**Ceruloplasmin**

Testing is usually done as a screen for Wilson’s disease. A study performed in Hong Kong using a Beckman Immage indicate that Wilson’s disease is highly likely when ceruloplasmin is < 0.14 g/L. Equivalent value for the Roche assay = 0.08 g/L Other causes of a low Ceruloplasmin includes: Menkes disease, aceruloplasminemia, liver failure and protein loosing conditions . Ceruloplasmin levels > 0.16 g/L (Roche) suggest Wilson’s disease is unlikely but inflammation can modestly increase levels for 4 – 20 days following the insult. Levels can be increased to > 0.16 g/L with acute liver failure. Ceruloplasmin levels are increased during pregnancy or when taking estrogen. 24 hr Urinary copper excretion in patients with Wilson’s disease can vary from mildly increased (1.5 times ULN) to 20 times ULN.

Approximately 95% of copper in blood is transported by ceruloplasmin. The main function of ceruloplasmin is to oxidise iron (Fe2+ to Fe3+). The main use of measuring ceruloplasmin is to screen for Wilson disease. Wilson disease is caused by a defect in copper transporter ATP7B, which prevents copper from being incorporated in ceruloplasmin. Low ceruloplasmin levels are suspicious of Wilsons disease, other even rarer causes of a low ceruloplasmin level include Menkes kinky hair syndrome (caused by a defect in copper transporter ATP7A, preventing absorption of copper in the GI system) and aceruloplasminemia. Other causes of a low ceruloplasmin level includes liver disease or protein loss. Increased levels are seen with acute phase response.

**New assay reporting:**

0 days onwards:

The reference intervals for this age group is not well defined.

Adult reference interval: Males 0.15 - 0.30 g/L, Females: 0.16 – 0.45

Ceruloplasmin levels are approximately 0.05 - 0.30 g/L in neonates, gradually rising to approximately adult levels (or slightly higher) by 1 year of age. Low levels are observed in most patients with Wilson's disease but may be within normal limits, especially in patients with a liver presentation. Low levels are also seen in 10 - 20% of carriers of Wilson's disease, or patients with aceruloplasminemia, Menkes kinky hair disease, advanced liver disease or with protein losing conditions. Ceruloplasmin is a positive acute phase protein.

1 Year onwards:

Adult reference interval: Males 0.15 - 0.30 g/L, Females: 0.16 – 0.45

Reference intervals in children are not well defined.

At 1 year levels are similar or slightly higher than in adults, then increase to peak at 2 - 3 years (approximately 1.5 x adult levels). Levels then gradually decline to adult levels at approximately 17 years of age.

Low levels are seen in most patients with Wilson's disease but may be within normal limits, especially in patients with a liver presentation.

Low levels are also seen in 10 - 20% of carriers of Wilson's disease, aceruloplasminemia, Menkes kinky hair disease, advanced liver disease and in protein losing conditions.

Ceruloplasmin is an acute phase protein. Stephen DuToit, Chemical Pathologist, 021 245 8322

17 years onwards:

Male: 0.15 – 0.30

Female: 0.16 – 0.45

(1) Age and gender specific pediatric reference intervals for aldolase, amylase,

ceruloplasmin, creatine kinase, pancreatic amylase, prealbumin, and uric acid. Sarah M. Clifford a, Ashley M. Bunker b, Jeffrey R. Jacobsen c, William L. Roberts a,b,⁎ Clinica Chimica Acta 412 (2011) 788–790

(2) <https://caliper.research.sickkids.ca/#/search>

(3) Mayo clinic [https://www.mayocliniclabs.com/test-catalog/overview/614504#](https://www.mayocliniclabs.com/test-catalog/overview/614504)

(4) Diagnostic Accuracy of Serum Ceruloplasmin in Wilson Disease: Determination of Sensitivity and Specificity by ROC Curve Analysis among ATP7B-Genotyped Subjects. Chloe M. Mak,1,2 Ching-Wan Lam,1\* and Sidney Tam2. Clinical Chemistry 54:8 1356–1362 (2008)