

**Non-ceruloplasmin copper and urinary copper in clinically stable
Wilson disease: Alignment with recommended targets and values in
healthy persons**

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Table S1.

Distribution Total Cu and NCC-Sp in healthy humans according to gender and ethnicity.

	Total Plasma Copper (µg/L)				NCC-Sp (µg/L)			
	N	Median	0.025-0.975 % range	<i>p</i>	N	Median	0.025-0.975 % range	<i>p</i>
All	75	1030	717-1731		75	86	46-213	
Gender								
Female	37	1030	730-1861	0.55*)	37	63	45-209	0.0007*)
Male	38	1030	718-1655		38	109	63-211	
Ethnicity								
Black	19	1170	791-1911	0.002**)	19	80	54-203	<10 ⁻⁵
Caucasian	23	941	692-1301		23	178	117-212	
Hispanic	31	1030	772-1605		31	63	44-164	
Other	2	1040	930-1150		2	192	208-209	

*) Mann-Whitney's test;

**) Kruskal-Wallis' test ("Other" not included because of less than 5 samples)

Fig. S1.

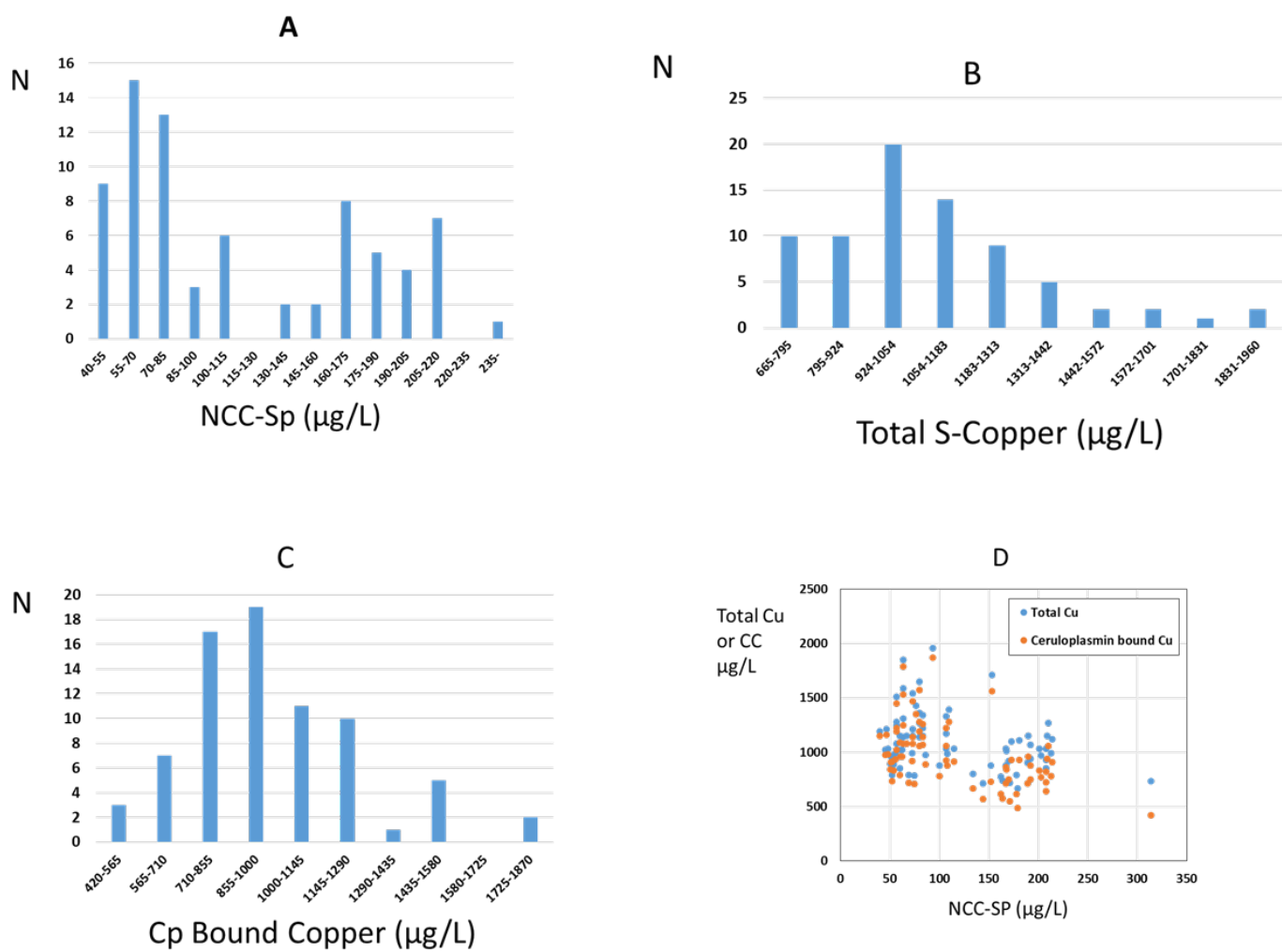


Fig. S1. Distribution of NCC-Sp, Total S-Cu, and Cp Bound Cu in 75 healthy volunteers.

Fig. S2.

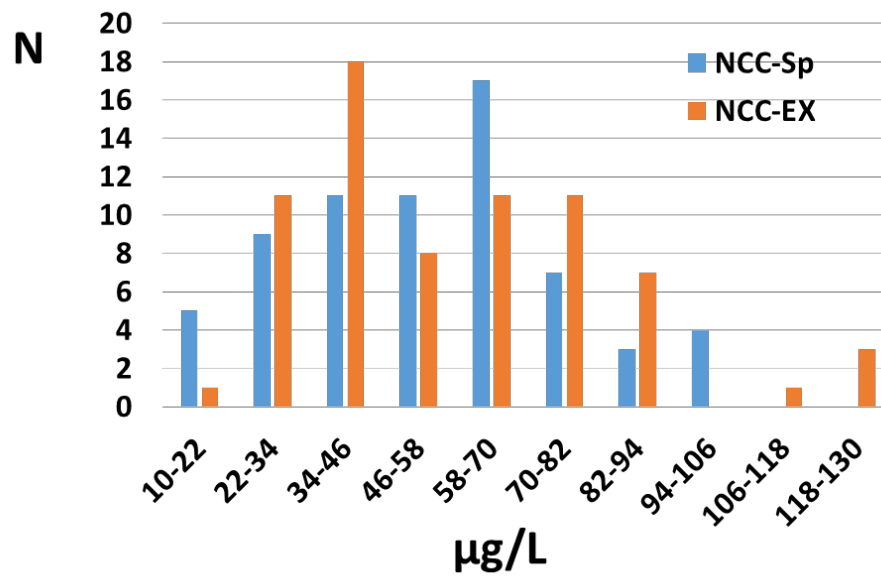


Fig. S2. Distribution of NCC-Sp in 69 patients and NCC-Ex in 71 WD patients at their first visit in the Chelate study (27).

Fig. S3.

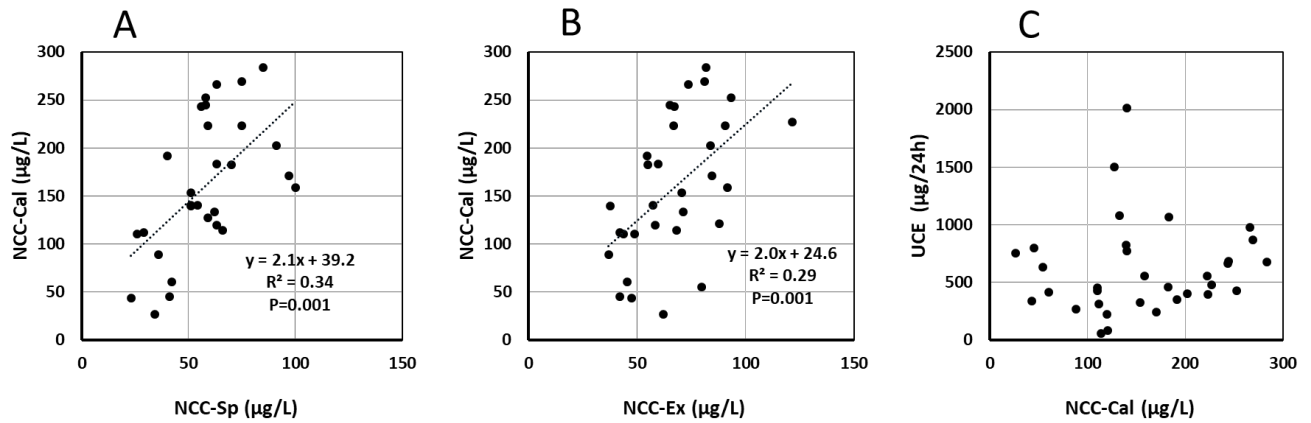


Fig. S3. Relation of NCC-Cal to NCC-Sp, NCC-Ex and UCE. NCC-Cal was calculated as Total Serum Copper (µg/L) – 3.14·Ceruloplasmin (µg/L). Values could only be calculated in 27, since only 33 patients had ceruloplasmin above the lower limit of quantification (LLOQ) of 52 mg/L and total copper not available in 6 of these. There was no covariation between NCC-Cal and UCE. Both NCC-Sp and NCC-Ex were positively correlated to NCC-Cal ($p=0.001$ in both cases), but NCC-Cal was clearly above the line of identity. NCC-Cal did not correlate with UCE.