

Nephrotic syndrome frequently relapsing

This infoKID topic is for parents and carers about children's kidney conditions. This leaflet has an overview only. Visit www.infoKID.org.uk to find more topics about conditions, tests & diagnosis, treatments and supporting information. Each topic starts with an overview followed by several sections with more information.

[Links to sections](#) in topic | [Other topics](#) available on website

In nephrotic syndrome, the kidneys leak too much protein into urine, leading to a drop in the levels of protein in the blood. This causes swelling in the body, especially in the face, legs and feet (oedema). Nephrotic syndrome is usually treated with a steroid medicine called prednisolone, and in most children the symptoms go away after a few weeks or a bit longer.

Many children have at least one relapse – when the symptoms come back after treatment, and they need to take steroid medicines again. Some children have relapses that happen very frequently. This is called frequently relapsing nephrotic syndrome. These children may need further treatment during their relapses, such as stronger medicines.

→ If this is the first time your child has nephrotic syndrome, please first read the infoKID topic: [Nephrotic syndrome](#)



Overview

About nephrotic syndrome

When children first develop nephrotic syndrome, they are usually admitted to hospital for monitoring and treatment. In many children, the steroid successfully treats the condition within a few weeks or a bit longer. Their kidneys stop leaking protein and their swelling goes down.

- This is called **steroid-sensitive nephrotic syndrome (SSNS)** because the nephrotic syndrome is 'sensitive to' (can be treated by) steroids.
- It may also be called **minimal change nephrotic syndrome** because there is no or very little change in the way the kidneys look.

In most children, the nephrotic syndrome comes back within a year – this is a relapse.

Frequently relapsing nephrotic syndrome

About half of children with SSNS have frequent relapses. This means that although the nephrotic syndrome gets better with steroids, it keeps coming back in a short space of time. It is called **frequently relapsing nephrotic syndrome** if this happens:

- two or more times in 6 months, or
- four or more times in one year.

Children with frequent relapses may continue to have them throughout childhood, though they tend to relapse less often as they grow into their teenage years. It is rare that SSNS continues to relapse in adulthood.

Monitoring for relapses

If your child has frequent relapses, you will need to test his or her urine for protein every morning. You will be given **dipsticks** – these are strips with chemical pads that change colour if there is protein in the urine. A nurse will show you how to use them.

Continue doing the urine tests even when your child is well, and keep a record of the results.

» More about urine testing at home

→ If the urine dipstick shows either 3+ or 4+ protein for three days in a row, the nephrotic syndrome has relapsed.

What happens next

By regularly testing your child's urine, you can quickly detect a relapse and your child can start treatment with steroids quickly, often without needing to go into hospital. If the urine tests are not used and your child has a relapse, he or she may become unwell and develop oedema, and is more likely to need to go into hospital.

Symptoms and complications

Oedema

The main symptom of nephrotic syndrome is **oedema** – swelling or puffiness in the body, especially around the eyes and in the legs and feet. It happens when there is too much fluid (water) in the body's soft tissues. This happens because the kidneys leak a lot of protein, which is normally in the blood, into urine. When there is more protein in the urine than normal, this is called proteinuria. There are different types of protein in the blood. The most common type that is leaked is a type called albumin. Albumin helps keep fluid in the blood. Fluid moves between the bloodstream and the body's soft tissues. When there is not enough albumin in the blood, fluid stays in the soft tissues.

However, if you detect a relapse with home urine testing and your child starts treatment soon after, he or she is less likely to develop oedema. If there is oedema, it will probably be less than the first time your child had nephrotic syndrome.

Complications

Complications – health problems that happen because of the nephrotic syndrome or treatment – are rare. They happen in a small number of children during the first episode of nephrotic syndrome or in relapses.

If your child has any of the below, especially in a relapse, contact your hospital doctor or seek medical advice:

- headaches, vomiting or blurred (fuzzy) vision – this may be a sign of hypertension (high blood pressure) – this may be a complication of taking steroids

- a large swelling of the abdomen (tummy). This is called ascites. It happens when fluid builds up in the area around the organs in the abdomen – the peritoneal cavity.
- feeling breathless – this can happen if fluid builds up in the area around their lungs
- severe pain in the tummy – which may be a sign of too little fluid in the blood supply to the bowels
- severe pain in the tummy and a fever (temperature above 38°C), and sometimes sweating more than usual or shivering – this may be a sign of an infection called peritonitis
- a swollen, red or painful leg – this may be a sign of a blood clot (when a clump of blood blocks a blood vessel).

Acute kidney injury

Rarely, the kidneys stop working as well as they should in relapses – this is called acute kidney injury (AKI). In nephrotic syndrome, this may happen when the body has a lower volume of blood than normal – this is called hypovolaemia. Many children with AKI get better after a few weeks, and some need to take medicines or have more intensive treatment.

» More about symptoms and complications in Nephrotic syndrome

Causes

Nephrotic syndrome happens in people of all ages. Steroid-sensitive nephrotic syndrome (SSNS) usually starts when a child is between 2 and 5 years old.

Many cases of nephrotic syndrome – including those that relapse often – are linked to the immune system, which normally protects the body against disease and infection.

» More about causes in Nephrotic syndrome

Tests

Your child will have had tests to **diagnose** nephrotic syndrome, including **urine tests**, **blood tests** and, sometimes, **imaging tests** (scans). Your child will not usually need further tests for relapses.

A **kidney biopsy** is not usually needed, but further into your child's treatment your doctor may recommend one to find out how much damage there is in his or her kidneys. This is especially important as some medicines that are used to treat frequently relapsing nephrotic syndrome may harm the kidneys.

A tiny piece of a kidney is removed from the body with a needle and examined under microscopes. Special medicines are used so your child will not feel any pain or can sleep through the procedure.

» More about tests and diagnosis in Nephrotic Syndrome – Steroid Sensitive: Tests and diagnosis



A child with swelling around his eyes

Treatment

→ It is important that you continue giving the medicines to your child as your doctor has told you, even if your child is getting better.

Referral to specialist services

Children with frequently relapsing nephrotic syndrome may be referred to a **paediatric renal unit** (a specialised unit that treats babies, children and young people with kidney conditions). They are looked after by a **paediatric nephrologist**, a doctor who treats children with kidney problems.

Steroids

If your child has frequent relapses, your doctor may prescribe a small 'preventer' dose of the steroid prednisolone, to be taken daily or every other day. Steroids **suppress** the immune system, or make it less active.

Prednisolone is available as tablets. Children who are unable to take medicines by mouth may be given a similar medicine called methylprednisolone, which is injected using a needle.

» [More about steroids - Giving steroids](#)

Immunosuppressants

If your child continues to have relapses, your doctor may prescribe other medicines known as **immunosuppressants**. There are different types of immunosuppressants, and your child may need to take more than one of these to try to treat the symptoms and prevent relapses.

Your doctor will work out the best medicines for your child. They may include:

- **levamisole**
- **mycophenolate mofetil** – also called MMF
- **cyclophosphamide**
- **ciclosporin**
- **tacrolimus**
- **rituximab**

Medicines that are more rarely used are:

- **rituximab**
- **vincristine**
- **chlorambucil**.

Side-effects of immunosuppressants

Immunosuppressants depress the immune system.

» [Read more about side effects of steroids on the next page](#)

Treatment: Read more about hospital visits and the healthcare team

Hospital visits

A team of healthcare professionals may:

- 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**.

Healthcare team

Your child's healthcare team may include a:

- **paediatrician** – a doctor who treats babies, children and young people
- **paediatric nephrologist** – a doctor who treats children with kidney problems
- **renal nurse** – a nurse who cares for children with kidney problems
- **renal dietitian** – a healthcare professional who advises what your child should eat and drink during different stages of a 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 to look 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
- **renal pharmacist** – some hospitals have a pharmacist who specialises in medicines for children and/or adults with kidney problems.

Risk of infection

Because these medicines work on the immune system, your child is more likely to get infections, or have more difficulty fighting off an infection. He or she may also recover less quickly from injury or after surgery.

→ Contact your doctor straight away if your child:

- has been in contact with somebody with measles or chicken pox (and has not had chicken pox before)
- are unwell and you are worried about an infection.

Risk for the kidneys

Although we use these medicines to try to treat nephrotic syndrome, some, especially tacrolimus and ciclosporin, may be harmful to the kidney. Your child will have regular

blood tests to check the levels of these medicines to try to avoid this. If they are on these medicines for a long time (more than 2 years), your doctor may do a kidney biopsy to check for kidney damage.

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 and in particular 'live' 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. The live attenuated influenza vaccine (Fluenz Tetra®) should not be given to children who are receiving high dose corticosteroids.

Steroids: Read more about side-effects

Side-effects you need to do something about

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, for more than 12 hours
- a rash or severe bruising
- bad stomach pain or repeated vomiting – sometimes steroids can cause ulcers in the stomach.

Other side-effects

Other side-effects that affect some children are below – speak with your doctor or nurse if you are concerned or need more support:

- nausea, vomiting, stomach pain and/or indigestion (heartburn) due to irritation to the lining of the stomach – your doctor may give your child some medicine to treat this
- behavioural problems – you may find that your child is more likely to have tantrums, be moody, feel depressed, have difficulty sleeping or have nightmares
- weight gain and larger appetite – you can help by making sure your child has lots of physical activity and eats fruits, vegetables and low-calorie food, rather than food that is high in calories (such as cakes, biscuits, sweets and crisps), and by reducing the portion size
- high blood pressure (**hypertension**) – your child's blood pressure will be regularly checked, and if it is too high, will need to be controlled by eating a diet with no-added salt.

There may, sometimes, be other side-effects that are not listed above. If you notice anything unusual and are concerned, contact your doctor.

Side effects with high doses or long courses

When steroids are needed over a long time, or in high doses, they may have other side-effects. If you have any concerns, talk to your doctor.

- All steroid medicines, including prednisolone, may affect the adrenal glands so that they produce less of a hormone called cortisol when the body is stressed (e.g. during illness or injury). This means that your child may have more difficulty fighting off an infection, or may recover less quickly from injury or after surgery. **If your child is unwell and you are worried about an infection, contact your doctor straight away.**
- Steroids can slow growth and affect puberty. They can also cause growth of body hair and irregular periods in girls. Your doctor will check your child's growth and development.
- Your child's skin may become thinner, and heal more slowly than usual. Acne (spots) may become worse, or your child may develop mouth ulcers or thrush (**candidiasis**).
- Your child may develop problems with their hip bones, or their bones may become weaker (**osteoporosis**). The muscles around their hips and shoulders may also become weaker. If your child has any difficulty walking or moving around, contact your doctor.
- Occasionally, steroids cause **diabetes**. If your child seems more thirsty than normal, needs to pass urine (wee) often, or starts wetting the bed at night, contact your doctor.

Other treatments

Some children need to take **antibiotic** medicines to help prevent or treat infections, or **diuretic** medicines to reduce swelling in their body. Most children need to eat a no-added salt diet to control their blood pressure.

» **More about treatment in Nephrotic syndrome**

About the future

Most children with frequently relapsing nephrotic syndrome, especially if it can be treated with a low dose of steroids, will have no long-term problems with their kidneys.

Impact on your child and family

Children can usually do the things that other children their age do, including during relapses. After a hospital stay, they should be able to continue going to school or nursery. They can play with other children and stay active.

If your child is in contact with measles or chicken pox, tell your hospital straight away.

Future relapses

In most children, there are fewer or no relapses as they get older. Over 8 in 10 children with nephrotic syndrome will *not* have relapses 10 years after the first episode. Some children will continue to have relapses into their adulthood.

Long-term problems

If your child needs to take immunosuppressants, he or she will have regular check ups and blood tests to monitor for side effects. With some immunosuppressants there may be a risk to kidney function or the blood count and your child will need regular blood tests to check for any abnormalities. Depending on the type of immunosuppressant, the drug levels may also be monitored. Occasionally there may be a risk of hypertension (high blood pressure) and your child's blood pressure will need to be monitored.

Some children develop **chronic kidney disease (CKD)** – the kidneys stop working as well as they should – this happens slowly, often over many years. These children may need further care. They will need to go back to the hospital or clinic for regular follow-up appointments.

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

Further information

Getting 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.**

This is the end of the information on frequently-relapsing nephrotic syndrome. 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.

Your notes and contact information

www.infoKID.org.uk



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