Clinical Guideline

INITIAL MANAGEMENT OF HIGH WHITE COUNT LEUKAEMIA (WCC>100)

SETTING  
Bristol Royal Hospital for Children- Haematology, Oncology and BMT Department and South West Paediatric Oncology Network Shared Care Hospitals

FOR STAFF  
Medical staff in hours and out of hours involved in the assessment of children with newly presenting leukaemia

PATIENTS  
Children (age<18) with a probable diagnosis of acute leukaemia and high initial white cell count (>100)

Guidance

The purpose of this document is to outline the initial management of high white count leukaemia patients; stabilisation, transfer from peripheral centre (POSCU, Paediatric Oncology Shared Care Unit) to Bristol (PTC, Primary Treatment Centre) and initial out of hours PTC management in the absence of a consultant paediatric haematologist.

It should be used in conjunction with the available febrile neutropenia, tumour lysis, SVC obstruction and other initial management guidelines as required.

Page 2 should be faxed/emailed to POSCU on referral of patient to aid their management.

These patients are at risk of: -

• Tracheo-bronchial and venous obstruction from mediastinal mass
• Tumour Lysis Syndrome (TLS)
• Leucostasis
• Coagulopathy

Note: the risk of leucostasis and coagulopathy is higher in AML, TLS and those with airway problems in ALL. Risk of leucostasis proportional to WCC ~ 100 for AML, 200 for CML, 500 for ALL.

Initial (Peripheral Hospital) Management of patient with WCC >100.

Important points in history/examination (all must be specifically documented)

• Evidence of respiratory distress (cough, wheeze, accessory muscles, flare, desaturation, orthopnoea)
• Evidence of bulky disease (LNss, HSM)
• Evidence of SVC obstruction (swollen face, red face, distended neck veins, collaterals)
• Evidence of leucostasis (headaches, hearing problems, visual disturbance, abnormal neurology)
Critical investigations (all must be done)

- FBC, X-match
- U+E, uric acid, calcium, phosphate, magnesium
- PT, APTT, Fibrinogen
- CxR (AP – add lateral if chest mass seen to identify any narrowing of airway, please arrange transfer of images to PTC). Consider cross-sectional imaging if timing allows (but avoid general anaesthetic/sedation).

Management prior to transfer

1. Obtain good venous access.
2. Hyperhydration. 3L/m². 0.45% sodium chloride/2.5% glucose, please note, potassium must NOT be added due to risk of TLS. This must be accompanied by strict fluid balance. In the context of chest mass or SVC obstruction this may need to be reduced to 2L/m². Discuss with PTC consultant. Use diuresis aggressively if required to maintain urine output.
3. Rasburicase – 0.2mg/kg stat IV. A single dose is often sufficient to prevent TLS. However, if in established tumour lysis, rasburicase should be continued at 0.2mg/kg OD dose (no max). (Jones et al, British Journal of Haematology, 2015, 169, 661–671)
4. Correction of coagulopathy – aim for platelets >10 (>50 if associated coagulopathy or leucostasis), fibrinogen >1, INR/APTR <1.5
5. Bloods every 4 hours to assess progress.
6. Do not transfuse red cells unless clinically symptomatic severe anaemia (increases risk of leucostasis, preferably discuss with PTC haematology consultant first).
7. See TLS protocol for more detail on management of TLS.

Inter-hospital Transfer

Patients in Peripheral Hospitals should be transferred at this point in their management and not before. It should take less than 12 hours from initial call to arrival of the patient in Bristol. If necessary, a medical escort or retrieval by the WATCh service may be required, and should be discussed between POSCU and PTC consultant at an early stage to avoid un-necessary delay.

During transfer

Continue IV fluids and fluid balance. A patient must NEVER be transferred without a prior CxR. If SVC obstruction, nurse upright, avoid lines in upper venous system if possible, reduce hydration rate if safe to do so (depends on TLS status).

If chest mass, consultation with PICU/WATCh, consultant paediatric anaesthetist, consultant haematologist and cardiothoracics is necessary prior to considering any anaesthesia. This should never be done in a peripheral hospital except in extremis. There is a significant risk of death on induction!

Subsequent PTC Management

1. Formal medical review of patient and discussion of the patient with the attending consultant essential within 30 minutes of arrival.
2. It is essential that the chest X-ray is reviewed by PTC staff.
3. Continue all initial management steps as outlined above.
Beyond this it is hard to be specific, but consideration may need to be given to:-

- Insertion of vascath to enable leucopheresis or dialysis.
- Institution of chemotherapy without formal diagnosis if hydration and rasburicase does not stabilize the patient (it usually does). If emergency treatment is required, morphological assessment by the most experienced morphologist available is essential. Save an EDTA sample, stored at 4°C for immunophenotyping.

For Acute Lymphoblastic Leukaemia, emergency initial therapy should be with oral steroids alone using dexamethasone 3mg/m²/dose BD or prednisolone 20mg/m²/dose BD. If significant risk of TLS (e.g. pre-existent renal impairment) 50% dose reduction may be appropriate.

For AML start stabilising treatment initially with cytarabine 100mg/m² or Hydroxycarbamide 15-30mg/kg/day

In the rare instance of CML and symptomatic leucostasis then leucopheresis should be the 1st line therapy, together with hydroxycarbamide, 15-30mg/kg/day. Allopurinol may suffice instead of rasburicase as TLS is rare.

Apheresis service can be contacted on 0117 342 1092 / 0117 988 2047 / 0117 912 5724 (OOH on call medic).

*Protocols can generally be found online or paper copies can be found in the doctor's office on Ocean Unit, Level 6 BRHC.

**REFERENCES**

Not applicable

**RELATED DOCUMENTS AND PAGES**

Not applicable

**AUTHORISING BODY**

Paediatric Oncology, Haematology and Bone Marrow Transplant Quality Assurance Forum (Quaf)

**SAFETY**

Not applicable

**QUERIES AND CONTACT**

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