

Number of cases per 100,000 inhabitants										
	2001	2002	2003	2010	2011	2018	2019	2020	Average	
AAC	13.2	13.2	13.7	12.7	12.7	12.1	11.4	13.0	13.1	
NAAC	12.8	13.4	11.6	6.6	6.0	7.0	7.0	6.2	7.7	
Difference between AAC & NAAC (%)	3.0%	-1.5%	15.3%	48.0%	52.8%	42.2%	38.6%	52.3%	41.2%	
NAAC Women with Public Insurance	10.0	9.4	10.9	5.9	5.7	7.1	7.5	7.1	7.0	
NAAC Women with Private Insurance	0.0	6.2	8.8	9.5	11.0	10.8	9.5	7.9	8.4	
Difference between Women with Public & Private Insurance in NAAC (%)	100.0%	34.0%	19.3%	-61.0%	-93.0%	-52.1%	-26.7%	-11.3%	-20.0%	
NAAC Men with Public Insurance	15.5	15.6	13.7	7.1	4.7	5.3	5.6	5.8	7.8	
NAAC Men with Private Insurance	0.0	2.4	10.2	7.0	10.9	12.5	9.2	5.8	8.5	
Difference between Men with Public & Private Insurance in NAAC (%)	100.0%	84.6%	25.5%	1.4%	-131.9%	-135.8%	-64.3%	0.0%	-9.0%	
AAC Women with Public Insurance	6.5	6.0	6.8	5.8	6.0	5.7	5.7	5.9	6.3	
AAC Women with Private Insurance	0.1	1.8	2.4	2.0	2.0	1.1	1.6	1.2	1.7	
Difference between Women with Public & Private Insurance in AAC (%)	98.5%	70.0%	64.7%	66.5%	66.7%	80.7%	71.9%	79.7%	73.0%	
AAC Men with Public Insurance	26.01	24.8	27.1	23.5	23.1	23.2	22.2	26.9	25.2	
AAC Men with Private Insurance	0.0	4.7	8.0	9.3	9.3	7.1	5.4	5.9	6.5	
Difference between Men with Public & Private Insurance in AAC (%)	100.0%	81.0%	70.5%	60.4%	59.7%	69.4%	75.7%	78.1%	74.2%	
AAC 15-19 y	0.2	0.4	0.1	0.1	0.5	0.2	0.1	0.2	0.2	
NAAC 15-19 y	0.3	0.3	0.2	0.4	0.4	1.3	1.4	0.6	0.7	
Difference between 15-19 y AAC & NAAC (%)	-50.0%	25.0%	-100.0%	-300.0%	-20.0%	-550.0%	-1300.0%	-200.0%	-250.0%	
AAC 20-29 y	1.0	1.0	1.5	0.8	1.5	0.7	0.9	0.8	1.1	
NAAC 20-29 y	1.0	1.1	1.3	0.8	1.4	1.7	1.5	1.0	1.1	
Difference between 20-29 y AAC & NAAC (%)	0.0%	-10.0%	13.3%	0.0%	6.7%	-142.9%	-66.7%	-25.0%	0.0%	
AAC 50-59 y	44.1	42.3	37.7	31.3	31.4	32.5	29.0	35.4	35.7	
NAAC 50-59 y	37.1	39.6	30.0	15.8	14.1	15.5	13.0	12.3	18.9	
Difference between 50-59 y AAC & NAAC (%)	15.9%	6.4%	20.4%	49.5%	55.1%	52.3%	55.2%	65.3%	47.1%	
AAC 60-69 y	56.8	53.0	56.1	46.9	48.2	36.9	35.4	38.3	47.6	
NAAC 60-69 y	62.4	66.4	59.7	26.6	26.2	21.7	27.5	20.5	33.3	
Difference between 60-69 y AAC & NAAC (%)	-9.9%	-25.3%	-6.4%	43.3%	45.6%	41.2%	22.3%	46.5%	30.0%	

Table 1: Number of cases per 100,000 inhabitants in the corresponding group under sturdy

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P-9 SURVIVAL ANALYSIS OF A BRAZILIAN COHORT OF PATIENTS WITH WILSON DISEASE

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Conflict of interest: No
Introduction and Objectives: Wilson's disease (WD) is a rare autosomal recessive disease leading to impairment in copper excretion and subsequent accumulation primarily in the liver and brain. Long-term survival appears to be similar to that of the general population, depending on early diagnosis and treatment. However, studies evaluating WD survival are scarce. We aimed to evaluate the long-term survival of a cohort of WD patients followed at a single Hepatology Center in Brazil.

Patients / Materials and Methods: Demographic characteristics, age at diagnosis, type of manifestation (isolated hepatic, isolated neuropsychiatric or associated hepatic and neuropsychiatric) of WD patients, followed between 1999 and 2023, were evaluated. The average survival and its relationship with the predominant disease phenotype were determined.
Results and Discussion: 34 patients were evaluated, 53% female, mean age at diagnosis 21 ± 9 years (6 to 42), disease duration of 11 ± 7 years (1 to 25) until outcome. The majority (47%) had the isolated hepatic manifestation, 38% hepatic neuropsychiatric manifestations and 15% isolated neuropsychiatric disorder. The overall population mortality rate was 36%. The mean age at the time of death was 33 ± 12 years (11 to 55). The estimated average survival at 5 years was 85%, at 10 years 70% and at 20 years 40%, from diagnosis. There was a tendency towards lower survival in patients who presented neuropsychiatric manifestations, either alone or in association with liver disease, compared to patients with isolated liver manifestations (50% vs. 80%; p= 0.07). There was no difference in survival curves according to gender (p=0.82) or considering the age at onset of the disease above or below 20 years (p=0.54).
Conclusions: Despite being potentially treatable, WD still presents high mortality, affecting young patients, whose survival rate is greatly reduced. WD patients with neuropsychiatric symptoms had worse survival. These data highlight the need for earlier diagnosis and easier access to treatment.

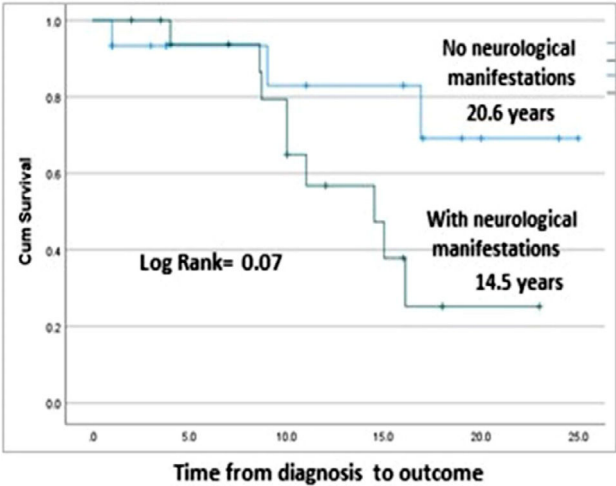


Figure 1. Survival curve according to clinical manifestation after WD diagnosis

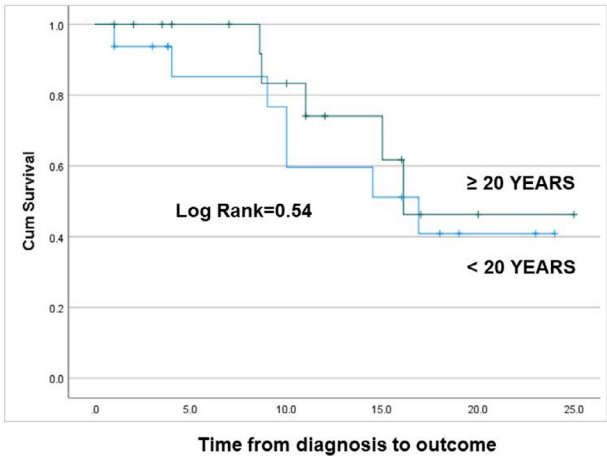


Figure 2. Survival curve considering the age at onset of the disease? or < 20 years

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