



## FACTORS ASSOCIATED WITH RENAL AND PATIENT SURVIVAL IN WEGENER GRANULOMATOSIS

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**OBJECTIVE:** Wegener granulomatosis (WG), is a rare multisystem autoimmune disease of unknown etiology. WG, mainly affects blood vessels in the nose, sinuses, ears, lungs, and kidneys. Renal disease is present in 17% of patients at initial diagnosis and is usually asymptomatic. Renal failure occurs in 11% at presentation. More than 50% of patients with WG recover renal function and are able to become dialysis independent with aggressive therapy for active disease. Our aim was to investigate demographic, clinic and laboratory factors related to renal and patient survival in WG.

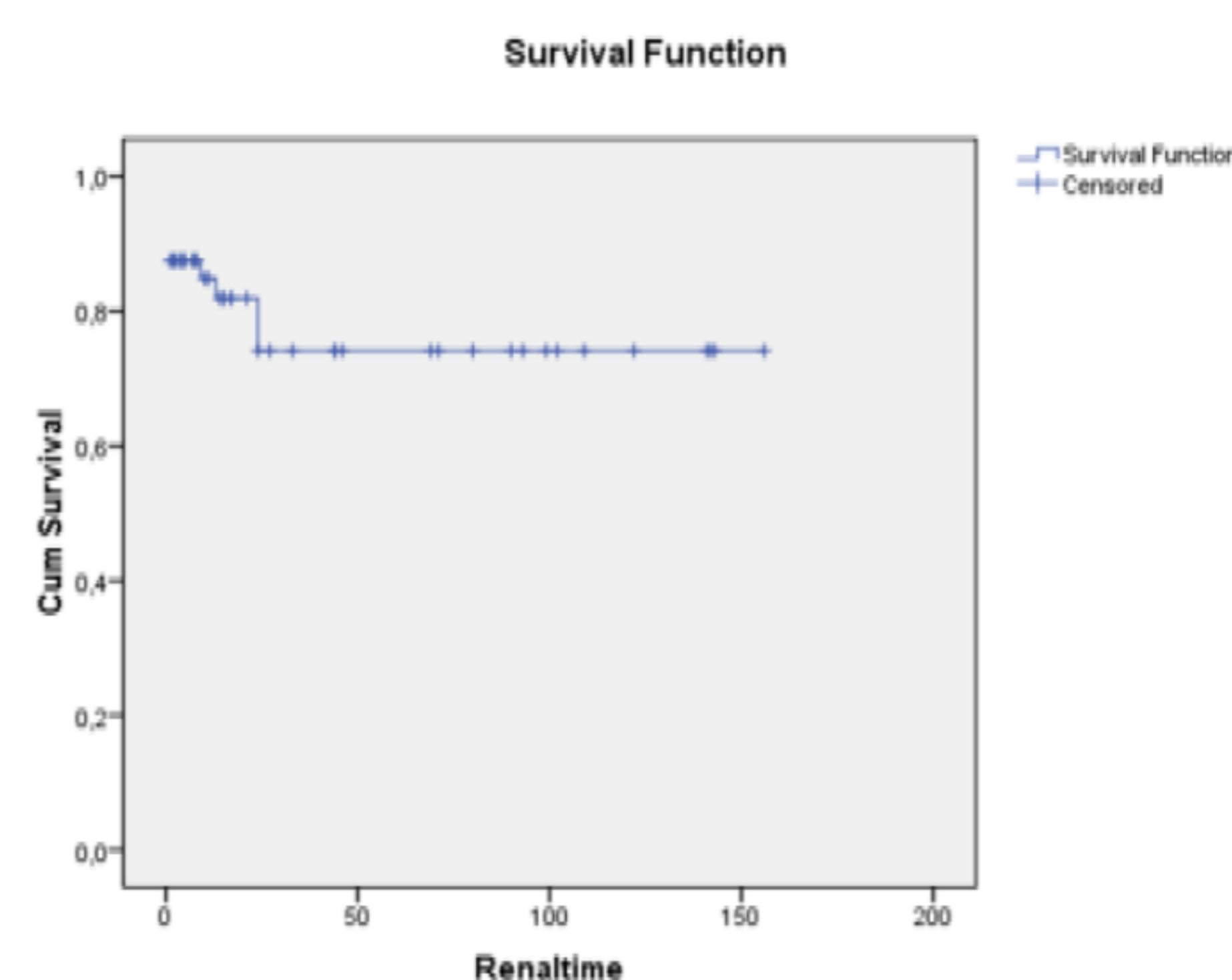


Figure 1. Renal survival in all patients.

**METHODS:** Patients who were diagnosed with WG, between January 2000 and January 2014, were examined retrospectively based on the registry files. Age, sex, time between onset of symptoms and diagnosis, disease activity (BVAS) and damage (VDI) scores, cause of death, renal and patient survival were investigated. Endpoint for renal survival was defined as being on hemodialysis (HD). Symptoms, physical findings, urine analysis, whole blood count, erythrocyte sedimentation rate (ESR), 24 hour proteinuria, glomerular filtration rate (GFR), anti-neutrophil cytoplasmic antibody (ANCA), anti-proteinase-3 and myeloperoxidase results were recorded.

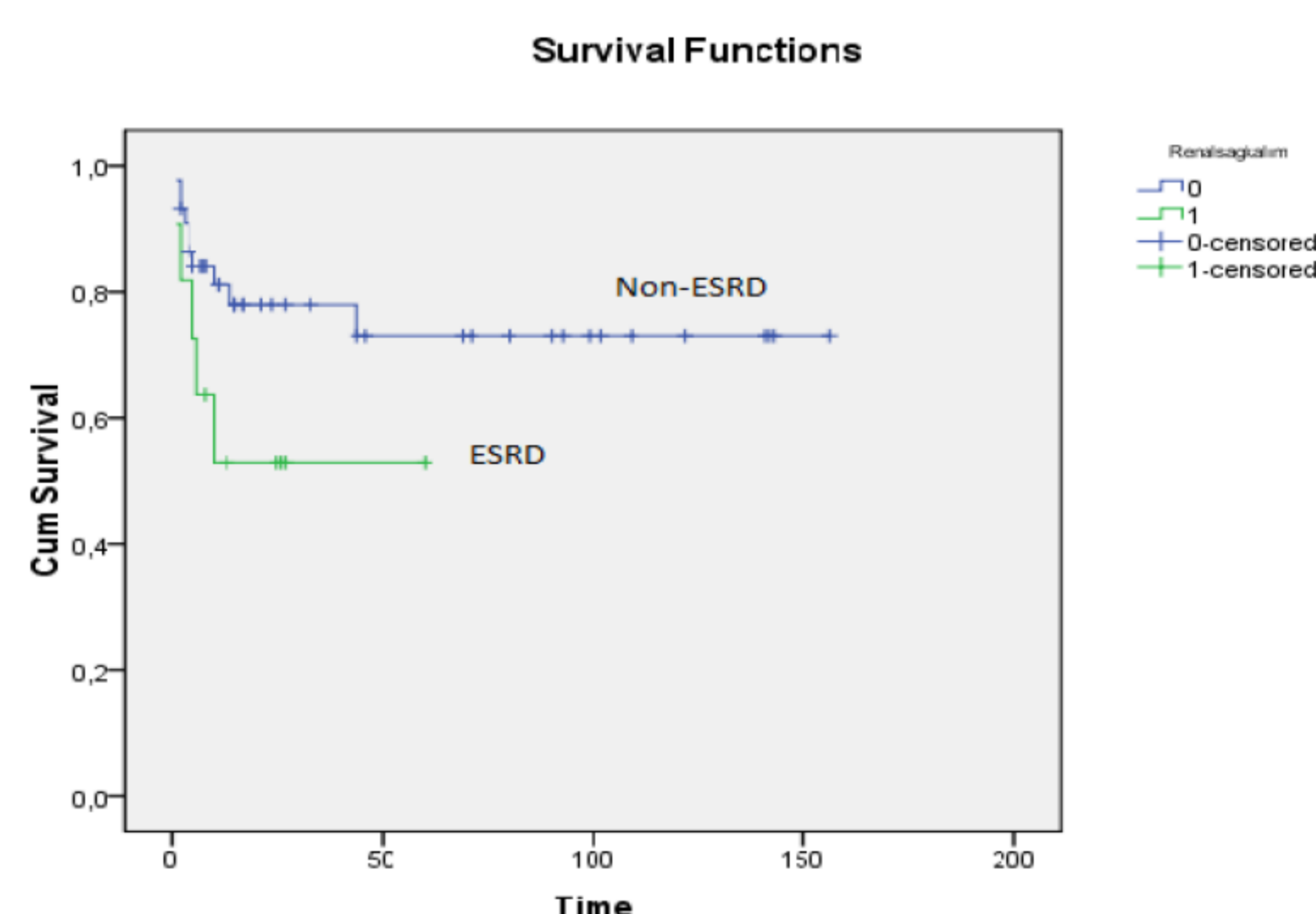


Figure 2. Survival of patients with and without ESRD.

**RESULTS:** Clinical and laboratory findings of WG patients (n:56, Female: 24, Male:32) are shown in Table 1. Thirty one patients had renal involvement and 24 of these patients had chronic kidney disease (42.8%). One, 5 and 10 year renal survival rates were 84.8%, 74.1% and 74.1%, respectively. Renal survival was 118.2 ± 10.4 months in all patients (Figure 1). Eleven patients had end-stage renal disease (ESRD) and 10 patients underwent HD (19.6%). There were no significant differences regarding patient survival between patients with or without ESRD (34.1 and 117.4 months, respectively; Log Rank test p=0.1) (Figure 2). There was a negative correlation between time to HD and renal survival (r=-0.435, p=0.001). However there was a positive correlation between renal survival and plasmapheresis and HD treatment (p<0.001, r=0.633 and 0.885, respectively) (Table 2).

Urea, creatinine and proteinuria were significantly higher and GFR was lower in patients who were dead compared to who survived. HD treatment was found to be more prevalent in the former group. Age was the only factor which effected mortality (p=0.026). The most prevalent cause of death was respiratory failure (53.3%).

**CONCLUSION:** Renal impairment was significantly more frequent in patients who were dead. ESRD was not associated with patient survival. However, the prolonged time period before the onset of HD was associated with reduced renal survival. HD and plasmapheresis treatment affected renal survival positively.

Age(years)	45.7±17.0
Female/Male	24/32
Time from onset of symptoms to diagnosis (months)	12.3±21.8
Hematocrit(%)	35.5±5.9
ESR(mm/hour)	66.0±65.0
CRP(mg/L)	66.4±25.0
Creatinine(mg/dL)	2.7±0.9
Albumin(g/dL)	3.2±0.6
Hematuria(%)	48
Proteinuria(mg/day)	765.2±1067.7
GFR(ml/min/1.73 m <sup>2</sup> )	69.1±43.0
ANCA positivity(%)	94
PR3/MPO ANCA positivity(%)	65/23.9
BVAS/WG	2.1(0-10)
VDI	3.8(1-9)
BVAS/WG: Birmingham vasculitis activity score/Wegener granulomatosis; VDI: Vasculitis damage index	

Table 1. Clinical and laboratory findings of patients.

	Correlation coefficient(r)	p
Age	-0.31	NS
Sex	0.247	NS
Time to diagnosis	-0.234	NS
Lung/Upper respiratory system involvement	0.078/-0.154	NS
Nervous system/Eye involvement	-0.155/-0.096	NS
Arthritis	0.215	NS
Skin involvement	0.026	NS
Initial ESR	0.039	NS
Last ESR	0.392	0.003
Initial/Last CRP	0.003/0.176	NS
Initial/Last ANCA	-0.028/0.047	NS
BVAS/WG	0.071	NS
VDI	0.289	0.021

Table 2. Clinical and laboratory findings associated with renal survival.