Understanding the clinical course and complications in adults with ADPKD

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INTRODUCTION:
Autosomal dominant polycystic kidney disease (ADPKD) is a multi-system disease characterised by a gradual progressive decline in kidney function; many affected patients will reach ESRF and require transplantation and/or dialysis. The clinical phenotype is heterogenous, with wide variation in renal and extra renal complications, including after ESRF. We sought to describe the frequency of renal and extra renal complications in our ESRF ADPKD cohort to help identify potential opportunities for research and service improvement.

METHODS:
All adult patients with a diagnosis of ADPKD with ESRF were identified from our tertiary centre renal database, Renalware, between 1996 and 2018. Renalware is a comprehensive clinical database with high data validity. Data extracted include cohort demographics and the frequency of complications including recurrent UTI, intracerebral aneurysm, renal cell carcinoma, nephrectomy, hypertension, diabetes & CVD (coronary artery disease, myocardial infarction, valvular disease, cardiomyopathies, aortic dissection and peripheral vascular disease). Descriptive statistics were used to report the frequency of co-morbidities and complications related to ADPKD.

RESULTS:
178 patients were identified as reaching ESRF requiring long-term RRT. During follow-up, 36.5\% of patients died; the mean age of ESRF was 54 (34-89) years. The median age of death was 68 years. The most frequent coded cause of death was withdrawal from life-supporting treatment (16.9\%). Cardiovascular comorbidity was common - hypertension (97.2\%) and cardiovascular disease (34.3\%). Diabetes mellitus was recorded in 16.9\%, intracerebral aneurysms were detected in 16.3\%.

Records showed 69.1\% of ESRF patients were prescribed ACEi or ARBs as part of their cardiovascular risk management.

Recurrent UTI (10.7\%) and pyelonephritis (6.2\%) were common; renal cell carcinoma was diagnosed in 2.2\%. 38/178 patients (21.3\%) had undergone nephrectomy. Of these, 20/38 patients had a pre-transplant nephrectomy, 6/38 nephrectomy at the time of kidney transplantation and 9/38 patients had a post-transplant nephrectomy. The timing of nephrectomy in relation to transplantation was not known for 3 patients.

The most common indication for native nephrectomy pre-transplant was mass effect related to enlarged kidneys (35.0\%), followed by urine infections including recurrent pyelonephritis (40.0\%). The most common reason for peri-transplant nephrectomy was as part of simultaneous liver transplant (50\%). The most commonly observed reason for post-nephrectomy was renal cell carcinoma (22.2\%), followed by infection (11.1\%) and transplant failure (11.1\%). In 4 cases the data was not available.

DISCUSSION:
Cardiovascular co-morbidity is common in patients with ESRF and ADPKD as expected. Nephrectomy in our cohort was fairly prevalent - primarily in relation to preparation for transplantation.
We have developed a specialist clinic for patients with ADPKD. In addition, in conjunction with colleagues at the local transplanting centre, we have developed a specialist MDT involving urology, transplant surgeons, nephrology, radiology and pain colleagues to optimise holistic care for complex ADPKD patients.