



# Sickle Cell and Thalassaemia Centre 0121 507 6040

Sandwell and West Birmingham Hospitals



NHS Trust

## What is Thalassaemia?

Thalassaemia is the name given to a family of conditions where less haemoglobin than normal is produced. Of these the most serious are alpha thalassaemia major (which is incompatible with life and babies do not usually survive the pregnancy) and beta Thalassaemia major (BTM) – where a defect in the normal haemoglobin gene prevents the body from producing haemoglobin.

This is a **quantity** problem that results in life threatening anaemia. People with BTM need regular blood transfusions and life-long treatment to clear excess iron, known as chelation. The highest prevalence of BTM is among Pakistanis, Cypriots, Italians, Greeks, Indians, Bangladeshis, Chinese and other South East Asian groups.

Thalassaemia Trait or carrier status does not cause health problems for the individual.

## **Treatment for Thalassaemia**

1. Blood transfusions give normal red blood cell (rbc) to the person with BTM. The rbc have a limited lifespan. So, transfusions normally have to be repeated every 3-4 weeks.
2. Chelation helps the body get rid of excess iron. With thalassaemia, the body gets overloaded with iron. This is from blood transfusions, and also because the thalassaemia itself makes the body absorb more iron from food. This treatment is **really important**; if excess iron is not removed, it can damage internal organs and cause complications.

**Thalassaemia is mostly managed as an outpatient, patients may need an inpatient admission because of:**

**Complications of anaemia and transfusions** Untreated anaemia can affect growth and bone development. Anaemia can also cause an enlarged spleen.

**Complications of iron overload** eg with the endocrine system, heart or liver.

**Complications of chelation** Chelation treatments have various possible side-effects that could involve the blood, liver, kidneys, vision, hearing, and bones.

**Infections** People with BTM can be more prone to serious bacterial infections; it is important that doctors and nurses know about the patient's thalassaemia and the treatment. Non-specialist doctors should contact your thalassaemia specialists for advice. Presence of CVAD or portacath, patients on chelation therapy and patients who have had a splenectomy are all at high risk for bacterial infections.

**Bone problems** for example osteoporosis (thinning of bones) can occur at a younger age than usual.

Refer to the:

[Trust guidelines on the management of non transfusion dependent patients with thalassaemia](#)

[Trust guidelines on the management of transfusion dependent thalassaemia](#)