

A Case of Late Presenting Congenital Diaphragmatic Hernia-Anaesthetic Management

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Abstract

Late presenting Congenital Diaphragmatic hernia beyond the neonatal period though rare, mostly misdiagnosed, mistreated and end up in life threatening complications. CDH can occur at any older age and usually present as mild respiratory symptoms to as severe as gastric obstruction or acute obstructive respiratory problems. Perhaps the error at reaching a definitive diagnosis is simply because of the fact, the possibility of CDH at an older age is totally neglected.

Keywords: Congenital Diaphragmatic Hernia; Acute respiratory distress.

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Introduction

Congenital Diaphragmatic Hernia refers to the defect in the diaphragm, through which the intra-abdominal organs extrude into the thoracic cavity.¹

Though CDH usually presents in the first few hours of life with respiratory distress (from mild dysnoea to cyanosis) it is quite common to have late presentation at an older age. Majority of cases present with nonspecific respiratory and gastrointestinal (GI) symptoms in childhood or early adult life. The prognosis of late-presenting CDH is usually favourable than the CDH presenting in the neonatal period, only if properly diagnosed and surgically corrected.

Case Report

A 9 yr old boy weighing 19kg was brought to the emergency room of our hospital with progressive worsening of dysnoea over 2 days.

In the Emergency room, the boy was in severe respiratory distress with a respiratory rate of 58/min and oxygen saturation of 72% in room air. Child was put on high flow O₂ and his saturation improved to 90 - 92%. Systematic examination showed decreased to no air entry on left side, decreased air entry on right side in basal area with rhonchi and creps. Mediastinum was markedly shifted to right side with bowel sounds in thorax. Heart sounds were shifted to right. Past h/o similar symptoms were there since 5yrs and was treated

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as bronchial asthma and was on conservative management. Neonatal history was uneventful and didn't suggest any acute respiratory distress at birth.

A CXR taken at emergency room, showed bowel and stomach herniated to left side with subnormal lung expansion on left with air and fluid in left hemithorax, there was no liver herniation. (Figure 1).



Fig. 1



Fig. 2

A CT chest taken, showed Direct discontinuity of left hemidiaphragm with intrathoracic extension of abdominal contents diaphragmatic structure with hernia. Collapse consolidation of posterior segment of right upper lobe and superior segments bilateral lower lobes medial segments. And Provisional diagnosis of Left diaphragmatic hernia was made.

After getting written high risk informed consent, child was posted for emergency repair of diaphragmatic hernia. Preoperative investigations were nearly normal and preop ABG showed:

PH:7.381, PO₂ :70, PCO₂ :35.4, HCO₃ :20.6, SO₂ : 93.6.

On arrival to OT, child's vitals noted as-Heart rate:124/min, BP: 80/60 mmHg, SPO₂: 92%, Respiratory rate:50/min. 100% oxygen was given via facemask, a nasogastric tube and a urinary catheter were placed.

Pre-op monitors ECG, NIBP and Pulse oximetry were attached. Child was premedicated with Inj. perinorm 4mg, Inj.gylcopyrrolate 0.2mg and Inj. Fentanyl 40mcg. A Modified Rapid sequence Induction was done with Inj Propofol 40mg IV, skeletal muscle relaxant, Inj Rocuronium 20mg was given and at 1 minute child was intubated with ETT of size 5.5 mm and fixed at 16 cm. Anaesthesia was maintained with Air: oxygen at 1:1 to maintain end tidal isoflurane between 0.8 to 1 MAC. Patient was ventilated in PCV mode with settings TV: 120ml, PEEP: 5, RR: 33/min, A/W pressure: 30.

After Initially attempts of laparoscopic reduction of abdominal organs, the diaphragmatic elements were found to be very thin and minimal, surgeon proceeded with open diaphragmatic hernia repair. Intraoperatively patient was stable with SPO₂ at 93%, HR- 120/min, BP- 100/60, ETCO₂ - 52. Diaphragmatic repair was closed and hypoplastic lung was released and ICD was placed on left side. Duration of surgery lasted for 4 hrs with an intraoperative blood loss of about 200ml and had urine output - 50ml. Intraoperative fluid Mx was done with DNS as per Holiday Segar formula.

There was no episodes of hypotension, hypoxia, and hypothermia in intraoperative and immediate postoperative period.

CXR was done in the immediate postoperative period showed improved air entry into left lung. (Fig 2).

On POD1, child developed fever, tachycardia and hypotension with subcutaneous emphysema extending to the neck region. Chest X ray shows features suggestive of pneumomediastinum and pneumothorax.

ABG was done

ABG s/o respiratory acidosis (Ph - 7.32, PCO₂ - 51, PO₂ - 76, HCO₃ - 25). Urine output decreased. Child was managed with dopamine, I/V fluids, and antibiotic was changed to Meropenem. ICD was repositioned. Over the next few days, child improved and child was extubated on 5th postoperative day but was continued on high flow oxygen for two more days. ICD was removed and child got discharged on 14 th postoperative day.

Discussion

The CDH can be a challenging case as it needs a teamwork of pediatrician, surgeon as well as anaesthesiologist. Clinical manifestations of late presenting CDH are so diverse that gastrointestinal symptoms-vomiting, abdominal pain and respiratory symptoms-dyspnea, cough, cyanosis can present alone or in combination. Chest x-ray, which is performed routinely in patient with these symptoms can mimic pleural effusion, pneumonia, or pneumothorax, which can lead to misdiagnosis and serious iatrogenic complications like gastric perforation by chest tube thoracotomy. When chest X-ray images are not diagnostic, spiral CT and MRI should be done for accurate diagnosis and management. If surgical treatment would not be followed promptly after accurate diagnosis for late presenting CDH, various disastrous complications such as intestinal strangulation, necrosis of herniated organs, hypersplenism, cardiopulmonary dysfunction, and short bowel syndrome, gastric volvulus, and death can be inevitable. If timely diagnosed and correctly repaired, their outcomes are mostly excellent because they have little or no lung hypoplasia.

Patient with CDH should be considered as full stomach because of possible gastrointestinal obstruction, and, therefore, these patients required aspiration prophylaxis. Nasogastric tube should be inserted and aspirated before induction. Large gauge intravenous access is necessary to manage any hemodynamic instability.

Rapid sequence induction with cricoid pressure must be the induction of choice, but when difficult airway is anticipated, awake fiberoptic intubation is the gold standard technique. Any event which increases intraabdominal pressure especially

during induction, intubation, and extubation is detrimental. Positive pressure ventilation with potential gastric insufflation and expansion of compressed lung may decrease venous return and cardiac output. For the same reason protective ventilation strategies that avoid further injury to damaged lung tissue must be executed. The CDH EURO Consortium advocates aiming for the limitation of peak inspiratory pressures to 25 cm H₂O with PEEP kept at 3-5 cm H₂O and allowing permissive hypercapnia. Nitrous oxide may also worsen mass effect should, therefore, be avoided.

Conclusion

Late presenting CDH can have a wide spectrum of clinical manifestation. As it can progress to life threatening conditions such as CDH with gastric volvulus. A high index of suspicion is vital for the timely diagnosis and management.

As survival improves, further studies are required to ensure that the patient have a high quality of life after discharge, which can only occur with regular follow ups by multi disciplinary team.

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Erratum

Article Titled: "Intrathecal Bupivacaine vs Bupivacaine and Clonidine in Paediatrics Age Group: A Comparative Evaluation"

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The original published version of this Article contained errors in the designation and Institution name of the first author mentioned. Now Designation and Institution name of the first author in article re-added as

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