Nephrotic Syndrome

What is nephrotic syndrome?
Nephrotic syndrome is a disease where the kidneys leak too much protein into the urine. The kidney works a lot like a spaghetti strainer: it keeps the good things in (the "spaghetti": red blood cells, proteins) and lets the waste (extra water, extra salt, waste products) through to make urine. In nephrotic syndrome, the glomerulus, which is the filtering part of the kidney, is leaky. It lets proteins leak through into the urine.

Some of the proteins that are lost work in the body like sponges. They help keep water inside the blood vessels. When the proteins are gone from the blood because they have all left in the urine, water leaks out of the blood vessels. This fluid then leaks into the skin and causes swelling. Common body parts that swell are around the eyes, ankles, and belly.

People with nephrotic syndrome have three main problems.

- Protein in the urine, and
- Swelling, and
- Low protein in the blood.

What causes nephrotic syndrome?
- Many kidney diseases cause nephrotic syndrome. The most common cause in children is called **minimal change disease.** No one knows what causes minimal change disease. We think it might involve the immune system. The immune system normally fights infections. In some people it may also attack the kidneys. Nephrotic syndrome is not contagious. You can't "catch" it from someone else. Most children (80%) respond to treatment which is wonderful. ***This disease can be very frustrating however because the majority of children who respond to therapy will relapse. These relapses can sometimes occur multiple times a year.***

How is nephrotic syndrome treated?
- The main treatment is a drug called **prednisone** (in tablets) or **prednisolone** (in a liquid). It belongs to a class of drugs called corticosteroids. This is not the same as the steroids that bodybuilders take! This is a medicine that calms down the immune system.
- Sometimes doctors prescribe a **diuretic** (water pill) to help with the swelling. These drugs make you pee off all the extra fluid in the body. Most children can stay at home, but a few children have to stay in the hospital for a few days to reduce the swelling with intravenous (IV) medicines.
- Your child will take these medicines until the protein is gone from the urine. Then he or she will take smaller doses of the medicine for a few more weeks. You can test your child's urine at home for protein. See instructions at the bottom of this document. It's easy to do!
- Your child should not get certain vaccines (shots) while taking this treatment. He or she should not get "live" vaccines such as varicella ("chicken pox"), measles-mumps-rubella ("MMR"), or the intra-nasal flu vaccine. Most other vaccines are okay. Make sure your doctor knows your child is on prednisone (steroids) before your child gets any vaccines.

Staying away from salty foods will help reduce the swelling. Don't add salt to your child's foods. Try to give your child foods that are low in salt, like fruits, vegetables, and fresh meats. Avoid salty foods, like chips, lunch meats, pizza, cheese, ramen noodles, basically most things that come in a can or a package. Children taking predniso(lo)ne may want to eat a lot. Give your child healthy, low-fat snacks. Your child should eat a normal amount of protein. Your doctor or dietician can tell you more about low-salt foods and healthy snacks.

- Children with nephrotic syndrome have a higher risk of getting infections, both because of the nephrotic syndrome, and because of the prednisone (steroids). You must call your child's doctor **right away** if your child:
- Has stomach pain
- Has a fever (temperature greater than 101° F or 38.5° C)
- Is around someone who has chicken pox

Will it come back?

- Most children with nephrotic syndrome will have several episodes during their lives. When nephrotic syndrome comes back, it's called a **relapse**. These relapses are treated with medicine much like the first time. Relapses often happen when the child gets sick with a cold or another illness. Sometimes they happen when the child is well. We don't know what causes relapses. Relapses usually get better with treatment, the same way the first episode did. Most children with nephrotic syndrome will stop having relapses during their teenage years. A small number of children continue to have relapses as adults.

Will my child need a kidney biopsy?

- A biopsy is when a kidney doctor uses a long hollow needle that goes through the skin of the back to take out a tiny piece of the kidney. This piece is then looked at under a microscope. Most children with nephrotic syndrome **do not** need a kidney biopsy. Your doctor may suggest a kidney biopsy if:
- Your child has many relapses
- Your child does not improve with treatment
- Your doctor thinks your child might have a disease other than minimal change disease

Will my child always have to take medicine?

- Most children can stop taking prednisone after they get better. You should only stop the medicine when your doctor tells you to. It's very important to take the medicine the way the doctor prescribed it. If you stop giving prednisone suddenly, your child can become very ill. If you need more medicine, call the doctor's office and we can re-prescribe it right away.
- Your child will have to start taking prednisone again if he or she has a **relapse** in the future.
- If your child has many relapses, he or she may need a different treatment to help prevent relapses. There are several medicines available that your kidney doctor will tell you about.

Does nephrotic syndrome cause kidney failure?

- Most children with nephrotic syndrome never develop kidney failure. Most never need dialysis or a kidney transplant. But a small number of children do. It's important to

see the doctor regularly. He or she will watch for any early signs of kidney trouble (high blood pressure, not growing tall enough), and tell you about possible treatments.

Test strips are $14 for 100 strips on Amazon. Search for “urine reagent strips for urinalysis” on the amazon.com website.

You will only be looking at the fourth square from the end, the one that matches up to the word “PRO” on the bottle.

If it’s yellow, there is no protein.
If it’s very light green, there is “15” protein, or “trace”
If it’s a little darker green, there is “30” protein, or “1+”
If it’s a little darker green, there is “100” protein, or 2+
If it’s the darkest green, there is “300” protein, or 3+

Here is a video with a doctor describing how to do urine testing at home:
Search youtube for “testing urine for protein and glucose”