Port J Nephrol Hypert 2007; 21 (3): 211-217

Paediatric nephrology patients moving to adult age: a difficult transition to manage. The experience of a paediatric-to-adult out-patient clinic

Sofia Jorge[1], Fernando C. Neves[1], Erica Mendonça[2], Rosário Stone[2], Margarida Almeida[2], Mateus Martins Prata[1]

[1] Renal Transplantation and Nephrology Department
[2] Paediatric Nephrology Unit
Hospital de Santa Maria. Lisbon, Portugal.

Received for publication: 07/01/2007
Accepted in revised form: 22/03/2007

ABSTRACT

The continuity of care of patients whose disease process begins in childhood poses a fresh challenge for medicine and one which nephrology and other medical specialisations must face. Hospital de Santa Maria has had a transition clinic based in the paediatric unit to orientate this transition from paediatric to adult nephrology since 1998. We retrospectively analysed the clinical outcome of the patients referred to this clinic, studying adhesion to transition, renal function and need for renal replacement therapy.

Sixty-nine patients have been referred for transition since 1998 and five of these have never attended a transition visit. The prevailing pathologies were glomerulonephritis (28 patients), tubulopathies (13 patients), uropathies (19 patients) and other diagnosis in 4 patients. Mean follow-up time was 17.6 years and current mean age is 23.3 years.

Fourteen of the 64 successfully transferred patients (38 males, 26 females) left the clinic. Four transferred to other hospitals, nine progressed to end-stage renal failure and needed renal replacement therapies and one abandoned the clinic.

We discuss the results obtained, the efficacy of our transition model and particular aspects of the prevailing pathologies, particularly outcome of renal function.

Key-Words:
Adult nephrology; paediatric nephrology; renal failure.

INTRODUCTION

Chronic diseases beginning in childhood present doctors with a challenge; the task of managing patients' transition to adult health care services. Renal disease is no exception, and the particular aspects of this specialisation influence this transition process.

The disease's physical and psychological consequences and the need to adapt to transition cause difficulties for patients and parents alike and constitute obstacles to the success of this task[1-6]. An important aspect to bear in mind is the individual personal and family circumstances of each patient as transition has to be adapted to these circumstances[1]. Adolescents with chronic kidney disease are frequently over-protected by their parents and

sometimes also by health caregivers, which compromises their authonomy[2,3], especially for those who grew up in hospital nurseries. The adolescent patient faces the difficulties of transfer of care just when he or she is already struggling with problems of identity, independence, self-image, burgeoning sexuality and the need to define his/her goals in life, against a background of the additional burdens of his or her chronic illness and its multivarious treatments and restrictions[7].

The broad spectrum of nephrological pathologies also affects patients whose clinical conditions require annual visits, such as after an episode of haemolytic uraemic syndrome or Henoch-Schönlein purpura, and those who have variable degrees of proteinuria, hypertension or renal failure. While these patients have a weaker bond to paediatric services, making them less resistant to transition to adult care, they can also easily abandon medical care because they feel and look well. Although the management of those in clear need of continued care remains paramount, it is not apparent which of these patients should be referred to a nephrologist or to a general practitioner, and in addition guidelines for their follow-up management are lacking[4].

We considered it vital for an adequate follow-up of these patients that they remained in the same hospital they were in before transition.

PATIENTS AND METHODS

The patients transferred to adult clinic are those referred by the paediatrician who have kidney disease needing continuous care in adult age, but are not yet on renal replacement therapy. Patients who already reached end-stage renal failure before adulthood and are on renal replacement therapy when they reach this age are referred directly by the paediatrician to peritoneal dialysis or transplantation units at our hospital or to extra-hospital haemodialysis units. They are not referred to our clinic and were excluded from our analysis.

Patients are usually transferred to an adult clinic when they are eighteen years old. In our clinic, transferral occurs through the organisation of a specific visit. This takes place at the paediatric unit where the patient has always met the paediatrician, with the paediatrician introducing the patient and the parents to the future doctor. In that meeting the adult's nephrologist is informed of the patient's medical condition, and the next visit is scheduled at the adult clinic. The patient and parents are also informed of the new rules, including the fact that from that date on the parents' presence at the medical visits depends on the patient's say-so.

There is prior preparation for this transition by the paediatrician, who explains its reasons, relevance, and clarifies patient and parents' doubts or difficulties. The length of this preparation depends on the patient and family´s individual behaviour and needs.

This new model was adopted in 1998 and since then the transition meetings have taken place at the paediatric clinic on a weekly basis.

Patients from the paediatric departments of other hospitals are also referred to this clinic, either by their paediatrician or general practitioner.

We retrospectively studied 64 out of 69 patients referred for transition from 1998 to 2005, aiming to evaluate adhesion to transition and to characterise the clinical evolution of the patients transferred, especially renal function.

RESULTS

Sixty nine patients were referred for transition, with five of them never attending the visit. Sixty four patients were transferred; 26 females, 38 males. Mean age at diagnosis was 6.7±4.3 years, mean age at transfer time was 18±3.2 years and the mean age at present time is 23.3±3.8 years. Mean medical follow-up time is 17.6±4.8 years; 13.7±5.3 of which are at the paediatric clinic and 3.9±2.9 at the adult clinic.

The prevailing pathologies were glomerulonephritis (28 patients), tubulopathies (13 patients), uropathies (19 patients) and 'other' in four patients. Eighteen renal biopsies were performed on thirteen patients, with only two performed at the adult clinic. All patients submitted to renal biopsy had glomerulonephritis.

After transferral, three patients needed hospital care; deep venous thrombosis (one patient) and nephrotic syndrome relapse (two patients).

Renal function at transfer was normal in 46 patients (72%), but eighteen (28%) had chronic kidney disease (CKD) K/DOQI classification[8]; 9 patients CKD stage 1 and 9 patients CKD stage 4. Fourteen patients left the adult clinic. One left medical care, four were transferred to other clinics and nine patients developed end-stage renal failure. Fifty patients remain at the adult clinic. Renal function remains normal in 41 patients and nine patients have CKD stage 2. In those patients who progressed to end-stage renal failure (14 %), the cause of renal failure was glomerulonephritis in five patients, uropathies in three cases and unknown in one patient.

At the present moment, four of these patients are on haemodialysis, one is on peritoneal dialysis and four have received a renal transplant.

Glomerulonephritis

Twenty-eight patients were transferred; 16 female, 12 male. Most of the glomerulonephritis were primary nephrotic syndromes (PNS) (Table I).

Table I

Prevailing glomerulopathies. PNS: Primary nephrotic syndrome, IgA/HSN: IgA nephropathy, Henoch-Schönlein purpura; SLE: Systemic Lupus Erythematosus.

Glomerulopathy	Nr of patients
PNS	15
IgA/HSN	6
SLE	5
Alport´s disease	2

Mean age at diagnosis was 8.2±4.3 years, mean age at transfer was 19.3±2.8 years and mean age at the present is 22.7±4.2 years.

The mean follow-up time is 14.7±4.3 years; 10.7±5.0 years at the paediatric clinic and 4±2.2 years at the adult clinic. The thirteen patients submitted to renal biopsy had a relatively rare type of glomerulonephritis (Table II).

Table II

Histological diagnosis in the patients submitted to renal biopsy.

Histological diagnosis	Nr of patients
FSGS	1
Minimal change disease	1
Membrano-proliferative GN	2
IgM nephropaty	1
Mesangial Proliferative GN	1
IgA nephropathy	4
Lupus nephritis (WHO class II)	1
Alport´s disease	2

At transfer six patients had chronic kidney disease, one at CKD stage 1 and five at CKD stage 4. Five patients left the adult clinic when they reached end-stage renal disease and needed renal replacement therapy, four patients were referred to other clinics and one patient abandoned our clinic. There are currently 18 patients remaining at our clinic and all have stable renal function.

In those patients who progressed to end-stage renal failure, only three had histological diagnosis: one membranoproliferative glomerulonephritis type I, one IgA nephropathy and one Alport´s disease.

Six patients with primary nephrotic syndrome (PNS) were submitted to renal biopsy in the paediatric clinic with the following histological diagnosis: two minimal change disease, two membranoproliferative glomerulonephritis, one mesangial proliferative and one IgM nephropathy. After transferral, these patients experienced relapses, less than one per patient per year; all patients were cortico-sensitive and three patients were on cyclosporine when they relapsed. All were treated with corticosteroids with remission. Renal biopsies were repeated in two patients with a previous histological diagnosis of minimal change disease. This was confirmed in one of them and in the other the second renal biopsy revealed focal segmental glomerulosclerosis (FSGS).

Three patients with PNS progressed to CKD stage V; two without histological diagnosis and one with membranoproliferative glomerulonephritis type 1.

Of the four patients with IgA nephropathy and the two with Henoch-Schönlein purpura, three had frequent relapses of haematuria-proteinuria and one patient progressed to end-stage renal failure.

Tubulopathies

Thirteen patients with tubulopathies were referred for transition, seven male and six female (Table III). Mean age at diagnosis was 3.6±2.4 years, mean age at transition was 23.7±4.9 years and at the present moment mean age is 27±9.2 years.

Table III

Prevailing tubulopathies.

Tubulopathy	Nr of patients
Renal tubular acidosis	3
Bartter´s syndrome	3
Hypophosphatemic rickets	4
Idiopatic hypercalciuria	2
Non-specified tubulopathy	1

Mean follow-up time was 21.9 years, 18 years in paediatric nephrology and 3.9 years in adult nephrology. There was no drop-out.

All prevailing co-morbidities were present prior to the transition (Table IV).

Table IV

Prevailing co-morbidities in the patients with tubulopathies

Comorbidity	Patients	Diagnosis
Nephrocalcinosis	5	Distal renal tubular acidosis (2)
		Bartter´s syndrome (1)
		Idiopathic hypercalciuria (1)
		Non-specified tubulopathy (1)
Growth retardation	4	Hypophosphataemic rickets
Skeletal deformities	2	Hypophosphataemic rickets

In two cases of hypophosphataemic rickets, the patients were female, and descendents presented the disease, which may lead us to conclude that these were hereditary cases, autosomal dominant or X-linked.

There have been no medical complications such as renal function deterioration or admission to hospital up to the present day. All patients remain at the adult clinic with no need for therapy change, and the follow-up requires one to four visits a year.

Uropathies

Nineteen patients with nephropathy secondary to urologic problems were transferred to our clinic, 14 male and 5 female.

Mean follow-up time is sixteen years, mean age at diagnosis was 4.8±4.6 years, at transfer was 17.7±2.6 years and mean age at the present is 22.7±3.3 years.

The most prevailing uropathies are vesico-ureteral reflux (eight cases, five of which were caused by posterior ureteral valves) and complex non-specified uropathies (five cases). Table V.

Table V

Prevailing uropathies.

Diagnosis	Patients
Posterior ureteral valves	8
Pieloureteral duplicity	2
Junction syndrome	1
Prune Belly syndrome	1
Oxalosis	1
Dysplasic kidney	1
Complex uropathy	5

Most of these patients (10/19) underwent more than one surgical procedure to correct the anatomical abnormalities. Hypertension was highly prevalent in this population (17/19 patients) and all received ACE inhibitors and when necessary other anti-hypertensive drugs to control blood pressure. There was no proteinuria in these patients.

Renal function remains stable in sixteen patients, ten of which had normal renal function and six of which were already suffering from chronic kidney disease CKD stage 3. Three patients progressed from CKD stage 4 to stage 5 (oxalosis and two complex uropathies) and are now on renal replacement therapies (2 received a renal transplant and one is on haemodialysis).

DISCUSSION

Non-adhesion to health care transition is one of the most important obstacles that we face. Changes are always seen as destabilising the fragile balance of these patients´ lives. However, transition can be seen as a challenge through which these adolescents gain maturity, autonomy, self-esteem and self-confidence[9]. Choosing the most appropriate moment for transferring these patients is crucial to the success

of transferral. In our experience, this moment is defined by patients' age, but individually adapted to the patients' personal and family circumstances. We receive patients from two different paediatric services which use different age criteria for transition, reflecting the absence of a consensus position towards continuity of care.

One of the worrying aspects of transition for patients and family, especially parents, are doubts about their future participation in health care[10,11]. This is why we explain to them at the transition meeting that from that time on, the parents' presence depends on the patient's say-so.

The differences between the two medical cultures are also an obstacle, because of the gap between the family-like and protective attitude of the paediatric services and the coldness and formality of the adult medical services. Neither patients, family, or doctors know how to handle these differences[10-12]. We try to humanise transition in this first meeting, with the presence of the paediatrician making it a more familiar event. Obviously, the paediatrician's presence also serves another purpose, which is the cessation of relations with paediatric services, and the promotion of a trusting relationship with the future doctor. Medical information is transmitted in the presence of the patient and parents, which allows them to see that no information is lost in transition. According to the literature, some paediatricians may fear transition as they lack trust in the future doctor[5], but this is not something that we have ever had to deal with.

Our transition model only provides for a single meeting with the adult nephrologist and it could be debated that this may be insufficient for patients to feel familiar with the future doctor. It is undeniable that the creation of any relationship requires more time, but this was the solution that we found for a problem that includes the limited time healthcare professionals, patients and parents have available.

Obviously, there is the serious and gradual work of the paediatrician in preparing the patient and family for the need of transfer to the adult clinic, which is fundamental to decrease anxiety, clear doubts and allow a smooth and gradual transition. This cannot be reduced to one simple meeting and a mere "say hello, wave goodbye".

In the literature, we find references to different models of transition[1]; the type of clinic adopted appears to depend on local circumstances, considering the fact that the number of patients transferred yearly in each centre will usually be small. While some centres promote a series of pre-transfer meetings at the transition clinic between the adult nephrologist and the young person, in most instances there is a single transfer clinic at some distance from the regional paediatric unit. This latter arrangement demands preparatory work in the giving of information and competency checklist verifying[1].

The parents' presence at the first meeting is also seen as useful to diminish the family´s anxieties[1].

While we found references to these models and the ideal methodology, detailing all the difficulties felt by patients, parents and caregivers, we found no description of any transition clinic's experience and feel the models of transition should be discussed and validated. In describing our experience, we are reflecting on and critiquing our model, although some other aspects should be assessed to properly evaluate it, such as patients´ and parents´ opinions.

In this group of patients, five of the 69 referred for transition never attended their transition meeting. These were patients with normal renal function, whose medical condition allows them to feel and look well.

In the 64 patients who were successfully transferred, there was only one case of drop-out, which was related to the patient's poor social and economic conditions.

These results support the fact that in this model, the most delicate moment of healthcare transition is the moment of transfer and not the adaptation to the adult clinic, which apparently succeeds.

These numbers may reflect the efficacy of our model of transition, but also the selection of patients who are motivated and conscious of the importance of continuity of medical care. The patients who transferred to other clinics did so because they changed address, not because they were dissatisfied with their medical care. Only one patient left consultation and this was due to poor social conditions and lack of familial and social support. We did not gauge patient

or family satisfaction with transition and the continuing care at the adult clinic, which could be fundamental to a thorough evaluation and validation of this model. However, our results, with only one patient abandoning medical follow-up after transition, may reflect at least minimal satisfaction with access to continuing medical care. Moreover, patients and families frequently mention the importance of medical care, the need to understand their disease and to be stakeholders in their healthcare. Patient and family motivation is a characteristic of this population.

There were, however, five patients who never attended the transition visit. These were patients who feel and apparently look well and who consequently believe they do not need medical care.

The incidence of CKD in our population was 14%, with the main aetiology being glomerulonephritis. 18% of the patients with glomerular diseases progressed to end-stage renal failure despite medical follow-up.

We interpret these results as reflecting a prior selection of patients referred for transition and not because glomerulonephritis in our paediatric population were particularly severe. In fact, considering not only glomerulonephritis, but also tubulopathies and uropathies beginning in childhood we must consider that the most severe forms progress to end-stage renal disease before adult age, and these patients never arrive at our adult clinic, but instead are referred for renal replacement therapies and are followed in the specific clinics (haemodialysis, peritoneal dialysis and renal transplantation). Consequently, there is a clear bias in the prevailing pathologies in the young adults that are referred to our adult clinic; we cannot say that we receive the most ill patients or the healthiest ones, but those in between these two extremes. Neither can we interpret our results as reflecting the main aetiologies of CKD stage V in children or young patients, because we only follow some of them. In fact, in some series, uropathies frequently followed in glomerular diseases are given as the most prevalent causes of CKD (all NKF CKD stages) in children[13].

In sum, what we can say is that in our experience, patients with glomerulonephritis are older than those with tubulopathies and uropathies at the time of diagnosis (mean age 8.2 *vs.* 4 and 4.8 years respectively). Glomerulonephritis have a more unpredictable relapsing-remitting course with possible deterioration of renal function, requiring more follow-up. The duration of cyclosporine treatment and the possibility of using new immunosupressors should be discussed.

Uropathies have a variable clinical course but present several medical problems during follow-up and a high prevalence of CKD (48%, 9/19), particularly with progression to ESRD (15.8%, 3/19). Patients with tubulopathies are transferred at a later age, reflecting that the complications of the disease onset during the adolescent growth period and stabilise thereafter. Timely diagnosis of tubulopathies may allow adequate therapy and diminish the consequences of the disease. There are no references to the occurrence of arrhythmias in our population, despite the known risk of sudden cardiac death in patients with tubulopathy and serious hypokaliaemia. This can be explained by the efficacy of therapy or the reduced size of our population. However, literature documenting other series shows the same doubt over the incidence of this complication[14].

Among tubulopathies, hypophosphataemic rickets X-linked poses a problem in the patient's follow-up, especially female patients. The disease appears to stabilise with the end of adolescence, so the efficacy of maintaining therapy thereafter is questionable, bearing in mind the adverse effects of therapy. In addition, there is no evidence that during pregnancy phosphorus supplements must be offered, though in theory it is reasonable to do so because maternal supply is the only source of this ion to the foetus.

The medical follow-up of these patients allows us to have contact with specific and unusual nephrological pathologies. Therefore, it seems important to centre follow-up under a single nephrologist who can devote more care to this particular area.

CONCLUSIONS

The results obtained in adhesion to transition to adult nephrology allow us to say that this is a successful model in our population.

Despite tight follow-up, nine patients progressed to CKD stage 5. Several questions must be raised, including length of therapy with cyclosporine.

The specificity of CKD beginning in childhood and its particular aspects deserve attention from health care professionals, always bearing *kaizen*, or continuous improvement, in mind.

Acknowledgements

The authors would like to express their respect and acknowledgment to Professor Fernando Coelho Rosa, pioneer of Paediatric Nephrology in Portugal and to its beginnings at Hospital Santa Maria.

Conflict of interest statement. None declared.

References

[1] Watson AR. Problems and pitfalls of transition from paediatric to adult renal care. Pediatr Nephrol 2005;20:113-117

[2] Reiss JG, Gibson RW, Walker LR. Health care transition: Youth, Family, and Provider Perspectives. Pediatrics 2005;115:112-120

[3] Olsen DG, Swigonski NL. Transition to adulthood: the important role of the paediatrician. Pediatrics 2004;113:159-162

[4] Cameron JS. The continued care of children with renal disease into adult life. Pediatr Nephrol 2001;16:680-685

[5] Fox A. Physicians as barriers to successful transitional care. Int J Adolesc Med Health 2002;14: 3-7

[6] Brumfield K, Lansburry G. Experiences of adolescents with cystic fibrosis during their transition from paediatric to adult health care: a qualitative study of young Australian adults. Disabil Reahabil 2004;18:223-234

[7] Cameron JS. Transition of children with renal diseases into adulthood. In: Hogg R (ed): Kidney disorders in children and adolescents. A global perspective of clinical practice. London and New-York: Taylor & Francis, 2006: 253-261

[8] National Kidney Foundation. K/DOQI clinical practice guidelines for chronic kidney disease: evaluation, classification, and stratification. Am J Kidney Dis 2002; 2 Suppl 1: S1-266

[9] Pinzon JL, Jacobson K, Reiss J. Say goodbye and say hello: The transition from pediatric to adult gastroenterology. Can J Gastroenterol 2004;18:735-742

[10] Rosen DS. Transition of young people with respiratory diseases to adult health care. Paediatric Respiratory Review 2004;5:124-131

[11] Telfair J, Ehiri JE, Loosier PS, Baskin BS, Baskin ML. Transition to adult care for adolescents with sickle cell disease: Results of a national survey. Int J Adoles Med Health 2004;16:47-64

[12] Sawyer SM, Blair S, Bowes G. Chronic illness in adolescents: transfer or transition to adult services? J Paediatr Child Health 1997;33:88-90

[13] Wong H, Mylrea K, Feber J, Drukker A, Filler G. Prevalence of complications in children with chronic kidney disease according to KDOQI. Kidney Int 2006, 70: 585-590

[14] Cortesi C, Bettinelli A, Emma F, Fischbach M, Bertolani P, Bianchetti MG. Severe syncope and sudden death in children with inborn salt-losing hypokaliaemic tubulopathies. Nephrol Dial Transplant 2005; 20: 1981-1983

Correspondence to:
Dr Sofia Jorge
Serviço de Nefrologia e Transplantação Renal
Hospital de Santa Maria
Av. Prof. Egas Moniz
1649-035 Lisboa, Portugal
E-mail:sofiacjorge@sapo.pt