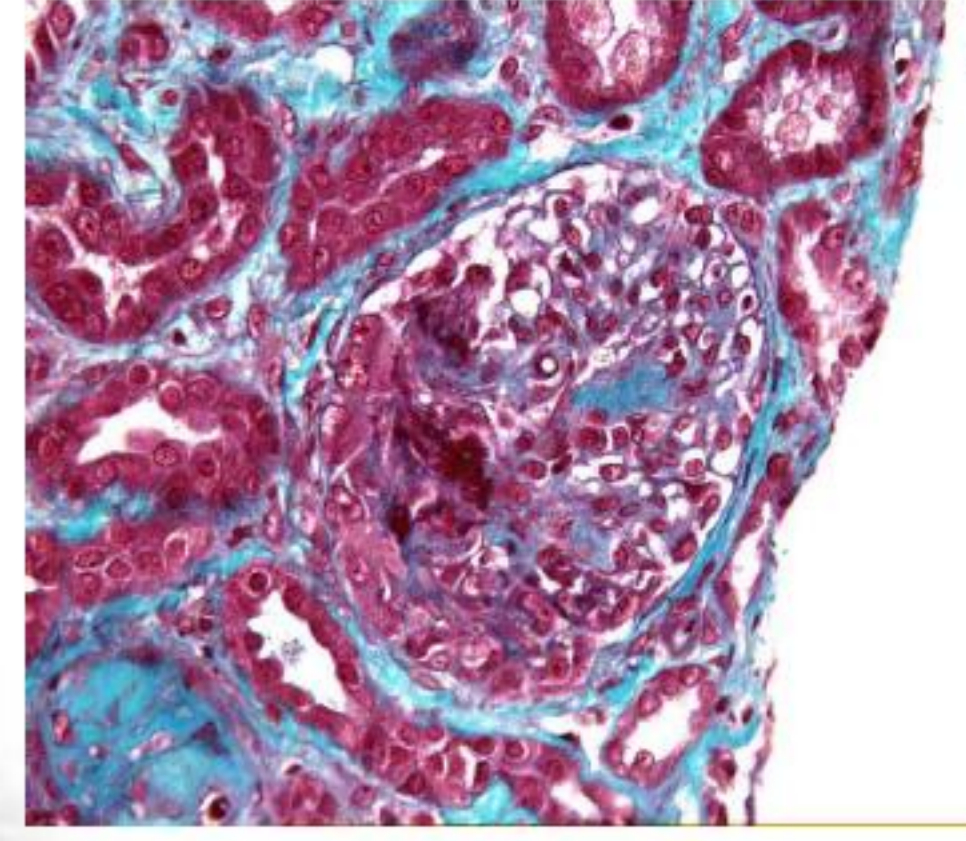


# ANTINEUTROPHIL CYTOPLASMIC AUTOANTIBODY-NEGATIVE SMALL VESSEL VASCULITIS WITH RENAL INVOLVEMENT: A 25-YEAR RETROSPECTIVE STUDY

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## INTRODUCTION

- Pauci-immune renal vasculitis with focal glomerular necrosis and crescent formation is usually associated with antineutrophil cytoplasmic antibodies (ANCA). However, ANCA's are absent in 10 to 33% of cases.
- The prognosis of small vessel vasculitis depends on the precocity of the effectiveness and complications of immunosuppressive therapy.
- Prevalence, clinical manifestations, histopathology and outcomes of ANCA-negative small vessel vasculitis remain controversial.
- We compare a group of ANCA-positive patients versus a group of ANCA-negative patients with vasculitis with renal involvement.

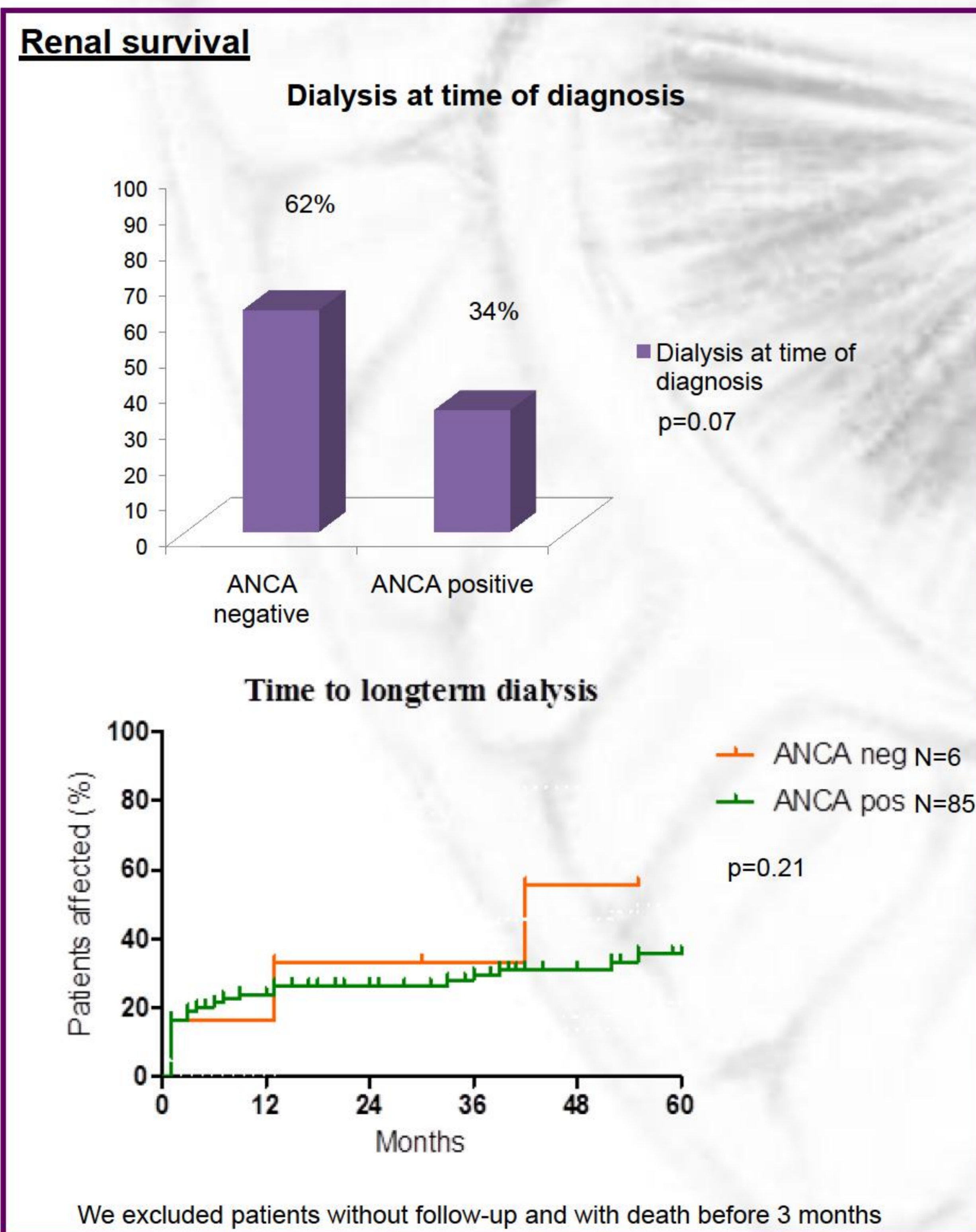
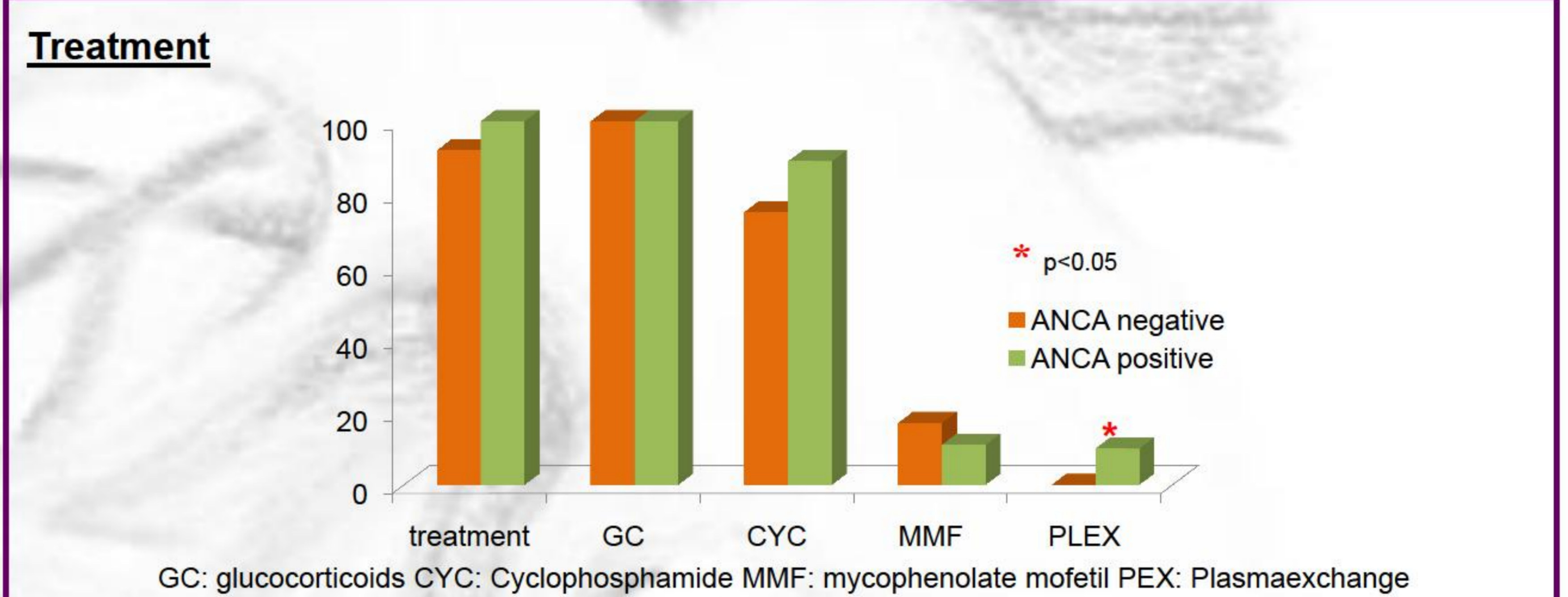
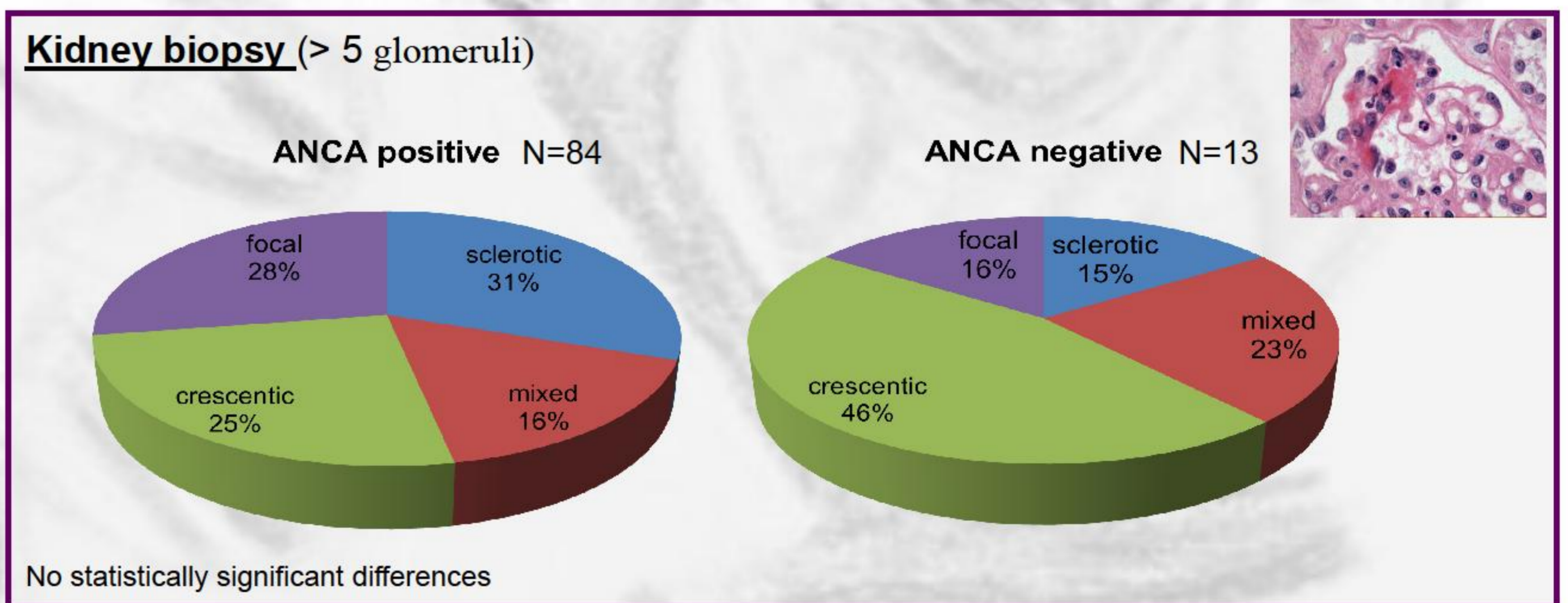
## METHODS

- Retrospective analysis of 128 cases of small vessel vasculitis with renal involvement assessed between 1985 and 2010 in two Spanish centers (Fundació Puigvert (N=104) and Germans Trias i Pujol (N=24)).
- Clinical and laboratory variables, presence and type of ANCA, kidney biopsy, immunosuppression therapy and renal/patient survival were evaluated.
- Proportions of patients were compared using Fisher's exact and serum creatinine by a t-student test. Patient and renal survival was analysed using Kaplan-Meier survival analysis.

## RESULTS

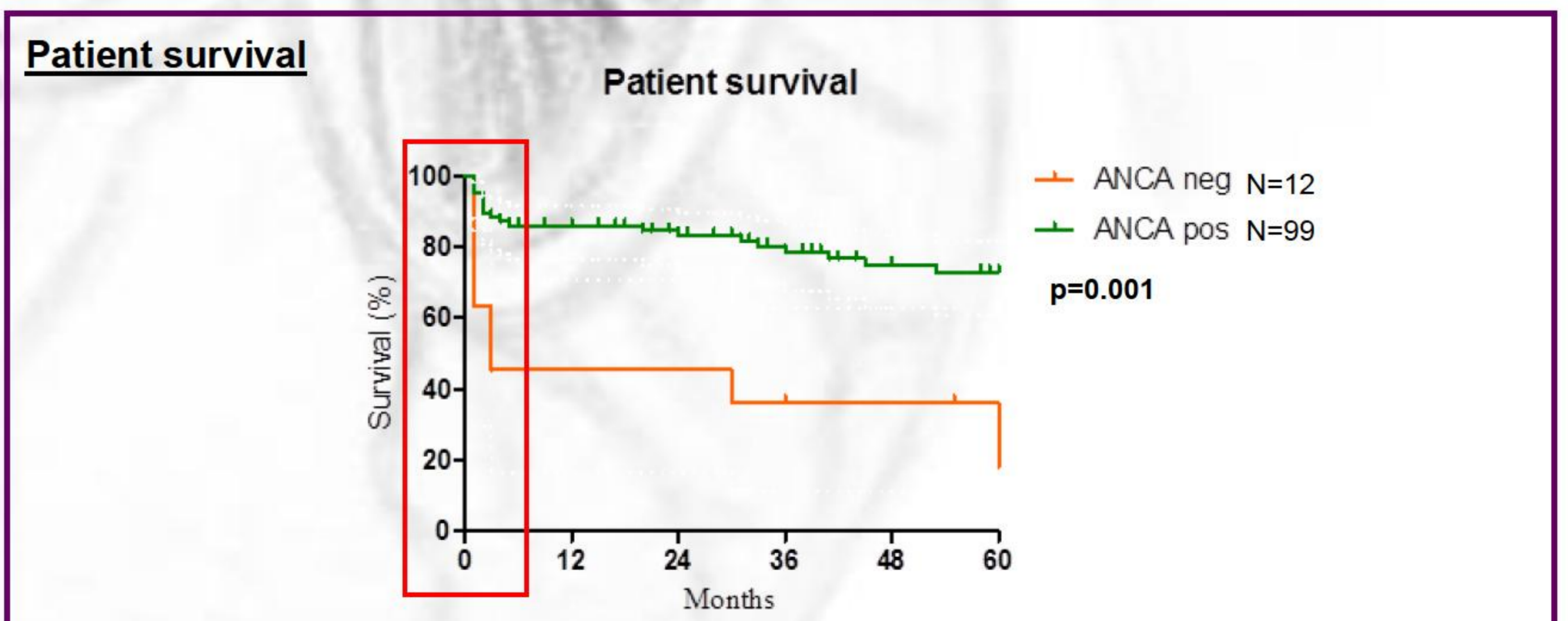
Patient characteristics			
	ANCA-positive 113/128 (88%)	ANCA-negative 15/128 (12%)	
Mean age (years)	63.78 +/-1.33	70.87 +/-2.37	0.09
<b>Diagnosis</b>			
MPA	104 (92%)	15 (100%)	0.59
GPA	9 (8%)		
<b>ANCA positive</b>			
MPO	94 (83%)		
PR3	16 (14%)		
MPO+PR3	3 (3%)		
<b>Other manifestations</b>	2/14 (14%)	26/102 (25%)	0.51
<b>Mean creatinine (µmol/L) at time of diagnosis</b>	422.07 +/-22.04	541 +/-102.1	0.09
<b>Time between clinic symptoms and diagnosis (days)</b>	42 +/-37 (N=67)	59 +/-11 (N=12)	0.09
<b>Median follow-up (months) (N=104)</b>	60.1 +/-5.06 (N=99)	29.6 +/-5.06 (N=12)	<b>0.04</b>

MPA: Microscopic Polyangiitis GPA: Granulomatosis with polyangiitis ( Wegener )



**Follow-up**

	ANCA-positive	ANCA-negative	
<b>Infections</b>			
% patients	47/105 (44%)	4/13 (30%)	0.38
No. events	56	4	
	8 opportunistic	1 opportunistic	
<b>Relapse</b>	21/105 (20%)	0%	0.12
<b>Deaths At 6months</b>	27/99 (27%) 12/99 (12%)	9/13 (69%) 6/13 (46%)	<b>0.004</b> <b>0.006</b>



## CONCLUSIONS

- In our cohort of patients with ANCA-negative vasculitis (12%), they had tendency to higher creatinine levels and higher tendency to require treatment with dialysis at diagnosis.
- The increase in mortality highlights the importance of early diagnosis and treatment of this disease despite the ANCA-negative to improve renal and patient survival.

