DISABILITY RISK FACTORS FOR THALASSAEMIA

The following factors should be considered for the disability assessment of people with thalassaemia:

RISK FACTORS	ASSESSMENT CRITERIA		
Age		45-50 years old 30-45 years old < 30 years old	In countries where quality health care is provided, thalassaemia syndromes are entering a new era of prolonged survival. Thalassaemia is, however, a disability-inducing disease, with comorbidities appearing as age progresses.
Pre-transfusion Hb		<7 mg/dl 8.0-9.0 g/dl 9.5-10.0 g/dl	A pre-transfusion haemoglobin of 9.0-10.5 g/dl has been shown to promote normal growth, allow normal physical activities, adequately suppress bone marrow activity and minimise transfusional iron accumulation.
Serum Ferritin		>3000 mg/L 2000-3000 mg/ <2000 mg/L	Serum ferritin (SF) generally correlates with body iron stores, and is relatively easy and inexpensive to determine repeatedly. Serum ferritin is most useful in identifying trends: an increasing SF trend implies an increasing iron burden that requires further clinical investigation.
Cardiac MRI T2*		<10 ms 10-15 ms >20 ms	T2* is an MRI technique to measure liver and cardiac iron, recommended as part of yearly monitoring of multi-transfused patients at risk of developing myocardial or liver iron loading. It requires shorter acquisition times and allows the early identification of patients at high risk of developing organ deterioration and complications.
Liver Iron Concentration		>11 mg/gdw 7-10 mg/gdw <7 mg/gdw	Liver iron concentration (LIC) is measured to identify whether body iron is adequately controlled, as it is linked to the risk of hepatic/ extrahepatic damage. Sustained high LIC (above 15-20 mg/g dry wt) have been linked to worsening prognosis, liver fibrosis progression or liver function abnormalities. The most common methods to measure LIC is through biopsy, SQUID and MRI.
Splenectomy		Yes Yes No	If the clinical management of transfusion-dependent thalassaemia is inadequate, the increased destruction of red blood cells results in splenomegaly. Splenectomy, the surgical removal of the spleen, is a treatment option with major adverse effects, including Sepsis, Thrombophilia, Pulmonary hypertension and Iron overload, affecting both the survival and quality of life of patients.
Comorbidities		2+ 1-2 | Well managed None	Iron overload causes a number of co-morbidities including vascular disease, pulmonary hypertension, heart cardiomyopathy leading to heart failure; pituitary damage, growth retardation; endocrine complications; liver disease. Bone disease is common at all ages, causing pain, fractures (osteopenia, osteoporosis) and limiting mobility. The presence of these co-occurring conditions should be regarded as causing disability.

THALIA

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