KSN 2019
Seoul, Korea · May 23 - 26

Abstract Type : Oral
Abstract Submission No. : OR-1419

Clinical characteristics and outcomes in patients with autosomal dominant polycystic kidney disease on renal replacement therapy

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Objectives: Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent inherited disorder of the kidney and accounts for up to 10% of patients in end stage of renal disease (ESRD). However, approaches to the prevention and treatment of ADPKD vary widely and there is no established treatment to slow disease progression. In addition, there are few reports about clinical outcomes in ADPKD patients on renal replacement therapy (RRT).

Methods:

We retrospectively analyzed 288 patients with ADPKD at Keimyung university Dongsan medical center between January 1989 and August 2018. We investigated risk factors for disease progression and survival rates of patients receiving RRT.

Results: The mean age at diagnosis was 43.1 ± 14.1 years and 146 patients (50.7%) were male. Among 288 patients, 197 patients (68.4%) had hypertension and 11 patients (3.8%) had cerebral aneurysm. During the follow-up period (117.1 ± 102.1 months), 28 patients (9.7%) were dead. Most common cause of mortality was infection (42.9%), followed by sudden cardiac death (25.0%). One hundred thirty-two patients (45.8%) progressed to ESRD and 104 patients (36.1%) received RRT. The 10-year survival rate of patient and kidney was 80.9% and 66.3%. The overall survival of patients with kidney transplantation was significantly longer than patients receiving other modalities of RRT by the Kaplan-Meier method (p = 0.001). In multivariable analysis, the independent risk factors for ESRD were age at diagnosis (OR, 1.038; 95% CI, 1.018-1.058; p < 0.001), hypertension (OR, 2.267; 95% CI, 1.283-4.008; p = 0.005) and cyst infection (OR, 4.362; 95% CI, 1.712-11.112; p = 0.002).

Conclusions:

Age at diagnosis, hypertension and cystic infection were independent risk factors of poor renal prognosis in ADPKD patients. To prevent progression to ESRD in ADPKD patients, early diagnosis and strict blood pressure control are important. Kidney transplantation can be preferred option for ADPKD patients receiving RRT.

Patient survival rate according to RRT modality

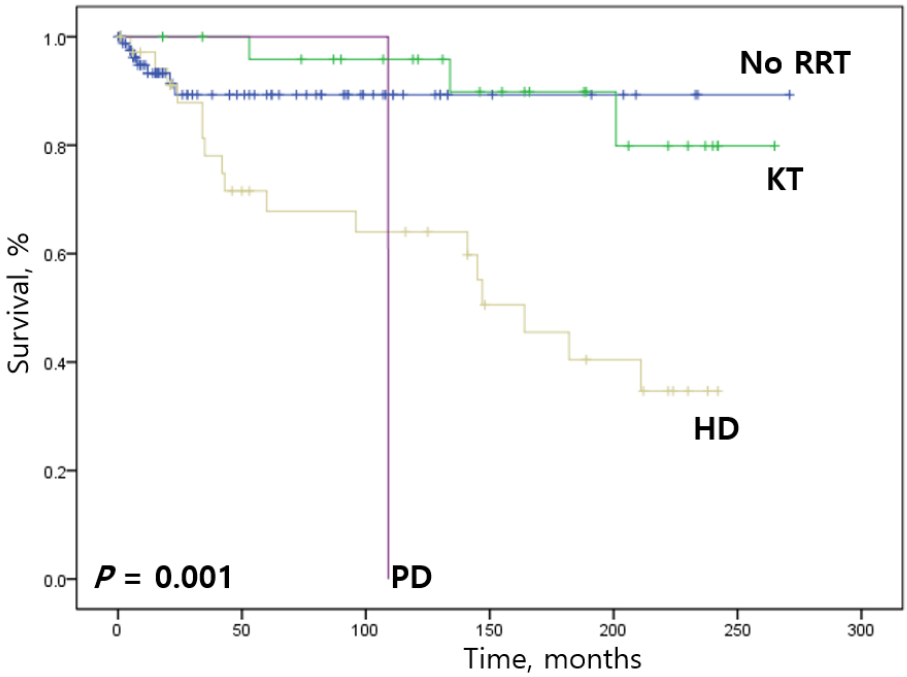

Figure 1. Patient survival rate according to RRT modality