Chapter 16: Report from the Paediatric Renal Registry

Summary

- Paediatric data collection has continued across the UK from the 13 regional tertiary paediatric nephrology units. This has provided information on an almost complete cohort of children under the age of 15 years on renal replacement therapy. Although the incidence of established renal failure in children is fairly constant, there has been a significant increase in the prevalence over the past 10 years. Established renal failure in children therefore continues to be a cumulative service: this has significant implications for the need for resources.

- There is a significant overrepresentation of patients from the Asian subcontinent on renal replacement therapy, and these patients are geographically concentrated in a few units. This has implications for workforce and resource planning in these areas.

- Established renal failure presents at all ages in childhood, with a slowly increasing incidence with age. Males significantly outnumber females in early childhood, but by the teenage years, more females are presenting than males.

- Renal dysplasia and obstructive uropathy remain the most common causes of renal replacement therapy in childhood. Patients with these problems present with established renal failure at all ages throughout childhood. Reflux nephropathy becomes more frequent in the older groups as renal dysplasia reduces, and the overall incidence of renal dysplasia plus reflux nephropathy is constant, suggesting that they are different ends of the spectrum of one condition.

- The majority of paediatric renal replacement therapy patients have a functioning transplant, but the rate of cadaveric transplantation is falling. This is, however, balanced by an associated trend of an increase in live related transplants, allowing a constant transplant rate overall.

- The data in this report relate to the under-15-year-old age group, which represents only 64% of the total workload of the paediatric nephrology units. Many children with chronic conditions do not transfer to the adult services until the age of 18.

Introduction

In the first report from the paediatric arm of the Renal Registry in 1999, the incidence and prevalence of established renal failure (ERF) in children across the UK was reported. At the time the report was compiled, the Registry data collection mechanism was in its infancy, and the data represented a cross-sectional analysis of the population as it appeared at that time. Since this first report, data collection has continued, and the Registry is now in a position to

look at sequential data sets for a 5 year period. This report looks at the paediatric population as it stood on the 1 April 2001 and centres around an analysis of the incidence and prevalence of renal replacement therapy (RRT) in children in the UK, taking particular account of the breakdown according to diagnosis, age of presentation, ethnic origin and current management.

The 1999 report included the paediatric RRT population for both the UK and Eire. Data for Eire were not available for this report, and appropriate adjustments to the 1999 report figures have therefore been made to allow for meaningful comparisons.

The paediatric RRT population

As discussed in previous paediatric reports, any assessment of the size of the paediatric RRT population is hampered by the variable referral and treatment of teenage patients between paediatric and adult units. The Renal Registry is rapidly expanding towards 100% capture of data from adult units across the UK, and with this a complete analysis of the split of treatment in teenage patients will become possible. Until then, complete assessment is possible only for the population under the age of 15 years, who virtually are all looked after in paediatric units. Patients on RRT who are under the age of 15 but are not being seen within a designated paediatric renal unit will be the small number with multiorgan problems receiving combined transplants in specialised units and 11 patients in Scotland who did not, at that time, attend the one designated unit there.

The total number of patients being cared for in the 13 paediatric nephrology centres providing RRT treatment on 1 April 2001 was 800, with a gender ratio of 1.54:1 males to females. Of these 800 patients, 753 were under the age of 18 years. This represents a growth in the prevalent patient population of 3.8% from the 725 patients under the age of 18 years in 1999. Table 16.1 gives a breakdown of this patient group by age and gender. As might be expected, through the survival of young patients on RRT and the continuing uptake of new patients on RRT throughout the age spectrum, there is an increasing number of patients in each age group until a fall-off occurs from the transfer to adult units. This is shown graphically in Figure 16.1, where the population has been split into equal 4 year age bands to allow ease of comparison between these groups.

Age group (years)	Males	Females	Total (%)
0–1.9	9	4	13 (1.6)
2–4.9	38	18	56 (7.0)
5–9.9	94	52	146 (18.3)
10–14.9	183	118	301 (37.6)
15–17.9	135	102	237 (29.6)
≥18	28	19	47 (5.9)
Total	488	316	800

Table 16.1: Age and gender distribution of the paediatric RRT population

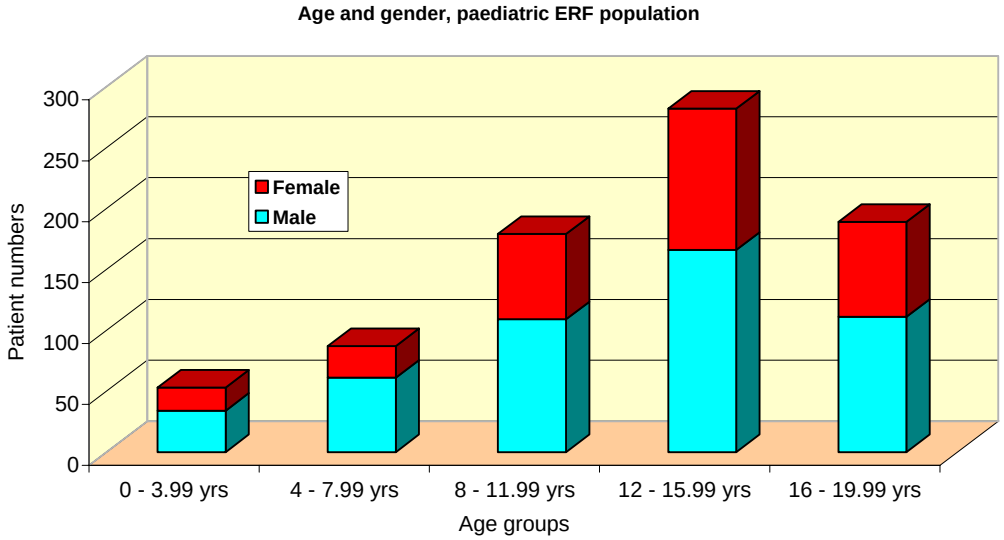

Figure 16.1: Age and gender distribution of the paediatric RRT population

Table 16.2 shows the change in prevalent patient population for those under the age of 15 years. Data from the Paediatric Registry for 1999 and 2001 have been compared with those published in the 1995 British Association of Paediatric Nephrology (BAPN) document *Report on the Provision of Services in the United Kingdom for Children and Adolescents with Renal Disease*. Between 1992 and 2001, there has been a 20% increase in the number of patients under the age of 15 years receiving ERF management. Some of this increase may be the result of incomplete data collection for the 1995 report. Although patient prevalence has continued to increase, the rate of increase appears recently to have slowed.

Age group (years)	1986	1992	1999	2001
0–1.99		16	18	13
2–4.99		55	46	56
5–9.99		150	151	146
10–14.99		208	293	301
Total	263	429	508	516
Per annum increase in prevalence		27.67	11.29	4.00

Table 16.2: Changes in the under-15-year-old prevalent RRT population, by year

Figures collected for 1992 and published in the 1995 working party report were based upon a total childhood population under the age of 15 years of 11.05 million in the UK at that time. Based on this, the prevalence of ERF in children under the age of 15 years in the UK in 1992 was 38.8 per million and the incident rate of new ERF patients 9.7 per million. For this report, population statistics were taken from the UK Government website (www.statistics.gov.uk), which quoted figures from the Office for National Statistics and General Register Office for Scotland. Figures given were for the projected population at mid-2000. Table 16.3 shows a breakdown of the population according to age and gender, and also shows the prevalence of RRT in children in each of these groups. As expected, prevalence rises with age. Within all age groups, there is an excess of males, which is similar to what is found in the adult population. Assuming the projected figures for childhood population are correct, there has been a significant increase in prevalence, to 47.5 per million, since 1992, matching the increase in current patient population.

Age group (years)	UK population (millions)			ERF population			Prevalence (pmp)		
	Males	Females	Total	Males	Females	Total	Males	Females	Total
0–4.99	1.7711	1.6854	3.4565	46	23	69	25.97	13.65	20.25
5–9.99	1.9140	1.8208	3.7348	94	52	146	49.11	28.56	39.36
10–14.99	1.9246	1.8250	3.7496	183	118	301	95.08	64.66	80.81
All <15	5.6097	5.3312	10.9409	323	193	516	57.76	36.76	47.53
UK population	28.6270	29.4309	58.0579				11.32	6.66	8.96

Table 16.3: Prevalence of ERF in children under 15 years of age in the UK
pmp, per million population.

Table 16.4 shows the uptake of new RRT patients over the past 5 years. This is further broken down according to age at start of RRT and gender. It can be seen that the average uptake of new patients is 81 patients per year. There is some year-to-year variation but no trend in this regard. The relative take-on rate for males to females is 1.8:1 for those starting RRT in the first 5 years of life. This falls to 1.2:1 for those starting RRT between 5 and 10 years and reverses to 1:1.03 for the 10–15-year-olds. This relates to the differing patterns of disease causing ERF at differing ages and is discussed in more detail later. The uptake of new patients under the age of 15 years in 1992 was 106 patients. These data suggest that the annual incidence rate of children presenting with ERF is currently stable and certainly not rising at a rate of 5–6% per annum, as was suggested in 1992 based upon the 1992 figures and data from 1984 and 1986. This may well be secondary to an upsurge in the number of patients being treated in the late 1980s and early 1990s as units began to accept patients with ERF starting at birth and in early infancy. Since the initial inclusion of these patients, the numbers have remained stable.

Age group (years)	New patients starting ERF treatment, by year (April to April)															Average		
	1996–97			1997–98			1998–99			1999–2000			2000–2001					
	M	F	T	M	F	T	M	F	T	M	F	T	M	F	T	M	F	T
0–4.99	8	6	14	26	5	31	17	11	28	10	6	16	11	12	23	14.4	8.0	22.4
5–9.99	12	6	18	11	11	22	14	12	26	14	4	18	6	13	19	11.4	9.2	20.6
10–14.99	22	22	44	21	13	34	23	20	43	14	28	32	13	13	26	18.6	19.2	37.8
All <15	42	34	76	58	29	87	54	43	97	38	38	76	30	38	68	44.4	36.4	80.8

Table 16.4: Patients under the age of 15 years entering the RRT programme

Due to the year-to-year variability seen when dealing with a relatively small population of new RRT patients, the incidence rate has been calculated using the average figures over the past 5 years. This is shown with the overall population statistics in Table 16.5. Overall, the new patient rate is 7.4 per million children under the age of 15 years, compared with 9.7 in 1992. For the 1999 report, the age distribution of the RRT population at the start of dialysis was compared with that of the population at the time of the report. These two graphs are shown side by side in Figure 16.2.

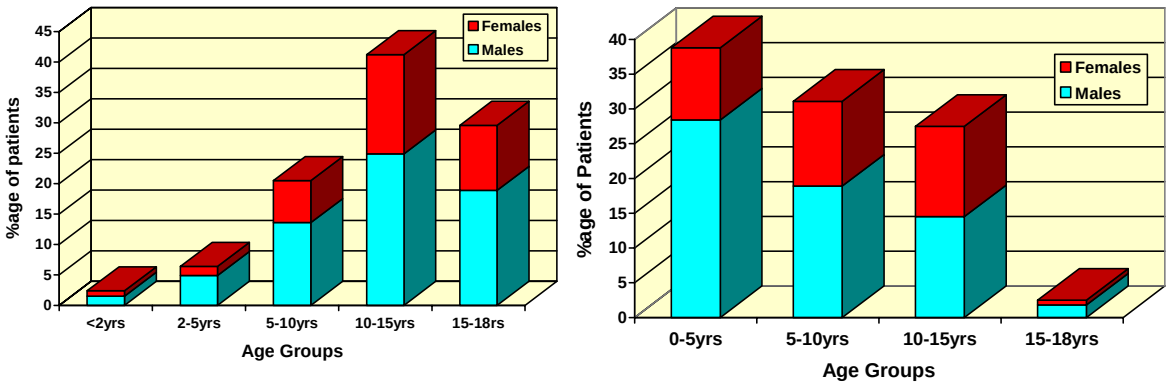

Figure 16.2: Age distribution of the population in 1999 compared with the start of RRT

With the majority of patients commencing dialysis early in life, it was concluded that the most common age of presentation was in early childhood and that the current age distribution was a result of improved life expectancy among this cohort. With the current data, the new patient rate has been analysed by age and gender (Figure 16.3). From these data, it is clear that presentation with ERF occurs throughout childhood, and, if anything, more patients present with ERF over the age of 10 years than below the age of 5.

The current age distribution of the population, as shown in Figure 16.1 above, is not significantly different from that shown in Figure 16.2 for 1999, and similarly there is no real change in the age of commencement of ERF of the current population. The explanation for the apparent discrepancy between the age of starting ERF management for the current patient cohort and the age of presentation with ERF over the past 5 years lies in the regular transfer of older patients to adult units, leaving a disproportionately large cohort of patients commencing ERF management early within the paediatric clinic.

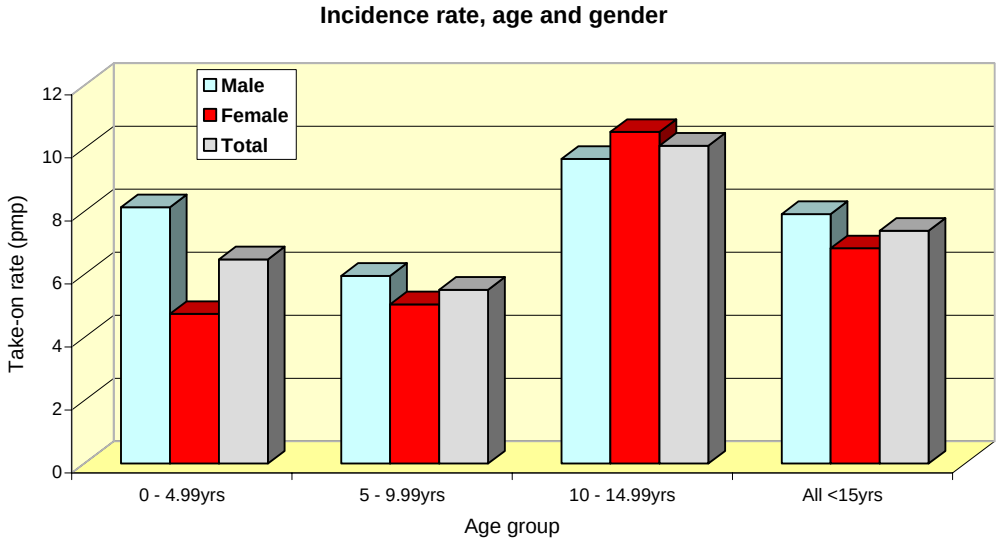

Figure 16.3: Average take-on rate of new ERF patients per year, by age and gender

	UK population (millions)			New patients (average)			Take-on rate (pmp)		
Age group (years)	**Males**	**Females**	**Total**	**Males**	**Females**	**Total**	**Males**	**Females**	**Total**
0–4.99	1.7711	1.6854	3.4565	14.4	8.0	22.4	8.1	4.7	6.5
5–9.99	1.9140	1.8208	3.7348	11.4	9.2	20.6	6.0	5.1	5.5
10–14.99	1.9246	1.8250	3.7496	18.6	19.2	37.8	9.7	10.5	10.1
All <15	5.6097	5.3312	10.9409	44.4	36.4	80.8	7.9	6.8	7.4
UK population	28.6270	29.4309	58.0579				1.6	1.2	1.4

Table 16.5: Incident rate of children under 15 years of age on RRT in the UK

The 1999 report highlighted the relative excess of patients from the Asian subcontinent among the childhood RRT population when compared with the general population. To study this further, the prevalence and new patient rate were analysed by ethnic origin, breaking this down into four groups: White, Black, Asian (defined as patients from the Asian subcontinent of India, Pakistan and Bangladesh) and Other. When looking at data on ethnic distribution, account has to be taken of different age distributions of the population between ethnic groups. Thus, for the White population, 20% of the population is under 16 years of age. For the Indian population, this figure is 23%, and for the Pakistani and Bangladeshi communities it is 37%.

Using these data on age and ethnic distribution, Table 16.6 below shows the number of children in the UK according to ethnic origin, and the prevalence of RRT and incidence rate of new cases of ERF for these populations. It can be seen that both the prevalence and incidence rate for the Asian population was well in excess of that of the other ethnic groups. These data are shown graphically in Figure 16.4. The distribution of ethnic minority groups around the UK is not even, and it might be expected that a concentration of these groups in certain areas would lead to an increased workload and an increased prevalence of ERF for units serving these locations. This would have implications for the distribution of resources and manpower.

Ethnicity	**Population**	**Prevalence (pmp)**	**Take-on rate (pmp)**
Asian	539,386	102.0	23.4
Black	401,531	27.4	4.5
Other	235,229	59.5	9.4
White	9,764,753	44.7	6.6
All <15	10,940,900	47.2	7.4

Table 16.6: Prevalence of ERF in children and take-on rate of new patients by ethnicity

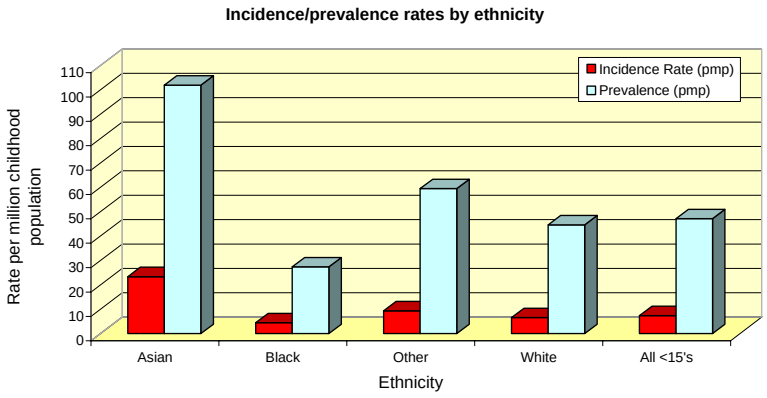

Figure 16.4: Prevalence of RRT in children and take-on rate of new patients by ethnicity

Table 16.7 shows the ethnic distribution of patients below the age of 15 years presenting to the 13 UK paediatric nephrology units over the past 5 years, together with the ethnic distribution of the current patient prevalence under the age of 15 years. Also shown is the approximate childhood population served by each unit, and from this the prevalence and new patient rate for each unit have been calculated.

It can be seen that there are vast differences between the units with regard to the proportion of ethnic minority patients being treated. Although there is a general trend, with regard to both new and existing patients (Figures 16.5a and 16.5b), there is currently no significant correlation between the proportion of the patients on the RRT programme in individual units and the prevalence of RRT in the area covered or the new patient rate. This is likely to be due to the presence of other factors influencing the new patient rate (such as the high prevalence of certain inherited diseases in specific areas) and the imprecision of our current ethnic breakdown. Other evidence suggests that, among patients from the Asian subcontinent, there are wide ranges of disease patterns. Thus, areas with a large Indian population may have a very different incidence and prevalence of childhood ERF from those in areas with a large Pakistani population. This may be studied by a more precise analysis of ethnic origin within the database.

	New patients <15 years of age, 96 – Apr 01					Patient prevalence 15 years age in Apr 01					Population <15 years	Prevalence	TOR
Centre	**White**	**Asian**	**Black**	**Other**	**Total**	**White**	**Asian**	**Black**	**Other**	**Total**	**served**	**(pmp)**	**(pmp)**
Belfast	13	0	0	0	13	18	0	0	0	18	381,468	47.19	6.82
Birmingham	25	10	1	0	36	42	9	1	1	53	1,238,900	42.78	5.81
Bristol	32	1	0	0	33	31	0	0	0	31	881,800	35.16	7.48
Cardiff	7	0	0	0	7	15	0	0	0	15	342,084	43.85	4.09
Glasgow	19	0	0	0	19	40	0	0	0	40	936,147	42.73	4.06
GOS	38	15	5	4	62	64	17	6	8	95	1,780,400	53.36	6.96
Guys	35	6	2	5	48	41	6	4	4	55	1,432,200	38.40	6.70
Leeds	23	13	0	0	36	31	7	0	0	38	756,800	50.21	9.51
Liverpool	15	1	0	0	16	23	1	0	0	24	494,342	48.55	6.47
Manchester	34	14	0	0	48	46	13	0	0	59	919,900	64.14	10.44
Newcastle	23	0	0	0	23	29	1	0	0	30	535,800	55.99	8.59
Nottingham	44	4	0	1	49	47	1	0	1	49	1,122,700	43.64	8.73
Southampton	13	0	0	1	14	9	0	0	0	9	359,000	25.07	7.80

Table 16.7: Prevalence and take-on rate (TOR), by centre and ethnicity

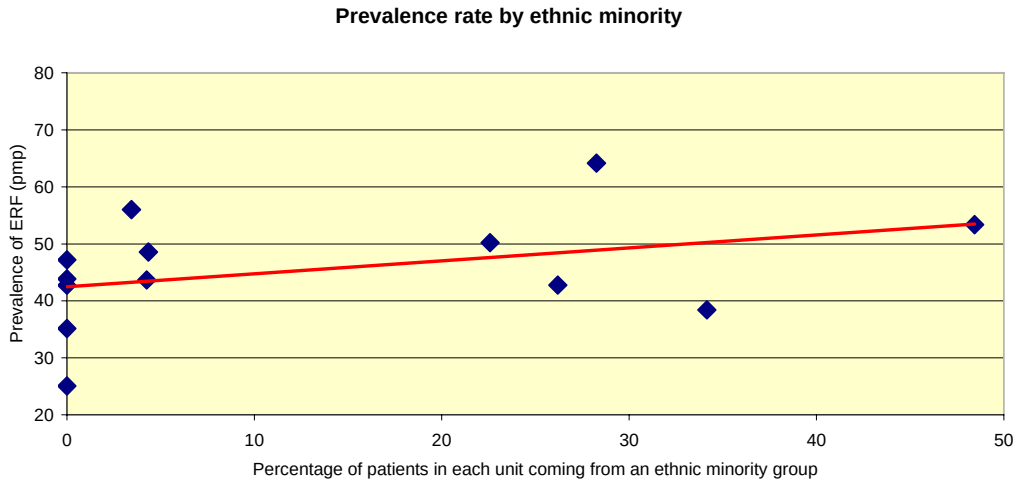

Figure 16.5a: Relationship between take-on rate and % from ethnic minority groups, by centre

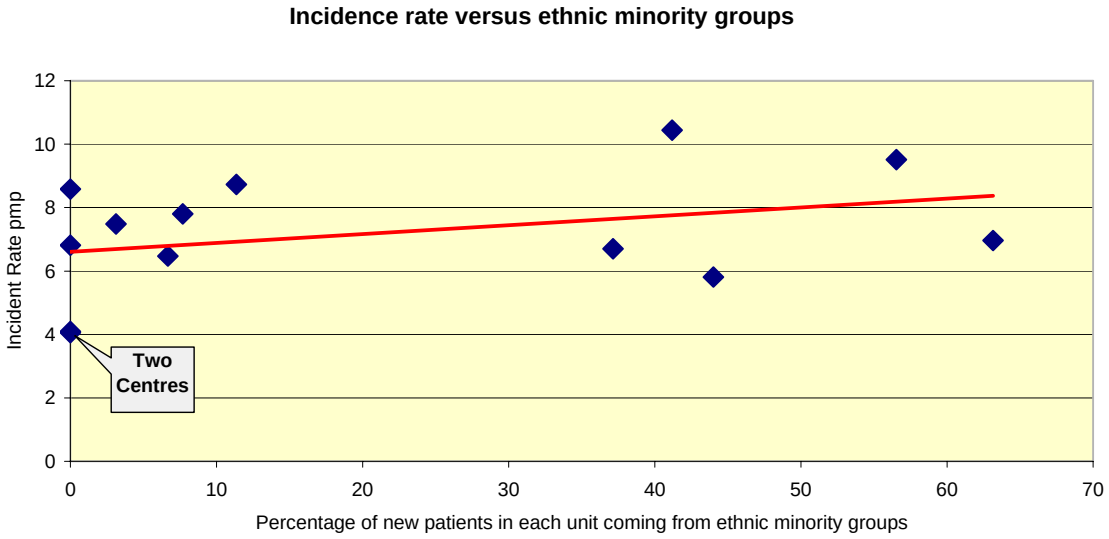

Figure 16.5b: Relationship between prevalence of RRT and ethnic minority group, by centre

Causes of ERF in childhood

The primary cause of ERF was available for 759 of the 800 patients (95%) being cared for in paediatric units on 1 April 2001. The diagnoses are listed alphabetically in Table 16.8 and grouped into disease bands in Table 16.9. Renal dysplasia, either isolated or associated with a syndromal diagnosis, remains the predominant cause of ERF in this population, accounting for 27.7% of the total. Obstructive uropathy is the next largest group, at 19.2%; 73% of this group were boys with posterior urethral valves. Glomerulonephritides account for 18.5% of childhood ERF, with primary focal segmental sclerosis accounting for 38.5% of these cases. The number of patients with reflux nephropathy has fallen slightly from 7.2% to 5.3% of the total. There has been little change in the proportions of patients on RRT secondary to conditions in the other diagnostic headings.

Superficial analysis of the causes of ERF in the childhood population through cohort analysis, as given above, is prone to giving an incorrect picture or disease incidence and prevalence. Diseases that cause ERF in early childhood will create a group of patients who have a prolonged stay on the paediatric unit, whereas patients with diseases presenting late in childhood might be common but underrepresented as their stay with a paediatric unit is limited. To overcome this problem, the primary ERF diagnosis in all the children registered with the BAPN registry in whom both an ERF diagnosis and an age of ERF onset was available has been analysed. Many of these patients are from the current cohort, but some will have been transferred to adult units and some will have died. Only patients with an age of onset of ERF before the age of 15 years were considered so that an accurate picture of pathology in the paediatric age range could be obtained without the data being skewed by patients being referred primarily to adult units.

Diagnosis	Males	Females	Total	%
Acquired obstructive uropathy	2	0	2	0.26
Alport's syndrome	4	2	6	0.79
Anti-GBM disease	0	2	2	0.26
Autosomal recessive polycystic kidney disease	9	5	14	1.84
Barrter's syndrome	0	0	0	0.00
Branchio-oto-renal syndrome	3	1	4	0.53
Chronic renal failure – uncertain aetiology	6	7	13	1.71
Cis-platinum nephrotoxicity	0	0	0	0.00
Congenital nephrotic syndrome (DMS)	5	1	6	0.79
Congenital nephrotic syndrome (Finnish)	10	11	21	2.77
Congenital nephrotic syndrome (FSGS)	3	5	8	1.05
Congenital nephrotic syndrome (unspecified)	7	15	22	2.90
Congenital obstructive uropathy – bladder outlet obstruction (not PUV)	10	2	12	1.58
Congenital obstructive uropathy (not BOO)	5	4	9	1.19
Congenital obstructive uropathy – posterior urethral valves	107	0	107	14.10
Cortical necrosis	8	8	16	2.11
Crescentic glomerulonephritis	1	3	4	0.53
Cyclosporin nephrotoxicity	4	2	6	0.79
Cystinosis	11	11	22	2.90
D-pos haemolytic uraemic syndrome	9	16	25	3.29
D-neg haemolytic uraemic syndrome	2	1	3	0.40
Drug nephrotoxicity (unspecified)	0	0	0	0.00
Glomerulonephritis (unspecified)	2	3	5	0.66
Henoch Schoenlein nephritis	7	9	16	2.11
IgA nephropathy	1	3	4	0.53
Lawrence–Moon–Biedl syndrome	2	2	4	0.53
Megacystis megaureter	2	0	2	0.26
Mesangio-capillary glomerulonephritis type 1	3	2	5	0.66
Mesangio-capillary glomerulonephritis type 2	1	4	5	0.66
Mesoblastic nephroma	1	0	1	0.13
Microscopic polyarteritis nodosa	0	1	1	0.13
Mitochondrial cytopathy	1	1	2	0.26
Multicystic dysplastic kidneys	5	4	9	1.19
Nephrocalcinosis	0	1	1	0.13
Nephronophthisis	23	18	41	5.40
Neuropathic bladder	7	9	16	2.11
Other cytotoxic drug nephrotoxicity	0	2	2	0.26
Polycystic kidney disease (other)	4	1	5	0.66
Primary focal segmental glomerulosclerosis	24	30	54	7.11
Primary hyperoxaluria type 1	2	1	3	0.40
Primary interstitial nephritis	6	4	10	1.32
Proliferative glomerulonephritis	1	1	2	0.26
Prune belly syndrome	14	0	14	1.84
Reflux nephropathy	15	25	40	5.27
Renal artery stenosis	2	1	3	0.40
Renal artery thrombosis	1	1	2	0.26
Renal dysplasia	110	56	166	21.87
Renal hypoplasia	5	6	11	1.45
Renal trauma	0	1	1	0.13
Renal tubular acidosis	2	0	2	0.26
Renal vein thrombosis	6	3	9	1.19

Diagnosis	Males	Females	Total	%
Systemic lupus erythematosus	0	3	3	0.40
Tuberous sclerosis polycystic kidney disease	0	1	1	0.13
Tubular disorders (other)	1	0	1	0.13
Vasculitis (unspecified)	1	3	4	0.53
Wegner's granulomatosis	0	1	1	0.13
Wilms' nephropathy	1	0	1	0.13
Wilms' tumour	5	5	10	1.32

Table 16.8: Primary ERF diagnosis for the paediatric RRT population on 1 April 2001

Diagnostic group	Males	Females	Total	% of total
Renal dysplasia and related conditions				
Renal dysplasia	110	56	166	21.87
Prune belly syndrome	14	0	14	1.84
Renal hypoplasia	5	6	11	1.45
Multicystic dysplastic kidneys	5	4	9	1.19
Branchio-oto-renal syndrome	3	1	4	0.53
Lawrence–Moon–Biedl syndrome	2	2	4	0.53
Megacystis megaureter	2	0	2	0.26
Total with primary renal dysplasia	141	69	210	27.67
Obstructive uropathy				
Posterior urethral valves	107	0	107	14.10
Neuropathic bladder	7	9	16	2.11
Congenital bladder outlet obstruction (not PUV)	10	2	12	1.58
Congenital obstructive uropathy (not BOO)	5	4	9	1.19
Acquired obstructive uropathy	2	0	2	0.26
Total with obstructive uropathy	131	15	146	19.24
Glomerulonephritis, vasculitis and glomerulopathy				
Primary focal segmental glomerulosclerosis	24	30	54	7.11
D-pos haemolytic uraemic syndrome	9	16	25	3.29
Henoch Schoenlein nephritis	7	9	16	2.11
Alport's syndrome	4	2	6	0.79
Glomerulonephritis (unspecified)	2	3	5	0.66
Mesangio-capillary glomerulonephritis type 1	3	2	5	0.66
Mesangio-capillary glomerulonephritis type 2	1	4	5	0.66
Crescentic glomerulonephritis	1	3	4	0.53
IgA nephropathy	1	3	4	0.53
Vasculitis (unspecified)	1	3	4	0.53
D-neg haemolytic uraemic syndrome	2	1	3	0.40
Systemic lupus erythematosus	0	3	3	0.40
Anti-GBM disease	0	2	2	0.26
Proliferative glomerulonephritis	1	1	2	0.26
Microscopic polyarteritis nodosa	0	1	1	0.13
Wegner's granulomatosis	0	1	1	0.13
Total with glomerular disease	56	84	140	18.45
Reflux nephropathy and CRF of uncertain aetiology				
Reflux nephropathy	15	25	40	5.27
Chronic renal failure – uncertain aetiology	6	7	13	1.71
Total with reflux nephropathy and CRF of uncertain aetiology	21	32	53	6.98
Primary tubular and interstitial disorders				
Nephronophthisis	23	18	41	5.40
Primary interstitial nephritis	6	4	10	1.32
Renal tubular acidosis	2	0	2	0.26
Tubular disorders (other)	1	0	1	0.13
Barrter's syndrome	0	0	0	0.00
Total with primary tubular and interstitial disorders	32	22	54	7.11
Congenital nephrotic syndrome				
Congenital nephrotic syndrome (unspecified)	7	15	22	2.90

Diagnostic group	Males	Females	Total	% of total
Congenital nephrotic syndrome (Finnish)	10	11	21	2.77
Congenital nephrotic syndrome (FSGS)	3	5	8	1.05
Congenital nephrotic syndrome (DMS)	5	1	6	0.79
Total with congenital nephrotic syndrome	25	32	57	7.51
Renal vascular disorders				
Cortical necrosis	8	8	16	2.11
Renal vein thrombosis	6	3	9	1.19
Renal artery stenosis	2	1	3	0.40
Renal artery thrombosis	1	1	2	0.26
Renal trauma	0	1	1	0.13
Total with renal vascular disorders	17	14	31	4.08
Metabolic diseases and drug nephrotoxicity				
Cystinosis	11	11	22	2.90
Cyclosporin nephrotoxicity	4	2	6	0.79
Primary hyperoxaluria type 1	2	1	3	0.40
Mitochondrial cytopathy	1	1	2	0.26
Other cytotoxic drug nephrotoxicity	0	2	2	0.26
Nephrocalcinosis	0	1	1	0.13
Cis-platinum nephrotoxicity	0	0	0	0.00
Drug nephrotoxicity (unspecified)	0	0	0	0.00
Total with metabolic diseases and drug nephrotoxicity	18	18	36	4.74
Polycystic kidney disease				
Autosomal recessive PKD	9	5	14	1.84
Tuberous sclerosis PKD	0	1	1	0.13
Polycystic kidney disease (other)	4	1	5	0.66
Total with polycystic kidney disease	13	7	20	2.64
Malignant and related diseases				
Wilms' tumour	5	5	10	1.32
Mesoblastic nephroma	1	0	1	0.13
Wilms' nephropathy	1	0	1	0.13
Total with malignant and related diseases	7	5	12	1.58

Table 16.9: Primary ERF diagnoses, grouped by disease category

For this analysis, there were 882 patients with data available. These patients were divided into three groups, consisting of 324 patients on RRT commencing before the age of 5 years, 257 patients on RRT commencing between the ages of 5 and 10 years, and 301 patients on RRT commencing between the ages of 10 and 15 years. Figure 16.6 shows the gender distribution of these three groups. As expected, there is a massive preponderance of boys in the under-5 age group, which reduces in the 5–10-year-old group. Interestingly, there are still more boys than girls in the 10–15-year-old group, contrary to the trend over the past 5 years shown in Figure 16.3 above, where girls presenting with ERF outnumber boys at this age. This difference was not statistically significant.

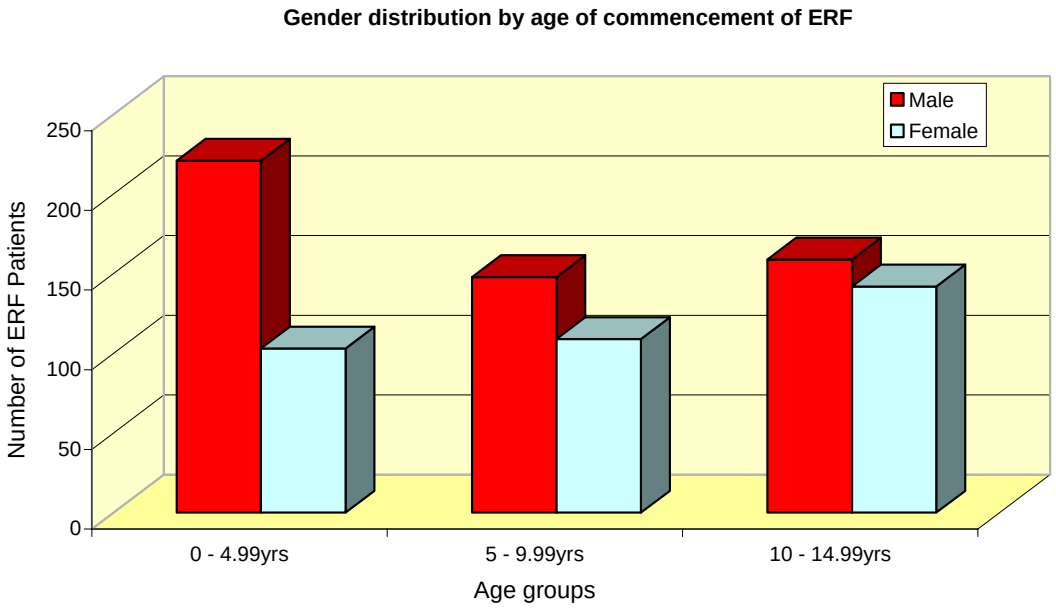

Figure 16.6: Gender distribution of patients, by age at start of RRT

Table 16.10 shows the diagnostic groupings used for the ERF population and the percentage of patients belonging to each group for the current cohort of patients and the cohorts of patients presenting at different ages. The data from the three age cohorts are shown graphically in Figure 16.7. It is clear that there are some diagnostic groups, such as the congenital nephrotic syndromes, in which the vast majority of patients enter ERF early in life, some entering in the middle years of childhood, and in which the commencement of ERF after the age of 10 years is rare. Polycystic kidney diseases and malignant and related disorders follow a similar pattern. Other diseases, for example glomerular diseases, tubular and interstitial disorders and metabolic diseases, become increasingly common with age.

Diagnostic group	Percentage of children on RRT			
	Current patient cohort	ERF start <5 years	ERF start 5–10 years	ERF start 10–15 years
Primary renal dysplasia	27.52	31.79	28.40	17.61
Obstructive uropathy	19.13	22.22	17.12	16.28
Glomerular disease	18.35	8.64	21.40	26.91
Reflux nephropathy and CRF of uncertain aetiology	6.95	3.09	6.23	17.61
Primary tubular and interstitial disorders	7.08	3.70	7.78	10.30
Congenital nephrotic syndrome	7.47	15.74	3.89	0.33
Renal vascular disorders	4.06	5.56	4.28	1.99
Metabolic diseases and drug nephrotoxicity	4.72	0.93	9.34	6.98
Polycystic kidney disease	2.62	5.25	1.17	1.00
Malignant and related diseases	1.57	3.09	0.39	1.00

Table 16.10: Percentage of patients, by diagnostic group and age at start of RRT
CRF, chronic renal failure.

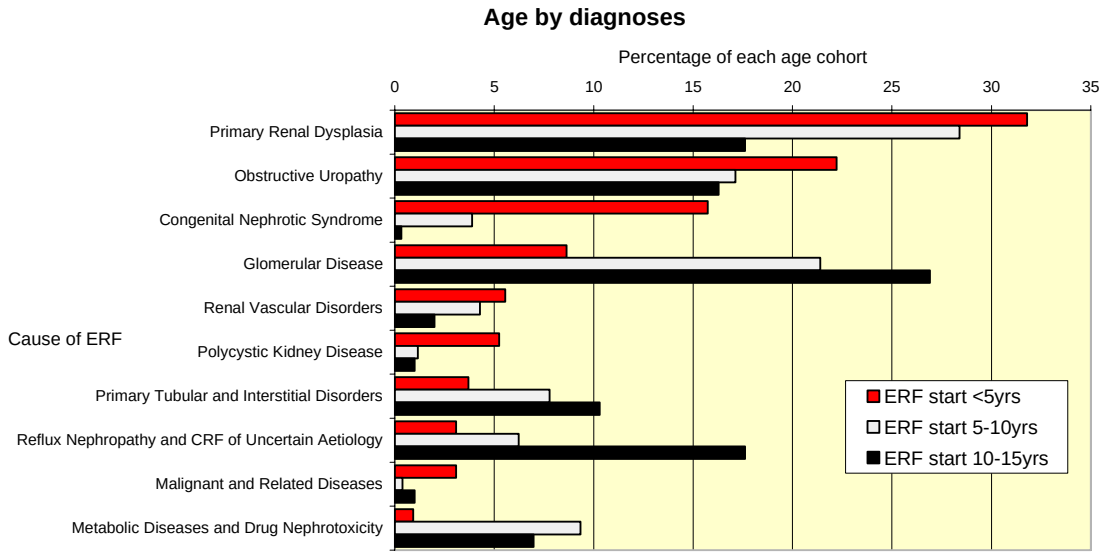

Figure 16.7: Percentage of patients in each diagnostic group, by age at start of RRT

The importance of primary focal segmental glomerulosclerosis (FSGS) as a cause of ERF in childhood is shown in Figure 16.8. Here, the number of children in each age group on RRT secondary to a glomerulopathy is shown, together with the number of these that are secondary to primary FSGS. Primary FSGS accounts for almost 50% of ERF from glomerular disease in the first decade of life. Other major contributions to this disease category are Henoch Schoenlein nephritis and ERF secondary to haemolytic uraemic syndrome. All other forms of glomerulonephritis and systemic vasculitis causing ERF are rare in childhood.

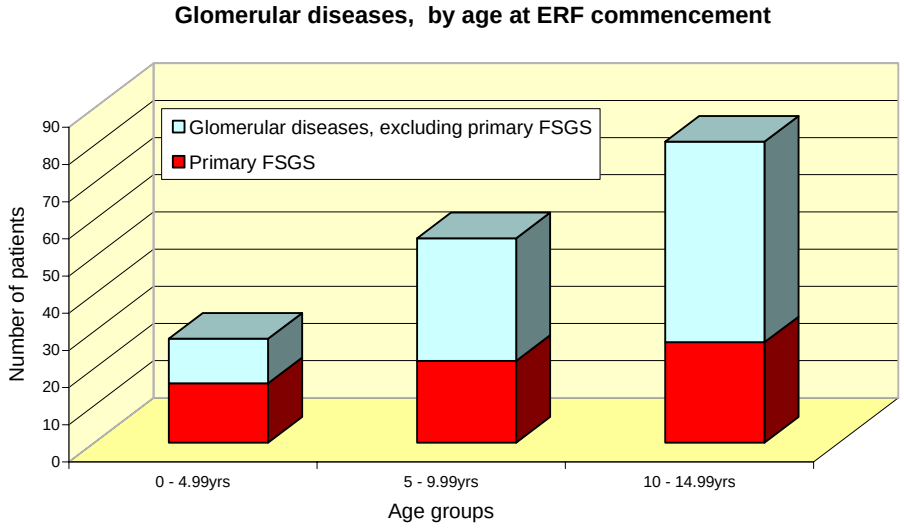

Figure 16.8: Glomerular diseases causing ERF in childhood, by age at start of RRT

Obstructive uropathy is one of those conditions that is very common in those presenting with ERF at a young age and reduces in frequency as a cause of ERF with increasing age. The reduction is, however, not as dramatic as with other conditions, even though the vast majority of patients have congenital lesions. Figure 16.9 shows this graphically. Posterior urethral

valves account for almost all ERF resulting from obstructive uropathy in the first 5 years of life and for 59% of ERF arising from obstructive uropathy in patients presenting between the ages of 10 and 15 years. Other congenital abnormalities causing obstruction account for 12.7% of patients on RRT from obstructive uropathy. The only significant acquired form of obstructive uropathy in childhood is neuropathic bladder, which accounts for 9.7% of these patients. These data emphasise the need for a close follow-up of all patients with congenital obstructive uropathies and also the requirement for the early diagnosis and active management of neuropathic bladder in childhood.

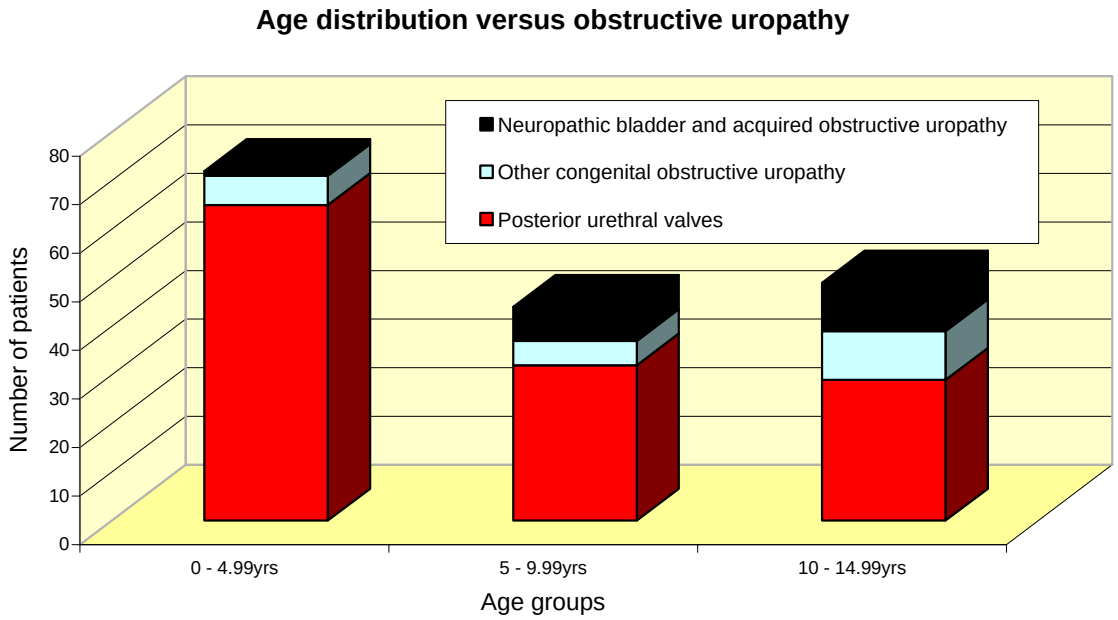

Figure 16.9: Age distribution and type of obstructive uropathy in children on RRT

Renal dysplasia, the predominant cause of ERF in the childhood population, shows a similar and more dramatic fall in incidence with age. There are several syndromal conditions associated with renal dysplasia (such as prune belly syndrome and branchio-oto-renal syndrome), but, as can be seen from Figure 16.10, the majority of cases have primary dysplasia or hypoplasia. From Figure 16.7 above, it can be seen that reflux nephropathy and chronic renal failure of uncertain aetiology follow the opposite distribution, steadily rising in frequency with age. Figure 16.11 shows that this is accounted for by an increasing incidence of reflux nephropathy, with only a small cohort throughout having chronic renal failure of uncertain aetiology. This is important as one of the achievements of paediatric nephrology has been the apparent reduction of ERF caused by reflux nephropathy. In those presenting with ERF between 10 and 15 years of age, however, reflux nephropathy still accounts for 14.6% of all renal failure. What is intriguing is the relationship between renal dysplasia (in which vesico-ureteric reflux is often an associated feature) and reflux nephropathy. Paediatric nephrologists are well accustomed to patients with vesico-ureteric reflux who develop numerous urinary infections but do not progress to a decline in renal functional. Similarly, patients presenting in ERF with what is felt to be reflux nephropathy often have no preceding history of urinary sepsis.

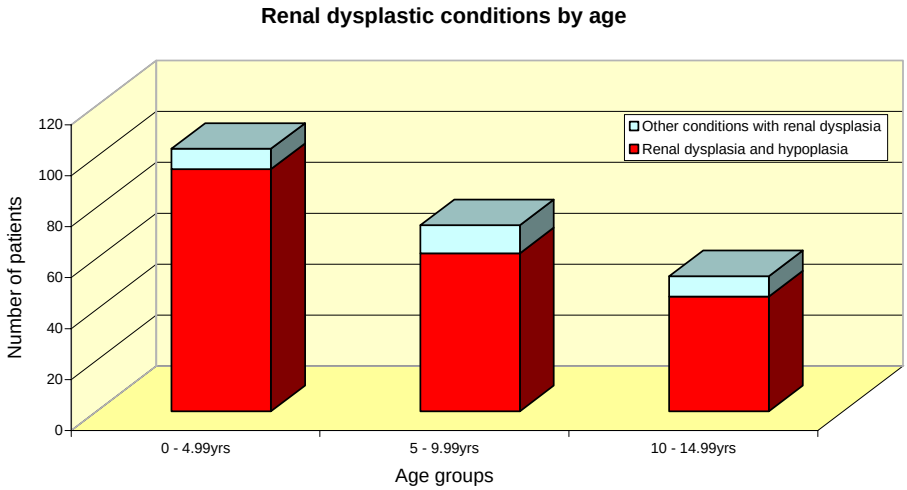

Figure 16.10: Patients with renal dysplastic conditions causing ERF, by age at start of RRT

Figure 16.12 shows the percentage of patients with ERF from renal dysplasia and that of patients with reflux nephropathy summated. It is clear that the overall incidence of either reflux nephropathy or renal dysplasia as a cause of ERF is virtually constant throughout childhood. There is, however, a shift from the use of renal dysplasia as a diagnostic category to reflux nephropathy with age. This raises the possibility that this is a single condition (renal dysplasia with or without vesico-ureteric reflux) with a varied time progression to renal failure. History has dictated that patients presenting in late childhood with ERF, small shrunken kidneys and vesico-ureteric reflux are labelled as having reflux nephropathy. Treating all these patients as children with renal dysplasia and the eventual exhaustion of a reduced and abnormal nephron mass would explain the ongoing presentation of reflux nephropathy at a time at which the awareness of urinary infection is increased. It also explains the absence of a history of urinary infection in patients presenting in ERF.

One unexplained feature of the 1999 report analysis was the excess of boys with renal dysplasia, which had not been seen before or reported in Registry reports from other countries. The ratio of boys to girls with renal dysplasia as a cause of ERF remains high, at 1.9:1 (2:1 in the 1999 report). When these are combined with patients with reflux nephropathy, the ratio falls to 1.4:1. For both renal dysplasia and reflux nephropathy, girls present at a later age than boys (Figure 16.13).

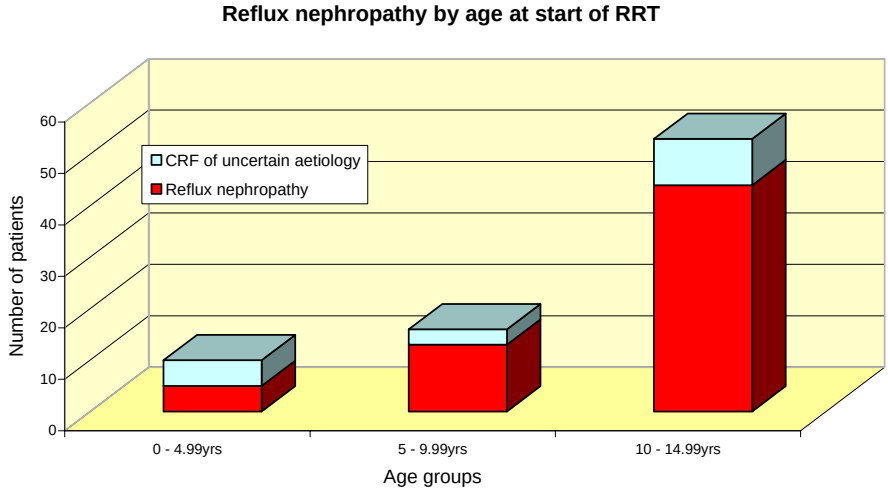

Figure 16.11: Patients with reflux nephropathy causing ERF, by age at start of RRT

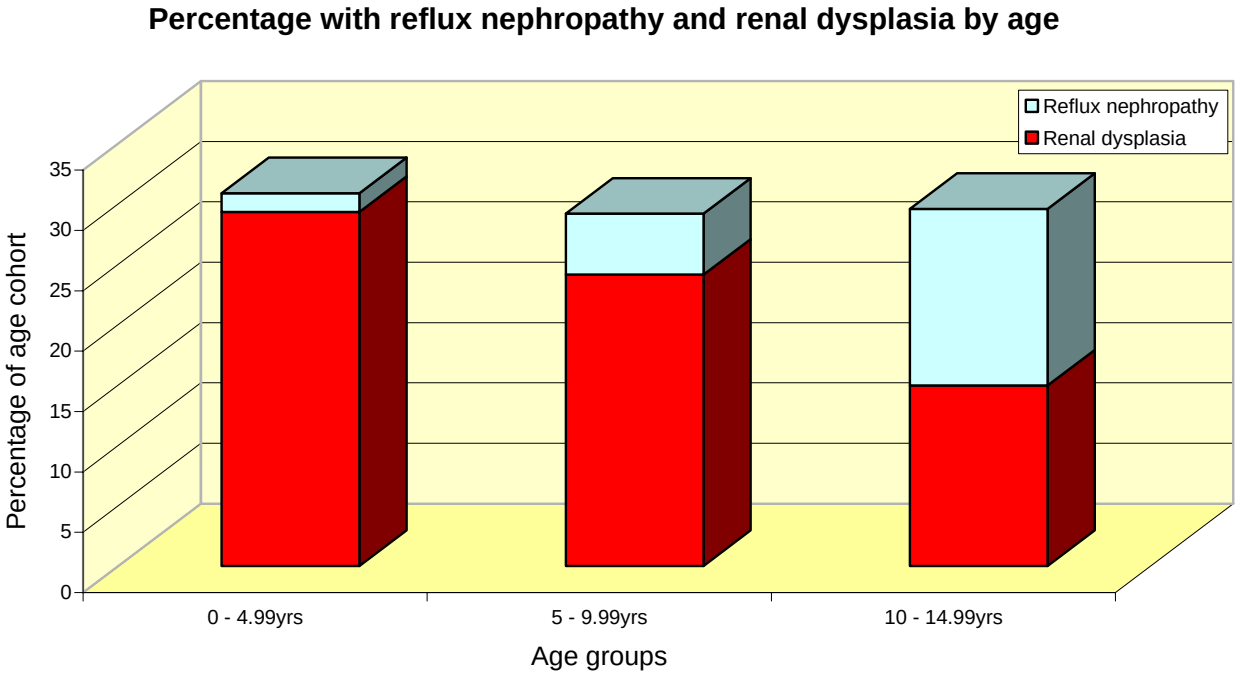

Figure 16.12: % of patients with renal dysplasia and reflux nephropathy causing ERF, by age group

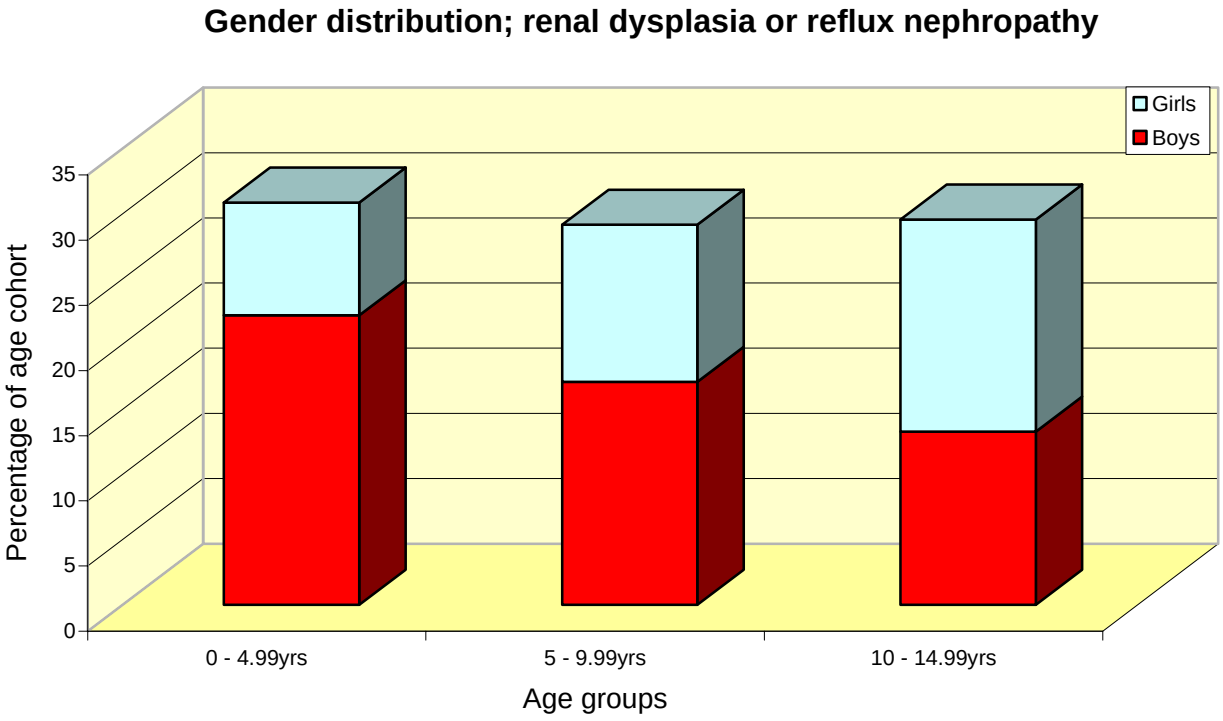

Figure 16.13: Gender distribution of children with renal dysplasia or reflux nephropathy, by age at start of RRT

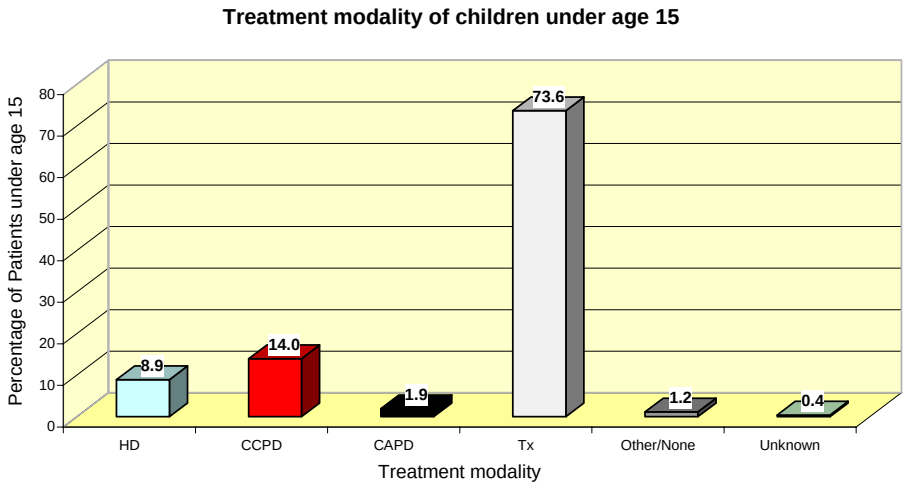

Figure 16.14: ERF management of children under 15 years of age in the UK

Current management of ERF in childhood

As shown in the 2000 annual report of the UK Renal Registry, the majority of children on RRT in the UK have functioning renal allografts. Data from April 2001 are shown in Figure 16.14; it is clear that this is still the case, 73.6% of patients cared for in the 13 regional paediatric units having a functioning renal allograft in April 2001. For those on dialysis, cycling peritoneal dialysis was the most popular form. Peritoneal dialysis was being used in 64% of patients and haemodialysis in 36%. Of those who were on peritoneal dialysis, 88% were having overnight cycling dialysis, with or without a daytime dwell, and just 12% were being treated with continuous ambulatory peritoneal dialysis (CAPD).

Although there has been a small fall in the proportion of patients with a functioning allograft, from 76% in 2000 to 73.6% currently, there has also been a reduction in the rate of transplantation. These percentages are protected by the buffer effect of having many young children, transplanted early, who remain with functioning allografts throughout their stay with the paediatric unit. Figure 16.15 shows the number of transplants performed in children under the age of 15 years in the 11 UK centres for paediatric renal transplantation over the past 6 years. It can be seen that there has been a trend towards a reduced number of cadaveric allografts over the past 5 years. The effect on the total number of transplants has been reduced by the increasing number of living related donations.

With the high flux of paediatric patients, because of their presentation at varying times in childhood and then their movement a few years later into an adult unit, this reduction in the rate of transplantation has an effect on the workload of the paediatric renal units. Looking at the population of children with renal transplants in 2000, 22% received a pre-emptive allograft, avoiding a preceding period of dialysis. Figure 16.16 shows the proportion of new patients presenting in each of the 5 years from 1996 to 2001 who received a pre-emptive transplant. It can be seen that the recent trend is towards a decreasing number of pre-emptive cadaveric allografts, and, as there is no trend towards increasing pre-emptive living related donations, there is an overall reduction in the total number of pre-emptive transplants.

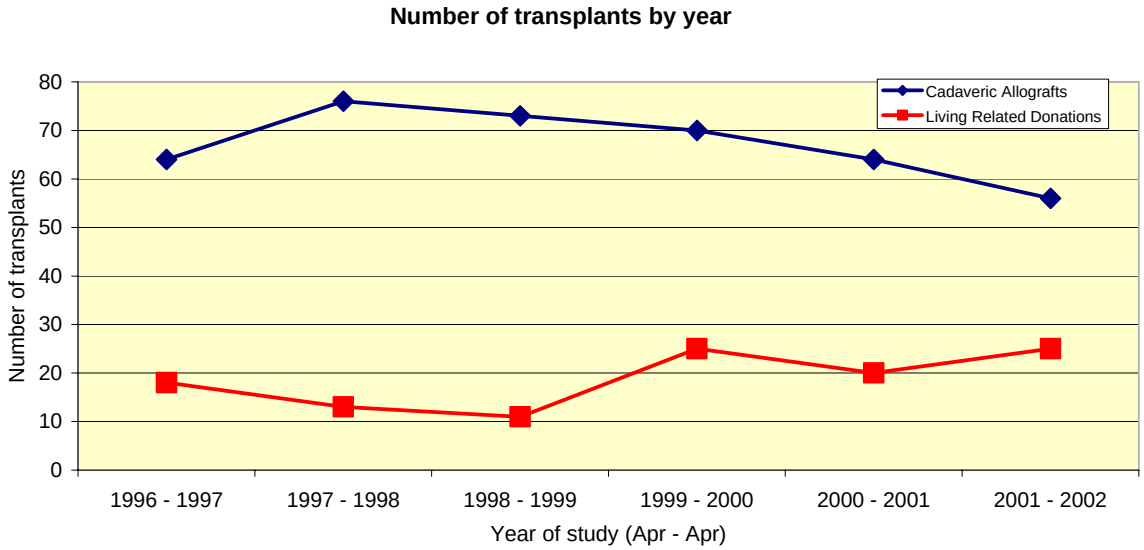

Figure 16.15: Transplant activity in patients aged <15 years over the past 6 years

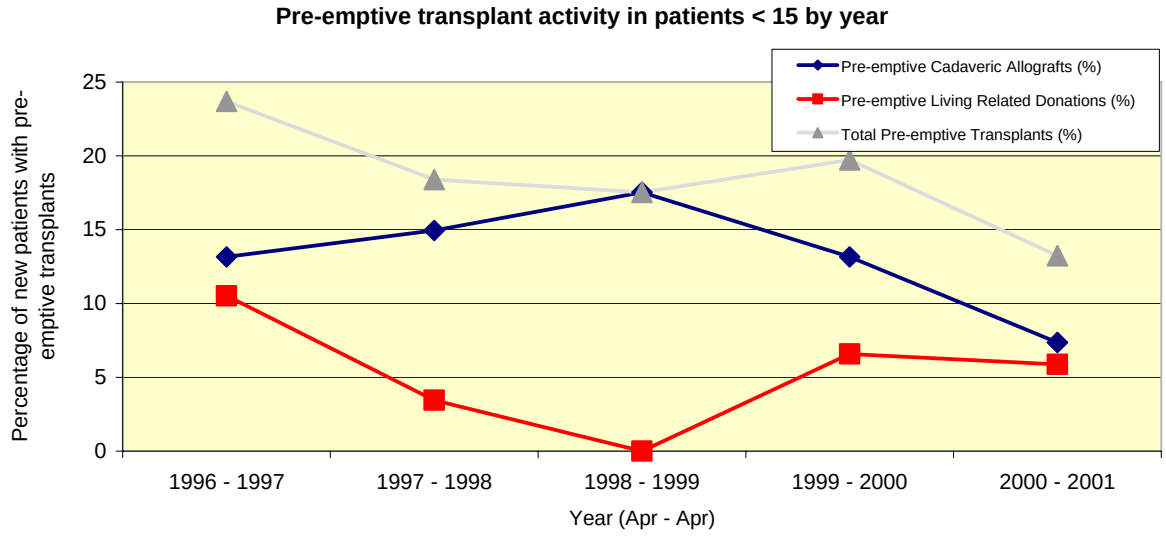

Figure 16.16: Pre-emptive transplants in children under the age of 15 years

As there has been no significant change in the number of patients presenting to paediatric renal units with ERF over the past 5 years, the result of the above is an increase in the dialysis workload for the individual units. Figure 16.17a shows the number of under-15-year-old patients on RRT in the 13 paediatric units in the UK, divided according to whether they have a functioning allograft or are on dialysis. Figure 16.17b shows these same data but divides the patients according to the proportion on each treatment modality rather than the absolute number. It can be seen that there is a large variability between units in their dialysis workload, ranging from just 7% of patients being on dialysis to 43% being on dialysis. Table 16.11 shows these data by individual renal units in greater detail. In addition to the variable proportion of patients with a transplant, it is clear that there is great variability in the

proportion receiving haemodialysis. All this has major implications for the provision of resources. Staffing levles in units with a high proportion of dialysis patients, particularly those with many haemodialysis patients, will need to increase. Paediatric haemodialysis patients often require one-to-one nursing during dialysis sessions, and a further analysis of the current trends over the next few years will be important for both manpower planning and the provision of appropriate training.

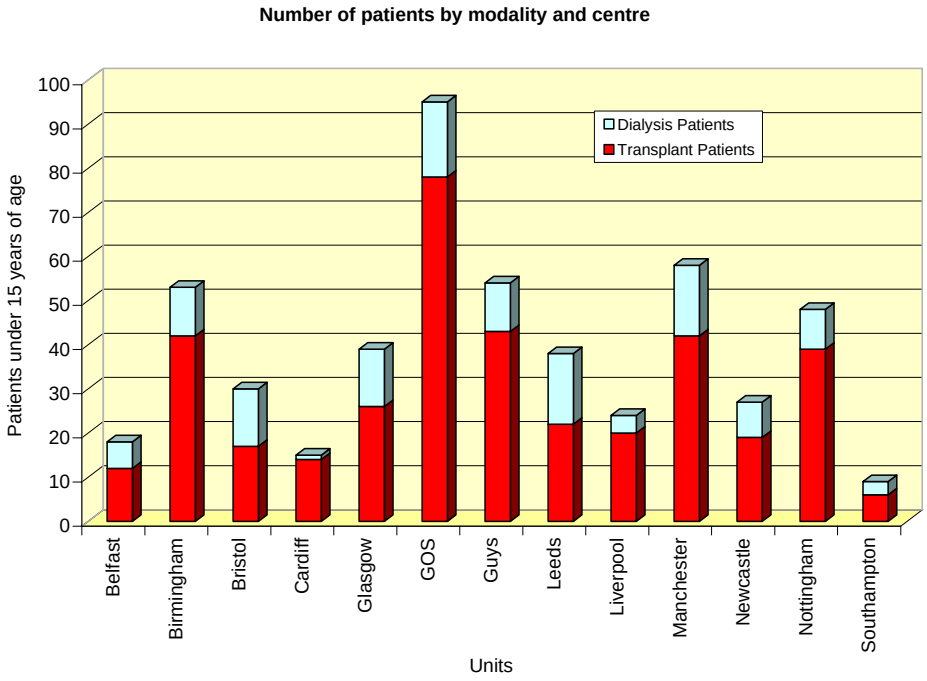

Figure 16.17a: Patients aged <15 years on dialysis or with a functioning allograft, by centre

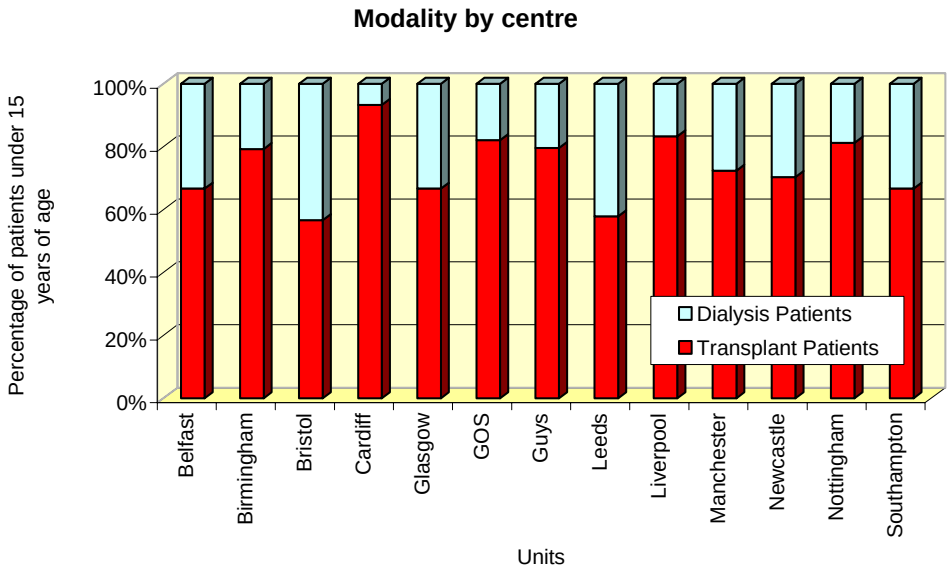

Figure 16.17b: Proportion of patients under 15 years of age, by modality and centre

	Treatment modality						
Centre	**HD**	**CCPD**	**CAPD**	**Tx**	**Other/none**	**Unknown**	**Total**
Belfast	2	4	0	12	0	0	18
Birmingham	5	5	1	42	0	0	53
Bristol	5	8	0	17	1	0	31
Cardiff	0	1	0	14	0	0	15
Glasgow	2	11	0	26	0	1	40
GOS	9	6	2	78	0	0	95
Guys	5	6	0	43	0	1	55
Leeds	6	9	1	22	0	0	38
Liverpool	2	2	0	20	0	0	24
Manchester	3	7	6	42	1	0	59
Newcastle	5	3	0	19	3	0	30
Nottingham	2	7	0	39	1	0	49
Southampton	0	3	0	6	0	0	9
Total	46	72	10	380	6	2	516

Table 16.11: Modality of patients under the age of 15 years, by centre, on 1 April 2001
HD, haemodialysis; CCPD, continuous cycling peritoneal dialysis.

Conclusion

Although there has been a gradual increase in the total number of patients being cared for in paediatric renal units in the UK, the overall incidence of ERF in children appears to be fairly stable. The local incidence and prevalence of ERF are affected by the ethnic mix of the population served, and the high rate of ERF in children with ethnic origins in the Asian subcontinent may have implications for the provision of services locally and the resources that need to be made available. ERF presents at all ages in childhood, with a slowly increasing incidence with age. Males significantly outnumber females in early childhood, but by the teenage years more females are presenting than males.

The causes of childhood ERF vary with age. Renal dysplasia and obstructive uropathy are the predominant causes in the first 5 years of life. For those presenting between 5 and 10 years of age, renal dysplasia is still the most common cause, but glomerular diseases are the second leading cause. For those presenting after the age of 10 years, glomerular diseases become the most common cause, relegating renal dysplasia to second place. Within this latter age group, reflux nephropathy remains a significant cause of renal failure, accounting for 15% of all cases. The combination of renal dysplasia and reflux nephropathy accounts for just under 30% of all cases of childhood ERF across the age ranges, raising the possibility that these are different ends of the same spectrum of disease. Indeed, it may well be better to relabel the two diagnostic entities together as ‘congenital renal malformation with or without vesico-ureteric reflux but without obstruction’.

Transplantation remains the overwhelming treatment of choice for children on RRT, and 73% of children have functioning allografts. The rate of cadaveric transplantation appears to be decreasing, however, offset slightly by an increasing rate of living related donations. Fewer pre-emptive transplants are now being performed, which is leading to an increasing workload for paediatric dialysis units. If this trend continues, there will be significant resource implications. For those on dialysis, 64% are on peritoneal dialysis and 36% haemodialysis. Cycling peritoneal dialysis is the modality of choice, with only a small number of patients on CAPD.

Acknowledgements

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The report was reviewed by the Paediatric Renal Registry Subcommittee, comprising: Dr Jane Tizard, Dr Chris Reid, Dr William van't Hoff, Prof Adrian Woolf, Dr Rodney Gilbert and Dr Malcolm Lewis.

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