Journal of Pediatric Surgery 51 (2016) 1917–1920

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Acute gastrointestinal compromise in neonates with congenital diaphragmatic hernia prior to repair☆

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ARTICLE INFO

Article history:
Received 4 August 2016
Accepted 12 September 2016

Key words:
Congenital diaphragmatic hernia
Acute gastrointestinal compromise
Neonatal
Volvulus
Neonatal surgery
Lactic acidosis

ABSTRACT

Background: Congenital diaphragmatic hernia (CDH) affects 1 in 3000 live births. Modern management strategies include delayed repair of the diaphragm to permit pre-operative optimization of cardiorespiratory status. We describe a cohort of neonates in whom early emergency operative intervention was required for potentially fatal intestinal compromise.
Methods: A retrospective review was performed of all neonatal CDH patients managed at a tertiary center in an 8-year period (2005–2012).
Results: A total of 126 CDH patients were managed during the 8-year period. Five neonates (male – 1; gestation 37 + 4–39 + 7; birth weight 2.9–3.7 kg; left CDH – 5) required emergency operative intervention for presumed gastrointestinal compromise. All five neonates demonstrated systemic hypotension despite inotropic support, raised serum lactate (>2 mmol/L), and abnormal radiographic findings. Operative intervention occurred within 3 days of birth (1–3 days). Findings included gastric volvulus, jejunal volvulus, and perforated caecum. All patients underwent primary diaphragmatic repair without a patch. Temporary ileostomy was required in 1 patient. All patients remain alive.
Conclusion: Gastrointestinal compromise is a rare, but potentially catastrophic, complication of CDH. Emergency operative intervention may be required in a select cohort of patients. Early deterioration following birth should alert clinicians to the possibility of significant intestinal pathology.
Level of evidence: Level IV case series with no comparison group.

Congenital diaphragmatic hernia (CDH) affects approximately 1 in 3000 live births. Despite continued advances in treatment, it remains a significant cause of neonatal morbidity and mortality. Initial clinical management is aimed at treatment of respiratory insufficiency and pulmonary hypertension. Modern management strategies incorporate use of "gentle" ventilation techniques and pulmonary vasodilators, combined with delayed repair of the hernia defect to allow optimization of cardiorespiratory status pre-operatively.

Congenital diaphragmatic hernia may involve herniation of a variety of abdominal viscera, including stomach, small intestine, colon, spleen and liver, through diaphragmatic defects of highly variable size. Acute gastrointestinal complications in the neonatal period in CDH are considered to be rare [1]. We describe five neonates with acute gastrointestinal compromise (GIC) in conjunction with CDH, managed over a relatively short time period in a large tertiary pediatric center. We have highlighted the early diagnostic indicators and presenting features of GIC in CDH, and have described the surgical management and outcome of this rare but potentially life-threatening complication of CDH.

1. Methods

We conducted a retrospective case-note review of neonates with CDH admitted to the newborn intensive care unit (NICU) at our tertiary institution between January 2005 and December 2012. We identified all neonates who developed acute GIC prior to operative repair. The institution's research ethics and governance approved the research protocol.

All neonates were out born in our state and transferred to our center by a dedicated neonatal transport service. Neonates were managed with

☆ Conflicts of interest: None.

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http://dx.doi.org/10.1016/j.jpedsurg.2016.09.012
0022-3468/

Table 1
Demographic data for GIC-CDH group and all-CDH group.

Demographic	GIC-CDH group (n5)	All-CDH group (n126)	*P* value
Gestation	38.9 (37.4–39.7)	38.4 (29.6–41.6)	0.573
Gender (M:F)	1:4	73:53	0.166
Weight	3.2 kg (2.9–3.7)	3.0 kg (1.0–4.4)	0.280
Antenatal diagnosis	3/5 (60%)	72/126 (57%)	0.361
Left hernia	5/5 (100%)	102/126 (81%)	0.02

reference to a departmental CDH guideline. The strategy was to minimize lung injury by incorporating permissive hypercapnia, early use of high frequency oscillation and jet ventilation, and use of pulmonary vasodilators, including inhaled nitric oxide, based on clinical and echocardiographic assessment of pulmonary hypertension.

Decisions relating to timing of operative CDH repair were made jointly between the pediatric surgical and neonatal teams, based on comprehensive assessment of clinical status and stability. It was usual practice to consider delaying repair to optimize pre-operative clinical status. In each case of CDH diagnosed with GIC (GIC-CDH group) the following data were collected from case-note review:

1. demographic data: gender, gestation, birth weight, laterality and type of hernia, antenatal diagnosis
2. presenting features in the first 72 h of life: radiographic findings, volume and presence of bile in nasogastric (NG) aspirates, peak plasma lactate, presence of thrombocytopenia, coagulation profile (plasma APTT and INR), systemic hypotension requiring inotropic support
3. operative findings: defect size, herniated viscera, requirement for patch repair
4. outcome data: age at first operation/CDH repair, age at full enteral feeds, length of stay, survival to discharge

Antenatal measures of lung volume were not consistently recorded and were therefore not included in analysis. For comparison, corresponding demographic, operative and outcome data were obtained from the departmental database for all cases of CDH (all-CDH group) managed during the same time period. Age at full enteral feeds and defect size were not recorded routinely in the database and were therefore not available for this group. Continuous data for the all-CDH group were summarized as median and range. Comparison between GIC-CDH and all-CDH groups was performed by unpaired t-test and chi-square for continuous and categorical data, respectively. A p value <0.05 was considered significant.

2. Results

There were five cases of acute GIC from a total of 126 neonates with CDH admitted within the eight-year study period. Demographic data and presenting features in the first 72 h are provided for each of the neonates in the GIC-CDH group and summarized for the all-CDH group (Tables 1 and 2). There were no significant differences in gestation, gender or weight between the two groups. An antenatal diagnosis of CDH was made in 3/5 (60%) GIC-CDH and 72/126 (57%) all-CDH. All neonates in the GIC-CDH group had a left-sided CDH, compared to 102 (81%) of the all-CDH group ($p = 0.02$).

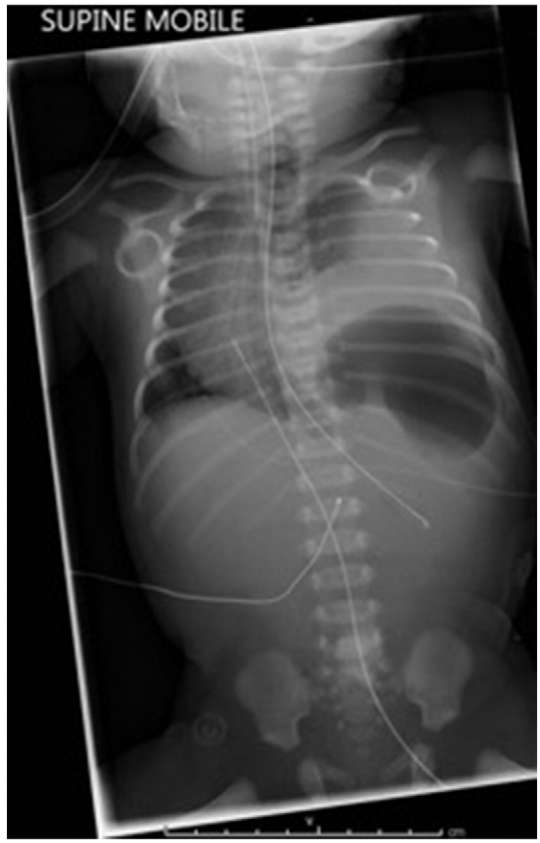

Fig. 1. Chest radiograph demonstrating left congenital diaphragmatic hernia with a prominent gastric shadow in left hemithorax.

The commonest presenting features, observed in all five neonates in the GIC-CDH group, were abnormal radiographic findings, elevated plasma lactate (≥2 mmol/L) and systemic hypotension requiring inotropic support. Initial radiographs demonstrated abnormally distended loops of intestine and/or a prominent gastric bubble (Figs. 1 and 2). Of note, these abnormal findings were persistently present on serial radiographs in all affected neonates. Two neonates had bilious NG aspirates. Three neonates were coagulopathic.

The median age at operative repair was 2 days in the GIC-CDH group and 4 days in the all-CDH group. Operative findings and procedures and outcomes are summarized (Table 3). All neonates in the GIC-CDH group had a small type-A (Boston classification) defect. All defects were closed by primary repair (no patch) at first operation. Two neonates had obstructed, ischemic proximal large intestine with cecal perforations. The terminal ileum, cecum and transverse or ascending colon were resected in both of these patients, with a terminal ileostomy formed in one patient. One neonate had an obstructed loop of large intestine herniating through the defect, but required neither resection nor stoma formation. One neonate had evidence of malrotation and volvulus, with an ischemic proximal jejunum, which was resected and primarily anastomosed. The final neonate had a gastric volvulus, with

Table 2
Presenting features.

Pt	X-ray changes	NG aspirates bile/vol (ml/kg per day)	Peak lactate (mmol/L)	Platelets ($\times 10^9$/L)	APTT/INR ref range APTT 27–65 INR 0.8–1.5	Low BP
1	Dilated bowel loop, abnormal stomach bubble in chest	No/13	2.1	242	46/1.3	Y
2	Prominent stomach bubble	Yes/6	6.1	296	44/1.2	Y
3	Distended loops with pneumatosis	No/6	2.0	315	39/1.2	Y
4	Distended stomach and bowel, air in pleural cavity	No/4	2.7	186	55/1.8	Y
5	Dilated bowel loops	Yes/2	3.0	158	>180/1.6	Y