

Congenital paraesophageal hernia in infancy and childhood

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ABSTRACT

Objectives: Congenital paraesophageal hernia is a rare condition in the pediatric age group. The symptomatology of these patients is usually non-specific in the form of repeated attacks of chest infection and/or recurrent attacks of vomiting but can be associated with serious complications such as intrathoracic gastric volvulus.

Methods: Between 1989 and 1997, 6 children with paraesophageal hernia were treated at our hospital.

Results: Six children (4 males and 2 females) were treated for congenital paraesophageal hernia. Their age at presentation ranged from 2 days to 2½ years (mean 1.3 years). Two presented with recurrent chest infection, while 3 others had recurrent attacks of vomiting with fullness and pain in the epigastrium in one of them. One of our patients presented acutely immediately after birth with respiratory distress while another was found to have intrathoracic gastric volvulus. Chest x-ray was suggestive of paraesophageal hernia in all of them but the diagnosis was confirmed by Barium swallow and meal. Intraoperatively there was a hernial sac in all of them. The surgical treatment consisted of excision of the hernial sac

after reducing the stomach and tightening of the crura of the esophageal hiatus. Nissen's fundoplication was added in 3 patients, but in one of them this was dismantled because of tight repair. Anterior and fundal gastropexy was added in one patient, while 2 had tightening of the crura only.

Conclusions: Congenital paraesophageal hernia, although rare in the pediatric age group, can present acutely with respiratory distress or intrathoracic gastric volvulus. Physicians caring for these patients should be aware of such a presentation and complication and paraesophageal hernia should be included in the differential diagnosis of children with repeated attacks of chest infection and/or vomiting. The rarity of this condition in children makes it difficult to evaluate the true necessity of adding an antireflux procedure in these patients. We feel some form of gastropexy may be a more appropriate procedure to be added to the repair.

Keywords: Paraesophageal hernia, congenital, children, intrathoracic volvulus.

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Herniation through the esophageal hiatus is of 2 types. In sliding hiatus hernia, the esophago-gastric junction and a small portion of the stomach herniates through the esophageal hiatus.¹ Sliding hiatus hernia is seen now with increasing frequency as a result of the extensive investigation of infants and children with suspected gastroesophageal reflux. Congenital paraesophageal hernia on the other hand is rare in infants and children.² In paraesophageal hernia the stomach or sometimes other parts of the

gastrointestinal tract herniates into the chest through the esophageal hiatus but the gastroesophageal junction remains in its usual position.³ Mixed varieties of sliding and paraesophageal hernias have also been described. The presentation of congenital paraesophageal hernia is either in the form of repeated attacks of vomiting or chest infection, or rarely as an incidental finding. Awareness of such a presentation is of paramount importance for early diagnosis and to obviate the danger of intrathoracic

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volvulus.^{4,5} The purpose of this paper is to describe 6 children with paraesophageal hernia outlining their presentation and aspects of management.

Methods. Between June 1989 and July 1997, 6 children were treated at our hospital for congenital paraesophageal hernia. The charts of these patients were reviewed for: age at presentation, sex, symptomatology and duration of symptoms, radiological investigations, associated anomalies and method of treatment. The site of hernia as well as the operative procedures were obtained from the operative notes.

Results. Over a period of 8 years, 6 children were treated at our hospital for congenital paraesophageal hernia. There were 4 males and 2 females. Their age at presentation ranged from 2 days to 2½ years (mean 1.3 years). Their clinical features are summarized in Table 1. Two of our patients presented with recurrent chest infection, while 3 other had recurrent attacks of vomiting with fullness and pain in the epigastrium in one of them. One of our patients (Patient no. 4) presented acutely immediately after birth with respiratory distress. This patient was premature, with a birthweight of 1.8kg and had congenital heart disease and chromosomal abnormalities. Patient No. 5 was a 2½ year old male child who presented with recurrent attacks of cough and vomiting of 1 years duration. This patient had repair of esophageal atresia and tracheoesophageal fistula at 1 day old. He was found to have a large left paraesophageal hernia on barium swallow. Upper gastrointestinal endoscopy showed only mild esophagitis with no stricture or stenosis. In all of them the diagnosis was suspected on chest x-ray (Figure 1) and confirmed by barium swallow and meal (Figure 2). One of our patients (Patient No. 6) who presented at the age of 3 months with



Figure 1 - Chest x-ray showing a rounded shadow in the right lower chest.



Figure 2 - Barium meal showing the stomach herniating on the right side.

Table 1 - Clinical features.

Patient's No.	Age	Sex	Symptomatology	Duration of symptoms	Associated anomalies	Site	Treatment
1	1 year	F	Recurrent vomiting, pain and fullness in the epigastrium	3 months	Sickle cell disease	Right	Repair of crura of diaphragm + Nissen's fundoplication
2	2½ years	M	Recurrent chest infection	6 months	Right inguinal hernia	Right	Repair of crura of diaphragm
3	1 year & 9 months	M	Recurrent chest infection	Since 1½ months old	-	Right	Repair of crura of diaphragm + Nissen's fundoplication
4	2 days	F	Respiratory distress	Since birth	Premature, 34 weeks gestation, 1.8kg birthweight, congenital heart disease, trisomy	Right	Repair of crura
5	2½ years	M	Recurrent attacks of cough and vomiting	1 year	Esophageal atresia and tracheoesophageal fistula	Left	Repair of crura of diaphragm + Nissen's fundoplication
6	3 months	M	Recurrent attacks of vomiting	1 week	-	Left	Repair of crura of diaphragm + gastropexy

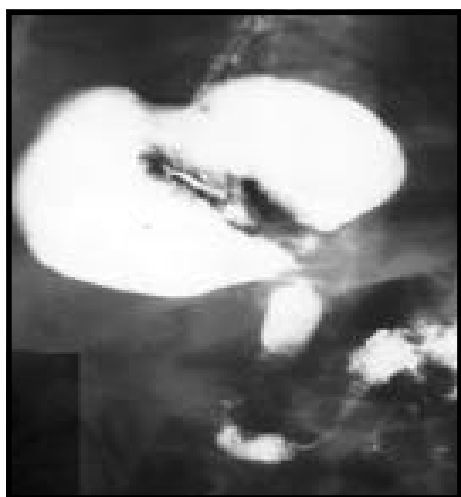


Figure 3 - Barium meal showing a large paraesophageal hernia with intrathoracic volvulus.

recurrent attacks of vomiting of one weeks duration, was found on barium meal to have a large left sided paraesophageal hernia with intrathoracic gastric volvulus (Figure 3). This patient was operated on as an emergency. He was found to have a large left sided paraesophageal hernia with almost the whole of the stomach inside the chest. The stomach was reduced and found to be congested but viable.

All patients were explored through a supraumbilical midline incision. In all, the stomach was found herniating into the chest and in all of them there was a hernial sac. After reducing the stomach, the hernial sac was excised and tightening of the crura of the esophageal hiatus was carried out. Nissen's fundoplication was carried out in 3 patients. For patient no. 4 and because of her critical condition, as she developed cardiac arrest twice during surgery, only repair of the crura of diaphragm was carried out, while patient no. 6 had, in addition to repair of the crura, anterior as well as fundal gastropexy. Post-operatively, patient no. 4 died on the 2nd post operative day because of respiratory distress and congenital heart disease. Patient no. 1 developed early dysphagia and vomiting because of excessive narrowing of the hiatal opening. This was corrected by removal of 2 of the crural stiches and undoing the Nissen's fundoplication, following which the patient did well. At a mean follow-up of 3.3 years (3 months - 7.6 years), the surviving 5 patients were symptom free and none of them developed recurrence.

DISCUSSION. Herniation of the stomach into the chest can occur due to 3 separate conditions: (1) Acquired shortening of the esophagus as a result of severe esophagitis or following repair of esophageal atresia and tracheoesophageal fistula. Herniation of the stomach in patient no. 5 in our series is most

likely due to shortening of the esophagus following repair of esophageal atresia and tracheoesophageal fistula as this patient did not show significant reflux and esophagitis on endoscopy that could contribute to shortening of his esophagus. (2) Rarely the stomach may herniate into the chest as a result of a congenital short esophagus. Congenital short esophagus is an extremely rare condition and the criteria for its diagnosis were outline by Skandelakis et al.⁶ (3) The majority of paraesophageal hernias are the result of herniation of the stomach through a congenital abnormally large esophageal hiatus. Whereas acquired paraesophageal hernia as a complication of Nissen's fundoplication is not an uncommon condition with a reported incidence of 6% to 17%, true paraesophageal hernia on the other hand is a rare condition in infancy and childhood.^{3,7}

Anatomically, the normal stomach is fixed by the gastrophrenic, gastrohepatic, gastrosplenic and gastrocolic ligaments as well as peritoneal fixation of the duodenum. So, for a paraesophageal hernia to develop 2 factors are required: (1) Congenital anatomical weakness with dilatation of the esophageal hiatus, which results from a fault during embryogenesis of the lumbar part of the diaphragm. (2) Absence or laxity of the normal anatomical gastric anchors. Add to this, the negative intrathoracic pressure which inances the upward pull of the stomach.⁴ This is supported by the fact that in the majority of these patients, a large portion if not most of the stomach herniates into the chest. This amount of gastric herniation is not possible if the stomach is anatomically fixed even in the presence of the anatomical defect. This gastric laxity is also a contributing factor for the acute development of intrathoracic volvulus as happened in one of our patients.^{4,5} In these patients the clinical symptoms depend upon the extent and degree of gastric rotation. If the degree of gastric rotation is severe, gangrene and gastric perforation may occur. The mortality from untreated gastric volvulus has been estimated as 80%.⁸ To obviate this disastrous complication, children with paraesophageal hernia should be diagnosed early and treated aggressively. Physicians taking care of these patients should be aware of such a presentation and complication. Since the presentation of these patients is either respiratory in the form of repeated attacks of cough and chest infection, or gastrointestinal in the form of repeated attacks of vomiting, careful history taking is of paramount importance and if this is suggestive a plain chest x-ray followed by barium swallow and meal should be ordered. Paraesophageal hernia should be included in the differential diagnosis of children with repeated attacks of chest infection or repeated attacks of vomiting, or both.

The surgical treatment of paraesophageal hernia is standardized and consists of reduction of the hernial contents, dissection and excision of the hernial sac

which is present in the majority if not all of these patients and repair of the hiatus by suturing the crura of diaphragm together. During the procedure care should be taken not to injure the vagus nerve or make the crura repair too tight as this is likely to cause esophageal obstruction as happened in one of our patients. There is still however no consensus as to whether an antireflux procedure or gastropexy should be added to the repair to prevent recurrent herniation of the stomach or to overcome associated reflux.^{1,3} Reflux is not rare in these patients, but the mere evaluation of associated reflux radiologically is not enough to justify the addition of an antireflux procedure with its attendant complications. Add to this the fact that reflux in these patients is due to distortion of the hiatus by the herniating stomach and once the anatomy has been restored surgically, reflux which is not present in all these patients is likely to disappear. In adult patients with paraesophageal hernia, the reported incidence of endoscopically confirmed esophagitis was 13-47%.⁹⁻¹¹ Pakic et al in a series of 40 adult patients with paraesophageal hernia based the addition of fundoplication to the repair on the presence of preoperative endoscopic evidence of esophagitis in these patients.¹² The rarity of this condition in children makes it difficult to evaluate the true necessity of adding an antireflux procedure in these patients as the reported series are small. Our series is like others, small, but 3 of our patients without an antireflux procedure are asymptomatic on follow-up, and until a proper

evaluation of this point, and to overcome the associated absence or laxity of the normal anatomical gastric anchors, some form of gastropexy may be a more appropriate procedure to be added to the repair.

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