and change the way blood vessels work. This reduces oxygen delivery and causes kidney cells to die. This leads to the kidney issues seen in SCN.

WHAT ARE THE SYMPTOMS?

Some symptoms of chronic kidney disease include:

- Frequent urination
- Nighttime bedwetting
- High blood pressure (hypertension)
- Bloody urine
- Nausea and vomiting
- Fatigue and weakness
- Shortness of breath and chest pain

People with SCD often have a low blood pressure. This means that a relatively “normal” blood pressure may sometimes actually be a sign of hypertension (high blood pressure) and kidney disease.

These are not all the symptoms of kidney disease, and many of these symptoms can be caused by other complications of SCD. If you experience any symptoms of kidney disease, talk to your doctor about getting tested for kidney function.

ALBUMINURIA (TOO MUCH PROTEIN IN THE URINE)

People with SCD often have albuminuria, or too much protein in the urine (possibly an early sign of kidney disease). A simple urine test done every year will help your provider monitor the amount of protein in your urine. Your provider may prescribe medicines that lower the amount of protein in your urine to help protect your kidneys.

WORSENING ANEMIA RELATED TO CHRONIC KIDNEY DISEASE

In people with SCD, chronic kidney disease can cause worsening anemia, a condition in which the body does not make enough red blood cells. Share with your provider if you have symptoms of anemia like worsening fatigue, generalized weakness, pale skin, dizziness, and more.

Understanding KIDNEY PROBLEMS

and Sickle Cell Disease

People with sickle cell disease (SCD) are at greater risk than the general population for kidney complications. The purpose of this brochure is to provide basic understanding of kidney problems and Sickle Cell Disease.

GET IN TOUCH!

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WHAT IS SICKLE CELL DISEASE?

Sickle cell disease (SCD) is an inherited blood disorder (a blood disorder that runs in families). People with SCD produce an abnormal type of hemoglobin (called hemoglobin S (HbS) or sickle hemoglobin). Hemoglobin is a protein in red blood cells that carries oxygen from the lungs to the organs and tissues in the body. The abnormal hemoglobin in SCD can cause the red blood cells to have a sickle or banana shape under certain conditions. People with SCD often have a decreased number of red blood cells, a condition called anemia,which can cause lack of energy, breathlessness, and pale color of the skin and lips.

WHAT KIDNEY PROBLEMS DO PEOPLE WITH SCD EXPERIENCE?

Chronic kidney disease is common in SCD and shows a range of conditions, called sickle cell nephropathy (SCN).

The kidney filters waste from our blood and produces urine. It also makes urine concentrated by reducing the amount of water needed to remove waste.

When the kidney does not get enough oxygen, kidney cells begin to die. The kidney gradually loses its ability to filter out waste. This may cause proteins that are normally saved by the kidney to be lost in urine (proteinuria).

Without oxygen, the kidney also cannot absorb water from urine, leading to urine with too much water in it (hyposthenuria). This can lead to excessive and nighttime urination.

Kidney problems can also cause bloody urine (hematuria). However, hematuria may be caused by other SCD-related complications, such as urinary tract infections and a kidney cancer called renal medullary carcinoma.

Children and adults with SCD should be regularly tested for kidney disease. Doctors will perform lab tests on blood and urine to determine if you have kidney problems, such as proteinuria, hyposthenuria, or hematuria. They may also analyze how much creatinine is in your blood and urine. Creatinine is a waste product released into urine by the kidneys. Creatinine concentrations in blood and urine can tell doctors how well the kidney is filtering blood.

Acute kidney injury (AKI) is also common in children and adults with SCD. This is a sudden rise in blood creatinine levels. Most people recover kidney function after an episode, but repeated episodes may lead to chronic kidney disease.

WHY DOES SCD CAUSE THESE PROBLEMS?

Sickle cell neuropathy leads to chronic kidney disease in 1 out of every 3 to 4 adults with SCD. People with sickle cell anemia (HbSS) or sickle beta zero thalassemia have a higher risk than people with other types of SCD. About 1 in 6 people with SCD die because of chronic kidney disease. On average, people with SCD who have kidney failure live about 12 years shorter than people with SCD who do not.

SCD causes kidney problems when sickle cells block blood flow in the kidney. The blood vessels leading to the innermost part of the kidney are most susceptible to sickling. This is because it is a relatively acidic area with low oxygen. In this environment, sickle hemoglobin proteins are more likely to form rigid strands inside red blood cells and produce sickle-shaped cells.

Sickle cells in the kidney block flow