

CORRESPONDENCE

To the Editor:

In this issue of the Journal, Longaker et al (pp 411-414) describe two cases of nephron-sparing surgery in metachronous (secondary) bilateral Wilms' tumor, in the kidney remaining after a previous unilateral nephrectomy. The rationale for this delicate in situ, as well as ex vivo, surgery is twofold. First, the investigators state that the outcome of renal transplantation (RT) in bilateral Wilms' tumor is poor. Second, they state that, as much as possible, renal tissue has to be saved in order to prevent early functional deterioration of the remnant renal mass. We would like to comment on these two statements.

The outcome of renal replacement therapy in these children is affected by various factors. The 3-year survival rate of patients having synchronous bilateral Wilms' tumor is about 80%,¹ but prognosis is poor in metachronous bilateral tumors.² Until 1980, experience with RT in relatively young children was limited, and the outcome generally was poor, irrespective of the cause of the renal failure. More recently, survival rates of children undergoing transplantation at an early age have improved considerably.³ The outcome of RT, in cases of bilateral nephrectomy for Wilms' tumor, is further worsened by the effects of radiation and chemotherapy, which in combination with the antirejection therapy, resulted in a high death rate due to sepsis.^{4,5} Therefore, the present policy is to wait till at least 2 years after the tumor removal before RT is done. Single centre⁶ and European Dialysis and Transplant Association data⁷ show that RT is then possible with a good outcome.

However, if possible, nephron-sparing surgery remains the treatment of choice. In this way, the devastating effects of renal failure on young infants and children can be avoided or reduced. Saving enough renal tissue to keep glomerular filtration rate (GFR) at an adequate level of greater than 50 mL per minute per 1.73 m² is preferred to early dialysis. The minimal amount of renal tissue needed to obtain this level would be about 20% to 25% of the total, ie, 40% to 50% of one kidney. In rats, progression into renal failure after unilateral nephrectomy at a young age,⁸ and more severe forms of initial loss of renal function^{9,10} have been demonstrated.

Little information is available on the progression into chronic or terminal renal failure of remnant kidneys in humans. Recent follow-up data from Paris⁶ on 36 cases, with various amounts of renal tissue left, indicated that three went into end stage renal failure (GFR <10 mL/min/1.73 m²) and three into chronic renal failure (GFR 10 to 40 mL/min/1.73 m²). With a single exception, it took 6 to 17 years to reach this stage of functional deterioration. There was no clear correlation between the amount of renal tissue removed and the time necessary to develop renal failure. This contrasts with data obtained from rats, but it should be kept in mind that in humans, biological variation is larger and environmental factors are much more variable than in experimental animals.

A complicating factor is the irradiation of the remnant part of the kidney. Although radiation therapy may be necessary, it could be harmful to the remaining part of the kidney. We have shown that the growing kidneys of young rats are more susceptible to irradiation than those of adult rats.¹¹ Young rats develop hypertension, proteinuria, and chronic renal failure earlier than adults. This may be true in children. Therefore, children with an irradiated remnant kidney, after a nephron-sparing surgery for bilateral Wilms' tumor, are at an even higher risk of developing chronic renal failure. Regular control of renal function is indicated, if possible, with a more adequate assessment than a simple plasma creatinine determination.

In conclusion, in cases of bilateral Wilms' tumor, conservative surgery remains the best choice if the tumor can be completely removed and enough renal tissue is left to guarantee the adequate renal function necessary for the physical and mental development of these children. With time, renal failure may develop, but RT is then possible if the patient has remained without a Wilms' tumor for at least 2 years.

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