# UNDERSTANDING FOCAL SEGMENTAL GLOMERULOSCLEROSIS (FSGS)

| | |
|---|---|
| **Focal** | Some |
| **Segmental** | sections |
| **Glomerulo-** | of the kidney's filter units |
| **Sclerosis** | are scarred over time |

TRAVERE THERAPEUTICS

## What is FSGS?

**Focal segmental glomerulosclerosis (FSGS) is a rare kidney condition. It affects the kidney's filter units (called glomeruli). They stop filtering the blood properly and protein spills into the urine (this is called proteinuria). Over time, this may lead to kidney failure.**

## PHONETICS

**Focal segmental glomerulosclerosis:**
<FOE-cul seg-MEN-tal glo-MER-u-low-scle-ROW-sis>

**Proteinuria:**
<proh-tee-NUR-ree-uh>

**Podocyte:**
<POD-oh-site>

**Nephrotic:**
<neh-FROT-ik>

**Edema:**
<ih-DEE-muh>

**Idiopathic:**
<id-ee-oh-PATH-ik>

**Angiotensin:**
<an-jee-oh-TEN-sin>

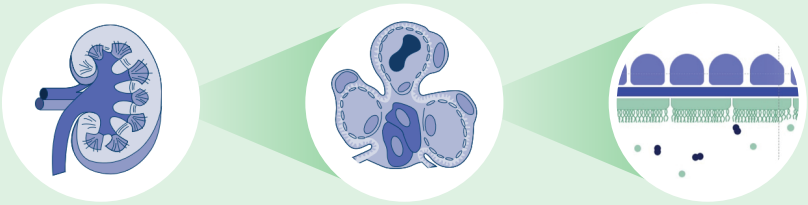
## How does FSGS affect the kidney?

Within the glomeruli are blood vessels and cells called podocytes. In a healthy kidney, podocytes form a barrier between the blood vessels and the urine.

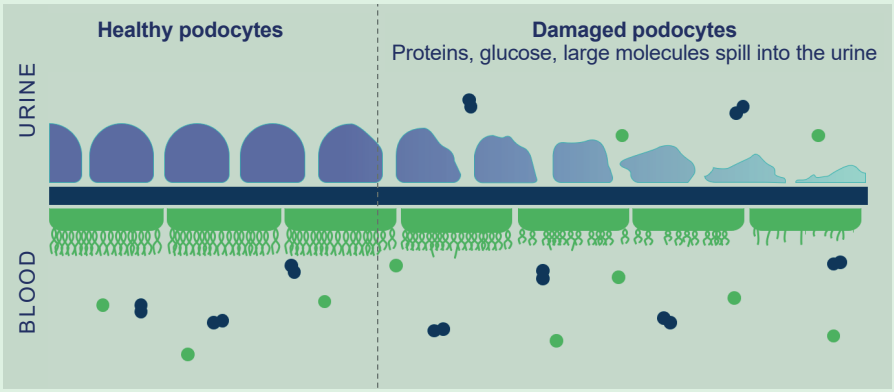
The kidney

A glomerulus – the filter unit of the kidney

Podocytes – cells that make up part of the filtration barrier that prevent proteins and blood from going into the urine



These cells are damaged in FSGS. This allows proteins, sugar, and other substances to pass from the blood into the urine.




## Who does FSGS affect?

Globally, FSGS affects **1 in a million children** and **8 in a million adults** each year

Among people with nephrotic syndrome (a collection of symptoms that show kidney damage), FSGS is the cause in up to **20% of children** and **40% of adults**

FSGS is more common in people of African ancestry

**Numbers of people affected by FSGS are RISING,** but no one knows why

## What are the most common symptoms of FSGS?

- Foamy urine
  Protein in the urine
- Swelling in the body (edema):
  Especially eyes, hands and feet, and abdomen
- High blood pressure
- Low blood albumin
  High cholesterol
  High creatinine (a waste product produced by muscles)

## How is FSGS diagnosed?

### Kidney biopsy

- Biopsy means taking a small piece of tissue from the kidney. This is done to see if the glomeruli have been damaged
- As FSGS only affects some sections of the kidney, biopsies can be unclear or miss the areas of FSGS
- Biopsies should sample several different glomeruli to make sure they are as accurate as possible

### Blood and urine tests

- Other blood and urine tests look at how well the kidney is working
- These include tests to measure how much protein is leaking into the urine

## FSGS is divided into different types, depending on the cause:

### Genetic FSGS

- Caused by abnormal genes that are inherited from a parent
- Also called familial FSGS, as it can run in families

### 1 Primary FSGS

- This type of FSGS happens on its own and has no clear cause
- Also called idiopathic FSGS, which means that the cause is unknown

### 2 Secondary FSGS

This type of FSGS has a known cause, such as:

- Viral infections
- Certain medications
- A type of cancer affecting immune cells, called lymphoma
- Changes to the kidney caused by other conditions or surgery

## How is FSGS treated?

**Currently, there are no medicines specifically approved to treat FSGS**

Treatment aims to reduce protein loss into the urine. **People receive combinations of medicines based on the cause of FSGS, their age, and any other medical conditions they may have such as diabetes or obesity.**

Medicines that help protect the kidney work on a system in the body called the "renin-angiotensin system". These types of medicines are called **angiotensin-converting enzyme (ACE) inhibitors** and **angiotensin II receptor blockers (ARBs)**. They are sometimes given with medicines that suppress the immune system, including steroids such as prednisone.

## TALK TO YOUR DOCTOR

**if you have any questions about FSGS or your treatment**



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