The ESPN/ERA-EDTA Registry

ARPKD - COMBINED LIVER KIDNEY TRANSPLANTATION MIGHT NOT BE THE IDEAL STRATEGY.

Djalila Mekahli[1], Karlijn J. van Stralen[2], Kitty J. Jager, Benetti, N Godefroid, V Edvarson, JG Heaf, A Jankauskiene, Puteo, T Seeman, A Zurowska, S Marinova, Franz Schaefer[3], Jaap Groothoff[4] on behalf of the ESPN/ERA-EDTA Registry.
For more information: k.j.vanstralen@amc.uva.nl

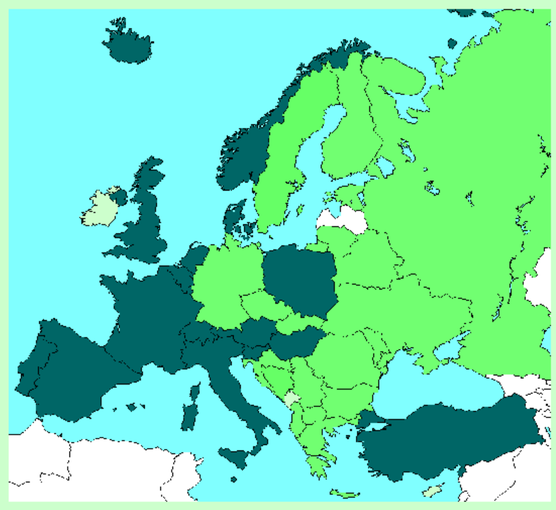
Background

- Autosomal recessive polycystic kidney disease (ARPKD) occurs in 1:20,000 live births and is the most common cystic kidney disease in childhood.
- Combined liver kidney transplantation (CLKT) has been recommended over kidney alone (KT) to benefit from a potential immunological advantage, but may be associated with increased mortality.
- We aimed to analyse the characteristics and outcomes of different transplantation strategies in children with ARPKD

Methods

- ESPN/ERA-EDTA registry
- 239 incident ARPKD patients who started RRT since 1995.
- Data included date of birth, gender, age at RRT, treatment modality of RRT, type of transplantation (KT or CLKT), changes in RRT modality during follow-up, survival and cause of death.

Countries with ARPKD patients

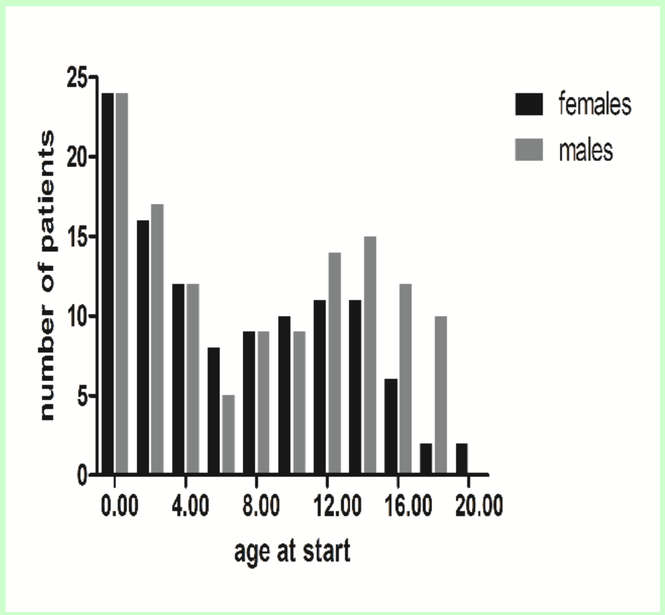


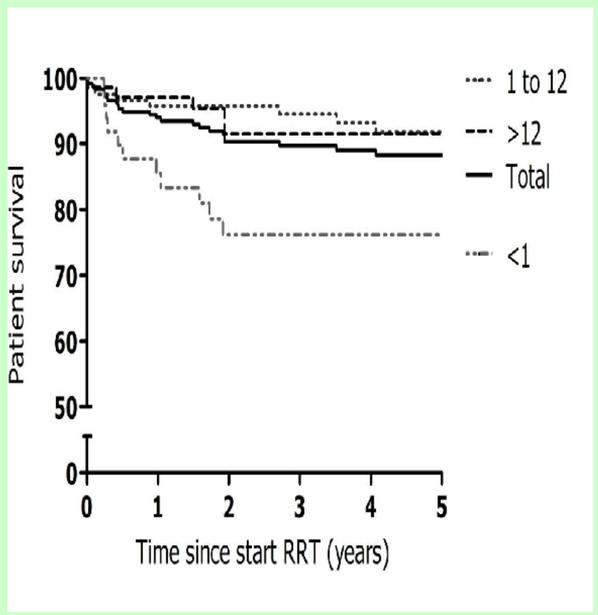
Results

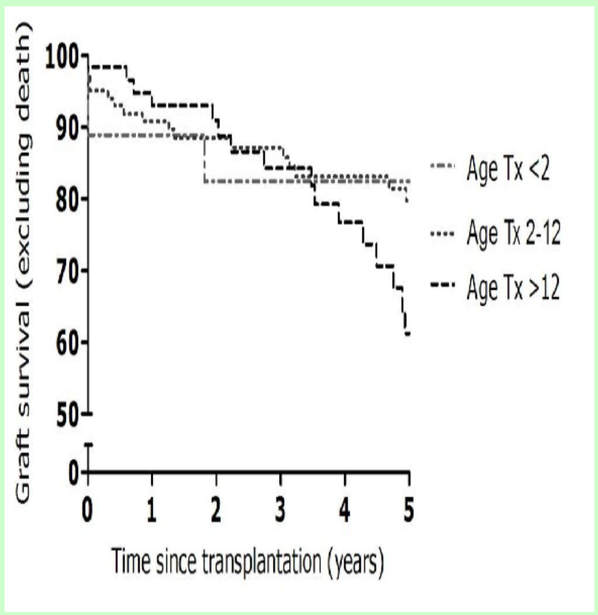
Differences between age groups

45% of the children were below 5year at start of RRT, with an equal gender distribution. Among the patients starting in puberty, there were significantly more boys. Patient survival was significantly worse among those starting before the age of 1, while there were no differences between the older age-groups. Graft survival was significantly worse among the oldest patients.

	ARPKD patients
	N=239
Age at start, median (p5-p95)	7.2 (0.01 to 17.3)
Duration of follow-up, median (p5-p95)	3.9 yrs (0.18 to 11.8)
Female gender	111 (46.4%)
RRT treatment at start	
HD	65 (27.2%)
PD	101 (42.2%)
Tx	69 (28.9%)
Dialysis/Unknown	4 (1.7%)

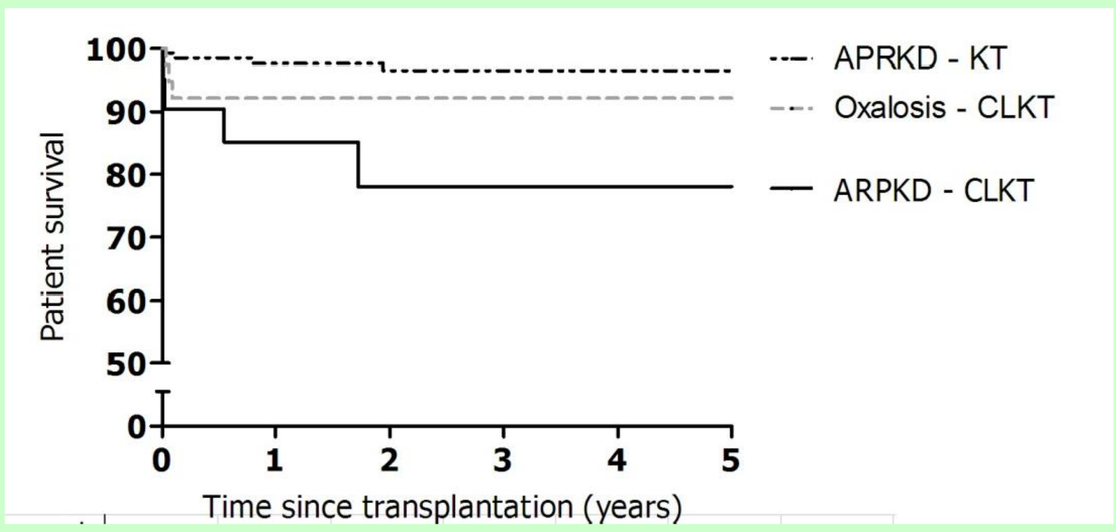


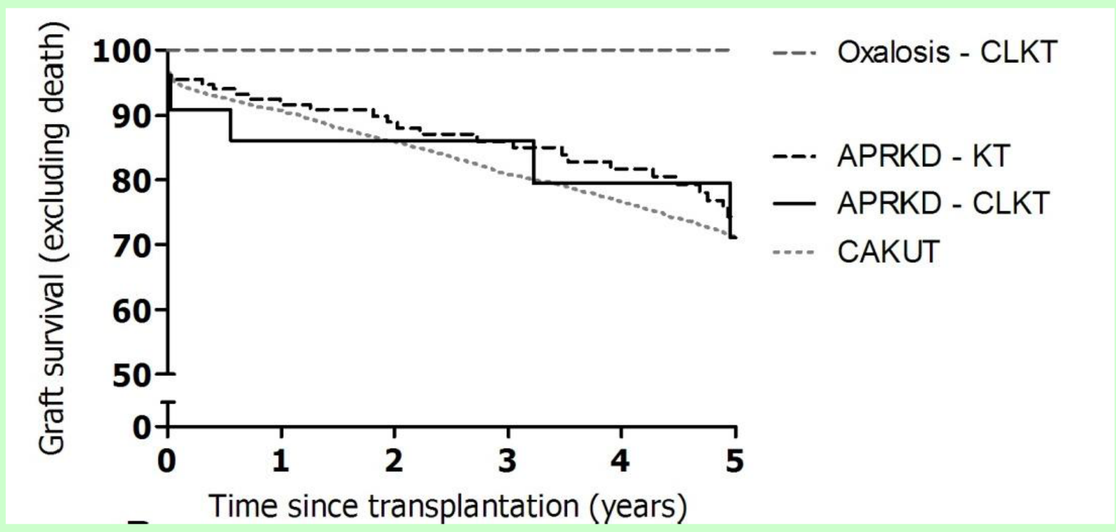




Combined liver-kidney versus kidney alone transplantation.

There was no significant difference between kidney-alone (KT) versus combined liver kidney transplantation (CLKT) with respect to graft survival (excluding death). However there was a significant difference in patient survival (including death after graft loss). Patients survival in CLKT was very similar between patients with oxalosis and ARPKD, with most deaths shortly after transplantation.





Conclusions

The need for RRT in the first year of life for ARPKD adversely impacts survival.
Combined liver-kidney transplantation is a significant risk factor for mortality on short-term, and is not associated with better 5-year graft survival. A potentially reduced risk of infections, and consequently better patient and graft survival are not well balanced against the high post-operative risk.

A limitation of our study is the lack of data on the reason for the CLKT. Therefore patients in the need for a CLKT might be in a worse condition than patients in the need for a KT only, thereby possibly explaining part of the difference in survival. However, most of the deaths seemed to be related to the transplantation itself as they occurred shortly thereafter.

Still our data suggest that whenever the CLKT is not immediately needed because of liver problems, it might be best to post-pone a liver transplantation as long as possible.

WE WOULD LIKE TO THANK ALL FOR CONTRIBUTING TO THE ESPN/ERA-EDTA REGISTRY

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