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Review Article

Perinatal Management of Congenital Diaphragmatic Hernia

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Abstract

Congenital diaphragmatic hernia (CDH) carries significant mortality and pulmonary morbidity. CDH morbidity is mainly due to herniation of abdominal viscera into the chest cavity leading to underdevelopment of the lungs which is a major contributor to the development of pulmonary hypertension. Pulmonary hypertension is a major contributor to both morbidity and mortality in CDH.

The aim of this review is to discuss the management of CDH including prenatal, delivery room, and postnatal periods. The later period includes a discussion of the use of a fraction of inspired oxygen, surfactant therapy, gentle ventilation, mode of ventilation, medical management of pulmonary hypertension, the utilization of extracorporeal membrane oxygenation, and the timing of surgical repair.

Keywords: Congenital Diaphragmatic Hernia; Persistent Pulmonary Hypertension

Introduction

Congenital diaphragmatic hernia (CDH) is relatively a rare birth defect that affects around 1 in 3000 live births. It occurs 90% of the time in the posterolateral aspect of the diaphragm known as a Bochdalek hernia. Over 85% of CDH is occurring on the left side. Most studies have not observed a sex association or any association with maternal age [1, 2].

Despite improvements in both prenatal and postnatal management, CDH is still associated with significant neonatal mortality, with rates varying between 30% and 50% and the majority of deaths occurring in the first year of life [3].

Approximately 50 to 70 percent of cases of CDH are isolated. Pulmonary hypoplasia, intestinal malrotation, and cardiac dextroposition are considered part of the CDH sequence [4, 5]. While 30 to 50 percent of CDH cases are called "complex," "nonisolated," or "syndromic". These cases are associated with additional abnormalities, including major structural malformations, chromosomal abnormalities, and/or single gene disorders. Congenital heart defects (25%–40%), urogenital anomalies (18%), musculoskeletal anomalies (16%) and CNS anomalies (10%) occur respectively in CDH [4, 5].

Discussion

Pathophysiology

The whole mark of CDH is failure of normal closure of the pleuroperitoneal folds during the 4th to 10th weeks postfertilization allows herniation of viscera into the thoracic cavity [6]. This interferes with normal lung development and has several potential adverse consequences, including reduction in bronchiolar branching and loss of pulmonary mass, surfactant system dysfunction, over-muscularization of the pulmonary arterial tree, all together can lead to persistent pulmonary hypertension.

CDH can also lead to hypoplasia of ipsilateral cardiac structures [6].

Survival

The postnatal survival rate at tertiary centers has improved, with reported rates of 70 to 90 percent. These data represent the survival rate of cases of CDH that were full term infants born or transferred to tertiary care centers with available skilled personnel and access to advanced technology (eg, extracorporeal membrane oxygenation [ECMO]) [7].

Postnatal survival appears to be less likely in the setting of [8, 9]:

- Abnormal microarray or findings suggestive of a fetal syndrome
- Serious associated anomalies
- Liver herniation
- Lower fetal lung volume
- Cardiac abnormalities
- Persistent and Severe Pulmonary Hypertension
- Prematurity
- Site of care and need for transport
- Defect size
- Right- versus left-sided lesion

Marisa E. Schwab et al showed in a study that survival rate with liver herniation is 45 percent versus without herniation is 74 percent [8]. In two studies using prenatal MRI to evaluate the lung volume showed postnatal survival is poor when fetal lung volume measured by MRI is less than 30 percent of expected lung volume for gestational age and especially when less than 15 percent [10, 11].

Ultrasound examination is used to calculate the lung area to head circumference ratio (LHR), defined as the area of the contralateral lung at the level of the 4-chamber view of the heart divided by the head circumference [8]. Observed [o]/expected [e] LHR is directly related to Survival rates: extreme low lung volume defined as LHR<15%, while severe 15-25%, moderate 26-35% and mild 36-45% has survival rate of zero, 20%, 30-60% and 75% respectively [12].

Prenatal Management Of CDH

Fetal Endoscopic Tracheal Occlusion (FETO):

Occluding the trachea of fetuses with CDH increases lung volume, decreases herniation of abdominal viscera, and improves postnatal lung function [13].

Fetal assessment:

Serial ultrasound examinations, biophysical profiles, and estimating fetal weight are recommended for all pregnancies with fetuses with CDH

Antenatal glucocorticoids:

Antenatal glucocorticoids are administered when preterm birth is anticipated [5].

Timing and Mode of Delivery:

It is suggested that a planned induction of labor at 39 weeks of gestation to minimize complications from early delivery. Cesarean birth is performed for standard obstetric indications; there is no evidence that routine cesarean birth is beneficial [5, 7, 14].

Delivery Room Management Of CDH

Commencing resuscitation and initiating ventilation while the infant is still attached to the placenta is feasible in infants with CDH. The procedure may support the cardiorespiratory transition at birth in infants with CDH [15, 16].

However, The Canadian and EURO CDH guidelines recommend intubating infants with prenatally diagnosed CDH immediately after birth as a standard of care [1, 5].

Nicu Management Of CDH

Naso or Orogastric Tube Insertion

The CDH EURO Consortium Consensus recommends immediate placing of an oro- or nasogastric tube with continuous or intermittent suctioning in order to prevent bowel distension and any additional ipsilateral lung compression [5].

Surfactant Administration

Although administration of surfactant therapy has been suggested in treating infants with CDH, it does not appear that surfactant administration improves outcomes. For that surfactant should be administered in preterm neonates with chest radiographic findings suggestive of respiratory distress syndrome (RDS) [1, 5].

Echocardiography

Echocardiography is performed early to detect any associated cardiac anomalies, establish the presence and severity of persistent pulmonary hypertension (PPHN) and shunting, and assess ventricular function, as these factors impact management decisions [1, 5, 18]. The echocardiographic signs of PPHN include poor contractility of the right ventricle, enlarged right heart chambers, pulmonic and tricuspid valve regurgitation, and presence of ductal shunting [18].

Ventilation

Ventilation strategy is aimed at minimizing lung trauma to hypoplastic lungs, which contributes to mortality and morbidity [1, 17]. Ventilation management should aim to use the minimal settings to maintain [1, 17]:

1. Preductal saturation 85% - 95%.
2. Arterial PaCO2 between 45 and 60 mm Hg
3. PH between 7.25 and 7.40

Type of ventilation

Conventional mechanical ventilation (CMV) management consists of pressure control ventilation with target PIP less than 25 cm H2O and target PEEP of 5 cm H2O, aiming for lung volumes of 4-6 ml/kg [1, 17]. Hyperventilation, hypocarbia, and alkalosis may decrease ductal shunting and control pulmonary hypertension in CDH, but at the expense of increased barotrauma. Permissive hypercapnia has been used in neonates with CDH, with reports of increased survival compared with hyperventilation and alkalization [1, 17].

High Frequency Ventilation

High-frequency Ventilation should be considered for neonates who continue to have hypoxia and hypercarbia (PaCO2 >65 mmHg) refractory to CMV [1, 17].

Settings:

- Mean airway pressures (MAPs) of 1-2 cm H2O above the MAP on the CMV.
- Frequency 6 to 8 Hertz, with adjustments made based on gestational age.
- Amplitude 2-2.5 of MAP.

Paralysis and Sedation

Sedation should be provided to all mechanically ventilated newborns with CDH & PPHN. Deep sedation and neuromuscular blockade should be provided selectively to those with greater ventilation or oxygen [1, 17].

Hemodynamic Support

Treatment of poor perfusion (capillary refill > 3 s, lactate > 3 mmol/L, urine output < 1 mL/kg/h) and blood pressure below norms for age should include [1, 5]:

- judicious administration of crystalloids, generally not exceeding 20 mL/kg
- inotropic agents such as dopamine or epinephrine
- hydrocortisone.

Management Of Persistent Pulmonary Hypertension In CDH

There is no universally effective single agent for treating cardiac function and persistent pulmonary hypertension in CDH [1, 5].

- iNO is indicated for confirmed suprasystemic pulmonary arterial hypertension without left ventricular dysfunction, provided lung recruitment is adequate.
- Sildenafil should be considered in patients with refractory pulmonary hypertension (i.e., unresponsive to iNO) or as an adjunct when weaning iNO.
- Prostaglandin E1 can be used to maintain ductus arteriosus patency and reduce right ventricular afterload in patients with pulmonary hypertension with right ventricular failure.
- Milrinone (19)

Milrinone is phosphodiesterase 3 (PDE3) inhibitor. The indications of milrinone in pulmonary hypertension include:

1. Ventricular dysfunction:

- It has inotropic (enhance cardiac contraction) and lusitropic (augment cardiac relaxation) effects.

- Milrinone causes pulmonary and systemic vasodilation and reduces afterload on both ventricles.

2. As an adjuvant to iNO to promote pulmonary vasodilation.

Extracorporeal Membrane Oxygenation (ECMO)

There are no randomized controlled studies that have demonstrated the efficacy of ECMO in infants with CDH. ECMO, if available, is considered for infants who cannot be managed with maximal conventional medical therapy (maximal ventilatory support, inotropic support for BP and iNO), as these patients would not survive without ECMO [5, 20].

Criteria to Consider ECMO (5,20)

1. Inability to maintain preductal O2 saturations >85 percent or postductal PaO2 >30 mmHg.
2. Inadequate oxygen delivery with persistent metabolic acidosis
3. PIP >28 cm H2O or mean airway pressure (MAP) >15 cm H2O
4. Hypotension that is resistant to fluid and inotropic support

Inclusion Criteria for ECMO (5,20)

1. Birth weight (BW) >2 kg
2. Gestational age (GA) >34 weeks; determined on a case-by-case basis 32-34 weeks
3. Absence of intracranial hemorrhage greater than grade I
4. Absence of chromosomal anomalies

Surgery

Timing of surgery With a better understanding of the pathophysiology and variation in the degree of pulmonary impairment, the timing of surgery has shifted from early surgical intervention to delaying surgical correction until the patient has been stabilized medically. However, the timing of repair remains controversial without any definitive study that demonstrates superiority of early (within 24 hours) or delayed repair [1, 2].

The following physiologic criteria should be met before surgery [1, 5]:

- Urine output > 1 mL/kg/h
- FiO2 < 0.5

- Preductal oxygen saturation between 85% and 95%
- Normal mean arterial pressure for gestational age
- lactate < 3 mmol/L
- Estimated pulmonary artery pressures less than systemic pressure. Failure to meet these criteria within 2 weeks should prompt consideration of either attempted repair or a palliative approach [1, 5].

Acute Complications Post-repair of CDH

The most serious complication post-repair of CDH is persistent pulmonary hypertension (PPHN). Some patients may require extracorporeal membrane oxygenation (ECMO). Other complications early in the postoperative course include hemorrhage, chylothorax, and infection (patch infection, sepsis, and urinary tract infection) [21].

Conclusion

The care of neonates with congenital diaphragmatic hernia (CDH) remains challenging with no standard of care across institutions. Although many studies are conducted to evaluate treatment strategies, no large, multi-institutional studies exist to help determine the best practice. Development of multi-institutional treatment guidelines with prospective data collection and review may be the only way to determine the best practice in management of CDH which could protect the hypoplastic lungs from damage and improve survival rates among infants with CDH.

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Conflicts of Interest

The author declares no conflicts of interest.

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