University Hospitals of Leicester NHS Trust

Nottingham University Hospitals NHS Trust

# Oncology Emergencies:
# Tumour Lysis Syndrome, Hyperleukocytosis and Anterior Mediastinal Mass

**This guideline is for use by healthcare staff, at CoMET undertaking critical care retrieval, transport and stabilisation of children, and young adults.**

**CoMET is a Paediatric Critical Care Transport service and is hosted by the University Hospitals of Leicester NHS trust working in partnership with the Nottingham University Hospitals NHS Trust.**

**The guidance supports decision making by individual healthcare professionals and to make decisions in the best interest of the individual patient.**

**This guideline represents the view of CoMET, and is produced to be used mainly by healthcare staff working for CoMET, although, professionals, working in similar field will find it useful for easy reference at the bedside.**

**We are grateful to the many existing paediatric critical care transport services, whose advice and current guidelines have been referred to for preparing this document. Thank You.**

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| **Approved By:** | **UHL Policy and Guideline Committee** |
| **Date of Latest Approval:** | **16 August 2024** |
| **Version:** | **(1)** |
| **Next Review Date:** | **August 2026** |

## Education and Training

**1.** Annual Transport team update training days

**2.** Workshops delivered in Regional Transport Study days/ Outreach

## Monitoring Compliance

| What will be measured to monitor compliance | How will compliance be monitored | Monitoring Lead | Frequency | Reporting arrangements |
|---|---|---|---|---|
| Incident reporting | Review related Datix | Abi Hill – Lead Transport Nurse<br>abi.hill@uhl-tr.nhs.uk | Monthly | CoMET Lead Governance Meeting |
| Documentation Compliance | Documentation Audit | Abi Hill – Lead Transport Nurse<br>abi.hill@uhl-tr.nhs.uk | 3 Monthly | CoMET Lead Governance Meeting |

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# Management of Tumour Lysis Syndrome (TLS)

- Form rapid cell lysis in haematological malignancies /lymphoproliferative (LPD) conditions.
- Life threatening electrolyte imbalance and renal failure.
- Can occur prior to chemotherapy- for instance a child presenting with stridor (from undiagnosed mediastinal mass) may be treated as croup and receive dexamethasone.

**Discuss urgently with the local haematology/oncology consultant on call and make COMET referral if likely for PICU admission**

## Airway

- Intubate if airway needs protection due to deteriorating neurology, or if respiratory distress warrants, **alert local anaesthetic team early.**
- Discuss with COMET consultant prior to intubation if there is a suspicion of a mediastinal mass
- Prepare for cardiorespiratory instability post intubation

## Breathing

- Use a cuffed ETT as pulmonary oedema will result in "stiff lungs" and may need relatively high peak inspiratory pressures
- If intubated for pulmonary oedema use a high PEEP ventilation strategy
- CXR to confirm ETT position
- Consider discussion with ECMO if remains hypoxic

## Circulation

- Cardiovascular compromise may occur due to **vascular compression** compromising cardiac output
- Ensure the child is well filled (No potassium in fluid)
- **Position the child sitting**. This will reduce the pressure of the mass off the great vessels in the event of arrhythmias
- Start treatment of hyperkalaemia without delay
- Risk of Hyperuricaemia, hyperphosphataemia, hyperkalaemia and hypocalcaemia – start treatment without delay (See COMET electrolyte emergency guideline)- include hyperlink to this

## Transport Considerations

- In the setting of a mediastinal mass causing TLS, **always transport in a sitting position**
- Prepare for CVS compromise due to electrolyte disturbance
- Transport with defibrillator pads attached

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# Management of Hyperleukocytosis

Defined as symptomatic (dyspnea, hypoxia or altered neurology) with: • WCC > 100x10 9
• In monocytic AML (FAB type M5), > 50x 10 9 (Large cells, can aggregate and cause coagulopathy more readily).

**Discuss with the local consultant, local haematology/oncology consultant on call and plan for prompt transfer to QMC or LRI (make COMET referral if needs PICU admission)**

**Medical emergency if symptomatic – Need rapid cyto-reduction with TLS prophylaxis**

## Airway and Breathing

- May need to be protected after cytotoxic therapy is started because of deterioration in respiratory status and neurology. **Alert local anaesthetic team early.**
- Use a cuffed ET tube as respiratory distress can occur from:
  - Leukostasis (causing sludging in lung microvasculature)
  - Fluid overload (from hyper hydration)
  - Pulmonary haemorrhage or Infection

## Circulation

- **Avoid red cell transfusion if possible** - may exacerbate leukostasis
- Only transfuse after discussion with haematology-oncology consultant
- If transfusing - administer no more than 5ml/kg over 4 hours
- If a second transfusion is required, re-discuss with haematology/oncology consultant
- Aim platelet count >30 or in the presence of active bleeding or coagulopathy aim for >50
- If **active bleeding**, FFP (15ml/kg) and cryoprecipitate (5ml/kg) should be given according to the clotting parameters – discuss with consultant haematologist
- If **not bleeding** with prolonged PT or APTT (>3 seconds above normal range) – give 10-15ml/kg FFP and maintain Fibrinogen > 1g/L with cryoprecipitate (5ml/kg)

**Recheck coagulation post transfusion**

## Disability

If clinical signs of CNS leukostasis are evident (Abnormal neurology, Raised intracranial pressure):

**Discuss urgently with local anaesthetic team**
**Discuss with COMET and haematology consultants**
**Need urgent neuroimaging and the images should be sent**

*Please note the above signs may indicate infarction or haemorrhage*

## If very high count (suspected AML):

- Consider urgent transfer to PICU for exchange transfusion or leukopheresis
- Prior to giving furosemide, discuss with haematology/oncology consultant as it may worsen hyper viscosity

## Transport Considerations

- May need high pressures to oxygenate
- Use high PEEP in setting of pulmonary haemorrhage/oedema
- If neurological concerns- neuroprotective measures

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# Management of Anterior Mediastinal Mass

**This is a haemato-oncological emergency and requires experienced senior supervision due to the risk of rapid deterioration, airway compromise and cardio-respiratory arrest.**

**Discuss with the local consultant, local haematology/oncology consultant on call and refer to CoMET for urgent transfer to specialist centre**

| Do | Don't |
|---|---|
| Manage and stabilise using an ABCDE approach as a priority over diagnostic investigations | Give hyper-hydration when signs of SVC obstruction evident as this can lead to cerebral oedema and death |
| Stabilise and transfer early with a low threshold for CoMET referral | Commence steroid treatment without discussion with a haematology consultant |
| Avoid sedation / anaesthesia for imaging and diagnostic procedures due to a high risk of airway compromise or respiratory arrest | Perform a CT chest in a child with respiratory or cardiac compromise |
| Continually reassess for signs of airway compromise, respiratory distress, SVC obstruction or neurological compromise | Struggle alone! |

## Airway

**A major risk is loss of airway – check for:**

- Stridor or wheeze
- Positional changes – may not like lying flat, or prefer one side or prone
- If the patient has airway compromise discuss management with COMET
- Only intubate for life threatening airway compromise
- **Ensure senior anaesthetic and ENT support is available prior to intubation**
- If intubation is required use a reinforced and cuffed tube where possible
- Intubate the patient in the position in which their breathing is most comfortable – often sitting up

## Breathing

- If unable to ventilate consider lateral or prone positioning
- Will require high peak pressures and need to be kept sitting post intubation.

## Circulation

**A major risk is loss of cardiac output from vascular compression – check for:**

- Facial or neck swelling or enlarged neck veins – Signs of SVC obstruction
- Raised HR, narrow "pulse pressure" on non-invasive BP – Signs of pleural/pericardial effusion/ tamponade
- Be prepared for significant cardiovascular compromise peri and post intubation
  - Have fluid boluses prepared as filling may help
  - Have peripheral adrenaline infusion running prior to induction

## Transport Considerations

- Ideally transport self-ventilating in the position of maximum comfort
- Have multiple fluid boluses prepared and keep the patient well filled
- Check electrolytes (especially K) immediately prior to and if possible, during transport as these children are at risk of TLS – Be prepared to treat hyperkalaemia en-route

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## References

1. Porcu P, Cripe LD, Ng EW, et al. Hyperleukocytic leukemias and leukostasis: a review of pathophysiology, clinical presentation and management. Leuk Lymphoma 2000; 39:1.
2. Green K, Behjati S, Cheng D. Arch Dis Child Educ Pract Ed 2019;104:298–303.