Guy's and St Thomas' NHS
NHS Foundation Trust

Clinical Guidance

Guidelines for Management of Acute Presentation in Adults with Thalassaemia

Summary
Guidance for management of the rare acute complications that patients with Thalassaemia may present with. Including sepsis, liver decompensation and cardiac decompensation. These comprehensive guidelines are intended for use as a reference for medical, nursing staff and all health care professionals.

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Background

All patients with a haemoglobinopathy diagnosis on a regular transfusion program will be under regular review in the haematology outpatient department and well known to the haemoglobinopathy team. Should any such patient be admitted acutely unwell out of hours please inform the Haematology SpR on call.

Fever & Sepsis

Infection is encountered in thalassaemic and other transfusion dependent patients for a variety of reasons including the fact that these patients may have significant organ iron deposition, a long-term indwelling vascular access device such as a port-a-cath or a peripherally inserted central catheter (PICC) line, they may have other conditions such as diabetes which may affect their ability to mount an adequate immune response.

Common causes of fever and sepsis a thalassaemic patient include:

- Acute sepsis cause unknown
- Gram negative sepsis (*Klebsiella, E.Coli, Salmonella sp*)
- Lower respiratory tract infection
- Urinary tract infection
- Osteomyelitis
- Biliary sepsis
- Viral infections
- Central venous catheter related infection (Port-a-cath etc)

NB*: Yersinia enterocolitica* is a very rare cause of infection in these patients, particularly those who are severely iron loaded or on chelation with Desferrioxamine.

Management

All patients with pyrexia should undergo clinical assessment for infection. Assess for signs of circulatory collapse and sepsis. [NB: Fever may not always be present, just a high CRP (C-reactive protein) and white blood cell count]. It is important to consider neutropaenic sepsis and send a full blood count (FBC) off urgently as these patients may be on Hydroxycarbamide or Deferiprone both of which may cause neutropaenia.

Investigations:

- FBC
- CRP
- Urine Dipstix
- Other investigations as dictated by clinical features
- MRSA swabs as per trust policy including invasive live sites

If temp > 38°C

- Blood/Urine/Stool culture
- Chest X-ray
- Other investigations e.g. abdominal ultrasound as dictated by clinical features.

Antibiotics

A non neutropaenic patient with signs suggesting infection with a clear source must be treated as per Trust antibiotic guidance

If there are signs suggesting infection but no clear source, clinical assessment of severity should determine whether antibiotics should be started. If empirical antibiotics are thought appropriate, start Co-amoxiclav, adding a single dose of Gentamicin 5mg/kg (see Trust Guidelines for dosing in overweight patients) if there is evidence of sepsis or a recent hospital admission.

If neutropaenic treat as per Trust guidance for neutropaenic sepsis and stop any drugs which may be causative for neutropaenia such as Hydroxycarbamide or Deferiprone.

CARDIC COMPLICATIONS

We have very few patients with significant cardiac iron loading. All such known patients are very closely monitored and appropriately commenced on aggressive chelation regimes. A rare patient may due to either poor compliance or an intercurrent infection present with acute cardiac decompensation.

Any such patient must be referred urgently to the cardiologist on call and also brought to the attention of the Haematology SpR on call who in turn must inform the haematology consultant on call.

Presentation symptoms are usually as for heart failure with:

- ankle oedema
- abdominal distension
- shortness of breath and
- palpitations.

Investigations at time of admission should include:

- ECG,
- CXR,
- Echo within 24 hours (on weekdays)

Management must be under the direction of a cardiologist in a Coronary Care Unit/ High Dependency unit/ Intensive Care Unit with support from haematology.

- **All patients should be on a continuous intravenous Desferrioxamine dosed at 50mg/kg/day, 24 hours a day 7 days a week as this provides a cardioprotective effect.**

If a cardiac T2* MRI assessment has not been done in the last 6 months then arrange urgently to assess the severity of the cardiac iron loading and Ejection Fraction.

Once the patient is stable, chelation therapy and its mode of administration will be reviewed to optimise compliance.

Endocrine complications:

Endocrine complications including short stature, delayed puberty, hypogonadotropic hypogonadism, hypothyroidism, hypoparathyroidism, and diabetes mellitus are common in thalassaemia major, and are primarily due to iron overload of endocrine glands.

As all our thalassaemia patients are regularly followed up in the OPD, and screened for all expected complications, therapy is usually commenced promptly for all encountered problems including the endocrine abnormalities which can occur even in our optimally treated patients. It is rare for such patients to present as an emergency with an acute endocrinopathy, however should this occur please treat as per trust guidance and inform the haematology SpR on call of the admission.

The most likely presentations will be Diabetic patients with hypo or hyperglycaemic emergencies, and a rare patient may present with acute onset hypocalcaemia due to hypoparathyroidism. These patients should all be managed under direction of the Endocrine team with haematology support as required.

Liver complications:

Liver disease is a common complication in older thalassemia patients, causes of liver disease include transfusion-related viral hepatitis (Hepatitis B, C), iron overload, drug toxicity, and biliary disease due to gallstones.

Due to close monitoring these patients will rarely present with acute liver decompensation, unless they are known to have cirrhosis in which case they will be under the care of a hepatologist. They are also at risk from potentially life threatening variceal bleeds.

Should a patient present with symptoms of decompensated liver disease or a variceal bleed they should be managed by the gastroenterologist with support while an inpatient from the haematology team.

References:

Cheema AN et al. Early detection of cardiac dysfunction by BNP in beta-thalassaemia major patients. Acta Cardiol. 2012 Jun;67(3):331-5.

Thalassaemia major and other rare anaemia acute presentation; clinical guideline for The Whittington hospital NHS Trust

http://www.thalassemia.ca/wp-content/uploads/Thalassemia-Guidelines_LR.pdf