Megaesophagus in congenital diaphragmatic hernia

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ABSTRACT

A newborn with megaesophagus associated with a left sided congenital diaphragmatic hernia is reported. This is an under recognized condition associated with herniation of the stomach into the chest and results in chronic morbidity with impairment of growth due to severe gastro esophageal reflux and feed intolerance. The infant was treated successfully by repair of the diaphragmatic hernia and subsequently by fundoplication. The megaesophagus associated with diaphragmatic hernia may not require surgical correction in the absence of severe symptoms.

Key words: Congenital diaphragmatic hernia, megaesophagus

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CASE HISTORY

Congenital diaphragmatic hernia (CDH) commonly occurs through the posterolateral defect of Bochdalek and left sided hernias are more common than right. The incidence and variety of associated malformations are highly variable and may be related to the side of herniation. The association of CDH with megaesophagus has been described earlier and clinical symptoms managed without surgery but with compromised growth. We describe a newborn with left sided CDH and megaesophagus who required early surgical correction due to severe gastro esophageal reflux (GER) and feed intolerance.

A male baby weighing 2400 g, born by spontaneous vaginal delivery at 36 weeks gestation to a primigravida mother was found to be cyanosed with severe respiratory distress at birth. The antenatal period was uneventful and a second trimester antenatal scan was reported as normal. A clinical diagnosis of left sided diaphragmatic hernia was made and the neonate immediately intubated and ventilated. His vital signs improved dramatically with positive pressure ventilation and he received antibiotics, sedation, muscle paralysis and inotropes to stabilize his general condition. A plain radiograph of the chest and abdomen revealed a left sided diaphragmatic hernia with the stomach and intestines located in the left hemithorax (Figure 1). Echocardiogram revealed a dextroposed heart with persistent foramen ovale, interatrial septum bulging into left atrium, hypoplastic left pulmonary artery and mild tricuspid regurgitation.

He underwent diaphragmatic repair on the fourth day of life via a left subcostal incision. During surgery a large posterolateral defect was noted, with herniation of stomach, spleen and small and large intestines into the thoracic cavity. The left lung was small and hypoplastic. After reduction of the viscera the diaphragmatic defect was repaired. The infant was extubated on the eighth day of life but remained tachypnoeic and was given total parenteral nutrition. The nasogastric aspirate was initially greenish with large volumes and associated with occasional vomiting. Gradually it became less and nonbilious and he was started on small volume of milk feeds (1 ml/h). He passed normal stools and did not have any abdominal distension. However he did not tolerate any further increase in feed even after 2 weeks of starting nasogastric feed.
An upper gastrointestinal contrast study done at this time showed a dilated lower esophagus with massive reflux and a sub diaphragmatic stomach with delayed emptying (Figure 2). Although malrotation was noted there was no evidence of distal obstruction. A laparotomy was done on the 22nd day of life, which showed a megaesophagus without hiatus hernia. The diaphragmatic repair was intact and extensive adhesions and malrotation were noted. Nissen fundoplication, correction of malrotation and release of all adhesions were done. The infant was extubated on the following day and nasogastric feed started a day later with progressive and rapid increase to reach full feed. He was discharged at 1 month of age on exclusive breastfeeds and gaining weight. On review after 6 weeks the infant had gained 1000 g in weight and continues to thrive well at 6 months of age.

**DISCUSSION**

Congenital diaphragmatic hernia occurs due to failure of the pleuroperitoneal membrane to develop adequately and close around the eighth week of gestation. The abdominal viscera then enter the pleural cavity and cause poor lung development leading to pulmonary hypoplasia. Over 90% involve a posterolateral defect and occur on the left side.\(^1\) High risk factors for morbidity and mortality in CDH include antenatal diagnosis, early onset of symptoms after birth, prematurity and any associated anomalies. The mortality of CDH is directly related to the degree of lung hypoplasia and pulmonary hypertension. Besides chromosomal anomalies, malformations associated with CDH may involve cardiovascular, neurological, skeletal, genitourinary and gastrointestinal systems. Gastrointestinal anomalies known to occur include pyloric stenosis, malrotation of gut, intestinal atresia, imperforate anus, Meckel’s diverticulum and Hirschsprung’s disease.\(^2\)

Most cases of CDH are detected by antental ultrasonography and most neonates present with severe respiratory distress within the first few hours of life. A plain radiograph of the chest is helpful in the initial evaluation of the type of defect and its contents. A nonaerated CDH can appear as a solid mass and an aerated hernia needs to be differentiated from other congenital cystic lung lesions. In a left sided CDH the presence of the tip of the nasogastric tube in the left hemithorax or at the level of the esophago gastric junction are indicative of gastric herniation.

Feed intolerance is commonly observed after CDH repair and besides postoperative sedation, paralysis and ileus, the associated gastrointestinal anomalies often add to its causation. Esophageal dilatation causing GER is increasingly recognized as a cause of both acute and chronic morbidity in infants surviving CDH repair. The association of CDH with megaesophagus has been described earlier\(^3,4\) and clinical symptoms managed without surgery but with compromised growth. There are many factors, which contribute to the esophageal dilatation.\(^5\) The mediastinal shift and compression associated with CDH may disturb the normal development and mobility of the intrathoracic oesophagus, leading to chronic ectasia and impairment of lower esophageal sphincter function. The relative esophageal obstruction due to kinking at the gastro esophag-
geal junction may lead to in utero polyhydramnios and contribute to development of esophageal dilatation. Abnormality of the gastro esophageal junction, a shortened intra-abdominal esophagus and an obtuse angle of His are other postulated reasons. The increased intra-abdominal pressure after repair and deformed crus due to tight repair of the hemidiaphragm are other contributory causes. In a left sided CDH with intrathoracic stomach these factors result in esophageal dilatation in utero and produce GER and feed intolerance after surgical repair. Since primary repair of a large defect without a prosthesis has been thought to be one of the reasons for an obstructive mechanism at the gastro esophageal junction, some centres routinely use them for repair of for very large defects. We chose not to use the above as our preference was for anatomical closure. The use of prosthesis during repair has not been conclusively shown to prevent GER.

The preoperative fluoroscopic examination did not show evidence of distal obstruction. Hence we believe that the feed intolerance was due to severe GER, rather than malrotation, which is an associated finding in CDH. The megaesophagus in CDH may resolve spontaneously with conservative management. Fundoplication was required for the above infant in the neonatal period itself, due to severe GER and inability to continue enteral feeding even after 2 weeks of CDH repair. We did not attempt a trial of prolonged nasoduodenal or nasojejunal feeding due to the severity of symptoms. A recent review looking at nutritional morbidity in survivors of CDH suggests antireflux fundoplication operation for pathologic GER. The subsequent relief of symptoms with rapid increase to reach full oral feed and the remarkable weight gain observed during follow-up justified the necessity for an early antireflux surgery, rarely done at this age.

To conclude esophageal dilatation with GER is a cause of acute and chronic morbidity in infants surviving repair of congenital diaphragmatic hernia. An early gastrointestinal contrast study is indicated in presence of persistent feed intolerance postoperatively. If left untreated it results in significant and long-term impairment of growth and development of these infants. The megaesophagus associated with diaphragmatic hernia may not require immediate surgical correction in the absence of severe symptoms.

REFERENCES