A PARENT'S HANDBOOK

Childhood Nephrotic Syndrome

Developed by

Pediatric Nephrology Program
British Columbia Children's Hospital
British Columbia Provincial Renal Agency
Vancouver, British Columbia, Canada

3RD EDITION • OCTOBER 2016

INFORMATION

When and how to contact us

If you think your child needs to be seen or if they are having problems we recommend that you first try to contact your family doctor or pediatrician. If you are not able to reach your doctor, you can contact the Renal Program at BC Children's Hospital.

During business hours, please call **604 875 2272** and we will connect you with one of the team members.

Outside business hours, please call the hospital switchboard at **604 875 2345** and ask for the kidney doctor (nephrologist) on call.

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Table of Contents

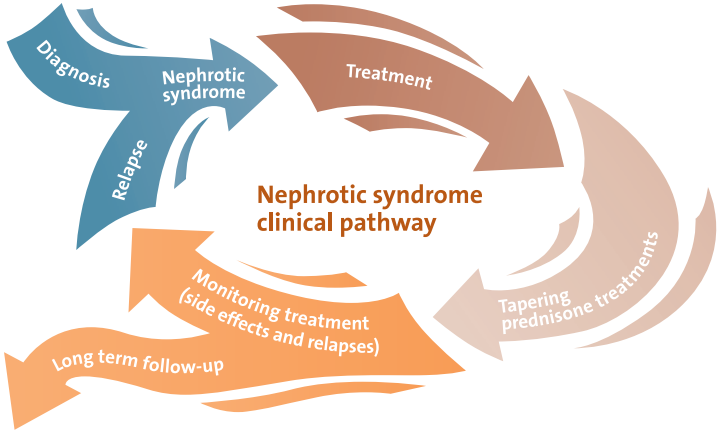
SECTION 1

Introduction

The purpose of this handbook is to provide information to parents of children with nephrotic syndrome. We present an organized approach for the diagnosis and management of this condition. **We call this approach our nephrotic syndrome clinical pathway.** It is the result of the work of our group of doctors, students, dietitians, nurses, and families of children with nephrotic syndrome.

This handbook provides general information about the condition and its treatments. The accompanying *Worksheets* are practical, easy-to-use, and can be filled in at home. These worksheets are used to keep track of prednisone dosing, urine dipstick results, relapses, possible side effects or complications, and diet intake. Recording your child's progress and therapy will help us provide the best care for your child.

Our pathway recommendations are based on the best and most current evidence, our own experience, and the advice of our expert team and families who care for children with nephrotic syndrome. Welcome to the team!



The Shared Care Renal Program at BC Children's Hospital

Nephrotic syndrome is one of the most common kidney problems in children. In our clinic at BC Children's Hospital we treat up to 20 new patients every year. We continue to follow close to 100 patients in our renal clinic.

Children with nephrotic syndrome require shared and ongoing care. Families, family doctors, pediatricians, and kidney specialists all play an equally important role. At BC Children's Hospital, all of our kidney doctors (nephrologists) are responsible for your child's care. However, in clinic you may only see a few of them depending on the day of your appointment. Eventually you will get to know many of our doctors and they will get to know you. This is particularly useful if you or your family doctor or pediatrician have to call the hospital after hours or in an emergency.

When you come to clinic you will meet various members of the renal clinic team as needed *(Table 1)*. We will answer any questions you may have, make our recommendations, and arrange for your follow-up visit before you leave.

TABLE 1

Members of the renal clinic team

Booking clerk
Nurse's assistant
Renal clinic nurse

Doctor in training (student, resident, fellow)
Nephrologist

Dietitian
Pharmacist
Social worker
Researcher

SECTION 2

Diagnosis of nephrotic syndrome

Your child has been diagnosed as having nephrotic syndrome because they have swelling, protein losses in the urine, and low blood protein levels.

Swelling (edema)

Swelling is the most common feature that brings children with nephrotic syndrome to medical attention. It may come and go for weeks prior to the first medical visit. This swelling, particularly around the eyes, is often mistaken for an allergic reaction *(Figure 1)* and may have been treated unsuccessfully with allergy medications. Most children and their parents will notice puffy eyelids first, followed by more generalized swelling of the feet and legs, and swelling of the belly (abdomen) *(Figures 2, 3a, 3b)*. In severe forms, there is swelling of the genitals (scrotum or labia).

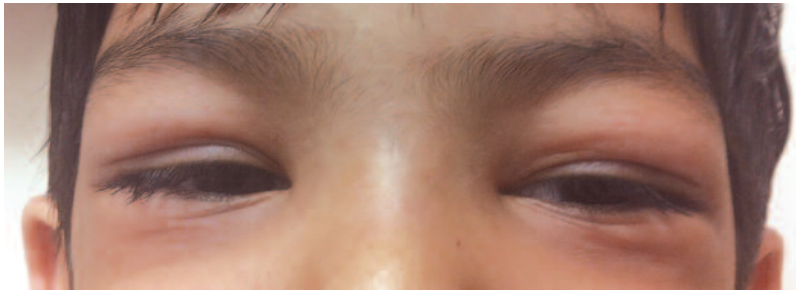
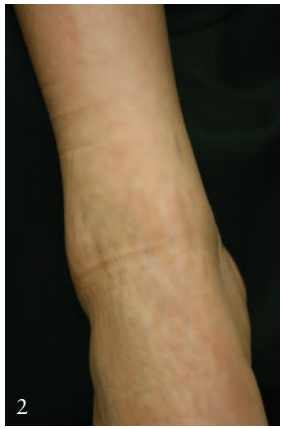
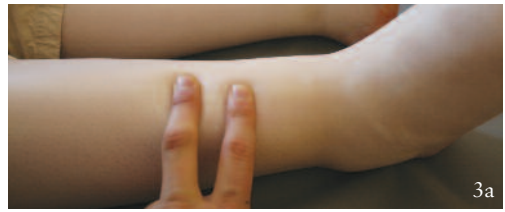


Figure 1. Patient has moderate swelling around the eyes.

The swelling may develop gradually over the course of a few weeks and may shift from one area of the body to another, depending on the time of day. For example, you may notice more swelling in your child's face early in the morning after sleeping. During the day, the swelling becomes more noticeable in the legs, feet, and ankles.





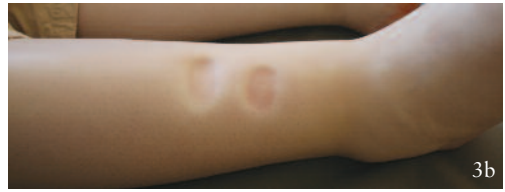


Figure 2. Patient has mild swelling of the feet and ankles (note the stocking lines).
Figure 3a. Patient has severe swelling of the lower leg that hides ankle definition.
Figure 3b. Gentle pressure with the finger tips leaves a temporary imprint on the lower leg.

Protein losses in the urine (proteinuria)

Large losses of protein (albumin) in the urine is an important feature of nephrotic syndrome. This may appear as frothy (foamy) urine in the toilet, but is confirmed by dipping the urine with a dipstick *(Figure 4* below and the *Urine protein testing at home* section of the *Worksheets)* and a urine protein test sent to the lab.

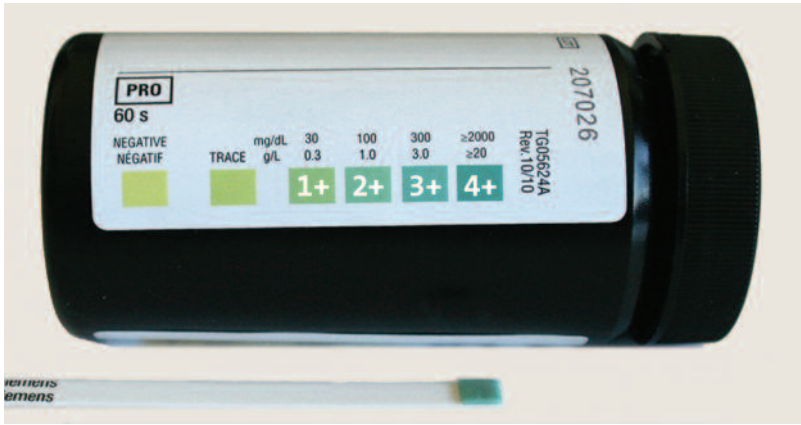


Figure 4. This is an example of a urine dipstick test. The dipstick at the bottom of the picture has turned a shade of green after 60 seconds. The green shade would be read as 4+ according to the scale shown, NOT ≥2000 or ≥20 as indicated on the bottle. The + indicators (1+ to 4+) do not actually appear on the bottle.

Low blood protein levels (hypoalbuminemia)

Large losses of protein in the urine cause low levels of protein (albumin) in the blood. Low blood protein levels lead to fluid leaking from the blood into the body tissues, causing swelling *(Figure 5)*. Low blood protein levels are found by a simple blood test that measures the concentration of albumin.

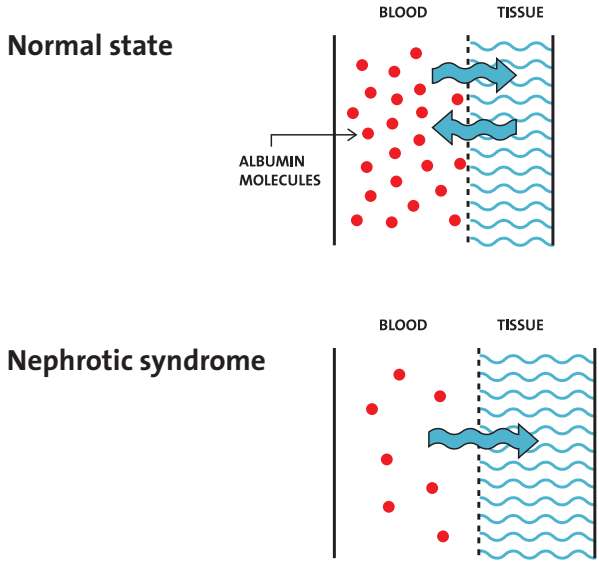


Figure 5. Diagram of water movement in normal state and in nephrotic syndrome leading to swelling (arrows show water movement).

SECTION 3

Types of nephrotic syndrome

There are different types of childhood nephrotic syndrome. One of the best ways of classifying nephrotic syndrome is by the response to prednisone. Prednisone, is a common "steroid" medicine used in several childhood diseases. How a child with nephrotic syndrome responds to prednisone can often predict the course of illness and the long-term outcome.

Steroid sensitive nephrotic syndrome

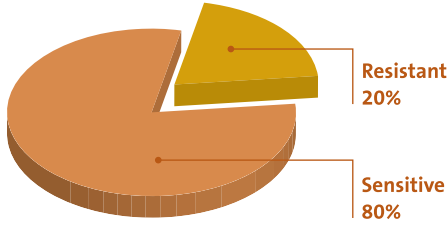
Children have **steroid sensitive** nephrotic syndrome if they respond to prednisone within 6 weeks. This means that by 6 weeks of starting prednisone the swelling has improved and there is no more protein in the urine. About 80% of children with nephrotic syndrome have steroid sensitive nephrotic syndrome *(Pie Chart 1)*. A response to prednisone means your child has nephrotic syndrome likely due to **minimal change disease.** With minimal change disease the kidneys look normal when examined under a microscope. **This is the best type of nephrotic syndrome to have.** If your child has steroid sensitive nephrotic syndrome, there is usually no need for a kidney biopsy as most children who respond to prednisone have minimal change disease *(Pie Chart 2)*.

Steroid resistant nephrotic syndrome

Children have **steroid resistant** nephrotic syndrome if they do not respond to prednisone after 6 weeks. This means that by 6 weeks of starting prednisone there is still some swelling and/or protein in the urine. About 20% of children with nephrotic syndrome have steroid resistant nephrotic syndrome *(Pie Chart 1)*. They will require a kidney biopsy to diagnose the specific type of nephrotic syndrome as they are less likely to have minimal change disease. Steroid resistant nephrotic syndrome often requires different medicines. Children with steroid resistant nephrotic syndrome are at higher risk of long-term kidney problems such as reduced kidney function and kidney failure.

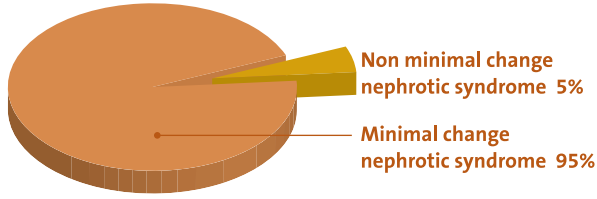
PIE CHART 1

Childhood nephrotic syndrome: response to prednisone



PIE CHART 2

Types of nephrotic syndrome in children who respond to prednisone



SECTION 4

Your child's first visit to the doctor

Your child's first visit may take place in the doctor's office, medical clinic, or emergency room. At the first visit, the doctor will ask you about your child's complete medical history, perform a thorough physical examination, and order the appropriate laboratory testing. In particular they will check your child's blood pressure, kidney function, and urine. The results of these evaluations will identify any problems that need to be dealt with and help predict the long-term response to treatments.

Checking blood pressure

High blood pressure is occasionally seen in children with nephrotic syndrome at their first visit. Sometimes high blood pressure may be related to the underlying form of nephrotic syndrome. Other factors such as pain and anxiety may also cause temporary increases in blood pressure. It is therefore important to repeat this measurement when your child is calm and relaxed. If blood pressure stays high after the first visit, treatment may be required.

Monitoring kidney function

Many children with nephrotic syndrome have a temporary decrease in kidney function, which may be due to swelling or being dehydrated. This usually improves once prednisone treatment is started and the swelling decreases, but your child may need to be checked again if the kidney function was found to be abnormal initially. Kidney function can be measured with a blood test (creatinine).

Testing urine for blood

At the first visit, the urine test your doctor performs checks for both blood and protein. If your child has significant amounts of blood in the urine (hematuria) at the first visit this may require careful attention since it is less common in minimal change disease.

SECTION 5

Complications of nephrotic syndrome

Now that your child has been diagnosed with nephrotic syndrome and started on treatment, it is important to watch for complications that might occur either because of the illness itself or because of the treatment.

If you think your child needs to be seen or if they are having problems we recommend that you first try to contact your family doctor or pediatrician. If you are not able to reach your doctor, you can contact the Renal Program at BC Children's Hospital. During business hours, please call **604 875 2272** and we will connect you with one of the team members. Outside business hours, please call the hospital switchboard at **604 875 2345** and ask for the kidney doctor (nephrologist) on call.

Points to remember

Contact your doctor if your child:

1. Has swelling that interferes with normal daily function or is associated with decreased urine production
2. Develops signs of infection (fever, skin rash, sore throat)

Contact your doctor **immediately** if your child:

1. Develops a fever AND belly (abdominal) pain
2. Has recently been exposed to chickenpox (varicella) or shingles
3. Develops signs of blood clotting (severe headaches, worse swelling in one leg, swelling with leg pain, difficulty breathing)

Swelling (edema)

If severe enough, swelling can lead to breathing difficulties, belly (abdominal) pain, skin changes, and infection. Swelling in the legs can make shoes tight and walking difficult. Severe swelling of the

genitals (scrotum or labia) can be uncomfortable and distressing. Most often attention to sodium and fluid intake will be enough to prevent further swelling while waiting for the prednisone to work *(*see *Section 7 Diet)*. Severe swelling can affect your child's kidney function and can lead to a decrease in the amount of urine produced. Sometimes the swelling can be so severe that admission to hospital is needed for further treatment. **If swelling interferes with normal daily functions or there is a drop in urine production while swollen, your child should be seen by a doctor.**

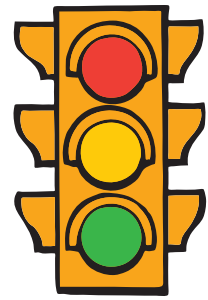
Infection

Your child's immune system is weaker than normal because of the large losses of protein in the urine with nephrotic syndrome. The medicines used to treat the nephrotic syndrome including prednisone make this worse. This puts your child at risk for severe infections. Good hand hygiene, regular visits to the doctor and dentist, and ensuring your child's routine immunizations are up-to-date will help lower their chance of getting sick. **If signs of a possible infection develop, including fever, skin rash, or sore throat, your child should be seen by a doctor. If a fever AND belly pain develops, your child should be seen by a doctor immediately. If your child has recently been exposed to someone with active chickenpox (varicella) or shingles, contact your doctor immediately.**

Blood clotting

Your child is at risk of developing clots in blood vessels because of nephrotic syndrome. This is an uncommon problem but can be serious, often requiring treatment as soon as possible. If it occurs it usually does so around the time of diagnosis, when your child is having a relapse, or if your child has one of the more severe types of nephrotic syndrome. Symptoms of blood clotting include severe headaches, worse swelling in one leg, swelling with leg pain, and difficulty breathing. **If you are concerned about signs of blood clotting, your child should be seen by a doctor immediately.**

SECTION 6



Treatment of nephrotic syndrome

Most children diagnosed with nephrotic syndrome do not require admission to hospital. They are typically first seen in the family doctor's or pediatrician's office or the emergency room. The information provided in this section and the accompanying *Worksheets* will help you navigate your child's course from the first day of being diagnosed with nephrotic syndrome and through any relapses that might occur in the future.

Prednisone treatment for initial episode

Once your child is diagnosed with nephrotic syndrome the next step is to start treatment with a medication called prednisone. Prednisone is the best medication for the initial treatment of nephrotic syndrome. Its benefits are greater than any potential risks. It is very important to give prednisone exactly as it is prescribed. Failure to give prednisone correctly may delay your child going into remission, putting them at higher risk of complications. If prednisone is not given as prescribed, your child may be incorrectly diagnosed as steroid resistant *(see Section 3 Types of nephrotic syndrome)*. This may then lead to your child being placed on other medications for the wrong reason.

We treat the **initial or first episode** of nephrotic syndrome with 6 weeks of daily prednisone. These 6 weeks of daily prednisone are followed by 6 weeks of alternate day (or every other day) prednisone. The total duration of initial prednisone treatment adds up to 12 weeks *(Figure 6)*.

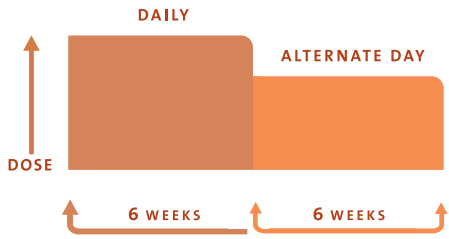


Figure 6. Time course of prednisone treatment for initial episode

The starting dose of prednisone will be based on your child's body surface area. Body surface area is a measure of your child's size in square meters (m^2), which is based on height (cm) and weight (kg). A prednisone schedule outlining the starting dose of prednisone and when to change from daily to alternate day prednisone will be given to you. The prednisone dosing for the initial episode is outlined in *Table 2*.

TABLE 2

Prednisone dosing for your child's initial episode of nephrotic syndrome

DURATION	DOSE BY CHILD'S SIZE (mg/m^2)*
6 weeks	60 mg/m^2 daily (max 60 mg total)
6 weeks	40 mg/m^2 alternate day (max 40 mg total)
STOP	

*Once daily in the morning on the days prednisone is given

What is a remission?

A **remission** is defined as 3 days in a row of negative or trace protein on first morning urine dipstick testing at home. In other words, remission means that for 3 consecutive days urine protein losses stop, swelling decreases, and blood protein levels return to normal. Your child has steroid sensitive nephrotic syndrome if, with prednisone treatment, they go into remission. The majority of children who are going to respond to prednisone do so within 2 weeks of treatment. Almost all the children who are going to respond to prednisone do so by 6 weeks of starting treatment. Some older teenagers take longer. Details about monitoring urine protein levels with dipsticks are included in the *Urine protein testing at home* section of the *Worksheets*.

What will happen after your child starts on prednisone for the initial episode of nephrotic syndrome?

After starting prednisone, one of three things will happen:

Your child's condition will get better: If your child goes into remission within 4 weeks of initial treatment, you should continue the prednisone. Your child's condition will be reviewed in clinic at the 4-week mark of starting initial treatment. This is expected in most cases.

Your child's condition will stay the same: If by 4 weeks after starting initial treatment your child has not gone into remission, so long as there are no other major health concerns you should continue the prednisone. Your child will be seen in clinic at the 4-week and 6-week mark of starting initial treatment to be sure there are no developing concerns. If your child does not go into remission by 6 weeks, they may require a kidney biopsy. This situation occurs less frequently.

Your child's condition will get worse: If while waiting to respond your child becomes more swollen or experiences any of the other complications (see *Section 5 Complications of nephrotic syndrome*), you should continue the prednisone and contact your doctor for further advice. This situation also occurs less frequently.

What is a relapse?

A **relapse** is defined as 3 days in a row of 3+ or 4+ protein on first morning urine dipstick testing at home — after previously being in remission (see *What is a remission?*).

Most children with nephrotic syndrome have relapses that require repeated prednisone treatment, so do not be alarmed or discouraged if this happens to your child. Approximately 90% of children with nephrotic syndrome will have a relapse. Most relapses are triggered by an infection, such as a cold or flu. Even very minor infections, dental cavities, or bug bites can sometimes trigger a relapse. A relapse will often require another course of prednisone treatment. Your child may not have swelling at the time a relapse is diagnosed; however, if left untreated, they will often develop swelling.

If a relapse happens you should contact either your family doctor or pediatrician. If you are having problems contacting them, give us a call.

If your child has multiple relapses requiring repeated courses of prednisone they may need other forms of treatment.

Prednisone treatment for relapse

It is very likely that your child will have one or more relapses after their initial episode. Unfortunately we cannot predict the timing or frequency of relapses. Urine dipsticks are used to confirm a relapse at home *(Figure 4)*. Relapses will be treated with daily prednisone until your child's first morning dipsticks are negative or trace for 3 days in a row (remission). Once back in remission, the prednisone dose will be reduced to alternate day treatment for 2 weeks as outlined in *Figure 7* and *Table 3*. The treatment of relapses is shorter than the initial treatment.

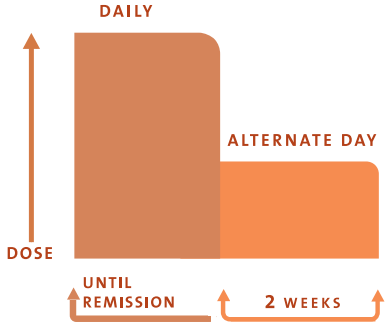


Figure 7. Time course of prednisone treatment for relapses

TABLE 3

Prednisone dosing for your child's relapse of nephrotic syndrome

DURATION	DOSE BY CHILD'S SIZE (mg/m²)*
Until remission	60 mg/m² daily (max 60 mg)
2 weeks	40 mg/m² alternate day (max 40 mg)
STOP	

*Once daily in the morning on the days prednisone is given

What will happen after your child starts on prednisone for a relapse of nephrotic syndrome?

After starting prednisone, one of three things will happen:

Your child's condition will get better: If your child goes into remission within the first 2 weeks of relapse treatment, you should contact your doctor to discuss the details of decreasing the prednisone. You do not have to be seen in clinic at this point.

Your child's condition will stay the same: If by 2 weeks after starting relapse treatment your child has not gone into remission, you should contact your doctor. Your child will be seen in clinic at the 4-week mark of starting relapse treatment to be sure there are no concerns.

Your child's condition will get worse: If after starting relapse treatment your child becomes more swollen or develops any of the other complications (see *Section 5 Complications of nephrotic sydnrome*), you should contact your doctor as your child may need to be seen promptly.

Different forms of prednisone

Prednisone can be given in both tablet and liquid form. Tablets come in different strengths (1, 5 and 50 mg). We will use whatever tablets are most convenient for your child to help make taking prednisone quick and easy for them. The 5 mg tablets are generally small and easier to swallow *(Figure 8)*.

The liquid form is called prednisone or prednisoLONE. The concentration of liquid prednisone can vary from 1 mg/mL to 5 mg/mL depending on the pharmacy, so it is important to be aware of the exact concentration you are provided.

Figure 8. 5 mg prednisone tablets are close in size to the end of a pencil.

Tips on how to give your child prednisone at home

Tablet prednisone (50 mg, 5 mg, 1 mg)

Have your child swallow the tablet(s) quickly. If it sits on the tongue it begins to dissolve and has a bitter taste. You may crush the tablet(s) and give with a small spoonful of soft food such as jam, ice cream, yogurt, pudding, jello, custard or similar. Put the powdered tablet onto the food, and ensure the entire amount is swallowed.

Liquid prednisone

Shake well before measuring the dose in the plastic syringe provided. Give the prednisone as is (do NOT dilute). Place the oral syringe into your child's cheek toward the back of the mouth and gently squeeze the prednisone into their cheek. Blow on your child's face as it stimulates them to swallow. You may need to give the dose of prednisone in repeated smaller amounts over 10 or 15 minutes, especially if the amount is large. After the prednisone, you may give your child water or a strong tasting food or drink, but NOT their favorite as they may learn to dislike it. You may need to swaddle your child in a towel or hug them tightly to give this medicine if they are resistant. You may even need to hold your child's nose to make them open their mouth.

Points to remember

DO give prednisone with food. Prednisone can cause upset stomach. This can be reduced by giving your child their prednisone with food. Giving prednisone in the morning with breakfast is recommended.

DO NOT try to hide prednisone in food, like a bowl of cereal or other food. Your child may not finish the food, they may develop a distrust of you and of food, or they may start to hate a previously loved food or drink.

DO NOT dilute the prednisone in a glass of liquid. Your child may not drink the whole amount and would not get the full dose.

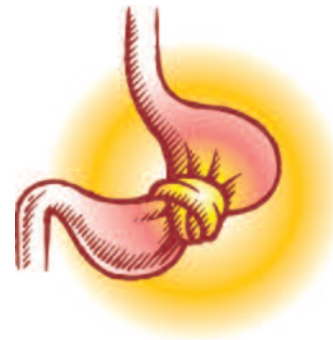
DO NOT freeze liquid prednisone. Freezing may weaken the prednisone so that it is not effective.

Prednisone side effects

Weight gain

Prednisone often causes an increase in appetite. Because of this, it is common for children on prednisone to gain extra weight. Making healthy food choices can help minimize extra weight gain *(see Section 7 Diet)*. Your child's appetite will likely normalize once they are off daily prednisone. Being physically active will also help minimize extra weight gain. All children taking prednisone are encouraged to remain physically active.

Stomach pain

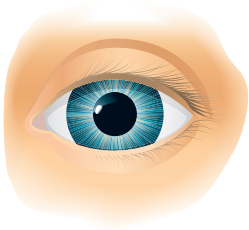


While on daily prednisone, your child may experience stomach pain (gastritis) or heart burn (esophagitis). These side effects get better as the prednisone dose is decreased. **If the stomach pain or heartburn continue, let your family doctor or pediatrician know as it can be treated with medications such as Ranitidine.**

High blood pressure

While on daily prednisone, your child may experience high blood pressure. The most common symptoms of high blood pressure are headache and sometimes dizziness and shortness of breath. However, because many children with high blood pressure do not have any symptoms at all it is important to record the blood pressure at each visit to the doctor. If necessary, high blood pressure will be treated. **If any symptoms of high blood pressure occur, let your family doctor or pediatrician know.**

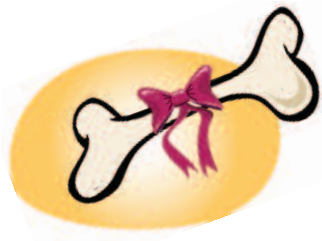
Eye problems



Long-term use of prednisone can cause eye problems. The two most common eye problems seen are cataracts and glaucoma. Cataracts may present as blurred vision and glaucoma may present as eye pain. Symptoms are infrequently seen with either condition. Your child will be referred for a check-up with an eye doctor (ophthalmologist) during their initial prednisone treatment and then later if needed.

Bone problems

Prednisone may cause bone problems, including decreased bone density (osteopenia) and increased risk of bone fractures. These are more of a concern in children who require repeated courses of prednisone over time due to frequent relapses of their nephrotic syndrome. It is important to meet Health Canada's recommended daily intake for calcium and vitamin D. Staying active and maintaining a healthy weight are also important in helping reduce the risk of bone disease and fractures.



To ensure children with nephrotic syndrome get enough calcium and vitamin D, we recommend supplementing 500 mg elemental calcium and 800–1000 IU vitamin D on top of their diet *(see Section 7 Diet)*. **If your child develops bone pain or has a bone fracture, contact your family doctor or pediatrician.**

Cosmetic changes

While on prednisone, your child may experience extra hair growth on their face and body (hirsutism), hair thinning, acne, skin streaks (striae), an enlarged belly, or a moon-shaped face (Cushingoid facies). These are well described side effects of prednisone that will get better and usually disappear as the prednisone dose is decreased.

Behaviour changes

While on prednisone, your child may experience mood changes, difficulty sleeping, or hyperactivity. These are well described side effects of prednisone that will get better and usually disappear as the prednisone dose is decreased.

Low adrenal gland function

The adrenal glands help the body respond to stress, like illnesses or accidents. The use of prednisone can lower the function of the adrenal glands and make it harder for the body to respond to stressful events. The most common symptoms of low adrenal gland function are tiredness (fatigue), low appetite, weight loss, and muscle weakness. These side effects usually get better once your child is completely off prednisone for a period of time. If they do not get better then further treatment is sometimes needed. Your doctor may recommend certain tests to monitor for this.

SECTION 7

Diet

Diet plays an important role in the management of your child's nephrotic syndrome. Our recommendations for your child's diet are particularly important around the time of diagnosis and during relapses. Making healthy food choices will help to reduce swelling and control blood pressure, prevent excessive weight gain during prednisone treatment, and keep bones healthy. Your child's diet will be reviewed at initial diagnosis, then again at the 4-week and 12-month clinic visits and more frequently as needed. Use the 3-day Food Intake Record sheets provided in the *Worksheets* to track your child's intake at 4-weeks and 12-months.

The dietitians at BC Children's Hospital are available to answer any of your questions. In situations where a dietitian is not available, you can also call HealthLink BC (dial 8-1-1).

Sodium and Fluid

We have two approaches to help reduce swelling associated with nephrotic syndrome. **First, we recommend a diet limited in sodium for the duration of prednisone treatment. Second, we also recommend a daily fluid limit until your child is in remission.** Reducing the amount of sodium in your child's diet makes your child less thirsty, making it easier to stay within their prescribed fluid allowance. Our recommendations for the amounts of sodium and fluid are based on your child's gender and weight.

Table 4 shows the maximum amount of sodium in milligrams (mg) your child should consume each day while they are taking prednisone. *Table 5* shows the maximum amount of fluid in milliliters (mL) your child should consume each day until remission.

For example, if your daughter weighs 20 kg (44 pounds) and she has a relapse of her nephrotic syndrome, we recommend maximum 1,300 mg of sodium per day for the time she is taking prednisone

(Table 4) and 800 mL of fluids per day until she goes into remission *(Table 5)*. Careful attention to the sodium content of foods you are offering, as well as dividing the fluid allowance across the day will help your child to adhere to the recommended sodium and fluid limits.

Decreasing sodium and fluids beyond these recommendations is not necessary. Recommendations for your child's diet will be reviewed with you at your visits to the clinic.

TABLE 4

Recommended maximum daily sodium intake while on prednisone

WEIGHT (kg)	SODIUM INTAKE (mg/day)*	
	BOYS	GIRLS
10–19	800	800
20–29	1300	1300
30–39	1600	1400
40–49	1700	1600
50–59	2000	1700
60–69	2300	1800
70 and above	2400	1900

*While your child is on prednisone

TABLE 5

Recommended maximum daily fluid intake until remission

WEIGHT (kg)	FLUID INTAKE (mL/day)*
10–19	500
20–29	750
30–39	900
40–49	1000
50–59	1100
60–69	1200
70and above	1300

*Until your child is in remission

* Provide the following foods with caution as they are made up of a large amount of water: pudding, yogurt, jello, popsicles, watermelon and pureed fruits

To help achieve the recommendations for sodium and fluid, extra attention to your child's diet will be needed. Choosing foods that fit can be a challenge! Here are some tips to get you started:

What is sodium and where is it found?

Sodium is a mineral found in table salt, baking soda and many food preservatives. Small amounts of sodium occur naturally in foods while foods that have been processed may have high amounts of sodium. About 80% of the sodium that Canadians consume comes from salt that has been added to foods during processing.

Table 6 illustrates how the sodium content of foods in their natural form increases as they are processed.

TABLE 6

Sodium content of foods

HEALTHY CHOICE	LESS HEALTHY CHOICE	UNHEALTHY CHOICE
Cucumber 7 mg	Cucumber with Ranch dressing 145 mg	Dill pickle 1 medium 928 mg
Tomato 1 small 14 mg	Tomato ketchup 2 packages 140 mg	Tomato sauce ½ cup 640 mg
Chicken ½ breast 69 mg	Chicken nuggets + sauce (4 nuggets) 670 mg	Chicken luncheon meat 3 ounces 1059 mg
Pork 3 ounces 59 mg	Bacon 4 slices 548 mg	Ham 3 ounces 1,114 mg
Roast beef 3 ounces 54 mg	Hot dog weiner 1 regular 487 mg	Pepperoni 3 ounces 892 mg
Cheddar cheese 1 ounce 173 mg	Processed cheese slice 1 slice 390 mg	Cheese spread 2 tablespoons 540 mg
Cooked oatmeal 1 cup <1 mg	Bread 2 slices 298 mg	Instant noodles 1 serving 2,200 mg
Shredded Wheat 1 round <1 mg	Rice Krispies® 1 cup 298 mg	
Steamed rice/cooked pasta 1 cup <1 mg		
Olive oil 1 tablespoon <1 mg	Salted butter/margarine 1 tablespoon 81/70 mg	Soy sauce 1 tablespoon 1,029 mg
Lemon juice 1 tablespoon 1 mg		Salt 1 teaspoon 2,325 mg
Unsalted butter/margarine 1 tablespoon 2/0 mg		

As you move from left to right in *Table 6* you will notice that foods have increasing amounts of sodium. The preferred foods would be those on the left in the green boxes as they have less sodium. Foods listed on the far right in the red boxes contain the most amount of sodium and should be avoided.

Sample menu

The menu plan in *Table 7* was created for a 10-year-old child being treated with prednisone for nephrotic syndrome. The menu suggestions are low in sodium, relatively low in fat, and contain adequate calcium, all important recommendations for children with nephrotic syndrome. A diet low in sodium can be healthy and enjoyable for ALL family members.

TABLE 7

Sample menu

MEAL	LOW SODIUM SUGGESTION
Breakfast	1 cup Shredded Wheat/Mini Wheats® 1 cup 1% milk (½ on cereal + ½ to drink) 1 medium banana
Morning snack	1 orange
Lunch	1 jam sandwich 2 graham crackers ½ cup yogurt 1 small apple
Afternoon snack	3 cups unsalted popcorn Carrot sticks
Dinner	2 oz chicken breast 1 cup steamed rice 1 cup green salad 1 tablespoon oil + balsamic vinaigrette ½ cup steamed broccoli ½ cup corn niblets 1 cup of 1% milk
Evening snack	½ cup applesauce 1 cup 1% milk

In order to choose the right foods for your child, it is important to read and understand food labels. Be sure to check labels for the sodium content of any packaged foods. See *Reading food labels* for some additional information to help you determine how much sodium is in the packaged foods you choose.

Reading food labels

In Canada, packaged foods are required to have a Nutrition Facts box on the label. By law, sodium is one of 13 items that is required to be reported on the package label. When reading labels, follow these simple instructions to determine whether the food item is appropriate for your child with nephrotic syndrome:

1. **Note the number of calories provided by the serving size listed.**
2. **Note the number of mg of sodium in the serving.**
3. **Choose food items that have LESS sodium mgs than calories.**

Figure 9 and *Figure 10* are two examples where reading the labels helps make the right choice:

Nutrition Facts

Per 125 mL (87 g)*

Amount	% Daily Value*
Calories 80	
Fat 0.5 g	**1%**
Saturated 0 g + Trans 0 g	**0%**
Cholesterol 0 mg	
Sodium 0 mg	**0%**
Carbohydrate 18 g	**6%**
Fibre 2 g	**8%**
Sugars 2 g	
Protein 3 g	

Vitamin A	2%	Vitamin C	10%
Calcium	0%	Iron	2%

Figure 9. This nutrition label shows the food item provides 80 calories with 0 mg of sodium. The number of mg of sodium is less than the number calories, therefore, an appropriate low-sodium choice.

Nutrition Facts
Valeur nutritive

Per 1 tray (212 g)/Pour 1 plat (212 g)

Amount Teneur	% Daily Value % valeur quotidienne
Calories/Calories 170	
Fat/Lipides 2.5 g	**4%**
Saturated/Saturés 0.5 g + Trans/Trans 0 g	**3%**
Cholesterol/Cholestérol 25 mg	**8%**
Sodium/Sodium 620 mg	**26%**
Carbohydrate/Glucides 25 g	**8%**
Fibre/Fibres 2 g	**8%**
Sugars/Sucres 2 g	
Protein/Protéines 12 g	
Vitamin A/Vitamine A	**4%**
Vitamin C/Vitamine C	**15%**
Calcium/Calcium	**2%**
Iron/Fer	**8%**

Figure 10. This nutrition label shows a food that contains 170 calories with 620 mg of sodium. The number of mg of sodium is nearly four times the number of calories, and is NOT an appropriate low-sodium choice.

Calcium and vitamin D

An important side effect of nephrotic syndrome and being treated with prednisone is long-term bone problems. We therefore recommend a minimum daily calcium and vitamin D intake, which depending on how much prednisone your child needs, may vary from time to time (see *Section 6 Treatment of nephrotic syndrome*). **To ensure children with nephrotic syndrome get enough calcium and vitamin D, we recommend supplementing 500 mg elemental calcium and 800–1000 IU vitamin D on top of their diet while they are on prednisone.**

What foods provide calcium?

Calcium is found in many foods. Your child can get recommended amounts of calcium by eating a variety of foods, including the following: milk and milk alternatives such as yogurt, cheese and fortified plant-based beverages (such as fortified soy beverages); dark green vegetables such as broccoli, kale and spinach; fish with soft bones that are eaten, such as canned salmon or sardines.

What foods provide vitamin D?

The major sources of vitamin D are vitamin D-fortified foods, including cow's milk and margarine. Goat's milk, plant-based beverages such as soy, and some calcium-fortified orange juices may not be fortified with vitamin D. Cheese and yogurt can be made with vitamin D-fortified milk, but the final product does not contain as much vitamin D as fluid milk alone. The only natural sources of vitamin D in the Canadian food supply are fatty fish and egg yolks.

Other important diet modifications

Prednisone is an important medication in the treatment of nephrotic syndrome. This medication can have side effects such as an increased appetite which may lead to weight gain and it can also cause high blood sugar. Limiting intake of sugary foods and choosing the right type and amount of fat will help to reduce weight gain.

Protein

Even though your child may be losing protein in the urine, we do not recommend increasing dietary protein.

Sugars

Limit the sugar your child eats by avoiding fruit juice or limiting intake to a maximum of 1 cup/day. Avoid beverages with added sugar such as fruit drinks, soft drinks, pop, fruit punch, iced tea and lemonade.

Encourage your child to eat fruit and to drink water or milk as prescribed. Limit candies, cookies, ice cream and pastries.

Fats

Strategies to help limit excessive weight gain include limiting foods with saturated fats and cholesterol, cooking with oils that are liquid at room temperature, and using soft unsalted margarines in place of butter or stick margarines. Serve low-fat or nonfat milk and yogurts. Cut the visible fat off of meat and poultry. When choosing packaged foods, look for foods made without saturated or trans fats.

SECTION 8

Immunizations and infections

Because of your child's nephrotic syndrome and the prednisone used to treat this condition, your child will be unable to fight infections properly. They will be at greater risk of getting sick. Viral infections, including the seasonal flu and chickenpox (varicella) may be severe. Your child is also at higher risk for developing bacterial infections of the belly (peritonitis), skin (cellulitis), and lungs (pneumonia).

Good hand hygiene, regular visits to the doctor and dentist, and ensuring your child's immunizations are up-to-date will help lower their chance of getting sick. If possible, they should also minimize contact with noticeably sick people. However, drastic changes to their normal day-to-day routine are not recommended. With responsible monitoring, they can still go to school, join activities, and play with friends. If you have any questions or concerns about infections, contact your doctor.

Points to remember

1. Contact your doctor if you have any concerns about infection.
2. Ensure your child's immunizations are up-to-date.
3. Avoid live vaccines while your child is in relapse and/or is on prednisone.
4. Bring your child's immunization records to your clinic visit and each time they get a new vaccination.

Live vaccines to avoid

Live vaccines are vaccines that contain a weak but living form of virus. Since children with nephrotic syndrome are at greater risk of getting sick, live vaccines should be avoided while they are in relapse and/or on prednisone. Live vaccines to avoid include MMR (Measles, Mumps, Rubella), varicella (chickenpox), intranasal influenza (FluMist®),

yellow fever, and BCG (Bacillus Calmette-Guerin). Note that intranasal influenza, yellow fever, and BCG are not part of the routine British Columbia immunization schedule *(www.immunizebc.ca)*.

Influenza

Your child can have a severe course of influenza A or seasonal flu infection. These infections can often trigger a relapse of nephrotic syndrome. We recommend annual influenza immunizations regardless of whether your child is in remission, in relapse, or if they are taking prednisone. To further reduce the risk of infection to your child we also recommend immunizing all family members during flu season, which typically starts in the fall.

A live influenza vaccine is now available as a nasal spray (FluMist®). However it may not be as effective and we do not recommend its use.

Chickenpox (varicella)

Typical chickenpox starts with a fever followed by a red rash that often develops into little blisters *(Figures 11a, 11b)*. The chickenpox virus spreads through the air (by coughing and sneezing) and by direct contact with mucus, saliva, or fluid from the blisters. Chickenpox is contagious from about 2 days before the rash appears and until all the blisters are crusted over.

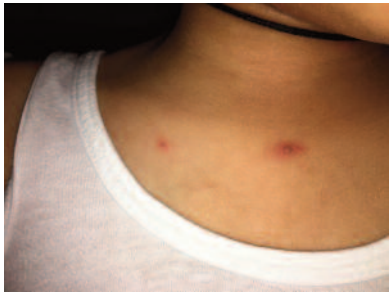


Figure 11a. Patient with varicella blisters on the neck resembling "dew drops on rose petals."

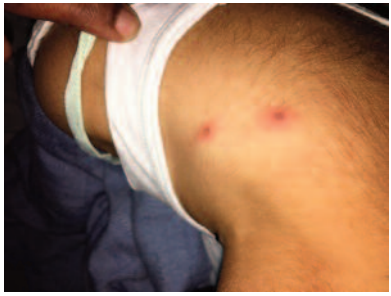


Figure 11b. Patient with varicella blisters on the shoulder.

Your child should be immunized with the varicella vaccination. In British Columbia, chickenpox immunization is given at 12 months and again at 4–6 years of age. We consider children to be likely immunized if they had at least one chickenpox immunization. Severe chickenpox infections are a real risk for all non-immunized children

with nephrotic syndrome. If your child has not been fully immunized for chickenpox (has not had 2 immunizations), they should get their vaccination once they are in remission AND off prednisone for more than 4 weeks. To further reduce the risk of infection to your child, we recommend that all the members of your family also be immunized if they have not had chickenpox or have not been immunized.

If your child has not had chickenpox, has not been fully immunized AND is exposed to anyone who has chickenpox or shingles, contact your doctor immediately. If your child develops signs of chickenpox, even if they have been previously immunized, contact your doctor immediately.

Pneumococcus

Your child is at risk for developing severe infections from a bacterium called pneumococcus. These infections include severe belly infections (peritonitis), severe lung infections (pneumonia), and severe skin infections (cellulitis). Therefore it is important that your child receives all their pneumococcal immunizations according to the provincial schedule *(www.immunizebc.ca)*. In British Columbia, standard pneumococcal immunizations are given at 2 months, 4 months, and 12 months of age. Most children who present with nephrotic syndrome are fully immunized by the time that they are diagnosed. However, due to the higher risk of infection with specific types of pneumococcus that are not covered with the standard schedule, we recommend an additional one time pneumococcal vaccination (Pneumovax™) that can be arranged through the family physician or closest public health clinic.

SECTION 9

Ongoing care

Follow-up during the first year

After your child's initial diagnosis they will need to be followed closely over the first year. The goals of follow-up during this time are to document their response to prednisone and to look for complications of nephrotic syndrome or for possible side effects from prednisone treatment. We recommend clinic visits within 1 week from initial diagnosis, then at 4 weeks, 12 weeks, 6 months, 9 months, and 12 months after diagnosis (see *Table 8 Schedule of ongoing care*).

The 1-week clinic visit will be used for teaching. We want to make sure that your child is properly started on prednisone after diagnosis and that you are comfortable with home monitoring, such as using dipsticks and knowing when to call your doctor about complications.

The 4-week visit will be used for more teaching and to check the response to prednisone treatment. Your child should also have a check up with an eye doctor (ophthalmologist) around this time. This is to check for possible eye problems related to prednisone. Your doctor may also request other tests for your child at this time. These tests may be done to review bone health or adrenal gland function, as needed.

An extra 6-week visit will only be necessary if your child has still not yet responded to prednisone.

At 12 weeks, if all has gone according to plan, your child will be off prednisone. The 12-week visit will focus on monitoring the effects of prednisone treatment.

At 6, 9, and 12 months, visits will focus on your child's growth and the education around treatment and management of future relapses. More visits may be required based on your child's individual needs.

At all visits, general good health practices such as routine dental check-ups and up-to-date immunizations will be reviewed. A complicated course or unusual presentation may require more attention, including ongoing support from our dietitian or other health care workers.

Table 8 summarizes when the visits in the first year will be scheduled and what will be reviewed.

TABLE 8

Schedule of ongoing care

TEST/REVIEW	CLINIC VISIT						
	0 w*	4 w	6 w**	12 w	6 m	9 m	12 m
Confirm and explain diagnosis	✓						
History							
• History of swelling	✓						
• Review immunization status	✓	✓	✓	✓	✓	✓	✓
• Document date and time to remission		✓	✓				
• Review Handbook Worksheets		✓	✓	✓	✓	✓	✓
• Review prednisone schedule		✓	✓	✓	✓	✓	✓
• Review medication history	✓	✓	✓	✓	✓	✓	✓
Physical examination							
• Blood pressure, heart rate and temperature	✓	✓	✓	✓	✓	✓	✓
• Record growth (height, weight)	✓	✓	✓	✓	✓	✓	✓
• Check for swelling	✓	✓	✓	✓	✓	✓	✓
• Other signs of prednisone side effects (eyes, bone, skin)		✓	✓	✓	✓	✓	✓
Lab testing							
• CBC, albumin, urea, creatinine, Na^+, K^+, Cl^-, HCO_3^-	✓	No regular blood testing needed					
• Urinalysis, microscopy, and PCR	✓	✓	✓	✓	✓	✓	✓
Monitoring							
• Review fluid restriction	✓	✓					
• Review dietary recommendations	✓	✓		✓			✓
• Review home monitoring (urine dipsticks)	✓	✓	✓	✓	✓	✓	✓
• Eye clinic evaluation		✓					
• Family given handbook & worksheets	✓						
• Schedule follow-up appointment	✓	✓	✓	✓	✓	✓	✓

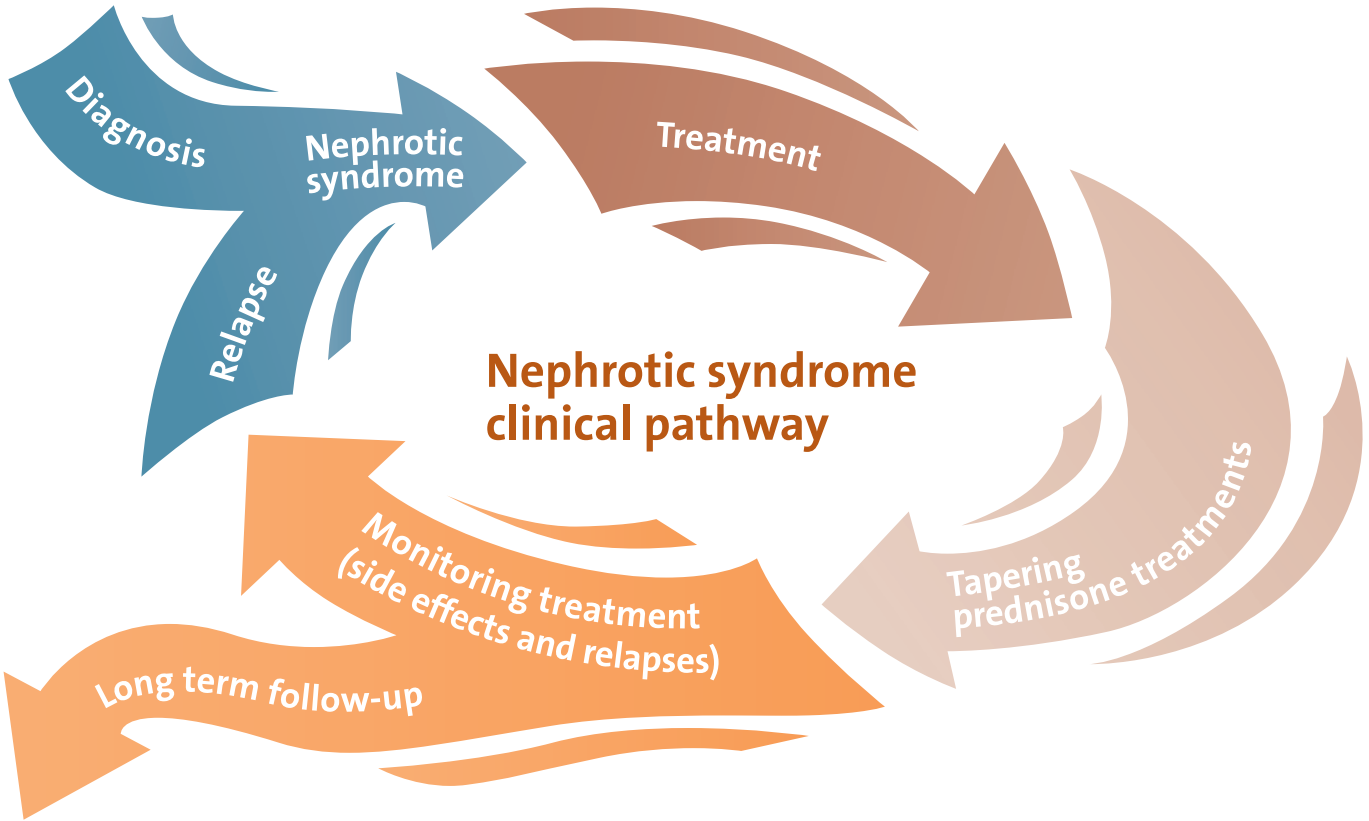
* Visit at which prednisone is started. A subsequent clinic visit within 1 week is likely needed to complete all necessary tests and teaching

** A six-week visit is required if your child is not in remission at 4 weeks

Follow-up after the first year

After the first year the frequency of your child's visits will depend on how well they are doing and how often they relapse. Most of the time relapses can be dealt with over the phone with your family doctor or pediatrician. No extra lab tests will be needed. If your child has complications during a future relapse, such as severe swelling or no response to prednisone, you may need to return to the clinic for more treatment.

If your child continues to have relapses of their nephrotic syndrome, they will need to be seen more frequently to be sure they have responded properly to treatment and that they have not developed side effects of treatment. Regardless of course, most children will need to be seen by a kidney doctor (nephrologist) at least once a year.


Diagnosis
Nephrotic syndrome
Relapse
Treatment
Tapering prednisone treatments
Monitoring treatment (side effects and relapses)
Long term follow-up
Nephrotic syndrome clinical pathway

Appendix

Commonly used terms

Albumin	Protein made by the liver that makes up nearly half of the total blood proteins
Ascites	Accumulation of fluid in the abdominal cavity
Biopsy	Sample of body tissue or cells for testing including microscopic review
Cellulitis	Infection of the skin, causing localized swelling, redness and pain
Childhood onset nephrotic syndrome	Nephrotic syndrome presenting between 1 and 18 years of age
Creatinine	Waste product of muscle that is filtered by the kidneys (used as a marker for kidney function)
Frequent relapsing nephrotic syndrome	Nephrotic syndrome with 4 or more relapses in any 12 months or 2 or more relapses in the first 6 months after presentation
Glomerulus	Filter unit of the kidney
Hematuria	Blood in the urine — may be either visible to the eye (macroscopic) or only under the microscope (microscopic)
Hypertension	High blood pressure
Immunocompromised	State of decreased resistance to infections because of disease or the use of certain drugs
Minimal change disease	The most common type of nephrotic syndrome in children who respond to prednisone. It is characterized by normal looking kidney filters (glomeruli) when examined under a microscope
Nephrologist	Doctor specializing in kidney disease
Nephrotic syndrome	Clinical diagnosis consisting of swelling, low blood albumin and heavy protein losses in the urine
Peritonitis	Infection of the layers lining the abdominal cavity
Proteinuria	Loss of proteins in the urine, usually discovered by a urine dipstick
Relapse	Three days in a row of 3+ or 4+ protein on urine dipstick
Remission/response	Three days in a row of negative or trace protein on urine dipstick
Steroid dependent nephrotic syndrome	Nephrotic syndrome responsive to prednisone, but relapses occur while on steroids or within 14 days after stopping steroids
Steroid resistant nephrotic syndrome	Nephrotic syndrome not responding to prednisone after 6 weeks of daily prednisone therapy

Notes

Notes

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BC CHILDREN'S HOSPITAL
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