Expression and functional characterization of *AKU* alleles in *E. coli.*

TABLE1. Enzyme activities of mutant HGO proteins

HGO protein	Specific Activity (nmol/min mg)	%wt
WT	20932 ± 1394	100
E42A	5972 ± 787	29
W60G	13,2	0.06
Y62C	4708 ± 834	22.5
H80Q	19275	95
W97G	22.5	0.1
A122D	6842 ± 1760	33.5
D153G	6408 ± 1966	32.7
G161R	204 ± 80	1
S189I	700 ± 43	3.4
I216T	1.4	0
R225H	23 ± 9	0.1
F227S	20.45	0.1
P230S	850 ± 306	4
P230T	210	1
D291E	0	0
V300G	400	1.9
R330S	93	0.4
M368V	7715 ± 1749	37
H371R	0	0

(AKUdatabase; http://www.alkaptonuria.cib.csic.es/)

(from: Rodríguez et al. 2000, Hum Mol Genet 9:2341-2350)

TABLE 2. Steady-state kinetic parameters of AKU HGO proteins showing significant activity

Mutant protein change	Vmax (nmol/min mg)	Km (mM)	Vmax/Km	% wt
wt	22300	6,2	3597	100
E42A	6692	25,4	263	7,3
Y62C	4896	6	816	22,7
A122D	8382	9,2	911	25,3
D153G	6309	7,1	889	24,7
M368V	7400	14,4	514	14,3