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# Management of Children with Transfusion Dependent Thalassaemia and Non-febrile Acute Illness

| Document Information | | | |
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| **Stakeholders/ committees involved in guideline development :** | | | |

| For Child Health Clinical Guidelines Groups' use only | |
|---|---|
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**Document History**

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**Consultation distribution (before ratification)**

| Sent to | Version | Date | Actions taken as a result |
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| Professor David Rees | 2 | June 2014 | No amendments |
| Dr Sue Height | 2 | June 2014 | Minor amendments agreed |
| Sandra O'Driscoll | 2 | June 2014 | |

**Reviews and updates (including CGG comments)**

| Date | New version no. | Summary of Changes | Author of change/s |
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**Dissemination schedule (after ratification)**

| Target audience(s) | Method | Person responsible |
|---|---|---|
| South Thames paediatricians | Via network meetings and email | Professor Rees |
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**Management of Children with Transfusion Dependent Thalassaemia and Non-febrile Acute Illness**

Background

This guideline covers the management of children with transfusion dependent thalassaemia and non-febrile acute illness including liver, cardiac and endocrine failure. It is aimed at the paediatric, haematology and nursing members of the multi-disciplinary team. It should be noted that there is also a guideline on the management of fever and infection in children with transfusion dependent thalassaemia

Contents of Guideline:

**Management of Children with Transfusion Dependent Thalassaemia and Non-febrile Acute Illness**

This guideline covers the management of children with transfusion dependent thalassaemia and non-febrile acute illness including liver, cardiac and endocrine failure. It is aimed at the paediatric, haematology and nursing members of the multi-disciplinary team. It should be noted that there is also a guideline on the management of fever and infection in children with transfusion dependent thalassaemia

**General considerations**

Children with thalassaemia presenting with acute illness should be assessed urgently and the paediatric haematology team informed at an early stage of their admission. History and examination should be focused on complications of thalassaemia, but not ignore the possibility that the child could have unrelated problems, including surgical complications such as appendicitis.

**History**

History should establish the approximate number of transfusions received, the use of iron chelation, and previous history of cardiac, liver or endocrine problems.

Specific enquiry should be made about:

- Palpitations
- Fainting and collapse
- Dyspnoea, including orthopnoea and paroxysmal nocturnal dyspnoea
- Chest pain
- Weight gain
- Increasing jaundice
- Abdominal pain
- Polyuria and polydipsia
- Muscle pains and spasms
- Menarche (if appropriate)
- Previous cardiac T2* MRI scans

**Examination**

- Full examination should be performed on every patient with particular attention to
    - Cardiac rate and rhythm
    - Evidence of left and/or right biventricular failure
    - Presence of hepatomegaly and signs of liver disease
    - Signs of hypothyroidism, hypocalcaemia

**Investigation**

- Investigations will be determined by the findings on history and examination.

- All acutely unwell patients with transfusion-dependent thalassaemia should have
    - Full blood count with reticulocytes
    - Renal function tests
    - Liver function tests including ALT
    - INR, APTR, Clauss fibrinogen
    - Calcium and phosphate
    - Serum ferritin
    - Blood glucose
    - Thyroid function tests
    - Blood and urine cultures
    - Urine dip-stick for ketones
    - Chest X-ray
    - ECG
    - Pulse oximetry
- If there is evidence of cardiac problems:
    - Troponin levels
    - Organise echocardiography – discuss initially with HDU/PICU consultant. Older patients may have the echo performed at KCH, whilst younger patients will need to be assessed by the paediatric cardiologists at the Evelina Children's Hospital.
- If there is evidence of acute liver failure
    - Split bilirubin
    - LDH
    - Serum amylase
    - Ultrasound of the liver, bile ducts, gall bladder, pancreas and spleen
    - If there is acute hepatitis (ALT>110 IU/ml) request 'New Patient - Core Investigations >3 years' on EPR which includes ammonia, lactate, alpha-feto protein, caruloplasmin, zinc, copper, creatinine kinase, lipid profile, hepatitis A-E serology,EBV serology, autoantibodies, immunoglobulins.
- If there is hyperglycaemia, glycosuria or ketonuria
    - Capillary or venous blood gases

**Management**
Management will depend on the initial diagnosis, and will be in conjunction with the general and specialist paediatric teams.

*Management of Acute Cardiac Problems*

- The patient should be admitted to HDU.
- Continuous ECG monitoring should be started.
- Acute treatment for cardiac dysrhythmias or ventricular failure may be necessary, and should be started following discussion with the HDU/PICU consultant.
- The patient should be discussed with the on-call paediatric cardiologists at the Evelina Children's Hospital, and may need to be transferred.
- When the patient is stabilised, continuous treatment with intravenous desferrioxamine at a dose of 40mg/kg/day should be started. If the child is on deferasirox (Exjade), this should be stopped for 24 hours before starting the desferrioxamine infusion.
  (please refer to the iron chelation guideline)

*Management of Acute Liver Failure*

- The patient should be managed in conjunction with the paediatric Hepatology team, usually on Rays of Sunshine ward.
- Specific management will depend on the diagnosis and will be determined by the paediatric hepatologists.
- Iron chelation should be stopped until the diagnosis is established. Desferrioxamine should be used in preference to deferasirox in the presence of liver failure.
  (please refer to the iron chelation guideline)

*Management of Diabetic Ketoacidosis*

- Management should take place on HDU with cardiac monitoring.
- The child should be managed according to the 'Guidelines for the Management of Children with Diabetic Ketoacidosis' available on Cliniweb, in conjunction with the paediatric endocrinologists.
- Iron chelation should be stopped until the patient is stable.

Moira Dick
June 2014