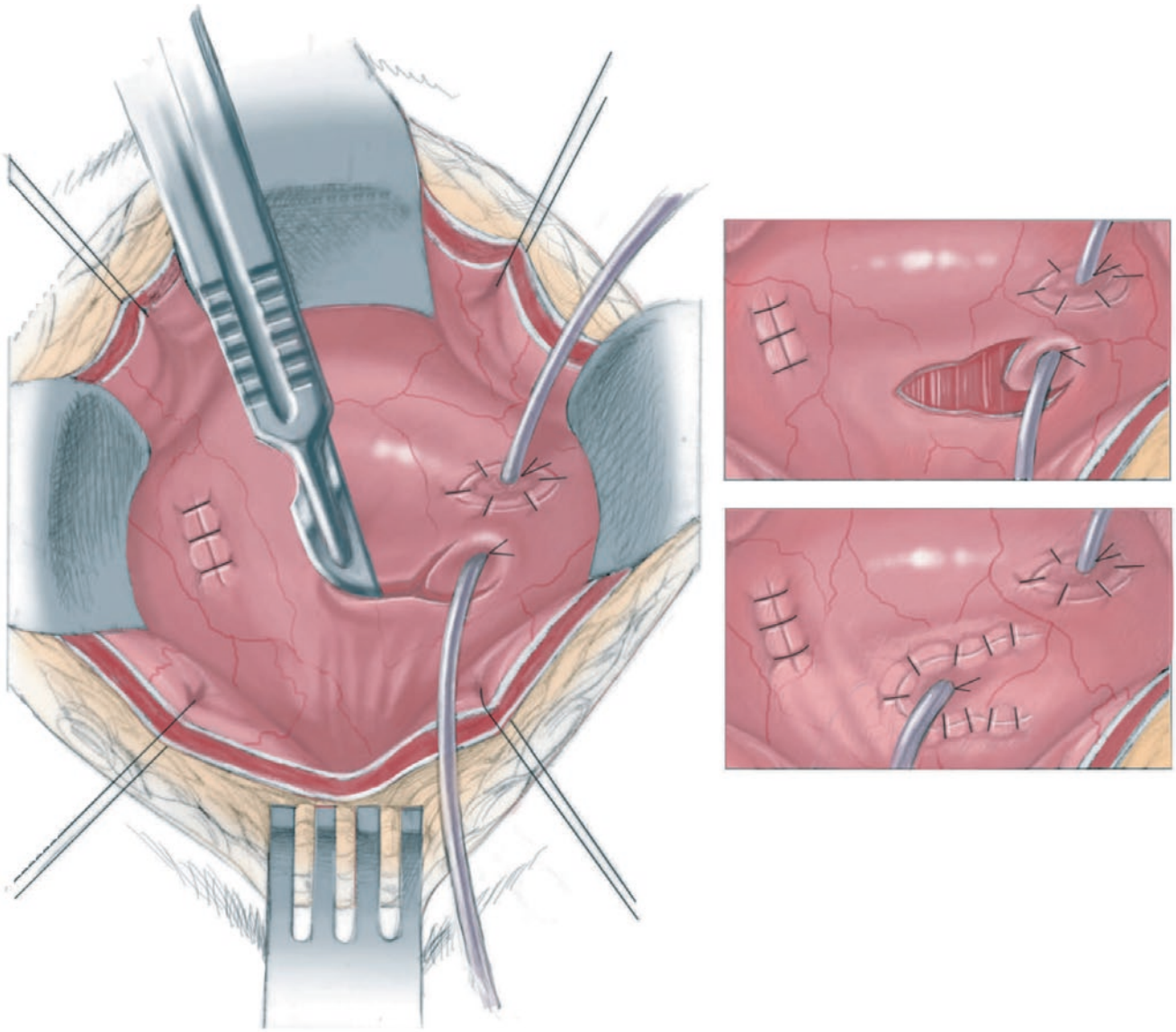

Fig. 61.10 Performing a contralateral Gil-Vernet ureteral reimplant (contralateral ureteral meatal advancement)

61.3.2 Detrusorrhaphy

The ureter may be reimplanted using an extravesical technique, termed *detrusorrhaphy*. This technique evolved from the Lich-Gregoir procedure. The success rate is identical to that of intravesical procedures. Following unilateral detrusorrhaphy, the incidence of contralateral VUR is less than 5%. With bilateral detrusorrhaphy, there is a small but significant risk of temporary (or even permanent) atonic bladder requiring clean intermittent catheterization. Consequently, many use this procedure only for unilateral reflux.

It is often helpful to perform cystoscopy and insert a ureteral catheter into the refluxing ureter(s). This manoeuvre facilitates identification of the ureter after the bladder is exposed.

A tremendous asset for this procedure is the robot retractor, which attaches to the operating table and holds retractors placed to retract the bladder and peritoneum, allowing exposure of the ureter.

The urethral meatus should be included in the operative field. A Foley catheter should be inserted at the beginning of the procedure and the bladder should be filled to a moderate degree manually.

For bilateral cases, the bladder should be exposed through a Pfannenstiel incision as described above. For unilateral cases, a unilateral 5-cm inguinal (modified Gibson) incision may be made.

The lateral wall of the bladder is mobilized by blunt dissection and muscular traction sutures are placed using 3/0 absorbable sutures. These traction sutures allow the bladder to be "rolled" medially, facilitating identification of the ureterovesical junction. The bladder may need to be emptied partially to facilitate this dissection. A Deaver retractor is inserted to retract the bladder medially. If the ureter is not immediately apparent, the obliterated umbilical artery is identified, ligated, and divided with 3/0 absorbable sutures. The ureter is just deep to the obliterated umbilical artery.

The ureter is isolated with a vessel loop (Fig. 61.11). By blunt dissection, the ureter is followed to its junction with the detrusor, termed the ureterovesical junction (UVJ). PGA 3/0 traction sutures are placed distal to the UVJ.

A right-angle clamp is inserted into the plane between the detrusor and bladder mucosa (Fig. 61.12), and the detrusor may be incised with the cautery. It is important to keep the cautery tip away from the mucosa. The junction of the ureter with the bladder mucosa is dissected out circumferentially in this manner.

The detrusor is separated from the mucosa inferomedially and incised with the cautery. A submucosal tunnel is developed superior to the hiatus for several centimetres, to a length four or five times as long as the ureteral width (Fig. 61.13). If the underlying bladder mucosa is cut inadvertently, interrupted 6/0 or 5/0 absorbable sutures should be placed through the open mucosal defect. Interrupted 3/0 absorbable traction sutures should be placed on either side of the detrusor incision. The bladder is emptied further and the ureteral catheter should be removed.

The ureter should be anchored inferiorly to stabilize the UVJ during bladder filling. One or two 4/0 PGA "U" stitches are placed from the distal detrusor muscle, proximally through the inferior edge of the UVJ, and distally through the detrusor (Fig. 61.14). These sutures are tied down.

The ureter is then laid into the trough created by opening the detrusor (Fig. 61.15), and the detrusor is brought together over it with interrupted 3/0 absorbable sutures (Fig. 61.16). The sutures should be tied down as they are placed.

Periodically a right angle clamp should be placed superficial to the intramural ureter to be certain that the tunnel is not too tight (Fig. 61.17). When the tunnel is completed, a suture should be placed between the detrusor muscle and the muscular layer of the ureter as it enters the tunnel, to prevent it from everting during bladder filling. The Foley catheter is then drained.

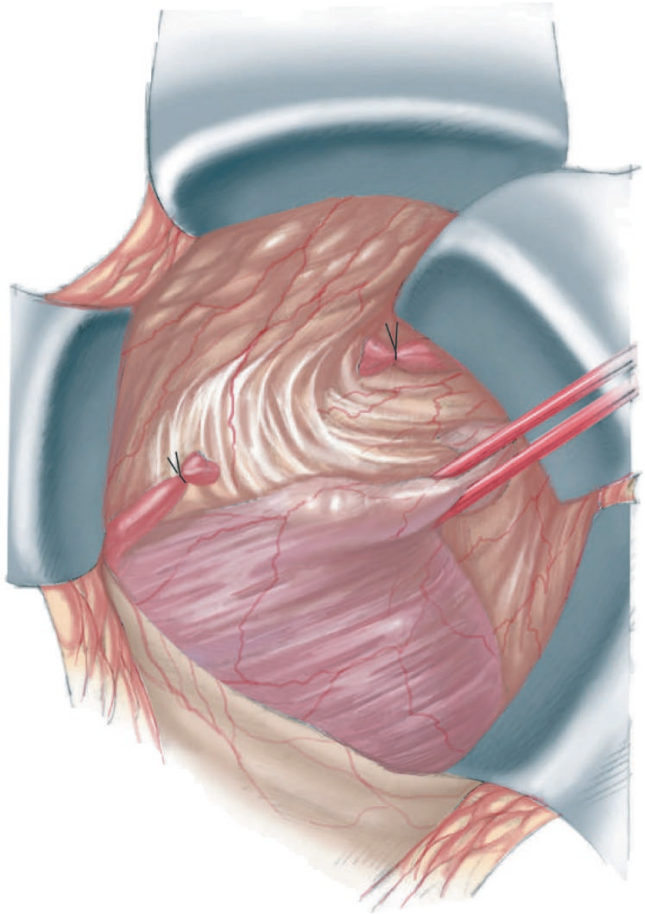

Fig. 61.11 Isolating the ureter with a vessel loop after division of the umbilical artery

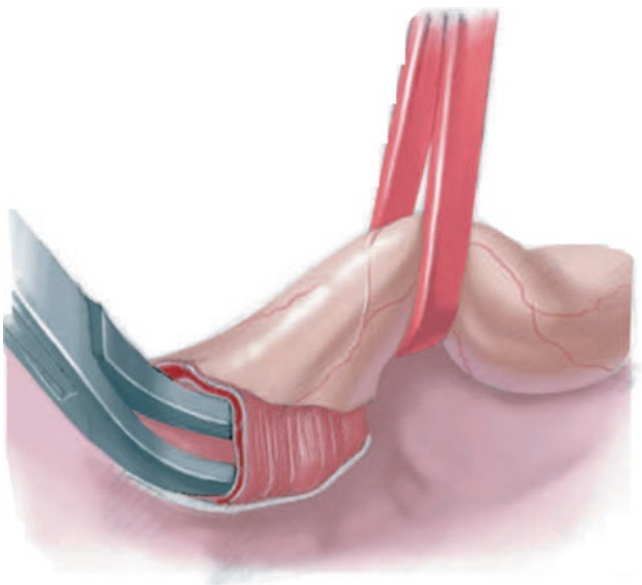

Fig. 61.12 Using a clamp to dissect out the junction of the ureter with the bladder mucosa

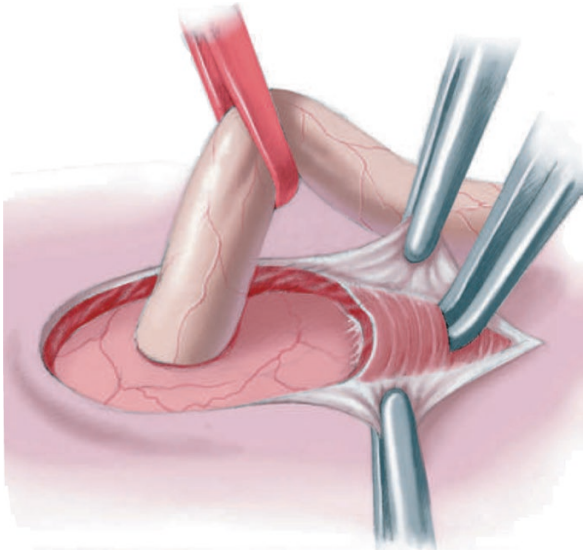

Fig. 61.13 Creating a submucosal tunnel

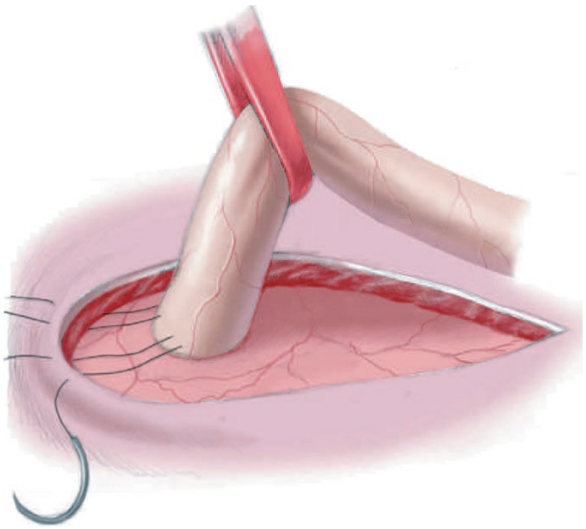

Fig. 61.14 Anchoring the ureter inferiorly

Fig. 61.15 Laying the ureter into the trough

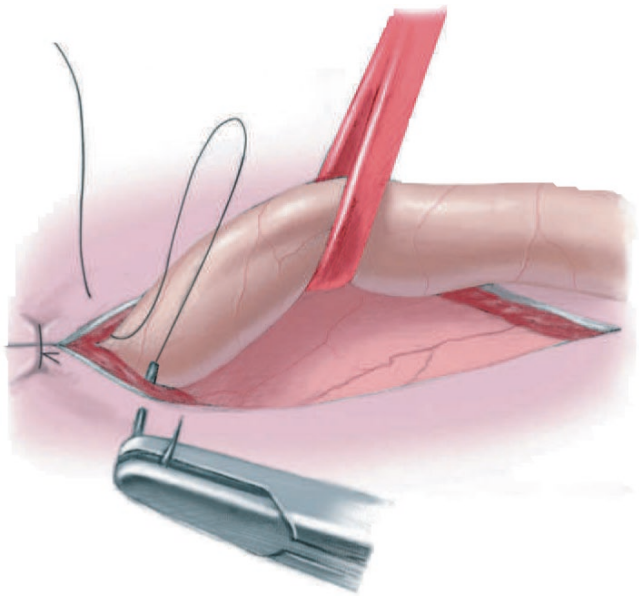

Fig. 61.16 Bringing the detrusor together over the ureter

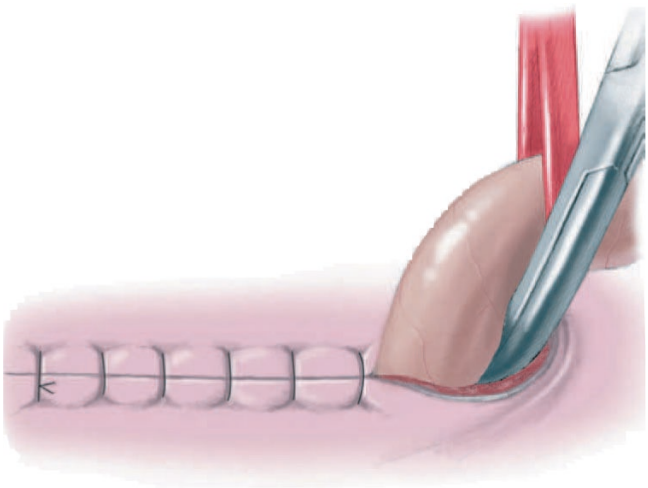

Fig. 61.17 Ensuring that the tunnel is not too tight

61.3.3 Politano-Leadbetter (P-L) Procedure

This technique is another form of intravesical antireflux surgery. It may also be performed as a combined intravesical/extravesical procedure. The operation involves creating a new ureteral hiatus superiorly in the bladder and bringing the ureteral opening near its original location.

The bladder is opened and the ureters are mobilized identically as described above. A vein retractor or small Army-Navy retractor is placed in the medial wall of the hiatus. The peritoneum is teased away with either a large right angle clamp or a Kitner dissector (Fig. 61.18, *top*). A new position for the hiatus should be made in a fixed portion of the bladder base several centimetres superior to the original hiatus.

From outside the bladder, the right angle clamp is used to indent the bladder, the clamp is opened slightly, and the overlying bladder mucosa is cauterized, exposing the tip of the clamp (Fig. 61.18, *bottom*). The right-angle clamp is then opened to create a new hiatus of satisfactory size.

A second right-angle clamp is passed from the inside of the bladder outside through the new hiatus, the feeding tube in the ureter is grasped (Fig. 61.19), and the ureter is brought into the bladder. It is important for the ureter to travel in a relatively straight direction.

At times it is necessary to perform an extravesical dissection also. If so, the Denis Browne retractor must be taken out and the outside wall of the bladder retracted medially. The obliterated umbilical artery should be identified; it is a firm white structure extending from the dome of the bladder toward the hypogastric artery. The artery is ligated and divided with 3/0 absorbable suture. The bladder may then be mobilized further. Beneath the obliterated umbilical artery is the ureter. This extravesical dissection facilitates the establishment of a new hiatus with minimal risk of bowel injury.

After the ureter is brought into the bladder, the original hiatus should be closed with three or four 3/0 absorbable sutures placed through the detrusor.

A submucosal tunnel is created. Tenotomy scissors are used to incise the mucosal attachment off the underlying detrusor in the old hiatus, and then the submucosal tunnel is created toward the new hiatus by gently spreading the tenotomy scissors between the mucosa and detrusor. The width of the tunnel should be approximately twice as long as the ureteral width, and the length is four to five times as long as the width.

When the new hiatus is reached, a right-angle clamp is passed through the tunnel and the feeding tube is grasped. The submucosal tunnel may be extended distally toward the bladder neck if necessary to create a tunnel of sufficient length. The ureter is pulled through the new submucosal tunnel. The feeding tube should be removed and the distal aspect of the ureter resected. The ureter is then spatulated slightly.

With the feeding tube in place, the ureter is sutured to the bladder mucosa with interrupted 5/0 or 6/0 absorbable

sutures; the distal apical suture should be placed through the bladder muscle also, to help fix the ureter in place. There should be no tension on the ureter. Small mosquito clamps are placed for traction on the two apical sutures to allow easy identification of the new ureteral orifice. The feeding tube should be removed and then reinserted into the ureter; the feeding tube should pass easily into the kidney. It is unnecessary to leave a feeding tube in the ureter postoperatively. After the ureter(s) is fixed in place, the bladder mucosa is closed with running 5/0 absorbable sutures (Fig. 61.20). The bladder is then closed as described in the above section.

The success rates of the P-L and the Cohen (transtrigonal) techniques are similar. The advantage of the P-L is that the ureter is much easier to catheterize for retrograde pyelography and ureteral endoscopy because the ureteral opening of the Cohen is on the opposite side of the bladder. The disadvantage is that in creating the new ureteral hiatus, there is a blind spot behind the bladder, and a peritoneotomy or even bowel injury may occur, particularly in reoperative cases.

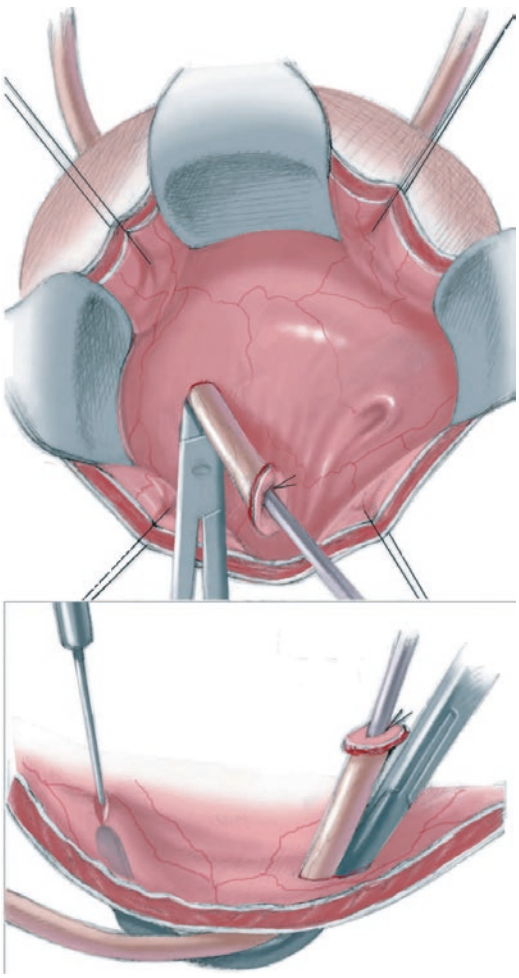

Fig. 61.18 Teasing away the peritoneum (*top*) and creating a new hiatus (*bottom*)

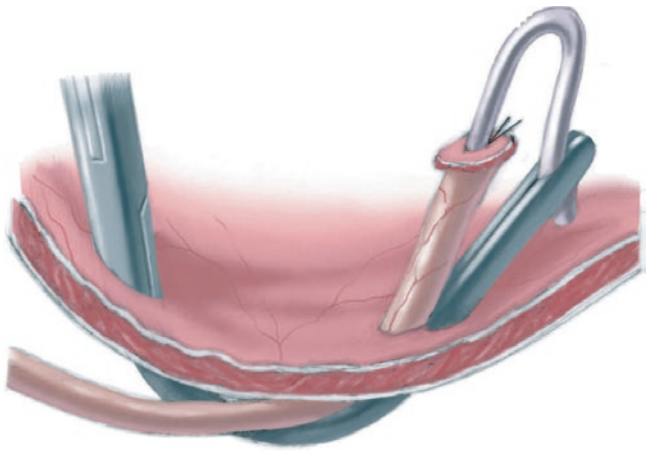

Fig. 61.19 Preparing to bring the ureter into the bladder

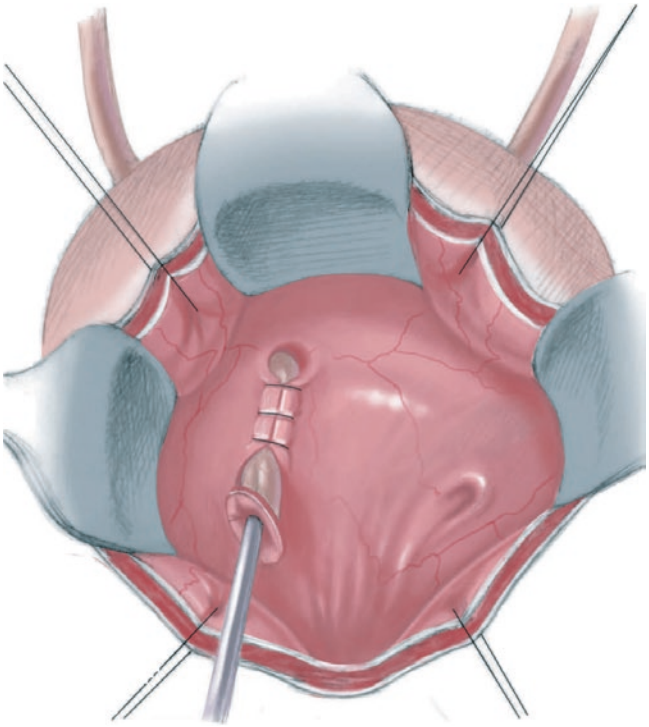

Fig. 61.20 Completing the P-L procedure

61.3.4 Laparoscopic/Robotic Extravesical Ureteral Reimplantation

The child is placed in modified lithotomy position with Allen stirrups. One setup is used. Cystoscopy is performed and the refluxing ureter(s) is cannulated with a 4F ureteral catheter. A Foley catheter is inserted and the bladder is drained. The Foley and ureteral catheter(s) are kept in the sterile field. The child is placed in Trendelenburg position. The camera port is inserted at the umbilicus and 5-mm ports are placed at the level of the anterior superior iliac spine just lateral to the rectus muscle. A 5-mm assistant port is placed lateral to the 5-mm port, contralateral to the refluxing ureter. Maryland grasping forceps and a hook cautery are used. In girls, the ureter is identified in the pelvis just inferior to the fallopian tube. The ureter is mobilized inferior to the uterine artery to its junction with the bladder. An attempt should be made to identify the pelvic plexus, particularly in children undergoing bilateral extravesical ureteral reimplantation. The tissue plane between the medial aspect of the ureter and the internal genitalia is bluntly dissected from the ureter. The ureter is then retracted medially, and the tissue medial and caudal to the ureter is retracted laterally and anteriorly, to expose the nerves from the pelvic plexus. The ureter is then dissected circumferentially from the detrusor, keeping the nerves in sight and away from the dissection.

The bladder is distended with 50–100 mL of saline; some use carbon dioxide to distend the bladder. The detrusor is incised just anterior to the ureterovesical junction. Using the hook dissector, the detrusor muscle is incised around the ureter down to the level of the mucosa (Fig. 61.21). A submucosal tunnel four or five times as long as the width of the ureter is then planned. A 3–0 or 4–0 Vicryl "hitch" stitch on an RB-1 needle is placed percutaneously through the abdominal wall. It is placed through the detrusor just above the planned submucosal tunnel. The detrusor is incised along this pathway down to the level of the mucosa. Lifting the muscle fibers and then cauterizing is useful. If the mucosa is incised inadvertently, it is closed with a 6–0 Vicryl on an RB-1 needle. The tunnel should be sufficiently wide to allow the ureter to be placed comfortably against the mucosa. The ureteral catheter is removed. The ureter is lifted and can be fixed to the tip of the submucosal tunnel with a 4–0 Vicryl stitch. Next, the ureter is pushed into the mucosa and the seromuscular layer at the UVJ is fixed to the detrusor with a 4–0 Vicryl stitch. This suture is left long; the assistant can grasp it for traction. The assistant places tension on the external detrusor hitch stitch and the stitch at the ureterovesical junction, which creates a diagonal or horizontal submucosal tunnel to close. The detrusor is closed over the ureter with a running or interrupted 3–0 or 4–0 Vicryl suture (Fig. 61.22). It may be useful to place the distal stitch through the seromuscular layer of the ureter to fix it in place. The bladder is drained.

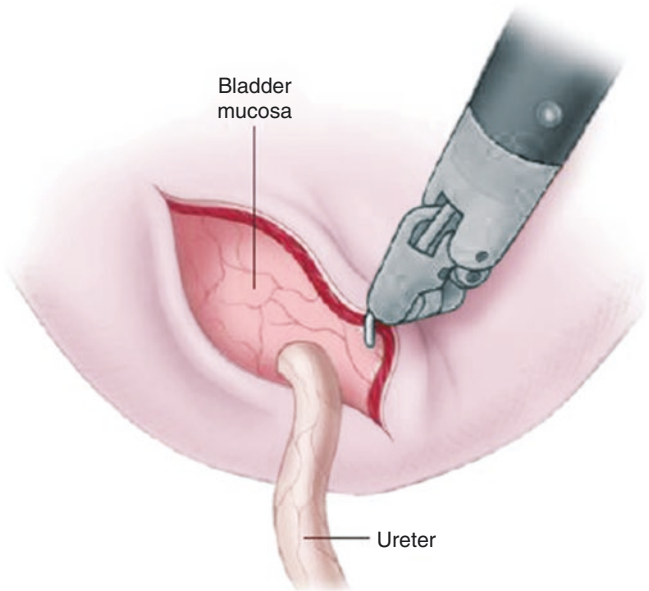

Fig. 61.21 Robotic extraperitoneal exposure of the ureterovesical junction

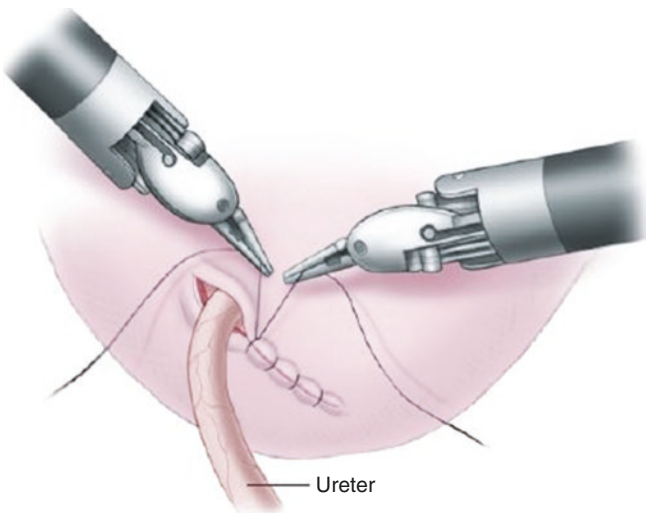

Fig. 61.22 Robotic closure of the ureterovesical junction

61.3.5 Ureteral Tapering (Tailoring)

When the refluxing ureter is too wide to reimplant it into the bladder with a 4:1 or 5:1 length:width ratio, the ureter should be narrowed. The most common surgical technique is with ureteral tapering, in which the ureteral wall is excised. Tapering the ureter is performed on its lateral aspect, because the main collateral blood supply is along its medial wall. Further, when the suture line for tapering is closed, it should be placed posteriorly next to the detrusor muscle when the ureter is reimplanted into the bladder, thus reducing the likelihood that a fistula into the bladder will form. Using sharp, straight scissors, the strip of ureter is removed; it must not be too wide (Fig. 61.23). Trimming the ureter excessively will make it too narrow and jeopardize its blood supply. The ureteral closure is performed with a running, imbricating 4–0 absorbable suture. The dilated ureter is long, and part of the distal end will be excised. When approximately two thirds of the ureteral closure has been performed, the remaining closure should be performed with interrupted sutures, to avoid having to transect part of the tapered ureter that was closed with a running suture. The closure should be performed over an 8F or 10F catheter. The ureter should be implanted with at least a 3:1 or 4:1 length-to-width ratio, using any of the techniques described above. In selected cases, the superolateral corner of the bladder may be fixed to the psoas muscle (termed a psoas hitch) to facilitate a straight ureter, avoiding ureteral kinking. A temporary double-J stent should be left.

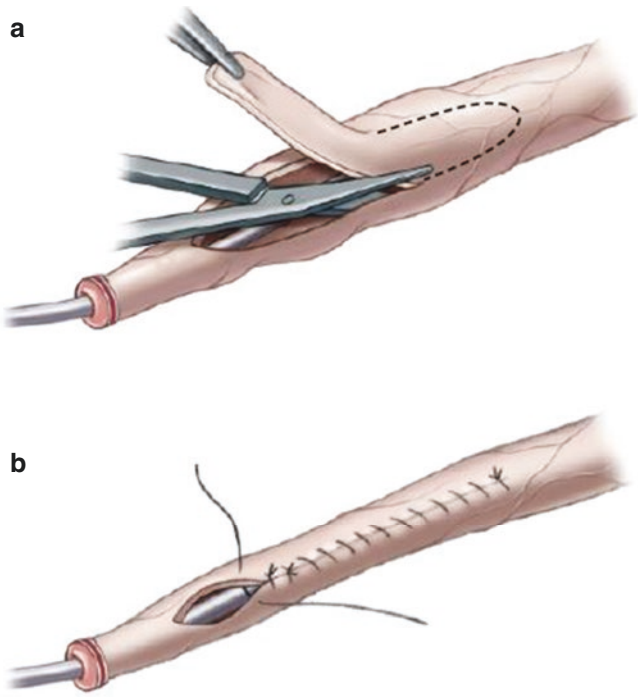

Fig. 61.23 (**a**) Excision of ellipse of ureteral wall over catheter to reduce its diameter. (**b**) Closure of ureter with running absorbable suture proximally and interrupted sutures distally

61.4 Postoperative Management

The most common complications of extravesical ureteroneocystostomy are persistent VUR and ureteral obstruction. Persistent VUR results if the submucosal tunnel is too short or if the ureter is not laid against the bladder mucosa. Ureteral obstruction may result if the ureter sustains a thermal injury during mobilization, if the submucosal tunnel is not straight, or if there is undiagnosed distal ureteral narrowing. An atonic or hypotonic bladder may occur following bilateral extravesical ureteroneocystostomy if the ureters are overmobilized near the pelvic plexus. In these children, a urethral catheter should be left for 4–7 days.

Urine output is measured in the post-anaesthesia care unit and the patient should be hydrated until the output is 1–2 mL/kg per hour. Pain control is established: (1) A caudal block is performed at the beginning and the end of the procedure; (2) Intravenous ketorolac is administered at a dosage of 0.5 mg/kg (maximum 30 mg) during wound closure and is continued at a dosage of 0.25 mg/kg every 6 h for 48 h; and (3) Intravenous morphine is administered at 0.1 mg/kg every 3 h, or a patient-controlled analgesic (PCA) pump can be used for children over 6 years. A regular diet may be prescribed. The Foley catheter is removed the day following the surgical procedure. Discharge is appropriate when the patient is comfortable and afebrile. Following intravesical ureteroneocystostomy, the child may experience moderate or significant bladder spasm, and oral administration of oxybutynin chloride three times daily for 10–14 days often is helpful.

The child should continue to take prophylactic antibiotics for 4 weeks after surgery, at which time a renal sonogram should be performed. Whether to perform a postoperative VCUG depends on the surgeon's experience. Because the success rate of ureteroneocystostomy is over 95%, many surgeons choose to perform a postoperative VCUG only if the child has a febrile UTI suggestive of pyelonephritis or if there is hydronephrosis suggestive of obstruction or persistent reflux.

61.5 Results and Conclusions

The goal of surgical correction of VUR is to minimize the risk and complications of upper tract infection, including new renal scarring, reduced renal function, impaired somatic growth, and complications of pregnancy. In the International Reflux Study, medical and surgical therapy was compared for grades III and IV VUR. The incidence of new renal scarring (approximately 15%) was similar between the two groups, but the incidence of pyelonephritis was 2.5 times higher in the medical group. In the European arm, many of the surgical patients who experienced complications were not operated on by full-time pediatric urologists; no surgical morbidity occurred in the United States arm. More contemporary series with high surgical success rates have shown that the incidence of new renal scarring is probably around 1–2%. The RIVUR Trial demonstrated a benefit of antibiotic prophylaxis in children with VUR, but did not address the role of surgical treatment. Ureteral reimplantation continues to be indicated for children with VUR when medical therapy fails.

The success rate for ureteroneocystostomy is generally over 95% for grades I to IV VUR, regardless of technique. Consequently, many surgeons do not perform a routine postoperative VCUG unless the child develops an upper tract UTI; instead, they monitor their patients with serial renal sonograms. Even with successful surgical correction, however, approximately 5–10% will develop a febrile UTI over the following 10 years.

The late sequelae of ureteroneocystostomy continue to be studied. A disadvantage of the transtrigonal technique is that subsequent endoscopic ureteral manipulation is difficult if an upper tract ureteral calculus occurs, whereas the ureteral orifice is in normal position with the detrusorrhaphy and Politano-Leadbetter techniques.

Suggested Reading

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Ureteric Duplication

62

Angela M. Arlen and Andrew J. Kirsch

Ureteric duplication is one of the most common malformations of the urinary tract, affecting 1 in 125 children (0.8%). The combination of routine prenatal ultrasound and improving sonographic technology has resulted in increased detection of organ anomalies, and urologic abnormalities are among the most commonly reported findings. Ureteral duplication may be associated with ectopia or ureterocele, as well as corresponding vesicoureteral reflux (VUR) or hydronephrosis, but it is also compatible with a normally functioning collecting system in cases of partial duplication or when both ureters enter the bladder in an orthotopic location.

Duplication can be complete or partial. A bifid renal pelvis includes only a single ureter, but confluence of the upper and lower ureters below the ureteropelvic junction (UPJ) and above the bladder constitutes partial duplication of the ureter or bifid ureter. Partial duplication does not predispose to VUR, and surgical correction is based on the same indications as a single refluxing ureter. In contrast, complete ureteric duplication may be associated with UPJ obstruction, VUR, ureteral ectopia, and/or ureterocele.

62.1 Associated Anomalies

Conditions that routinely affect the single-system kidney are those that affect the lower pole, including UPJ obstruction and VUR. The upper pole is more commonly affected by conditions secondary to abnormal ureteral formation, including ectopia and ureterocele. The evaluation and management of lower pole UPJ obstruction is similar to that of single-system UPJ obstruction. Multiple operative possibilities exist for reflux in ureteric duplication, depending on associated anomalies and whether the VUR affects the lower pole ureter or both ureters. Therapeutic considerations for surgical correction of VUR associated with duplication are similar to those for a single refluxing ureter, though spontaneous resolution is less likely to occur. Nuclear scintigraphy or magnetic resonance urography (MRU) with differential function of the upper and lower pole moieties may be performed to assist with operative planning.

62.1.1 Ureteral Ectopia

Ureteral ectopia exists when the ureteric orifice is located outside the bladder. Ectopia is associated with duplication in approximately 80% of cases, particularly in females. In males, the ectopic ureter is more likely to be associated with a single collecting system. When associated with a duplex system, the upper pole parenchyma related to the ectopic ureter often has minimal function, and dysplasia is frequently present. In such cases, preservation of the upper pole does not contribute significantly to total renal function, and it is likely to remain a source of infection.

62.1.2 Ureterocele

Ureteroceles are congenital cystic dilatations of the submucosal terminal ureter within the detrusor muscle. As with ureteral ectopia, ureteroceles can be found in either single or duplex systems; in duplicated systems, they are associated with the upper pole moiety. The position of the ureteric orifice allows for classification of intravesical (simple) and extravesical (ectopic) ureterocele—an important distinction,

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as management may differ. The intravesical ureterocele is entirely within the bladder and above the bladder neck. In the extravesical ureterocele, the orifice is located at the bladder neck or within the urethra.

62.2 Treatment Options

The main goals of ureterocele management include prevention of renal damage associated with obstruction or VUR and urinary tract infection (UTI), maintenance of continence, and minimizing surgical complications with the fewest number of procedures possible. Voiding cystourethrogram is essential in the evaluation of ureterocele because of the high incidence of concomitant ipsilateral (50%) and contralateral (25%) VUR. Optimal management of ureterocele remains controversial; options include observation, endoscopic ureterocele puncture, upper pole nephrectomy, high or low ureteroureterostomy, and excision of the ureterocele with ureteral reimplantation. The surgical approach is dependent upon the clinical scenario (patient age, ureterocele size and anatomy, presence of concomitant VUR, infection, and function of involved renal moieties) as well as surgeon and parental preference.

A conservative, nonoperative approach may be considered in select cases. Observation may be employed in those with a nonfunctioning, dysplastic upper pole, and in those with a normal upper pole as long as there is no evidence of obstruction or infection. In neonates with sepsis, preliminary decompression may be necessary, and simple endoscopic incision of the ureterocele is advisable. Transurethral ureterocele puncture may also be indicated in infants with uninfected ureteroceles discovered on prenatal ultrasonography, to allow for preservation or recovery of satisfactory renal function. Upper pole heminephrectomy may be clinically indicated when the upper pole is nonfunctioning, and associated with UTI in the absence of high-grade lower pole VUR. When obstruction of the upper pole moiety is the only feature encountered with duplex system ureteroceles, anastomosis between the upper and lower pole ureters may be an appealing approach, although risk to the more normal lower pole moiety remains a rare but distinct possibility. A total, single-stage complex reconstruction includes complete excision of an ectopic ureterocele with ureteral reimplantation of the lower pole ureter as well as well as upper pole nephroureterectomy, with care taken to preserve the lower pole ureteral blood supply. In rare cases, a total nephroureterectomy is indicated when the lower pole has minimal function owing to obstruction or reflux nephropathy.

62.3 Surgical Procedures

62.3.1 Reimplantation of an Ectopic Ureter

Through a Pfannenstiel incision, the bladder is exposed and a self-retaining retractor is placed. A combination of extravesical and intravesical dissection results in adequate mobilization, allowing for a 5:1 ratio of submucosal tunnel to ureteral diameter. As both ureters are bound together distally in a common sheath, they are treated en bloc as a single unit in order to prevent devascularization (Fig. 62.1). A common sheath ureteral reimplantation is performed, most commonly using Cohen cross-trigonal or Politano-Leadbetter techniques (Fig. 62.2). In some cases, the upper pole ectopic ureter may be distinct from the lower pole ureter and can be reimplanted while leaving a nonrefluxing lower pole ureter intact. Open bladder or extravesical approaches (Lich-Gregor) may be performed. Ureteral stenting and the need for postoperative bladder drainage depends upon clinical factors such as patient age, ureteral tapering, or whether the ureter is dismembered, as well as surgeon preference.

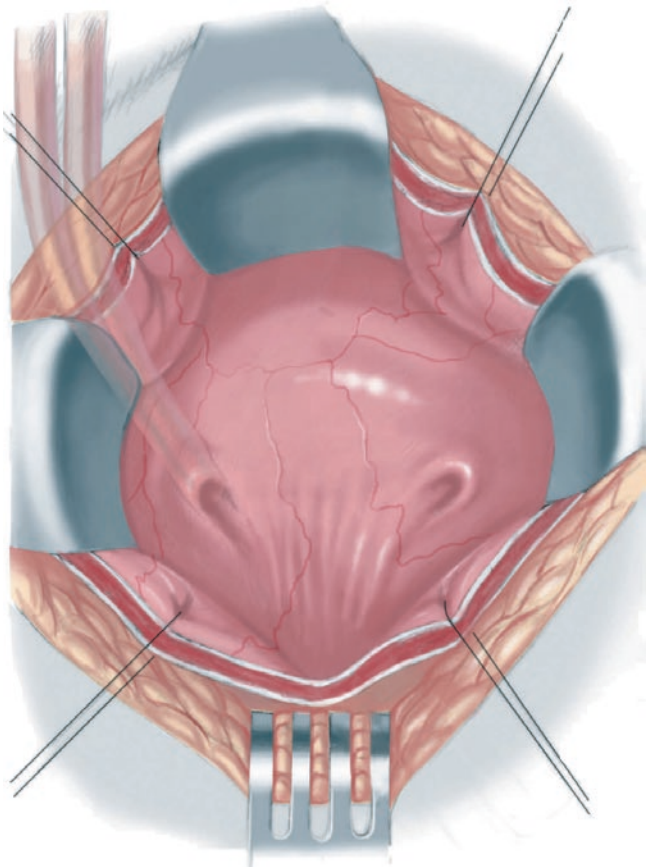

Fig. 62.1 Complete ureteral duplication with a common sheath ureteral reimplantation

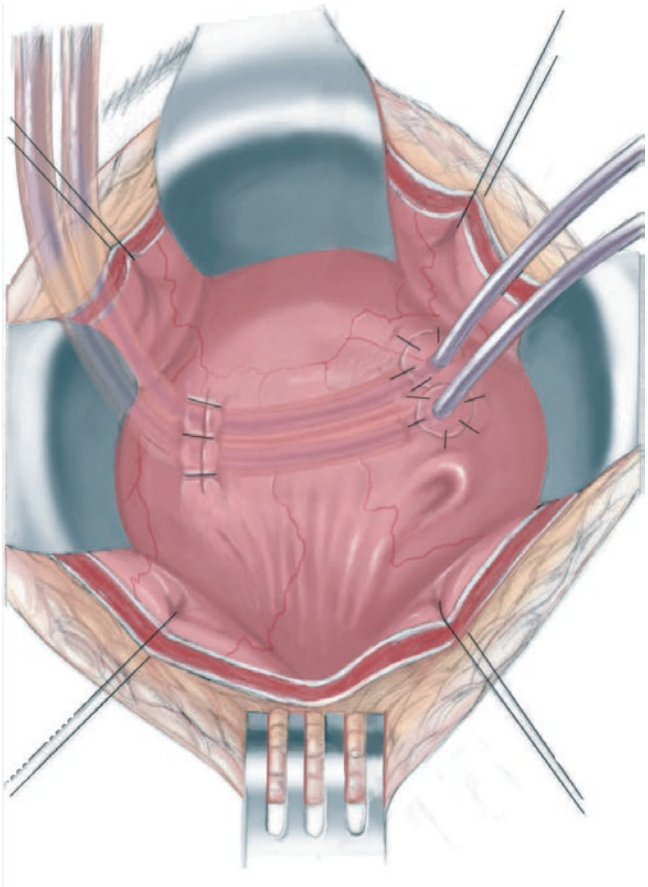

Fig. 62.2 Common sheath ureteral reimplantation of only the upper pole ectopic ureter

62.3.2 Ureteroureterostomy or Ureteropyelostomy

Ureteroureterostomy or ureteropyelostomy can be considered when obstruction of the upper pole moiety is the only feature encountered with a duplex system. This can performed via a flank, dorsal lumbotomy, or laparoscopic/robotic approach. Mobilization is directed towards the upper pole ureter, in order to avoid distortion of the adjacent lower pole ureter. The upper pole ureter is often dilated, necessitating generous spatulation of the ureters to allow for an appropriate caliber end-to-side anastomosis. When performing an ureteropyelostomy, the excess abnormal ureter is excised. A large end-to-side anastomosis between the upper pole ureter and lower pole renal pelvis is performed (Fig. 62.3), mimicking a bifid renal pelvis without functional obstruction. The anastomosis should be stented for several weeks.

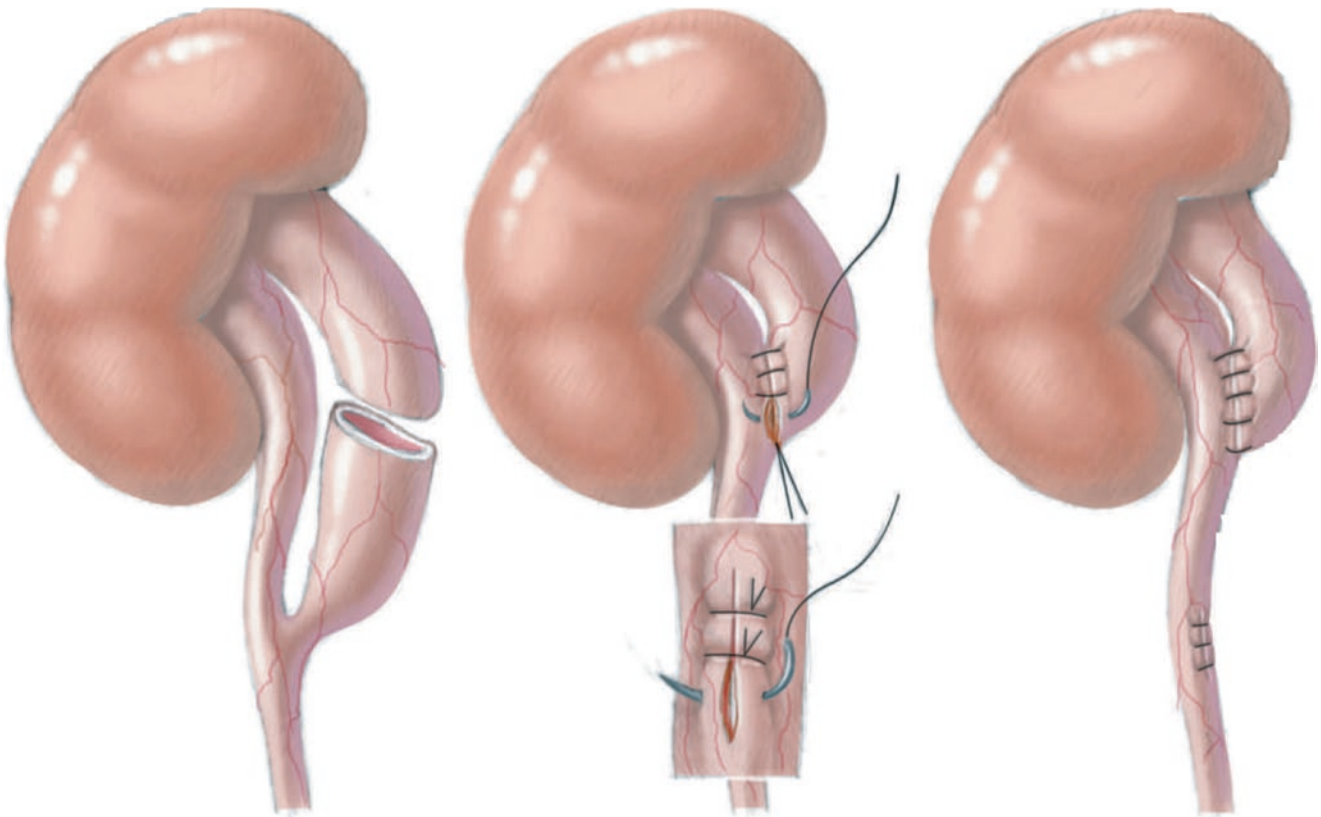

Fig. 62.3 Ureteropyelostomy

62.3.3 Upper Pole Heminephrectomy

Upper pole heminephrectomy can be performed via a subcostal, flank, or laparoscopic/robotic approach. The dysplastic upper pole moiety and the upper pole ureter are excised (Fig. 62.4). The renal capsule of the upper pole is incised and later utilized to oversew the renal parenchyma. A clear demarcation is often visible between the nonfunctioning upper pole and the normal lower pole parenchyma. It is preferable to follow this cleavage plane regardless of surgical approach (Fig. 62.5). Vascularization of the upper pole is quite variable. Upper pole arteries may separate from the main artery close to the renal sinus, making clamping of these vessels potentially hazardous, as it can result in ischemia and eventual atrophy of the remaining lower pole renal segment.

Heminephrectomy is completed by suturing the parenchyma of the remaining lower pole, if indicated, and then closing the capsule (Fig. 62.6). In cases with a completely non-functioning upper pole and obvious dissection plane, suturing the remnant renal parenchyma may not be necessary.

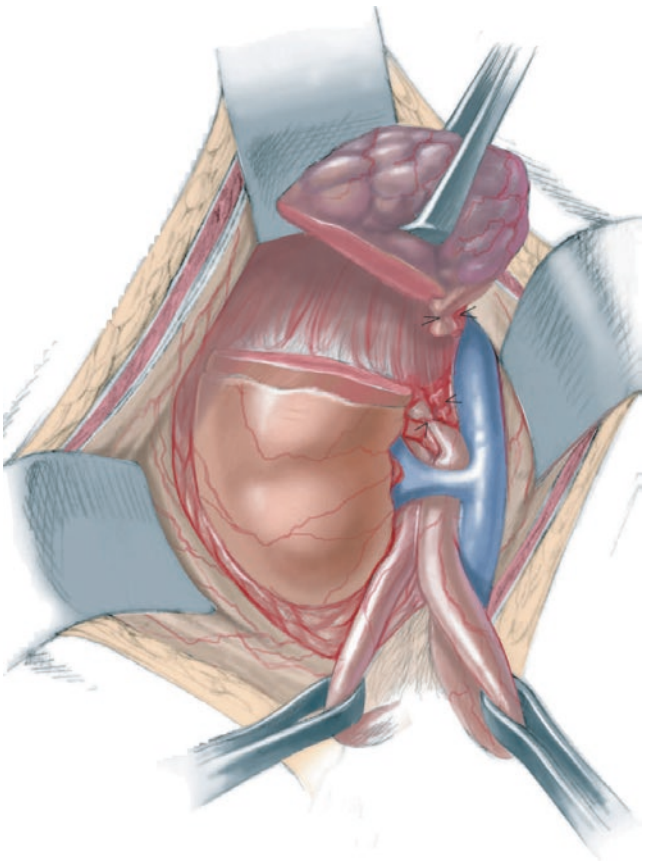

Fig. 62.5 Cleavage plane separating nonfunctioning and normal renal parenchyma

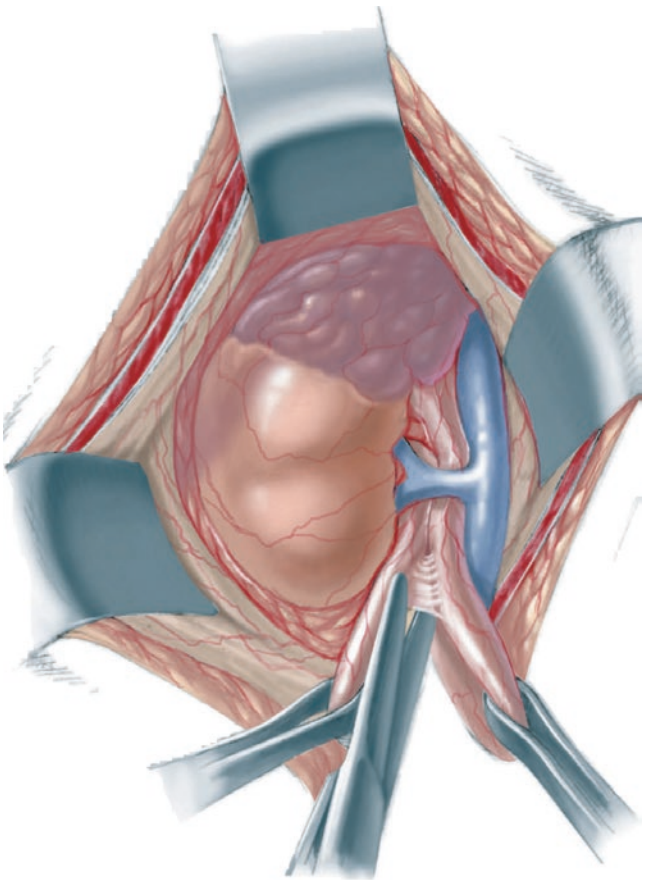

Fig. 62.4 Excising the upper pole moiety and the upper pole ureter

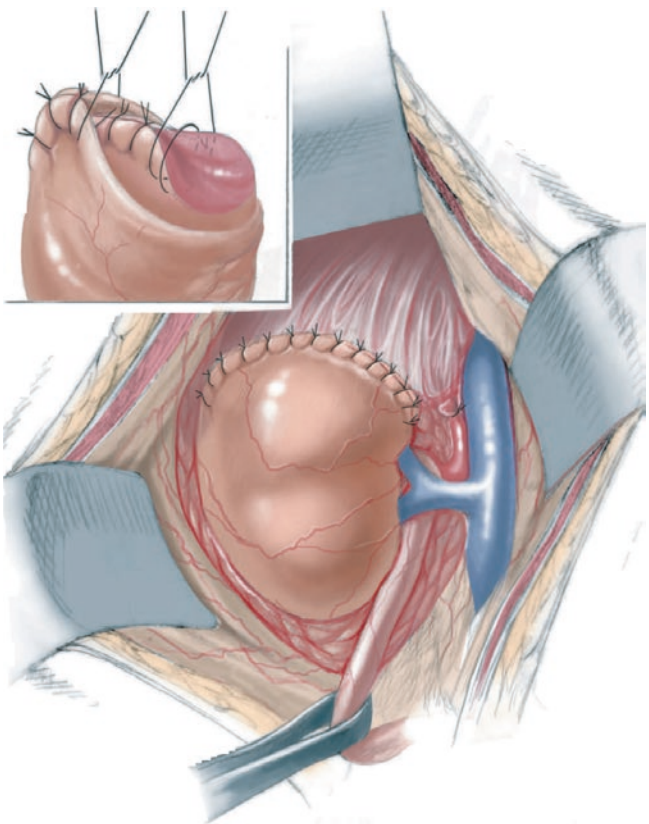

Fig. 62.6 Suturing the remnant renal parenchyma

62.3.4 Endoscopic Ureterocele Puncture

A *duplex system ureterocele* is when the ureterocele is associated with the upper pole ureter of a completely duplicated collecting system and a *single system ureterocele* is when the ureterocele is attached to a single ureter draining the kidney. If the ureterocele is located entirely within the bladder, the term *intravesical* or *simple* is applied. If the ureterocele and its orifice extend beyond the trigone to the bladder neck or urethra, the term *ectopic* or *extravesical* is used. Intravesical ureteroceles are usually associated with single systems, whereas ectopic ureteroceles are often associated with duplex systems. In rare cases, a single system ureterocele may be associated with an ipsilateral nonfunctioning, multicystic, dysplastic kidney and may require total nephrectomy.

The goal of endoscopic puncture is to decompress the ureterocele in a minimally invasive fashion while minimizing the risk of post-puncture VUR and the need for further urinary tract reconstruction. Management of the orthotopic ureterocele is straightforward. The ureterocele is punctured using pure cutting current at the leading edge of the ureterocele, just above its junction with the bladder (Fig. 62.7). As the Bugbee electrode is passed through the ureterocele wall, it may be moved slightly laterally in either direction to enlarge the opening. One may readily observe deflation of the ureterocele. In some cases, IV fluid bolus with or without Lasix and manual compression of the flank over the hydronephrotic kidney is helpful in distending the ureterocele prior to puncture.

A similar technique is employed for an ectopic ureterocele. The puncture is made at the base of the ureterocele in an intravesical location, with care taken to avoid the lower pole and contralateral ureteral orifices. Because the ureterocele may extend distally beyond the bladder neck, the location of the puncture must be clearly visualized (Fig. 62.7). Adequacy of puncture can be confirmed by the presence of a urine jet, or visualization of urothelium within the ureterocele. Additional procedures are more likely to be needed after puncture of ectopic ureteroceles than for simple ureteroceles.

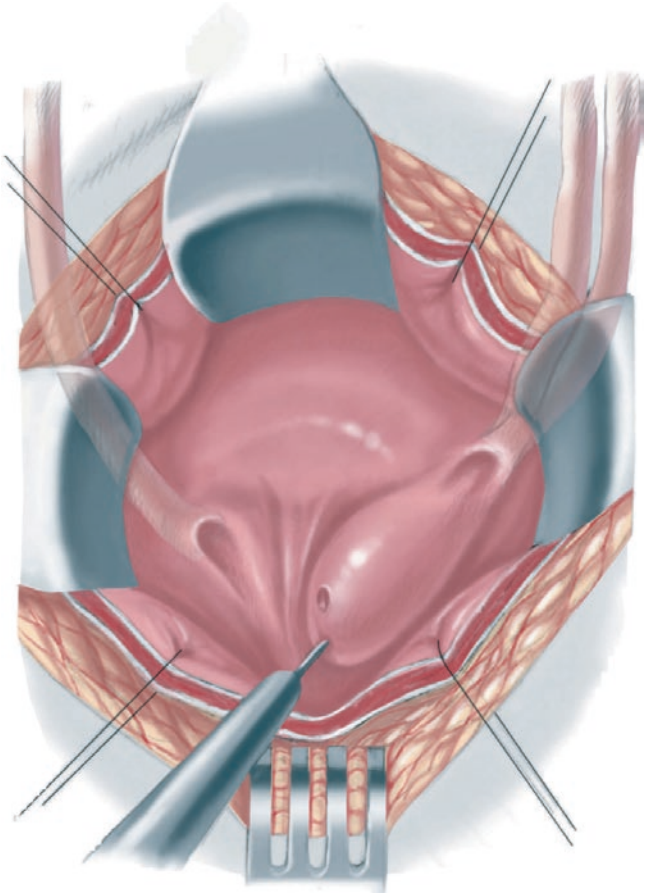

Fig. 62.7 Puncture of ureterocele

62.3.5 Further Ureterocele Treatment Options

If VUR persists following endoscopic ureterocele puncture, treatment options include endoscopic injection of bulking agents (*see* Chap. 60) or open reimplantation of duplex ureters (*see* Chap. 61).

Ureteral reimplantation involves excision of the ureterocele with intravesical common-sheath reimplantation. The ureters are cannulated with feeding tubes, and a circumferential incision is made around the perimeter of the ureterocele. The ureterocele and its associated upper pole ureter are dissected away from the bladder, incorporating the lower pole ureter (Fig. 62.8). The ureters are mobilized within their common sheath to avoid damage to the vascular supply. Care must be taken to avoid damage to the sphincter during distal dissection of the ureterocele.

After excision of the ureterocele, the hiatus may be quite large. The hiatus is closed, again treating the two ureters as a single unit. A common sheath ureteral reimplantation is performed, often in a Cohen cross-trigonal fashion (Fig. 62.9) or a Politano-Leadbetter fashion.

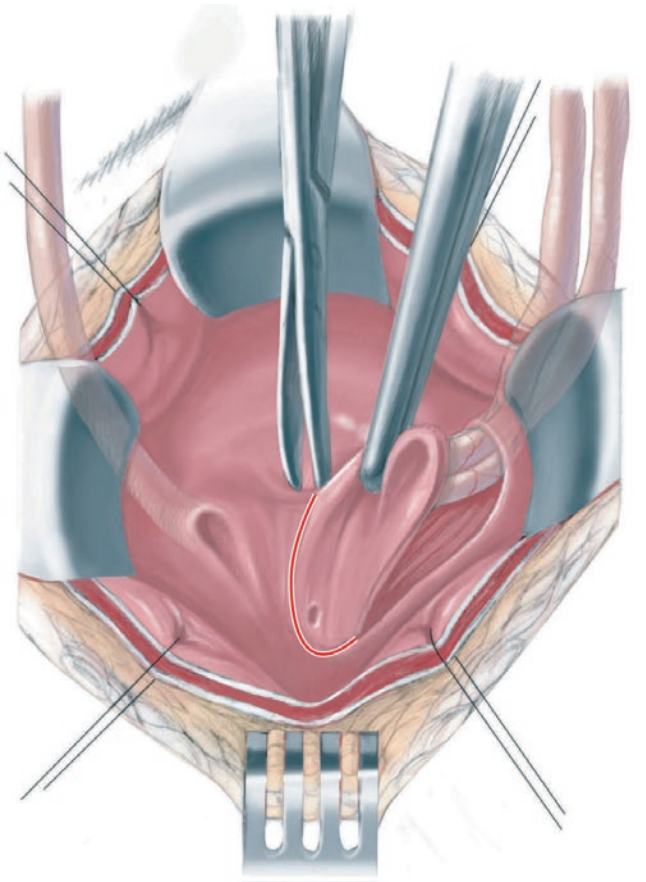

Fig. 62.8 Dissection of the ureterocele and upper pole ureter from the bladder

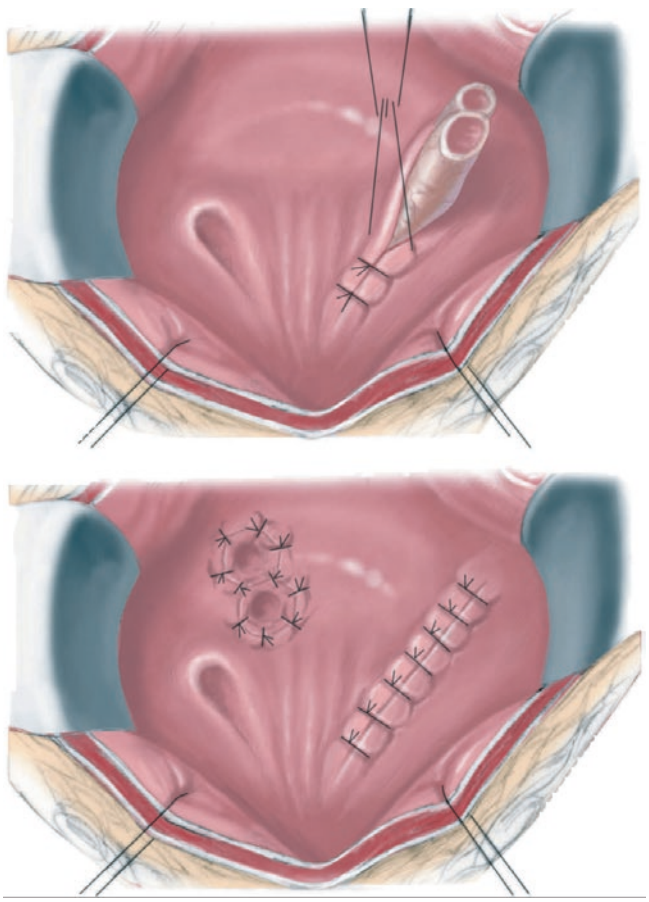

Fig. 62.9 Cohen cross-trigonal ureteral reimplantation

Conclusions

Ureteral duplication is a common anomaly, and is often an incidental finding on prenatal sonography. Multiple treatment strategies exist for duplex collecting systems associated with reflux, ureteral ectopia, and ureteroceles. An individualized, risk-based approach that takes into consideration a multitude of demographic, radiographic, and clinical factors should guide management.

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Posterior Urethral Valves

63

Abhishek Seth, Chester J. Koh, and David A. Diamond

Posterior urethral valves (PUV) represents the most common etiology of congenital urethral obstruction in boys and the most common urologic cause for end-stage renal disease in children. The birth prevalence of PUV—approximately 11 per 100,000 live births—has remained stable over the past 12 years.

Historically, four types of PUVs have been described. However, only types I and III are generally acknowledged as obstructing valvular lesions. Type I valves, the most common, occur in 95% of cases. They typically arise from the posterior and inferior edge of the verumontanum, and radiate distally toward the membranous urethra, where they insert and fuse anteriorly near the proximal margin of the membranous urethra. Type II valves are not considered obstructive lesions; they usually represent hypertrophy of the superficial trigonal muscle in response to distal urethral obstruction. Type III valves, which occur in 5% of cases, usually consist of an obstructing circumferential membrane that sits distal to the verumontanum at the level of the membranous urethra. These may form long folds that prolapse distally and lead to a windsock appearance. Type IV valves are usually seen in prune belly syndrome, where there is a kinking of the flabby, poorly supported prostate.

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63.1 Presentation and Diagnosis

Patients with PUV can present with varying degrees of urethral obstruction, the severity of which is usually dependent upon the age at diagnosis. Newborns may present with signs of severe bladder outlet obstruction, such as palpable abdominal masses from a distended bladder or hydronephrosis, and ascites, or they may have respiratory distress from pulmonary hypoplasia. Toddlers usually present with signs of urinary infection. School-age boys commonly present with signs of voiding dysfunction, such as urinary frequency and incontinence.

More recently, PUVs have been increasingly diagnosed on prenatal ultrasound. In one series, 10% of fetal hydronephrosis cases discovered on ultrasound were attributed to PUVs. On ultrasound, the characteristic findings include bilateral hydronephrosis, a distended and thickened bladder, and a dilated prostatic urethra. In terms of renal echotexture, the parenchyma may appear normal or echodense, which usually suggests the presence of renal dysplasia. In addition, the loss of corticomedullary differentiation by ultrasound postnatally is associated with renal insufficiency and a poor prognosis.

On voiding cystourethrogram, a distended bladder with prostatic urethral dilatation is usually seen with a linear filling defect coursing from the verumontanum to the membranous urethra, which represents the obstructing valve. Other radiographic features may also be present, such as massive, unilateral vesicoureteral reflux; large bladder diverticula; and urinary ascites. All of these findings appear to be associated with improved prognosis, as they indicate the presence of a "pop-off valve" mechanism. These are thought to reduce intraluminal pressures, thereby allowing the fetal renal parenchyma to develop more normally.

In the past, many infants with PUV presented with dehydration, severe electrolyte abnormalities, urosepsis, and renal insufficiency. In these situations, a small feeding tube should be placed transurethrally to allow for adequate vesical

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drainage during the initiation of parenteral antibiotics and rehydration. In placing the tube, one must avoid coiling in the prostatic urethra due to the elevated bladder neck and dilated prostatic urethra. Currently, however, most patients are diagnosed by prenatal ultrasound, avoiding the severe illness previously associated with this anomaly.

63.2 Treatment

63.2.1 Endoscopic Ablation

With the introduction of infant-size cystoscopes and miniature electrocautery, endoscopic ablation of the valves has become the treatment of choice for neonates with PUV. Endoscopic valve ablation involves three essential steps: the assessment of the urethral caliber, the delineation of the anatomy, and the ablation of the valve tissue.

Assessment of the Urethral Caliber: Most newborn urethras will comfortably accommodate a 6F or 7.5F endoscope. For older infants and children, a 9F or 10F instrument can be used. The caliber of the urethral meatus and the fossa navicularis tend to be the limiting anatomic factors in passing an instrument. To determine the maximum capacity of the urethra, pediatric Campbell's sounds should be passed into the anterior urethra only. Calibration of the anterior urethra should occur to one size larger than the instrument to be used, with care taken to avoid overdilation of the urethra.

Delineation of the Anatomy: The anatomy can most easily be determined by passing the endoscope under direct vision into the bladder, then carefully withdrawing the scope. The bladder neck may be quite high, which will require positioning of the endoscope almost vertically to enter the bladder. Then, with the irrigation flowing, the endoscope is gradually withdrawn. In the case of a type I valve, a prominent lip will appear posteriorly just distal to the bladder neck. Then one can visualize prominent urethral folds radiating upward from the verumontanum (Fig. 63.1). The sail-like folds of the type I valve then emanate distally and laterally from the lower portion of the verumontanum and join anteriorly at the 12 o'clock position (Fig. 63.2). The folds can most clearly be seen at the 5 and 7 o'clock positions, and one can see the external sphincter just distal to the valve leaflets.

To best visualize the valves, the bladder should be full, and the end of the cystoscope should be placed at the level of the external sphincter. The lower abdominal wall should then be pressed from above in a Crede fashion, with the drainage channel open on the cystoscope to create a urinary flow. This flow should fill the valve leaflets, which usually snap into an obstructing position.

In the case of a type III valve, which appears like a narrow obstructing diaphragm in the posterior urethra, the passage of the endoscope through the aperture in this diaphragm will often disrupt the valve.

Valve Ablation: The optimal anatomic location for valve ablation is still the subject of debate. The 12 o'clock position has been referred to as the most critical area to ablate, as this is where the anteriorly fused membrane lies. Others have argued that fulguration at the 5 and 7 o'clock positions is optimal because the leaflets are best visualized and most safely fulgurated at these positions. Our preference is to ablate the valve leaflets at the 5 and 7 o'clock positions, although we acknowledge that ablation of the valves at one location alone may disrupt the ring-like obstruction and be curative.

The 7.5F infant resectoscope allows one to safely ablate the valve under direct vision. Once the bladder and urethra have been inspected with the cystoscope, the resectoscope sheath can be introduced after gentle urethral calibration. The sheath of the resectoscope should be passed into the anterior urethra only with the use of the obturator; then the working element can be placed to direct the instrument into the bladder under vision. The visibility with the resectoscope is usually slightly inferior to that with the cystoscope because of the reduced flow of irrigation, so one should reestablish landmarks at this point.

With the use of the 0° lens, the leaflets can best be visualized at the 5 and 7 o'clock positions. Once the valves are in view, the resectoscope loop can be used to hook the leaflets and draw them into the resectoscope sheath (Fig. 63.3). Applying a pure cutting current at 20–30 W will lead to ablation of the valve tissue. In addition, the instrument can be rotated 180° to visualize and ablate the obstructing valve tissue at the 12 o'clock position. One advantage of the resectoscope is its ability to distinguish insignificant leaflets, which do not produce enough resistance to be hooked by the loop.

If the fossa navicularis cannot accommodate the resectoscope sheath comfortably, one may use the 6F cystoscope and the 3F Bugbee electrode. Ablation with the Bugbee electrode is performed by engaging the medial edge of the leaflet with the electrode and then gently pushing toward the bladder and applying the current (Fig. 63.4). The cautery settings should be at 25 W on pure cut. Ablation at the 5 and 7 o'clock positions should render the valve incompetent. The remnant leaflets should flutter with the expressed urine. Repeat fulguration may be necessary if residual obstructive tissue is noted.

Following valve ablation, an indwelling catheter is left in place for 24 h, although this may not always be necessary.

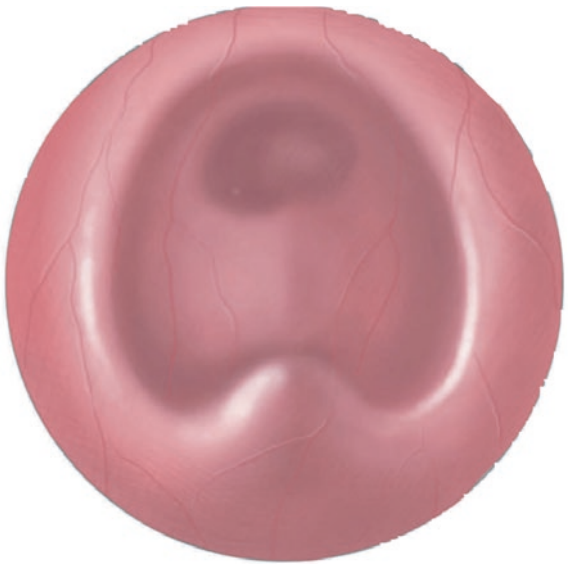

Fig. 63.1 Urethral folds radiating upward from the verumontanum in a type I PUV

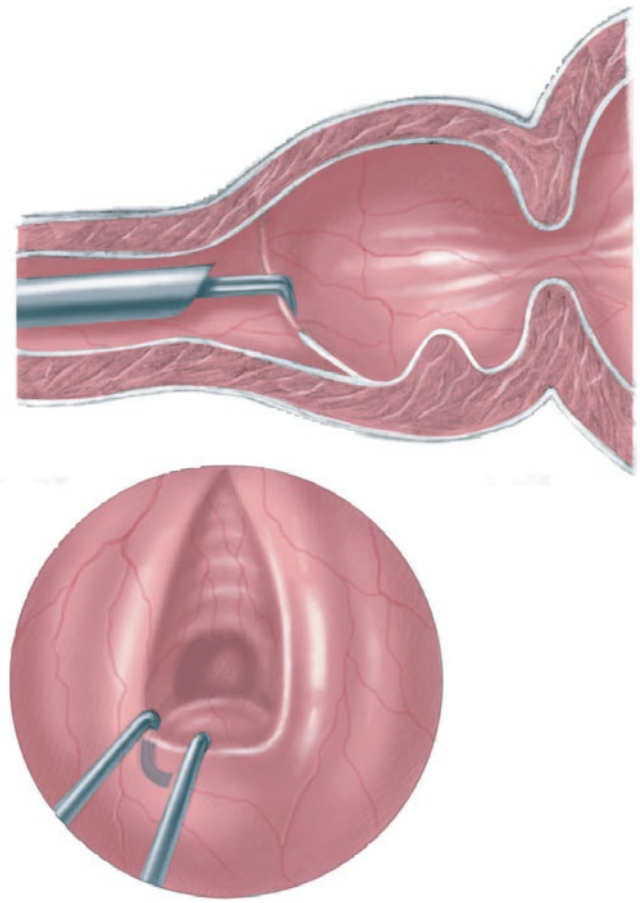

Fig. 63.3 Using the resectoscope loop to hook the leaflets

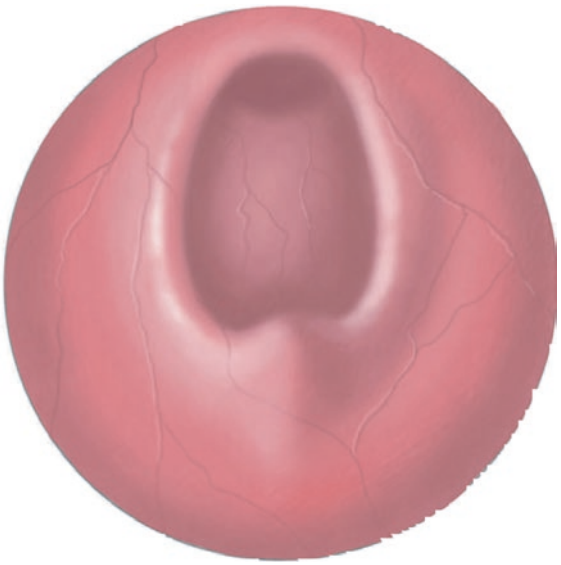

Fig. 63.2 Sail-like folds joining at the 12 o'clock position

Fig. 63.4 Leaflet ablation using a Bugbee electrode

63.2.2 Other Procedures

For smaller infants, an alternative means of valve ablation may be necessary, such as the use of a 3F ureteric catheter and a wire stylet. The urethras of these children may only allow this smaller catheter to be passed through or alongside the 6F infant cystoscope while permitting the flow of irrigant during the procedure for 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 catheter itself acts as the insulator to the wire. The catheter is usually advanced through the cystoscope to the level of the valve before the wire is advanced. Then the wire should be advanced 2–3 mm out of the catheter and pushed just into the valve leaflet. Fulguration can occur in a similar fashion as with the larger Bugbee electrode, but several short bursts of current may be necessary to effectively ablate the valves. Care should be taken to avoid overzealous fulguration, as this complication can lead to injury to the prostatic urethra and external sphincter.

In select cases, alternative techniques may be useful. In developing countries without miniature equipment, or in premature infants in whom safe ablation is limited by urethral caliber, ablation under fluoroscopic guidance with a Fogarty balloon catheter may be advantageous. In similar settings, the Whitaker insulated hook provides an alternative approach.

As the ability to prenatally diagnose PUV has developed over time, so has the temptation of in utero intervention to relieve urinary obstruction secondary to PUVs. The long-term benefits of in utero intervention are currently unclear, however, and they must be weighed against the significant risks to the mother and fetus. In the recently published PLUTO trial, Smith *et al*. attempted to assess the effect of vesicoamniotic shunting in prolonging survival in fetuses with evidence of lower urinary tract obstruction, compared with conservative management. Although this trial showed a small but statistically significant improvement in short-term (28 days) and long-term (1 and 2 years) survival in the analysis based on treatment, no such improvement was seen in an intention-to-treat analysis. Furthermore, the findings from this trial also showed that regardless of whether vesicoamniotic shunting is performed, the probability of infants surviving with normal renal function is very low. Therefore, the benefit of vesicoamniotic shunting could not be definitively proven. In light of the morbid nature of vesicoamniotic shunting and its unclear benefit, the most compelling scenario for proceeding with prenatal intervention is when a male fetus with signs of infravesical obstruction is initially noted to have normal amounts of amniotic fluid but subsequently develops oligohydramnios, while having evidence of good renal function based on serial aspirations for urine electrolyte.

63.3 Results

Transurethral ablation usually leads to relief of urethral obstruction and often resolution of reflux via a minimally invasive approach that entails minimal risk. The benefits of ablation include decompression of the urinary tract and the tendency toward maximum recoverability of overall renal function during a period of substantial renal growth. In addition, transurethral ablation is believed to allow the bladder to fill and empty on a regular cycle, which is thought to lead to more normal voiding patterns and to decreased collagen deposition and perhaps a reduction in the risk of bladder noncompliance.

Certain prognostic factors have been reported in the literature. At age 12 months, a nadir creatinine of 0.8 mg/dL or less is associated with normal long-term renal function and improved prognosis. On the other hand, the presence of daytime urinary incontinence after age 5 years is associated with abnormal long-term renal function.

Nevertheless, bladder dysfunction (described as the *valve bladder syndrome*) is always a distinct possibility even after successful fulguration of valves. In general, management of the valve bladder consists of anticholinergic medications to reduce uninhibited detrusor contractions, clean intermittent catheterization to ensure adequate bladder emptying, or bladder augmentation to improve bladder compliance and bladder volume.

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Hypospadias

64

Mariette Renaux-Petel, Pierre-Yves Mure,
Daniela-Brindusa Gorduza, and Pierre Mouriquand

Hypospadias is one of the most common urogenital anomalies, occurring in 3 in 1000 births. It is better defined as an arrest of the development of the genital tubercle (GT) during week 6 and week 16 of gestation, leading to hypoplasia of the tissues forming the ventral aspect (ventral radius) of the penis beyond the division of the corpus spongiosum. It is characterized by a ventral triangular defect whose summit is the division of the corpus spongiosium, whose sides are represented by the two pillars of atretic spongiosum, and whose base is the glans itself.

In the middle of this triangle, from the tip to the base of the penis, sit a widely open glans, the urethral plate (which extends from the ectopic urethral meatus up to the glans apex), the ectopic meatus, and a segment of variable length of atretic urethra (not surrounded by any spongiosum), which starts where the corpus spongiosum divides. Tissues sitting inside this triangle are dysplastic and do not grow at the same pace as the rest of the GT.

There are two main types of hypospadias:

- The hypospadias with a distal division of the corpus spongiosum with little or no ventral curvature when the penis is erected.
- The hypospadias with a proximal division of the corpus spongiosum, with a marked degree of hypoplasia of the tissues forming the ventral radius, associated with a significant degree of ventral curvature.

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64.1 Causes of Hypospadias

The causes of hypospadias remain essentially unknown. Several avenues have been explored to explain this congenital defect of the genital tubercle:

- Some endocrine disorders have been described in relation to hypospadias, mainly an insufficient secretion of androgens or insufficient response by the target tissues. These disorders can be demonstrated in very few cases, however.
- Some genetic disorders could explain why hypospadias may be found in several members of the same family.
- Young and old mothers are more prone to carry a baby with hypospadias. Low-birth-weight babies and twins also have a higher risk of presenting with a hypospadias. This risk could be explained by a placental insufficiency.
- The possible and controversial increase of hypospadias in the population over the past 20 years suggests a role for possible environmental factors such as oestrogenlike molecules, pesticides, and fertilizers.
- Abnormal or insufficient growth factors could also be responsible for these penile anomalies and could also explain the significant complication rate met in this surgery.

64.2 Surgical Techniques

Three surgical steps characterize hypospadias surgery: (1) Degloving of the penis and deep dissection of the glans wings; (2) Repair of the urethra (urethroplasty); (3) Reconstruction of the ventral radius of the penis.

Degloving of the penis and a deep dissection of the glans wings: In most cases, the penis is straightened and the common ventral glans tilt is corrected. In some midshaft and proximal hypospadias cases, the residual curvature

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demonstrated by an erection test is due to asymmetrical corpora cavernosa and requires a dorsal corporeoplasty (dorsal shortening of the albuginea of the corpora cavernosa). In most cases, the urethral plate can be preserved, although there are situations in which the urethral strip is very poor and the two corpora writhe around it. In these cases, the urethral plate is usually sacrificed.

Repair of the urethra (urethroplasty): Once the GT has been fully dissected, the length of urethra to be reconstructed can be precisely evaluated. The repair technique chosen depends on the size and quality of the urethral plate:

- If the urethral plate is wide and healthy, some will choose to tubularize it following the Thiersch-Duplay technique.
- If it is too narrow to be tubularized, the Snodgrass open urethrotomy is a popular option, or additional tissue can be laid on the urethral plate using a rectangle of pediculized preputial mucosa (onlay urethroplasty) or a flap of ventral penile skin (Mathieu procedure).
- If the segment of urethra to replace is short (<1.5 cm) and if the distal urethra is not hypoplastic, a complete mobilisation of the whole penile urethra may be adequate to bridge the defect. This technique (Koff) has the advantage of avoiding the use of nonurethral tissue.
- If the urethral plate cannot be preserved, a tube must be made to replace the missing urethra, using either a pediculized rectangle of preputial mucosa (Asopa-Duckett technique) or buccal mucosa.
- In major hypovirilization of the GT, the Koyanagi procedure, mobilising the tissues of the ventral and lateral aspects and dorsal prepuce with the blood supply, is a reliable option to reconstruct the missing urethra. Two-stage procedures (Cloutier-Bracka procedure) are an alternative for long urethroplasties using either preputial mucosa or buccal mucosa.

Reconstruction of the ventral radius of the penis: Once the urethra has been repaired, reconstructive steps are taken:

- Meatoplasty, trying to create a slit-shaped meatus.
- Glanuloplasty, by stitching together the two glans wings over the reconstructed urethra to reconstruct the ventral aspect of the glans.
- Creation of a preputial collar around the glans.
- Coverage of the reconstructed urethra (spongioplasty), using the lateral pillars of spongiosum or the pedicle of the onlay flap.
- Skin cover with a redistribution of the skin shaft, bringing the excess dorsal skin to the ventrum.
- Reconstruction of the foreskin, or circumcision.

The patient's age at surgery for primary hypospadias repair is usually between 6 and 24 months.

Pre-operative hormonal stimulation of the penis using beta human chorionic gonadotrophin (βHCG), testosterone, or dihydrotestosterone is sometimes indicated in case of a small penis (dorsal radius <25 mm long or glans width <15 mm in the first year of life) or in case of redo surgery. It remains unclear how safe these treatments are on a long-term basis. Some publications have pointed out their detrimental action on the healing process.

General anaesthetic is the rule, often associated with caudal or penile anaesthesia. Magnification is commonly used in this surgery. Coagulation is often not needed in this surgery when the tourniquet is used followed by a slightly compressive dressing. Other surgeons prefer bipolar coagulation or adrenaline injection prior to incision.

Urine drainage via a suprapubic catheter, a transurethral bladder catheter, or dripping urethral stent varies from one surgeon to another; some do not drain at all.

Antibiotic protocols also vary greatly from one centre to another, and their efficacy needs to be proven. A dressing is essential after this surgery. We prefer the so-called daisy dressing because it is very comfortable for the patient and contains postoperative bleeding, but some others prefer OPSITE dressing, Silastic foam dressing, or Tegaderm dressing.

Postoperative pain control is essential, using morphine instillations, anti-inflammatory medications, an anticholinergic (oxybutynin chloride), and diazepam to reduce bladder spasms caused by the bladder catheter.

64.3 Specific Surgical Procedures

From the tip to the base of the penis, the ventral aspect of the glans is widely opened. The urethral plate extends from the apex of the glans down to the ectopic meatus (Fig. 64.1). Behind the ectopic meatus sits a segment of hypoplastic urethra not surrounded by any spongiosum. The division of the corpus spongiosum marks the proximal limit of the malformation. It defines a triangular defect whose summit is the division of the corpus spongiosum, whose base is the glanular cap and whose sides are represented by the two lateral pillars of atretic spongiosum.

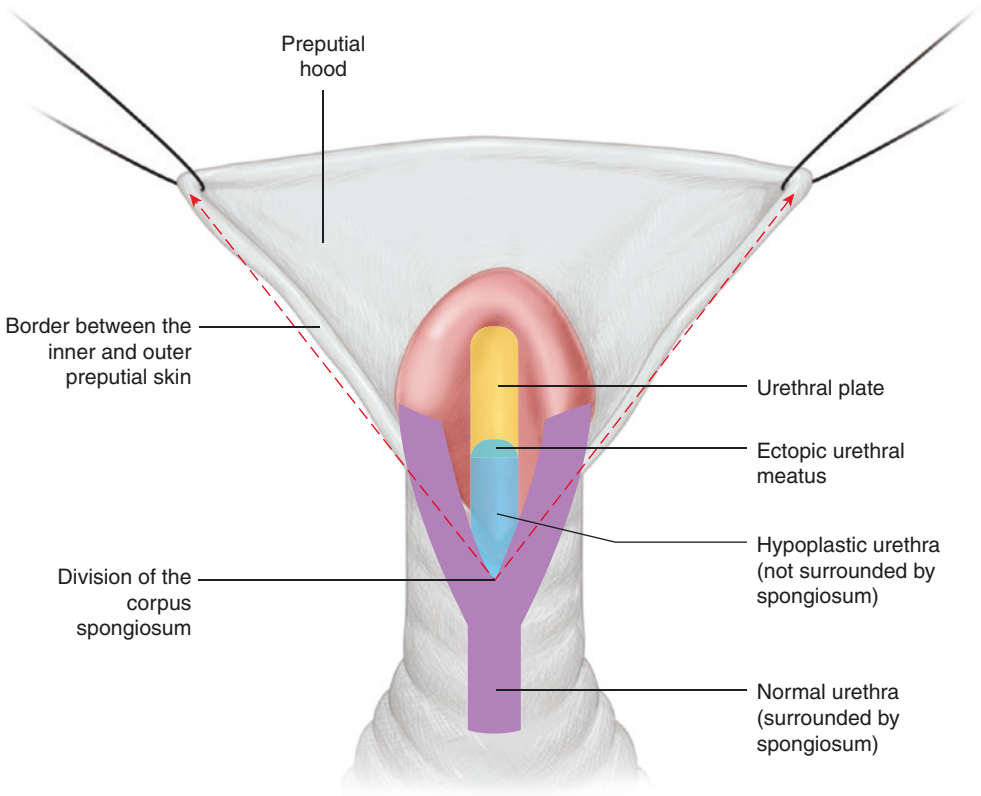

Fig. 64.1 Anatomy of the defect

64.3.1 Correction of Ventral Curvature: TAP Procedure

Most penises are fully straightened after skin shaft degloving and extended dissection of the glans wings. In hypospadias with a proximal division of the corpora cavernosa, a complementary corporeoplasty may be needed to complete the straightening, either by tunica albuginea plication (TAP procedure), plicating the dorsal albuginea of the corpora cavernosa on the dorsal midline (Fig. 64.2), or by using the Nesbit procedure. Some suggest grafting the ventral aspect of the corpora to extend its length.

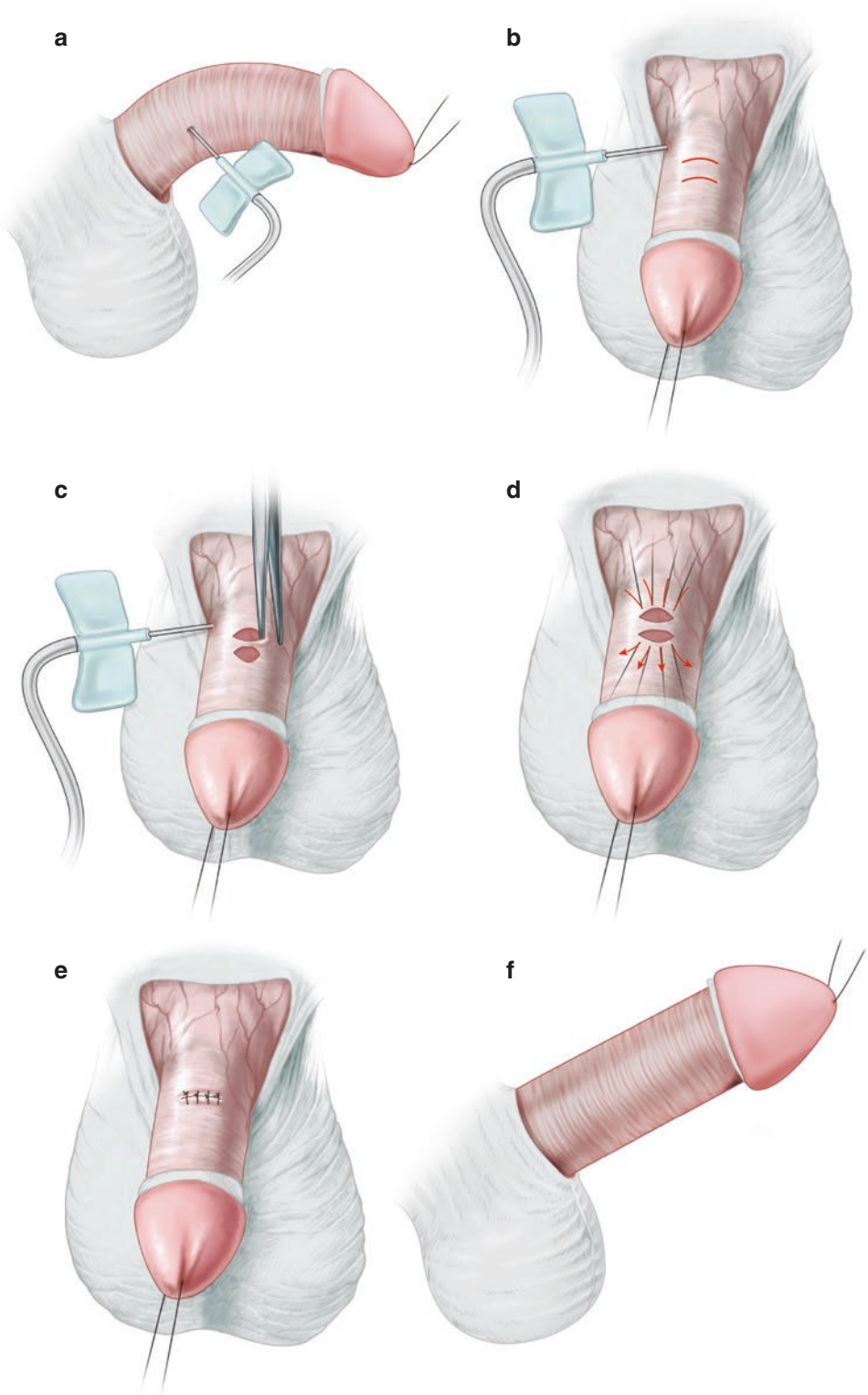

Fig. 64.2 (**a**–**f**) Tunica albuginea plication (TAP procedure) for correction of ventral curvature

64.3.2 Thiersch-Duplay Procedure

If the urethral plate seems to be wide and healthy, it can be tubularized following the Thiersch-Duplay technique. Its best indication is in the glanular hypospadias with intact prepuce (megameatus), where the urethral plate is thick, deep, and healthy. The incision lines follow each side of the urethral plate from the tip of the glans down to the division of the corpus spongiosum (Fig. 64.3). The two wings of the glans are dissected deeply and laterally until the corpora are clearly identified. The urethral plate is tubularized around an 8F catheter for children under 3 years of age, using a 6/0 or 7/0 absorbable running suture. The neourethra is then covered by two wings of the glans in one or two layers.

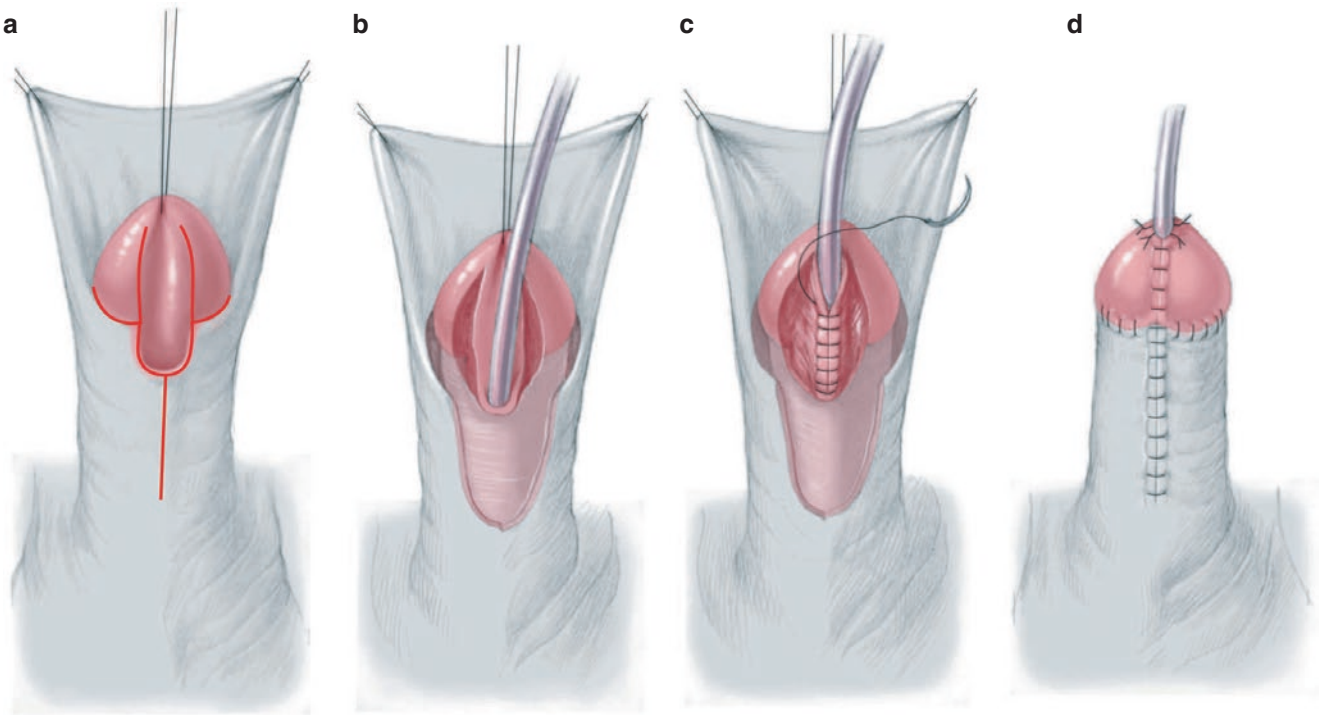

Fig. 64.3 (**a**–**d**) The Thiersch-Duplay technique for hypospadias repair

64.3.3 Snodgrass Procedure

In the Snodgrass procedure (Fig. 64.4), the urethral plate is incised longitudinally on its midline from the ectopic meatus up to the glans and subsequently tubularized around an 8F catheter. This procedure leaves a dorsal raw area in the urethra, which is subsequently epithelized. Some suggest grafting the raw area with either foreskin or buccal mucosa; the technique is then called the inlay urethroplasty or "Snodgraft" procedure.

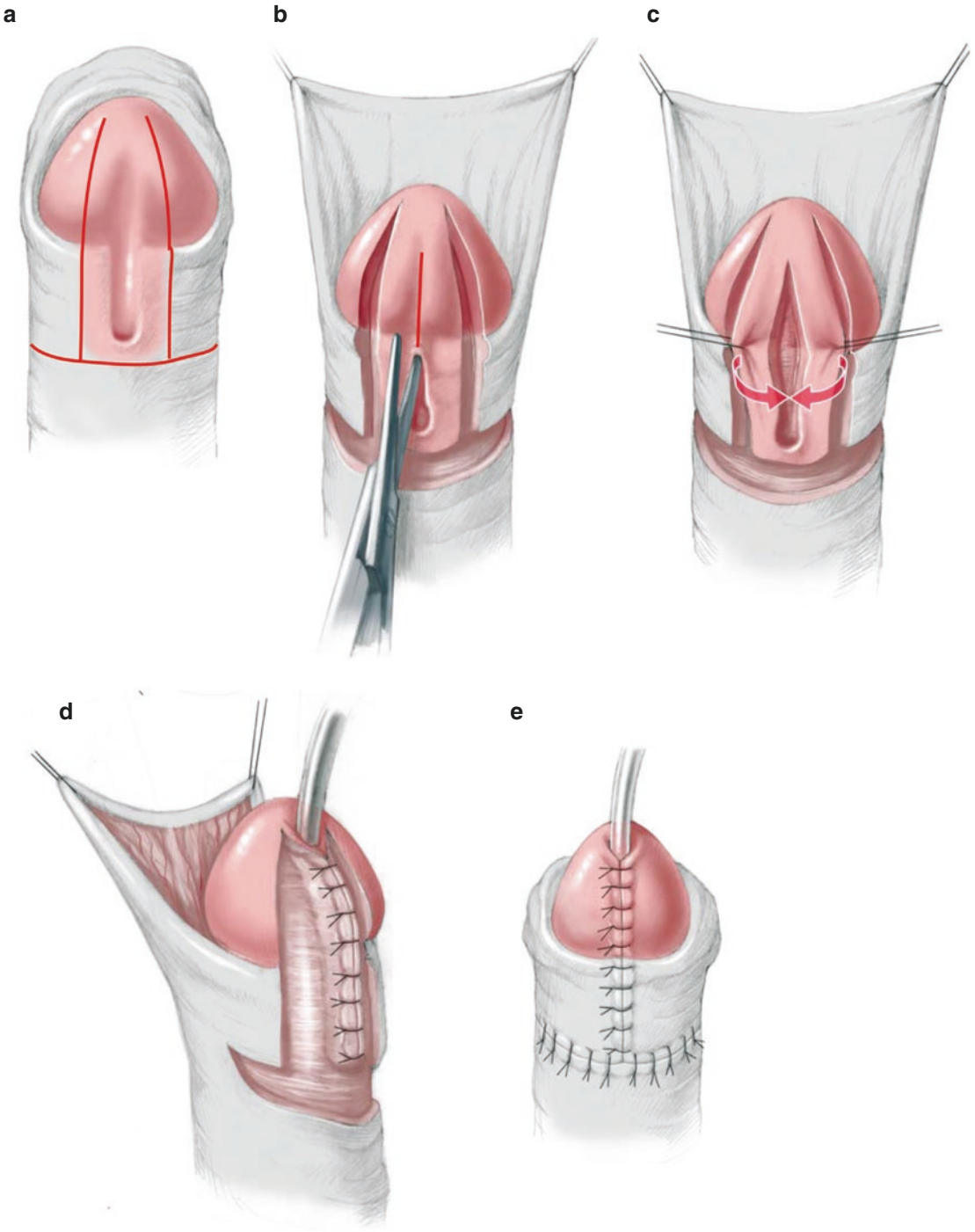

Fig. 64.4 (**a**–**e**) Snodgross procedure for hypospadias repair

64.3.4 Onlay Procedure

In the onlay procedure (Fig. 64.5), a rectangle of preputial mucosa is pediculized down to the base of the penis and transferred to the ventrum of the penis to be layed on the urethral plate using interrupted 6/0 or 7/0 or a running suture.

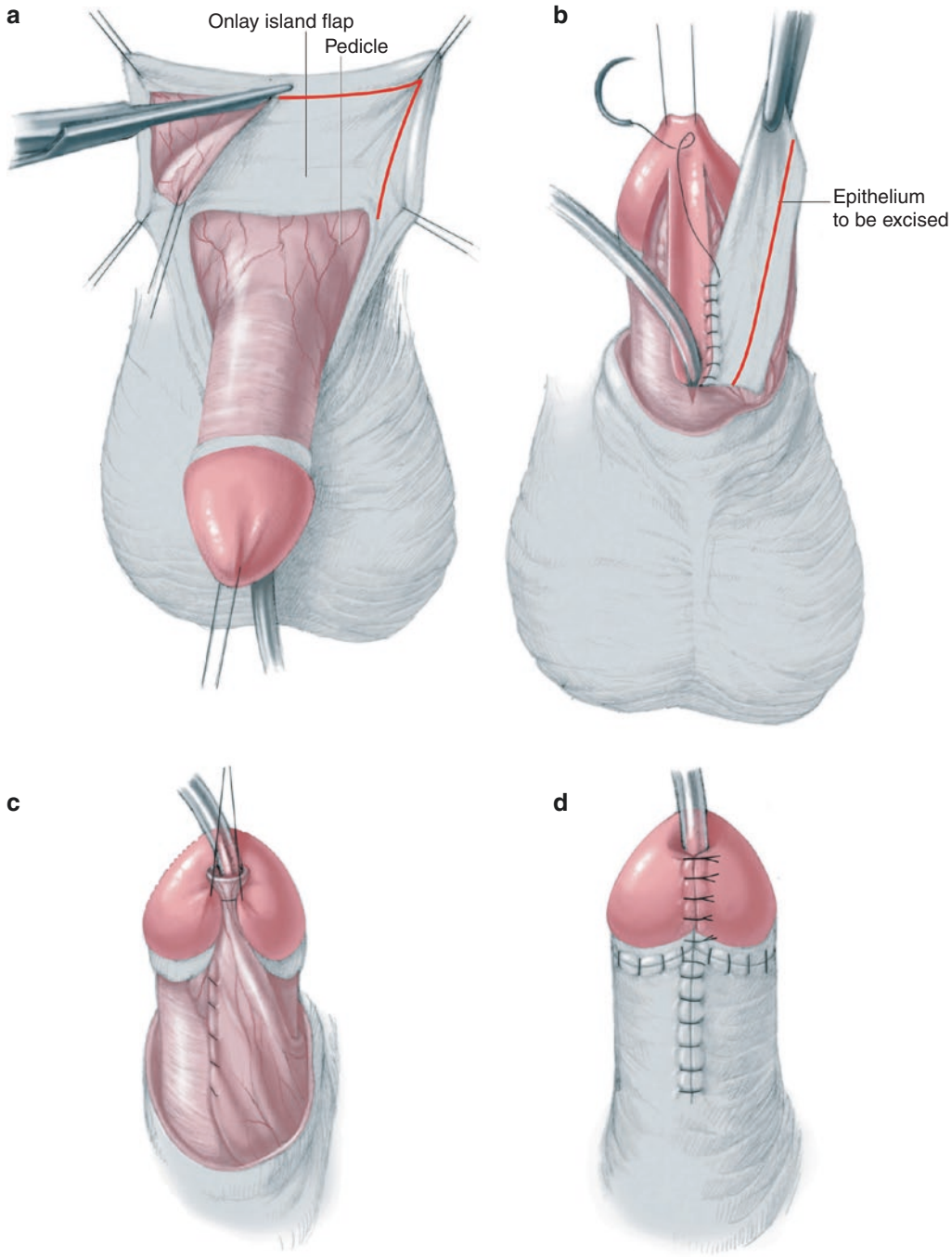

Fig. 64.5 (**a**–**d**) Onlay procedure for hypospadias repair

64.3.5 Mathieu Procedure

In the Mathieu procedure (Fig. 64.6), two parallel incisions are made on either side of the urethral plate up to the tip of the glans and deep down to the corpora cavernosa. The incision line delimits a perimeatal-based skin flap that is folded over and sutured to the edges of the urethral plate. The lateral wings of the glans are generously dissected from the corpora cavernosa and approximated together, producing a conical shape of the glans.

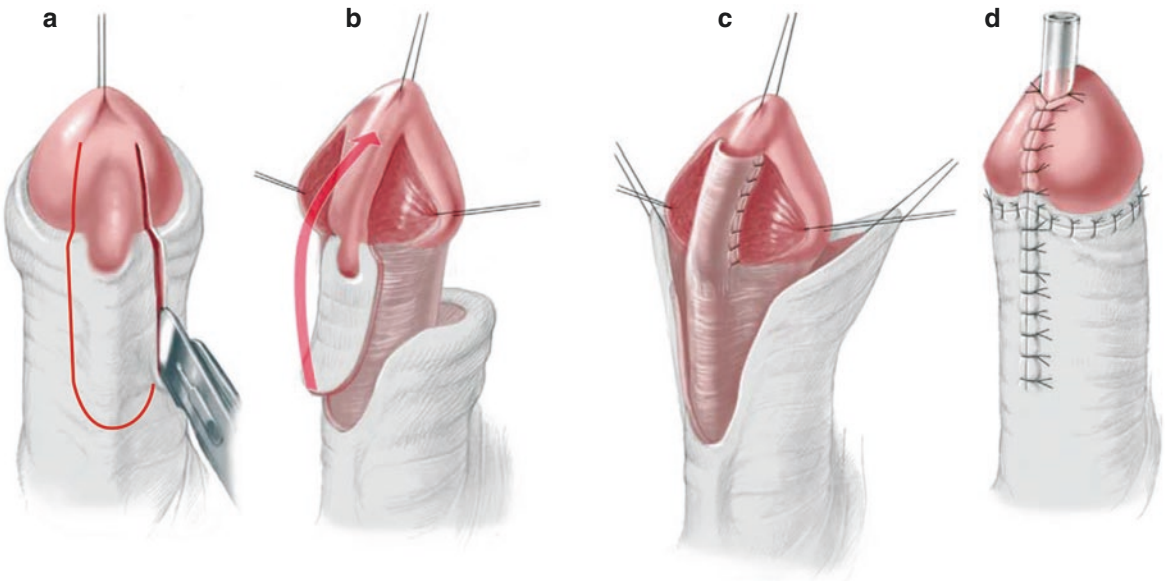

Fig. 64.6 (**a**–**d**) Mathieu procedure for hypospadias repair

64.3.6 Koff's Urethral Mobilisation

When the segment of the urethra to reconstruct is short (<1.5 cm) and when the distal urethra is healthy, a full mobilisation of the penile urethra can be performed following Koff's technique (Fig. 64.7). In these cases, the penile urethra is detached down to the base of the penis, and then it is moved upward to bring the meatus to the tip of the glans. The gain of length may be up to 15 mm. It is important to reattach the freed urethra to the corporeal surface using interrupted 7/0 resorbable stitches to avoid a ventral tilt of the glans and an iatrogenic curvature.

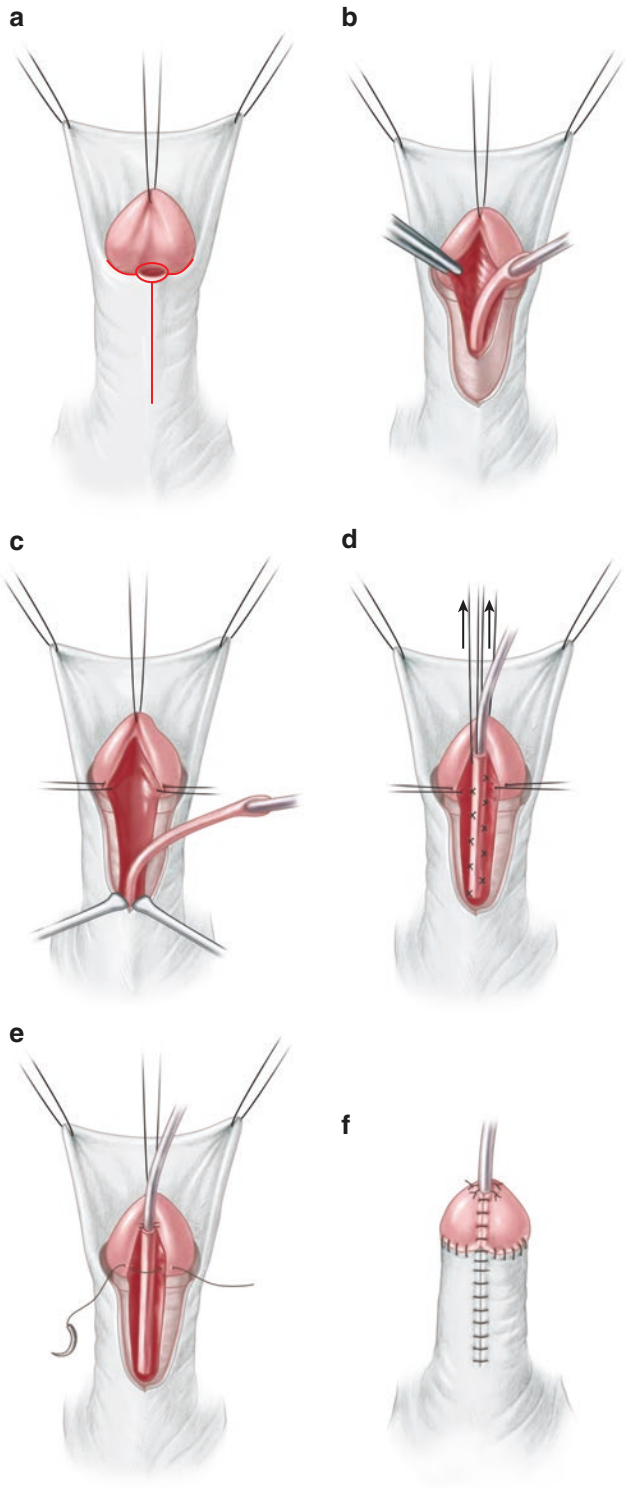

Fig. 64.7 (**a**–**f**) Koff's uretheral mobilisation technique for reconstruction of a short segment

64.3.7 Koyanagi Procedure

In the Koyanagi procedure, the initial incision lines follow a coronal circumference with a transection of the urethral plate just below the glans (Fig. 64.8). Then a second, racket-shaped incision allows complete mobilisation of the urethral plate and the adjacent tissues with the inner aspect of the preputial hood. A long, wide strip of well-vascularized tissue is freed with its blood supply down to the base of the penis. The whole graft is transferred to the ventral aspect of the penis after the midline dorsal division of the inner aspect of the preputial hood. The skin flaps are joined together to reconstitute the basement of the future urethral plate, which is then duplay-ed from the ectopic meatus up to the glans. In the Hayashi modification, dorsal preputial skin is left intact to improve the blood supply of the distal graft. In the original Koyanagi procedure, the dorsal preputial skin is divided on the midline as shown in Fig. 64.8.

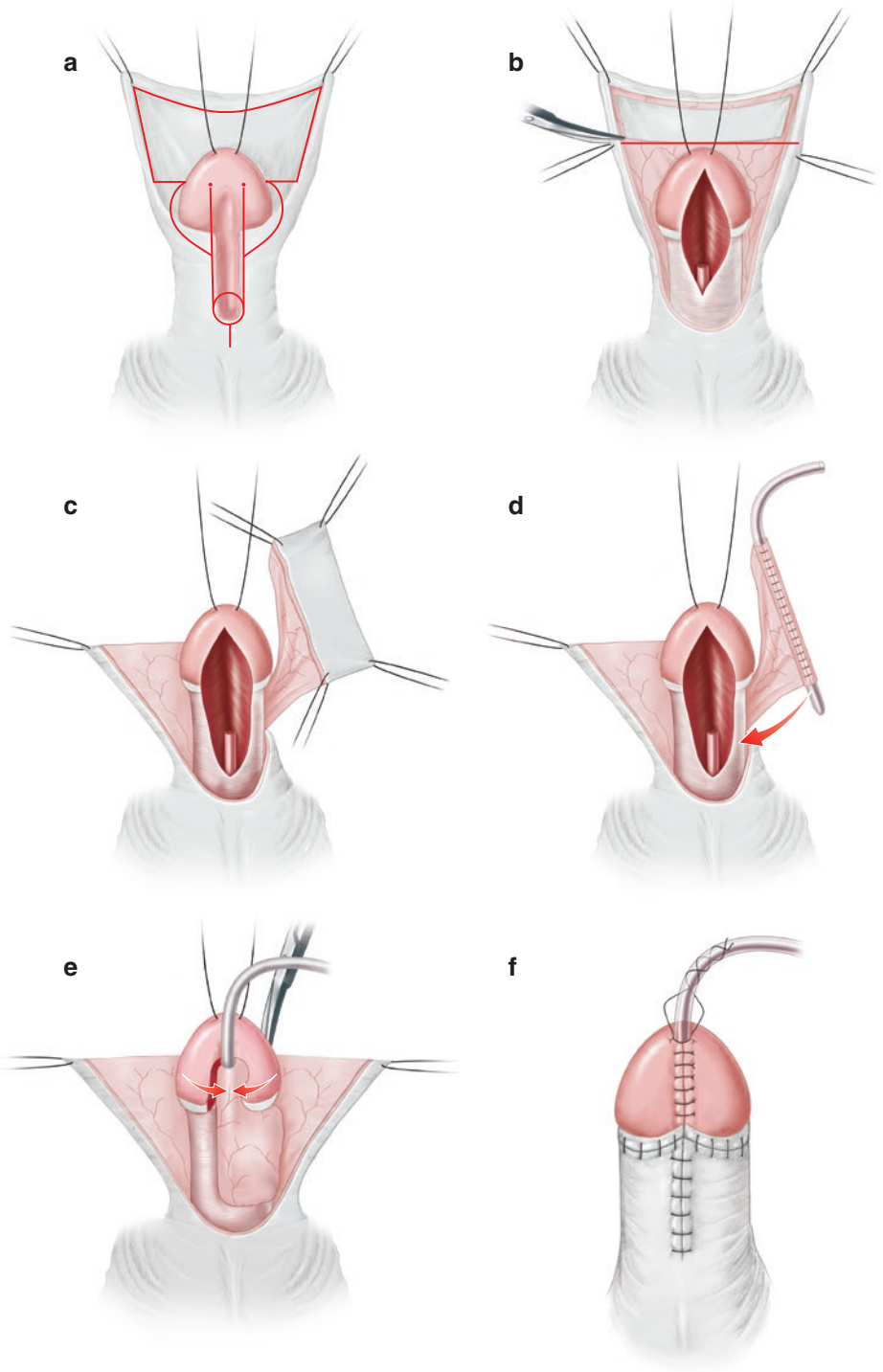

Fig. 64.8 (**a**–**f**) The Koyanagi procedure, used when a long urethroplasty is required

64.3.8 Bracka-Cloutier Procedure

Repair of some proximal hypospadias or some redo hypospadias may benefit from a two-stage procedure: The first stage aims at bringing fresh tissue (the internal aspect of the preputial hood or buccal mucosa) onto the ventrum of the genital tubercle (Fig. 64.9). These free grafts need to find adequate blood supply to survive. The graft is stitched flat on the ventrum of the corpora from the ectopic meatus to the glans wings. It will be subsequently duplay-ed 3 months later once the graft take is confirmed.

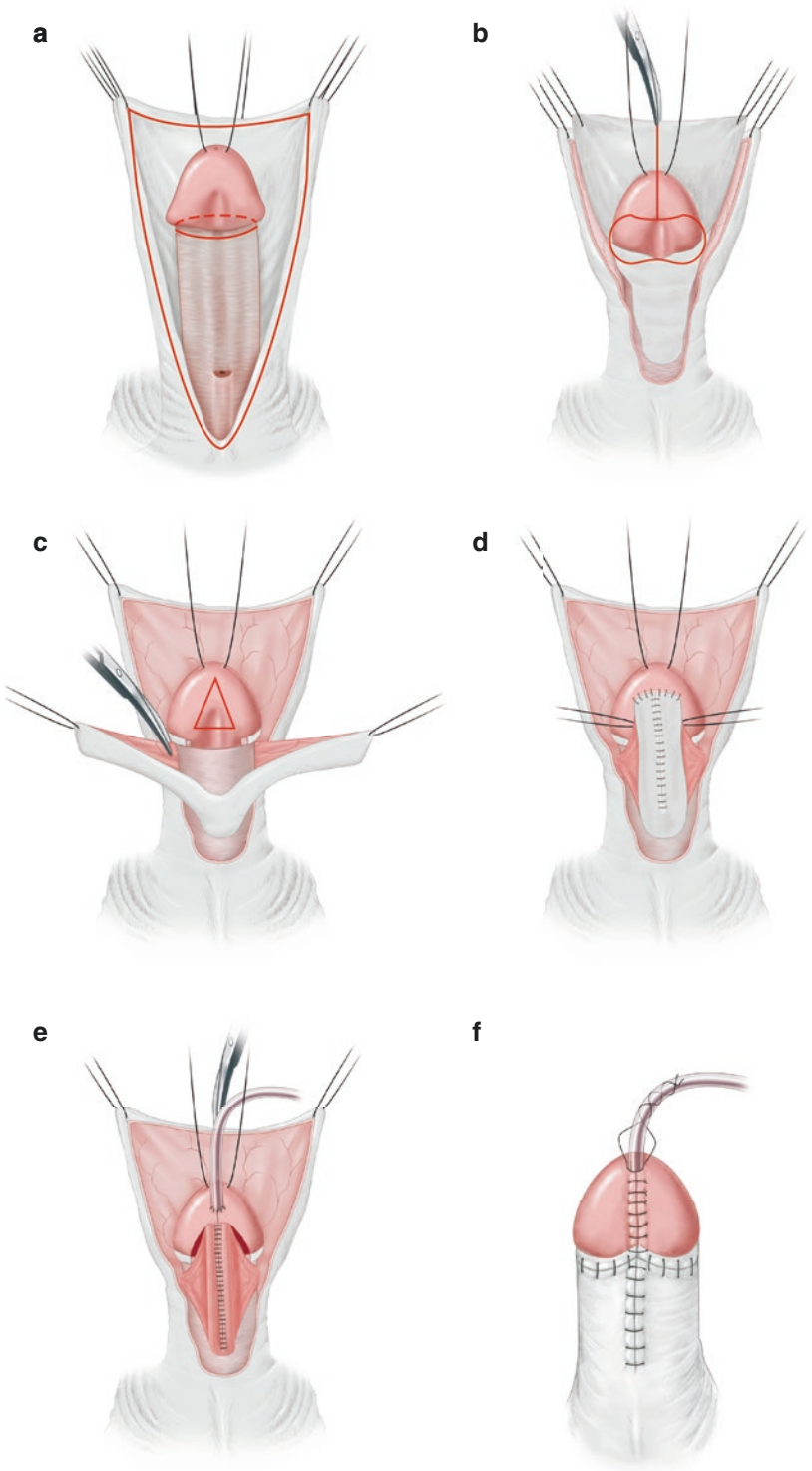

Fig. 64.9 (**a**–**f**) The two-stage Bracka-Cloutier procedure using a free graft for hypospadias repair

64.4 Results and Conclusions

Hypospadias surgery remains a difficult challenge, as several factors of success remain unknown. One of the most intriguing aspects is the variation in the healing abilities of patients. With the development of tissue engineering, it is hoped that urethral substitution using the patient's own urethral tissue might be a future avenue to resolve current difficulties. Long-term follow-up of these patients appears to be crucial to assess and validate the various techniques currently available. The problem is how to follow these patients. Clinical examination of the penis is highly subjective, and assessment of the urine stream is difficult, as urine flow studies are very often abnormal after urethral reconstruction even if the caliber of the neourethra is correct. At the end of the day, the experience and honesty of the paediatric urologist remain the two most important factors for progress in hypospadiology.

Parents should be clearly informed that a significant number of hypospadias repairs will require further surgical attention during the patient's life. Because minor hypospadias are much less common than believed, this surgery should always be performed by experienced paediatric urologists. Collaboration with paediatric endocrinologists is also important, to increase the chances of surgical success. Preoperative and postoperative treatment may be helpful to improve each patient's healing abilities.

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Phimosis and Buried Penis

65

Navroop Johal and Peter Cuckow

The term *phimosis*, derived from the Greek word meaning *to muzzle*, is a descriptive term referring to the natural conical shape of the foreskin in early life. This shape prevents its retraction and keeps the glans of the penis covered. Additionally, the inner surface of the prepuce is initially fused with the outer surface of the glans. Widening of the narrow tip of the prepuce, combined with separation of its inner adhesions, occurs during childhood to allow full retraction and uncovering of the glans by puberty.

Studies of normal infants and children have shown that the rate of this process is very variable—to the extent that in some newborns the prepuce is already fully retractile, while it can remain "phimotic" in about 20% of 5-year-olds. The failure to recognize this normal process of development has led many doctors to label nonretractile foreskins abnormal and refer patients for circumcision. The vast majority of prepuces are normal, however, and will become fully retractile by puberty.

65.1 Foreskin Abnormalities and Complications

In the 1% of males who do not become retractile, most are subject to a pathological process called balanitis xerotica et obliterans (BXO), which causes clinically apparent scarring at the preputial tip. BXO is usually cured by circumcision and is the only absolute indication in childhood, but it is rarely seen in children younger than 5 years old.

Other foreskin problems may be related to its development and nonretraction, but these are usually transient rather than long-term problems. Thus ballooning during micturition and infection of the foreskin or balanitis will resolve and cause no discernible damage, but severe symptoms from these conditions do represent a relative indication for circumcision. By applying these principles, several groups of British pediatric surgeons have reported circumcising only about 25% of boys referred for consideration of the operation. In the United States, circumcision is a common procedure, generally performed during the newborn period. A policy statement by the American Academy of Pediatrics Task Force on Circumcision identified specific benefits, which included prevention of urinary tract infections, penile cancer, and transmission of some sexually transmitted infections, including HIV.

There is a minority of boys whose foreskins remain narrow or tight on retraction even though no obvious scarring can be seen. In these patients, preputial plasty or widening of the narrow tip of the prepuce can be a helpful and more conservative alternative to circumcision.

A wide range of complications of circumcision is reported, although there are few prospective long-term studies. Early complications include haemorrhage, retention of urine, infection, and meatal ulceration. Meatal stenosis is the most significant long-term complication seen in 2 and 35% of cases. These patients can develop voiding difficulties and even bladder dysfunction if the outflow obstruction is severe. This stenosis is managed by meatotomy. Other long-term complications are harder to define. Cosmetic appearance is subjective, but significant numbers are reported.

Penile abnormality is a contraindication for routine circumcision. Patients with hypospadias require a more complex reconstruction if they are to achieve a more normal "circumcised" appearance; this procedure is discussed elsewhere in this book (Chap. 64).

The buried penis is an abnormality of peno-scrotal fusion, in which the penile corpora are also tethered to the deep fascia of the lower abdominal wall. It is associated with phimosis, and the appearance of the external skin suggests that the penis is small or even absent. Often the inner preputial space is enlarged and balloons during voiding, with dribbling from the preputial orifice—referred to as megaprepuce.

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Circumcision of the visible external prepuce in these cases does not achieve emergence of the penis and may compromise the eventual reconstruction of a more normal circumcised appearance. An operation for this condition, to release the tethered corpora and remodel the shaft skin, is presented later in this chapter.

65.2 Penile Surgery Techniques

Penile surgery should always be performed under general anaesthesia. Local anaesthetic techniques can also be used to provide good intraoperative and postoperative analgesia; a caudal epidural is the standard. This is probably not required for meatotomy or preputial plasty. Sutures for this surgery should be absorbable monofilaments such as Monocryl. We always use round-bodied or taper-point needles.

65.2.1 Circumcision

The prepuce is fully retracted behind the glans and any residual adhesions to the glans are separated carefully. If the tip is narrow or scarred, a haemostat is inserted into the preputial orifice and opened in order to dilate it and allow retraction. For this to be done, it may be necessary to make a small dorsal incision in the tip of the prepuce with scissors. In a patient with severe BXO (rare in those under 5 years of age), adhesions to the glans may be dense and their separation can be traumatic.

With the skin held under tension, retracted back over the penile shaft, a circumferential incision is made with a size 15-blade scalpel, 8 mm proximal to the glans on the dorsal surface (Fig. 65.1). This incision is completed ventrally on each side, parallel to the coronal sulcus, where the frenulum and its artery are divided. Care is taken not to damage the urethra, which is quite superficial at this point.

The foreskin is replaced over the glans. A clip is placed on its edge in the midline anteriorly (usually the position of the midline raphe). A second clip is placed opposite this dorsally and the foreskin is held forward, under slight tension. The second circumferential incision is made just proximal to the corona of the glans, whose profile can be seen and felt through the foreskin. Care is taken to ensure that enough skin is retained to cover the shaft. The incision is angled distally towards the ventral surface, which allows for the natural angle of the coronal sulcus with the penile shaft.

The clips are repositioned on the dorsal edge of the foreskin on either side of the midline, and a dorsal slit is made between them with scissors (Fig. 65.2). This joins the inner and outer incisions. Scissors are used to divide the subcutaneous layers circumferentially and remove the sleeve of foreskin.

The remaining cylinder of penile skin is retracted proximally to expose the raw shaft and bleeding vessels, which are coagulated with bipolar diathermy. Attention is paid to the frenular aspect of the penis and coagulation of the distal end of its artery beneath the ventral skin cuff.

A fine absorbable suture (5/0 or 6/0 Vicryl or Monocryl) on a round-bodied needle is used to approximate the shaft skin to the cuff of distal skin (Fig. 65.3). A box stitch is used to reconstitute the frenulum. A second stitch is placed in the midline dorsally and the penis is suspended between them. Interrupted sutures are placed at 5-mm intervals along each side to complete the anastomosis (Fig. 65.4).

Following the operation, the penis is wrapped in gauze and squeezed for 2 min to exclude haematoma and aid haemostasis, before the boy is awakened. Any bleeding at this point can be stopped with a circumferential gauze dressing, which can be removed before the patient goes home, but with effective haemostasis this dressing should usually be unnecessary. Patients may be given topical antibacterial cream to apply for a few days postoperatively. This cream may reduce local infection and prevents the exposed wound from sticking to underwear or bedclothes. Some surgeons use tissue glue to approximate wound edges in circumcision.

In many centers circumcision is carried out either under general anesthesia or with EMLA crème under local anesthesia by the Plastibell method first published in 1956. The device consists of a translucent plastic bell with a handle to position it over the glans once the foreskin is retracted. The bell has a deep groove and once the prepuce is brought over the foreskin is tightly tied by a non-absorbable thread over the device in this groove. The distal part of the foreskin is resected. The plastibell device will fall off between 4 and 12 days.

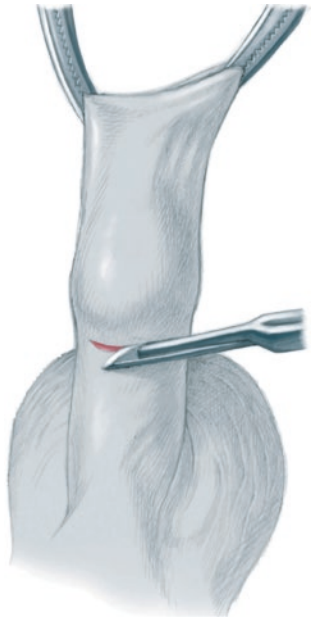

Fig. 65.1 Beginning the first circumferential incision

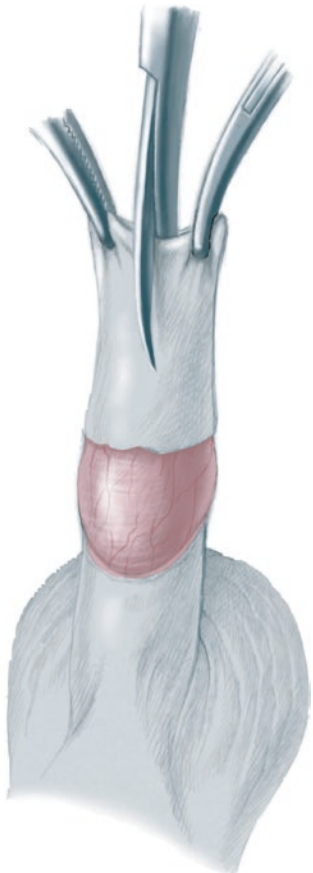

Fig. 65.2 Making a dorsal slit to join the inner and outer incisions

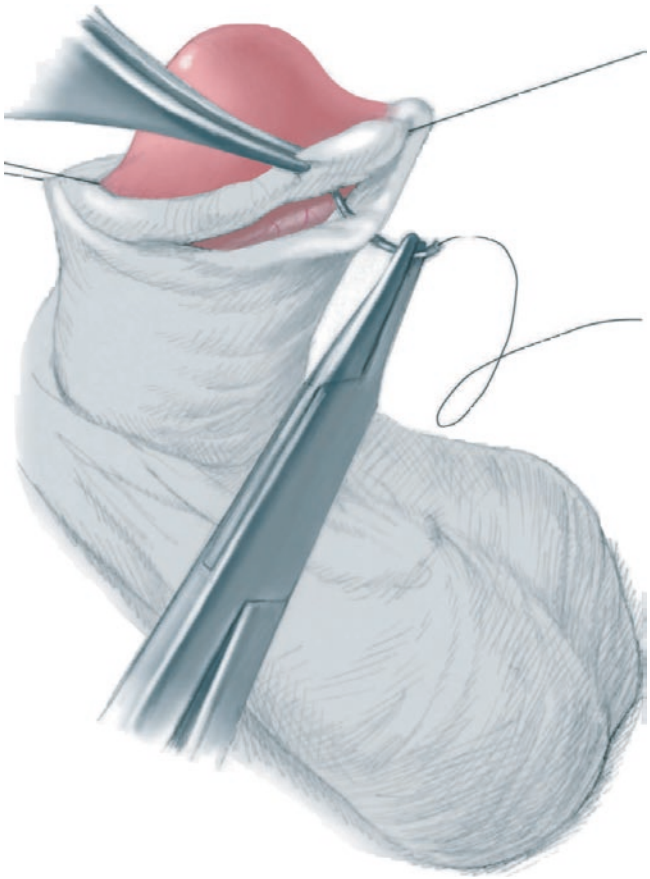

Fig. 65.3 Suturing the shaft skin to the cuff of distal skin

Fig. 65.4 The completed anastomosis

65.2.2 Meatotomy

The meatus is identified and a small lachrymal probe can be inserted to demonstrate it. The orifice is dilated sufficiently to allow one blade of a small haemostat to be inserted and directed proximally. The thin ventral tissue below the meatus is crushed in the midline by closing the haemostat. This usually incorporates at least 5 mm of tissue but does not extend more than halfway towards the corona.

The crushed tissue is now divided carefully with a fine pair of sharp pointed scissors (Fig. 65.5). The crushed tissue maintains haemostasis at this point. The neomeatus is further secured with fine monofilament absorbable sutures (6/0 Monocryl). These are placed at its apex and on either side (Figs. 65.6 and 65.7).

The new meatus should calibrate easily to at least 14F. If it does not, the ventral incision can be extended by repeating the above steps. Postoperatively, patients are supplied with chloramphenicol eye ointment. The small nozzle on the applicator can be inserted gently into the meatus to apply ointment within, twice daily for the next week. This gently opens the meatus and reduces inflammation as it heals.

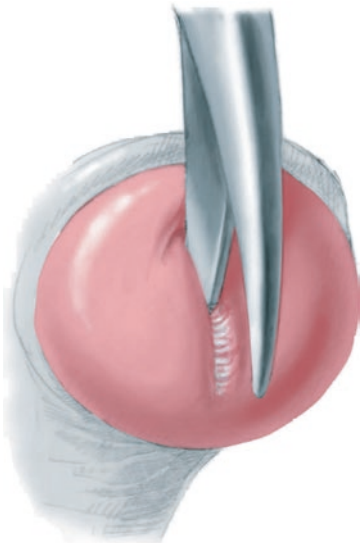

Fig. 65.5 Division of the ventral tissue below the meatus, previously crushed by a small haemostat

Fig. 65.6 Securing the neomeatus with fine, absorbable sutures

Fig. 65.7 Neomeatus with completed sutures

65.2.3 Repair of Buried Penis

Abnormality of peno-scrotal fusion and tethering of the penile corpora to the deep fascia give rise to the anomaly of buried penis (Fig. 65.8). The external prepuce is small and phimotic, and there may be an associated inner-preputial sac, sometimes referred to as a megaprepuce.

Pinching the tip of the prepuce and holding it forward identifies the line of demarcation between the scrotal and penile shaft skin. Deep palpation reveals normal-sized corpora and glans. A curved line is drawn along this line of demarcation to the apex of the scrotum. This marks the extent of what is to become the penile shaft skin. Care must be taken to ensure that this area is broad enough to envelop the penile shaft without tension.

Using a cutting diathermy needle, an incision is made along this line and deepened through the dartos fascia on to the deep fascia of the penile shaft. The shaft skin is lifted off the dorsum of the penis and dissection is continued around it using diathermy and blunt dissection with a gauze swab (Figs. 65.9 and 65.10). It is important to expose the deep fascia of the penile shaft and to extend this proximally to its base, in order to completely free it.

The preputial sac is opened by ventral incision into the orifice along the ventral raphe.

This allows retraction of the prepuce and exposure of the glans. There is a variable cuff of inner prepuce, which may be quite extensive and rugose. A glans suture is placed at this point to aid in retraction.

The inner prepuce is pulled back along the penile shaft and circumcised 6–8 mm below the glans, parallel to the corona (Fig. 65.11).

The skin is lifted from the penis by opening the cuff of inner prepuce ventrally (Fig. 65.12). The dorsal flap of skin is thinned by removing some of its subcutaneous fat layer with sharp scissors. This leaves a quadrilateral flap of skin from which skin cover is obtained.

The base of the flap is secured using a deep suture of 5/0 PDS (polydioxanone), which also picks up the deep fascia over the urethra (Fig. 65.13). This suture is placed at the apex of the original incision on each side.

Ventral closure of the skin is continued distally with interrupted 6/0 Monocryl sutures. The excess skin is trimmed dorsally with a scalpel. The skin suturing is completed circumferentially. The scrotal raphe is sutured to the proximal end of the shaft suture line, using a box stitch (Fig. 65.14). The scrotal defects on each side are closed with interrupted sutures.

Postoperatively, a hypospadias dressing is used, with a dripping stent, and left in situ for 1 week. Patients are given oral antibiotics and oxybutynin during this time.

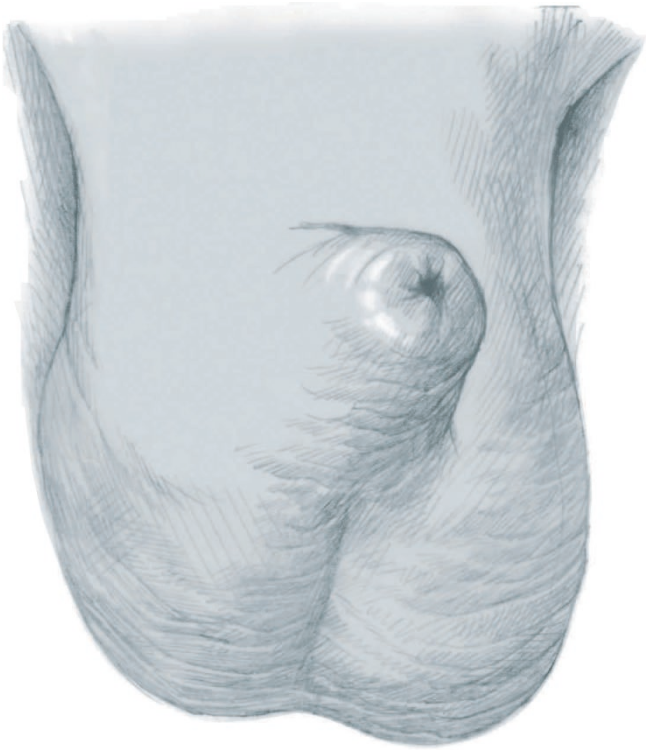

Fig. 65.8 Buried penis before surgery

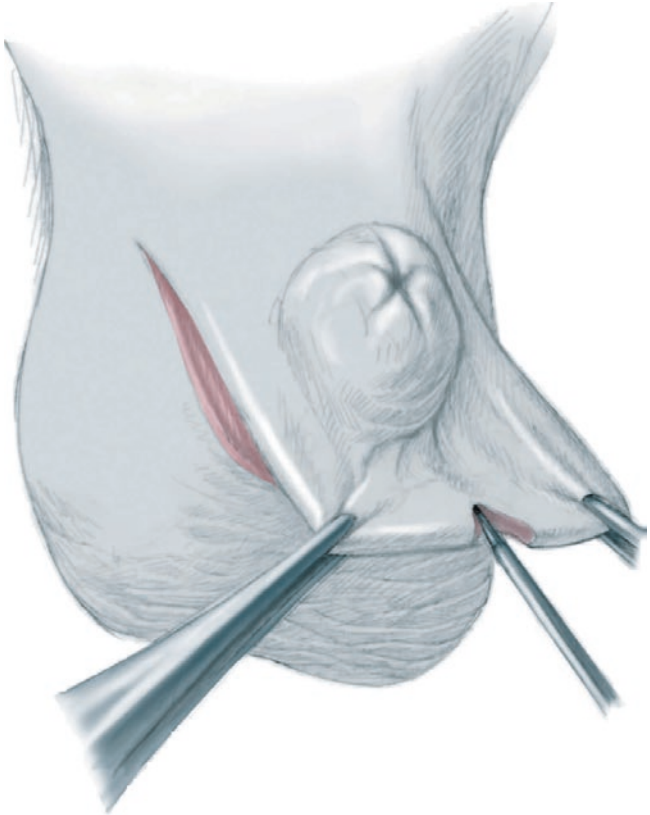

Fig. 65.10 Continued dissection

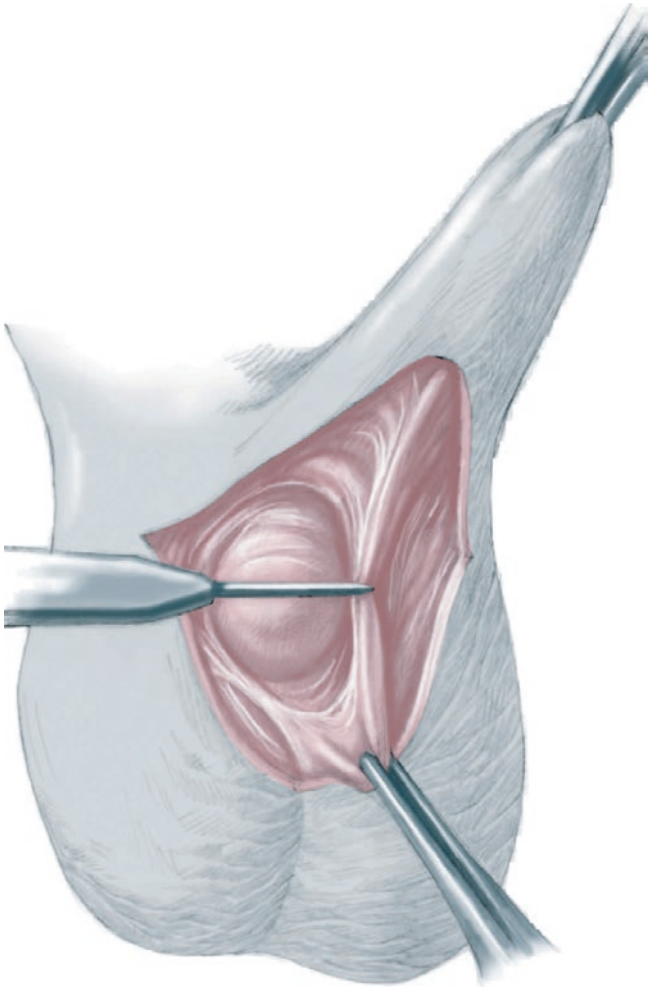

Fig. 65.9 Dissection of the shaft skin

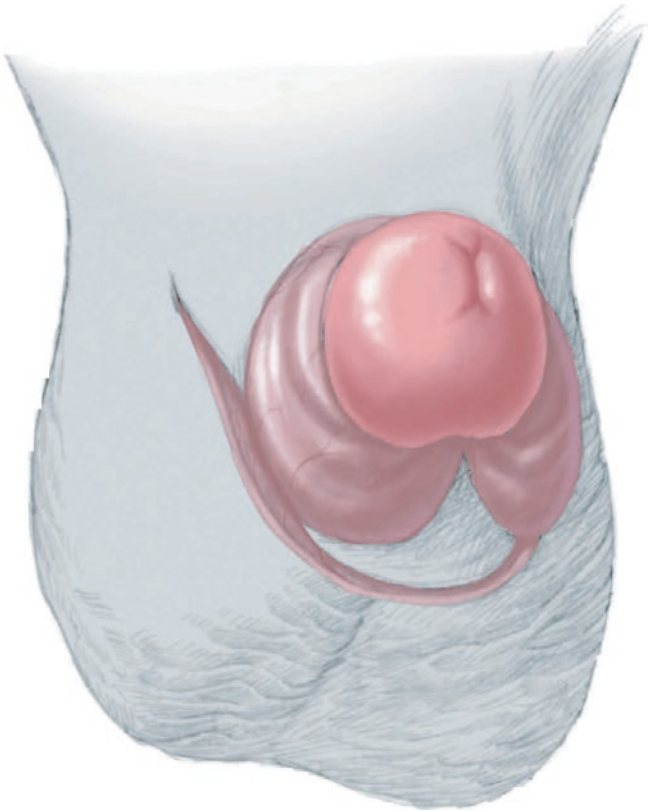

Fig. 65.11 The glans following circumcision

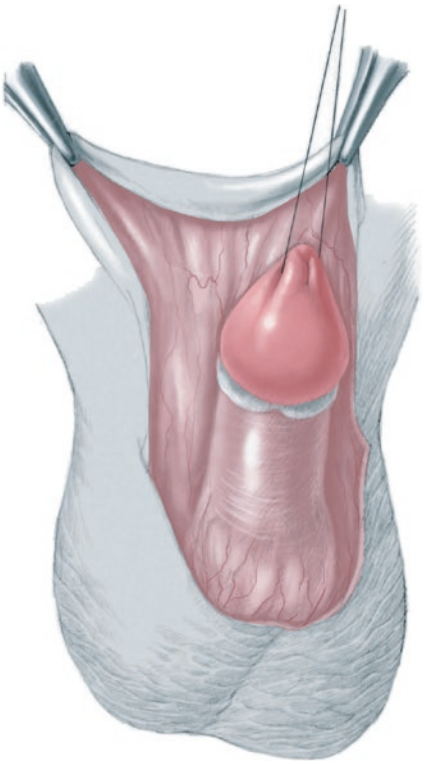

Fig. 65.12 Lifting the skin from the penis

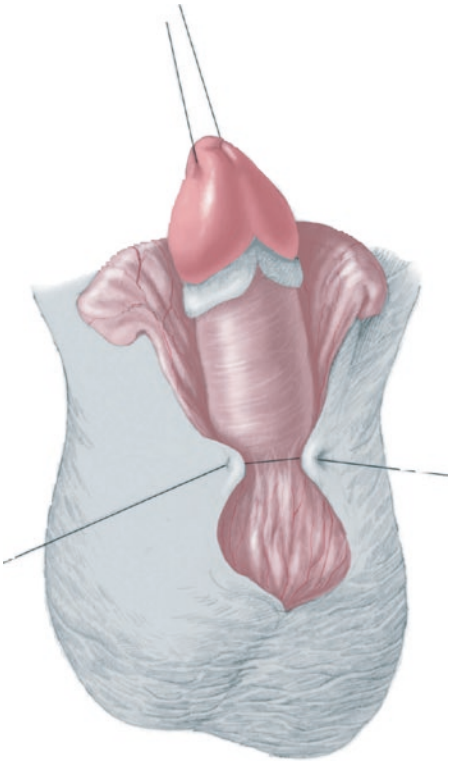

Fig. 65.13 Securing the base of the flap with a deep suture

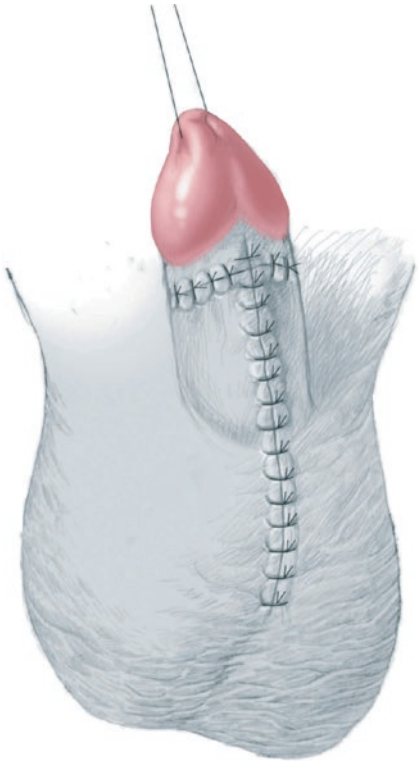

Fig. 65.14 The completed repair

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Orchidopexy

66

John M. Hutson

Undescended testis (UDT) is one of the commonest abnormalities in male infants. In preterm infants, the incidence may be 20% or more, as the final stage of descent from the groin to the scrotum normally occurs at about 25–35 weeks of gestation. About 4%–5% of males have undescended testes at birth, but more than half of these will continue to descend in the first 12 weeks postnatally. By 3 months postterm, the incidence of congenital cryptorchidism is 1%–2%.

There is considerable controversy about whether the subsequent testicular dysfunction is primary or secondary. Some authors have proposed that germ cell loss postnatally is secondary to a primary defect of the hypothalamic-pituitary-gonadal axis. Alternatively, there is strong evidence that postnatally high temperature interferes with normal testicular function, leading to germ cell depletion and risk of cancer in adulthood. Consensus is building that the crucial step in postnatal germ cell maturation is transformation of neonatal gonocytes into type A spermatogonia in the second 6 months after birth; hence the current recommendation for orchidopexy is at 6 months of age. Prevention of germ-cell loss is the aim of surgery, although this effect remains unproven.

The testis fails to remain in the scrotum in a significant number of older boys. They appear to have acquired cryptorchidism, which has been called ascending testis, gliding testis, or pathologically retractable testis. The abnormality is likely secondary to failure of the spermatic cord to elongate in proportion to the boy himself. (The spermatic cord length doubles from 5 cm to 8–10 cm in the first 10 years after birth.) At surgery, the major finding is a fibrous remnant of the obliterated processus vaginalis, linking acquired UDT with inguinal hernia. Occasionally the processus is still patent as a latent hernia. The indication for surgery in this acquired group is failure of the testis to remain in the scrotum without traction.

66.1 Surgical Technique

66.1.1 Standard Inguinal Operation

The patient is placed supine, with legs slightly apart, under full general anaesthesia (day surgery). Skin preparation extends from the umbilicus to below the scrotum. For congenital cryptorchidism, a standard inguinal operation is preferred. A transverse skin crease incision is made over the inguinal canal (one finger breadth above pubic tubercle) from level with the tubercle to the mid inguinal point (Fig. 66.1).

The incision is deepened through fat to expose the white fibrous layer of the superficial fascia (of Scarpa), which is opened formally (diathermy or scissors). The superficial inferior epigastric vein is swept aside or coagulated and divided formally.

Beneath Scarpa's fascia, the external oblique muscle is distinguished by the oblique orientation of its fibres. Square retractors under Scarpa's fascia reveal the lower edge of the muscle and the inguinal ligament; sweeping movements with the closed scissors are effective to reveal the spermatic cord where it emerges from the external inguinal ring.

The inguinal canal is opened either with a scalpel incision in external oblique aponeurosis parallel to the inguinal ligament, or by opening the external inguinal ring with scissors and slitting the aponeurosis laterally (Fig. 66.2). Small artery forceps on the edges of the external oblique control them and allow easy identification at the end of the procedure. The ilioinguinal nerve is identified on the surface of the cremaster fascia and avoided.

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https://doi.org/10.1007/978-3-662-56282-6_66

The spermatic cord is separated from the external oblique aponeurosis and the cord and testis (usually just outside the external ring) are mobilized out of the wound by blunt dissection (Fig. 66.3). The distal gubernacular attachment is divided carefully (diathermy or scissors), avoiding any long-looping vas deferens. A small clamp on the gubernaculum or distal tunica vaginalis allows traction on the spermatic cord. The spermatic cord is dissected by first isolating and stripping off with blunt forceps any cremaster muscle fibres. Occasionally the cremasteric artery (deep to the cord) needs diathermy.

The remnant of the processus vaginalis is usually obvious in cryptorchidism as a shiny white, translucent inelastic layer covering the vas deferens and testicular vessels. The hernial sac is grasped with blunt forceps and the vas and vessels are carefully swept off its posterior surface *en masse* (Fig. 66.4). The vas deferens must be identified formally during this dissection, as it is closely adherent to the posterior surface of peritoneum. The vas deferens and gonadal vessels are retracted away from the sac, which then can be clamped and divided.

The hernial sac is pulled cranially (Fig. 66.5), allowing the vas deferens and vessels to be separated bluntly from the posterior surface right up to the internal inguinal ring. At this point the vas and vessels begin to diverge and the peritoneum becomes more opaque.

The vas deferens hooks medially around the edge of the transversalis fascia and adjacent inferior epigastric vessels, whereas the spermatic vessels pass laterally and cranially into the retroperitoneal space. A small Langenbeck retractor is placed behind the peritoneum and the retroperitoneal space is opened with blunt dissection. Any lateral fibrous attachments to the vessels are identified by traction and then divided (Fig. 66.6). This should provide adequate length to allow the testis to reach the scrotum. If there is inadequate length of the vas deferens, the inferior epigastric vessels can be isolated and a hole made in the posterior wall of the inguinal canal medial to the vessels. Once the vas deferens has been mobilized carefully, the testis is redirected medial to the vessels: further traction on the testis will identify any remaining fibrous strands that need division. The processus vaginalis is twisted to exclude intraperitoneal contents and is transfixed and ligated with absorbable suture.

A finger is introduced through the wound and bluntly pushed down to the scrotum. The scrotal skin is immobilized between the internal finger and the thumb and then a skin incision is made (either horizontal or vertical in the midline) (Fig. 66.7). A subcutaneous pouch is developed with scissors or small artery forceps with the finger still in place (Fig. 66.8). Bleeding is managed by meticulous diathermy to avoid subsequent scrotal haematoma.

A small artery forceps is passed into the pouch and pressed against the deep fascia stretched over the finger tip, which then guides the forceps back out through the inguinal incision.

The gubernaculum or tunica vaginalis is grasped by the forceps and the testis is gently drawn down to the scrotum (Fig. 66.9) and out through the lower incision. At this point the tunica can be opened and everted and any testicular appendages are excised. The testis can be held in the scrotal pouch by a small suture through the lower septum and the tunica albuginea. Alternatively, the neck of the scrotum can be tightened with a suture that gathers the fibro-fatty tissue around the spermatic cord. Sometimes the buttonhole in the tissues at the neck of the scrotum is small enough to hold the testis without any suture.

The testis is nestled into its new subcutaneous pouch and the scrotum is closed with subcuticular suture (Fig. 66.10). The inguinal incision is closed with a running suture in the external oblique aponeurosis (the artery forceps placed on the edges at the start make identification easy). Scarpa's fascia is closed with one or two sutures, and the skin is closed with subcuticular suture. A waterproof dressing is applied to both wounds.

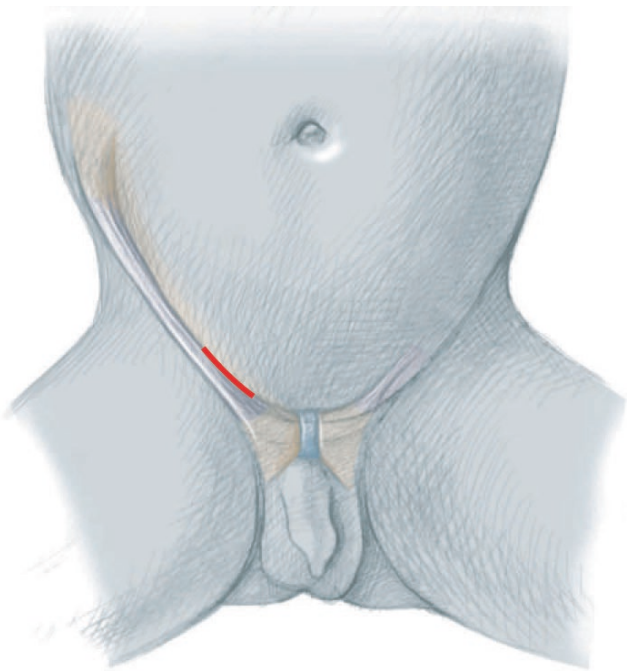

Fig. 66.1 Transverse skin crease incision

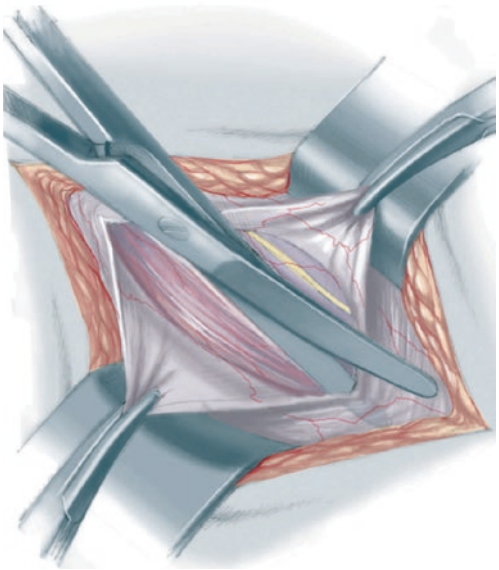

Fig. 66.2 Opening the inguinal canal

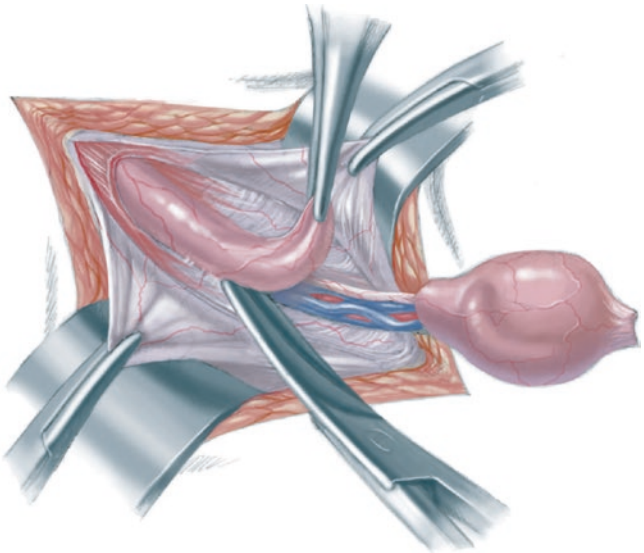

Fig. 66.4 Grasping the hernial sac and sweeping the vas and vessels off its posterior surface

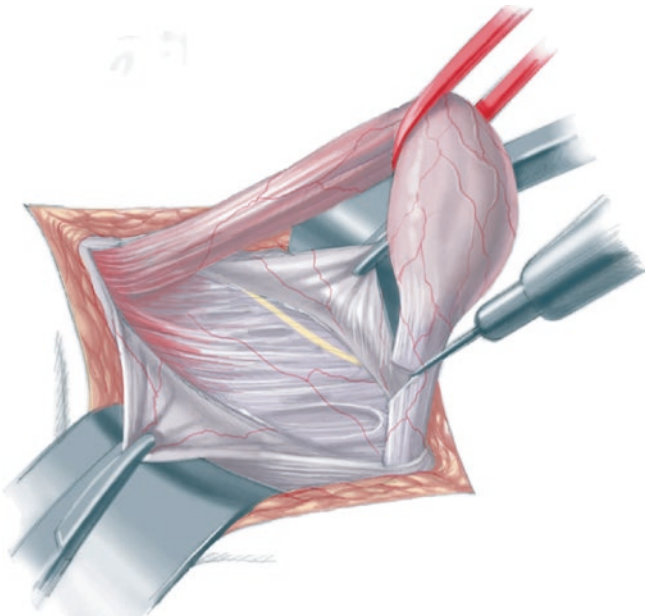

Fig. 66.3 Mobilizing the testis and dividing the distal gubernacular attachment

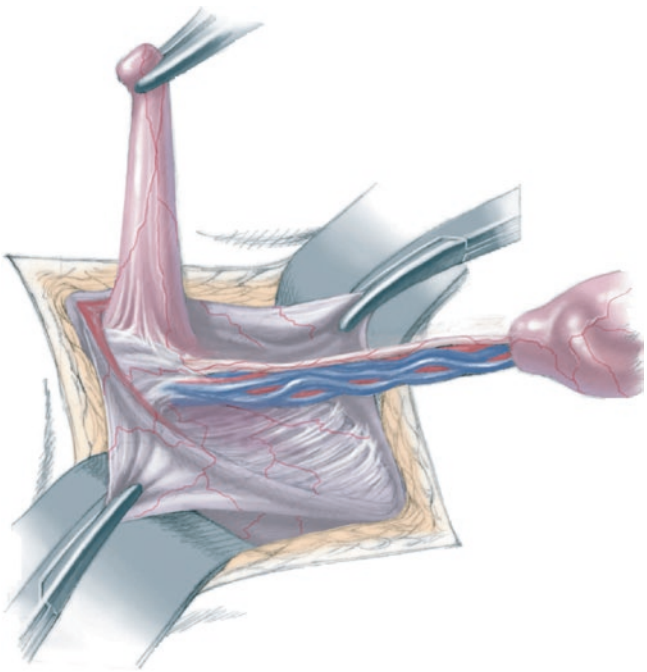

Fig. 66.5 Pulling the hernia sac cranially

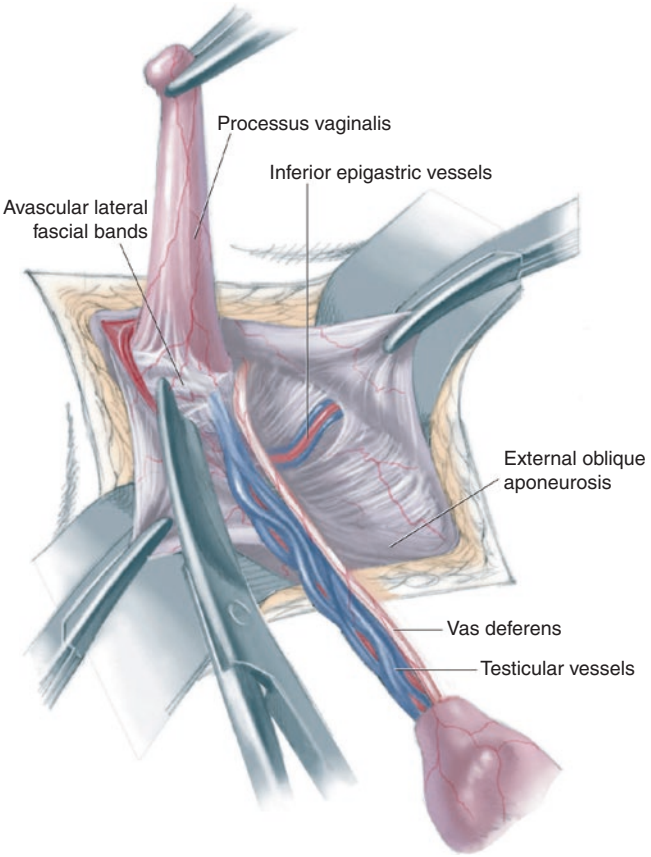

Fig. 66.6 Dividing lateral fibrous attachments to the spermatic vessels

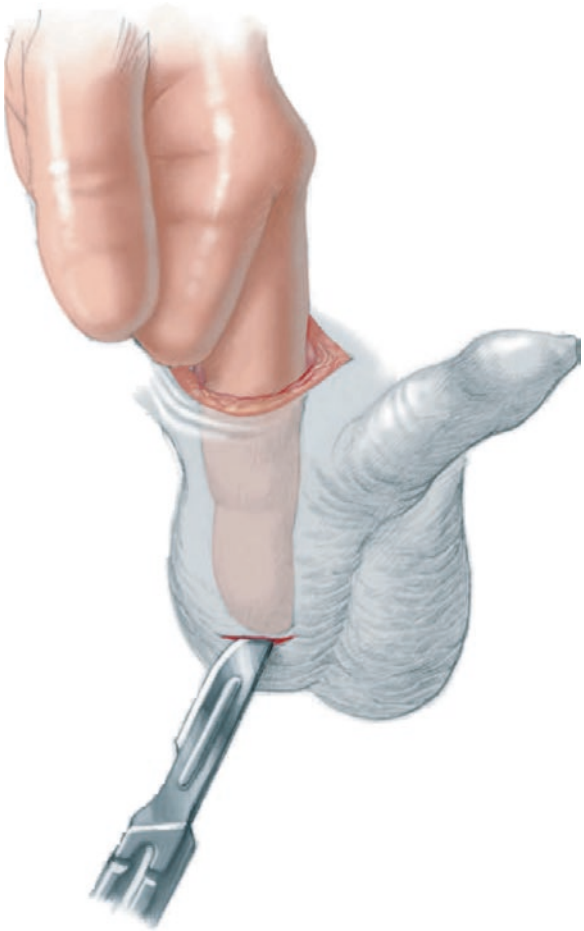

Fig. 66.7 Making a skin incision in the scrotum

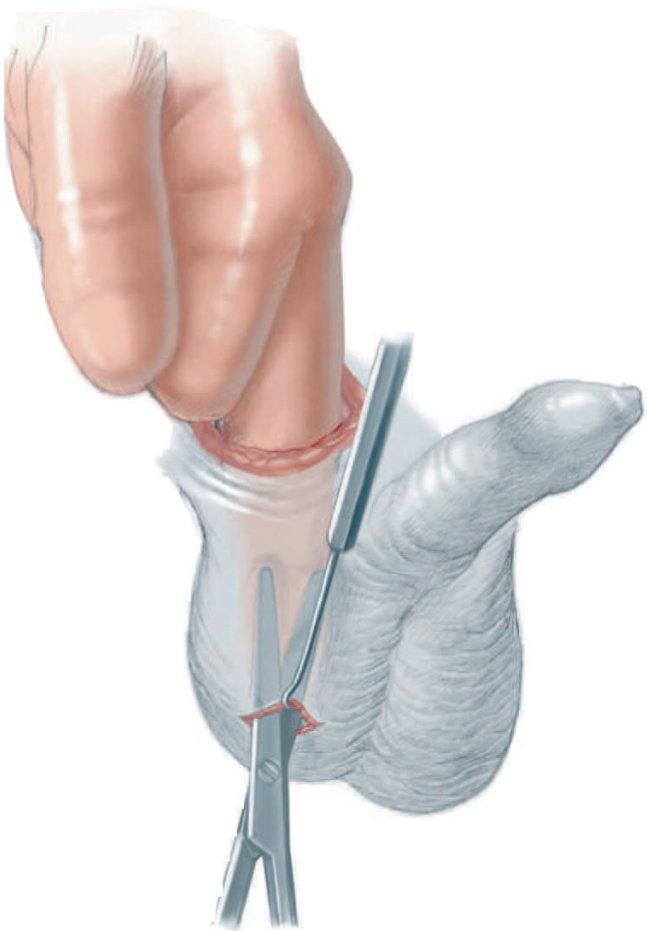

Fig. 66.8 Developing a subcutaneous pouch

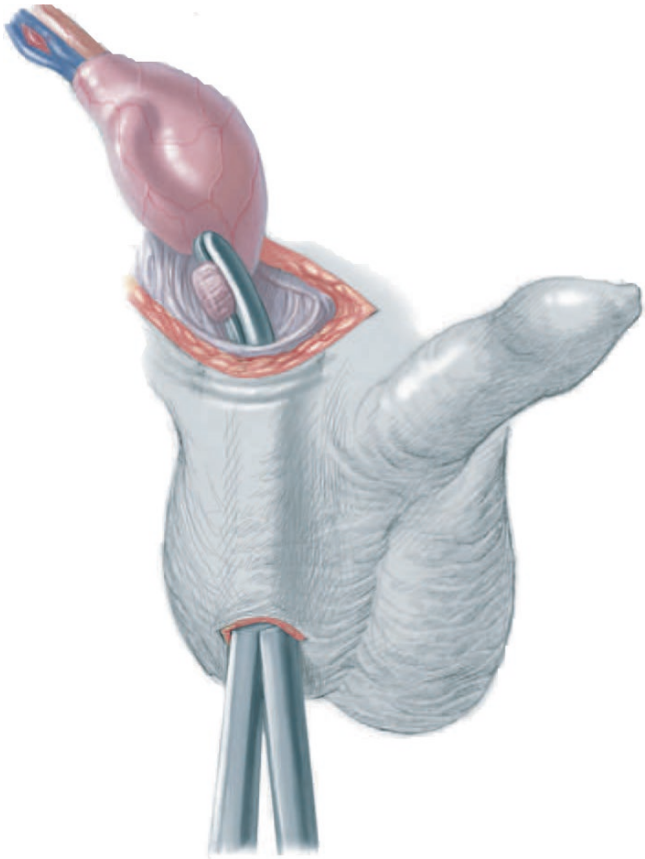

Fig. 66.9 Drawing the testis down to the scrotum

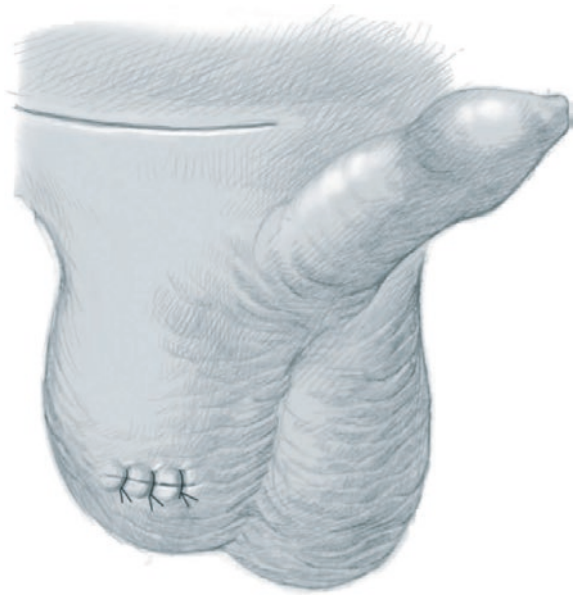

Fig. 66.10 Sutures closing the wounds

66.1.2 Diagnostic Laparoscopy

Diagnostic laparoscopy (Fig. 66.11) is the first step in the management of a nonpalpable testis. There are three likely findings at laparoscopy:

- *Blind-ending vas and vessels (vanishing testis)*: Viable testicular remnants have been noted in 6%–20% of vanishing testes. It has been suggested that inguinal exploration should be carried out in these patients to remove testicular "nubbins" to prevent future risk of malignancy. Contralateral fixation orchidopexy may be indicated.
- *Cord structures entering the internal ring*: Inguinal exploration is carried out, and if a viable testis is found, the testis is relocated to the scrotum. If a remnant "nubbin" is found, it is excised *in toto* to remove the nidus of germ cells in order to prevent later malignancy.
- *A viable intra-abdominal testis*: The factor limiting the relocation of an intra-abdominal testis to the scrotum is the length of the gonadal vessels. The testis is supplied by three arteries—the main testicular, the vasal, and the cremasteric arteries. The decision to perform a Fowler-Stephens orchidopexy or one of its modifications must be made before any extensive inguinal exploration that disrupts the collaterals. The viable intra-abdominal testis is treated by laparoscopic clip ligation of the main spermatic vessels, and orchidopexy (based upon vasal vessels and the collaterals) is performed at a later stage.

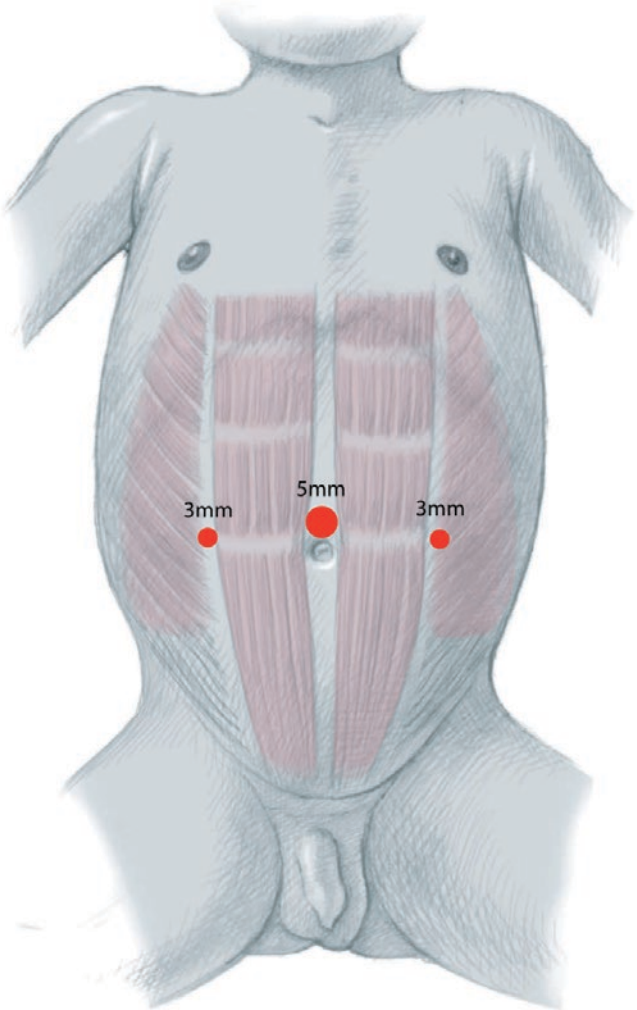

Fig. 66.11 Port positions for laparoscopy in a child with a nonpalpable testis

66.1.3 Orchidopexy

Intra-abdominal testes are usually close to the internal inguinal ring. Evidence that a one-stage orchidopexy is feasible is obtained by grasping the testis and seeing if it can be pulled to the opposite internal inguinal ring (Fig. 66.12). If so, then there will be adequate length of the testicular vessels without the need to divide them, and the testis can be relocated into the scrotum via a hole in the conjoined tendon medial to the inferior epigastric vessels. By open inguinal exposure or a grasper placed up the inguinal canal from a small scrotal incision, the grasper is pushed through the posterior inguinal canal wall (under direct laparoscopic vision to ensure that the inferior epigastric vessels are preserved) and the mobilised testis, along with a generous cuff of peritoneum covering the testicular vessels, vas deferens, and adjacent collaterals, is pulled down to the scrotum and anchored there with a suture.

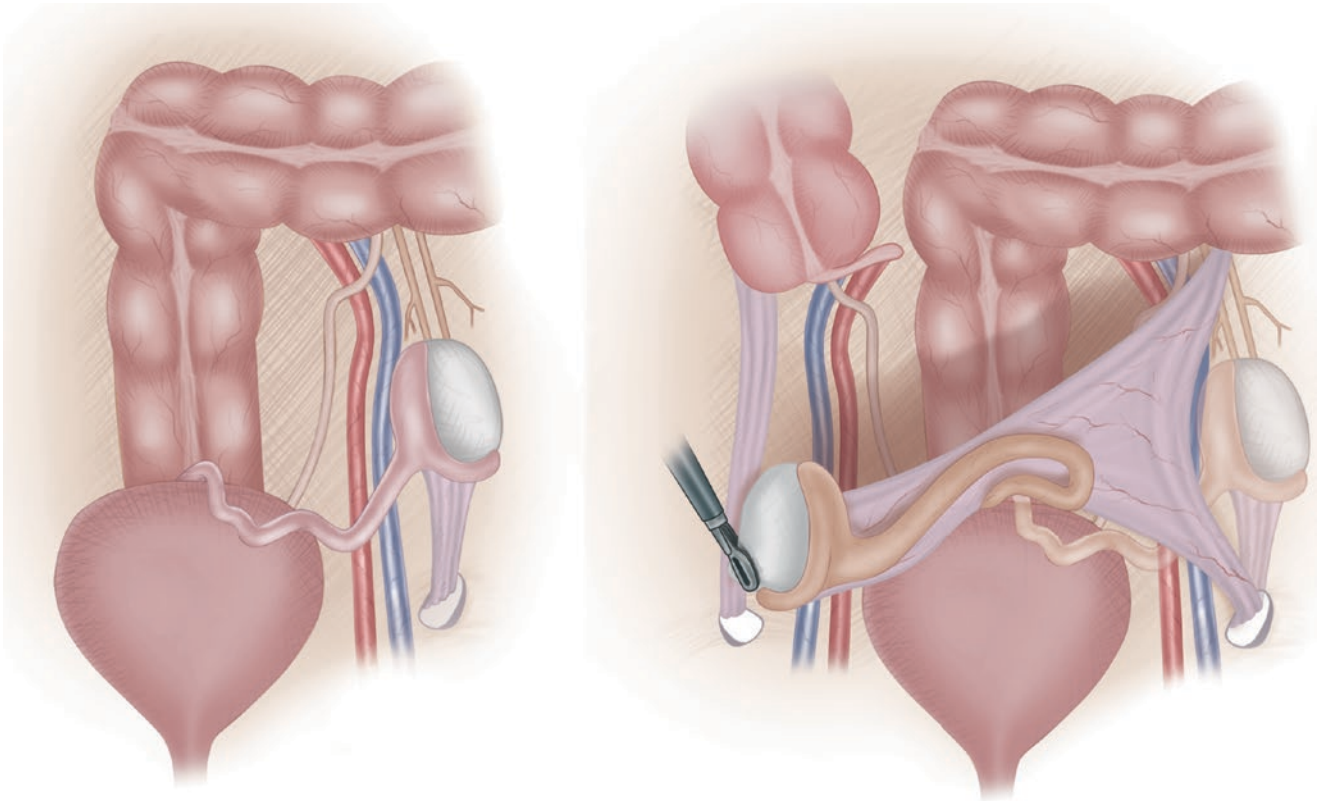

Fig. 66.12 *Left*, The intra-abdominal testis is usually close to the internal inguinal ring (IIR). *Right*, The testis can be pulled to the opposite IIR to gauge how much mobility there is in the mesorchium: orchidopexy should be OK if testis reaches opposite IIR

66.1.4 Fowler-Stephens Orchidopexy

If the intra-abdominal testis is higher, the colon is mobilised to expose the gonadal vessels proximally, so that they can be tied or clamped far enough away from the testis to preserve the collateral circulation around the head of the epididymis. This is the first stage of the two-stage Fowler-Stephens operation, which is based on the principle that the testicular artery, which is too short to reach the scrotum, supplies only 60%–70% of the gonadal blood supply, so it can be divided safely as long as the collateral supply remains intact (Fig. 66.13).

Six months later, after adaption and enlargement of the artery of the vas, the peritoneum around the testis and vas deferens is divided so that it is fully mobilised. The scrotum is then opened and a grasper is inserted up to and in the external ring; it is pushed through the conjoined tendon medial to the inferior epigastric vessels to enter the peritoneal cavity. The testis, along with the peritoneum containing the collateral vessels, is then grasped and pulled down to the scrotal incision. It is usually necessary to ensure that the new tunnel in the back of the inguinal canal is stretched up adequately beforehand so that the testis will fit through comfortably without compressing the tissue or constricting the vessels. The testis is fixed in the scrotum as in a normal orchidopexy (Fig. 66.14).

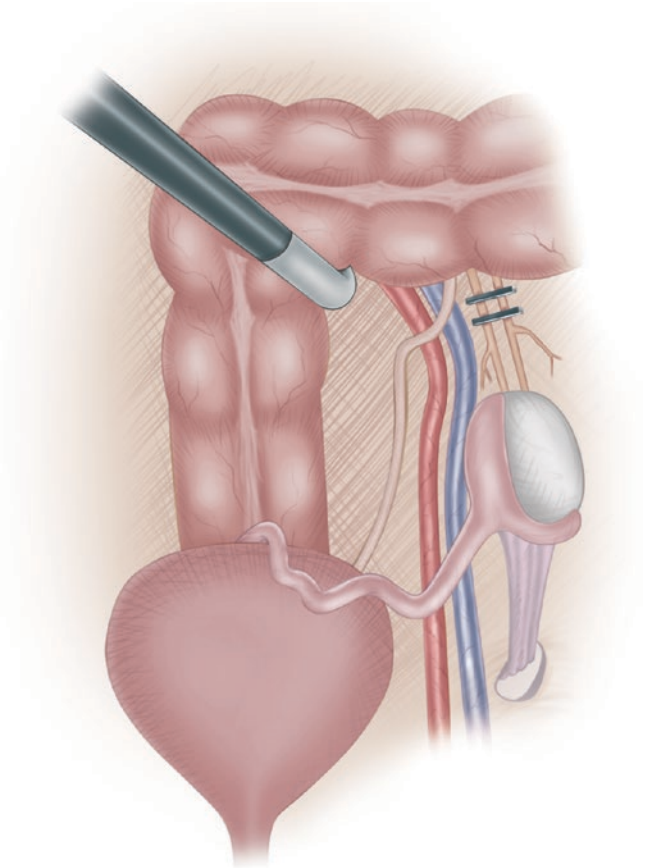

Fig. 66.13 *Left*, Creating a peritoneal flap the shape of a tennis racket to preserve any collateral vessels around the testis and epididymis. *Right*, Once testis has been mobilised on the peritoneal flap, it can be pulled to the scrotum, often through the back wall of the inguinal canal medial to inferior epigastric vessels, as in the Prentiss manoeuvre

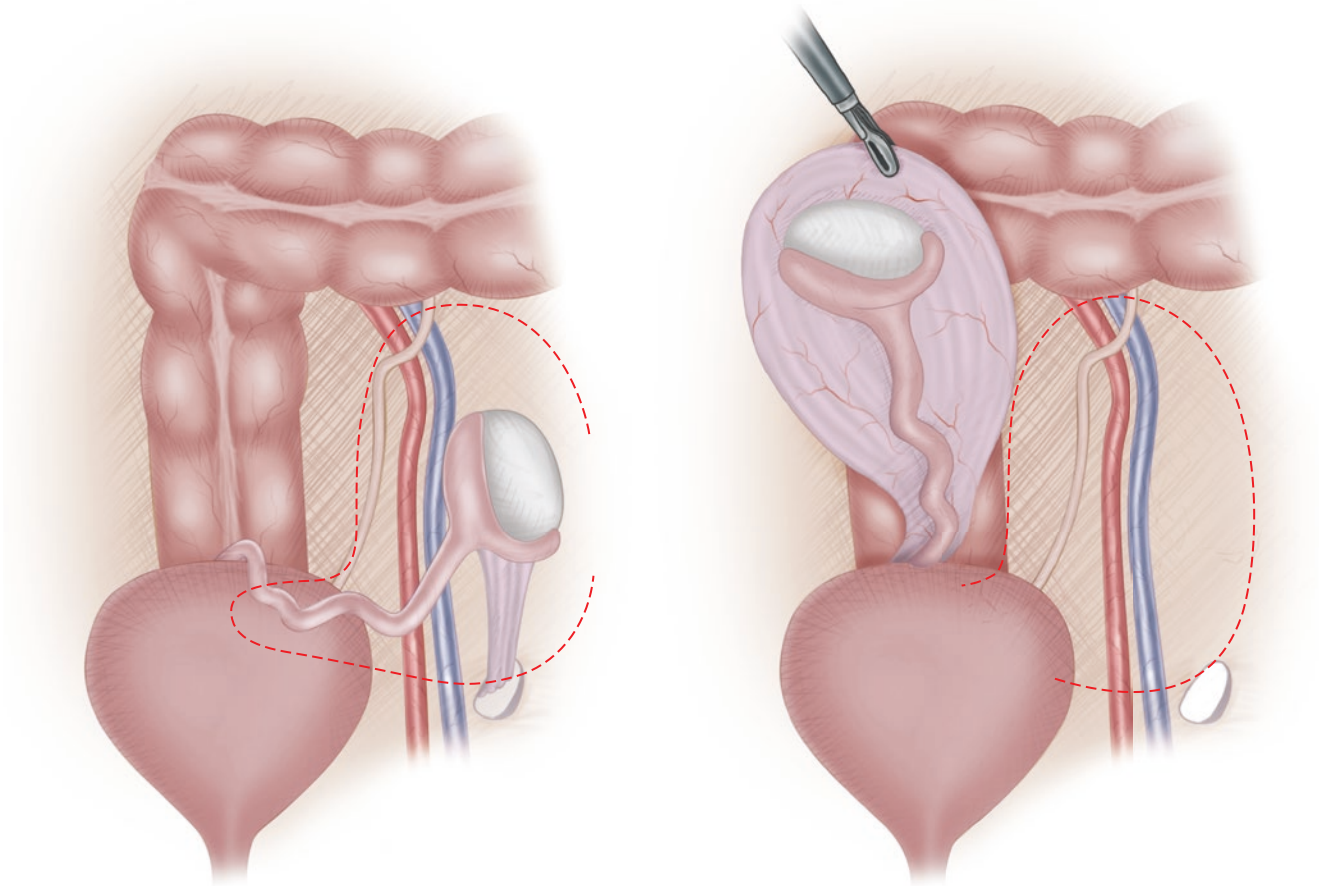

Fig. 66.14 In two-stage Fowler-Stephens operation, the colon is mobilised to enable the testicular vessels to be clipped or ligated far enough away from the epididymal head to avoid damage to the collateral blood supply

66.2 Results and Complications

Postoperative recovery from all orchidopexy procedures is rapid, with return to full activity within a few days. The infant is reviewed again in 6–12 months to ensure that atrophy has not occurred. Boys with primary maldescent (especially bilateral) and those with impalpable testes would be advised to return at 14 years of age for review of pubertal development and discussion about prognosis for cancer and fertility.

The most frequent complications are wound infection or haematoma, both of which can be avoided by meticulous haemostasis at operation and leaving the waterproof dressing in place for at least a week. The risk of testicular atrophy should be less than 5%; in most series it is 1%–2%. Depending on the method used to fix the testis in the scrotum, there is a small risk of retraction of the testis back into the groin, requiring a secondary orchidopexy.

In the past, the estimated risk of testicular cancer (within 15–40 years) has been approximately 5–10 times higher than in a normal testis, but most paediatric surgeons anticipate that orchidopexy in early infancy (<1 year of age) may avoid this danger.

The estimated risk of infertility is about 30% for bilateral undescended testes; the risk is lower (but not normal) for unilateral cases. Whether these risks will disappear with early surgery remains unknown at this time. Because epididymal anomalies are commonly associated with cryptorchidism, it is likely that a small number of boys may be infertile subsequently because of epididymal-testicular dissociation, even if germ cell maturation is normal.

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Varicocele

67

Michael E. Höllwarth

Testicular varicocele is characterized by variceal dilatation of the veins in the pampiniform venous plexus secondary to incompetent valves in the testicular vein. Varicoceles are almost always localized on the left side, supposedly because the left testicular vein drains via the left renal vein, which offers higher resistance to the bloodstream than the right testicular vein, which enters the vena cava directly. The age group most frequently affected is older boys and adolescents. Symptoms are rare. Sometimes, an ill-defined discomfort in the scrotum (such as a dragging sensation) is reported.

On physical examination, grade III varices may be seen through the scrotal skin. The characteristic soft nodular mass, which is described as feeling like "a bag of worms", is palpable, but not visible, in grade II varicoceles. It becomes more prominent with increased venous filling due to gravity (in erect position) or to venous outflow obstruction by an intra-abdominal pressure surge. This pressure can be provoked by a Valsalva manoeuvre, which is necessary to render a grade I varicocele obvious. In contrast, the varicocele can be neither seen nor felt in the supine patient (particularly in a Trendelenburg position), as the veins empty.

Clinical relevance derives from the fact that varicoceles can result in testicular atrophy and infertility. The pathophysiological mechanism has not yet been elucidated. Production of antibodies in reaction to increased temperature within the scrotum or to a leak in the blood-testis barrier has been suggested to explain bilateral damage. Semen analysis in 30%–50% of affected men show abnormalities that may improve significantly after resolution of scrotal venous hypertension, provided that the treatment is not delayed until the damage has become irreversible. Treatment of varicoceles is therefore indicated in children and adolescents to prevent such damage.

Various surgical methods are available, the most popular of which have been described by Palomo and Ivanissevich. Palomo's method consists of high mass ligation with or without resection of 3 cm of the testicular veins and artery. This procedure can be done by an open surgical retroperitoneal approach via an incision in the left iliac fossa, or by transperitoneal laparoscopy. Ivanissevich popularized dissection and ligation of two thirds of the veins of the pampiniformis plexus via an inguinal exploration. We prefer the minimally invasive method described by Tauber (Tauber and Johnsen 1994), as described in the following pages. This method, which is simple and equally effective, can be performed under local anaesthesia in most patients (Figs. 67.1, 67.2, 67.3, 67.4, and 67.5).

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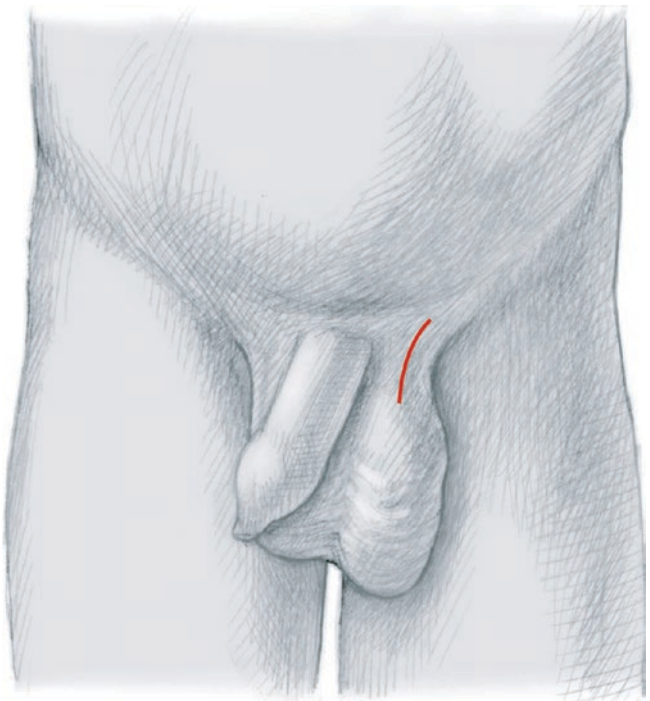

Fig. 67.1 The patient is positioned supine on the operating table. The region from the left external inguinal ring to the upper third of the scrotum is shaved. After adequate regional infiltration of anaesthesia, a 3-cm longitudinal incision is made in the direction of the spermatic cord in the uppermost part of the scrotum

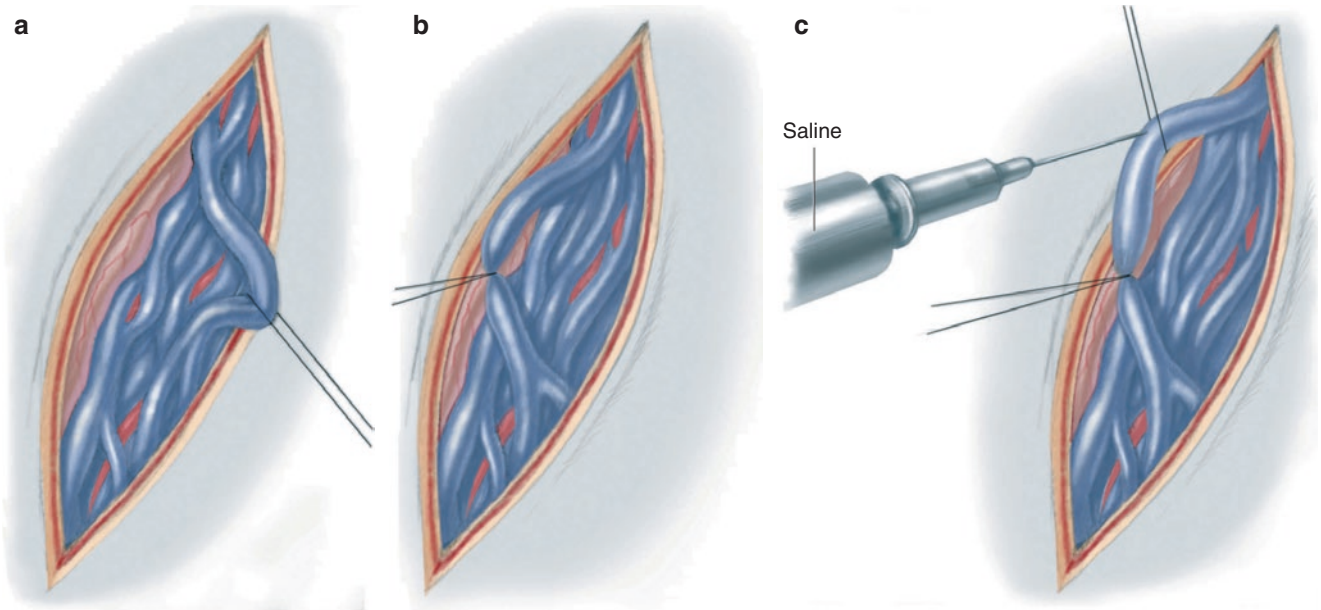

Fig. 67.2 (**a**–**c**) The subcutaneous tissue is divided and the outer fascia of the cord is opened longitudinally. With fine forceps and a right-angled clamp, one major vein is isolated from the pampiniform plexus over a distance of about 1.5 cm. The vein is ligated distally; a second suture is pulled around it proximally. The vein is then cannulated with an 18-gauge needle in cephalad direction. If saline can be injected without problems, the proximal thread is held just firmly enough to maintain the cannula in the vein

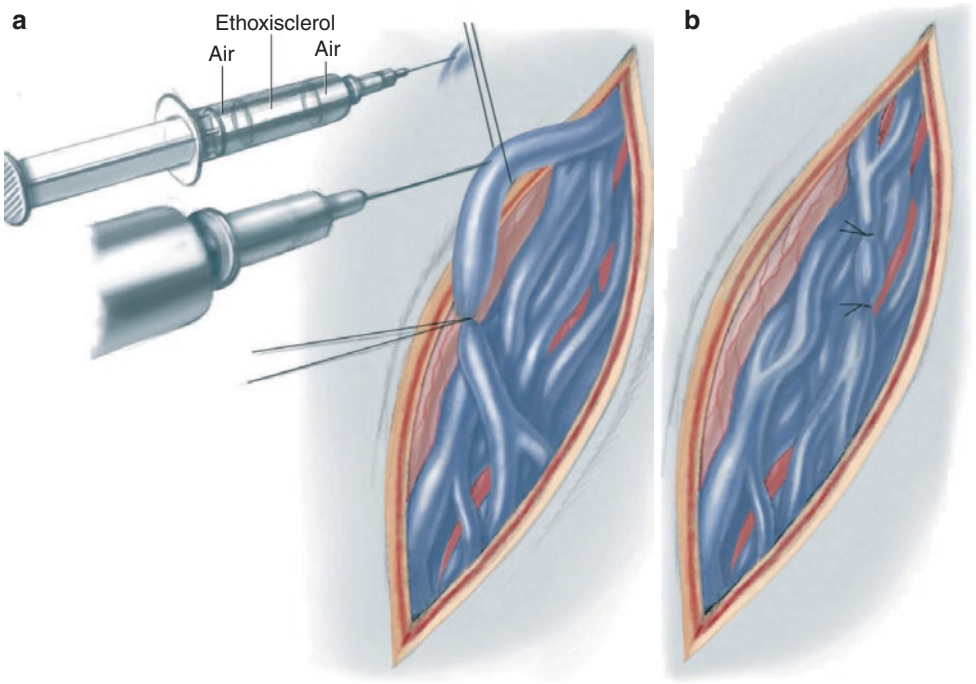

Fig. 67.3 (**a**, **b**) Diluted contrast medium is injected under fluoroscopic control to ascertain that the cannulated vein drains into the left renal vein via the testicular vein without aberrant flow into pelvic veins. If pelvic collaterals are outlined, the needle must be removed and the vein ligated. Then another vein is punctured and tested for flow exclusively towards the renal vein in the same way. Once an appropriate vein has been found, 3 mL of a sclerosing agent (*e.g.*, ethoxysclerol 1–3%) and 2 mL of air are aspirated into a 5-mL syringe. While the anaesthetist increases the inspiratory pressure to simulate a Valsalva manoeuvre, the surgeon injects 1 mL air, followed by the whole amount of the sclerosing material, and then the remaining 1 mL of air. The Valsalva manoeuvre is continued for approximately 30 s. Subsequently, the needle is removed and the plexus vein is ligated proximal to the access site. The incision is closed with absorbable subcutaneous 5/0 and subcuticular 6/0 sutures

Fig. 67.4 The high retroperitoneal ligature of the testicular vein or veins, as introduced by Palomo. The procedure was originally performed by a left lower-quadrant extraperitoneal laparotomy (McBurney on the left side). When the peritoneum is reached, it is gently pushed away so that the left retroperitoneal space can be reached. The testicular vessels are identified and the vein (or two or three veins) is ligated and resected over a distance of 2–3 cm. Sometimes it seems difficult to identify the vein or separate it from the artery. Therefore, some authors prefer to ligate all vessels, including the artery. Recently, the laparoscopic minimally invasive procedure has been the preferred method. The surgeon stands on the right side of the patient. A 5-mm laparoscope for the optic is inserted through the umbilicus. Two other instruments (2-mm or 5-mm) are inserted, one lateral to the left rectus muscle margin and the other above the bladder

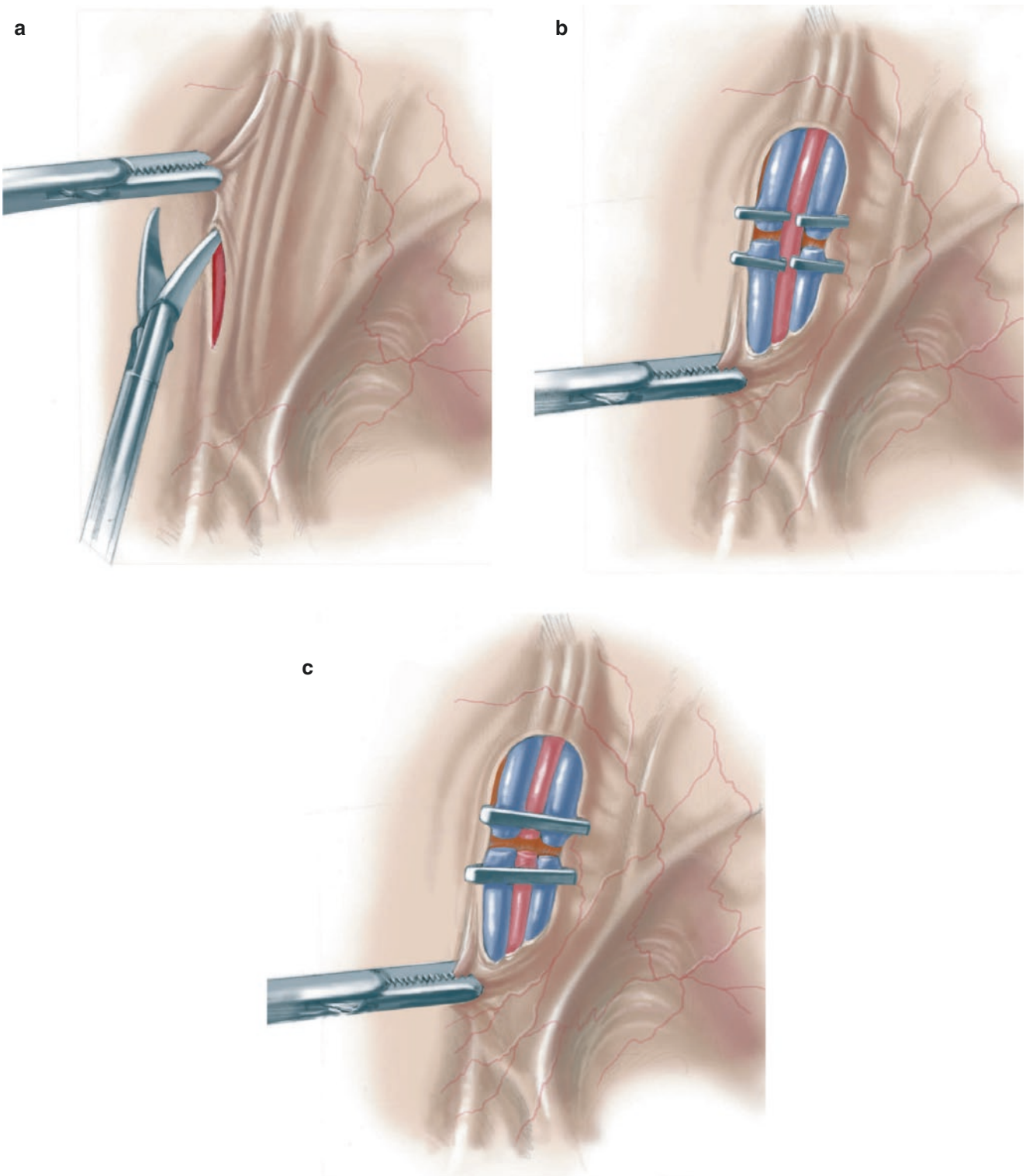

Fig. 67.5 Adhesions to the sigmoid colon can be transected. The peritoneal reflection over the vessels is incised (**a**) and the veins are identified and transected between clips (**b**, **c**). Some authors resect approximately 1 cm of the vessels in order to prevent any recurrence. Electrocautery should be used very carefully because underlying nerve fibres may be damaged, leading to dysaesthesia at the left thigh. If bleeding occurs while the veins are being separated, the only option is to ligate all vessels without sparing the artery. Testicular blood flow is provided through the collateral vessels from the vas. Recently, a lymphatic-preserving procedure close to the internal inguinal ring has been advocated to reduce the incidence of postoperative hydrocele

67.1 Results and Conclusions

Long-term follow-up studies of treated varicoceles have shown a small but consistent recurrence rate. With the artery and vein ligature technique, performed either open or by laparoscopy, the recurrence rate is between 5% and 16%. The rate is significantly lower (about 1.5%) when the testicular artery is ligated together with the vein. Tauber's antegrade sclerotherapy, sparing the testicular artery, is also a minimally invasive method that is even easier and faster to perform than the laparoscopic procedure. In our hands, the recurrence rate after 1 year has been 6%, but repeated sclerotherapy can be easily performed. Temporary hydrocele occurs rarely after retrograde sclerotherapy, indicating that the pathophysiologic mechanism may be driven not only by the ligation of lymphatic vessels running along the spermatic artery and vein but also by a temporary increase of intravenous pressure until collaterals along the vas drain the venous blood adequately.

Suggested Reading

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Genitoplasty for Congenital Adrenal Hyperplasia

68

Martin A. Koyle and Richard S. Hurwitz

The finding of ambiguous genitalia in the newborn creates the need for a team of experts, the DSD (disorder of sexual differentiation) team, to be assembled in order to determine the genetic sex, identify any underlying biochemical abnormalities, and provide education and support for the family. This team may include a neonatologist, a pediatric endocrinologist, a geneticist, a pediatric surgeon or pediatric urologist, a social worker, and a psychiatrist. Whereas DSD was once considered to be a medical and surgical emergency, today it represents more of a social urgency, but prompt medical evaluation is also necessary because of potential metabolic and electrolyte disturbances.

Congenital adrenal hyperplasia (CAH), a form of 46XX DSD, is the most common cause of masculinization of the female newborn. CAH is a disorder of adrenal steroidogenesis in which an enzyme deficiency is present in the cortisol pathway, leading to reduced cortisol production and the subsequent overproduction of adrenal androgens. The most common of these deficiencies is an autosomal recessive disorder, 21-hydroxylase deficiency. As many as 75% of patients will have a life-threatening salt-losing metabolic condition due to deficient aldosterone production. In utero exposure of the 46XX female fetus to adrenal androgens results in varying degrees of virilization of the external genitalia and the distal vagina. Müllerian precursors of the internal genitalia, fallopian tubes, uterus, and proximal vagina develop normally in the absence of müllerian inhibiting substance.

Feminizing genitoplasty is composed of clitoroplasty, vaginoplasty, and labioplasty. From a surgical perspective, it is important to emphasize that CAH represents a spectrum disorder in which the ultimate reconstruction should be specifically tailored to each individual patient. This chapter focuses on feminizing surgery for patients with CAH.

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68.1 Perioperative Evaluation

Individual evaluation is essential to determine the type of vaginoplasty best suited to each patient. The goal of the perioperative evaluation is to understand the anatomy of the urogenital sinus (UGS). The key information needed is the length of the UGS and the specific location of the vaginal confluence in relation to the bladder neck and the UGS orifice. The size of the vagina should also be determined. Vaginal confluences have been described as high, intermediate, and low, but in reality there exists a true spectrum from the very difficult suprasphincteric confluence to simple posterior labial fusion. Most of these patients will have had a pelvic ultrasound as part of the evaluation of their ambiguous genitalia, but this study is rarely helpful in the planning of genitoplasty surgery. Genitography, although not always definitive, is still a standard procedure in many centers performing feminizing genitoplasty. Whether to perform it depends on the discretion of the surgeon and the center's philosophy.

Endoscopy sometimes can be the surgeon's best friend for defining complex UGS anatomy. It helps to document the anatomy precisely and to determine the surgical approach. In the past, endoscopy was often done under a separate anesthetic, but today it is usually performed coincidentally with the reconstructive surgery. Rink *et al*. described the "PVE" classification, in which the length and width of the phallus (P) is measured in centimeters, the location of the vaginal confluence (V) is recorded in centimeters from the bladder neck and from the UGS opening, and the degree of masculinization of the external genitalia (E) is estimated with a Prader number of 1–5. The confluence is not always readily apparent, especially in very masculinized patients who have a UGS resembling a male urethra. In these patients, one must actively look for a verumontanum-like structure with a small "utricular" opening in the proximal urethra. This is the confluence. A guidewire or ureteral catheter should be inserted into this communication through the cystoscope, and then the scope can be passed over the guide into the vagina. In almost all cases, we find it helpful to place a Fogarty catheter into the vagina and inflate the balloon (Fig. 68.1). Even in the smallest neonate, this can be done by passing the Fogarty catheter through the cystoscopic sheath and clamping the catheter with a hemostat after the balloon is inflated.

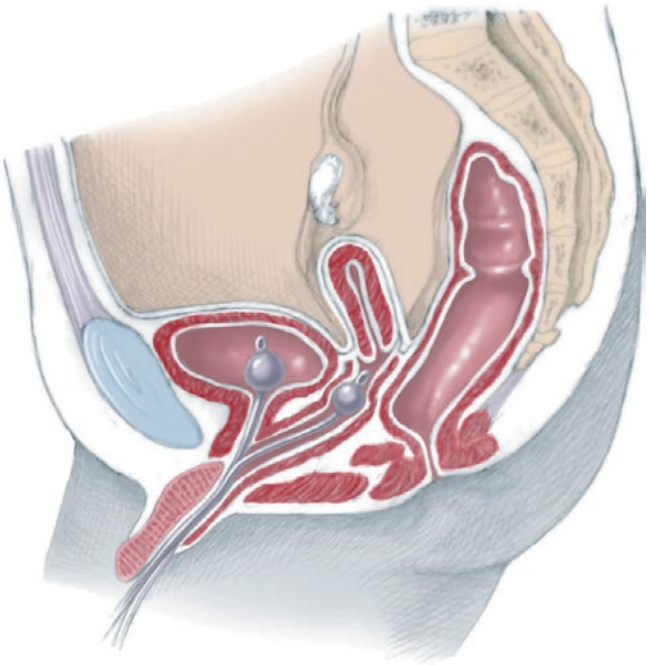

Fig. 68.1 There is a spectrum of vaginal confluence locations in urogenital sinus anomalies. This diagram shows a mid-level confluence

68.2 Timing and Perioperative Considerations

The issue of timing of the genital reconstruction has become a subject of a debate that is beyond the scope of this chapter. From the newborn period to the time of surgery, it is imperative that the family members (parents) have undergone counselling and education, so that the process of informed consent is assured and they indeed are making the final decision regarding any surgery offered versus other alternatives. We favor a single-stage surgical procedure during the first year of life, assuming that the patient has had appropriate endocrine control and is fit to undergo major surgery under general anesthesia. In 1995, de Jong and Boemers advocated performing a one-stage operation in the neonatal period. It has been suggested that prenatal maternal estrogen stimulation persists into the first 3 or 4 weeks of life, facilitating an easier vaginal pull-through due to a larger vagina with more robust tissue. The 2006 consensus paper on DSD stated: "There is inadequate evidence currently in relation to establishment of functional anatomy, to abandon the practice of early separation of the vagina and the urethra." It also has been suggested that surgery is best avoided between age 1 and puberty.

68.3 Preparation and Positioning

Most of these children can have their surgery performed as a same-day admission and are discharged when medically stable, often within 24–48 h. A bowel prep is up to the discretion of the surgeon; we do not employ it routinely. Perioperative antibiotics are administered.

The patient is placed in either a lithotomy or supine position, depending on the surgeon's preference. She is prepped from the level of the nipples to the toes, including the buttocks and back, so that she can be turned to the prone position if necessary for the posterior or anterior sagittal approach [4]. The legs are wrapped in a towel, which is then covered with stockinette. The lower body of the child is then placed through the aperture in the drapes. This position allows complete access to the perineum and abdomen. It also allows the child to be easily turned to the prone position for a posterior sagittal approach for the higher confluence. The supine position also allows the assistant to have excellent vision for teaching purposes. Allen stirrups or candy cane supports can used for the lithotomy position. The legs are well padded and positioned carefully to be certain there are no points of compression. Hip and knee positioning is checked and a small pad is placed under the sacrum. If the procedure is lengthy, the pressure points should be checked periodically and the legs repositioned. This initial positioning allows excellent vision for the surgeon for a perineal approach.

68.4 Proposed Incisions

Regardless of the procedure, the same basic incisions are used. A subcoronal circumcising line is drawn, which may be carried down to the UGS meatus in the midline, or vertical lines may be drawn to leave the urethral plate. An omega-shaped line is drawn from the inferior aspect of the UGS opening, extending inferiorly on each side. When the labia are fused into a scrotum, the omega flap will be positioned more inferiorly as shown in the diagram (Fig. 68.2). U-shaped lines are drawn around the inferior portion of each labia majora. The surgical area is infiltrated with 0.5% lidocaine with 1:200,000 epinephrine for hemostatic purposes.

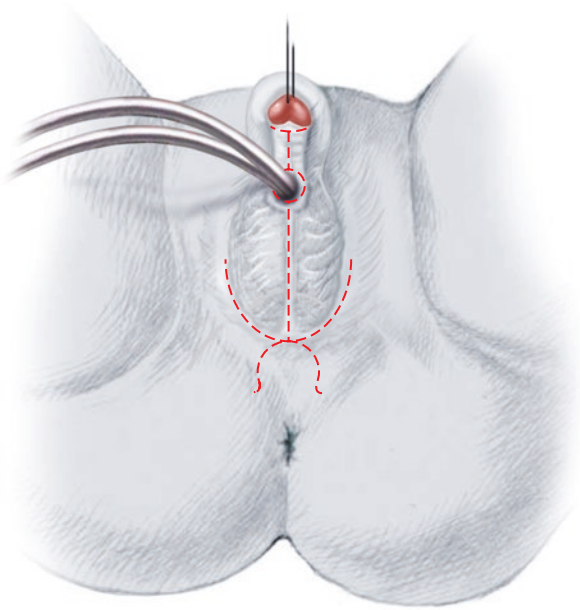

Fig. 68.2 Basic incisions

68.5 Specific Procedures

68.5.1 Clitoroplasty

Virtually all current techniques are based on the subtunical reduction of erectile tissue described by Kogan *et al*. in 1983, leaving the untouched neurovascular structures sandwiched between Buck's fascia and the dorsal tunica albuginea. Any incision into the corpora should be only on the very ventral aspect. To maximize sensitivity, we are cautious about glans reduction.

When the clitoris is very large, rather than using a tourniquet, two overlapping absorbable sutures are placed through the base of the clitoris for hemostasis as shown (Fig. 68.3). Care is taken to avoid the neurovascular structures. These sutures tie off about one half to three fourths of the erectile tissue. Longitudinal ventral incisions are made in each corporal body distal to these hemostatic sutures (Fig. 68.4).

The erectile tissue is teased off the inside of the tunica albuginea on each side both laterally and dorsally. The thin septum is punctured proximally and a suture ligature is passed through and tied ventrally. This tie is slightly distal to the initial hemostatic sutures and captures almost the complete circumference of the erectile tissue and the ventral tunica albuginea. The erectile tissue and ventral tunica albuginea are divided just distal to this tie, and dissected distally along the septum until all the erectile tissue has been removed, leaving the glans attached to the thick dorsal pedicle (Fig. 68.5).

As mentioned above, we prefer to maintain the integrity of the glans and leave it untouched, but when the glans is very large, it can be reduced ventrally (Fig. 68.6). The glans or the tunic just proximal to the glans is then sutured to the stumps of the corpora cavernosa. If previously detached, the ventral mucosal strip is reattached to the glans inferiorly.

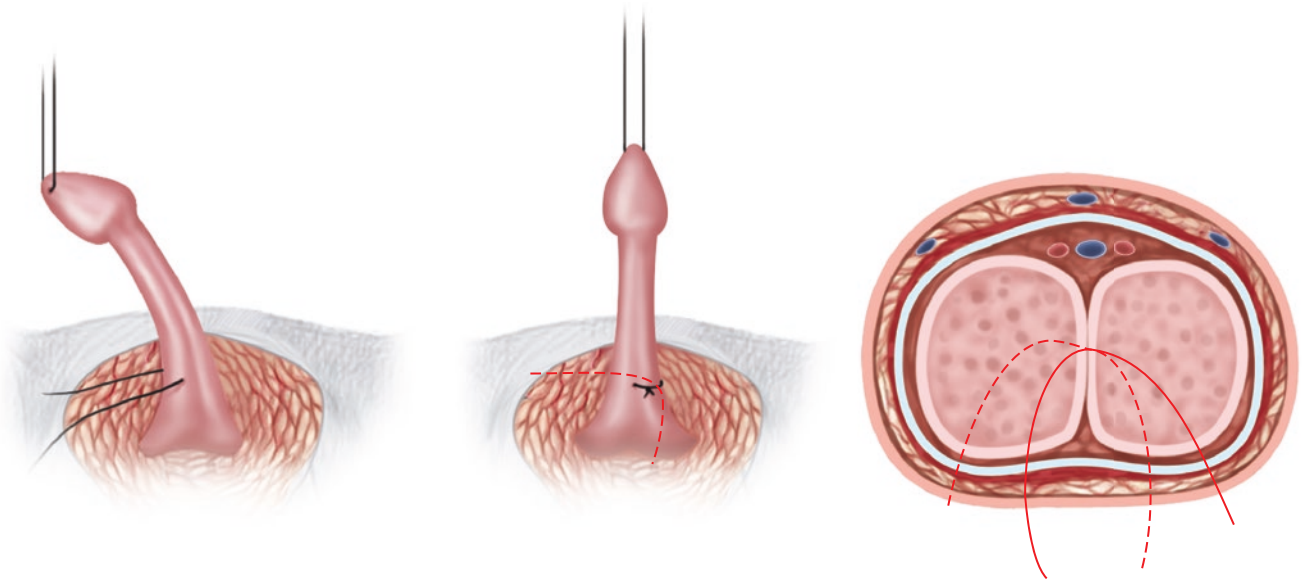

Fig. 68.3 Clitoroplasty: sutures placed for hemostasis

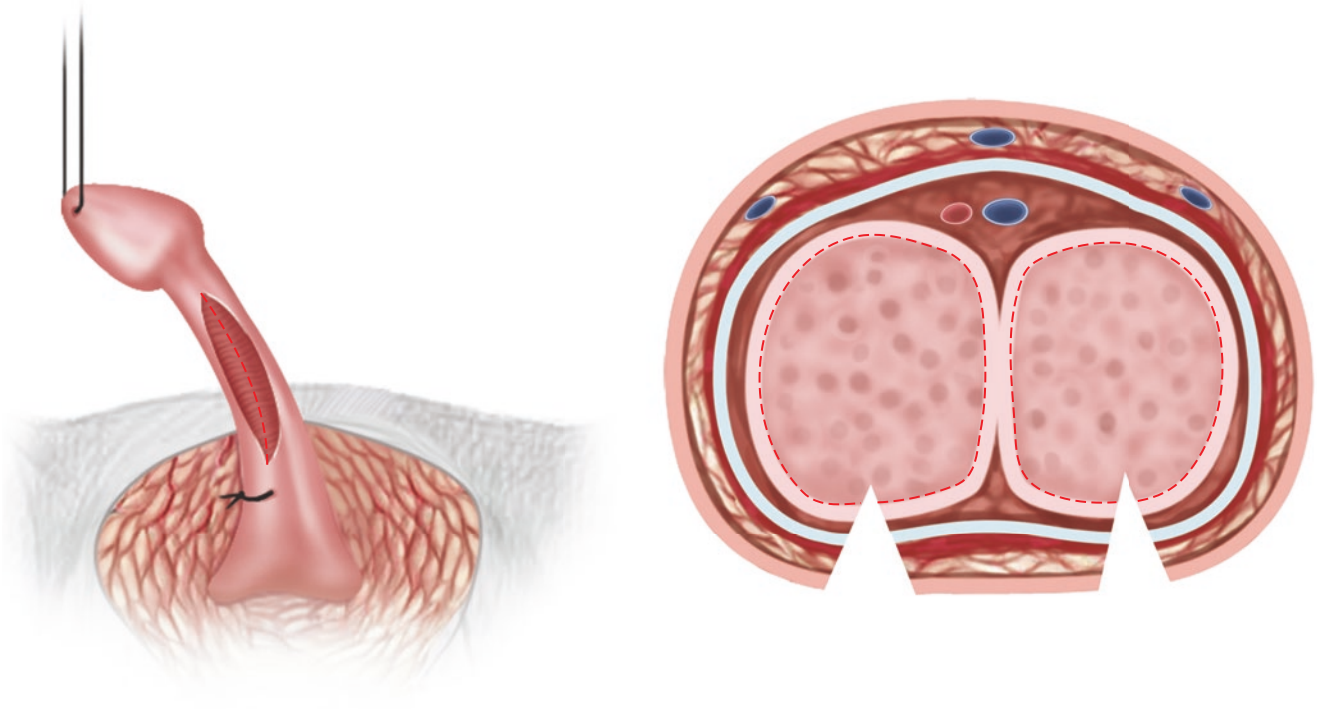

Fig. 68.4 Ventral incisions; erectile tissue teased off the inside of the tunica albuginea

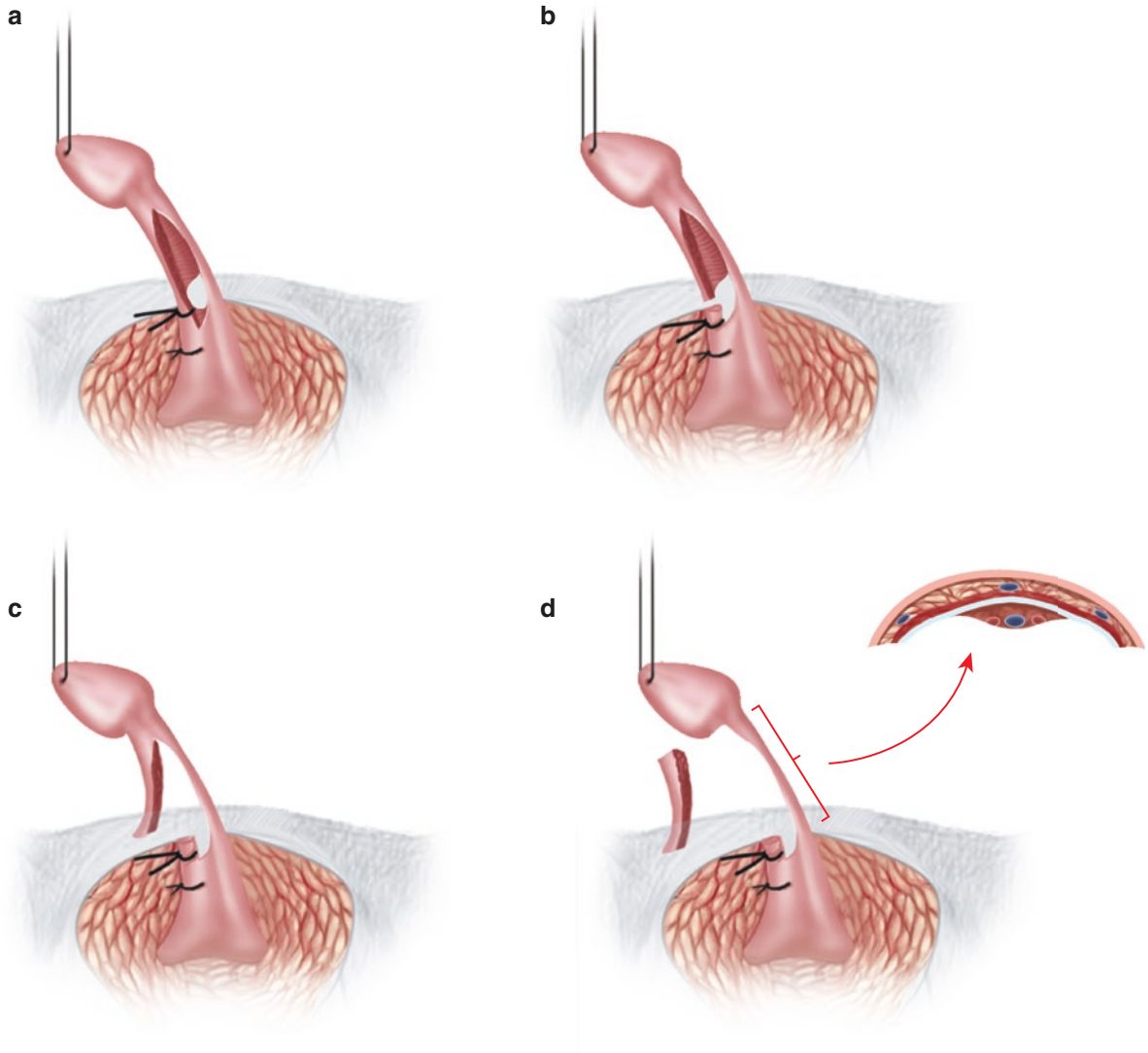

Fig. 68.5 (**a**), Ligature tied ventrally. (**b**–**d**), Removal of the erectile tissue leaving the glans attached to the thick dorsal pedicle

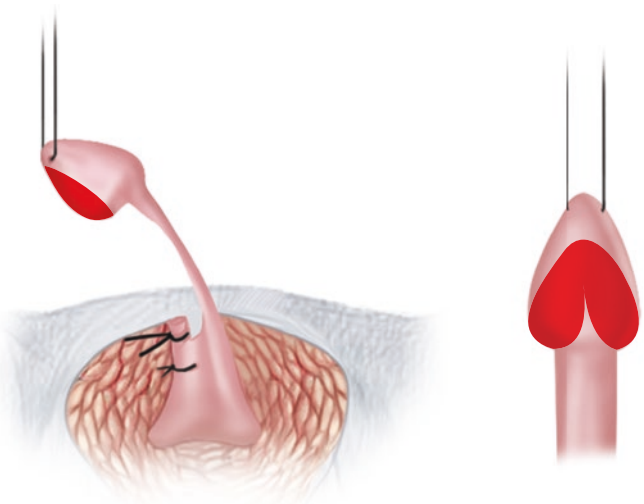

Fig. 68.6 Ventral glans reduction for the very large clitoris

68.5.2 Vaginoplasty

Labial fusion can be easily treated by a "cut-back" vaginoplasty. (This is the only time a cut back should be used.) For low to mid confluence with the UGS, a "flap" vaginoplasty should be used. A "pull-through" vaginoplasty is necessary for higher confluences.

To perform a *posterior flap vaginoplasty*, an omega flap incision, a modification of the "U-shaped" incision described by Fortunoff *et al*. in 1964, is made through the skin and partway through fat. The midline spongiosal tissue is avoided. The UGS is incised vertically in the midline for 5–10 mm at a time while placing full-thickness figure-of-eight fine absorbable sutures laterally into each cut edge. These sutures provide symmetry, hemostasis, and mild traction. The midline incision is continued into the vagina for about 2 cm, until the normal-caliber vagina is reached. At this point, the posterior flap can be anastomosed to the posterior vaginal incision.

68.5.3 Modified Urogenital Mobilization for Mid to High Confluence

After circumscribing the common meatus, the UGS is dissected from the corporal bodies. A plane of dissection is created between the UGS and the rectum. While retracting the UGS dorsally, dissection is carried laterally between the UGS and the crurae of the corpora. Finally, the dissection proceeds anteriorly to the level of the pubis. Although Peña has favored total urogenital mobilization where the puboure-thral ligament is transected, in most instances this dissection is not necessary. As such, the partial urogenital mobilization described by Rink *et al*. is applicable to the vast majority of patients (Fig. 68.7).

The posterior wall of the vagina is then exposed. The continuous palpation of the balloon in the vagina enables accurate and safe dissection. The vagina is opened between stay sutures (Fig. 68.8). A decision is made at this point. If the vagina can be brought to the perineum without tension, a posterior flap vaginoplasty is performed. The posterior wall of the vagina is then incised deeply through the narrow dysplastic distal vagina into the normal elastic vaginal tissue, usually for about 2 cm in babies. The omega flap is then inserted (Fig. 68.9). If the vagina will not reach the perineum, it must be detached from the urogenital sinus and exteriorized by laying in skin and urogenital sinus tissue flaps. Before the vagina is exteriorized, the communication with the UGS and urethra can be identified and closed from the vaginal side in two layers (Fig. 68.10). The distal UGS then remains as a tube and is brought below the clitoris as the urethra.

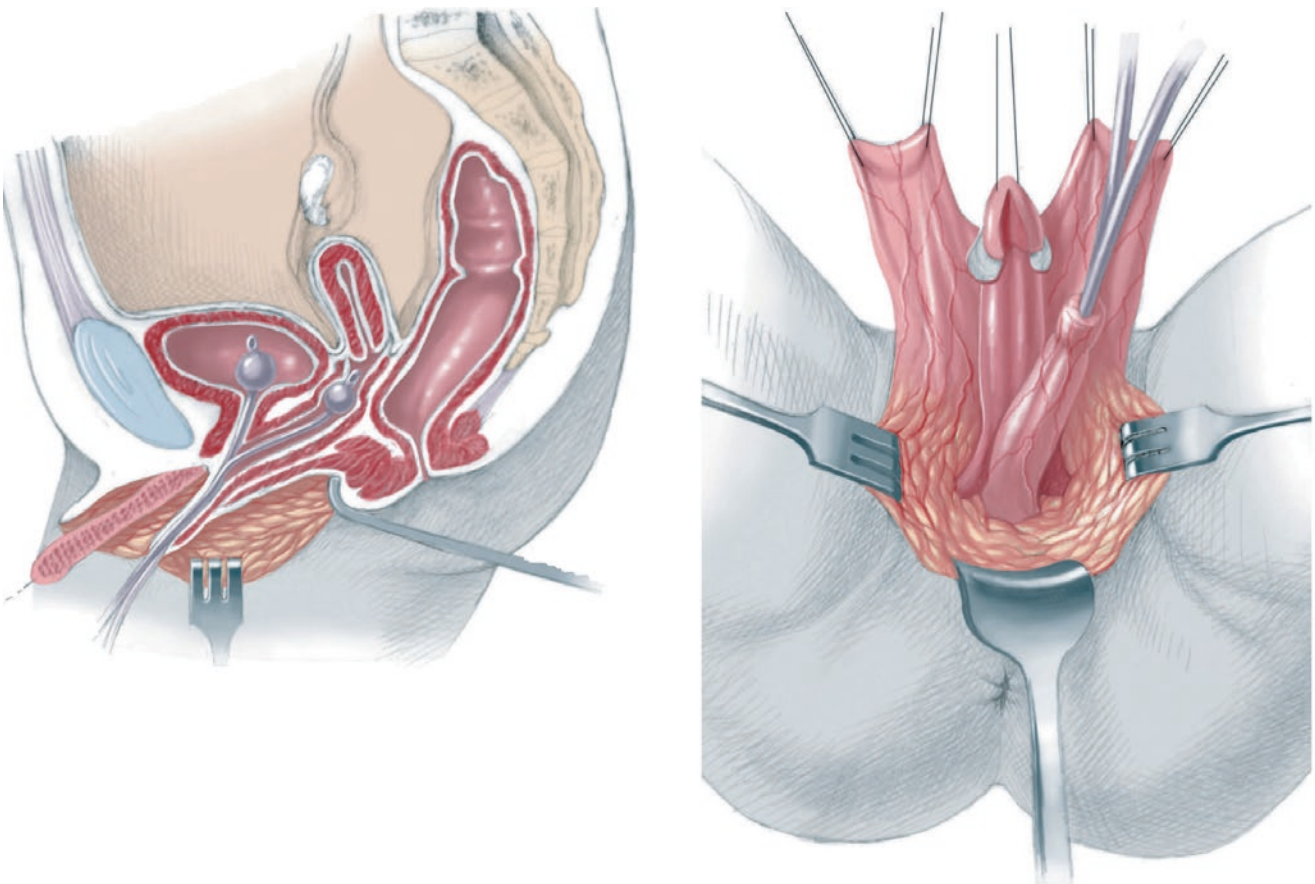

Fig. 68.7 Modified urogenital mobilization

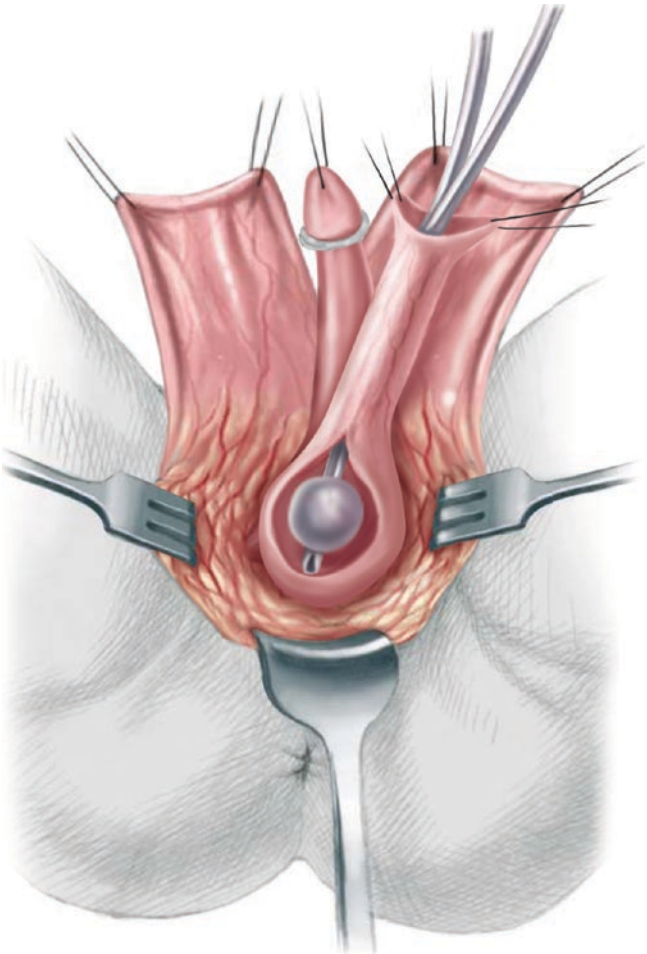

Fig. 68.8 Vagina opened over Fogarty catheter balloon

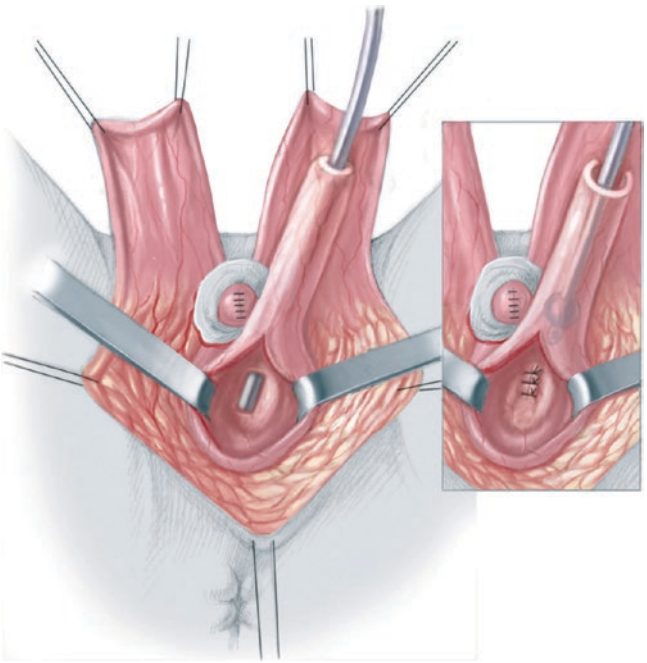

Fig. 68.10 Closure of the communication from the vagina to the urogenital sinus and urethra

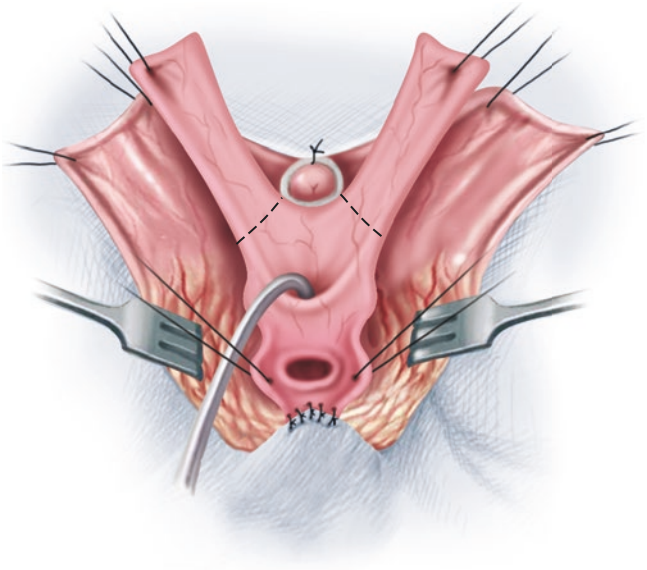

Fig. 68.9 Posterior flap vaginoplasty

68.5.4 Modified Passerini Technique for Mid to High Confluence

The Passerini procedure is ideal for the patient with a high vagina and a long UGS. The advantages of the Passerini procedure include a natural vulvar appearance with a mucosal lining of the introitus and less risk of urethral stenosis. The procedure can be performed in one stage at any age.

The key to the Passerini procedure is a meticulous perineal dissection through the urogenital diaphragm to create a working space and expose the vagina. The vagina is detached from its entry point into the UGS and then the anterior wall of the vagina is carefully dissected off of the overlying urethra and bladder neck. The tissue is very thin anteriorly and there is no good anatomic plane between the vagina and proximal urethra. The connecting "fistula" to the UGS is closed. Next, the UGS is dissected off of the corpora cavernosa down to where the urethral meatus is to be located. At this point, the reduction clitoroplasty is performed using the subtunical approach described above, modified from Kogan *et al*.

The UGS is then opened vertically along its dorsal surface. At the very proximal part of this incision, a small V-shaped flap is formed. This flap is sewn to the clitoris so that the ventral surface of the clitoris is covered with UGS mucosa. The phallic skin is unfurled so the maximum length can be obtained. This is split down the middle (Fig. 68.11).

Each of these flaps is sutured around the clitoris and joined to the upper part of the opened UGS tissue on each side, either before or after the UGS tissue is connected to the anterior vaginal wall (Fig. 68.12). Passerini [9] has pointed out that often the opened UGS is too long. If left redundant, it will cause a bulging of the vulvar wall between the urethral and vaginal openings. In this case, he recommends incising the distal end of the opened UGS vertically in the midline for about 1 cm. This also further enlarges the vaginal anastomosis. The apex of this incision is then sewn to the 12 o'clock position of the anterior vaginal wall.

After the UGS flap is sewn into place anteriorly, the distal ends of the phallic skin flaps are sutured to the lateral vaginal wall and to the UGS tissue medially (Fig. 68.13). Finally, the posterior flap is sutured into the vertically incised posterior vaginal wall (Fig. 68.14).

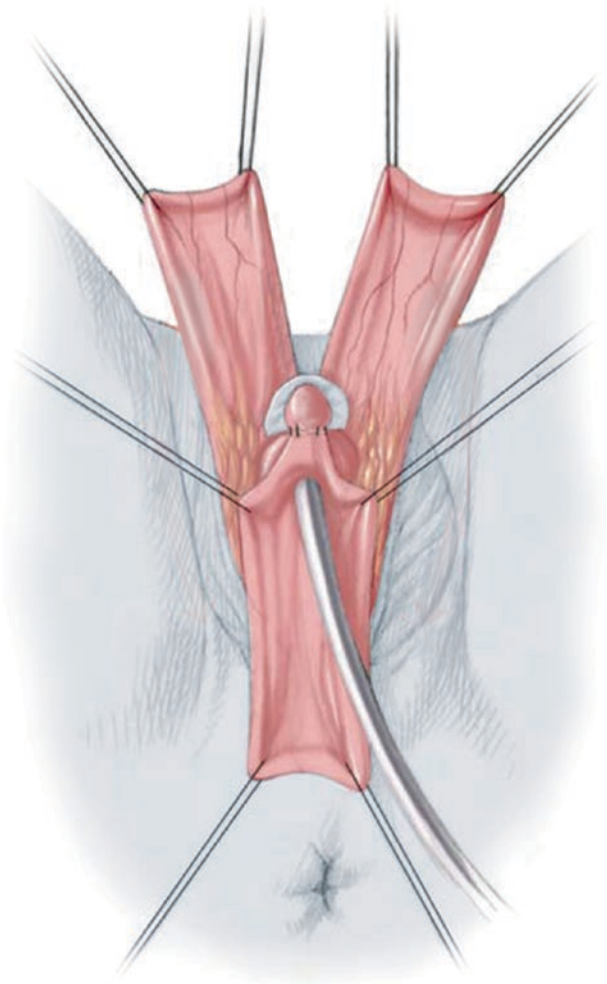

Fig. 68.11 Opened urogenital sinus shown with phallic skin flaps

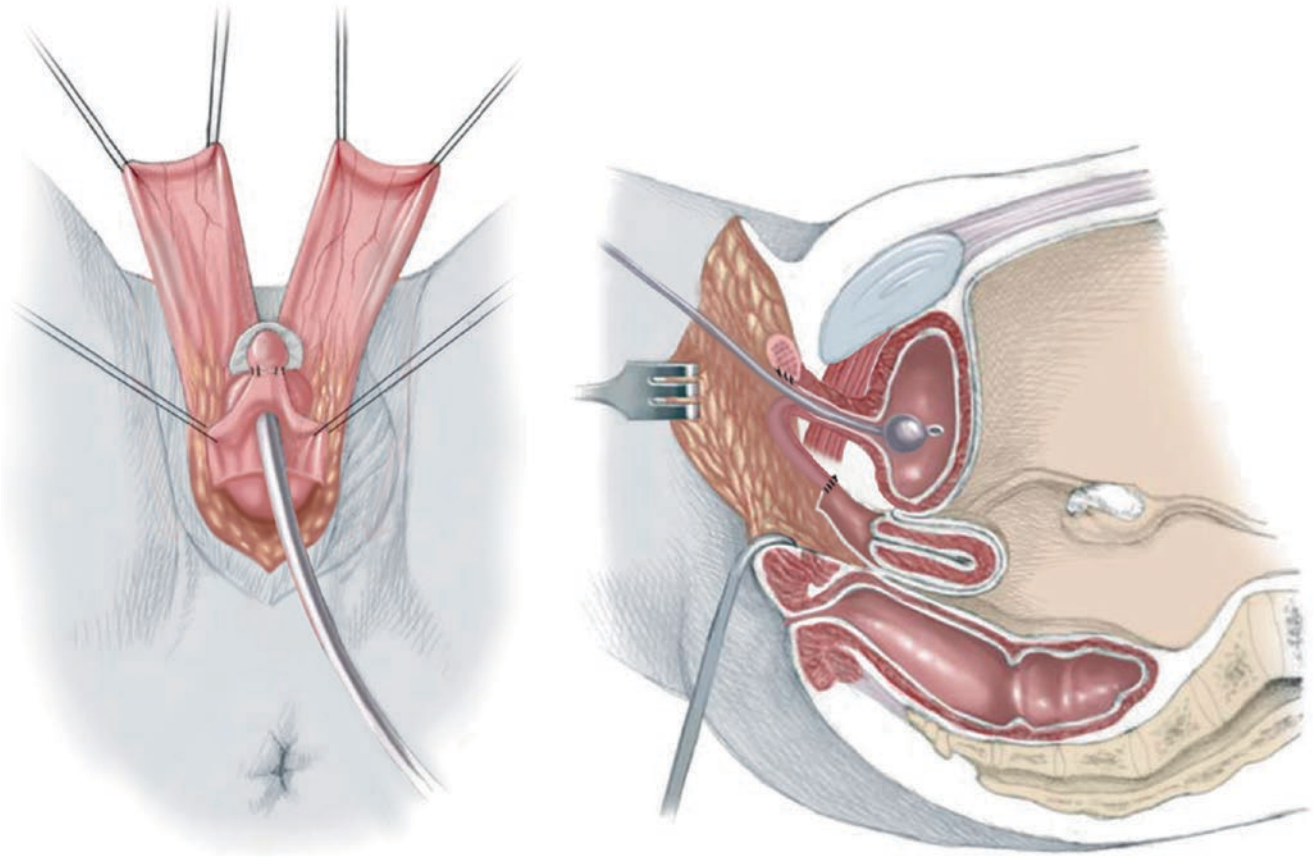

Fig. 68.12 Opened urogenital sinus tissue turned in and connected to the anterior vaginal wall

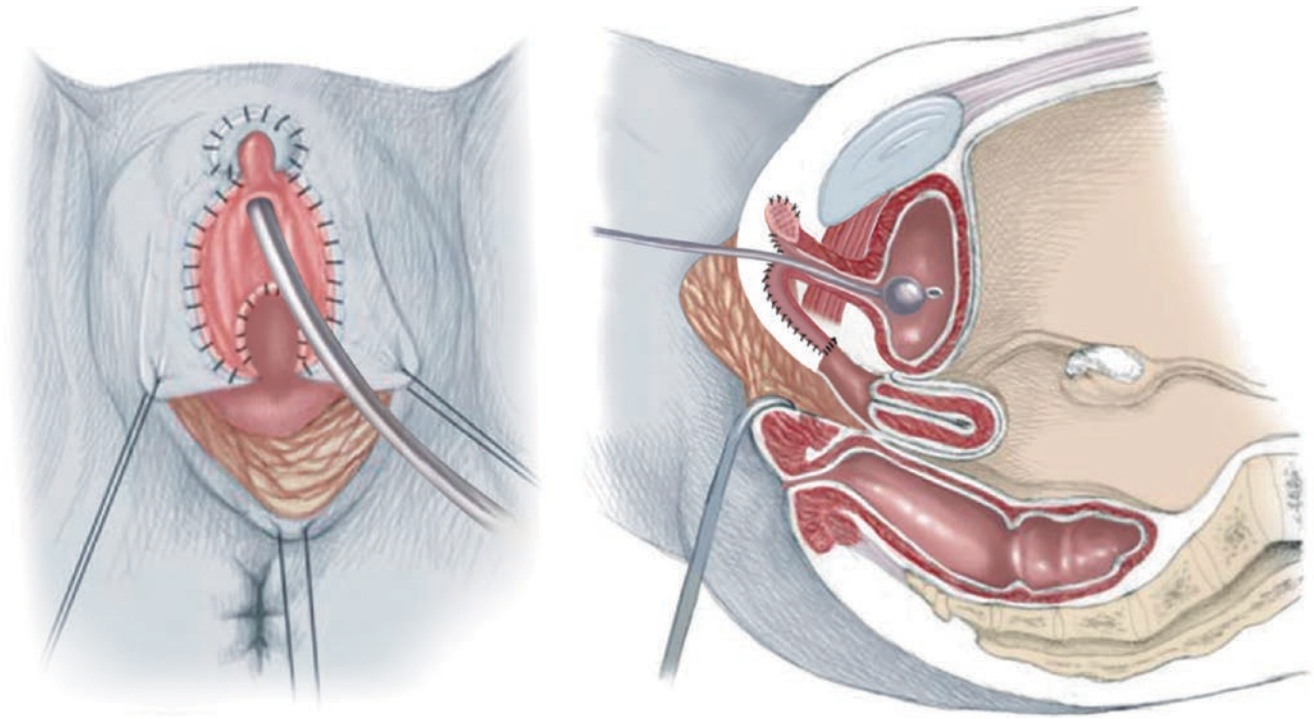

Fig. 68.13 Phallic skin flaps sutured to the lateral vaginal wall and urogenital sinus tissue

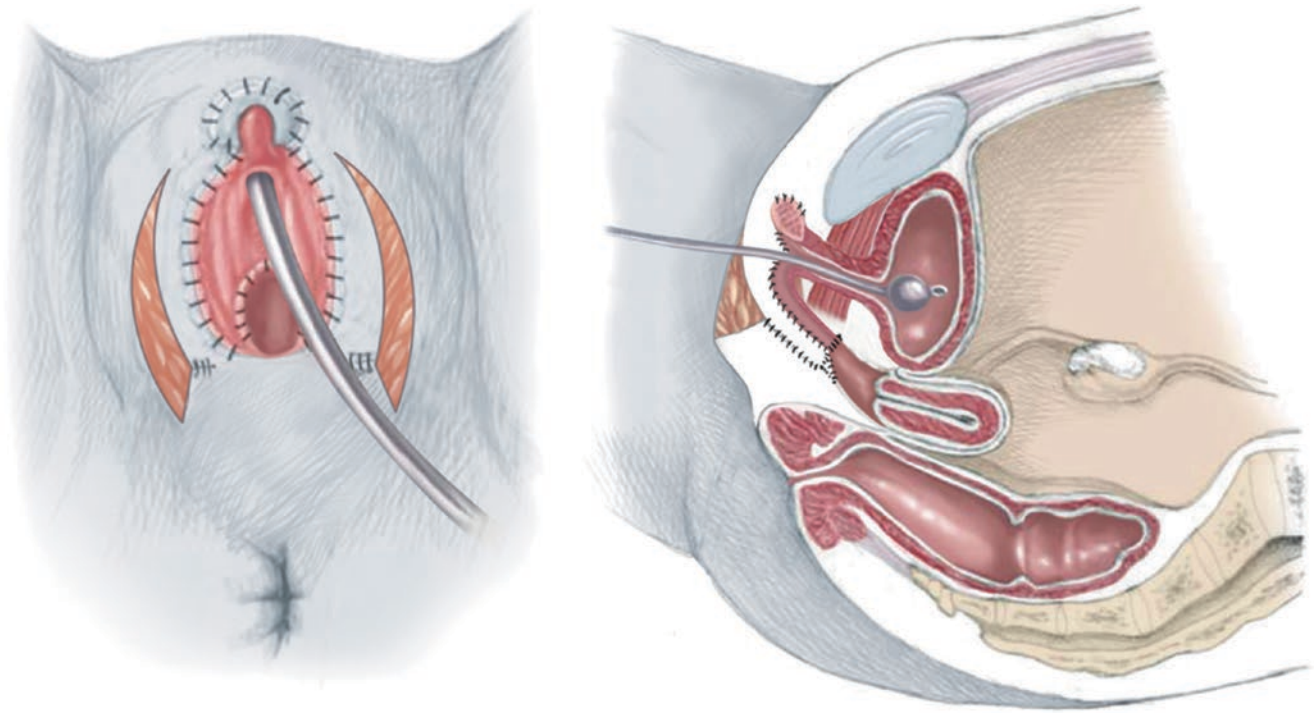

Fig. 68.14 Posterior flap sutured to the posterior vaginal wall

68.5.5 Labioplasty

The preputial skin is divided in the midline creating phallic skin flaps. For a posterior flap vaginoplasty, the flaps are sutured around the clitoris and to the opened urogenital sinus tissue medially and to the labioscrotal skin laterally. For the Passerini procedure, the phallic skin flaps tend to be longer and can therefore also be used to help exteriorize the detached vagina. The phallic flaps recreate the labia minor. The lateral labioscrotal skin can be brought down with a Y-V plasty and sutured into the corners between the posterior flap and the lateral skin to form the labia majora (Fig. 68.15a, b). This cosmetically brings the vagina into the introitus and the labia beside the vagina in the normal location.

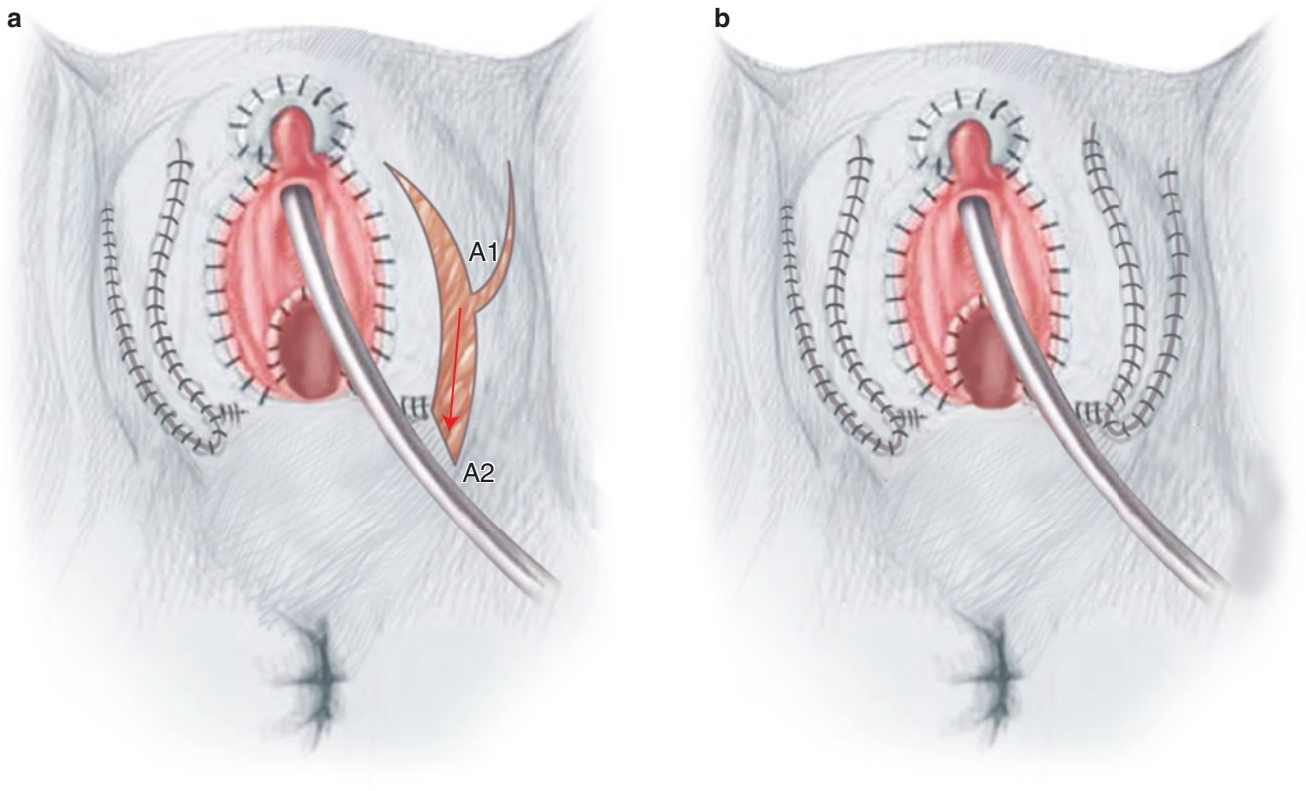

Fig. 68.15 Forming labia majora from the mobilized lateral labioscrotal skin

68.6 Postoperative Care

Vaginal drains and packs are at the discretion of the surgeon. A mild compression dressing is left in place for 24–48 h. A Foley catheter is left in place for 2 days following a posterior flap vaginoplasty and for 5–7 days after a Passerini-type of procedure involving vaginal detachment and fistula closure. Caudal anesthesia aids immediate postoperative pain control.

Conclusions

It should be emphasized that the techniques illustrated in this chapter are primarily applicable to patients with CAH especially with mid to high confluences. Many approaches that are beyond the scope of this chapter are described in the literature and have been used by other surgeons. There have been tremendous advances in the reconstructive techniques for patients with UGS and ambiguous genitalia. As a result, cosmetic appearances have been improved, but long-term functional results are still to be determined. Because this is a spectrum disorder about which there is much controversy, it is necessary that the parents have complete engagement and total understanding before repair is begun. An involved, dedicated team approach is preferred in dealing with these complex disorders.

Suggested Reading

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Bladder Exstrophy and Epispadias

69

Ezekiel E. Young and John P. Gearhart

Bladder exstrophy and epispadias are rare and complex urogenital malformations predominantly occurring in males. The defect can be accurately diagnosed with modern prenatal ultrasound, but the definitive diagnosis is made after examining the newborn at birth. Classic bladder exstrophy is characterized by an open abdominal wall, bladder, and urethra and a wide diastasis of the symphysis pubis, caused by a 30% bony deficit of the anterior pubic rami in combination with a 12° external rotation of the posterior aspect of the pelvis and an 18° external rotation of the anterior aspect. Girls present with a bifid clitoris and a short vagina; boys have a 50% shortening of the anterior corpora cavernosa and an upward deviation of the penis. Ureteral reflux in various degrees is seen in 100% of cases after closure. A preoperative ultrasound evaluation of the otherwise usually unaffected upper tracts is mandatory to determine the presence of two normal kidneys. The magnitude of the defect and the complexity of the treatment require transfer of the affected child to a specialized centre. Only the multidisciplinary care of surgeons, anaesthesiologists, psychologists, and nursing staff can guarantee the most favourable outcome for these otherwise healthy children.

69.1 Principles of Surgical Treatment

Before Hugh Hampton Young performed the first recorded successful primary closure of a female exstrophy patient in 1942, bladder exstrophy was primarily treated by covering the defect with skin flaps. Modern exstrophy closure, based on the pioneering work of Jeffs et al. and Cendron, has been significantly modified in the past decade and is performed in many major centres worldwide. Depending on the surgeon's experience, some centres perform the type of repairs described by Grady and Mitchell or Kelly. Regardless of the method of repair, the primary principles in surgical management are a secure initial abdominal closure, the reconstruction of a functional and cosmetically satisfactory external genitalia, and the achievement of urinary continence with preservation of renal function. Though other forms of repair have been promoted, this approach provides the longest follow-up data and most favourable outcome in treating children with this complex malformation. The technique includes early closure of the bladder, posterior urethra, and abdominal wall, usually with pelvic osteotomy in the newborn period, subsequently followed by an early epispadias repair at 6 months to 1 year of age after testosterone stimulation by intramuscular injection. Around age 4–5 years, when adequate bladder capacity is reached and the child is ready to participate in a very structured preoperative and postoperative voiding program, a competent bladder neck is reconstructed, with bilateral ureteral reimplantation.

Achieving urinary continence with a sufficient bladder capacity is strongly dependent on initial successful closure of the bladder and the posterior urethra as well as the size of the bladder template. Therefore, the first step of the reconstruction is conversion of bladder exstrophy into a penile epispadias with incontinence with a balanced posterior outlet resistance that preserves renal function but stimulates bladder growth. In very select cases, newborn exstrophy closure can be combined with epispadias repair (see below). This approach can also be performed in delayed primary closure and reoperative exstrophy repairs, but success requires a good urethral plate and a reasonable bladder template. Additionally, pelvic osteotomies are performed if the patient is older than 72 h, for a symphyseal diastasis of more than 4 cm or if a tension-free closure cannot be achieved. In those

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https://doi.org/10.1007/978-3-662-56282-6_69

cases, osteotomies are crucial to ensure tension-free approximation of the bladder, posterior urethra, and abdominal wall, preventing dehiscence or bladder prolapse. Furthermore, it places the urethra deep within the pelvic ring, enhances bladder outlet resistance, and ensures alignment of the large pelvic floor muscles to support the bladder neck.

Combined exstrophy and epispadias repair: In a few selected cases, newborn exstrophy closure can be combined with epispadias repair. This approach requires good phallic length, a deep urethral groove, and an adequate amount of penile skin, however, and it should be attempted only by experienced exstrophy surgeons, as the complications can be severe. Combined exstrophy and epispadias repair is best applied in the patient undergoing delayed primary or reoperative exstrophy closure. The preoperative use of intramuscular testosterone in reoperative exstrophy patients will allow for improved vascularity and more penile skin for the reconstruction.

The operations are performed with general anaesthesia, with the patient in a supine position even for the osteotomies. A tunneled epidural catheter is placed when possible to reduce the intraoperative amount of anaesthetic agents and for postoperative pain control. It is typically left in place for several weeks after the closure. Care must be taken to create a latex-free environment in the operation room, as many children with bladder exstrophy are prone to latex allergies. Perioperative broad-spectrum antibiotics are administered and continued throughout the first postoperative week.

69.2 Surgical Procedures

69.2.1 Combined Bilateral Transverse Innominate and Vertical Iliac Osteotomies

With the patient in the supine position, the pelvis is exposed from the iliac wings inferiorly to the pectineal tubercle and posteriorly to the sacroiliac joint. The periosteum and sciatic notch are carefully elevated, and a Gigli saw is used to create a transverse innominate osteotomy, exiting anteriorly at a point halfway between the anterosuperior and the anteroinferior spines (Fig. 69.1). Unlike the Salter osteotomy, this osteotomy is more cranial, to facilitate placement of external fixator pins in the distal segment. To correct the posterior malrotation, the posterior ilium is incised by creating a closing wedge osteotomy vertically and just lateral to the sacroiliac joint. Note the intact proximal posterior iliac cortex, serving as a hinge when the pubic bones are brought together in the midline.

Two fixator pins are placed in the inferior segment and the wing of the superior ileum on each side (Fig. 69.2). In newborns, often only one pin can be placed in the inferior fragment because of the lack of cancellous bone. After radiographs confirm the correct placement of the pins, the urological part of the procedure is performed. Afterwards, placing a suture between the two pubic rami concludes the pelvic closure. External fixators are applied and the child is placed in light horizontal Buck traction for 4 weeks to stabilize the pelvis and avoid ureteral and suprapubic tube displacement. Once good callus formation is confirmed on pelvic radiographs (around week 6), the pins are removed at the bedside under light sedation.

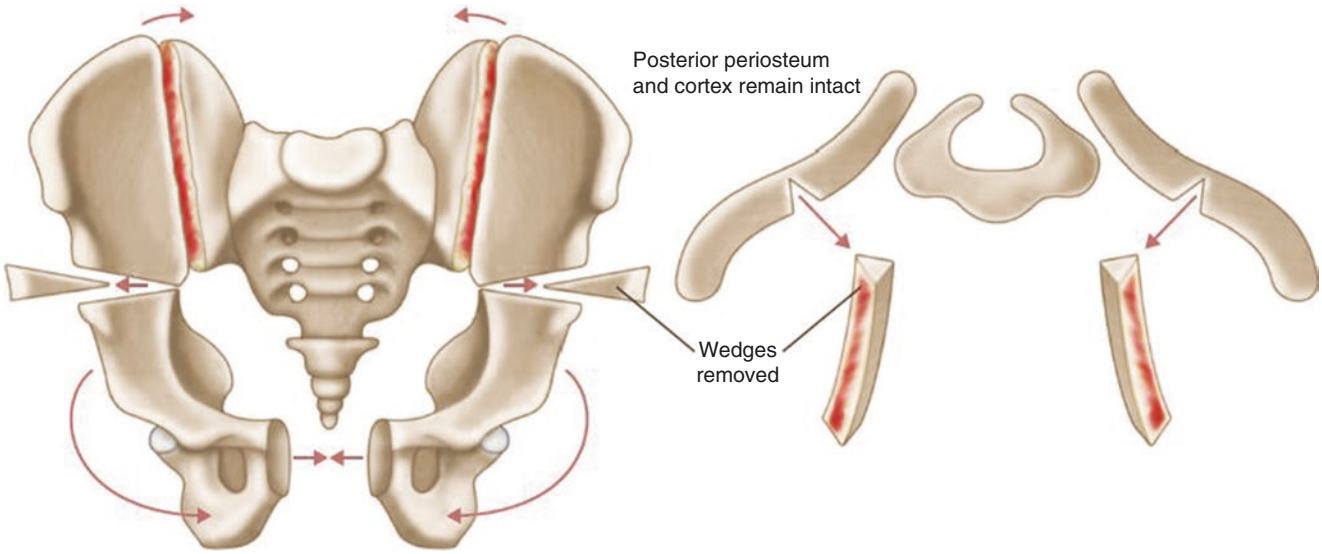

Fig. 69.1 Osteotomies

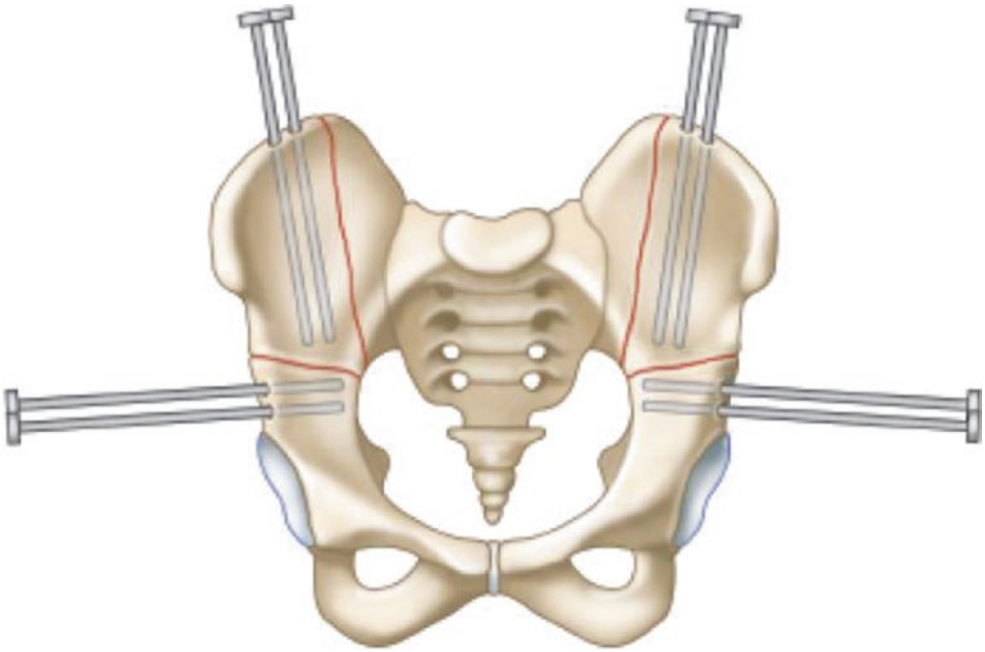

Fig. 69.2 Placement of fixator pins

69.2.2 Bladder, Posterior Urethral, and Abdominal Closure

For bladder, posterior urethral, and abdominal closure, anatomical outlay of the malformation before outlining the incision is important. The size of the bladder template is best evaluated by inverting the plate with sterile gloved fingers. With this manoeuvre, a previously unrecognized part of the bladder can be found behind the fascia in some cases with small bladder templates. To achieve proper retraction, a nylon suture is placed through the ventral glans (Fig. 69.3).

The complete incision is outlined with a blue marking pen. The skin is incised using a no.15 blade above the umbilicus, around the junction of the bladder and the paraexstrophy skin, to the level of the urethral plate. The remainder of the incision is performed with electrocautery. For the prostatic and posterior urethral reconstruction, a 2-cm wide mucosal strip from the distal trigone to below the verumontanum out onto the midshaft of the penis is outlined and incised. The foreskin is preserved for later distal epispadias repair. In females, the incision is carried out down to the vaginal orifice and the clitoral halves are denuded for the complete reconstruction of the outer genitalia along with the bladder and posterior urethra. The skin incision leaves an inner mucosal line for closure of the bladder and an outer skin rim for the closure of the skin.

The plane between the rectus fascia and the bladder is found and entered just above the umbilicus. The dome of the bladder is separated from the peritoneum and the retropubic space behind the bladder becomes developed (Fig. 69.4). The attachments connecting the rectus sheath and muscle to the bladder are released sharply and the umbilical vessels are freed, transected and doubly ligated. Taking down the caudal rectus attachments and peritoneum from the dome leaves the cephalad part of the bladder completely mobilized, therefore allowing the bladder to be placed deeply into the pelvis, where it will begin to fold on itself.

At this point the urogenital diaphragm can be recognized connecting the pubic bone with the posterior urethra and bladder neck. Placing a skin hook in the pubic tubercles allows for lateral retraction, revealing the urogenital diaphragm completely. It is crucial to radically incise the diaphragm using electrocautery completely all the way down to the levator hiatus to allow deep placement of the vesicourethral unit into the pelvis prior to pubic apposition. Care is taken to release the fibrous band sharply at the subperiosteal level bilaterally (Fig. 69.5). This manoeuvre cannot be emphasized enough; incomplete dissection of the diaphragm fibres will create anterior tension and often causes failed closure.

By applying gentle traction on the glans caudally, the insertions of the corporal bodies on the lateral inferior aspect of the pubic bone can be visualized. Releasing the attachments of the suspensory ligaments to the corpora bilaterally at this level results in some penile lengthening by bringing the congenital shorter corpora further out of the pelvis.

The ureters are stented because swelling and the increased intravesical pressure can cause temporary obstruction. A Malecot catheter drains the bladder, and all tubes are brought through the bladder wall and attached to the inside and outside of the bladder with 4/0 absorbable sutures. The mucosa and muscle of the bladder and bladder neck area are closed with a running 3/0 absorbable suture in the midline anteriorly. The urethral plate is closed with an interrupted 5/0 absorbable suture from just distal to the bladder neck down to the midshaft penis (Fig. 69.6). The closure covers the ejaculatory duct and the proximal two thirds of the posterior urethra. After urethral closure, a 12F sound should be easily passable through the orifice into the bladder. The correct choice of bladder outlet resistance is of critical importance. Creating bladder outlet obstruction would ultimately lead to increased intravesical pressure and upper tract changes, but the outlet resistance must be high enough to promote bladder adaptation and growth and prevent bladder prolapse. A second layer of interrupted sutures is placed and the posterior urethra and bladder neck are buttressed to the second layer of local tissue if possible. A piece of decellularized cadaveric human dermis (AlloDerm) is placed over the posterior urethra bladder neck area as a buttress under the intrapubic stitch. The urethra is sounded again to ensure that the second layer did not add additional obstruction.

Following closure of the bladder, the suprapubic tube and ureteral stents are exteriorized through the neoumbilicus (Fig. 69.7), which is created by a V-shaped flap of abdominal skin, tacked down to the abdominal fascia in the correct anatomical position. The ureteral stents are left in place for 10–14 days, and the suprapubic tube is removed 4 weeks postoperatively, after calibrating the bladder outlet to warrant free drainage. Note that the urethra is not stented at the end of the operation to avoid pressure necrosis, infection, and secretion accumulation. The pelvis is approximated in the midline by gently applying pressure over the greater trochanters bilaterally. Horizontal mattress sutures of #2 nylon are placed in the pubis (Fig. 69.7, inset).

It is important to tie the knot away from the neourethra to avoid material migration into the posterior urethra. A second stitch of #2 nylon is used at the most caudal insertion of the rectus fascia onto the pubic bone for added security, if it can be easily done and does not compromise the first stitch.

69.2.2.1 Epispadias Repair

The modified Cantwell-Ransley epispadias repair is begun by placing a nylon suture through the ventral glans for traction. A circumcising incision is made and the ventral penile skin is taken down to the level of the scrotum to deglove the penis (Fig. 69.8). Holding sutures are placed into the ventral prepuce. The ventral mesentery between the corpora is left intact for the blood supply to the urethral plate. The base of

this mesentery is located where the corporal bodies diverge on the ventral aspect of the corpora.

A deep vertical incision in the distal urethral plate is performed and closed transversely with 6/0 polyglycolic sutures to flatten the distal urethra and advance it to the tip of the penis for later closure (Fig. 69.9). On the dorsum of the penis, an 18-mm-wide urethral mucosal strip from the midshaft urethral meatus to the tip of the glans is outlined with a blue marking pen and incised with a no. 15 scalpel. Lateral skin flaps are mobilized and undermined.

The neurovascular bundles situated between Buck's fascia and the corporal bodies can be visualized at the lateral aspects of the corpora (Fig. 69.10). Thick glandular wings are developed sharply off the corpora, and triangular mucosal areas are excised to bring denuded glans together at time of closure. At the dorsal base of the phallus, a Z-incision is performed in the suprapubic area to release tension from old scar tissue of the initial closure. Any remaining suspensory ligament tissue is exposed and divided to gain penile length.

The dissection of the urethral plate is continued from the ventral side. By strictly dissecting on the surface of Buck's fascia, the plane is followed in a circumferential fashion between the corpora spongiosum and cavernosa towards the dorsal side. The incisions from the dorsal and ventral side are joined, followed by the dissection of the contralateral side in the same way. Loops are passed around the corpora and the plane is extended proximally to the level of the prostate and distally to the junction of the glans with the corporal bodies. Care is taken never to leave the surface of Buck's fascia to avoid injury of the corpora and neurovascular bundles. The urethral plate is now completely freed from the corporal bodies except for the distal 1-cm attachment of the mucosal plate to the glans penis. This degree of mobilisation is necessary to rotate the corporal bodies over the urethra at the level of the corona. The urethral plate is now tubularized, beginning at the level of the prostate, over an 8F soft silicone stent, using a running 6/0 polyglycolic suture (Fig. 69.11).

If the rotating the corpora over the urethra alone does not satisfactorily straighten the penis, corporal incisions are performed at the point of maximum curvature (Fig. 69.12). This procedure is usually performed in older patients. To ascertain that the neurovascular bundles are properly protected, they are dissected free from the corporal bodies and secured with vessel loops. The corpora can now be easily rotated over the neourethra.

If corporal incision took place, the diamond-shaped defects are sutured to each other over the neourethra with two 5/0 polydioxanone running sutures (Fig. 69.13).

If no incision was necessary, the corpora are rolled over and closed over the neourethra with interrupted 4/0 polydioxanone sutures (Fig. 69.14). This manoeuvre deflects the penis downward and provides some increase in penile length. Additional sutures of 4/0 polyglycolic acid are placed between the corporal bodies to bury the urethra further, especially at the point where the urethra emerges between the corpora and at the level of the corona, to avoid fistula development.

The glans is now closed in two layers of interrupted sutures, using 5/0 polyglycolic acid sutures for the subcutaneous layer and 6/0 polyglycolic acid sutures for the epithelial layer (Fig. 69.15). The reconstructed phallus now has a ventrally displaced urethra, and the corporal bodies (with the neurovascular bundles running on the lateral sides) reach the reconstructed glans.

The mobilized ventral skin is brought up and sutured to the ventral edge of the corona. The dorsal side is covered with the skin flaps by bringing them together in the midline of the dorsal phallus with interrupted 5/0 or 6/0 polyglycolic acid sutures (Fig. 69.16). The urethral stent is secured at the tip of the glans with a nylon stitch and left in place for 10–12 days.

At the end of the operation, the penis is covered with a plastic occlusive dressing, which will stay on until it falls off by itself. Postoperative pain and bladder spasm control must be provided to keep the child comfortable and prevent urinary extravasation and fistula formation.

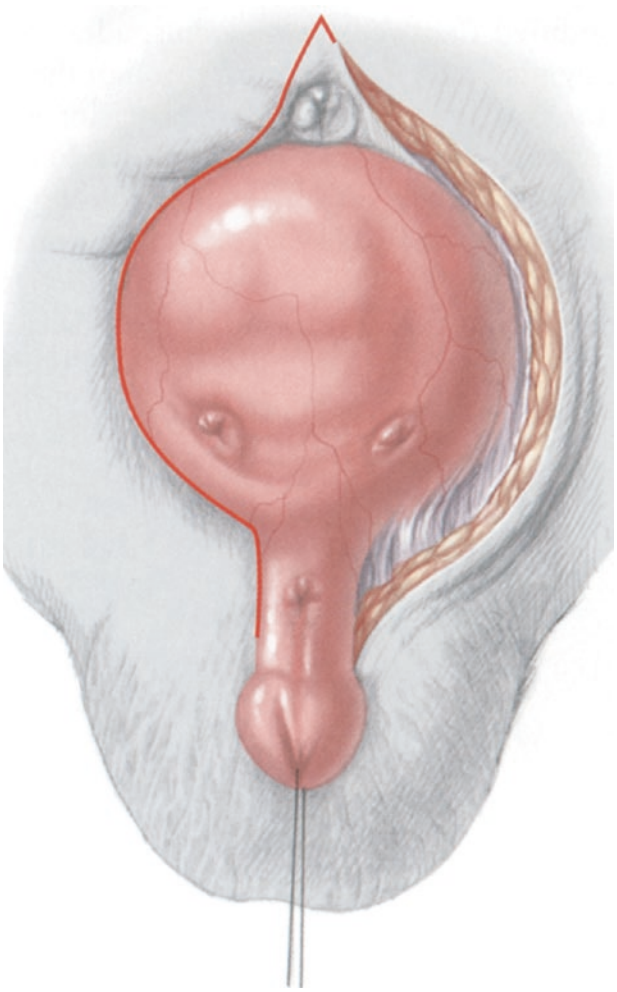

Fig. 69.3 Beginning the incision for closure

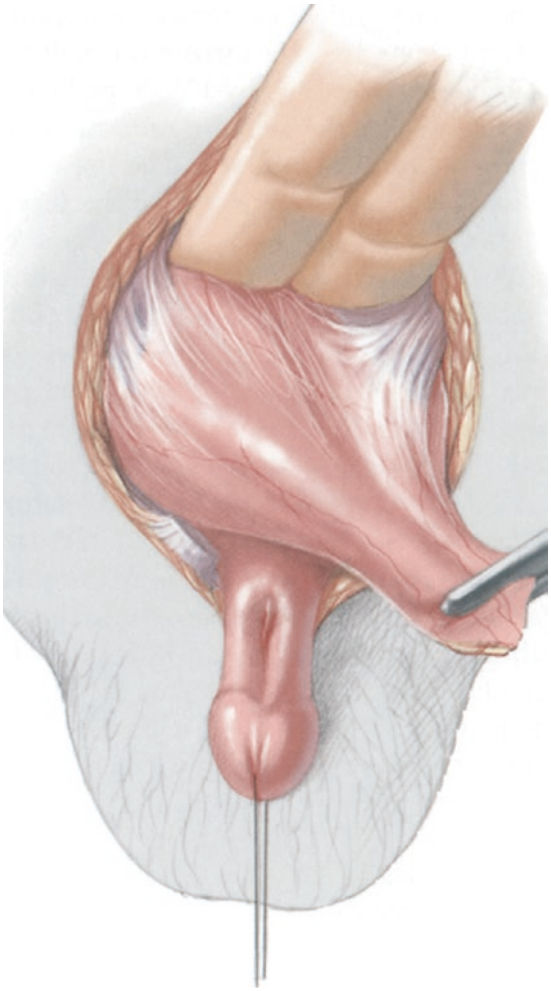

Fig. 69.4 Developing the retropubic space behind the bladder

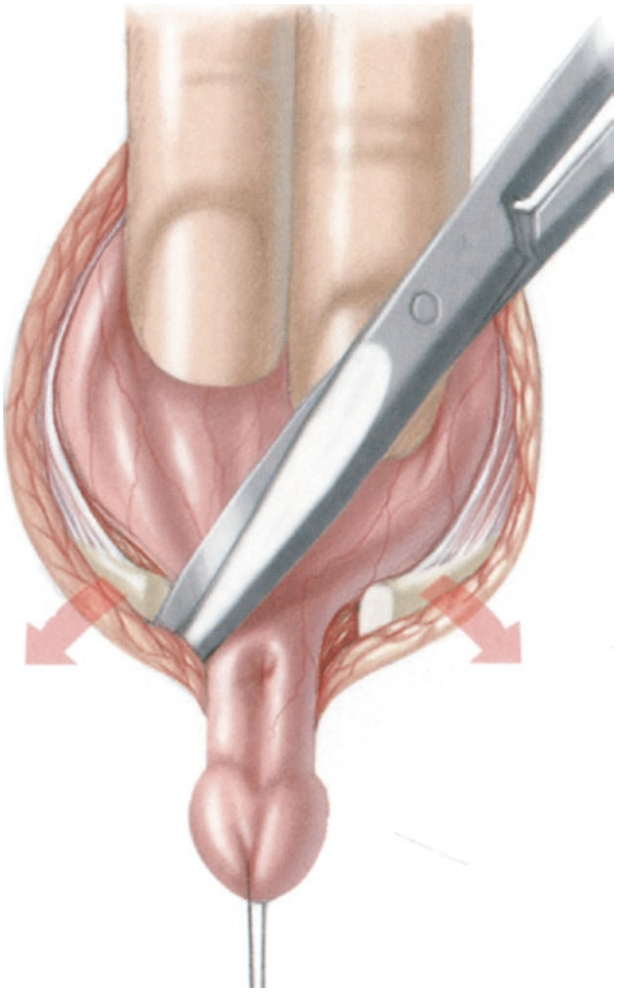

Fig. 69.5 Releasing fibrous bands

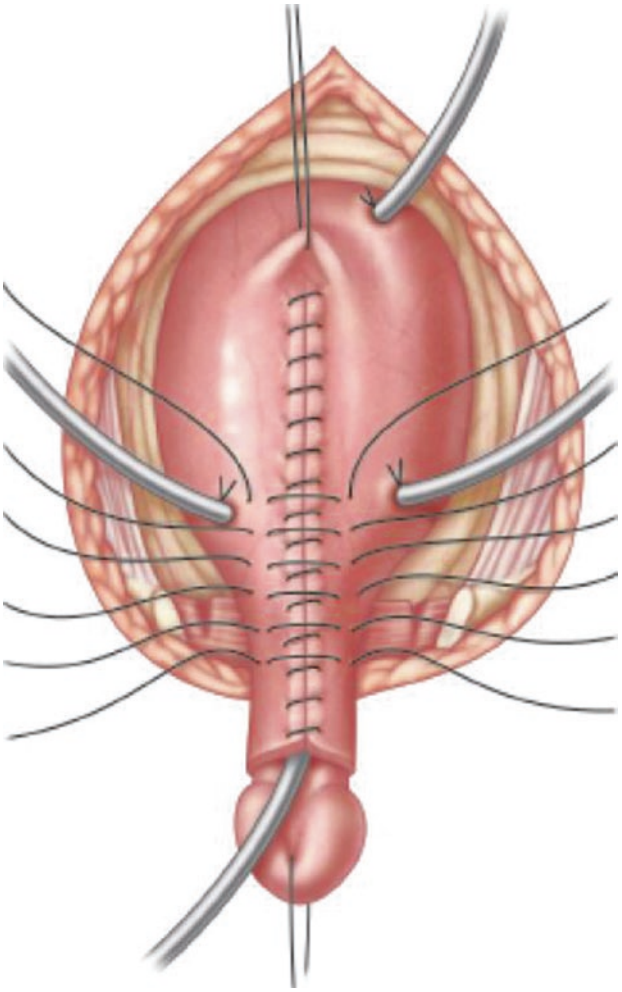

Fig. 69.6 Closure of the urethral plate

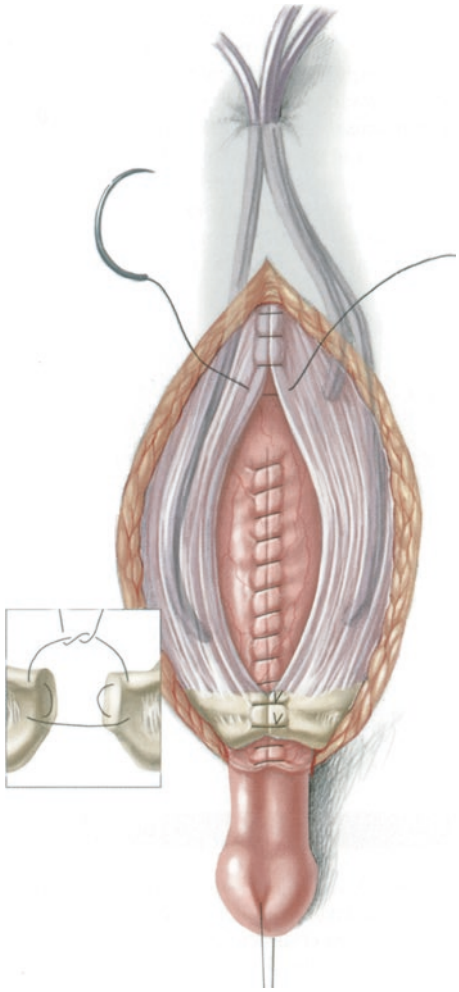

Fig. 69.7 Closure of abdominal fascia approximation of the pelvis

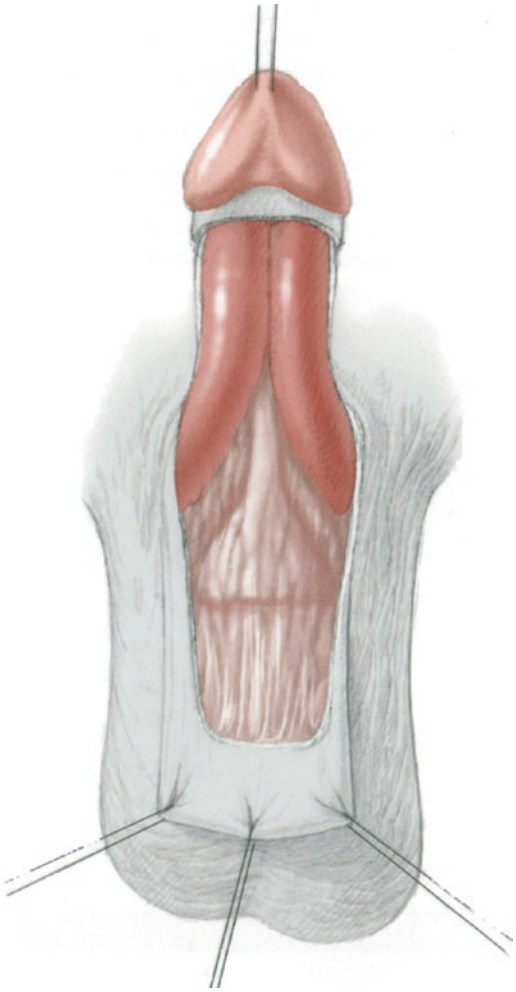

Fig. 69.8 Degloving the penis

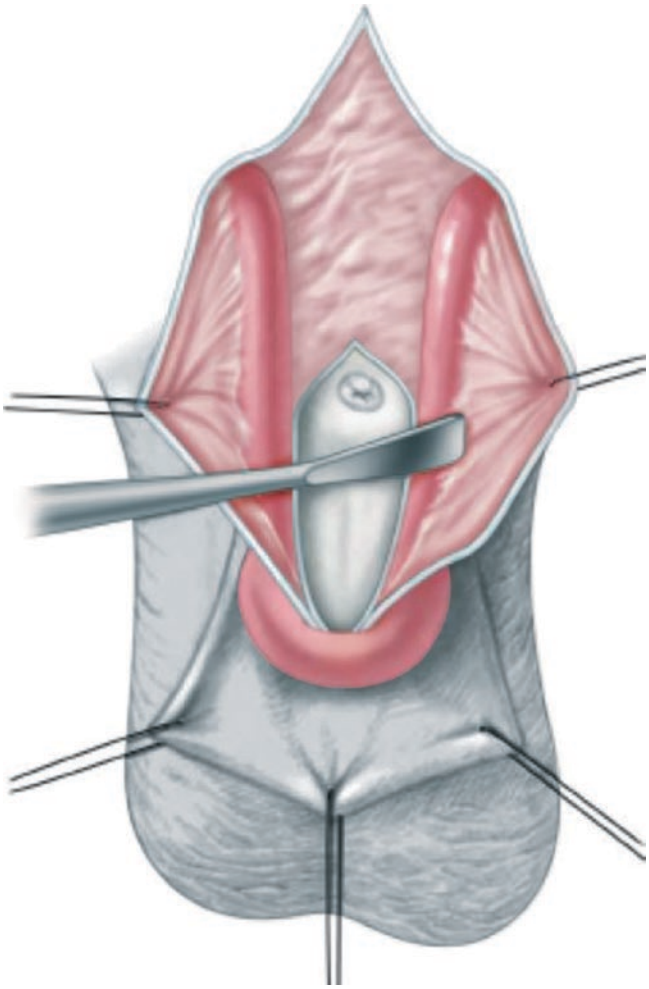

Fig. 69.9 Continuation of a modified Cantwell-Ransley epispadias repair

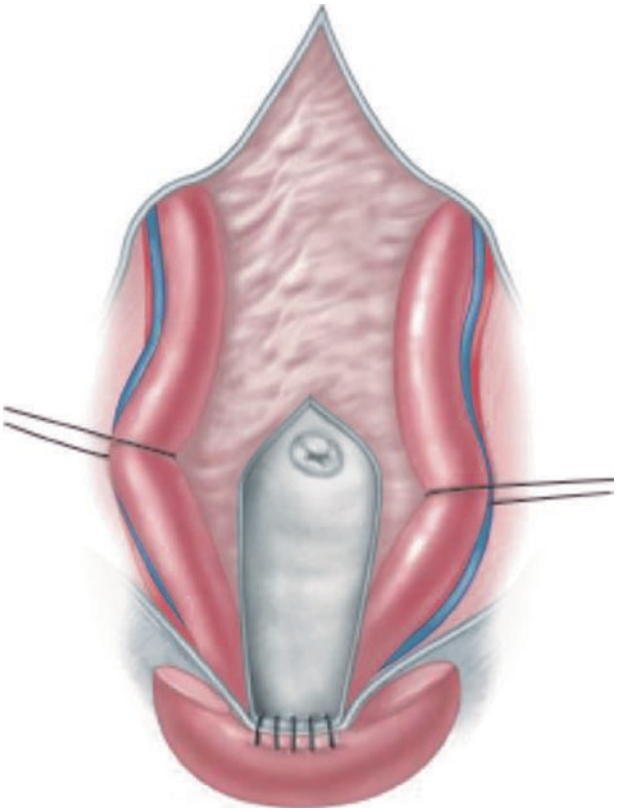

Fig. 69.10 Visualising the neurovascular bundles

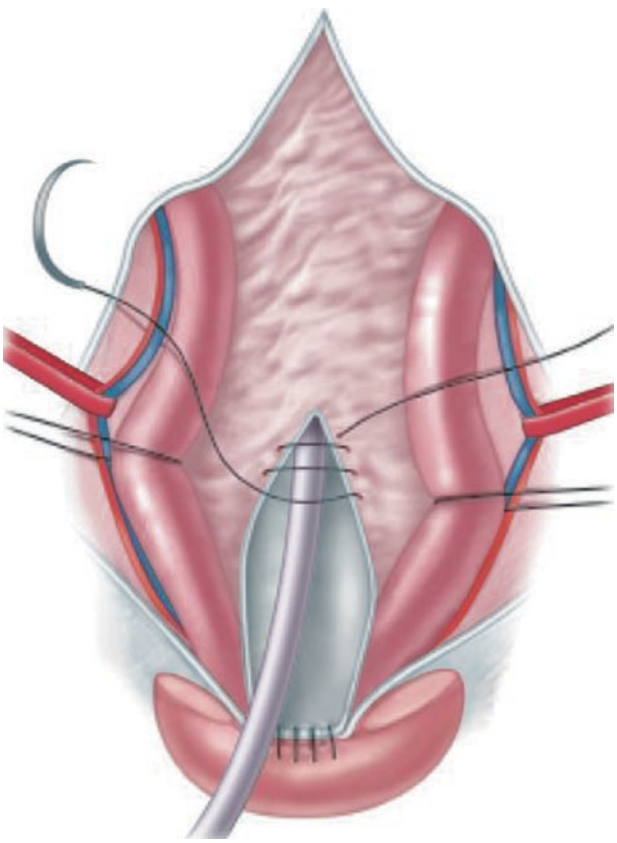

Fig. 69.11 Tubularising the urethral plate

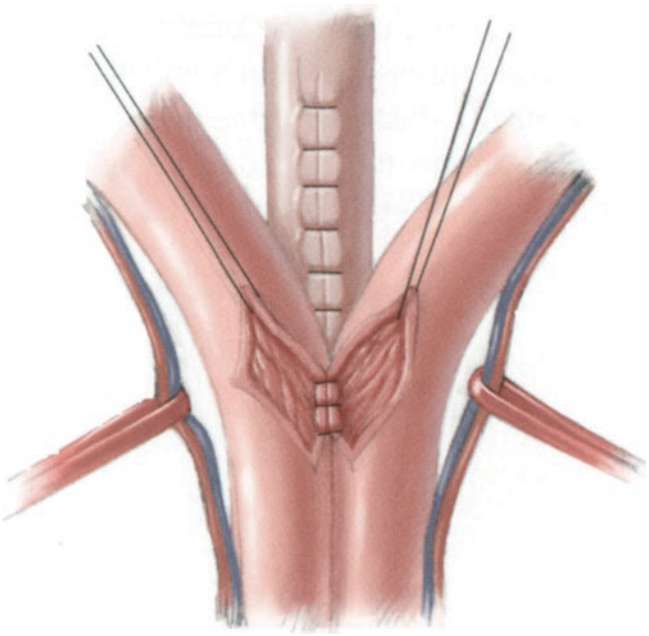

Fig. 69.12 Corporal incisions to correct curvature of the penis

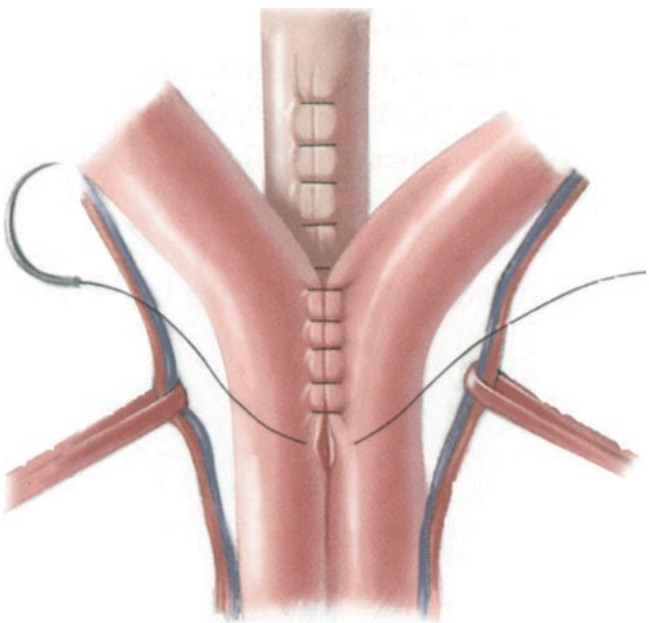

Fig. 69.13 Suturing of the corporal incisions

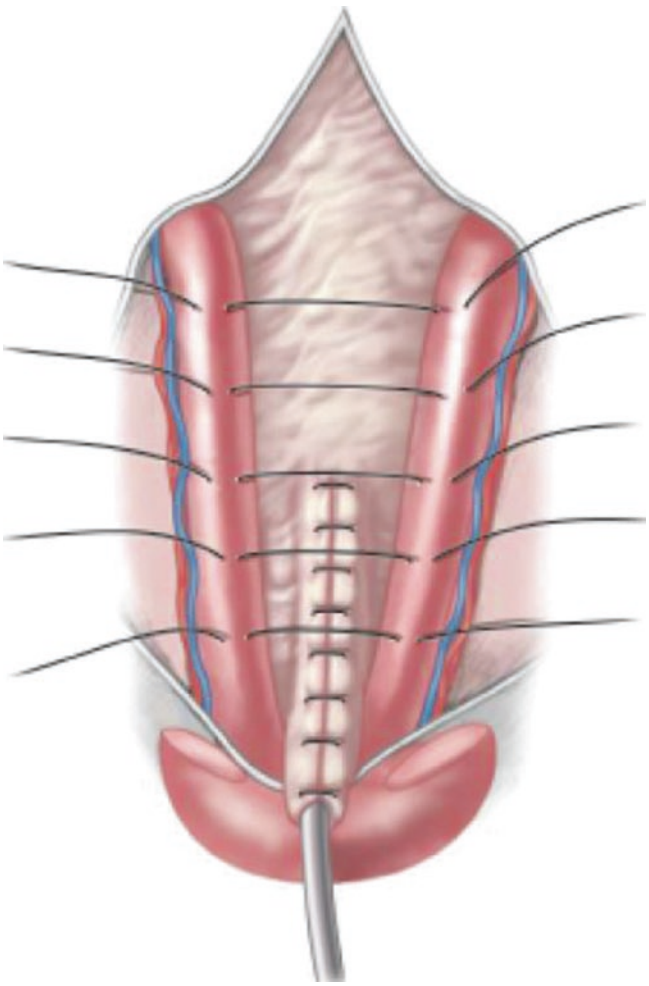

Fig. 69.14 Closing the corpora over the neourethra

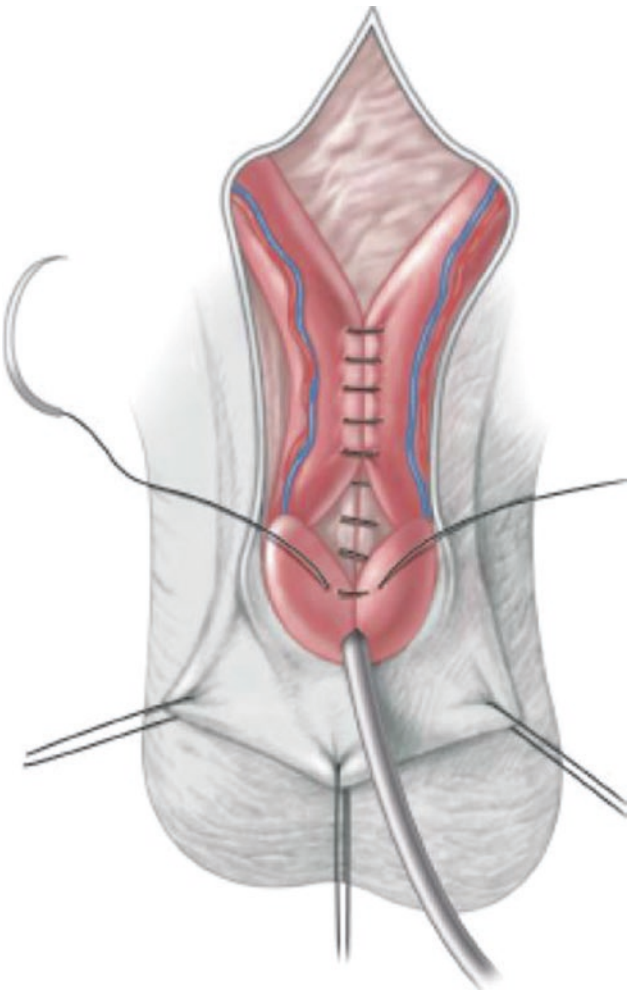

Fig. 69.15 Closing the glans

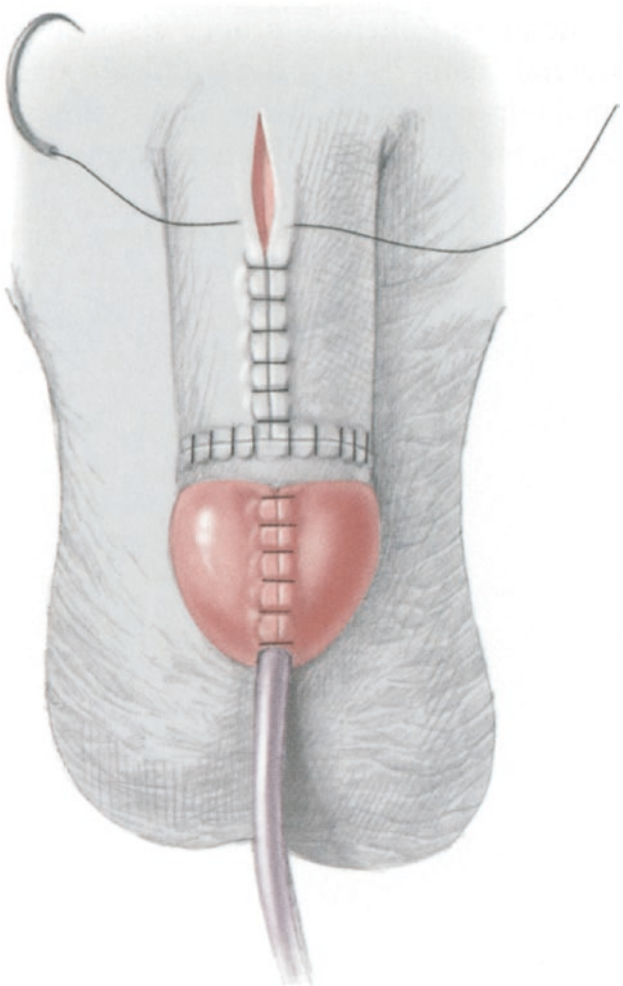

Fig. 69.16 Completing the reconstruction

69.2.3 Bladder Neck Reconstruction

Before the operation, the bladder capacity is measured annually by gravity cystograms with the child in anaesthesia. A bladder capacity of 100 mL or more is necessary to undergo bladder neck reconstruction. All children undergo an intense voiding training program, along with urodynamic evaluation, prior to the reconstruction.

The abdomen is accessed through a Pfannenstiel incision. The bladder is incised from the dome to the bladder neck with an additional vertical incision (Fig. 69.17). This type of incision will narrow the bladder neck at the time of midline closure, while enlarging the vertical dimension of the bladder. A very radical dissection of the bladder, the bladder neck, and the posterior urethra in the pelvis as well as behind the pubic bar is critical. If the posterior urethra cannot be visualized, one should not hesitate to cut the intrasymphyseal bar to gain exposure and access. Enough mobility of the vesicourethral complex is necessary to create an adequate narrowing and tightening of the bladder neck to achieve postoperative continence. The symphysis is closed afterwards with 0-PDS.

The ureters are reimplanted using standard Cohen's transtrigonal technique. If the ureteral hiatus is too close to the trigone, a cephalotrigonal reimplantation directs the ureters away from the trigone to ensure proper distance between the reconstructed bladder neck and the reimplants, preventing obstruction of the upper tracts. Ureteral stents are placed and brought through the bladder wall (Fig. 69.18).

A posterior mucosal strip 15 mm wide and 30 mm long, extending from the midtrigone to the prostate or posterior urethra, is outlined with a blue marking pen and incised using a no. 15 blade. Note that the transverse incision is only at the level of the mucosa and does not include the muscle (Fig. 69.19). Muscular incision at this level bears a high risk of denervation and ischaemia, leading to failure of the procedure.

The bladder muscle lateral to the mucosal strip is denuded of mucosa and covered in 1:200,000 adrenaline-soaked sponges to control bleeding for better visualization. The denuded lateral muscle triangles (Fig. 69.20) are tailored by multiple small incisions using electrocautery to allow the area of reconstruction to assume a more cephalic position.

The previously outlined mucosal strip is tubularized over an 8F urethral stent using interrupted 4/0 polyglycolic acid sutures (Fig. 69.21). The denuded muscle flaps are overlapped tightly and sutured firmly in place with 3/0 polydioxanone sutures to reinforce the bladder neck. The result is a mucosa-lined tube inside a muscular funnel narrowing from the bladder neck towards the posterior urethra. Three of the muscular sutures are left long and are brought through the rectus fascia as suspension sutures.

The urethral stent is removed after the bladder neck reconstruction. A suprapubic tube is brought through the bladder and secured with 4/0 polydioxanone sutures.

The bladder is closed in two layers (Fig. 69.22) and the tubes are brought through the skin and secured with a nylon stitch. The suspension sutures are elevated and tied on the rectus fascia, increasing outlet resistance, which is estimated intraoperatively by water barometer. Note the absence of a urethral catheter. The ureteral stents are removed after 21 days. The suprapubic tube is clamped for the first time 3 weeks postoperatively and is removed once the child empties the bladder without residual urine, as confirmed by ultrasound.

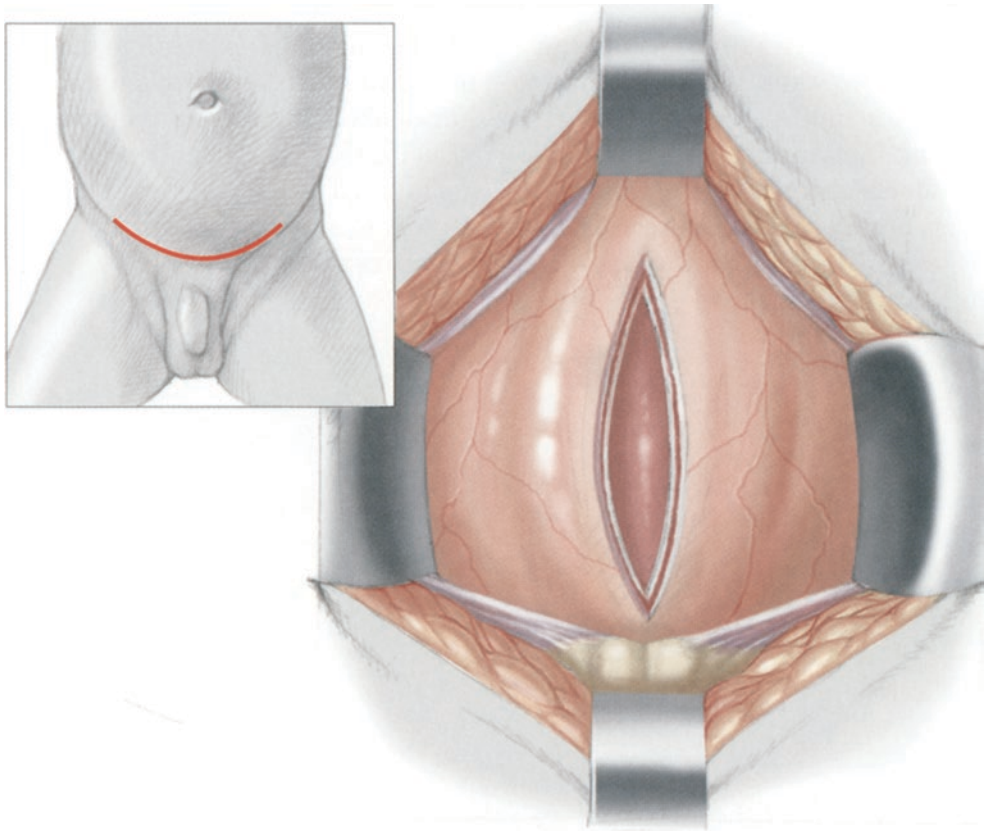

Fig. 69.17 Incisions for bladder neck reconstruction

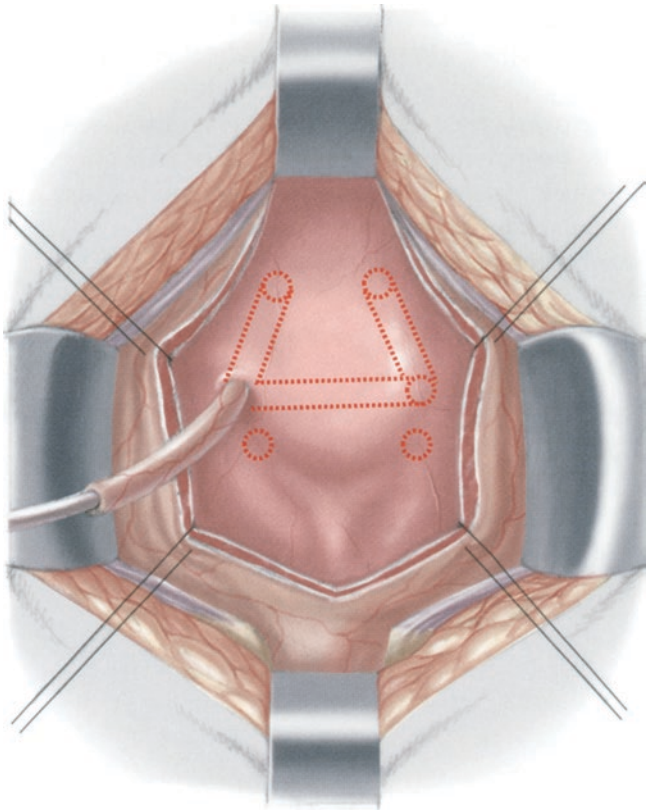

Fig. 69.18 Ureteral stents

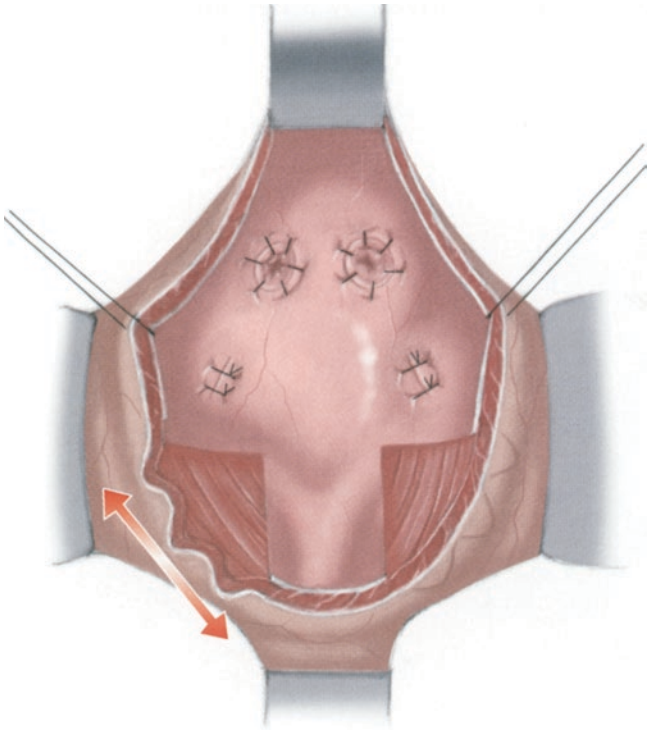

Fig. 69.20 Lateral muscle triangles

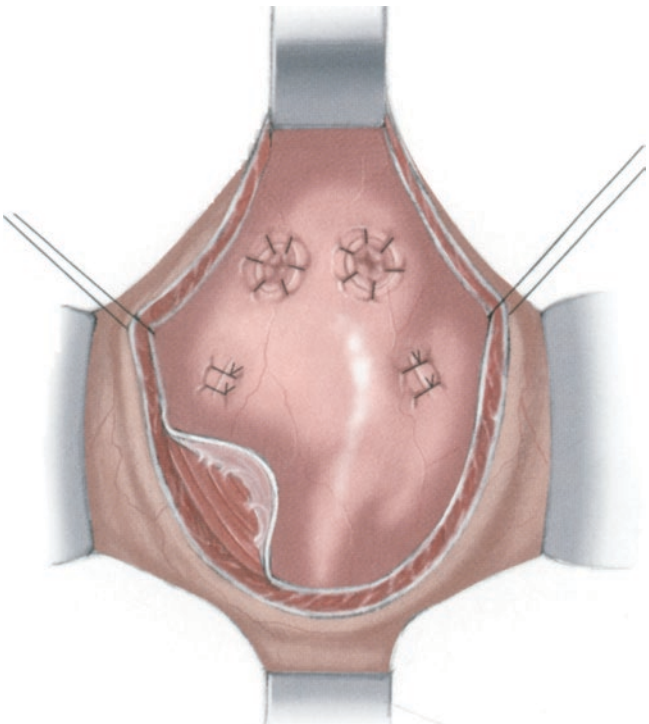

Fig. 69.19 Outlining a posterior mucosal strip

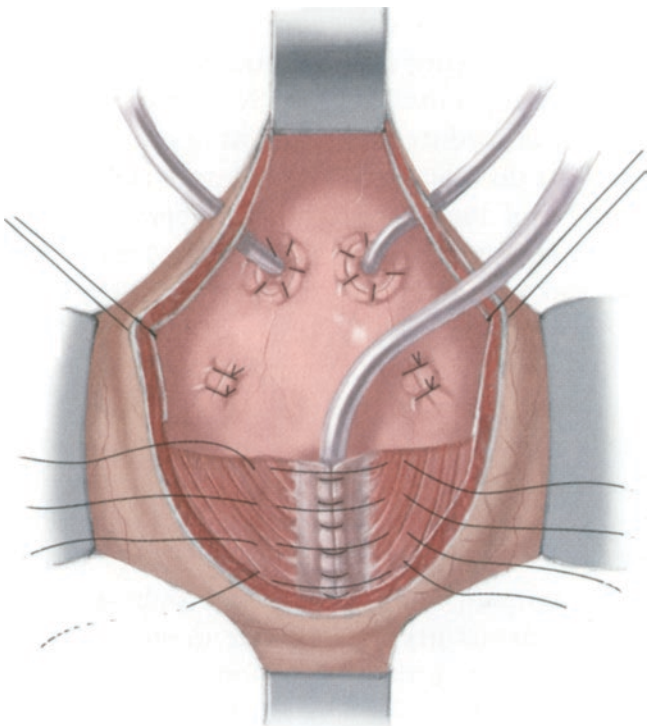

Fig. 69.21 Creating a mucosal tube inside a muscular funnel

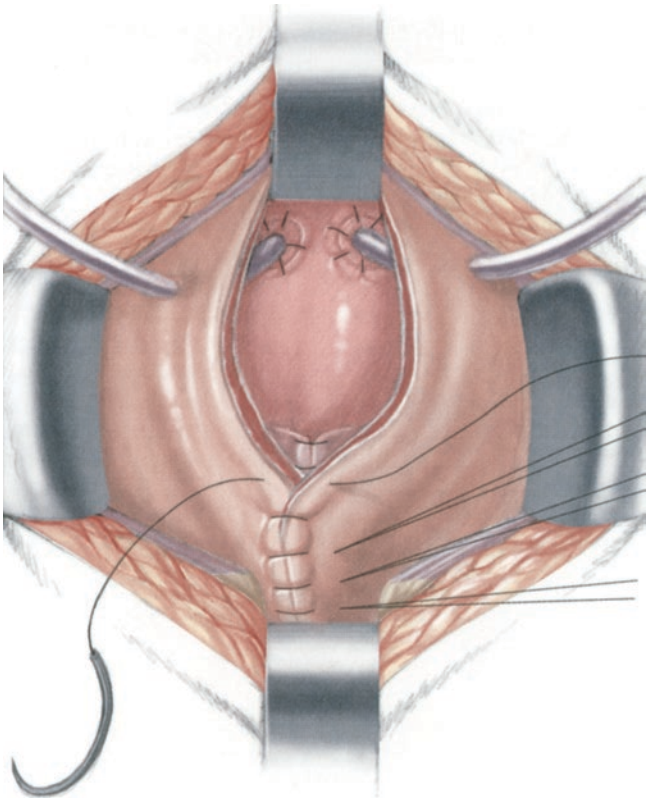

Fig. 69.22 Closing the bladder

69.3 Results and Conclusions

Successful initial closure of the bladder and posterior urethra is the most important factor for achieving urinary continence and sufficient bladder capacity. In the past decade, the procedure described here has been significantly modified from its original form, leading to a dramatic increase in the rate of success. Several series since then have shown the success and applicability of our method for the treatment of bladder exstrophy. Strict criteria for the selection of patients who are suitable for this approach have been defined, however. The fragile mucosa and the detrusor function are best preserved by closing the bladder in the newborn period, but the size and the functional capacity of the detrusor muscle are important considerations for the outcome. Therefore, in the rare presence of a small, fibrotic bladder patch without elasticity or contractility, the operation should be deferred until adequate growth of the bladder template occurs. The risk of bladder neck failure is higher for the group with smaller bladder capacities (<100 mL). If sufficient size is not reached 4–6 months after birth, alternative options like creation of a colon conduit or ureterosigmoidostomy must be employed. Later in life, the former urinary diversion can be converted into a continent catheterizable pouch bladder or augmented bladder if the template is still intact.

In our current database of 1278 patients, those who were treated entirely at our institution who underwent initial modern staged closure demonstrate a 72% rate of daytime continence and 83% rate of social continence. Continence is defined as being dry for more than 3 h. Socially continent patients achieve that goal during the day but have bedwetting incidents during the nighttime, often requiring treatment with desmopressin or anticholinergics.

The described approach for the functional closure of bladder exstrophy has developed in recent decades out of the experience and insight of committed researchers and surgeons in the field of the bladder exstrophy–epispadias complex. Pooling of experience in specialized centres is critical as surgeons, orthopaedists, anaesthesiologists, psychiatrists, researchers, nurses, child life experts, social workers, and the active exstrophy groups work together on a daily basis. The collaboration of these groups is the best basis to achieve the optimal outcome for each individual born with the malformation.

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Cloacal Exstrophy

70

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Cloacal exstrophy is the most severe anomaly in the spectrum of the exstrophy-epispadias complex. It is extremely rare, occurring in approximately 1 in 200,000 to 400,000 live births, with males affected twice as often as females. Cloacal exstrophy often presents with the OEIS complex, including associated omphalocele, exstrophy, imperforate anus, and spinal dysraphism. The exact embryology is still debated, but the defect is thought to be due to abnormal mesodermal migration during development of the lower abdominal wall, the urogenital sinus, and the anorectal canal.

70.1 Prenatal Diagnosis

With the advent of routine prenatal imaging, more patients are now diagnosed by prenatal ultrasound. Proposed ultrasound criteria include nonvisualization of the bladder, a large midline infraumbilical anterior wall defect or cystic anterior wall structure (persistent cloacal membrane), omphalocele, and lumbosacral anomalies. Proposed minor criteria include lower extremity defects, renal anomalies, ascites, widened pubic arches, narrowed thorax, hydrocephalus, and solitary umbilical artery. Additionally, the "elephant trunk-like image" due to prolapsing of the terminal ileum has been described as unique to cloacal exstrophy. Despite these predictive criteria, renal ultrasound remains a poor predictor of diagnosis, with less than 3% diagnosed by prenatal ultrasound.

Fetal MRI has been more recently used to identify cloacal exstrophy. Case series have described the presence of a cystic abdominal mass, abdominal wall defect, omphalocele, single umbilical artery, absence of meconium in the bowel, and nonvisualized bladder in the setting of normal amniotic fluid levels as suspicious for the diagnosis.

70.2 Associated Anomalies

Associated neurologic, skeletal, genitourinary, and gastrointestinal anomalies are common in patients with cloacal exstrophy. Spinal dysraphism is present in about 67% of patients and is associated with significant neurologic morbidity. Additionally, 31% have associated hydrocephalus, Chiari malformation, or other intracranial pathology, with increased risk of poor mobility. Skeletal system anomalies such as hemivertebrae and limb deformities are seen in over one fourth of patients.

Associated genitourinary anomalies are also common in these patients; renal anomalies including pelvic kidney, supernumerary kidney, and renal agenesis are seen in one third of all patients. Genital anomalies are also associated with cloacal exstrophy. In girls, duplication of the uterus and vagina is common, but the ovaries are generally histologically normal. In boys, up to 30% have associated diminutive or absent phallus requiring post-pubertal reconstruction. Cryptorchidism is also common. Although the testes appear histologically normal, no cases of paternity have been documented.

The OEIS complex commonly includes multiple gastrointestinal anomalies, including omphalocele, exstrophy of the cecal plate, blind-ending hindgut, and imperforate anus. Additionally, shortening of the small bowel, colonic duplication, duodenal web, malrotation, and duodenal atresia may be seen. Because of the associated risk of malabsorption in cases of short bowel syndrome, infants with cloacal exstrophy are at risk of impaired growth, with up to half of infants below the fifth percentile for weight. Despite the initial risk of short bowel syndrome, however, many of these children are ultimately able to adapt, with improved nutritional absorption over time.

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70.3 Postnatal Management

Cloacal exstrophy should be managed by a multidisciplinary team comprising a pediatric urologist, pediatric surgeon, neonatologist, pediatric neurosurgeon, and a pediatric orthopedic surgeon at a tertiary care center with interest and experience in the management of these complex anomalies. Prior to definitive management, a cling-film dressing should be used to cover the exposed bladder plate and the hindgut. This film minimizes fluid losses and reduces mucosal damage. The umbilical cord should be ligated with a nonabsorbable suture to prevent the umbilical clamp from abrading the bladder plate or the hindgut. Preoperative imaging including renal ultrasound and spinal imaging with ultrasound or MRI is necessary to identify potential associated anomalies requiring early intervention.

Intravenous access should be obtained in the upper limbs to avoid the associated lower limb anomalies and to allow for complete surgical access to the lower half of the infant. Routine preoperative labs (including a karyotype) should be drawn. Preoperative counseling should include discussion of gender assignment and anticipated needs for future surgery.

During initial reconstruction, the hindgut should be tubularized and all bowel segments, including the appendix, should be preserved to reduce the risk of associated fluid and electrolyte imbalances, or of malabsorption due to short bowel syndrome, and to preserve potential tissue for future genitourinary reconstruction. Data regarding the potential benefit of a single-stage exstrophy repair are mixed. Studies have suggested comparable urologic outcomes with either closure of the hindgut and creation of a diverting ostomy at a first stage with subsequent repair of the bladder exstrophy-epispadias complex at a second stage, or an integrated repair of both the gastrointestinal and genitourinary systems in a single stage. As in classic bladder exstrophy, osteotomies should be considered to reduce tension on the closure in cases of staged repair or wide pubic diastases. Close postoperative monitoring in the neonatal intensive care unit is necessary to ensure adequate immobilization and nutritional support in the early postoperative period.

70.3.1 Reconstruction Procedure

Figure 70.1 shows an anatomic example. The foreshortened hindgut or cecum is seen between the two hemibladders. The orifice of the terminal ileum, rudimentary hindgut, and appendix are seen on the cecal surface. The ileum may be prolapsed. The pubic symphysis is widely separated and the hips are externally rotated and abducted. The phallus is separated into right and left halves with the adjacent labium or scrotal half. Superiorly, there is an omphalocele containing small bowel and sometimes liver. There is a large variation between patients, and a classification system has been proposed.

The size of the omphalocele and the hindgut plate largely determine the extent of the initial closure. The order of the repair is closure of the omphalocele first, followed by separation of the hemibladders from the hindgut plate, GI reconstruction, and then bladder closure. Omphalocele closure is not always possible, and a silo may be required in some cases, as shown in Chapter 20.

Two 5F ureteric catheters are placed in the two ureteric orifices and secured with 5/0 absorbable sutures. Dissection commences first superiorly, and the umbilical vessels are doubly ligated and divided. The exstrophied section is separated from the skin and the adjacent hindgut with diathermy, taking care to avoid the ureters, which can be felt medially once the stents are inserted. The two hemibladders are then separated from the exstrophied hindgut.

Once this is performed, the length of the available hindgut is measured and the exstrophied hindgut is tubularized to recreate the ileocolonic valve. The terminal part of the colon is fashioned into a colostomy (Fig. 70.2). The appendix, where possible, is preserved.

The two hemibladders are approximated and the bladder is closed according to standard principles applied for primary bladder exstrophy closure. The urethra is tubularized over an 8F catheter, either completely (in girls) or partially (in boys).

The abdominal wall is closed in layers with interrupted 3/0 absorbable sutures. The pubic symphysis is approximated with a 0/0 suture. Osteotomies may be performed to assist closure. Skin closure is performed with subcuticular 5/0 absorbable suture, and the stents may be brought out through the suture line or via the neourethra (Fig. 70.3).

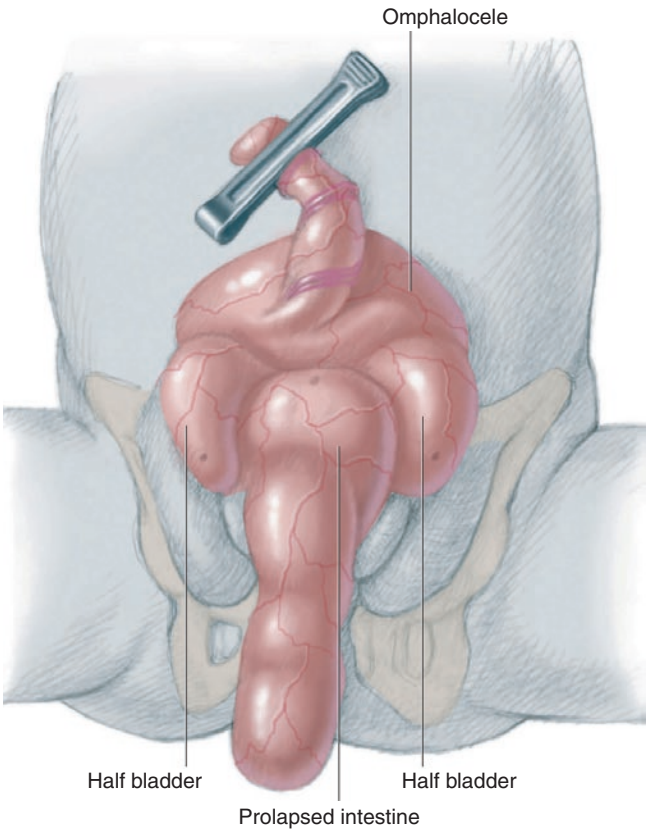

Fig. 70.1 Anatomy of cloacal exstrophy

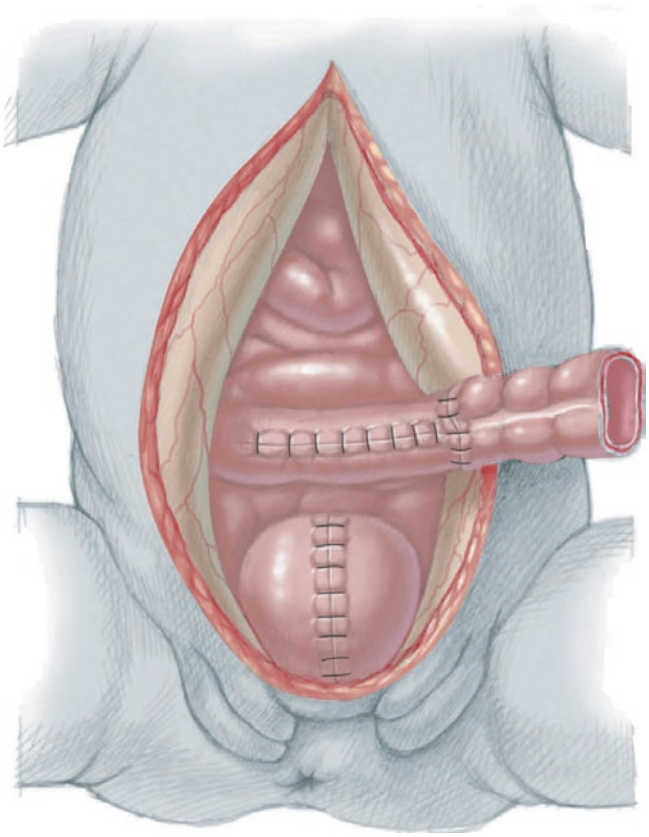

Fig. 70.2 Fashioning of a colostomy and closure of the bladder

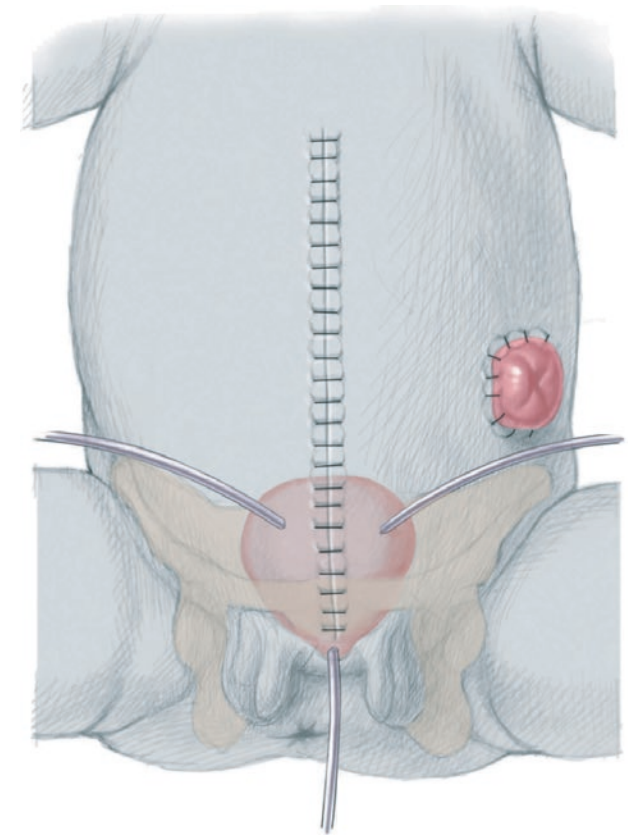

Fig. 70.3 Skin closure and stents

70.4 Urologic and Gastrointestinal Outcomes

Bladder outcomes appear to be less favorable than in classic bladder exstrophy. Prior series have reported more than 75% persistent incontinence after bladder neck reconstruction in these patients. As a result, most children will require additional urinary reconstruction with a catheterizable channel and possible augmentation cystoplasty to achieve continence. This decreased continence and need for catheterization may be due in part to a smaller bladder plate after closure and to the association with underlying spinal abnormalities. In their series of 23 patients, Husmann et al. found that presence of a clinically significant neurologic anomaly was associated with significantly lower continence rates (7% vs. 40%).

As in classic bladder exstrophy, patients undergoing bladder neck reconstruction or closure may be at risk for upper tract deterioration. In a series of 57 patients who underwent bladder neck procedures for exstrophy, 32% had evidence of hydronephrosis, with almost one fourth having evidence of renal scarring or severe hydronephrosis and two having evidence of renal insufficiency or failure. As a result, patients with cloacal exstrophy undergoing bladder neck reconstruction or ureteral reimplantation should be closely followed to minimize the risk of associated renal deterioration.

A considerable proportion of patients have morbidity related to the GI tract, and due attention should be paid to treating underlying bowel dysfunction. Because of concerns about adequate fluid and electrolyte absorption, preservation of the hindgut at the time of primary repair has been recommended. Patients with cloacal exstrophy have traditionally been managed with diverting ostomy owing to associated spinal cord abnormalities and the absence of normal sphincter mechanisms. In a series of 53 patients in which 27 underwent anal pull-through, however, 85% had bowel continence and 10% had occasional soiling with voluntary bowel movements. To maintain continence, a constipating diet, antidiarrheal medications, and management with an antegrade enema may be necessary. Similarly, in a series of 50 patients, another group found that 76% of patients who underwent a pull-through were able to be continent, with the majority requiring daily enemas to avoid fecal soiling. As a result, rectal pull-through should be considered as an option for definitive bowel management in motivated patients with adequate colonic length.

70.5 Gender Assignment and Quality-of-Life Outcomes

Gender reassignment in males with cloacal exstrophy remains a subject of debate. A study of cloacal exstrophy children aged 5–18 years suggests that children with cloacal exstrophy and genital ambiguity requiring gender assignment did not have significantly different social or behavioral competence or perceived quality of life when compared with children who had cloacal anomalies and no associated genital ambiguity. As a result, many pediatric urologists, especially older providers, continue to support female gender assignment owing to concerns about sexual function.

In a recent survey, however, younger pediatric urologists favored male gender assignment despite concerns about adequate phallic size and function in males who often have diminutive or absent phallic structures. This shift away from gender assignment is in part based on concerns about prenatal imprinting, especially given that these patients have normal testes. Prior studies have also suggested a significant discordance of gender identity with neonatal gender assignment in genetic males assigned to female gender. These findings suggest that a shift away from gender reassignment in these patients may be warranted despite concerns about future sexual function.

Long-term quality-of-life studies are limited in these patients. In a series of 23 adult females with bladder and cloacal exstrophy, over half were working, with 82% satisfied with their professional life. Over three fourths of women reported a satisfactory social life and were active sexually; over half were married or in a stable relationship. Of note, urinary incontinence and poor body image were significantly adversely associated with quality of life.

Conclusions

With advances in perinatal management and surgical reconstruction, most patients with cloacal exstrophy now survive. The emphasis has shifted from trying to achieve survival to providing a better quality of life to patients in the long term. Psychological well-being and stable gender identity are paramount when considering the quality of life for patients with cloacal exstrophy. Because of the rarity of this anomaly and the lack of data regarding long-term function and quality of life in these patients, the optimal short-term and long-term management remains unclear, so treatment plans should be individualized to the needs of the patient, in close consultation with the family.

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Augmentation Cystoplasty and Appendicovesicostomy (Mitrofanoff Principle)

71

Boris Chertin

There are two types of cystoplasty: augmentation cystoplasty, in which the bladder is enlarged, and substitution cystoplasty, in which the bladder is replaced. Augmentation cystoplasty is now commonly performed at most pediatric urological centres. Bladder augmentation has three major goals: to provide a compliant bladder reservoir, to limit bladder contractility, and to increase bladder capacity. Augmentation cystoplasty should allow the urinary tract to remain intact while preserving renal function and providing urinary continence. Various substrates are utilized to augment the bladder; the most commonly used is a segment of ileum, but stomach and large bowel also have been used. Ileum has been demonstrated to be the least contractile segment and therefore has become the tissue most often used for bladder augmentation. Sufficient augmentation should lead to effective bladder capacity.

Clean intermittent self-catheterization (CISC) has become a universal procedure that ensures effective emptying of the bladder after augmentation, but although CISC is simple and easy for mobile patients to perform, there are a relatively high number of failures in wheelchair patients who have difficulties in self-catheterization. To overcome those difficulties, different techniques of continent diversion have been proposed. The technique of appendicovesicostomy, which is also well known as the Mitrofanoff principle, has gained a wide popularity.

The indications and detailed descriptions of the different types of augmentation cystoplasty and continent diversion are outside the scope of this chapter. This chapter addresses the technical details of augmentation cystoplasty utilizing either ileal patch (ileocystoplasty) or ileum segment folded in a U shape and also describes the technique of appendicovesicostomy.

71.1 Preoperative Preparation

Children are admitted to the hospital 2 days prior to the surgery, and a clear fluid diet is started. The bowel preparation using GoLYTELY® (PEG-3350 and electrolytes) is administered, with an enema performed the night before surgery. (Recent trends are changing some of this preparation to the outpatient setting.) Special attention should be paid to the bowel preparation of patients with neurogenic dysfunction, who often have chronic constipation. Parenteral antibiotics such as gentamycin, ampicillin, and metronidazole are administered at the induction of anaesthetic.

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71.2 Surgical Procedures

71.2.1 Augmentation Cystoplasty

A lower midline incision is made from pubis to umbilicus. Some surgeons recommend a lower Pfannenstiel incision, but in our opinion this incision may limit exposure. In contrast, a lower midline incision can be easily extended and allows access to the entire abdomen. Following fascia incision, the pyramidalis muscles and recti are exposed. The retropubic space is widely opened in order to expose the surgical field all the way around the front and both lateral aspects of the bladder. The urachus is defined at the dome of the bladder, and is ligated and divided. The peritoneum is opened to expose the dome of the bladder with its peritoneal covering. Using a traction suture at the urachus, the plane cleavage along lateral bladder margin down to the ureters and superior vesical pedicle on each side is performed (Fig. 71.1, *dotted line*).

Following maximal bladder exposure, incision of the bladder wall is performed in either a circumferential or midline fashion. If the surgeon prefers circumferential incision, the bladder is exposed from the point of 2 cm from the ureteric orifice. In the case of midline bladder incision, the bladder is opened 1 cm from the bladder neck on each side of the bladder. For many years we have used the circumferential incision shown in Fig. 71.2, which gives maximal exposure without jeopardizing the ureters. Safe incision is achieved by starting on the bladder dome and opening the bladder using diathermy point down one side at a time. The use of ureteric catheters is an option at the time of incision. Tape or tubing is then used to measure the circumference of the bisected bladder, in order to define the required length of the bowel segment.

A convenient segment of the terminal ileum is isolated 15 cm proximal to the ileocecal valve (Fig. 71.3). The segment should be equal in length to the measured maximal circumference of the bisected bladder, and it should be supplied by a well-defined vascular pedicle. Two-layer ileoileostomy using GIA staplers or hand-sewn sutures is performed, and the ileal segment is then opened on the antimesenteric aspect to produce a patch.

The ileal patch is inset into the bisected bladder and is sewn in place (Fig. 71.4). To prevent overlap of the ileal edge and bladder edges (because of the tendency of the bladder wall to contract during the procedure), we halved and thereafter quartered the suture line with stay sutures. The ileal patch is anastomosed to the posterior bladder wall first, using 3/0 absorbable continuous sutures, locking the stay sutures as they are encountered. If the appendix is to be implanted for the Mitrofanoff principle, that should be done before the patch is sewn to the other side of the bladder.

If more volume of the bladder is required, the ileal segment can be significantly longer, allowing it to be folded in a U shape (Fig. 71.5) or even into an S or W configuration.

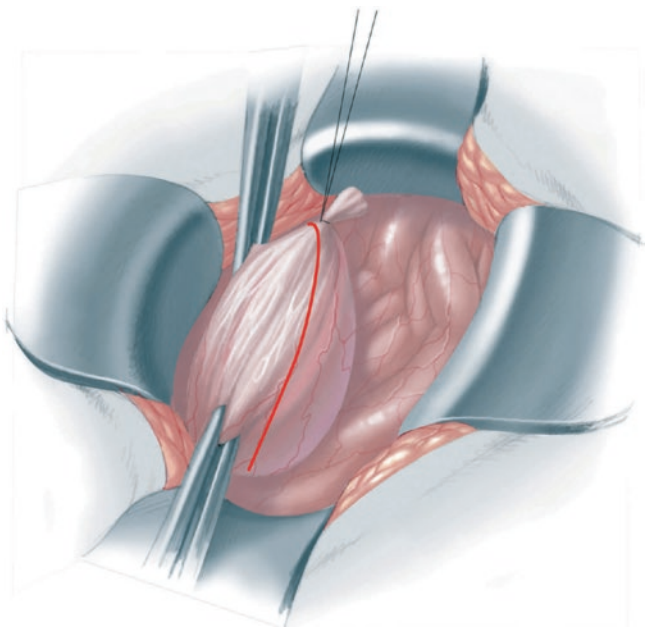

Fig. 71.1 Line of plane cleavage along the lateral bladder margin

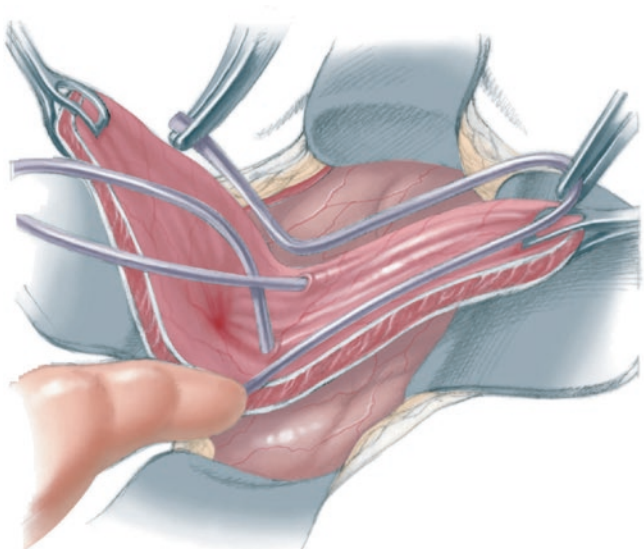

Fig. 71.2 Circumferential incision for bladder exposure

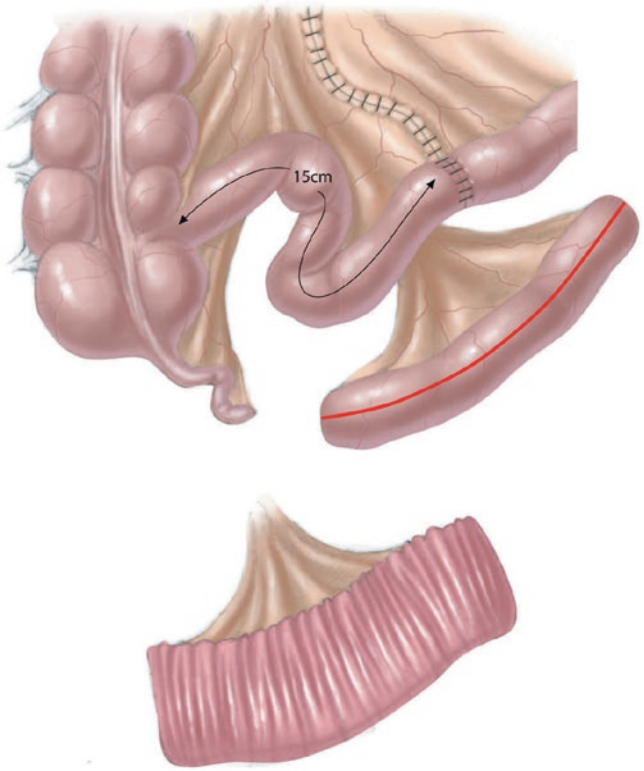

Fig. 71.3 Choosing and preparing the ileal segment

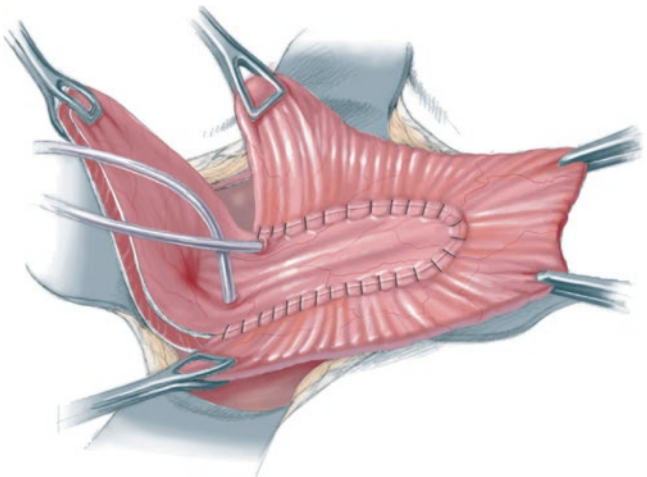

Fig. 71.4 Sewing the ileal patch to the posterior bladder wall

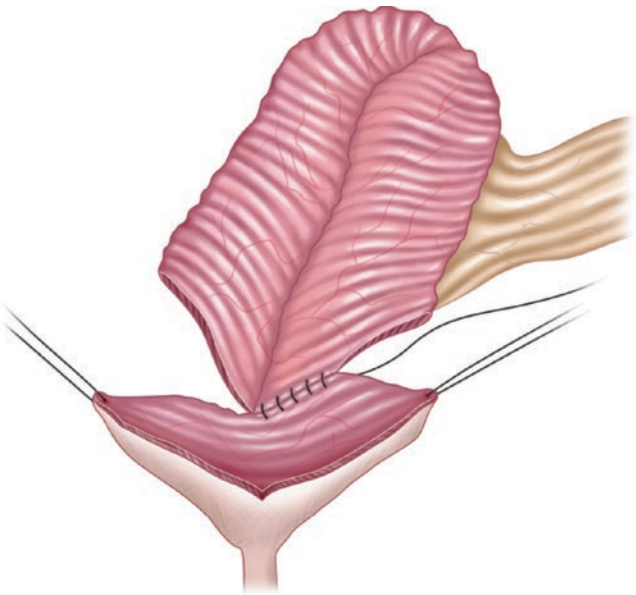

Fig. 71.5 Folding a U-shaped patch for greater volume

71.2.2 Mitrofanoff Procedure

If a catheterizable Mitrofanoff procedure is to be performed, it should be done at this time. The caecum and appendix are widely mobilized, and the appendix is isolated with its vascular pedicle and a small cuff of caecum (Fig. 71.6). The caecum is closed.

The appendix is opened at the tip. The appendix is implanted in the bladder above the trigone in a transverse or oblique submucosal tunnel (Fig. 71.7). The tunnel should be at least 2.5–3.0 cm in length, with sufficient hiatus in the detrusor to avoid constricting its blood supply.

When the appendicovesicostomy is completed, the ileal patch is flipped over and the procedure is repeated in order to seal the anterior bladder anastomosis (Fig. 71.8). The anastomosis is performed as before, but this time the anastomosis is performed outside the bladder. At the end of the procedure, a suprapubic catheter (big enough not to be obstructed by ileal mucus) is left in the bladder. The catheter is brought out through the native bladder wall rather than through the suture line between the bladder and the ileal patch. A wound drain is left in the retropubic space.

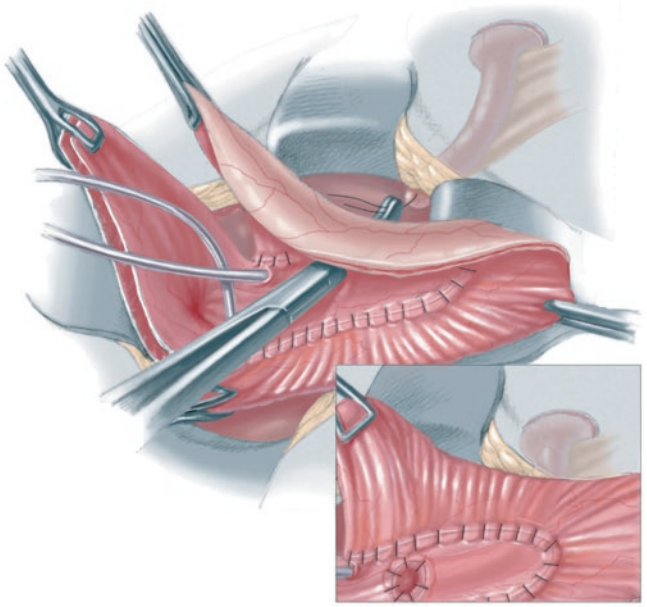

Fig. 71.7 Implanting the appendix in the bladder

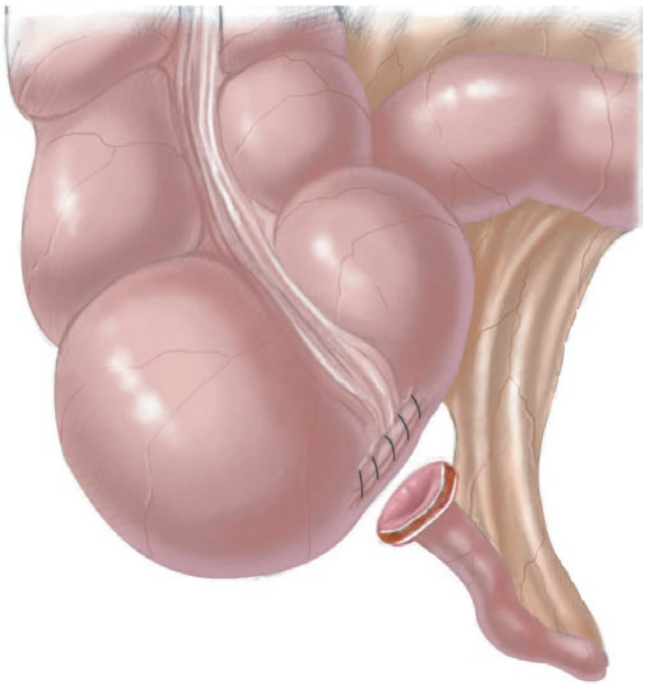

Fig. 71.6 Isolating the appendix and its pedicle

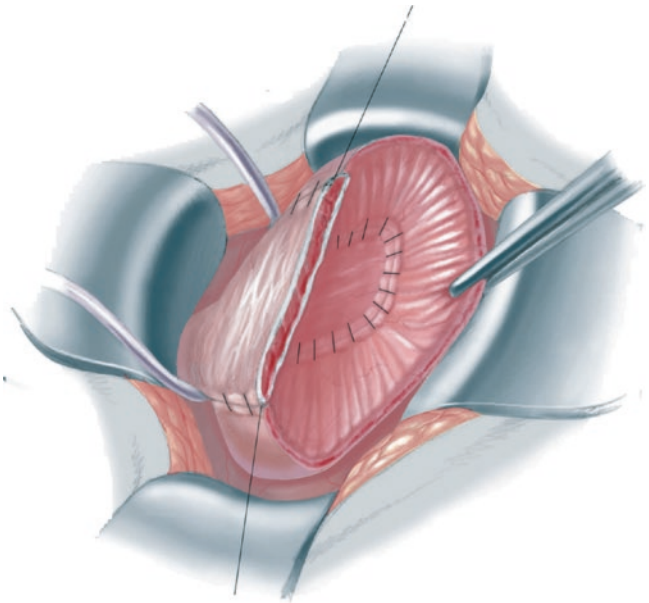

Fig. 71.8 Completing the bladder anastomosis with the ileal patch

71.2.3 Extravesical Appendicovesicostomy

In some patients, we have found it extremely helpful to implant the appendix into the bladder in an extravesical fashion, allowing us to choose the precise place for the appendicovesicostomy and to create a straight channel for future intermittent catheterizations. Following appendix isolation and completion of bladder augmentation, the bladder is filled out with saline to 60% of its estimated volume. The appendix then is implanted in the lateral bladder wall in the same fashion as extravesical reimplantation, avoiding any ureteral twisting or kinks between the bladder and the anterior abdominal wall. The detrusor is incised with low current electrical cautery to create a submucosal tunnel. Then tenotomy scissors are used to divide the detrusor fibers along a newly created tunnel. Care should be taken to incise all detrusor fibers without injuring underlying mucosa. Following dissection, the mucosa is opened and the bladder is decompressed. The appendicovesicostomy is completed and the detrusor is reapproximated using interrupted 3/0 absorbable sutures (Fig. 71.9). After creation of the appendicovesicostomy is completed, a 10 F or 12 F Foley catheter is passed through the appendix to ensure a free passage and the absence of urinary leaking or kinks.

The base of the appendix is drawn to the skin through a large hole in the abdominal muscles, avoiding any kinking in its passage. At this stage, the bladder is fastened to the abdominal muscle, taking care to avoid strangulation of appendix and its pedicle. The site of the stoma on the skin is selected entirely for the patient's convenience. The umbilicus provides a good passage and the best cosmetic result, but it is not an option in exstrophy patients. With patients in wheelchairs, the spine tends to become twisted with time, causing progressive abdominal compression, and the abdomen becomes hidden from the patient's field of vision. For these patients, a stoma site close to the xiphisternum is the best option. The cutaneous end of the appendix is spatulated, and a triangular skin flap (2–3 cm) is sutured into the spatulation with absorbable sutures, leaving a small portion of mucosa visible in order to prevent any risk of delayed stenosis (Fig. 71.10). If the conduit is short, the triangular skin flap is raised and rolled over the catheter to make a tube, which is sutured to the spatulated end of the conduit to lengthen it. A 10F or 12F Foley catheter is passed through the appendix and is left in the bladder for least 2 weeks.

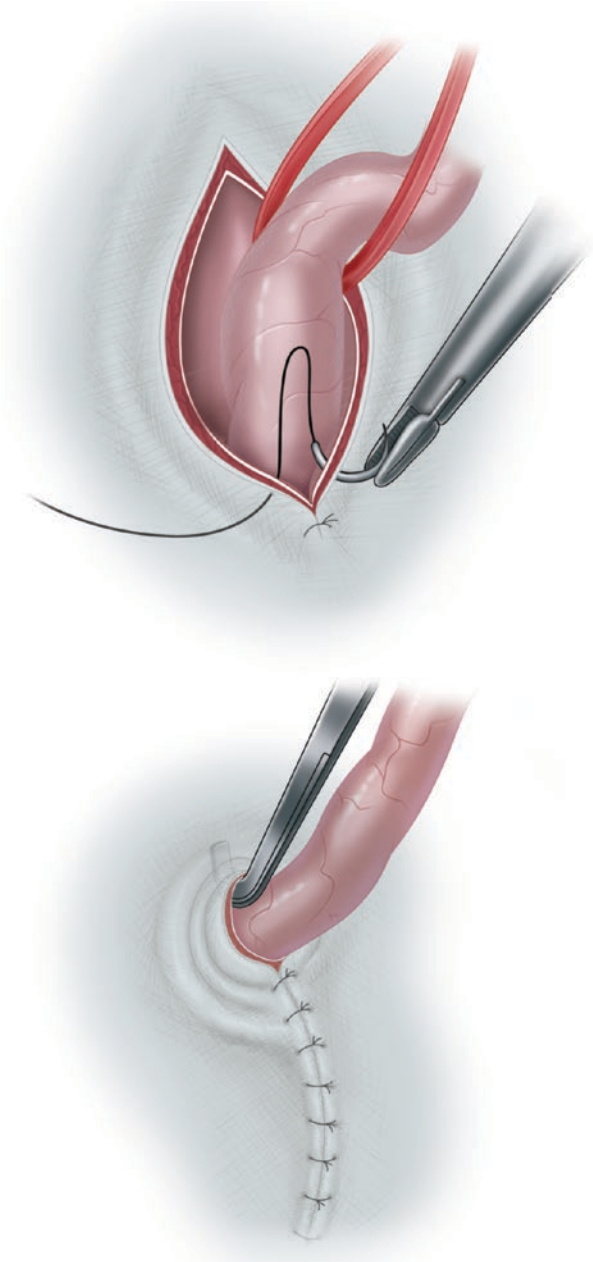

Fig. 71.9 Completing the extravesical appendicovesicostomy

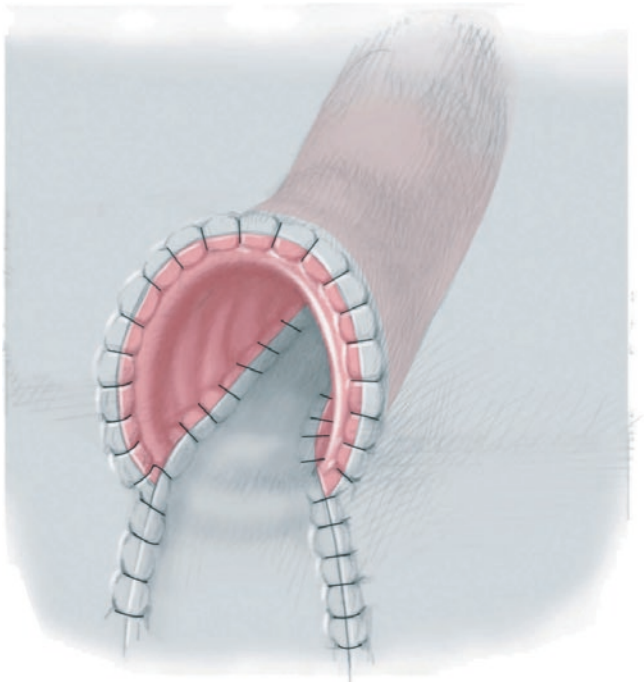

Fig. 71.10 Creation of the cutaneous stoma

71.3 Postoperative Care

The wound drain is removed when it stops draining. The suprapubic catheter is clamped about 8 days after surgery. When the suprapubic catheter is clamped, the residual urine volume is checked out. The catheter is removed 24 h after satisfactory voiding. In children being managed by intermittent catheterization, we have kept the urethral indwelling catheter for 48 h to ensure a complete healing of the suprapubic site prior to commencement of the intermittent catheterizations.

71.4 Complications

We have used augmentation cystoplasty (with either an ileal patch or a U-shaped ileal segment) for years with a negligible complication rate and satisfactory results with respect to bladder compliance and capacity in the long-term follow-up. None of our patients have needed secondary augmentation because of increased contractile activity.

Even after the ileal segment is isolated from the gastrointestinal tract, it retains its absorption and secretory properties. As a result of the electrolyte exchange, hyperchloremic metabolic acidosis may develop. The extent of the electrolyte exchange depends on the amount of intestinal surface area in contact with urine and the time that the urine remains in contact with the intestine. Patients with normal renal function prior to augmentation have adequate compensatory mechanisms and do not have significant problems with acute metabolic changes. Patients who develop persistent, severe metabolic changes must be evaluated for insufficient bladder emptying. Another concern in these patients is a possible increased incidence of urinary tract infection (UTI), as bacterial flora from the augmentation may colonize the urinary tract. In recent years, we have used in our patients daily irrigation of the augmented bladder with 30 mL of 120 mg gentamicin per 250 mL of normal saline, and have found a low incidence of UTI with no evidence of gentamicin absorption or renal function deterioration.

Suggested Reading

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The MACE (Malone Antegrade Continence Enema) Procedure

72

Frank J. Penna and Martin A. Koyle

Since its development in 1989, the ACE procedure has become widely accepted as a valuable addition to the therapeutic regimens available for treating intractable faecal incontinence associated with conditions such as myelomeningocele and anorectal malformations. Thousands of patients around the world have undergone ACE procedures with success rates in excess of 80% reported.

The original procedure described disconnecting the appendix from the caecum, amputating its tip, reversing it, and reimplanting it into a submucosal tunnel on the anterior wall of the caecum to produce a continence mechanism; the stoma was usually sited in the right iliac fossa. Since the original description, numerous modifications have been proposed and introduced, so it is now incorrect to talk about the ACE procedure; and it would be more accurate to use the term *ACE principle.*

Because a failure rate of 20% exists, it is useful to perform an initial therapeutic trial of antegrade washouts by minimally invasive means before proceeding to the definitive procedure. Under either radiologic or colonoscopic control (as one would perform a percutaneous endoscopic gastrostomy, PEG), a colonic catheter can be inserted percutaneously into any part of the large bowel and used to administer the enemata. If constipation is the major problem, the catheter is best placed in the distal descending colon, but in the absence of constipation the caecum remains the best site. If the washouts are successful, the patient has a choice: to keep the catheter, exchange it for a colonic button (identical to a gastrostomy button), or have a conduit constructed at open surgery. In our experience, most patients ultimately opt for a conduit, as there is a tendency for leakage of flatus, stool, or washout fluid to occur around the side of the buttons. Using this approach, it is also possible to test in which part of the colon the conduit will work best. Increasing numbers of conduits are now placed in the distal descending colon because the time taken to perform the washout is reduced.

In practical terms, there are now two types of ACE, the original *caecal ACE* and the new *left colonic ACE.* For the caecal ACE, many surgeons now advocate simply amputating the tip of the appendix and bringing the open end onto the abdominal wall without constructing any continence mechanism. This procedure is being increasingly performed laparoscopically. In fact, since the first description of the laparoscopic approach to the ACE (LACE), the laparoscopy-assisted approach has been applied with good success to many reconstructive urologic procedures, including bladder augmentation, appendicovesicostomy, ileovesicostomy, and ACE procedures. The benefits of laparoscopic surgery in children include decreased morbidity with smaller incisions, better surgical cosmesis, less potential for intra-abdominal adhesion formation, less pain, shorter hospital stay, and quicker convalescence. For the surgeon, it provides easier access to difficult anatomy (i.e. malrotated caecum) and better accommodation to altered body habitus (obesity, exaggerated lordosis).

The procedure has, in fact, been reported in situ with good success without caecoplication. Caecoplication has been thought to be a potentially unnecessary step because of the inherent valve mechanism of the appendix. The procedure can be performed completely intracorporeally or extracorporeally with similar results. A combined procedure (Mitrofanoff and ACE) using a robot-assisted laparoscopic approach with a divided appendix also has been described; in experienced hands, this approach has the potential to result in shorter operative times.

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P. Puri, M. E. Höllwarth (eds.), *Pediatric Surgery*, Springer Surgery Atlas Series,
https://doi.org/10.1007/978-3-662-56282-6_72

72.1 Surgical Procedures

72.1.1 Basic ACE Procedure

The patient is placed supine on the operating table. The caecum is mobilized, the tip of the appendix is amputated, and a stay suture is inserted in the open end to apply traction. A 10–12 Ch catheter is passed via the appendix into the caecum, to confirm that it is catheterizable (Fig. 72.1). The stretched mesentery is inspected and fenestrated between the vessels. Doing so allows the caecum to be wrapped around the appendix through the mesenteric windows, to produce the continence mechanism without compromising the blood supply. Stay sutures are inserted into the caecum alongside the anterior taenia to keep it under tension whilst a submucosal tunnel is made. The serosa and muscle are initially incised using a diathermy (Fig. 72.1) and the trough is widened to expose the submucosa by spreading a mosquito artery forceps as one would perform a pyloromyotomy (Fig. 72.2). It is important that this incision includes the base of the appendix, as it allows this area to be buried in the caecum, reducing angulation and making catheterization easier.

The appendix is folded along the length of the submucosal tunnel and the caecum is wrapped around it, as one would perform a Nissen fundoplication, to produce the continence mechanism (Fig. 72.3). This is done using an absorbable 4/0 suture. The first suture is placed at the base of the appendix picking up the caecum–appendix–caecum to ensure that the appendix is firmly secured in the tunnel, preventing movement and kinking and facilitating easy catheterization. Further sutures are progressively placed along the length of the appendix in a similar fashion, bringing the caecal wrap through the mesenteric windows that were created earlier. The entire appendix is wrapped within the caecum until only a sufficient length is left to bring it out through the abdominal wall when the stoma is constructed (Fig. 72.4). The antimesenteric end of the appendix is spatulated to allow a V flap of skin to be inlaid during construction of the stoma to reduce the incidence of stomal stenosis. It is important to anchor the caecum to the posterior aspect of the anterior abdominal wall using absorbable sutures so that it is not hanging on the appendix and at risk of torsion.

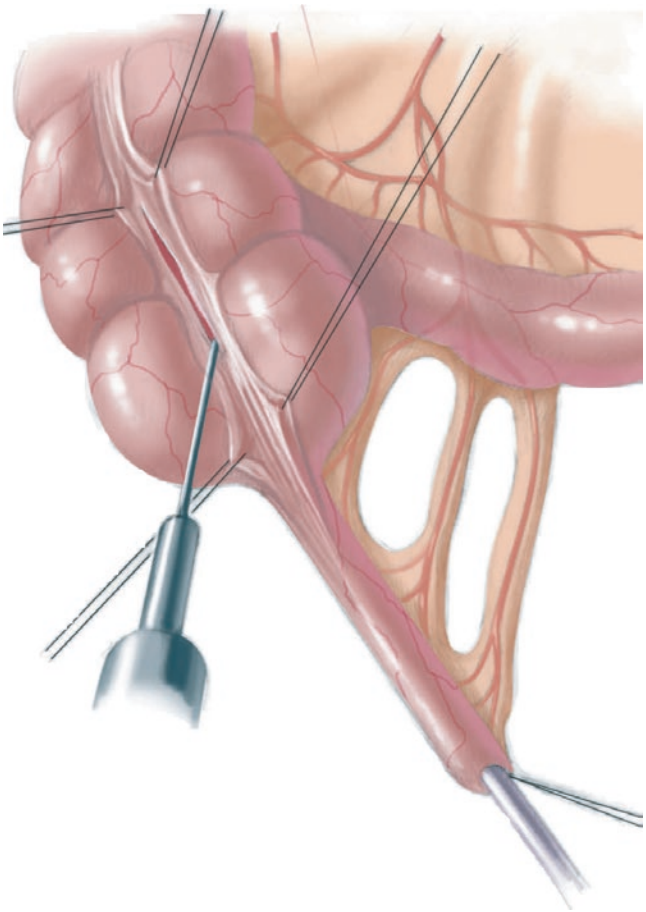

Fig. 72.1 With stay sutures in place, incision of the caecal serosa and muscle

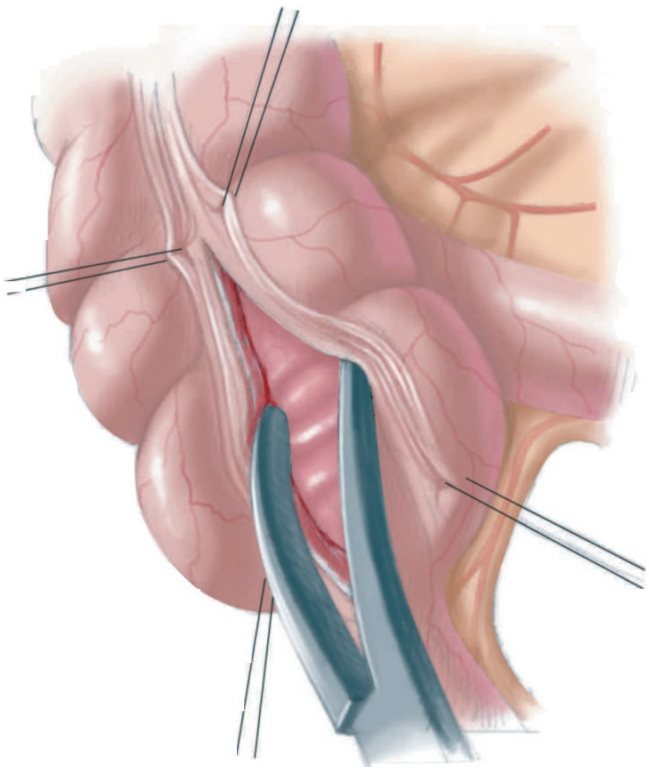

Fig. 72.2 Widening the trough

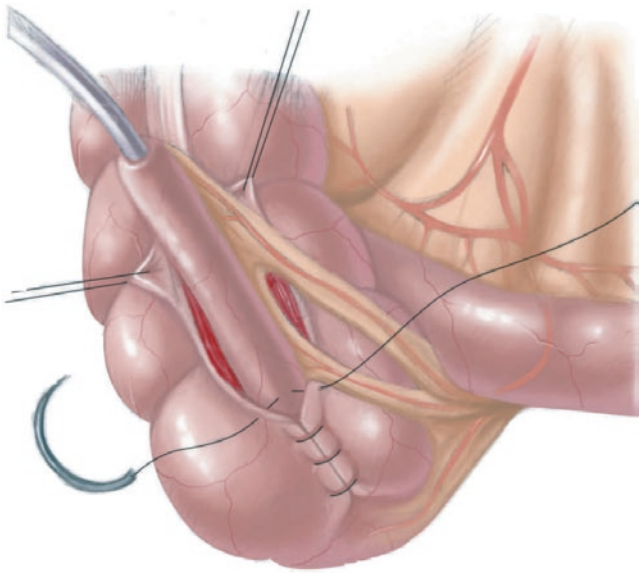

Fig. 72.3 Wrapping the caecum around the folded appendix

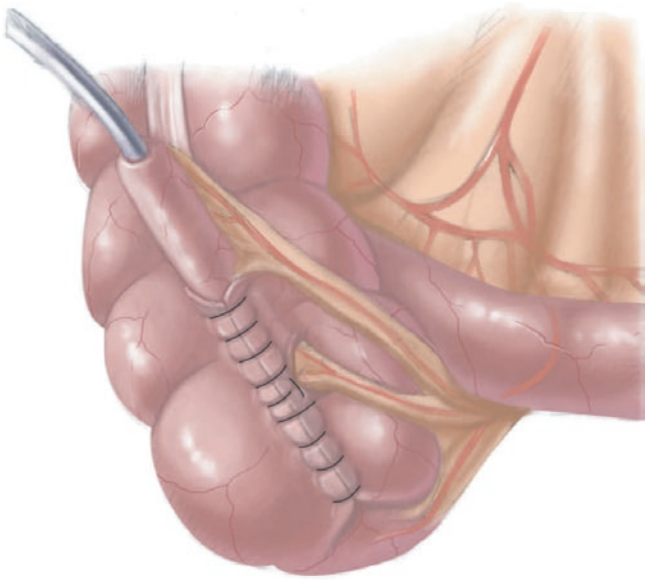

Fig. 72.4 The completed caecal wrap

72.1.2 Monti ACE

The Monti ACE is required when the appendix is absent or required for a Mitrofanoff conduit, or when a left colonic ACE is constructed. A 2-cm segment of ileum is isolated on its vascular pedicle (Fig. 72.5). Straight noncrushing bowel clamps are applied to either end of the isolated segment and the bowel is then divided using a knife. An end-to-end ileal anastomosis is performed using interrupted extramucosal 4/0 absorbable sutures and the mesenteric defect is closed. The bowel is detubularized by opening it along its antimesenteric border in the midline (Figs. 72.6 and 72.7) using scissors or diathermy.

The Monti tube is constructed using a single layer of interrupted extramucosal 6/0 monofilament absorbable sutures over a 12 Ch catheter (Fig. 72.8). Initially it is helpful to place stay sutures at either end and in the middle, in the region of the mesentery, to keep the tube straight and under some tension. Care should be taken to ensure that the lumen of the conduit remains symmetrical throughout to avoid subsequent catheterisation difficulties. It is also helpful not to tubularize the conduit all the way to the end where the stoma is to be constructed, allowing for a defect into which a V flap of skin can be inserted in an attempt to reduce postoperative stomal stenosis.

Following completion of the conduit, two segments are left at either side of the mesentery, one for insertion into the submucosal tunnel in the bowel and the other for bringing through the abdominal wall to the stoma.

The next step is to create the continence mechanism. At least four stay sutures are placed in the colon on either side of a suitably placed taenia. The seromuscular layer is divided using diathermy and the trough created is widened using mosquito artery forceps, until it is sufficiently wide to be able to close it over the Monti tube with no tension. The mucosa is opened at the end of this trough and the conduit is anastomosed to it, end-to-side fashion, using an absorbable suture (5/0) (Fig. 72.9). Following this anastomosis, it is important to check that the conduit remains easy to catheterize. The seromuscular layer is closed over the conduit using a 4/0 absorbable suture, picking up each side of the colon and the conduit to ensure that it does not shift in the tunnel (Fig. 72.10). It is vital to ensure that the vascular pedicle is not compromised during closure of the tunnel. The length of the conduit outside the tunnel should be just long enough to reach the skin of the abdominal wall.

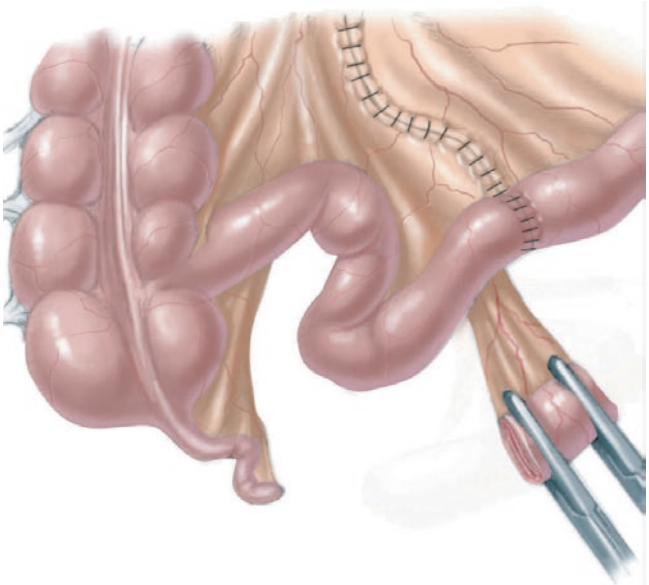

Fig. 72.5 Isolating an ileum segment for Monti ACE

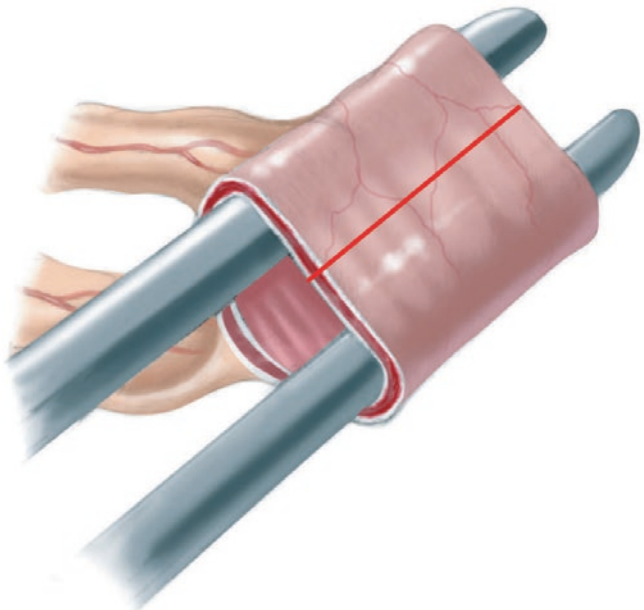

Fig. 72.6 Incision line for detubularizing the ileum segment

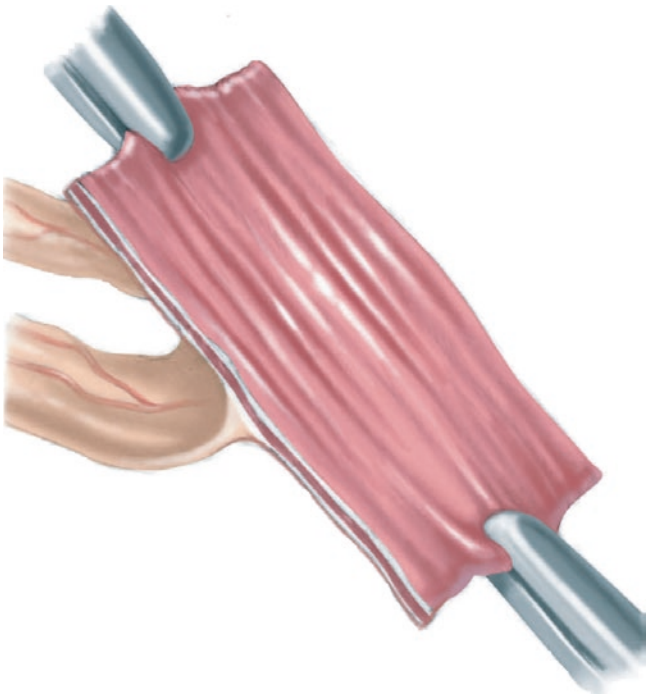

Fig. 72.7 The detubularized segment

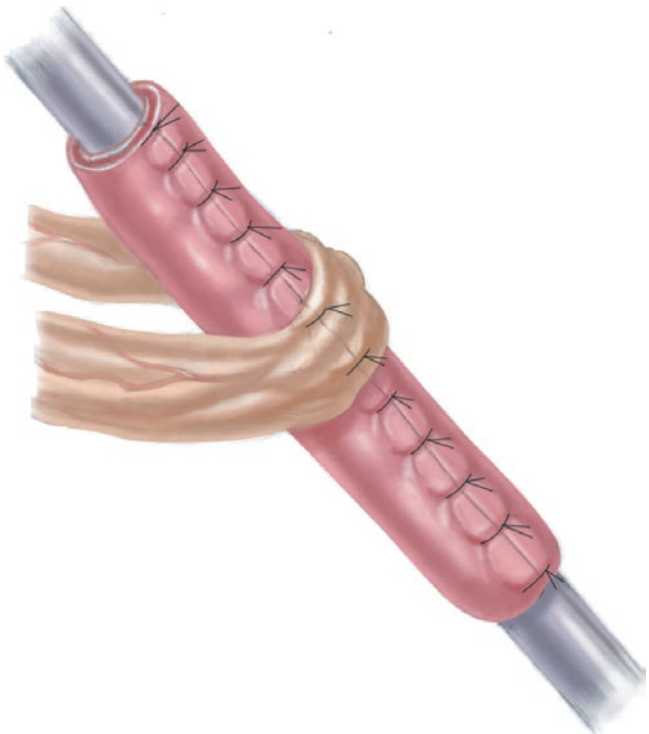

Fig. 72.8 The Monti tube

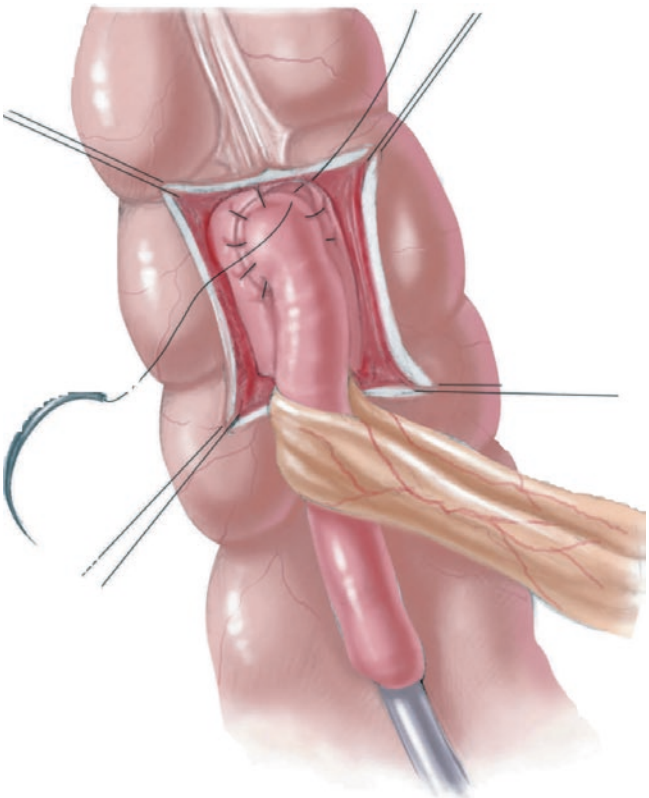

Fig. 72.9 Anastomosing the Monti tube to the colon

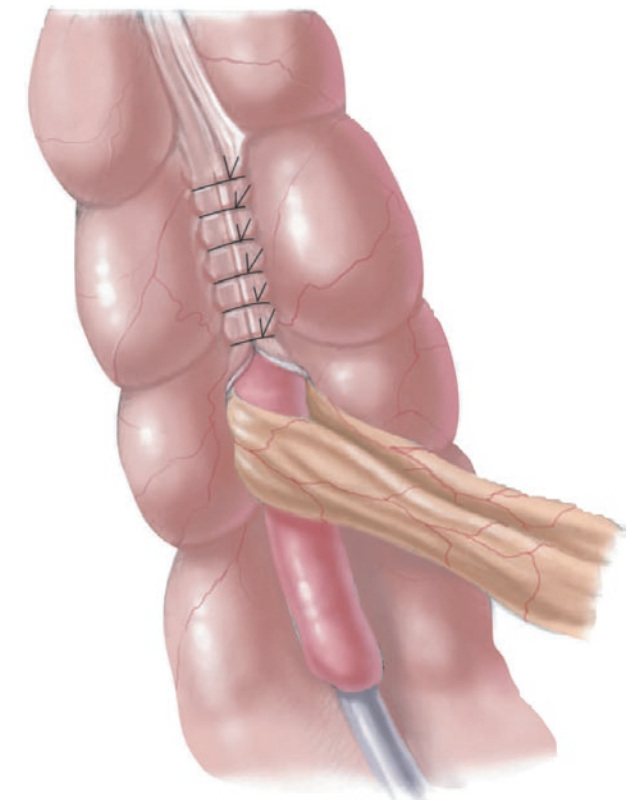

Fig. 72.10 Closure of the tunnel

72.1.3 Stoma Creation

The stoma may be positioned at any convenient place on the abdominal wall, including the umbilicus. Regardless of where the stoma is placed, a V flap of skin should be laid into the spatulated end of the conduit. This chapter illustrates the more complex VQC stoma, because studies have shown that it has a lower risk of developing stomal stenosis.

The skin flaps are initially marked (Fig. 72.11). It is important to ensure that the centre of the V flap directly overlies the fascial defect through which the conduit is brought. It is also important to ensure that the skin and abdominal wall are stretched at this stage using Kocher forceps, because if they are not the channel for the conduit may not be straight when the abdominal wound is closed. The Q flap and V flap are then mobilized (Fig. 72.12).

The V flap is sutured into the spatulated end of the conduit using an absorbable suture. We favour interrupted 5/0 Maxon sutures, as the needle is tapered, atraumatic, and strong; it goes through the skin easily but does not damage and pull through the conduit. The knots are placed on the outside so they will not snag on the catheter as it is passed to test the conduit and stoma during its construction. It is vital to test the ease with which the conduit catheterises after each separate step of its construction. The V flap is sutured until there is enough conduit left to be anastomosed to the edge of the Q flap (Fig. 72.13). The Q flap is rolled over the anterior aspect of the conduit, anastomosing its inferior edge to the V flap while its medial edge is anastomosed to the anterior margin of the conduit (Fig. 72.14). This is best done with a 12–14 Ch catheter in place. The Q flap is anastomosed to the whole of the anterior edge of the conduit, and the superior defect between the Q and V flaps is also closed with interrupted 5/0 absorbable sutures. This then leaves a C-shaped skin defect. This skin defect is usually easy to close without tension by running a simple subcuticular 5/0 absorbable suture on a cutting needle (Fig. 72.15). If there are abdominal scars following previous surgery, it may be necessary to perform a relieving Z-plasty to facilitate tension-free closure.

The catheter is left in place for 4 weeks prior to the commencement of intermittent catheterization, but washouts can commence as soon as the patient has recovered from the postoperative ileus, usually on day 5.

72.1.4 Laparoscopy-Assisted MACE Creation

The steps for the laparoscopic approach are essentially the same as for the open approach with or without caecoplication. When caecoplication is performed, the appendix is kept in situ and is retroverted. The colonic serosa is then imbricated around it in an antirefluxing fashion.

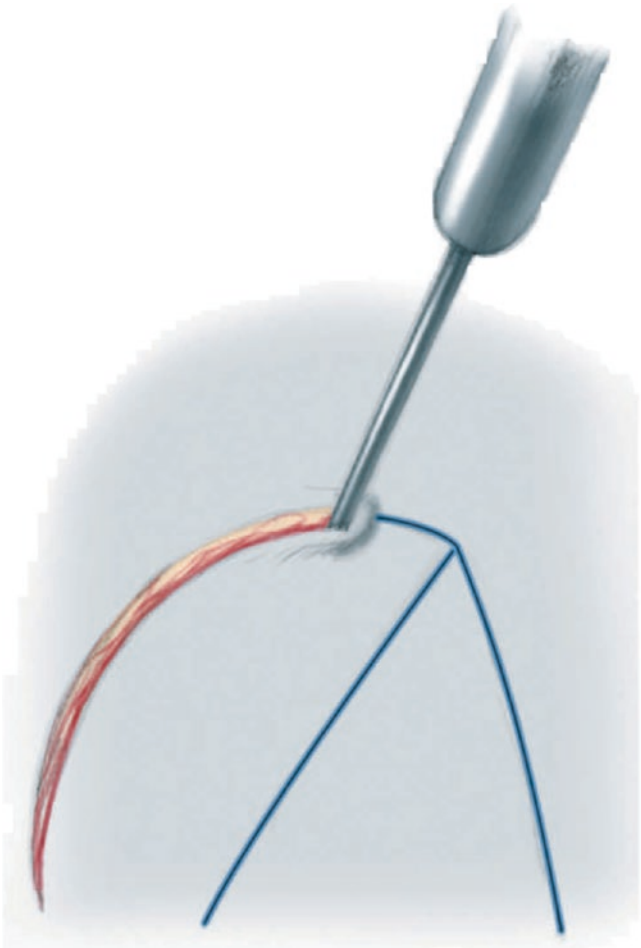

Fig. 72.11 Skin flaps marked to create the stoma

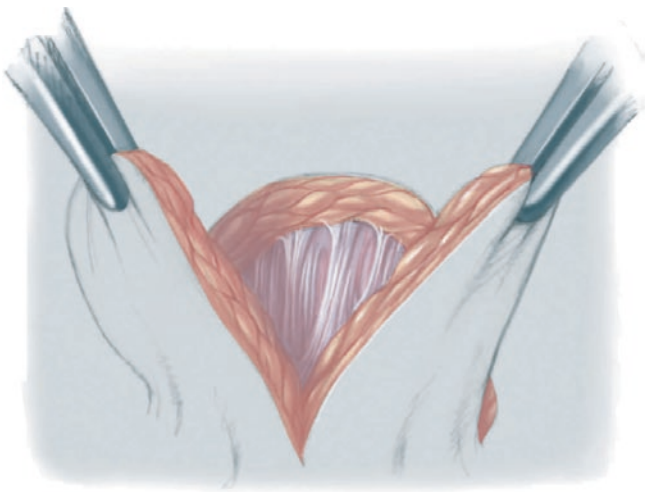

Fig. 72.12 Mobilizing the Q and V flaps

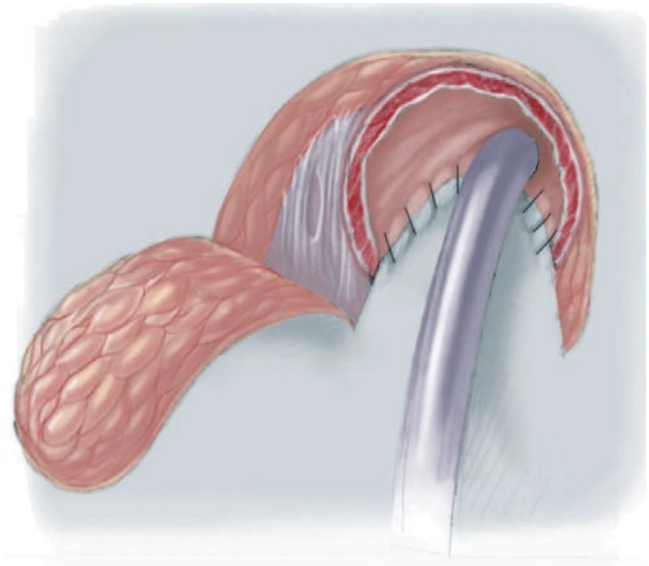

Fig. 72.13 The sutured V flap with the test catheter

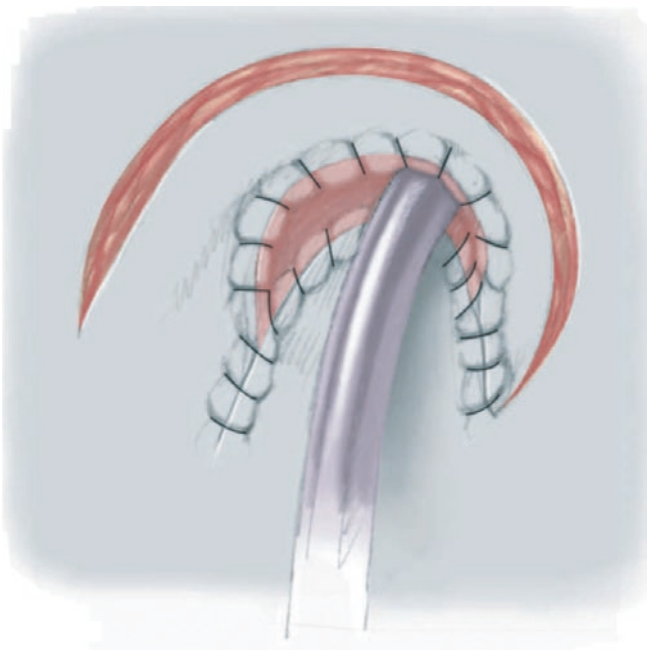

Fig. 72.14 Attaching the Q flap to the V flap and conduit

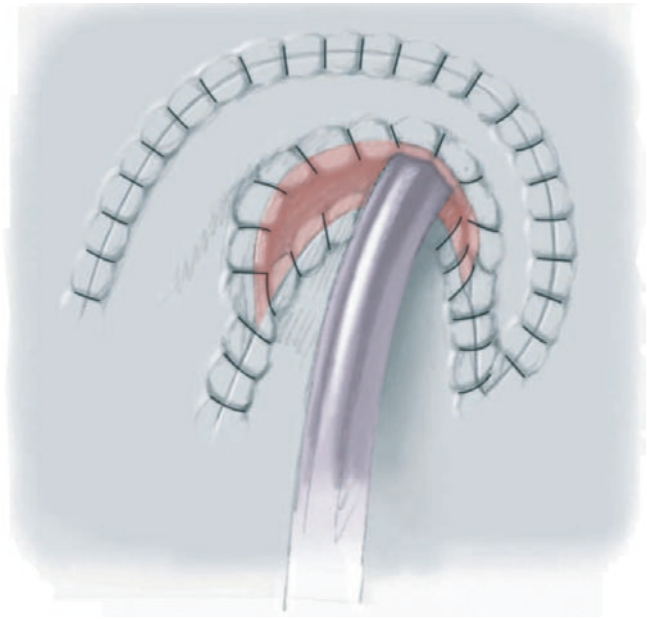

Fig. 72.15 Closure of the skin defect

72.2 Results

All conservative measures should be tried first before resorting to the ACE procedure. The underlying diagnosis is important, as it influences the success rate. In the Southampton experience, patients with a neuropathic bowel or an anorectal malformation had a success rate of 73%, compared with 38% for patients with chronic idiopathic constipation. The age at operation is also important. In Southampton, there was a 70% failure rate for patients under 5 years of age, compared with 24% for those over 5. This difference was independent of the underlying diagnosis and probably reflected the inability of a child under 5 years of age to sit on a toilet for up to 1 h before emptying is complete.

The commonest operative complication encountered is stomal stenosis, which occurs in up to 30% of cases. Half of these patients require revisional surgery. The VQC stoma described in this chapter has considerably reduced the incidence of stenosis. The site of the stoma and the type of conduit used makes no difference to the stenosis rate.

72.3 Establishing a Washout Regimen

Patient and caregiver motivation is vital in determining success. A lack of compliance with the washout regimen was a major contributory factor to failure in a number of series. Detailed preoperative counselling and continued postoperative support, ideally provided by a nurse specialist, are essential to ensure adequate and continued motivation, without which the ACE is doomed to failure. It is an advantage to introduce the potential patient to a child and family with a functioning ACE prior to the surgical procedure. The initial therapeutic trial that has recently been introduced by the use of the percutaneously placed tube is a significant step forward, as it enables the surgeon to test the family's motivation and their ability to cope with the regimen before proceeding to the definitive procedure. It also provides the opportunity to test a number of washout regimens to see which one is most suitable.

The washout regimen is usually established by trial and error, and it can take up to 6 months before a stable situation is reached. Many different washout regimens are in use around the world. In Southampton, a phosphate enema is generally used. Initially 50 mL of enema solution is diluted up to 100 mL with water and rapidly instilled, followed by approximately 500 mL of water. (In the original description, saline was used, but this is not necessary; it is safe just to use tap water.) The regimen is adjusted depending on the response, increasing or reducing the volume of phosphate and/or water until a stable situation is reached. Care must be taken when using the phosphate, as toxicity can occur if the

enema is retained. Some units do not use a stimulant of any kind, simply relying on large volumes of water for the washout.

Several problems have been encountered during the establishment of the ACE. The most common is pain during the washout, which is reported in up to 60% of patients. This usually settles spontaneously during the first 3 months but can be helped by reducing the concentration of the phosphate, reducing the rate of the infusion, or using an antispasmodic prior to the enema (such as Colofac, Solvary, UK). Despite regular washouts, patients may still become constipated, which also produces pain and should always be excluded. One of the other problems encountered is the time taken for the enema to pass and achieve a result; this has been a significant contributory factor to failure. The placement of the conduit in the distal descending colon has improved this situation considerably. Patients in whom the standard caecal ACE did not work have achieved excellent results with the left colonic ACE.

Conclusions

Despite all the problems associated with the ACE, there is little doubt that it can achieve social continence and avoid a colostomy in patients with intractable incontinence, a situation that did not exist prior to the development of the ACE. It has also been shown to significantly improve the patient's quality of life.

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Hydrometrocolpos

73

Devendra K. Gupta and Shilpa Sharma

Hydrometrocolpos is a condition in which the uterus and vagina are grossly distended with retained fluid other than blood, usually in the presence of distal vaginal obstruction. Diagnosis and treatment are now more efficient, as the condition is being diagnosed more often prenatally with the use of ultrasound and fetal MRI.

Hydrometrocolpos presents at the two extremes of childhood: initially during the neonatal period, when there is a high level of maternal hormones, and then at early puberty, when the patient herself begins to have production of estrogenic hormones. The distal vaginal obstruction is mostly due to imperforate hymen (in two thirds of cases), followed by a transverse vaginal septum and less commonly, vaginal atresia (with or without persistence of a urogenital sinus or cloaca).

Associated anomalies are common and quite often severe. These include anorectal malformations and unilateral or bilateral agenesis of the kidneys, ureters, and trigone. Ultrasonography, micturating cystourethrography (MCU), and MR urography can help to evaluate the associated genitourinary anomalies.

73.1 Classification

Hydrometrocolpos has been classified into five types (Fig. 73.1) on the basis of the type and level of obstruction:

1. Low hymenal obstruction.
2. Mid-plane transverse membrane or septum.
   (a) Without communication.
   (b) With a small orifice as communication.
3. High obstruction with distal vaginal atresia.
   (a) Without perineal swelling.
   (b) With perineal swelling.
4. Vaginal atresia with persistence of the urogenital sinus.
5. Vaginal atresia with cloacal anomaly.

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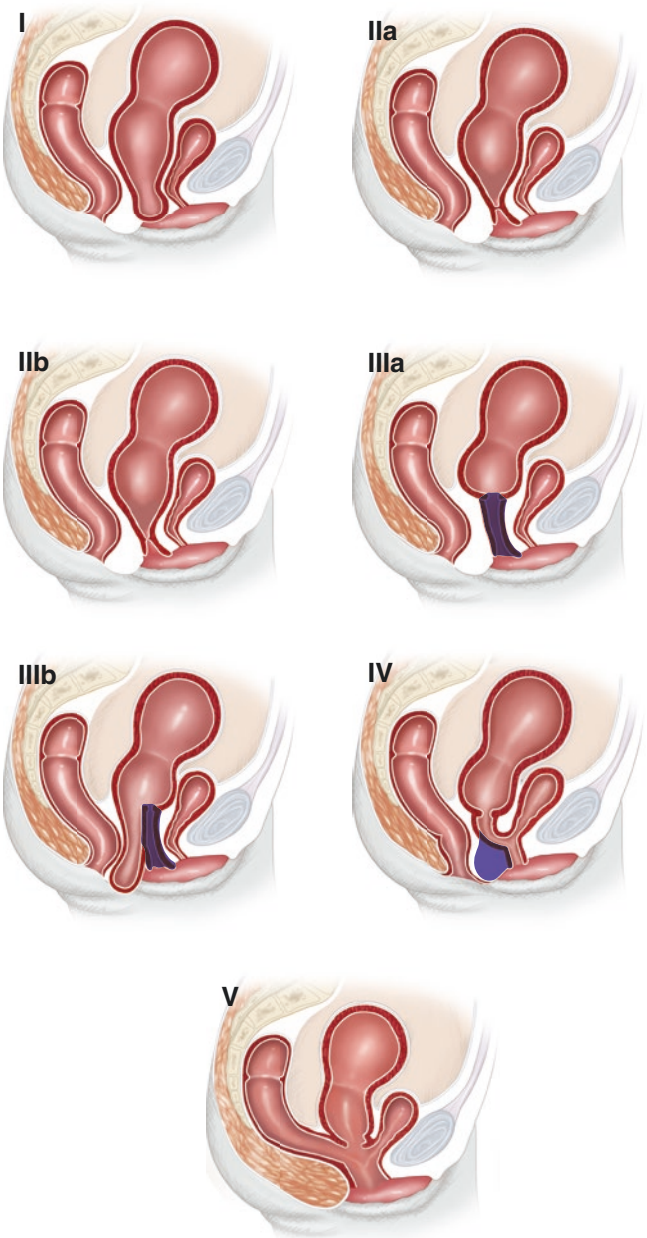

Fig. 73.1 Classification of the types of hydrometrocolpos

73.2 Treatment Principles and Initial Approach

The main treatment of hydrometrocolpos is surgical. Medical management is required to build the baby and make her fit for the surgery. The operative procedure and timing of surgical intervention depends upon the severity of the condition, the type of anomaly, and the age at presentation. Early surgery in the neonatal period is indicated when a grossly distended hydrometrocolpos presents with a bulging hymen, quite often associated with complications such as abdominal mass, urinary obstruction, constipation, sepsis, and dehydration. Laparotomy is indicated in patients with high vaginal obstruction, abdominal complications, or associated anomalies.

The baby will need resuscitation and stabilization during the acute stage, with antibiotics and intravenous fluids. A temporizing procedure like aspiration of the turbid, infected fluid (or an abdominal tube or a flap vaginostomy) is indicated in the neonatal period to drain the infected material from the vagina. A percutaneous nephrostomy may also be needed in patients presenting with severe bilateral hydroureteronephrosis leading to urinary obstruction, uraemia, and sepsis.

Figure 73.2 suggests an algorithm for management of hydrometrocolpos [1]. Newborns presenting with complications should be managed in the intensive care unit with systemic antibiotics, intravenous fluids, decompression of the gastrointestinal tract by nasogastric aspiration, and administration of oxygen. Fluid and electrolyte imbalances should be corrected. A Foley catheter should be inserted in the bladder for urinary drainage. In the presence of a huge, distended hydrometrocolpos in a sick neonate, preliminary drainage by puncturing the vagina under ultrasonographic guidance may be done for 24–48 h prior to corrective surgery. Alternatively, the vaginal septum (type II anomaly) can be incised safely under ultrasonographic guidance or under vision in experienced hands.

The management options depend upon the type of obstruction. The management is simple with low Type I and II anomalies, but patients with the Type III, IV, and V anomalies are usually obstructed and infected. Moreover, type IV anomaly has a valvular opening between the urethra and the vagina, allowing urine flow to the vagina during the act of micturition and vice versa. In such cases, drainage of the infected material collected in the vagina should be attempted by a suprapubic route (catheter or a flap vaginostomy as a first-stage procedure) and allowing the dilated vagina to shrink. A definitive procedure (vaginal pull-through) follows later, as a second-stage procedure. Earlier attempts to drain the vagina and simultaneously reconstruct the vaginal tract in newborns carried a very high mortality and is thus not recommended.

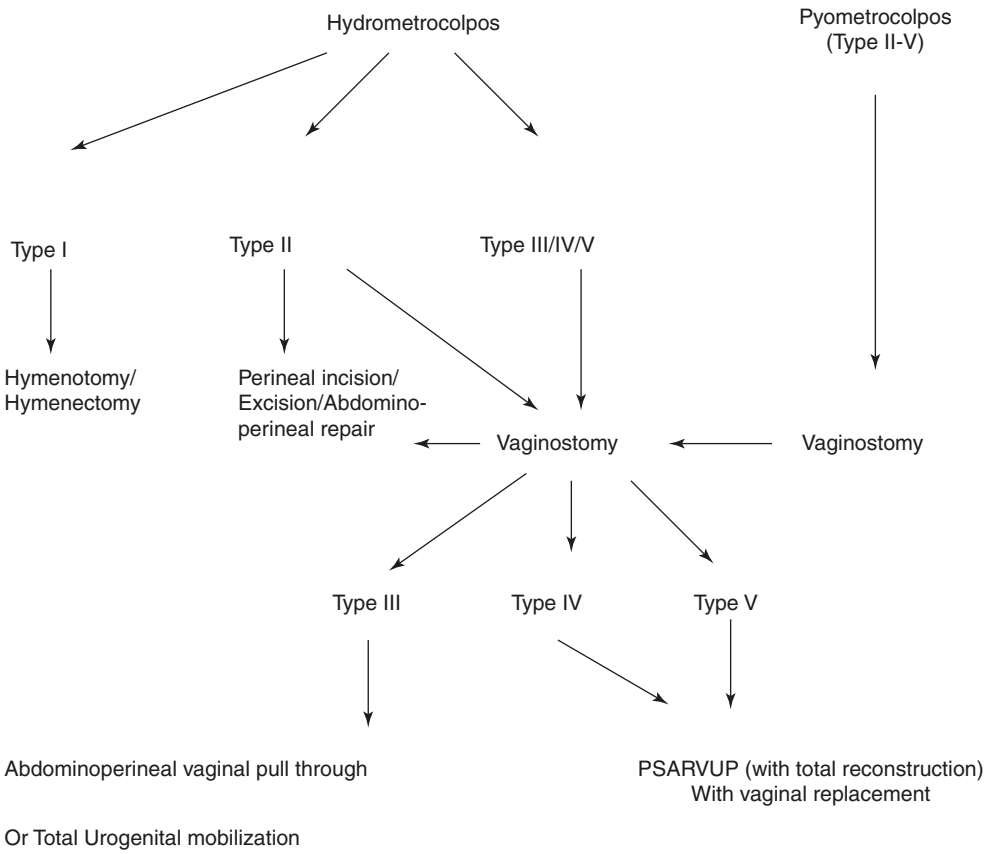

Fig. 73.2 Algorithm for the management of hydrometrocolpos. PSARVUP—posterior sagittal anorectovagino-urethroplasty

73.3 Drainage Procedures

The advantages of early drainage in neonates include drainage of the infected material to reduce sepsis and disconnection of the communication and retrograde flow of urine to the vagina. The inflamed vagina and uterus are then allowed to shrink to near-normal anatomical positions and size, assisting the planning of definitive surgery. In the prepubertal age, drainage allows natural passage for menstrual flow and creation of a passage for future sexual activity and fertility. The drainage procedure through the perineum may be the only definitive treatment required in Type I and II anomalies.

73.3.1 Hymenotomy and Hymenectomy

A bulging membrane in an infant with imperforate hymen or transverse septum of the vagina may be incised without anesthesia, but excision is preferable if the hymen is thickened or the patient is an adolescent. Hymenotomy (Fig. 73.3) may resolve the acute renal failure caused by hydrometrocolpos. In all cases, it is desirable to maintain the patency of the opening by the initial use of a drain, followed by repeated dilatations.

In the lithotomy position, the bulging hymen becomes visible as a grayish membrane. If necessary, the abdomen may be compressed to make the hymenal bulge more prominent. Before hymenectomy, a Foley catheter is inserted into the urinary bladder to decompress it, as well as to identify the urethra during the surgery (Fig. 73.4). A series of stay sutures is placed at the center of the hymen, and the vaginal fluid is aspirated and sent for microscopic examination and culture. A circular hymenal segment is excised using a no. 11 blade. The cut margin is oversewn and retracted with vertical mattress sutures. The sutures are tied, exposing the vaginal cavity. A soft Silastic catheter is inserted into the vagina. A contrast study is performed to delineate the internal anatomy. The vagina is drained for a couple of days. This procedure is simple and can be performed at the bedside or in the intensive care unit in a sick baby after assessing the depth with a needle puncture and ultrasonography. If there is doubt, the procedure may be done safely in the operating room through the abdominal route, after performing a dye study. A Hegar dilator or the finger is placed through the vaginostomy opening as a guide, to remove a disc of the septum, which may be thick. The edges are then oversewn and a drain is placed. Antibiotics should be given for 5–7 days postoperatively.

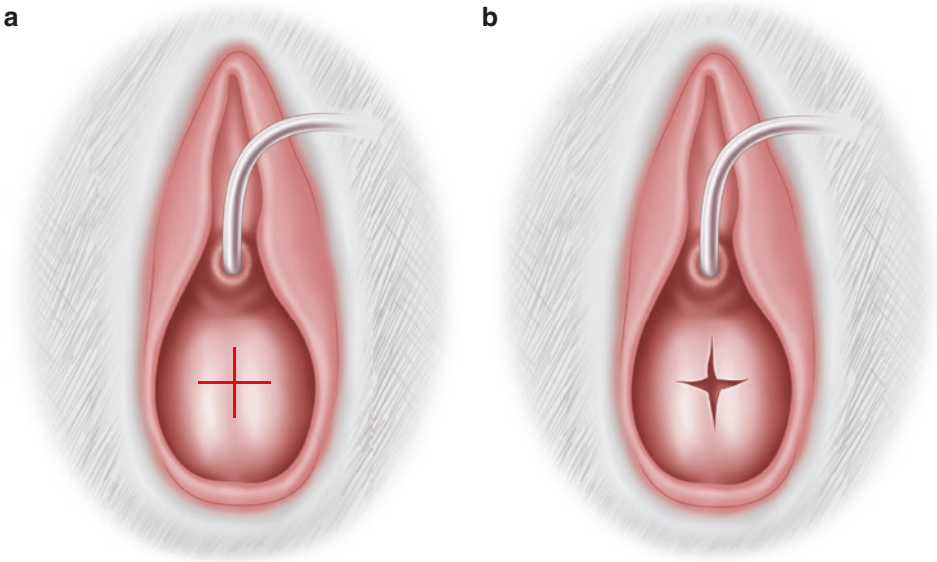

Fig. 73.3 (**a** and **b**) Hymenotomy

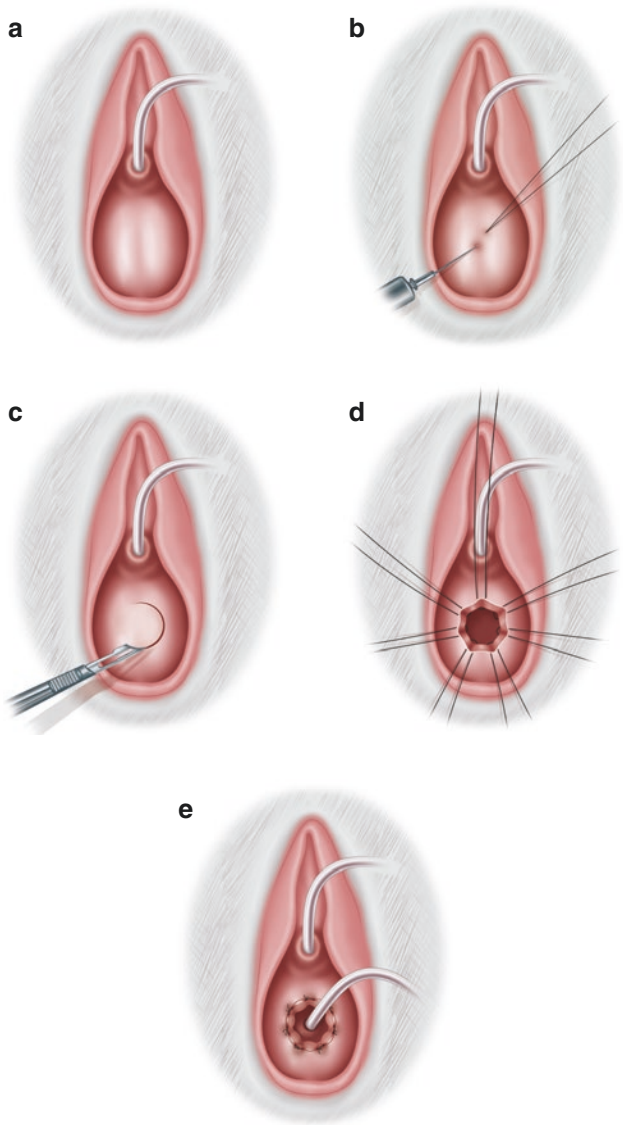

Fig. 73.4 Hymenectomy (with excision of the membrane disc). (**a**) Foley catheter. (**b**) Stay sutures and aspiration of vaginal fluid. (**c**) Circular hymenal segment. (**d**) Vertical mattress sutures in the cut margin. (**e**) Tied sutures and insertion of a catheter

73.3.2 Perineal Vaginostomy

A vaginostomy (Fig. 73.5) serves as a temporizing drainage procedure for cases with infected fluid. It may be done through the perineal route in Type II after confirmation of the diagnosis with a needle aspiration. When a low transverse vaginal septum is present (between the lower one third and upper two thirds of the vagina), it presents as a bulging membrane, which can be excised by the perineal route to permit drainage. In some patients, a minute vaginal orifice may be visible that may be surgically enlarged and oversewn, with the placement of a catheter in the vagina to establish drainage.

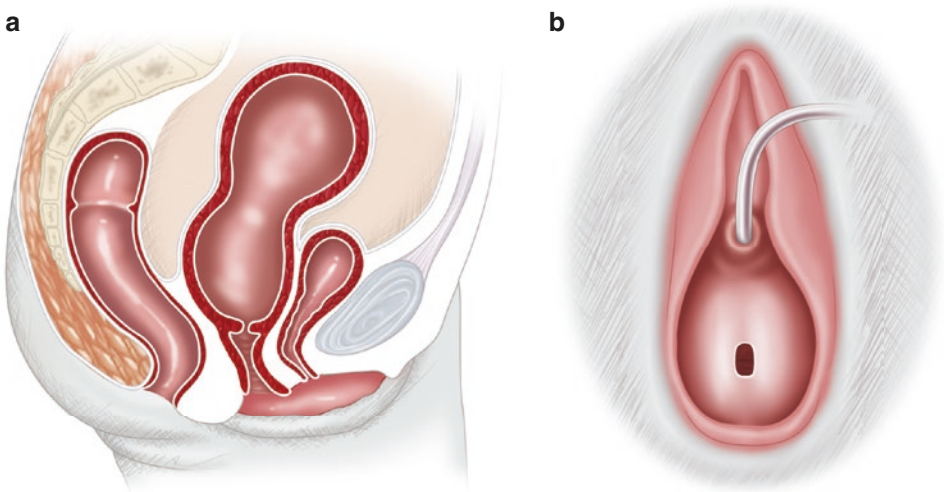

Fig. 73.5 Perineal vaginostomy. (**a**) Low transverse vaginal septum. (**b**) A minute vaginal orifice

73.3.3 Abdominoperineal Repair

Even in babies with a Type II anomaly, a perineal procedure sometimes should be deferred, as suprapubic drainage by catheter or a vaginostomy is necessary because of secondary infection or complex anomalies. The proximal diversion stoma not only helps in decompressing the vagina but also provides a portal for detailed radiographic studies to delineate the anatomy. If the transverse septum in Type II cases is more than 1 cm thick, it is wiser to perform a laparotomy and define the proper anatomy, incising the septum precisely under direct vision and then draining the vagina. This technique is used to prevent injury to the urethra and rectum during the perineal dissection.

Figure 73.6 illustrates abdominoperineal repair of Type II hydrometrocolpos. A low transverse abdominal incision is made, the hydrometrocolpos is delivered from the abdominal wound and a purse-string suture is applied on the vaginal wall. Fluid is drained out; a Kelly's forceps is then introduced through the vaginostomy opening and the tip is advanced towards the most dependent part of the dilated vagina in the perineum. The vaginal septum is then incised, while the outer vaginal orifice is spread open by a nasal speculum and the transverse septum is pushed downwards by the Kelly's forceps. The septum is incised and the Kelly's forceps is pushed down and out. A Silastic catheter drains the vagina from below. The hydrometrocolpos is also drained by an indwelling catheter from above for 5–7 days.

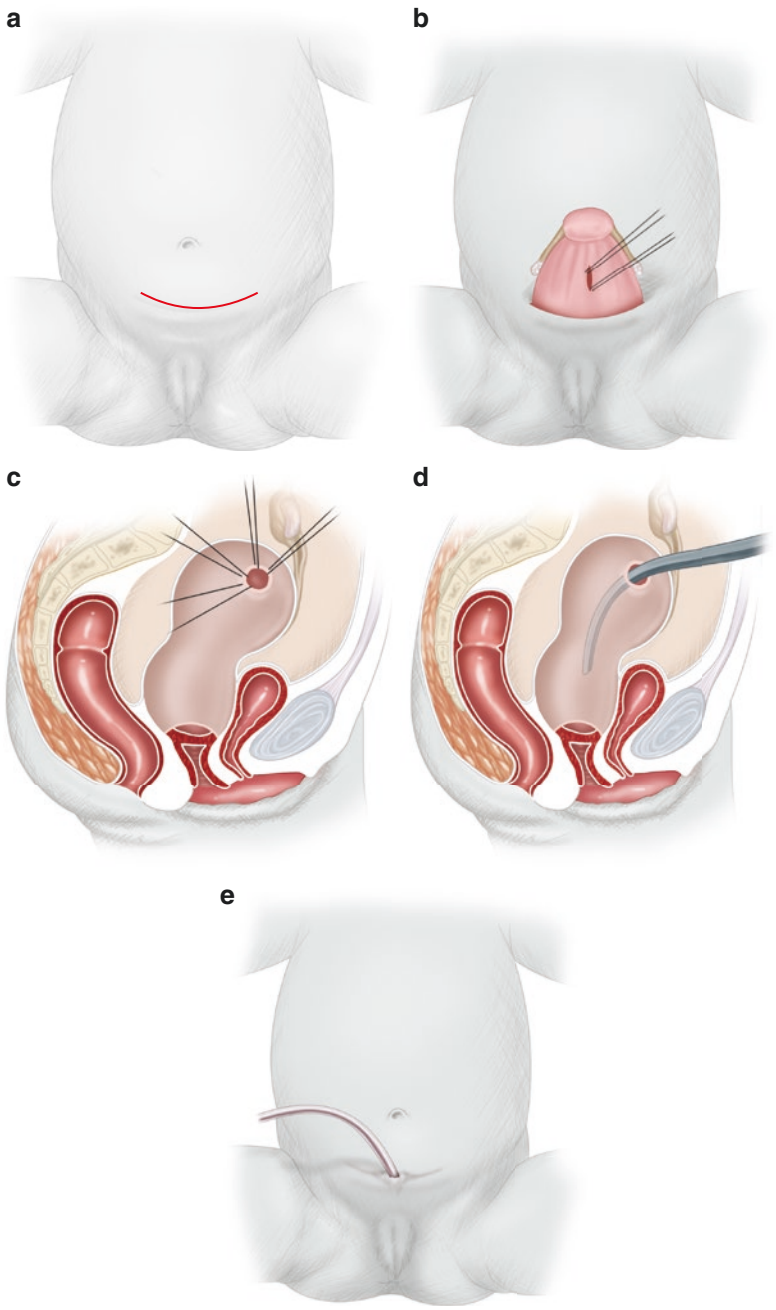

Fig. 73.6 Abdominoperineal repair of Type II hydrometrocolpos. (**a**) Low, transverse abdominal incision. (**b**) Purse-string suture applied on vaginal wall. (**c**) Incising the vaginal wall. (**d**) Catheter draining the vagina from below. (**e**) Indwelling catheter for drainage from above

73.3.4 Abdominal Wall Vaginostomy

For Types III, IV, or V hydrometrocolpos with atresia of the lower two thirds of the vagina or common cloaca with hydrometrocolpos, an abdominal route is preferred for the vaginostomy. If the atretic lower portion of the vagina has been retracted up into the pelvis, it may be desirable to open the vagina through a laparotomy incision to avoid damage to the urethra, bladder, and rectum.

Abdominal vaginostomy may be of two types: vaginostomy with an indwelling catheter and tubed vaginostomy. For both types, the abdomen is opened with a Pfannenstiel incision, the dilated vagina is identified, and stay sutures are applied. The initial steps are the same as for the abdominoperineal repair just described, through the incision of the vaginal septum.

Vaginostomy with an indwelling catheter (*see* Fig. 73.6) is easy to perform, but it has certain disadvantages like infection or encrustation. Another disadvantage is the need to keep the tube in situ; the requirement for frequent changes in an inconvenience to the patient.

In the tubed vaginostomy procedure (Fig. 73.7), after the initial steps, a U-shaped flap of the vagina is made to shape a tube to provide drainage until the time of definitive surgery. A U-shaped incision creates a flap that is made into a tube, and the edge of the tube is fixed to the abdominal wall and the skin. A tubed vaginostomy provides effective drainage while avoiding the long-term use of an indwelling catheter. It also provides easy access for the dye studies performed to outline the anatomy prior to definitive surgery.

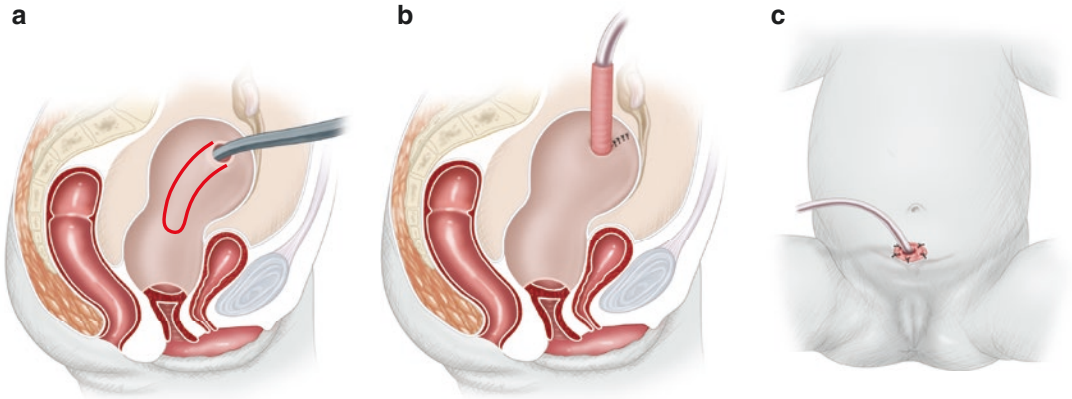

Fig. 73.7 Tubed Abdominal wall vaginostomy. (**a**) U-shaped incision. (**b**) Creation of the tube. (**c**) Fixation of the tube to the abdominal wall and skin

73.3.5 Transverse Colostomy for Common Cloaca

Type V hydrometrocolpos associated with common cloaca and a common channel more than 3 cm long requires a vaginostomy and also a diverting colostomy (Fig. 73.8). This procedure is done keeping a sufficiently long segment of sigmoid bowel available for both vaginal replacement and bowel pull-through in the future. Thus a transverse colostomy is preferred.

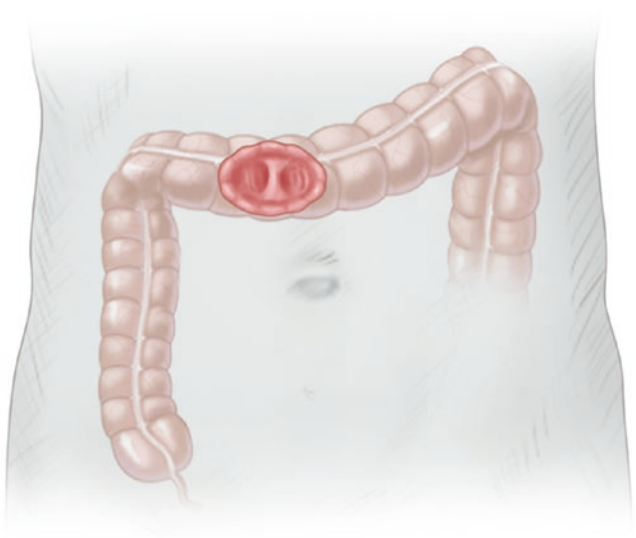

Fig. 73.8 Location of transverse colostomy for common cloaca

73.3.6 Total Urogenital Mobilization

In Type IV hydrometrocolpos, if the common channel of the urogenital sinus is less than 2.5 cm long (as confirmed by endoscopy at surgery), the fistula is disconnected, followed by total urogenital mobilization (TUM), in which the vagina may be exteriorized onto the perineum and the urogenital sinus can be made to function as the main urethra (Fig. 73.9). With the patient in lithotomy position, stay sutures are applied around the urogenital sinus so that the sutures lie within the line of incision. An incision is made all around the urogenital sinus, with a midline posterior incision in the sinus that extends posterolaterally into the perineum for adequate exposure and mobilization. As the meticulous dissection continues, the urethra and the vagina become visible externally as two separate openings, and the tissues are approximated. If the vaginal introitus is narrow, it can be widened by placing a Barrow's skin pedicle flap in its posterior wall (*see below*).

At times, the vagina may open into the bladder as a fistulous communication. If the length of the urogenital sinus is more than 2.5 cm, vaginal replacement will be required. The fistulous communication between the urethra and the vagina is divided and suture repaired, and the vagina is brought to the perineum by using the posterior sagittal route, bisecting the rectum to approach the vagina directly from behind (posterior sagittal anorectovagino-urethroplasty, PSARVUP). This procedure is a major undertaking; achievement of the best results requires that it be performed by a team of the experienced surgeons, preferably in a tertiary care center.

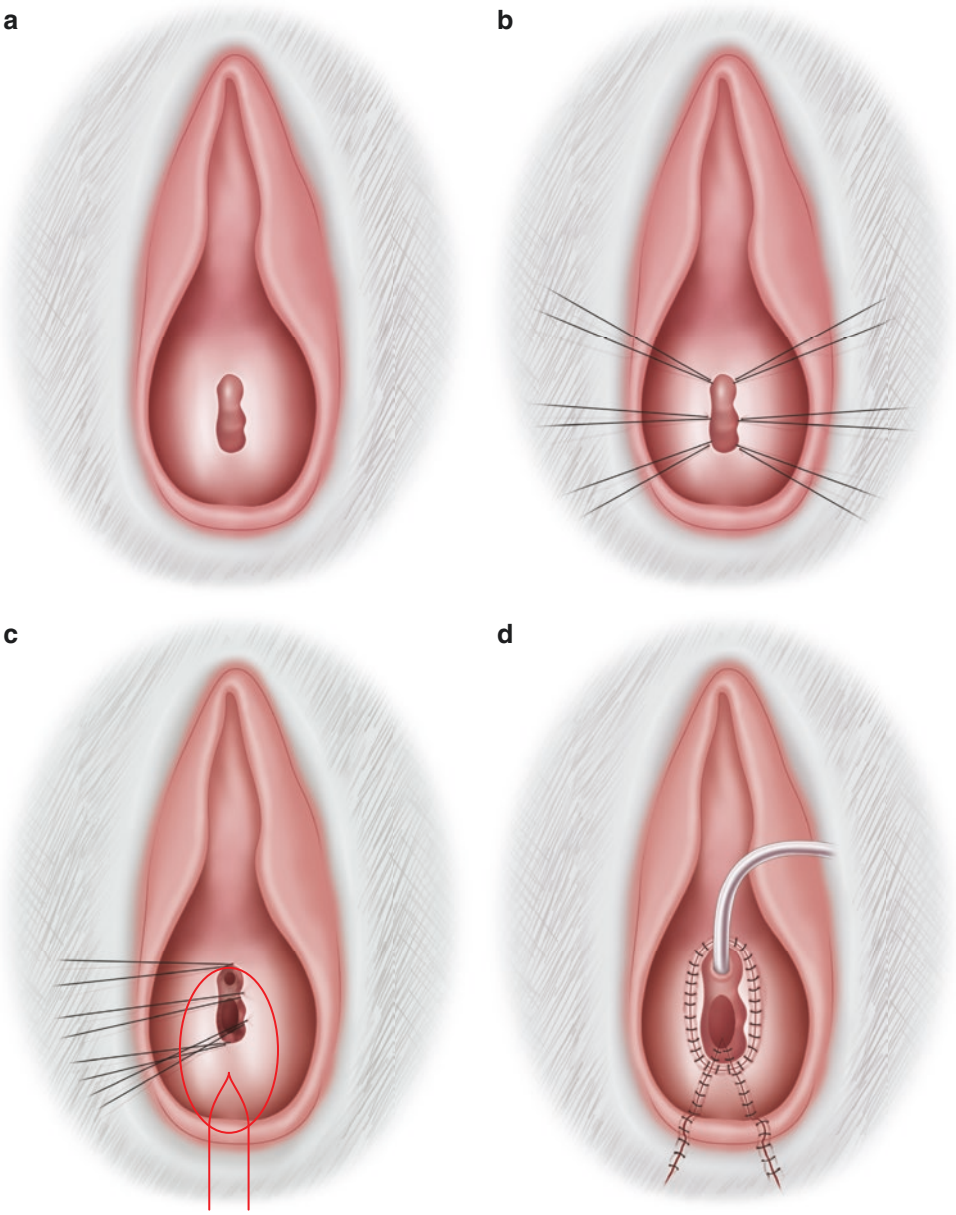

Fig. 73.9 Total urogenital mobilization (TUM). (**a**) Exterior view of the urogenital sinus. (**b**) Application of stay sutures. (**c**) Incision around the urogenital sinus, with a midline posterior incision extending posterolaterally in the perineum. (**d**) Separate openings revealed by dissection; placement of a skin flap to widen the vaginal introitus

73.3.7 Vaginal Replacement with Bowel Segment

The common channel in Type V hydrometrocolpos is usually longer than 3 cm, mandating a vaginal replacement. In Types III, IV, and V with pyometrocolpos, it may be difficult to separate the vagina from the surrounding structures because of severe inflammation and dense adhesions. In such situations, it would be better to replace the vagina using bowel vaginoplasty, using a loop of sigmoid colon or ileum (Fig. 73.10). An abdominoperineal approach is adopted. The abdomen is opened with a Pfannenstiel incision with extension on the left side in a hockey stick manner. The vaginal space between the rectum and the urethra is carefully created in the perineum and also from the abdominal side. The length of the bowel required for vaginal replacement is assessed. An adequate length of the colonic bowel segment on its mesentery with adequate blood supply is selected for the pull-through. The upper end is anastomosed at the proximal end of the vagina from the abdominal side and the distal end of the bowel is anastomosed to the perineal skin at the proposed vaginal introitus, taking care not to cause any torsion to the bowel segment or a traction on the blood supply.

73.3.8 Barrow's Flap Vaginoplasty

If the distal vagina is narrow or if the vagina has retracted following its repair, flaps of perineal skin may be used to contribute to the distal vaginal segment. A Barrow's flap is the most commonly used procedure (Fig. 73.11). An inverted Y incision is performed, with the vertical limb of the Y going inside the vaginal introitus for a centimeter or so in the posterior wall. A V-shaped flap of perineal skin with intact blood supply is created and mobilized sufficiently. It is then advanced into the vagina and sutured to the margins of the incision edges in the introitus.

Many surgeons prefer the use of a skin graft over a vaginal mold, a cylinder of a prosthetic patch, or a buccal mucosa graft with a mesh to form a neovagina. The results are variable, with retractions and graft contraction being not uncommon. Expertise is needed to perform such a surgery. This kind of replacement is better performed at puberty or before marriage, as the replacements are prone to shrinkage and need repeated dilatations.

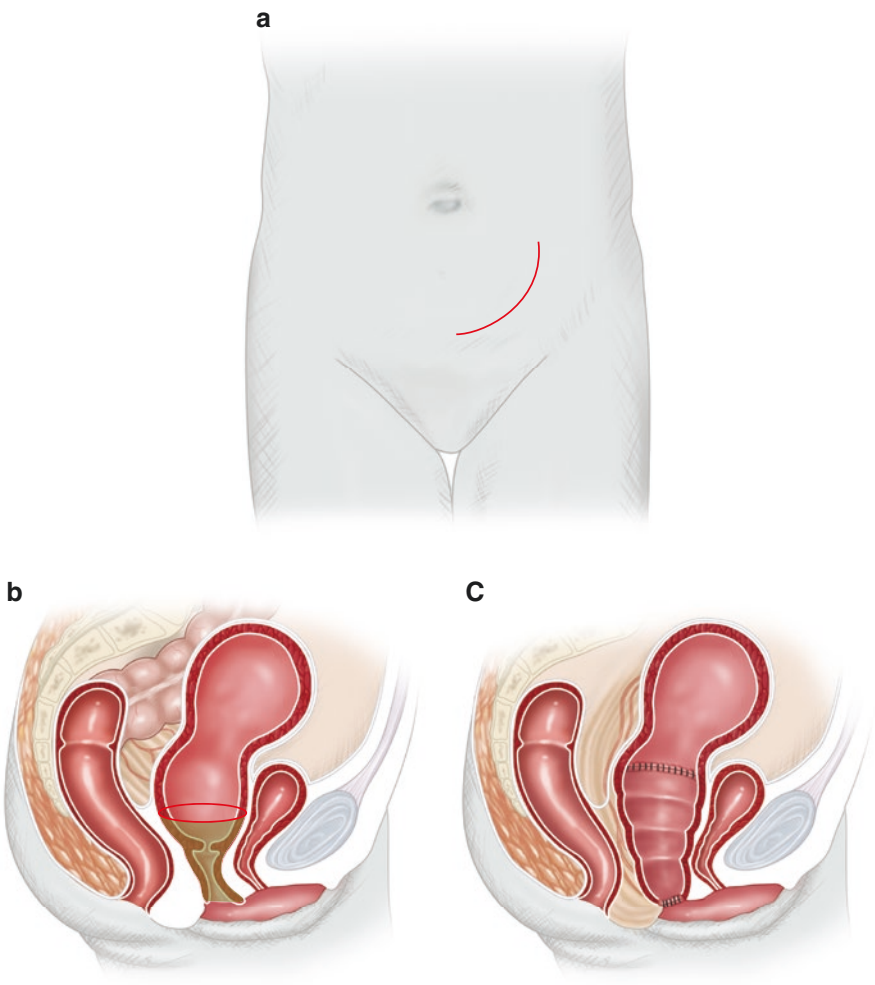

Fig. 73.10 Bowel vaginoplasty. (**a**) Pfannenstiel incision with extension on the left side. (**b**) Resection of the atretic part of vagina. (**c**) Anastomosis of the upper end of the bowel segment to the vagina and the distal end of the bowel to the perineum

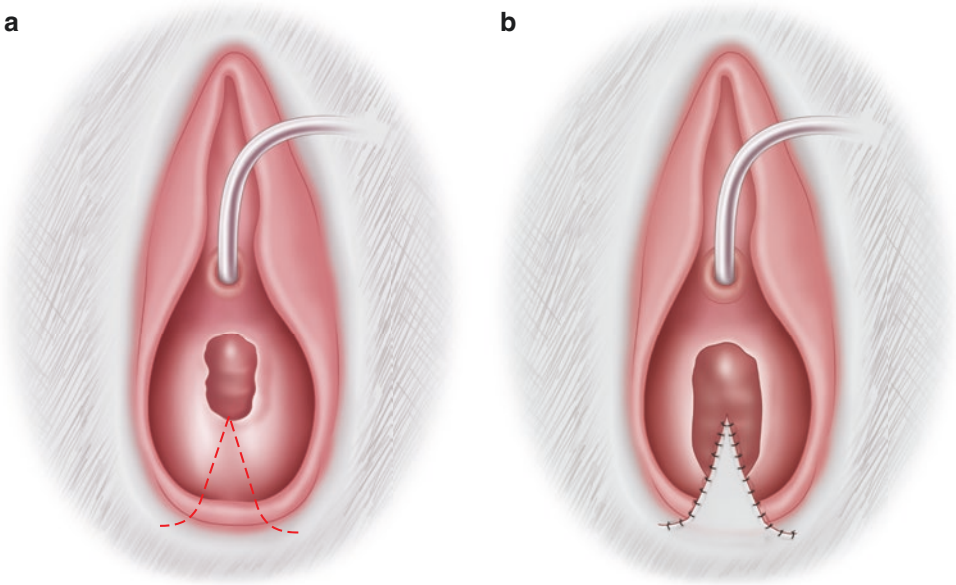

Fig. 73.11 Barrow's flap vaginoplasty. (**a**) Inverted Y incision entering the posterior wall of the vagina. (**b**) V-shaped flap of perineal skin sutured to the incision edges in the introitus

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Venous and Peritoneal Access in Renal Failure

74

Marcus D. Jarboe and Ronald B. Hirschl

There are numerous options for chronic dialysis, including peritoneal dialysis (PD), intravenous catheter-based dialysis, and arteriovenous fistulas (AVFs). The choice of dialysis depends on many factors such as patient age, social situation, opportunities for kidney transplantation, and options for venous access. With the promotion of the Fistula First National Vascular Access Improvement Initiative (www.fistulafirst.org), however, there have been increased efforts to create AVFs as the primary hemodialysis access in children.

The management of access for dialysis begins from the moment that renal insufficiency is recognized. In many cases, access for acute dialysis is required early in a hospital course, whereas in others the progression of renal failure is insidious, with the requirement for dialysis identified well in advance of its need. Procedures for such access should be performed with the recognition that peripheral venous fistula formation may be required in the future. Our protocol calls for immediate bilateral upper extremity venous evaluation in order to identify the preferred arm for a future fistula. Central venous access for acute dialysis is preferably performed in the upper extremity least favorable for fistula formation, and almost always uses the internal jugular vein rather than the subclavian vein. Intravenous catheters and peripheral intravenous central catheters (PICCs) are avoided in veins that are candidates for fistula formation, such as the cephalic and basilic veins.

74.1 Peritoneal Dialysis

74.1.1 Preoperative Planning

Age is not a limiting factor for peritoneal dialysis (PD); in fact, we have successfully placed catheters in patients as young as a few days old. The presence of sufficient peritoneal surface area for dialysis is a factor that determines success, however. Adequate dialysis may be difficult to achieve in patients with numerous adhesions due to prior abdominal procedures. In patients of adequate size and in whom adhesions do not present too much risk, a laparoscopic approach may be entertained. Plans should be made to excise the omentum, because it is frequently associated with catheter occlusion. Studies have demonstrated that catheter infections are reduced when a double-cuff catheter is used and when the catheter exits from the abdomen in a downward direction. Therefore, swan neck catheters of the following sizes should be used:

- *Infant 5–10 kg:* Infant Double Cuff Swan Neck Coiled Catheter (38.9 cm) (Covidien, Mansfield, MA).
- *Pediatric patient 10–30 kg:* Pediatric Double Cuff Swan Neck Coiled Catheter (42 cm).
- *Pediatric patient 30–45 kg:* Small Adult Double Cuff Swan Neck Coiled Catheter (57 cm).
- *Pediatric patient > 45 kg:* Adult Double Cuff Swan Neck Coiled Catheter (62.2 cm).

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74.1.2 Open Peritoneal Dialysis Catheter Placement

From an infectious standpoint, the catheter should be positioned so that both the intraperitoneal end of the catheter and the exit site are facing downward or caudal (Fig. 74.1a). (In infants, the exit may be lateral.) To determine the exact locations of the incisions, the catheter should be placed on the abdomen and the positions of the internal cuff, tunnel cuff, and exit site should be marked on the skin with the catheter lying naturally. The position of the tunnel cuff or exit site should avoid the belt line in children and teens. The distance from the tunnel cuff to the exit site should be at least 1.5 cm, to reduce the risk of extrusion.

A transverse incision is made in the upper quadrant with a #15 scalpel. The rectus fascia and peritoneum are incised. An omentectomy is performed, with the omentum being delivered through the incision and excised using 3–0 polyglactin ties. The peritoneal end of the swan neck peritoneal dialysis catheter is placed into the peritoneal cavity with the curled end in the pelvis. The peritoneum/posterior rectus fascia is closed with a running polypropylene suture, with the cuff maintained just external to the posterior fascia so that it sits within the rectus sheath. The fascial suture should be placed such that it tightens the fascia around the catheter. In the open setting, a purse-string suture can be placed through the cuff and the posterior rectus fascia, further sealing and securing the cuff to the posterior rectus fascia. These maneuvers help

Fig. 74.1 (**a**) Anterior/posterior view of catheter layout with both ends in the caudal direction. (**b**) Side view of catheter, showing relationships with the anterior and posterior rectus sheath

to seal the space where the catheter traverses the posterior rectus fascia and thus reduce the risk of leakage once dialysis is started. The external aspect of the catheter is tunneled 1–2 cm superiorly in the rectus sheath and exits through a separate incision in the anterior rectus sheath. A minimal incision is made for the catheter skin exit site lower in the abdomen, and a tendon passer is used to create a subcutaneous tunnel. Again, it is important that the swan neck catheter be tunneled caudally so that the exit site is in the lower abdomen and the exit site faces downward. The second cuff lies in the subcutaneous tunnel. This configuration is demonstrated in Fig. 74.1a, b.

The function of the catheter is now tested. The titanium Luer lock adapter is placed into the end of the catheter. The transfer set is placed on the end of the Luer lock adapter (Fig. 74.2). The function of the peritoneal dialysis catheter is tested by infusing 30 mL/kg of saline from an intravenous setup with sterile connectors and then draining some of the infused saline by dropping the IV bag below the patient. A sterile betadine minicap is then placed on the end of the transfer set.

The skin is closed with running 4–0 absorbable monofilament suture. No sutures are placed at the exit site. Mastisol® liquid adhesive (Ferndale Laboratories, Ferndale, MI), and ½″ Steristrips (3 M Healthcare, St Paul, MN) are placed to reinforce the incision, and a Tegaderm™ dressing (3 M Healthcare, St Paul, MN) with a pad is used at the catheter exit site.

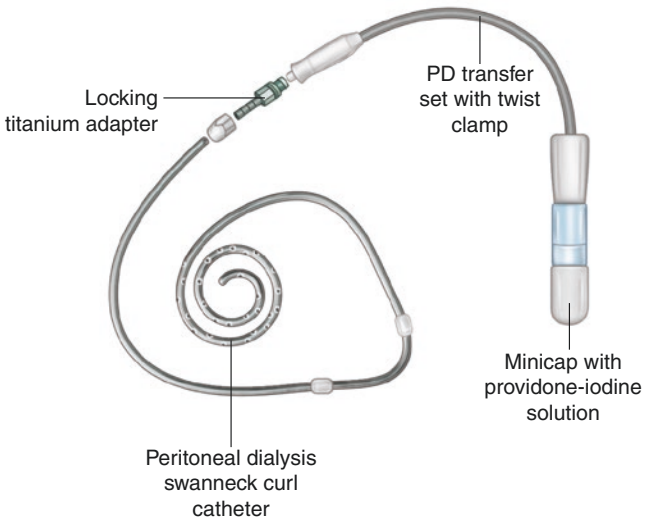

Fig. 74.2 Peritoneal dialysis setup

74.1.3 Laparoscopic Peritoneal Dialysis Catheter Placement

The catheter should be positioned and marked in a similar fashion as described above for the open procedure. A periumbilical incision is made with a scalpel. The fascia is incised and a Veress needle and sheath are placed into the peritoneal cavity. The abdomen is insufflated with CO_2. Except in infants, a 12-mm umbilical port is placed, followed by a 5-mm, 30-degree scope. An additional 5-mm port is placed at the intended peritoneal entry site of the catheter.

As with the open technique, an omentectomy should always be performed. The scope is placed through the 5-mm port and a grasper is used to grab the omentum through the 12-mm port. The omentum is delivered through the port site as the port is removed. The omentum is ligated with 3–0 polyglactin ties or an energy source. The port is then replaced. This procedure is done repeatedly until the omentum is completely excised (Fig. 74.3). Alternatively, a third 5-mm port can be placed in the left side of the abdomen and a surgical energy device can be used to perform the omentectomy intracorporeally. The specimen can then be removed through the umbilical site.

The scope is then placed through the 12-mm port, and the 5-mm port is removed. Under direct vision, the catheter can be placed through the port site into the pelvis. Alternatively, a 16 Fr peel-away sheath can be placed through the 5-mm port site incision and angled inferiorly so that a tangential passage is created through the rectus sheath. Insertion of the peel-away sheath is facilitated by placing an 18- or 19-G needle through the incision and tangentially through the abdominal wall. The needle is exchanged for an 0.035-stiff guidewire. Over the guidewire, the peel-away sheath is placed into the abdomen (Fig. 74.4). Under direct visualization with the camera, the peritoneal end of the swan neck peritoneal dialysis catheter is placed through the sheath and directed into the pelvis (Fig. 74.4). The peel-away sheath is removed.

A small incision is made for the catheter skin exit site lower in the abdomen and a tendon passer is used to create the subcutaneous tunnel. Again, it is important that the swan neck catheter be tunneled caudally so that the exit site is in the lower abdomen and facing downward. One cuff is located just above the rectus fascia. The second cuff resides in the subcutaneous tunnel (about 1.5 cm away from the exit site). The fascia is closed at the umbilicus with 2–0 or 0 polyglactin on a UR-6 needle.

The function of the catheter is now tested and the wounds are closed and dressed as delineated for the open technique.

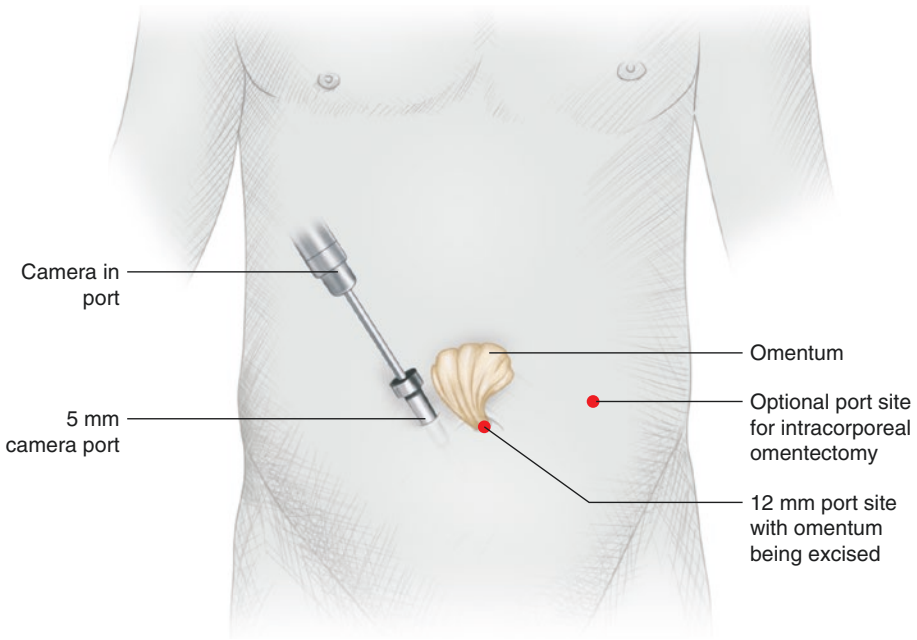

Fig. 74.3 The scope is placed through the 5-mm port and a grasper is used to grab the omentum through the 12-mm port. The omentum is delivered through the port site as the port is removed. The omentum is ligated with 3–0 polyglactin ties or an energy source, and the port is then replaced

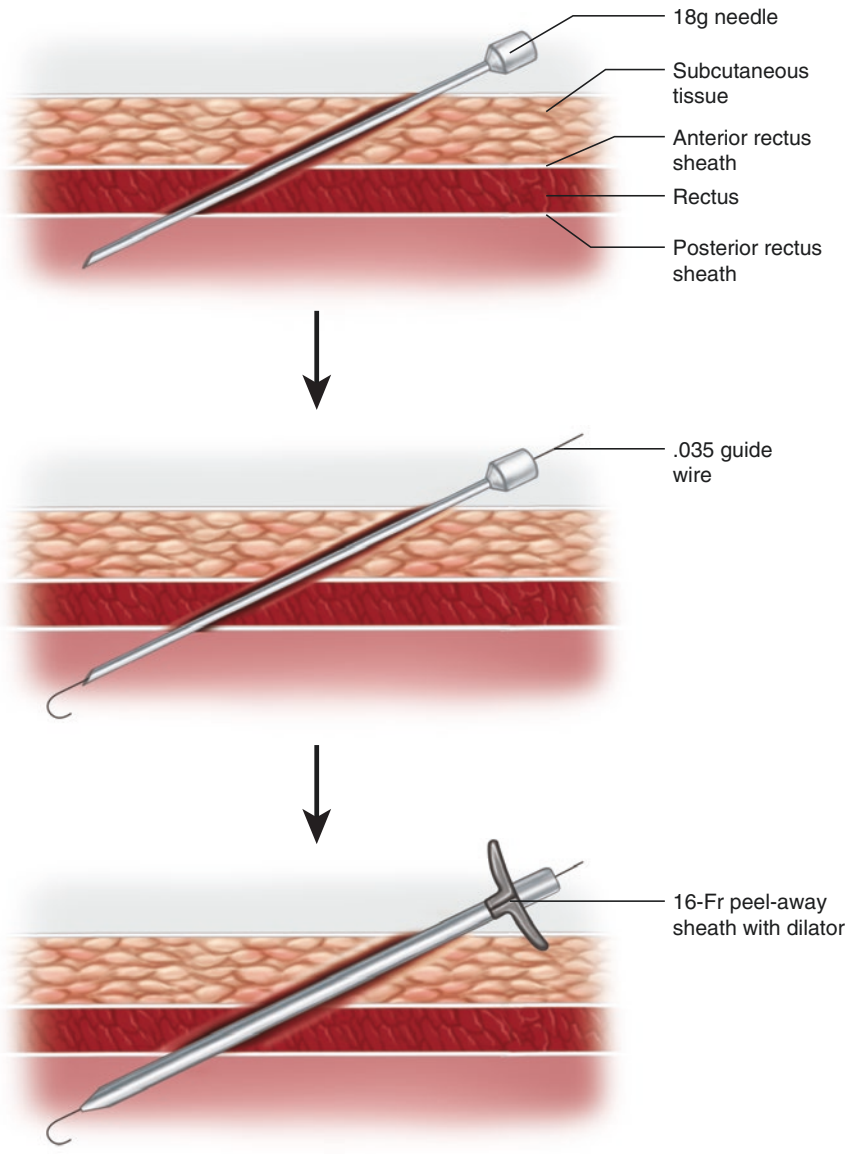

Fig. 74.4 A 16 Fr peel-away sheath can be placed through the 5-mm port site incision and angled inferiorly so that a tangential passage is created through the rectus sheath. This passage can be facilitated by an 18- or 19-G needle and a 0.035-inch guide. Under direct visualization with the camera, the peritoneal end of the swan neck peritoneal dialysis catheter is placed through the sheath and directed into the pelvis. The peel away sheath is removed

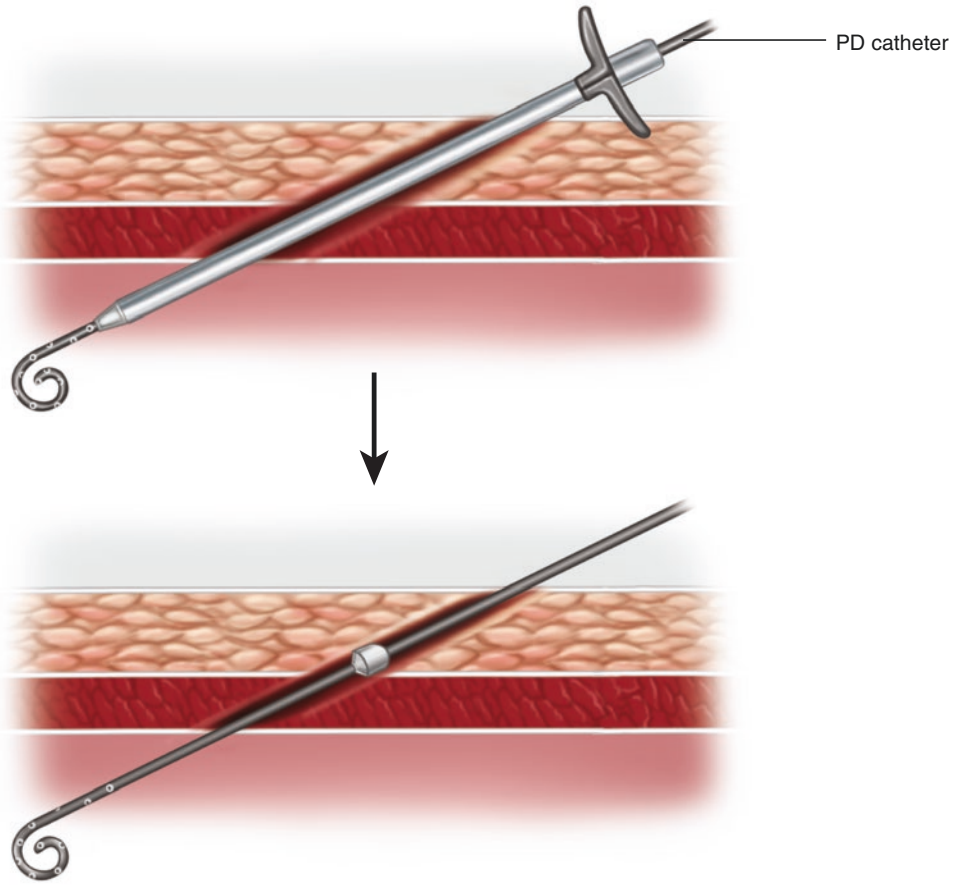

Fig. 74.4 (continued)

74.1.4 Postoperative Care and Complications

If it is necessary to perform peritoneal dialysis immediately, then low volumes (10 mL/kg per dialysate infusion exchange) are used, increasing by 10 mL/kg every 2 weeks until full-volume peritoneal dialysis at 40 mL/kg per exchange is achieved. If the patient's situation allows, the catheter is not used for 4 weeks, after which peritoneal dialysis is performed with 20 mL/kg exchanges, increasing by 10 mL/kg every 2 weeks until full peritoneal dialysis (40 mL/kg) is achieved. Antibiotics are administered in the operating room, but not postoperatively.

The most frequent complication in the first month is outflow failure, which is usually caused by omentum or remnant portions of the omentum obstructing the catheter. If there are inflow problems as well, the trouble may be due to malposition. Laparoscopic exploration, repositioning, and cleaning out of the catheter can be successful, but replacement is necessary in most cases. Leakage is also common immediately after placement. It can be treated by decreasing the amount of dialysate and increasing the number of exchanges. Nearly all of these leaks will seal with conservative management unless the inner cuff has been dislodged. Intra-abdominal bleeding postoperatively can also be a problem. If the hematocrit of the effluent is less than 2 or the red cell count is less than 60,000/cm^3, then the bleeding is insignificant. Peritonitis is a common late complication of peritoneal dialysis and is a major source of morbidity associated with the catheters. Peritonitis will likely be clinically evident on examination. The effluent may be cloudy and will have a white cell count greater than 100/cm^3 with a neutrophil predominance. The most common organism is coagulase-negative *Staphylococcus*. Catheter infections can sometimes be treated with antibiotics placed through the catheter. If this fails, then removal of the catheter may be necessary. Infections of the tunneled portion of the catheter can also cause peritonitis.

74.2 Central Venous Dialysis Catheter Placement

74.2.1 Preoperative Planning

It is important to perform ultrasound evaluation of central vein patency in those patients in whom numerous access devices previously have been placed. All anticoagulants should be discontinued and the platelet count and clotting factors should be normalized.

74.2.2 Central Venous Dialysis Catheter

An ultrasound is used to identify the vein (Fig. 74.5). Access to the internal jugular vein is performed via a low (just above the clavicle) and posterior approach behind the sternocleidomastoid muscle.

The needle should go in at a longitudinal orientation to the ultrasound probe, to allow for continuous guidance of the needle with the ultrasound (Fig. 74.6). This approach allows for a smooth curve to the catheter, which is *critical* for optimal dialysis flow as well as for avoidance of carotid artery puncture and pneumothorax. A 21 G needle is used to gain access, with a 0.017-inch guide wire. Once the 0.018 wire is in place, it should be exchanged for a 0.035-inch guide wire using a 3–4 exchange dilator (a 3 Fr dilator seated inside a 4 Fr dilator.

Using a #11 scalpel, a small incision is made in the skin to enlarge the wire's insertion site. The catheter is placed on the skin and lined up with the wire in the superior vena cava with fluoroscopy in order to determine the appropriate location of the exit site incision. This site is placed in a position that leaves a gentle curve in the catheter and allows the tip of the catheter to sit at the junction of the superior vena cava and the right atrium. This junction can best be estimated with fluoroscopy as a distance of 1.5–2 vertebral bodies below the level of the carina (Fig. 74.7).

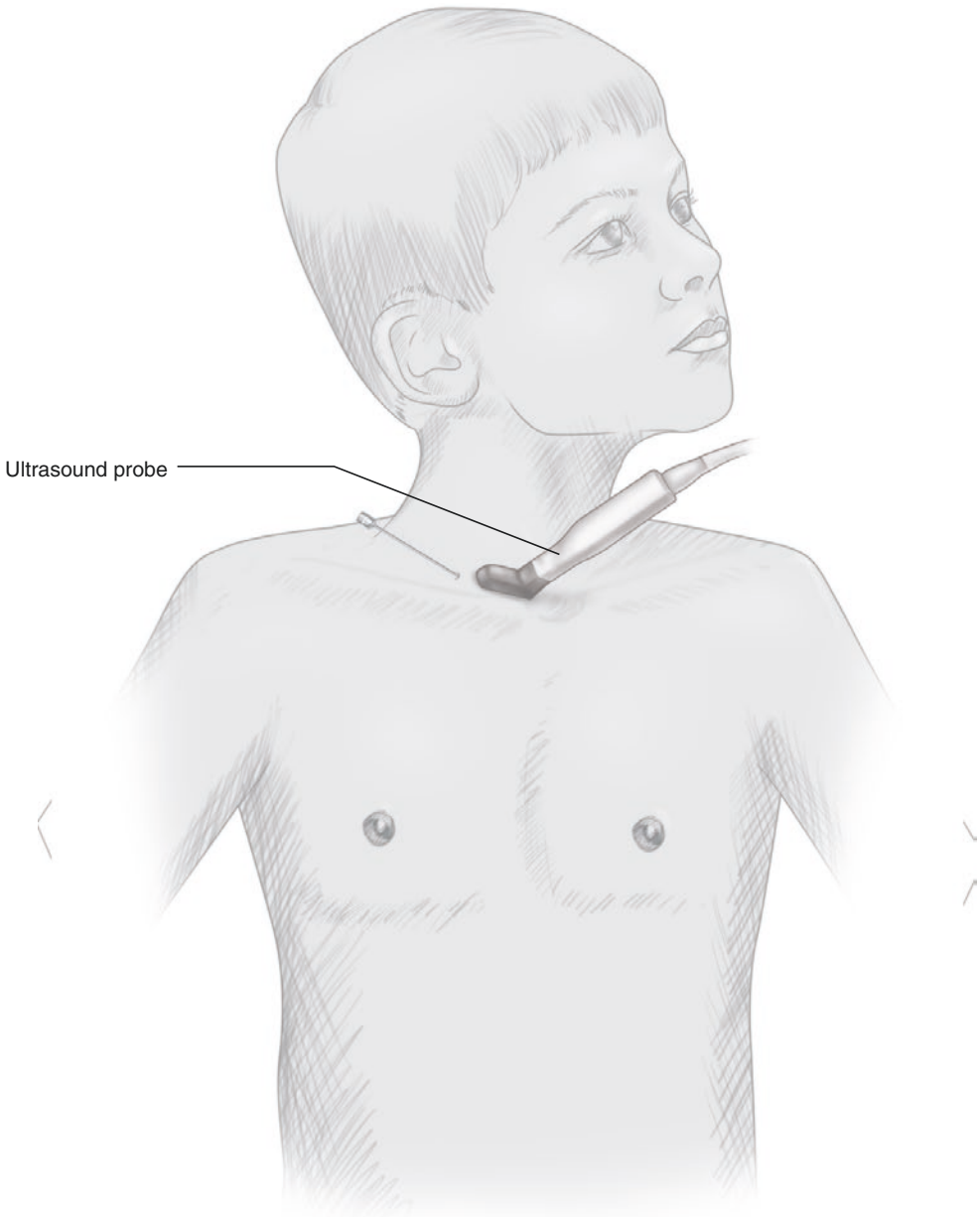

Fig. 74.5 An ultrasound is used to identify the vein for placement of a central venous dialysis catheter. Access to the internal jugular vein is performed via a low (just above the clavicle) and posterior approach behind the sternocleidomastoid muscle

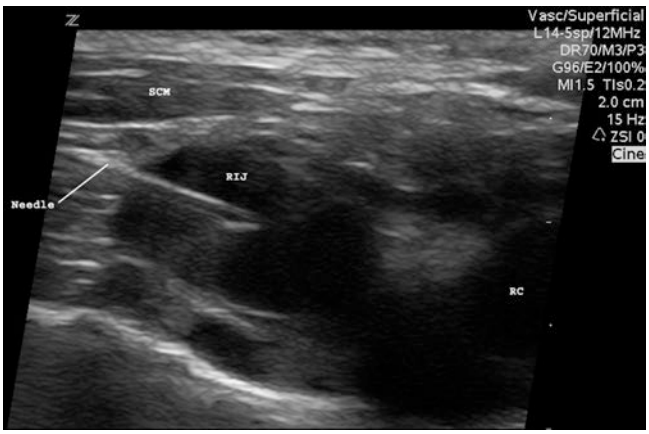

Fig. 74.6 Ultrasound guidance of needle entering laterally into the right internal jugular vein (RIJ) under the sternocleidomastoid muscle (SCM); the right carotid artery (RC) is on the right of the screen

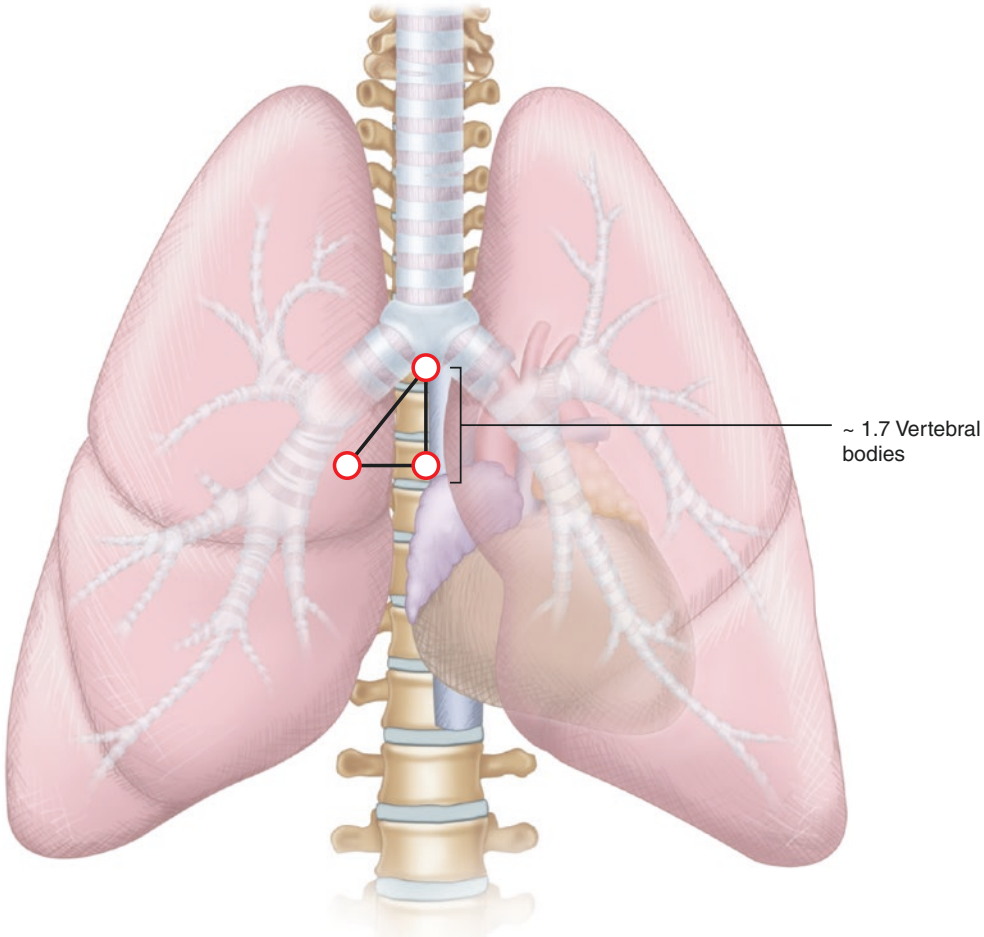

Fig. 74.7 The junction of the superior vena cava and the right atrium can best be estimated with fluoroscopy as a distance of 1.5–2 vertebral bodies below the level of the carina

A tendon passer is used to pass the catheter between incisions. Marcaine (0.25%) is infiltrated into the tissues surrounding the tunnel *before* the catheter is pulled through. A dilator and peel-away sheath are placed over the wire under fluoroscopic guidance, followed by placement of the catheter into the peel-away sheath with forceps (Fig. 74.8). The peel-away sheath is removed and fluoroscopy is used to document placement. The catheter is flushed with heparinized saline, 10 U/mL. (Note that most nephrologists use 1000 U/mL heparin for catheters after dialysis, but in the perioperative period our group uses only 10 U/mL heparin.) The neck incision is then closed with an absorbable suture or skin glue. The catheter is anchored with a 2–0 nylon suture, and a sterile dressing is placed over the exit site.

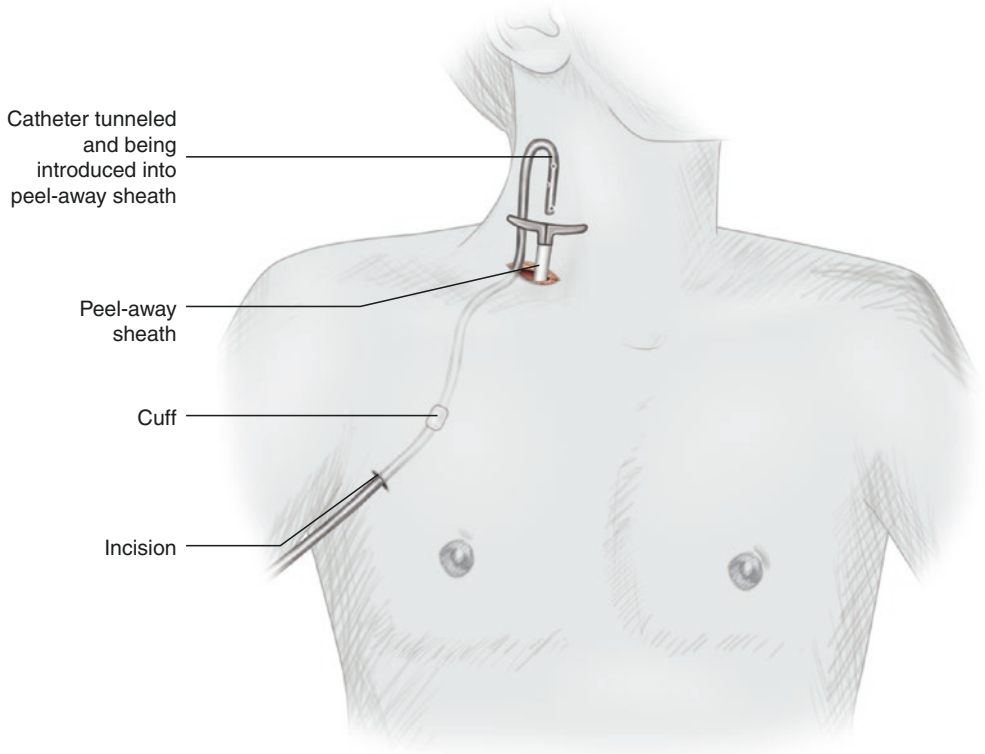

Fig. 74.8 A dilator and peel-away sheath are placed over the wire under fluoroscopic guidance, followed by placement of the catheter into the peel-away sheath with forceps

74.2.3 Postoperative Care and Complications

A postoperative chest radiograph is obtained to confirm catheter placement. Bleeding is a rare, but important, complication. If ultrasound guidance is used for the lateral/posterior approach as described above, pneumothorax is extremely rare.

74.3 Arteriovenous Fistulas

74.3.1 Preoperative Planning

In preparing for placement of an AVF, ultrasound evaluation of both arms for venous and arterial patency is critical, with assessment of cephalic and basilic vein diameter. In general, contrast or CO_2 venography is required because of previous line placements and potential areas of stricture. An Allen's test should be performed when a radiocephalic AVF is planned. The nondominant arm should be used first if there are no contraindications.

In concept, the approach to fistula construction is to perform the most distal access first and to utilize autogenous rather than synthetic material. Thus, a typical order for access would be the following:

1. Radiocephalic (Brescia-Cimino) followed by brachiobasilic in the nondominant arm first, and then the dominant arm.
2. Brachiocephalic graft followed by brachioaxillary graft in the nondominant arm first, followed by the dominant arm.

We have successfully performed brachiobasilic vein AVF procedures in patients as young as 2 years, but performance of an AVF in a vein smaller than 2 mm is unlikely to be successful. As a result, we often prefer a brachiobasilic AVF as the first option in young children. The multi-incision technique initially used, with subcutaneous tunneling of the basilic vein between the two incisions, has been further modified to a two-staged procedure. The two stages allow arterialization of the vein before transferring it to a superficial location. We prefer the two-stage elevation technique because the creation of the arteriovenous anastomosis without dissection of the proximal vein at the first stage minimizes trauma to the small-caliber vein that might predispose it to spasm, kinking, and subsequent thrombosis.

74.3.2 Basilic Vein Transposition

Basilic vein transposition (BVT) fistulas may be created using a single-stage or two-stage transposition technique. In the latter, an incision is performed just proximal to the antecubital fossa (Fig. 74.9) and a second incision is made in the mid upper arm. Through the lower incision, the basilic vein and brachial artery are identified and dissected, with all venous side branches tied off. The vein is transected as distally as possible and is delivered into the upper incision. The anterior surface is marked (with a surgical marking pen) to monitor and avoid twisting as it is pulled through a gently curved subcutaneous tunnel. An end-to-side anastomosis between the basilic vein and the brachial artery is then performed.

Our preference is to perform a two-stage BVT fistula in children. The first stage entails the identification of the basilic vein just proximal to the antecubital fossa, with division of the vein as distally as possible. Care is taken to avoid injury to the median antebrachial cutaneous nerve, which surrounds the basilic vein. The brachial artery is found in the middle of the arm, lateral to the median nerve, usually surrounded by the brachial veins. The artery and vein are controlled with vessel loops. Intravenous heparin (100 U/kg) is administered by the anesthetist. A Heifetz clamp or a small bulldog is placed proximally on the vein, although the pres-

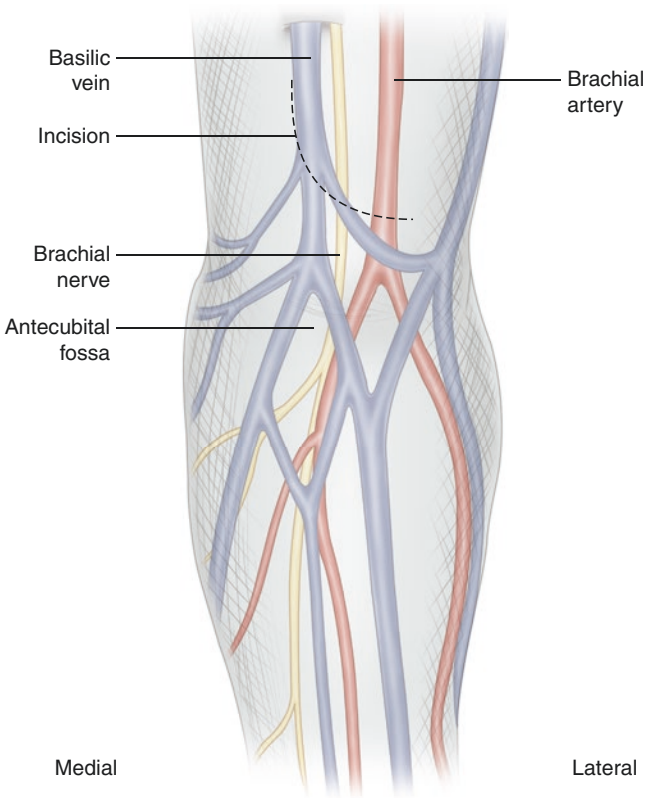

Fig. 74.9 Incision for two-stage basilic vein transposition fistula

ence of a valve may prevent backbleeding and preclude the need for a clamp. In most cases, ligation of the basilic vein is performed just distal to a branched vein (Fig. 74.10). The vein is opened at a branch point to provide a spatulated hood for an end-to-side anastomosis. Before division, the vein is marked with ink on the anterior surface to prevent twisting. The basilic vein is dissected proximally only enough to allow a smooth curve to the brachial artery. Care should be taken to make the vein short enough to avoid kinking. Occasionally, some additional venous branches need to be ligated.

The artery is occluded with proximal and distal Heifetz or small bulldog clamps. An arteriotomy approximately 1 cm in length is made, and an arteriovenous anastomosis is performed with 6–0 or 7–0 polypropylene. The anastomosis is initiated at the heel of the vein and the proximal aspect of the arteriotomy. Next, the suture is run on the posterior aspect of the anastomosis, continuing three fourths of the way around the anastomosis, onto the anterior aspect. The other end of the suture is then run to meet the previous suture and complete the anterior aspect of the anastomosis (Fig. 74.11). In small children, the anastomosis is completed with one or two interrupted sutures to allow for growth. Visual magnification is also used in small children, as well as a microvascular instrument set. The anastomosis is flushed just prior to completion. The clamps are removed and the fistula is checked for Doppler presence of flow and

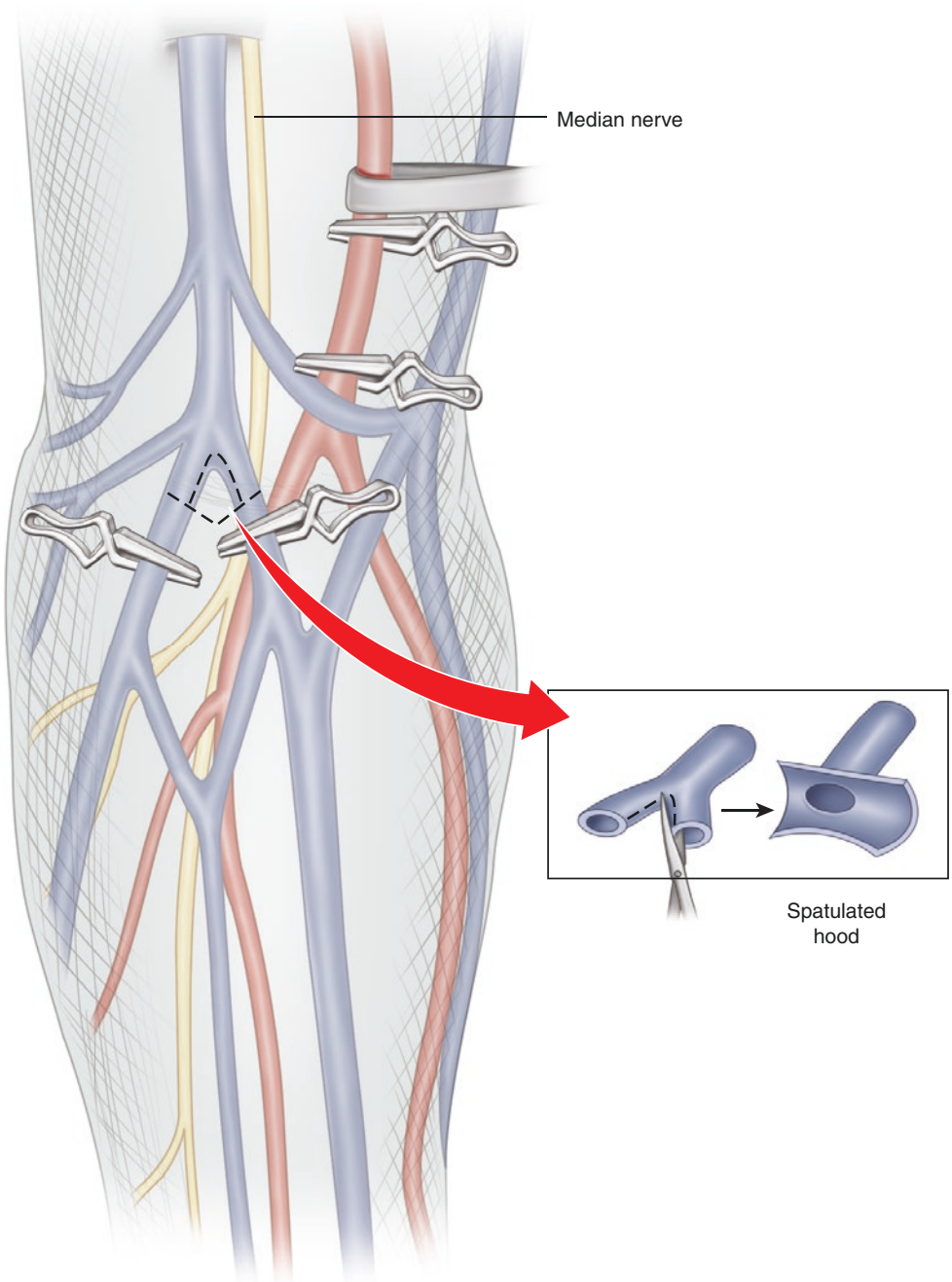

Fig. 74.10 Dissection of the brachial artery and division of the basilic vein just below a branching point. The vein is opened along its inferior aspect to create a spatulated hood (*inset*)

a palpable thrill. The incision is closed with interrupted 3–0 polyglactin suture for the subcutaneous tissues and 5–0 absorbable, monofilament suture for the skin.

Over the ensuing weeks, the vein is examined by serial ultrasound studies until it is confirmed that the caliber has increased to greater than 0.6 cm. Once sufficient vein size is confirmed, fistula elevation is undertaken. The second stage involves an incision along the medial upper arm over the basilic vein (Fig. 74.12). Heparin (100 U/kg) is administered and the vein is dissected free up into the axilla. All side branches are ligated with suture ligatures (Fig. 74.13). The brachial fascia and subcutaneous tissues are closed deep to the vein (Fig. 74.14). Subcutaneous flaps are developed and the incision is closed directly over the vein.

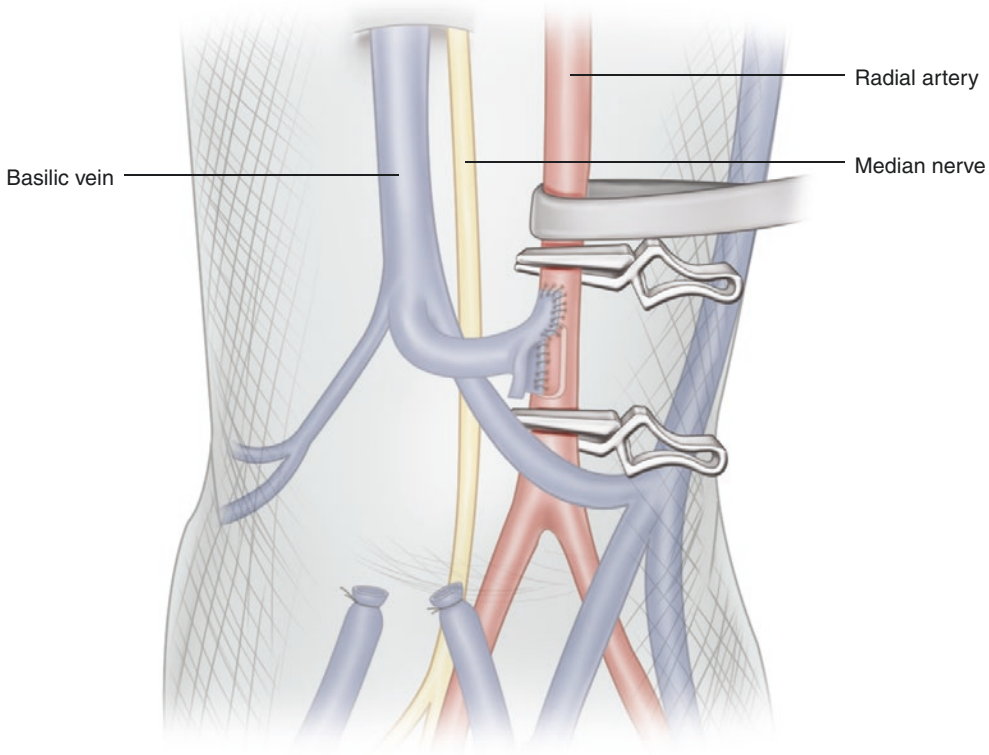

Fig. 74.11 The anastomosis is performed

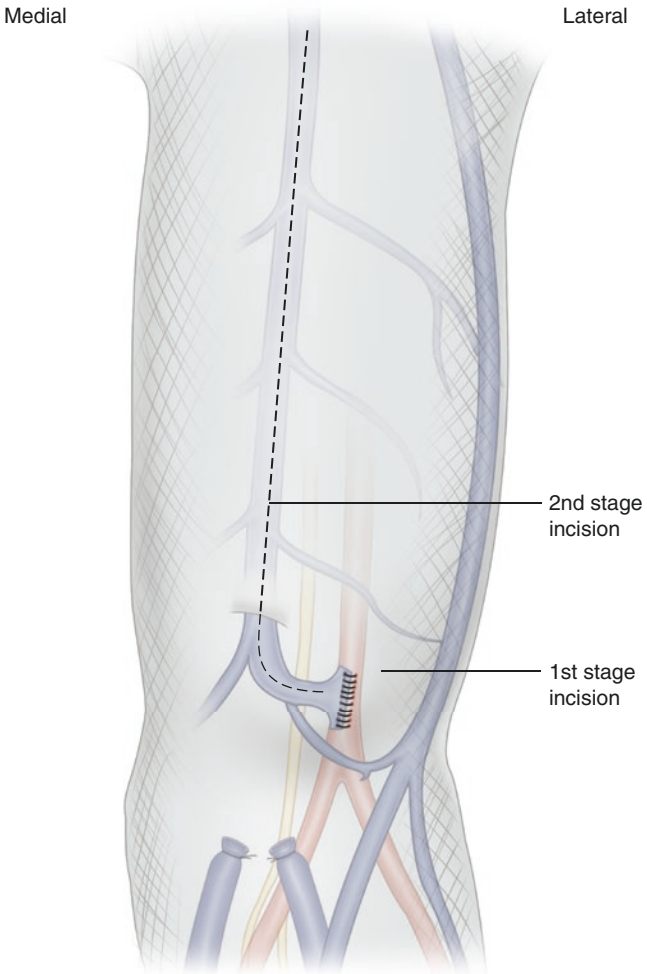

Fig. 74.12 Incision for the second stage of basilic vein transposition. The first-stage incision is continued up the arm over the basilic vein

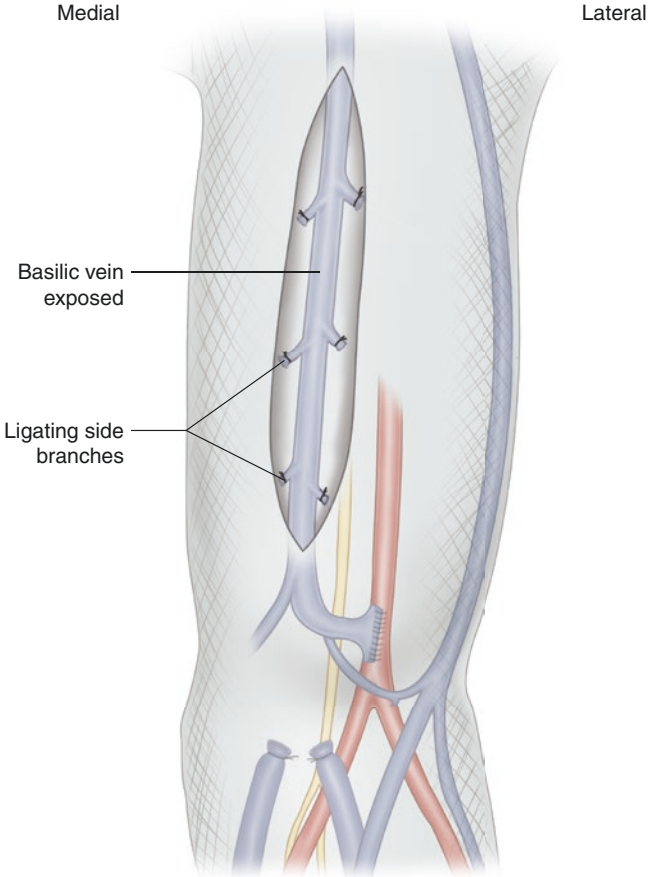

Fig. 74.13 The basilic vein is exposed and dissected free up to the axilla, with side branches ligated

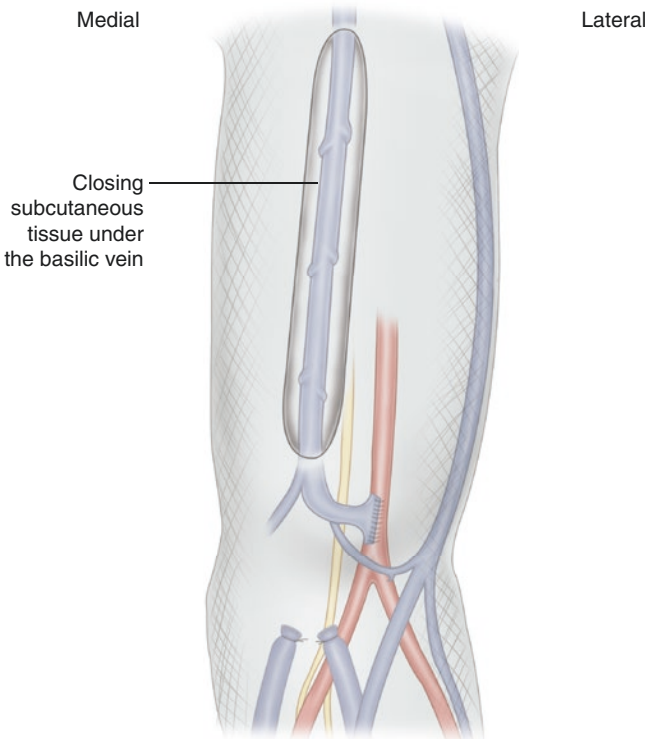

Fig. 74.14 The subcutaneous tissues are closed under the basilic vein, thus elevating it to a position just under the skin

74.3.3 Postoperative Care and Complications

Postoperatively, we use a thromboprophylaxis protocol for all pediatric patients undergoing an AVF. In essence, a continuous infusion of heparin at 10 U/h is administered starting in the operating room when the anastomosis is complete, followed by low-molecular-weight heparin (full anticoagulation dosing) on postoperative day 1. The low-molecular-weight heparin is continued until maturation is documented approximately 2–3 weeks after the second-stage procedure.

Fistula failure occurs in 7% of the two-stage BVT group, compared with 59% using other AVF approaches. Other complications include thrombosis (33%), bleeding (10%), hematoma formation (14%), cellulitis (5%), development of stenosis (12%), and steal (2%). The use of a two-stage BVT in children results in a higher rate of success in creating an AVF that is used for dialysis (87% vs. 48%). Overall, mean patency (or duration of use) of AVF in children has been reported to be as low as 6 months and as high as 30 months.

Thrombosis is the most common postoperative complication with fistulas. Early thrombosis (within the first 3 months) is usually secondary to technical errors in fistula construction, such as twisting of the vein, small-sized anastomosis, or low flow secondary to compression or hypotension. Late thrombosis is usually caused by stenosis secondary to intimal hyperplasia or repeated access punctures. Physical examination, ultrasound, and angiography are all useful in evaluating potential problems. Interventions include thrombectomy, angioplasty, and revision of the fistula.

Conclusion

Chronic dialysis can be accomplished in three ways: peritoneal dialysis, catheter-based hemodialysis, and fistula-based dialysis. Each has strengths and weaknesses and the appropriate application. Each can provide life-saving therapy, but care must be taken in technique and postoperative care.

Suggested Reading

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