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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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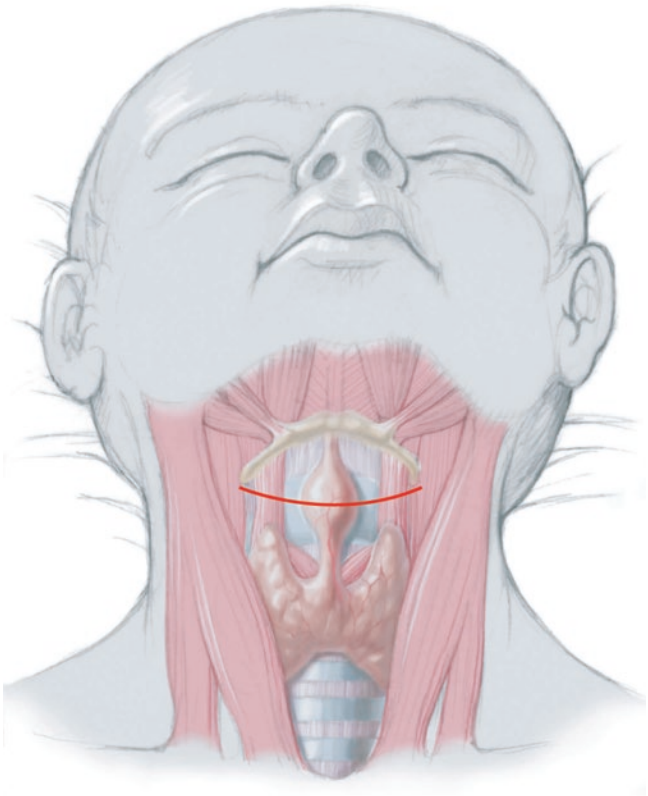

Fig. 1.1 Following induction of general anaesthesia with endotracheal intubation, the neck is hyperextended by placing a sandbag or towel roll beneath the shoulders. A horizontal skin incision is made over the cyst. In case of a fistula, the cutaneous orifice is circumcised in a horizontally oriented, 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

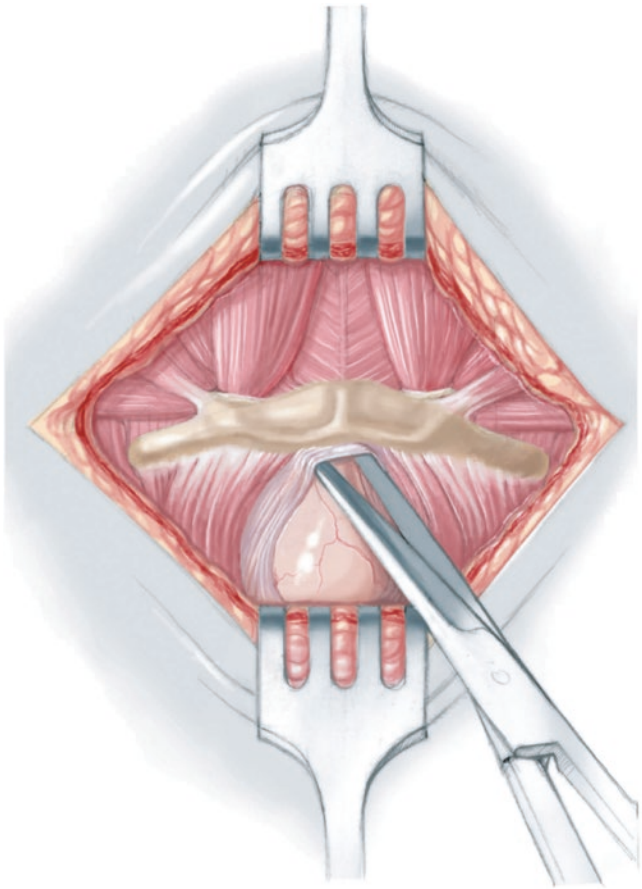

Fig. 1.2 The duct is attached to the cyst, running in a cephalic direction between the sternohyoid muscles to the body of the hyoid bone. It is usually not possible to recognize whether the duct perforates the hyoid body or passes across its anterior or posterior surface. The central part of the hyoid 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

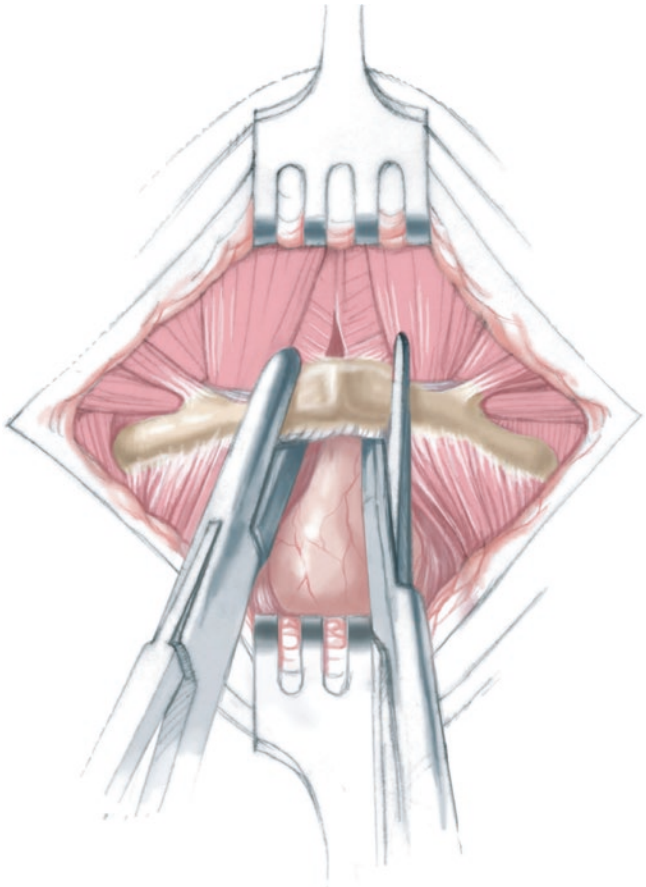

Fig. 1.3 The exposed hyoid bone is then stabilized with strong Kocher forceps on one side, clearly lateral to the median line, and the central segment is excised with strong Mayo scissors

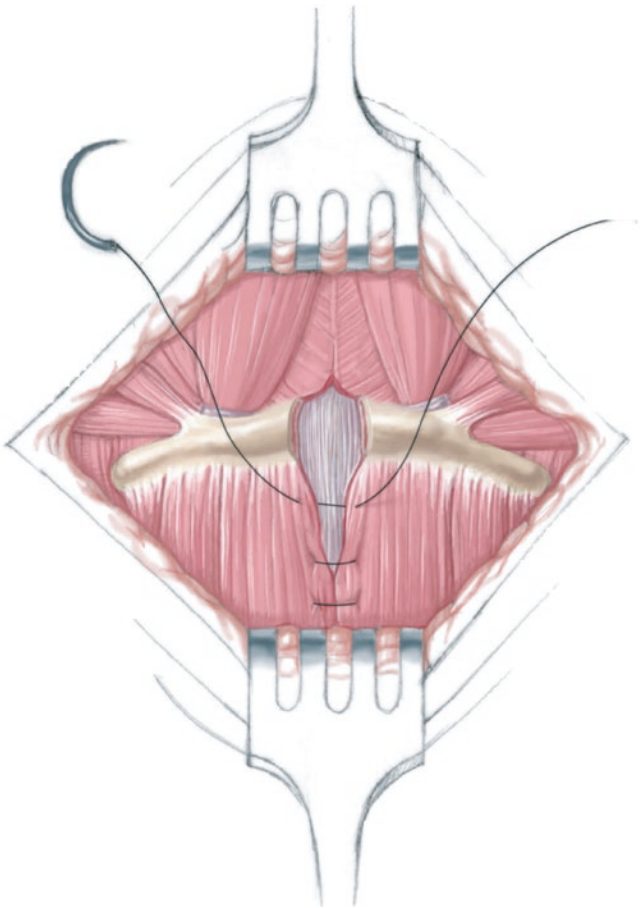

Fig. 1.4 If the duct extends beyond the posterior aspect of the hyoid bone, it is followed upward and divided close to the base of the tongue with a 5/0 absorbable transfixation ligature. If the floor of the mouth is entered accidentally, the mucosa of the tongue is closed with interrupted plain absorbable 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

El Gohary Y, Gittes G. Congenital cysts and sinuses of the neck. In: Puri P, editor. Newborn surgery. 3rd ed. London: Hodder Arnold; 2011.

Foley DS, Fallat ME. Thyroglossal duct and other congenital midline cervical anomalies. Semin Pediatr Surg. 2006;15:70–5.

Horisawa M, Niiomi N, Ito T. Anatomical reconstruction of the thyroglossal duct. J Pediatr Surg. 1991;26:766–9.

Peretz A, Leibermann E, Kapelushnik J, et al. Thyroglossal duct carcinoma in children: case presentation and review of the literature. Thyroid. 2004;14(9):777–85.

Waldhausen JHT, Tapper D. Head and neck sinuses and masses. In: Ashcraft KW, editor. Pediatric surgery. Philadelphia: WB Saunders; 2000. p. 987–99.

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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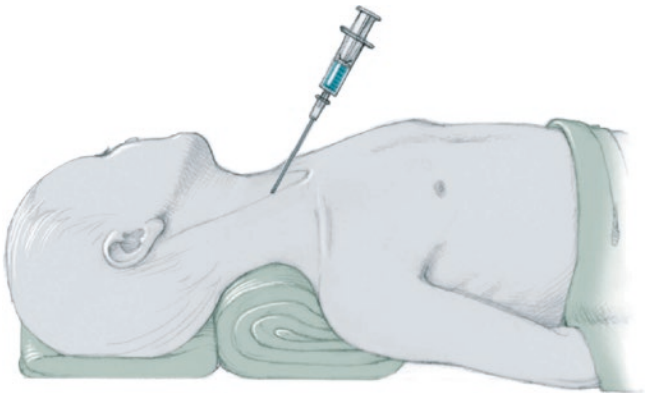

Fig. 2.1 For excision of the most common second branchial pouch remnant, the patient is placed in a supine position. Following induction of general anaesthesia with endotracheal intubation, the head is turned to the side. A sandbag is placed beneath the shoulders to expose the affected side. Instillation 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

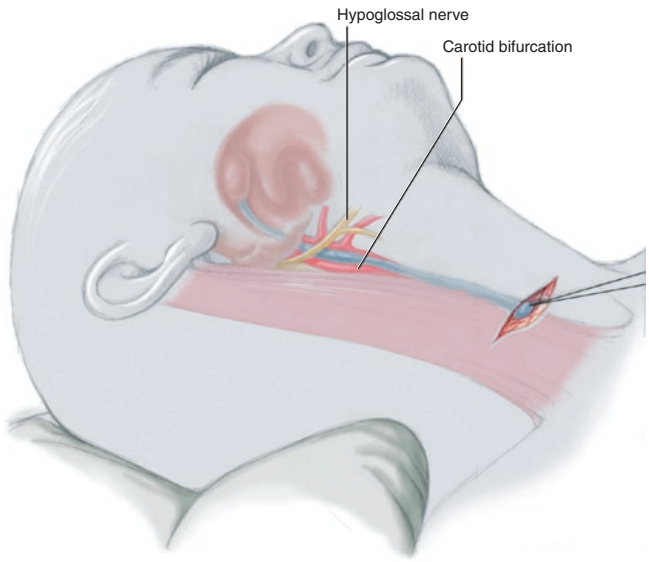

Fig. 2.2 In patients with a branchial cyst, the incision is made over the cyst along the Langer's lines. An elliptical incision is made around the sinus. A traction suture is applied to it just underneath the skin for manipulation during further dissection

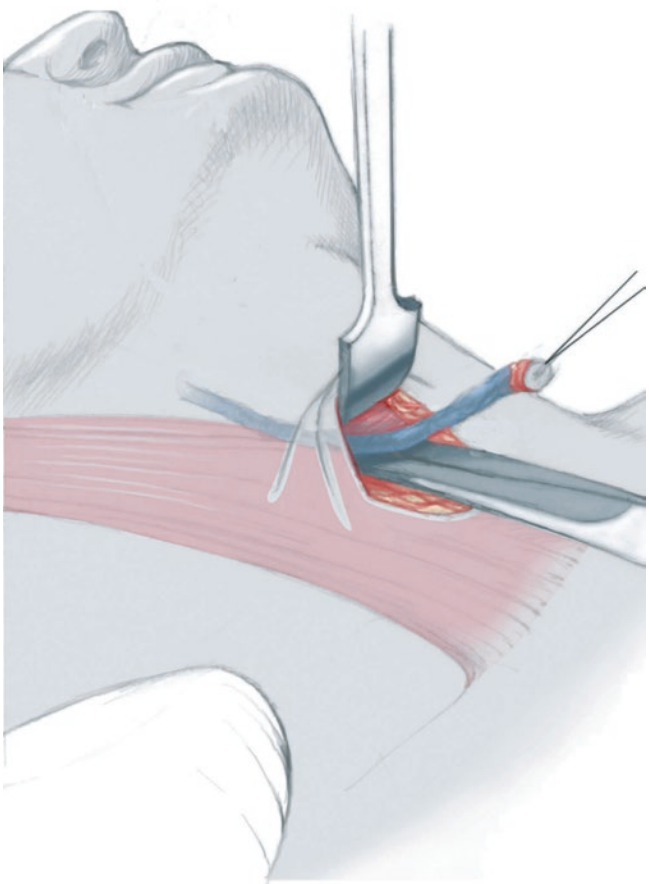

Fig. 2.3 Subcutaneous tissue and platysma are divided until the sinus tract is reached; the tract is easily palpable when the traction suture is gently tensed. Mobilization of the sinus continues in a cephalad direction as far as possible with gentle traction. The operation can usually be done through 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

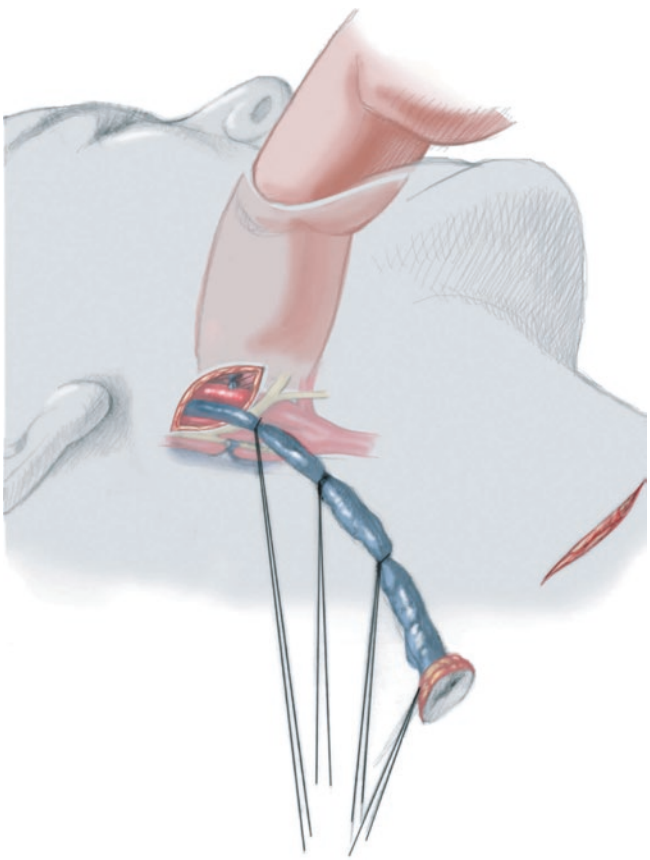

Fig. 2.4 In adolescents, a second transverse (stepladder) incision, made approximately 4–5 cm above the first, may be necessary to completely excise the sinus tract. Both incisions are closed with absorbable interrupted fine subcutaneous (5/0) and subcuticular (6/0) sutures

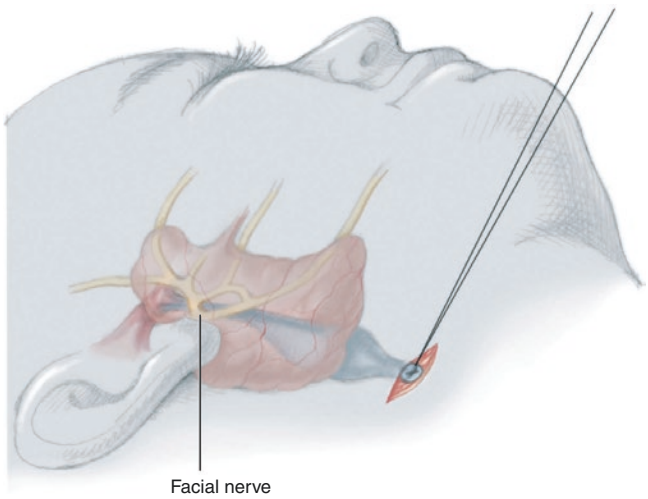

Fig. 2.5 For first branchial pouch remnants, the opening of the fistula is circumcised with an elliptical skin incision. Careful dissection liberates the subcutaneous layer of the embryological remnant, which is now transfixed with a stay suture. This suture is used for traction on the duct, which facilitates 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.

Suggested Reading

El Gohary Y, Gittes G. Congenital cysts and sinuses of the neck. In: Puri P, editor. Newborn Surgery. 3rd ed. London: Hodder Arnold; 2011.

Magdy EA, Ashram YA. First branchial cleft anomalies: presentation, variability and safe surgical management. Eur Arch Otorhinolaryngol. 2013;270:1917–25.

Nicoucar K, Giger R, Jaecklin T, Pope HG Jr, Dulguerov P. Management of congenital third branchial arch anomalies: a systematic review. Otolaryngol Head Neck Surg. 2010;142:21–28.e2.

Nicoucar K, Giger R, Pope HG Jr, Jaecklin T, Dulguerov P, et al. Management of congenital fourth branchial arch anomalies: a review and analysis of published cases. J Pediatr Surg. 2009;44:1432–9.

Sadler TW. Langman's medical embryology. 12th ed. Philadelphia: Lippincott Williams & Wilkins; 2006.

Waldhausen JHT. Branchial cleft and arch anomalies in children. Semin Pediatr Surg. 2006;15:64–9.

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.

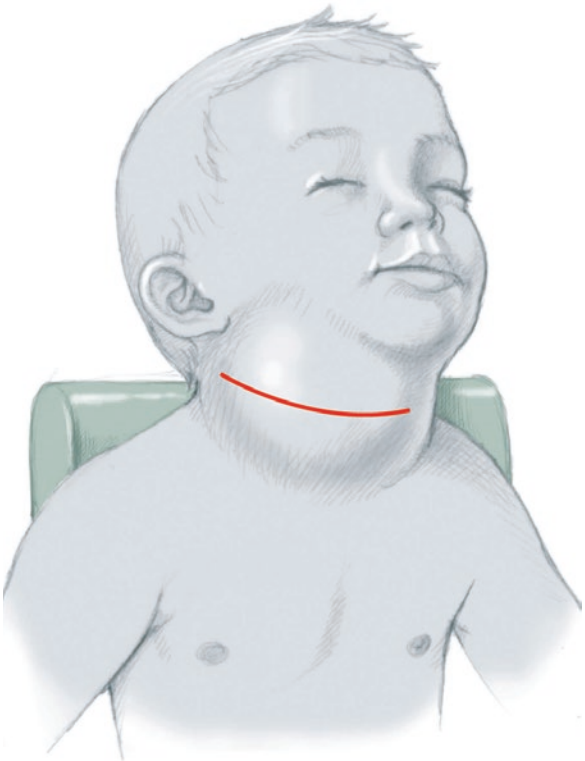

Fig. 3.1 For the most common (cervical) lesions, a transverse skin crease incision extending the length of the mass is placed in Langer's lines

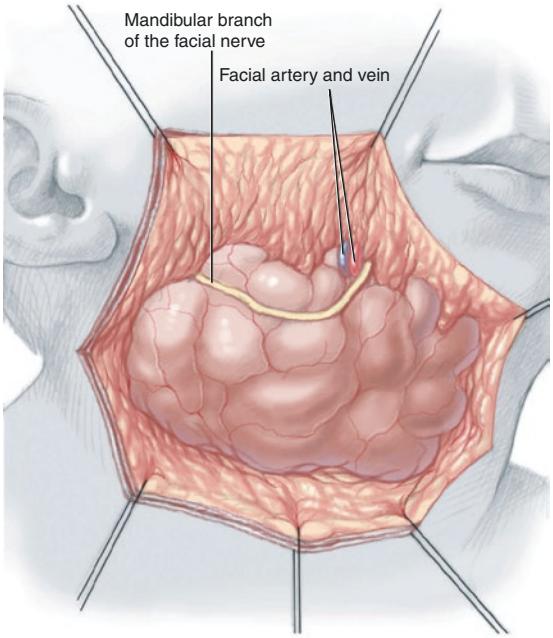

Fig. 3.3 Dissection of cervical lesions begins at the superior margin of the mass, near the ramus of the mandible. Upward reflection of the facial artery and vein allow the precise visualization necessary to preserve the marginal mandibular branch of the facial nerve. Bipolar cautery may be used and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

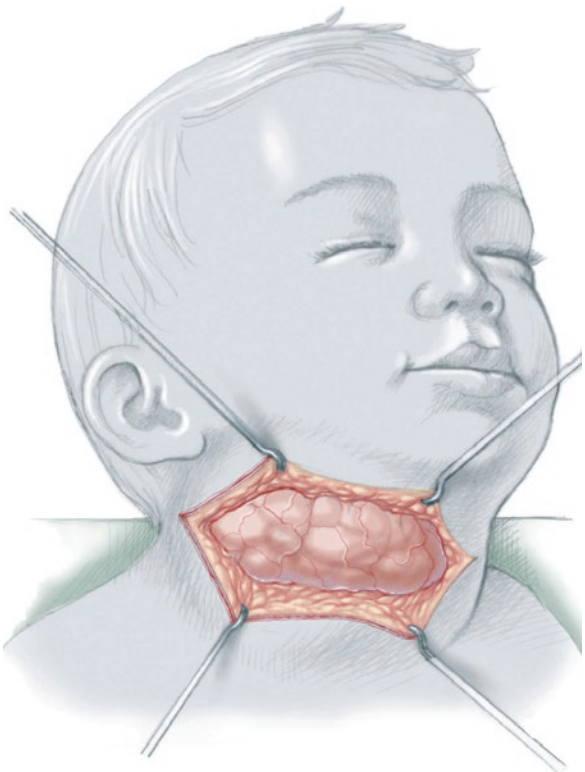

Fig. 3.2 If the lymphatic malformation demonstrates dermal infiltration, an ellipse of skin is removed. Otherwise, generous sub-platysmal skin flaps are raised. The external jugular vein and ansa cervicalis are not considered essential and may be sacrificed

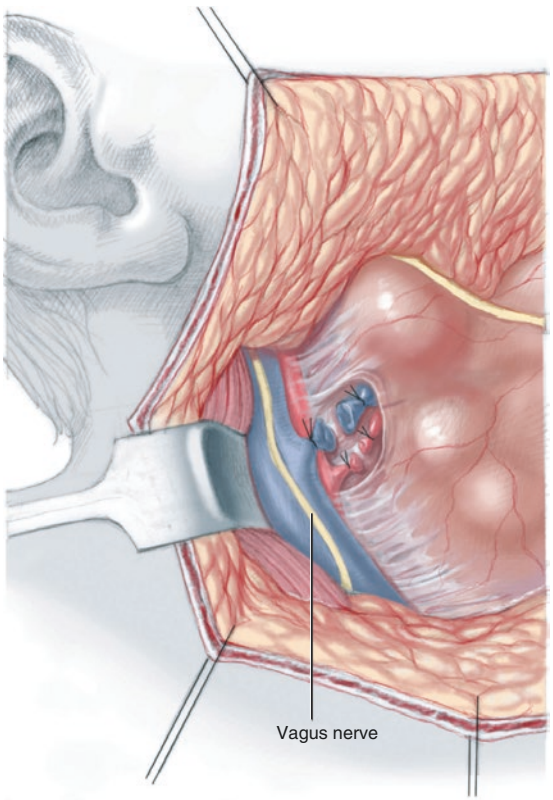

Fig. 3.4 The dissection proceeds medially, lifting the cyst from the surrounding alveolar tissue. It may be necessary to divide the middle thyroid vein and artery as the carotid sheath is approached. Deep dissection frequently involves the contents of the carotid sheath and sometimes the following nerves: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

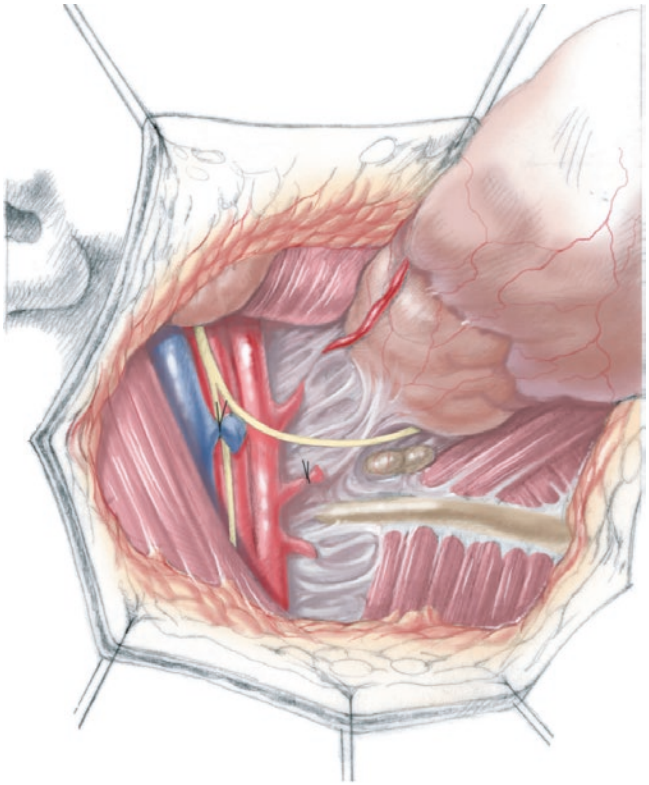

Fig. 3.5 Care is taken to preserve the hypoglossal nerve as it passes through the bifurcation of the carotid artery. The mass must then be freed from the hyoid bone and submandibular gland. It is rarely necessary to remove the submandibular gland en bloc with the mass, sacrificing the facial artery. 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

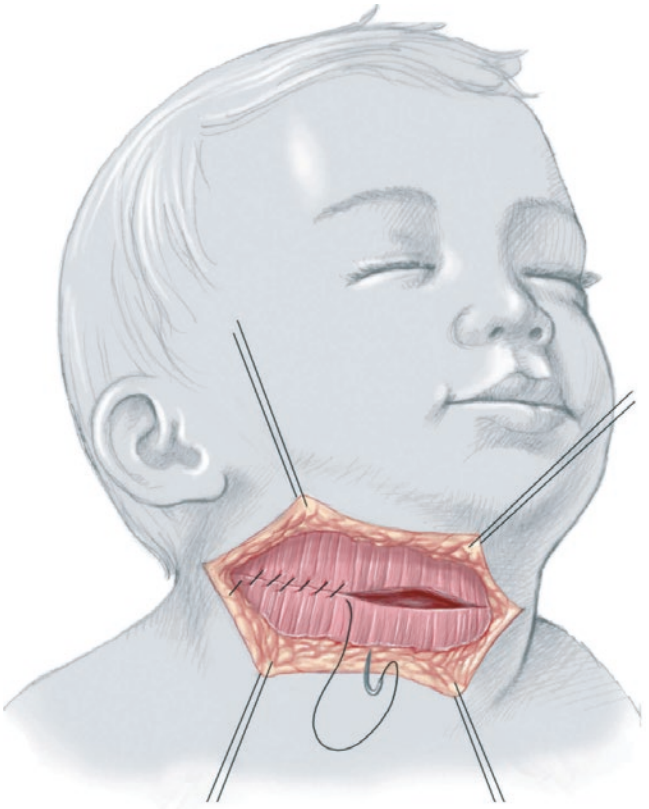

Fig. 3.6 The platysma is reapproximated with fine absorbable sutures, and the skin is closed with subcuticular sutures of similar material. Closed suction drainage is used for most lesions

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.

Suggested Reading

Adams DM, Trenor III CC, Hammill AM, et al. Efficacy and safety of Sirolimus in the treatment of complicated vascular anomalies. Pediatrics 2016;137:e20153257

Azizkhan RG. Complex vascular anomalies. Pediatr Surg Int. 2013;29:1023–38.

Charabi B, Bretlau P, Bille M, Holmelund M. Cystic hygroma of the head and neck--long-term follow up of 44 cases. Acta Otolaryngol Suppl. 2000;543:248–50.

Lackner H, Karastaneva A, Schwinger W, et al. Sirolimus for the treatment of children with various complicated vascular anomalies. Eur J Pediatr 2015;174:1579–1584

Lazar DA, Olutoye OO, Moise KJ Jr, Ivey RT, Johnson A, Ayres N, et al. Ex-utero intrapartum treatment procedure for giant neck masses--fetal and maternal outcomes. J Pediatr Surg. 2011;46:817–22.

Nehra D, Jacobson L, Barnes P, Mallory B, Albanese CT, Sylvester KG. Doxycycline sclerotherapy as primary treatment of head and neck lymphatic malformations in children. J Pediatr Surg. 2008;43:451–60.

Swetman GL, Berk DR, Vasanawala SS, Feinstein JA, Lane AT, Bruckner AL. Sildenafil for severe lymphatic malformations [Case Reports Letter]. N Engl J Med. 2012;366:384–6.

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.

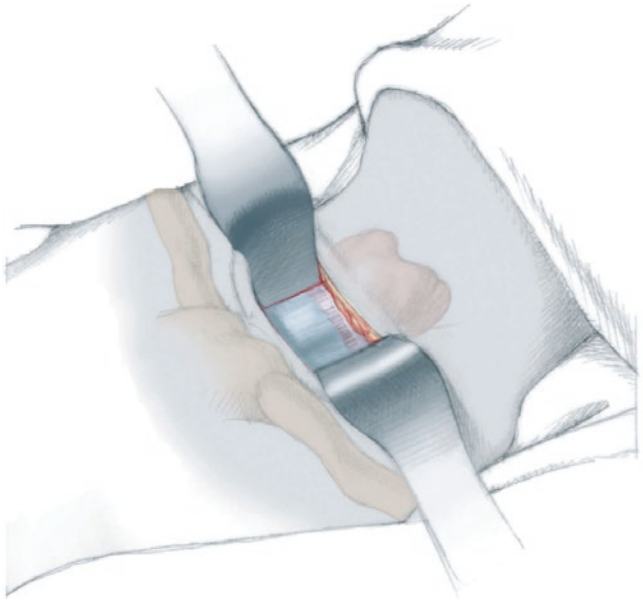

Fig. 4.1 The tracheostomy begins with a horizontal incision after local anaesthetic with adrenaline for infiltration is administered midway between the cricoid cartilage and the sternal notch. The incision is extended to the subcutaneous tissues and platysma muscle (superficial cervical fascia). Excess 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

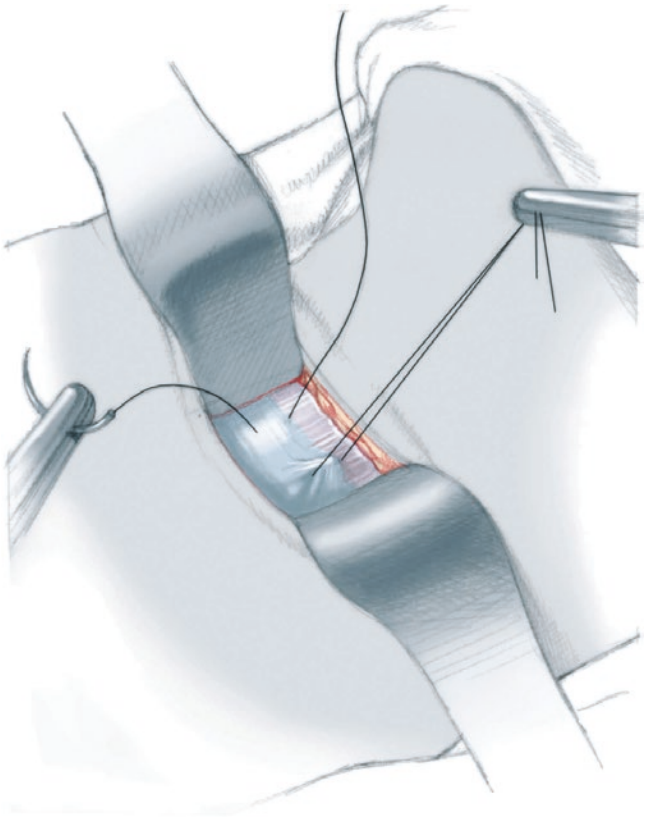

Fig. 4.2 The trachea is now exposed. The proposed tracheostomy cannula should be opened and its outer diameter visually checked against the exposed trachea. The pretracheal fascia is scored vertically with bipolar cautery to coagulate any vessels on the trachea in the midline. A suture of 4/0 nonabsorbable 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