1. Introduction

The use of splenectomy in thalassaemia has declined in recent years. This is partly due to a decreased prevalence of hypersplenism in adequately transfused patients. There is also an increased appreciation of the adverse effects of splenectomy on blood coagulation. In general, splenectomy should be avoided unless absolutely indicated.

Indications of Splenectomy in the transfusion-dependent patient:

- When hypersplenism increases blood transfusion requirement and prevents adequate control of body iron with chelation therapy.

- An enlarged spleen—without an associated increase in transfusion requirement—is not necessarily an indication for surgery. Patients with hypersplenism may have moderate to enormous splenomegaly, and some degree of neutropaenia or thrombocytopaenia may be present.

- Annual transfusion volume exceeding 225 to 250 mL/kg per year with packed red blood cells (haematocrit 75 percent) may indicate the presence of hypersplenism. The volume calculation should be corrected if the average haematocrit is less than 75 percent. The possible development of alloantibody should also be ruled out.

- Splenectomy should be avoided unless there is an inability to maintain iron balance with optimal chelation or if there are clinically significant complications such as pancytopenia and marked enlargement.

- Often, hypersplenism develops because of low pre-transfusion haemoglobin. Increasing the pre-transfusion haemoglobin to between 9.5 and 10 may reverse hypersplenism.

- The indications for splenectomy in haemoglobin H–Constant Spring patients are different than in beta-thalassaemia disorders.

- Transfusion-dependent infants with haemoglobin H–Constant Spring respond rapidly to splenectomy but require prophylactic anticoagulation because of a high incidence of serious thrombosis.
Types of splenectomy:
- If a decision to perform surgery is made, partial or full splenectomy is the option.

- Partial splenectomy is a complicated surgery utilized to preserve some splenic function. It should be reserved for infants requiring splenectomy.

- Full splenectomy can usually be performed by laparoscopic technique. However, open procedure is necessary in cases of marked splenomegaly.

Precautions pre and post splenectomy:
- Patients must receive adequate immunization against Streptococcus pneumoniae, Haemophilus influenza type B, and Neisseria meningitides at least 2 weeks prior to surgery.

- Splenectomy should be avoided in children younger than five years because of a greater risk of fulminant post splenectomy sepsis.

- After splenectomy, patients should receive oral penicillin prophylaxis (250 mg twice daily) and be instructed to seek urgent medical attention for a fever over 38.5°C.

- Post-splenectomy thrombocytosis is common, and low dose aspirin should be given during this time.

- Another complication following splenectomy is the development of a thrombophilic state. Venous thromboembolism, more common in thalassaemia intermedia and haemoglobin H–Constant Spring, can develop following splenectomy.

- Patients should have annual echocardiographic measurement of the pulmonary artery pressure to monitor for development of pulmonary hypertension.

3. References
Standard of Care Guidelines for Thalassaemia (Children’s Hospital & Research Center Oakland (2008))
Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK (2008)

4. Legal Liability Guideline Statement
Guidelines issued and approved by the Trust are considered to represent best practice. Staff may only exceptionally depart from any relevant Trust guidelines providing always that such departure is confined to the specific needs of individual circumstances. In healthcare delivery such departure shall only be undertaken where, in the judgement of the responsible healthcare professional, it is fully appropriate and justifiable - such decision to be fully recorded in the patient’s notes.