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CASE REPORT

A Challenging Case of Congenital Diaphragmatic Hernia Managed Successfully: A Case Report

Saurabh Jitendra Tiwari, Apoorva Kulkarni,* Geeta Kekre, Abhaya Gupta, Shalika Jayaswal, Paras Kothari

Lokmanya Tilak Municipal Medical College and Sion Hospital, Mumbai, India

ABSTRACT

Despite advances in antenatal diagnosis and perinatal care, mortality rate remains elevated in patients with congenital diaphragmatic hernia (CDH). We report a case of left sided CDH in a term baby with persistent gastroesophageal reflux (GER) after repair, and persistent pulmonary hypertension requiring multiple vasodilators. Baby required high frequency oscillatory ventilation for 6 weeks. After failed medical management, bedside gastrostomy in neonatal intensive care unit was done. Nissen's fundoplication was done later. After 2.5 months child was discharged asymptomatically.

Key words: Congenital diaphragmatic hernia; Gastroesophageal reflux disease; High frequency oscillatory ventilation; Fundoplication; Pulmonary hypertension

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a life-threatening congenital anomaly with an incidence of 1 in 2,500 live births. Few studies focused on associated chronic morbidity and long-term outcomes.[1] Gastroesophageal reflux (GER) is a well-recognized complication in postoperative patients with CDH which may occur in 30–70% of patients [2], but an incidence of up to 80 % has been reported in patients treated with ECMO (extracorporeal membrane oxygenation) before CDH repair. It is also reported that 21-60% of CDH patients remain symptomatic after medical treatment, and thus require anti-reflux surgery.[3] Herein, we report a case of CDH complicated by refractory GERD.

CASE REPORT

A full-term male baby, weighing 2.3kg, antenatally diagnosed case of CDH, was admitted for the management of CDH. The baby was vitally stable with decreased air entry on left side of chest. Rest of the systemic examination was normal. On day one of life, open CDH repair was done. There was herniation of small bowel, transverse colon, spleen, left lobe of liver and stomach. Postoperatively, the patient was put on ventilatory and ionotropic support. The baby had convulsions on postoperative day 2, thus anticonvulsants were started. The baby was extubated on postoperative day 3, however, the baby developed pulmonary hypertension (as detected on echocardiogram) and was started on multiple vasodilators like milrinone, sildenafil, Prostaglandin E1, bosentan.

The baby was reintubated on day of life (DOL) 8 due to repeated episodes of desaturation and put on high frequency oscillatory ventilation for a period of 6 weeks. The feeds were started on DOL 13, but there were repeated episodes of gastroesophageal reflux and ventilator associated pneumonia. GER was treated with dietary thickeners, head-up positioning, prokinetic agents, and histamine type-2 receptor antagonists, but failed. Feeding gastrostomy (GT) under anaesthesia cover was done at bedside in NICU (neonatal intensive care unit) on DOL 40. The infant was shifted on conventional ventilator mode on DOL 52.

As GER persisted, Nissen's fundoplication was also done on DOL 54. Postoperatively, feeds were started and gradually increased which were well tolerated. The baby required prolonged antibiotics and was extubated on DOL 64, and was on room air on DOL 73. In view of the persistent irritability, psychiatry opinion was taken, and the baby was diagnosed with benzodiazepine withdrawal syndrome for which carbamazepine was started, which was

Correspondence*: Apoorva Kulkarni, Lokmanya Tilak Municipal Medical College and Sion Hospital, Mumbai, India
E mail: apookool@gmail.com

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tapered and stopped over 15 days. The patient was discharged on full feeds on DOL 90. At one month follow up, patient was asymptomatic and thriving well. The baby has been on regular follow up since then.

DISCUSSION

GER is one of the major sequelae in infants with congenital diaphragmatic hernia. Symptoms include coughing or choking, vomiting, chronic respiratory distress, growth failure, sleeping problems, and apnea. Anatomic abnormalities include a small, vertically oriented stomach, an obtuse angle of His, and the lack of an intra-abdominal esophagus. Indications for fundoplication include recurrent aspiration pneumonia, apneic episodes, bradycardia, apparent life-threatening events, bronchopulmonary dysplasia, severe vomiting, growth failure, esophagitis, and esophageal stricture.[4]

The incidence and the severity of GERD, however, decrease after the first year of life. Relaxation of the lower esophageal sphincter is the main event for the occurrence of reflux; therefore, meticulous attention must be paid to the diaphragmatic crura during surgical repair.[5] Verla et al, pointed several possible factors for GER; slow pulmonary expansion of the affected side, size of defect, malposition of the stomach, use of mechanical ventilation, or extracorporeal membrane oxygenator.[6] Marseglia et al, proposed increased intra-abdominal pressure after closure of the abdominal defect, motility disturbance of the upper gastrointestinal tract, or damage of esophageal peristaltic pump, as possible causes of GERD in cases of CDH.[7] There was a correlation between GER and early detection of CDH in prenatal studies and the presence of polyhydramnios. Patients with GER also required longer artificial ventilation and longer hospitalization.[8]

GERD can be recalcitrant to the medical management and necessitate alternate feeding routes for nutritional rehabilitation such as continuous tube feedings. Similarly, in the index case, on failure of medical management of GER, we had to perform feeding gastrostomy. However later, the patient ultimately required fundoplication. The outcome of fundoplication in our patient was favourable as symptoms disappeared completely and full enteral feeding was possible.

In conclusion, GER is a known association of CDH. Constant surveillance and follow-up are necessary for timely intervention to prevent the ravages of untreated GERD complications. Prompt intervention with a combination of medical and surgical management will prevent disabling long-term nutritional and pulmonary problems. Better communication between the neonatologist and surgeon is essential to individualize treatment. Fundoplication is safe and effective and can be recommended for infants with severe GERD not settling on medical management.

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Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material (if any used), from the legal guardian of the patient with an under-standing that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

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