CHYLOTHORAX MANAGEMENT – CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- This document is an evidence-based guideline for diagnosis and management of chylothorax in children which is most commonly seen as a complication of cardiothoracic surgery
- Patients with single ventricle physiology are at higher risk of developing chylothorax, and this group of patients require individually tailored management strategies developed within the multi-disciplinary collaborative care provided by cardiology, cardiac surgery, intensive care and neonatology as appropriate
- Development of chylothorax has significant associated detrimental effects and complications impacting patient morbidity and mortality
- Accurate diagnosis and early treatment are vital for achieving favourable outcomes
- Chylothorax management varies widely internationally
- Nutrition therapy plays a major role in the treatment of chyle leaks
- Dietitian referral must be made for all patients with chylothorax– dietitian involvement is essential to ensure optimal nutrition delivery for appropriate growth, development and recovery
- This guideline aims to streamline management for patients with chylothorax at CHW based on current evidence and available resources - a management algorithm is included to be adapted

This document reflects what is currently regarded as safe practice. However, as in any clinical situation, there may be factors which cannot be covered by a single set of guidelines. This document does not replace the need for the application of clinical judgement to each individual presentation.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st March 2022	**Review Period:**
Team Leader:	Dietician and Fellow	**Area/Dept:** ESW and PICU

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This Guideline may be varied, withdrawn or replaced at any time.

CHANGE SUMMARY

- N/A - New document

READ ACKNOWLEDGEMENT

- All staff caring for children who require management of chylothorax are to read and acknowledge having read this guideline.

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1 Definition & Overview

- Chylothorax is the accumulation of chyle in the pleural spaces
- Chyle is a milky fluid that is transported via the lymphatic system and consists of lymphocytes, electrolytes, protein, triglycerides, cholesterol, fat soluble vitamins, enzymes and immunoglobulin
- Chyle is formed in the intestinal lacteals during digestion of fat - it plays a role in the absorption of fat-soluble vitamins and reabsorption of proteins lost through capillary leakage
- Chylothorax in children is most commonly seen as a complication of cardiothoracic surgery, either due to damage to the thoracic duct, increased pressure in the systemic venous circulation, or central vein thrombosis
- Patients at increased risk of chylothorax include those with:
    - prior chylothorax or repeat sternotomy
    - elevated venous pressure
    - right heart dysfunction
    - venous thrombus
    - cavopulmonary anastomosis
    - genetic syndromes (e.g. Noonan's syndrome, Turner's syndrome)
- Nontraumatic chylothorax is less common and typically due to infection, malignancy and congenital or idiopathic disorders of the lymphatic system
- Reported incidence of chylothorax post operatively is low (approximately 3-4%) but is associated with a significant increase in morbidity and mortality
- Associated effects (immunological, hematologic, respiratory) & complications:
    - impaired cell-mediated immunity from lymphocyte depletion
    - increased risk of infections and sepsis
    - increased risk of coagulopathy and thromboembolic events
    - restrictive lung disease contributing to respiratory insufficiency and respiratory support requirement
    - malnutrition
    - hypoalbuminemia
    - delayed recovery
    - tripled hospital length of stay
- Accurate diagnosis and early treatment are vital for achieving favourable outcomes
- Management varies widely internationally

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2 Single ventricle patients

- Chylothoraces are common in patients undergoing single ventricle palliation consequent to multiple issues:
  - elevation of central venous pressure following both bidirectional Glenn and completion of the total cavopulmonary connection (Fontan pathway)
  - venous thrombosis due to central venous access
- Maintain a high index of suspicion for chylothorax in the post-operative period following any stage of single ventricle palliation
- Due to the unique nature of the circulation and associated complex underlying anatomy and physiology in single ventricle patients, management of chylothoraces should be guided by multi-disciplinary collaborative care provided by cardiology, cardiac surgery, intensive care and neonatology as appropriate
- Additional investigations and management pathways may be necessary in these patients
- Management decisions may not necessarily follow the algorithms suggested in this document due to the need to tailor individual management in this subgroup of patients

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3 Investigations & diagnosis

- Suspect chylothorax if
  - Fluid from chest drain looks milky/cloudy
  - Drain output >5mL/kg/day after 4 postoperative days irrespective of appearance
- Send chest drain fluid to lab for analysis as below
- Diagnosis of chylothorax confirmed if ***one*** of the following criteria are met:
  - Chylomicrons *
  - Triglycerides >1.1mmol/L *
  - Absolute cell count >1000 cells/uL with lymphocyte fraction >80%

** May be falsely negative in the absence of enteral feeding with fat and so in this context consider cell count and lymphocyte fraction*

- Consider the following investigations to determine aetiology
  - ECHO and US Doppler of head and neck vessels
  - Central venous pressure measurement using in situ lines
  - Genetic investigation or follow up (e.g. Turners syndrome or Noonan Syndrome)
- If no response to conservative management after 10-14 days or output >20mL/kg/day for 2-6 days consider further lymphatic imaging for the following circumstances:
  - underlying aetiology remains unknown
  - anomalous thoracic duct anatomy is suspected
  - site of chyle leak is unknown and an intervention is planned to resolve lymphatic leakage
  - recurrence of chylothorax after thoracic duct ligation
- Consideration of cardiac catheterization to diagnose or address clot +/- measure pressures
- Choice of lymphatic imaging modality is individualised and influenced by the suspected underlying aetiology as follows:
  - Chest CT
  - **Lymphangiography** - contrast-enhanced study of the lymphatic system that can delineate thoracic duct anatomy and identify a potential site of chyle leak
  - **Magnetic resonance lymphangiography** - non-contrast high resolution imaging which provides detailed visualisation of the entire thoracic duct (can demonstrate the relationship of lymphatics to adjacent structures to assist preoperative planning)

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4 Biochemical monitoring

Recommended minimum laboratory monitoring if drain output > 10mL/kg/day or clinical compromise:

Daily	Blood gas Electrolytes Protein/Albumin
At diagnosis then Every 2-3 weeks	FBC LFTs Immunoglobulin Protein C and S AT3
Weekly	Thyroid Function Test Vitamin D if not supplemented
As per dietitian advice	Nutrition bloods
If on anticoagulant titration	Coagulation profile

5 Dietary management

Overview

- Nutrition therapy plays a major role in the conservative treatment of chyle leaks
- Diet modification reduces intestinal lymphatic flow and decreases chyle production, allowing the thoracic duct to heal
- Following diagnosis, patients will be commenced on a low long chain triglyceride (LCT) diet, with the addition of medium chain triglycerides (MCT) as an alternative fat and energy source
- LCT's are absorbed and transported via the lymphatic system as chylomicrons, whereas MCTs are absorbed directly into the portal system and do not stimulate an increase in lymphatic flow
- Feeding route is dependent on various factors including age, pre-morbid route of feeding and chest drain outputs
- These patients have increased nutritional requirements due to lost nutrients from chylous drain output and in cases of cardiac disease, the related increased metabolic demands and post-operative recovery
- These patients are at increased risk of poor growth and micronutrient deficiencies
- Refer ***all*** patients to dietitian – dietitian involvement is essential to ensure optimal nutrition delivery for appropriate growth, development and recovery

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Feeding methods

Enteral feeds

- In the absence of contraindications, commence enteral feeding using a low LCT formula (most commonly Monogen at CHW) - see Appendix for formula information
- Internationally some centres have reported the safe use of skimmed breast milk for enteral nutrition for infants with chylothorax however this is not currently practiced in Australia
- Note: it is important to ensure that the patient receives the adequate amount of essential fatty acids throughout treatment

Parenteral nutrition (PN)

- Indicated when drain output remains high (>20mL/kg/day) after a trial of 5-7 days on low LCT enteral feeds
- May be required before 5-7 days of low LCT enteral feeds if:
  - drain outputs are high (>20mL/kg/day),
  - serum albumin is <20g/L
  - high central venous pressure
  - poor nutrition status at baseline
  - enteral feeding is otherwise contraindicated
- Consider transition back to low LCT enteral feeds once drain outputs are <20mL/kg/day
- Associated with risk factors including infection, thrombosis and liver disease hence aim to transition to enteral feeds as soon as clinically appropriate
- Regular monitoring of biochemistry required as per Parenteral Nutrition Practice Guideline.
- Note: it is important to ensure that the patient receives the adequate amount of essential fatty acids throughout treatment

Oral diet

- Diet code = “Minimal LCT, MCT” (ensure any other dietary modifications, allergies or intolerances also included)
- Foods consumed are to be <2-3g of fat/100g serving depending on food group/type or alternatively that LCT fat limited to 1g per year of life
- Nutrition education on how to eliminate fat from the diet and obtain adequate protein from fat free foods or supplements will be provided by dietitian
- MCT supplementation may enhance energy intake – consider MCT oil and MCT supplements
- For introducing solids into infant diets, a minimal LCT formula will continue to form a major part of nutritional intake up until 1 year of age

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- A low-fat diet can be difficult to maintain and requires the patient and family to be extremely motivated and compliant
- Oral nutritional support:
  - < 5 years old encourage a low LCT supplement drink (e.g. Monogen) *
  - > 5 years old skimmed cow's milk should be used *
  - Low fat flavouring can be added if needed to improve palatability
  - MCT fat (e.g. MCT Procol or Liquigen) can be added to provide additional calories
  - Consider a nasogastric tube If there are concerns with growth or dietary intake or oral nutrition supplements are poorly tolerated

* Monogen and skimmed milk are not appropriate in presence of cow's milk protein allergy - please seek advice from dietitian

- Note: it is important to ensure that the patient receives the adequate amount of essential fatty acids throughout treatment

Nutrition monitoring

Anthropometry:

- Measure weight at baseline and a minimum of twice weekly - Monday and Thursday
- < 2 years – measure length and head circumference once weekly
- > 3 months where weight is an unreliable measure due to fluid status, consider measuring mid upper arm circumference (MUAC) once monthly

Nutritional biochemistry:

- Consider in patients with:
  - Poor growth
  - Prolonged high drain outputs (>10mL/kg/day for > 5-7 days)
  - On long term or oral dietary modification >4 weeks (in particular those not on Monogen supplementation)
- Can be ordered as subset on powerchart under 'Nutritional Biochemistry' (includes EUC/CMP, LFT's, Albumin, FBC, CRP, Iron studies, Folate, B12, Zinc, Selenium and Vitamins A, D, E & C)

Note: Measurement of Essential Fatty Acid's is not possible at CHW - if required, a sample can be sent to the Mater Hospital in Brisbane for analysis which will take 1-3 months to process and obtain results.

Supplementation

- Additional supplementation not usually required when on full enteral feeds as Monogen is a complete infant formula with added EFA's
- For those requiring long duration dietary LCT restriction >4 weeks on an oral diet (without Monogen) supplementation of EFA's (e.g. with walnut oil) and fat soluble vitamins may be required

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Diet duration & transitioning off

- Limited prospective evidence is available to support the optimal duration of LCT restriction required and the process of transition back to standard diet/feeds
- General recommendation is to continue for 2-6 weeks from chest drain removal if there is no recurrence of chylothorax
- CHW recommendation is transition back to standard feeds/diet 4 weeks after resolution of chylous drainage and drain removal (may vary depending on individual case and is to be discussed between treating teams and clearly documented)
- After resolution of chylothorax there is no further preventative effect to remaining on low LCT enteral feeds/diet – it is not nutritionally appropriate and may be harmful in the long-term
- CHW recommendation for transition back to standard feeds/diet is over 1-3 day. In some cases may require transition over a longer period of 1-7 days
- Monitor closely for signs of chylothorax recurrence, consider chest x-ray surveillance

6 Chest drains & volume replacement

- Consider replacement fluid if drain output is >20mL/kg/day or patient demonstrating clinical compromise:
  - Assess volume status and replacement requirement minimum 4th hourly (clinical need may necessitate greater frequency)
  - Use caution with high volume replacements and consider if volume replacement is potentially driving outputs
  - Replacement product choice = FFP
    4% albumin
  - Consider alternating replacement products depending on clinical need
- Special circumstances to consider:
  - **Concentrated 20% albumin** aiming to maintain serum albumin > 30g/L
  - **Intravenous Immunoglobulin** if serum level < 200 mg/dL (400 if acute infection /immunocompromised):
    - discuss with Immunology team
    - dose as advised by immunology team
    - risk of systemic vasodilation, transfusion reaction or anaphylactic/oid reactions
    - consider pre-treatment with paracetamol and antihistamine
  - **AT3 replacement:**
    - discuss with haematology
    - incorporate serum activity levels, anticoagulant doses and coagulation results
    - consider implications on anticoagulation dosing and risk of bleeding (FFP is alternative)

- Consider removal of chest drains once volume output < 3-5mL/kg/day on 2 consecutive days with Chest X-ray to confirm no collection with drain blockage

7 Additional medications

Consider if drain outputs remain > 20mL/kg/day following trial of Parenteral Nutrition for 5-7 days

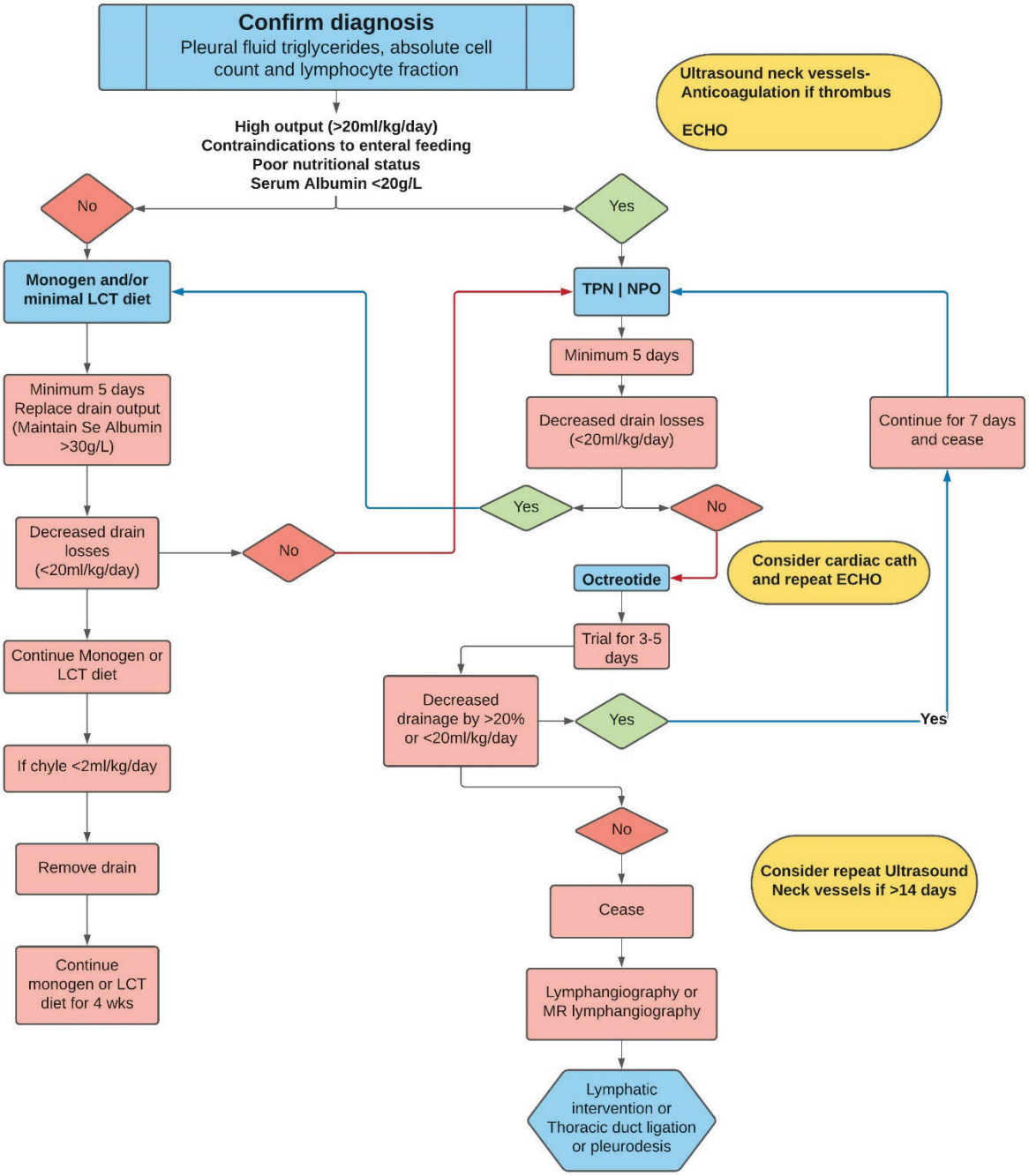
- **Octreotide (somatostatin analogue):** http://webapps.schn.health.nsw.gov.au/meds4kids/browse/O
  - Mechanism of action:
    - Mild vasoconstriction of splanchnic vessels, including hepatic venous flow, causing a decrease in gastric, pancreatic and intestinal secretions.
    - This leads to reduction in intestinal absorption and chyle flow and thereby healing of the lymphatic injury
    - Effect usually seen 5-6 days after commencement, clinician to titrate dose as required. Continued for 7-14 days depending on response
  - Dose:
    - continuous infusion 5microg/kg/hour, to a maximum of 10microg/kg/hour as required (short acting octreotide ONLY)
  - Presentation:
    - 50microg in 1mL, 100microg in 1mL, and 500microg ampoules in 1mL (short acting octreotide). Stored at 2-8 degrees (refrigerated)
  - Administration:
    - Dilute required dose in 50-200mL sodium chloride 0.9%
  - Monitoring:
    - hypertension, thrombocytopenia, hyperkalaemia, hyper/hypo-glycemia, gastrointestinal side effects, elevated liver transaminases, and thyroid function
  - Wean:
    - if response seen wean by 1microg/kg/hour every day, and if no response may withdraw by halving rate every 6-12 hours

Other medications may be considered under specific circumstances and depending upon presentation. Limited evidence available.

8 Surgical management considerations

- Consider when persistent and or high drain output not responsive to conservative management within 4 weeks
- Available interventions include thoracic duct ligation and pleurodesis
- Discuss with appropriate teams – Cardiothoracic surgeons, Paediatric surgeons and/or Interventional Radiology

9 Management algorithm



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10 Appendix – Low LCT High MCT formula information

Monogen

- Nutritionally complete powdered formula
- Contains whey protein, carbohydrate and fat
- High in medium chain triglycerides (MCT) at 84%
- Low in long chain triglycerides (LCT) at 16%
- Also contains vitamins, minerals, trace elements and EFA's
- Can be ordered from the formula room 8am-5pm
- After hours stock available
- Suitable for use from birth
- Concentrations:
  < 12 months - standard concentration 16.8% (75kcal/100mL)
  - can be gradually increased to max. 22.3%/1.33 conc (100kcal/100mL)
  >12 months - commence 22.3%/ 1.33 conc (100kcal/100mL)
  - can be increased as required to max 33.5%/2.0 conc (150kcal/100mL)

Modular formula

- For those with a cows' milk protein allergy
- Made up in the formula room
- Combination of Carb Plus Powder, Beneprotein, Calogen (LCT fat), Liquigen (MCT fat) and paediatric servatit (powdered vitamin, mineral and trace element mixture)

Skimmed breast milk

- Fat from expressed breast milk (EBM) can be removed using a refrigerated centrifuge
- Requires addition of calories using low LCT formula (e.g. Monogen) and modular supplements (e.g. liquigen and beneprotein) to ensure adequate calories provided
- Not currently in use at CHW

Lipistart

- Nutritionally complete powdered formula similar to Monogen
- Not currently available at CHW

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