Imaging Sickle Cell Disease in the Emergent Setting

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LEARNING OBJECTIVES

1. Summarize key epidemiological data regarding sickle cell disease
2. Understand the pathophysiology and imaging findings of the *most common* manifestations of sickle cell disease a radiologist can expect to encounter in the emergent setting
3. Recognize the pathophysiology and imaging features of *less common* complications of sickle cell disease in the emergent setting

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Children's Healthcare of Atlanta

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Epidemiology of Sickle Cell Disease (SCD)

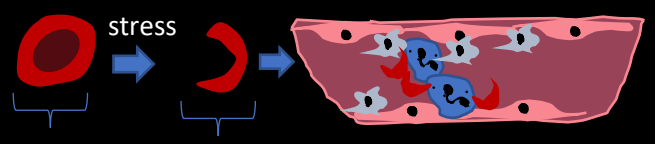
100,000
Americans are currently living with SCD

$13,000
Approximate mean annual healthcare expenditure per patient with SCD in 2005

57%
Of all patients with sickle cell disease will have an at least one ED visit in one year

27
Estimated number of imaging studies a child with sickle cell disease will have by age 18

Pathophysiology



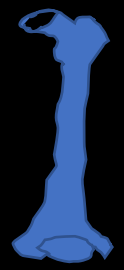
6-8 µm 6-10 µm Vaso-occlusion

Tissue hypoxemia (often induced by stress/infection) leads to abnormal polymerization of RBC proteins, leading to sickling and hemolysis → this in turn leads to aggregation of sickle red blood cell (RBC) with neutrophils and endothelial cells and platelets causing vaso-occlusion. → inflammatory tissue factors are released leading to further vaso-occlusion through increased adhesion.

Compared to normal RBC, (sickle cells have decreased vibroelasticity the ability to change morphology when traveling through vessels that are smaller than the RBC) and decreased O_2 capacity.

52% of patients with SCD will have an brain infarction by adulthood. This frequently occurs in watershed territories. Children with SCD are chronically anemic, exhausting cerebrovascular compensatory mechanisms. Therefore, when there is an insult requiring increased O_2 to the brain, watershed territories are at heightened risk for ischemia. *Moyamoya vasculopathy*, *aneurysms*, & *intimal hyperplasia* within the intracranial carotid and middle cerebral arteries are more common in SCD, resulting in large territory infarctions.

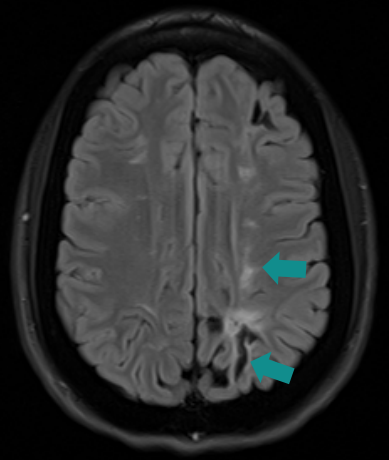
Repeated pulmonary vaso-occlusive crises can lead to endothelial damage and areas of *lung fibrosis*, most often in the dependent portion of the lungs. Additionally, endothelial damage can lead to pulmonary hypertension.



Vaso-occlusive crises in the bone can lead to micro-infarctions. Additionally, chronic anemia leads to extramedullary hematopoiesis and reconversion of yellow marrow into red marrow.

NEUROVASCULAR ANOMALIES

INFARCTION



FLAIR MRI demonstrates areas of gliosis & encephalomalacia (blue arrows).

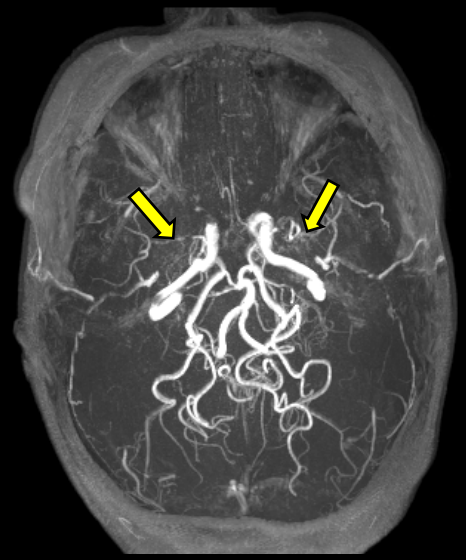
Lacunar and watershed territory infarctions involving white matter tracts are common in sickle cell patients. White matter gliosis can be seen in asymptomatic patients as "silent infarcts."

Silent infarctions are associated with decreased cognitive function over time.

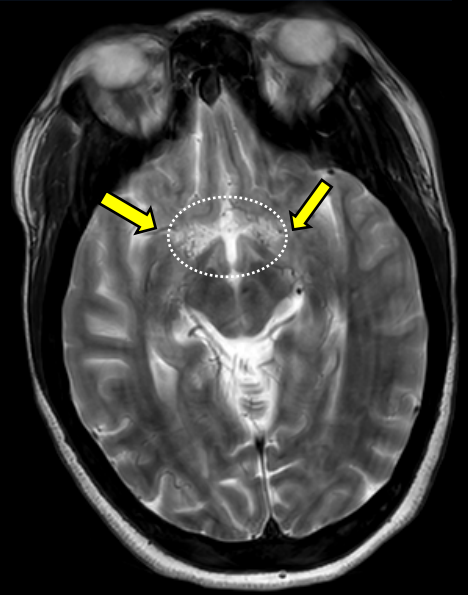
MOYAMOYA SYNDROME

Moyamoya syndrome from SCD is distinguished from the idiopathic type, termed *moyamoya disease*. This vasculopathy is an uncommon, but very important entity in SCD. Moyamoya syndrome can result in large territorial infarctions or intraparenchymal hemorrhage.

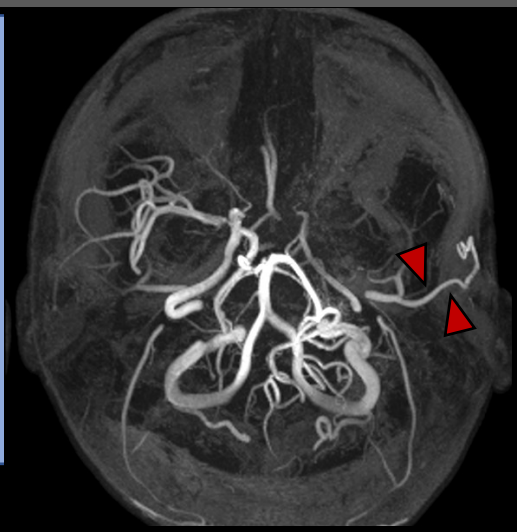
In SCD patients with cerebral vascular accidents, diagnosing moyamoya syndrome early can reduce morbidity. "Moyamoya" = Japanese for "puff of smoke" based on the appearance of tangles of collateral arteries compensating for diseased arteries.

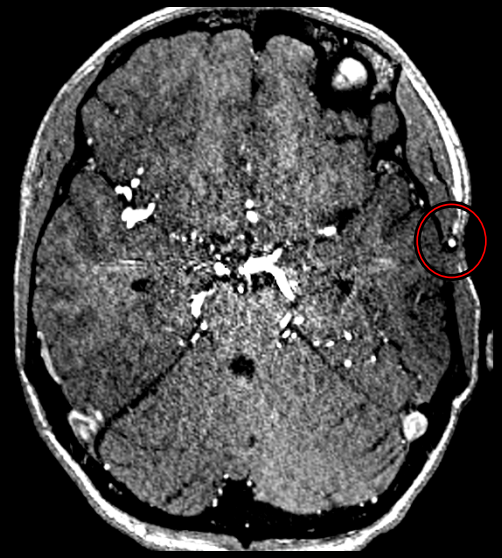


MRA shows severely narrowed vessels in the MCA territory, right greater than left (yellow arrows).



T2-weighted images show abnormally narrowed MCAs (yellow arrows) and *"puff of smoke" appearance of collaterals (dotted line).*





MRA showing patient status post EDAS -left temporal artery internalized to provide perfusion to the left hemisphere **(circle, arrowheads)**.

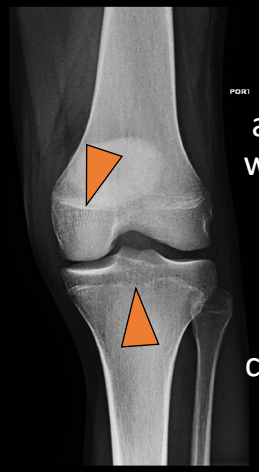
Encephaloduroateriosynangiosis (EDAS) can be performed to revascularize the affected brain parenchyma. A superficial artery (usually temporal artery) is internalized intradurally to revascularize the affected territories.

EDAS outcomes include *return to normal activities & reduction of strokes.*

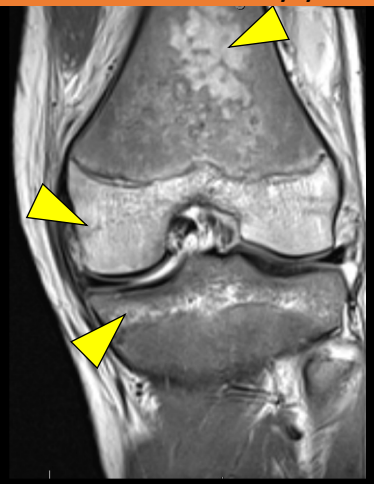
MUSCULOSKELETAL ANOMALIES

MARROW RECONVERSION

Marrow reconversion = process of reconverting yellow marrow to red marrow in anemic states. Red marrow appears dark on T1. Fatty yellow marrow is bright.

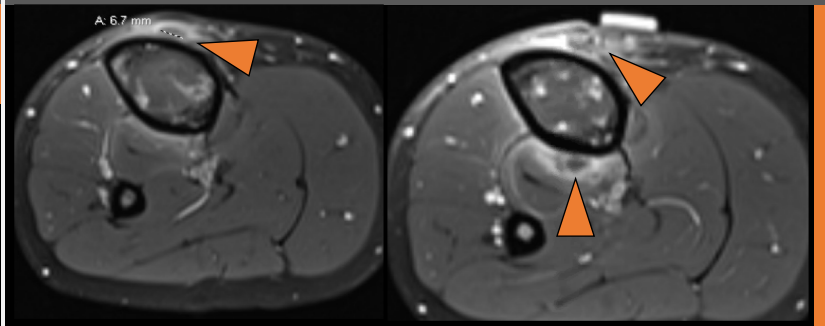


Normal knee radiograph in an 18-year-old with knee pain. Epiphyseal scars (orange arrowheads) are present, consistent with patient's age.



T1-weighted MRI: Normal fatty yellow marrow (yellow arrowheads) in epiphyses and focally in distal femoral shaft. Elsewhere, hypo-intense regions reflect conversion to red marrow.

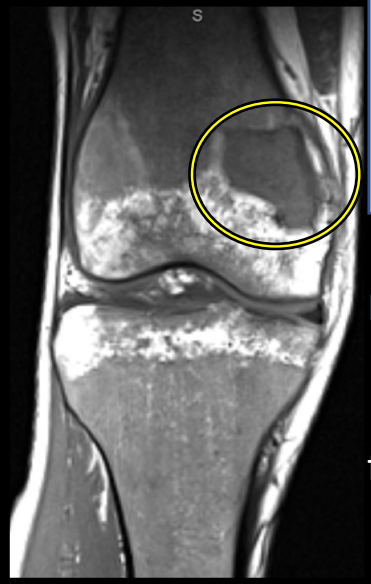
OSTEOMYELITIS



Bone infection with Salmonella is rare overall; but relatively common in SCD.
In the setting of bone pain out of proportion to usual crisis, consider MRI.

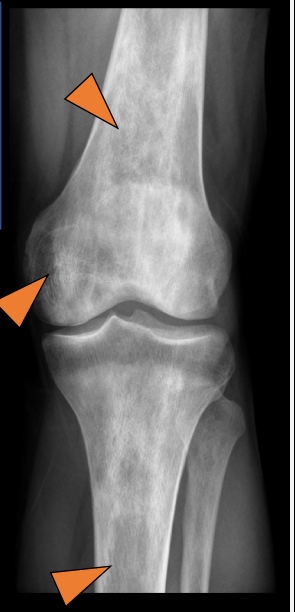
T1-weighted post-contrast with fat-saturation: Rim-enhancing fluid collections abutting anterior & posterior tibial cortices (orange arrowheads),consistent with subperiosteal abscesses, which were occult on radiography. Blood culture revealed *Salmonella*.

BONE INFARCTION



Vaso-occlusive crises in the bone can lead to patchy areas of infarction throughout the axial and appendicular skeleton.

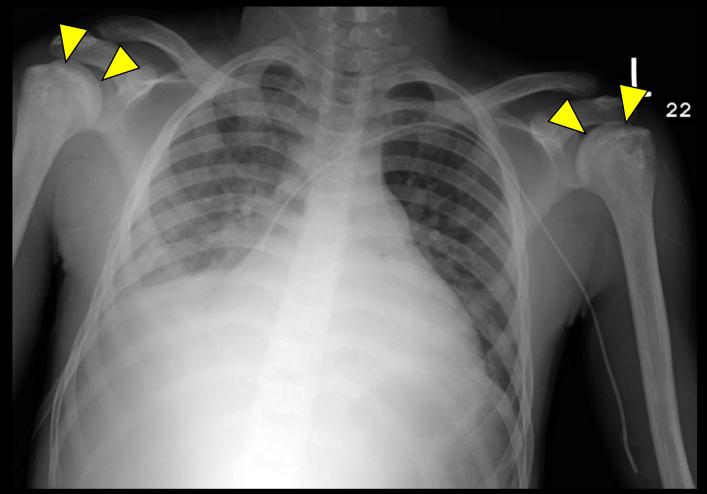
T1-weighted MRI: Large area of bone infarction in the medial distal femoral condyle (yellow circle).



Radiograph: lucent areas (arrowheads) on a background of serpentine pattern of sclerosis = infarcts

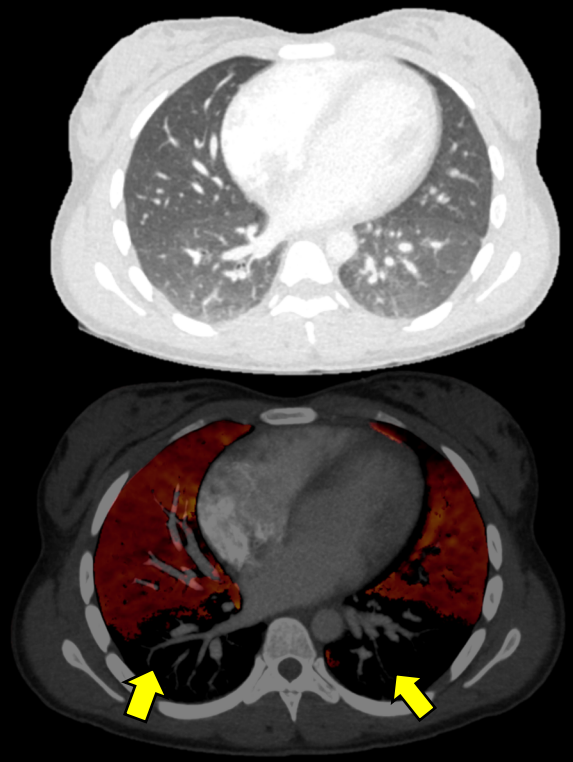
AVASCULAR NECROSIS

A feared, yet common complication, *avascular necrosis* typically occurs at the femoral and humeral heads. In this patient with acute chest syndrome, there is abnormal sclerosis of the humeral heads with crescent sign (yellow arrowheads) consistent with avascular necrosis. This is a type of bone infarct that occurs in the subchondral bone.



CHEST AND ABDOMEN ANOMALIES

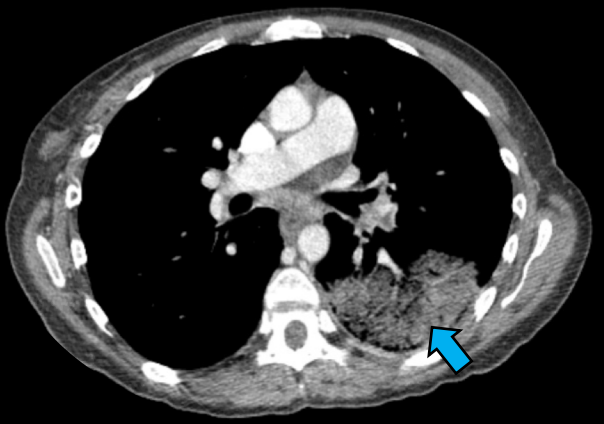
ASEPTIC HYPOXEMIA



In SCD patients with chest pain and hypoxemia, contrast-enhanced CT is often used to exclude acute *pulmonary embolism (PE)*; however, in the sickle cell population, rates of PE are actually **LOWER.**

Dual-energy CT demonstrates areas of decreased perfusion (yellow arrows), which do not correspond to any parenchymal disease (top). Hypoperfusion is likely secondary to chronic microvascular occlusion from sickling, resulting in scarring.

ACUTE CHEST SYNDROME



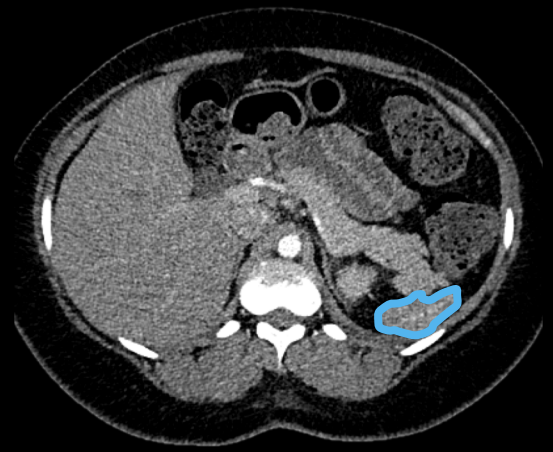
ACUTE CHEST SYNDROME = symptoms (such as hypoxemia or chest pain) **+** consolidation on radiograph or CT.

Contrast- enhanced axial CT of the chest: Left lower lobar consolidation (blue arrow) consistent with acute chest syndrome.

SPLENIC AUTO-INFARCTION

Repeated splenic infarctions in sickle cell patients results in a small (often hyperdense) spleen, termed *auto-infarction.*

In this young adult with SCD, the spleen is ~4cm (outlined). Normally the spleen measures between 9 and 12 cm.



SPLENIC SEQUESTRATION

Rarely, a crisis will result in splenic sequestration, which is a life-threatening emergency!

Imaging hallmarks include, enlarged spleen (outlined) and hypoenhancement of the spleen relative to the liver (on portal venous phase CT).
These findings + *acute decrease in hematocrit* should increase clinical suspicion of sequestration.