Guidelines for

Splenectomy in Haematological Diseases

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GUIDELINES : ELECTIVE SPLENECTOMY

CONTENT:

Indications of Elective Splenectomy

POSSIBLY INDICATED
1. Hereditary Spherocytosis
2. Immune Thrombocytopenia
3. Pyruvate Kinase Deficiency
4. Splenic Abscess
5. Splenic Marginal Zone Lymphoma
6. Splenomegaly (Massive or symptomatic)
7. Transfusion dependent thallasemia
8. Warm Autoimmune hemolytic anemia

RARELY INDICATED
1. Chronic lymphocytic Leukemia
2. Hairy Cell Leukemia
3. Primary Myelofibrosis
4. Splenic Infarction
5. Splenic sequestration crisis in sickle cell disease
GENERALLY CONTRAINDICATED
1. Hereditary Stomatocytosis
2. Hereditary Xerocytosis
3. Paroxysmal cold hemoglobinuria
4. Cold Agglutinin Disease
5. Autoimmune lymphoproliferative syndrome
6. Thrombocytopenia in hepatic cirrhosis
7. Gaucher Disease

Reference: Indications for splenectomy, Am Surg 2006;72(7):565

PREOPERATIVE-CARE

1. Vaccinations:

Elective Splenectomy: At least TWO weeks prior to surgery.

Adults

Hib MenACWY PPV	Every 5 Years PPV + MenACWY	Yearly Influenza vaccine

Children

Age	Month 0	Month 1	Later
Under 2 years	Complete according to national routine childhood schedule, including booster doses of **Hib/MenC and PCV13**	A dose of **MenACWY** conjugate vaccine should be given at least 1 month after the Hib/MenC and PCV13 booster doses	After the second birthday, one additional dose of **Hib/MenC and a dose of PPV** should be given
Over 2 years and under 5 years (previously completed routine childhood vaccinations with PCV7)	**Hib/ MenC booster +PCV13**	**MenACWY** conjugate Vaccine	**PPV** (at least 2 months after PCV13)
Over 2 years and under 5 years (previously completed routine childhood vaccinations with PCV13)	**Hib/MenC booster+ PPV**	**MenACWY** conjugate Vaccine	
Over 2 years and under 5 years (unvaccinated or previously partially vaccinated with PCV7)	**HibMenC** vaccine First dose of **PCV13**	**MenACWY** conjugate vaccine	Second dose of **PCV13** and then **PPV** (at least 2 months after PCV13)

Source: Davies et al., 2011 [Br J Haem 155, 2011, 308–17].

Special consideration:

Chemotherapy/ Irradiation/Immunosuppressive agents	Vaccinations should be given at least TWO weeks before initiation of treatment. Where it is not possible to vaccinate beforehand, splenectomy, chemotherapy or radiotherapy should never be delayed. If it is not practicable to vaccinate TWO weeks before the initiation of chemotherapy and/or radiotherapy, immunization can be delayed until at least THREE months after completion of therapy in order to maximise the response to the vaccine, whilst ensuring adequate antibiotic cover is prescribed in the interim
Pregnancy/Breast feeding	All of the vaccines may be given during pregnancy and breast-feeding when protection is required without delay.
Travel	Patients should be educated as to the potential risks of particularly with regards malaria and unusual infections, for example those resulting from animal bites and tick bites. Patients travelling for Hajj will require MenACWY conjugate booster dose.

2. Optimizing Haemoglobin and Platelet Count before surgery:

I. **Hemoglobin – >** 10g/dl

II. **Platelet count** – > Ideally 50 x 10^9/l. In patients with ITP, this may be achieved using Single donor or Random donor platelets before and after the surgery. In most of the clinical situation, target platelet count > 50 x10^9/l may not be achieved ; then procedure to be undertaken under platelet transfusion cover.

N Engl J Med. 2011 Dec;365(26):2453-62. Epub 2011 Dec 14

Medication Adjustment:

i. **Glucocorticoids:** If patients are still on a high dose of corticosteroids prior to surgery, reduce that dose prior to surgery. Steroids should not be stopped abruptly before surgery. Dose of hydrocortisone to maintained at 50-100mg/24 hours.

ii. **TPO-RAs** -Care should be taken in adjusting the dose postoperatively in order to avoid thrombocytosis or thrombocytopenia. Attention to venous thromboembolism (VTE) prophylaxis should be given.

Ther Adv Hematol. 2019;10:2040620719841735. Epub 2019 Apr 11
Can J Surg. 1992 Aug;35(4):432-6

VENOUS THROMBOEMBOLISM PROPHYLAXIS

Splenectomy is associated with **the risk of venous thromboembolism (VTE), including portal and splenic vein thrombosis , deep vein thrombosis (DVT) and pulmonary embolism (PE) and has around** 2 fold higher risk of VTE as compared to other surgeries. Pharmacologic thromboprophylaxis post- surgery to be offered assessing the risk factors including BMI, immobilization, previous history of VTE and malignancy, **excluding ITP.**

Blood. 2009;114(14):2861. Epub 2009 Jul 27.
Blood (2013) 121 (23): 4782–4790
International Journal of Surgery,Volume 24, Part A, December 2015, Pages 27-32

Choice of pharmacologic thromboprophylaxis:	Enoxaparin/ dalteparin S/C weight based Riivoroxaban if not able to self-inject post discharge
Timing of starting thromboprophylaxis:	Give weight based dose enoxaparin/dalteparin SC at least 12 hours before surgery followed by 5,000 units of UFH fixed dose after surgery (6 hours post wound closure), to continue on the day after surgery with the weight-based prophylactic doses, as long as there is no significant active bleeding Dose of rivaroxaban thromboprophylaxis: 10 mg orally, daily, starting 6 to 10 hours post-operatively

Duration of Thromboprophylaxis:	Active cancer disease: Extended prophylaxis for 28 Days Non cancer disease: Minimum 7 days (VTE risk assessment to be done) Patients with previous history of VTE and not already taking long term anticoagulation: 6 weeks of pharmacological and mechanical thromboprophylaxis. Patients who develop thrombosis extending beyond the splenic vein: therapeutic doses of LMWH followed by oral anticoagulation for 3-6 months. Treatment of symptomatic Thromboembolism of splenic and portal vein) is mandatory and a protracted course of oral anticoagulation is suggested
Mechanical Thromboprophylaxis:	All surgical patients deemed to be at risk of VTE on their VTE risk assessment should be offered mechanical thromboprophylaxis

*Reference: (Krauth et al, **2008**).*

SURGICAL APPROACH

Open versus laparoscopic procedure

First choice - Laparoscopic

●Lower surgical mortality
●Shorter hospitalization and faster recovery
●Reduced complications

Settings in which an open procedure may be preferred include the following:

- Massive splenomegaly with concern about the ability to remove the spleen via a laparoscopic procedure.
- Local expertise favoring an open procedure, cost or lack of support or equipment for laparoscopy.
- Ability to search more thoroughly for an accessory spleen.

- Cancer surgery or adhesion of the spleen to adjacent organs requiring laparotomy.

Histopathology: Pathologic sampling of the resected spleen should be done promptly after resection

Simultaneous cholecystectomy

This is to be performed if there is history of pigment gallstones, which can develop as a complication of chronic hemolysis, such as with an inherited hemolytic anemia.

POSTOPERATIVE PERIOD

Thrombocytosis Post splenectomy

If Platelet >600 x 10^9/l	Aspirin
Platelet count > 1000 x 10^9/l with evidence of arterial or venous thrombosis	Aspirin + cytoreductive agents such as hydroxyurea or anagrelide with close monitoring of platelet counts.

1. PROPHYLAXIS ANTIBIOTICS

- High risk patients remain on lifelong antibiotic prophylaxis.
- All patients are at high risk of infection in the immediate post-operative period – antibiotic prophylaxis should be started immediately post-operatively.

High risk patients include:

• Patients under 16 or over 50 years of age.

• Patients with poor or no response to pneumococcal vaccination.
• Patients who have had a previous episode of invasive pneumoccal disease.
• Patients undergoing splenectomy for haematological malignancy particularly in the context of on-going immunosuppression; those who have received splenic irradiation or who have ongoing GvHD.

Age at start of prophylaxis	Duration	First line	If Penicillin allergic
Children (< 16years)	Continue until 16years of age Minimum 2 years	Amoxicillin 1month-5 years:125mg BD 5-12 years: 250mg BD 12 years: 500mg BD	Clarithromycin/Azithromycin 1month-5 years:125mg BD 5-12 years: 250mg BD 12 years: 500mg BD
Adults >16years	Ideally lifelong Minimum 2 years	Amoxicillin 500mg BD	Clarithromycin /Azithromycin 250 mg BD

I. **Malaria chemoprophylaxis:** Patients who are residents of malaria endemic areas should consider taking lifelong anti-malarial prophylaxis.

Chloroquine: 500mg once weekly
Doxycycline: 100mg Daily administration
Mefloquine: 250 mg Weekly administration
Primaquine : 15mg daily

Recognition & Management of Sepsis Post Splenectomy:

Clinical Features of sepsis in this patient group typically presents after a short prodrome of fever, chills, pharyngitis, muscle aches and vomiting / diarrhoea.
Suggested initial investigations:

- Full blood count,
- DIC screen,
- Urea & electrolytes,
- C-reactive protein (CRP)
- Blood cultures
- Other microbiological samples guided by clinical features (e.g. CSF etc)
- EDTA blood sample for urgent Malaria film and antigen screen

Management of Sepsis Post Splenectomy:

Immediate Self-Treatment by Patient Due to the potential for rapid deterioration, self-administration of a single dose of antibiotic by the patient at the first sign of a suspicious illness is advised. The single oral dose of “rescue” antibiotics is as follows:

- Amoxicillin 3g sachet PO stat OR,
- Cefixime 800mg PO stat (if non-severe penicillin allergy) OR,
- Azithromycin 1g PO stat (if severe penicillin / beta-lactam allergy)

Management of Hospitalised Patients:

Once recognised as sepsis, the administration of prompt antibiotics (within the hour) is vital and is the responsibility of the attending doctor.

	Standard Regimen	**Cephalosporin Allergy/ Intolerance**
Adults	**Ceftriaxone** 2gm 12 hourly OR **Cefatoxime** 2gm 4-6 hourly **PLUS Vancomycin** (25-30mg/kg loading followed by 15-20mg/kg 8 to 12 hourly)	**Meropenem** 2gmIV 8 hourly **PLUS** Vancomycin (25-30mg/kg loading followed by 15-20mg/kg 8 to 12 hourly)

Children	**Ceftriaxone** 50gm/kg12 hourly OR **Cefatoxime** 75mg//kg 6 hourly **PLUS** Vancomycin (15 mg/kg every 6 hourly)	**Meropenem** 40mg/kg IV 8 hourly **PLUS** Vancomycin (15 mg/kg every 6 hourly) (25-30mg/kg loading followed by 15-20mg/kg 8 to 12 hourly)

Management of Influenza:

Patients who appear clinically to have influenza may be offered antiviral therapy (Oseltamivir or Zanamivir)

Reference: Review of guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen: prepared on behalf of the British Committee for Standards in Haematology by a Worology party of Haematology-Oncology Task-Force 12-10-2011

CHECKLIST FOR SPLENECTOMY

Indication:	
Consent:	
Vaccination:	
Target blood cell parameters	

Type of procedure	
Patient information leaflet	
Post splenectomy thromboprophylaxis	
Post-Splenectomy antibiotic prophylaxis	
Post splenectomy anti-malarial prophylaxis	
Red alert / Bracelet issued	
Local General Physician information leaflet	
Self-care	
Travel advices	
Vaccination booster dose	