AN EPIDEMIOLOGICAL STUDY ANALYSING SURVIVAL RATES AMONGST ANCA NEGATIVE PATIENTS

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Introduction

The introduction of ANCA assays has improved the diagnosis of small vessel vasculitis (SVV). Literature suggests that approximately 10% of patients with pauci-immune glomerulonephritis (PIGN) are ANCA negative[1]. Information regarding outcomes of ANCA negative patients is limited. Eisenberger (2005) *et al,* found that ANCA negative vasculitis had a comparable prognosis to that of ANCA positive vasculitis. However, specific renal and patient survival rates of ANCA negative vasculitis is sparse.

Objectives

The objectives of this study were to analyse the outcomes of ANCA negative vasculitis by;

- Analysing demographic variables between the two groups.
- Comparing patient survival of ANCA positive and negative patients
- Comparing renal survival of ANCA positive and negative patients
- Comparing cumulative cyclophosphamide (CYP) doses for each group.

Methods

All patients with renal biopsy confirmed PIGN, within a regional centre, between 1988 and 2010 were included in this study. Renal and patient survival outcomes were compared between those with ANCA positive and ANCA negative SVV. Furthermore, the cumulative CYP doses, for each ANCA status, was calculated.

Results

A total of 314 patients were identified, of which 238 patients had a recorded ANCA status. Overall, 163 (68%) patients were ANCA positive and 75 (32%) were ANCA negative. Both sexes were as likely to develop PIGN as each other.

Both ANCA negative and ANCA positive patients had comparable level of renal injury at time of diagnosis; however, the age at presentation was significantly lower in the ANCA negative group. The cumulative CYP dose did not differ significantly between the two groups. (Table-1).

Table-1. Patient demographics, clinical information and survival rates

	ANCA Positive (n= 163)	ANCA Negative (n= 75)	P Value
Male:Female Ratio	88:75	45:30	0.4027
Age at Diagnosis	60	50	<0.0001
Presenting Creatinine (µmol/L)	483.62	527.04	0.4418
Average CYP Dose (mg)	4231	3469	0.2935
Patient survival			
5 Year	70%	66%	0.5026
10 Year	52%	52%	0.5488
Renal survival			
5 Year	59%	43%	0.0261
10 Year	41%	30%	0.0352
Renal survival (death censored			
5 Year	65%	51%	0.0261
10 Year	56%	40%	0.0209

Figure-1. A comparison of renal survival rates

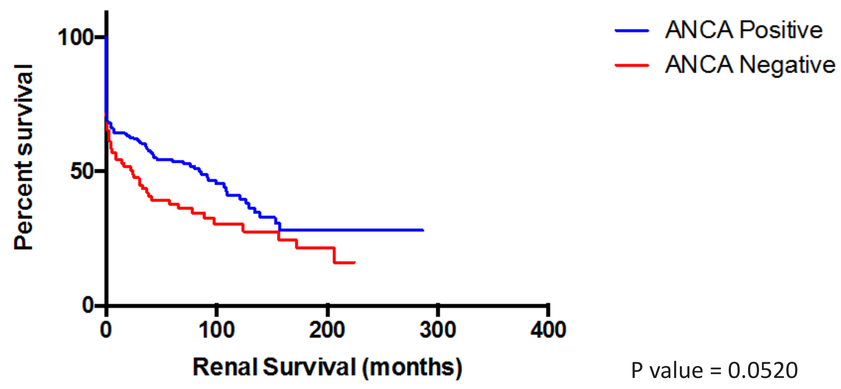
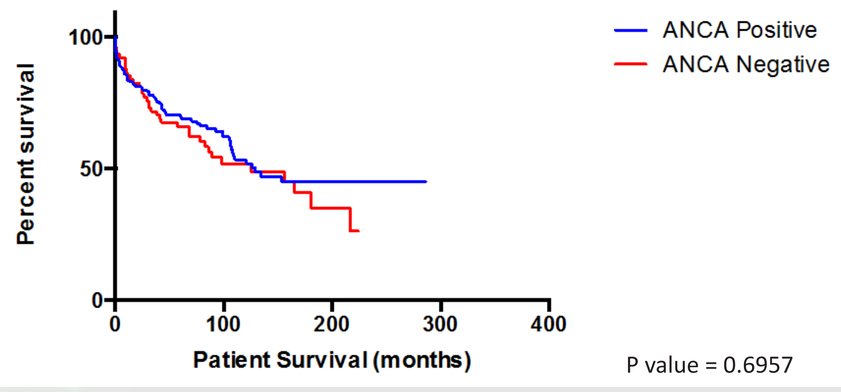


Figure-2. A comparison of patient survival rates



Discussion

A high percentage of ANCA Negative Vasculitis were identified compared with current literature. The incidence of ANCA negative vasculitis remained stable (at approximately 30%) throughout the 22 years, despite using the most up-to-date assays available.

Overall, ANCA negative patients trended towards worse renal survival rates, when compared with ANCA positive vasculitis. However, the patient survival rates were comparable.

ANCA negative vasculitis less commonly involves extra-renal organs[1]. The non-specific symptoms of early renal disease (e.g. lethargy) could explain late presentations.

Despite differences in the age at presentation, the cumulative CYP dose for each group was similar. The protocol used (table-2) to determine the dose of CYP given to patients results in older patients receiving less CYP. As our data shows the younger patient group (ANCA negative) received less CYP, it can be inferred we are under treating ANCA negative vasculitis. An uncertainty regarding diagnosis and no disease activity markers could explain the reason for the lower dose.

Table-2. CYP dosing protocol

Age (years)	Creatinine <300 µmol/L	Creatinine >300 µmol/L
<60	15mg/kg/pulse	12.5mg/kg/pulse
60-70	12.5mg/kg/pulse	10mg/kg/pulse
>70	10mg/kg/pulse	7.5mg/kg/pulse

Conclusions

Our study suggests,

1. ANCA negative patients present earlier in life than their ANCA positive counterparts.
2. ANCA negative vasculitis has a noticeably higher incidence in the regional centre studied, when compared to current literature[1].
3. Despite a similar standard of immunosuppressive treatment, ANCA negative patients have a poorer long-term renal survival.

References:

1. Harris AA, Falk RJ, Jennette JC: Crescentic glomerulonephritis with a paucity of glomerular immunoglobulin localization. *Am J Kidney Dis* **32** :179 –184, 1998
2. Eisenberger U, Fakhouri F, Vanhille P, Beaufils H, Mahr A, Guillevin L, Lesavre P, Noel LH: ANCA-negative pauci immune renal vasculitis: Histology and outcome. *Nephrol Dial Transplant* 20: 1392-1399, 2005

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