

Heart Disease in-a-Dish: Cardiomyopathy Patient-Specific iPSC Lines with BAG3 Gene Mutations

- Genetic mutations in the *BAG3* gene (BCL2-associated athanogene 3) cause inherited cardiomyopathy, primarily dilated cardiomyopathy (DCM).
- BAG3* gene encodes for a co-chaperone protein highly expressed in cardiac and skeletal muscle.
- BAG3* protein roles: sarcomere maintenance, protein quality control and anti-apoptosis.
- High age-dependent penetrance: 80% penetrance by age 40.
- BAG3* Mutations = Aggressive Heart Disease Progression: high risk of rapid progression to end-stage heart failure, requiring heart transplant or ventricular assist device (LVAD).
- Greenstone Biosciences' iPSC Biobank is offering ten (10) patient-specific *BAG3* mutant cell lines (Table 1). These cells are available for Disease in-a-Dish assays and Clinical Trials in a Dish for discovery of new treatments and/or for cardiac safety screening.

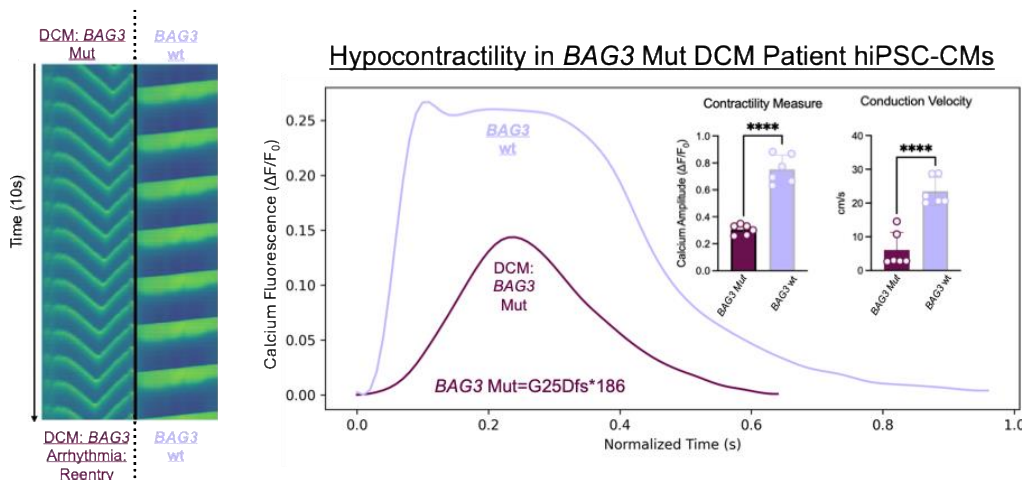


Figure 1. Disease Phenotype-*BAG3* Mutant iPSC-CMs.

BAG3 Mut Phenotype:

- Optical mapping
- Calcium flux
- Spontaneous arrhythmia (Rotor)
- Reduced contractility
- Slow impulse velocity

Cell Line	Disease	<i>BAG3</i> mutation	Sex	Age	Ejection Fraction	BMI
GSB-L194	DCM	G25Dfs*186	M	48	0.12	27.97
GSB-L394	DCM	A479V	F	35	0.56	27.05
GSB-L574	DCM	Y233*	F	61	0.39	22.78
GSB-L599	HCM	S185L	F	72	0.65	21.46
GSB-L747	LVNC	R218Q	F	47	NA	35.67
GSB-L815	DCM	T34Afs*21	M	42	0.51	23.34
GSB-L2235	DCM	P23S	M	77	NA	24.67
GSB-L2788	Other	R218W	F	46	0.65	50.35
GSB-L2795	Other	A164S	F	49	0.53	41.32
GSB-L3371	DCM	Q251*	F	41	0.20	21.14

Table 1. Greenstone Biosciences' *BAG3* Mut Patient Derived hiPSC-CM Lines.

HCM=Hypertrophic Cardiomyopathy, DCM=Dilated Cardiomyopathy, LVNC=Left Ventricular Non-Compaction