

## **ANTENATAL EVALUATION AND MANAGEMENT OF CONGENITAL DIAPHRAGMATIC HERNIA**

### **Initial Diagnosis of Congenital Diaphragmatic Hernia**

Congenital diaphragmatic hernia (CDH) occurs in 1 per 3,000-5,000 live births. Over half of the cases are diagnosed antenatally. In addition to confirmation of the diagnosis, parents should receive information on how the condition will affect their baby after birth. In order to facilitate an accurate, individualized prognosis and counselling of all current management options by a multidisciplinary team, parents should be offered referral to the regional specialist fetal medicine service within five days. It is also recognized that this practice will offer the potential advantage of a “second opinion”, which may benefit many families.

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### **Key Points at Initial Antenatal Diagnosis**

- Following antenatal diagnosis of CDH, in addition to consideration of differential diagnoses and assessing for other fetal anomalies, the woman should be offered referral to the regional specialist fetal medicine service within five days.
- An information leaflet is available from the Scottish Diaphragmatic Hernia Clinical network (SDHCN) that can be used for counselling at this stage. If leaflets are not available locally, a copy can be downloaded from the SDHCN website ([www.sdhcn.scot.nhs.uk](http://www.sdhcn.scot.nhs.uk)).

### **Initial Evaluation at Fetal Medicine Department**

At the initial appointment with fetal medicine the following will be assessed; side of defect, liver and stomach position. Factors linked with good prognosis are; left-sided CDH, liver in abdomen, and isolated CDH with no other anomalies. A fetus with liver herniation has a poorer prognosis (see table in next section). Associated anomalies can occur in 30-60% cases, these include cardiac defects (52%), genitourinary (23%), gastrointestinal (14%) and central nervous system anomalies (10%). Following ultrasound assessment, the woman/family will be counselled regarding CDH and the options of continuing with or terminating the pregnancy will be discussed. Karyotyping will usually be offered (if not already performed locally). The SDHCN antenatal information leaflet should be provided at this stage.

### **Key Points at Initial Evaluation at Fetal Medicine Department**

- At the initial appointment with fetal medicine, the side of defect, liver and stomach position will be assessed
  
- The woman will be counselled regarding CDH, and will be offered karyotyping (if not already performed)
  
- Options for management of pregnancy will be discussed and will be influenced by parental preference, scan findings, and presence of associated anomalies.
  
- SDHCN antenatal information leaflet should be provided. If leaflets are not available locally, a copy can be downloaded from the SDHCN website ([www.sdhc.scot.nhs.uk](http://www.sdhc.scot.nhs.uk)).
  
- Relevant clinical and sonographic details should be recorded on the SDHCN database by the local coordinator.

## Review until 32 Weeks Gestation

Detailed ultrasound examinations will be performed at the regional specialist fetal medicine department between 22–28 weeks gestation in order to assess lung head ratio, liquor volume, to look for associated structural anomalies, and to perform fetal echocardiography. Fetal anatomical surveys are ideally performed around 22–24 weeks gestation for cardiac and structural assessments. Further scans to check on fetal growth and wellbeing may be organised locally depending on individual cases and following discussion between clinicians.

In order to evaluate fetal lung volume, lung head ratio (LHR) is calculated. This is obtained by measuring the size of the contralateral lung and considering it in proportion to the body by also measuring the head circumference. In the U.K, two different methods of lung measurements are used currently. Therefore, the method of calculating the LHR should be documented i.e. longest axis or AP diameter method (see appendix, adapted from <http://www.totaltrial.eu>). It is worth noting that most European studies have used the longest axis method.

Because the lung and head do not grow at the same rate, observed to expected lung head ratio (O/E LHR) indicates the difference between LHR in the fetus with CDH to that expected in a normal baby. This is expressed as a percentage and is a predictor of neonatal morbidity and mortality and can be used to counsel women on prognosis (see table 1).

Degree of Lung Hypoplasia	O/E LHR	Liver Position	Survival
Extreme	<15%	-	<5%
Severe	15-25%	Up	10%
	15-25%	Down	25%
Moderate	26-35%	Up	30%
	26-35%	Down	60%
	36-45%	Up	60%
Mild	36-45%	Down	75%
	>45%	-	>95%

Table 1. Prognostic values for left sided CDH (Based on data from the Antenatal-CDH-Registry Group in Europe).

In addition to the published relationship between severity of lung hypoplasia

and survival, O/E LHR measurements are also predictive of early neonatal morbidity. There is a relationship between LHR and oxygen demand, days on conventional ventilation, start of enteral feeding and days in NICU. The severe lung hypoplasia group often require a patch at surgery, which is in itself a predictor of late morbidity.

Ultrasonography is the main imaging modality for antenatal evaluation of CDH, however fetal magnetic resonance imaging (MRI) may have a role in providing prognostic information to aid parental decision making. There is currently no evidence that fetal MRI is superior to ultrasound at predicting outcome, however, fetal MRI for lung volume may yield additional useful information, and it can be offered at 24 and 34 weeks gestation.

Multidisciplinary counseling by obstetrician, neonatologist and paediatric surgeon should ideally be planned to occur during two separate sessions in the second and third trimester, but not necessarily with all three specialists during one visit.

In selected continuing pregnancies with normal karyotype, the TOTAL (**T**racheal **O**ccclusion **T**o **A**ccelerate **L**ung growth) trial may be discussed. Fetal intervention by occluding the trachea has been shown to stimulate lung growth in experimental studies. The occlusion causes entrapment of lung fluid which leads to increased pressure within the lung. The stretch on the airway stimulates epithelial proliferation, but at the expense of differentiation. Periodic tracheal occlusion has been shown to address this by mimicking fetal breathing airway movement thereby increasing the number of alveolar epithelial type 2 cells, improving surfactant production and lung compliance in an experimental sheep model (Nelson *et al*). Early clinical trials of tracheal occlusion in America and Europe demonstrated varied success in neonatal survival at the expense of a high incidence of preterm prelabour rupture of membranes (PPROM) and preterm delivery.

The TOTAL trial is recruiting fetuses with normal karyotype and isolated left CDH that are considered “high-risk”. Two different high-risk groups are being recruited into separate studies; “severe lung hypoplasia” group (O/E LHR <25%) and “moderate lung hypoplasia” group (O/E LHR 26-35% or O/E LHR 36-45% with liver in thorax). Recruited mothers will be reassessed at a participating centre and randomized to either expectant management or fetal

endoscopic tracheal occlusion (FETO) via a balloon prior to birth. FETO is performed at 27-29+6 weeks gestation for the severe group and 30-32+6 weeks gestation for the moderate group. Elective balloon removal is performed around 34 weeks gestation with a delivery planned at 38 weeks gestation.

There needs to be careful evaluation before referring for this trial since the financial aspects are uncertain at present. Not only will the mother travel abroad for both balloon insertion and removal, she may encounter prolonged stay abroad due to the risk of PPRM and preterm delivery. Further information about the TOTAL trial can be found at: <http://www.totaltrial.eu>.

### **Key Points for Review until 32 Weeks Gestation**

- Detailed ultrasound examinations will be performed between 22–28 weeks gestation in order to assess O/E LHR, liquor volume, to look for other structural anomalies, and to perform fetal echocardiography.
- The method of calculating the O/E LHR should be documented i.e. longest axis or AP diameter method. The longest axis method may be preferred.
- Fetal MRI for lung volume may be offered at 24 and 34 weeks gestation to provide further prognostic information.
- Multidisciplinary counselling by obstetrician, neonatologist and paediatric surgeon should ideally be planned to occur during two separate sessions in the second and third trimester, but not necessarily with all three specialists during one visit.
- In selected continuing pregnancies with normal karyotype, the TOTAL trial may be discussed.

### **Review from 32 Weeks Gestation to Delivery**

At the 32–34 week follow-up scan with the fetal medicine service, growth and liquor volume will be assessed as well as any other anomalies. Further counselling by the multidisciplinary team may occur if the woman has not already seen all three specialists. Fetal MRI may be offered again at 34 weeks gestation as it may provide further objective information on prognosis and give an estimation of possible postnatal lung volume.

Glucocorticoids are known to stimulate surfactant production by type 2 pneumatocytes and induce maturation of the alveolar septum in premature neonates with respiratory distress syndrome when administered prenatally. There are beneficial effects of antenatal glucocorticoid exposure on various aspects of lung development in animal models of CDH. In addition, corticosteroids may exert protective effects through mechanisms not yet fully understood. At present, the role of prenatal steroid therapy in the human fetus with CDH remains unclear with small studies showing conflicting results. Since a single course of antenatal corticosteroids is unlikely to cause harm, its administration should be considered around 34 weeks gestation to aid lung maturation.

### **Key Points for Review from 32 Weeks Gestation to Delivery**

- At the 32–34 week follow-up scan with fetal medicine service, growth and liquor volume will be assessed as well as any other anomalies.
- Maternal steroid administration should be considered around 34 weeks gestation.

## **Delivery**

In order to ensure that experienced staff and appropriate equipment are present, delivery should be planned around 39 weeks gestation in a tertiary centre. This is achieved by either induction of labour or elective Caesarean section. The mode of delivery is dependent on obstetric indications and ensuring neonatologist presence, as there is no difference in neonatal outcomes between either mode of delivery provided that there is optimal neonatal support. Other practical factors that can influence delivery planning include; geographical location of the woman, risk factors for preterm labour such as polyhydramnios, and past obstetric history. The woman may be asked to stay near the tertiary centre if her usual residence is too remote or rural.

### **Key Points for Delivery**

- Delivery in a tertiary centre should be planned around 39 weeks gestation.
- If the diagnosis of CDH has been made prenatally, consultant neonatal staff should be in attendance at the delivery (see SDHCN Inpatient Management Guidelines for delivery room management of the newborn with CDH).

## Summary

- Following antenatal diagnosis of CDH, in addition to consideration of differential diagnoses and assessing for other fetal anomalies, the woman should be offered referral to the regional specialist fetal medicine service within five days
- An information leaflet is available from the SDHCN that can be used for counselling. If leaflets are not available locally, a copy can be downloaded from the SDHCN website ([www.sdhc.scot.nhs.uk](http://www.sdhc.scot.nhs.uk)).
- At the initial appointment with fetal medicine, the following will be assessed; side of defect, liver and stomach position.
- The woman will be counselled regarding CDH and will usually be offered karyotyping.
- Options for management of pregnancy will be discussed and will be influenced by parental preference, scan findings, and presence of associated anomalies
- Relevant clinical and sonographic details should be recorded on the SDHCN database by the local coordinator.
- Detailed ultrasound examinations will be performed between 22–28 weeks gestation in order to assess O/E LHR, liquor volume, to look for other structural anomalies, and to perform fetal echocardiography.
- The method of calculating the lung head ratio should be documented i.e. longest axis or AP diameter method (<http://www.totaltrial.eu>).
- Fetal MRI for lung volume may be offered at 24 and 34 weeks gestation to provide further prognostic information.
- Multidisciplinary counselling by obstetrician, neonatologist and paediatric surgeon should ideally be planned to occur during two separate sessions in the second and third trimester, but not necessarily with all three specialists during one visit.
- In selected continuing pregnancies, the TOTAL trial may be discussed.
- At the 32–34 week follow-up scan with fetal medicine service, growth and liquor volume will be assessed as well as any other anomalies.
- Maternal steroid administration should be considered around 34 weeks gestation.
- Delivery in a tertiary centre should be planned around 39 weeks gestation.
- If the diagnosis of CDH has been made prenatally, consultant neonatal staff should be in attendance at the delivery (see SDHCN Inpatient Management Guidelines for delivery room management of the newborn with CDH).





## Reference

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*This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.*