

Human disease		Branching Enzyme (EC 2.4.1.18)										Debranching Enzyme (EC 3.2.1.33)		
		GSD4					GSD4/APBD					GSD3		
Human disease mutation position		224	257	329			515	524	545	628		1147	1448	
Human disease mutation		L → P	F → L	Y → S	R → C/H	R → Q	intra-domain					R → G	G → R	
		Protein Acc.	Gene	Alpha-amylase						A-a_C	Protein Acc.	Gene	GDE_C	
Animals	<i>Homo sapiens</i>	Q04446	GBE1	L	F	Y	R	R	H	H	P35573	AGL	R	G
	<i>Mus musculus</i>	Q9D6Y9	Gbe1	L	F	Y	R	R	H	H	F8VFN4		R	G
	<i>Danio rerio</i>	XP_687620.6		L	F	Y	R	R	H	H	NP_001166124.1		R	G
	<i>Caenorhabditis elegans</i>	Q22137		L	F	Y	R	R	H	H	O62334	agl-1	R	G
	<i>Drosophila melanogaster</i>	A1Z992	AGBE	V	Y	Y	R	R	H	H	E1JGQ5/Q9W2H8		R	G
	<i>Amphimedon queenslandica</i>	I1FQH3		L	H	Y	R	R	H	H	I1FDE0		R	G
Choanoflagellida	<i>Monosiga brevicollis</i>	A9URY2		L	F	Y	R	R	H	H	A9V544		R	G
Ichthyosporaea	<i>Capsaspora owczarzaki</i>	E9C2E3		L	F	Y	R	R	H	H	E9CDY8		R	G
Filisterea	<i>Sphaeroforma arctica</i>	SARC_09810T0		I	F	Y	R	R	H	H	SARC_05895T0		R	G
Fungi	<i>Saccharomyces cerevisiae</i>	P32775	GLC3	L	F	Y	R	R	H	H	Q06625	GDB1	R	G
Amoebozoa	<i>Dictyostelium discoideum</i>	Q555Q9	glgB	L	F	Y	R	R	H	H	Q54K94	agl	R	G
Land Plants		O23647	SBE2.1	L	F	Y	R	R	H	H				
	<i>Arabidopsis thaliana</i>	Q9LZS3	SBE2.2	L	F	Y	R	R	H	H				
		Q01401	SBE1	L	F	Y	R	R	H	H				
	<i>Oryza sativa japonica</i>	Q6H6P8		L	F	Y	R	R	H	H				
Green Algae		D8TIE8	glgB6	L	F	Y	R	R	H	H				
	<i>Volvox carteri</i>	D8U9K6	glgB7	L	F	Y	R	R	H	S				
Red Algae	<i>Cyanidioschyzon merolae</i>	CMH144C		L	F	Y	R	R	H	H				
Alveolata	<i>Paramecium tetraurelia</i>	A0DXF8		L	F	Y	R	R	H	H	A0BWA0		R	G
	<i>Tetrahymena thermophila</i>	Q23TC5		L	Y	F	K	R	S	N	I7M1C6		R	G
Apusozoa	<i>Thecamonas trahens</i>	AMSG_09093T0		L	F	Y	R	R	H	H	AMSG_02577T0		R	G
Bacteria	<i>Bacteroides thetaiotaomicron</i>	Q8A9P4		L	F	Y	R	R	H	N	Q89ZS3		E	G
	<i>Escherichia coli</i>	P07762	glgB	V	Y	Y	P	N	Q	T				
	<i>Nosfoc punctiforme</i>	B2J3N1	glgB	I	Y	Y	P	N	Q	T				