

Childhood Nephrotic Syndrome



A guide for children and families on the treatment and management of Nephrotic Syndrome

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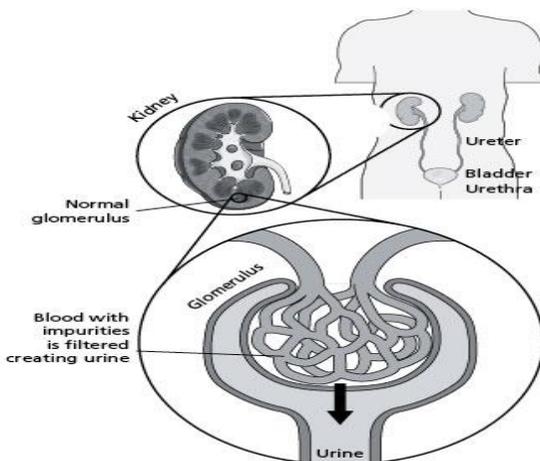
Introduction:

Your child has recently been diagnosed with Nephrotic Syndrome. This booklet aims to help you understand the treatment and management of nephrotic syndrome and to answer any questions that you may have.

The Kidneys:

Most people have 2 kidneys, situated on either side of the mid-spine. Kidneys although small keep the environment in the body balanced by filtering our blood to remove waste products and excess fluid from our body.

The kidneys are bean shaped organs which are made up of millions of tiny filtering glomeruli which filter the blood like a sieve. The kidneys remove waste products from the blood, whilst preventing loss of useful components like blood cells and protein.





What is Nephrotic syndrome?

Nephrotic Syndrome is a problem with the sieve mechanism of the kidney. The holes within the sieve enlarge, allowing large amounts of protein to be lost from the blood stream into the urine. As a result the levels of protein (albumin) in the blood stream fall.

Why do we need protein in the blood?

Protein in the blood acts like a magnet. It keeps fluid in the blood vessels where it is meant to be. When protein levels fall, fluid seeps out of the blood vessels and into the tissues of the body causing oedema (swelling).

What causes Nephrotic Syndrome?

Nephrotic Syndrome often presents between the ages of 2 years and 5 years, affecting twice as many boys as girls. We would expect to see about 50 children who develop nephrotic syndrome each year. In most cases there is no known cause for Nephrotic Syndrome. It is linked to a disruption in the immune system and may occur after a cold or a virus. In nephrotic syndrome, the immune system can interfere with the tiny filters in the kidneys, known as glomeruli.

Types of Nephrotic Syndrome

There are different types of nephrotic syndrome, which depend on whether they can be treated with steroids. It is often not possible to find out which type your child has until after they have taken a course of steroids. Some causes of nephrotic syndrome can only be diagnosed (identified) in a kidney biopsy in which a tiny piece of one kidney is removed from the body with a needle and examined under special microscopes. Not all children with nephrotic syndrome require a biopsy.

- About 9 in 10 children with nephrotic syndrome will have a type that can be treated with steroids – this is called steroid-sensitive nephrotic syndrome (SSNS) or minimal change nephrotic syndrome (MCNS).
- In some children the nephrotic syndrome keeps coming back, even after successful treatment with steroids – this is called frequently relapsing nephrotic syndrome.
- A few children (about 1 in 10) have a type that cannot be treated by steroids alone – this is called steroid resistant nephrotic syndrome (SRNS) because it is “resistant” (does not respond) to steroid treatment. Focal segmental glomerulosclerosis (FSGS) and IgM nephropathy are causes of steroid-resistant nephrotic syndrome.
- Occasionally, babies are born with congenital nephrotic syndrome.

What is a kidney biopsy?

Children who are not responding to steroid treatment and continue to have large amounts of protein in their urine may need a kidney biopsy. A kidney biopsy allows us to investigate in detail what is happening in the kidney. Kidney biopsies are performed in the X-Ray department under anaesthetic. A tiny sample of a kidney is removed from the body with a needle – special medicines are used so your child will not feel any pain and will sleep through the procedure. The sample is sent to laboratory where it is looked at under microscopes.



What are the signs and symptoms of Nephrotic Syndrome?

Large amounts of protein appear in the urine - this can make the urine appear frothy.

- ✚ Your child will appear puffy around the eyes - this is due to fluid in the tissues (also called oedema).



- ✚ Swelling of the legs is common - sometimes swelling can affect the abdomen or the groin.

- ✚ Swelling, including inside the body can cause shortness of breath or abdominal pain and diarrhoea.
- ✚ You may notice that your child's clothing has become tighter than usual and they have gained weight rapidly.

How is Nephrotic Syndrome diagnosed?

Your doctor can diagnose nephrotic syndrome by doing a physical exam and asking about symptoms.

A urine test called a urinalysis will show large levels of protein in the urine.

A blood test will show low levels of protein (albumin) in the blood.

Sometimes a kidney biopsy will need to be performed



How is Nephrotic Syndrome treated?

The safest and most effective treatment for nephrotic syndrome is a course of oral steroids. Steroids are hormones which are naturally produced by the body. We use a steroid called Prednisolone to treat nephrotic syndrome. Prednisolone is a synthetic hormone which can be given by mouth. Most children respond well to steroid treatment. Most children respond to steroids within about 2 weeks. Research has shown that steroids need to be given for several months for the first episode to give children the best chance of only having one episode of nephrotic syndrome.

Remission

Remission is when protein disappears from the urine and the swelling is lost from the face and body.



Relapse

If protein comes back in the urine we call this a 'relapse'. Almost 70% of children with nephrotic syndrome will have a relapse at some point in the future.

Relapses are frequently triggered by coughs, colds or infection. It is not unusual to have + or ++ of protein in the urine every now and then.

A relapse is 3+ or 4+ of protein in the urine for MORE THAN 3 DAYS in a row.

Relapses are also treated with steroids. However, a different steroid weaning plan may be used and other medicines may be added once in remission.

Steroids

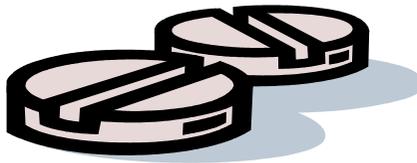
The first line of treatment for nephrotic syndrome is a steroid treatment called Prednisolone.

Steroids are made in the body but can also be made as medicines. It is important that you follow your doctor's instructions about when and how much to give. Continue to give the medicine to your child as your doctor has told you, even if he or she is getting better.

Do not stop this medicine suddenly.

What are the side effects of steroids?

Steroids have some unwanted side effects and this depends on the dose your child requires. Some side effects your child may experience include:



Weight gain:

- Steroids increase the appetite.
- Avoid sugary foods and high calorie drinks.
- Use healthy alternatives-fruit, vegetables, sugar free drinks, brown bread instead of white.
- You may meet with a dietician to discuss this.

High blood pressure:

- Steroids can increase your child's blood pressure due to increased salt and water retention.
- Avoid processed or salt containing foods if blood pressure is raised.

- We recommend regular blood pressure checks when on high dose steroids.
- Please see the diary for suggested times at which blood pressure should be checked.
- High blood pressure can be treated with medication.

Stomach irritation:

- Steroids irritate the lining of the stomach.
- Steroids should be given on a full stomach.
- Please let us know if signs of stomach irritation occur - for example heart burn or tummy pain.
- Stomach irritation can be treated with medicines such as Ranitidine or Omeprazole.

Altered mood or behaviour:

- Steroids can make children moody or upset
- This effect tends to be short lived once children are weaned from their steroid course.
- Try to avoid giving steroids late at night as they can disturb sleep.

Reduced immunity:

- Steroids suppress the body's natural immune responses.
- Children on steroids are at increased risk of developing serious bacterial infection
- If your child has a temperature >38 C they should be reviewed urgently by a doctor
- We will do a blood test to check whether your child is immune to chickenpox and measles.
- If your child is not immune, then children with these illnesses should be avoided.

- If your child accidentally comes into contact with chickenpox or measles when they are not immune you should contact us within 24 hours if possible as treatment may be needed.
- Coughs and colds cannot be avoided, therefore unless your child is unwell, they should go to school as normal.



High blood sugar levels:

- Steroids can increase the levels of sugar in the blood.
- Please let us know if your child develops excessive thirst; passes urine more frequently or shows glucose in their urine on dipstick testing.

Skin changes:

- Steroids may make your child's face rounder and their cheeks appear rosy.
- Steroids can also cause stretch marks if weight is put on quickly.

Poor growth:

- Over a period of a year or more, high dose steroid treatment can result in poor growth.
- We will monitor your child's height and weight in clinic to look for signs of poor growth.

Thinning of the bones:

- Steroids interfere with the body's ability to build strong bones
- A dexa scan may be considered in children who are dependent on long-term steroids.

Special precautions whilst on steroid treatment:

Vaccinations:

- Please speak to your nurse or doctor if your child is due a vaccination.
- Your child should not be given any 'live' vaccines when on steroids or second line agents such as Cyclophosphamide, Tacrolimus, Cyclosporin or Rituximab. Live vaccines include the BCG (for TB); MMR, Varicella and Yellow fever.

Chickenpox and measles:

- If your child is not immune to chickenpox or measles, then they should avoid contact with children with chickenpox and measles whilst on steroids.
- Ask your child's crèche or school to inform you if there is a contact.
- If your child is in direct contact with a child who has chicken pox or measles, you must contact the hospital immediately.
- Chicken pox or measles can be more severe in children who are taking steroids or second line agents.
- Your child may need an infusion of immunoglobulin to protect them. This needs to be given as soon as possible, please contact a member of the renal team.

- If your child develops chicken pox or measles you must contact the hospital immediately as they will require intravenous treatment with an antiviral drug.



Adrenal suppression:

- Treatment with steroids suppresses the body's natural ability to produce steroid hormones during periods of stress or illness.
- **STEROID TREATMENT SHOULD NEVER BE STOPPED ABRUPTLY**
- If your child has an accident or needs an operation whilst taking steroids then A BOOSTER DOSE OF STEROID MAY BE NEEDED
- Always carry a steroid warning card-available from your pharmacy.

What do I have to do at home?

1. Check your child's **first** morning urine for protein and record the results in the DIARY.
2. Once things have settled down, the urine can be checked every second to third day.
3. Remember to check a urine specimen whenever your child has a cough, cold, is unwell or looks puffy.

4. Only the first urine passed each morning should be tested. The results of dipstick test taken later in the day are unreliable and will show more protein than is really there.
5. You should also check for blood and glucose when you look at the urine dip stick
6. It is important to keep a good record of the results of urine tests and the doses of medication given. Bring the diary with you to your clinic visits. This will show your doctor if your child is responding to the prescribed treatment.
7. Here is an example of how to fill the Diary in

| Date | Day | Prednisolone | Protein | Comments |
|------------|-------|--------------|---------|-----------|
| 14/05/2016 | Mon | 45mg | 4+ | Head cold |
| 15/05/2016 | Tue | 45mg | 4+ | BP 105/64 |
| 16/05/2016 | Wed | 45mg | 3+ | |
| 17/05/2016 | Thurs | 45mg | 3+ | |
| 18/05/2016 | Fri | 45mg | 2+ | |
| 19/05/2016 | Sat | 30mg | T | Trace |
| 20/05/2016 | Sun | - | T | |
| 21/05/2016 | Mon | 30mg | N | Negative |
| 22/05/2016 | Tues | - | N | Well |
| 23/05/2016 | Wed | 15mg | N | |
| 24/05/2016 | Thurs | - | T | |
| 25/05/2016 | Fri | 15mg | N | |

CHECK THE NUMBER AND STRENGTH OF TABLETS CAREFULLY TO MAKE THE CORRECT DOSE IS BEING GIVEN.

8. When taking steroids every day we often recommend splitting the dose into two to reduce stomach irritation e.g. 60 mg could be taken as six x

5 mg (30mg) tablets in the morning and six x 5 mg (30mg) tablets at lunch time.

Remember to store all medicines out in a safe place out of reach of children.

Clinic visits

Once your child has been diagnosed with nephrotic syndrome, they will be followed up at regular intervals in the out-patient clinic.

Even if your child is well it is important to attend these clinic appointments so that the doctor can assess your child's progress.

At each clinic visit, we will check your child's height, weight and blood pressure. It is important that you remember to bring your child's treatment diary and an early morning sample of urine. You can get urine jars from your pharmacy.

What other treatments may be necessary?

Sometimes it can be difficult to manage nephrotic syndrome on steroids alone therefore, other treatments may need to be considered.

- Cyclophosphamide- a cytotoxic drug given in small doses over 8-12 weeks.
- Cyclosporin- An immunosuppressive drug taken twice daily.
- Tacrolimus- An immunosuppressive drug taken once or twice daily.
- Mycophenolate- An immunosuppressive drug taken twice daily.

These drugs may help your child remain in remission for longer periods, therefore reducing the amount of steroids that your child needs to take.

These medications will be discussed in more detail should the need arise.

Is there a special diet?

You will meet a dietician who will discuss the following:

- Eating a healthy diet
- A 'No added salt' diet
- Vitamin D Supplements while on steroids.
- Adequate calcium intake to ensure healthy bones

Recommendations regarding healthy eating and a no added salt diet are a good idea for all members of the family.



Can my child lead a normal life?

Yes - we would encourage you to think of your child as being normal and healthy.

Children with Nephrotic Syndrome do not need to stay off school even when they relapse unless told otherwise.

There is no need to restrict your child's activities unless they are unwell.

Many children manage to continue a full range of activities even during a relapse.



What about holidays?

Unless your child is having complications it is possible to have a normal family holiday.

Let us know that you will be away.

Remember to bring a supply of steroids with you - just in case.

You can ring us from abroad if you need advice. Bring a recent medical letter with your medical history with you in case you have to present at a local hospital

Always check that you have adequate insurance cover before travelling abroad.



GET IN TOUCH WITH US IF YOU NEED HELP!

PLEASE LET US KNOW URGENTLY IF:

1. Your child starts to suffer with tummy pain when they have protein in their urine.
2. Your child starts to suffer from headaches which do not settle with paracetamol.
3. Your child is puffy and unwell.
4. If your child has diarrhoea and vomiting
5. Your child comes into contact with Chickenpox or Measles and is not immune.

Please let us know if:

1. Your child's urine, after having cleared of protein, shows 3+ or more, for more than 3 days in a row. This is a relapse.
2. Your child's urine dipstick shows glucose
3. Your child's urine dipstick shows blood that was not there previously.
4. Your child has a temperature of 38 degrees or above.
5. Your child is due any live vaccinations.

Who to call:

Clinical Nurse Specialists:

Please phone _____ and ask for bleep _____.

Medical team:

Please phone _____ and ask for the renal registrar or the renal SHO.

Renal Ward-(out of Office hours):

OLCHC- Nephro-Urology Ward: _____

CUH- Michael's C Ward: _____

Useful telephone numbers

| | |
|-----------------------------|-------|
| G.P | |
| Pharmacy | |
| Hospital switch board | |
| Renal Registrar | |
| Renal SHO | |
| Clinical nurse specialist | |
| Nephro-urology Ward (OLCHC) | |
| Michael's C Ward (CUH) | |
| Dietician | Bleep |
| Medical social worker | Bleep |
| | |
| | |
| | |
| | |
| | |
| | |

To bleep a member of staff:

1. Phone the hospital switch board on _____ and ask for bleep number

Useful websites:

www.infokid.org.uk

www.gosh.nhs.uk