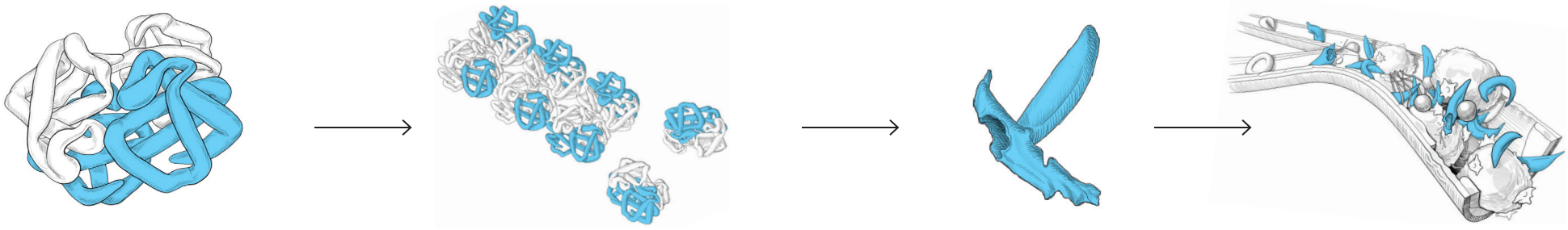
Understanding and addressing the COMPLICATIONS *of* SCD

Sickle cell disease (SCD) is a lifelong *genetic disease* caused by a single mutation in the *HBB* gene and is characterized by hemolytic anemia, vasculopathy, and episodic vaso-occlusive events (VOEs)[1-3]



When glutamic acid in the sixth position of adult hemoglobin is replaced by valine, the result is hemoglobin Sickle (HbS)[3]

Under low oxygen conditions, HbS molecules undergo a conformational change, which causes HbS molecules to bind to one another and form polymers; HbS polymerization correlates with high concentrations of HbS within red blood cells (RBCs)[2-4]

HbS polymerization distorts the RBC, leading to the distinct sickle shape, increased fragility, aggregation, and hemolysis[3,4]

Sickled RBCs induce chronic hemolytic anemia, vasculopathy, and episodic VOEs[2,3]

Vaso-occlusions manifest as both unpredictable *acute and ongoing chronic complications*

Acute complications may occur suddenly and resolve quickly, but the underlying damage associated with acute complications may occur and worsen over time. Recurrent VOEs can lead to serious and chronic complications and organ damage[1,2]

ACUTE COMPLICATIONS[1,5-8]

Acute pain

Acute chest syndrome

Infection (including sepsis and parvovirus)

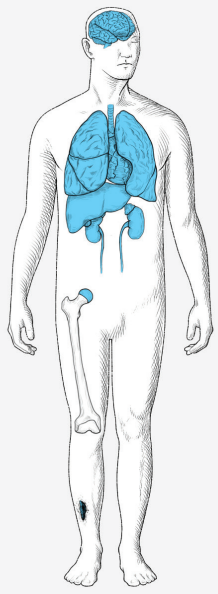
Priapism

Overt thrombotic and hemorrhagic stroke

Cerebral vasculopathy

Splenic sequestration

Although the occurrence of acute complications in SCD is unpredictable, the chronic damage that results from the disease is expected and deserves careful monitoring and management[1,2]



CHRONIC COMPLICATIONS[1,7,9-14]

Sickle cell retinopathy

Pulmonary hypertension

Cardiovascular complications

Cerebral vasculopathy

Cholelithiasis

Renal dysfunction

Avascular necrosis of femoral head

Recurrent, chronic leg ulcers

Silent stroke (asymptomatic cerebral infarction)

Depression and anxiety

End organ disease

Chronic complications can affect any organ in the body and may lead to progressive organ damage and even organ failure[1,14]

Beyond the physiological effects of SCD, patients often experience a psychosocial burden attributed to the disease[13]

Physicians can take the following steps to optimize treatment for patients with SCD

Individualize pain management for each patient

The published guidelines recommend an individualized protocol for pain management

Monitor and manage for chronic complications and organ damage

Consult the published guidelines for information about the management of chronic complications of SCD

Initiate a discussion with patients about any struggles they face beyond SCD

Work with patients to understand what support you can provide and help to connect them to mental health resources, community, and sickle cell organizations

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