# Chylothorax Clinical Guideline

**Inclusion Criteria**: Postoperative cardiac surgical patient. Diagnosis of chylothorax confirmed as per diagnostic criteria listed.
**Exclusion Criteria:** Patients with allergy to MCT formula.

This document serves to aid the clinical team in the prevention and management of chylothorax in infants and children with congenital heart disease. It should be used as a guide to identify high-risk patients for preoperative and intraoperative considerations and to manage chylothorax if diagnosed postoperatively. Clinical teams should use their own judgement and assessment whether the protocol needs to be adapted to meet the needs of the individual patient.

***This guideline was developed in conjunction with UCLA Mattel Children's Hospital as part of the joint CHOC-UCLA Congenital Heart Program.***

**Preoperative consultation** with the Lymphatic Team should be considered for patients with the following diagnoses:

- Noonan's syndrome
- Turner's syndrome
- Trisomy 21
- Prior history of chylothorax
- Generalized Lymphatic Anomaly (GLA)
- Kaposiform Lymphangiomatosis (KLA)

***Coordinate with Cardiology for Lymphatic Team Consultation**

**Intraoperative considerations** for patients at high risk for chylothorax (same diagnoses as above) include:

- Avoid thymus and lymph node dissection/excision if not necessary.
- Avoid dissection and snare control of vena cavae. If necessary, limit the dissection to within the pericardial reflection.
- Take a few minutes at the end to look for lymphatic leaks in likely locations and fix leaks, if needed.

**Page 2:**
Initial Evaluation for Chylothorax *and*
MCT/Minimal Fat Diet Algorithm

**Page 3:**
NPO/TPN Algorithm

**Page 4:**
Medication Algorithm

Approved Evidence Based Medicine Committee
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## Initial Evaluation for Chylothorax and MCT/Minimal Fat Diet Algorithm

- If output is serous or milky, send pleural fluid for triglyceride (TG) concentration, cell count, serum TG:
  - **<u>Chylothorax diagnosis is:</u>**
    - lymphocytes > 80% or TG > 110 mg/dL
- If output is **<u>severe</u>** (> 40 mL/kg/day) for 2 days, consider early lymphatic intervention.
- Coordinate with Cardiology for Lymphatic Team Consultation for all chylothorax diagnoses.
- Obtain ultrasound of upper extremities and neck veins to rule out thrombus.
- Daily chest x-ray (CXR) is *not* necessary.

**Enter Minimal Fat Diet Algorithm**

- Consult Dietitian
- Initiate age appropriate diet.
- If patient already on MCT or minimal fat diet, go directly to NPO/TPN Algorithm (page 3).

- Monitor output for 5 days and on day #6:
  - **Consider earlier transition on days 3 - 4 to NPO algorithm if output > 20 mL/kg/day or high-risk patient.*

Output ≥ 10 mL/kg/day

Output < 10 mL/kg/day

Output not trending down after 3 additional days

**Enter NPO/TPN**

If chest tube (CT) output is ***<u>trending</u>*** down, discontinue CT when < 3 mL/kg/q12h

Continue MCT/Minimal fat diet for 4 weeks
(6 weeks for high-risk patients)

**High-Risk Patients**

Those with the following diagnoses should be moved through this algorithm faster (earlier NPO + medication and earlier consideration for lymphatic intervention):

- Noonan/Turner's syndrome
- Trisomy 21

**<u>Newborn/Infant</u>**

- Consider spun down breastmilk.
- Enfaport at 20 - 24 kcals/oz, plus baby fruits, vegetables and grains - no meats, no avocados.
- If milk-protein allergy, consider full strength Vivonex pediatric formula (24 kcal/oz) or similar formula.

**<u>Toddler/School-Age Children</u>**

- 1 gm fat/kg/day, max 10 gm fat/day, plus 1 - 2 boxes of Ensure Clear supplement.
- If patient is on tube feeds may use Enfaport at 30 kcals/oz.

**<u>Adolescent</u>**

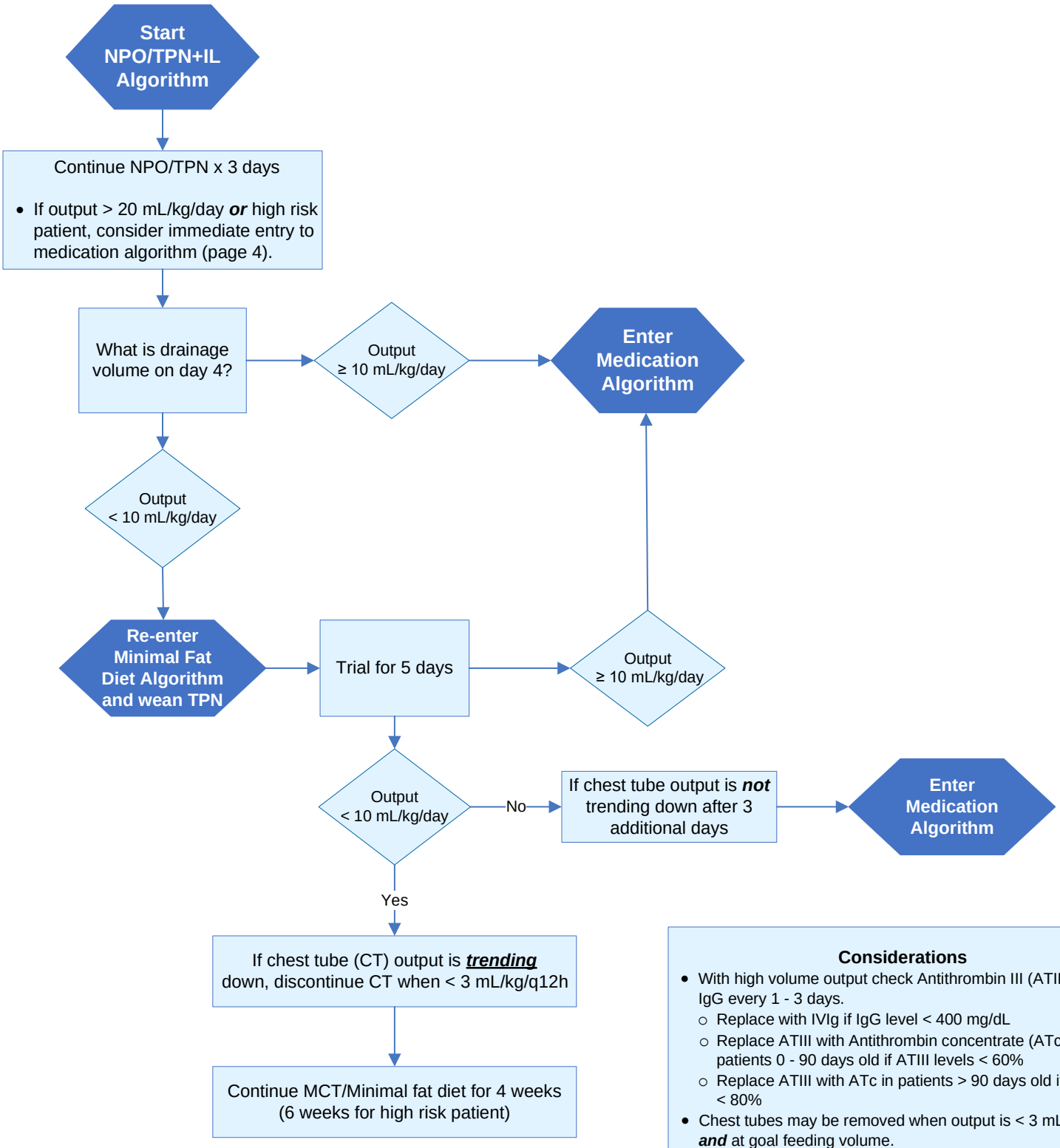
- Max 10 gm fat/day, plus 3 boxes Ensure Clear supplement.
- If patient is on tube feed's may use Enfaport at 30 kcals/oz.

**Considerations**

- With high volume output (≥ 10 mL/kg/day) check Antithrombin III (ATIII) and IgG every 1 - 3 days.
  - Replace with IVIg if IgG level < 400 mg/dL
  - Replace ATIII with Antithrombin concentrate (ATc) in patients 0 - 90 days old if ATIII levels < 60%
  - Replace ATIII with ATc in patients > 90 days old if ATIII < 80%
- Monitor albumin, fibrinogen, INR, thyroid studies closely.
- Chest tubes may be removed when output is < 3 mL/kg/q12h ***and*** at goal feeding volume.
- If chest tube output rapidly declines, obtain CXR to assess for kink or occlusion in chest tube.
- If inpatient on minimal fat diet, they may need weekly Intralipid starting with 1 gm fat/kg and advancing as tolerated to max of 3 gm fat/kg, as applicable (meaning it would be appropriate for infants but not adolescents).
- If outpatient, may need Essential Fatty Acids (EFA) supplementation to prevent EFA Deficiency (EFAD) complications (discuss with nutrition).

# Chylothorax Clinical Guideline

## NPO/TPN Algorithm




### Considerations

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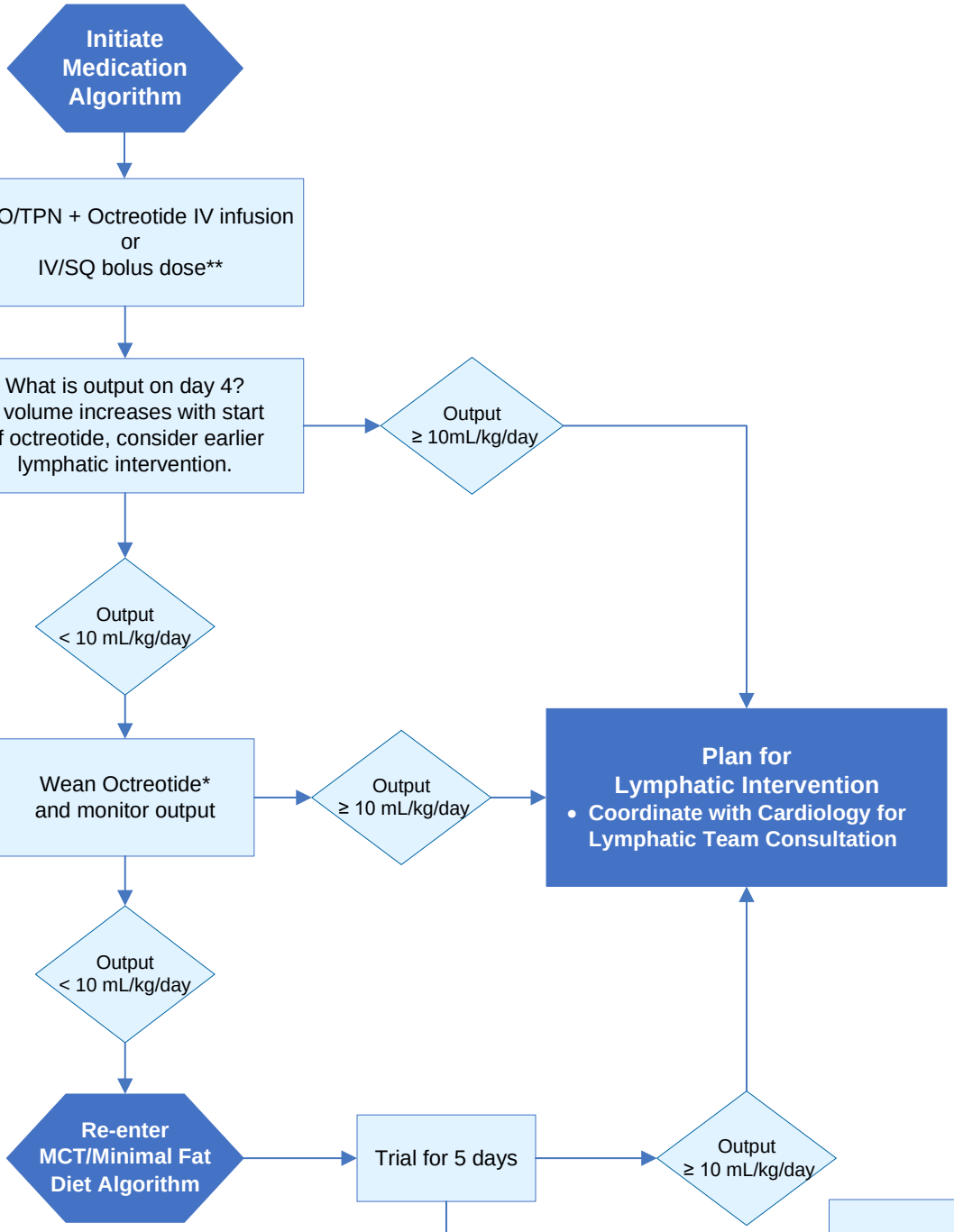
- Chest tubes may be removed when output is < 3 mL/kg/q12h ***and*** at goal feeding volume.
- If chest tube output rapidly declines, obtain CXR to assess for kink or occlusion in chest tube.
- Intralipids should be provided daily while on TPN.
  - There is no contraindication to intralipids with chylothorax.

# Chylothorax Clinical Guideline

## Medication Algorithm




- If output < 10 mL/kg/day and trending down:
  - Discontinue CT when < 3mL/kg/q12h.
  - Continue MCT/Minimal fat diet for 4 weeks (6 weeks for high risk patient).

***OR***

- If not trending down after 3 days:
  - Use clinical discretion whether to retrial Medication Algorithm or go directly to lymphatic intervention.

**Considerations**

If going forward with intervention, consider reinitiating feeds to fill lymphatic system.

**Octreotide**

** IV infusion dose:
- Start at 1 mcg/kg/hr to max of 5 mcg/kg/hr

** SQ dose:
- 5 mcg/kg/dose IV/IM q8h and increase by 5 mcg/kg/dose every 24 hours to max of 40 mcg/kg/day
- Count first day of treatment when at full dosing.

* Weaning should occur over 4 days.
- Decrease by 25% each day.

## *Chylothorax Clinical Guideline*
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