Rapidly progressive glomerulonephritis (RPGN) is an important cause of morbidity and mortality in ANCA- associated vasculitis (AAV). Studies have specifically addressed the outcome of patients presenting with severe renal injury, traditionally defined as dialysis dependence or a serum creatinine of over 500 micromoles/l and in particular the benefit of plasma exchange. Recent research has challenged the benefit of plasma exchange in this patient set. Here we describe long term patient and renal outcomes in a large cohort of patients treated consistently with a regime including plasma exchange at a single centre over 40 years.

Methods

This is a retrospective analysis of patients treated at our centre between 1977 and 2017 with newly presenting ANCA associated vasculitis and severe renal dysfunction. Patients were classified as being independent of dialysis but with a serum creatinine concentration ≥ 500 µmol/L or an estimated glomerular filtration rate of less than 15 ml/minute or dialysis dependent. Dialysis-dependent renal failure was defined as the need for dialysis within 72 hours of admission to the hospital. Patients with a concomitant anti-glomerular basement membrane (GBM) antibody were excluded. Clinical, laboratory and histopathological data were collated. Patients were treated consistently with steroids, cyclophosphamide, plasma exchange (+/- rituximab post 2011) and azathioprine as first line maintenance.

Results:

Data were obtained for 181 patients. The 149 patients who presented with dialysis dependence, had a one year survival of 71% and a five year survival of 50%. At 1 year, 58% of these patients had regained independent renal function. There was no significant difference in terms of demographics or ANCA type when comparing the patients who came off dialysis with those who remained on. There was also no significant increase in the proportion of patients with a biopsy classification of sclerotic disease in the group who did not regain function. At 5 years, 48% had independent renal function. Patient survival was 71% and 50 % at 1 and 5 years respectively. Both renal and patient survival were better in the dialysis independent patients (please see figure 1). There was a non significant trend to improved renal and patient outcomes in patients presenting after 2000.

Conclusion:

This a large cohort of patients with severe renal dysfunction with a treatment regime including plasma exchange at a single centre. Both renal and patient outcomes were comparable with other studies. Histological classification was not of prognostic value in this cohort.