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Best practice guidelines

Congenital diaphragmatic hernia

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ABSTRACT

There is a paucity of level 1 and level 2 evidence for best practice in surgical management of CDH. Antenatal imaging and prognostication is developing. Observed to expected lung-to-head ratio on ultrasound allows better predictive value over simple lung-to-head ratio. Based on 2 randomised studies, the verdict is still out in terms the best group and indication for antenatal intervention and their outcome. Tracheal occlusion is best suited for prospective randomised studies of benefit and outcome. Only one pilot randomised controlled study of thoracoscopic repair exists, suggesting increased acidosis; blood gases and CO_2 levels should be closely monitored. Only poorly controlled retrospective studies suggest higher recurrence rates. Randomised studies on the outcome of thoracoscopic repair are needed. Careful selection, anaesthetic vigilance, monitoring and follow-up of these cases are required. There is no evidence to suggest the best patch material to decrease recurrences. Evidence suggests no benefit from routine fundoplication based on the one randomised study. Multi-disciplinary follow-up is required. This can be visits to different specialities, but may be best served by a multi-disciplinary one-stop clinic.

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1. Introduction

Congenital diaphragmatic hernia (CDH) is characterised by a spectrum developmental defects in the diaphragm caused by disordered embryogenesis, resulting in incomplete fusion of elements giving rise to the diaphragm. Ninety percent of CDH cases are found in a posterolateral defect (Bochdaleck hernia), and 9% are found in an anteriomedial defect (Morgagni hernia). The remainder of cases comprise the relatively rarer forms of total absence of the diaphragm, absence of the central portion of the diaphragm, and oesophageal hiatal hernia. Most of classic CDH diagnosed at birth comprise the Bochdaleck type with a varying degree of the size of the defect. The incidence of CDH is 1 in 2,500 to 1 in 3,500 live births. Left-sided CDH is more common than right-sided, with a ratio of 6:1. Bilateral lesions are reported, but they are invariably fatal.

Antenatal diagnosis and prognostication is increasing with sophisticated imaging techniques. Non-antenatally diagnosed cases can present in the early postnatal period with respiratory distress. Associated chromosomal abnormalities, lung hypoplasia, pulmonary vascular abnormality

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and cardiac abnormalities lead to a high mortality. Prompt neonatal management is the most important influence on outcome. Surgical correction has become a non-emergent secondary intervention.

Lung hypoplasia plays a significant role in determining clinical course and outcome. The development of type II alveolar cells that produce surfactant is also inhibited, resulting in relative surfactant deficiency. Antenatal prediction of outcome in isolated CDH is based on this associated lung hypoplasia. The abnormal development of the pulmonary vasculature leads to pulmonary hypertension and increased pulmonary vasculature reactivity. The neonate is prone to episodes of hypoxia and hypercapnia, which in turn further increase the pulmonary hypertension and cause persistent foetal circulation. Persistent foetal circulation further worsens the hypercapnia and hypoxia. This viscous positive cycle can lead to severe physiological consequences in those most affected. The lung hypoplasia along with pulmonary hypertension is a detrimental patho-physiological process that affects outcome.

Surgical management has changed. Antenatal assessment and intervention has been used to surgically treat patients with predicted poor outcome. Surgical morbidity is associated with the size of the defect and the need for patch repair. The advent of minimally invasive approach to surgical repair has developed but requires expert management of acidosis and oxygenation. Long-term morbidity (affecting multiple systems) is also high and can be related to both the underlying diagnosis as well as treatment. Feeding difficulties is common and gastro-oesophageal reflux can need surgical management.

This review will summarize the available evidence for best practice in surgical management of CDH; antenatal imaging and intervention, operative approach, type of patch repair, surgical management of reflux and address long-term follow-up.

2. Antenatal imaging and prognostication

Routine antenatal ultrasound scanning detects approximately 50–85% cases of CDH. Lung-to-head ratio (LHR: contralateral lung area to head circumference) measured by antenatal ultrasound is used to predict the severity and outcome in CDH and to predict patients with poor outcome for antenatal intervention. There are reports suggesting a good correlation, although some reports highlighted inconsistency in the predictive value of measured LHR [1]. This may be due to the 'learning curve' associated with the fast developing area of antenatal imaging, interpretation and prognostication. Part of the discrepancy may be due to different means and timing of estimating LHR. A uniform technique to avoid inter-operator variation and to unify data has been suggested by Jani et al. [2]. By standardising the methods of measurement using the longest diameter, the anteroposterior diameter at mid clavicle or a tracing method (preferably the latter as it seems to be most reproducible), errors in measurements can be avoided and unification of technique should allow better correlation. There are also problems inherent on using isolated LHR, as LHR changes with gestational age [2] and tends to underestimate the severity of the defect [3].

Greater consistency and accuracy in predictive value is obtained by using the observed to expected LHR (O/E LHR), and magnetic resonance imaging (MRI) total lung volume (TLV). The O/E LHR was developed in response to the observation that lung growth is 4 times that of head growth in the 3rd trimester [3]. The Antenatal-CDH-Registry Group [4] demonstrated that the O/E LHR (by taking a transverse section of the foetal chest demonstrating the four-chamber view of the heart and multiplying the contralateral lung area's longest diameter by the longest perpendicular to it) eliminates the effect of gestational age. In foetuses with both left- and right-sided CDH, the measurement of the O/E LHR provides a useful prediction of subsequent survival. The O/E LHR is lower in foetuses with CDH compared to normal foetuses, and lower still in babies who die with CDH than those who survive [2]. There was however some overlap in values between survivors and non-survivors. The survival for left sided lesion related to O/E LHR with liver down was as follows: ≤25%:30% survival; 26–35%:62% survival; 36–45%:75% survival; 46–55%: 90% survival; and >55%: 85% survival [4].

Although LHR is predictive of mortality in CDH, it is not strongly correlated with morbidity.

2.1. MRI

Foetal MRI may have a role in providing more specific information to aid prognostic decision, and can be offered at 24 and 34 weeks gestation. Contralateral lung volume/TLV on MRI strongly correlates with lung area measured on US. There is currently no evidence that foetal MRI is superior to ultrasound at predicting outcome. In one study, foetal MRI TLV permitted calculation of lung volumes, but these volumes were not predictive of outcome [5]. This may depend on the precise method used. Alternatively, foetal MRI for lung volume may yield additional useful information (e.g. % of liver herniation) and give better receiver operator curves for prediction [6]. O/E TLV obtained by MRI scan correlates with US derived LHR but without the operator dependant nature of measurements and maternal and foetal motion artefacts [6].

Best practice: Use of O/E LHR on ultrasound allows better predictive value over LHR alone. Standardisation of the method of measurement should allow better reproducibility.

3. Antenatal intervention

Although antenatal trachea occlusion (TO) was suggested as a potential inducer of increased foetal lung growth in animal models, clinical benefits are not always clear from the research done to date. Initial investigation showed mixed results in terms of both lung growth and survival in cohort studies. Further advance on the technique and management has allowed units to offer antenatal foetal endoscopic tracheal occlusion (FETO) by endoscopic plugging in management of severe cases of CDH. There have been a few randomized trials reporting outcome of TO.

Harrision et al. [1] randomized 24 patients with left CDH, liver up and LHR <1.4 and assessed the survival effect of TO performed via maternal laparotomy. Early termination of trial enrolment was needed because survival in the control group was better than anticipated and benefit was not evident from the plugging. There may have been some modest improvements in lung function but no discernible clinical benefit and no differences in neurodevelopmental, respiratory, surgical, growth and nutritional outcomes at 1 and 2 years of age.

By contrast, Ruano et al. [7] randomised 41 foetuses (any side; LHR < 1.0, liver herniation and no other detectable anomalies; 38/41 received allocated treatment) and found a significant 6-month survival advantage associated with antenatal plugging (received treatment analysis, 10/19 (52.6%) infants in the FETO group and 1/19 (5.3%) controls survived; relative risk 10.0; 95% CI, 1.4–70.6).

There are differences in both studies which may account for the difference in outcome between them. There were differences in the LHR which allowed inclusion into both studies. The more severely affected foetuses in the latter study may have therefore benefited from TO, whereas the effect on less severely affected foetuses was not marked. There were differences in technique (maternal laparotomy versus fetal endoscopic approach), which may have influenced incidence of preterm labour, resulting in a 5-week difference in the gestational age at delivery between both studies, with obvious confounding effects. Further studies are needed to clarify the patient groups that may benefit from this intervention.

Cohort studies demonstrate that there is an antenatal response (pre- and post-FETO O/E LHR or TFLV) to plugging in foetuses with CDH which may provide independent prediction of postnatal survival. Jani et al. [8] reported on multicentre study of 210 FETO cases. They found a survival advantage when compared with expected survival as predicted by O/E LHR. In addition to the obstetric complications of FETO, the complication to the foetus includes tracheal changes (dilation and