

Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric Guideline

Monitoring and management of Endocrine complications in children with Thalassaemia

Endocrine complications are mainly due to iron overload and are difficult to reverse. The aim should be prevention with adequate chelation therapy. Endocrine failure should be screened for regularly and managed jointly with a paediatric endocrinologist.

Growth and puberty

Monitoring

- Biannual height and weight and assess with growth chart
- Annual assessment of puberty with Tanner staging from age 10
- LH/FSH and oestradiol if menstrual disturbance develops
- Annual morning testosterone from age 14 with LH/FSH/SHBG if low

Management

- Declining height velocity should be investigated and desferrioxamine toxicity and growth hormone deficiency considered
- Delayed puberty should be investigated and hormone therapy considered

Bone health

Monitoring

- Calcium, phosphate, alkaline phosphatase 6 monthly from age 12
- Check PTH if deranged bone profile
- Vitamin D annually from age 2 at latest
- DEXA scan from age 16

Management

- Replace vitamin D and /or calcium as per local guidelines
- Encourage patients to have an active lifestyle and diet rich in calcium

Other

- Random glucose 6 monthly
- GTT annually from puberty or age 10 if family history of diabetes
- Morning cortisol annually from age 14
- Annual thyroid function tests from age 10

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.