Congenital Chylothorax

**Antenatal management**

Refer to a tertiary maternity unit: Consider thoracentesis or pleuro-amniotic shunts to prevent pulmonary hypoplasia

**After Birth**

Provide respiratory support as necessary

Investigations: CXR shows pleural effusion. Consider ultrasound to estimate the size of the effusion

If there is significant respiratory compromise (RR>60, O2 Saturations<92%, FiO2 >40%, requiring CPAP or ventilation), insert a chest drain to drain pleural fluid

Biochemical analysis of fluid indicates chylothorax: Triglycerides > 1.1 mmol/L and total cell count of > 1000 cells/ml with > 80% lymphocytes. If infant is not feeding triglycerides and chylomicrons may be normal, total cell count & lymphocyte count may be used for diagnosis.

Check FBC with differential

**Ongoing Management**

Refer to a tertiary neonatal unit/childrens hospital

Investigations: Echocardiogram, chromosomal analysis, TORCH

Medium chain triglyceride formula (Monogen) or TPN for 4-6 weeks

If clinical deterioration or failure to resolve after 24 hours consider insertion of further chest drain

After 5 days conservative management Octreotide (1-10ug/kg/hr IV) may be considered (case reports only). When stopping wean over 48 hours Refractory cases (without resolution after 2 weeks conservative management) require cardiothoracic surgical assessment. Investigations include lymphangiography, lymphoscintigraphy & video assisted thoracoscopy. Surgical interventions include pleurodesis, thoracic duct ligation and pleuroperitoneal shunt.

Administer immunisations as normal
References:


This care pathway has been produced by the National Paediatric and Neonatology Clinical Programme. It is aimed at medical, nursing and allied health professionals working in Irish neonatal units.

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Disclaimer

This algorithm has been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach.

Information in this algorithm is current at time of publication.

Clinical material offered in this algorithm does not replace or remove clinical judgement or the professional care and duty necessary for each specific patient case.

Clinical care carried out in accordance with this algorithm should be provided within the context of locally available resources and expertise.

This algorithm does not address all elements of standard practice and assumes that individual clinicians are responsible to:

- Discuss care with consumers in an environment that is culturally appropriate and which enables respectful confidential discussion.
- Advise consumers of their choice and ensure informed consent is obtained
- Provide care within scope of practice, meet all legislative requirements and maintain standards of professional conduct
- Apply standard precautions and additional precautions as necessary, when delivering care
- Document all care in accordance with mandatory and local requirements