

Focal segmental glomerulosclerosis (FSGS) and IgM nephropathy

Overview



infoKID

Information for parents and carers about children's kidney conditions

This leaflet has an overview of this topic.

Go to www.infoKID.org.uk to find out more about this topic, or to find other topics for parents and carers about children's kidney conditions.

This topic is about the conditions focal segmental glomerulosclerosis (FSGS) and IgM nephropathy, which cause nephrotic syndrome. This causes swelling in the body, especially in their face, legs and feet.

The main treatment for nephrotic syndrome is medicines called steroids. But in some children, the nephrotic syndrome may keep coming back. Your doctor may arrange some tests to find out if the nephrotic syndrome is caused by FSGS or IgM nephropathy – children with these conditions need further treatment.

The information covers:

- what FSGS and IgM nephropathy are, and how they lead to nephrotic syndrome
- how your doctor can find out whether your child has FSGS or IgM nephropathy
- treatment that your child needs
- long-term effects of FSGS and IgM nephropathy.

You may also wish to read the infoKID topic **Nephrotic syndrome**, which covers symptoms and signs, tests and diagnosis, and early treatment of nephrotic syndrome. Go to www.infoKID.org.uk



About nephrotic syndrome

There are different types of nephrotic syndrome. About 9 in 10 children with nephrotic syndrome have a type that can be treated with steroids – this is called **steroid-sensitive nephrotic syndrome** (SSNS) (it is “sensitive” to steroids).

In some children, the nephrotic syndrome keeps coming back after treatment – this is called **frequently relapsing nephrotic syndrome**.

About FSGS and IgM nephropathy

About 1 out of 10 children with nephrotic syndrome have a type that is caused by FSGS or IgM nephropathy. These conditions affect the kidneys' tiny filters called **glomeruli**.

In focal segmental glomerulosclerosis (FSGS), some of the glomeruli are damaged, and have tiny scars. The name describes how the glomeruli are scarred:

- **focal** – some, not all, of the glomeruli are affected
- **segmental** – a segment (part) of each glomerulus is scarred
- **glomerulosclerosis** – there is scarring on the glomeruli (“sclerosis” means scarring).

IgM is short for **immunoglobulin M**. This is a type of protein that the body makes to fight infection. In IgM nephropathy, this protein gets trapped in the glomeruli.



Symptoms and complications

Symptoms and signs

The symptoms and signs are similar for all types of nephrotic syndrome. The main symptom is swelling or puffiness in different parts of the body, especially around the eyes, legs and feet. This is called **oedema**.

The swelling happens because the kidneys leak a lot of protein in the urine (which is called **proteinuria**). You cannot usually see the protein, though it can be found by a simple **urine test**.

In FSGS and IgM nephropathy, there may also be:

- blood in the urine (**haematuria**) – you cannot always see the blood, but if there is a lot, the urine may be coloured red or dark brown (like blackcurrant squash or a cola drink)
- urinating less often or passing smaller amounts
- rarely, other symptoms such as a very swollen tummy or breathlessness.

Complications

Complications, or further problems, are rare. Because of the treatment, some children are more likely to get infections.

→ If your child has any of the below contact your doctor:

- severe pain in his or her tummy
- fever (temperature above 38°C) – your child may sweat more than usual or be shivery.
- swollen, red or painful leg – this may be a sign of a **blood clot**.

Causes

FSGS and IgM nephropathy are rare. About 1 in 50,000 children get nephrotic syndrome each year, but only about 1 in 10 of these cases are due to these conditions. Occasionally, FSGS can be caused by a **genetic mutation** that may run in families. **Genetic mutations** are problems in the genes (which are in each of our body's living cells), which are passed on by the parents.

It is not always known why some children get FSGS or IgM nephropathy. It is often considered to be linked to a problem with the **immune system**, which is the body's way of protecting itself from infection.

Some children who have had frequently relapsing nephrotic syndrome can develop FSGS or IgM nephropathy after many years.

Tests and diagnosis

If your doctor thinks that your child has FSGS or IgM nephropathy, he or she may be referred to a **paediatric renal unit**, a special unit for children with kidney conditions, which may be in a different hospital to your own. A **paediatric nephrologist**, a doctor who treats children with kidney problems, can advise on further tests or treatment.

Your doctor may suspect FSGS or IgM nephropathy if your child has symptoms of nephrotic syndrome *and*:

- has visible blood in his or her urine (**haematuria**)
- is urinating less often or smaller amounts
- has high blood pressure (**hypertension**)
- blood tests show that his or her kidneys are not working properly
- has a disease in another part of his or her body, such as the skin or joints of the arms and legs
- is a baby, infant or teenager, or
- has nephrotic syndrome that keeps coming back (**frequently relapsing**) in spite of taking extra medicines.

A **kidney biopsy** can confirm FSGS or IgM nephropathy. A tiny piece of one kidney is removed from the body with a needle and examined under microscopes. Special medicines are used so your child does not feel any pain or can sleep through the procedure.

If your doctor thinks your child has FSGS due to a **genetic mutation**, you may be referred to genetic testing and counselling.

Treatment

In FSGS or IgM nephropathy, the symptoms of nephrotic syndrome cannot be treated by steroids alone. Your child may need to take stronger **immunosuppressant** medicines, which make the immune system less active so it does not harm the kidneys.

Some children need to take medicines to prevent or treat infections, or to treat complications.

→ It is important that your child follows any treatment plan outlined by your doctor.

Monitoring

You will need to test your child's urine for protein every morning while your child is on steroids or immunosuppressants. A nurse will show you how to do this.

- If there is very little or no protein in the urine and the swelling goes down, the condition has gone into **remission**.
- If there is protein in the urine again, this is a **relapse**. You will need to contact your doctor, and your child may need another course of steroids or immunosuppressant medicines.

About the future

Follow up

All children with FSGS or IgM nephropathy will need to go back to the hospital for follow-up appointments to check for any health problems.

Long-term effects

Your healthcare team will speak with you and your child about any long-term effects your child might have with FSGS or IgM nephropathy. Some children have no long term problems with their **kidney function** (how well their kidneys are working). If the condition goes away, this is called **remission**.

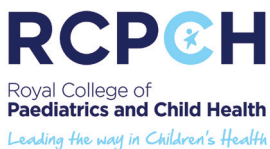
In some children, the kidneys stop working as well as they

should – this can happen slowly, over many years. This is called **chronic kidney disease** (CKD). These children will need to go back to the hospital or clinic for regular follow-up appointments. You and your child will learn more about this condition over time and about what to expect and how to help manage the condition.

» [Read more about FSGS and IgM nephropathy on www.infoKID.org.uk](http://www.infoKID.org.uk)

Your notes and contact information

www.infoKID.org.uk



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For details on any sources of information used in this topic, please contact us through our website at www.infoKID.org.uk.

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