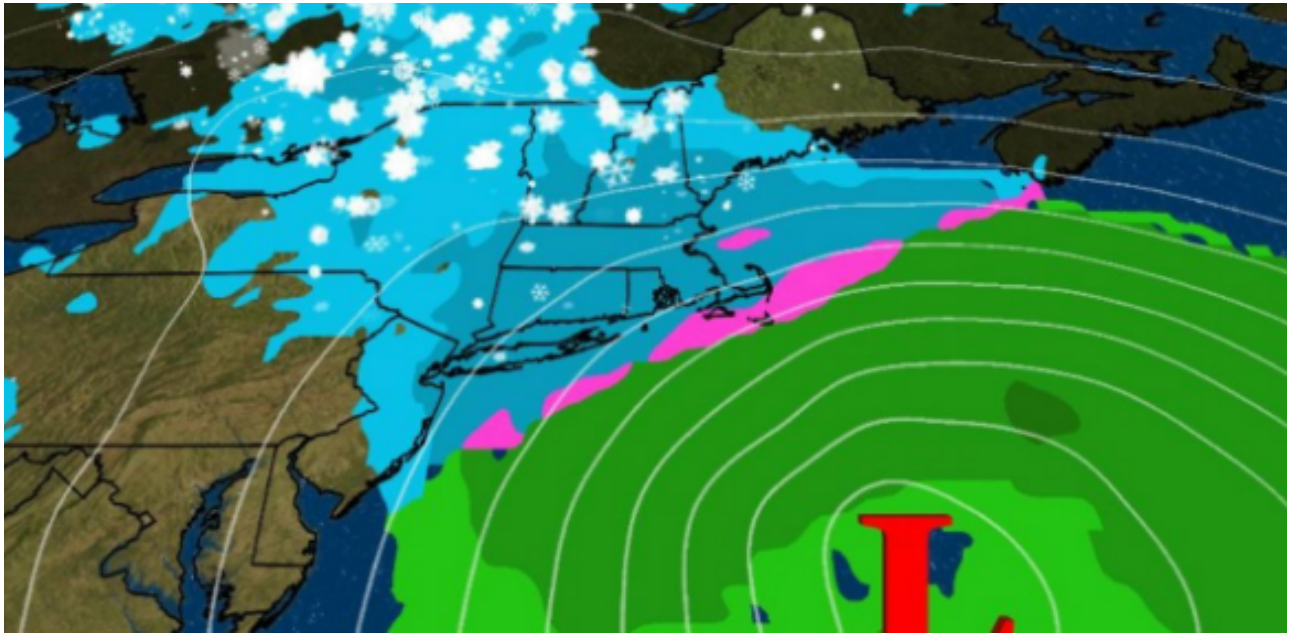
Heme Questions and Derivatives for the USMLE Step One Exam

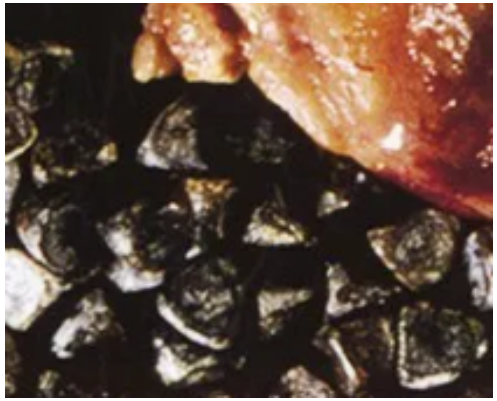


Winter Storm Skylar Edition

Howard J. Sachs, MD
Howard@12DaysinMarch.com
www.12DaysinMarch.com

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder. He is taken to OR for cholecystectomy. Specimen shown.

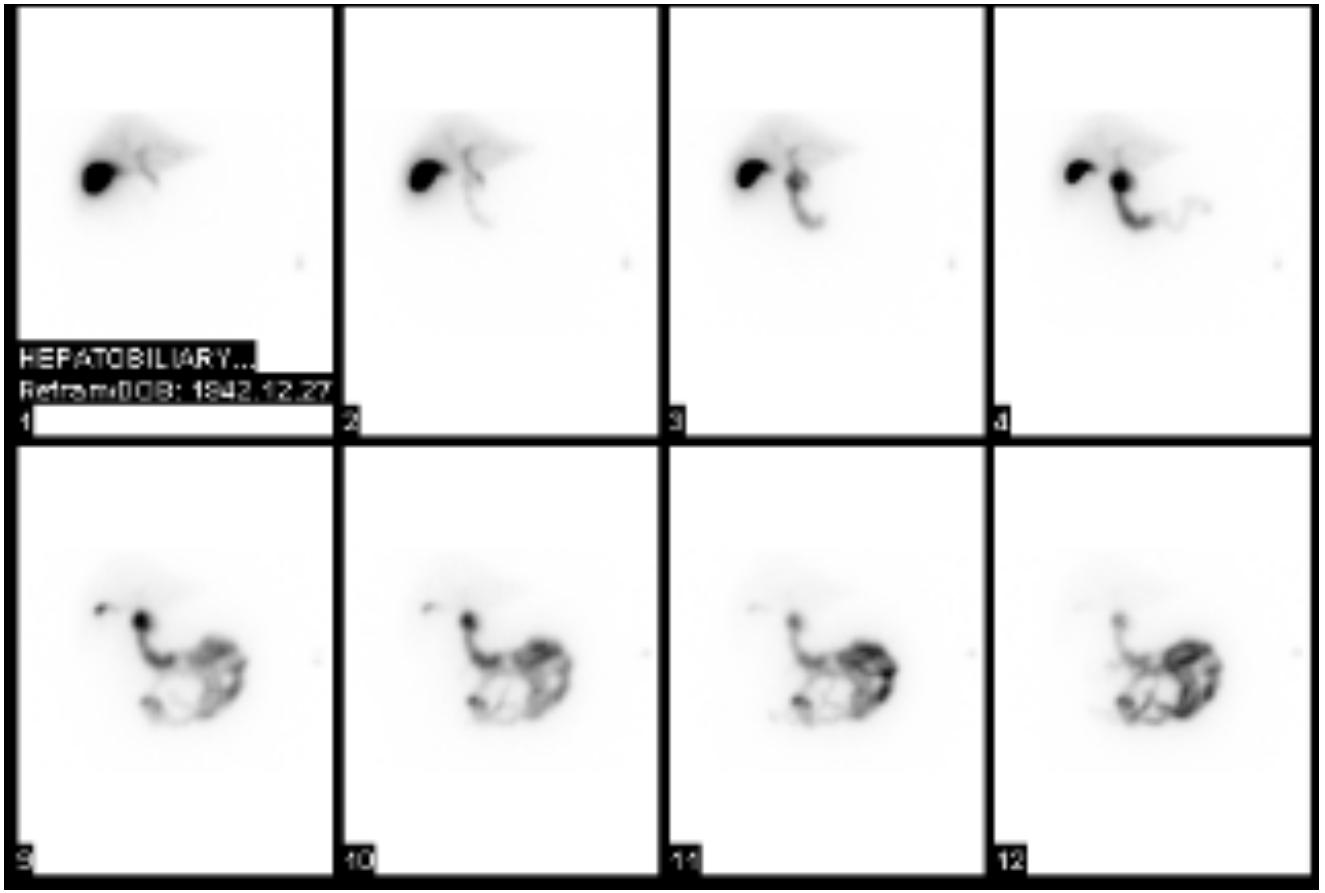
Which of the following patterns would be most suggestive of the predisposing condition?



	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
1	35	76	No	32	8
2	35	76	Yes	38	24
3	35	105	Yes	36	24
4	35	105	No	36	24
5	55	76	Yes	32	8

MCHC, normal 33-36 g/dL

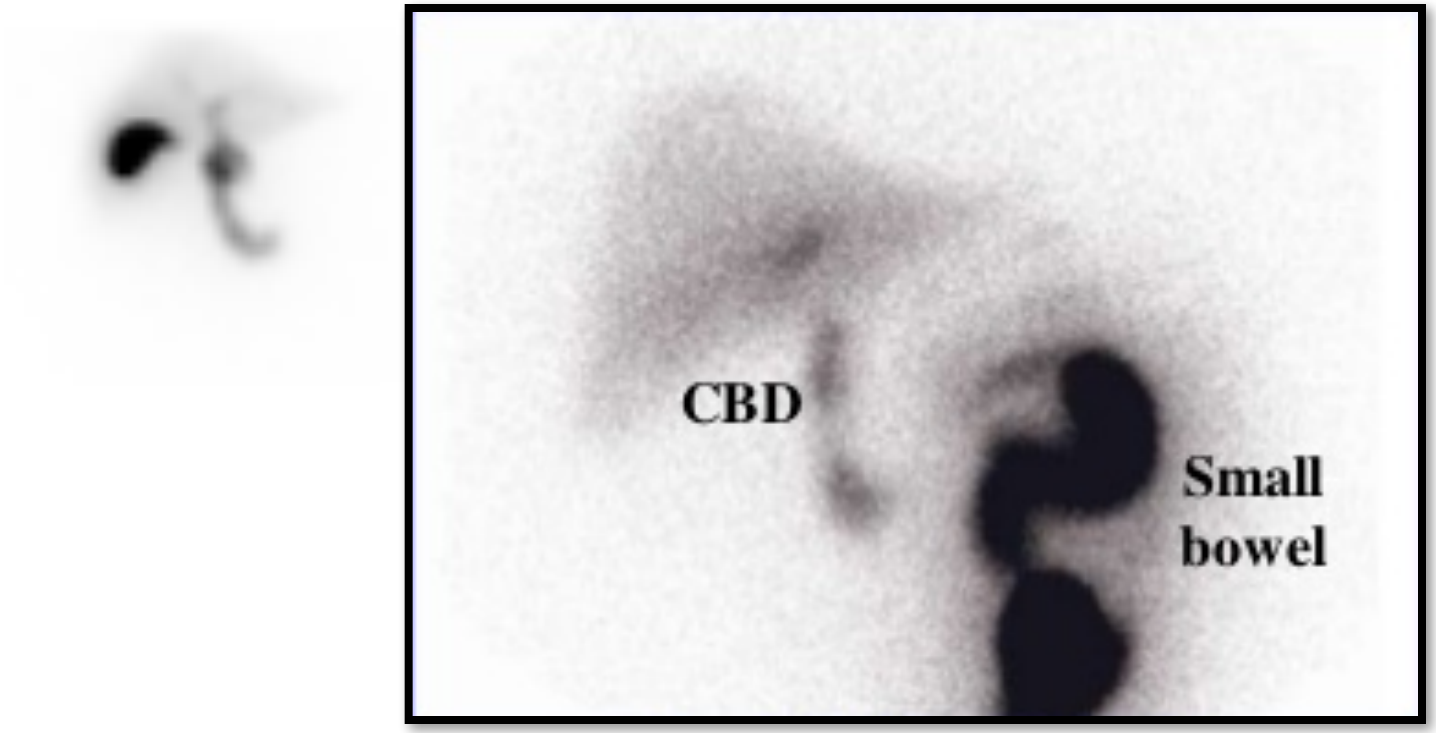
Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder.



HIDA: Hepatobiliary Iminodiacetic Acid

Patient presents with RUQ pain. ~~HIDA~~ scan fails to visualize gallbladder.

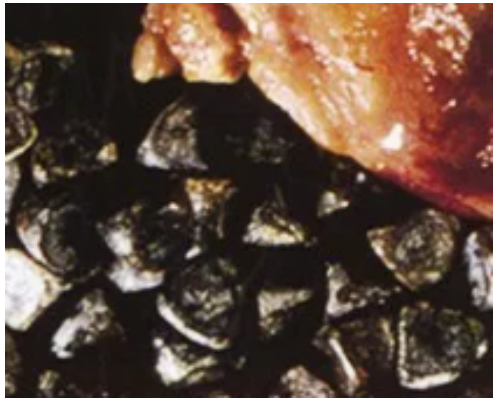
Radionuclide biliary scan



Gallbladder not visualized.
Cystic duct is obstructed.
Acute Cholecystitis

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder.
He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?

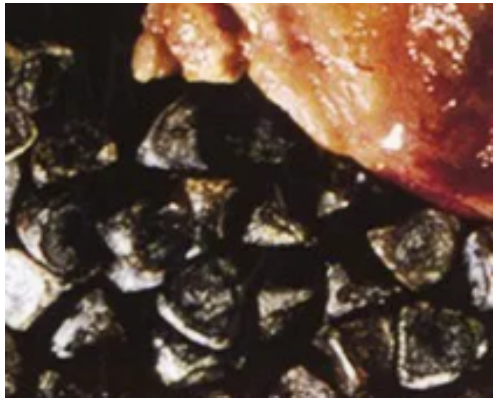


Pigment stones is the language of Hemolysis

Pigment (calcium bilirubinate) = Increased bilirubin in the bile

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder. He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?

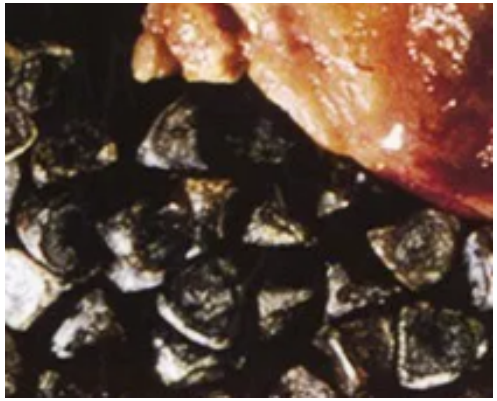


	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
1	35	76	No	32	8
2	35	76	Yes	38	24
3	35	105	Yes	36	24
4	35	105	No	36	24
✖ 5	55	76	Yes	32	8

MCHC, normal 33-36 g/dL

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder. He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?

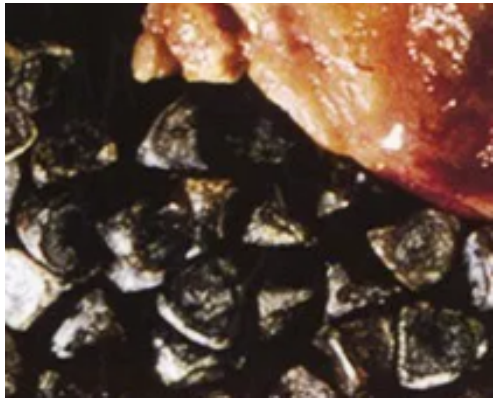


	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
1	35	76	No	32	8
2	35	76	Yes	38	24
3	35	105	Yes	36	24
4	35	105	No	36	24
5	55	76	Yes	32	8

MCHC, normal 33-36 g/dL

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder. He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?



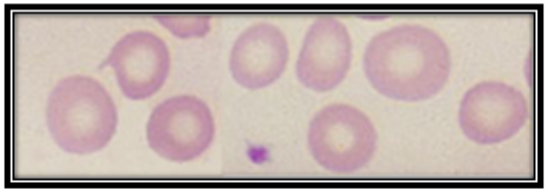
		HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
✗	1	35	76	No	32	8
	2	35	76	Yes	38	24
✗	3	35	105	Yes	36	24
✗	4	35	105	No	36	24
✗	5	55	76	Yes	32	8

MCHC, normal 33-36 g/dL

Microcytic Anemia: Iron Deficiency (IDA) and ACD*

IDA: iron deficiency anemia

*ACD: anemia of chronic disease



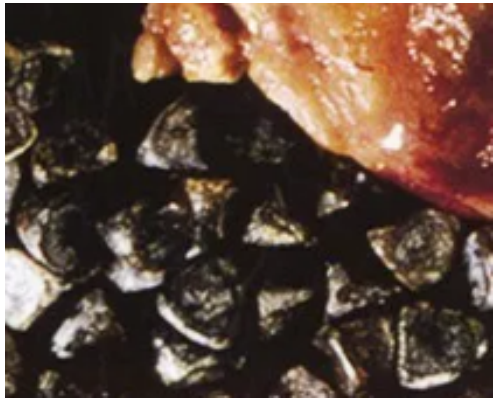
Howard J. Sachs, MD
www.12daysinmarch.com

	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
1	35	76	No	32	8
2	35	76	Yes	38	24
3	35	105	Yes	36	24
4	35	105	No	36	24
5	55	76	Yes	32	8

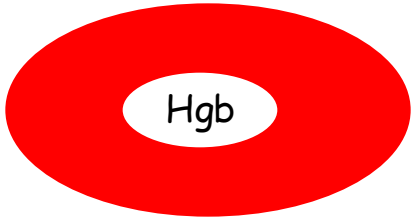
MCHC, normal 33-36 g/dL

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder.
He is taken to OR for cholecystectomy. Specimen shown.

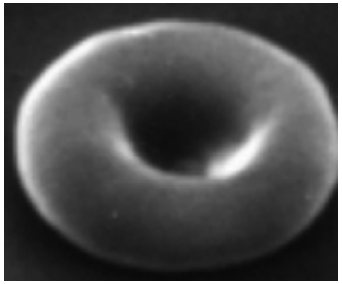
Which of the following patterns would be most suggestive of the predisposing condition?

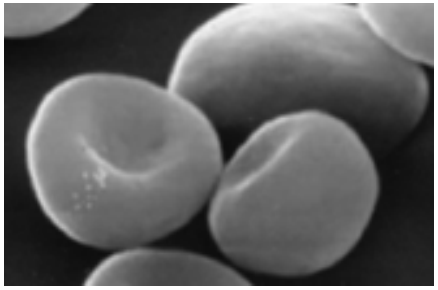


	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
2	35	76	Yes	38	24



Normal membrane
Normal MCH(C)

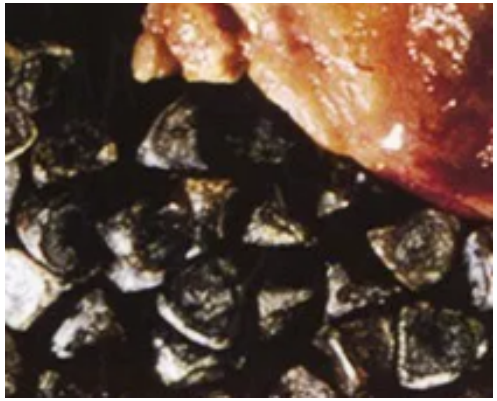




Membrane loss
MCH(C) ↑↑

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder. He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?

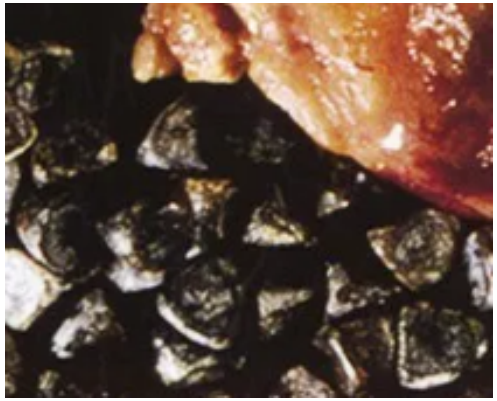


	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
1	35	76	No	32	8
B-12 or Folate Deficiency secondary to defective DNA synthesis					
4	35	105	No	36	24
5	55	76	Yes	32	8

MCHC, normal 33-36 g/dL

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder. He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?

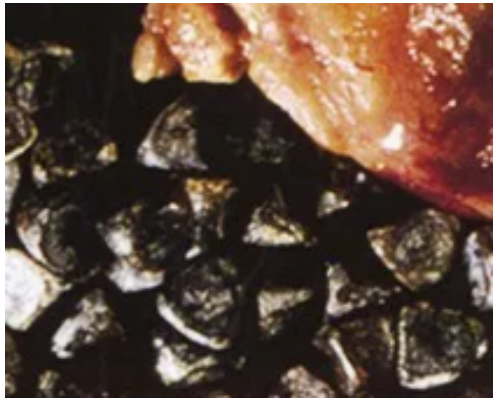


	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
Chronic liver disease with Portal HTN (and splenomegaly).					
3	35	105	Yes	36	24
4	35	105	No	36	24
5	55	76	Yes	32	8

MCHC, normal 33-36 g/dL

Patient presents with RUQ pain. HIDA scan fails to visualize gallbladder.
He is taken to OR for cholecystectomy. Specimen shown.

Which of the following patterns would be most suggestive of the predisposing condition?



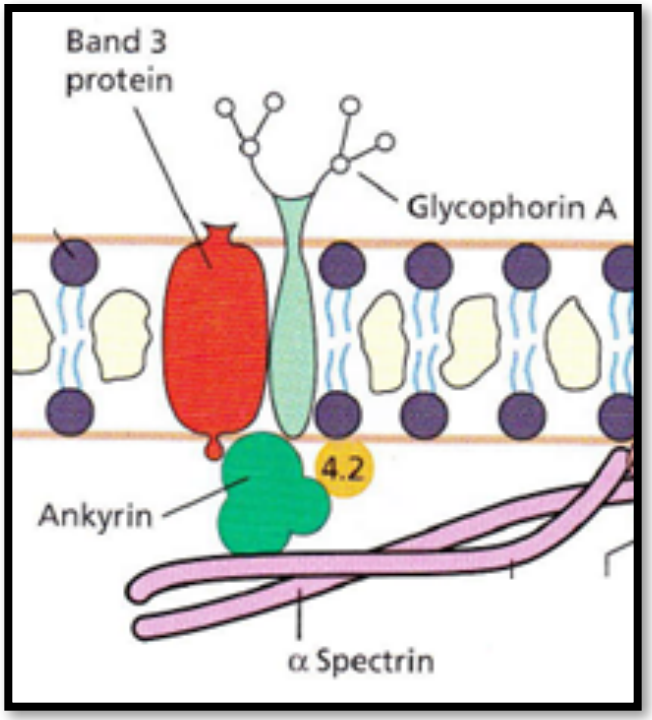
	HCT	MCV	Splenomegaly	MCHC	Transferrin Saturation (%)
2	35	76	Yes	38	24

Hereditary Spherocytosis

Hereditary Spherocytosis

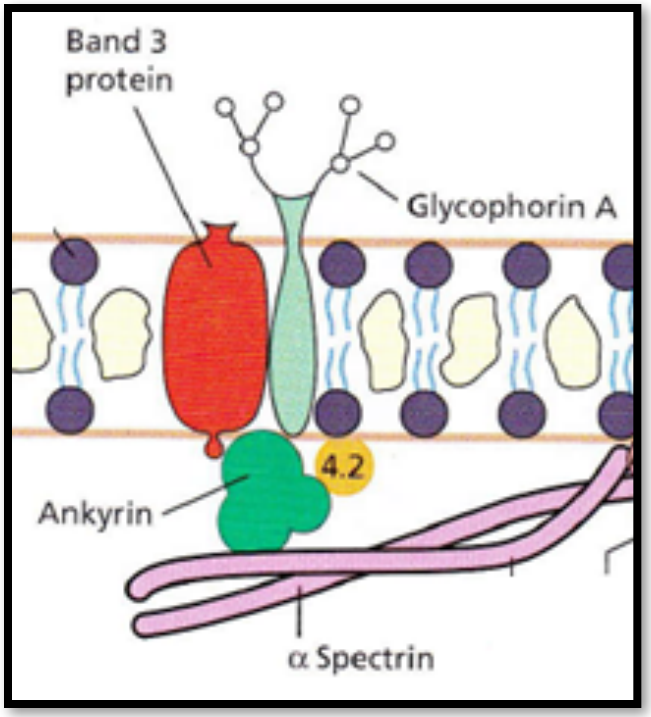
- Background
  - Hemolytic anemia 2° to membrane defect; shortened RBC lifespan due to loss of deformability and sequestration within spleen
  - AD inheritance with failure to produce tethering proteins

Be familiar with those proteins



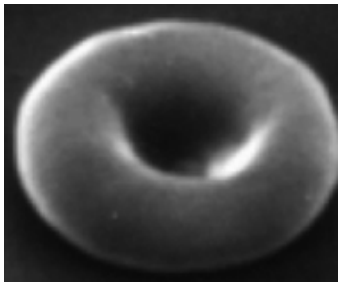
Hereditary Spherocytosis

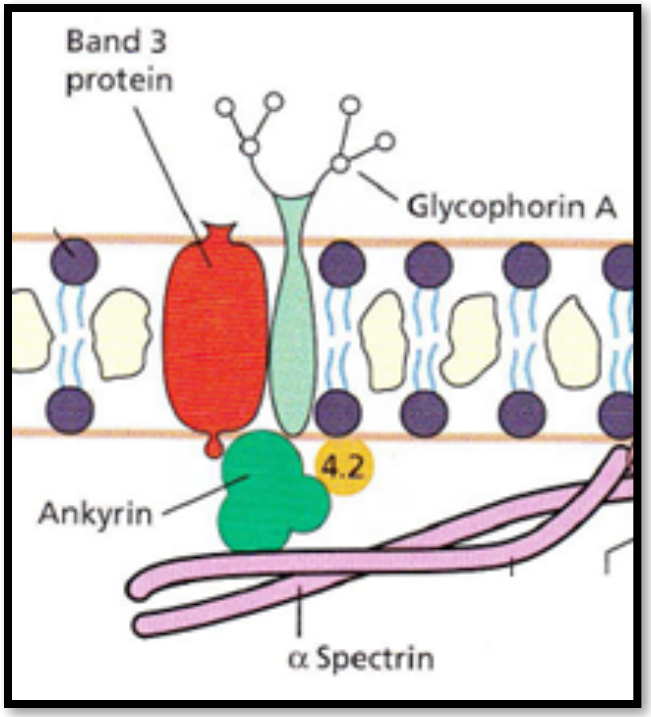
- Pathogenesis
  - Defects in the vertical interactions of the cytoskeleton and the lipid bilayer.
  - Marrow produces 'normal' cells. They become abnormal circulating through the spleen (2° to ↓ deformability)

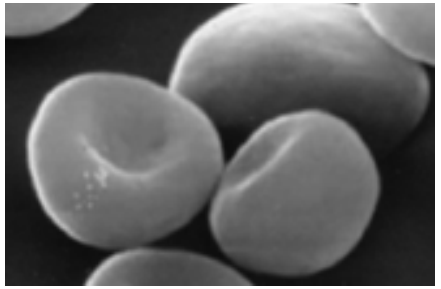


Hereditary Spherocytosis

- Pathogenesis
  - Defects in the vertical interactions of the cytoskeleton and the lipid bilayer.
  - Marrow produces 'normal' cells. They become abnormal circulating through the spleen (2° to ↓ deformability)

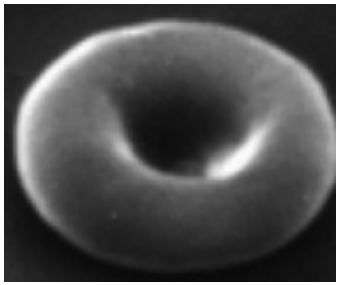




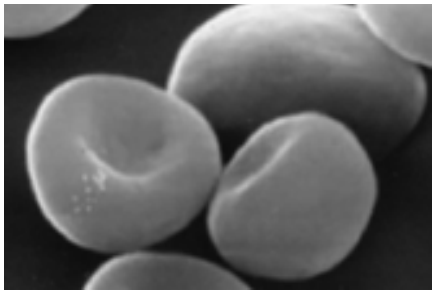


Hereditary Spherocytosis

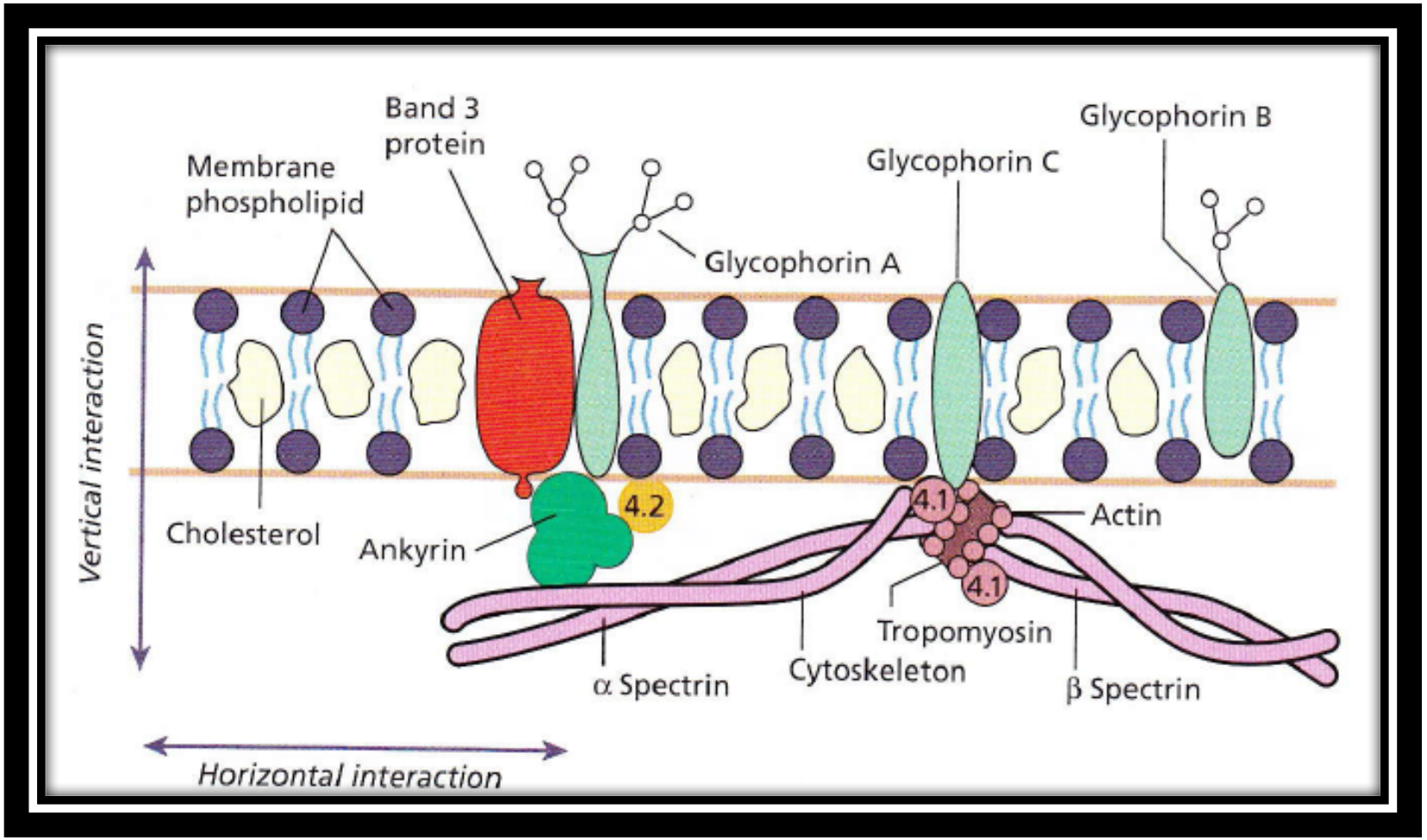
- Pathogenesis
  - Defects in the vertical interactions of the cytoskeleton and the lipid bilayer.
  - Marrow produces 'normal' cells. They become abnormal circulating through the spleen (2° to ↓ deformability)



Birth

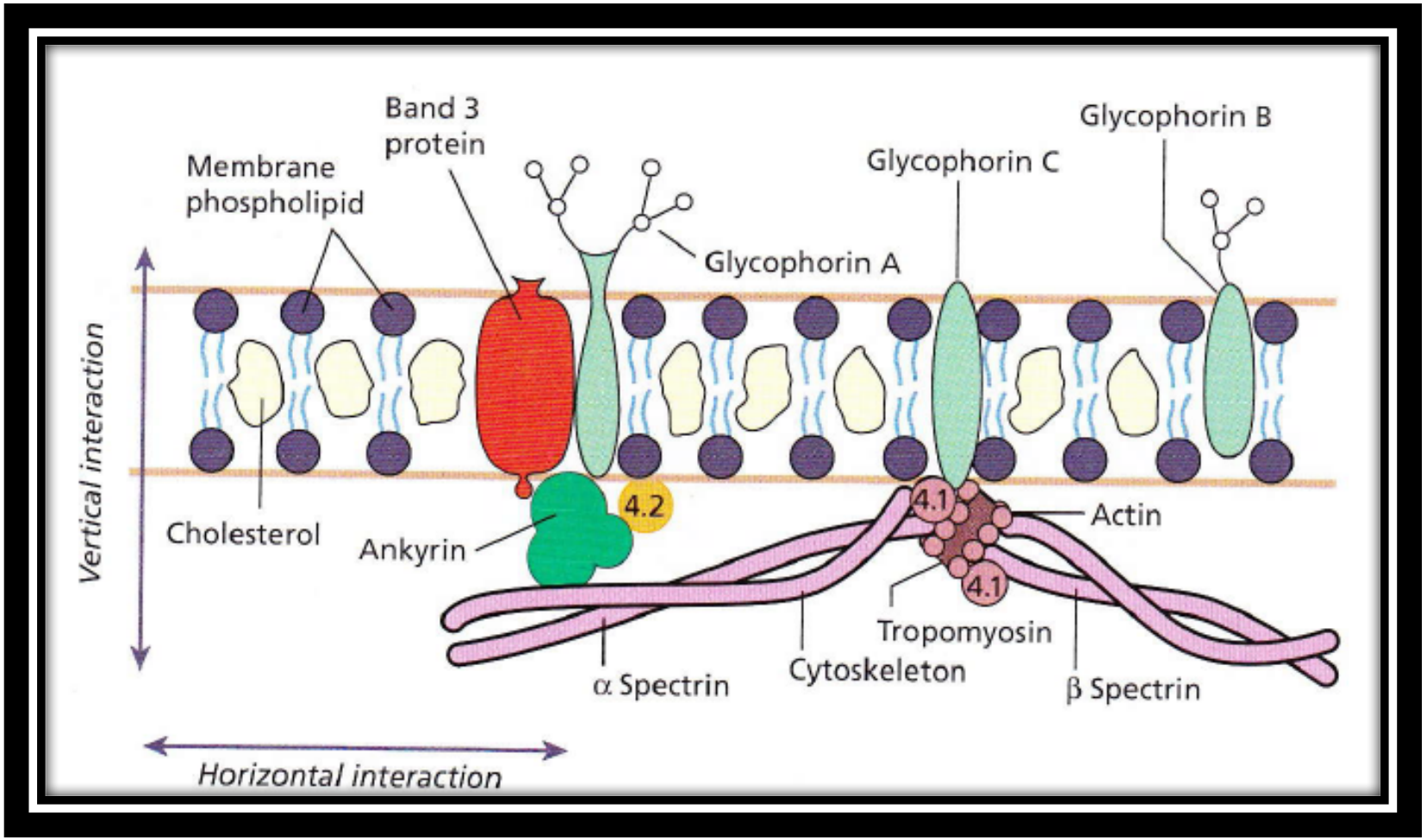


After a few passes through spleen



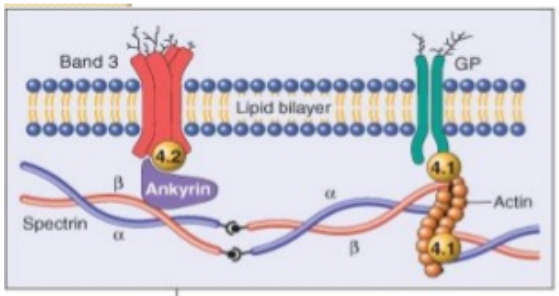
Defect in Ankyrin (vertical) and/or Spectrin (horizontal)

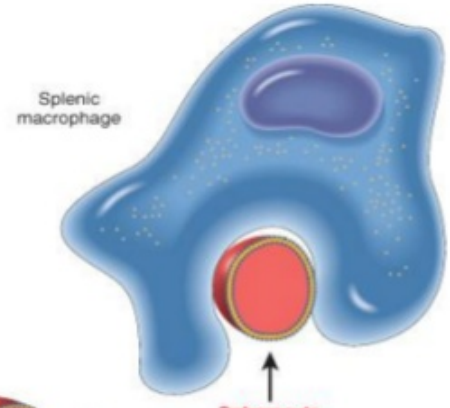
Spherocytes ⟷ Elliptocytes

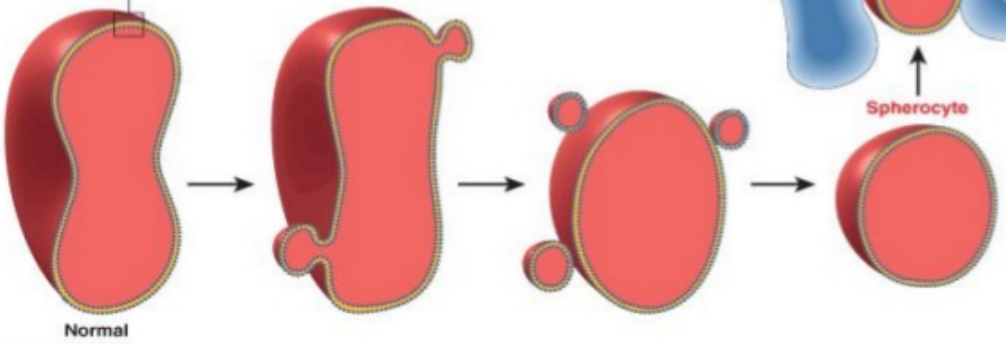


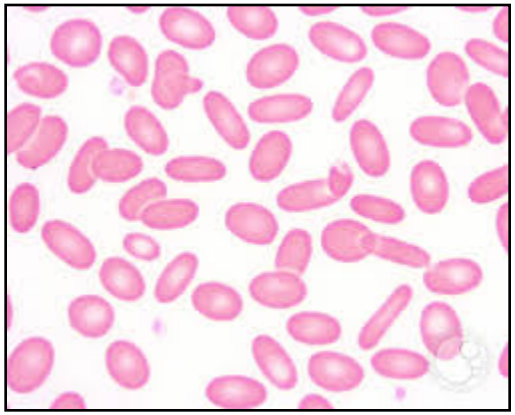
Cytoskeleton and Membrane Defect

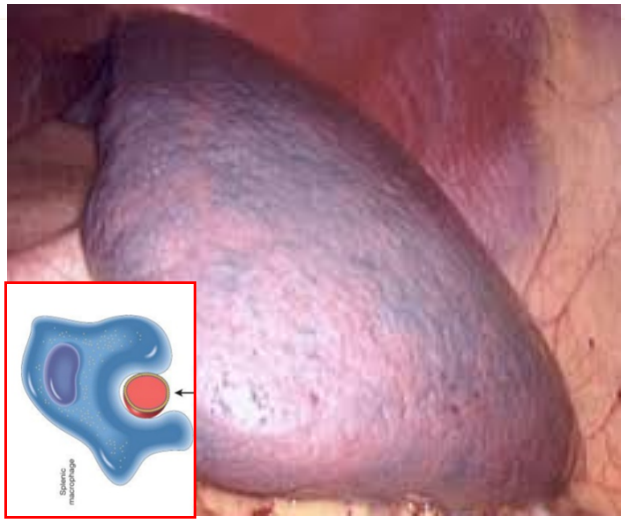
Mercifully, this is not a hemoglobinopathy.
Less Headaches.

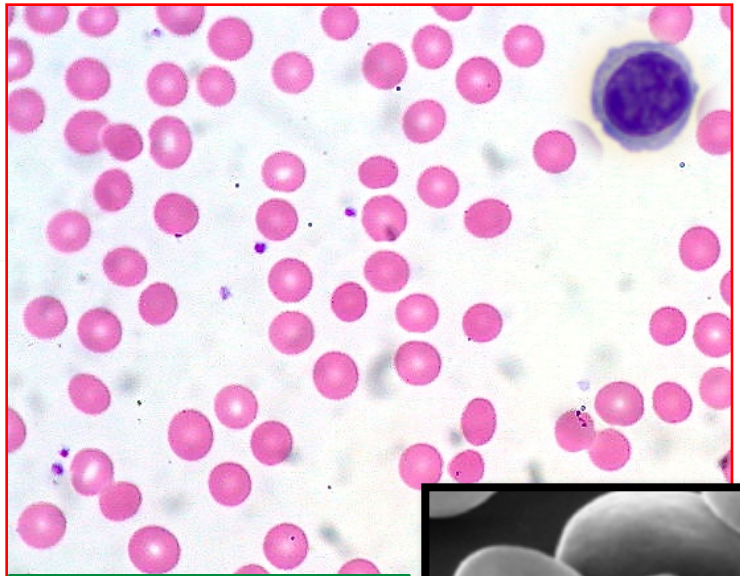




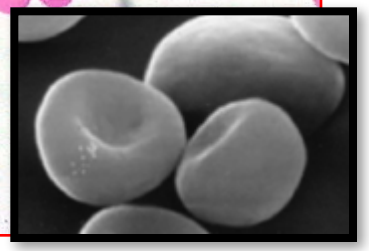








Where is the central pallor?

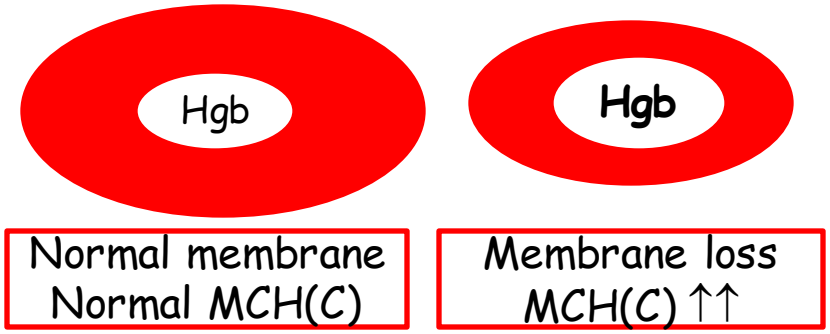


Hereditary Spherocytosis

- Clinical Presentation
  - Features of anemia, hemolysis (intermittent jaundice), splenomegaly
  - Complications of hemolysis: pigment stones (calcium bilirubinate)

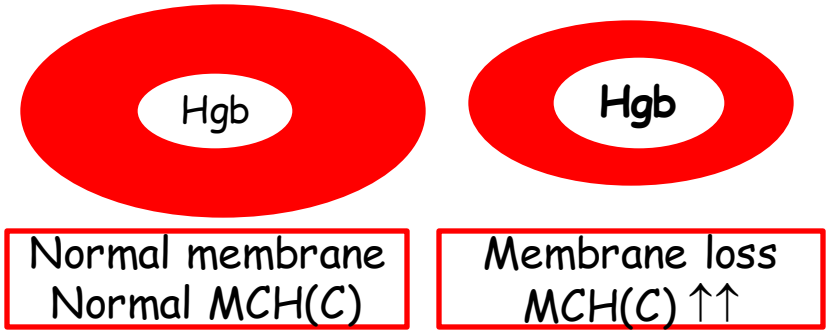
Hereditary Spherocytosis

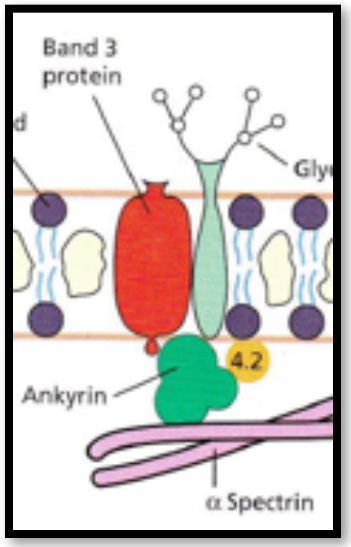
- Diagnosis
  - Smear: spherocytes (microcytic, ≠ hypochromic ∴ ↑ MCHC)
  - Hemolysis labs:
    - Reticulocytosis, LDH ↑/↓ haptoglobin/↑ bilirubin; Coombs (-)
  - Osmotic fragility test (dilute saline → hemolysis)



Hereditary Spherocytosis

- Diagnosis
  - Smear: spherocytes (microcytic, ≠ hypochromic ∴ ↑ MCHC)
  - Hemolysis labs:
    - Reticulocytosis, LDH ↑/↓ haptoglobin/↑ bilirubin; Coombs (-)
  - Osmotic fragility test (dilute saline → hemolysis)
  - Eosin-5-maleimide binding test (EMA)
    - Flow cytometry: measures binding of dye (EMA) to band 3 protein
  - Bone marrow: erythroid hyperplasia





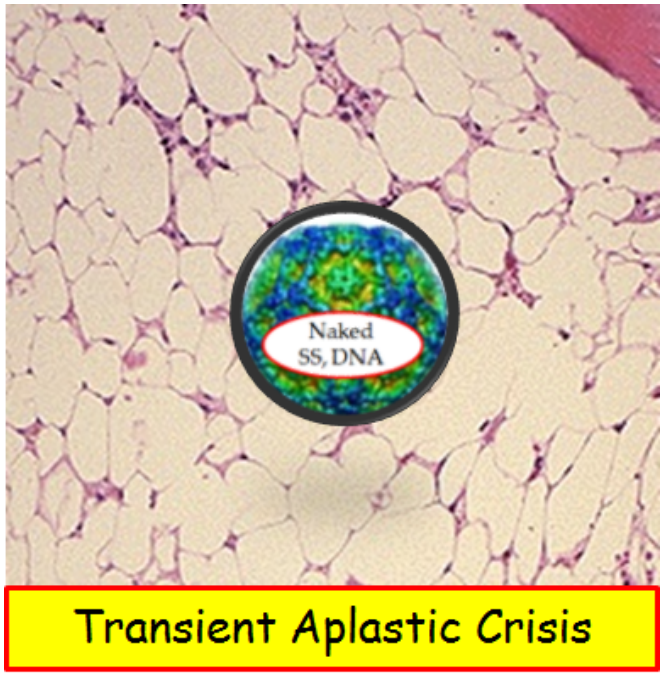
Hereditary Spherocytosis

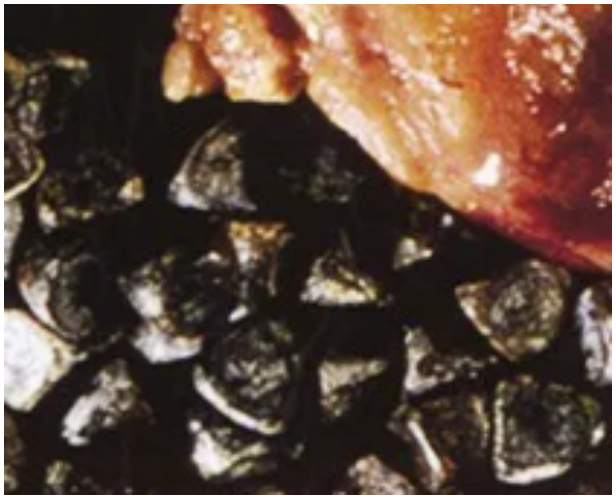
Which viral infection can shut down erythrocyte production in a jiffy?

- Bone marrow: erythroid hyperplasia

Hereditary Spherocytosis

- Special Notes:
  - Parvovirus: transient aplastic crisis
  - Rx: splenectomy when indicated



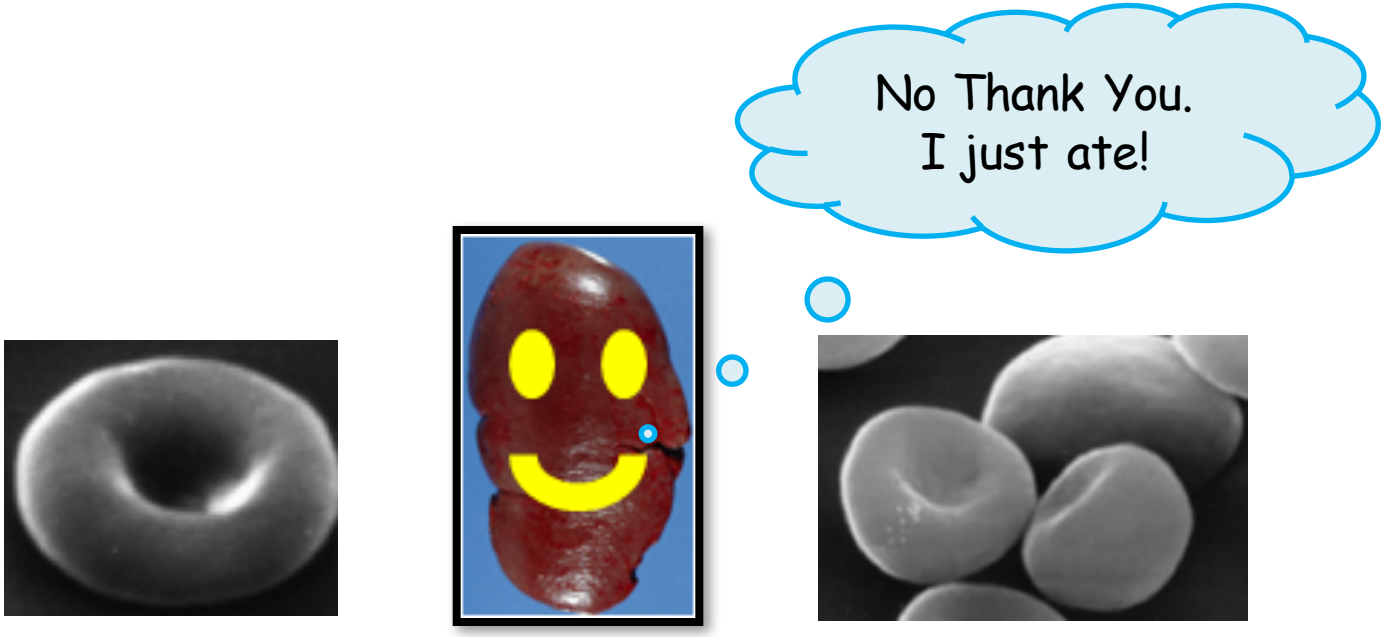


Pigment Stones:
A gateway condition to hemolytic anemia and associated derivatives.

Spherocytosis Derivatives:

1. Microcytosis with elevated MCHC/hyperchromic cells
2. Smear: loss of central pallor
3. Abnormal anchoring protein: Ankyrin (Spectrin)
4. PE: big spleen, mild jaundice
5. Dx: Osmotic Fragility Test/Flow cytometry
6. Complication: TAC (Parvovirus), pigment stones
7. Rx: Splenectomy

Heme Questions and Derivatives for the USMLE Step One Exam



Howard J. Sachs, MD
Howard@12DaysinMarch.com
www.12DaysinMarch.com