

Targeting chloride transport in autosomal dominant polycystic kidney disease

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent inherited kidney disease. Transepithelial fluid secretion is one of the key factors of cystogenesis in ADPKD. Multiple studies have suggested that fluid secretion across ADPKD cyst-lining cells is driven by the secretion of chloride, essentially mediated by the CFTR channel and stimulated by increased intracellular levels of 3',5'-cyclic adenosine monophosphate. This review focuses on the pathophysiology of fluid secretion in ADPKD based on the pioneering studies of Jared Grantham and colleagues, and on the follow-up investigations from the molecular level to the potential applications in ADPKD patients. Altogether, the studies of fluid and chloride transport in ADPKD paved the way for innovative therapeutic targets to prevent cyst volume expansion and thus, kidney disease progression.