

Childhood Nephrotic Syndrome



*A guide for
parents on the
management
and treatment
of Childhood
Nephrotic
Syndrome*



The foundation of kidney care.

Acknowledgments

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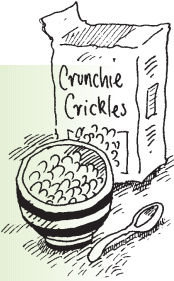
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Introduction

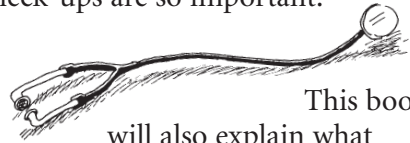
You have recently learned that your child has **nephrotic syndrome**. You probably have many questions about this condition and how it is treated. This booklet has been prepared to help answer some of those questions.

After you have read this booklet, you will know what nephrotic syndrome is and what treatments are used to manage it. You will also learn how you can play an important role in helping your child, such as by choosing healthy foods and encouraging your child to take part in normal childhood activities.



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You will learn why regular medical check-ups are so important.

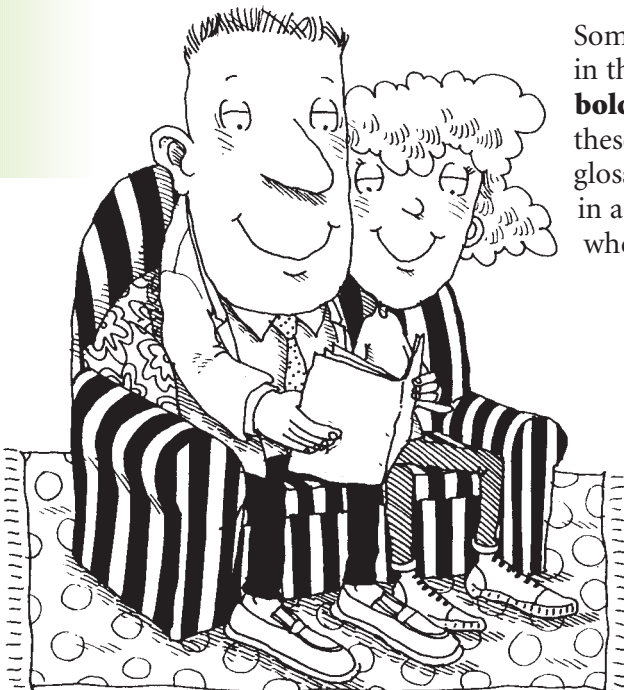


This booklet will also explain what physical signs to watch for and when to call your child's doctor. Finally, this booklet will tell you where you can obtain more information about nephrotic syndrome.

It is also important for you to remember that you have the full support of your child's healthcare team. You are not alone in managing your child's treatment and recovery.



Some words used in the text appear in **bold lettering** and these are explained in the glossary at the back. Keep this booklet in a handy place so you can refer to it when you need to.



What is nephrotic syndrome?

■ An overview...

Nephrotic syndrome is also called **nephrosis**. These two terms describe a condition in which the **kidneys** leak large and abnormal amounts of protein into the urine. When protein is lost in the urine, this leads to puffiness or swelling (**edema**), often of the eyelids, feet and ankles, and eventually the abdomen. If left untreated, this can lead to problems with breathing, eating and infections. More than 90% of children with nephrotic syndrome are successfully treated with **steroids**. (You'll learn more about steroids later.) Other treatments are available for more difficult cases.

■ A bit more detail...

Part of the job of the kidneys is to clean the blood of waste substances and to help regulate the amount of water and salt (sodium) in the body. In nephrotic syndrome, when the kidney filters (**glomeruli**) are cleaning the blood, too much protein leaks out of the blood through the filters and into the urine.

This causes the level of protein in the blood to fall.

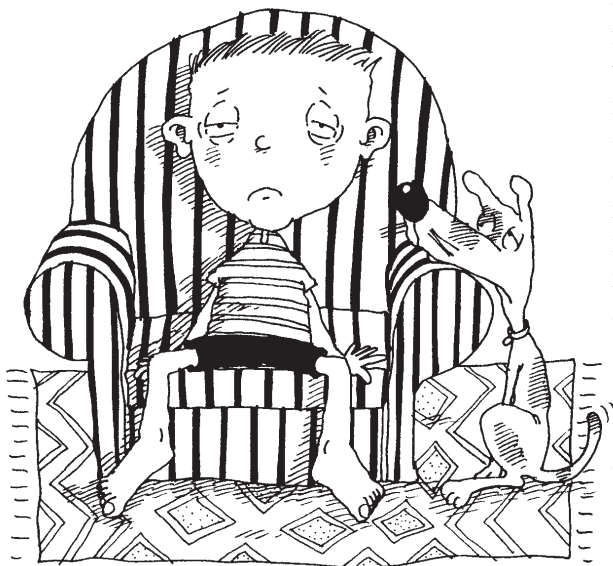
The blood needs protein to help keep water within the blood vessels (circulation). When the protein levels in the blood become very low (because a large amount of protein is leaking out into the urine), water can't be kept within the blood vessels and leaks into the tissues that surround very small blood vessels (capillaries). This water in the tissues appears as swelling (also called edema).

Nephrotic syndrome is diagnosed by recognising three findings:

1. swelling (edema)
2. high levels of protein in the urine (**proteinuria**) and
3. low levels of protein (**albumin**) in the blood (**hypoalbuminemia**).

■ What is the course of the disease?

The onset or first “attacks” of nephrotic syndrome can be a disturbing experience for you, as parents, and for your child. Because the swelling tends to develop slowly, you may not recognize it right away. By the time a diagnosis is made by a doctor, your child may be very swollen and may need to be hospitalized. Most children respond very well to treatment of nephrotic syndrome (see page 5) and, although most children have further “attacks” of the disease (see page 9), the long-term prognosis for most children is very good (see page 10).





■ Who is affected?

Childhood nephrotic syndrome can start at any age, but usually begins between the ages of two and five years. It is a rare condition that affects about 16 out of every 100,000 children at any given time, and it affects more boys

than girls. It is extremely unlikely that other children in your family will also have nephrotic syndrome. While there are a few types of nephrotic syndrome which do run in families, these are very rare.



What causes nephrotic syndrome?

The exact cause of nephrotic syndrome is not known and it cannot be prevented. However, research into this condition is ongoing and researchers are trying to develop increasingly effective treatments.

What we do know is that nephrotic syndrome is usually caused by an imbalance, from time to time, of the body's immune system. This imbalance

causes certain chemicals to disturb the filters of the kidneys. These filters begin to allow proteins to leak into the urine. All the successful treatments for nephrotic syndrome work on the immune system in some way.

Many, but not all, attacks of nephrotic syndrome are brought on by something that stimulates the immune system, such as a cold, flu or other infection.

What investigations are required?

Relatively few tests are usually required. These may include:

- Urine tests to assess the amount of proteinuria
- Blood tests to measure blood protein levels and kidney function

A **kidney biopsy** is not usually needed at the time of diagnosis in most children. However, if your child is very young (less than a year of age)

or relatively old (usually more than 10 years old) at the time of the first “attack” of nephrotic syndrome, a biopsy may be required. See page 8 for more information.

Rarely nephrotic syndrome affects more than one person in an extended family tree. In these cases, sometimes genetic testing may be recommended by your healthcare team.

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What is the treatment?

The following is a general description of the treatment for nephrotic syndrome. Of course, *specific treatments may vary from child to child* or from community to community. Please feel free to ask your child's healthcare team about any details of your child's treatment that you do not understand.

■ Steroid drugs (prednisone)

When your child is first diagnosed with nephrotic syndrome, the doctors will usually prescribe a medication called **prednisone** or **prednisolone**. Prednisone is a steroid, and most children respond very well to this drug treatment.





However, it does take some time for the steroids to work, so your child may not immediately appear to be getting better. Usually, within one to two weeks the protein in the urine disappears and the swelling in the tissues goes away. This stage is called **remission**.

A child that responds to steroids (steroid sensitive nephrotic syndrome) by going into remission is usually thought to have a clinical diagnosis of **minimal change disease**. Minimal change disease usually has a good prognosis (see page 10 for more details).

Your child must always take the steroids *exactly* as directed by the doctor. If your child *suddenly* stops taking the steroids, he or she may have complications or become very ill.

You should also know that the kinds of steroids used to treat nephrotic syndrome are *not* the same as the anabolic steroids that are sometimes abused by some athletes.



Sometimes, until the steroids have a chance to work, **diuretics** may also be given to help control severe swelling. Diuretics are a type of medication which helps the body to get rid of extra salt and water. Also, infusions of albumin (a blood product) are sometimes necessary to control severe swelling.





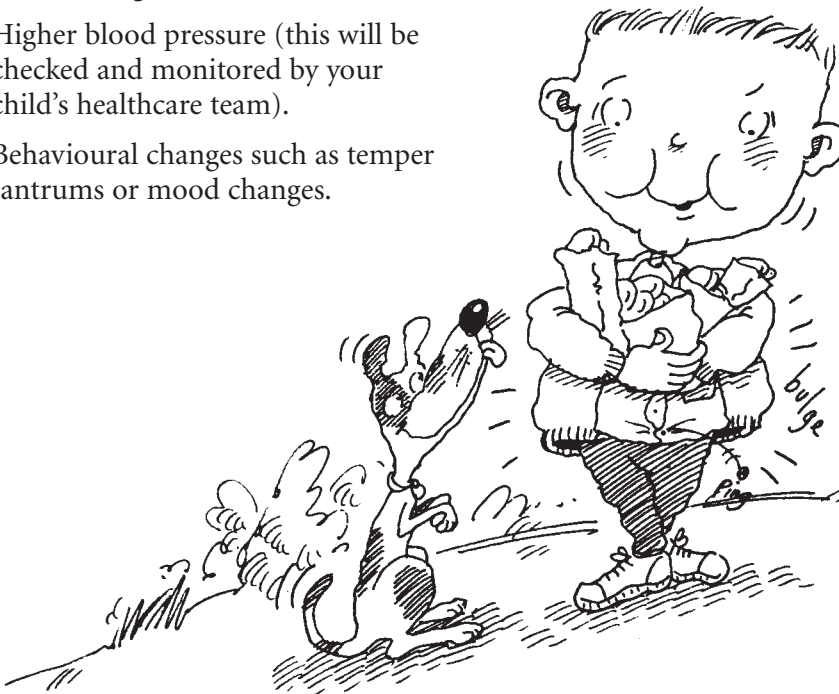
■ Possible adverse effects of steroids

There are usually no serious or permanent adverse effects when steroids are prescribed for short periods of time. However, sometimes adverse effects *may* occur and these are listed below:

- Increased appetite which may lead to rapid weight gain (see page 14 for helpful information on ways to minimize this).
- Flushed, swollen cheeks.
- Stretch marks on the skin.
- Decreased resistance to infections, such as coughs or colds.
- Higher blood pressure (this will be checked and monitored by your child's healthcare team).
- Behavioural changes such as temper tantrums or mood changes.



Important: If your child develops diarrhea, vomiting, severe abdominal pain or a high fever while taking steroids, contact your child's doctor right away.



■ Does steroid treatment always work?

There is a very small group of children who do not respond to steroid treatment (steroid resistant nephrotic syndrome), who continue to have lots of protein leaking into their urine, or who show serious adverse effects from prednisone. These children may need to have a kidney biopsy to find out what is going on in their kidneys. If a biopsy is necessary, your child's doctor will discuss this with you thoroughly.

■ What is a kidney biopsy?

A kidney biopsy is a procedure in which a very tiny tissue sample is removed from one kidney using a special needle. This procedure is carried out in the hospital. Older children are usually given a medicine to make them sleepy and some local anesthetic (freezing). Sometimes, young children are put completely to sleep using a general anesthetic. The procedure is more uncomfortable than painful, and usually has no long term effect on the kidney.

During the procedure, your child lies flat on his or her stomach. The doctors may use an ultrasound scan to find out where the kidneys are and where to insert a needle. The doctors use the needle to remove the tissue sample and send it to be examined under a microscope. It may take one to two weeks before the results of the biopsy are available.





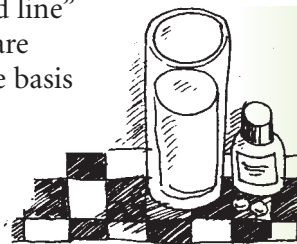
After the biopsy, your child must rest for a while, but can then be up and about quite quickly. Your child may be able to go home from the hospital the *same* day but this might not be possible if the biopsy is done later in the day.

It is usually recommended to restrict heavy exercise for one to two weeks after your child is released from hospital. If your child is of school age, he or she may even be able to return to school the day after release from the hospital. Specific instructions for your child will be given to you by your healthcare team.

■ Other treatment

Unfortunately, some children have nephrotic syndrome for many years, and despite taking steroids, have many relapses. A **relapse** means protein is leaking into the urine again, that may cause swelling again (see page 10 for more about relapses). Some children who have frequent relapses or have troublesome adverse effects from the steroid treatment may require other treatment prescribed by their nephrologist.

These “second line” medications are chosen on the basis of the child’s condition and after discussion of the



likely benefits and possible adverse effects of the various choices of second line medications.

The choice of medications may include **cyclophosphamide, chlorambucil, levamisole, tacrolimus** or **cyclosporin**.

These medications may help these children to have a long remission (see page 10).

If your child needs treatment with these or other medications, you will be given more information about them from your child’s healthcare team.

How long will my child have nephrotic syndrome?

Most children will have at least one relapse or recurrence of protein in the urine. Each relapse will need a further course of steroid treatment.

In general, steroid treatment for a relapse is for a shorter time than the initial treatment at the time of diagnosis of nephrotic syndrome.

If a child has fairly frequent relapses then they might take a small dose of steroid medication on alternate days to prevent relapses. This is usually called maintenance treatment.

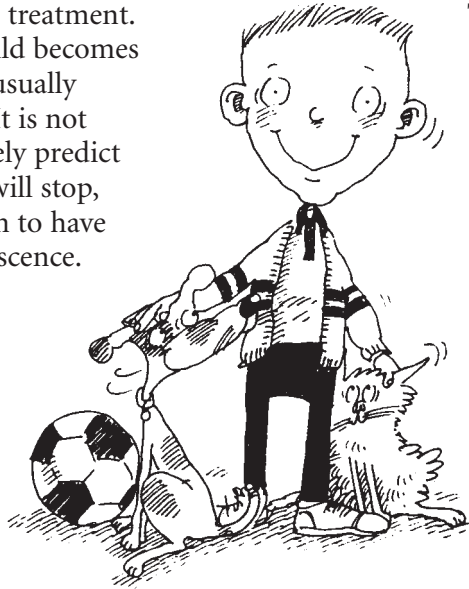
However, as the child becomes older, the relapses usually happen less often. It is not possible to accurately predict when the relapses will stop, but it is uncommon to have relapses after adolescence.

It is also uncommon to have a relapse after the child has been free of protein in the urine (in remission) for five years.

It is important to remember that children with the usual type of nephrotic syndrome (minimal change disease) have *no* risk of kidney failure.

Usually, as your child grows older, the nephrotic syndrome will become less troublesome. In fact, the statistics are on your child's side.

The great majority of children will eventually outgrow the problem and become teenagers and adults with normally functioning kidneys.



How can I help in my child's treatment?

There are many ways in which you can play an important role in helping to manage your child's treatment. These include making sure your child receives regular medical check-ups, follows healthy eating patterns and maintains a normal, active family lifestyle. Sometimes you will also be asked to test your child's urine at home (to find out if there is protein in it). Each of these points will be discussed in some detail in the following sections.

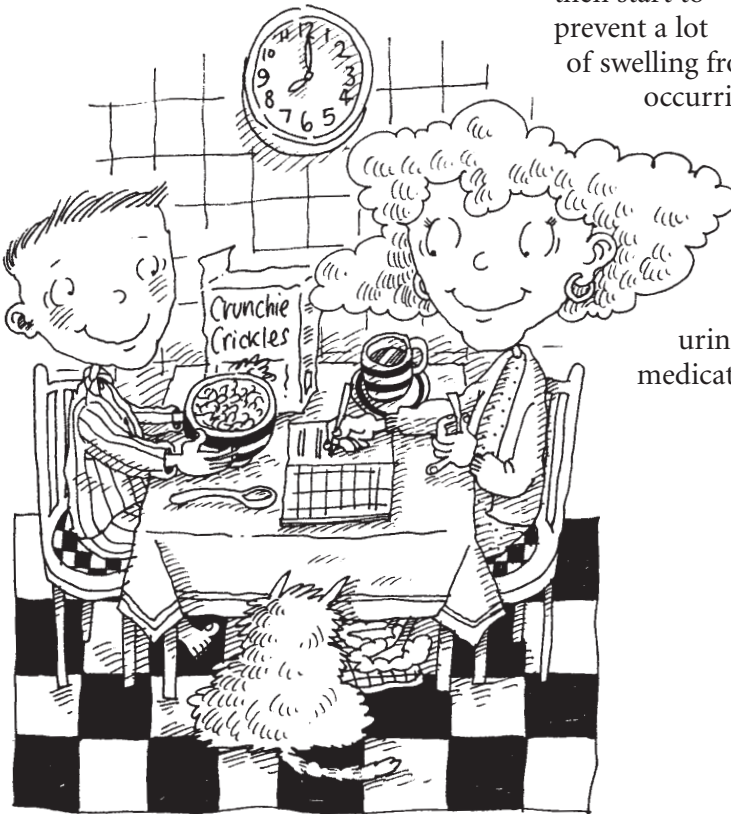


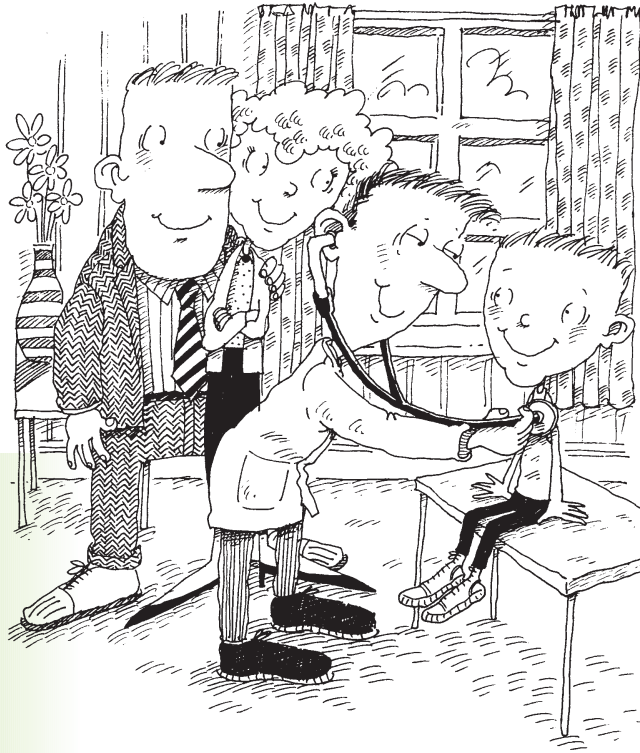
■ Do home urine testing

Soon after the diagnosis of nephrotic syndrome is made, your child's healthcare team will show you how to check the morning urine for the presence of protein. (The best urine sample to test is the first one obtained in the morning.) You will be advised how often this testing should be done. At times, there is an advantage to daily urine testing. The advantage of testing the urine at home is that you will be able to spot a relapse early before it has caused swelling. Treatment can then start to prevent a lot of swelling from



occurring. There are other times when you will only need to do urine testing when your child has a "cold". You will also be shown how to keep a record book of urine testing results and the medications you give your child.





■ Take your child for regular check-ups

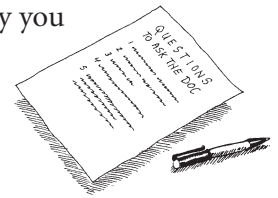
Your child will need to go for regular check-ups with the doctor, and perhaps other members of the healthcare team, such as the dietitian. Even if your child is well, it is very important that these regular visits occur because they help to monitor your child's overall progress.

During each check-up your child will receive a physical examination and have his or her height, weight and blood pressure recorded.

The urine will be analyzed as well. Less often, blood tests may be required, but these are performed only when necessary. If your child is very worried about blood tests (because of the needle), the healthcare team can apply an anaesthetic cream in advance so that needle injections are less painful.



It is a good idea to write down any questions you may have before going to the check-up. This way you will know what to ask your child's doctor, and other members of the healthcare team.



Finally, remember to take care of your child's teeth. Good dental care is essential, so it is important to take your child for regular check-ups at the dentist as well.

■ Choose healthy foods

Another way you can help your child is to make sure they follow a sensible and healthy eating plan.

This will help your child to maintain an ideal body weight and to feel better.

Children taking steroids tend to be ravenously hungry. If they are allowed free access to food, they will gain a great deal of weight. Your dietitian can help you devise tasty snacks which can help to lessen some of the hunger.



In fact, it is a good idea for the whole family to choose nutritious foods. Here are some guidelines to help you.

■ **No added salt.** Do not add salt to food at the table. Try not to use too many processed, prepackaged or “fast” foods. They contain a lot of hidden salt.



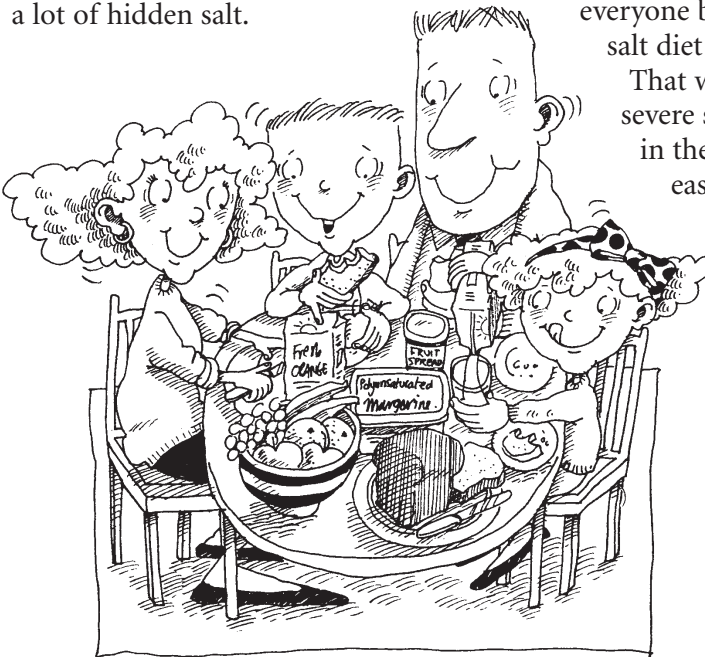
Examples of high-salt foods:

Processed meats: wieners, salami, ham
Canned foods: soup, pasta, vegetables
Prepackaged foods: macaroni and cheese dinners, dried soup mixes
Other foods: potato chips, pretzels, snack crackers, pickles, ketchup

Important: It is best to strictly *reduce salt intake* when your child is leaking protein into the urine (i.e. during a relapse) or when taking prednisone. Salt will make your child thirsty and want to drink more fluids. It will also make your child hold extra fluid which will make the swelling or puffiness worse.

Ultimately, it is much easier on your child and your entire family if everyone becomes used to a lower salt diet from the beginning.

That way, you can help prevent severe swelling from occurring in the first place. This is much easier on your child than the treatments needed to reduce swelling.





- **Reduce sugar intake.** Try not to add sugar to food and drinks. Replace soft drinks and “kool-aid” type drinks with water or sugar-free varieties. Cut down as much as possible on sweets such as candies, cakes, cookies and chocolate.
- **Eat more fiber rich foods.** Try whole-wheat or multi-grain breads and breakfast cereals. Choose more fresh fruits and vegetables. They make excellent snacks!
- **Limit high fat foods.** Choose foods that are baked, broiled or barbecued rather than fried or deep-fried. Try not to add a lot of extra fat, such as margarine or gravy, to foods. Choose soft-tub margarines and vegetable oils rather than hard margarines, shortening or lard.

If you would like more advice on healthier eating, there are many good sources available. Your dietitian will be able to suggest some for you to read.





■ Plan sports and leisure activities

Sports and exercise are important for the whole family. Your child should participate in all sports and leisure activities that are within his or her capabilities, including those at school. If you have any questions about certain sports, ask your child's doctor. In fact, the doctor may recommend that your child exercise regularly to help counteract many of the adverse effects that steroids sometimes cause. Also, being active can help prevent your child from gaining too much weight.



■ Consider MedicAlert® identification

Some families feel more secure if their child wears a *MedicAlert*® bracelet at all times. These bracelets help in providing immediate identification of your child's medical conditions and medications. This information may be useful in emergency situations. For example, children who have recently been on steroids may require *increased* doses of steroids if they are involved in an accident, or if they require an operation. To find out how you can obtain a *MedicAlert*® bracelet, you can contact your local office of The Kidney Foundation of Canada.

Family outings and holidays should also be possible. Unless your child is having complications, these activities usually pose no problems. Be sure to tell your healthcare team about any travel plans you have. It is wise to do this well in advance of traveling, especially if you are going to areas that require special vaccinations.



When should I get in touch with the doctor?

There are some situations when you should call your child's doctor right away.

- If your child is in close contact with *chickenpox* or *red measles*. Call the doctor within 24 hours because your child may need a special injection to boost his or her resistance to these infections.
- If your child is puffy or their urine test shows a large amount of protein in the urine.

- If your child is unwell with a fever, vomiting or abdominal pain.
- If your child is due for immunization or vaccination, discuss potential problems with your child's doctor.

For any worry it is probably best to phone your child's doctor. At the back of this booklet there is a place for you to write down important phone numbers.



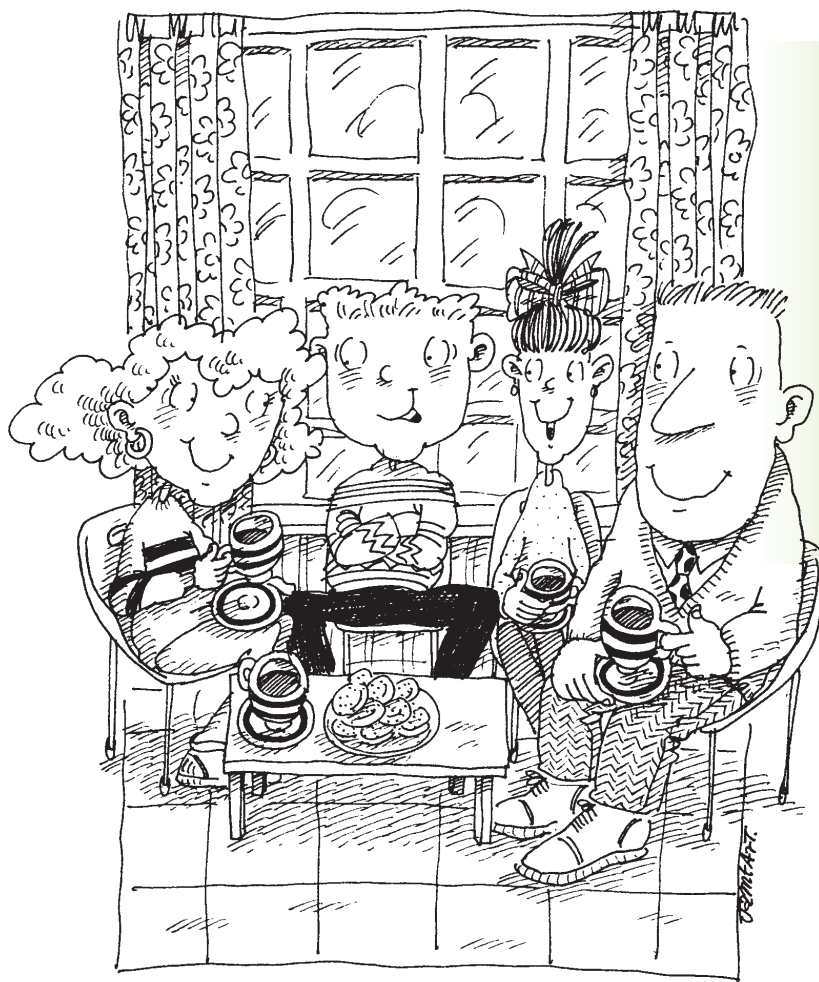
Are there any support groups?

The Kidney Foundation of Canada offers a wide range of support services for people living with kidney disease. There may also be other families in your area who have had to learn about nephrotic syndrome, and contact with these families can be helpful and reassuring. To find out what services are available in your area, it is best to consult your local chapter or branch of the Foundation.



You can also ask the members of your healthcare team, such as the nurses or renal social worker, about local support services.

Nurses and renal social workers are important members of your child's healthcare team. Their role is to help support you and your child during all stages of treatment.



Where can I obtain more information?

This booklet has probably not answered all your questions. The best sources to provide you with more information about nephrotic syndrome, and about living with kidney disease, are the members of your child's healthcare team and The Kidney Foundation of Canada.

There are many educational booklets available simply by calling The Kidney Foundation. Or, if you have access to the Internet/World Wide Web, you can visit The Kidney Foundation's Web site at www.kidney.ca.

Useful telephone numbers

Family Doctor

Pediatrician

Pediatric Nephrologist

Clinic Nurse

Pediatric/Renal Dietitian

Social Worker

Local Kidney Foundation Office

Other



Albumin

The most common type of protein normally found in blood. Albumin, as a blood product, can be used as an intravenous medication to help control severe edema (swelling).

Chlorambucil

Medication used to treat many diseases, including, as a second line medication, nephrotic syndrome.

Cyclophosphamide

Medication used to treat many diseases, including, as a second line medication, nephrotic syndrome.

Cyclosporin

Medication used, as a second line medication, to treat nephrotic syndrome (also used to prevent rejection of organ transplants).

Diuretics

Medication given to help the body eliminate extra water and salt.

Edema

Swelling of the body tissues due to salt and water retention.

Hypoalbuminemia

Low levels of albumin protein in the blood. Albumin is the most common protein found in blood.

Kidney

One of the two “fist-sized” organs located at the back of the abdominal cavity on each side of the spinal column.

Kidney Biopsy

A procedure to remove a tiny piece of kidney tissue for special examination under the microscope. Often an ultrasound will be used to guide the needle to the kidney.

Levamisole

Medication used, as a second line medication, to treat nephrotic syndrome and many other conditions.

Minimal Change Disease

This describes the “minimal changes” in the kidney, eg the lack of any chronic damage, shown by a kidney biopsy that would be seen in most children with nephrotic syndrome.

Most often minimal change disease is clinically diagnosed (without a kidney biopsy) after a remission due to steroids since most children do not have to have a kidney biopsy performed.

Nephrosis

Another term for nephrotic syndrome.

Nephrotic Syndrome

A collection of findings (syndrome) found by clinical examination/laboratory tests. The findings are swelling (edema), proteinuria and low blood albumin (hypoalbuminemia).

Prednisolone

Also called prednisone. Steroid medication used to treat nephrotic syndrome and many other conditions.

Prednisone

Steroid medication used to treat nephrotic syndrome and many other conditions.

Proteinuria

Increased amount of protein in urine. Normally the urine contains only very small amounts of protein.

Relapse

An episode of nephrotic syndrome characterized by protein in the urine followed by swelling. Usually defined by 3-5 consecutive days of increased proteinuria.

Remission

When a relapse is finished. Usually defined by 3-5 consecutive days of no proteinuria.

Steroid

Medication which reduces inflammation and is used to treat many kidney diseases. (These kinds of steroids are not the same as the anabolic steroids that are sometimes abused by some athletes.)

Tacrolimus

Medication used, as a second line medication, to treat nephrotic syndrome (also used to prevent rejection of organ transplants).

The Kidney Foundation

OUR VISION

Kidney health, and improved lives for all people affected by kidney disease.

OUR MISSION

The Kidney Foundation of Canada is the national volunteer organization committed to reducing the burden of kidney disease through:

- funding and stimulating innovative research;
- providing education and support;
- promoting access to high quality healthcare; and
- increasing public awareness and commitment to advancing kidney health and organ donation.

Since 1964, our fundraising campaigns have allowed us to contribute millions of dollars to research, and to provide services to individuals living with chronic kidney disease and related conditions.

For further information, or if you wish to help us in our efforts, please contact The Kidney Foundation of Canada office in your area. You can also visit our Web site at www.kidney.ca.