CHAPTER 5

Oesophageal Atresia

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INTRODUCTION

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 within the fourth week of gestation, during which 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, however, occur sporadically without evidence for either hereditary or specific environmental teratogenic causes. The incidence approximates to 1:4,500 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 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 a polyhydramnios in the second half of pregnancy. Polyhydramnios is an unspecific 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 12 F feeding tube into the stomach is not possible, oesophageal atresia is almost certain. Immediate oro- or naso-oesophageal insertion of a Replogle tube as soon as the diagnosis is established is mandatory for continuous or intermittent aspiration of saliva in order to prevent aspiration. The baby should be nursed propped up in order to prevent aspiration of gastric contents in to the tracheobronchial tree.

Prior to surgery, the type of atresia should be determined. 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 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.

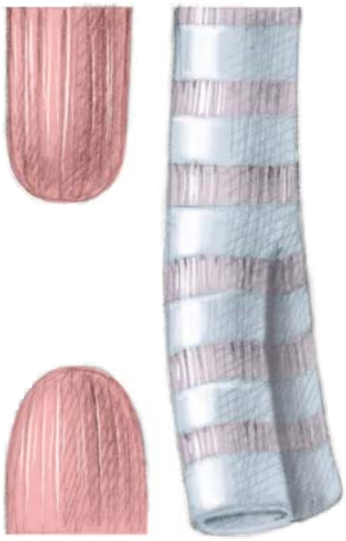
Additional malformations are looked for. Every neonate is checked for visible anomalies such as anal atresia or limb malformations. The thoraco-abdominal radiography may reveal duodenal or lower intestinal atresia, a diaphragmatic hernia and/or skeletal anomalies. Ribs and vertebrae must be counted and carefully examined for deformations. Usage of contrast medium is rarely indicated. Cardiologic assessment, including echocardiography, forms part of routine pre-operative 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 searching for urinary tract anomalies is performed routinely.

The baby is nursed in the intensive care unit (ICU). Immediate surgery is rarely required, so that all above-mentioned investigations can be performed step by step. Intubation and ventilation is only necessary in cases of respiratory distress, severe pneumonia or severe associated malformations demanding 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 if a high gastrointestinal atresia is associated.

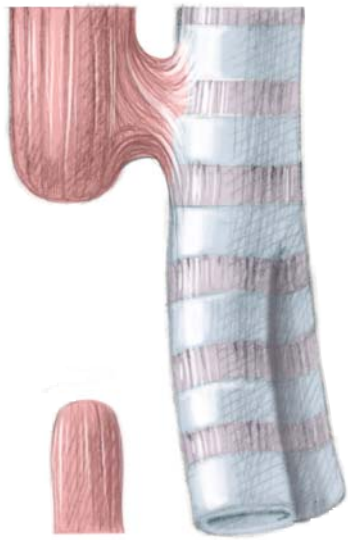
Figure 5.1a–e

Classifications usually take their orientation on 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' 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. The 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 – H-type fistula – as type 4/E (3%), although it belongs to a different spectrum because the oesophagus is patent. In Gross' classification, congenital oesophageal stenosis constitutes type F.

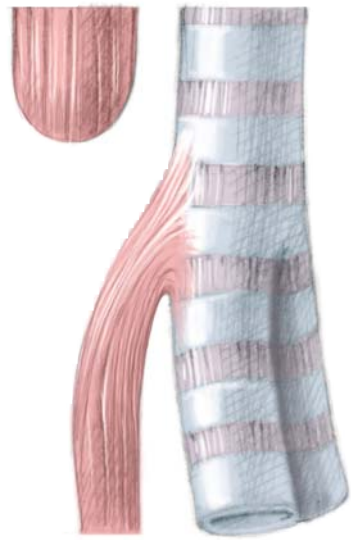
Figure 5.1a–e



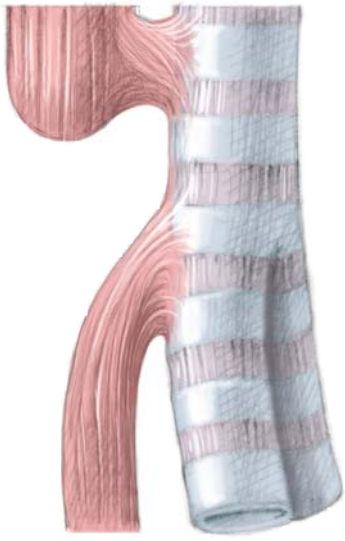
2/A



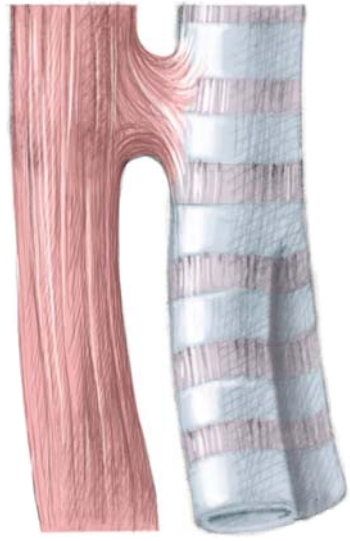
3a/B



3b/C



3c/D



4/E

Figure 5.2

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 intra-operatively. Broad-spectrum antibiotic prophylaxis is administered on induction. We routinely start with a tracheo-bronchoscopy using a rigid 3.5 mm endoscope. Trachea and main bronchi are briefly inspected, and the fistula to the oesophagus is localized, which is usually approximately 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 with 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.

Figure 5.3

The standard approach for repair of an oesophageal atresia is a right latero-dorsal thoracotomy. If a right aortic arch is diagnosed pre-operatively, a left-sided thoracotomy is recommended. However, if an unsuspected right descending aorta is encountered during surgery, the procedure can be continued in most cases, 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 underneath the feet, or the limbs wrapped with cotton wool, which protects against heat loss at the same time. 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 – due 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 rectracted, 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.

Figure 5.2

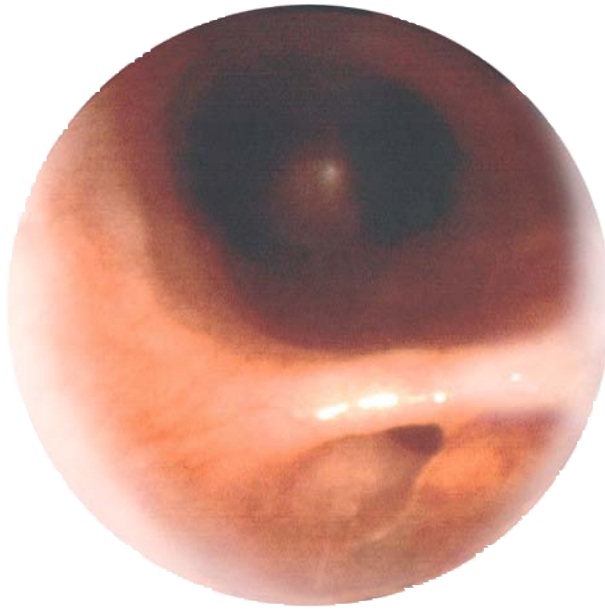


Figure 5.3

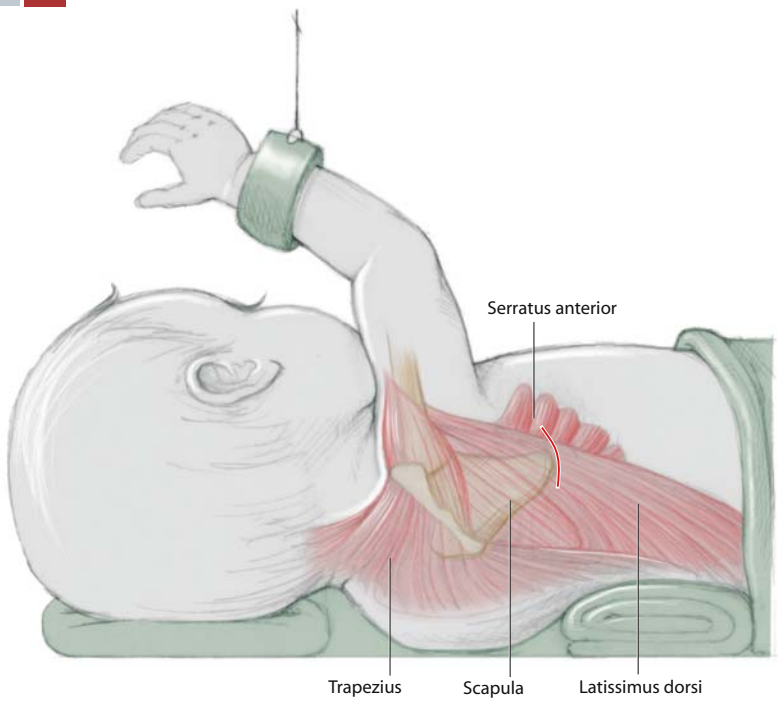


Figure 5.4–5.6

The intercostals 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, the other to proceed with the dissection. An inadvertent tear in the pleura can be closed with a fine (6/0) monofilament absorbable suture.

Figure 5.4

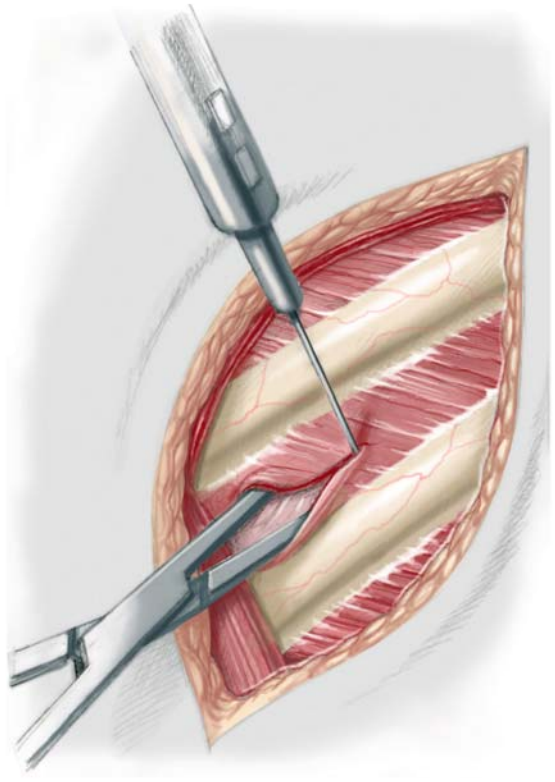


Figure 5.5

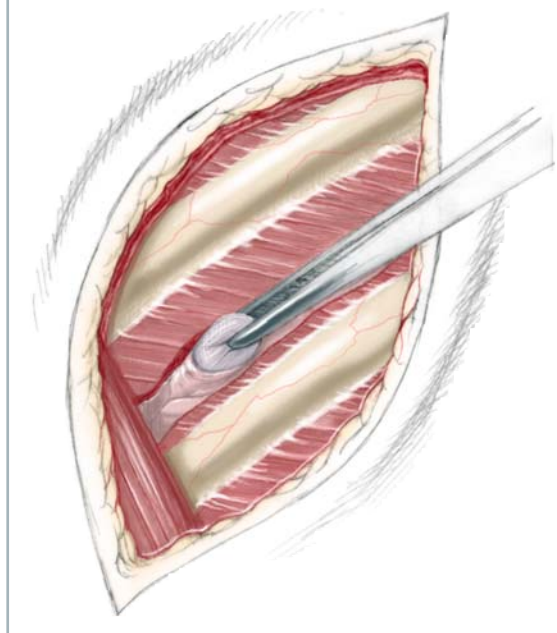


Figure 5.6

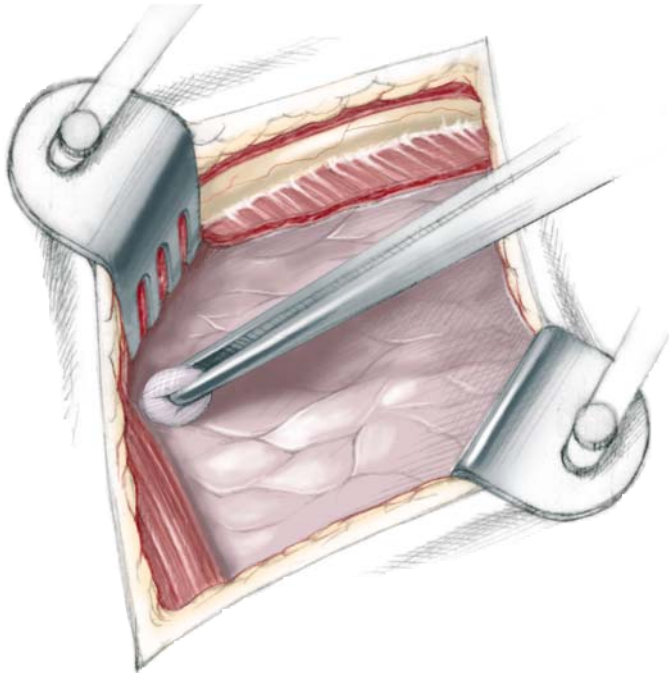


Figure 5.7

The azygos vein is mobilized with right-angled forceps and divided in 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 towards the lower oesophagus. The lower oesophagus 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. Preservation of all vagal fibres supplying the lower oesophagus is also aimed for. Denudation invariably entails a significant motility disorder and may cause severe gastro-oesophageal reflux.

Figure 5.8, 5.9

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. This facilitates identification of tracheo-esophageal fistula, which is now freed from surrounding tissue.

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

Figure 5.7

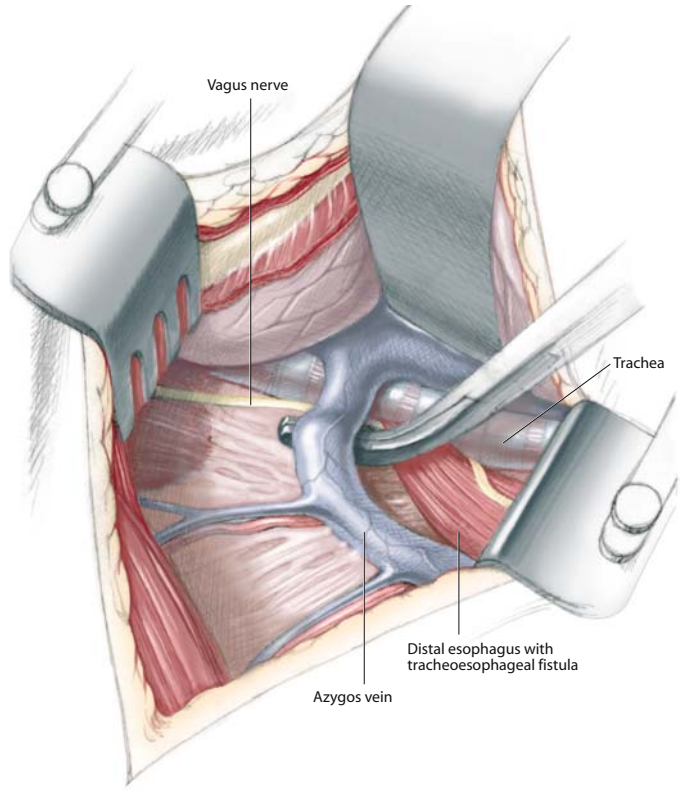


Figure 5.8

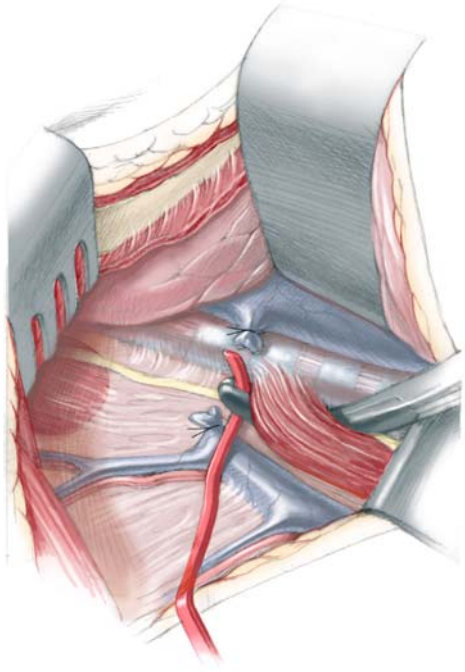


Figure 5.9

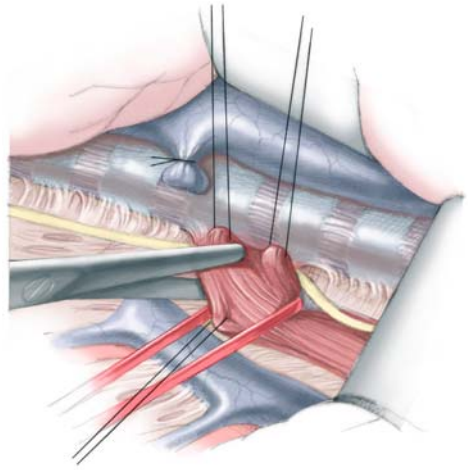


Figure 5.10

At this stage, the fistula is divided and closed with a continuous absorbable monofilament 6/0 suture. Some authors prefer interrupted stitches, others apply transfixation stitches. The level of division must be as close to the trachea as possible without risking a narrowing of the airway. Since 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.

Figure 5.11

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 firm connective tissue layer. 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. Contrary to 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.

Figure 5.12

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 pre-anastomotic outpouching. 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.

Figure 5.10

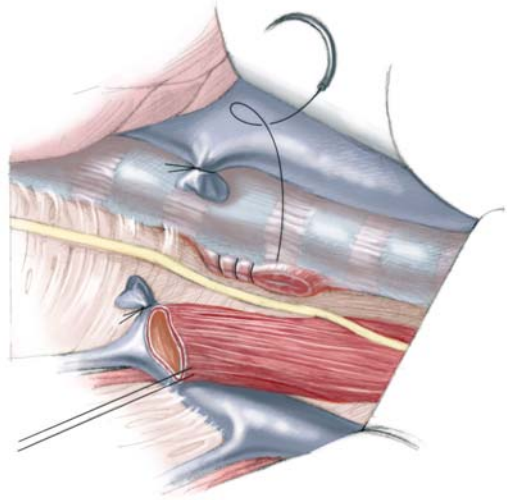


Figure 5.11

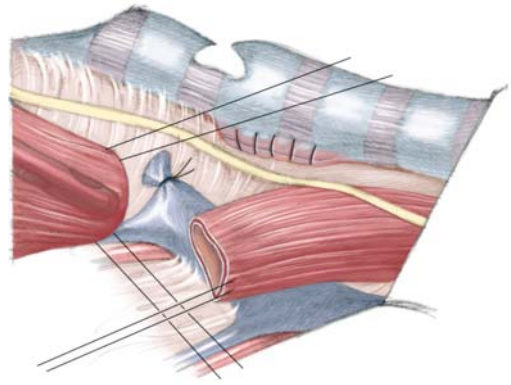


Figure 5.12

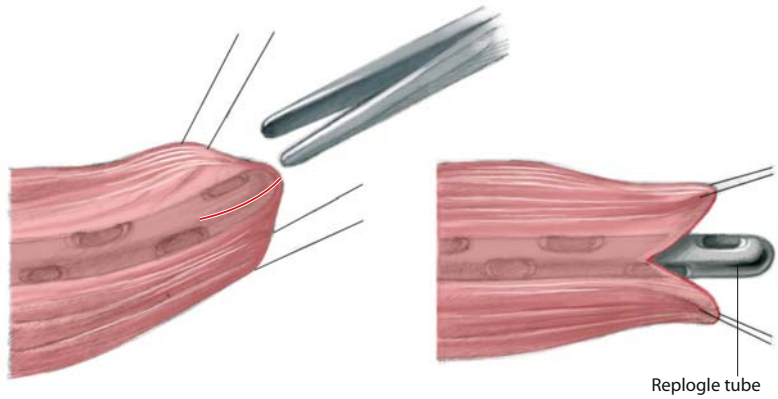


Figure 5.13, 5.14

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 muscular tissue together with the mucosal layer. The latter tends to retract upwards in the upper pouch as soon as it 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 transanastomotic splint for drainage of saliva.

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.

Figure 5.15, 5.16

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 according to Livaditis. 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 and 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, either in a circular or in a spiral fashion. The mucosal layer of the upper pouch is rather thick so that mucosal tears can usually 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 (outpouching of the mucosa through the established gap in the muscle layer) after circular myotomy has been described.

Figure 5.13

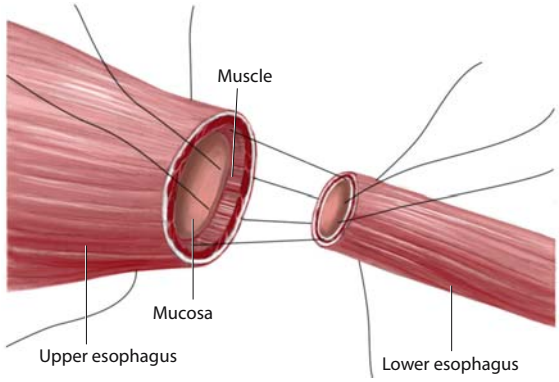


Figure 5.14

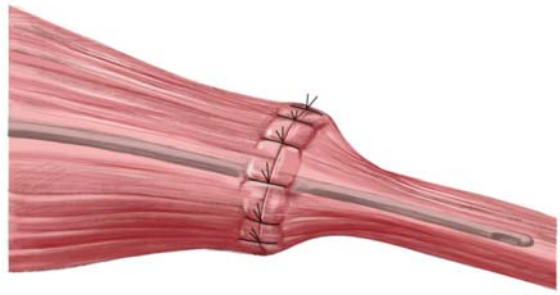


Figure 5.15

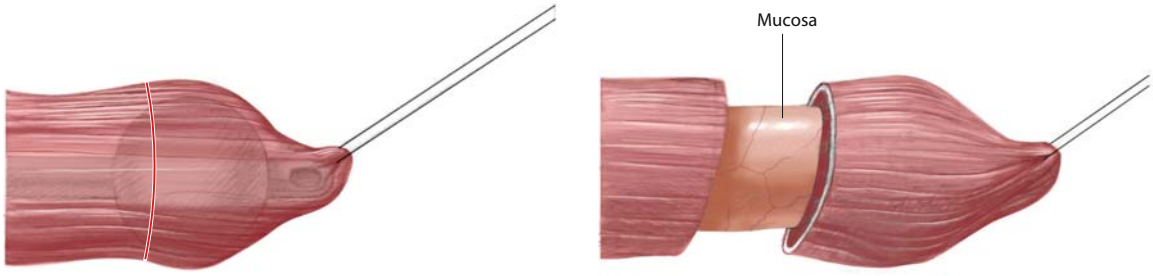


Figure 5.16

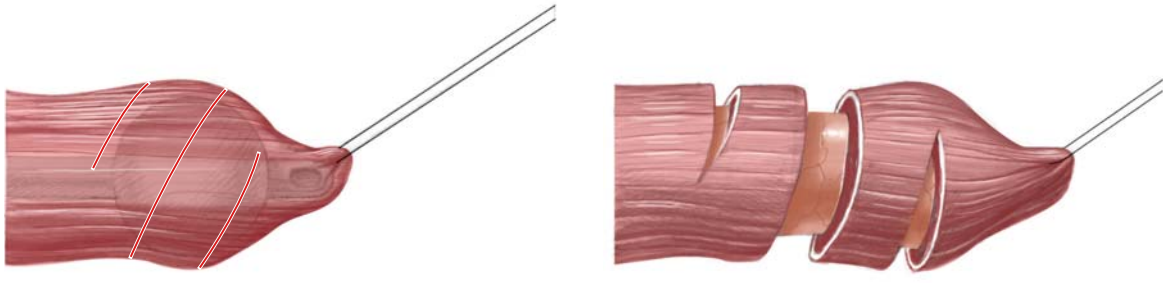


Figure 5.17, 5.18

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.

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 placed near the anastomosis. Before closure, the lungs are fully expanded by forced ventilation until all collapsed regions are well aerated again.

The ribs are approximated with two or three pericostal sutures. 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. In those cases in whom wound margin adaptation remains unsatisfactory, a continuous subcuticular monofilament 5/0 suture is applied, which is pulled after a few days.

Figure 5.17

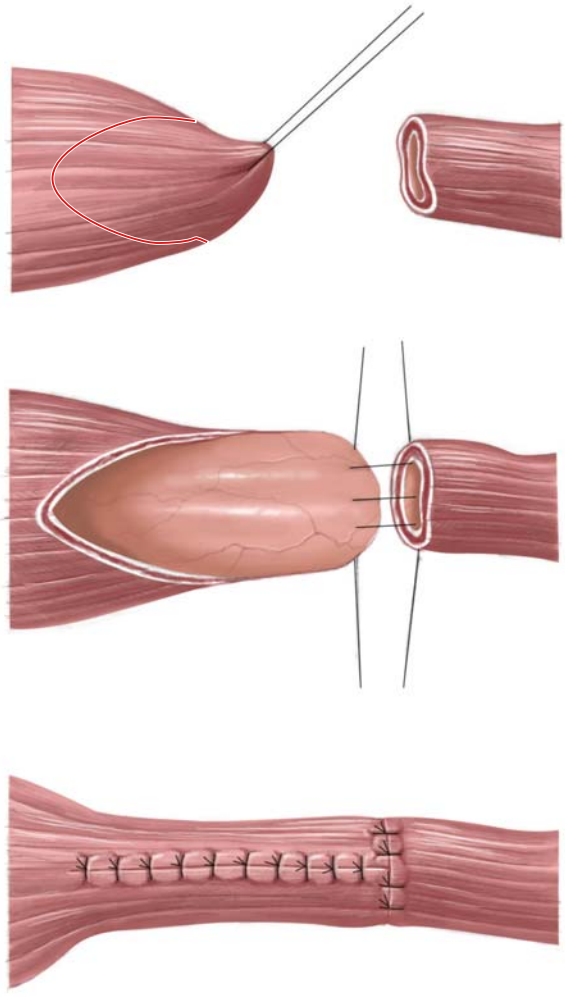


Figure 5.18

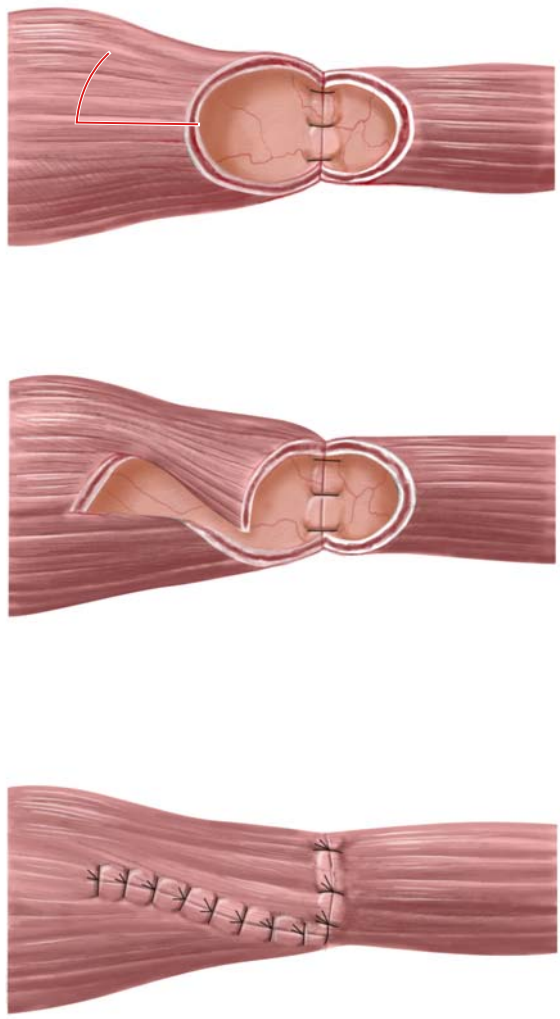


Figure 5.19

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 due to the long distance between the oesophageal pouches.

Two basic surgical strategies are available in cases of long-gap oesophageal atresia: either preservation of the patient's own oesophagus or oesophageal replacement. Three opinions exist concerning preservation and delayed repair of the native oesophagus in the absence of a lower fistula. The first is to await spontaneous growth, which is more pronounced in the upper stump. As experience tells us, it takes 8–12 weeks on the average until a safe anastomosis is feasible. Second, one can attempt to promote elongation of the upper oesophageal segment by regular longitudinal stretching. Third, approximation may be further accelerated by additional bouginage of the lower pouch. The latter is our preferred method, permitting one to anastomose the two segments after 3–5 weeks.

A primary gastrostomy is essential for enteral feeding in all long-gap oesophageal atresia cases. It is also used for estimation of the length of the gap as well as for the distal elongation manoeuvre.

A transverse incision is made in the left epigastric area at a level midway between umbilicus and costal angle. We favour a Stamm gastrostomy with two circular 3/0 absorbable purse-string sutures close to the gastric angle on the lesser curve. The stomach wall is incised in the centre of the purse-string sutures. If stretching of the lower pouch is not desired, a proper gastrostomy tube is introduced, the purse-string sutures are tied and fixed to the parietal peritoneum within the incision. If, however, a longitudinal bouginage from above and below is planned, a jejunostomy for feeding is fashioned in the first jejunal loop with a separate exit below the abdominal incision and with a single 3/0 purse-string suture that is anchored on the internal aspect of the abdominal wall. The feeding tube is advanced deep into the jejunum. Enteral feeding may be started after 24 h.

Figure 5.20

If mechanical elongation of the lower pouch is planned, the gap is assessed in the following way: a 8F–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 anaesthetist introduces a radio-opaque device into the upper pouch. Both probes are pushed towards 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, more forceful pressure in the upper pouch. Leaving the manoeuvre in the same experienced hands throughout has saved us from ever causing a perforation. 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.

Figure 5.19

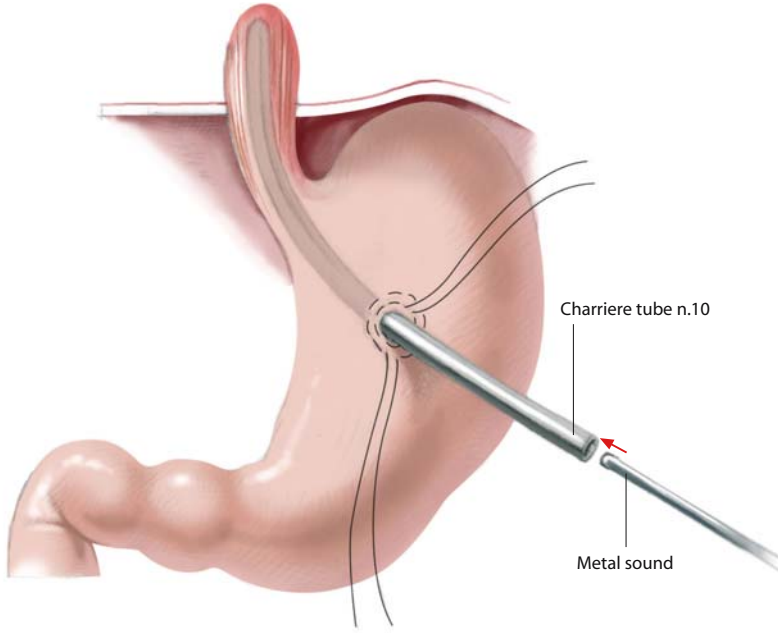


Figure 5.20

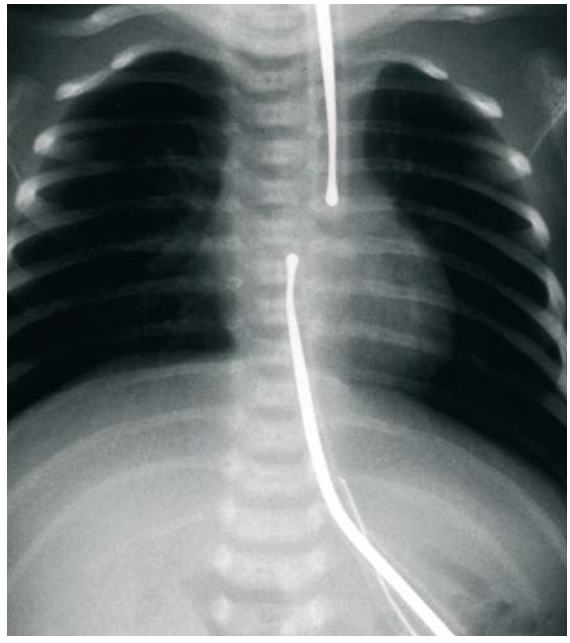


Figure 5.21

H-type fistulas without atresia account for about 3% of the tracheo-oesophageal anomalies. Presentation is usually more protracted and sometimes 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 trachea and oesophagus which are situated medial and posterior to 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 trachea and oesophagus close to the fistula. It must be clearly identified and protected from any injury.

Figure 5.22–5.24

The plane between 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.

Figure 5.21

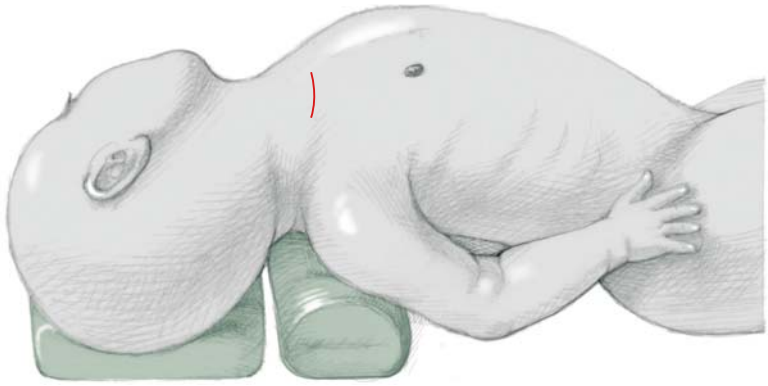


Figure 5.22

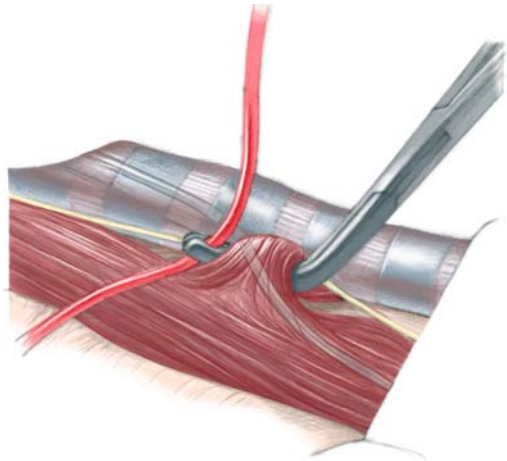


Figure 5.23

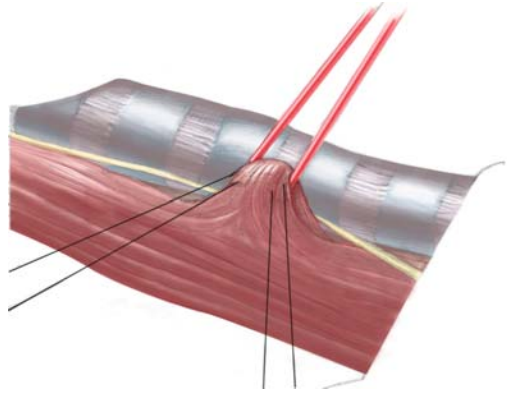
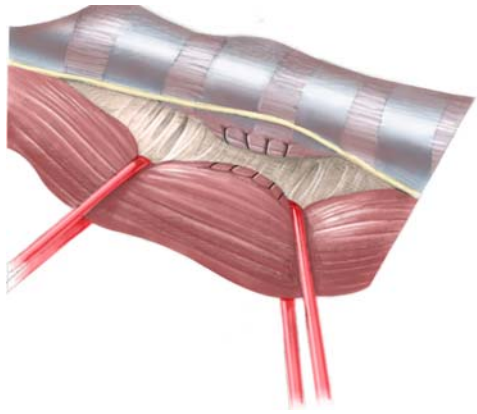


Figure 5.24



CONCLUSION

The first successful primary repair of an oesophageal atresia was achieved by Cameron Haight in 1941. Mortality remained, however, high in the following decades. The outcome was influenced by birth weight, severity of additional malformations, and development of aspiration pneumonia due to delayed diagnosis. Nowadays, the diagnosis is, in most cases, 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. Thus, severe associated anomalies have become the main factor determining outcome. While basic surgical management has become uniform for the most common type of oesophageal atresia with a lower fistula, the best strategy for babies with long-gap oesophageal atresia has remained 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 propagate 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 sequel. 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 may be iatrogenically worsened by damage of vagal nerve fibres during dissection of the distal oesophageal pouch. However, severe swallowing problems with dysphagia are rare, but impaction of foreign bodies, most often bread, meat or fruit pieces, may be partially attributable to the motility disorder.

An anastomotic stricture can be either 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 due to the high incidence of gastro-oesophageal reflux disease that exceeds 40% in patients with oesophageal atresia.

Atypically shaped cartilaginous C-rings and a wide intercartilagineous membrane within the region of the former fistula may be underlying causes of another common complication: tracheomalacia with an incidence around 20%. The anterior-posterior diameter of the trachea is reduced and 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, however, yet to be found and long-term results are awaited.

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