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DEVELOPMENT OF NEPHROTIC SYNDROME IN INFANTS

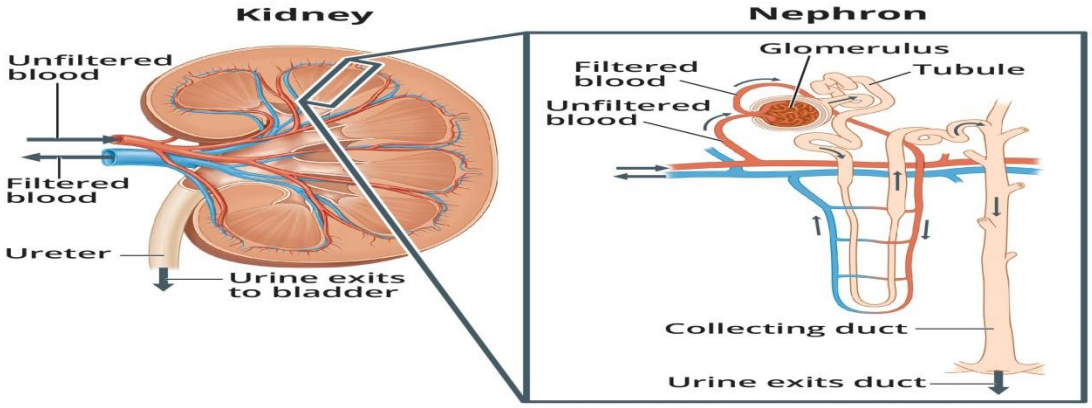
Temirova Oydin
Andijan State Medical Institute, Uzbekistan

Annotation:
The kidneys are made up of about a million filtering units called nephrons. Each nephron includes a filter, called the glomerulus, and a tubule. The glomerulus filters the blood, and the tubule returns needed substances to the blood and removes wastes and extra water, which become urine. Nephrotic syndrome usually happens when the glomeruli are damaged, allowing too much protein to leak from the blood into the urine.

Keywords: Nephrotic syndrome, called edema, health

Nephrotic syndrome is a group of symptoms that indicate the kidneys are not working properly. These symptoms include

- too much protein in the urine, called proteinuria
- low levels of a protein called albumin in the blood, called hypoalbuminemia
- swelling in parts of the body, called edema
- high levels of cholesterol and other lipids (fats) in the blood, called hyperlipidemia



Health care professionals use different terms to refer to nephrotic syndrome in children, depending on

- how old the child is when symptoms begin
  - congenital nephrotic syndrome—birth to 3 months
  - infantile nephrotic syndrome—3 to 12 months
  - childhood nephrotic syndrome—12 months or older
- the cause of nephrotic syndrome
  - primary nephrotic syndrome—the syndrome is caused by a kidney disease that affects only the kidneys
  - secondary nephrotic syndrome—the syndrome develops because of other causes, such as diseases that affect other parts of the body, infections, and medicines

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Nephrotic syndrome is not very common in children. On average, fewer than 5 in 100,000 children worldwide develop nephrotic syndrome each year.[1]

Four types of kidney disease can cause primary nephrotic syndrome in children and adolescents.[2]

- Minimal change disease *NIH external link* (MCD). MCD is the most common cause of nephrotic syndrome in young children. The disease causes very little change to the glomeruli or nearby kidney tissue. The changes in the kidney can only be seen using an electron microscope, which shows tiny details. Although the cause of MCD is unknown, some health care professionals think the immune system may be involved.
- Focal segmental glomerulosclerosis *NIH external link* (FSGS). This disease can cause some of the kidney's glomeruli to become scarred. FSGS may be caused by genetic variants *NIH external link*, or changes in genes present at birth.
- Membranous nephropathy *NIH external link* (MN). MN is an autoimmune disease that causes immune proteins to build up in the kidney's glomerular basement membrane. As a result, the membrane becomes thick and does not work properly, allowing too much protein to pass into the urine.

Other causes of primary nephrotic syndrome are uncommon.

Secondary nephrotic syndrome

Causes of secondary nephrotic syndrome in children include[3]

- diseases that involve many organs or the whole body, called systemic diseases. Examples include IgA vasculitis (also known as Henoch-Schönlein purpura) and lupus.
- infections, including hepatitis B and C, HIV *NIH external link*, and malaria *NIH external link*.
- diseases of the blood, such as leukemia *NIH external link*, lymphoma *NIH external link*, and sickle cell disease *NIH external link*.
- some medicines and drugs, such as nonsteroidal anti-inflammatory drugs, and some medicines used to treat mood disorders, bone loss, or cancer.

Children respond to medicines and treatments differently than adults. The way to get the best treatments for children is through research designed specifically for them.

We have already made great strides in improving children's health outcomes through clinical trials—and other types of clinical studies *NIH external link*. Vaccines, treatments for children with cancer, and interventions for premature babies are just a few examples of how this targeted research can help. However, we still have many questions to answer and more children waiting to benefit.

Congenital nephrotic syndrome

Among newborns and infants younger than 12 months old, the two most common causes of nephrotic syndrome are[4] genetic variants, which account for most cases of congenital nephrotic syndrome

Corticosteroids. Corticosteroids, or steroids, are the medicines most often used to treat children with primary nephrotic syndrome. These medicines suppress the immune system, reduce the amount of protein passed into the urine, and decrease swelling. In most children, treatment with corticosteroids will make nephrotic syndrome improve—also called "remission." If symptoms return, called a "relapse," the health care professional may prescribe a shorter course of corticosteroids until the disease goes into remission again. Although children may have multiple relapses, they often recover without long-term

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kidney damage. In most cases, relapses happen less often as children get older.[3] Although corticosteroids effectively treat nephrotic syndrome in many children, using these medicines for long periods of time can cause side effects, such as impaired growth, obesity, high blood pressure, eye problems, and bone loss.[3] Other common side effects include anxiety, depression, and aggressive behavior. These problems are more likely to develop with larger doses and longer use. In some cases, nephrotic syndrome may not improve with corticosteroids. Cases of nephrotic syndrome that don't respond to corticosteroids are more difficult to treat than those that do. They are also more likely to progress to end-stage kidney disease.[3] Other medicines that suppress the immune system. If corticosteroids are not working or are causing harmful side effects, your child's health care professional may prescribe other medicines that reduce the activity of the immune system. In some cases, your child may take these medicines together with low-dose corticosteroids.

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