

# Focal segmental glomerulosclerosis (FSGS) and IgM nephropathy



infoKID

Information for parents and carers about children's kidney conditions

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Each topic starts with an overview followed by several sections with more information.

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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](http://www.infoKID.org.uk)



## Overview

### 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**.

» **More about symptoms and complications**

## 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.

» **More about causes**

# 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.

» **More about tests and diagnosis**

## 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.

» **More information about treatment**

## 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.

» **More about monitoring**

## 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.

» **More about the future**

## Symptoms and complications

### Common symptoms and signs

The main symptoms and signs for FSGS and IgM nephropathy are similar to those in other types of nephrotic syndrome.

#### Swelling in the body

Children have swelling or puffiness in different parts of their body – this is called **oedema**. The oedema is normally around the eyes in the morning, and in the legs and feet later in the day. After a while there may be swelling throughout the day. Boys may also have oedema of the scrotum.

It is **pitting oedema**. This means that when the affected area is gently pushed, there is a small dent for a few moments.

→ **If your child has swelling in the body, seek medical advice as soon as possible.**

In FSGS and IgM nephropathy, the swelling happens because there is too much fluid in the body's soft tissues.

This is caused by the kidneys leaking a lot of protein, which is normally in the bloodstream, into the urine. An abnormal amount of protein in the urine is **proteinuria**. You cannot usually see the protein. Your child's doctor can find proteinuria on a urine test.

A lot of the protein that is leaked is a type called **albumin**. Albumin helps keep fluid in the blood. Fluid moves between the blood and the body's soft tissues, and when there is not enough albumin in the blood, fluid stays in the soft tissues.

#### Problems urinating

Some children urinate less often or smaller amounts.



A child with swelling around her eyes

#### Blood in the urine

Some children also have **haematuria** (blood in their urine), which happens when **red blood cells** leak through the kidney's filters into the urine. Sometimes you cannot 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).

### Rare symptoms and signs

#### Swelling in the tummy and breathlessness

- A few children get a large swelling in their abdomen (tummy area). This is called **ascites**. It happens when fluid builds up in the area around the organs in the abdomen – the **peritoneal cavity**.
- A very small number of children feel breathless. This happens when fluid builds up in the area around their lungs.

→ **If your child has FSGS or IgM nephropathy and a very swollen tummy or feels breathless, contact your doctor as soon as possible.**

#### Other rare symptoms

The symptoms below are occasionally found in children with FSGS or IgM nephropathy:

- tiredness, low energy or difficulty concentrating
- decreased appetite (not wanting to eat)
- nausea (feeling sick), vomiting (being sick), or diarrhoea
- pain in the abdomen (tummy)
- headaches that keep coming back or that do not go away
- pain, stiffness or swelling of the joints.

## Complications

Some children have complications – health problems that happen because of the condition or its treatment. These are very rare but are more likely to happen if the oedema lasts a long time.

### Risk of infections

- Children with FSGS and IgM nephropathy are treated with medicines that suppress, or damp down, their immune system, which normally protects them from infections.
- If your child is unwell and you are worried about an infection, contact your doctor straight away.

## Other complications

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

- severe pain in his or her tummy and/or a fever (temperature above 38°C) – this may be a sign of an infection called peritonitis
- swollen, red or painful leg – this may be a sign of a blood clot
- headaches, vomiting or blurred (fuzzy) vision – this may be a sign of hypertension (high blood pressure)
- feeling more tired than usual, having sleep disturbances such as difficulty getting to sleep, becoming more forgetful or gaining weight – this may be a sign of low levels of thyroid hormone

### Complications: Read more about complications

#### Peritonitis (tummy pain and fever)

Peritonitis is inflammation (like a swelling) of the **peritoneum**. This is a thin layer that lines the inside of the abdomen (tummy area). It surrounds and supports organs such as the stomach and liver.

Peritonitis is caused by a bacterial infection. Children with peritonitis have severe pain in their tummy and a fever (temperature above 38°C), and may sweat and/or seem shivery.

#### Thrombosis (blood clot)

In nephrotic syndrome, blood may be thicker than normal. This happens when some of the water in the blood leaks out of the blood vessels. The blood is more likely to form a **blood clot** (like a clump of hardened blood).

If a blood clot is large enough to block a vein, it can cause **thrombosis**. The leg, or occasionally another part of the body, may look swollen and may look red and feel painful.

#### Hypertension (high blood pressure)

**Hypertension** is **blood pressure** that is too high. In some children, this leads to headaches, vomiting or blurred (fuzzy) vision. Hypertension that lasts a long time can also increase the risk of getting other diseases in adulthood.

If your child has hypertension, he or she may need to make changes to his or her diet, or take medicines to get their blood pressure in a healthy range.

#### Low levels of thyroid hormone

The **thyroid** is a gland in the neck that makes thyroid hormone. This **hormone** (a chemical that is carried in the blood to send messages to other parts of the body) controls how the body uses energy.

Some children with nephrotic syndrome have low levels of thyroid hormone in their body – this is called **hypothyroidism** (“hypo” means low or not enough). This causes many of the body’s functions to slow down. Some children with hypothyroidism may:

- be more tired than usual or have sleep disturbances such as difficulty getting to sleep
- become more forgetful
- gain weight
- have dry hair, skin and nails
- have changes in mood (more sad than normal)
- be constipated (difficulty doing a poo)
- feel breathless
- feel sensitive to cold
- get cramps in their muscles, or feel weak.

#### Diarrhoea

A few children have swelling of the lining of their gut. This can cause diarrhoea.



## High levels of fat in the blood

Rarely, children with FSGS and IgM nephropathy have too much of some fats in their blood – even when they eat a healthy diet and are active. This is more likely in children who have proteinuria for long periods of time.

## When the kidneys stop working suddenly

In serious cases, the kidneys stop working as well as they should, over a short period of time – this is called **acute kidney injury (AKI)**. Some children get better after a few weeks, and some need to take medicines or have more intensive treatment.

### Complications: Read more about high levels of fat in the blood

Nephrotic syndrome can lead to a high level of fats in the blood, which is called **hyperlipidaemia** (“hyper” means too much). These fats are made in the body and are in many foods. Two types of fat are:

- **triglycerides** – a high level of these is called hypertriglyceridaemia
- **cholesterol** – a high level of this is called hypercholesterolaemia.

Eating a healthy diet and being active usually helps keep these fats at a healthy level. But in nephrotic syndrome, there is a lower level of some proteins, especially **albumin**, in the blood because they are leaked into urine. This causes the levels of fats to increase.

## Causes

**FSGS and IgM nephropathy lead to nephrotic syndrome – the kidneys leak a lot of protein into urine. This leads to a drop in the levels of protein in the blood, which causes swelling in the body (oedema).**

» [Read more about how the kidney works and protein leak on the next page](#)

### Who gets FSGS and IgM nephropathy?

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.

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

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.

### The immune system

Many germs – including bacteria and viruses – can make us sick if they get into the body. The immune system can kill these germs. However, if the immune system is not working properly, it can start to cause problems.

» [Read more about the immune system on the next page](#)

### Other causes

Some cases of FSGS and IgM nephropathy are due to other reasons, though these are more rare.

- **Genetic mutation** – some children have a genetic mutation that affects the structure of part of the glomerulus in the kidney, which causes the protein leak. Genetic mutations are problems in the genes (which are in each of our body’s living cells), which are passed on by the parents. Your doctor will talk to you about this, and may arrange for a genetic test to be done.

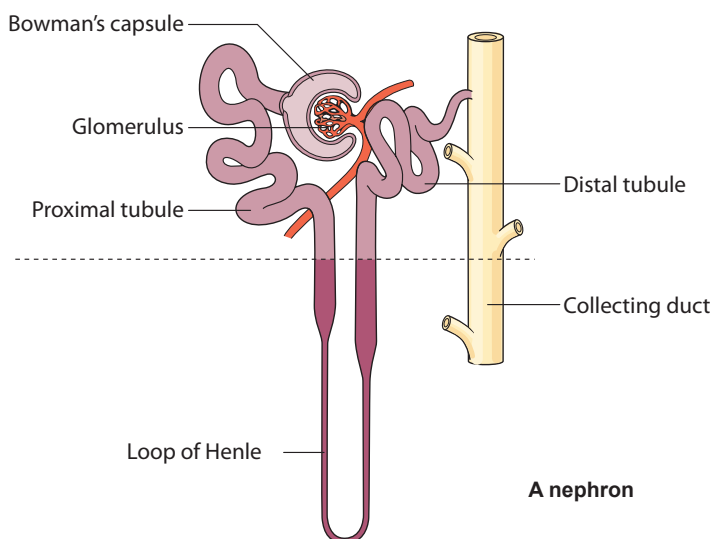
### Will it affect other family members?

It is very rare for FSGS, and even rarer for IgM nephropathy, to run in families. Your child’s doctor will ask about any family history of these conditions, and it is usual to arrange for siblings to have a urine test to check for protein.

### How the kidney works

Inside each kidney, there are about one million **nephrons**. Each nephron is made up of a **glomerulus** (when we talk about more than one glomerulus, we say glomeruli), and a **renal tubule**.

- Each glomerulus acts like a sieve, helping to remove extra water and waste from the body, and holding on to blood cells and protein, which the body needs.
- Blood flows into the kidneys and to each glomerulus.
- Most of the water and some other substances in the blood pass through the glomeruli.
- This liquid flows into the renal tubule. Most of this liquid moves back into the bloodstream. The rest of it becomes urine.
- The urine leaves the kidney by the ureters and goes into the bladder, where it is stored until we are ready to go to the toilet.



### » More about what the kidney does

#### Protein leak

Protein is an important part of our diet and is in most foods. When we eat protein, it is digested (broken down) in the stomach and intestines and taken into the blood. It is important that we have the right amounts of protein in our body to stay healthy.

Normally, most protein stays in the blood and does not get removed by the kidneys into the urine. This is mostly because proteins are too big to pass through the glomeruli.

In nephrotic syndrome, there is a change in the glomeruli. They become more leaky, and so some proteins pass through, leaving the body in urine.

The main protein that leaks out is **albumin** which is quite a small protein and more likely to leak through the glomeruli.

## Causes: Read more about the immune system

The immune system protects the body against germs such as bacteria and viruses that can cause illness. These germs can enter the body in lots of ways, such as by the nose and throat or the urinary system. If we get a cold or flu, this means that a virus germ has got into the body and started to infect some of our body's cells.

### Germs

Germs have special 'markers' that are different from the markers on our own body's cells. This means that the immune system can recognise that they are germs and kill them. We often feel sick for a few days or a few weeks while this is happening.

### Parts of the immune system

The immune system has many different ways to protect the body against disease. Some of the parts include the below.

- **White blood cells** are living cells in the blood. Often, the number of white blood cells found in a blood test can give information about someone's immune system. The two main types of white blood cells in the immune system are **neutrophils** and **lymphocytes**.
- **Antibodies or immunoglobulins** recognise the germs that have come into the body, and can bind (stick) to them. There are five types: immunoglobulin A (IgA), immunoglobulin D (IgD), immunoglobulin E (IgE), immunoglobulin G (IgG) and immunoglobulin M (IgM).
- The **complement system** is made up of other proteins that float in the blood. These proteins work with (complement) other parts of the immune system to help kill germs or cells infected by germs. Normally the body controls when complement is activated, so it does not attack the body itself.

### When the immune system does not work properly

Sometimes the immune system does not work as expected and can cause problems. For example, sometimes the immune system thinks that the body's own cells are invaders like germs, and may attack them.

# Tests and diagnosis

## Diagnosing nephrotic syndrome

Your doctor may have diagnosed (identified) nephrotic syndrome before suspecting FSGS or IgM nephropathy.

» **To find out how nephrotic syndrome is first diagnosed, go to Nephrotic syndrome – steroid-sensitive: Tests and diagnosis.**

## When FSGS or IgM nephropathy is suspected

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

- has visible blood in his or her urine (**haematuria**) – the urine may be coloured red or dark brown (like blackcurrant squash or a cola drink)
- is urinating less often or smaller amounts
- has high blood pressure (**hypertension**)
- blood tests show that the 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.

## Tests for FSGS or IgM nephropathy

Your child will need further tests to find out more about his or her health, the effect of any medicines and to make a diagnosis. These tests may include:

- an examination – for example, to check for early signs of **oedema** (swelling of the body)
- **blood pressure** measurements – to check for **hypertension**
- **urine tests** – to check the urine for protein and other substances such as blood cells
- **blood tests** – to check the blood for levels of protein, how well the kidneys are working, and the levels of medicines your child is taking

- a **genetic test** and **genetic counselling** – to check if your child has a faulty gene (genetic mutation).. Genetic testing usually involves getting a sample of blood or body tissue, which can be checked for a specific gene. **Genetic counselling** is a service that can give you information and guidance about conditions caused by genetic mutations

- kidney biopsy
- imaging tests (scans).

### Kidney biopsy

A **kidney biopsy** can usually confirm whether a child has FSGS or IgM nephropathy or any other abnormalities. It can also give more information about how much damage there is in the kidney.

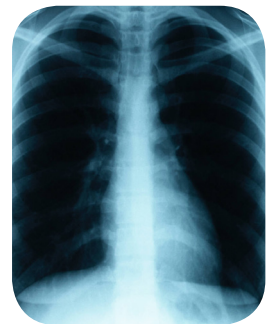
A tiny piece of one kidney is removed from the body with a needle – special medicines are used so your child does not feel any pain or can sleep through the procedure. The sample is sent to a laboratory where it is looked at under microscopes. It can also be checked for proteins related to the immune system.

### Imaging tests

**Imaging tests** use special equipment to get images (pictures) of the inside of your child's body.

- **Ultrasound scan** – looks at the shape and size of kidneys and other parts of the urinary system. A small handheld device is moved around your child's skin and uses sound waves to create an image on a screen.
- **Chest x-ray** – for children with breathing problems, this test checks for any fluid (liquid) around the lungs. Your child sits or lies still for a few seconds while a machine takes x-ray images.

Occasionally, other imaging tests are needed.



A chest X-ray of the lungs

# Treatment

In FSGS or IgM nephropathy, the symptoms of nephrotic syndrome usually cannot be treated by steroids alone. Your child may need to take stronger immunosuppressant medicines, which reduce the effect of the immune system on the kidneys.

Some children need treatment for any complications – to prevent or treat infections, reduce oedema (swelling in the body) or reduce the levels of fats in the blood.

## Where will my child be treated?

Once your child is diagnosed with **FSGS** or **IgM nephropathy** – usually after a **kidney biopsy** – your doctor will decide on the best treatment.

### Treatment: Read more about what happens in hospital

A team of healthcare professionals will:

- check your child's kidney function (how well his or her kidneys are working)
- measure how much your child is drinking and how much urine he or she is passing
- test your child's **urine** with a dipstick
- check your child's **blood pressure**
- weigh your child to help find out if he or she has too much or too little fluid in their body

#### Healthcare team

Your child's healthcare team may include:

- **paediatrician** – a children's doctor
- **paediatric nephrologist** – a doctor who treats children with kidney problems
- **renal nurse** – a nurse who cares for children with kidney problems
- **paediatric dietitian** – a professional who advises what your child should eat and drink during different stages of their kidney condition
- **renal social worker** – a professional who supports you and your family, especially with any concerns about money, travel and housing related to looking after your child with kidney disease
- **renal psychologist** – a healthcare professional who supports your child and family, especially with emotional stresses and strains from having or looking after a child with kidney disease
- **play specialist** – a professional who uses dolls and other toys to help your child prepare for procedures, such as blood tests and dialysis

## Steroids and immunosuppressants

### Steroids

The first treatment for nephrotic syndrome is a steroid medicine, usually prednisolone. Steroids are chemicals that are made naturally in the body, but can also be made as medicines. They suppress the immune system, or make it less active. Your doctor will give you more information about how and when your child should take the medicine.

→ It is important that you continue giving the steroids to your child as your doctor has told you, even if your child is getting better. Stopping steroids suddenly can make your child very unwell, so your child should only stop taking steroid medicines on the advice of your doctor.

### Immunosuppressants

However, children whose nephrotic syndrome is caused by FSGS or IgM nephropathy may need stronger medicines that are known as **immunosuppressants**. There are different types of immunosuppressants, and your child may need to take more than one of these medicines, sometimes in combination, to try to treat the condition. They include:

- **ciclosporin** – also known as cyclosporin
- **tacrolimus**
- **mycophenolate mofetil** – also called MMF.

More rarely, the medicines include:

- rituximab
- vincristine
- cyclophosphamide
- chlorambucil.



## Vaccines

It is important that your child has the **vaccines** (immunisations) that he or she needs to reduce the risk of some diseases. Speak with your doctor about these – some vaccines are not safe to give while a child is taking steroids or immunosuppressants.

→ It is important that your child has the flu vaccine each year.

## Infections

Children taking these medicines are more likely to get infections. Speak with your doctor or nurse, and your child's teacher or daycare manager, about how you can help prevent infection.

Contact your doctor straight away if your child:

- has been in contact with someone who has chicken pox (if they have not had this illness before) or measles (if they have not had the MMR vaccine, which protects against measles, mumps and rubella); or
- is unwell and you are worried about an infection.

If your child does get an infection, he or she may need to take an **antibiotic** medicine, which kills the germs that cause infection. Your child must take the medicine for the number of days that the doctor has told you, or until all of the medicine has been taken.

## Side-effects

We use medicines to help children get better, but sometimes they have effects we do not want – **side-effects**. Contact your doctor straight away if your child has any of the following while on steroid medicines:

- fever (temperature above 38°C), with a sore throat or a cough
- a rash or severe bruising
- bad stomach pain or repeated vomiting – sometimes steroids can cause ulcers in the stomach.

» **More about side-effects of steroids in Nephrotic syndrome**

## Risk to kidney

Although we use these medicines to try to treat nephrotic syndrome, some, especially tacrolimus and ciclosporin, may have an effect on kidney function (how well the kidneys work). Your child's healthcare team will closely monitor the levels of these medicines with regular blood tests.

## Changes to eating and drinking

Your child may need to make temporary changes to what he or she eats. For example, he or she may need to eat less salt while they have oedema or are taking steroids. This is because higher levels of salt in the body cause the kidneys to keep more water.

Your doctor, nurse or dietitian will help you and your child make these changes.

## Treating complications

Some children need further treatment for complications, such as:

- preventing or treating infections, such as **peritonitis**, which affects the area inside their abdomen (tummy) – your child may need to take **antibiotics**, medicines that kill bacteria
- reducing oedema
- reducing the level of fats in their blood
- treating low levels of thyroid hormone.

## Managing reduced kidney function

For some children, the kidneys do not work as well as normal during this illness – they have **reduced kidney function**. Most children do not need any special treatment, but will need long term follow-up because kidney function may slowly get worse over years.

A small number of children with FSGS, and a very small number of children with IgM nephropathy will have more serious kidney problems and will need further treatment. This may include a period of time on **dialysis**, which uses special equipment to 'clean' the blood, removing waste products and extra water and salts.

## Questions to ask the doctor or nurse

- What treatment will my child need?
- How will the treatment help my child?
- How long will my child be in hospital?
- How can I help my child prepare for procedures and treatments?
- If the first treatment does not work in my child, what happens next?
- How will I know if we need to go back to the hospital or see the doctor?
- Will the conditions come back? How will I know?

### Preventing or treating infection

- Your child may be at higher risk of getting infections when he or she is being treated for nephrotic syndrome. This is because the medicines suppress the immune system, so it cannot work as effectively to fight germs, and the oedema lets germs grow more easily. One important infection is **peritonitis**, which affects the **peritoneum**, the area around the organs in the abdomen (tummy area).
- Antibiotics are medicines that kill bacteria. Your child may need to take antibiotics to prevent or treat infection – this is often the medicine **penicillin**.
- To prevent infections, you may need to give a small amount of antibiotic twice a day while your child has oedema.
- If your child gets an infection, you may need to give a full course of this or another antibiotic to treat it.

→ It is important that you follow your doctor's instructions about when and how much to give.

» Medicines for Children – Penicillin V for prevention of pneumococcal infection

» Medicines for Children – Penicillin V for bacterial infection

» Medicines for Children – General advice about antibiotics

### Reducing oedema

Occasionally children with these conditions need to take **diuretics**. These medicines help reduce oedema by removing extra water and salt from the body, and by increasing the amount of urine that is passed. Your doctor will examine your child for oedema and measure your child's weight while they are taking diuretics, to check how well the medicine is working.

Common diuretics are: furosemide, spironolactone and metolazone. Other diuretics that may be used are: amiloride, bendroflumethiazide and chlorothiazide.

### Controlling levels of fat in the blood

Some children need medicines to treat **hyperlipidaemia**, or high levels of fats, including cholesterol and triglycerides. Some children who have high levels of protein in their urine for a long time may be at risk of this.

Some medicines include:

- **statins** – which reduce the amount of cholesterol the body makes
- fish oil supplements, such as MaxEPA – because some types of fish are thought to help reduce the level of fats.

### Low levels of thyroid hormone

The thyroid is a gland in the neck that makes thyroid hormone. This hormone (a chemical that is carried in the blood to send messages to other parts of the body) controls how the body uses energy.

Some children with nephrotic syndrome have low levels of thyroid hormone in their body – this is called hypothyroidism ("hypo" means low or not enough). This causes many of the body's functions to slow down. Some children with hypothyroidism may:

- be more tired than usual or have sleep disturbances such as difficulty getting to sleep
- become more forgetful
- gain weight
- have dry hair, skin and nails
- have changes in mood (more sad than normal)
- be constipated (difficulty doing a poo)
- feel breathless
- feel sensitive to cold
- get cramps in their muscles, or feel weak.

# Monitoring

## Testing urine at home

While your child is on steroids or immunosuppressants, you will need to **test your child's urine** for protein every morning. Your doctor will let you know how often to test the urine and when you can stop.

### When to do the urine tests

Some children, even when they do not have nephrotic syndrome or another kidney problem, have a very small amount of protein in their urine after standing or sitting upright for a few hours. For this reason, it is best to get a sample of your child's **first morning urine**. This is the urine that your child passes when they first wake up in the morning.

### How to do the urine tests

You will be given **dipsticks** – these are strips with chemical pads that change colour depending on what substances are in the urine. A nurse will show you how to use them.

Older children can urinate over the dipstick. For younger children, you can get a sample of urine from a potty.

- Put the dipstick in the urine sample, remove it and read the result. Use the instructions included in the package.
- Record the result in a diary – make sure you include:
  - the date
  - the urine test result
  - any medicines taken (e.g. steroid and dose, or amount)
  - any comments about your child's health (e.g. feeling unwell)
- Bring the diary to your next clinic visit.

## Remission

If the urine dipstick shows either "trace" or no protein for three days in a row, and the swelling goes down, this means that the condition has gone into remission.

Most children go into remission within 6 months of starting the immunosuppressants. Some have a lower level of proteinuria – this is called **partial remission**.

## Relapse

If the urine dipstick shows either 3+ or 4+ protein for three days in a row, this means that there has been a **relapse**. The kidneys have started leaking protein again, and the symptoms may come back.

Daily testing of your child's urine will allow you to quickly detect a relapse. Your child will be able to start treatment, often without needing to go into hospital. If the urine tests are not used and your child has a relapse, they may become unwell and develop oedema, and are more likely to need to go into hospital.

→ **If the dipstick shows a relapse, contact your doctor as soon as possible. Your child will need another course of steroids or other immunosuppressants.**

## Measuring blood pressure at home

You may be asked to measure your child's blood pressure, using a blood pressure monitor that you can get from your doctor or a pharmacy.

» **More about measuring blood pressure at home in Blood pressure**

# About the future

Your healthcare team will speak with you and your family about any long-term effects your child might have with FSGS and IgM nephropathy. Your child will have follow-up appointments and you may need to monitor him or her at home.

Some children fully recover from these conditions – when the symptoms go away, this is called remission. Others may experience relapses – when the kidneys start leaking protein again and the symptoms reappear. These may be triggered by an infection, such as a cold.

A few children have long-term problems with their kidneys.

## Follow-up appointments

Your child will need to have follow-up appointments at the hospital or clinic to check for any health problems.

It is important to go to these appointments, even if your child seems well – remember to bring the results from your home urine testing. You will also have the opportunity to ask any questions. At these appointments, your child may have:

- his or her height and weight checked
- a physical examination
- **urine tests** – to check for blood, protein and other substances in his or her urine
- **blood tests** – to check for the amount of protein and other substances in his or her blood, and measure his or her kidney function
- his or her **blood pressure** measured.

## Long-term effects

### Remission and partial remission

Most children with FSGS and IgM nephropathy will go into remission within 6 months of starting the immunosuppressant medicines. Of those children who do not go into remission, some will have less proteinuria – this is called partial remission.

### Frequent relapses

Most children have at least one relapse. A few children have frequent **relapses** – this is called **frequently relapsing nephrotic syndrome** – and may need further treatment.

### Long-term problems

Some children with these conditions may develop a low level of **proteinuria** (an abnormal amount of protein in the urine).

Some children may have **hypertension** (high blood pressure), especially later in life.

A few children develop long-term problems with their kidneys – this may be due to the problems with the condition or be a result of the treatment, such as some of

the immunosuppressants that are needed. The kidneys stop working as well as they should. This is called **chronic kidney disease (CKD)**. A very small number of children need **dialysis** and **transplantation** as a result of FSGS. FSGS may recur (come back) in the transplanted kidney. Your healthcare team will talk to you about this.

If your child has CKD, you will learn more over time about how to help manage the condition, and what to expect.

## What you can do

### After treatment

Children who have been successfully treated for FSGS or IgM nephropathy can usually do the things that other children their age do. They should be able to continue going to school or nursery. They can play with other children and stay active.

### Living healthily

Your child can help protect his or her kidneys by leading a healthy lifestyle throughout their child and adult years. This includes:

- eating a healthy diet – with at least five servings of fruit and vegetables a day, taking care not to eat too much salt, sugar and fats (especially saturated fats)
- getting plenty of exercise
- not smoking.

## Further support

This can be a difficult and stressful experience for your child and the whole family, including other children. If you have any concerns or need additional support, speak with your doctor or nurse.

## Further information

This is the end of the information on FSGS and IgM nephropathy. If you would like to read more about other kidney conditions, tests and diagnosis, treatment or supporting information, you can find a list of topics covered on the infoKID website at [www.infoKID.org.uk](http://www.infoKID.org.uk).

## Your notes and contact information

[www.infoKID.org.uk](http://www.infoKID.org.uk)



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