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Pediatric Nephrology

Ask the expert*

A 3-year-old girl had one episode of isolated macroscopic hematuria. One week later, urine examination was negative for protein and red blood cells. Intravenous pyelography was not conclusive, but renal ultrasound examination showed one cyst of 1 cm in the right kidney. Ultrasound examination of the mother, age 42 years, showed one cyst in each kidney. There is no known history of renal disease in the grandparents. The father of the girl was not available for examination. What is the diagnosis? Are there other clinical investigations to be performed in this case?

Key words: Hematuria – Renal cyst – Polycystic kidney disease

Two questions are raised by this patient: (1) the diagnosis of the cystic lesion and (2) the causal relationship between the cyst and the hematuria. At the age of 8 years, a simple renal cyst is very uncommon [1]. Therefore such a diagnosis can only be made after a cystic renal disease has been ruled out. In the present case, the finding of one cyst in each of the mother's kidneys may be suggestive of autosomal dominant polycystic kidney disease (ADPKD), in which cysts are often unilateral in childhood. However, at age 42 years two cysts in *each kidney* are required for the diagnosis [2], at least in PKD1, the most frequent genetic form (and, until now, the only one where cysts are detectable in childhood). Some small cysts, however, may be missed by ultrasonography, and a computed tomographic scan of the mother's kidneys could be useful to disclose other smaller cysts which could support the diagnosis of ADPKD. If available, ultrasonography of the grandparents would also be very useful. Some other autosomal dominant diseases may be associated with renal cysts, such as tuberous sclerosis, in which extrarenal symptoms may be lacking or insignificant. The girl and her mother should be evaluated for cutaneous or neurological symptoms.

The causal relationship between the renal cyst and macroscopic hematuria is also questionable. Hematuria is a classical symptom of renal cysts in ADPKD [3], the mechanism of which is not always clear. Lesions of the cyst wall caused by stones which have developed inside the cyst or traumatic rupture of a cyst are the two most convincing causes of cyst hemorrhage. However these two factors are not always found. Microlithiasis, which is not apparent on X-rays or ultrasonography, may explain some hematurias, and urinary crystals and predisposing factors such as insufficient water intake, hypercalciuria, hyperoxaluria, etc. should be looked for.

The presence of a renal cyst does not preclude a glomerular origin of hematuria. The absence of proteinuria and microscopic hematuria 1 week later does not completely exclude an IgA nephropathy, for example. Microalbuminuria should be measured and, if hematuria recurs, a morphological analysis of fresh urinary erythrocytes would be required to eliminate a glomerular origin. If all these investigations are normal, no definitive diagnosis is possible and serial ultrasonography of the girl and her mother will be necessary over several years.

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* The editors invite questions for this section

Book review

The replacement of renal function by dialysis
Edited by C. Jacobs, C. M. Kjellstrand, K. M. Koch, and J. F. Winchester, Kluwer, Dordrecht, 1996, pp 1535, US $ 325.00, ISBN 0792336100

The fourth revised edition of *The replacement of renal function by dialysis* represents an extensive revision of a classic textbook. Since the book first appeared in 1978, it has set high standards for excellence and distinctive scholarship in each chapter. The current edition is virtually a new book. In the 7-year interval between the third and the fourth editions there has been an explosion of new information. This has necessitated extensive rewriting and a much larger text.

The authorship represents an international collaboration. There are 139 contributors! The organization of the text is excellent. The chapters are grouped into eight sections entitled: "pathophysiology of the uremic syndrome – principle and biophysics of dialysis, technology of dialysis and associated methods, quantification and prescription of dialysis, complications of dialysis, pharmacological considerations, special clinical situations, organ system and metabolic complications in chronic dialysis," and "organization and results of chronic dialysis." Each chapter has been extensively rewritten. With few exceptions, tables, figures, and radiographs have been carefully selected and enhance major points in the text. Although the authors devoted one chapter to pediatric dialysis and transplantation, the book generally ignores differences between adults and children. Growth and nutritional management of children undergoing dialysis treatment and developmental aspects of the kinetics of peritoneal dialysis in children are largely unmentioned. A section on developmental renal physiology would highlight important changes which occur in the kidney during the first years of life.

Overall, this book represents an authoritative, valuable, and comprehensive resource which will be essential for anyone who takes care of patients with end-stage renal disease. Although not a pediatric text, it is a must for the nephrologist's shelf.

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