

Score de Leipzig

Objetivo: Estabelecer a probabilidade diagnóstica de doença de Wilson

Variáveis, Cálculo e Interpretação:

Typical clinical symptoms and signs		Other tests	
KF rings		Liver copper (in the absence of cholestasis)	
Present	2	>5x ULN (>4 µmol/g)	2
Absent	0	0.8-4 µmol/g	1
Neurologic symptoms**		Normal (<0.8 µmol/g)	-1
Severe	2	Rhodanine-positive granules*	1
Mild	1	Urinary copper (in the absence of acute hepatitis)	
Absent	0	Normal	0
Serum ceruloplasmin		1-2x ULN	1
Normal (>0.2 g/L)	0	>2x ULN	2
0.1-0.2 g/L	1	Normal, but >5x ULN after D-penicillamine	2
<0.1 g/L	2	Mutation analysis	
Coombs-negative hemolytic anemia		On both chromosomes detected	4
Present	1	On 1 chromosome detected	1
Absent	0	No mutations detected	0
TOTAL SCORE	Evaluation:		
4 or more	Diagnosis established		
3	Diagnosis possible, more tests needed		
2 or less	Diagnosis very unlikely		

*If no quantitative liver copper available, **or typical abnormalities at brain magnetic resonance imaging. KF, Kayser-Fleischer; ULN, upper limit of normal.