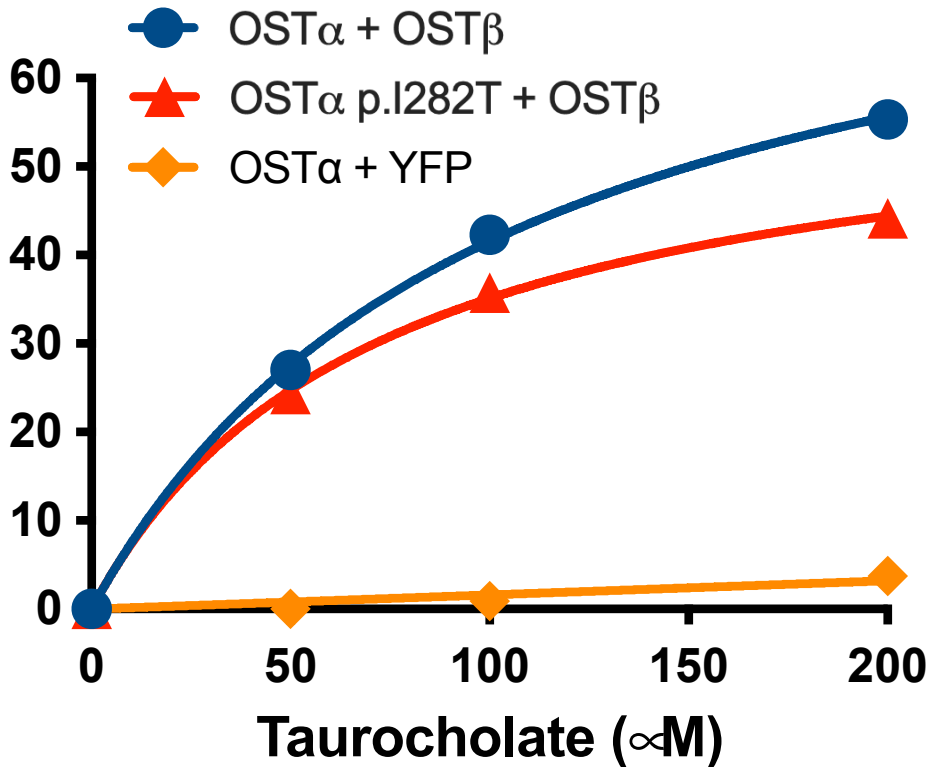


Taurocholate Uptake
(pmol/mg protein)



Supplemental Table 1. Serum Bile Acids

Bile Acid (nmol/L)	Patient 1	Patient 1 UDCA	Patient 2	Patient 2 UDCA
TCA	232.8	8.7	229.0	16.1
GCA	155.1	56.3	582.9	90.3
TCDC	200.1	10.5	55.0	10.7
GCDCA	366.0	171.4	222.2	157.1
T/G DCA	0.0	0.0	0.0	0.0
T/G LCA	0.0	0.4	0.4	1.3
CA	49.5	64.5	13.7	128.9
CDCA	387.5	358.7	53.3	264.1
DCA	0.7	0.5	0.8	1.0
HCA	110.4	4.2	33.2	14.8
TUDCA	2.7	35.1	0.3	19.3
GUDCA	14.5	1751.0	7.1	1365.6
UDCA	13.8	1761.3	2.6	1017.8
iso-UDCA	57.1	392.3	22.3	488.0
Other	19.9	36.5	8.3	17.1
Total	1610	4647	1198	3577

Serum bile acid species in patients off and on UDCA treatment. The concentration of taurine (T) and glycine (G) conjugated and unconjugated bile acids are shown as nmoles per liter. Minor bile acid species, which include HDCA, MDCA, α MCA, β MCA, ω MCA are summed and listed as Other. Cholic acid (CA), chenodeoxycholic acid (CDCA), deoxycholic acid (DCA), hyodeoxycholic acid (HCA), ursodeoxycholic acid (UDCA), hyodeoxycholic acid (HDCA), murideoxycholic acid (MDCA), muricholic acid (MCA)