



Primary paraesophageal hernia in children

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Abstract

Background: Paraesophageal hernias are uncommon in children and are distinctively different from the more common sliding hiatus hernias and those occurring after antireflux surgery in anatomy, pathology, symptoms, complications, and management. We reviewed a single institution's experience with the pathology of paraesophageal hernias.

Methods: We conducted a retrospective analysis of patient records.

Results: Fifty-nine children with paraesophageal hernias were treated during a 42-year period. Their mean age at presentation was 23.4 months (range, 1 day to 11 years). Presenting complaints were recurrent chest infections (n = 32), vomiting (n = 24), symptomatic anemia (n = 20), failure to thrive (n = 18), and dysphagia (n = 6). Five children were asymptomatic, and their diagnosis was established when they were investigated for incidentally noted asymptomatic anemia (n = 3), scoliosis (n = 2), and mumps (n = 1). Radiology typically showed cystic masses in the posterior mediastinum in the right lower chest and occasionally had an air-fluid level in the cystic mass or a dilated esophagus. None presented with strangulation or hematemesis. All patients were operated on. Surgical findings included a peritoneal lined sac and herniation through a widened diaphragmatic hiatus, containing the stomach and at times the transverse colon, spleen, and small bowel. The hernial sac usually occurred on the right. Principles of surgery included reduction of the contents, partial excision of the sac, crural approximation, and a fundoplication in 39 patients. No fundoplication was done in the earlier years in 20 patients, of whom 12 had recurrent reflux symptoms. Postoperative complications were bowel obstruction (n = 6), intussusception (n = 3), dysphagia (n = 3), breakdown of the repair (n = 3), and pneumothorax (n = 1). There was one mortality caused by preoperative aspiration.

Conclusion: Paraesophageal hernias in children are uncommon and most likely caused by a congenital defect. They are associated with considerable morbidity. Strangulation is not a feature. Principles of repair are well established and should include an antireflux procedure.

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Primary paraesophageal hiatal hernias are uncommon in childhood and are distinctively different from the more common sliding hiatus hernias and those occurring after antireflux surgery in anatomy, pathology, symptoms, complications, and management. These hernias can present at birth and are most likely caused by anatomical predis-

positions. They are usually symptomatic because the mobile components of the stomach have migrated cephalad into the posterior mediastinum through a large sacculated hiatal defect. Organoaxial rotation of the stomach may *theoretically* cause partial or complete gastric obstruction associated with incarceration and complicated by strangulation or mucosal ulceration and anemia. With time, during the evolution of the hernia, the gastroesophageal junction may

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Table 1 Symptom complexes

Symptoms	n
Recurrent respiratory tract infections	32
Vomiting	24
Symptomatic anemia	20
Failure to thrive	18
Dysphagia	6
Early satiety	2
Asymptomatic	
Asymptomatic incidental anemia	3
Scoliosis	2
Mumps	1

also migrate proximally, thereby constituting a combined paraesophageal and sliding hiatus hernia [1].

In contrast to the adult literature on paraesophageal hernias, only few reports on these hernias with an emphasis on etiology, symptoms, and management have been published in the pediatric literature. The principal points of controversy in the pediatric/adult literature concern the etiology, either congenital or acquired, and the need for a concomitant antireflux procedure during surgical repair. This article reports on our analysis of our experience with 59 children with large paraesophageal hernias.

1. Patients and methods

Between 1962 and 2004, 59 children with large paraesophageal hernias were managed at the Red Cross War Memorial Children's Hospital. Excluded from the study were all children with a sliding hiatus hernia, paraesophageal hiatus hernias after antireflux surgery (Nissen fundoplication and 180° anterior oblique fundoplication/Boix-Ochoa fundoplication), and other congenital or traumatic hernias through the diaphragm.

Children were investigated radiologically with plain chest radiographs, upper gastrointestinal contrast studies, and endoscopy. Preoperative abnormalities were corrected, and the hernias were repaired electively.

2. Results

Over a 42-year period, we studied 59 patients. The group consisted of 27 girls and 32 boys with an average age of 23.4 months (range, 1 day to 11 years).

The symptom complexes were varied; these are listed in Table 1. Many of the patients had more than one of the listed symptoms. Only 6 patients were asymptomatic, detected incidentally while being investigated for other illnesses.

3. Investigation

Investigation of these patients was primarily radiologic. Chest radiographs were usually the initial means by which patients were diagnosed (Fig. 1).

The results of these radiographs were reviewed by a pediatric radiologist. Herniated contents were seen in all cases as a cystic mass on the plain chest radiograph. This was found both on the left and the right of the midline in the posterior mediastinum in 47% of the case patients, was only on the right side in 51.4%, and was visualized on the left of the midline in the remaining 1.5%.

The sizes of the cystic mass as measured by the greatest diameter on the lateral chest radiographs were on average 4.7 cm (range, 2.5-9.5 cm). This had minimal correlation to age because many of the biggest cysts were present in the youngest children.

Other features were an air-fluid level in the cystic mass (32%) and a dilated esophagus (31%).

The presence of an isolated paraesophageal hernia and those with an associated sliding component (ie, migration of the esophagogastric junction) were noted with specific

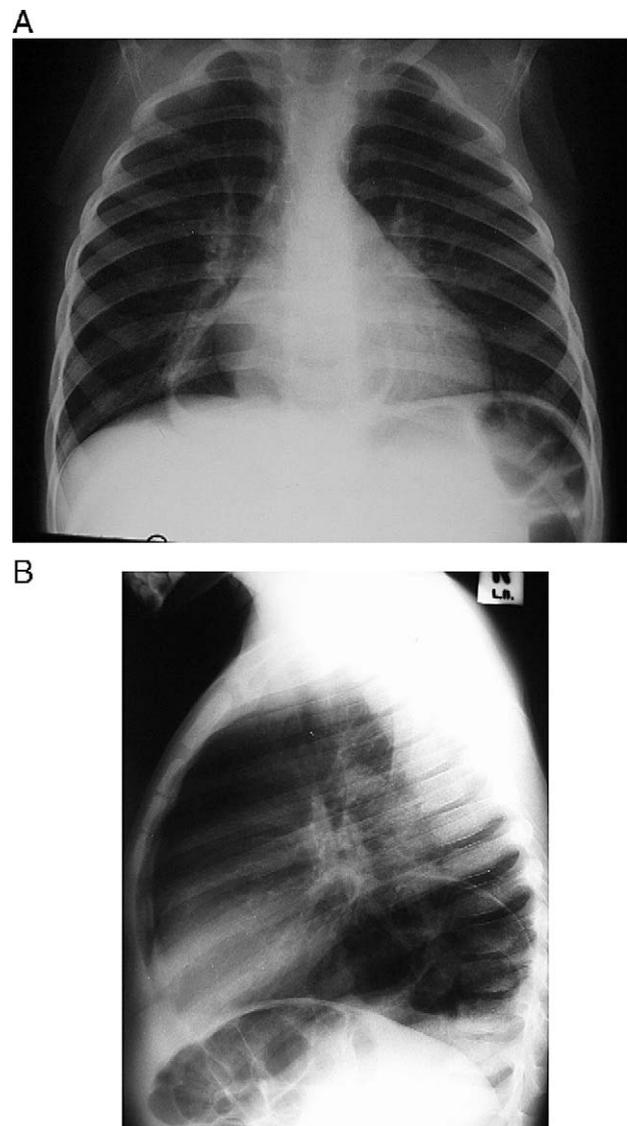


Fig. 1 A, Anteroposterior and B, lateral chest radiographs showing cystic shadow in the right posterior mediastinum.



Fig. 2 Contrast study showing the stomach in the paraesophageal hernia.

reference to patient age. This was done to determine if the former condition occurred in younger children with a progression to the latter condition in older children. The average patient age for isolated paraesophageal hernia occurrence was 2 years 2 months (range, 1 month to 8 years). The combined group had a similar average, 2 years 10 months (range, 2 months to 11 years).

Contrast studies were performed to confirm the diagnosis, which showed a contrast-filled stomach in the posterior mediastinum, often with organoaxial volvulus (Fig. 2). These were not useful in determining the full extent of the hernial contents, which often contained the colon, spleen, and small bowel.

In one patient, a computed tomographic scan was used to investigate a mediastinal mass. It identified a fluid-filled structure in the posterior right mediastinum.

Esophagogastroscopy was only performed in the 6 patients who presented with dysphagia and showed fundal herniation without evidence of gastroesophageal reflux.

Gastroesophageal reflux was not specifically evaluated in these patients once a paraesophageal hernia had been identified. If gastroesophageal reflux was seen on the contrast study, it was noted; however, it did not alter our management because an antireflux procedure is part of our surgical repair.

Differential diagnosis for both solid and cystic masses should be taken into account, which includes a large sliding

hiatus hernia, lung abscess, congenital lung cysts, hydatid disease, pericardial cysts, foregut duplication cysts, contained perforations, and epiphrenic diverticulum.

4. Surgical repair

An abdominal approach was used in all patients, except for 3 who were operated on through a right thoracotomy during the early phase of the retrospective study. Celiotomy either through a midline or through a left subcostal incision adequately exposed the subdiaphragmatic space. After confirmation of the pathology, the left lobe of the liver was mobilized by dividing the left triangular ligament. Reduction of herniated content was always performed with ease, and there was no need to enlarge the hiatal orifice. Contents usually included the stomach and transverse colon, but proximal small bowel and spleen were also found in many instances. A hernial sac was found in all children, which on histology was similar to the peritoneum—differing only in the increased dense connective tissue with a lax muscle tissue component. This was found anteriorly, extending predominantly into the right posterior mediastinum. The sac was opened and excised partially or completely after division of adhesions to the right parietal pleura and pericardium. Identification of the vagal nerves, especially posteriorly, was problematic and often not visualized. The esophagus was always easy to mobilize irrespective of whether the gastroesophageal junction was found cephalad to its normal position. Approximation of the diaphragmatic crura posterior to the esophagus was at times complemented by anterior stitches to the arch of the hiatal defect to prevent undue angulation of the esophagus at this site. No diaphragmatic relaxation incision was required, and no prosthetic graft was used to reinforce the crural repair. An antireflux procedure (Nissen fundoplication, $n = 20$; Boix-Ochoa fundoplication, $n = 13$; transabdominal Belsey, $n = 3$; transthoracic Belsey, $n = 3$) was performed during the initial procedure.

The initial 20 patients treated were managed with reduction of the hernial contents, partial sac excision, and crural approximation but no antireflux procedure. Follow-up of these initial patients revealed significant symptomatic gastroesophageal reflux in 12 (60%) of the case patients, necessitating further management. This is in contrast to

Table 2 Complications

Morbidity	n
Bowel obstruction	6 (1 child \times 2)
Intussusception	3
Dysphagia	3 (2 revised, 1 dilated)
Breakdown of repair	3
Pneumonia	2
Pneumothorax	1
Wound sepsis	2

6 (15%) of the 39 case patients in whom an antireflux procedure was performed at the initial surgery.

5. Complications

There were several complications related to the surgical repair of the paraesophageal hernias (Table 2).

There was only one mortality: in a patient who was in a moribund state on admission and died preoperatively of aspiration pneumonia.

6. Discussion

The typical paraesophageal hernias encountered in infancy and childhood have several components. The defect is within the esophageal hiatus and is lined by a peritoneal sac, usually extending anteriorly and to the right of the esophagus as well as into the posterior mediastinum. The esophagogastric junction is assumed to be in the normal intraabdominal position, and the mobile components of the stomach (fundus, body, antrum, greater curvature, and pylorus) subsequently migrate cephalad through the defect and come to lie predominantly in the posterior mediastinum and right extrapleural thoracic space. During this migrating process, the stomach tends to rotate around its organoaxial axis, which can theoretically lead to partial or complete gastric obstruction between the distal esophagus above and the duodenum below.

The incidence of paraesophageal hernia reportedly varies between 3.5% and 5% of all operated hiatus hernias [2,3]. It is predominantly seen in adults older than 40 years and is prone to complications, such as incarceration, mucosal ulceration, necrosis, and perforation.

In adults, paraesophageal hernia is postulated to be an acquired condition requiring immediate surgical repair because of its high complication rate [1-6]. The severity and incidence of the complication, as quoted in these studies, are based on hospital-based audits and do not account for case patients who may have demised before presenting for surgical repair. The accepted management of these patients is surgical repair, involving hernial reduction, excision of the hernial sac, crural approximation, and an antireflux procedure where indicated [1,6]. The urgency and necessity of asymptomatic paraesophageal hernia repair have been recently questioned, and a wait-and-watch approach has been advocated [7].

However, there are substantial differences between paraesophageal hernias encountered during childhood and those presenting in later life.

Paraesophageal hernias in childhood are thought to be caused by a congenital abnormality that can be explained on an embryologic basis. During the development of the human diaphragm, 2 small coelomic spaces, called pneumoenteric recesses, develop on either side of the midline in the

mediastinum. With the fusion of the sides of the pleuro-peritoneal canals, the larger recess becomes isolated as the infracardiac bursa [8]. The recess on the left side is transitory. Persistence of the recess on the right creates a flattened and elongated mesothelial lined space within the esophageal hiatus [9] (ie, the postulated congenital predisposition for the development of a paraesophageal hernia). The space is approximately 1 cm long at birth and may become obliterated during adulthood [10]. A similar serous sac is found within the porcine esophageal hiatus and is situated to the right and ventral to the esophagus [11]. The development of the recess must be abnormal in the cases under discussion.

Alternatively, an aberration of embryonic development of the lumbar component of the diaphragm, which originates from mesodermal cells around the aorta, may be responsible. The hernias have been diagnosed prenatally and shortly after birth and in siblings, further supporting the theory that these patients have an abnormal anatomical predisposition [12].

We could find no evidence to support the view that these hernias represent large sliding hiatus hernias. Unlike symptoms of sliding hiatus hernias that are primarily a result of deranged physiology, those of paraesophageal hernias are of a mechanical nature. Paraesophageal hernias can surprisingly be asymptomatic (8%); however, significant morbidity was experienced in virtually all our patients. With time, symptoms of postprandial discomfort, breathlessness, nausea, vomiting, and pain may develop. Many of these symptoms are vague in nature, with no guide as to their origin, the diagnosis often made incidentally or on review, in which there is no improvement in the symptoms on empirical treatment. Obstruction caused by gastric torsion can occur at the gastroesophageal junction, mid body of the stomach, or duodenum. Symptoms and signs will be determined by these factors and are due primarily to displacement of the stomach with entrapment of gas and food in the intrathoracic segment. Dysphagia is uncommon. In true anatomical paraesophageal hernia, the esophagogastric junction remains in its normal position, and the sphincter mechanism therefore remains intact. However, with cephalad displacement, the sphincter may become dysfunctional and pathologic reflux will result. Other structures, such as the colon, small bowel, duodenum, and even spleen, may accompany the stomach's migration to complicate the situation even further.

All our reported paraesophageal hernias were clinically incarcerated. None of the case patients was strangulated or presented with evidence of complete bowel obstruction despite the fact that symptoms were present for longer than 6 months. Of the 16 children with iron deficiency anemia, it is assumed that it was caused by blood loss, the consequence of either mucosal ulceration within the incarcerated stomach or reflux esophagitis.

The presence of an abnormality is established by chest radiograph. In our series, all patients had an easily

identifiable cystic mass in the posterior mediastinum. An air-fluid level was seen in 32% of the patients. An upper gastrointestinal contrast study confirmed that the cystic mass was in fact the gastrointestinal tract, thus differentiating it from other causes of a cystic mass. On the chest radiograph, the differential diagnosis of an air fluid-filled cyst includes a lung abscess, sliding hiatus hernia, congenital lung cysts, or hydatid disease. Fluid-filled cysts need to be differentiated from a posterior mediastinal mass, hydatid disease, pericardial cysts, foregut duplication cysts, contained perforations, and epiphrenic diverticulum. Occasionally, a malpositioned nasogastric tube can be observed in the posterior mediastinum.

Diagnosis was always made on a barium meal examination. This showed various degrees of prolapse and distortion of the stomach. However, it was difficult to determine the position of the esophagogastric junction, the length of the esophagus, whether gastroesophageal reflux was present, and the exact anatomical configuration of the prolapsed stomach.

There is a contention that all cases initially began exclusively as paraesophageal hernias defined by a normally positioned esophagogastric junction; with progression and gradual displacement of this junction, a sliding component was introduced and mixed hernia was formed. However, we could not confirm this because the age ranges and averages of the 2 groups were very similar.

Computed tomography is reported to be helpful in that it documents the widened esophageal hiatus; hiatal size and content; the orientation of the stomach; and the hernia's position, to the right, straddling, or more to the left of the midline [13]. We found no use for this as a diagnostic method, and computed tomography was only performed in one patient before referral to our service. Only plain chest radiographs and a contrast study were required to establish the diagnosis.

In the absence of dysphagia, endoscopic examination is not essential in children but may help exclude other disorders and determine the presence or absence of esophagitis. Esophagoscopy should be done immediately before surgery.

Gastric emptying was not assessed preoperatively or postoperatively. It is well established [14] that a large percentage of children with gastroesophageal reflux will have gastric stasis but that most of them would improve postoperatively, thus rendering a concomitant gastric drainage procedure unnecessary. The validity of extrapolating these findings to patients with paraesophageal hernias can be argued, but none of the patients developed clinical gastric stasis postoperatively.

Complications are more prevalent in paraesophageal hernias than in sliding hiatus hernias, and elective repair is advocated on diagnosis, even in asymptomatic cases. Medical therapy is ineffective. This is a mechanical defect that will progress. An abdominal surgical approach is usually preferred. The principles of repair consist of

reduction of the hernia, partial or complete excision of the accompanying sac to prevent recurrence and cyst formation, and crural approximation—which is predominantly posterior but occasionally anterior if the defect is very large—followed by an antireflux fundoplication. The hiatal defect in the diaphragm is in our experience wide at its base and arches over the esophagus anteriorly. Great care should be taken to prevent injury to the vagal nerves during dissection and resection of the sac.

Repair of the crura without the use of prosthetic material should be done if possible. It was not needed in our series because the limbs of the hiatal defect could always be approximated without resorting to its use or releasing incisions in the diaphragm.

We believe that an antireflux operation is an essential component to the repair. Because hiatal attachments are disrupted with the cephalad movement of the stomach and the esophagogastric junction into the posterior mediastinum and after operative sac removal and mobilization of the esophagus, normal physiologic and anatomical mechanisms for the prevention of gastroesophageal reflux are disturbed. The high incidence of gastroesophageal reflux in paraesophageal hernias is well supported in the literature [15]; in a series similar to this, it was found to be 68.4% [16]. Furthermore, the occurrence of symptomatic reflux in 12 (60%) of the 20 patients who did not undergo an antireflux procedure would support the addition of an antireflux procedure to the surgical repair of these patients. A Stamm gastrostomy for gastric fixation was not used in our series although it is advocated by others.

Laparoscopic repair of paraesophageal hernias is now commonly practiced in adults and more recently advocated for infants and children. This approach must be balanced against the traditional method and relative risks involved [17,18].

Paraesophageal hernias in children do not carry the same morbidity and mortality as do those in adults. In the reported children, despite the common findings of incarceration or radiographic organoaxial gastric volvulus, none of the hernias was complicated by strangulation or significant obstruction as reported in the adult literature [1-6]. These studies, like most in the adult literature, only assessed patients presenting for medical care and may have missed patients who died before presentation. Routine postmortem examination of children with sudden death is not undertaken; thus, the stated statements and conclusions can only be based on patients under our care. Despite their relative benign presentation in children, however, these hernias should be electively repaired on diagnosis.

7. Conclusion

Paraesophageal hernias are uncommon in children. They are thought to be caused by a congenital anatomical predisposition, the presence of a persistent right pneumo-

enteric recess. These hernias do not carry the same life-threatening risk of strangulation as seen in adult cases despite the radiographic demonstration of organoaxial gastric volvulus and the operative findings of incarceration. However, they give considerable morbidity and will not resolve with nonoperative management. The operation is best performed via an abdominal approach and includes sac resection and an antireflux procedure.

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