

## Diaphragmatic Hernia Information Sheet

Definition:	Herniation of abdominal contents into chest cavity	
Incidence	1:2000-5000 births	
Diagnosis:	Cystic mass in chest and absence of stomach bubble Peristalsis in chest Abnormal cardiac axis	
Location	Left sided	80-90%
	Right sided	10%
	Bilateral	<5%

Differential diagnosis: Cystic adenomatoid malformation (CAM)

Diagnosis may be made at any time in pregnancy, however can also be missed especially if stomach is not in chest. Is one of the structural causes of a thickened NT in first trimester.

Overall diagnosis currently carries a very poor prognosis with 60-80% of fetuses / babies dying. Single greatest reason for death is pulmonary hypoplasia.

At time of diagnosis needs careful ultrasound assessment for other anomalies especially within the CNS (30%) and cardiac 20%. Fetal heart often difficult to evaluate due to abnormal position therefore all require fetal echocardiography.

There is a significant risk of chromosome anomalies (16-37%) and therefore karyotyping must be offered to all. The risk in an isolated diaphragmatic hernia is probably relatively low, but still warrants at least an offer of a test.

Overall we currently suggest that the overall mortality is 80% at diagnosis which falls to 60% if we have excluded a chromosome anomaly, another structural anomaly or a cardiac defect. In the presence of one of these mortality approaches 100%

85% of cases have liver in chest which carries a worse prognosis.

Other poor prognostic signs for an isolated lesion are polyhydramnios and early diagnosis

As stated above cause of death is usually pulmonary hypoplasia. Greatest problem to date has been to identify those with normal lungs from those with hypoplastic ones. Current data suggests that a lung head ratio (LHR) may be beneficial in some cases.

LHR > 1.4 good prognosis

LHR <1.0 poor prognosis

(Note: this is only for cases where the liver is within the fetal chest)

### **Measurements of LHR**

- Area of contra-lateral lung divided by head circumference
- Obtain chest image at level of 4 chamber view. Measure two orthogonal cross sectional lung measurements and multiply to calculate lung area.
- Measure head circumference
- For example lung measurement 23 and 17 mm =  $391\text{mm}^2$
- Head circumference 250mm
- $\text{LHR} = 391/250 = 1.56$  which would potentially carry a good prognosis.

This data is not sufficiently robust to offer termination of pregnancy based on the results but may give an indication and aid in counselling.

### **Termination of pregnancy**

We would discuss TOP with all women with this diagnosis but would be happy to support any who wished to continue. Majority of couples with isolated lesion opt to continue.

### **Care pathway**

*20 week detailed anomaly scan, offer karyotyping and fetal echocardiography.*

If wish to continue for *review during pregnancy by paediatric surgeon.*  
(In Leeds all cases of lung anomalies including CDH are seen by Mr David Crabbe in his clinic on a Thursday morning)

*Plan follow up ultrasounds at 26, 30 and 34 weeks.* These can be carried out in the local hospital. The purpose of these assessments are to check fetal growth, identify significant hydramnios and provide on going support to the parents.

If significant hydramnios may need to consider amniodrainage. This would be performed if the mother was distressed by the degree of the hydramnios or there was felt to be a significant risk of a very preterm delivery. Amniodrainage is not without risk and can cause preterm labour

*Delivery should be in a centre with paediatric surgery.*

*Aim for a vaginal delivery* whenever possible. There is no benefit to either the mother or baby from a caesarean section.

A paediatrician will be available at delivery who will electively intubate the baby and pass a NG tube to keep the stomach empty.

If the baby is stable surgery is normally performed sometime after the first 48 hours. Those children who survive to surgery usually have a favourable outcome.