NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

# Scottish Paediatric & Adult Haemoglobinopathy Network

## Adult Guideline
## Thalassaemia Outpatient Management

This guideline is designed to support the clinician when meeting a patient for the first time, or when performing an Annual Review.

**NOTE**
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

## Annual Review

The following should be considered at initial visit and each annual review visit:

- General wellbeing
- Transfusion issues: frequency, adequacy of pre-transfusion Hb, venous access issues, transfusion reactions
- Chelation issues: drugs, dose, compliance, monitoring as per chelation guideline
- Episodes of hospitalisation
- Current medications, side effects and compliance issues
- Immunisation status: Hep B status, post-splenectomy vaccines, if appropriate
- Outcomes of other specialist reviews e.g. endocrinology, cardiology, ophthalmology, audiology
- Upcoming surgical procedures
- Any other issues of concern for the patient
- Family planning and screening including partner testing and pre-pregnancy counselling if appropriate

## Examination

- Height
- Weight
- Cardiovascular examination
- Abdominal examination for organomegaly

## Once only investigations to be performed at initial visit

- Alpha and beta globin genotype – via Edinburgh Genetics lab
- Red cell genotyping (will presumably have been recently transfused and therefore phenotyping not an option)
- Notification of special transfusion requirements to local blood bank

# Regular Investigations (See table below)

| TESTS | FREQUENCY | INDICATION |
|---|---|---|
| FBC | • Pre-transfusion<br>• If infective symptoms or signs on DFP | Pre-transfusion Hb monitoring; ANC monitoring, if on deferiprone (DFP) |
| Calcium*, phosphate and ALP | • 6 monthly | Bone mineral density assessment |
| Vitamin D | • Annually | |
| DEXA | • 2-3 yearly | |
| LFTs | • As per iron chelation guidelines | Monitoring of iron chelation (see iron chelation guidelines) |
| Ferritin | | |
| U&E | | |
| TFTs | • Annually, or if symptomatic | Monitoring endocrine function |
| Morning cortisol level | • Annually | |
| Morning testosterone levels** (males) | • Annually | |
| Glucose tolerance test | • Annually | |
| HepB sAg, Hep B core abs; HCV antibody | • Annually | Monitoring for liver disease |
| AFP, US/CT/MRI Liver, if cirrhosis | • 6 monthly | HCC surveillance |
| MRI (Liver) – see iron chelation guidelines for further information | • 6 -12 monthly | < 3 mg/g dw or rapidly falling |
| | • Annually | > 7 mg/g dw |
| | • 1 – 2 yearly | 3 – 7 mg/g dw |
| Pure tone audiometry | • Annually | DFX, DFO or combination DFO & DFP |
| ECG | • Annually (16-25 years old) | Asymptomatic, stable and well chelated patients with previously normal results |
| | • 2 yearly (> 25 years old) | |
| | • Consider more frequently | • Poorly chelated patient, without heart failure or impaired LV function AND patients recovered from an episode of acute heart failure AND those with impaired LV but no symptoms<br>• Pregnancy at first visit |
| ECHO | • Annually (16-25 years old) | Asymptomatic, stable and well chelated patients with previously normal results |
| | • 2 yearly (> 25 years old) | |
| | • 6 monthly | Poorly chelated +/- LVSD or heart failure |
| | • Consider more frequently | Pregnancy |
| MRI (Cardiac T2*) - – see iron chelation guidelines for further information | • 3 - 6 monthly | < 10 ms |
| | • Annually | 10-20 ms |
| | • 2 yearly | > 20 ms |

*Parathyroid hormone level if calcium low
** LH/FSH and sex hormone binding globulin level if morning testosterone low

## Specialist review

- Cardiology
  - Pre-pregnancy; at 12 & 28 weeks gestation; 3 months post-partum
  - If clinical concerns otherwise
- Endocrinology
- Ophthalmology/Optometry/Audiometry as per chelation guidelines
- Audiology, if on desferrioxamine or deferasirox

## SPECIAL CIRCUMSTANCES

**If considering pregnancy**

- Cardiology review with ECHO and Cardiac T2* MRI
- Glucose Tolerance Test
- If diabetic, fructosamine to measure glycaemic control (aim < 300 nmol/L)
- Thyroid function tests
- Hepatitis B serology – ensure immune
- Ensure vaccinations up to date
- Bone density assessment - DEXA scanning
- NB iron chelation generally contraindicated in pregnancy (maybe used in special circumstances)

## References

Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK, 3rd edition (2016) http://ukts.org/standards/Standards-2016final.pdf