Portuguese familial amyloidotic polyneuropathy (FAP) type I is a systemic amyloidotic disease due to an amyloidogenic transthyretin (TTR) protein, in which an amino acid substitution of methionine for valine at position 30 of TTR molecule is present. Although peripheral nervous system is primarily affected, renal involvement is common. In this disease, all patients had amyloid deposits in the kidney, but only one third will develop CKD and 10% will progress to stage 5.

The aims of this study was to evaluate the incidence of renal dysfunction post OLT and its impact on patient’s survival.

RESULTS

185 OLT recipients:
- Male gender 59%
- Mean age: 36.8±9.5 years
- Diabetes in 2 patients; Hypertension in 17 patients
- Mean follow up time 3.6±3.7 years, 28.6% > to 5 years

AKI n=57 (26.3%)

Defined by RIFLE criteria

RRT in 28% (n=16)
Death in 23% (n=13)
Renal recovery in 3.5% (n=2)

AKI pos OLT: more common in women, and associated with acute RRT necessity, CKD development, and high mortality.

In multivariate analysis, adjusting for age and for presence of renal dysfunction pre OLT, AKI was correlated with development of CKD.

AKI post OLT, was correlated with development of CKD post OLT.

CKD n=75 (34.6%)

CKD stage 3 in 23.5% (n=51)
CKD stage 4 in 6% (n=13)
CKD stage 5 in 5% (n=11)

Age, renal dysfunction pre OLT and AKI post OLT were risk factors for CKD development in all the 3 stages. Tacrolimus seemed for age, gender.

Mortality n=32 (14.7%),
Mean follow up time 1.8±3.2 years

Mortality was associated with age, retransplantation, and with any kind of renal dysfunction namely renal dysfunction pre OLT, AKI post OLT (specially F class), and CKD development (particularly stage 5).

CONCLUSIONS

These results demonstrate that renal complications are important prognostic tools in FAP patients submitted to OLT. Careful assessment of pre transplant renal function is essential. Combined liver kidney transplantation should be proposed for patients at higher risk of renal dysfunction post transplant.