Understanding Childhood Nephrotic Syndrome

Childhood nephrotic syndrome is a disease that affects the kidneys. This handout will explain childhood nephrotic syndrome, the symptoms of nephrotic syndrome, how we test for it and how we treat it.

WHAT IS CHILDHOOD NEPHROTIC SYNDROME?
Childhood nephrotic syndrome, or simply nephrotic syndrome, is a disease that damages the glomeruli in the kidneys. It can happen at any age, but it happens most often in babies and children ages 1-5. Nephrotic syndrome tends to affect boys more often than girls. There are 2 types of nephrotic syndrome, which are minimal change disease (MCNS) and focal segmental glomerulosclerosis (FSGS). The first type, MCNS, is more common. The second, FSGS, is less common, but more serious than MCNS.

WHAT ARE GLOMERULI?
Glomeruli are small vessels in the kidneys that help filter blood and protein throughout your body. Protein in the blood helps keep fluids from leaking into the body’s tissues. When the glomeruli are damaged, they leak large amounts of protein into urine. This leaves little protein behind for the kidneys to keep fluid from leaking into your body’s tissues. This leaking fluid causes swelling throughout your body.

WHAT ARE THE SYMPTOMS OF NEPHROTIC SYNDROME?
The most common sign of nephrotic syndrome is swelling throughout the body, but other symptoms include:
- Swelling around your child’s eyes, belly and legs
- Urinating less often
- Weight gain from excess water
- High levels of protein in the blood (proteinuria)
- Low levels of protein in the blood from loss of protein in the urine (hypoalbuminemia)
- High cholesterol levels in the blood (hyperlipidemia)

WHAT CAUSES NEPHROTIC SYNDROME?
In many cases, we do not know what causes nephrotic syndrome. We think it might be triggered by allergies, a cold, the flu or other viral illnesses. Sometimes, your child can relapse, which means he or she can develop nephrotic syndrome again shortly after recovering from it. We think relapses are caused by viral illnesses or infections.

HOW DO WE DIAGNOSE NEPHROTIC SYNDROME?
We diagnose nephrotic syndrome in 3 ways, which are:
- A complete review of your child’s medical history
- A physical exam by your child’s nephrologist (kidney doctor)
- Diagnostic testing

There are 4 diagnostic tests, but your child will most likely not have all 4. We will talk with you about which tests are most appropriate. These tests are:
- Urine testing to check for protein. We might test multiple samples of your child’s urine over a 24-hour period to get more exact measures of the protein.
- Blood testing to see how well your child’s kidneys are working and to check cholesterol and albumin levels
- An ultrasound of the kidneys to check the kidneys’ sizes and structures
- A kidney biopsy, in which we remove a small sample of your child’s kidney and check it under a microscope. We do this only if your child’s nephrologist thinks it is needed.
HOW DO WE TREAT NEPHROTIC SYNDROME?
We treat nephrotic syndrome in 3 ways:

- With a medication called **corticosteroid**
- By limiting the amount of fluid and salt your child has in his or her diet
- With a medication called a **diuretic** (water pills), which will help reduce the swelling in your child’s body

Corticosteroids and limiting salt and fluids in your child’s diet are more common treatments than water pills. Your child’s nephrologist will tell you if your child needs water pills.

The chart below helps explain some of the different treatments we use for nephrotic syndrome. Not all treatments are listed. We will talk with you if your child needs treatment that isn’t on the chart.

THE CORTICOSTEROIDS DID NOT HELP. WHAT OTHER TREATMENTS ARE THERE?
Sometimes, the corticosteroids do not help as well as they should or at all. This is true if your child relapses often or has bad side effects from the corticosteroids. If this happens, we will give your child a medication called a **cytotoxic agent** for 8-12 weeks.

If a cytotoxic agent does not help OR if your child has bad side effects, we will give your child a medication called a **calcineurin inhibitor**. Your child will take this for about 6 months. If it doesn’t work after 6 months, your child will stop taking it and we will talk with you about other treatment options.

Your child’s nephrologist might do a kidney biopsy before giving your child a calcineurin inhibitor. This is to make sure that your child’s kidneys are healthy enough because the medication can cause interstitial fibrosis (kidney scarring). We will talk with you if we think your child needs a kidney biopsy.

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Help</th>
<th>Side Effects</th>
<th>Duration</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corticosteroids AND Low-Salt, Low-Fluid Diet With or without water pills</td>
<td>Helps without bad side effects</td>
<td>Continue with treatment until symptoms go away</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does not help OR Your child has bad side effects</td>
<td>We try a cytotoxic agent</td>
<td>Helps without bad side effects</td>
<td>Continue treatment for 8-12 weeks (2-3 months)</td>
<td></td>
</tr>
<tr>
<td>Does not help OR Your child has bad side effects</td>
<td>We try a calcineurin inhibitor</td>
<td>Helps without bad side effects</td>
<td>Continue treatment for up to 6 months OR until symptoms go away</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Does not help</td>
<td>We will talk about other treatment options</td>
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</tbody>
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