



## First Pediatric Nephrology Appointment: Tips for Parents

*The following information was prepared by Alport Syndrome Foundation (ASF) with the guidance of Medical Advisory Committee member Dr. Bradley A. Warady (Children's Mercy Hospitals and Clinics in Kansas City, MO).*

*For further information, you are strongly encouraged to watch ASF's accompanying [Pediatric Alport Patient Care: What You Need to Know Right Now](#) video.*

### **Achieving Optimal Care**

The "Team Approach" (parent or guardian/child/health care provider working together) allows for the best quality of life for the child and preservation of kidney function. Parents are critically important advocates for children diagnosed with Alport syndrome. It's of the utmost importance to make sure affected children are age-appropriately educated about their disease and are following the recommendations of their health care providers. Parents are encouraged to work with the nephrologist and larger health care team to most appropriately educate children of all ages affected by Alport syndrome.

The experiences of children and teens with Alport syndrome can vary widely. Typical commonalities include daily medications, ongoing appointments with specialists, diet adjustments, regular blood/urine tests, and living with a label of "rare disease," which can lead to feelings of isolation.

Children with Alport syndrome must develop a progressive understanding of their disease. Age-appropriate education and modeling of open communication with a nephrologist can be very helpful for these children to develop a sense of empowerment over their disease. It's important for them to increasingly become advocates for themselves over time. Self-management is key, especially as they approach adolescence – a time when adherence to treatment medications is strained but critical. Alport syndrome doesn't need to control your child's life but rather become something the child can help control.

Over the past decade, research has led to a much greater understanding of Alport syndrome and treatment. [There is increased data and understanding of the value of treating Alport patients of all ages, including children, with ACE/ARB medications to delay the progression of kidney disease.](#) This intervention can begin in patients as young as 12 to 24 months of age with liquid forms of the medications. ACE/ARB therapy is usually initiated at a low level and progressively increased to achieve a maximum tolerable dose – a level that works ideally in decreasing protein in the urine but also does not cause adverse effects on the well-being of the child. The "team approach," when coupled with ongoing treatment, blood pressure control, and dietary maintenance, helps increase the likelihood of best outcomes in Alport patients.

## **Alport Syndrome Key Terminology**

To communicate with a nephrologist most effectively, it's important to understand some common medical terminology associated with Alport syndrome.

**Hematuria:** The presence of blood in the urine. Microhematuria refers to blood in the urine that is only visible under a microscope; it is often the first symptom for many patients. Gross hematuria refers to the presence of red blood cells in the urine resulting in a visible pink, red, or brown urine color (observed in diapers of babies, or in urine stream of school-aged children). *This term will not be found on a lab report.*

**Proteinuria:** The presence/amount of protein in the urine. The onset of [Alport syndrome treatment](#) for your child may be based on the time of your child's diagnosis, genetic type of Alport syndrome, and/or the amount of albumin (protein) observed in their urine. *This term will not be found on a lab report.*

**Serum creatinine:** Measured via a blood test, nephrologists use this value to assess the degree of kidney impairment. A lower value = better kidney function; a higher value = increased kidney injury. Levels can fluctuate due to medications, hydration, blood pressure, and other factors. This value is used to help calculate eGFR (defined below). *This term can be found on a lab report.*

**Estimated glomerular filtration rate (eGFR):** A measure of overall kidney function that is calculated using your serum creatinine level, age, gender, and other factors. Scores are reported on a scale of 1 to 100, with 100 indicating full kidney function. eGFR determines your chronic kidney disease (CKD) stage from 1 to 5. Stage 1 indicates strong renal function; stage 5 requires dialysis/transplant. *This will likely appear on your comprehensive metabolic panel report.*

## **Results to Track Over Time**

*Note: Blood and urine tests may be ordered in advance of, or following, your child's first visit with a pediatric nephrologist. Together, these tests provide information regarding kidney function.*

*Tip: Parents are encouraged to 1) track lab values by always asking for a printed or digital copy of blood/urine test results and 2) maintain a personal record of key values. Parents have shared ways of storing this information such as a dedicated notebook, notes on their cell phone, an excel sheet, or notetaking apps. It's important to keep organized records and have them readily available as needed.*

Suggested values to track:

- Blood pressure at each appointment: This can also affect kidney health.
- Urine protein and urine protein to creatinine ratio: A measure indicating the amount of protein in the urine. This ratio can indicate renal injury.
- Potassium: One of many electrolytes monitored in renal patients. The use of ACE/ARB medications can lead to hyperkalemia (high potassium levels), so routine monitoring is key. High potassium can put patients at risk for serious side effects.
- Serum creatinine (defined above)
- eGFR (as defined above): Your physician may need to calculate and provide to you, if not on a lab report.

## **Recommended Questions to Ask the Pediatric Nephrologist:**

*TIP: It is recommended to bring a list of questions with you to nephrology visits, either on paper or on a cell phone, and a way to track the nephrologist's answers. This way, you won't forget important questions you have or information learned during the appointment.*

***TIP: Is it strongly recommended to ask your child's pediatric nephrologist about blood pressure reading results, serum creatinine, and the presence/amount of protein in the urine (proteinuria) during the first visit and all follow-up care visits.***

Have you treated other patients with Alport syndrome before?

Are you aware of and/or do you follow the [latest \(2020\) recommended guidelines for treating pediatric Alport syndrome](#), or do you have a different threshold for starting a preventive medication?

How often would you like to see my child for follow-up? When is my child's next appointment?

Will any blood or urine tests be required prior to the next appointment with you? Am I leaving with printed lab orders or will they be electronically sent to the testing location?

Is future lab work performed prior to, or the morning of, my child's next nephrology appointment?

Will I be able to view my child's lab results electronically or can I have a printed copy for my records?

Do you recommend any changes to my child's diet? Can you refer us to a renal dietician so that we build a relationship and educate ourselves as my child grows?

What physical symptoms should I be mindful of in my child (Ex. Color and presentation of urine, fatigue, dizziness)? Are there any symptoms that would warrant a call to your office?

What time of day should my child take the ACE/ARB medication? If it makes my child sleepy/dizzy, could it be taken only at bedtime to mitigate the effects of low blood pressure?

How long will my child be on this treatment protocol for Alport syndrome?

How much water should my child drink each day for proper hydration and to mitigate side effects of treatment medication(s)?

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Additional information on Alport syndrome treatment can be found [on the ASF website](#). The ASF website also includes numerous [resources for Medical Professionals](#), including [published research](#) and a free, [60-minute CME/CNE](#) (Continuing Medical Education online course for healthcare providers).