Recommendations from the International Pediatric Nephrology Association for the Diagnosis and Management of Children with Steroid-Resistant Nephrotic Syndrome (SRNS)

**Diagnosis**

Steroid-resistant nephrotic syndrome (SRNS) is the persistence of protein in the urine after 4 weeks of treatment with prednisone/prednisolone. It can lead to decreased kidney function and/or kidney failure.

Genetic testing and a kidney biopsy should be considered in all children with SRNS without a clearly identified cause.

Genetic causes are identified in up to 1/3 of children. If a genetic cause is identified, medications that act on the immune system are not effective and should be discontinued.

**Treatment**

Use of medications that decrease the amount of protein in the urine and protect the kidneys are recommended once the diagnosis of SRNS is confirmed.

Once the diagnosis of SRNS is confirmed, treatment with cyclosporine or tacrolimus (or alternatives) for at least 6 months, should be started. If there is no response after 6 months, they should be discontinued. Medications to manage chronic kidney disease may be used.

Kidney transplant is recommended to all children who reach kidney failure, recognizing that there is a risk of recurrence of nephrotic syndrome in the new kidney. Removal of one or both kidneys in a patient may be needed prior to transplantation.

**General measures**

Encourage physical activity and healthy nutrition. Excessive salt intake should be avoided.

Routine vaccinations including the annual flu shot should be given. Live virus vaccines require caution in children taking immunosuppressive medications. Speak with your doctor before vaccinations.

Different medications may be needed to compensate the loss of proteins in the urine (hormones, vitamins, calcium).

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