Editorial

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Polycystic Kidney Disease and its Effects

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Editorial

Cystic kidneys are common causes of end-stage renal disease, both in children and in adults. Autosomal Dominant Polycystic Kidney Disease (ADPKD) and Autosomal Recessive Polycystic Kidney Disease (ARPKD) are cilia-related disorders and the two main forms of monogenic cystic kidney diseases. ADPKD is a common disease that mostly presents in adults, whereas ARPKD is a rarer and often more severe form of Polycystic Kidney Disease (PKD) that usually presents perinatally or in early childhood. Cell biological and clinical research approaches have expanded our knowledge of the pathogenesis of ADPKD and ARPKD and revealed some mechanistic overlap between them.

In patients with Polycystic Kidney Diseases (PKDs), the kidneys contain multiple fluid-filled cysts, although other organs may also be affected. Although PKD is inherited monogenically, it is phenotypically, genically and allelically heterogeneous. Autosomal Dominant PKD (ADPKD) is the most common form of PKD and is generally an adult-onset, multisystem disorder that is characterized by gradually growing renal cysts that start to develop in utero and can originate from all areas of the kidneys, although cysts usually form in the distal regions of the nephron and the collecting duct. Progressive fibrocystic renal disease in ADPKD is often accompanied by hepatobiliary changes or other extra renal abnormalities, such as intracranial arterial aneurysms.

To ascertain the exact prevalence of a hereditary disease, several factors must be considered the geographic and ethnic composition and the size of the population, the choice and mode of calculation of epidemiologic measurement, the screening policies and other characteristics of the health-care system, the disease definition and diagnostic criteria that are used, the sources of ascertainment and possible causes of under-ascertainment and the period of time during which events are counted (ascertainment period). Thus, variability in study design and ascertainment might partially explain the large differences in estimates of the prevalence of ADPKD that have been reported in some studies. For example, two clinical surveys in Europe.

Early in the course of ADPKD when renal function is still normal, most patients already develop arterial hypertension that contributes considerably to the increased cardiovascular morbidity and mortality observed in patients with ADPKD. Although various pathogenic mechanisms are known and activation of the renin–angiotensin–aldosterone system (RAAS; a hormone system that regulates blood pressure and fluid balance) is clearly the most prominent, other mechanisms have also been described, including increased activity of the sympathetic nervous system and disturbances in the fine-tuning of vascular tone through the action of endothelin, Nitric Oxide (NO) and intracellular calcium.

Epithelial cell proliferation, abnormal fluid secretion and excessive extracellular matrix deposition are the major characteristics of cystic epithelial cells. These changes are accompanied by alterations in the pericystic blood and lymphatic microvas-culature. Most cysts detach from the tubules from which they form and fill with fluid by transepithelial secretion. Cyst enlargement also compresses the surrounding nephrons, interstitium and vasculature. The obstructed nephrons eventually form atubular glomeruli and apoptotic proximal tubules.

These events are accompanied by the production of chemokines, cytokines and growth factors by epithelial cells, interstitial fibroblasts and inflammatory cells, such as macrophages. Abnormal cytokine-mediated crosstalk between epithelial and inflammatory cells promotes more inflammation and fibrosis, new cyst formation and disease progression, which result in massively fibrotic kidneys at end-stage disease. Increased inflammation is probably an early event in disease progression, and the selective depletion of macrophages in the kidneys resulted in considerable amelioration of the cystic phenotype and improvement in renal function

n the personalized medicine approach, it is no longer sufficient to treat the disease instead, clinicians must treat their patients. Consequently, disease management paradigms should include biological, physical and psychological assessments, which seems to be particularly important for patients with multifaceted diseases such as ADPKD and further underscores the importance of including patient-reported outcomes in routine clinical care. In parallel, sensitive and appropriate kidney disease measures should be developed alongside other established tools to assess the burden of ADPKD in an individualized, personalized manner. Health-Related QOL (HRQOL), which encompasses physical and mental health, social issues as well as pain and discomfort, facilitates patient-centered care and encourages shared decision-making among medical professionals and patients. The specific issues of QOL

have not been systemically studied in patients with ARPKD or their families accordingly, we focus on only ADPKD.