

Triglycerides (N<1.7 mmol/L)	1.69	0.69	0.87	0.55	0.97	0.75	0.89	0.54
LDL-cholesterol (N<3.5 mmol/L)	2.35	2.39	2.30	1.89	2.31	2.91	1.98	2.07
Alpha-1-antitrypsin (N>0.9 g/L)	1.46	1.34	1.37	1.74	1.26	1.46	1.54	1.49
Ceruleoplasmin (N>0.15 g/L)	0.47				0.24	0.33		
Serum copper concentration (N>13 µmol/L)	28.1				17.3	18.5		

Supplementary Table 2. Main clinical and biological characteristics of patients with A20 haploinsufficiency and liver involvement in the literature.

First author ^{Ref}	Age at first symptoms	Age at the diagnosis of liver involvement	Sex	Mutation	Hepatomegaly	Splenomegaly	Hepatic cytolysis	Liver biopsy	Other description
Tsuchida ⁴	12 years	29 years	Female	c.1434C>A, p.Cys478*	Yes	Yes	NR		
Kadowaki ⁵	5 years	5 years	Female	c.2209delC, p.Gln737Serfs*79	NR	NR	NR		Chronic hepatitis
Kadowaki ⁵ , Takagi ⁷	7 months	1 year	Male	c.1245_1248del, p.Lys417Serfs*4	Yes	Yes	2 x normal	Interface hepatitis with spotty focal necrosis associated with giant multinucleated hepatocytes and marked CD8 ⁺ T cell infiltration	Liver dysfunction
Li ⁶	7 years	7 years	Female	c.559C>T, p.Gln187*	Yes		NR	Liver fibrosis	Liver dysfunction
Hautala ¹⁴	4 years	30 years	Female	c.271A>T, p.Lys91*	NR	NR	2 x normal	Hepatosteatorosis, iron load in Kupffer cells and sinusoidal macrophages	
	7 years	43 years	Female	c.271A>T, p.Lys91*	NR	NR	8 x normal		Liver cirrhosis with liver dysfunction
Schwartz ¹⁵	8 years	8 years	Female	p.Leu626Valfs*45	NR	NR	3 x normal	Autoinflammatory hepatitis with fibrosis	
Duncan ¹⁷	10 years	10 years	Male	c.1466_1467delTG, p.Val489Alafs*7	NR	NR	NR		Hepatitis
Gans ¹⁸	Late childhood	27 years	Male	c.912del, p.Glu305Serfs*3	Yes	Yes	NR		
He ¹⁹	6 years	6 years	Female	c.258_261del, p.Cys86Trpfs*8	NR	NR	NR		Autoimmune hepatitis
Kim ²⁰	2 months	2 months	Female	c.492G>A, p.Trp164Ter	NR	NR	20 x normal		
Taniguchi ²¹	3 weeks	3 weeks	Female	c.305A>G, p.Asn102Ser	Yes	Yes	NR		

NR: not reported.