

Transplantation and Autosomal Recessive Polycystic Kidney Disease

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Renal transplantation is a lifesaving treatment for children with kidney failure, including children with autosomal recessive polycystic kidney disease (ARPKD). ARPKD occurs in 1 out of 20,000-40,000 live births. In addition to renal abnormalities, extra-renal manifestations lead to severe morbidity and mortality in ARPKD. In the neonatal period, pulmonary disease is the most common cause of mortality. Later in life, many patients develop liver disease, which varies in severity and onset. Previously thought to be a fatal condition, the prognosis for children with ARPKD has improved dramatically. Twenty years ago only half of the children born with the disease survived to their 10th birthday, but now that percentage has increased to 85%. Despite this improvement in outcome, there are unique issues that affect not only survival, but also the growth and development of children with ARPKD that must be considered to guide these children safely to renal transplantation.

The immediate life-threatening issue for infants with ARPKD is the severity of lung immaturity. Lung immaturity is caused in part by insufficient amniotic fluid, which is produced by the kidneys, due to the poor prenatal renal function.

Patients with ARPKD often develop kidney failure at a young age. However, immediate transplantation is often not possible because of the infant's small size. Most patients with ARPKD experience growth retardation. The infant's weight can be misleading because of the disproportionate weight of the massive kidneys, which can weigh more than 10 ounces each. For a successful renal transplant, the goal is to provide adequate nutritional support to allow the child to grow to approximately 22 pounds. At this size, the child achieves the capacity to handle an adult sized kidney thus limiting the chance of rejection

If the patient needs dialysis before they are large enough to undergo transplantation, peritoneal dialysis is typically used. During peritoneal dialysis, fluid is infused into the abdomen to remove toxins that would otherwise be filtered from the blood. This additional fluid in the abdomen can further compress the stomach, leading to reflux and poor growth. Therefore, early unilateral or bilateral nephrectomy can help patients maintain adequate nutrition and grow, allowing them to reach a sufficient size for safe renal transplantation more quickly.

Despite receiving transplants at a younger age, children with ARPKD enjoy similar survival as other kidney transplant recipients. Their grafts last for the same amount of time and they have the same number of rejection episodes as other kidney transplant recipients. Since the ability to manage the complications of kidney disease in small children has improved, children are living longer, and as a result, the liver disease associated with ARPKD is becoming more evident. Progressive scarring of the liver leads to portal vein hypertension and gastrointestinal bleeding in approximately 80% of patients who receive a renal transplant for ARPKD.

Infections are another serious concern for this transplant population. Children who had received a kidney transplant due to ARPKD are twice as likely to die from overwhelming bacterial infection than children receiving kidney transplants for other indications.

Overall, the prognosis for children with ARPKD has improved substantially during the last few decades. Early initiation of peritoneal dialysis and nephrectomy when indicated has contributed to improved patient survival. With careful attention to nutrition, sufficient growth can be achieved to allow for successful kidney transplantation. Although ARPKD kidney recipients do as well as children receiving kidneys for other reasons, some of these patients die from infection despite good kidney function. Continued improvements in transplant care will hopefully allow more children to celebrate not only their 10th birthday, but many more beyond.



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