

## **Vesalius SCALpel™ : Spleen (see also: abdomen => spleen folios)**

### **Anatomy**

15-30% accessory spleens: 80% hilum, splenocolic ligament, omentum, pelvis, small bowel mesentery (scrotum male)

white zone: lymphatic nodules & germinal centers

red zone: thin walled sinuses, sinusoids, clear old, damaged RBCs

4-5 discrete vascular territories

marginal zone: interface

splenic cyst: true or post-traumatic (pseudocyst)(75%):

25% unknown cause

LUQ pain, US, CT: symptomatic: aspirate (recurrence, bleeding), unroof, partial splenectomy

congenital: epidermal, simple

parasitic: echinococcal most common; liver > lung > spleen in frequency

neoplastic: resect

dermoid: 3 layers, rarest, congenital, resect

neoplastic, echinococcal and dermoid resect; congenital and traumatic if symptomatic

no percutaneous aspiration, can bleed, recur

### **Splenic artery aneurysm**

3<sup>rd</sup> most common abdominal aneurysm after aorta, iliac

CT scan Dx

most near tail of pancreas, multiparous female, 5-10% incidence rupture

indications for surgery:

> 2cm asymptomatic

all pregnant women or of childbearing age

rupture during pregnancy maternal and fetal mortality 80%

if proximal can ligate proximally and distally, if distal may require splenectomy

embolization option: splenic infarction very painful

### **Function**

clear abnormal RBCs, platelets, cellular debris

RBC life cycle normally ~120d, 20cc removed daily by spleen

RBC membrane defect: spherocytosis (splenectomy), elliptocytosis

sickle cell autosplenectomy; increased risk of infection

should not see Howell-Jolly (residual nuclear chromatin), Heinz or Pappenheimer bodies

with normally functioning spleen, cleared by spleen

if splenectomy is complete (no accessory) will see the above on smear; if not still

have residual splenic tissue, accessory spleen

miss rate for accessory spleen lap-scope = open

platelets 10d lifespan, 1/3 of platelets pooled in spleen, can rise to 80% with splenomegaly

wbc 6h half life

abnormal function splenic antibodies (IgM) cause excess cell destruction, bind to platelets

opsonins

properdin: initiates alternate pathway of complement activation

tuftsin: binds granulocytes to promote phagocytosis

## **ITP**

Primary or secondary

Dx of exclusion:

SLE, antiphospholipid syndrome, immunodeficiency, lymphoproliferative, HIV,

hepatitis C, heparin, drug related antibodies (quinidine), thyroid disease

peripheral smear to R/O pseudothrombocytopenia, inherited giant platelet syndrome & other hematological disorders

immune disorder with IgG antibodies, F:M 3:1

IgG antiglobulin on platelets verifies

can follow upper respiratory infection

spleen is the major source of IgG in ITP, increased 5-6X

antibodies bind to platelets which are destroyed by spleen

platelets <50k, normal bone marrow

30-50K bruising

10-30K spontaneous ecchymosis

<10K internal bleeding

bleed: vaginal, mucosal, UGI, nose

usually sporadic, increasing with AIDS, SLE

spleen rarely palpable. if palpable consider Dx of hypersplenism

50% of cases children ~5yo, M=F

child good prognosis, 80% recovery without treatment < 6mo (usually within a few wks), do not do splenectomy, rare (<1%) intracranial hemorrhage

adults require Rx at presentation, 50% platelets < 10K

treatment

initial trial of steroids, 1mg prednisone/kg/d

3-6w, if responds wean, 50-75% success

initial response to steroids suggests good response to splenectomy

IV immune globulin for internal bleeding, platelets < 5K despite steroids, extensive progressive purpura

80% respond, but common relapse

if no response, requirement for high dose (10-20mg/d) or recurrent drop platelets:

elective splenectomy, 85% success

splenectomy

surgery 92% response v 30% medical Rx, surgery treatment of choice

predictors of response to surgery

young age, most common positive predictor

short interval diagnosis to surgery

initial response to steroids

HIV+

high pre-op platelets  
40K platelets OK to proceed  
don't give platelets unless untoward bleeding post op  
look for accessory spleen: 30% in hematological disorder v 20%  
plasmapheresis takes 4-5d for response, not beneficial in acute crisis  
emergency splenectomy only for neurologic crisis (intracranial bleed, pl < 10K)  
recurrence  
accessory spleen, observe  
search for other causes of thrombocytopenia: hemolytic disorder, thyroid, pregnancy,  
infection  
moderate, 40-50K try cytoxan, steroids, plasma exchange  
persistent: reoperate after CT, US, Tc scan looking for accessory spleen; gamma probe with  
indium labeled platelets

## **TTP**

autoimmune response to endothelial cell antigen (arterioles, small capillaries)  
disease of arteries with diffuse platelet trapping in small vessels (arterioles and capillaries)  
platelet aggregation, hyaline deposits in/under endothelium  
normal size spleen  
pentad: fever, purpura (thrombocytopenia), hemolytic anemia, neurologic abnormalities, renal failure  
profound thrombocytopenia, elevated WBC, elevated bilirubin (hemolysis), hematuria, protein casts  
peak 20-30yo, F>M  
may be initiated by viral or bacterial infection, pregnancy, drugs (BCP)  
untreated 10% 1y survival  
Rx steroids, plasmapheresis, rarely splenectomy

## **Hereditary spherocytosis**

autosomal dominant, defect in RBC membrane structural protein spectrin  
shortened RBC lifespan, osmotic fragility  
anemia, reticulocytosis, jaundice, splenomegaly, pigment gallstones (94% by 13)  
increased osmotic fragility, high sequestration, destruction by spleen  
wait until at least age 4 before splenectomy, cholecystectomy at same time

## **Sickle cell disease**

HbA replaced by HbS (valine substitution for glutamic acid 6<sup>th</sup> position on beta chain of Hb)  
with decreased O<sub>2</sub>, RBC elongates and distorts causing increased viscosity, stasis, crenation, clotting,  
worsening hypoxemia, cycle  
mortality from recurrent infection, renal failure, heart failure  
spleen autoinfarcts, rarely need splenectomy  
Howell-Jolly bodies seen  
splenic infarction may result in splenic abscess

## **Thalassemia**

autosomal dominant defect in Hb synthesis  
presents early in life  
persistence of HbF (fetal) and decreased HbA  
accumulation of intracellular material causes structural abnormality RBC  
need recurrent transfusion, can't maintain Hb > 10  
need for splenectomy  
high risk OPSS

**Primary hypersplenism** : rarely responds to steroids

## **Secondary hypersplenism**

portal hypertension may result in splenic enlargement, anemia, leucopenia,  
thrombocytopenia  
treat portal hypertension, no need for splenectomy

## **Splenic vein thrombosis**

acute or chronic pancreatitis, pancreatic tumor  
splenic enlargement, trapping  
normal liver (thrombosis may propagate into portal vein)  
isolated gastric varices, no esophageal varices  
gastric varices lower incidence of bleeding than esophageal, not amenable to banding  
splenectomy cures gastric varices and hypersplenism

**Acquired immune hemolytic anemia:** medical Rx 1<sup>st</sup>

**Hairy cell leukemia:** high recurrence after splenectomy; now treated with alpha2 interferon

## **Portal vein thrombosis**

hypercoagulable state, stasis, (long splenic v stump?)  
abdominal pain 1-2w post splenectomy, may result in dead gut  
anticoagulate, thrombolytic: heparin to coumadin X 6mo

## **Felty's syndrome**

rheumatoid arthritis, neutropenia, recurrent leg infection/ulcer, splenomegaly  
splenectomy may be beneficial if medical Rx fails

## **Sarcoidosis**

lung and liver disease, 20% incidence splenomegaly  
splenectomy may be beneficial

## Gaucher's

disorder of lipid metabolism  
splenectomy may be beneficial

## Myeloid metaplasia

progressive marrow fibrosis  
peripheral extramedullary hematopoiesis  
immature precursors in peripheral blood  
highest incidence of portal vein thrombosis  
splenectomy

## Splenic abscess

chills, fever, LUQ tenderness, splenomegaly  
contiguous spread, hematogenous spread, immunocompromise, (sickle infarct)  
splenic enterococcus abscess may seed diseased mitral valve  
staph, salmonella, e.coli, enterococcus (ICU pts), fungus  
salmonella increased in ICU pts, sickle cell disease, typhoid, immunocompromised  
poultry, turtle sources

Dx: US, CT  
percutaneous drainage 20-30% success for unilocular  
splenectomy may be necessary for multiple

## Trauma

penetrating LUQ with intraabdominal bleeding requires surgery  
blunt

FAST exam has replaced peritoneal lavage, go to CT scan if positive or suspicious for injury  
CT grading more accurate in pediatric than adult  
grade

- I < 10% of surface, < 1cm deep
- II non-expanding subcapsular hematoma 10-50% of surface, non-expanding  
intraparenchymal hematoma < 2cm, bleeding capsular tear or parenchymal  
laceration 1-3cm deep without trabecular vessel involvement
- III expanding subcapsular or intraparenchymal hematoma, bleeding subcapsular  
hematoma > 50% of surface, intraparenchymal hematoma > 2cm,  
parenchymal laceration > 3cm deep or trabecular vessel involvement
- IV ruptured intraparenchymal hematoma with active bleeding, laceration involving  
segmental or hilar vessels resulting in major (> 25% of volume)  
devascularization

V completely shattered or avulsed, hilar laceration with total devascularization  
non-op management (grade I-III)

stable patient, grade I-III, ability to do serial exams (even on vent), <2U blood loss related to spleen (v pelvic, femur fx)  
low failure rate (80% of blunt trauma, 90% success)  
incidence of missed injuries ~2%  
failure: hemodynamic decompensation, new or increased abdominal pain (other visceral injury), dropping Hct  
contrast blush on angio indicates active bleeding, poor prognosis, to OR  
24h ICU observation, 3-4d bed rest, minimal activity 1-2w, no contact sports 3 mo  
no need for CT or US f/u  
indications for splenectomy: hemodynamic instability, peritoneal signs, ongoing blood loss  
splenorrhaphy (those who are candidates with isolated splenic injury don't go to the operating room anymore)  
blood loss <500cc, minimal associated injuries, no hilar involvement, minimal-moderate splenic disruption, normal coag, no associated injuries  
suture, cautery, surgical, hemostatic glue, partial splenectomy, mesh wrap

### **Spontaneous rupture**

malaria most common cause worldwide, mono in US  
sarcoid, leukemia, delayed rupture from blunt

### **Overwhelming post-splenectomy sepsis (OPSS)**

avoid splenectomy < 4  
highest risk 1<sup>st</sup> 2y post splenectomy (60% of adult cases, 80% of child)  
the earlier the infection, the higher the mortality  
risk varies with indication for splenectomy  
accessory spleen not enough to confer immunity  
greatest risk children, less common in adults  
hematologic disease higher risk than trauma  
highest risk thalassemia, lymphoma, Hodgkins, don't do as well  
strep pneumonia (70%), h. flu, neisseria (encapsulated BT)  
pneumococcal vaccine (covers 73% of strains, 40% of strains penicillin resistant), h. flu, n. meningitis vaccines  
give early, 2w before elective, otherwise prior to discharge, revaccinate @5y  
prophylactic antibiotics 6mo-1y, penn, amoxicillin, erythromycin  
child with febrile illness after splenectomy take to ER