

## Case Management Session: Disorders of the Spleen

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### Anatomy

- Develops from dorsal mesogastrium
- Present by 6<sup>th</sup> week gestation
- LUQ of abdomen
- Diaphragm superiorly, lower thoracic cage anteriorly
- Associated with : pancreas, stomach, left kidney, colon, diaphragm

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### Anatomy

- Suspensory ligaments
  - Splenorenal
  - Gastrosplenic
  - Splenocolic
  - Splenophrenic
- Blood Supply
  - Splenic artery
  - Splenic vein
  - Short gastric arteries

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## Anatomy

- Weight 75-150 gm
- Size patient's fist
- Receives 5% cardiac output (350 l/day)
- Accessory spleens in 10-30%
  - Splenic hilum
  - Splenocolic ligament
  - Gastrocolic ligament
  - Splenorenal ligament
  - omentum

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## Physiology

- Functions
  - Fetal Hematopoiesis: usually ceases by birth
  - Filtration of blood
  - Immune modulation: production of opsonins and clearance of opsonized particles to battle encapsulated organisms

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## Case # 1

- 13 year old female with complaints of fatigue, and vague, intermittent abdominal pain.

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### Case # 1

- What other questions would you like to ask?

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### Case # 1

- Pain is in upper abdomen, not associated with eating
- No history of bleeding/bruising
- No nausea/vomiting
- FH-father none, mother was adopted
- PMH
  - normal growth/development
  - Menarche 12 ½ years

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### Case # 1

- What are you looking for on physical exam?

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### Case # 1

- Scleral icterus
- Yellow nail beds
- 2/6 systolic ejection murmur
- Mass in LUQ

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### Case # 1

- What is your differential diagnosis?

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### Case # 1

- Labs
  - Hgb 8.2, spherocytes on smear, positive osmotic fragility test
- Radiographic Studies
  - US/CT show enlarged spleen

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## Case # 1

- Diagnosis
  - Hereditary spherocytosis
- Ddx
  - Elyptocytosis
  - G6PD deficiency
  - Sickle cell anemia with hypersplenism

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## Case #1

- Plan of Treatment
- Vaccination for *S. pneumoniae*, *N. meningitidis*, *H. influenzae*
- Splenectomy
  - Laparoscopic
  - open

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## Case # 1

- For what other hematologic disorders might splenectomy be indicated?

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### Case # 1

- Hereditary spherocytosis
- Sickle cell anemia
- Idiopathic thrombocytopenic purpura
- Thalassemia
- Leukemia/Lymphoma
- Gaucher's Disease
- Hypersplenism

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### Case # 1

- Sickle Cell Anemia
  - Substitution in beta chain of Hgb A resulting in Hgb S
  - RBC's become rigid with decrease in O2 saturation causing occlusion of capillaries
  - Eventually leads to autoinfarction of spleen
  - Can lead to sequestration crisis requiring splenectomy

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### Case # 1

- Idiopathic Thrombocytopenic Purpura
  - Anti-platelet antibodies (IgG) bind with platelets leading to destruction of RES
  - Treatment
    - corticosteroids,
    - IVIG
    - splenectomy
  - Childhood ITP usually self-limited and acute
  - Splenectomy only indicated for chronic cases

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### Case # 1

- **Thalassemia**
  - Abnormal production of alpha or beta chains of Hgb
  - Most severe form Thalassemia major
  - Splenic enlargement and sequestration
  - Splenectomy decreases need for transfusion

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### Case # 1

- **Gaucher's Disease**
  - deficiency of B-glucocerebrosidase
  - Excessive glucocerebroside in macrophages
  - Severe splenomegaly and hypersplenism
  - Recurrence high after partial splenectomy

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### Case # 1

- **Hypersplenism**
  - Decreased platelets
  - Decreased Hgb
  - Decreased WBC
  - Enlarged spleen
  - Primary or secondary

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### Case # 1

- What are the postoperative complications of splenectomy?

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### Case # 1

- Bleeding
- Gastric paresis
- Overwhelming post-splenectomy sepsis (OPSI)
  - Decreased clearance of encapsulated bacteria
  - Increased 60-100 fold age < 5 years
  - Incidence 0.13%-8.1% age < 15 years
  - 0.28-1.9% adults

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### Case # 1

- Overwhelming post-splenectomy sepsis
  - Mortality 1.8% overall
  - 60% fatal infections and 50% all infections due to *S. pneumoniae*
  - 32% mortality due to *H. influenzae*
  - Fatal OPSI
    - 3.77% children
    - 0.39% adults

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### Case # 1

- Rate of infection related to age at splenectomy
  - 13.8% age < 5years
  - 0.5% age > 5 years
- Post-splenectomy Immunizations
  - S. pneumo
  - H. flu
  - N. men
- Immunize 2-3 weeks prior to splenectomy

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### Case # 1

- Prophylactic antibiotics
  - Recommendations unclear
  - Highest rate OPSI in first 2 years after splenectomy
  - Lifelong PCN?
  - PCN for first 10 years?

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### Case # 2

- 24 year old male on motorcycle hit cement median on expressway. He had helmet in place. He was found awake but combative on scene. He is brought to ER on backboard and in c-collar.

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## Case # 2

- What do you want to know?

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## Case # 2

- AMPLE History
  - Allergies
  - Medications
  - Past medical history
  - Last meal
  - Events

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## Case # 2

- What are you going to do and in what order?

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## Case # 2

- Airway
  - Breathing
  - Circulation
  - Disability
  - Exposure
- Airway patent, bilateral breath sounds, R 28, BP 120/85, heart rate 130/regular, GCS 13, moving RUE, LUE, RLE, temp 37 rectal

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## Case # 2

- Secondary Survey
  - Tenderness LUQ and costal margin, no distention
  - Deformity left thigh
  - Unstable pelvis

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## Case # 2

- What do you think has been injured?

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### Case # 2

- Ribs
- Spleen
- Pelvis
- Femur
- Possibly lung, head, neck

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### Case # 2

- What xrays do you want to get?

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### Case # 2

- CXR
- Lateral c-spine
- Pelvis
- Left femur, hip, knee
- FAST
- CT abdomen and pelvis
- CT head

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### Case # 2

- CXR –fracture ribs 9 and 10 on left
- Cpsine-negative
- Pelvis-fracture both pubic rami on left
- Femur-fracture of femoral neck left
- FAST- fluid in LUQ and pelvis
- CT head-negative
- CT abdomen/pelvis-grade 3 spleen laceration, free fluid in peritoneal cavity, left pubic rami fracture

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### Case # 2

- What are your management options?

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### Case # 2

- Operative management of spleen
- Non-operative management of spleen
- Orthopedics consult

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## Case # 2

- Operative Management
  - Laparotomy or laparoscopy
  - Total splenectomy
  - Partial splenectomy
  - Splenorhaphy

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## Case # 2

- Non-operative management
  - Bedrest
  - Hemodynamic monitoring
  - Serial physical exams
  - Serial Hgb
  - Possible role for angiography

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## Case # 2

- Must be hemodynamically normal and stable
- No suspicion for bowel injury
- If need for transfusion 2 units PRBC's then risk of splenectomy less than non-operative

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