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GSTFT Clinical Practice Guideline

Management of Children with Sickle Cell Disease on a Chronic Transfusion Programme

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Overview:

Some specific complications of sickle cell disease have been shown to benefit from maintenance chronic blood transfusions. The goal is to suppress **erythropoiesis** sufficiently and to provide enough normal red blood cells to maintain the percentage of the patient's cells (i.e. haemoglobin S) at less than 30%. Experience has shown that this approach significantly reduces the risk of recurrent stroke. Such transfusions also reduce the incidence of other sickle-related complications such as vaso-occlusive pain and acute chest syndrome. It may also prevent the progression of chronic organ damage and even reverse some pre-existing organ dysfunction. This has been shown most clearly in patients with Hb SS and functional asplenia, some of who show improved splenic reticuloendothelial function after receiving chronic transfusions. Many children with sickle cell disease treated with chronic transfusions also experience an increased sense of well being, with improved energy levels, exercise tolerance, growth velocity and sexual development. Thus, transfusions to chronically replace sickle cells with normal erythrocytes can be considered a specific therapy that markedly ameliorates the disease.

Indications:

- Stroke
- Abnormal Transcranial Doppler scan
- Transient Ischaemic Attacks with abnormal cerebro-vascular neuro-imaging
- Severe Acute Chest Syndrome
- Persistent Priapism

Indications in Selected Patients:

- Severe debilitating pain
- Following splenic sequestration (as alternative to observation or early surgical splenectomy)
- Recurrent priapism
- Chronic organ failure
- Intractable leg ulcers
- Severe chronic anaemia with high output cardiac failure

Blood Transfusion:

- PRBC 10-15 ml/kg (minor-antigen-matched, sickle-negative, leukocyte-depleted) given over 3-4 hr with standard monitoring. Minor -antigen matching for Rh (C, D, E,) and Kell should be provided for all patients.
- More extensive matching for patients with previous allo-immunisation. Frequency of transfusions (usually 3-4 weeks) is adjusted to maintain Hb S below 30% (typically with nadir Hb >9-10 gm/dl).
- For patients receiving chronic transfusions for stroke who have had no recurrent Neurologic events for 3 years, consider decreasing frequency of transfusions to maintain Hb S below 50%.

Record:

- Volume of RBC transfused.
- Serial erythrocytapheresis is an alternative and it is associated with substantially less iron loading.
- Patients should be immunised to hepatitis A and B.
- Continue prophylactic penicillin.

Iron Chelation:

Initiation of Chelation with after ³ 1 year of chronic transfusions and / or when:

- Serum ferritin is increased to ³ 1000 mg/L.

- Hepatic iron content >4 mg /gm dry wt liver tissue (as determined by liver biopsy) also has been used as an indication for beginning iron Chelation.
- Initial dose desferrioxamine 25 mg/ kg/ d s.c. in at least 8-10 ml sterile water infused over 10-12 hr, 5-6 nights per week.
- Exjade (oral iron chelation therapy) may be started at 20mg/kg and may increased by 5mg/kg every three months to maximum of 30mg/kg before review for a possible higher dose protocol, second line indication
- GFR Assessment using eGFR / SDMA
- Skin irritation can be reduced by diluting the desferoxamine in a larger volume (12cc H_20/gm desferoxamine) and/or adding 5 mg hydrocortisone to infusate.
- Use of serial erythrocytapheresis for chronic transfusions may delay or limit the duration or avoid entirely the need for Chelation.
- Counsel regarding avoidance of excess dietary iron.
- Consider vitamin C supplementation, 100-250 mg/d, only at start of each dose of desferoxamine.

Monitoring:

Monthly

- FBC, HbS%, Reticulocyte count, type and cross, antibody screen, serum ferritin
- Height, weight, history, physical exam
- Renal Profile
- Hb electrophoresis, ferritin, LFTs
- Liver function tests. Consider Hepatitis C, HIV, alkaline phosphtase, thyroid profile, fasting glucose, and other endocrine studies as indicated.
- Assess acceptance and compliance of patient and family with desferoxamine therapy.

Yearly Monitoring

- Audiology evaluation
- Ophthalmology examination
- Ophthalmology consultation for any new visual symptoms
- Consider CXR, ECG, echocardiogram / Cardiac MRI T2*
- Liver MRI T2* or Ferriscan.
- Consider CNS evaluation including TCP, MRI, MRA, and/or neurocognitive testing for patients with stroke

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