

# SPLENOMEGALY AND SPLENECTOMY

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# Splenomegaly and splenectomy

## Introduction

- **Increased destruction** of RBC in the spleen, together with **extramedullary hematopoiesis**, results in **splenomegaly**
- **Splenectomy**, is **more** commonly **needed** in **TI** than **TM**
- The **main reason** for splenectomy in **TM** is to **decrease transfusion** and resultant **reducing iron** overload

# Introduction

- Current transfusion guidelines reduced incidence of splenomegaly and need for splenectomy
- The probability of undergoing surgery for patients with TM within the first 10 years of life is
  - 57% if born in 1960s
  - 22% if born in 1970s
  - 6% if born in 1980s and
  - 7% if born in 1990s

# Introduction

- Spleen **Size** in TM should be checked by **PhE** and as needed, by **ultrasonography**
- **Splenomegaly** due to **under-transfusion** may be **reversible**
- **Before** considering **splenectomy** in **this** situation, patient should be placed on **adequate transfusion** program and then **re-evaluated**
- The **spleen** may **increase** in **size** during **pregnancy** and these patients require careful **F/U**

# Steps to reduce or to delay the splenomegaly

- Maintain pre-transfusion Hb  $>9$  g/dl
- Sustained splenomegaly despite this regimen may be reversible by increasing transfusion, at the cost of increased iron load
- Use of the fresh PC ,less than 2 weeks old

# Indications for Splenectomy

- All agree to **restrict it** due to increased **risk** of:
  - venous **thrombosis**
  - **pulmonary** hypertension
  - Overwhelming **infections** after splenectomy
- Splenectomy should be **avoided** in children **<5** years because of a considerably greater risk of fulminant **sepsis**

**Table 1.** Indications for Splenectomy in Thalassaemia Major.

INDICATION	COMMENT
Increased blood requirements that prevent adequate control with iron chelation therapy, having made sure that the increased requirements are not due to allo/auto-antibodies or blood loss.	Annual transfusion volume used to flag an increased blood requirement (200-275 ml/kg/yr red cells)*. However, if effective chelation therapy continues to be maintained despite increased transfusion, splenectomy may not be necessary.
Hypersplenism	Cytopenias causing clinical problems
Symptomatic splenomegaly	<ul style="list-style-type: none"><li>• Accompanied by symptoms such as left upper quadrant pain or early satiety.</li><li>• Massive splenomegaly causing concern about possible splenic rupture.</li></ul>

# Splenectomy and Peri-operative Complications

- Surgery should include a careful **search** for **accessory** spleens
- There are **four approaches** to splenectomy:
  - **Open** total splenectomy
  - **Laparoscopic** total splenectomy,
  - **Partial** splenectomy and
  - Reduction of splenic tissue by **embolization**



# Splenectomy and Peri-operative Complications

- **Laparoscopic** splenectomy is associated with:
  - **Reduction** in postoperative **mortality**,
  - **Shorter** hospital **stay**, and
  - **Fewer** pulmonary, wound and infectious **complications**
- Also, **superiority** of **laparoscopic** splenectomy in patients with **massive** spleen **demonstrated**

# Splenectomy and Peri-operative Complications

- Partial splenectomy is used to preserve of the immune function while reducing the degree of hypersplenism
- There is no conclusion about effectiveness of partial splenectomy *VS* total splenectomy
- The long-term success of this approach is still undergoing evaluation
- Splenic re-growth and the volume of splenic tissue required to preserve immune function are two main questions

# Splenectomy and Peri-operative Complications

- Splenic embolization is less invasive
- The exact splenic volume necessary is undefined, optimal reduction is about 50%- 70%
- Common complications are fever, nausea, pain and possible need for a subsequent total splenectomy
- Significant complications include abscess formation, pleural effusion, portal vein thrombosis and liver failure
- Serious complications occur when the volume embolized is 70% or greater
- Splenic embolization has not gained wide acceptance
- Embolization does not permit a search for accessory spleens

# Concomitant Cholecystectomy

- An evaluation for **gallstones** should be performed **prior to splenectomy**,
- **Especially** if patient has **related** symptoms
- **Positive** findings will lead to **cholecystectomy** at the **same time**
- Removal of the **appendix** at the time of splenectomy **may prevent** later **problems** in distinguishing infection with **Yersinia** from appendicitis
- Splenectomy is a **opportunity** for **liver biopsy**

# Adverse Events of Splenectomy

## Peri-operative complications

- Bleeding
- Atelectasis
- Subphrenic abscess

## Major adverse effects are

- Sepsis
- Thrombosis and
- Pulmonary hypertension

# Sepsis

- The **major** long-term **risk** after splenectomy is **sepsis** because:
- **Spleen** has important **defense functions** by **removing** antigens and synthesizing **antibodies**, tuftsin, and **Igs**, principally IgM
- **Removal** of the **spleen** is associated with **predisposition** to severe **infections** and **mortality**

## **The risk of overwhelming post-splenectomy infection varies with:**

- **Age**: risk is greatest in children **<16** and in adults **> 50 years**
- **Time since splenectomy**: the **risk** of sepsis is **life-long** and reported as much as 25-40 years after splenectomy,
- However, the **greatest** risk is in the period **1-4 years** after surgery

# Sepsis

## status of patient

- Frequent pathogens are *Streptococcus pneumoniae*, *H-influenzae* type B, and *Neisseria meningitidis*, with high mortality rate
- Other organisms are *E-coli*, *Pseudomonas aeruginosa*, *Salmonella*, and *Klebsiella*
- Vaccination and prophylactic antibiotics can prevent pneumococcal infections in first 2-4 post-splenectomy yrs
- *Babesia* have been implicated in a fulminant haemolytic febrile state in splenectomized patients
- Malaria is more severe in asplenic people with an increased risk of death

# Sepsis

## status of patient

- Sudden onset of fever and
- Chills,
- Vomiting and
- Headache
- The illness rapidly progresses to hypotensive shock, and is commonly accompanied by DIC
- The mortality rate is approximately 50%,
- Therefore, early intervention based on suspicion, even in the absence of many of the above findings, is critical



**Table 2.** Immune prophylaxis in splenectomised patients (modified from Davies 2011)

VACCINE*	SCHEDULE	COMMENT
Streptococcus Pneumoniae Pneumococcal polysaccharide (23 valent) vaccine [Pneumovax 11] 1 dose 0.5ml IM <sup>1</sup>	4-6 weeks pre-splenectomy or at least 2 weeks in advance <sup>2</sup> . Repeat 5 yearly thereafter.	Rate of protection is 70-85% • The immune response is poor in children less than two years of age. In this age group PCV7 may be used
Haemophilus influenzae type B	4-6 weeks pre-splenectomy or at least 2 weeks in advance <sup>2</sup> .	There are currently no recommendations for repeat vaccinations
Meningococcal Group C conjugate vaccine	4-6 weeks pre-splenectomy or at least 2 weeks in advance <sup>2</sup> . MenACWY 2 months later Repeat 5 yearly thereafter.	
Influenza virus vaccination	Yearly	This should also be offered to non-splenectomised patients

\* Children vaccinated under the age of two should be re-vaccinated at age two.

<sup>1</sup> CDC (USA) guidelines additionally advise that a second pneumococcal vaccine, PCV13, should also be given 2 months prior to the 23 valent vaccine.

# Chemoprophylaxis in splenectomized patients

- Risk of infection after splenectomy is lifelong, but is highest in the first few years post-operatively
- A broad spectrum antibiotic should be given pre- and continued post-operatively
- There are no definite guideline regarding the length of time post-splenectomy penicillin should be continued
- A policy is the use of life-long prophylactic penicillin
- One guidelines recommend to life-long prophylactic antibiotics for patients at high risk of pneumococcal infection using penicillins or macrolides

# Chemoprophylaxis in splenectomised patients

- **Another guideline** recommend penicillin prophylaxis post-splenectomy **up to the age 16 years and over the age of 50 years**
- **Others** recommend prophylaxis in **children for 5 years and adults 2 years** post-splenectomy
- **Antibiotic** prophylaxis should be prescribed when a splenectomized patient **undergoes invasive dental** procedures and should be applied for pre- or post- of **any other invasive** procedure

# Chemoprophylaxis in splenectomized patients

- The importance of **compliance** with prophylactic antibiotics should be **stressed** to patients and parents
- However, Patients and parents should recognize that **chemoprophylaxis** does **not prevent all** cases of post-splenectomy sepsis
- The **risk of death** from febrile illnesses remains **high**, and **rapid evaluation** of febrile episode is essential
- Patient and parents must **report febrile** illnesses and **seeking** immediate **medical** attention

## For febrile episodes, the physician should consider

- Evaluating the patient with **PhE, B/C** and other cultures as indicated
- Starting **antibiotics** against Streptococcus, Neisseria and Klebsiella **while waiting** for cultures
- **Liver abscess** should be screened, **particularly** in **diabetic** patients
- Patients need to be **aware** of the potential for **travel-related** infections such as **malaria**
- Patients should be given a **card** with their splenectomy status and **contact number** of their primary treatment centre/team

# Hypercoagulability

- **Thromboembolism** is common both in the **peri-operative** period and **postsplenectomy**
- TM patients, particularly those with splenectomy have tendency for **both venous** and **arterial** embolism
- One of the **main** factors is the **procoagulant effect** of **phospholipids** on the surface of **altered RBC**, as the number of these cells is dramatically **increased** after **splenectomy**
- In **post-splenectomy** patients, markers of thrombin generation such as **D-dimer** should be assessed **annually**,
- **Portal vein thrombosis** post-splenectomy can also occur despite prophylactic anticoagulation

# Hypercoagulability

- Following splenectomy, **thrombocytosis** is **common**, with PLT often reaching **1,000,000**
- The **UK** thalassemia **guidelines** state that peri-operative **thromboprophylaxis** should be **routinely** given and continued **until** the patient is **fully mobile**
- It is recommended that all post-splenectomy patients receive **aspirin** as long as there are **no** **contraindications**
- This is **particularly** important for patients with a **history** of previous **thrombosis** or other risk factors

# *Pulmonary hypertension*

- This complication is **more frequent in TI**, but is also increasingly identified in TM
- **Advancing age** and a **history of splenectomy** are major **risk factors** in this population
- **Pulmonary HTN** is a consequence of delayed of **pulmonary artery thromboembolism**
- It is shown that **pulmonary HTN** is **associated** with **splenectomy**



# Iron overload

- Spleen is a major organ of iron storage in TM
- Following splenectomy, the total body iron storage capacity is reduced
- Splenectomy causes major changes in the status of iron overload and toxicity in TM patients
- Iron will be redirected and in the liver, heart and other organs, unless effective chelation is introduced
- Splenectomized patients had higher incidence of myocardial iron load (48%) compared to non-splenectomized patients (28%)

# Summary and Recommendations

- Splenectomy is the **recommended** intervention to **reduce blood** requirement and consequent severe **iron** overload
- But **splenectomy** has a **high** disease **burden**
- **Current** transfusion **regimens** and **chelation** have **reduced** the incidence of **splenomegaly** and **iron overload** in TM patients

# Summary and Recommendations

1. It is **not recommend** splenectomy as a **standard** procedure in TM
  - Splenectomy is **linked** to a variety of **complications** such as **pulmonary** HTN, brain **infarcts**, venous **thrombosis** and **sepsis**

**But should be considered in **three** clinical scenarios**

- **Increased** blood **requirement** that **prevents** iron control
- **Hypersplenism** and
- **Symptomatic splenomegaly**

# Summary and Recommendations

2. **Laparoscopic** splenectomy seems to be the **most favorable**
3. Most frequent **pathogens** in splenectomised patients are **Streptococcus**, **Neisseria**, Haemophilus influenzae **type B**
  - **Immune prophylaxis** is recommended against these agents at **least 2 weeks** prior to the operation and **repeated** per guideline
  - **Annual influenza** vaccination is encouraged

# Summary and Recommendations

4. Chemoprophylaxis with penicillin depends on the age of the patient and physician's opinion
  - Life-long antibiotics should be offered to patients considered at high risk of pneumococcal infection using oral penicillins or macrolides
5. Due to new transfusion and chelation protocols, fewer splenectomies are seen than before

# Summary and Recommendations

- **ADHERE** to current transfusion guidelines to prevent or delay splenomegaly
- If splenectomy cannot be avoided, make sure **IMMUNIZATION** protocols are followed
- Be aware of the dangers of **THROMBOSIS** in the peri- and post-operative period
- Discuss **CHEMOPROPHYLAXIS** with patient/family and make sure they are aware of the dangers of post-splenectomy sepsis
- **START IV** antibiotics in case of suspected sepsis while awaiting culture results



THANK YOU