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Pediatrics

PROGNOSIS OF INFANTS AND CHILDREN WITH DELAYED CONGENITAL DIAPHRAGMATIC HERNIA

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ABSTRACT

Background: The infants and children with Congenital diaphragmatic hernia (CDH) presenting beyond the neonatal period are classified as delayed congenital diaphragmatic hernia. The prognosis of this subset of patients is reported to be better than the neonatal CDH patients.

Methods: A retrospective review of the medical records between January 2014 and December 2017 of the infants and children admitted with congenital diaphragmatic hernia with delayed presentation beyond the neonatal period at our institution was performed.

Results: A total of 98 patients of CDH were identified and 11 patients presented after the neonatal period. The common presentation was respiratory distress. Three children presented with volvulus of the stomach. There were two deaths (18%) and a total of nine patients (82%) had survived.

Conclusion: The prognosis of the patients with delayed CDH is significantly better than the CDH patients presenting in the neonatal period. Gastric volvulus remains the most dangerous complication.

KEYWORDS : Congenital diaphragmatic hernia, delayed presentation.

1.INTRODUCTION

Congenital Diaphragmatic Hernia commonly presents in the neonatal period with either an antenatal diagnosis or with respiratory distress after birth. Rarely, the CDH presents later in the infancy or in the early childhood. The incidence of this delayed CDH presentation is reported to be between 5 – 45%[1,2]. The pathophysiology of this subset of the patients with delayed CDH is different from their neonatal counterparts with the incidence of pulmonary hypertension being significantly less in the delayed CDH patients.

2.MATERIALS AND METHODS:

The medical records of all the infants and children with CDH who were treated at the Institute of Child Health and Hospital for Children, Madras Medical College, Chennai were retrospectively reviewed. The study period was for 48 months from January 2014 to December 2017. All the infants and children with CDH confirmed during surgery were included in the study. The neonatal CDH patients and children with eventration were excluded from the study.

The medical records were reviewed for various patient parameters including age, sex, symptom at presentation, surgical procedure, and overall survival. All the patients underwent a Chest X-ray and some patients underwent CT scan of the Chest to rule out other pathologies. An echocardiogram was done to rule out pulmonary hypertension. The need for mechanical ventilation was decided depending on the clinical condition of the patient in an individual basis.

3.RESULTS:

A total of 98 patients with Congenital Diaphragmatic Hernia were identified during the study period and 11 patients who presented beyond the neonatal period were included in the study. The incidence of delayed CDH in our study was 11.22%. There were nine male children and two female children. Of the 11 children, four were infants, five children were less five years of age and two children were above five years of age. Ten patients had left-sided defect and one child had right CDH. The clinical and the operative details of each of the patients is summarised in the Table 1.

A total of nine children presented with respiratory symptoms. The respiratory distress was severe in two children with intra-thoracic gastric volvulus and required mechanical ventilation. The distress was less severe in the three patients who had pneumonia and bronchiolitis. The remaining four patients had upper respiratory tract symptoms (URI) without distress and were diagnosed with CDH/eventration in the screening chest X-ray. One child had failure to thrive and one child had recurrent URI. One infant presented with intestinal obstruction and a normal Chest X-ray. During laparotomy, a knuckle of the transverse colon was found trapped in a small 2 x 1 cm circular defect in the left posterolateral diaphragm. The child underwent resection anastomosis and closure of the defect.

Three patients presented with gastric volvulus. One of them had intraabdominal gastric volvulus and presented with abdominal pain and severe retching. The X-ray chest and abdomen was reported as left eventration with prominent stomach. During surgery, a 3 x 3 cm left diaphragmatic defect with herniated small bowel and a mesentericoaxial volvulus of the stomach was found. A CDH repair and an anterior gastropexy was done. The other two patients had intra-thoracic gastric volvulus. One child presented with gangrene of the intra-thoracic stomach with severe shock. The child died due to a cardiac arrest on table during the emergency surgery. Another patient with intrathoracic stomach and gastric volvulus with severe respiratory distress on ventilator was wrongly diagnosed as left pyo-pneumothorax based on the chest X-ray and an intercostal drain was inserted. Later the CT Chest was performed and a diagnosis of left CDH was made and the child was operated. The child required prolonged post-operative ventilation and underwent re-operation for burst abdomen and finally died of ventilator associated pneumonia.

The infants and children who were stable were evaluated with an Echocardiogram. An Echo was done for ten patients. None of them had pulmonary hypertension. One child had small ASD. Three children underwent CT chest to confirm the diagnosis. Eventration was the most common differential diagnosis and the diagnosis was confirmed only during the surgery.

All the children underwent laparotomy and repair of the diaphragmatic defect. The defect was small less than 3 cm in six patients and larger than 3 cm in the remaining patients. All of them had a tension-free anatomical repair of the diaphragm. Perioperative mechanical ventilation was required in two patients who presented with severe respiratory distress and intra-thoracic gastric volvulus. None of the remaining nine patients required ventilatory support. An intercostal drain was placed in all patients and was typically removed after 48 to 72 hours.

The two children with intra-thoracic gastric volvulus who died during the treatment contributed to the 18.2% mortality in our series. The remaining nine patients contributed to the 81.8% survival rate. All the surviving children are on regular follow-up and there were no documented recurrence in any of the patients.

Table 1: Clinical features other variables of the Children with delayed CDH

S.no	Age/Sex	Symptom	Surgery	Mech vent	Echo	Complications	Survival
1.	3/12, M	Intestinal obstruction	CDH repair/ colon Resection	No	N	Nil	Yes
2.	4/m	Abd. Gastric volvulus, pain	Left CDH repair	No	N	Prolonged NG aspirate	Yes
3.	7/m	Intrathoracic Gastric volvulus	Left CDH repair	yes	Not done	Gangrene stomach, shock	No
4.	8/12/M	Incidental/URI	Left CDH repair	No	N	Nil	Yes
5.	6/M	Intrathoracic Gastric volvulus	Left CDH repair	yes	N	Prolonged post op ventilation, burst abdomen.	No
6.	11/12/f	Incidental /URI	Left CDH repair	No	N	Nil	Yes
7.	1 ½ m	Pneumonia	Left CDH repair	No	N	Nil	Yes
8.	7/12/f	Bronchiolitis	Left CDH repair	No	N	Nil	Yes
9.	1 3/12 m	Recurrent URI	Right CDH repair	No	N	Nil	Yes
10.	2 ½ m	Pneumonia	Left CDH repair	No	N	Nil	Yes
11.	4 / m	Failure to thrive /URI	Left CDH repair	No	N	Nil	Yes

DISCUSSION

The estimated survival of neonatal CDH during the same study period was 34%, while the survival of delayed CDH reached 81.8%. The difference is significant with the fisher exact test statistical value of 0.0095 and a p value of <0.05. Male children predominated in our study with a ratio of 9:2 and left-sided defect predominated in 10:1 ratio. Associated anomalies were found in only one patient who had an ASD.

Respiratory symptoms remained the most common presentation in delayed CDH patients. However, the reason for the respiratory distress in the delayed CDH patients was usually an underlying pneumonia or rarely due to gastric volvulus rather than the pulmonary hypoplasia and pulmonary hypertension which are common in the neonatal CDH patients.

The cause of death in our study was due to the intra-thoracic gastric volvulus. The presentation of gastric volvulus can be acute in an apparently normal child[3]. The Chest X-ray can be confusing because the dilated intrathoracic stomach typically mimics a lung cyst or a pyo-pneumothorax[4] necessitating a CT scan of the Chest. A CT chest is the gold standard test to identify delayed CDH and especially an intra-thoracic gastric volvulus. Gastric volvulus that presents acutely is a surgical emergency and immediate surgery is indicated if the diagnosis is confirmed with a chest X-ray. Early identification and timely intervention will prevent morbidity and mortality. Apart from this small subset of the patients who presented acutely with gastric volvulus, the remaining patients had excellent prognosis. Hence the survival directly corelated with the absence of pulmonary hypoplasia and pulmonary hypertension.

The etiopathology of the delayed CDH is different from the neonatal CDH. Various hypothesis has been proposed to explain the phenomenon behind the occurrence of delayed CDH. In 1991, Suresh et al[5] published the occurrence of delayed CDH secondary to group B streptococcal infection. Several reports were later published linking the group B streptococcal infection with delayed CDH possibly due to associated diaphragmatic necrosis[6].

Acquired congenital diaphragmatic hernia[7] is another interesting presentation where the chest X-ray taken at birth or in the early neonatal period had documented the appearance of a normal diaphragm and the child at the time of presentation has a diaphragmatic hernia suggesting a possible acquired etiology. However, this acquired phenomenon may also be due to the presence of a small congenital defect in the diaphragm without initial herniation. The herniation may occur later in the post-natal period during increased intraabdominal pressure.

To conclude, delayed CDH has an excellent survival rate owing to the favourable lung parameters including absence of lung hypoplasia and pulmonary hypertension. However, a small subset of patients presents acutely with gastric volvulus and need expedite treatment to avoid morbidity and mortality.

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