

Title:	Initial stabilisation and transfer of a baby with congenital diaphragmatic hernia		
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Description

Congenital diaphragmatic hernia is a developmental defect in the diaphragm and lung tissue that allows abdominal viscera to herniate into the chest cavity. It is a serious congenital abnormality associated with:

- Pulmonary hypoplasia
- Structural and functional lung immaturity
- Pulmonary hypertension
 - A reduction in pulmonary arteriolar cross sectional area
 - Muscular hyperplasia of remaining pulmonary arterioles

Majority (85%) are left sided. An association with other major anomalies, chromosomal and non-chromosomal, in up to 20% of cases. Foetal LHR (lung:head ratio) of <1, right sided herniation and the presence of liver in thoracic cavity are associated with poor prognosis. Survival rate for all diagnosed cases despite treatment is about 60%. Among the babies supported by ECMO, the mortality is 49%. Higher mortality if associated cardiovascular or other anomalies are present.

Resuscitation at birth

- Dependent on condition of baby at birth
- Minimise/Avoid bag and mask ventilation.
- Consider sedation and paralysis as soon as possible after birth to avoid any inhalation of air and distension of GI contents.
- Immediate intubation with large ET tube.
- Pass large bore NG tube (size 10F) on free drainage with regular aspiration. Replogle tube may be used (refer to relevant guidelines).
- Monitor pre/post-ductal saturations. This will indicate if significant ductal shunting is occurring.
- Maintain temperature during resuscitation and transfer. Hypothermia can lead to pulmonary vasoconstriction and worsening of pulmonary hypertension.

Stabilisation

- Urgent vascular access, umbilical venous and arterial lines.
- Sedation and muscle relaxation as continuous infusion.
- Nurse with the unaffected side slightly elevated to allow maximum chest expansion.
- Achieve acceptable gas exchange. HFOV and Inhaled Nitric Oxide (NO) may be required.
- Surfactant use is generally not indicated, but can be discussed on a case by case basis with the Consultant
- Aim for respiratory parameters:
 - Preductal saturations >90% & PEEP 5-6. Consider HFOV if needing high pressures.
 - pH >7.30 and maintain normal PCO₂.

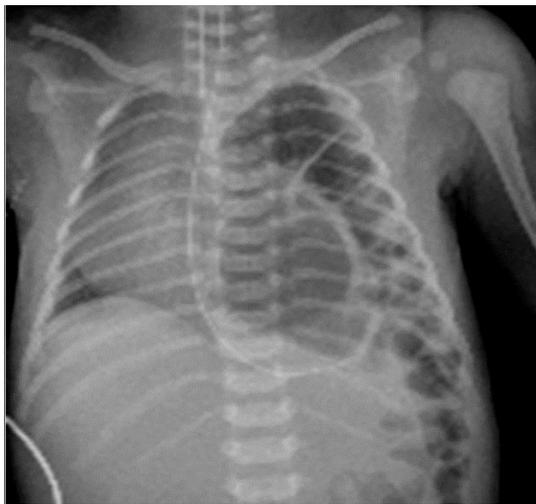
- Monitor invasive blood pressure and assess the circulation in order to determine the need for volume and inotropic support. Supranormal blood pressures may be required for the management of evolving PPHN. Monitor HR, perfusion, pH, lactate and urine output.
 - Consider iNO if postductal PO₂ (arterial) remains < 10 and / or clinical and echo signs of pulmonary hypertension
 - Monitor Oxygen Index

$$OI = \frac{F_iO_2 \times MAP(cmH_2O) \times 100\%}{P_aO_2(mmHg)}$$

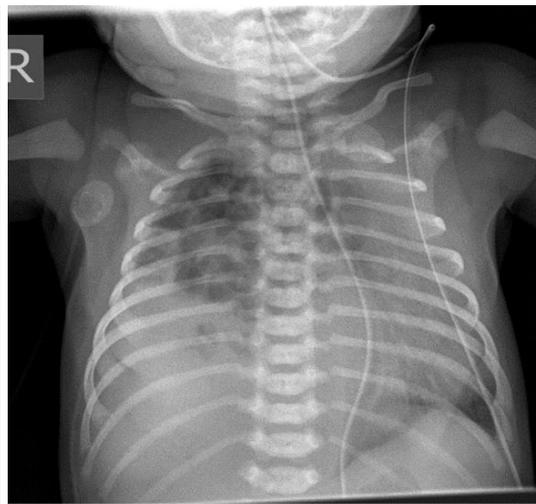
UK gas machines usually give PaO₂ in kPa so to convert PaO₂ (mmHg) = PaO₂ (kPa) x 7.5

- If OI >25 despite optimal cardio-respiratory treatment, consider discussing with ECMO centre.

• CXR and Abdominal XR



Left sided CDH



Right sided CDH

- If possible, arrange urgent echocardiography to assess pulmonary pressures and to rule out any structural anomalies. Ensure adequate preload and assess cardiac function.
- Dopamine and Adrenaline / Noradrenaline are the vasopressors of choice. If possible avoid counter-productive tachycardia.
- Correct any electrolyte disturbances. Maintain glucose and calcium homeostasis.
- Cranial USS to exclude IVH - implications for iNO and ECMO referral.
- Early joint medical and surgical management is essential, hence the need for urgent transfer to a surgical centre.
- Surgery is to be considered when FiO₂ is <0.5 and cardiovascular status is stable off all inotropes for at least 24 hours.