	Up to Age 20				Up to Age 30				Above the Age of 30			
	Not meeting AHA	Meeting AHA	Exceeding AHA	p	Not meeting AHA	Meeting AHA	Exceeding AHA	p	Not meeting AHA	Meeting AHA	Exceeding AHA	p
N	37	17	50		37	22	28		53	7	21	
Phenotype Neg	3	5	8	0.13	2	5	3	0.25	6	1	2	0.94
Average Age at Phe+ (mean)	40.6 (16.2)	37.0 (23.8)	33.1 (14.0)	0.51	42.8 (15.5)	40.2 (18.1)	36.3 (11.7)	0.64	41.3 (15.8)	38.7 (10.5)	39.4 (14.8)	0.61
LAD (mm, mean)	42.3 (7.1)	37.7 (5.7)	41.0 (6.0)	0.03	42.2 (7.1)	39.7 (6.9)	41.1 (5.8)	0.28	41.5 (6.8)	41.0 (11.6)	40.3 (4.5)	0.90
MWT (mm, mean)	18.0 (5.1)	17.1 (6.7)	15.8 (4.9)	0.40	18.1 (5.1)	16.1 (6.2)	16.5 (5.2)	0.13	17.2 (5.8)	16.4 (6.2)	17.6 (4.7)	0.79
LV Massi (g, mean)	67.5 (30.1)	55.3 (19.9)	61.5 (18.3)	0.14	64.3 (30.5)	56.4 (16.5)	62.3 (19.2)	0.40	64.2 (28.1)	60.7 (20.7)	60.7 (20.0)	0.94
LGE % (median)	3.2 (6.1)	3.1 (5.4)	7.7 (10.8)	0.16	6.0 (6.2)	6.7 (8.5)	9.7 (12.9)	0.84	9.3 (10.6)	8/9 (4.2)	2.9 (4.2)	0.67

Figure 1: Phenotypic expression stratified by exercise volume through different age groups. Exercise volume is divided into those that were not meeting, meeting and those exceeding AHA guideline recommendations for exercise. (433.33 – 866.66 MET-hours per year).

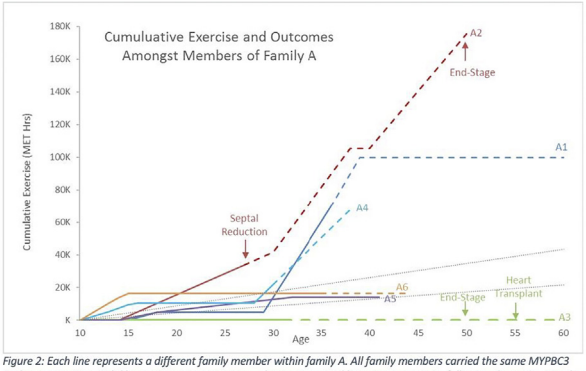

Figure 2: Each line represents a different family member within family A. All family members carried the same MYPBC3 pathogenic variant. Solid lines represent time prior to diagnosis. Dotted lines represent time following diagnosis. Grey lines represent AHA recommended range for physical activity of 500-1000 MET-minutes of moderate-to-vigorous physical activity per week. (The 2018 Physical Activity Guidelines Advisory Committee Scientific Report.

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BALLISTOCARDIOGRAPHY TO CHARACTERIZE PULMONARY ARTERY PRESSURE IN ADVANCED HEART FAILURE PATIENTS AND HEALTHY ADULTS

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BACKGROUND: Remote monitoring to manage heart failure (HF) at home can reduce readmission rates. Notably the implantable CardioMEMS device that measures pulmonary artery pressure (PAP) has proven effective but is both invasive and costly. Our goal is to develop smart home sensors that can monitor HF patients non-invasively and without requiring active patient engagement. The objective of this study is the development and evaluation of ballistocardiography (BCG) as a candidate technology for non-invasive, autonomous monitoring of PAP for HF patients.

METHODS AND RESULTS: BCG records the recoil force generated by the heart during ejection. The force generated by rapid ejection of the blood with each cardiac cycle yields micro-oscillations in the recorded BCG signal. Force and acceleration are equivalent in BCG given that the mass of the blood is assumed constant. Figure-A shows a healthy BCG signal in acceleration. A method for recording BCG has been developed that does not require electrodes, skin contact, or interaction with external devices. In healthy subjects, the dominant vector of acceleration is recorded as the J-wave, which represents the rapid increase in blood velocity with ejection (Figure-B). Fragmentation of J-wave (Figure-C) has been reported to emerge due to dyssynchronous ejection between the left and right ventricles (Figure-D). Increased afterload due to elevated PAP may increase dyssynchronous biventricular ejection and increase fragmentation of the J-wave. Five patients with advanced HF were recruited to undergo BCG recording on the same day as they underwent clinically indicated right heart catheterization. BCG was recorded while standing for 90 seconds. Mean PAPs of the patients were 19, 41, 30, 40, and 43 mmHg. Six healthy controls were also recruited to undergo BCG recordings. J-wave fragmentation was observed in HF patients with elevated PAPs (Figure-E). Compared to healthy controls (Figure-F), we also observed increased beat-to-beat variation and prolongation of signal duration based on visual inspection. These behaviours were not observed in the HF patient with normal PAP and healthy controls.

CONCLUSION: Preliminary observations support the possibility that J-wave fragmentation of BCG signal recordings may identify increased PAP in HF patients.

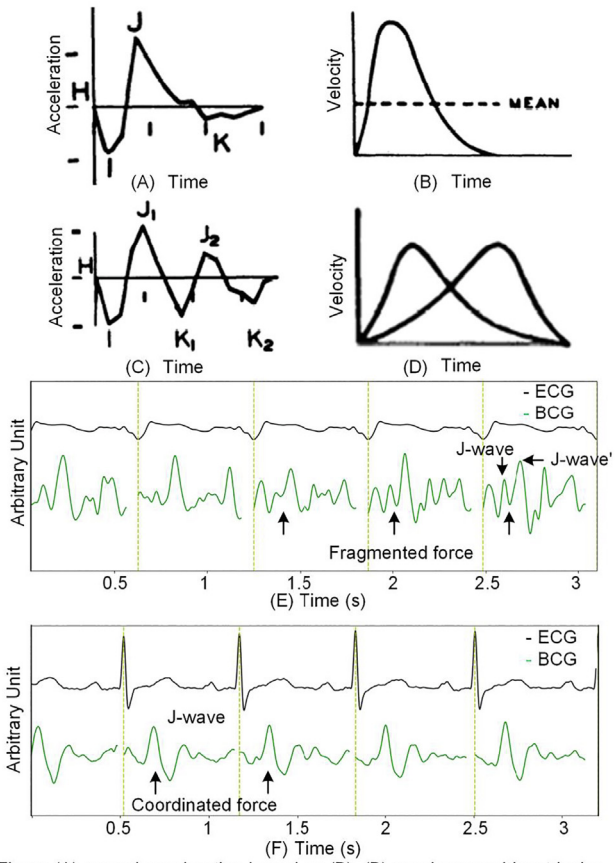

Figure (A) normal acceleration based on (B), (B) synchronous biventricular velocity generation, (C) fragmentation of the J-wave, (D) dyssynchronous biventricular velocity generation, (E) BCG tracing from a HF patient with pulmonary hypertension (75Y, M, mean PAP: 41mmHg), (F) BCG tracing from a healthy control (24Y, F). (E) and (F) have the same vertical scale.

Canadian Institutes of Health Research - Fellowship, University