

Management of congenital diaphragmatic hernia

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Abstract

Congenital diaphragmatic hernia (CDH) is associated with high mortality due to lung hypoplasia, pulmonary hypertension and co-existent anomalies. This review highlights recent progress in the perinatal management of CDH and addresses long-term outcome issues for survivors indicating the need for multidisciplinary follow up.

Keywords congenital diaphragmatic hernia; fetal therapies; minimally invasive surgery; outcomes

Introduction

Definition and aetiology

Congenital diaphragmatic hernia (CDH) is a defect in the fetal diaphragm allowing the contents of the abdominal cavity to protrude into the thorax. CDH has an incidence of 1 in 2500 live births in the UK. The birth defect may be associated with other major anomalies and the lesion may become apparent in the fetus, newborn or older child. Some forms of CDH remain asymptomatic and may not present until adulthood.

The majority of defects are sporadic and isolated (70%) with a number of gene mutations identified, e.g. Deletions of 4p, 8q, 15q. Links have been made to environmental factors such as: thalidomide, nitrofen and vitamin A deficiency. CDH may be associated with other chromosomal abnormalities e.g. Fryn's and Pallister Killian syndromes.

Pathology

The precursors of the diaphragm begin development during the 4th week of gestation in the form of the septum transversum and lateral folds of mesenchymal tissue. These partition the abdominal and thoracic compartments with the formation of the pleuroperitoneal membrane occurring by the 8th week. As development continues muscle fibres migrate into this membrane. Failure of these stages of development will result in either a 'true' diaphragmatic defect (CDH) or a complete, yet hypoplastic, diaphragm resulting in a diaphragmatic eventration.

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Closure of the right hemidiaphragm occurs before the left, which may account for the higher incidence of left sided diaphragmatic defects (84%). Right diaphragm defects comprise 13% of lesions and 2% are bilateral defects, the remaining cases comprising the rarer variants such as complete diaphragmatic agenesis. The commonest lethal form of CDH is the lesion occurring in the posterolateral aspect of the developing diaphragm notably the Bochdalek hernia with other less common defects e.g. in the anterior-lateral diaphragm notably the Morgagni hernia.

Outcome in CDH remains highly variable with regard to prognosis in comparison to other neonatal surgical conditions, with reported mortality rates varying between 20 and 60% in centres worldwide. The morbidity and mortality of CDH is traditionally related to the mechanical compression of the herniated viscera on the developing lung leading to pulmonary hypoplasia and pulmonary hypertension. The histological changes seen at post mortem in the underdeveloped ipsilateral lung on the affected side of the defect are also mirrored in the contralateral lung suggesting that whatever insult is responsible for the failure of the diaphragm to develop may also have a global impact on the primordial development of the respiratory system.

Antenatal care (fetal CDH)

Antenatal diagnosis

Although not always identified in the antenatal period all cases detected prenatally should ideally be referred early to a specialist centre. Antenatal diagnosis allows counselling by a multidisciplinary team (paediatric surgeons, neonatologists and obstetricians) as well as allowing treatment and delivery planning. Antenatal diagnosis has been reported as early as 11th week of gestation, however, in the UK, it is more often diagnosed at the 20 week anomaly scan. Within the wider European region a multicentre study found that 60% of cases of CDH were identified antenatally with a mean gestational age of 24.2 weeks at diagnosis. Recent studies suggest that care in specialist, high volume centres (more than 5 cases per annum) achieves an improved survival rate compared to low volume centres.

Associated anomalies

Antenatal investigations should attempt to identify other associated anomalies, to permit more accurate counselling, as survival in newborns with associated anomalies remains dismally poor (less than 10%). Amniocentesis is recommended to identify chromosomal anomalies, present in up to 10% of cases, e.g. Trisomies 13, 18 & 21, Donnai-Barrow and Fryns syndromes. A thorough sonographic evaluation of the urinary, gastrointestinal and central nervous system should be performed to detect further structural malformations (33%). Echocardiography is utilised to detect cardiac anomalies, present in up to 18% of cases. It may also usefully be deployed to measure pulmonary artery diameters, which have been found to be predictors in some series of postnatal pulmonary hypertension and mortality.

Predicting outcome

Defining antenatal predictors of outcome in CDH has been the subject of intense study over the last few years. Most current methods rely on indirect techniques, e.g. US/MRI to make an

assessment of fetal lung volume. Measurement of the contralateral fetal lung area to head circumference ratio (LHR), using 2D ultrasound (US) imaging, as a predictor of outcome was first proposed by Harrison's group based at UCSF, San Francisco in the mid 1990s. LHR gained popularity as a good prognostic marker and led to a randomised clinical trial that guided decision making for fetal intervention in CDH. A systematic review and meta-analysis conducted in 2007 suggested that further refinement with regard to LHR criteria was needed to more accurately predict fetal outcome. More recently LHR measurements have been improved to account for the four fold relative increase of lung area to head circumference that occurs between 12 and 32 weeks in the fetal period. Jani and colleagues (antenatal-CDH registry group) utilizing the observed to predicted (O/E) LHR indicated more accurate prognostic scoring in 354 cases of unilateral, isolated CDH.

Aside from determining fetal lung volumes with ultrasound other useful potential markers guiding prognosis antenatally include the presence of liver herniation and the role of fetal MRI lung volumes. Studies have shown that fetuses with an increased volume of liver within the thoracic cavity, designated "liver up" cases, are associated with a larger hernia defect, a greater need for prosthetic patch repair and decreased survival.

Fetal intervention

Initial interest in fetal intervention concentrated on open repair of the diaphragmatic defect following maternal hysterotomy. Trials were discontinued due to preterm labour with poor outcome. The dramatic changes seen in lung growth achieved by occluding/'plugging' the fetal trachea, led to refinements in fetal surgical techniques for CDH. In 2003 Harrison et al., reported a randomised controlled trial of fetal endoscopic tracheal occlusion (FETO) showing equivalent survival to a group of 'high risk' fetuses managed by conventional postnatal care in specialist CDH centres. This study led to further efforts to improve better case selection for fetal intervention using LHR (O/E) and 'liver up' entry criteria to identify those with the very worst prognosis that may justly benefit from FETO procedures.

European fetal medicine programmes using selective entry criteria (O/E LHR less than 27–28% and liver herniation) have found a statistically significant improved survival rate in a FETO treated CDH group in comparison to same severity controls. Results from a European prospective randomised trial (currently in progress) are awaited.

Newborn management — postnatal care

Delivery

With an increasing antenatal detection rate of CDH expert opinion regarding the mode of delivery remains a subject of debate. In 2007 the CDH study group interestingly reported a marginal (non significant) survival benefit for elective delivery by caesarean section. Further randomised studies are needed to draw definitive conclusions.

In order to maximise pulmonary development delivery should be planned as near to term (more than 37 weeks) as possible. Delivery should be co-ordinated in specialist centres equipped with full neonatal intensive care facilities with ready access to paediatric surgeons. Elective intubation following birth and

'gentle' ventilation (avoiding barotrauma) is recommended. All babies should have a nasogastric tube promptly inserted to avoid gastric distension and vascular access secured to aid delivery of fluids and pharmacological agents. Following stabilization a full clinical examination is required to exclude associated anomalies. Chest X-ray confirming the diagnosis and echocardiogram is performed to screen for cardiac anomalies.

Postnatal diagnosis — 'late presenting CDH'

Despite antenatal imaging 30% of patients with CDH may remain undetected until after delivery. These cases may present in the immediate newborn period or first few days after birth whilst others may remain asymptomatic until later life. Symptoms may include mild respiratory distress or feeding problems. Delayed presentation may occur with small diaphragmatic defects in which there is little or no herniated bowel at birth. Herniation of intestinal viscera as a later dynamic event may follow an episode of increased intra-abdominal pressure seen with a respiratory tract infection.

Clinical examination may reveal bowel sounds on chest auscultation. There may be signs of decreased air entry on the affected side and rarely mediastinal shift. Diagnosis is often made on chest X-ray but may require an upper gastrointestinal contrast study for further confirmation.

Stabilization

'Gentle' ventilation — a major advance in the management of CDH in the last 20 years has been the introduction of 'gentle ventilation' strategies (permissive hypercapnea) to reduce iatrogenic lung injury from barotrauma. Wung et al., introduced this novel concept characterised by preservation of spontaneous ventilation, permissive levels of hypercapnea (paCO₂ 60–65 mmHg or 9 kPa) and avoidance of high inspiratory airway pressures (ideally not exceeding 25 cm H₂O). A number of specialist centres steadily report improving outcomes (more than 80% survival) with this approach together with a reduced need for ECMO. High frequency oscillatory ventilation (HFOV) has also been utilised in the perinatal management of CDH both as a 'rescue therapy' prior to extracorporeal membrane oxygenation (ECMO) and as a primary ventilatory modality in an attempt to reduce pulmonary barotrauma. There have been several reports of increased CDH survival with HFOV strategies.

Other therapies

ECMO has been deployed to treat respiratory failure and pulmonary hypertensive crisis in CDH following failure of conventional therapies. Throughout the 1980s and early 1990s ECMO gained steady momentum in the management of 'high risk' newborns with CDH. A UK multicentre randomised controlled trial and a Cochrane review however failed to demonstrate significant survival benefits for the use of ECMO in CDH. Currently with the introduction of permissive hypercapnea strategies achieving improved CDH survival there has been a steady decline in the use of ECMO across many centres. Many major institutions continue to deploy ECMO with variable reported success. Refined entry criteria (ECMO eligibility) may influence outcomes.

Further methods to treat pulmonary hypertension associated with CDH have included the use of inhaled nitric oxide (iNO)

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