

Supplementary Online Content

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This supplementary material has been provided by the authors to give readers additional information about their work.

eAppendix. Additional Study Assessment

1. CPET procedures and quality control

Cardiopulmonary fitness was evaluated using a unified pediatric CPET protocol consistent with our previous similar studies.¹⁻³ Exercise laboratories for each center followed a high-quality score rating (≥ 10), according to ATS/ACCP statement on CPET,⁴ employing adapted equipment and procedures: adapted pediatric face masks, calibration of gas analyzers, utilization of breath-to-breath measurement software, 12-lead electrocardiogram equipment, and a pulse oximeter, mask leak assessment before and during exercise with monitoring of VO_2/W ratio. Spirometry using a common gas device was systematically performed before the exercise test with a flow-volume curve and measurement of forced expiratory volume in 1s (FEV1), forced vital capacity (FVC), and the FEV1/FVC ratio (FEV1%), with normalization to theoretical values.⁵

Children in both groups underwent a cycle ergometer to obtain a homogeneous incremental overall duration between 8 and 12 minutes, including a) 1-minute baseline b) 3-minute warm-up (10-20 watts) c) fixed increments of 10-20 watts/min (3.5 W/kg in adolescent boys and 3 W/kg in girls and preadolescent boys); d) pedaling rate of 60-80 revolutions/min; e) 3-minute active recovery; f) 2-minute rest. CPET was considered maximal when one of the following four criteria was reached: (1) respiratory exchange ratio ≥ 1 , (2) maximum heart rate $>85\%$ of maximal age-predicted heart rate ($220 - \text{age}$), (3) limit of the patient's tolerance despite verbal encouragement, and (4) plateau of VO_2 despite increasing exercise intensity.

Upon compilation of data from all centers, most participants met at least two of the CPET maximal criteria, except for two patients (2.0%) and one control (0.5%).

2. Details on physical activity assessment

Data from the accelerometer were processed using a sampling rate of 30 Hz, an epoch of 15 s, and the non-wearing period was calculated using Choi's algorithm.⁶ Intensities were classified using the Evenson threshold: sedentary (0-100 counts/min), light physical activity (101-2295 counts/min), moderate physical activity (2296-4011 counts/min), vigorous physical activity (≥ 4012 counts/min), and moderate-to-vigorous physical activity (MVPA ≥ 2296 counts/min).⁷

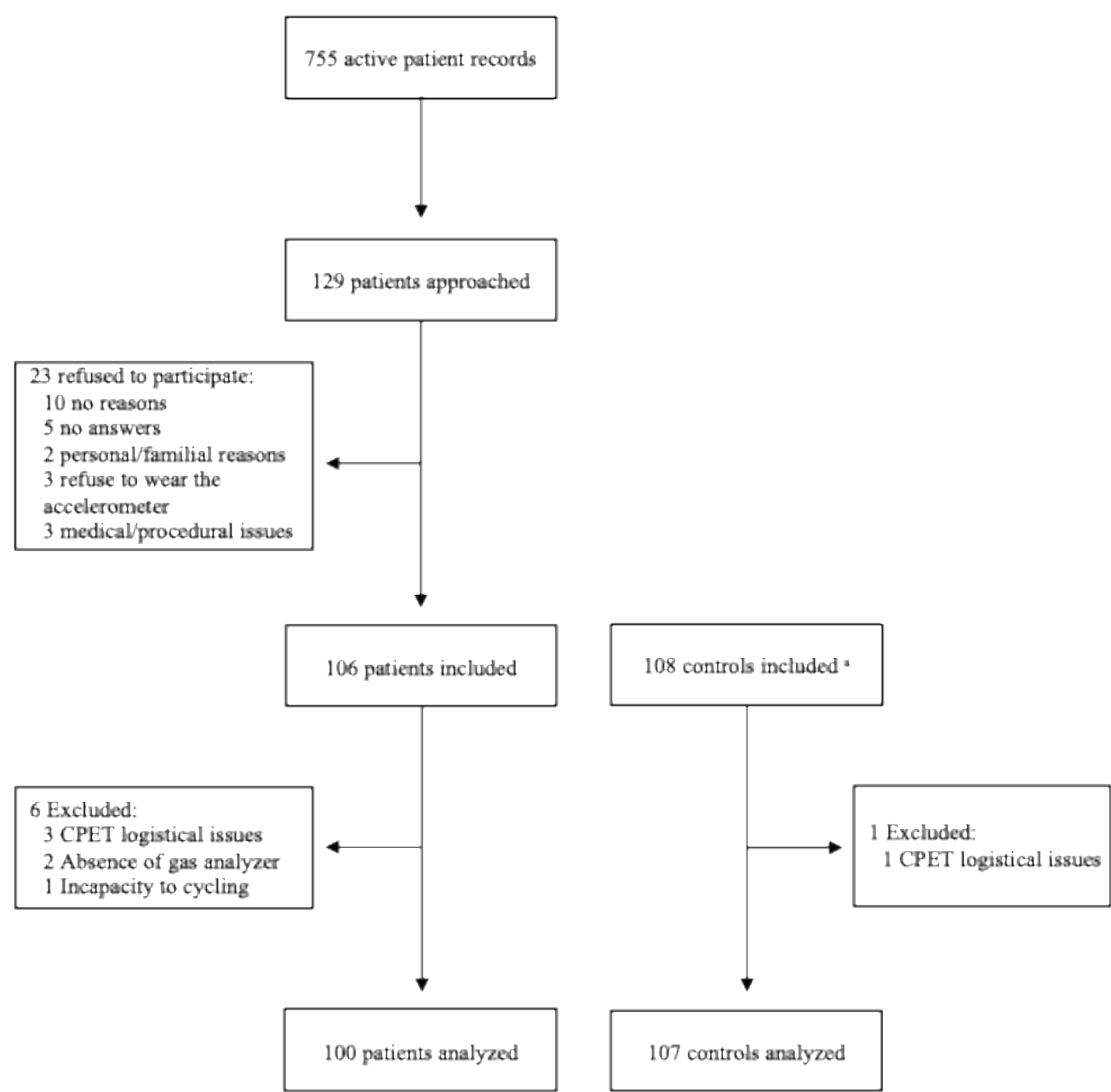
The physical activity questionnaire Ricci-Gagnon,⁸ previously used in pediatric cardiology,⁹ includes nine items, and scoring was performed if all items were collected: one for sedentary behaviors, four for sports and leisure activities, and four for daily life activities.

The motivation for health-oriented physical activity (EMAPS) questionnaire¹⁰ used includes an 18-item divided into three domains and was scored if all items within each domain were present: intrinsic motivation, observed when individuals adopt a behavior for the pleasure and satisfaction they derive from it; extrinsic motivation, observed when behavior is a means of achieving a positive result or preventing a negative one; and amotivation, observed when individuals feel that there is no way for them to obtain positive results from their actions.^{10,11}

eReferences.

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eFigure. Flow Chart of Participants Included in the Analysis



Footnotes: ^a Healthy children and adolescents were recruited upon voluntary participation following normal clinical check-up.

Abbreviations: CPET, cardiopulmonary exercise test.

eTable. Comparison of Z-Score VO_{2max} and VAT Across Subgroups of Patients and Controls

CPET Parameters	Control ^(a) (N=107)	Long QT syndrome ^(b) (N=47)	Hypertrophic cardiomyopathy ^(c) (N=24)	Dilated cardiomyopathy ^(d) (N=18)	ARVC + CPVT ^(e) (N=10)
VO _{2max} Z-score, Mean (SD)	-0.16 (±0.97) ^{b, c, e}	-1.49 (±1.47) ^a	-1.55 (±1.54) ^a	-1.11 (±1.62)	-1.95 (±1.24) ^a
VO ₂ at VAT Z-score, Mean (SD)	0.03 (±0.99) ^{b, c, e}	-1.33 (±1.45) ^a	-1.36 (±1.36) ^a	-0.70 (±1.64)	-1.61 (±1.63) ^a

Footnotes: Superscript letters indicate a significant difference (p<0.05) with the group. For pairwise comparisons, false discovery rate correction was applied. No significant difference was found between patients' subgroups.

Abbreviations: ARVC, Arrhythmogenic right ventricular cardiomyopathy; CPET, cardiopulmonary exercise test; CPVT, Catecholaminergic polymorphic ventricular tachycardia; VAT, ventilatory anaerobic threshold; VO_{2max}, maximal oxygen uptake.