CONGENITAL NEPHROTIC SYNDROME

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Definitions of Relevant Kidney Structures

- **Nephron**: unit of the kidney involved in excretion
- **Capillaries**: minute thin-walled blood vessels
- **Glomerulus**: clump of capillaries at the beginning of the nephron that passes protein-free filtrate from the blood into the surrounding capsule
- **Nephrin**: protein fund in the membrane of the glomerulus that provides a barrier during filtration
Figure 1. Nephrin in the slit membrane serves as a barrier to prevent blood proteins from passing into the surrounding capsule (top). In congenital nephrotic syndrome (bottom), nephrin is defective and not present. Hence, proteins pass through and end up in the urine (proteinuria). Loss of blood proteins causes swelling (see Fig. 2).

Figure 2. As shown in Fig. 1, the lack of nephrin leads to protein loss from the blood into the urine. Normally, proteins help retain fluid in the blood vessels (top). In congenital nephrotic syndrome (bottom), the loss of blood proteins leads to loss of fluid from the blood vessel so that the concentration of protein remains somewhat constant. This fluid leaks into surrounding tissues causing swelling (edema). Patients are given diuretics to help the body get rid of some of this fluid.