
Infant Dialysis

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Abstract

Although the number of infants needing end stage renal failure (ESRF) management is small their care continues to present many ethical and practical challenges. The mortality in children starting dialysis age less than 1 year is higher compared to older children, with co-morbid non-renal disease a highly significant risk factor. Families must be carefully counselled as to the possible management options. Peritoneal dialysis is the treatment of choice if the infant is to be actively managed. Most infants will need enteral feeding and frequent adjustment of dialysis prescription, medications and feed recipe are necessary. Haemodialysis can be undertaken but problems with access are very common. The prognosis for survival, linear growth and development in otherwise normal infants is encouraging. Infant dialysis can now be performed in most infants with ESRF with the aim of facilitating early renal transplantation.

Key words

End stage renal failure; Haemodialysis; Infants; Outcome; Peritoneal dialysis

Introduction

The decision to embark on a programme of end stage renal failure (ESRF) management in an infant continues to present an ethical and practical challenge for the paediatric nephrology team. This is a time of great emotional vulnerability for parents. With antenatal diagnosis, termination may have been discussed and if the pregnancy continues there may have been foetal interventions. During the pregnancy, parents may have had time to consider the possible treatment options for their infant, or unexpected perinatal events will confront parents with a situation for which there can be no preparation. Paediatric nephrologists and their multidisciplinary teams will have different attitudes to the care of the infant with ESRF^{1,2} but their

responsibility is to ensure families are fully informed about the management and possible outcome for their baby before starting long-term renal replacement therapy.

What Is the Scope of the Problem?

The true incidence of ESRF is unknown as terminations for renal anomalies and early deaths in infants with associated co-morbid conditions, never referred for ESRF management may not be reported. In the UK, the incidence of ESRF in children age less than 15 remains stable but the prevalence has increased over the last 10 years. Total numbers remain small with an average of 22.4 children under 5 years starting ESRF treatment each year since 1996 with 13 children, age 0-1.9 years prevalent in April 2001.³ This represents an incidence of 6.5 per million population age 0-4.99 years with the US renal data system reporting 11 incident patients per million age 0-4 years.⁴ Although infants are only 1-2% of the paediatric ESRF population they require a disproportionately greater input of resources and skill to establish and maintain successful dialysis.

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Prognostic Factors and Survival

The NAPRTCS 2001 study of 2,221 children age 0-21 starting dialysis between 1992-2000 gives a patient survival rate of 95%, 90.1% and 85.7% at 1, 2 and 3 years respectively.⁵ The mortality was higher in children starting dialysis age less than 1 year at 84.5%, 74.4% and 68.2% by 3 years. Further analysis of the NAPRTCS data by Wood et al identified co-morbid non-renal disease particularly pulmonary hypoplasia and anuria or oliguria if age less than 2 years as risk factors.⁶ Smaller studies have all emphasised the adverse impact of co-morbidity on survival including severe developmental delay, pulmonary hypoplasia and congenital heart disease.⁷⁻⁹ At Great Ormond Street Hospital for Children (GOS) the mortality of 101 children presenting at age <6 months with severe chronic renal failure (GFR less than 20 ml/min/1.73m²) or ESRF was 87% at 1 year and 78% at 5 years.¹⁰ The commonest diagnosis was renal dysplasia with or without reflux (54%) and posterior urethral valves (25%) followed by congenital nephrotic syndrome (11%). In addition 26 were preterm and 18 of low birthweight. There were 18 deaths in the first 2 years of life with co-morbidity a significant factor in thirteen. Although we can now feel more optimistic that intensive treatment of the infant with ESRF offers the best outcome for growth, development and subsequent transplantation, the long-term outcome for these children is still unknown.⁹⁻¹² An unpublished analysis of 77 children taken onto the dialysis programme at GOS for at least 3 months between 1984-1997 with a minimum of 5 years follow-up illustrates the continuing uncertainties faced by these children and their families. The overall patient survival was 77% at a median follow-up of 7.5 years. Sixty-nine of the children have had 99 renal transplants and 14 have been maintained on long-term dialysis (>5 years). Currently, 62% have functioning renal transplants and 16% are on dialysis. The majority of the 15 (19%) deaths on dialysis and post-transplant were again associated with other serious non-renal conditions.

What Are the Treatment Options?

Parents are faced with several choices for the care of their infant with ESRF; no active intervention/palliative care, supportive care to monitor the effect of time on renal function or intensive treatment to try and ensure optimum brain and somatic growth and facilitate early renal transplantation. However, if possible, treatment decisions

should not be rushed and must not be considered irrevocable.

Palliative Care

In some circumstances families may feel that their child will suffer without any certainty that they will have an acceptable quality of life. This is particularly likely if there are co-morbid conditions. In our series of 101 infants with ESRF, 13 were not actively managed of whom 10 died in the first year of life. Although 10 had severe co-morbidity including Jeune's and Down's syndrome, cystic fibrosis, blindness and developmental delay, 3 were otherwise normal.¹⁰ Families must be appropriately counselled and decisions taken in partnership with the multidisciplinary team.¹³

Supportive Care

Parents may be able to adopt a 'wait and see' policy as there can be spontaneous improvement in renal function during the first year of life. Infants with polyuria and severely compromised renal function can survive for many months as fluid overload and hyperkalaemia are rare. Relief of obstruction and prevention of urinary sepsis, correction of electrolyte disturbances and anaemia and attention to nutrition including early enteral feeding can contribute to stabilisation of renal function and improved growth.¹⁴ In a study of 11 infants with renal dysplasia, only those with a calculated GFR of <15 ml/min/1.73 m² at age 6 months had no improvement of renal function.¹⁵ However infants with growth failure and electrolyte disturbances despite supportive conservative management will need dialysis for an optimal outcome.

Intensive Management

Infant dialysis requires skilled input from medical, nursing and dietetic staff and unremitting commitment from families. The emotional and often financial burden should not be underestimated. Although peritoneal dialysis (PD) frees families from three times weekly hospital visits for haemodialysis (HD) it increases the responsibilities placed upon them at home. Most infants will require overnight feeds delivered by a pump and sleep is often disturbed by dialysis machine alarms. Hospital visits are frequent to adjust feeds, medications and dialysis prescription and readmission may be necessary to treat dialysis complications or surgery for the original condition. However PD is the treatment of choice as vascular access for HD is difficult and blood priming of lines may be necessary in small infants risking HLA sensitisation.

Peritoneal Dialysis

Most infants will require 12-14 hours of overnight continuous cycling peritoneal dialysis to remove the solute and water provided by the diet prescribed, to ensure adequate growth. Double-cuffed catheters with a swan-neck tunnel and downward pointing exit site are placed and if possible use is delayed for 2-3 weeks to allow healing. Catheter complications are common. An analysis of 20 infants on PD for up to 6 years in our centre found that only 2 infants did not require any catheter intervention and 12 needed at least one replacement.⁹ The peritonitis rate was 1.1 episodes/patient year but 70% of infants had only 0-1 episodes despite the presence of a urinary diversion in five infants. Although there is no robust data on adequacy of dialysis and outcome, the current recommendations are a fill volume of 1100-1400 mls/m² body surface area with a daytime dwell if possible, to provide optimal dialysis as tolerated.¹⁶ The majority of infants will need enteral feeding as spontaneous oral intake is rarely adequate in infants with ESRF who also frequently vomit due to a high incidence of gastro-oesophageal reflux.¹⁷ As growth is primarily nutrition dependent in the first 2 years of life, early intervention is vital to improve height prognosis. Calorie and protein requirements may be as high as 150 kcal/kg/day and 3 g protein/kg/day in term infants.¹⁸ Although in the very young infant feeds can be initially be delivered using a nasogastric tube, a gastrostomy can subsequently be placed; percutaneously if the infant is still conservatively managed or by an open procedure if established on PD.¹⁹ With slow delivery of small volumes of feed continuously overnight, supplemented by daytime boluses as necessary vomiting often improves although in some infants a fundoplication may be necessary.^{10,14} There can be continuing tubular losses of bicarbonate and particularly sodium requiring supplementation. The early prevention of renal bone disease is critical and our policy is to maintain parathyroid hormone levels within the normal range using phosphate binders and activated vitamin D. Erythropoietin beta is usually given by weekly subcutaneous injection to maintain the haemoglobin within the normal range for age.

Haemodialysis

Haemodialysis, although technically challenging and labour intensive may be the only option in the infant after abdominal surgery or PD failure. In a review of 18 children age less than 2 years on HD at GOS over 16 years, problems with access were very common with a revision ratio of 40%. Poor function due to malposition/thrombosis or infection each accounted for 50% of line changes. Sixteen children

were transplanted of whom 12 had been re-established on PD, emphasising the need for both dialysis modalities to be available in specialised centres.²⁰ Although there were subsequently four deaths, none occurred on HD.

Outcome

The aim of an early intensive approach to the treatment of ESRF is to facilitate successful renal transplantation which offers the best prognosis for normal growth, development and psychosocial adjustment. With nutritional support, infants on PD can achieve significant catch-up growth in both length and head circumference with the mean SDS increasing over a year from -1.8 to -1.1 and -1.9 to -0.9 respectively in our centre.⁹ In children with severe CRF presenting up to the age of 2 years requiring dialysis before transplantation, the mean Ht SDS was -1.54 at transplant (n=30) with significant improvement in Ht SDS one year post transplant.¹⁰ Another treatment option for infants with severe CRF is recombinant human growth hormone which gives similar results to those achieved with intensive nutrition and should therefore be reserved for those infants that do not respond to enteral feeding.^{10,21} The developmental outcome for survivors of an infant dialysis programme is encouraging. Warady et al found that in 28 survivors of 34 infants dialysed age <3 months, only one was significantly delayed on testing at one year of age and of 16 children ≥5 years, 15 were attending full time school in age-appropriate classes.¹¹ In 16 surviving infants of the PD programme at GOS, 87% had developmental scores within 2 SD's of the norm (mean IQ=86.6) but 50% demonstrated borderline/abnormal psychosocial adjustment scores.²² In three infants, developmental delay could be attributed to perinatal asphyxia in two and hypothyroidism in one.⁹ Impaired cognitive and educational attainment in adults with onset of ESRD age 0-14 years born before 1979, has been reported in a Dutch study.²³ However it is difficult to separate the effect of poor school attendance due to chronic illness and frequent hospitalisation on academic achievement, from intrinsic impairment of cognitive development. Provision of psychosocial and teaching support should be an integral part of the continuing care of infants and young children with ESRF.

Conclusion

Although the care of the infant with ESRF is demanding

for families and professionals, survival in otherwise normal infants is similar to older children and prognosis for growth and development is encouraging. Improvements in the provision of renal transplantation for infants and young children now give parents reasonable hope for a relatively normal quality of life for their child. Intensive treatment of the infant with ESRD is justified in most circumstances and should now be considered the standard of care.^{12,24}

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