# Case Presentation

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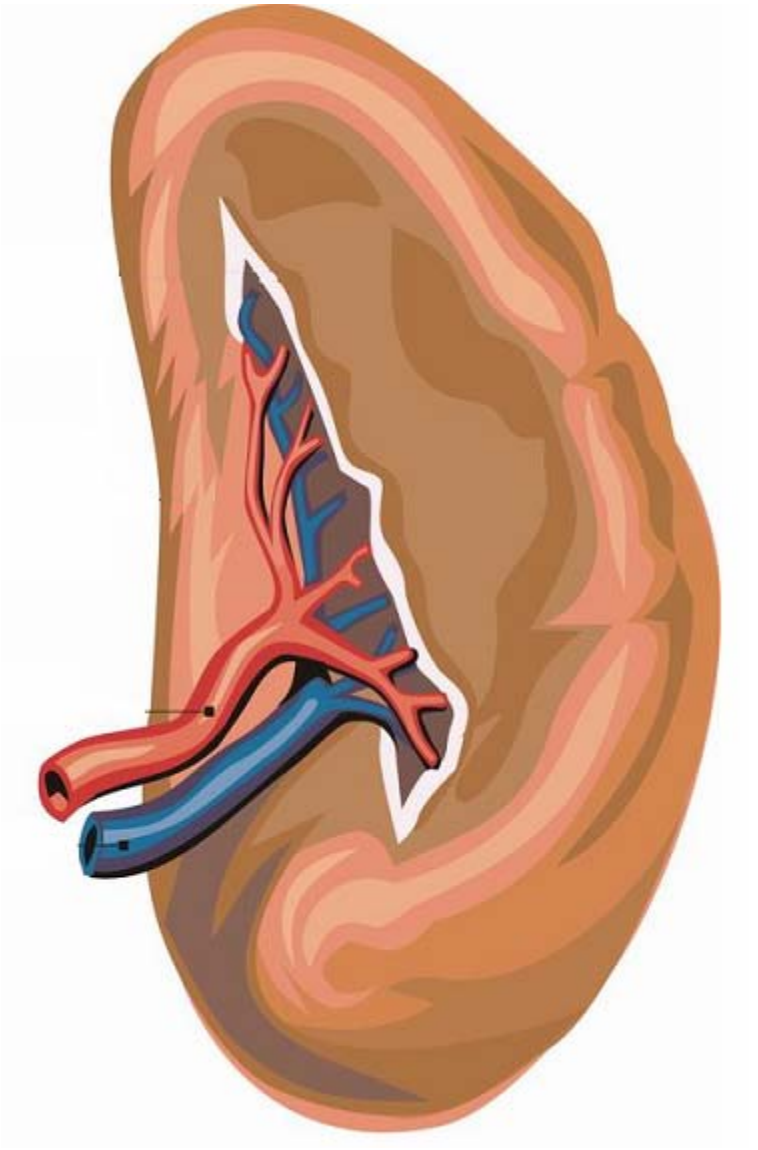
# Medical Indications for Splenectomy

# Definition

- A large, highly vascular lymphoid organ, lying in the human body to the left of the stomach below the diaphragm, serving to store blood, disintegrate old blood cells, and filter foreign substances from the blood



# History

- Erasistratus of Chios (304 - 250 BC)
  - Spleen maintains the symmetry of the abdomen
- Galen of Pergamum (130 - 200 AD)
  - “Splenum mysterii organon”
  - Source of black bile

# History

- 1777: William Hewson recognizes associations with the lymphatic system
- 1846: Rudolph Virchow demonstrates that the follicles in the spleen are related to the white blood cells
- 1885: Ponfick recognized the ability of the spleen to remove particles from the blood
- 1926: O'Donnell reports a case of "acute septicemia" in a 6-year-old boy 2 years after splenectomy

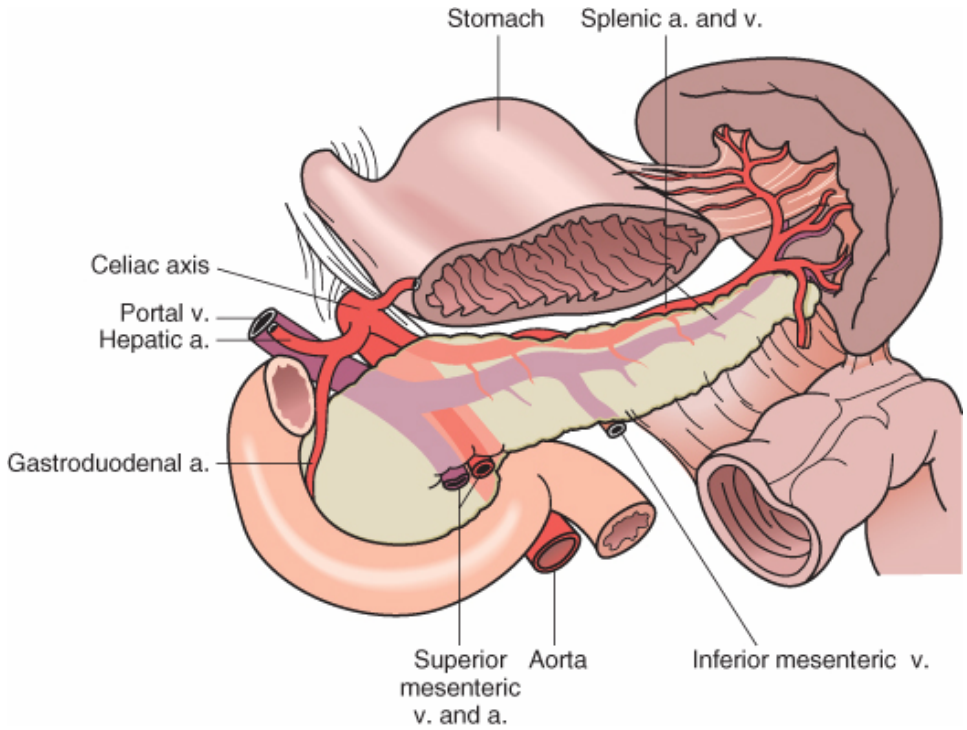
# History

- 1887: Sir Spencer Wells performs one of the first successful splenectomies[1]
  - Operated on a 24-year-old anemic female expecting to find a uterine fibroid
  - Instead found a “wandering spleen”
  - The patient had hereditary spherocytosis
- 1911: Ferdinando Micheli introduces the concept of splenectomy for hemolytic anemia[2]

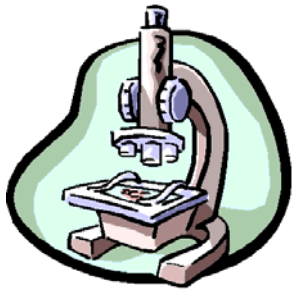
[1] Wells, T.S.: Remarks on splenectomy with a report of a successful case. Med. Chir. Trans. 71:255, 1888
[2] Micheli, F.: Effetti immediati della splenectomia in un caso di ittero emolitico splenomegalico acquisito tipo Hayem-Widal (ittero splenoemolitico). Clin. Med. Ital. 50:453, 1911

# Anatomy

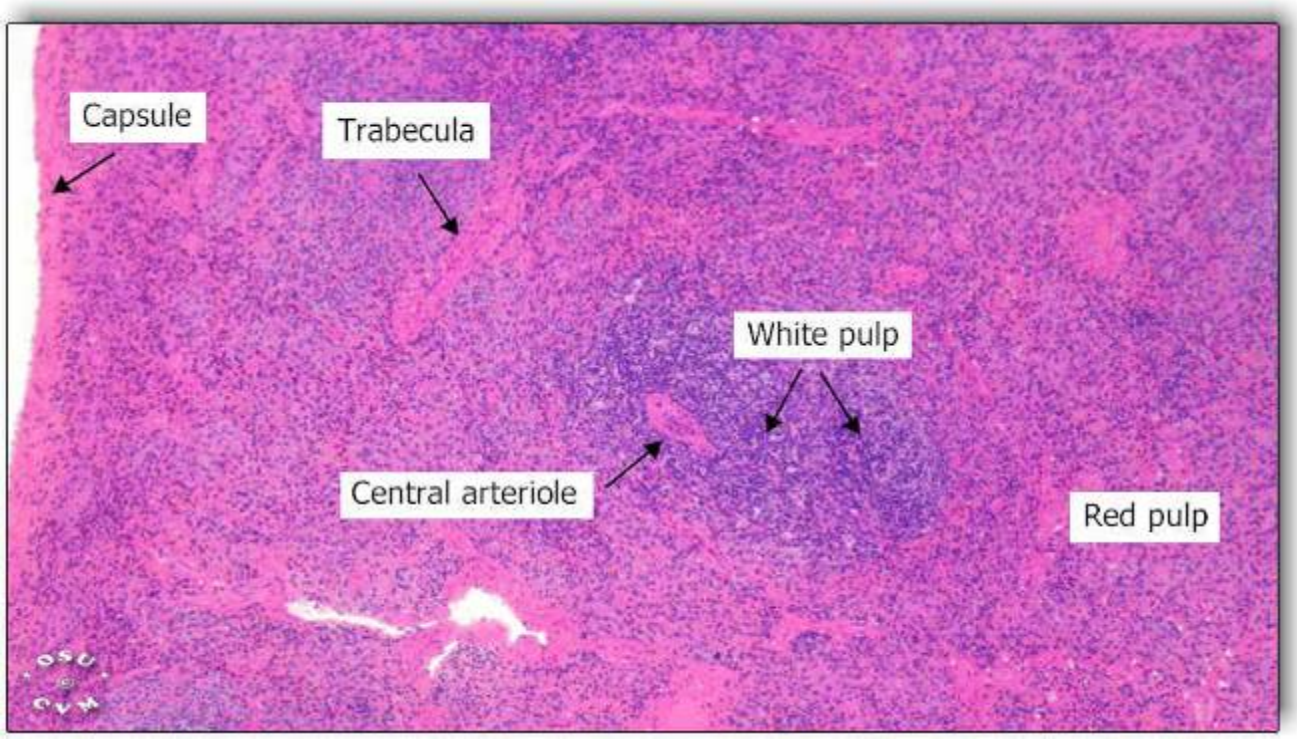
- 12 to 15 cm in length, 4 to 8 cm in width and 3 to 4 cm in thickness
- Average weight is about 140 to 180 g
- lies in the shelter of the 9th to 11th rib at the left side of the abdominal cavity



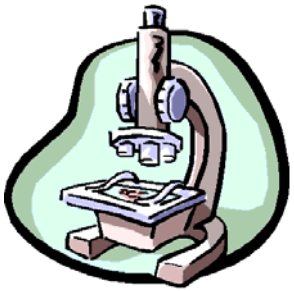
# Histology



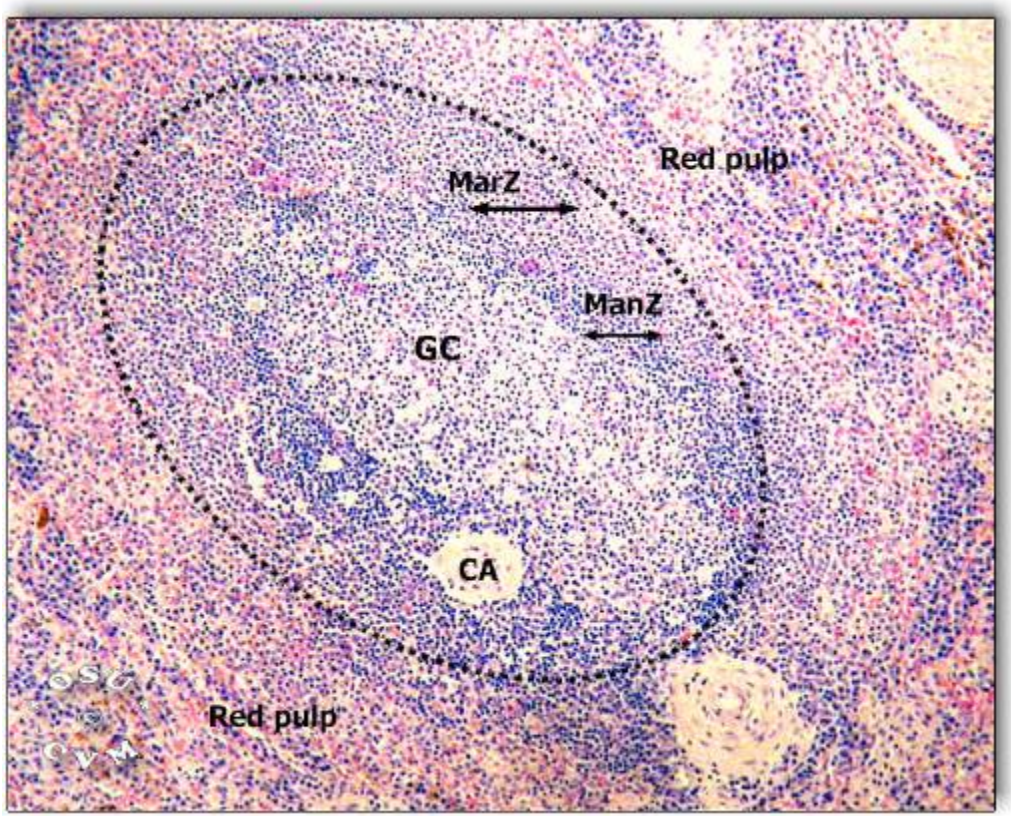
- Two general components:
  - White pulp (5-20%)
  - Red pulp (~85%)
  - Enclosed by capsule and interspersed with trabeculae



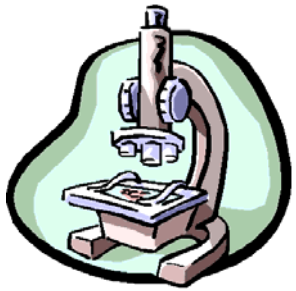
# Histology



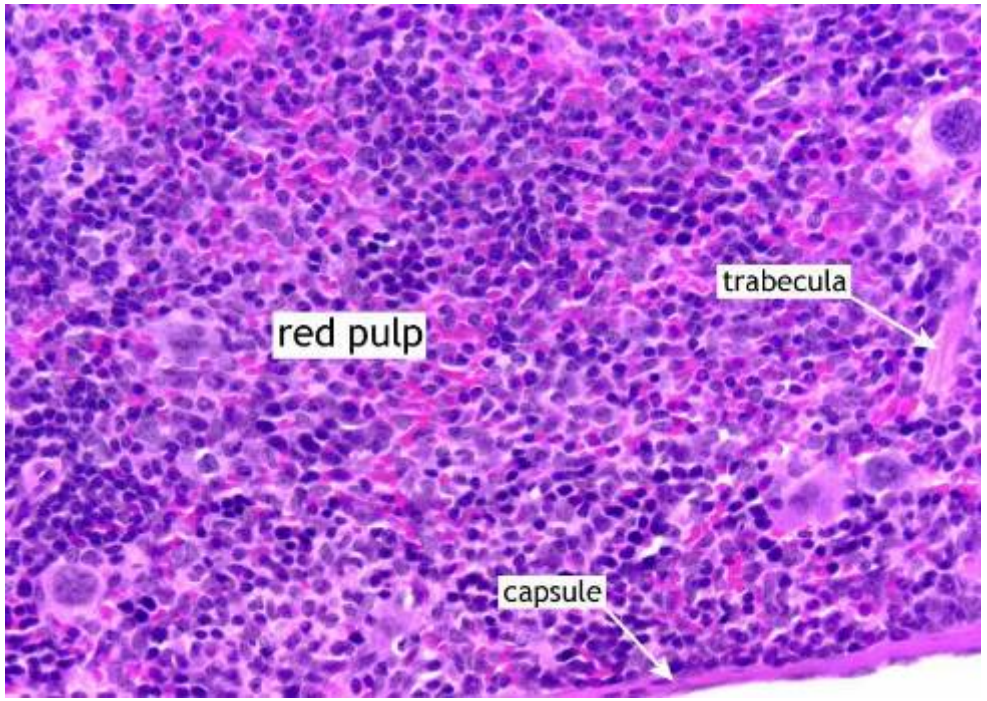
- White pulp – three major compartments
  - PALS
  - Lymphoid follicle
  - Marginal zone



# Histology

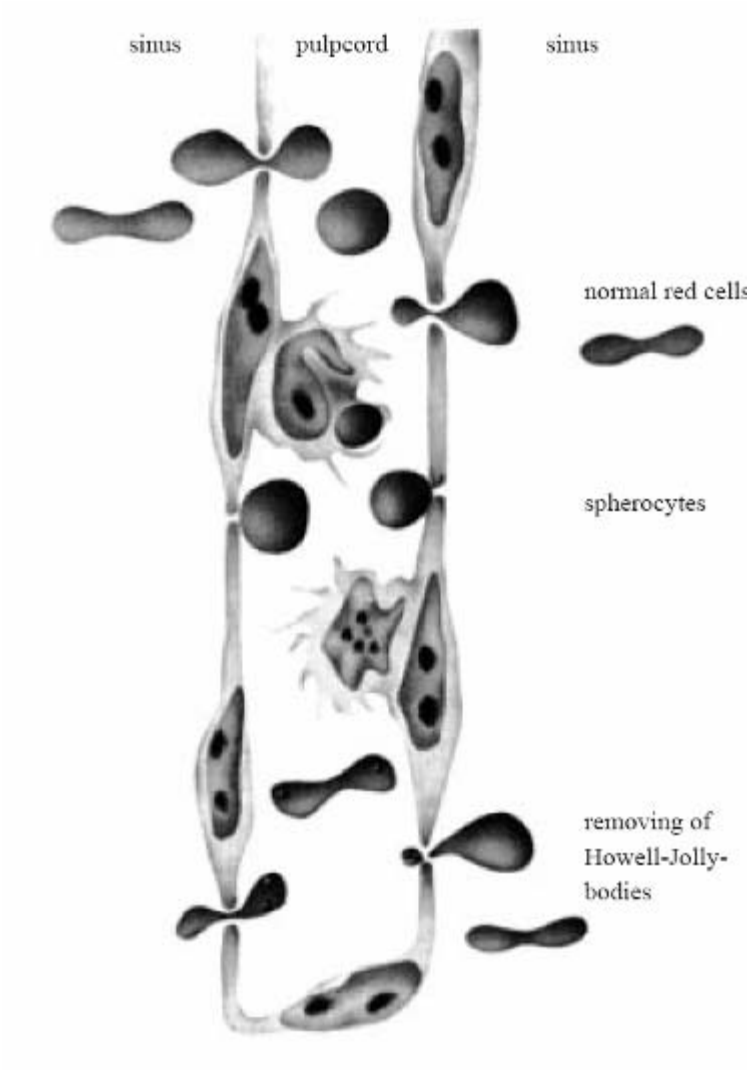


- Red pulp
  - Consists of a loose reticular tissue rich in capillaries and venous sinusoids
  - Sinusoids have an unique endothelium of longitudinally arranged cells – central to filtration function



# Physiology

- Filtration
  - Stiff or fragile RBCs cannot pass through interendothelial slits
- Immune function
  - Splenic phagocytes, together withmacrophages in the liver, synthesize the majority of components of the classical pathway of complement
  - Slow blood flow in the red pulp cords allows foreign particles to be phagocytosed without specific ligand-receptor interactions
  - Important in fighting early bacterial infection



# Physiology: Functions of the spleen

| White pulp | Red pulp |
| --- | --- |
| - Antibody synthesis<br>- Initiation of humoral response<br>- Reservoir of lymphocytes | - Filter function<br>- Phagocytosis (especially badly opsonized particles)<br>- Reservoir of thrombocytes and immature erythrocytes<br>- Haematopoiesis (fetal life)<br>- Tuftsin production<br>- Role in alternative complement pathway<br>- Positive effect on factor VIII<br>- Reutilization of iron<br>-Inhibition of angiotensin converting enzyme |

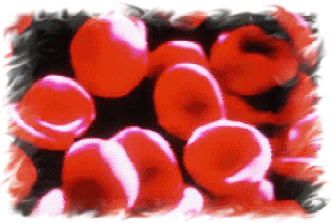
Lockwood CM. Immunological functions of the spleen. Clinical Haematology 1983; 12:449-465

# Nontraumatic indications for splenectomy

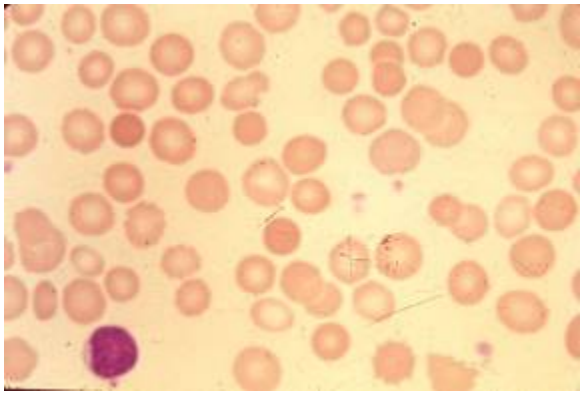
Table 1. Splenectomy for hematologic disorders: 1958–1995 (727 cases)

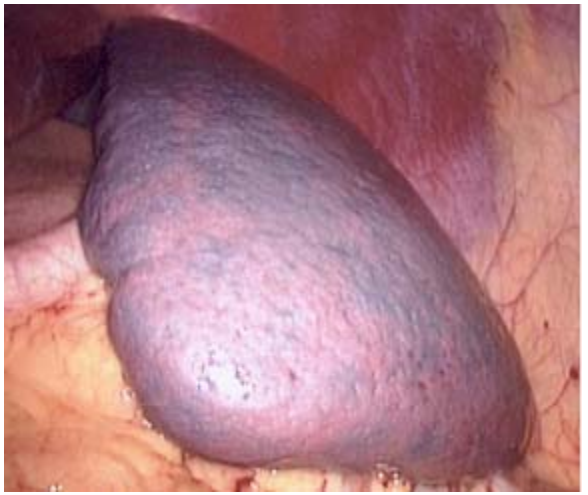
| Disorder | No. of splenectomies |
|---|---|
| Idiopathic thrombocytopenic purpura | 218 |
| Hodgkin's staging | 212 |
| Hereditary spherocytosis | 75 |
| Chronic lymphocytic leukemia and histiocytic lymphoma | 59 |
| Myeloproliferative disorders | 49 |
| Thrombotic thrombocytopenic purpura | 25 |
| Acquired hemolytic anemia | 20 |
| Thalassemia | 16 |
| Primary hypersplenism | 12 |
| Felty syndrome | 9 |
| Lupus erythematosus | 9 |
| Sarcoid | 7 |
| Evans syndrome | 5 |
| Gaucher's disease | 5 |
| Sickle cell | 2 |
| Kasabach-Merritt syndrome | 1 |
| Spontaneous rupture | 3 |

# Splenectomy for Hemolytic Diseases



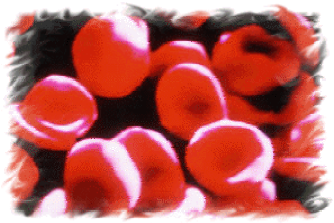
- Hereditary spherocytosis
  - Autosomal dominant trait
  - Most common hemolytic anemia for which splenectomy is advised
  - Defective erythrocyte membrane causes trapping and disintegration within the spleen
  - Presents with anemia, reticulocytosis, jaundice, and splenomegaly
  - Diagnosis made by peripheral blood smear
  - Splenectomy is the only therapeutic modality (wait until age 4 to 6)
  - Outcomes are excellent



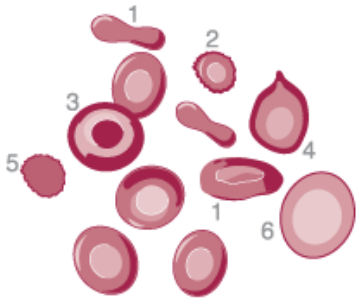


Schwartz, S.I.: Splenectomy for hematologic disease. Surg. Clin. North Am. 61:117, 1981

# Splenectomy for Hemolytic Diseases



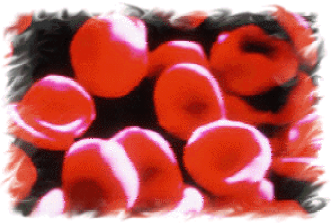
- Thalassemia
  - Autosomal dominant transmission
  - Defect in the synthesis rate of hemoglobin
  - Thalassemia major (homozygous) presents with pallor, retarded body growth, enlarged head, and intractable ulcers
  - Diagnosis made by nucleated RBCs (target cells) in smear
  - Splenectomy reserved for patients with markedly symptomatic splenomegaly, painful splenic infarction, and increased transfusion requirements
  - Greater risk of post-splenectomy sepsis



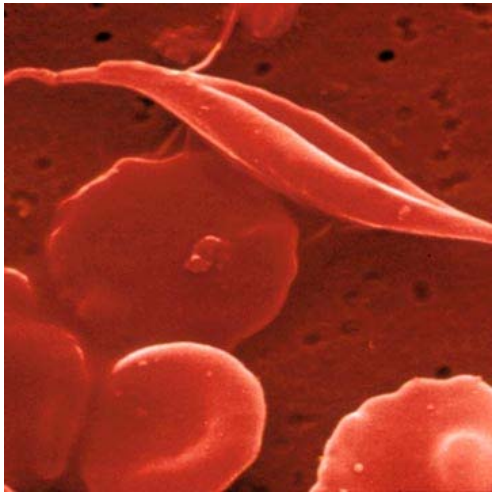

Poikilocytic red cells (elliptocytes[1], schistocytes[2], target cells[3], tear drop[4], spherocytes[5] & hypochromic[6]) usually present in Thalassemia Major.

Pinna AD, Argiolu F, Marongiu L, Pinna DC. Indications and results for splenectomy for beta thalassemia in two hundred and twenty-one pediatric patients. Surg Gynecol Obstet 167:109, 1988

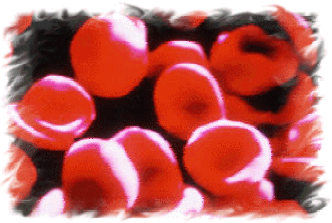
# Splenectomy for Hemolytic Diseases



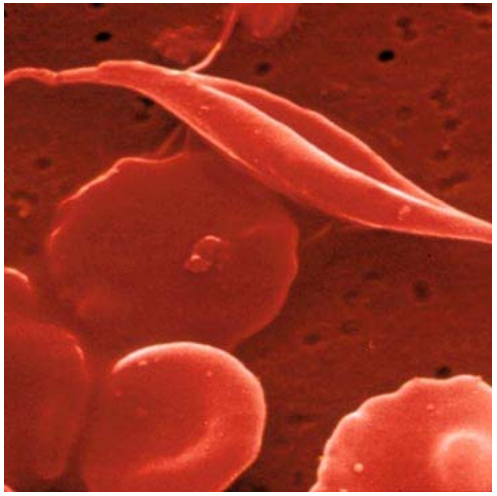
- Sickle Cell Disease
  - Due to homozygous inheritance of HbS
  - Single amino-acid substitution of valine for glutamic acid
  - Spleen commonly enlarged during the first decade of life but then undergoes progressive atrophy due to repeated attacks of vaso-occlusion and infarction
  - In general, splenectomy should be avoided in patients with SCD
    - Already immunocompromized



# Splenectomy for Hemolytic Diseases



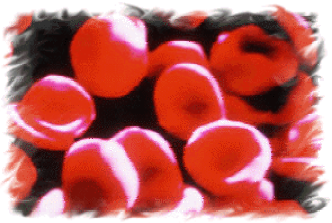
- Sickle Cell Disease
  - Main indications are:
    - Acute splenic sequestration crisis[1]
      - Circulatory collapse and sudden death from the rapid sequestration of red blood cells
      - Second-leading cause of pediatric deaths in patients with SCD
    - Hypersplenism
      - Splenectomy decreases transfusion requirements
      - Partial splenectomy may be an option[2]
    - Splenic abscess
      - Increased incidence of *Salmonella*




[1] Powell RW, Levine GL, Yang Y, Mankad VN. Acute splenic sequestration crisis in sickle cell disease: early detection and treatment. J Pediatr Surg 1992;27:215-9

[2] Witte CL, Van Wyck DB, Mitte MH, et al. Ischaemia and partial resection for control of splenic hyperfunction. Br J Surg 1982;69:531-5

# Splenectomy for Hemolytic Diseases



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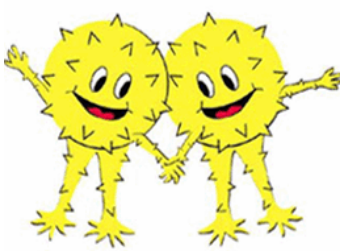
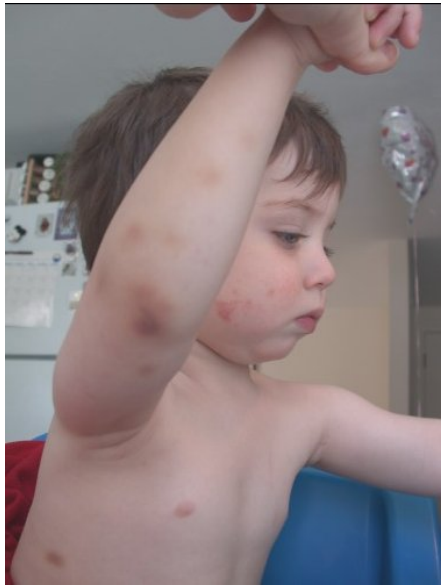
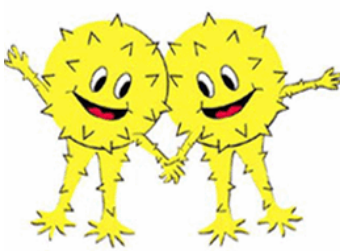
# Splenectomy for Platelet Disorders



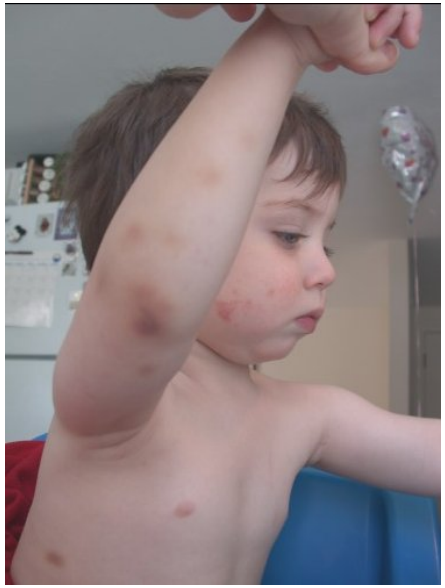
- Idiopathic thrombocytopenic purpura
  - Most common hematologic indication for splenectomy
  - Spleen is the source of circulating antiplatelet IgG
  - Also responsible for sequestration of sensitized platelets
  - Diagnosis made by thrombocytopenia with normal bone marrow
  - Usually an isolated phenomenon



# Splenectomy for Platelet Disorders

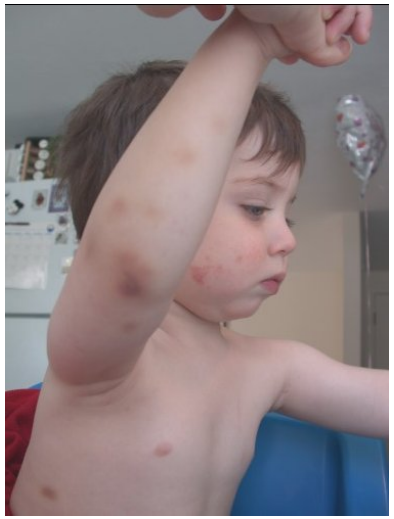


- Idiopathic thrombocytopenic purpura
  - Patients present with ecchymosis and purpura and at times there is excessive bleeding from the gums, vaginal bleeding, gastrointestinal bleeding, and hematuria
  - Platelet count characteristically less than 50,000/$mm^3$



# Splenectomy for Platelet Disorders

- Idiopathic thrombocytopenic purpura
  - First line of treatment medical
    - Steroids
    - IgG
    - Plasmapheresis
  - Medical treatment only curative in 15% of adults
  - Cure rates up to 87% reported with splenectomy[1]
  - Given risks of splenectomy, should be reserved for patients with platelet counts <25,000/mm$^3$ and bleeding



Outcome of patients with ITP treated by splenectomy or medical therapy

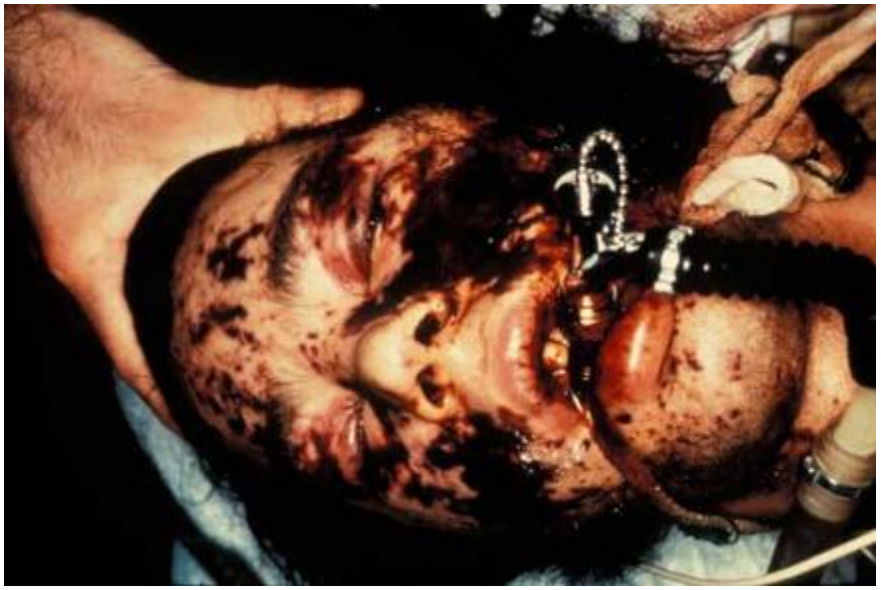
| | CR | PR | Failure | Lowest PC after therapy | Highest PC after therapy |
|---|---|---|---|---|---|
| Splenectomy | 33 (87%)* | 2 (5%)* | 3 (8%)* | 180 (150–200)* | 340 (270–425)* |
| Medical treatment | 15 (17%) | 12 (13%) | 64 (70%) | 80 (50–140) | 170 (100–230) |

The lowest and highest platelet count observed during the follow-up period is shown. Values expressed as medians (interquartile range).
* $P < 0.05$ versus medical treatment.
CR = complete response; PR = partial response; PC = platelet count (g/L).

[1] Goldenstatter M, Lamprecht B, Klingler A, et al. Splenectomy versus medical treatment for idiopathic thrombocytopenic purpura. Am J Surg 2002:184;606-610

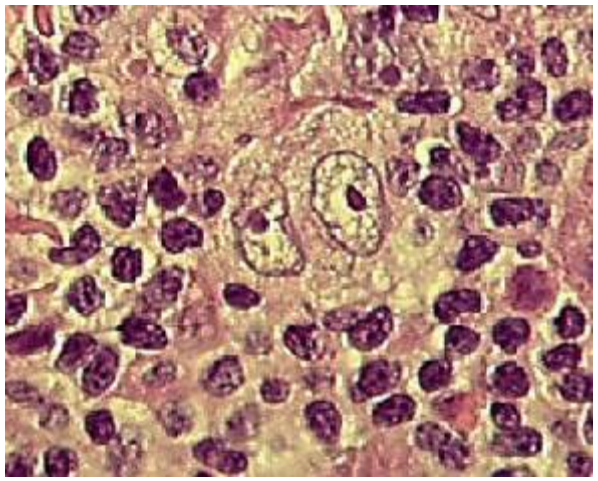
# Splenectomy for Platelet Disorders

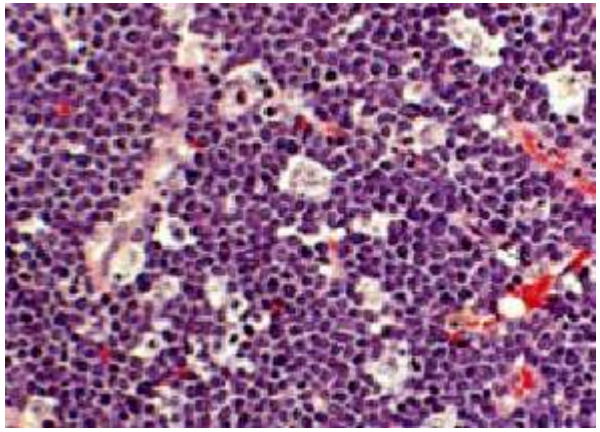
- Thrombotic thrombocytopenic purpura
  - Increase of subendothelial collagen in the arterioles and capillaries causing diffuse platelet trapping
  - Manifested by thrombocytopenia, hemolytic anemia, fever, neurologic manifestations, and renal disease
  - Primarily treated with high volume plasmapheresis (80 to 90% survival)
  - Splenectomy reserved for non-responders



# Splenectomy for Hematologic Malignancy

- Hodgkin's Disease
  - Historically, staging laparotomy with splenectomy were essential guides to treatment
  - Improvement in imaging and chemotherapy have minimized the role of the surgeon
- Splenectomy for nonhodgkin's lymphomas
  - Symptomatic splenomegaly
  - NHL confined to the spleen or with prominent splenic involvement (survival improvement from 24 to 108 months[1])
- Leukemias
  - Splenectomy indicated for symtpomatic splenomegaly

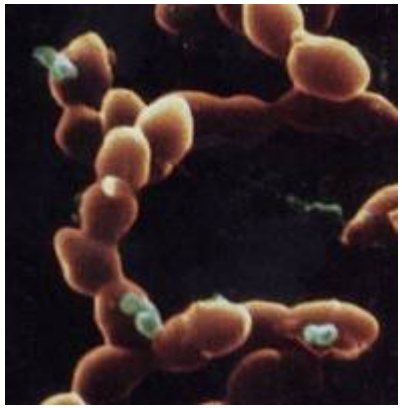




[1] Morel P, Dupriez B, Gosselin B, et al: Role of early splenectomy in malignant lymphomas with prominent splenic involvement (primary lymphomas of the spleen): A study of 59 cases. Cancer 71:207-215, 1993

# Consequences of Splenectomy

- Overwhelming post-splenectomy infection
  - 1952: King and Schumacker report fatal bacterial sepsis in five infants following splenectomy for hereditary spherocytosis[1]
  - Asplenic patients have a five-fold increased risk for fatal sepsis[2]
  - Risk varies with patient age and indication for splenectomy
  - May occur at any time
  - *Streptococcus pneumoniae* implicated in 50 to 90%; also *Haemophilus influenzae*, *Neisseria meningitidis*
  - Prophylactic immunization 2 weeks before elective splenectomy; otherwise immunization prior to discharge

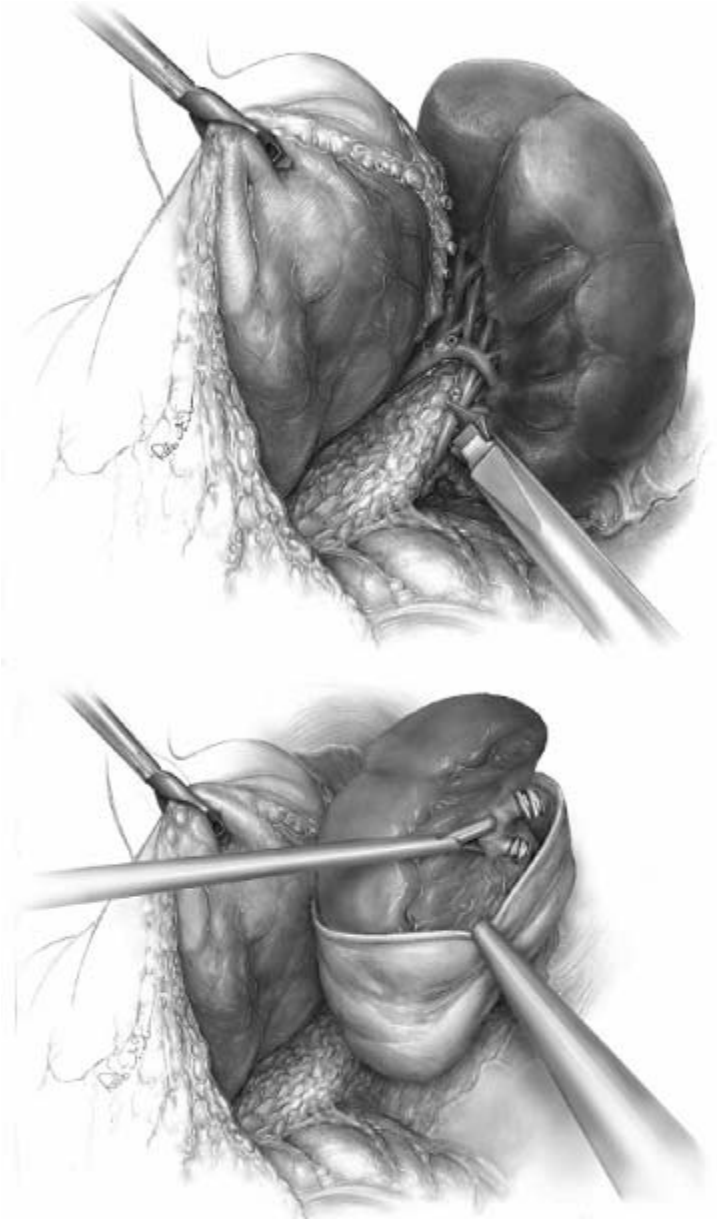


[1] King H, Schumacker HB. Splenic studies: susceptibility to infection after splenectomy performed in infancy. Ann Surg 1952;136:239-42

[2] Horowitz J, Smith JL, Weber TK, et al: Postoperative complications after splenectomy for hematologic malignancies. Ann Surg 223:290-296, 1996

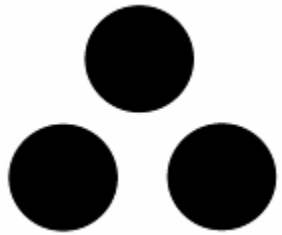
# Laparoscopic Splenectomy

- Benefits of laparoscopic splenectomy similar to those of other laparoscopic procedures
  - Decreased pain
  - Quicker return of bowel function
  - Shorter hospital stay
  - Improved cosmetic result
- Low conversion rate
- Morbidity similar to open
- Accessory spleens may be easier to miss



Minkes RK, Lagzdins M, Langer JC. Laparoscopic versus open splenectomy in children. J Pediatr Surg 2000;35:699-701

# Conclusions



- The spleen may become problematic during the course of nonsurgical illnesses by virtue of either its undesirable functionality or its physical obtrusiveness
- Asplenia is disadvantageous in its own right
- The surgeon must carefully weigh the risks and benefits of splenectomy on a case by case basis


Special thanks to Dr. Manuel Molina for making this presentation possible