

childhood NEPHROTIC SYNDROME



A guide to the treatment
and management of
Childhood Nephrotic Syndrome

CONTENTS

Introduction	1
What is Nephrotic Syndrome?	2
What is the cause of Nephrotic Syndrome?	4
What is the treatment?	5
Side-effects of prednisolone	7
What to do at home?	8
The importance of a diary	9
Keeping a diary	10
Clinic visits	12
Does the steroid treatment always work?	14
What is a kidney biopsy?	15
Side effects of Cyclophosphamide	16
Is there a special diet?	18
Immunisations	20
Will my child continue to suffer from Nephrotic Syndrome?	21
Sport and exercise	22
Holidays	23
When to call the doctor	24
The Parents Group	26
Useful telephone numbers	28




INTRODUCTION

You have recently learned that your child has Nephrotic Syndrome. This causes problems which require special attention. Hopefully as your child grows older the nephrotic syndrome will become less troublesome.

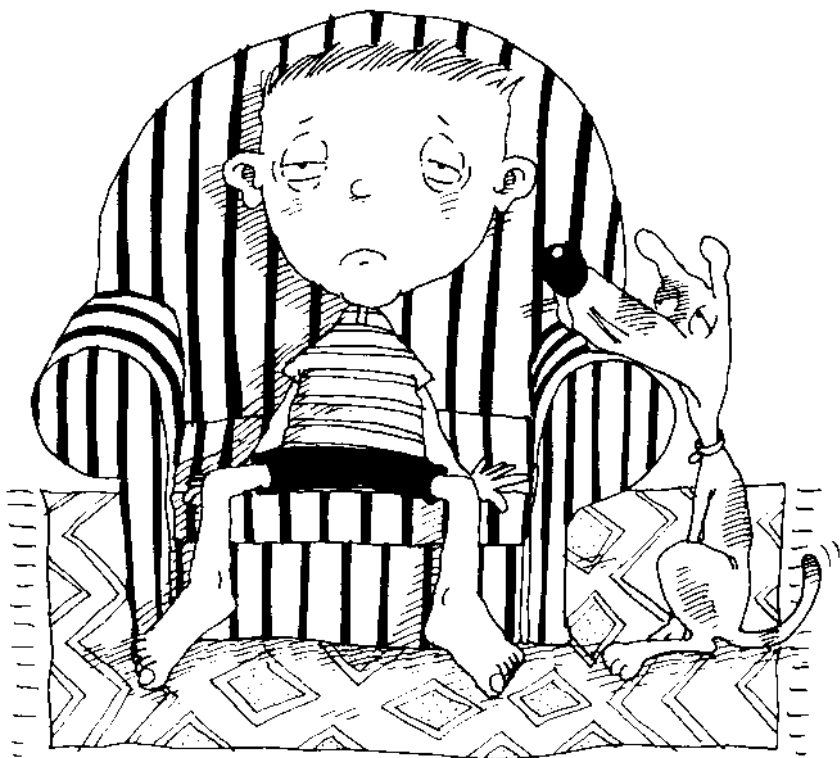
This booklet has been prepared to help parents understand the treatment and management of childhood Nephrotic Syndrome.



WHAT IS NEPHROTIC SYNDROME?



Nephrotic Syndrome occurs when the kidneys leak large amounts of protein (mostly albumin) into the urine. Protein lost in the urine results in a lower level of proteins in the blood. It is these proteins in the blood which help to hold water within the blood vessels. Therefore, when the protein level is very low, water passes into the tissues and this is recognised as swelling (oedema).

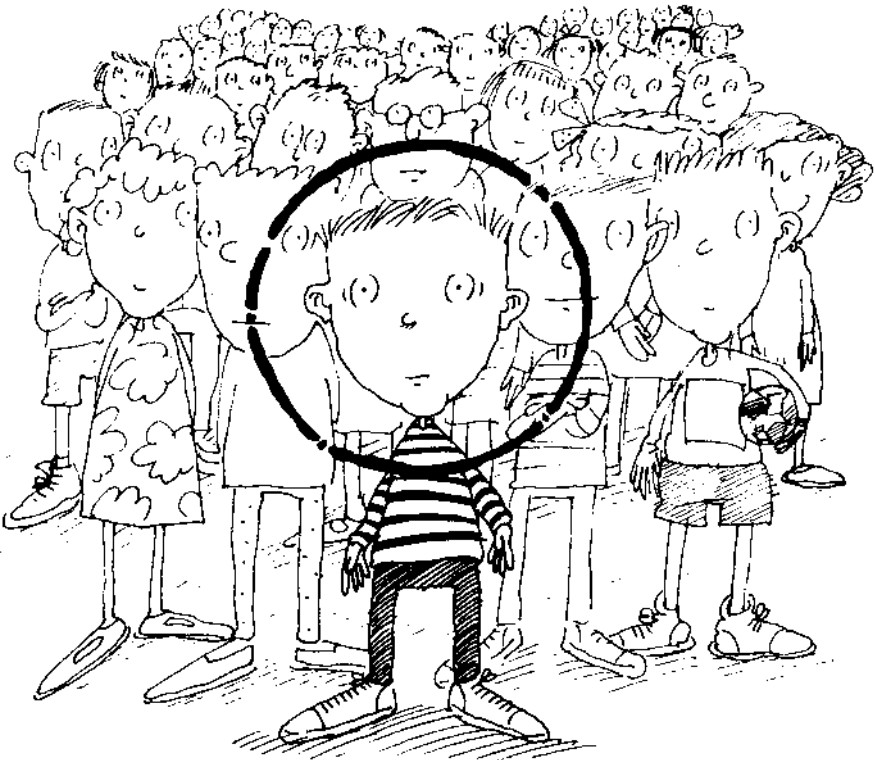


WHAT IS THE CAUSE OF NEPHROTIC SYNDROME?

The cause is still unknown but research is beginning to provide more information about the actual changes in the kidney.

Nephrotic Syndrome affects boys more than girls. The usual age at which the problem begins is 2 to 5 years.

It is a rare condition which affects approximately 1 in 50,000 children per year. The condition tends to be more common in families with a history of allergies.



WHAT IS THE TREATMENT?



Prednisolone (steroid) will be the drug prescribed for your child when he/she is first diagnosed.

Most children respond to this drug with disappearance of the protein in their urine and loss of swelling within 1-2 weeks (we call this REMISSION).

Diuretics (water tablets) may also be given to control the oedema (swelling) before the Prednisolone has worked.

Penicillin V may be prescribed if your child has a lot of swelling as excess fluid in the tissues provides a good environment for bacteria to grow.



AS A PRECAUTION



Children who are receiving steroid treatment should carry a steroid warning card with them at all times. Such steroid warning cards are available from the hospital pharmacy.

Children who have recently been on steroids may require increased doses if involved in an accident or requiring an operation. This is the reason for carrying the steroid warning card. You may wish to consider purchasing a Medic Alert pendant or bracelet (this provides emergency identification for medical problems).



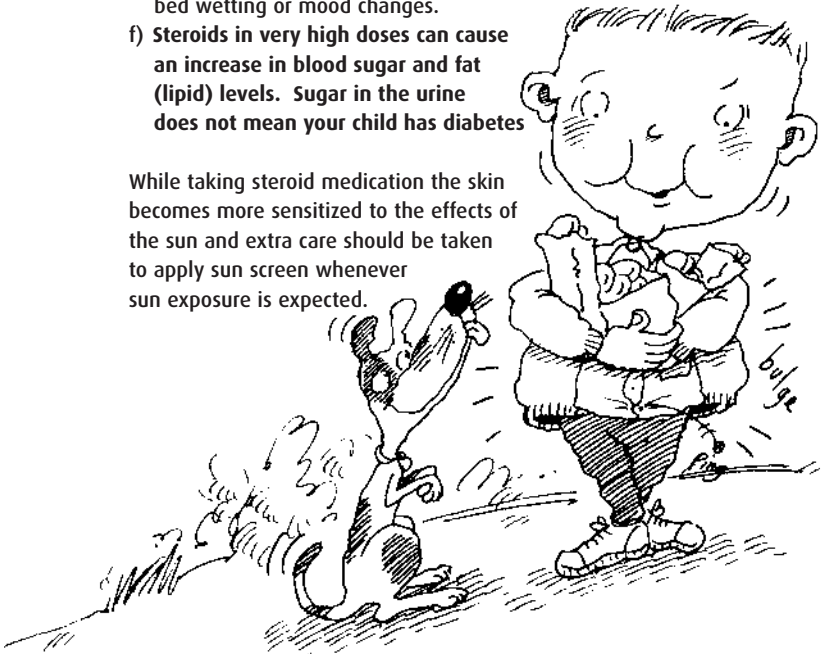
WHAT ARE THE SIDE-EFFECTS OF TREATMENT?

When Prednisolone is prescribed for short periods of time there are usually no serious or permanent side effects.

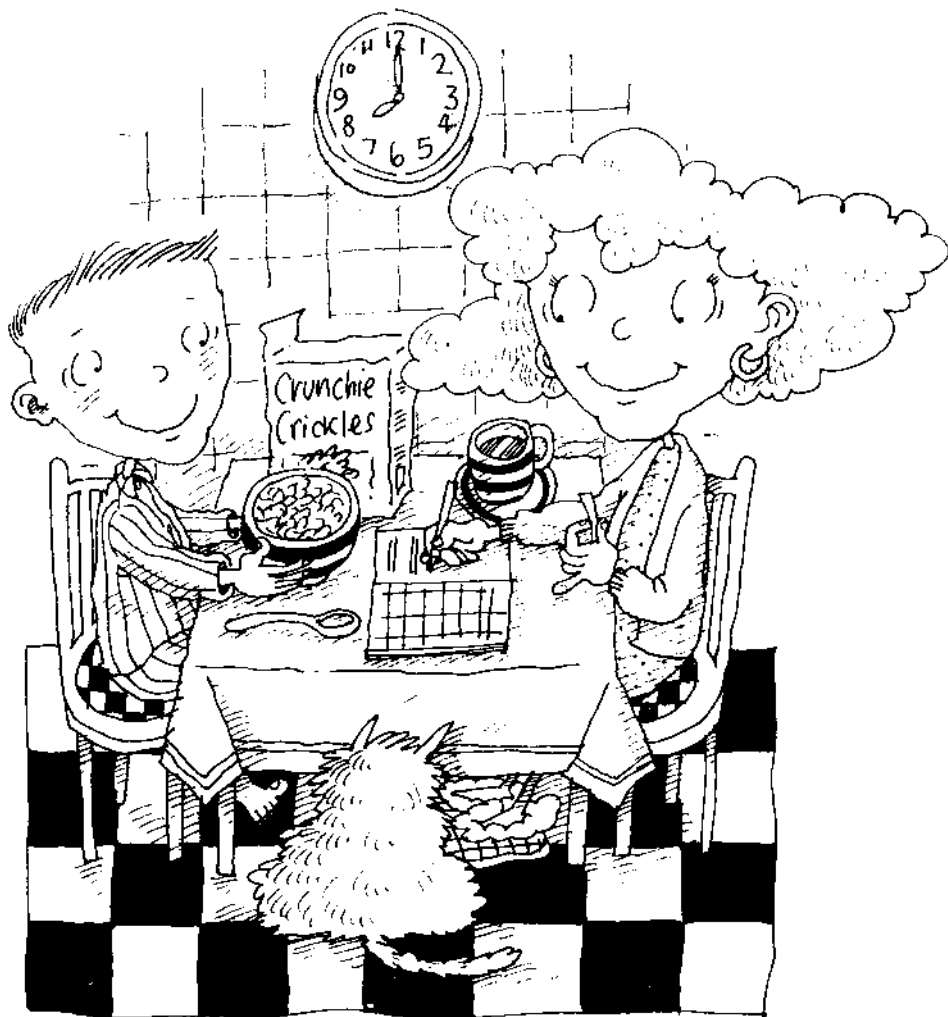
However, we have listed below the side effects which are most common

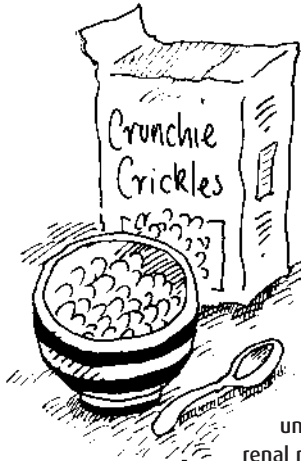
- a) Decreased resistance to infections such as coughs and colds
- b) Increase in appetite which may lead to a rapid increase in weight
- c) Flushed, swollen cheeks and stretch marks on the skin
- d) **Rise in blood pressure which will be monitored at the clinic or at home**
- e) Behavioural problems e.g temper tantrums, bed wetting or mood changes.
- f) **Steroids in very high doses can cause an increase in blood sugar and fat (lipid) levels. Sugar in the urine does not mean your child has diabetes**

While taking steroid medication the skin becomes more sensitized to the effects of the sun and extra care should be taken to apply sun screen whenever sun exposure is expected.



WHAT DO I HAVE TO DO
AT HOME?

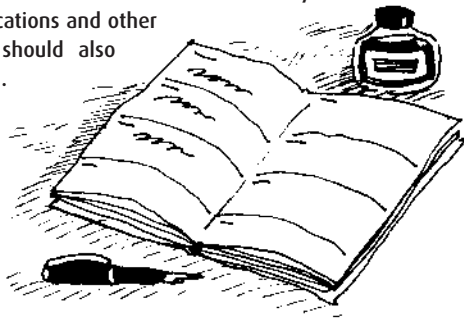




While your child is having treatment with steroids you will be taught to:

- Test your child's first morning urine for protein with the urine testing sticks on a DAILY basis, unless told otherwise by your doctor / renal nurse.

- Keep a diary of all urine results so that your doctor can review your child's progress in clinic. The dose of Prednisolone and any other medications and other comments should also be recorded.





WHY IS KEEPING A DIARY IMPORTANT?

The urine test results will show if your child is responding to the prescribed treatment. If your child has a large amount (2+) of protein in the urine for three days in a row this could mean there is a RELAPSE of the conditions and you should contact your doctor / renal nurse. If your child has NO protein in the urine for three or more days he / she is in REMISSION and should be well. Records that are complete and accurate give the doctor a good idea of your child's general health and progress and this record should be brought to each clinic visit.

Here is an example of how to record your child's progress.

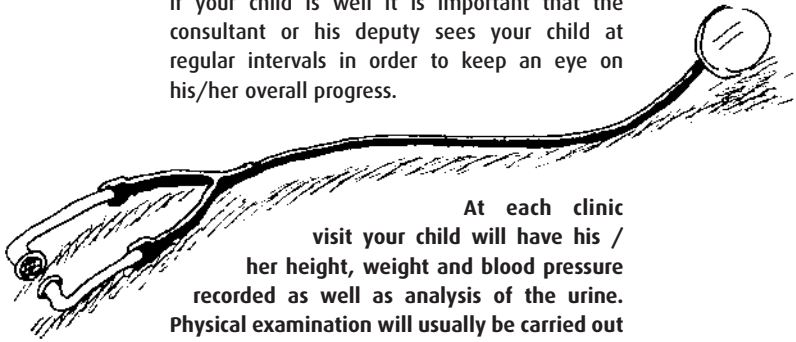
DATE:	URINE TEST	PREDNISOLONE DOSE	OTHER TREATMENT	COMMENTS
JULY 08				
1	N (negative)	10mg	-	Cold
2	0.3g (trace)	-	-	Well
3	N (negative)	10mg	-	Well
4	N (negative)	-	-	Well
5	N (negative)	10mg	-	Well
6	N (negative)	-	-	Well
7	1.0g (++)	10mg	-	Pale & not feeling well
8	3.0g (+++)	-	-	Unwell
9	3.0g (+++)	10mg	-	Unwell
10	>5g (++++)		-	PHONE DOCTOR

How you record the urine test will depend upon the testing sticks used.



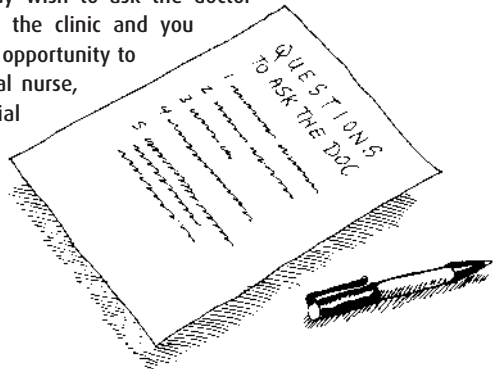
CLINIC VISITS

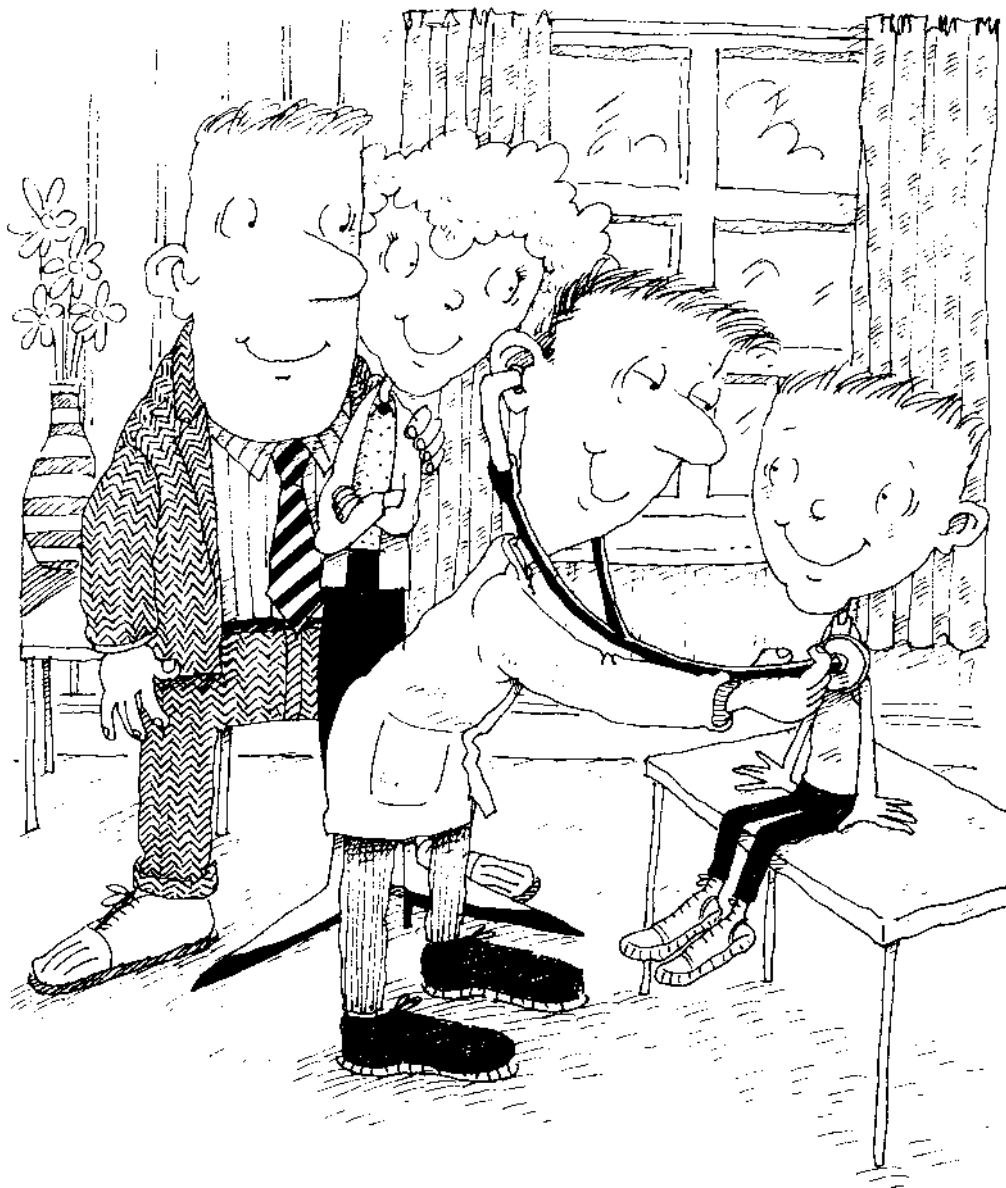
After your child has been diagnosed as having Nephrotic Syndrome he/she will be followed at regular intervals in the out-patient clinic. Even if your child is well it is important that the consultant or his deputy sees your child at regular intervals in order to keep an eye on his/her overall progress.



At each clinic visit your child will have his / her height, weight and blood pressure recorded as well as analysis of the urine. Physical examination will usually be carried out and only if it is necessary will blood tests be performed. If your child is very worried about blood tests then we can try using a special cream, or spray which makes this procedure less painful.

It is a good idea for you to write down any questions you may wish to ask the doctor before coming to the clinic and you may also have an opportunity to talk with the renal nurse, dietitian or social worker.



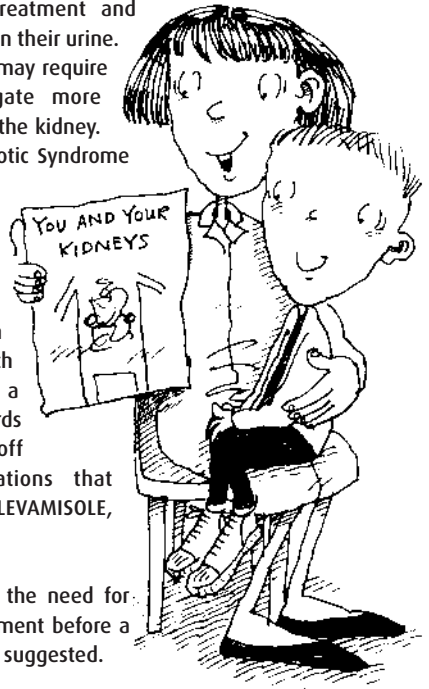


DOES STEROID TREATMENT ALWAYS WORK?

There is a very small group of children who do **not** respond to steroid treatment and continue to have lots of protein in their urine. It is this group of children who may require a kidney biopsy to investigate more thoroughly what is going on in the kidney. Also some children with Nephrotic Syndrome **RELAPSE** more often and begin to have side effects from Prednisolone.

It is for these children that we may decide to use a medication called **CYLCOPHOSPHAMIDE** which may help the child to have a long **REMISSION** (in other words urine free of protein and off Prednisolone). Other medications that may be considered include **LEVAMISOLE**, **CYCLOSPORIN**, **TACROLIMUS**.

The doctor will always discuss the need for further investigations and treatment before a biopsy or other medications are suggested.



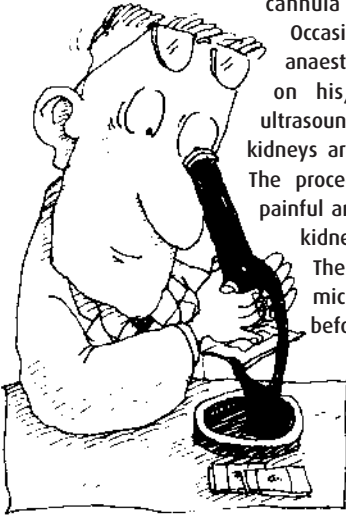
WHAT IS A KIDNEY BIOPSY?

This is a procedure which is carried out with the child sedated (medicine will be given through a cannula to make your child sleepy).

Occasionally in young children a general anaesthetic is required. The child lies flat on his/her stomach and we use an ultrasound scan to find out exactly where the kidneys are and where to insert the needle.

The procedure is more uncomfortable than painful and only a very small sample of one kidney is removed using a special needle.

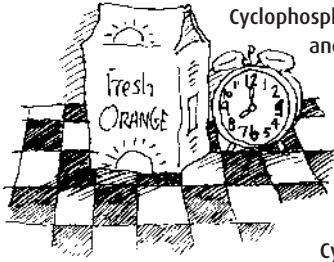
The sample is then examined under the microscope and it may take several days before the answer is available.



After the biopsy the child may rest for a while but then can be up and about quite quickly. Your child may be able to go home after the biopsy the same day.

The biopsy should have no long-term effect on the kidney. We generally restrict heavy exercise for one week after the hospital admission but your child can safely return to school.

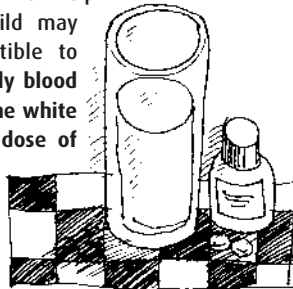
ARE THERE ANY SIDE EFFECTS FROM CYCLOPHOSPHAMIDE?



The course of treatment with Cyclophosphamide is usually 8-12 weeks and this will be discussed with you by the children's kidney specialist before commencing the drug.

There are usually no serious or permanent side effects from Cyclophosphamide when given at the low doses used to treat Nephrotic Syndrome. However we have listed the side effects which may occur.

1. Nausea and vomiting – anti sickness drugs can be given to reduce this
2. Thinning of the hair – this is usually only slight and the hair will re grow after the course of treatment.
3. Strange taste – your child may experience a strange taste in their mouth while receiving cyclophosphamide. This is only temporary.
4. Cyclophosphamide can affect the number of white cells in the blood which help to fight infection. Your child may therefore be more susceptible to catching infections. **A weekly blood test will be done to check the white cells are not low and the dose of Cyclophosphamide will be altered if necessary.**

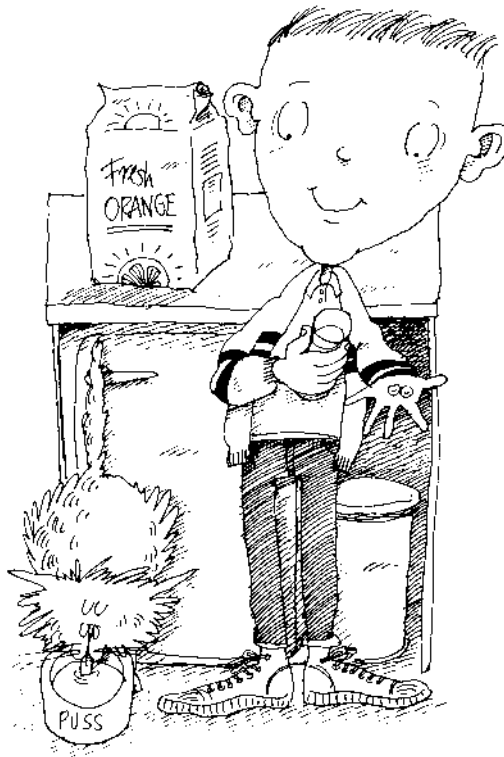


5. Future fertility – the short course of Cyclophosphamide that we now use should not have any long term effects on your child’s fertility. However, this point will be discussed fully with you by the specialist.

There are other less common side effects, these will be discussed with your doctor and may include:

1. A slight increased risk of developing some forms of cancer, this is more likely with individuals taking higher or longer term doses

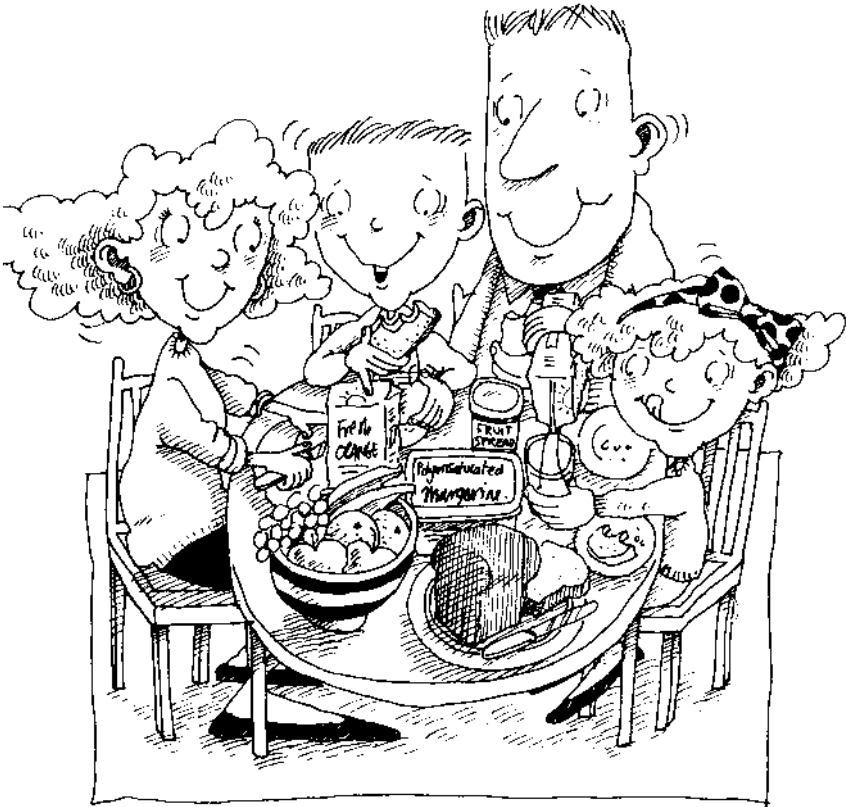
2. Irritation of the bladder wall, which may lead to blood in the urine.



IS THERE A SPECIAL DIET?

When in remission A 'Healthy Eating' diet is generally recommended for your child. It is a good idea for all members of the family to also follow a healthy eating plan.

At other times when your child may be in a Nephrotic state diet and fluid restrictions or guidelines may be in place – and should be followed.



GUIDELINES TO FOLLOW

- **no added salt** – avoid adding salt to food at the table and try to reduce the intake of processed foods e.g. crisps, tinned and packet foods.

- **ensure an adequate protein intake** – it is not usually necessary to take extra protein in the diet, a good healthy appetite will usually ensure an adequate intake.

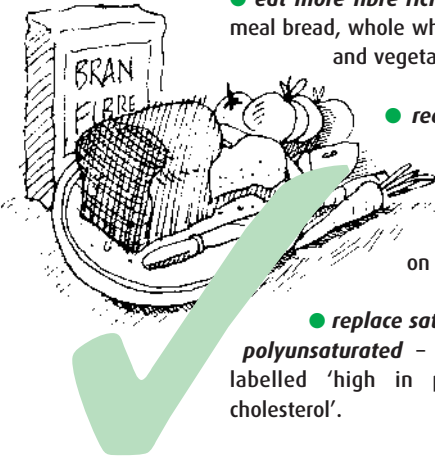
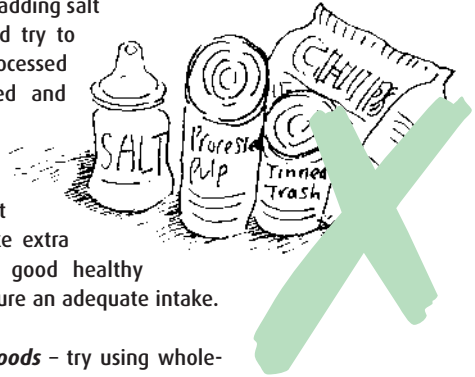
- **eat more fibre rich foods** – try using whole-meal bread, whole wheat breakfast cereals, fruit and vegetables

- **reduce sugar** – try to avoid adding sugar to drinks and food, replace fizzy drinks with sugar free varieties. Try to cut down on sweets and chocolate.

- **replace saturated fats with mono or polyunsaturated** – use margarines and oils labelled 'high in polyunsaturates, low in cholesterol'.

This advice should be treated as a HEALTHIER way of eating rather than as a diet.

Further dietary information and advice can be offered to you from the Paediatric Renal Dietitian whose number is in the back of this booklet.



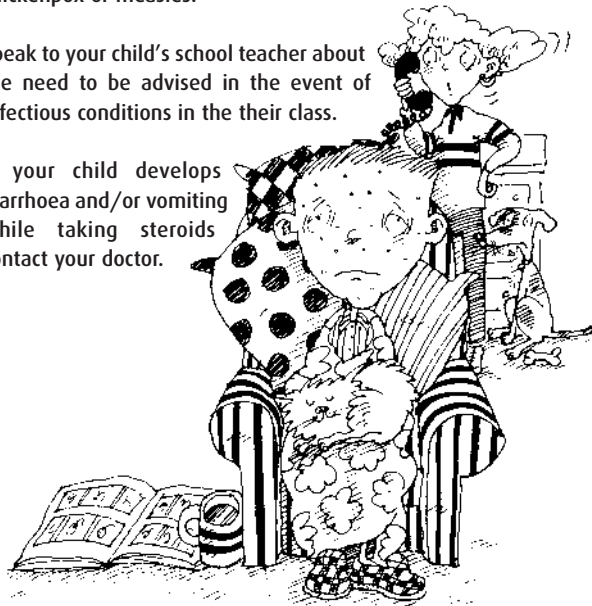
CAN MY CHILD RECEIVE IMMUNISATIONS WHILE TAKING PREDNISOLONE?

Not usually. Steroids can reduce the effectiveness of immunisations and relapses can sometimes be caused by the vaccination.

Children who are having steroid medications or Cyclophosphamide can become very unwell if exposed to CHICKENPOX or MEASLES. Let the hospital know if your child is in very close contact with another child who may have chickenpox or measles.

Speak to your child's school teacher about the need to be advised in the event of infectious conditions in their class.

If your child develops diarrhoea and/or vomiting while taking steroids contact your doctor.



WILL MY CHILD CONTINUE TO SUFFER FROM NEPHROTIC SYNDROME?

Most children will have at least one relapse, with significant protein in their urine (2 – 3+ for 3 days or more) and requiring a further course of steroid treatment.

If they have many relapses they will need to take their steroids for a longer period of time.

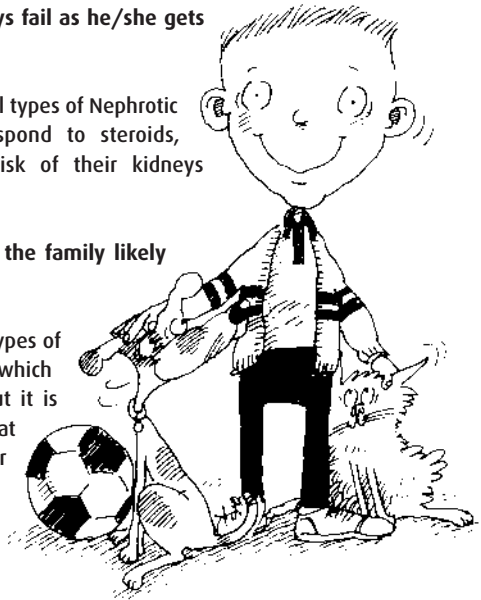
As the child becomes older the relapses usually become less frequent. It isn't possible to predict accurately when the relapses will cease but a relapse is less likely if the child has been free of protein in their urine for five years.

Will my child's kidneys fail as he/she gets older?

Children with the usual types of Nephrotic Syndrome, which respond to steroids, have no increased risk of their kidneys failing with time.

Are other children in the family likely to be affected?

There are a few rare types of Nephrotic Syndrome which do run in families, but it is extremely unlikely that your child will suffer from these.

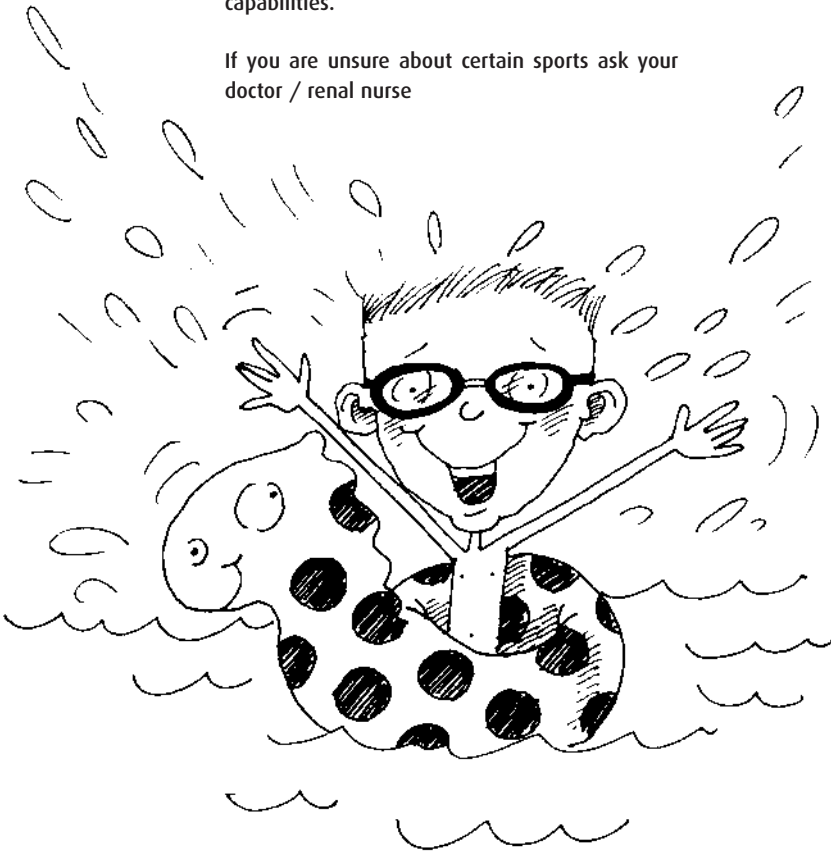


CAN MY CHILD CONTINUE TO PARTICIPATE IN EXERCISE AND SPORTING ACTIVITIES?

YES. Exercise is important for all the family.

The school should encourage the child to participate in all sports within the child's capabilities.

If you are unsure about certain sports ask your doctor / renal nurse



WHAT ABOUT HOLIDAYS?



Unless your child is having complications, a normal family holiday should be possible.

A copy of your latest clinic letter should accompany you and also an adequate supply of prednisolone in case of relapse.

Remember the need for extra sun protection when taking specific medications

Always ensure adequate hydration and movement if undertaking a long haul flight

Tell your doctor / renal nurse about your plans.

WHEN SHOULD I GET IN TOUCH WITH THE DOCTOR/RENAL NURSE?



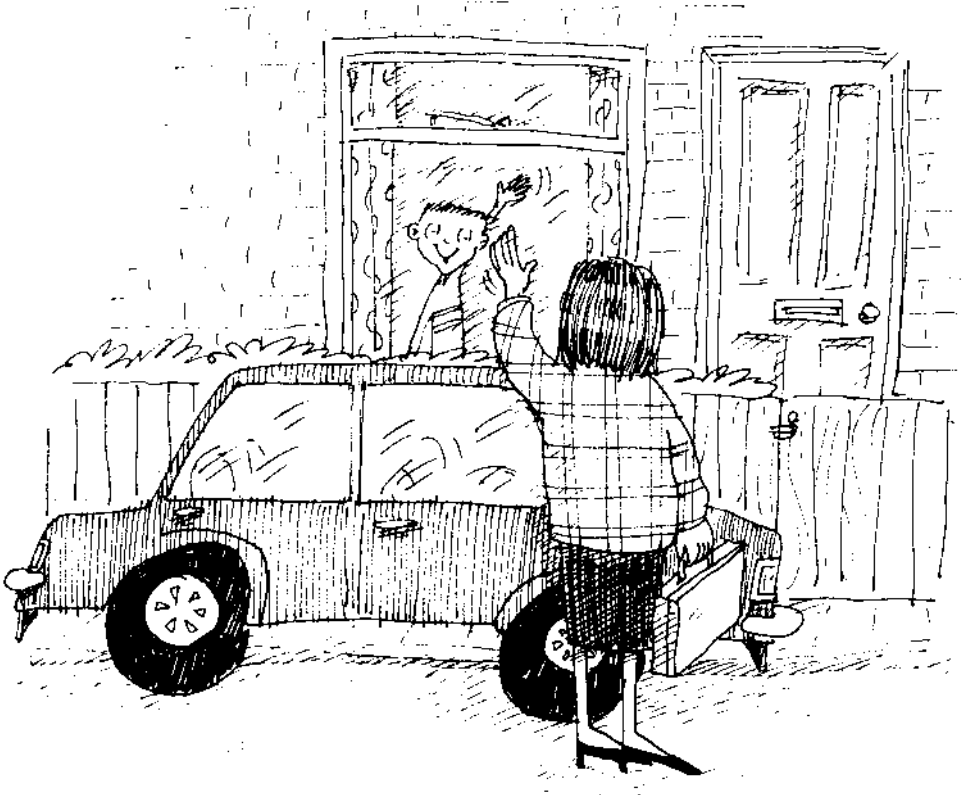
1. If your child is in close contact with chickenpox or measles contact the hospital unit within 24 hours as he/she may need a special injection to boost his/her resistance to these infections.
2. If your child has protein (++) or greater) in the urine for three days.
3. If your child is puffy, unwell or has a fever.
4. If your child has diarrhoea and vomiting.
5. For any small worry it is best to phone, no matter what time, as it may be important. The numbers you can contact if you have any worries are at the back of the booklet.



In some areas the Renal or Community Nurse or Health Visitor may visit you at home. If appropriate the renal nurse or community nurse may visit your child's nursery or school.

You will see the Renal Nurse at clinic visits and when admitted to the hospital

Discussion with the renal social worker may be helpful. It may be possible to receive practical assistance and more importantly support in dealing with your family's concerns.



THE PARENT'S GROUP

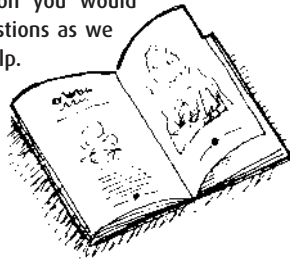
www.nephrotic-natter.org and www.nephrotic-natter.co.uk - An on-line resource for families and patients, providing an opportunity to share experiences, problems, and questions with others who are similarly affected.

Some hospitals have parent's groups and some try to have an occasional parents' evening during the year where parents can get together and discuss any worries. These are usually informal events and give an opportunity for everyone to share their experiences and common problems.

Even if there is no formal parents' group, your doctor, nurse, or paediatric social worker should be able to help you contact another parent who has a child with Nephrotic Syndrome.



This booklet will not answer all your questions or give all the information you would like, so please ask questions as we are always willing to help.





**HERE ARE SOME USEFUL
TELEPHONE NUMBERS:**



Family Doctor

Paediatrician

Paediatric Nephrologist

Paediatric (Renal) Sister

Paediatric (Renal) Dietitian

Paediatric Social Worker

Paediatric Community Sister

Hospital Ward

NOTES

Compiled by the members of the



Nottingham University Hospitals 
NHS Trust

Design: Room at the Top Design Nottingham • Illustrations: Jan Smith

©ARW 1988 • REPRINT 1997 • REVISED 2008