REVIEW ARTICLE

The Surgical Correction of Congenital Deformities

The Treatment of Diaphragmatic Hernia, Esophageal Atresia and Small Bowel Atresia

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SUMMARY

Background: More than half of all congenital deformities can be detected in utero. The initial surgical correction is of paramount importance for the achievement of good long-term results with low surgical morbidity and mortality.

Methods: Selective literature review and expert opinion.

Results: Congenital deformities are rare, and no controlled trials have been performed to determine their optimal treatment. In this article, we present the prenatal assessment, treatment, and long-term results of selected types of congenital deformity. Congenital diaphragmatic hernia (CDH) affects one in 3500 live-born infants, while esophageal atresia affects one in 3000 and small-bowel atresia one in 5000 to 10 000. If a congenital deformity is detected and its prognosis can be reliably inferred from a prenatal assessment, the child should be delivered at a specialized center (level 1 perinatal center). The associated survival rates are 60–80% after treatment for CDH and well over 90% after treatment for esophageal or small-bowel atresia. Despite improvements in surgical correction over the years, complications and comorbidities still affect 20–40% of the treated children. These are not limited to surgical complications in the narrow sense, such as recurrence, postoperative adhesions and obstruction, stenoses, strictures, and recurrent fistulae, but also include pulmonary problems (chronic lung disease, obstructive and restrictive pulmonary dysfunction), gastrointestinal problems (dysphagia, gastro-esophageal reflux, impaired intestinal motility), and failure to thrive. Moreover, the affected children can develop emotional and behavioral disturbances. Minimally invasive surgery in experienced hands yields results as good as those of conventional surgery, as long as proper selection criteria are observed.

Conclusion: Congenital deformities should be treated in recognized centers with highly experienced interdisciplinary teams. As no randomized trials of surgery for congenital deformities are available, longitudinal studies and registries will be very important in the future.

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Correcting congenital deformities is a challenge for pediatric surgeons. Up until 1940, successful surgical repair of congenital deformities was rare; the techniques of pediatric anesthesia and neonatal and pediatric intensive care were inadequate. The enormous progress made since then in pediatric intensive medicine and anesthesia, and improved surgical techniques, make it possible today to repair almost any malformation, and patient survival (with the exception of those with congenital diaphragmatic hernia) is almost taken for granted (1, 2). The requirement in terms of the quality of the repair, both on the part of those affected and of their doctors, has moved on from simple survival to improved quality of life (2–4, e1, e2). An essential prerequisite for this is multidisciplinary teams and standardized follow-up continuing into adulthood (3, 5). Malformations are rare; evidence-based treatment recommendations above the level of expert opinion do not exist (1, e3). With low case numbers, randomized studies are problematic and difficult to carry out for statistical reasons as well as for ethical reasons, because surgical correction is the only way to ensure survival (4–7). Reliable longitudinal studies only describe small cohorts (1, 3, 4, 6, 7, e3). Results based on registry data show continuous improvement in the quality of care accompanied by a fall in treatment complications and comorbidity in high-volume centers, thus reinforcing the demand for centralization (1, 3, 4, 6, 7, e3).

The present review, which is based on an up-to-date selective literature review (to December 2014), describes the prenatal diagnosis, treatment, and long-term outcome of selected congenital deformities.

Methods

The present article deals with the main principles of current methods of treatment and relates only to congenital diaphragmatic hernia (CDH) and esophageal and small-bowel atresia. It is the result of a selective literature search (focused on the years 2010–2014) and includes the authors' own experiences. The analysis is based on expert opinions and takes into account recent results in prenatal diagnosis, treatment, and long-term outcomes. Any relevant meta-analyses found were included in the analysis.

TABLE 1

Prognosis of congenital diaphragmatic hernia from GW 32 (from [6])

Prognostic values for the pregnancy from GW 32 onwards	
LHR (ultrasound) reference value	LHR 1.8–3.0
Probability that ECMO will be required	LHR <1.2
Probability of death (100%)	LHR <0.9
Bilateral lung volume on MRI	Volume 70 mL
Probability that ECMO will be required	Volume <25 mL
Probability of death (100%)	Volume <9 mL

GW = gestational week
LHR = lung-to-head ratio
ECMO = extracorporeal membrane oxygenation
MRI = magnetic resonance imaging

Pathogenesis of congenital deformities

Behind every malformation lies a genetic disorder, which is usually never completely clarified (6, e4–e7). Since most deformities are sporadic, the risk of recurrence in consecutive pregnancies is low (e2). The lack of evidence for Mendelian genetic transmission, coupled with indications that environmental or epigenetic factors may be in play, suggest a multifactorial process (e8). In 20% of cases at most, malformations are associated with syndromes (8, e8–e10). No environmental influences have been described for the malformations discussed in this article.

Prenatal diagnosis

The ultrasound scans around gestational weeks (GW) 10 and 22 have a screening function for congenital deformities and allow the place and mode of delivery to be decided. A qualified prenatal sonographer (at least DEGUM [Deutsche Gesellschaft für Ultraschall in der Medizin] level II) can identify direct or indirect indicators of deformities early on, and these must be very closely followed over the course of the pregnancy (6, 9, e8, e11, e12). Important signs relating to the three deformities are a low lung-to-head ratio (LHR), a small or absent gastric sac associated with polyhydramnios, and a distended gastric and duodenal sac or small-bowel dilatation transitioning into a hypoplastic large bowel. An MRI scan of mother and fetus will provide a greater level of detail (6, 9–12, e13). Interdisciplinary advice and counseling of the parents about the extent of the malformation, treatment options, and prognosis are standard medical practice (13). Alternative treatment methods and intrauterine interventions, if any, are discussed within the team and with the parents, and initiated if necessary (4, 6, e14). Delivery should if possible take place in a center with demonstrable experience in the treatment of the deformity, in order to allow the best possible repair (4).

Congenital diaphragmatic hernia

In 80% of cases, CDH is a left-sided defect in the diaphragm with prolapse of abdominal organs into the thoracic space and, if the defect is large, pronounced ipsilateral pulmonary hypoplasia (e4). Pulmonary hypoplasia, a hypoplastic left heart ventricle (in patients with large defects), and persistent pulmonary hypertension of the newborn (PPHN), with retention of the fetal circulation and a right–left shunt via the patent ductus arteriosus, lead to marked respiratory distress (13). Despite treatment in intensive care, survival is only 40% to 60% worldwide, or up to 80% in specialist centers (4, 7). Early intrauterine diagnosis is important (*Table 1*). Prognostic factors associated with a poor outcome are:

- Early detection (<GW 25)
- Intrathoracic parts of the liver
- Small lung volume
- Poor ventricular function
- Low birth weight (4, 7, 13).

Prenatal diagnosis can anticipate the need for specialist treatment and thus for delivery in a specialist center (9, 13, e15). Attempts at fetal surgical repair remain unsuccessful. On the other hand, attempts have been made to stimulate lung growth by trapping the pulmonary fluid secretions inside the lungs through temporary blockage of the fetal trachea using a minimally invasively introduced balloon (fetal endoscopic tracheal occlusion, FETO). The timing of the intervention, duration of tracheal occlusion, and the value of the procedure are still under debate (1, 6, 14, e15–e17).

Embryology, development, and incidence

The diaphragm develops between GW 4 and GW 8 by the formation of pleuroperitoneal folds in the coelom. This process takes longer dorsally and on the left side, and for this reason the defects are dorsolateral in more than 90% of cases and left-sided in more than 80%. Maternal vitamin A deficiency significantly increases the incidence of CDH, although the exact retinol/cholesterol pathway is unclear (15, e4–e9).

The prevalence of CDH is around 1:3500 births (200–250 babies per year in Germany). A hidden mortality rate (stillbirths) of 34,9% has been assumed (e18). Seventy percent are sporadic, 18% occur as part of multiple deformities, and 10% occur as part of a syndrome (13).

Clinical symptoms, diagnosis, and initial treatment

The main symptom is acute respiratory distress. Because air passes into the gastrointestinal system (meaning that mask ventilation is absolutely contraindicated), the hypoplastic lung does not expand and the respiratory distress rapidly intensifies (13). Other symptoms include a small abdomen, cyanosis, and signs of pulmonary hypertension. Chest radiograph shows the enterothorax with mediastinal displacement and pulmonary hypoplasia (*Figure 1*). Intestinal loops must not be mistaken for cysts and punctured (15).

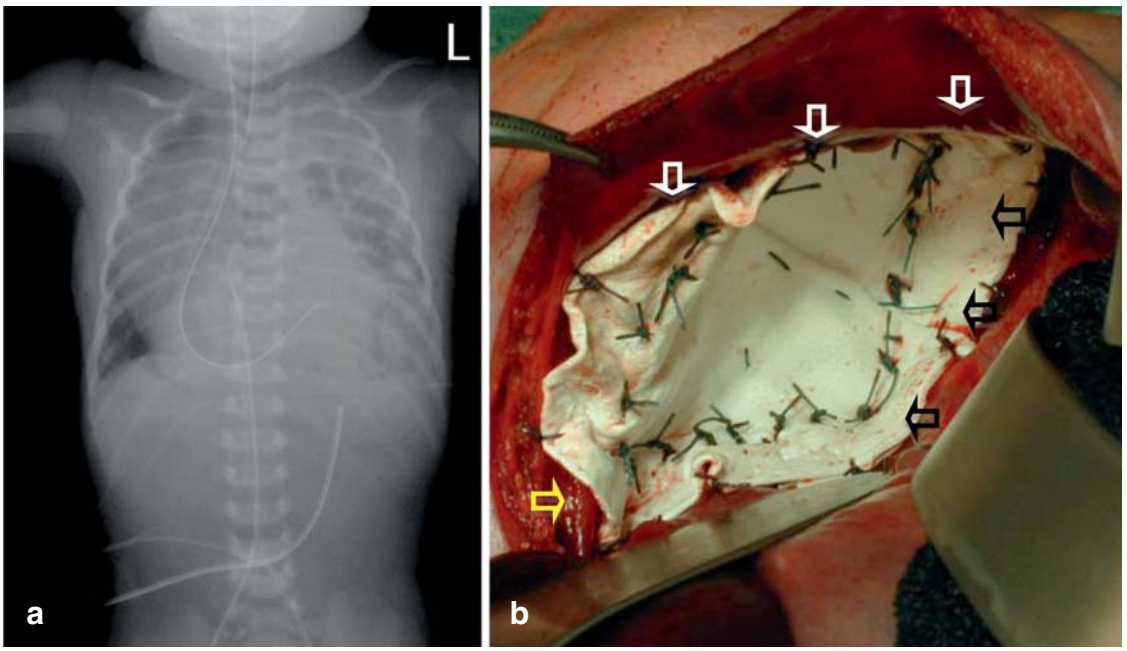

Figure 1: Radiological and intraoperative appearance of left-sided congenital diaphragmatic hernia.
a) Postnatal chest radiograph of a neonate with left-sided congenital diaphragmatic hernia. Note the left-sided enterothorax due to the defect in the diaphragm, with a barely visible left lung (hypoplasia), and the intrathoracic location of the gastric tube.
b) Intraoperative appearance after closure of a large, overlapping conical GoreTex patch. The conical shape has decreased the thoracic dead space and increased the abdominal space, reducing the risk of abdominal compartment syndrome. At the anterior margin the ventral diaphragmatic rim can be seen (white arrows, and medially the left crus of the diaphragm is visible attached to the esophagus (yellow arrow). Dorsally, the patch is attached around the ribs (black arrows)

Neonatal intensive care follows the guideline published by the EURO-CDH Consortium (a team of experts from large European centers with at least 10 cases a year) (13). If all neonatal ventilation options have been attempted and oxygenation is still inadequate with persistent hypercapnia and PPHN, extracorporeal membrane oxygenation (ECMO = artificial lung) must be considered (13). Surgical repair is performed only after hemodynamic stabilization has been achieved (4, 13).

Surgical repair

The technique used for surgery (open vs. thoracoscopic) depends on the size of the defect and on whether ECMO or some other form of ventilation is being used (1, 16). In babies with large defects where the diaphragmatic rim has remained incomplete, closure with a patch is obligatory and the risk of recurrence fundamentally higher (4, 15, 17, e19). The cardiopulmonary stability of a baby that has been on ECMO or is on high-frequency oscillatory ventilation (HFOV) is too poor for thoracoscopic repair to be carried out (1, 4, 17). The advantage of the abdominal approach is that the entire diaphragm remnant is in view, enabling secure anchoring of a patch. GoreTex Dualmesh has become internationally accepted as the best material for a patch (4, 7, 15, 16). Using absorbable material increases the recurrence rate significantly and is not recommended (1, 4, 6, 15, 17). The use of a conical patch reduces the dead space in the chest, increases the abdominal space, and reduces the recurrence rate (16). The advantage of thoracoscopy is the near-absence of scarring, but the disadvantage is the increased recurrence rate, up to 15% (1, 4, 6, 15, 17, e20).

Results and long-term outcome

Defects are classified by the Boston scale into four groups (A to D) and the associated mortality rates rise with the severity of the defect or concomitant malformations (from 2% in group A up to 61% in group D with concomitant malformations) (7, e21). Limiting variables for survival are defect size and PPHN; however, treatment complications, concomitant disease, and chronic lung disease continue to cause late morbidity into adulthood (*Table 2*) (2, 3, 12, 18–20, e22–e44). Published data regarding recurrences are heterogeneous. Often, only the neonatal period or the first 6 months of life are considered, although 20% of recurrences occur after the second year of life (4, 7, 16, e21).

Follow-up studies in children who underwent ECMO show cerebral morphological changes in two-thirds of the children (MRI) and a neurological deficit in 20% (e45). Longitudinal studies show airway disease, psychomotor retardation, chest deformities, gastroesophageal reflux, and failure to thrive persisting into adulthood (18, 19, e22, e46).

TABLE 2

Quality of life and morbidity of children and adults after repair of congenital diaphragmatic hernia, esophageal atresia, duodenal atresia, or an abdominal wall defect (after [2])

Malformation	Mortality	Residual morbidity	Data in %	References
Congenital diaphragmatic hernia	20–40%	Persistent pulmonary hypertension	5–20%	(11, 12, 17, 20, e43)
		Pulmonary hypoplasia	Up to 80%	(4, 15, e1)
		Lung function disorders (obstructive, restrictive, and mixed)	40–85%	(1, 2, 4, 11, 12, 15, 18, 19, 27, e4, e25, e27, e29, e46)
		Recurrence due to patch separation	5.4–50%	(4, 16, 17, e2, e21, e23, e36–e38, e41, e77)
		Chylothorax	4.6%	(e40)
		Failure to thrive	>60%	(19, 27, e35, e37, e42)
		Chest wall deformities	30%	(27, e37–39)
		Emotional and behavioral disturbances	Up to 80%	(18, 19, e23–e26, e29, e33, e41, e46)
Esophageal atresia	5–9%	Esophageal strictures	Up to 49%	(33, 34, e48, e53–e55, e60–e62, e67, e78)
		Esophagotracheal fistula recurrence	Up to 4%	(e56, e57)
		Impaired esophageal motility	100%	(e64, e65, e79)
		Gastroesophageal reflux, Barrett esophagus with risk of malignancy	Up to 50%	(29, e48, e53–e55, e63–e70, e78)
		Lung function disorders (obstructive, restrictive, and mixed)	Up to 43%	(28, e34, e53, e54, e80, e81)
		Recurrent upper airway infections	Up to 80%	(e44, e54, e80, e82, e83)
		Tracheomalacia	Up to 80%	(e34, e54, e80, e82, e83)
		Emotional and behavioral disturbances	Up to 80%	(e28–e31, e63–e65)
Small-bowel atresia	<5%	Bowel strictures	<5%	(36)
		Motility disorders	<5%	(37, 39, 40)
		Otherwise like the normal population	100%	(38, e76)
Abdominal wall defect	<5%	Concomitant malformations	Up to 30%	(e32, e84, e85)
		Otherwise like the normal population	100%	(e32, e84, e85)

Inclusion criteria for this review were: (1) original articles studying long-term outcomes and quality of life of children and adults that (2) were published in English between January 1990 and December 2014 and (3) were accessible on PubMed or the internet

Esophageal atresia

Esophageal atresia (EA) involves an interruption of the continuity of the esophagus, with a blind pouch in the upper mediastinum and associated esophagotracheal fistula (90% Vogt IIIb). In the German-speaking countries, the Vogt classification dating from 1929 is widely used.

Within the past 50 years, mortality has been reduced from 60% to between 5% and 9% (21, e46). The first successful repair was carried out by Haight in 1941 and the first successful thoracoscopic repair by Lobe in 1999 (e47–e49). Prenatal diagnosis reveals up to 50% of cases of esophageal atresia (polyhydramnios with small gastric sac). From GW 28 onwards, imaging will show the upper blind pouch with the help of MRI. In 44.7% of cases this is an isolated malformation, in 9.6% of cases it occurs as part of VACTERL, and in 31.6% it is accompanied by other deformities. Chromosomal anomalies are found in 8.3% of those affected (8). The VACTERL association involves the co-occurrence of at least three of the following deformities:

*V*ertebral anomalies, *a*nal atresia, *c*ardiac defects, *t*racheoesophageal fistula and/or *e*sophageal atresia, *r*enal anomalies, and *l*imb defects (8, 22).

Statistically significant factors for risk stratification of mortality in a series of 4168 cases of esophageal atresia were:

- Birth weight <1500 g (OR = 4.5)
- Surgery on first day of life (OR = 3.8)
- Gestational age <GW 28 (OR = 2.2), and
- Presence of ventricular septal defect (VSD) (OR = 3.8) (21).

Embryology, development, and incidence

Esophageal atresia results from failure of the esophagus to separate from the trachea during GW 3; exactly how this occurs is not clear (e8, e10).

The incidence is 1:3000 (corresponding to 220 cases of esophageal atresia per year in Germany). In 50% to 70% of those affected, concomitant malformations are present.

Clinical symptoms, diagnosis, and initial treatment

Postnatally, neonates show frothing at the mouth and nose because they are unable to swallow the saliva. Aspiration and pneumonia result. Chest radiograph shows a gastric tube looped inside the blind pouch in the upper esophagus and an air-filled gastrointestinal tract in the presence of a (distal) esophagotracheal fistula. If fistula is absent, the abdomen is free of air (Vogt type II). Contrast imaging is not necessary. Clinical and radiological diagnostic investigations (abdominal ultrasound, echocardiography) will mostly identify any important concomitant malformations (cardiac, abdominal, renal, and extremities). The upper blind pouch is continuously suctioned. Repair is undertaken on the 2nd to 4th day of life after vital signs have been stabilized, taking account of any chromosomal and cardiac anomalies.

Surgical repair

Preserving the original esophagus is the most important priority and is successfully achieved in 90% of all cases by primary anastomosis with closure of the esophagotracheal fistula. The preferred approach is through a right-sided thoracotomy (23). In the past 10 years, thoracoscopic repair has become standard in specialized centers (*Figure 2*). An international registry study showed that 10% of all cases are corrected by minimally invasive surgery and show treatment advantages compared to conventional surgery (*Table 3*) (24–27, e50–e52). However, no randomized prospective study has been carried out on this.

Selection criteria for minimally invasive repair (with the aim of reducing morbidity and avoiding conversion to open repair) are birth weight <2000 g, long-gap esophageal atresia, and concomitant severe cardiac malformations. In addition to cosmetic advantages, thoracoscopy offers reduction of the morbidities associated with thoracotomy (rib fusion, scoliosis, winged scapula) (e51). The possible comorbidities of CO_2 insufflation, leading to acidosis and cerebral damage, need to be evaluated (28). Several centers are investigating this question prospectively.

Placement of a transanastomotic tube, oral feeding starting between postoperative day 2 and 5, and contrast study on day 7 are recommended. Important complications are listed in the *Box*. Reoperation is required in 12% of patients.

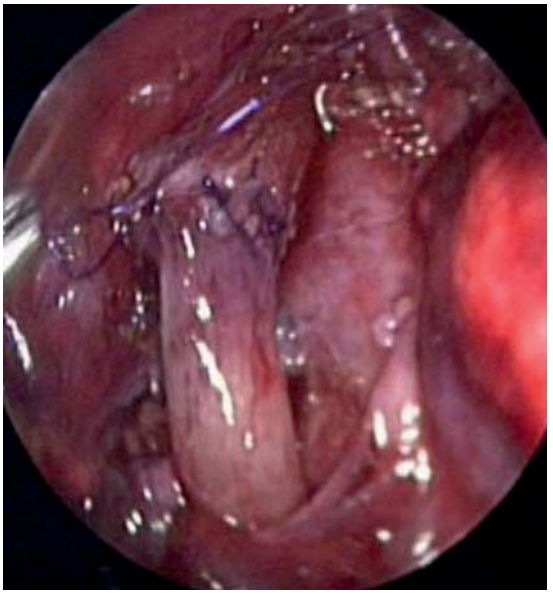

Figure 2: Thoracoscopically completed anastomosis in a child with esophageal atresia. Note the completed esophageal anastomosis; the purple sutures can be seen. Above and to the right, the esophagotracheal fistula oversewn at the trachea is visible

Long-gap esophageal atresia

The definition of long-gap esophageal atresia varies, and the distance between the upper and the lower blind pouch is given in centimeters (>2.5 cm) or number of thoracic vertebrae (>2). This is the form of esophageal atresia for which the debates over treatment are greatest, ranging from delayed anastomosis to esophageal replacement. Delayed anastomosis can be achieved through spontaneous growth of the ends of the esophagus or by means of an "elongation procedure." The most controversial of these at present is the Foker method (transthoracic traction of both blind pouches). This sometimes requires several thoracotomies; the

TABLE 3

Conventional versus minimally invasive repair of esophageal atresia

Study (year)	Leak after thoracotomy	Stricture after thoracotomy	Leak after thoracoscopy	Stricture after thoracoscopy
Lugo (2008)	14.3%	14.3%	19.2%	50%
Al Tokhais (2008)	17.4%	8.7%	13.6%	18%
Allal (2009)	0	21.4%	0	23.5%
Szavay (2011)	4%	0	3.1%	0
Total	8.7%	8.7%	9.3%	21.6%

Comparative meta-analyses of conventional (n = 97) versus thoracoscopic (n = 69) repair of esophageal atresia.
Operative times were similar (147 ± 20 minutes vs. 136 ± 31 minutes, p = ns), from (24)

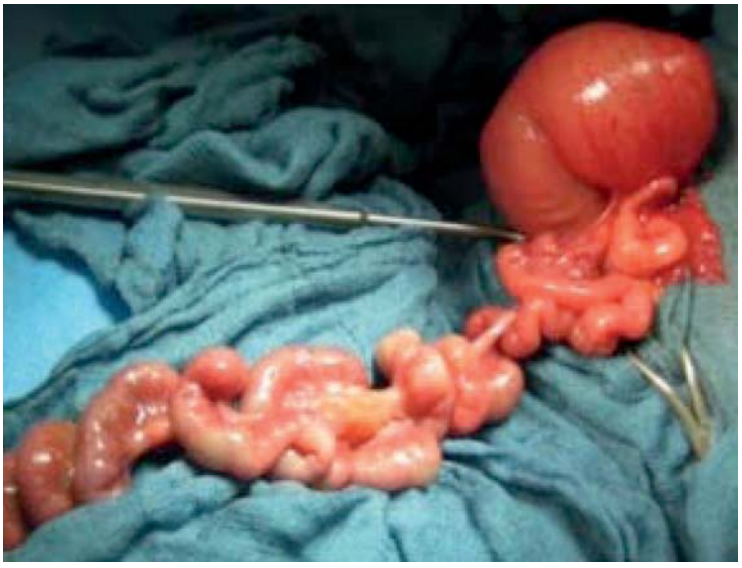

Figure 3: Appearance of type IIIb (apple-peel) small-bowel atresia. Children with apple-peel malformation (IIIb) and multiple atresias (IV) have the poorest prognosis. Note the central vessel around which the small bowel winds like pared apple peel, making bowel perfusion very fragile

BOX

Significant complications of surgical repair of esophageal atresia

- Anastomotic stricture (9% to 45% [29, e48, e53–e55])
- Recurrent esophagotracheal fistula (0% to 4% [e56, e57])
- Gastroesophageal reflux (22% to 50% [29, e44, e53])
- Anastomotic leak (0% to 30% [23, e57])

All these are usually treated conservatively.

main problems are mediastinitis, leakage, and anastomotic stricture (30, 31).

Various techniques are available as alternatives to elongation for esophageal replacement (gastric pull-up, colon or small-bowel interposition [32, e58]). Each of these methods has associated complications and comorbidities. At present, gastric pull-up is the method of choice. Due to numerous severe complications, the "gastric tube" surgical method is no longer used. Colon interposition is problematic because it leads to functional problems and refractory halitosis. Ileum interposition can have a good functional result but is extremely demanding technically (32, e59).

Results and long-term outcome

Anastomotic strictures are associated with anastomoses sutured under tension or with leaks, and are the most frequent problem (9% to 45% of cases) (29, 33, 34, e48, e53–e55, e60, e61). The wide range of incidence rates is due to the fact that esophageal atresia takes many forms and treatment approaches differ. Intermittent dilation (1 to 15 dilations, with a 0.1% to 0.4% risk of esophageal perforation) is the treatment of choice and is successful in 58% to 96% of cases; 50% of all dilations show lasting success in the first 6 months. In 30% of cases multiple dilations are necessary because of persistent stenosis. If success has still not been achieved after 10 dilations, surgical reintervention is recommended (e60). To treat therapy-refractory strictures, local application of mitomycin-C and esophageal stent placement are still under debate (33, 34, e61, e62).

Long-term studies have documented dysphagia and gastroesophageal reflux (29, e63–e68). Forty percent of all children need secondary fundoplication (e69). Twenty percent of all patients show metaplasia (Barrett esophagus) (e70). Currently, only eight cases of esophageal carcinoma after repair of esophageal atresia have been described (e70). Pulmonary symptoms often persist (*Table 2*) (29, 35, e63–e67). Satisfactory long-term results occur even after a complex clinical course or esophageal replacement (2, 3). Gastric pull-up shows the best results ((32, e59, e71, e72).

Small-bowel atresia (duodenum/small bowel)

Congenital defects of the continuity of the small bowel manifest as stenoses or atresia and can usually be diagnosed before birth (e73). The identification of any concomitant malformations has prognostic significance, especially in duodenal atresia (trisomy 21). Cystic fibrosis can occur in association with secondary small-bowel atresia in babies with meconium ileus (e74).

Embryology, development, and incidence

In 95% of cases small-bowel atresia is complete, occurring with a prevalence of 1:5000 to 1:10 000 live births; a third are in preterm neonates. Duodenal atresia occurs in 1:2500 to 1:5000 of neonates (36, 37).

The etiology of small-bowel atresia is unclear. There are two theories about how it arises:

- Failure of the embryonic bowel to recanalize correctly
- Selective obliteration of bowel segments due to vascular insufficiency (37).

For duodenal atresia, another possible cause in addition to failure to recanalize may be lack of rotation of the right pancreatic bud (annular pancreas) (37).

Clinical symptoms, diagnosis, and initial treatment

The typical prenatal presentation of small-bowel atresia shows the dilated stomach and variably dilated bowel loops. Depending on the level at which the obstruction has occurred, the mother experiences a pathological increase in the quantity of amniotic fluid (polyhydramnios) during the pregnancy. Duodenal atresia manifests as a classical widening of the stomach and duodenal bulb ("double bubble") (e73). Newborns with small-bowel atresia present with bile-stained vomit (always

highly pathological) (37). In babies with duodenal atresia the upper abdomen is domed forward and the lower abdomen is flat. In those with small-bowel atresia, the abdomen protrudes; the lower the atresia site, the greater the protrusion. Peritonism indicates a complication such as volvulus or peritonitis (after perforation).

Abdominal radiograph shows the atresia as a typical "double bubble" appearance (duodenal atresia) or multiple air–fluid levels in the bowel (small-bowel atresia). Surgery is performed electively on the 2nd to 4th day of life (36, 37). Relevant concomitant malformations are ruled out or repaired as needed, not least in order to prevent volvulus (malrotation).

Surgical repair

Repair of duodenal atresia requires a right upper abdominal laparotomy (or, optionally, laparoscopy) (e75) and consists of duodenoduodenostomy in the form of a diamond-shaped bypass anastomosis. If a windsock web is present, this is resected with preservation of the major duodenal papilla.

The incision for repair of small-bowel atresia is periumbilical or median. Repair of small-bowel atresia consists in resection of the atretic segment with anastomosis. "Apple-peel" small bowel (*Figure 3*) is a particular challenge, as in this syndrome significant parts of the small bowel are atretic and proper bowel function starts only after a long delay, leading to functional and actual short-bowel syndrome. In children with impaired bowel perfusion, volvulus, meconium ileus, or peritonitis, a double-barreled enterostomy is placed.

Results and long-term outcome

On the whole, the prognosis of small-bowel atresia is good (mortality <10%) (38). Postoperative complications include anastomotic leakage, stenosis, and infections. Prolonged impaired gastrointestinal motility is associated with secondary changes in innervation and the absence of interstitial cells of Cajal (39, 40). The long-term prognosis is determined by concomitant malformations or disease and is significantly poorer in patients with cystic fibrosis. The length of small bowel remaining has relevance for the severity or otherwise of short-bowel syndrome, if present (e76).

Conclusion

High rates of detection during prenatal diagnostic procedures mean that many deformities are identified early. The initial surgical treatment has a significant influence on long-term outcome, and for this reason treatment should if possible always be carried out in centers with demonstrable expertise and multidisciplinary teams, so as to reduce mortality and morbidity to a minimum. There is a role for minimally invasive surgery in carrying out repairs, so long as the selection criteria listed above are observed. Prospective studies—ideally multicenter or registry studies—are needed to provide a research basis for care provision in the future.

KEY MESSAGES

- Congenital malformations are rare.
- Prenatal detection is extremely important and allows the interdisciplinary team time to prepare the parents, explaining the extent of the malformation and its possible treatment and prognosis.
- The quality of the initial surgical treatment strongly affects the long-term outcome.
- Despite the good repair procedures available, in most cases morbidity persists into adulthood.
- Prospective registry studies and health services research are needed if the quality of care is to be further improved.

Conflict of interest statement
Professors Rolle, Wessel, and Fuchs declare that no conflict of interest exists.

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REVIEW ARTICLE

The Surgical Correction of Congenital Deformities

The Treatment of Diaphragmatic Hernia, Esophageal Atresia and Small Bowel Atresia

Lucas M. Wessel, Jörg Fuchs, Udo Rolle

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