

Adult Service**Deferiprone (Ferriprox) chelation in patients with haemoglobin disorders:
Clinical Guideline****Indication**

Deferiprone should be considered second line therapy for patients with major haemoglobinopathies requiring chelation therapy, especially if the cardiac MRI T2* <10ms.

It is most effective in removing cardiac iron, but can be useful in removal of hepatic iron in patients who have been splenectomised.

Prior to commencing treatment:

- Provide the patient with Patient Information Leaflet on Deferiprone (<https://www.whittington.nhs.uk/document.ashx?id=5981> From the Red Cell Network Thalassaemia HCC, with permission)
- All patients must be advised to report any symptoms indicative of infection such as fever, sore throat and flu-like symptoms and provided with local acute service contact details.

Dose Range

25mg/kg body weight, three times daily, to a total daily dose of 75 – 100mg/kg/day. The dose should be rounded down to the nearest half tablet (i.e. to the nearest 250mg). Consideration should be given to using 100mg/kg/day dose for all patients where it is being used as monotherapy. Doses above 100mg/kg/day are not recommended.

Relative contraindications

- History of unexplained neutropenia
- Pre-existing arthropathy
- Pregnancy or breastfeeding
- End stage renal failure
- Concurrent therapy with other agents associated with neutropenia (e.g. hydroxycarbamide),
- Immunocompromise, including HIV.

Baseline investigations

- FBC, reticulocyte count
- LFT, Renal, Bone, CRP, AFP, Urinalysis
- Ferritin, Iron, Transferrin saturation
- Cardiac T2* MRI, Ferriscan

Initial Dose

- 75mg/kg total daily dose
- Increase monthly up to maximum dose of 100mg/kg total daily dose

Monitoring

- Weekly FBC, for the first month then as per routine management
- Monthly – FBC, LFT, Renal, Bone, CRP, Urinalysis,

Adult Service

- 3 Monthly – Ferritin (stop treatment and monitor if ferritin <500microgram/l), Zinc
- Annually – Cardiac MRI T2*/Ferriscan while evidence of iron loading present, once normalised monitor by Ferritin.
- Audiometry annually, if used in combination
- Ophthalmology annually, if used in combination

Dose adjustments for Adverse Effects (incidence)

Agranulocytosis (1-2%)

- Stop the drug immediately and consider use of G-CSF
- Monitor neutrophil count at least weekly until recovery
- Do not re-challenge in the future
- Patients should be warned to report immediately any fever or sore throat

Neutropenia (4%)

- Temporary withdrawal of drug until resolved
- Monitor FBC at least weekly until recovery of neutrophils
- Patients should be warned to report immediately any fever or sore throat

Arthropathy

- Provide analgesia (NSAIDs), consider dose reduction
- Consider referral to Rheumatology / Orthopaedic surgery, and joint imaging under specialist guidance

Gastrointestinal side effects:

- Nausea, vomiting, diarrhoea and abdominal pain are more frequent at the beginning of therapy and usually resolve within a few weeks without the discontinuation of treatment.

Febrile Neutropenia

Manage as per Local Neutropenic Sepsis guidelines:

- All patients must be advised to report any symptoms indicative of infection such as fever, sore throat and flu-like symptoms.
- Stop the drug immediately
- Consider use of G-CSF

Documentation and acknowledgements:

Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK © Sickle Cell Society 2018

A. Victor Hoffbrand, Ali Taher, and Maria Domenica Cappellini. How I treat transfusional iron overload. Blood, 2012, Volume 120, Number18

Adult Service

United Kingdom Thalassaemia Society. Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK. 3rd Edition, 2016.

John Porter, Vip Viprakasit, and Antonis Kattamis. Guidelines for the Management of Transfusion Dependent Thalassaemia (TDT) [Internet]. 3rd edition Chapter 3. <https://www.ncbi.nlm.nih.gov/books/NBK269373/#APP.001>

NHSE. Clinical Commissioning Policy 16070/P: Treatment of iron overload for transfused and non- transfused patients with chronic inherited anaemias. 26 August 2016

Deferiprone (Ferriprox ®). Summary of product characteristics. Last Updated on eMC 27-Jun-2016.

Farrukh T. Shah, John B. Porter, Nandini Sadasivam, Banu Kaya, James C. Moon, Mark Velangi, Emmanuel Ako, Shivan Pancham, BJH guidelines, *Guidelines for the monitoring and management of iron overload in patients with haemoglobinopathies and rare anaemias*, 06 October 2021

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Review

Name	Revision	Date	Version	Review date
Wale Atoyebe	Pre-peer review	Jan 2013	1.0	Jan 2015
Wale Atoyebe	Pre-peer review	Feb 2013	1.1	Feb 2015
Deborah Hay	Routine review	Aug 2015	1.2	Jan 2017
Dr Magbor Akanni, MKUH. Nadjoua Maouche, Pharmacist Sandy Hayes, ANP	Review, additional referencing, ODN format	Jan 2017	2.0	Oct 2018
Dr Noemi Roy	Update, patient information link	June 2020	2.1	June 2022
Wale Atoyebe	Full review	October 2023	3.0	October 2025