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

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### Change History

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**STSTN - Management of Children with Transfusion Dependent Thalassaemia and Non-febrile Acute Illness 20 July 2020**

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Introduction

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.

Indications
Children with transfusion dependent thalassaemia and non-febrile acute illness including liver, cardiac and endocrine failure.

Contraindications
If patients are febrile please also consult the separate guideline on management of infection in children with transfusion-dependent thalassaemia.

Main Guideline

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* and Liver Ferriscan MRIs for iron loading quantitation

Examination
Full examination should be performed on every patient with particular attention to
- Cardiac rate and rhythm
- Evidence of left and/or right ventricular failure

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• 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 & reticulocytes
  • Renal function tests
  • Liver function tests including ALT and LDH
  • PT, APTT, Clauss fibrinogen
  • CRP
  • Blood Group and Antibody screen
  • Calcium, phosphate & magnesium
  • Venous/capillary gas & lactate
  • Serum ferritin and transferrin % saturation
  • 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–locally if available. Discuss the patient with the paediatric cardiologists at Evelina Children’s Hospital. Transfer to HDU/PICU may be required and STRS should be involved.

If there is evidence of acute liver failure – discuss with Paediatric Hepatology team (see below)
  • Split bilirubin
  • LDH
  • Serum amylase and lipase
  • Ultrasound of the liver, bile ducts, gall bladder, pancreas and spleen
  • If there is acute hepatitis (ALT>110 IU/ml) request additional investigations - ammonia, lactate, alpha-feto protein, caeruloplasmin, zinc, copper, creatinine kinase, lipid profile, hepatitis A-E serology, EBV serology, autoantibodies, immunoglobulins.
  • Serology for Parvovirus B19 IgG/IgM and DNA (‘send away’)
  • Adenovirus/CMV/EBV DNA

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If there is hyperglycaemia, glycosuria or ketonuria (discuss with Paediatric Diabetic team locally; otherwise contact the Endocrinology & Diabetic team at either KINGS OR EVELINA – see below)

- Capillary or venous blood gases
- Blood glucose monitoring

If there is diarrhoea, stool culture, including culture for Yersinia enterocolitica (speak to microbiology about need for specific culture).

Management
Management will depend on the initial diagnosis, and will be in conjunction with the general and specialist paediatric teams – contact the either KCH or ECH.

Management of Acute Cardiac Problems

- Discuss with on call paediatric cardiologist at Evelina Children’s Hospital. Paediatric Cardiology Registrar is 020 7188 7188 bleep 1344
- If transfer is needed, STRS should be involved. The patient should be admitted to HDU.
- Continuous ECG monitoring should be started.
- Acute treatment for cardiac dysrhythmias for 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, usually at a higher dose than recommended in the BNFc is used for acute cardiac toxicity (discuss with specialist centre). Deferiprone may also be added - please refer to the iron chelation guideline. In the BNFc there is a reference to concern about growth with use of Desferrioxamine at higher doses – this is not applicable in the acute treatment of severe iron overload with life-threatening organ impairment.
- Referrals for assessment of Pulmonary Hypertension in the specialist clinic at the Evelina should be emailed to Sadia.Quyam@gstt.nhs.uk (this is not an acute service)

Management of Acute Liver Failure

- The patient should be managed in conjunction with the King’s paediatric Hepatology team, Contact Paediatric Hepatology SpR 09.00-17.00 Switchboard 020 3299 9000 bleep 491 or Ext 37812, 17.00-09.00 or weekends/Bank holidays 07866-792368
- Specific management will depend on the cause of the liver failure 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 (Exjade FCT) in the presence of liver failure (please refer to the iron chelation guideline)

Management of Diabetic Ketoacidosis

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- Management should be with the local diabetic team or endocrinology/diabetic team at KCH, ECH involvement contact details:
  - Kings College Hospital: kch-tr.KingsPaediatricEndocrinology@nhs.net, MDT coordinator 02032999000 ext 31992
  - Queen Elizabeth Hospital: kch-tr.KingsPaediatricEndocrinology@nhs.net, MDT coordinator 02032999000 ext 31992
  - Croydon University Hospital: Tony.Hulse@gstt.nhs.uk
  - Guys & St Thomas Hospital: PaediatricEndocrineTeam@gstt.nhs.uk
- The child should be managed according to local protocols for the Management of Children and Young People presenting with Diabetic Ketoacidosis (DKA) in conjunction with the paediatric endocrinology and diabetes team.
- In severe DKA with concerns about conscious level or if <3 years old, admission to HDU with cardiac monitoring would be required.
- Iron chelation should be stopped until the patient is stable.

**Related Guidelines**

**References**
Standards of Clinical Care of Children and Adults with Thalassaemia in the UK 3rd Edition (2016)
Contact details:

KCH Paed sickle CNS: 02032994752
KCH Paed sickle consultants: david.rees2@nhs.net subarna.chakravorty@nhs.net sue.height@nhs.net
GSTT Paed sickle CNS: 0207 188 9432
GST Paed sickle consultants: Baba.Inusa@gstt.nhs.uk maria.pelidis@gstt.nhs.uk
QE Paed sickle CNS: 07741233556
QE Paed sickle consultants: julie.lord@nhs.net aruj.qayum@nhs.net
UHL Paed sickle CNS: 07741233556
UHL/QE Paeds sickle consultants: s.wilkinson6@nhs.net julie.lord@nhs.net
CUH Paeds sickle CNS: 0208 2517229
CUH Paeds sickle consultants: nazmachowdhury@nhs.net

Additional contacts can be found on the STSTN website (www.ststn.co.uk)

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