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Congenital Paraesophageal Hernia in Infants and Children: Our Experience

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Abstract

Background: Congenital Paraesophageal Hernia (CPH), though rare in the pediatric age group, can cause significant respiratory and gastrointestinal symptoms ranging from repeated attacks of chest infection and vomiting to serious complications like intrathoracic gastric volvulus. Our experience with five such cases over five years is presented in this study. *Material and Methods:* Five children (4 males:1 female) were diagnosed and managed in our department as congenital paraesophageal hernia from January 2013 through December 2017. The characteristics recorded included age at presentation, sex, duration of symptoms, investigation, associated anomalies, operative procedures, side and size of hernia were recorded from the operative notes. Follow up period ranged from 3 months to 4.5 years. *Results:* Age at presentation ranged from 6 months to 3years (mean 1.6 years). Two patients presented with recurrent vomiting, 2 had recurrent chest infection and one baby presented with respiratory distress. Duration of symptoms ranged from one week to eight weeks (mean 3 weeks and four days). One child had associated congenital heart disease and one had right inguinal hernia. Computed Tomography in 2 and upper GI contrast in 3 patients confirmed the diagnosis. Final diagnosis was confirmed intra-operatively, with right sided defect in all the 5 cases. Repair of the crura only was done in 2 patients while as Nissen's fundoplication and gastropexy were added in one and two patients, respectively. Herniation recurred in one case in the immediate post-operative period and was managed by redo-repair of hernia with gastropexy. All the patients were followed over a mean period of 2.5 years and are symptom free with optimal weight gain. *Conclusion:* Congenital paraesophageal hernia should be strongly suspected in case of repeated chest infection or repeated attacks of vomiting or respiratory distress. Chest x-ray followed by contrast upper GI study are simple investigations to confirm the diagnosis and computed tomography may rarely be needed. Gastropexy and/or an anti-reflux procedure, in select cases, may be added to surgical reduction of the hernia with repair of crura.

Keywords: Paraesophageal Hernia; Hiatus; Crura; Gastric Volvulus; Diaphragm.

Introduction

Congenital Paraesophageal Hernia (CPH) is an uncommon condition in the pediatric age group. Congenital paraesophageal hernia occurs secondary to the developmental defect in the right crus of diaphragm. Esophageal hiatal hernia are classified as: sliding hiatal hernia (type I), paraesophageal (type II), combined sliding and paraesophageal (type III) and complex paraesophageal [associated with hernia of other organs] (type IV). Type III and type IV are clinically classified as paraesophageal hernia [1].

Pediatric patients usually present with repeated attacks of chest infection and vomiting but can be associated with serious complications such as intrathoracic gastric volvulus. Awareness of such a presentation is important for early diagnosis and to prevent the risk of intrathoracic volvulus [2,3,4]. We aim to describe the clinical characteristics and management of CPH with our experience of five such cases over last five years.

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Material and Methods

Between January 2013 and December 2017, five children were managed at our hospital for congenital paraesophageal hernia. The clinical characteristics included age at presentation, sex of patient, duration of symptoms, investigation, associated anomalies and method of treatment. The site, size of hernia and the operative procedures were recorded from the operative notes. Patients were followed for varying periods ranging from 4 months to 4 years to look for the resolution of symptoms and any recurrence (Table 1).

Results

Five patients (4 males:1 female) were treated at our hospital for congenital paraesophageal hernia. Age at presentation ranged from 6 months to 3years (mean 1.6 years). Two of our patients presented with recurrent vomiting, 2 had recurrent attacks of chest infection and one baby presented with respiratory distress at 6 months of age. Duration of symptoms ranged from one week to eight weeks (mean 3weeks and four days). There were no major anomalies associated with the paraesophageal hernia except congenital heart disease in one patient and right inguinal hernia in one. Diagnosis was made on Computed Tomography in 2 patients (done elsewhere) and upper GI contrast study in other three (Fig. 1). Final diagnosis was confirmed intra-operatively in all the five cases. In three patients whole stomach was herniated in to the chest cavity and was partially herniated in two cases. Defect was on right side in all the 5 cases (Fig. 2). All the patients were explored through a midline supraumbilical incision. The stomach along with the hernial sac was found herniating into the chest cavity. Hernial sac was excised after reducing the stomach from the chest. Tightening of the crura of the esophageal hiatus

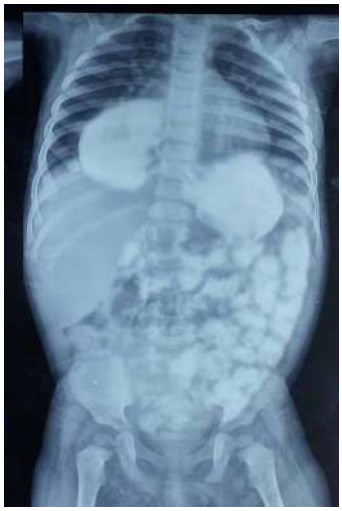

Fig. 1: Upper GI contrast study showing partial herniation of the stomach in to the right chest alongside the right of the esophagus

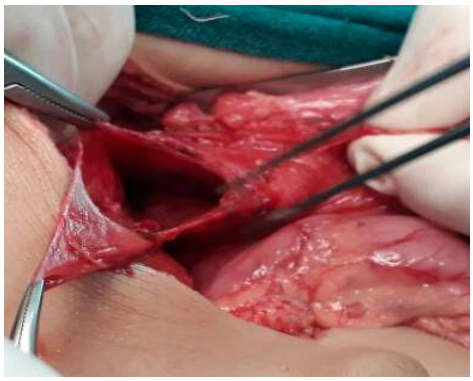

Fig. 2: Defect visible as wide gap between crura of esophageal hiatus after opening the peritoneal sac

Table 1: Clinical characteristics of the patients

Serial No.	Age	Sex	Symptoms	Duration of symptoms	Associated anomalies	Investigation(s)	Side	Treatment
1	1 year	M	Recurrent chest infection	4 weeks	congenital heart disease	Chest X-ray, Computed Tomography	Right	Repair of crura of diaphragm + gastropexy
2	3 years	M	Recurrent attacks of cough and vomiting	2 months	Right inguinal hernia	Chest X-ray Upper GI Contrast	Right	Repair of crura of diaphragm
3	1 year & 9 months	M	Recurrent chest infection	2weeks	-	Chest X-ray Upper GI Contrast	Right	Repair of crura of diaphragm + Nissen's fundoplication
4	6 months	F	Respiratory distress	1 week	-	Chest X-ray, Upper GI Contrast	Right	Repair of crura of diaphragm, Redo with gastropexy for
5	2 years	M	Recurrent vomiting	3 weeks	-	Computed Tomography	Right	Repair of crura of diaphragm

was performed with non-absorbable sutures (Fig. 3). Nissen's fundoplication was performed in one and gastropexy in two patients. Only repair of the crura of diaphragm was carried out in two patients. One patient (Patient No. 2) developed recurrence in the immediate post-operative period and was managed by re-exploration and redo-repair of hernia with gastropexy. All the patients were followed over a mean period of 2.5 years (3 months to 4.5 years). There was no mortality and no patient had significant reflux of esophagitis. All the 5 patients, till last follow up, are symptom free with optimal weight gain.

Fig. 3: Defect narrowed by crural repair with non-absorbable sutures

Discussion

Congenital diaphragmatic hernia includes Bochdalek hernia, Esophageal hiatal hernia, Morgagni hernia and Central part damage according to the location of the hernia. Esophageal hiatal has main two types sliding hiatal hernia and paraesophageal hernia [4]. Herniation of the stomach may occur as result of shortening of the esophagus, either congenital or acquired (repair of esophageal atresia and tracheoesophageal fistula) [5]. The majority of paraesophageal hernias are the result of herniation of the stomach through a congenital large esophageal hiatus. True paraesophageal hernia on the other hand is a rare condition in infancy and childhood [6]. This amount of gastric herniation is not possible if the stomach is anatomically fixed even in the presence of the anatomical defect.

As demonstrated in our series, symptoms are nonspecific ranging from vomiting and respiratory tract infection to volvulus. Infants present with respiratory distress and scaphoid abdomen [7]. Since the presentation of these patients is varied, careful history taking is of paramount importance and if it suggests, a plain chest x-ray, contrast study and/or CECT, if needed. A preliminary chest radiograph examination can show air-fluid level in the thoracic cavity. Definitive diagnosis needs contrast upper GI study as demonstrated in our cases. We preferred non-ionic water-soluble contrast (Sodium diatriazoate), although some authors use barium. A tomography is helpful to identify the accurate location of the defect and the herniated organs and/or a complication of herniated viscus like volvulus [8].

In complicated patients (gastric volvulus), the clinical symptoms depend upon the extent and degree of gastric rotation. In severe degree of gastric rotation, gangrene and gastric perforation may occur. Untreated gastric volvulus has been estimated to have 80% mortality [9]. To prevent this complication, children with paraesophageal hernia should be diagnosed early and treated aggressively. Physicians taking care of these cases should be cognisant of such a presentation and complication.

The surgical treatment of paraesophageal hernia consists of reduction of the hernia contents, dissection, excision of the hernial sac and repair of the hiatus by suturing the crura of diaphragm. Care should be taken not to injure the vagus nerve. Crura repair should not be too tight as this is likely to cause esophageal obstruction [10]. We treated all the cases by reduction of hernia with excision of sac with Nissen's fundoplication in one and gastropexy in two patients. There is still controversy as to whether gastropexy and/or anti-reflux procedure or should be added to the repair to prevent recurrent herniation of the stomach or to overcome associated reflux [11]. We did not demonstrate any significant reflux or esophagitis in the follow up, but reflux does occur in these patients. Reflux in these patients is due to buckling of the hiatus by the herniating stomach and once the anatomy has been re-established surgically, reflux which is not present in all these patients, is likely to disappear. In adult patients with paraesophageal hernia, the esophagitis has been reported in 13-47% of cases [12]. The rarity of this condition in children makes it difficult to evaluate the true necessity of adding an anti-reflux procedure in these patients as the reported series are small. Four patients in our series without an anti-reflux procedure were asymptomatic on follow-up.

Conclusion

Congenital paraesophageal hernia, though rare in infants and children, should be included in the differential diagnosis with history of repeated attacks of chest infection or repeated attacks of vomiting or respiratory distress. Chest x-ray followed by contrast upper GI study are simple investigations to confirm the diagnosis and computed tomography may rarely be needed. The optimal surgical treatment of these cases consists of reduction of the hernia contents, dissection and excision of the hernial sac and repair of crura of esophageal hiatus. Gastropexy and/or an anti-reflux procedure are needed in select cases.

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