VASO-OCCLUSIVE CRISIS (VOC)

TERM DEFINITION

A common painful complication of sickle cell disease characterized by acute episodes of pain due to erythrocyte microvascular occlusion and tissue hypoxia; also referred to as acute pain crisis or acute vaso-occlusive pain.

EPIDEMIOLOGY

Sickle cell disease (SCD) is an inherited blood disorder affecting about 100,000 individuals in the US. Nearly all such patients will experience a VOC during their lifetime. Some have 6 or more episodes per year, whereas others have episodes only at long intervals.

CLINICAL PEARLS

VOC in children typically presents as pain and swelling in hands and feet (dactylitis).

There are no tests to rule in or to rule out a VOC.

VOC is the leading cause of emergency department visits and hospitalizations for patients with SCD.

About 30% of patients develop at least one VOC per year requiring an inpatient stay. Each individual typically has a consistent pattern for pain distribution and crisis frequency.

Risk factors or triggers for VOC include infections, dehydration, cold weather, higher Hb levels, and extremes in humidity. High levels of HbF are associated with decreased frequency of VOC. Only a minority of pain episodes are associated with an identifiable precipitating factor.

PRESENTATION

SIGNS & SYMPTOMS

Pain crises begin suddenly, last several hours to days and often terminate as abruptly as they began. Pain most commonly occurs in the extremities, chest, and back, and may be accompanied by fever.

LABS

HEMATOLOGICAL FINDINGS

Hemoglobin typically drops by 1-2 g/dL in first 2 days.

Other hematological findings associated with pain crisis (vs. baseline):

- Elevated RDW
- Thrombocytosis
- Leukocytosis

OTHER LABS

Other labs increased vs. baseline:

- LDH
- ESR
- C-reactive protein

NOTE:
There are no specific tests for diagnosis of VOC.

DIAGNOSIS

Diagnosis of exclusion.

It is important to rule out other causes of pain, including osteomyelitis, acute chest syndrome, pulmonary embolus, papillary necrosis, acute abdomen or hepatic or splenic sequestration.

TREATMENT GOALS

- Control pain.
- Prevent progression to acute chest syndrome.
- Treat as an acute medical emergency.

PAIN CONTROL	Rapidly initiate (ideally within 30-60 minutes) analgesic therapy with NSAIDS (provided no contraindication) in those with mild to moderate pain and NSAIDS and opioids in patients with severe pain, using individualized care plan.
SPIROMETRY	Encourage incentive spirometry to reduce the risk of ACS.
SUPPLEMENTAL OXYGEN	Administer supplemental oxygen if oxygen saturation < 95% on room air.
IV FLUIDS	Provide IV hydration if patient unable to drink. No guideline recommendation of 0.45% vs. 0.9% saline.
TRANSFUSION	Do not administer transfusion unless there are other indicators for transfusion.

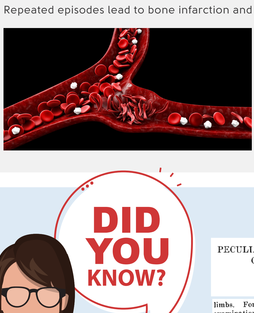
Note: *the patient with SCD and VOC who is able to drink and has good oxygen saturation or room air does not need IV fluids or supplemental oxygen.*

PROXIMATE MECHANISMS

A vaso-occlusive crisis occurs when the microcirculation is **obstructed by sickled red cells (SS-RBCs)**, causing **ischemic injury** and resultant pain. Proposed series of events:

1. SS-RBCs activate the endothelium.
2. Activated endothelial cells recruit adherent leukocytes.
3. Adherent neutrophils become activated.
4. Adherent, activated neutrophils interact with SS-RBCs.
5. Heterotypic cell-cell aggregates composed of SS-RBCs, adherent leukocytes, and possibly platelets cause vascular clogging.
6. Transit time increases, leading to increased formation of SS-RBCs.
7. Ischemia from the obstruction creates a feedback loop of worsening endothelial activation.

Repeated episodes lead to bone infarction and necrosis.



The schematic on the left shows the blockage of microvessel with sickled cells. This type of occlusion is responsible for the majority of acute and chronic complications of SCD, including VOC.

HISTORY OF MEDICINE

PECULIAR ELONGATED AND SICKLE-SHAPED RED BLOOD CORPUSCLES IN A CASE OF SEVERE ANEMIA

JAMES B. HERRICK, M.D.

CHICAGO

limbs. For the past five weeks he had been coughing. Two days prior to examination he had "taken cold," his cough had grown worse and he had had a slight chill, followed by fever. It was this cough and fever for which he wished treatment at the hospital, and of which he chiefly complained, though he mentioned also that he felt weak and dizzy, had headache and catarrh of the nose,

James Herrick's famous first report of sickle cell disease (*Arch Int Med.* 1910;6:517) seemed to describe a respiratory condition (acute chest syndrome?), not a pain crisis.

NOTES

ATTRIBUTIONS

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ENCYCLOPEDIA OF BLOOD