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Index Cases Of Intracranial Aneurysms In Autosomal Dominant Polycystic Kidney Disease: Longitudinal Experience From A Single Renal Transplantation Centre

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Introduction: The prevalence of intracranial aneurysms (ICAs) is higher in patients with autosomal dominant polycystic kidney disease (ADPKD), with suggestion that ICAs behave differently in ADPKD.

Methods: A retrospective review of prospectively collected clinical data for all patients who underwent nephrectomy/ies between 01/01/1995 and 31/12/2021 at CNARTS. Cases of ICAs were reviewed for clinical history, mode of diagnosis, patient age at diagnosis, family history and significant risk factors, temporal relationship between diagnosis and nephrectomy/renal transplantation.

Results: 111 nephrectomies were undertaken in 81 individual patients who had ongoing follow up. 49 patients had ADPKD and 32 had another primary nephrological diagnosis. There was no significant differences for gender and age at follow up, but ADPKD patients were more likely to have hypertension. 30 ADPKD patients and 15 non-ADPKD patients had neuroimaging.Seven of 49 ADPKD patients had ICAs (14.3% of all patients, 23.3% of those with neuroimaging), compared to two of 32 non-ADPKD patients (6.25% of all patients, 13.3% of those with neuroimaging). The mean age of ADPKD patients with ICA(s) was 48.9±12.9 years at time of diagnoses; all had hypertension. Six did not meet current guidelines criteria for ICA screening. Five had no known family history of ICA. Three patients suffered aneurysmal rupture, at a mean age of 39 years. Aneurysmal rupture was known to involve smaller ICAs in two of these cases. Cases demonstrated detectable vascular changes on early neuroimaging, hypoplastic anatomical variants, aneurysmal growth, de novo ICA formation, and association with other vascular abnormalities.

Conclusion: Early detection of ICA and pre-aneurysmal changes is possible. Our data confirmed risk for aneurysmal rupture at a younger age and with smaller aneurysms: this requires further investigation and clinician vigilance. It is vital for a multidisciplinary, patient-centred approach of ICA screening, surveillance and management for ADPKD patients and their families.