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Kidney disease progression and clinical outcomes in patients with autosomal dominant polycystic kidney disease

Seong SIK[1,2], Ha yeon PARK[1], Sang mok YEO[1], Woo YEONG[1,2], Kyubok JIN[1,2], Sung BAE[1,2], *Seungyeup HAN[1,2]

[1]Department of Internal Medicine, Keimyung University School of Medicine, Korea,South, [2]Keimyung University Kidney Institute, Korea,South

Objectives : Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary kidney disease in patients with renal replacement therapy (RRT). However, there is no established treatment to prevent the progression of ADPKD. In addition, there are few reports of kidney disease progression and clinical outcomes in ADPKD patients on RRT.

Methods : We retrospectively reviewed the medical records of 253 patients with ADPKD at Keimyung university Dongsan medical center between January 1989 and January 2017. We investigated disease progression risk factors and survival rates in ADPKD patients receiving RRT.

Results : The mean age at diagnosis was 43.2 ± 13.2 years (range, 10-75). Males were 129 (51%) and patients with a family history of ADPKD were 93 (37%). Average duration of follow-up was 76 ± 74.5 months (range, 1-280). Among the 253 patients, 125 (49%) progressed to CKD stage 5 and 100 (40%) received RRT. The mean duration from diagnosis to receiving RRT was 8.8 ± 7.8 years (range, 0-32). In a multivariate analysis, hypertension (HR 2.154, p=.016), cardiovascular disease (HR 4.325, p=.023), cyst infection (HR 5.317, p<.001) were independent risk factors for progression to end-stage renal disease (ESRD). The number of patient receiving hemodialysis, peritoneal dialysis, and kidney transplantation (KT) was 72, 2, and 26, respectively. Twelve (46%) of the 26 KT recipients (KTRs) were treated prior to transplantation. Four underwent simultaneous bilateral nephrectomy and 8 underwent renal artery embolization. The 5-year survival rate of KTRs was 96 ± 3.9%, which was higher than that of 82 ± 5.2% in hemodialysis patients (p=.040). The 5-year survival rate of allograft kidney was 88 ± 8.1%.

Conclusions : Hypertension, cardiovascular disease, and cyst infection were independent risk factors for kidney disease progression in ADPKD. In order to slow the progression to ESRD, best efforts to properly manage hypertension and cardiovascular disease are required. Nevertheless, when progressing to ESRD, KT can be recommended as treatment of choice.

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