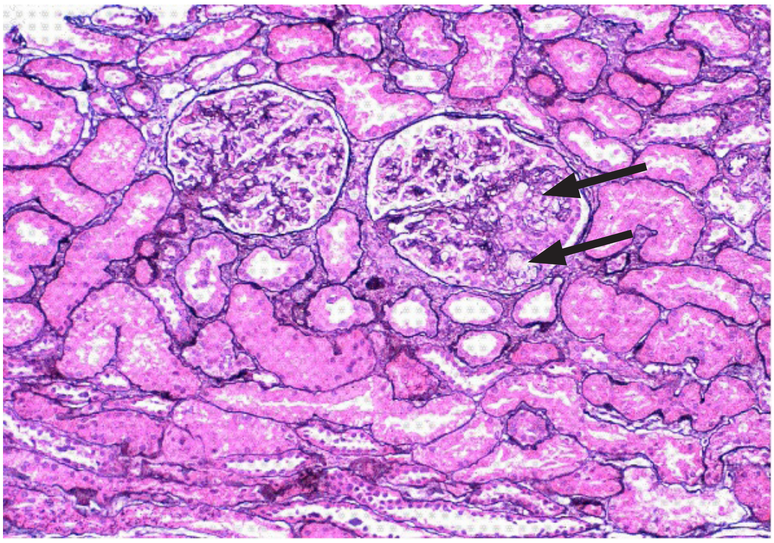
What is nephrotic syndrome?

Nephrotic syndrome is a condition that can happen in both adults and children. There are many different causes but they all cause the kidneys to leak **proteins**, particularly one called **albumin**, from the blood into the urine. Protein is lost from the body faster than the body can replace it. Albumin in the blood stream acts like a sponge to keep water in your blood vessels and without it fluid leaks out into the body tissues. Fluid accumulates in the tissue and the person starts to look puffy. Even though the kidneys are leaking protein, in other ways they are working normally, and it does not mean that your child's kidneys are failing.

What causes nephrotic syndrome?

Although there are different processes that affect the kidney leading to nephrotic syndrome, the most common form in children is one called **minimal change disease**. As the name suggests this form has very little sign of damage in the kidney when a small sample of kidney is looked at (see tests below). We don't know why children get this but it is related in some way to the immune system: some people have suggested it may be more common in children with a history of allergy. We know it is slightly more common in boys than girls and in children between the ages of two and ten years but it can happen at all ages. It can't be passed from one person to another and it may be related to genetic causes that as yet haven't been identified. Minimal change disease usually responds to medicine and children grow out of it over time. This form of nephrotic syndrome affects about 1 in 50,000 children.

There are other forms of nephrotic syndrome which are much rarer in children, but do sometimes happen. Many of these have had a genetic cause identified. One of these is called **congenital nephrotic syndrome** which is a form that you are born with. This can run in families. Another form is called **FSGS or focal segmental glomerulosclerosis**. In this form of nephrotic syndrome the kidney does become scarred or sclerosed which is where the name came from. These changes can be seen on kidney biopsy. These forms are more difficult to treat, and in some cases cause long term damage to the kidney leading it to kidney failure.



This picture from a kidney biopsy of a person with focal segmental glomerulosclerosis (FSGS) shows two kidney filters (glomeruli). The one on the left is normal and the one on the right has two small scars (arrows).

What is the usual treatment for nephrotic syndrome?

Most children are started on steroid therapy with a drug called prednisolone or prednisone. Most childhood forms of nephrotic syndrome respond to this drug. A response is called a remission and occurs when the protein disappears from the urine and the swelling goes away. It can take up to a couple of weeks to work and the first course of treatment is given for several months. Once the child is in remission the dose becomes smaller and can be given every other day. Side effects of the steroid can include a rise in blood pressure, an increase in appetite, weight gain, a slowing down of height growth, sleep disturbance and an angry and emotional child. All of these get better as the dose gets smaller.

Having the extra fluid on board can put a child at more risk of getting infections and so your child will be on a preventative antibiotic like penicillin.

While the doctors are waiting for the steroids to work they can try to help your child lose some of the extra fluid by giving replacement **albumin**. This helps get water out of the tissues back into the blood stream so the kidneys can get rid of the fluid. To help the kidneys do this your child might be given a medicine called frusemide which helps the kidneys make more urine. As the new albumin that is given is lost in the urine (just like the albumin the body produces), its effect may not last for long and it may need to be given repeatedly before nephrotic syndrome goes into remission.

What do I need to do at home to help my child?

You will be asked to learn how to do a dipstick test on your child’s urine first thing in the morning to check the protein level. Medication doses may need to change as the protein levels change; you should have a plan for this from your doctor, or know how to contact them. You may be asked to keep an eye on your child’s weight as sudden increases in weight in a relapse might mean that your child needs to be reviewed by a doctor. Some people find it easier to keep a record in a book every day.

Are there things my child should avoid?

We encourage children to lead as normal a life as possible including going to school and daycare. The school or daycare need to be aware that your child is more at risk of getting sick if they are exposed to illnesses like chickenpox. Ask them to let you know if there are illnesses going around, and check as to whether your child is immune to chickenpox in particular. There are some immunisations that may need to be delayed if your child is on higher doses of steroid, and your doctor can talk to you about this. It’s important to catch up with immunisations when the steroids are finished. If your child gets sick they may need a temporary increase in steroids to cover the stress on their body.

Do the steroids always work?

Most forms of minimal change disease will go into remission with prednisone (steroids) and so, unless your child's presentation looks like a different form of nephrotic syndrome, these will always be tried first. However, in some children steroids do not work. This does not mean that other medications will not work but does mean that they may have a different form of nephrotic syndrome (not minimal change disease). If the steroids have not worked after a few weeks, the doctors may need to take a closer look at your child's kidneys. This can require blood tests, or a special test called a **kidney biopsy**. There is information about having a renal biopsy on the Starship website and the **www.kidshealth.org.nz** website. This might be a time that your paediatrician discusses your child with a doctor who specialises in kidney problems in children (Paediatric Nephrologist).

What other medications may my child need?

Cyclophosphamide	This is a medicine that is used in children who go into remission with steroids, but have a lot of replases (over 3 per year) and/ or relapse (get protein leakage back again) whenever their steroids are stopped. As very long term steroids can have an effect on growth and bones this medicine can be used to reduce the amount of steroid given over time. It can stop relapses or reduce their number, or make them quicker and easier to treat. It was originally a chemotherapy medicine – it won't necessarily make your child sick or lose their hair but it does mean that there is a limit to the total dose that can be given, which is daily for 8 weeks. It is not a medicine that can be given several times. It does affect the production of white cells in the blood and these are checked weekly during treatment.
Cyclosporin	This drug was originally used in transplant medicine. It is used in children who continue to relapse after cyclophosphamide or in children who have FSGS. It is given twice a day and requires blood tests and checks on blood pressure. At the start regular blood drug levels are checked, but this becomes less frequent as the right level is reached.
Mycophenolate	This drug is used in a similar way to cyclosporin. It does not require a check on blood levels but can affect the white cells in the blood and can give some children diarrhoea.
Rituximab	This drug is a very strong immunosuppressant drug which is used in cases that have been resistant to all other forms of medicine. The drug is given into the vein (IV). If your child needs this drug it would be discussed in detail with you by the doctors

What does having nephrotic syndrome mean for my child long term?

Some children need one course of steroids and nephrotic syndrome stays away. Some children have relapses that respond to steroids. These relapses sometimes happen when they are sick for other reasons like coughs and colds. You will have a plan for what to do if your child gets sick, and if they have a relapse. For most children who have minimal change disease or steroid sensitive nephrotic syndrome there should be no long term consequences. Most children will catch up in height and the kidneys recover fully. We often find that children grow out of minimal change disease as they get older, but this can take a few years. Having relapses does not mean that children will not grow out of it. Sometimes children who have a lot of relapses need extra or alternative medication to steroids.

Children with FSGS or genetic forms of nephrotic syndrome do not grow out of it. They may need to be on more long term medication (see below). In some cases, the kidneys become damaged over time and eventually fail. This is rare in children, and children with these forms of nephrotic syndrome are seen by doctors who specialise in children's kidney disease and who can discuss the long term outlook on a case by case basis.

For the vast majority of children with nephrotic syndrome though the outlook is very good and they will grow out of it eventually.

Some terms you might hear or see in letters

Oedema – this is the fluid retention that you can see. One of the places parents notice this is round their child's eyes and eye lids in the morning, or collecting round their face. You will get to know where fluid tends to show up early in your child if they have relapses.

Proteinuria – this is the protein loss in the urine.

Remission – this is when the urine is clear from protein. Some children can come off all medicines when in remission while some need medicines to keep them in remission

Relapse – this is when the proteinuria comes back after a remission. Most children will have at least one relapse

Frequently relapsing – these are children who have a lot of relapses. They may be Steroid dependent which means that they need to stay on some steroids all the time.

Steroid sensitive – this describes a nephrotic syndrome that responds to steroids.

Steroid resistant – this describes all the forms of nephrotic syndrome that do not respond to steroids. Children with a congenital nephrotic syndrome and many children with FSGS fall into this group.

Glossary

◀ **Proteins**

Proteins form a large and essential part of the normal body. The health kidney filter ensures that protein is not lost through the kidney filters.

◀ **Albumin**

The most common protein in the blood.

◀ **Minimal Change Disease**

The most common form of nephrotic syndrome is called minimal change disease because under the ordinary light microscope the kidney biopsy appears normal. The condition most often affects children can occur in adults.

◀ **Congenital Nephrotic Syndrome**

A form of nephrotic syndrome passed down in families.

◀ **FSGS or Focal Segmental Glomerulosclerosis**

A type of glomerulonephritis that often causes nephrotic syndrome.

◀ **Kidney Biopsy**

The removal of a sample of tissue from the kidney the for purposes of diagnosis. Your kidney specialists can give you further information on this test.

PRINT

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CYCLOPHOSPHAMIDE (for renal patients)

(Cycloblastin®, Endoxan®, Cytoxan®)

What does it do?

Cyclophosphamide is used to treat some conditions affecting the kidneys.

How should you take it?

Cyclophosphamide tablets should be swallowed whole with food and a glass of water, usually in the morning. Do not break, crush or chew the tablets.
Cyclophosphamide injection is given as an infusion into a vein.

Wash your hands after handling *cyclophosphamide*

What if you miss a dose?

Take the missed dose as soon as possible and continue as directed. Do not take two doses at once.

Can you take other medicines?

Tell your pharmacist or doctor about <u>all medicines or treatments</u> that you may be taking including vitamins, herbal products (e.g. St John's wort, echinacea) or recreational drugs.

What side effects might you notice?

Side Effects	Recommended action
Increased risk of infection. Symptoms may include; fever, chills, sore throat aches and pains, tiredness, weakness boils or skin sores discoloured sputum generally feeling unwell Easy/unusual bruising or bleeding Difficult or frequent urination, blood in urine Signs of liver problems including; yellow skin or eyes, troublesome itching, dark urine, pale bowel motions Swelling of ankles or feet, shortness of breath, fast or irregular heart beat Skin rash, swelling, itching, breathing difficulties, dizziness	Tell your doctor immediately
Vomiting, loss of appetite, diarrhoea, nausea Mouth ulcers Hair loss or thinning Darkening of skin or fingernails Headache, dizziness, weakness, tiredness	Tell your doctor if troublesome

If you notice any other effects, discuss them with your doctor or pharmacist.

Other information:

- Reliable birth control for males and females is recommended while taking *cyclophosphamide*, and for 3 months after stopping. If you or your partner are planning to become pregnant, or find you are pregnant, please discuss this with your doctor.
- Drinking plenty of fluids may help prevent bladder damage – discuss this with your renal doctor.
- Tell your doctor if you have liver, bladder, blood or heart problems, or diabetes.
- Regular blood tests will be necessary while taking *cyclophosphamide* to monitor effects on your kidney function and blood count.
- Contact your kidney doctor if you come into contact with someone who has chickenpox or measles.
- *Cyclophosphamide* may reduce the response to some vaccines. Live vaccines, like rubella, should be avoided – discuss with your pharmacist or doctor.
- It is important to tell anyone who gives you medical or dental treatment that you are taking *cyclophosphamide*.

Prepared by the PILs Committee at Christchurch Hospital, Canterbury District Health Board, New Zealand. July 2007

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CICLOSPORIN (for transplant)

(Neoral®)

What does it do?

Ciclosporin is used with other medicines to prevent rejection after transplantation and treat other kidney conditions.

How should you take it?

It is important to take *ciclosporin* regularly at the same time each day – this will keep your blood levels constant. It is recommended that you take the morning dose with breakfast and the evening dose 12 hours later.

Ciclosporin capsules should be swallowed whole, with a glass of water. Store the capsules in the foil strips at all times – only remove them to take a dose.

Ciclosporin solution: carefully measure the dose using the syringe supplied, and add to a glass of apple or orange juice. This must be a glass cup – do not use plastic, paper or polystyrene. Always use the same type of juice. Stir the solution well and take immediately. Clean the syringe after use with a paper towel – do not rinse in water. Do not refrigerate the solution.

What if you miss a dose?

Take the missed dose as soon as possible and continue as directed. Do not take two doses at once.

Can you take other medicines?

Some medicines, available without prescription, may interact with *ciclosporin* including:

- anti-inflammatories such as; diclofenac (e.g. Voltaren Rapid®), ibuprofen (e.g. Nurofen®), mefenamic acid (e.g. Ponstan®), naproxen (e.g. Naprogesic®)

Tell your pharmacist or doctor about all medicines or treatments that you may be taking including vitamins, herbal products (e.g. St John's wort) or recreational drugs.

What side effects might you notice?

Side Effects	Recommended action
Increased risk of infection due to a reduced immune response. Symptoms may include; fever, chills, sore throat aches and pains, tiredness, weakness boils or skin sores painful urination, blood in urine discoloured sputum generally feeling unwell Easy/unusual bruising or bleeding Seizures	Tell your doctor immediately
Enlarged, tender or bleeding gums Altered vision Increased blood pressure Joint, muscle or bone aches and pains Frequent urination, increased thirst (may be signs of diabetes)	Tell your doctor
Increased body hair, acne or oily skin Fine tremor, tingling or numbness Headache, difficulty sleeping, mood changes, confusion Upset stomach, loss of appetite, constipation Skin rash, itching	Tell your doctor if troublesome

If you notice any other effects, discuss them with your doctor or pharmacist.

Other information:

- Do not stop *ciclosporin* unless your doctor at the clinic tells you to stop. You may have to take this medicine for the rest of your life.

- Regular blood tests will be necessary to measure the amount of *ciclosporin* in your blood, and your dose may be changed. On the morning of the blood test, do not take your regular dose of *ciclosporin* until after the blood has been taken. Other blood tests will also monitor your kidney function and cholesterol levels (*ciclosporin* can increase cholesterol).

- It is important to protect yourself from the sun while taking *ciclosporin* or any other immunosuppressive medicines. Immunosuppressive therapy may increase your risk of skin cancer. Always cover up and wear a good sunscreen (SPF30+) when outdoors.

- It is not advisable to get pregnant while taking *ciclosporin*. If you plan to become pregnant, or find you are pregnant, please discuss this with your doctor.

- Grapefruit, grapefruit juice and sour/Seville oranges should be avoided as they interact with this medicine. Discuss this with your pharmacist.

- Contact your kidney doctor if you come into contact with someone who has chickenpox or measles.

- Ciclosporin may reduce the response to some vaccines. Live vaccines, like rubella, should be avoided – discuss with your pharmacist or doctor.

Prepared by the PILs Committee at Christchurch Hospital, Canterbury District Health Board, New Zealand. June 2007

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MYCOPHENOLATE (for transplant)

(Cellcept®)

What does it do?

Mycophenolate is used with other medicines to prevent rejection after transplantation and to treat some other conditions affecting the kidney.

How should you take it?

It is important to take *mycophenolate* regularly, at the same time each day – this will keep your blood levels constant. It is recommended that you take the morning dose with breakfast and the evening dose 12 hours later.

Mycophenolate tablets and capsules should be swallowed whole (do not crush or chew), with a glass of water. Store the capsules in the foil strips at all times – only remove them to take a dose. Wash your hands after handling *mycophenolate*.

Mycophenolate suspension should be measured carefully and may be taken with a glass of water. Wash your hands after handling *mycophenolate* suspension.

What if you miss a dose?

Take the missed dose as soon as possible and continue as directed. Do not take two doses at once.

Can you take other medicines?

Some medicines, available without prescription, may interact with *mycophenolate* including:

- antacids (e.g. Mylanta®)
- calcium (e.g. Calci-tab®) or magnesium supplements

Do not take these medicines within 2 hours of taking *mycophenolate*.

Tell your pharmacist or doctor about all medicines or treatments that you may be taking including vitamins, herbal products (e.g. echinacea) or recreational drugs.

What side effects might you notice?

Side Effects	Recommended action
Increased risk of infection due to a reduced immune response. Symptoms may include; fever, chills, sore throat aches and pains, tiredness, weakness boils or skin sores painful urination, blood in urine discoloured sputum generally feeling unwell Easy/unusual bruising or bleeding Altered heart beat	Tell your doctor immediately
Severe diarrhoea, stomach pain, black bowel motions Cough, breathing difficulties Tiredness, dizziness, pale skin Altered vision Increased blood pressure, swelling	Tell your doctor
Nausea, constipation, indigestion Headache, difficulty sleeping, mood changes Acne, hair loss Fine tremor, tingling or numbness Skin rash, itching	Tell your doctor if troublesome

If you notice any other effects, discuss them with your doctor or pharmacist.

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RITUXIMAB

(Mabthera®)

What does it do?

Rituximab is used to treat certain types of nephrotic syndrome.

How is it given?

Rituximab is a clear liquid that will be given to you by infusion into a vein.
The drug will be given slowly at first but future doses may be infused over a shorter period of time.

Rituximab can cause a reaction while it is being infused. You may experience fever, chills, shivering, headache or breathing difficulties. Inform clinic staff immediately if any of these symptoms occur. This reaction is usually mild and can be controlled by giving the drug more slowly. You will also be given medicine shortly before the infusion to help control this reaction. Once the symptoms have gone away the speed of the infusion can usually be increased again.

What if you miss a dose?

If you are unable to attend an appointment for a dose of *rituximab,* contact the hospital clinic as soon as possible.

Can you take other medicines?

Tell your pharmacist or doctor about all medicines or treatments that you may be taking including vitamins, herbal products or recreational drugs.

What side effects might you notice?

Side Effects	Recommended action
Breathing difficulties Skin rash, itching, dizziness Swelling of the face, lips, mouth, tongue or throat Increased risk of infection. Symptoms may include: fever, sore throat, tiredness, aches and pains, boils or skin sores, pain, difficult or frequent urination, blood in urine, discoloured sputum and feeling unwell	Tell your doctor immediately
Joint, muscle or bone aches and pains Chest pain, palpitations	Tell your doctor
Stomach upset, diarrhoea, constipation	Tell your doctor if troublesome

If you notice any other effects, discuss them with your doctor or pharmacist.

Other information:

- Tell your doctor if you are pregnant, planning to become pregnant, or breast-feeding.
- Contact your kidney doctor if you come into contact with someone who has chickenpox or measles.

Prepared by the PILs Committee at Christchurch Hospital, Canterbury District Health Board, New Zealand. Jan 2007

IMPORTANT INFORMATION ABOUT MEDICINES

Take your medicines as directed

The instructions for how to use each medicine will be printed on the container. You should know when and how to take your medicines before you leave the pharmacy or doctor's office.

Complete the course

Continue taking your medicines as directed even if you start to feel better, unless your doctor says it is OK to stop.

Never share your medicines (this is illegal)

Your medicines have been prescribed for you and no one else. Do not let anyone else take your medicines. Do not take medicines intended for someone else.

Keep medicines in a cool, dry place

Heat, dampness and direct light can destroy some medicines. Store your medicines in an appropriate place.

Keep medicines out of reach of children

If necessary, store medicines in a locked cupboard.

Know your medicines

Take the time to learn about your medicines. Get to know their names, why you are taking them and what side effects they may cause. Your doctor or pharmacist can help by providing you with information such as written leaflets and an accurate record of all your medicines. Remember to bring this record, or your medicines, with you for all doctor and hospital visits.

Some common questions	What to do
What if you are pregnant, planning to become pregnant or breastfeeding?	Consult your doctor or pharmacist before taking any medicines when pregnant or breastfeeding, including over-the-counter remedies and herbal medicines.
What if the medicine is not working as you expected?	Talk to your doctor. The dose or type of medicine may need to be changed, or more time may be needed for you to notice the full effect.
How do you dispose of unwanted medicines?	Ask your pharmacist how to dispose of unwanted medicines. Do not throw medicines in the garbage or flush them down the toilet. Most regions in New Zealand have a medicines waste disposal service provided through community pharmacies.

This leaflet contains important information, but not ***all*** information about this medicine.
Discuss any questions with your health professional.

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