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ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

Treatment Considerations for Pediatric Patients With FSGS

Announcer:
Welcome to CE on ReachMD. This activity is provided by Medtelligence and is part of our MinuteCE curriculum.

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Dr. Lieberman:
This is CE on ReachMD, and I'm Dr. Kenneth Lieberman, a pediatric nephrologist. Today we'll discuss some important issues surrounding the pediatric presentations and treatment of FSGS.

In addition to the usual FSGS presentations that you're used to in adult medicine—namely, incidental finding of decreased kidney function, significant proteinuria, proteinuria plus hematuria, may or may not be accompanied by hypertension—we see more nephrotic syndrome presentations for our FSGS patients. And we come to them differently than you would in adult practice, and that is our patients get a trial of steroids before definitive diagnosis through a biopsy is made.

And so we only biopsy our patients if they have failed steroid treatment, which is a relatively small percent of our total. Of those patients who have been steroid resistant who we then biopsy, overwhelmingly, our most common biopsy finding is FSGS.

So at that point, we are on to secondary, second-line treatments.

Immunosuppressive treatments in this group of steroid-resistant patients have limited efficacy and are usually not beneficial. Occasionally, a monoclonal antibody will be used, but our mainstay at this point is going to be anti-RAAS treatment, ACEIs and ARBs, as would be the case in adult patients.

Our goal at this point is to minimize proteinuria, and in fact, nephrotic syndrome–level proteinuria is the most predictive of a rapid decline in kidney function. And so that will be our goal—to reduce the proteinuria and improve the quality of life of these children without exposing them, hopefully, to the consequences of some of the medications.

We've come to understand that FSGS is at least 3 major subsets, which is the primary or putative circulating factor—sometimes called idiopathic nephrotic syndrome—probably that is the most frequent in pediatrics. We know in pediatrics that we are facing an increased incidence of genetic FSGS as compared to adults. In peds, that incidence is probably more in the nature of 40%-50% of all-comers, rather than the around 10% or so that is true in the adult population.

And then the third category, which really is more frequent in adults, is the hemodynamic or maladaptive type of FSGS, generally due to reduction in renal mass by some mechanism or another. There is medication-induced FSGS. There's viral-induced FSGS. But these are smaller categories.

Because of this increased incidence of genetic FSGS in children, the importance of genetic testing has really increased. In my practice and many other pediatric practices, genetic testing is actually done before the biopsy, because if we can identify a known causative

variant, we can spare the child the biopsy.

The key takeaway points for this presentation are, number one, that children are more likely to present with the nephrotic syndrome. And at the time that we make the diagnosis by biopsy of FSGS, we have already established that they are steroid resistant.

Secondly, I would like to highlight the increased importance of genetics as an etiology for FSGS, especially in children.

And thirdly, I'd like to emphasize the emerging knowledge about the subsets of FSGS being primarily the primary or idiopathic or circulating factor FSGS, genetic FSGS, hemodynamic or maladaptive FSGS with 2 minor classifications: the drug-induced FSGS and viral causes.

That's all of our time for today. I hope you have found this presentation useful. It's been my pleasure presenting it.

Announcer:
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