Paediatric Intensive Care Unit

Chylothorax post cardiac surgery

Staff relevant to:	PICU staff
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1. Introduction and Who Guideline applies to

Chylothorax is the accumulation of lymphatic fluid (chyle) within pleural space. The most common cause in paediatric population is post-operative chylothorax, followed by less frequent congenital chylothorax and lymphatic malformations. (1) The reported incidence following congenital heart surgery varies from 0.25% to 9.2%. (2, 3, 18)
Loss of chyle by the drainage can lead to nutritional depletion, fluid and electrolyte loss, hypolipemia, and lymphocytopenia of T cells. Chylothorax is associated with the risk of prolonged mechanical ventilation, increased length of ICU and hospital stay; increased adjusted risk for in-hospital mortality and higher cost regardless of procedure complexity. (2, 4, 5, 6)

The mechanisms of chylothorax post cardiac surgery:

- a direct injury of the thoracic duct or its tributaries, altering lymphatic drainage
- a process that elevates the central venous pressure (e.g. thrombosis of subclavian vein, right ventricular diastolic dysfunction, Fontan physiology)
- lymphatic malformations, congenital syndromes

Related guidelines: C31/2021 Thrombolysis UHL Paediatric Intensive Care Guideline

Detection of following:

- Drainage > 15ml/kg on the day after sternal closure or
- Milky drainage (milky fluid might not be present in fasting patients) or persistent pleural drainage

Observe the drainage for 5 days before dietary restrictions are implemented (fat –modified diet trial)
Exception is Fontan circulation – test & change diet earlier if required.

Chylothorax confirmed: Triglycerides >1.1mmol/l
WBC > 1000 cells/ml with > 80% of lymphocytes

Chylothorax work-up

- ECHO – optimize haemodynamics (Milrinone, ACEi, diuretics)
- Doppler Head / neck ultrasound – look for venous thrombosis; venogram; commence Heparin / consider tPA, thrombectomy
- Can IJ or subclavian access be removed/ replaced?

Risk factors: previous chylothorax, elevated venous pressure, decreased function, thrombosis, residual lesion

1) Start FAT-MODIFIED DIET TRIAL for 72 hours [21]
monitor drain losses

NOT ABLE TO COMMENCE ENTERAL FEEDING
or
HIGH VOLUME LOSSES >20ml/kg/d

Low volume losses <20ml/kg/d

FAT-MODIFIED DIET (low LCT diet)
*Record losses daily
*chest drain losses > 20ml/kg/d on any day or > 10ml/kg/d after 5 days go to high volume arm

*Remove chest drain when losses **<2ml/kg/d**
*Continue diet for **4 weeks** from the start date of fat-modified diet
*Frequent dietician reviews – consider trace elements supplementation, carnitine, essential fatty acids, fat soluble vitamins especially, if diet is required longer than 2 weeks - **unnecessary if on Monogen** (nutritionally complete formula)

2) TPN trial for 7 days
If chest drain losses < 10ml/kg/d on any day go to low volume (rule out drain blockage)

NBM > 7 days & chest drain losses > 10ml/kg/d – REFRACTORY CHYLOTHORAX

1. **Replace losses;** usually 50-75% of losses >20-50ml/kg; alternating crystaloid (Plasmalyte) and coloid (FFP, albumin, IVIG)
2. **Consider therapeutic Heparin/ LMWH;** once thrombosis has been ruled out continue prophylactic dose; monitor Antithrombin III and replace FFP accordingly
3. **After 1-2 weeks of high losses**:
   - check and supplement if required trace elements, folate, ferritin, B12, fat soluble vitamins
   - check Ly - if lymphopenic, check Lymphocyte subsets (need for cotrimoxazol prophylaxis); consider repeating in 1 month
   - immunoglobulin levels

Additional therapies:
Octreotide: start 0.5 microgram/kg/h;
Gradually increase to 1-4 microgram/kg/h (max 10)
• effect within 6 days
• if not, wean by 25% daily over 4 days
Monitor glucose 6 hourly, TFT's on starting the treatment, watch for signs of NEC; prolonged QT.
Prednisolon 1mg/kg BD 7 days (6)
Propranolol (20)
Surgical interventions – pleurodesis, thoracic duct ligation

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2. Guideline standards and procedures

2.1 Risk factors:

- Bidirectional Glen operation, Fontan physiology, TOF repair with RV dysfunction, heart transplantation
- procedures in the vicinity of the thoracic duct including Systemic to pulmonary arterial shunt insertion, Coarctation and aortic arch repair, PDA ligation
- procedure complexity (longer bypass time, longer cross-clamp time, higher RACHS-1*), younger age, genetic syndromes
- central venous lines (IJ, subclavian) [5, 6,7]

* The Risk Adjustment for Congenital Heart Surgery (RACHS-1) method was created to allow a refined understanding of differences in mortality among patients undergoing congenital heart surgery

2.2 Diagnosis - pleural fluid testing

- presence of chylomicrons
- Triglycerides >1.1mmol/l
- fluid cell count - White Blood Cell count > 1000 cells/ml with > 80% of lymphocytes

The most consistent diagnostic marker is the predominance of lymphocytes. Poor enteral nutrition at the time of diagnosis may play a role in decreasing chylomicron and triglyceride levels in the pleural fluid. [8]
Differential diagnosis - pseudochylothorax doesn't contain chylomicrons and contains higher cholesterol level than chylothorax (pleural fluid to serum cholesterol level ratio in pseudochylothorax >1.0). [9]
Current literature reports that the median time from the operating room to chylothorax diagnosis ranges between 4 and 14 days [20]

2.3 Treatment

1. Relief of respiratory symptoms by drainage and placement an indwelling drain
2. Non - surgical treatment (successful in 80% cases) [4,5,6]
3. Surgical treatment (necessary in < 5% of cases) [14]

Dietary restriction of long-chain triglycerides (LCTs) is expected to decrease chylous leakage. That can be achieved by using fat-free or low fat formulas enriched with medium chain triglycerides (MCTs). There is growing evidence that fat-modified breast milk is also effective in resolving chylothorax, however, poor weight gain of infants on low LCT diet is remaining an issue (around 60% of expected age-appropriate weight gain in average). [10]

In responders, the drainage should decrease to < 10ml/kg/d in the first week and to minimal or no leak by the second week. [8]
The length of the treatment is recommended for 4 weeks with a conversion to regular nutrition within 7 days. Some authors report a faster conversion from low LCT diet to regular nutrition without chylothorax re-accumulation. [2]

There is no difference in clinical outcome reported between MCT enriched diet and PN especially when CVP is not raised, however, no RCT are available. If the low LCT fat diet with MCT supplementation is used long term, the essential fatty acids, fat soluble vitamins and trace elements will need to be monitored and supplemented (EFAs - walnut oil). (11) Monogen is nutritionally complete thus additional supplementation of EFAs, fat soluble vitamins, trace elements is unnecessary.

A more aggressive option is complete enteral rest by using parenteral nutrition. The risks are related to the central venous access need (thrombosis, infection, vascular injury). The advantage is that intravenous lipid emulsions contain essential fatty acids. (11)

Octreotide is a synthetic, long-acting somatostatin analog used with varying effect on chylothorax resolution. The exact mechanism of action is not clear.
There has been considered a reduction in intestinal blood flow by vasoconstriction, with reduction of lymphatic fluid production. The optimal timing of introduction and duration of treatment is unknown. (12)
Octreotide administration is associated with adverse effects such as arrhythmias (QT prolongation), vomiting, constipation or diarrhoea, hyperglycaemia, hypoglycaemia. Other adverse effects include transient impairment of liver function, biliary sludge and fat malabsorption, transient hypothyroidism and necrotizing enterocolitis. (13)
The recommended initial dose of octreotide is 0.5 mcg/kg/h and increasing in a stepwise fashion to usual dose 1 - 4mcg/kg/h with maximum of 10 mcg/kg/h. Significant effect - at least 50% reduction of chylous drainage - should be apparent up to 6 days. It's usually recommended to discontinue Octreotide after 1 week by reducing the dosing by 25% per day over 4 days.

Thoracic duct ligation or pleurodesis - Several authors advocate for early thoracic duct ligation or pleurodesis in patients failing conservative medical treatment (the effusion persists for > 2 weeks or persistent high output drainage) which is based on presumption that successful surgery can shorten hospitalisation and reduce the risks of malnutrition and immunosuppression.
There is no consensus on the timing of surgery; PHIS analysis reports median of 18 days (IQR 7 - 28 days) (1, 3, 5)
Patients with heterotaxy syndrome frequently have bilateral thoracic ducts, so bilateral thoracic ductal ligation was recommended in that population. (3)
In children with refractory chylothorax, the placement of a pleuro-peritoneal shunt can be considered. (1)
A minimally invasive alternative which recently emerged is percutaneous Thoracic duct embolisation.
A recent study from CHOP advocates for MRI imaging of lymphatic system before any of surgical interventions, such as thoracic duct ligation or pleurodesis are performed. There is a selected group of patients with abnormal central lymphatic flow where surgical intervention can worsen the condition, and conservative management should be continued. (19)

Monitoring and supplementation - Protein level monitoring and supplementation together with electrolytes and clotting factors (Antitrombin III especially) may be necessary to prevent complications related to a large volume losses of chyle. There is an evidence based recommendation regarding prevention of thrombotic complications of CVL indicating a prophylactic subcutaneous dose of LMWH daily in patients at high risk for thrombosis (grade C).
Despite the fact that the degree of lymphopenia worsens with longer duration of chylothorax, the infections characteristic for cellular immunodeficiency were not observed in increased number and the effect of these immunologic alterations on development of infections is

unknown. [16] There is no evidence that the supplementation of intravenous immunoglobulin has an effect on the frequency of sepsis, bloodstream infection or survival to hospital discharge in patients with chylothorax. [15, 16] Although IVIG is not approved for postoperative secondary immunodeficiency, there is category IIIC evidence that it might be beneficial in the setting of postoperative sepsis. [17]

Indication for Immunoglobulin supplement in patient with Chylothorax:

- associated septicaemia with positive blood culture
- low Ig level
- massive chest drain losses of > 100ml/year of age.

The Immunoglobulin regime: 1g/kg IVIG daily x 2 days with Ig level 24 hours after administration IVIG to see improvement compared to the level before as the levels may drop in the presence of massive chylous losses.

3. Education and Training

None

4. Monitoring Compliance

None identified at present

5. Supporting References

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6. Key Words

Chyle, Chylothorax, Chylomicrons, Lymphatic fluid, Post cardiac surgery, venous thrombosis

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The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) Julia Vujcikova - Consultant Paediatrician Updated by Saad Malik – Registrar	**Executive Lead** **Chief Medical officer**
Details of Changes made during review: **Added related documents and updated references** **Updated management flow chart** **Added statement regarding the median time from operating room to chylothorax.**	