



# NEPHROTIC SYNDROME



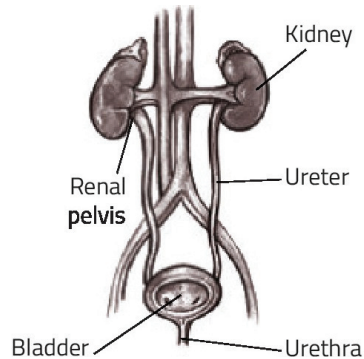
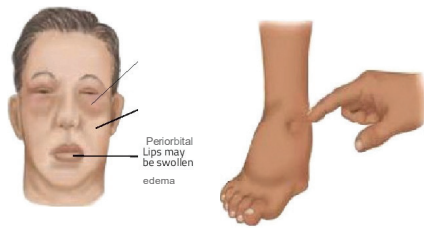
HUMAN KIDNEY CROSS SECTION

# Aster HOSPITAL

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### WHAT IS NEPHROTIC SYNDROME?

It is a clinical condition characterized by following the features: swelling of the legs and face and distension of abdomen. It is due to leakage of protein in the urine which results in low albumin levels in the blood. Patient usually has elevated serum cholesterol levels.

### WHAT ARE THE SYMPTOMS & SIGNS OF THIS DISEASE?

Patient presents with swelling in the legs, swelling around the eyelids, generalized swelling of the body and distension of the abdomen. These manifestations are due to fluid retention. Facial swelling is commonly seen early morning when the patient gets up from bed, and leg swelling is more during the latter part of the day. Fluid can get accumulated around the lungs (pleural effusion) and rarely may cause breathing difficulty. Children can have significant discomfort due to abdominal distension and can have abdominal pain sometimes.

### WHY DO CHILDREN DEVELOP SWELLING OVER THE BODY ?

Due to the leakage of protein in the urine, its levels in the blood gets significantly reduced resulting in accumulation of the fluid in between the cells, with accumulation of salt and water in the body.

### IS THIS A HEREDITARY DISEASE?

Only in a small percentage of the individuals, this disease runs in families. Otherwise in majority of the cases, it is acquired after birth.

### DOES THIS DISEASE AFFECT BOTH THE KIDNEYS?

Yes, it affects both the kidneys.

### WHAT ARE THE CAUSES OF THIS DISEASE CONDITION?

In majority of the children presenting with nephrotic syndrome, the exact cause is not known. There are some known causes of nephrotic syndrome which includes diabetes mellitus, Systemic Lupus Erythematosus (SLE), Malignancy, Lymphoma and drugs (commonly pain killers).

### HOW IS THIS CONDITION DIAGNOSED?

It can be diagnosed by blood and urine tests. Urine examination shows protein and sometimes red blood cells. Due to protein loss in urine, the amount of albumin in the blood is low. In addition, there will be increased cholesterol & triglyceride levels.

### WHAT IS KIDNEY BIOPSY?

It is a procedure which is done to know the exact cause of kidney disease causing nephrotic syndrome. It guides the clinician regarding management



(treatment options). Under ultrasound guidance, exact site of the kidney is determined and using a biopsy gun, a small core of kidney tissue is removed and sent for microscopic examination. Majority of the children with nephrotic syndrome would not need kidney biopsy.

### WHAT ARE THE COMPLICATIONS OF KIDNEY BIOPSY?

Complications are rare with kidney biopsy and is seen in 1 to 2% of the patients who undergo this procedure. Since kidney receives 20% of the blood pumped out by the heart every minute, bleeding (hematuria) can occur after the procedure. Commonly we see blood in the urine, which settles down over a period of time. Rarely gross hematuria can occur. These complications can usually be managed conservatively, and very rarely surgical intervention may be required. Hence following the kidney biopsy, patient is advised to take bed rest for at least 24 hours and avoid straining for at least a week.

### DO PARENTS NEED TO CHECK FOR PROTEIN IN URINE EVERY DAY?

Testing the urine for protein is not required every day. This would just add on to the anxiety of the parents. Urine dipsticks are commercially available. However, the frequency of urine testing should be based on the doctor's instructions.

### WHAT ARE THE TESTS TO BE DONE? HOW FREQUENTLY ARE THESE TESTS TO BE DONE?

Blood and urine tests are required to make the initial diagnosis. In adults, more laboratory investigations are required to determine the cause of nephrotic syndrome which includes screening for certain infections and malignancies, tests for SLE and diabetes mellitus. After the initiation of treatment (during subsequent follow up), urine albumin and albumin levels in the blood are checked. If expected improvement is not seen with treatment, detailed evaluation may be required. Self-testing and treatment should be discouraged, as it is not advisable.

### WHAT ARE THE MEASURES FOR THE MANAGEMENT OF NEPHROTIC SYNDROME?

Dietary modifications are important ; this includes dietary salt and fluid restriction. These patients are prone for infections and hence cleanliness and personal hygiene are of utmost importance. Specific medications are available for certain conditions which has to be taken as per doctor's advice. (E.g.: steroids for children with nephrotic syndrome).

### DOES THIS CONDITION NEED LONG TERM TREATMENT ?

Nephrotic syndrome in children usually presents with a relapsing and remitting course. Frequency of relapses may reduce as the age of the child advances and by 14-16 years of age, permanent remission is achieved in majority of the children. On every relapse, doctor has to be consulted and treatment has to be taken based on his advice.

### WHAT IS THE DURATION OF DRUG TREATMENT?

Duration of treatment depends on the cause of nephrotic syndrome. In children, usually it varies from 3 to 6 months.

### WHAT ARE THE PRECAUTIONS TO BE TAKEN WHILE ON MEDICINES FOR THIS DISEASE CONDITION?

Most importantly as per the doctor's advice, medicine has to be taken at the right dose and at the right time. Children's immunity can be reduced, and they are prone for infections. In case of fever, doctors should be immediately consulted. Child will also have increased appetite, increased food intake and weight gain. To avoid this, child should be on a proper diet. Carbohydrate rich diet (sweets, rice, roots) and fat rich diet (saturated fat, cooking oil) should be restricted. Child should be encouraged to involve in physical activity and play games to avoid weight gain.

### WOULD THESE MEDICINES CAUSE ANY SIDE EFFECTS?

Long term use of these medications can have certain side effects in a fraction of children which include stunted growth, osteoporosis (weakness of the bone), and cataract. Hence, medicines should be taken as per the doctor's advice (proper dose and duration).

### HOW CAN WE AVOID SIDE EFFECTS OF MEDICINES?

If the medicines are taken at the right time and in correct doses as per instructions of the doctor, the chances of relapses are less and thereby we can reduce the cumulative dose of the medicines that can result in adverse effects. The steroid medicine should be taken as a single dose after breakfast

which can reduce the chances of adverse effects.

### ARE THERE ANY MEDICINES OTHER THAN STEROIDS?

Many medicines are available for the treatment of nephrotic syndrome. Usually such medicines are used when the child is resistant or intolerant to steroids.

### ARE THERE ANY SIDE EFFECTS FOR PREDNISOLONE?

Most common side effect is acute gastritis presenting as pain abdomen, nausea and vomiting. Other side effects include excessive appetite, weight gain, fluid retention, acne (pimples) over the face and back, fat deposition over the face (moon face), back and abdomen. These patients are at increased risk of infections as their immunity is reduced. They can have bone related problems (osteoporosis) and cataract.

### CAN THE DOSE OF MEDICINES BE MISSED AT TIMES?

Even if a single dose is missed, we may not get the expected benefit. There is a risk of withdrawal symptoms on missing the dose. So, parents should make sure that medicines are given at the right time.

### WHAT IS RELAPSE AND REMISSION?

Disappearance of protein (albumin) in the urine is called 'remission'. Reappearance of protein (albumin) in the urine associated with swelling of face and feet is called 'relapse'.

### WHAT ARE THE CAUSES OF RELAPSE?

Upper respiratory infections (sore throat, running nose & cough), viral fever can precipitate a relapse. Cleanliness and

personal hygiene are the most important measures to prevent it. In some instances, no specific cause is found.

### DO WE NEED TO HAVE TREATMENT DURING REMISSIONS?

This disease requires long term follow up with the doctor. Even after achieving remission, it is better to consult the doctor at periodic intervals to know the possibility of relapse and institute treatment at the earliest if there are signs of relapse. Relapses are either frequent or infrequent and the treatment decisions are based on that.

### CAN THE CHILD BE GIVEN VACCINATION?



Vaccinations against certain infections like pneumonia and chicken pox should be given when the child is in remission. These vaccinations should not be given when the child is on medications for nephrotic syndrome.

### WHEN DOES THE CHILD NEED PLASMA INFUSIONS?

When the protein levels in the blood is significantly reduced, the child will develop edema of the legs, swelling of the abdomen and sometimes generalized swelling of the body leading to breathlessness. In such circumstances, albumin or plasma infusion can be given to the child for a temporary relief till

other medications which reduce the protein leak in the urine start acting.

### ARE THERE ANY DIETARY RECOMMENDATIONS?

Certain dietary instructions need to be followed in children with nephrotic syndrome. Salt and fluid restriction are of prime importance. Daily fluid intake should be restricted as per the advice of the doctor. Diet rich in fats and oily foods should be reduced. Fruits and vegetables can be taken. Protein rich foods like egg white and pulses need to be given.

### CAN THE CHILD BE GIVEN FISH & MEAT?

Egg white should be given to the child, as this helps to increase the protein level in the blood. Fish and chicken are also protein rich foods and should be given.

### DO WE NEED TO RESTRICT SALT AND WATER INTAKE? WHY?

When the child has swelling, salt and fluid has to be restricted. Salt needs to be restricted in children with high blood pressure. In the presence of nephrotic syndrome, increased dietary salt intake will further worsen the edema and can lead to complications like breathlessness and infections. Restriction of salt and fluid will facilitate reducing the swelling of the body.

### IS PROTEIN POWDER USEFUL?

If the child can take sufficient daily protein in the diet as recommended by the doctor, supplemental protein need not be required. Protein powder can be used in children with poor food intake.

### CAN FRUITS BE GIVEN TO THE CHILD?

If the kidney functions are normal, fruits can be given. If there is deterioration in

the kidney functions, certain fruits should be avoided. In nephrotic syndrome seen in most children, kidney functions are usually well preserved and hence fruits and vegetables can be given without any restrictions.

#### **WHY IS THE CHILD MORE SUSCEPTIBLE TO INFECTIONS?**

Children with this disease will have reduced immunity. Proteins that play an important role in immune mechanisms are lost in urine. Due to significant swelling, there is breach in the skin immunity and hence they are prone for infections. Commonly seen infections are peritonitis (infection of the abdominal cavity), cellulitis (infection of the skin & subcutaneous tissue) and gastrointestinal infections.

#### **WHAT ARE THE SIGNS OF INFECTIONS?**

When a child with nephrotic syndrome develops infections, the most common symptom is fever. Child can have cough with sputum production suggesting lung infection. Child can have pain abdomen, vomiting and loose stools suggesting peritonitis. Child can have increased frequency of urination and pain while passing urine suggesting urinary infection. Sometimes pain, swelling and redness over the legs and thigh are seen suggestive of cellulitis.

#### **WHAT ARE THE COMPLICATIONS OF NEPHROTIC SYNDROME?**

Generalized swelling of the body may cause abdominal discomfort and breathlessness (due to fluid accumulation of the lungs.) Child is at increased risk of infections as mentioned earlier, commonly includes peritonitis, lung infection, cellulitis and urinary infection. Rarely blockage (thrombosis) of arteries and veins are seen, manifesting

as stroke, pain and swelling of legs (secondary to deep vein thrombosis) and life-threatening pulmonary embolism (blockage of blood vessels in the lungs).

#### **WHAT ARE THE PROBLEMS THAT CAN BE SERIOUS?**

Infections can be serious and life threatening at times. Child can develop stroke due to thrombosis of vessels in the brain. Thrombosis of blood vessels in the lungs can present as breathlessness. Blood vessels in the kidneys can get blocked and they can present as kidney failure.

#### **IS THERE ANY NEED TO RESTRICT DAILY ACTIVITIES OF THE CHILD?**



It is recommended to restrict daily activities when the child is in relapse with significant swelling and also when the child has infections. Child can have normal activities when in remission.

#### **WHAT SHOULD BE DONE IF CHILD DEVELOPS FEVER?**

Fever is a sign of infection and child should be taken to a doctor immediately and his instructions followed.

#### **WOULD THIS CONDITION HAS ANY EFFECT ON CHILD'S FUTURE?**

This disease has a relapsing and remitting course in children. More than 80% of children will achieve sustained remission by the age of 14-16 years. However, a small percentage of the children can have relapses even after this age.

#### **CAN THE CHILD GO TO SCHOOL?**

This disease usually doesn't affect the scholastic performance. Child should be encouraged to go to school except when the child feels extremely tired and has significant generalized swelling of the body.

#### **WOULD THE KIDNEYS FAIL IN FUTURE? WOULD THERE BE A NEED FOR DIALYSIS?**

Nephrotic syndrome in children usually do not cause deterioration of the kidney functions. About 10% of the children can have progressive deterioration of kidney function and lead to chronic kidney failure. However, a higher percentage of the adults can have kidney failure.

#### **WHAT ARE THE DIRECTIONS FOR PARENTS OF A CHILD WITH NEPHROTIC SYNDROME?**

- Child should be taken to a doctor on regular basis and his/ her instructions followed.
- Parents need not be too anxious about the disease as it can be cured with medical management in majority of the children.
- Parents need not check the urine and blood very frequently. These tests should be done as per the instructions of the doctor. Daily testing of the urine using dipsticks will unnecessarily increase the anxiety of the parents rather than doing any good to the child.

- Don't treat the child as a patient and impose unnecessary restrictions on the child. Encourage the child to get involved in daily activities, go to school and to play games.
- Consult a doctor immediately if the child develops fever, cough with sputum production or any other signs of infection.
- Self-treatment of this condition should be discouraged; consult a doctor at the earliest if symptoms develop.
- Parents should not get disheartened with the disease state of the child, instead, they should work in liaison with the doctor and follow their instructions sincerely.
- Parents should have mental strength, trust the doctor and show perseverance in treating this disease condition.



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THE LOCATION

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