

# Nephrotic Syndrome

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## ***What is nephrotic syndrome?***

Nephrotic syndrome is a common kidney disease of children, although only 0.01% of children are affected. Most cases of nephrotic syndrome occur in children less than 4 years of age.

Children with nephrotic syndrome have leaky kidneys that allow excess protein to spill in the urine. This leads to low protein levels in the blood, which permits fluid to leak into the body and cause swelling. Areas that often swell include

the face, the belly (called ascites), the genitals, and the legs.

Most childhood nephrotic syndrome is idiopathic, meaning we do not know its cause. Some diseases can cause nephrotic syndrome, including cancers, infections, and others.

Nephrotic syndrome can be dangerous. Proteins lost in the urine include many that fight infections and some that keep the blood from clotting.

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## ***How do you diagnose nephrotic syndrome?***

First, your doctor will determine the amount of protein in the urine with a chemical dipstick and/or sending a urine specimen to the lab. The lab will measure the amount of protein and creatinine in the urine, and protein level will be divided by creatinine; results more than 0.2 are abnormal, and values above 2 often happen in nephrotic syndrome.

Blood tests will measure the amount of protein in the blood, the overall ability of the kidneys to clear wastes, and the possibilities of other disorders that cause nephrotic syndrome. Many doctors also study the kidneys with ultrasound.

In unusual cases, genetic tests and a kidney biopsy may be necessary to determine the cause of nephrotic syndrome and to guide treatment. Most children have a type called minimal change because the biopsy looks normal.

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## ***How do you treat nephrotic syndrome?***

Unless something unusual shows up on the exam or blood tests, most children with nephrotic syndrome will begin treatment with steroids like prednisone or prednisolone. About 85% of children quit spilling protein with this treatment. Most of these children have minimal change nephrotic syndrome. As long as a child responds to steroids, their long-term risk of permanent kidney damage remains very low.

steroid-dependent. Steroid dependent children can develop significant side effects of long-term steroid use. In both cases, doctors generally do a kidney biopsy and then treat with other drugs. These drugs include chemotherapy (cyclophosphamide), cyclosporine, tacrolimus, and mycophenolate. Other potent drugs that suppress the immune system may be used if these others do not produce a remission.

Your doctor may also prescribe a low-salt diet and diuretics (water pills) to reduce the swelling.

Children who respond easily to steroids often develop more episodes of nephrotic syndrome. Colds or other infections can trigger the protein spill, and the child will need to take steroids again. Most children with steroid-sensitive nephrotic syndrome outgrow it after a few years.

Long-term drug-resistant nephrotic syndrome may be treated with other agents as well. Drugs that inhibit a hormone called angiotensin may reduce the spill of protein. Cholesterol-lowering diet and drugs may be necessary. Drugs to inhibit blood clotting and improve immune function may also be prescribed.

Some children do not respond to the steroids or begin spilling protein very soon after steroids are stopped. These children are steroid-resistant and