Focal Segmental Glomerulosclerosis

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What is FSGS?

FSGS is a disease of the kidney. It can only be diagnosed by a kidney biopsy. Its name describes what is seen in the kidney:

Focal: Not all of the glomeruli (filtering units) are involved; scarring in even 1 can make the diagnosis.

Segmental: Scarring involves only a portion of the filtering unit. If the whole glomerulus were involved, the scarring would be global.

Glomerulusclerosis: This is the medical term for scarring of the filtration units (glomeruli) of the kidney.

Other findings may be present on the kidney biopsy that will help predict how your kidneys are going to do.

FSGS usually causes protein in the urine (proteinuria). This may be mild enough that it was noted on a routine urine test. FSGS commonly causes heavy proteinuria that produces swelling from the nephrotic syndrome.

No one knows what causes FSGS. There is nothing that could have prevented it. While it may respond to treatment, many cases do not. About half of children with FSGS will develop permanent kidney failure within 5 years of their diagnosis of FSGS.

FSGS is more common now than it was 30 years ago, perhaps because of rising rates of childhood obesity.

How do you diagnose FSGS?

FSGS is diagnosed by kidney biopsy in patients with unusual forms of nephrotic syndrome or heavy proteinuria with no obvious cause.

Some patients with FSGS have a protein in their blood that causes the kidney to leak protein.

Other patients may have gene mutations that contribute to the disorder. You may be asked to have blood tests to examine these possibilities.

How do you treat HUS?

The nephrotic syndrome can be treated with diuretics (water pills) such as hydrochlorothiazide (Diuril) or furosemide (Lasix) to control swelling. If very severe, a protein called albumin may be given through the vein with furosemide to remove swelling. While these may control the swelling, they do not treat the FSGS itself.

Some cases of FSGS will resolve with treatment with oral steroids (drugs such as prednisone). Often this does not work and other agents are used including chemotherapy drugs such as cyclophosphamide and immunosuppressant drugs such as cyclosporine (Neoral™) or tacrolimus (Prograf™). Control of high blood pressure is also very important in preventing kidney failure. Drugs such as ACE inhibitors or others may be used to control this complication.

Some newer drugs may produce remission in patients with FSGS. These include mycophenolate (Cellcept™), another drug that suppresses the immune system. Another drug, rituximab (Rituxan™), can be given by IV. It has produced long-lasting remissions in patients with FSGS, even when the disease occurs after transplant. This medication may not be covered by insurance at this time.

In about half of all patients, none of these treatments work to cure the FSGS. In this case, blood pressure medications are very important to keep the kidneys working as long as possible. Other drugs may be needed to control swelling and the fat content of the blood.

If the kidneys do fail, a kidney transplant can be done. Unfortunately, FSGS may affect the transplanted kidney and cause it to fail. This does not happen every time, and when it does there are therapies that may help delay or reverse FSGS in the transplant.