general surgery

Department of Pediatrics
Soochow University Affiliated Children’s Hospital
Congenital surgical abnormalities
The term congenital atresia of the esophagus describes a large group of variant malformations that share a defect of the esophageal continuity with or without a fistula to the trachea or to the bronchi.
Oesophageal atresia

Overview

- It is one of the most life-threatening anomalies in a newborn baby and the quality of survival depends on early diagnosis and appropriate therapy.

- The first successful surgery of a 12-day-old female baby was performed by Cameron Haight at the University of Michigan in 1941.
The history of surgical therapy of babies with esophageal atresia after Cameron Haight is a story of success starting with survival rates around 50%, but reaching nearly 100% today when associated life threatening malformations are excluded.
The incidence of an esophageal atresia with or without a fistula is approximately 1 in 3,000 to 4,500 births with a slight preponderance of males in the ratio of 3:2. Most cases occur sporadically.
The most frequent associated anomalies are:

- Musculoskeletal malformations: 20–70%
- Cardiovascular: 20–50%
- Genitourinary: 15–25%
- Gastrointestinal: 15–25%
- Chromosomal anomalies: 5–10%
Oesophageal Atresia (TOF)

VACTERL ASSOCIATION:

V  VERTEBRAL
A  ANAL / ALIMENTARY
C  CARDIAC
T  TRACHEO-OESOPHAGEAL
R  RENAL
L  LIMB
In 87% of cases with esophageal atresia, a tracheoesophageal fistula (TOF), connects the distal, blind-ending esophagus and trachea.
Oesophageal atresia

Presentation

♦ Antenatally:
  • polihydramnios
  • ultrasound (reduced intestinal fluid)
Oesophageal atresia

Presentation

♦ Postnatally:
  • excessive frothy saliva
  • respiratory distress at birth
  • coughing & cyanosis at first drink
  • inability to pass a N/G tube
Oesophageal atresia

Presentation

♦ Antenatally:
  • polyhydramnios
  • ultrasound (reduced intestinal fluid)
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.
Postnatally:

- excessive frothy saliva
- respiratory distress at birth
- coughing & cyanosis at first drink
- inability to pass a N/G tube
Oesophageal atresia

Presentation

Excessive frothy saliva
Recently, fetal magnetic resonance imaging (MRI) has gained more attention for prenatal diagnosis of congenital anomalies.
The H-type variant presents with recurrent aspiration and chest infections in later infancy and childhood.
Oesophageal atresia

Diagnosis

- The diagnostic step is to pass a 12 F (firm and X-ray visible) feeding tube into the stomach. If this is not successful, esophageal atresia is almost certain.

- inability to pass a N/G tube
However, small tubes must be avoided because they may curl up in the upper pouch thereby giving the illusion that they have been pushed forward into the stomach.

If an esophageal atresia is suspected, a physical examination of the entire body is performed to detect further associated malformations.

In this case, a very small feeding tube was used to diagnose esophageal atresia. It resulted in a curling up in the upper esophageal pouch misleading to the diagnosis of a normal esophagus. Therefore, a rather firm and thicker tube has to be used for this procedure.
Oesophageal atresia

Radiological Diagnosis

- The next step is to perform a plain X-ray including the neck, the thorax, and the abdomen. The approximate length of the upper pouch can be estimated by the length of the X-ray visible tube in it. Air below the diaphragm can be seen in the presence of a lower tracheoesophageal fistula and additional fluid levels indicate a duodenal or intestinal atresia.
Esophageal atresia without lower esophageal fistula indicating a long distance between esophageal segments.

A gasless abdomen indicates a pure esophageal atresia without a lower fistula.
Oesophageal atresia

Treatment

- In healthy infants without pulmonary complications, primary repair is performed within the first few days of life.
Oesophageal atresia

Treatment

- Repair is delayed in patients with low birth weight, pneumonia, or other major anomalies. Initially, treat patients conservatively with parenteral nutrition, gastrostomy, and upper pouch suction until they are considered to be low risk.
Oesophageal atresia

**Treatment**

- **Surgical Technique:**
  - bronchoscopy / oesophagoscopy to determine position of fistula
  - primary repair when possible
  - right thoracotomy
  - extrapleural approach
Oesophageal atresia

Treatment

- Repair is delayed in patients with low birth weight, pneumonia, or other major anomalies. Initially, treat patients conservatively with parenteral nutrition, gastrostomy, and upper pouch suction until they are considered to be low risk.
Duodenal atresia

INCIDENCE

Congenital duodenal obstruction (DO) is a frequent cause of congenital intestinal obstruction in the newborn, occurring in 1 per 5,000–10,000 live births, and affecting boys more commonly than girls.
More than 50% of patients with duodenal atresia or duodenal stenosis have associated congenital anomalies, especially Down syndrome, which occurs in about 30% of patients.
Table 39.1 Incidence of associated congenital anomalies (%) 
(N = 1,759 patients)

<table>
<thead>
<tr>
<th>Associated anomaly</th>
<th>%</th>
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<tbody>
<tr>
<td>Down syndrome</td>
<td>28.2</td>
</tr>
<tr>
<td>Annular pancreas</td>
<td>23.1</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>22.6</td>
</tr>
<tr>
<td>Malrotation</td>
<td>19.7</td>
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<tr>
<td>Esophageal atresia</td>
<td>8.5</td>
</tr>
<tr>
<td>tracheoesophageal fistula</td>
<td></td>
</tr>
<tr>
<td>Genitourinary</td>
<td>8.0</td>
</tr>
<tr>
<td>Anorectal</td>
<td>4.4</td>
</tr>
<tr>
<td>Other bowel atresia</td>
<td>3.5</td>
</tr>
<tr>
<td>Others</td>
<td>10.9</td>
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Duodenal obstruction is being increasingly diagnosed prenatally. The prenatal sonographic diagnosis of DO relies on the demonstration of the “double bubble” sign, which is due to the simultaneous distension of the stomach and the first portion of the duodenum. The dilated stomach and duodenum appear side by side across the midline of the upper fetal abdomen.
Ultrasonography (transverse view) of 24-week gestational age fetus showing the “double bubble” sign. S—stomach, P—pylorus, D—duodenum
Maternal polyhydramnios is also a common ultrasonographic finding observed in 20–75% of cases with duodenal atresia, mainly in the second half of pregnancy.

In all cases of combined polyhydramnios and “double bubble” sign, a detailed evaluation for other associated anomalies, especially cardiac anomalies, should be undertaken.

Amniocentesis for chromosomal analysis is helpful for counseling.
Duodenal atresia

Clinical

- About half of these patients are premature and of low birth weight. Vomiting and intolerance of attempted feedings are the most common symptoms and are usually present on the first day of life.
- Since 80% of the obstructions are located in the postampullary region of the duodenum, vomitus in the majority of cases is bile-stained.
An orogastric (OG) feeding tube in an infant with suspected duodenal obstruction typically yields a significant amount of bile-stained fluid.
**Clinical**

- In supra-ampullary atresia, it is nonbilious.
- There is no abdominal distention because of the high level of obstruction.
- Infants may pass some meconium in the first 24 h of life and thereafter constipation may develop.
- Dehydration with weight loss and electrolyte imbalance soon follows if fluid and electrolyte losses have not been adequately replaced.
Incomplete duodenal obstruction (e.g., duodenal membrane with a central aperture) usually leads to the delayed onset of symptoms, which may appear a few months or years after birth.
Duodenal atresia

Imaging Studies

The diagnosis of the duodenal atresia is confirmed on X-ray examination. An abdominal radiograph will show a dilated stomach and duodenum, giving the characteristic appearance of a “double bubble sign” (the stomach and the proximal duodenum are air filled), with no gas beyond the duodenum.
Duodenal atresia

Imaging Studies

Abdominal erect radiograph showing distended stomach and duodenum with a “double bubble” sign with no air beyond the duodenum.
Duodenal atresia
Imaging Studies

In partial duodenal obstruction, a plain film of the abdomen will show a “double bubble” appearance but there is usually some air in the distal intestine. Radiographic findings in the annular pancreas are usually indistinguishable from duodenal atresia and stenosis.
Duodenal atresia and stenosis are treated surgically. In patients with duodenal obstruction, a duodenoduodenostomy is the most commonly performed procedure. Some advocate duodenojejunalostomy, a procedure practiced by few because of its higher risk of long-term complications.
Small-bowel atresia

- Jejuno-ileal atresia has a prevalence rate of approximately 1:330–1:1,500 live births, with a third of infants either born prematurely or small for date.
- Associated chromosomal and extra abdominal anomalies (7%) are well documented but not as common as in duodenal atresias.
Classification

Table 41.1 Types of intestinal atresias seen at Red Cross Children’s Hospital 1959–2007

<table>
<thead>
<tr>
<th>Type</th>
<th>Jejunum</th>
<th>Ileum</th>
<th>Total (%)</th>
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<tbody>
<tr>
<td>Stenosis</td>
<td>21</td>
<td>13</td>
<td>34 (10.7)</td>
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<tr>
<td>Type I</td>
<td>57</td>
<td>15</td>
<td>72 (22.7)</td>
</tr>
<tr>
<td>Type II</td>
<td>18</td>
<td>13</td>
<td>31 (9.7)</td>
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<tr>
<td>Type III(a)</td>
<td>28</td>
<td>24</td>
<td>52 (16)</td>
</tr>
<tr>
<td>Type III(b)</td>
<td>59</td>
<td>0</td>
<td>59 (18.6)</td>
</tr>
<tr>
<td>Type IV</td>
<td>56</td>
<td>14</td>
<td>70 (22)</td>
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<tr>
<td>Total</td>
<td>239</td>
<td>79</td>
<td>318</td>
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</table>
Small-bowel atresia **Symptoms**

Jejunoileal atresia presents with:
- Bile-stained vomiting
- Abdominal distension
- Failure to pass meconium
Bile stained vomiting due to jejunal atresia
Small-bowel atresia

Abdominal radiograph of a newborn with jejunal atresia showing a few dilated proximal small bowel loops.
Abdominal radiograph of a newborn with distal small bowel stenosis showing large air filled intestinal loops.
Small-bowel atresia

Surgical therapy

- The operative management of intestinal atresia is individualized and determined by the pathologic findings, associated abnormalities, the length of the undamaged bowel and the general condition of the infant.
End to end or end to back single layer bowel anastomosis
Meconium ileus occurs when meconium becomes inspissated and obstructs the distal ileum. The condition is usually a manifestation of cystic fibrosis. Conversely, approximately 20% of infants with cystic fibrosis present with meconium ileus at birth. No other data have been reported regarding its incidence.
In meconium ileus, low or distal intestinal obstruction in results from the impaction of thick, tenacious meconium in the distal small bowel. In addition, complications such as ileal atresia or stenosis, ileal perforation, meconium peritonitis and volvulus with or without pseudocyst formation can occur in association with meconium ileus.
Meconium peritonitis
Meconium ileus

Infants with meconium ileus present with vomiting (usually bile stained), abdominal distension, and failure to pass meconium. Pulmonary manifestations, due to cystic fibrosis are not usually present at birth and develop shortly thereafter, typically resulting in a bronchiolitis-like picture. Although meconium ileus in the absence of cystic fibrosis is considered rare, it occurred in 21.6% of newborns in 1 series; these infants had no laboratory or clinical evidence of cystic fibrosis.
Supine outside abdominal film from the first day of life shows several dilated loops of bowel
Gastrografin enema study shows filling defects in the terminal ileum and cecum. Also note the microcolon (transverse and descending colon).
Enema study shows microcolon and contrast material outlining a terminal ileum packed with (meconium) filling defects
Thank You!