Polycystic kidney disease is a major cause of end stage renal disease in Europe. In this group of disorders fluid filled cavities and fibrotic tissue replace functional renal parenchyma resulting in progressive loss of kidney function

While Autosomal Dominant Polycystic Kidney Disease (ADPKD) is a very frequent monogenetic disease of adulthood Autosomal Recessive Polycystic Kidney Disease (ARPKD) is the rare and often severe paediatric form of cystic kidneys with an early impact on child health. Kidneys are often grossly enlarged at birth and up to 50% of the patients require renal replacement therapy in the first two decades of their life. In addition to renal disease, there is mandatory hepatic involvement. Other organs may also be affected.

The pathophysiology, clinical heterogeneity and long-term evolution of ARPKD remain poorly understood. There is currently no causative treatment and kidney-, liver- or combined liver and kidney transplantation may be required in case of renal and hepatic failure. No clinical classifications, clinical risk factors or evidence-based treatment guidelines have been established so far and experience remains sparse even in large European paediatric nephrology centers.

ARegPKD (www.aregpkd.org) is a multinational, mostly European, ARPKD registry study, which was initiated with the support of two clinical research consortia with an outstanding experience in paediatric nephrology studies, i.e. the German Paediatric Nephrology Association (GPN) and the European Study Consortium for Chronic Kidney Disorders Affecting Paediatric Patients (ESCAPE Network). In this project experienced and specialized clinical centers across Europe join forces with leading geneticists and pathologists in order to advance our understanding of ARPKD. We therefore invite the European paediatric nephrology centers to contribute to this initiative by introducing pseudonymized clinical patient data into our secure web-based database. Special interest is paid to the initial clinical presentation, pre- and perinatal history, hepatic and renal function, current treatment approaches and long-term courses as well as potential clinical overlap with other disorders presenting with cystic kidneys. Associated with the clinical database are a center for reference histology and an initiative for ARPKD-specific biobanking. Coordinators of ARegPKD are actively involved in different working groups of the European Society for Paediatric Nephrology (ESPN) thus ensuring rapid dissemination of the acquired knowledge to the involved professionals throughout Europe.

In summary, ARegPKD aims to provide an observational evidence base for unified clinical treatment concepts, to establish clinical or biochemical risk markers for disease progression and to lay the foundation for innovative translational research towards novel therapeutic targets and clinical trials for this severe disorder.

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European ARPKD registry ESPN 2014.2