

Copper levels and Ceruloplasmin

Estimation of serum copper, serum ceruloplasmin and 24-h urinary copper in suspected Wilson's Disease.

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Recommendations for Diagnostic Testing of Wilson's Disease

Ceruloplasmin

An extremely low serum ceruloplasmin level (<50 mg/L or <5 mg/dL) should be taken as strong evidence for the diagnosis of WD. Modestly subnormal levels suggest further evaluation is necessary. Serum ceruloplasmin within the normal range does not exclude the diagnosis (**Class I, Level B**).

Urinary Copper Excretion

Basal 24-hour urinary excretion of copper should be obtained in all patients in whom the diagnosis of WD is being considered. The amount of copper excreted in the 24-hour period is typically >100 micrograms (1.6 micromols) in symptomatic patients, but finding >40 micrograms (>0.6 micromol or >600 nmol) may indicate WD and requires further investigation (**Class I, Level B**).

Monitoring patients on pharmacotherapy

Serum 'Free' Copper can be calculated by subtracting ceruloplasmin bound copper ($3.15 \times$ ceruloplasmin in mg/l) from the total serum copper (in $\mu\text{g/l}$). It is of more value in monitoring pharmacotherapy than in the diagnosis of Wilson's Disease

Differential Diagnosis

Test	Wilson Disease	Copper Toxicity, Acute or Chronic	Menkes Disease (Kinky Hair Syndrome)
Copper, blood	Low but may be normal	High	Low
Copper, serum free	High	High	Low
Ceruloplasmin	Low but may be normal	High	Low
Copper, urine	Very high	High	Low
Copper, liver/hepatic*	Positive but depending on the site sampled, may be negative	High or normal	Low

*Excess copper in the liver is often unevenly distributed and may not be detected in a sample.

- Low blood copper concentrations along with increased urine copper levels, low ceruloplasmin levels, and increased hepatic copper are typically seen with Wilson disease.
- Increased blood and urine copper concentrations and normal or increased ceruloplasmin levels may indicate exposure to excess copper or may be associated with conditions that decrease copper excretion, such as chronic [liver disease](#), or that release copper from tissues, such as acute [hepatitis](#). Increased hepatic copper may be present with [chronic](#) conditions.
- Decreased blood and urine copper concentrations and decreased ceruloplasmin may indicate a copper deficiency.
- A normal hepatic copper test may indicate that copper metabolism is functioning properly or that the distribution of copper in the liver is uneven and the sample is not representative of the person's condition.

TEST DETAILS

Test Name	Copper Analysis ,Serum Copper Analysis ,Urine 24 hrs Copper Analysis , Urine Spot	Copper Analysis ,Serum	Copper oxidase (Ceruloplasmin) ,Serum
Technology	Atomic Absorption	Biochemical	Nephelometry
Detection	Wilson's Disease, Menkes Kinky hair Disease, Primary biliary Cirrhosis		
Starting Material	<ul style="list-style-type: none"> • 3 ml of Serum • 10ml of 24 hrs urine in acid washed, metal free container • 10ml of 24 hrs urine in acid washed, metal free container 	3 ml of Serum	2 ml of Serum
Turnaround Time	<ul style="list-style-type: none"> • 9 days • 3 days • 3 days 	3 days	2 days
Result & Interpretation			