

The kidney in Fabry disease

To the Editor:

We have read with great interest the article by Desnick and Brady¹ on Anderson-Fabry disease (FD) in childhood. As reviewed by the authors, FD is an X-linked glycosphingolipidosis as a result of the deficiency of the enzyme α -galactosidase A. The accumulation of globotriaosylceramide in the lysosomes of various tissues is the underlying known pathology causing the clinical symptoms. Until recently, most disease features had been reported in adults. Hemizygous adult males present with acroparesthesias, angiokeratomas, corneal opacities, hypohydrosis, and end-organ disease affecting the kidney, heart, and brain. Interestingly, years ago glycolipid storage was reported in fetal kidney, myenteric plexus,² and fetal cornea.³ Since then, pediatric observations of children with FD have been published,⁴ but during the last few years, the pediatric FD phenotype has been studied and reported more systematically.^{5,6} The review by Desnick and Brady¹ in *The Journal* emphasizes the importance of this disease in pediatric practice.

We report on the daughter of a kidney-transplanted patient with FD reported by Friedlaender et al.⁷ At 12 years of age, she had a kidney biopsy that demonstrated the typical changes of FD (Figure), including striking, diffuse foamy vacuolization of the glomerular epithelial cells, and at 14 years of age, bilateral cornea verticillata were observed. This observation emphasizes that significant kidney involvement can be present not only in males but also in heterozygous females, even as early as 12 years of age. Complaints of acroparesthesias in children should lead to urinalysis for proteinuria. Today, the pediatrician has to be aware of FD because early diagnosis is mandatory for accurate genetic counseling and timely enzyme replacement therapy with the hope of preventing complications of the disease.

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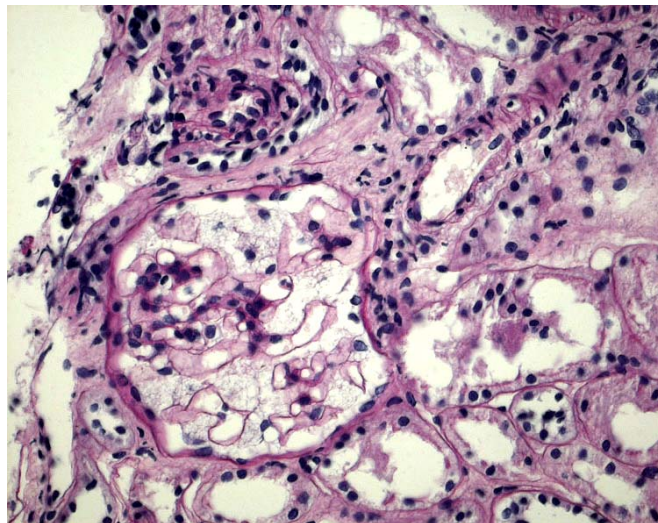


Figure. Renal biopsy showing enlarged podocytes with foamy cytoplasm that cause glomerular atelectasis. Periodic acid-schiff stain, original magnification. (Figure available in color online at www.us.elsevierhealth.com/jpeds.)

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Low birth weight and blood pressure: The role of neonatal factors in the "small-baby-syndrome"

To the Editor:

With regard to the phenomenological relationship observed between low birthweight and increased cardiovascular risk, Gillman et al published an important study on perinatal predictors of newborn blood pressure.¹ Interestingly, in contrast to data from long-term studies on the "small-baby syndrome," low birthweight is associated with *low* blood pressure in newborns. Launer et al show that the direction of the relation between birthweight and blood pressure reverses after 3 months of age.² We suggest that this reversal in early postnatal life strongly indicates the presence of neonatal factors acting critically, factors which are able to set those