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ABSTRACT

Congenital diaphragmatic hernia occurs 1 in 2500 births, when the abdominal contents protrude through the patent pleura-peritoneal canals in the diaphragm, leading to maldevelopment of the alveoli and pulmonary vessels. CDH is associated with high mortality, and the probability of survival was found to be low in the presence of any other associated congenital abnormality, worse with cardiac defects. We successfully managed a 2-day-old neonate born in our facility with left congenital diaphragmatic hernia presenting with immediate postnatal respiratory distress, and delayed repair of the defect was carried out - through an open trans-abdominal approach with good outcome; the neonate survived to hospital discharge without any complication.

Keywords: Left congenital diaphragmatic hernia, neonate, peri-operative management.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) occurs rarely, with an incidence of 1 in 2500 births. It occurs because of the abdominal contents protruding through the patent pleura-peritoneal canals in the diaphragm, leading to life-threatening maldevelopment of the alveoli and pulmonary vessels (abnormally responsive to vasoactive substances with resultant pulmonary hypertension).[1-3]

CDH accounts for 8% of all congenital anomalies, and part of a syndrome of genopathy, which is commonly associated with trisomies; 13-Patau’s, 18-Edward’s, and 21-Down’s syndromes.[3,4] In addition 40% of patients with CDH have associated major congenital abnormalities.[4] CDH can be classify into an anterior or Morgagni’s hernia-representing 1-6% of cases, the posteriolateral or Bochdalek in 90% of cases; of which 85% are left-sided, 15% right side; and 2% of cases are bilateral; the last two types is associated with the worst outcome.[1-4]

Until recently, CDH was considered as a surgical emergency to be treated under 24 hours, and immediate postnatal / early repair was the norm.[1, 4, 6]

We successfully managed a 2 days-old neonate with left congenital diaphragmatic hernia associated with poor Apgar score and neonatal respiratory distress with good outcome.

CASE REPORT

The anaesthetist's management of a 2 day-old neonate with radiological and clinical diagnosis of left posteriolateral congenital diaphragmatic hernia (LCDH) associated with poor neonatal apgar score 4 and 6 at 1 and 5 minutes, after birth respectively, and respiratory distress immediately necessitating endotracheal intubation and ventilation during resuscitation.

Preoperative assessment revealed: a term neonate female 40 weeks post conceptional age, weighing 2.6kg, length 50cm, head circumference 34cm. She had barrel shaped chest, diminished air entry on the left and shifted heart sounds to the right hemithorax - with normal echocardiography findings. Chest X-ray shows loops of bowels on the left hemithorax, the heart shadow on the right hemithorax suggesting CDH. The haematocrit was 44.9%, Haemoglobin 15.3 g/dl, platelet 447 x 10^3/mcl and the serum electrolytes values includes- Na 134 mmol/l low, K 4.2 mmol/l, CL 107 mmol/l, HCO3 17 mmol/l, and Urea 5 mg/dl, Creatinine 0.7 mg/dl, Ca2+ 8.4 low .TP 6.8 g/dl, alb 2.6 g/dl low. 75 mls of ORh-positive packed RBC’s was made available for surgery. Consent for anaesthesia was obtained.

The neonate was transported from NICU to the operating theatre incubated with size 3.5 endotracheal tube (ETT) connected to an oxylog transport ventilator- (parameter settings RR35, insp pressure 20 cm H20, and 3L/minute volume, fio21 and Spo2 100%). During pre-induction, the patient presented with three 22G cannula for intravenous access and maintenance fluid 5% dextrose in 0.225 saline 60 mls/kg/day. Monitors like pulse oximeter, NIBP, ECG and end tidal CO2 were attached, and baseline vital signs were taken. The baby was wrapped in warm gamgee and placed on the operating table with overlying electrically heated blanket for temperature control. The neonate’s ETT was connected to Mapleson-F
anaesthesia paediatric circuit, and she was manually ventilated, ensuring small tidal volume and high respiratory rates. The fresh gas flow was 3L/minute medical air oxygen mix to achieve 0.5 fio2. Anaesthesia was induced with intravenous (i.v) midazolam 0.3 mg, fentanyl 5 mcg, and maintained with sevofluorane 2% in 50% oxygen throughout. Temperature control was maintained by switching off the cooling system, electrically heated under mattress, and warmed fluids; 30 ml normal saline bolus was given to correct hyponatraemia, 40 mls 5% dextrose / 0.225 saline over the period of surgery- 75 minutes. The clinical parameters were stable without need for inotropic support. The surgical repair was carried out through the transverse upper abdominal incision, the contents of the hernia were reduced, exteriorised and anchoring sutures were used to repair the diaphragmatic defect. The intestines were repositioned, and the abdominal wall was closed in layers. Left sided thoracostomy tube was placed with under-water seal. The intra-operative period was well tolerated, Spo2 100%, sevofluorane was discontinued at the end of surgery, and the patient was transported with Draeger transport ventilator to the neonatal intensive care unit (NICU). The Perioperative ICU management- includes- sedation with 2 hourly I.V fentanyl 5 mcg and midazolam 0.3 mg to facilitate mechanical ventilation; Ventilator settings- fi02 0.5, RR varies initially from 60 bpm to 30 after normalization of arterial blood gas analysis by the end of day 2 as shown in [Table 1], (ABG-by radiometer ABL-800 Basic), Peep 5, Peak inspiratory pressure 15 cm H20, SP02-100%, Hb- 13.9, HCT 37.8% (without need for blood transfusion),and fluid therapy 5% dextrose / 0.225 saline in 60 – 90 mls/kg/24 hrs, antibiotic therapy i.v ampicillin 375 mg twice daily and gentamycin 10 mg once daily. The immediate postoperative X-ray done in the NICU shows re-expansion of the left lung and normal cardiac shadow. The patient had stable haemodynamics, and was extubated following tolerance of intermittent clamping of the thoracostomy tube- which was removed and patient survived and was discharged home in satisfactory condition, after 10 days.

**DISCUSSION**

Isolated left CDH (the defect located on the left side of the diaphragm) occurs in 85% of cases and is associated with high mortality of about 10 to 35% in liveborn;[1,4,5] A regional center with experienced personnel and NICU / PICU resources are required for the optimal care of the critically ill-neonate,[4] as in the management of this patient. CDH commonly presents in the newborn with life threatening respiratory distress requiring immediate intubation plus ventilation as part of resuscitation, and continuous nasogastric suctioning to aid improvement in the lung expansion by decompressing the abdominal contents,[1] as we did for our patient. The severity and outcome of Perioperative management depends on the extent of pulmonary hypoplasia, and pulmonary hypertension (PHT),[1,4] the index patient had respiratory and metabolic acidosis which could lead to the development of PHT; the blood gas results became normal following NICU intervention before surgery. It was reported that the ventilator settings was not found to have any significant impact on mortality.[1,4,6] Until recently CDH was considered as a surgical emergency to be treated under 24 hours, and immediate postnatal repair was the norm,[1,4,6] Emergency repair of the defect in CDH was not found to be accompanied by an improvement in gas exchange, but is often associated with a deterioration in thoracic compliance and Paco2,
and has been abandoned in favour of delayed surgery,[1,4,6] which was adopted in the management of our patient.

It has been reported that the timing of surgery alone does not affect survival, but optimisation or stabilisation of clinical and biochemical parameters before and after the surgical repair is crucial to ensure a good outcome.[4] Initial treatment strategy involves delayed surgical repair after 2 or 3 days of stabilisation deploying gentle mechanical ventilation advocated by Wung and colleagues,[1,7] they suggested maintaining low peak inspiratory pressure, not more than 25 cm H$_2$O plus permissive hypercapnia, along with spontaneous ventilation and SpO$_2$ above 80%. Additional treatment options includes HFOV (achieving mean airway pressures less than 15 cm H$_2$O, and to combat refractory hypercapnia when conventional ventilation fails), inhaled NO and ECMO.[1] Our patient responded well to conventional ventilation as shown by normalisation of ABG. Improved survival was found to occur in the last two decades from improved neonatal intensive care (NICU), neonatal anaesthetic care, surgical techniques, and surgical intervention after initial stabilisation in the NICU for 2 or 3 days – which is termed ”delayed surgical repair”.[4,6,8,9] Surgery may be delayed further if the infant has significant pulmonary hypertension despite appropriate treatment.[4,9]

About 30 to 50% of survivors from CDH develop long-term complications, recurrent respiratory tract infections (RSV infection / pneumonia), chronic pulmonary diseases, bronchial asthma, hypoxic brain injury, and pulmonary hypertension.[1,9] We successfully managed a 2-day-old neonate born in our facility with left congenital diaphragmatic hernia presenting with immediate postnatal respiratory distress, and delayed repair of the defect was carried out - through an open trans-abdominal approach with good outcome; the neonate survived to hospital discharge without any complication.

**CONCLUSION**

Perioperative gentle ventilation strategy and delayed surgical repair of left CDH in a critically ill neonate resulted in good clinical outcome.

**REFERENCES**


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