Valley Children's HEALTHCARE

Central California
Pediatrics

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Specialty information for physicians who treat children and expectant mothers.

# What to Know About Sickle Cell Disease

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## Understanding Sickle Cell Disease

Sickle cell disease is an inherited blood disorder affecting red blood cells. Normal red blood cells contain hemoglobin A. People with sickle cell disease have red blood cells containing mostly hemoglobin S, an abnormal type of hemoglobin. These red blood cells become sickle-shaped and have difficulty passing through small blood vessels. There are several different types of sickle cell disease; the most common types are homozygous sickle cell disease (SS disease), and sickle-cell beta thalassemia (Sß+ or Sß0 disease).

## How common is sickle cell disease?

Sickle cell disease is one of the most common diseases in the world, with approximately 300,000 babies being born each year with some form of the disease. Sickle cell disease is the most common disease detected by newborn screening efforts in the United States, as between 1 out of every 2500 and 1 out of every 2000 babies born in the U.S. has some form of the disease. Overall, it is estimated that approximately 100,000 persons in the U.S. are living with sickle cell disease.

## Who gets sickle cell disease?

Sickle cell disease can affect persons of any racial or ethnic background. In the U.S., African-Americans are most likely to have the disease, though it is found among many different racial and ethnic groups, including whites, Hispanics, Native Americans and Southeast Asians.

## What are the symptoms of sickle cell disease?

Sickle cell disease causes a number of health complications for the affected individual. Persons with sickle cell disease are more susceptible to infections and strokes. Sickle cell disease can cause progressive organ damage throughout the body, including the lungs, kidneys, and joints. Some people with sickle cell disease may develop neurocognitive deficits. The hallmark symptoms of the disease, however, are the episodes of severe acute pain, called vaso-occlusive crises (or sickle cell pain crises), that the individual can experience. These pain episodes can affect any part of the body. These episodes can be as short as a few hours in length, or they may last for days on end. In addition to this severe, acute pain, the disease is the source of chronic pain as well (in the hips, back, or other joints, for example). Sickle cell disease can cause early mortality, and even though people with the disease are living longer, it is estimated that persons with the most severe form of the disease (sickle cell anemia) have a median life expectancy approaching 50 years.

Diagnosis of sickle cell disease has been part newborn screening for all children in the United States since July 1, 1985. Comprehensive care includes early diagnosis, preventive measures, treatment of complications and ongoing patient education.

Patients with these conditions should be referred to specialized hematology centers for treatment. They need surveillance under the expert care of a hematologist

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### Preventing Infections

People with sickle cell anemia need to keep their immunizations up to date, including Haemophilus influenza, pneumococcal, meningococcal, hepatitis B, and influenza.

Some patients may receive antibiotics to prevent infections.

### When Should I Refer to a Hematologist?

Children should be referred to a hematologist after abnormal newborn screenings for hemoglobin disorders have been noted. Additionally, patients should be referred when S hemoglobin is noted instead of normal hemoglobin on hemoglobin electrophoresis, or when anemia and jaundice are noted with recurrent pain episodes.

## Upcoming CME Opportunities

**Physician Wellness Series Preventing Physician Suicide: How to Have Honest Conversations with Your Colleagues**
Presented by Michelle Grua, MD
Wednesday, November 2
12:15 p.m. - 1:15 p.m.

**Clinical Excellence & Innovations Series: Choosing Wisely in Pediatrics: When Less is More**
Presented by Dr. Jolie Limon & Dr. Shea Osburn
Thursday, November 3
12:15 p.m. – 1 p.m.

**Religious Beliefs & Practices Series: Native American Spirituality and Healing**
Presented by Delaine Bill
Friday, November 4
12:15 p.m. - 1:15 p.m.

**Pediatric Clinical Symposium Topic on Plastic Surgery**
Presented by Dr. Duncan Mackay
Wednesday, November 30
12:15 p.m. - 1:15 p.m.

Register for Valley Children's CME events through our CME Tracker, cmetracker.net/VCH

## Medical Staff News

The following pediatric specialist recently joined Valley Children's:

**Anesthesiology**
Michelle Grua, MD
Audesho Shlimun, MD

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**Child Advocacy**
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