

Supplementary Table S1: Comparison of the AGAL activities measured in this paper and those available in Benjamin et al. 2017.

	This paper nmol/(h mg) $\pm$ SD	Table 1S, Benjamin et al nmol/(h mg) $\pm$ SEM	Seeman et al 2020 nmol/(h mg) $\pm$ SD
	Stable transfections in patient-derived fibroblasts not expressing <i>GLA</i>	Transient transfections in HEK-293 cells	Patient-derived fibroblasts
Empty vector	0.35 $\pm$ 0.02	-	-
wt	5.20 $\pm$ 0.01	-	58.5 $\pm$ 31.1
D244H	0.65 $\pm$ 0.04	6129 $\pm$ 366	-
Q280K	0.34 $\pm$ 0.03	8019 $\pm$ 395	-
V269M	0.39 $\pm$ 0.02	1213 $\pm$ 40	-
L300F	0.70 $\pm$ 0.07	4746 $\pm$ 363	-
C56Y	0.47 $\pm$ 0.02	BLD *	-
E341D	0.34 $\pm$ 0.05	BLD *	-
A230T	0.60 $\pm$ 0.02	BLD *	-

AGAL activities expressed as nmol/(h mg) from the independent experiments reported in this paper were averaged and compared to those reported in Benjamin et al. The results highlight that our cell lines do not overexpress AGAL. \* BLD: Below the limit of detection