Chapter 13: Demography and Management of Childhood Established Renal Failure in the UK

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Summary

- The incidence and prevalence of ERF in children in the UK is relatively static at 8.0 and 47.7 per million population under the age of 15 years respectively.

- The prevalence of ERF in children from the South Asian community is almost 3 times that of the White population whilst the incidence is over 3 times that of the White population and similar to the increase seen in the adult population. The high incidence and prevalence are related to the high incidence of inherited diseases which cause ERF in the South Asian community.

- ERF in children is more common in males than females (male to female ratio 1.54:1). This is due to a preponderance of males with renal dysplasia and obstructive uropathy causing ERF. For the South Asian patients, the gender ratio is 1:1 as the inherited diseases are mainly autosomal recessive.

- Renal dysplasia is the single most common cause of ERF in childhood, followed closely by glomerular disorders and then obstructive uropathy.

- The majority of prevalent paediatric ERF patients (76%) have a renal allograft. Of these, 28% are from living donations.

- The proportion of patients from ethnic minority groups with a functioning allograft is significantly smaller than that in the White population ($p < 0.0001$). Despite this, the rate of living related donation is no higher in the ethnic minority population.

- In prevalent patients PD is twice as commonly used as HD with the majority managed with automated PD. For patients at one year from starting RRT, 49% are on PD, 10% on HD and 41% have a transplant.

Introduction

Knowledge of the demography of the ERF population is important both for the planning of service provision and for the development of preventative treatment programmes. This article covers the demography of ERF in children in the UK and their current modality of ERF treatment.

Paediatric ERF population

The paediatric arm of the Renal Registry currently holds data on some 1,800 patients who had ERF in childhood. A number of these patients have died and many have been transferred to adult units. The population of ERF patients being treated in paediatric units on 1st April 2005 stood at 768. This is a small fall on the number from 2004. The reasons for this probably lie with incomplete data returns from 3 units, together with variability of the population with the transfer of teenage patients to adult units.

Table 13.1 shows the prevalent population by gender and ethnicity together with the numbers who were under 18 years of age and 15 years of age on 1st April 2005. As in previous Reports, there are about 20 young people over the age of 18 years remaining in paediatric units. These patients are transferred between the age of 18 and 20 years. There are no patients over the age of 20 years in the current cohort. Reasons for delayed transfer include the management of specific paediatric co-morbidities and concerns over growth, development and education. The distribution of the population with regard to gender and ethnicity was unchanged from

Table 13.1: Prevalent patient population according to gender and ethnicity

	Patients	Male	Female	Ratio	% Total
Total	768	466	302	1.54:1	100.0
White	632	395	237	1.66:1	82.3
Asian	109	53	56	0.95:1	14.2
Black	14	9	5	1.80:1	1.8
Other	13	9	4	2.25:1	1.7
<18 years	748	456	292	1.56:1	97.4
<15 years	515	321	194	1.65:1	67.1

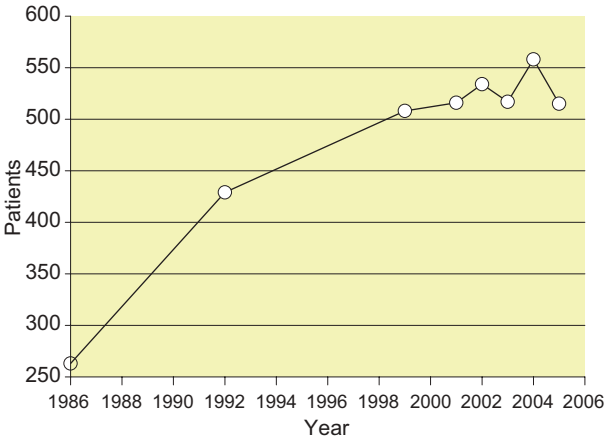

Figure 13.1: ERF patients below 15 years of age, by year of data collection

previous reports. There remains a predominance of males and just over 17% come from ethnic minority backgrounds.

Figure 13.1 shows the size of the population under the age of 15 years from 1986 to 2005. The apparent growth in this population seen in 2004 has not been maintained but this will be due to some missing data from units with incomplete submissions together with some variability year on year in presentation rates. The overall trend has been that of a slowing of the initially sharp increase in the population. This is supported by the data on incidence and prevalence presented below.

The age distribution of the population over a number of years is shown in Tables 13.2 and 13.3. The former gives the customary divisions by age and the latter shows the population divided into four year age bands for ease of comparison. Though there is year to year variability, the numbers have been fairly static of late, clearly showing a cessation of the rapid population growth seen after paediatric ERF treatment became available. Figure 13.2 shows the data in Table 13.3 graphically and clearly shows that over recent years there has been no significant change in the age distribution of the population.

Table 13.3: ERF population in 4 year age bands

	Patient population for the years of			
Age group (yrs)	2002	2003	2004	2005
0–3	49	39	41	36
4–7	94	103	112	108
8–11	185	176	173	152
12–15	294	291	297	321
16–19	171	164	179	151

Table 13.2: ERF population by age and year of data collection

	Patient population data for the years of							
Age group (yrs)	1986	1992	1999	2001	2002	2003	2004	2005
0–1		16	18	13	14	10	12	14
2–4		55	46	56	58	56	51	45
5–9		150	151	146	147	141	166	157
10–14		208	293	301	315	310	329	299
15–19			253	274	259	256	244	253
Total <15	263	429	508	516	534	517	558	515
Total <20			761	790	793	773	802	768

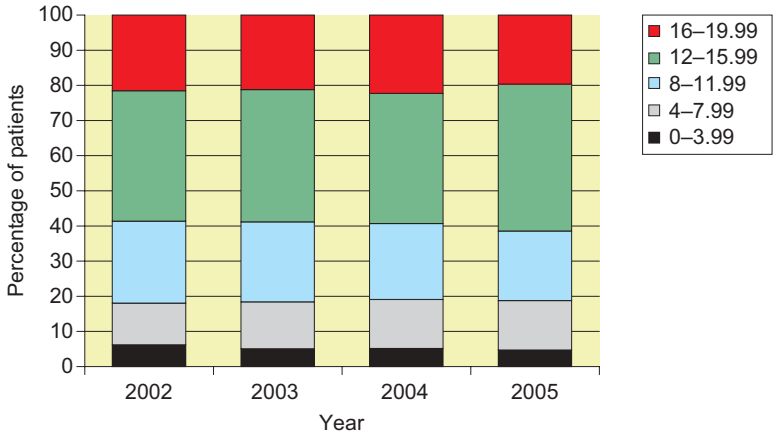

Figure 13.2: ERF population in 4 year age bands

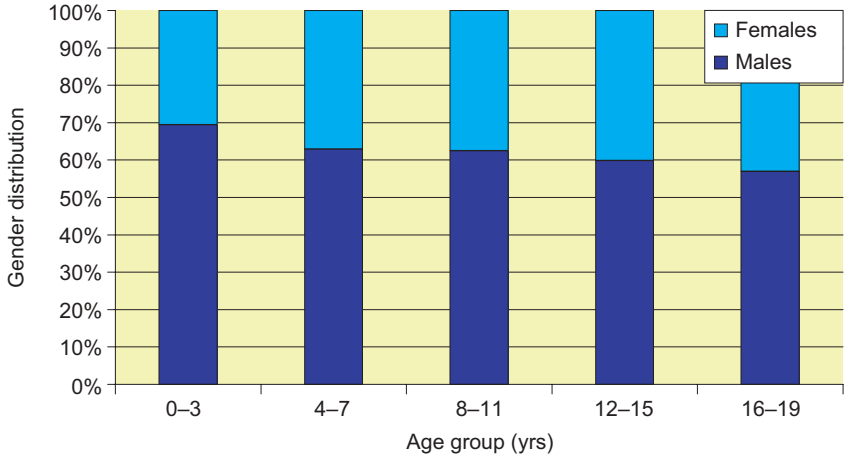

Figure 13.3: Gender distribution of the ERF population according to age

The gender distribution of the paediatric ERF population is shown in Figure 13.3. Throughout the age range, males predominate but there is a steady rise in the proportion of females in the population with increasing age.

Of the ethnic minority patients, the vast majority are of South Asian origin. The age and gender distributions of this cohort are somewhat different to that of the White population. This is secondary to the different causes of ERF in the South Asian community and is dealt with in detail below. Table 13.4 shows the age distribution of the population according to ethnicity. Although the difference in age distribution between the White and ethnic minority populations does not reach statistical significance the pattern is demonstrated in Figure 13.4.

The difference in gender distribution between the White and South Asian paediatric ERF populations is shown in Figure 13.5 which contrasts the proportion of the population in each age group who are male. In the under the age of 4 years group, 77% of White patients in this group are male. Thereafter, there is a fall in the proportion of males in the White population, with an increase in the proportion of males in the South Asian population, until in the young adults, both lie between 55 and 60%. There

Table 13.4: Age and ethnic distribution of the ERF prevalent population

Age group (yrs)	Ethnicity			
	White	South Asian	Black	Other
0–3	30	5	1	0
4–7	82	16	5	5
8–11	120	26	3	3
12–15	270	44	3	4
16–19	130	18	2	1
All <20	632	109	14	13

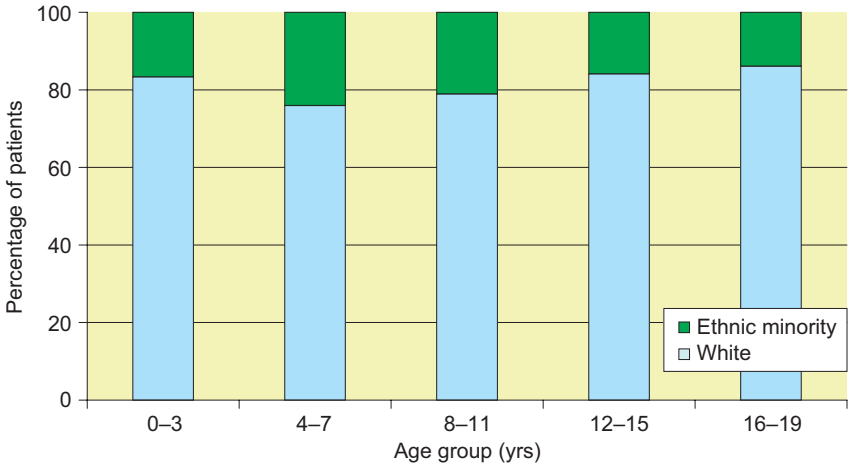

Figure 13.4: Age distribution of the White and Ethnic minority patients

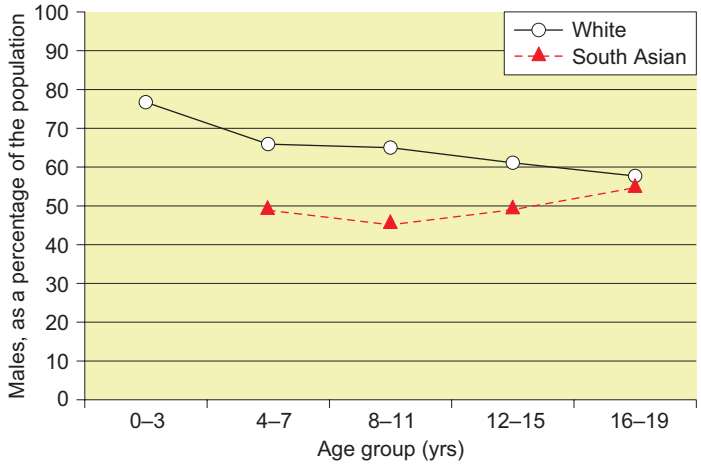

Figure 13.5: Gender distribution and ethnicity in the paediatric population

were only five Asian patients under age 4 so these have been removed from the graph.

Prevalence and take-on rate

Data on the UK population divided according to age and ethnic background was taken from the Office for National Statistics' Website (www.statistics.gov.uk). Data for this report is based upon current population estimates which themselves are extrapolated from the United Kingdom Census of 2001. Table 13.5 shows the prevalence of ERF per million childhood population for each age group. These figures have changed little since previous Reports[1–6] as one might expect from the stable population numbers. Figure 13.6 shows this graphically, clearly demonstrating the steady rise in prevalence with patient age until the fall in the over 16 year old

Table 13.5: Prevalence of ERF per million childhood population

	All patients		Males		Females	
Age group (yrs)	Patients	Prevalence	Patients	Prevalence	Patients	Prevalence
0–3	36	13.3	25	18.0	11	8.3
4–7	108	38.2	68	46.9	40	29.0
8–11	152	51.2	95	62.5	57	39.4
12–15	321	102.2	192	119.1	129	84.4
16–19	151	48.1	86	53.2	65	42.6
<15	515	47.4	321	57.6	194	36.6

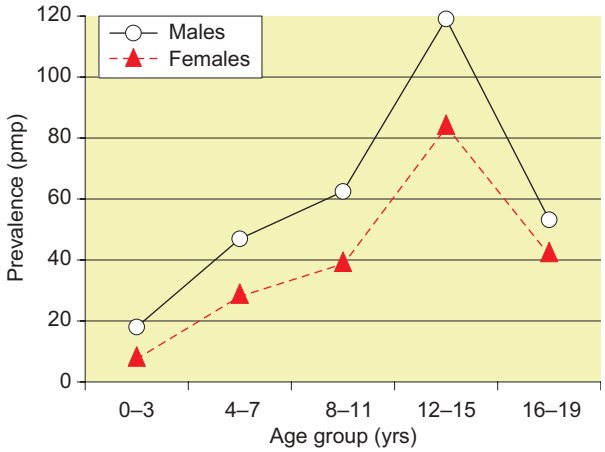

Figure 13.6: Prevalence of ERF according to gender

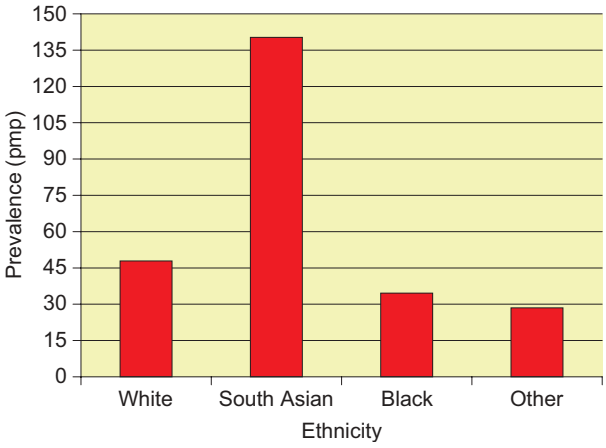

Figure 13.7: Prevalence of ERF according to ethnicity

group, secondary to transfer to adult units. The figures for prevalence of ERF in the UK are comparable with those presented in the USRDS and ANZDATA registries[7,8].

Whilst there is no mention of ethnicity in the most recent ANZDATA report the USRDS report does give an ethnic breakdown but not one which is specific to the paediatric age range. As the majority of the patients are adult and there are varying rates of glomerulonephritis, hypertensive and diabetic nephropathies amongst the different adult ethnic groups, it is impossible to extrapolate this published data to look at prevalence and ethnicity in children. As with previous reports from the UK paediatric registry the prevalence of ERF is much higher in the South Asian community, being almost three times that of the White population, whilst the prevalence of ERF in the Black population and those of other ethnic origins is a little below that of the White community. This is demonstrated in Figure 13.7. The reasons for this distribution lie in the varying causes of ERF with ethnicity and are discussed below.

The take on rates of patients starting RRT has been assessed looking at a 5 year period to even out the peaks and troughs seen with annual data collection when relatively small numbers are being analysed. This is demonstrated well by the undulant picture shown by the ANZDATA incidence chart. Looking at take on rate as a mean of consecutive 5 year periods, there is clearly little change in the incidence of ERF in children. Overall, the incidence of ERF in children in the UK is very similar to that of the Australian, New Zealand and US cohorts. These data are shown in 4 year age bands in Table 13.6 and graphically in Figure 13.8. There is a nadir of presentation of ERF in the 4 to 8 year old group following a peak in the first four years of life with the presentation of many children with obstructive uropathy and renal dysplasia. Following this there is a steady rise in incidence as the number of patients with glomerular diseases increases. As with the prevalence data, the take on rate of new patients with ERF in the South Asian community far outweighs that of the White community with an incidence per million childhood population 3.7 times that of the White population (Figure 13.9). This incidence figure will, over a number of years, lead to the proportion of the total population of children with

Table 13.6: Average 5 year incidence rate for patients with ERF per million childhood population

	All patients		Males		Females	
Age group (yrs)	Patients	Take on rate	Patients	Take on rate	Patients	Take on rate
0–3	22	8.0	13	9.4	9	6.7
4–7	15	5.2	8	5.2	7	5.1
8–11	24	8.0	13	8.4	11	7.6
12–15	35	11.3	19	11.8	16	10.7
<15	87	8.0	47	8.5	40	7.5

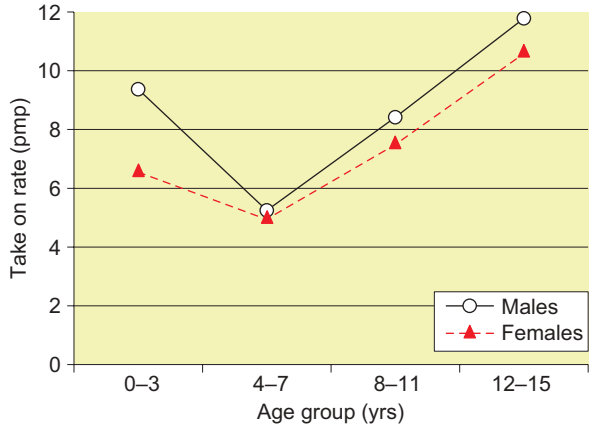

Figure 13.8: Average 5 year take on rate of children with ERF by gender

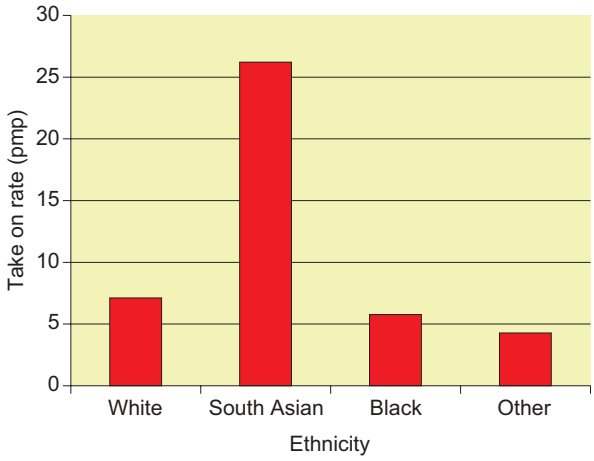

Figure 13.9: Average 5 year take on rate of children with ERF by ethnicity

ERF coming from the South Asian community rising still further. The distribution of the ethnic minority population (and consequently the ethnic minority children with ERF) around the UK is not evenly spread[6]. This has significant implications for resource management.

Causes of ERF in children

The causes of ERF have been analysed by looking at a total of 913 incident patients presenting with ERF before the age of 16 years, since the inception of the registry in 1996, for whom a primary diagnosis was stated. Diagnoses have been grouped into 12 bands. These are shown in Table 13.7 with a further breakdown of each of the groupings in Tables 13.8 to 13.17. Renal dysplasia remains the single most common diagnostic group comprising almost a quarter of the total cohort. There is a male predominance in patients with renal dysplasia, and this together with the male contingent with obstructive uropathy from posterior urethral valves, accounts for the overall gender distribution of the paediatric ERF population. The gender distribution of each diagnostic group is shown in Figure 13.10. Although there is no explanation for this, a high incidence of renal dysplasia in males has not only been noted in the UK registry reports but also in the NAPRTCS report[9]. Glomerular disease follows closely behind renal dysplasia, accounting for 22% of patients. Obstructive uropathy is the third most common cause accounting for 15%.

The nature and distribution of the diseases causing ERF in childhood have not changed significantly over the years that reports have been generated by the Registry. However, this will be due to the fact that a complete and

Table 13.7: ERF diagnostic grouping for 913 patients presenting after 1st April 1996

Diagnostic group	Patients	% of total	Males	Females	Ratio
Dysplasia	221	24.2	138	83	1.66:1
Glomerular diseases	205	22.5	91	114	0.80:1
Obstructive uropathy	136	14.9	121	15	8.06:1
Tubulo-interstitial diseases	73	8.0	36	37	0.97:1
Reflux nephropathy	69	7.6	34	35	0.97:1
Congenital nephrotic syndrome	46	5.0	18	28	0.64:1
Metabolic diseases	44	4.8	25	19	1.32:1
Renovascular problems	34	3.7	18	16	1.13:1
ERF of uncertain aetiology	29	3.2	12	17	0.71:1
Polycystic kidney disease	27	3.0	9	18	0.50:1
ERF from drug nephrotoxicity	19	2.1	13	6	2.17:1
Malignancy & associated disease	10	1.1	5	5	1.00:1

Table 13.8: Diagnoses for patients with renal dysplasia

Diagnoses in renal dysplasia group	Patients	Males	Females	Ratio
Renal dysplasia	184	114	70	1.63:1
Multicystic dysplastic kidneys	11	5	6	0.83:1
Prune belly syndrome	10	10	0	
Renal hypoplasia	8	3	5	0.75:1
Branchio-oto-renal syndrome	3	3	0	
Lawrence Moon Bardet Biedl syndrome	3	1	2	0.50:1
Megacystis megaureter	2	2	0	

Table 13.9: Diagnoses for patients with glomerular disease

Diagnoses in glomerular diseases group	Patients	Males	Females	Ratio
Primary focal segmental glomerulosclerosis	87	40	47	0.85:1
Diarrhoea positive HUS	18	8	10	0.80:1
Henoch Schoenlein nephritis	14	5	9	0.56:1
Diarrhoea negative HUS	12	3	9	0.33:1
GN (unspecified)	10	6	4	1.50:1
Alport's syndrome	9	8	1	8.00:1
IgA nephropathy	9	5	4	1.25:1
Mesangio-capillary GN type 1	9	4	5	0.80:1
Mesangio-capillary GN type 2	6	2	4	0.50:1
Crescentic GN	8	4	4	1.00:1
Proliferative GN	6	2	4	0.50:1
Systemic lupus erythematosis	6	1	5	0.20:1
Anti GBM disease	3	0	3	
Microscopic polyarteritis nodosa	3	1	2	0.50:1
Wegner's granulomatosis	3	2	1	2.00:1
Macroscopic polyarteritis nodosa	1	0	1	
Vasculitis (unspecified)	1	0	1	

Table 13.10: Diagnoses for patients with obstructive uropathy

Diagnoses in obstructive uropathy group	Patients	Males	Females	Ratio
Posterior urethral valves	103	103	0	
Neuropathic bladder	13	3	10	0.30:1
Bladder outlet obstruction (Not PUV)	11	9	2	4.50:1
Congenital obstructive uropathy (Not BOO)	7	4	3	1.25:1
Acquired obstructive uropathy	2	2	0	

(PUV = posterior urethral valves, BOO = bladder outlet obstruction)

Table 13.11: Diagnoses for patients with tubulo-interstitial disease

Diagnoses in tubulo-interstitial group	Patients	Males	Females	Ratio
Nephronophthisis	59	28	31	0.90:1
Primary interstitial nephritis	9	5	4	1.25:1
Bartter's syndrome	2	1	1	1.00:1
Nephrocalcinosis	1	0	1	
Renal tubular acidosis	1	1	0	
Tubular disorders (other)	1	1	0	

Table 13.12: Diagnoses for patients with congenital nephrotic syndrome

Diagnoses in congenital nephrotic syndrome group	Patients	Males	Females	Ratio
CNS unspecified	21	5	16	0.31:1
Finnish type	17	8	9	0.89:1
Diffuse mesangial sclerosis	5	4	1	4.00:1
Focal segmental glomerulosclerosis	3	1	2	0.50:1

Table 13.13: Diagnoses for patients with metabolic diseases

Diagnoses in metabolic diseases group	Patients	Males	Females	Ratio
Cystinosis	34	19	15	1.27:1
Primary hyperoxaluria type I	5	3	2	1.50:1
Mitochondrial cytopathy	4	2	2	1.00:1
Metabolic disease (other)	1	1	0	

Table 13.14: Diagnoses for patients with renovascular disease

Diagnoses in renovascular disease group	Patients	Males	Females	Ratio
Cortical necrosis	22	10	12	0.83:1
Renal vein thrombosis	8	6	2	3.00:1
Renal artery stenosis	2	1	1	1.00:1
Renal trauma	2	1	1	1.00:1

Table 13.15: Diagnoses for patients with polycystic kidney disease

Diagnoses in polycystic kidney disease group	Patients	Males	Females	Ratio
Recessive polycystic kidney disease	20	6	14	0.43:1
Polycystic kidney disease (other)	5	2	3	0.67:1
Dominant polycystic kidney disease	1	1	0	
Tuberous sclerosis with polycystic kidney disease	1	0	1	

Table 13.16: Diagnoses for patients with ERF from drug nephrotoxicity

ERF from drug nephrotoxicity group	Patients	Males	Females	Ratio
Calcineurin inhibitor nephrotoxicity	14	11	3	3.67:1
Cytotoxic drug nephrotoxicity	5	2	3	0.67:1

Table 13.17: Diagnoses for patients with malignant disease

Diagnoses in malignant disease group	Patients	Males	Females	Ratio
Wilms' tumour	7	3	4	0.75:1
Wilms' nephropathy	3	2	1	2.00:1

expanding cohort has been used to look at this distribution. Certainly the information provided by ANZDATA suggests a similar distribution of causes if one excludes the 15 to 20 year age group, which appears to be a complete cohort in ANZDATA and therefore dramatically expands the band of patients with glomerulonephritides. The USRDS data available does not give a specific diagnostic breakdown for children. The NAPRTCS report is more difficult

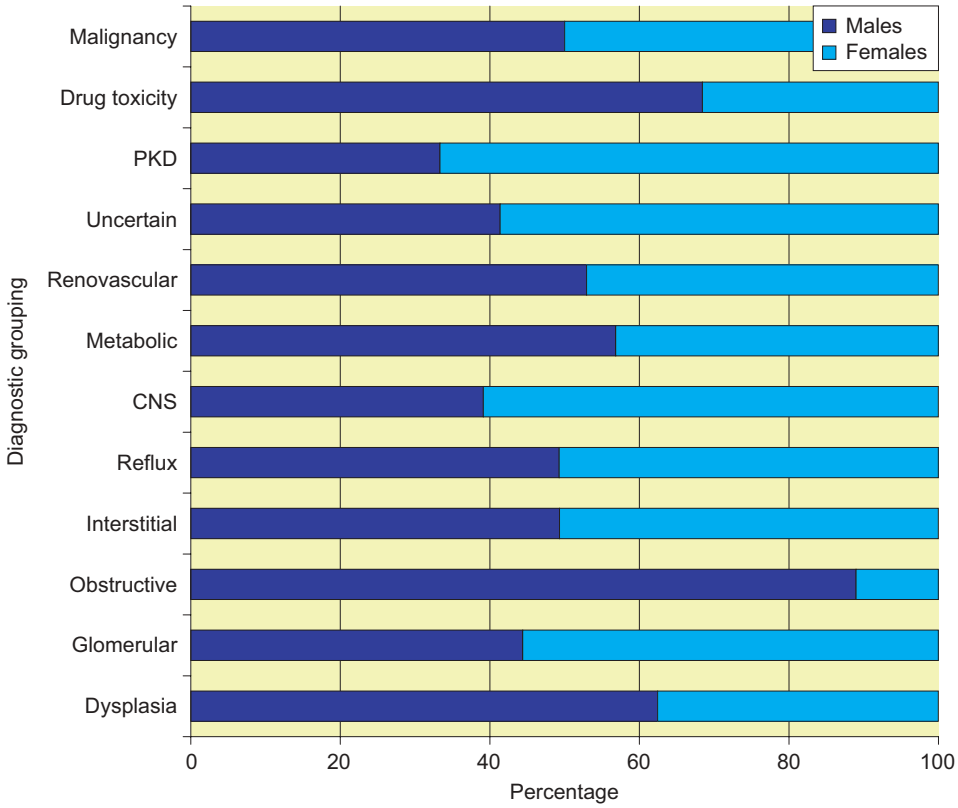

Figure 13.10: Gender distribution of the ERF population according to diagnostic group

to interpret as the analysis of transplant and dialysis patients is separate. Certainly it appears that for White and Hispanic patients, renal dysplasia leads in conjunction with obstructive uropathy. Unlike the UK data, glomerular diseases causing ERF appear less frequent in this population. This however, is offset by the high incidence of glomerular diseases causing ERF in the Black population. Differences in the patterns of primary pathology with ethnicity in the UK population are dealt with below.

To investigate whether there has been any change in the pattern of primary pathology causing ERF in children over the period the Registry has been collecting data, the distribution of diagnoses have been compared within the 12 main classifications in those patients presenting between 1996 (when data collection began) and 1999, with those patients presenting between 2002 and 2005. These data are shown in Table 13.18. There is no significant difference in the patterns of disease. The incidence of

Table 13.18: Comparison of diagnostic distributions 1996–1999 and 2002–2005

	Percentage of patients presenting	
Diagnostic group	**1996–1999**	**2002–2005**
Dysplasia	25.2	26.1
Glomerular diseases	21.1	21.2
Obstructive uropathy	17.1	13.8
Tubulo-interstitial diseases	6.7	9.9
Reflux nephropathy	9.4	5.7
Congenital nephrotic syndrome	6.0	4.2
Metabolic diseases	4.0	3.5
Renovascular problems	3.7	3.5
ERF of uncertain aetiology	2.3	5.3
Polycystic kidney disease	2.7	3.5
ERF from drug nephrotoxicity	1.0	1.4
Malignancy & associated disease	0.7	1.8

obstructive uropathy has fallen slightly and time will tell whether this is an ongoing trend. Reflux nephropathy has fallen and there has been a parallel rise in the incidence of ERF of uncertain aetiology. Knowing the difficulty in categorising patients who present with small kidneys, either in or near ERF, it is possible that this simply represents variability in classification. The incidence of tubulo-interstitial diseases has risen. Again, only time will tell whether this is a true trend, however, it is something that may be expected given the rising South Asian population and the increased frequency of these pathologies in this ethnic group.

As alluded to above and published in previous reports from the Registry, there is a significant difference in the pattern of diseases causing ERF in different ethnic groups. This is shown in Table 13.19. Whilst for the White population renal dysplasia predominates followed closely by glomerular diseases. In the South Asian population glomerular diseases predominate with a lower incidence of renal dysplasia. Tubulo-interstitial disorders, metabolic diseases and congenital nephrotic syndrome are much more common in the South Asian community. The overall difference in the distribution of diseases between the White and South Asian populations is highly significant ($\chi^2 = 40.2,\ p < 0.0001$). Interpretation of the distribution of diseases in the Black population and those from other ethnic backgrounds is more difficult because of the small numbers. Black patients with glomerular diseases contribute over 50% of the cohort and renal dysplasia is much less common with only occasional cases of other disorders appearing. Certainly data from NAPRTCS would suggest that this is not an unrepresentative pattern of disease.

Much of the difference between the patterns of disease in the South Asian patients compared to the White cohort can be explained by the high incidence of autosomal recessive inherited disorders in this population. Table 13.20 shows the pattern of inheritance of the primary cause of ERF in 913 patients presenting after 1996 and starting ERF before the age of 16 years for whom both details of primary diagnosis and ethnicity were available. Overall, 190 patients (20.8%) had diseases with a clear inheritance link, showing the major contribution of genetic

Table 13.19: Ethnic distribution of ERF diagnostic groups

Diagnostic group	White No (%)	South Asian No (%)	Black No (%)	Other No (%)
Dysplasia	193 (26.2)	23 (16.4)	5 (25.0)	0 (0.0)
Glomerular diseases	161 (21.8)	29 (20.7)	11 (55.0)	4 (26.7)
Obstructive uropathy	117 (15.9)	17 (12.1)	0 (0.0)	2 (13.3)
Tubulo-interstitial diseases	52 (7.1)	17 (12.1)	0 (0.0)	4 (26.7)
Reflux nephropathy	60 (8.1)	6 (4.3)	1 (5.0)	2 (13.3)
Congenital nephrotic syndrome	30 (4.1)	16 (11.4)	0 (0.0)	0 (0.0)
Metabolic diseases	31 (4.2)	13 (9.3)	0 (0.0)	0 (0.0)
Renovascular problems	31 (4.2)	2 (1.4)	1 (5.0)	0 (0.0)
ERF of uncertain aetiology	17 (2.3)	9 (6.4)	1 (5.0)	2 (13.3)
Polycystic kidney disease	20 (2.7)	5 (3.6)	1 (5.0)	1 (6.7)
ERF from drug nephrotoxicity	17 (2.3)	2 (1.4)	0 (0.0)	0 (0.0)
Malignant disease	9 (1.2)	1 (0.7)	0 (0.0)	0 (0.0)

Table 13.20: Ethnic distribution of inherited diseases

Disease inheritance	White	South Asian	Black	Other
Autosomal recessive	120	47	1	4
Autosomal dominant	5	0	0	0
Sex linked	6	2	1	0
Mitochondrial disease	3	1	0	0
Not directly inherited	604	90	18	11

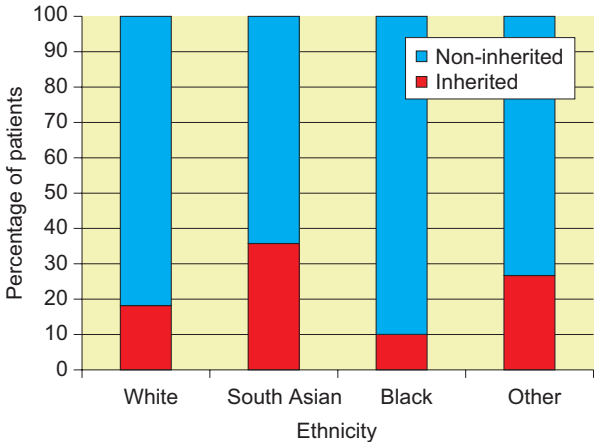

Figure 13.11: Percentage of inherited disease, by ethnicity

problems to childhood ERF. Of these, the vast majority (90.5%) were autosomal recessive diseases with just a small number of dominant, sex linked and mitochondrial disorders. These of course do not include patients with diseases that probably do have a strong genetic component that has not yet been clearly defined, such as isolated renal dysplasia. The proportion of each ethnic group with inherited disease as a cause of ERF is shown in Figure 13.11. This clearly shows the excess of inherited disease both in those of South Asian origin and in those of "Other" origin. Consanguineous marriage is more common in both of these groups compared to the White population. Although the small numbers of patients in the "Other" group make valid statistical analysis difficult, the increased proportion of inherited disease in the South Asian group compared to the White population is very significant ($p < 0.0001$, Fisher's exact test).

The age distribution of the paediatric ERF population is determined by both the survival of patients and the age of presentation with ERF. This in turn is often dependent upon the aetiology of ERF. The effect of diagnosis upon the population age distribution is shown in Figures 13.12, 13.13 and 13.14 below. For each of these figures the (a) pane shows the percentage of patients in a designated diagnostic group presenting in each age group, whilst the (b) pane shows the percentage of patients in each age group belonging to that diagnostic group. Thus, for patients with renal dysplasia, 32% present with ERF in the first 4 years of life and 32% present between the ages of 12 and 16 years whilst the remaining third present in the intervening 8 years.

The proportion of patients with renal dysplasia as a cause of ERF in each age group, account for 34% of those in the first four years of life but only 20% of those between the ages of 12 and 16 years because other causes of ERF have become more frequent in this latter age group. The pattern for obstructive uropathy is virtually identical to that for renal dysplasia. As with renal dysplasia, virtually all patients will have been born with their problem. The distributions of both these groups show the combined effect of the severity of the initial problem and the subsequent rate of decline of GFR with the stresses of growth and hyper-perfusion glomerulopathy. Reflux nephropathy rarely causes ERF

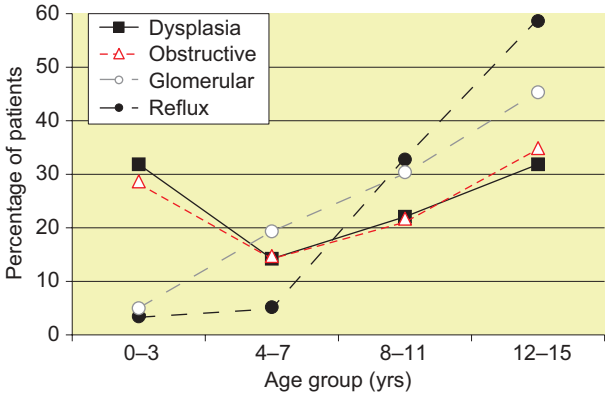

Figure 13.12a: Percentage of incident patients with renal dysplasia, obstructive uropathy, glomerular diseases and reflux nephropathy presenting in each age band

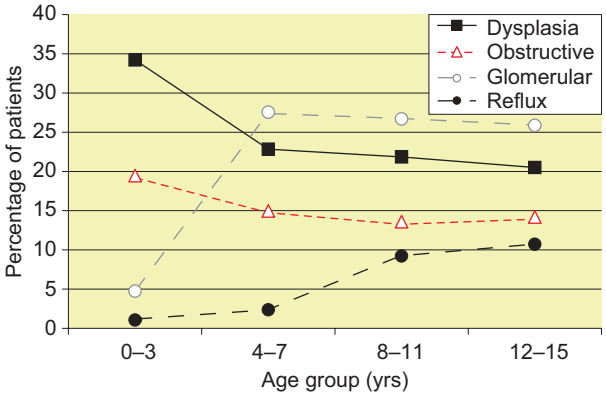

Figure 13.12b: Percentage of incident patients by age with renal dysplasia obstructive uropathy, glomerular disease or reflux nephropathy as the cause

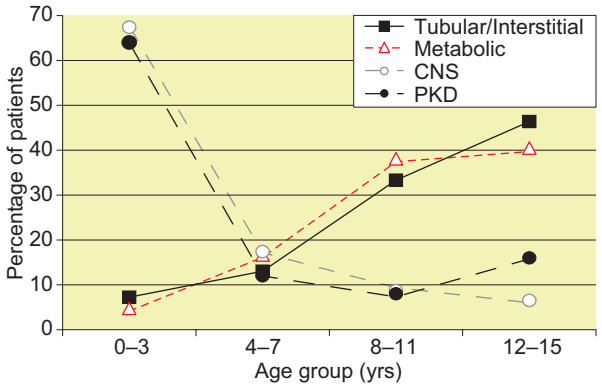

Figure 13.13a: Percentage of incident patients with tubulo-interstitial diseases, metabolic diseases, congenital nephrosis and polycystic disease by age band

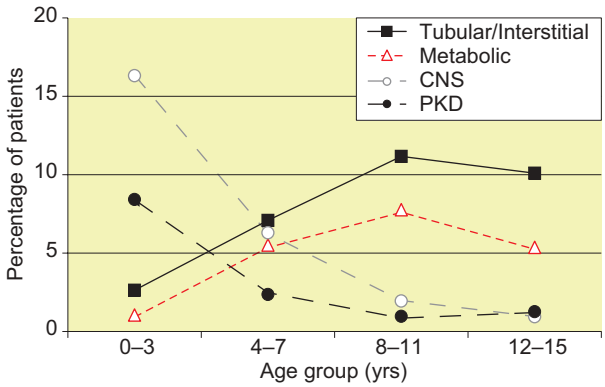

Figure 13.13b: Percentage of incident patients by age band with tubulo-interstitial or metabolic disease, congenital nephrosis or polycystic kidneys as the cause

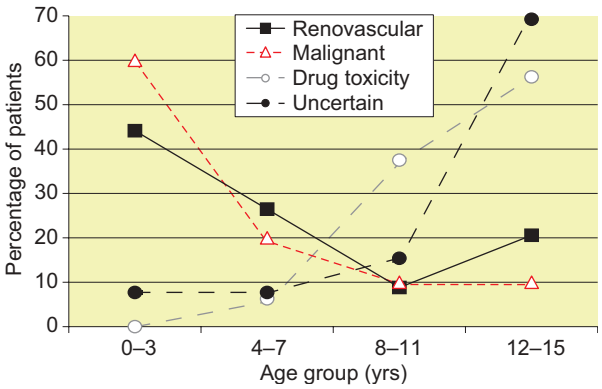

Figure 13.14a: Percentage of patients with renovascular diseases, malignant diseases, drug nephrotoxicity and uncertain aetiology presenting in each age band

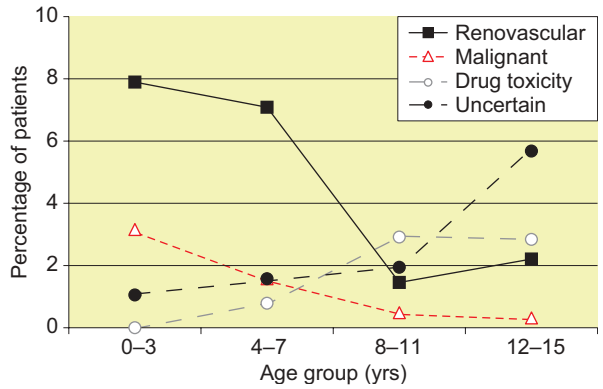

Figure 13.14b: Percentage of incident patients in each age band with renovascular or malignant disease, drug nephrotoxicity as the cause or uncertain aetiology

in the first 8 years of life and just over one third present between 8 and 12 years of life with almost 60% entering ERF between the age of 12 and 16 years. Even so reflux nephropathy only accounts for 11% of patients between the ages of 12 and 16 years with ERF. The addition of patients with renal dysplasia and reflux nephropathy together leads to a block accounting for a little under or over 30% of patients in each age group. In both conditions there is a high incidence of vesico-ureteric reflux and in both conditions there is likely to be congenital renal dysplasia. In view of the reduced frequency of urinary tract infections and clinical pyelonephritis in the older age groups, hyper-perfusion glomerulopathy is likely to play a major part in both conditions in determining the speed and timing of the decline into ERF. It is most likely therefore that reflux nephropathy and renal dysplasia share common origins. Glomerular diseases are rare in early childhood and 75% of children with these diseases will enter ERF beyond the age of 8 years. As glomerular diseases are the most common cause of ERF in Black children this explains the age distribution of this cohort. Whilst this is a small group within the UK, this observation is important with regard to the development of services in developing countries. Those with a predominantly Black population where consanguineous marriage is rare can expect their paediatric ERF population to come from the older childhood groups. This will limit the potential size of the paediatric unit, particularly if transfer to adult services is at a much younger age than is the norm in the UK and Europe.

The same data for the main disease groups with inherited diseases are shown in Figures 13.13(a) and (b). As one might expect, diseases

such as congenital nephrotic syndrome and polycystic kidney disease peak in the first 4 years of life, whilst the tubular and metabolic disorders peak later in childhood.

The final four groups are shown in Figures 13.14(a) and (b). Numerically these very different conditions account for only a small percentage of patients, both overall and in any one age band.

Current treatment of paediatric ESRF patients

Of the 768 patients, data on modality on the 1st April 2005 were available for 684 (89%). The distribution of modalities has changed little since previous reports with 76% of patients having a functioning allograft and for the remainder, peritoneal dialysis being a more common treatment than haemodialysis. For those with allografts, over two thirds have cadaveric grafts with 21% of the total population (28% of those with allografts) having a graft from a living donor. For those on peritoneal dialysis the vast majority are receiving automated PD with few centres using CAPD (Figure 13.15).

The proportion of engrafted patients, whose graft has come from a living (usually related) donor, rather than a cadaveric donor, is slowly but steadily increasing (Figure 13.16). This, in the face of a stable ERF population with a stable proportion whose management is with an allograft, highlights the shortage of suitable

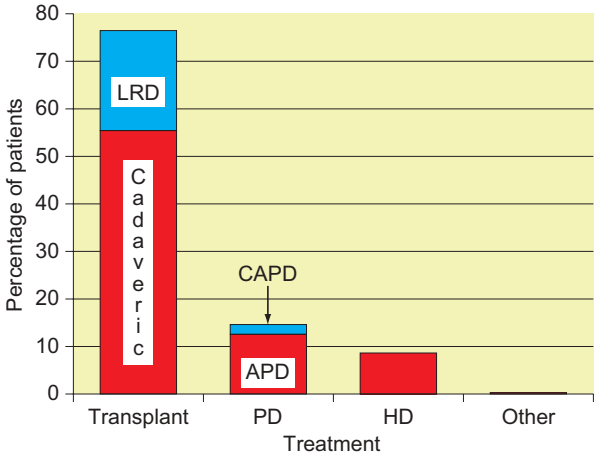

Figure 13.15: Distribution of RRT modality in paediatric patients on 1st April 2005

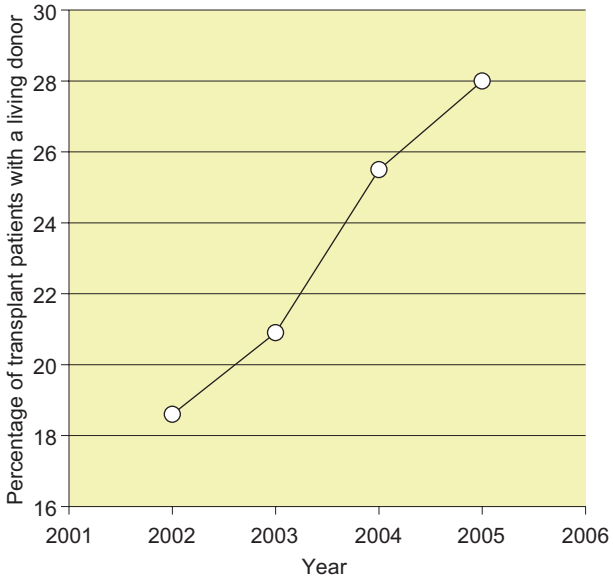

Figure 13.16: Percentage of patients with renal allografts whose graft came from a living rather than cadaveric donor

cadaveric organs, the need to use living donation to maintain the proportion of engrafted patients and the change in medical practice in the UK with a greater emphasis being placed upon the benefits of living donation.

The distribution of RRT modalities divided according to ethnic origin is shown in Figure 13.17. Whilst 80% of White patients have a functioning allograft only 63% of South Asian patients and 42% of Black patients have one. These populations therefore have proportionately larger numbers on dialysis. For all groups, peritoneal dialysis is the most frequent dialysis modality employed. The difference between ethnic groups in the distribution of treatment modalities is significant ($p < 0.0001$, $\chi^2 = 22.2$). Part of the explanation for the lower transplantation rates in ethnic minority groups is the lower rate of living donation. Certainly the proportion of South Asian patients with an allograft from a living donor is significantly lower than the proportion of White patients with one ($p = 0.0466$). This difference loses its significance if all ethnic minorities are compared to the White population. The ethnic minority population have a different distribution of tissue types and blood groups to the White population who form the vast majority of the donor pool. In these circumstances it is inevitable that there will be fewer offers of well matched cadaveric allografts for ethnic minority patients than White patients. In these

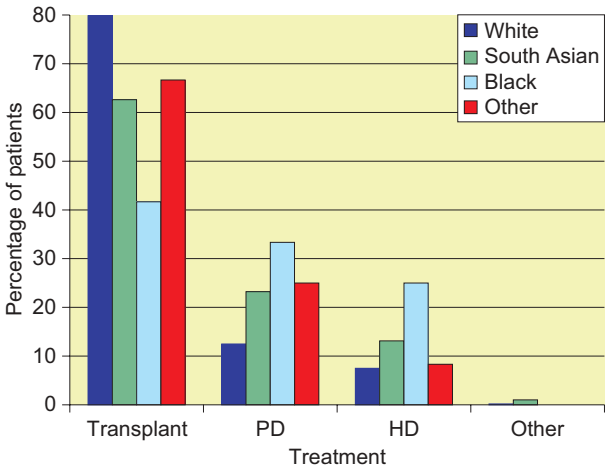

Figure 13.17: Distribution of RRT modalities according to ethnicity

circumstances only an increase in the number of live donors in the ethnic minority groups will allow the proportion with a functioning allograft in these groups to rise to that of the White population.

An important aspect of ERF management is treatment modality change with time. Figure 13.18 shows the distribution of patients according to whether or not their treatment modality had changed since the previous data collection in 2004. Clearly for the majority there was no change. Just under 11% of the cohort had had a change in treatment modality during the year whilst 77% did not. The remainder were new patients with no previous annual record.

For those who had had no change over the previous year, the vast majority (84%) had a functioning allograft. Nine percent were maintained on peritoneal dialysis and 7% on haemodialysis (Figure 13.19).

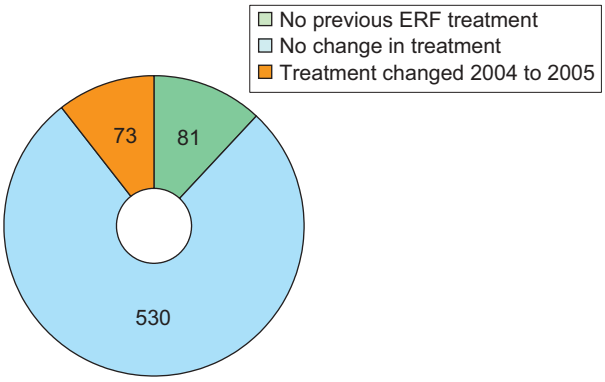

Figure 13.18: ERF modality changes from 2004 to 2005

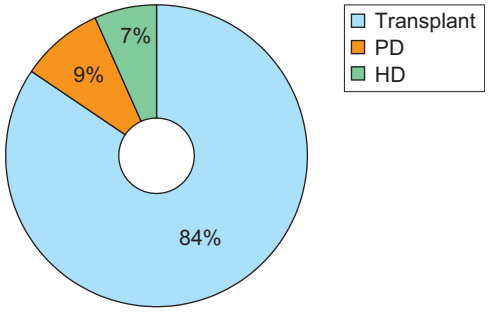

Figure 13.19: Distribution of modalities in those unchanged from 2004 to 2005

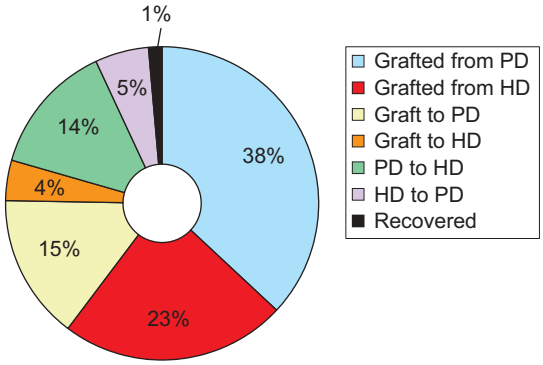

Figure 13.20: ERF modality changes from 2004 to 2005

For those who changed treatment modality over the course of the year the reason in most was because they were transplanted. 61% of this cohort received an allograft and the distribution of these between patients on peritoneal and haemodialysis was appropriate for the numbers on each modality. 19% lost grafts and started dialysis, 75% of these started peritoneal dialysis. 14% of the cohort moved from peritoneal to haemodialysis whilst only 5% of the cohort moved in the opposite direction. One patient recovered enough renal function to stop dialysis (Figure 13.20).

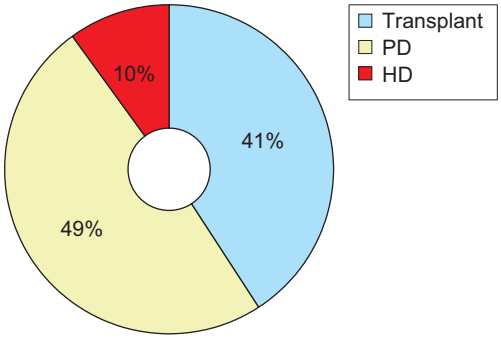

Figure 13.21: ERF modality in April 2005 for those starting after April 2004

The distribution of RRT modalities in April 2005 of the 81 patients starting ERF management during that year is shown in Figure 13.21. As expected, the single largest group accounting for 49% of the cohort were those on peritoneal dialysis. Just 10% were on haemodialysis whilst 41% had a functioning allograft. A proportion of this latter group would have had pre-emptive grafts whilst others will have received an allograft during the first year as a second treatment modality.

Conclusions

The incidence and prevalence of ERF in children in the UK has changed little over recent years. Similarly, analyses of the causes of ERF in childhood shows little change over the past decade. After an initial steep growth following the commencement of RRT services for children in the UK, the size of the paediatric ERF population is now relatively static. As with most paediatric and adult RRT studies there is a male predominance. In the paediatric population this is secondary to both the large proportion of patients with posterior urethral valves as a cause of ERF and the predominance of males with renal dysplasia as a cause of ERF.

The striking data is the high incidence and prevalence of ERF in the South Asian community in the UK. This is in part due to a high incidence of autosomal recessive inherited diseases causing ERF in this population. This could potentially lead not only to further growth of the ERF population over the next two decades, but also to a change in the pattern of disease causing ERF in the UK childhood population in addition to equalisation of the gender distribution of ERF.

The commonest RRT modality for children with ERF is transplantation, with 76% of the population having a functioning allograft. The paucity of cadaveric organs has led to an increase in the proportion of these patients with an allograft from a living donor. Living donation is less frequent in the South Asian community who by virtue of their tissue types and that of the cadaveric donor pool, are also less likely to receive a graft. This could lead to a growing number of patients on dialysis as the ethnic minority population grows. For those on dialysis the majority are managed with peritoneal dialysis and the vast majority of these patients receive APD rather than CAPD.

References

1. UK Renal Registry Report 1999. Chapter 15. UK Renal Registry, Bristol, UK. Editors Ansell D, Feest T.
2. UK Renal Registry Report 2000. Chapter 15. UK Renal Registry, Bristol, UK. Editors Ansell D, Feest T.
3. UK Renal Registry Report 2002. Chapter 16. UK Renal Registry, Bristol, UK. Editors Ansell D, Feest T.
4. UK Renal Registry Report 2003. Chapter 14. UK Renal Registry, Bristol, UK. Editors Ansell D, Feest T.
5. UK Renal Registry Report 2004. Chapter 13. UK Renal Registry, Bristol, UK. Editors Ansell D, Feest T.
6. UK Renal Registry Report 2005. Chapter 18. UK Renal Registry, Bristol, UK. Editors Ansell D, Feest T, Williams A, Winearls C.
7. U.S. Renal Data System, USRDS 2006 Annual Data Report: Atlas of End-stage Renal Disease in the United States, National institutes of Health, National Institute of Diabetes and Digestive and Kidney Diseases, Bethesda, MD, 2006.
8. Craig J, Henning P, McTaggart S, McDonald S, Chang S and Excell L. Paediatric Report; 143–153; ANZDATA Registry Report 2006, Australia and New Zealand Dialysis and Transplant Registry, Adelaide, South Australia.
9. NAPRTCS 2006 Annual Report. North American Pediatric Renal Trials and Collaborative Studies. http://web.emmes.com/ped/annlrept/annlrept2006.pdf

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