BANTAO Journal 3 (2): p 44; 2005

Inherited Kidney Diseases: the Necessary Transition from Pediatric to Adult Care

J.P. Grünfeld
Université Paris-René Descartes, Hôpital Necker, Paris, France.

Transition from pediatric to adult care is a common problem in chronic diseases that start in childhood and extend to adulthood, such as cystic fibrosis, congenital cardiopathies, renal diseases etc. Obviously it is a crucial issue in inherited kidney diseases. I will discuss not only why transition in medical care is necessary, but also why research should be shared, and training in these diseases should be offered to adult nephrologists. Finally I will underline how this transition is difficult, and should be prepared, organized and coordinated.

Many inherited kidney diseases are discovered in children, such as Alport syndrome, and progress to ESRD in adults. These patients (and their families) require extended follow-up (1). In autosomal recessive polycystic kidney disease, half the patients who survive the neonatal period, are not in ESRD at 15 years of age. Most of these will need renal replacement therapy and liver management in adulthood (2). Some rare inherited disorders may reveal late in life (whereas most patients are diagnosed in childhood) or may unexpectedly progress very slowly to ESRD: this is the case, for example, in Alagille syndrome (3) (characterized by paucity of intrahepatic bile duct responsible for chronic cholestasis, butterfly vertebrae, and renal involvement) or in Lowe syndrome (4) (X-linked oculo-cerebro-renal syndrome), respectively.

Surprisingly renal involvement may also develop in adulthood in some inherited metabolic diseases, such as type 1 glycogen storage disease (or von Gierke disease); many patients become proteinuric between 15 and 30 years of age, and develop progressive lesions of focal segmental glomerulosclerosis (5).

For these various reasons, transition between pediatric and adult care should be coordinated (6,7). "Transfer is an event but transition is a process" (A.R. Watson). There are many obstacles to transition coming from pediatric professionals, adolescents and their parents, and receiving adult services, often unprepared to this task. Successful transition requires tight cooperation and exchange of information between all partners. It should be programmed, prepared by "joint outpatient clinics". Transfer at times of crisis, particularly on the home front, should be avoided. The age range at transfer varies between 16 to 25 years, with most favouring 18 years. It depends mainly of the "maturity" of the young person to move from pediatric to adult care. In all cases transition should be prepared for a sufficient period of time before transfer, and information should be given to patients and families. Various procedures to facilitate transition have been proposed, including peer support from those who have already made the transition.

In **conclusion**, transition from pediatric to adult care is a major challenge for modern medicine. It requires new resources and adequate training of health professionals.

References

1. JP Jais, B Knebelmann, I Giatras et al. X-linked Alport syndrome: natural history in 195 families and genotype- phenotype correlations in males. *J Am Soc Nephrol* 2000;11: 649-657
2. C Fonck, D Chauveau, MF Gagnadoux et al. Autosomal recessive polycystic kidney disease in adulthood. *Nephrol Dial Transplant* 2001;16:1648-1652
3. M Schonck, S Hoorntje and J Van Hoofff. Renal transplantation in Alagille syndrome. *Nephrol Dial Transplant* 1998;13 :197-199
4. L Tricot, Y Yahiaoui, L Teixeira et al. End-stage renal failure in Lowe syndrome. Nephrol Dial Transplant 2003;18:1923-1925
5. SH Morgan and JP Grünfeld. Other inherited metabolic storage disorders with significant renal involvement. Oxford Textbook of Clinical Nephrology 3rd Edition 2005;3:2351-2355
6. R Viner Barriers and good practice in transition from paediatric to adult care. J R Soc Med 2001;94 (40):2-4
7. SD Callanan, R Feinstein Winitzer and Kenan P. Transition from pediatric to adult-oriented health care: a challenge for patients with chronic disease. Current Opinion in Pediatrics 2001;13:310-1

Correspondence to: Jean-Pierre Grünfeld, Université Paris-René Descartes, Hôpital Necker, Paris, France