CARDIOLOGY 2025

Encouraging Exercise in the Fontan Circulation: Yes! But How?

Saturday, Feb 22, 2025

Betsy Goldmuntz, MD

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THE PROBLEM

- 4 centers, n=321
- Age 21+/-9 years
- Fontan completion 7.1 =/- 5.9 years
- Included atriopulmonary as well as cavopulmonary anastomosis

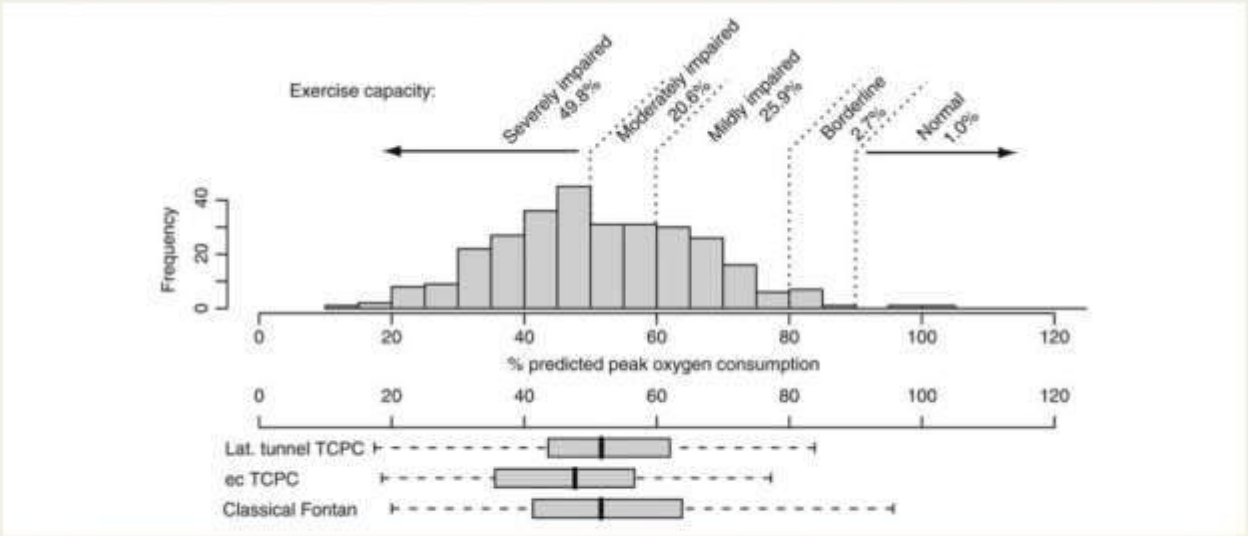

Figure 1 Distribution of % predicted peak oxygen consumption in patients after Fontan operation and its distribution in patients with different types of Fontan surgery.

Diller et al., Eur Heart J 2010

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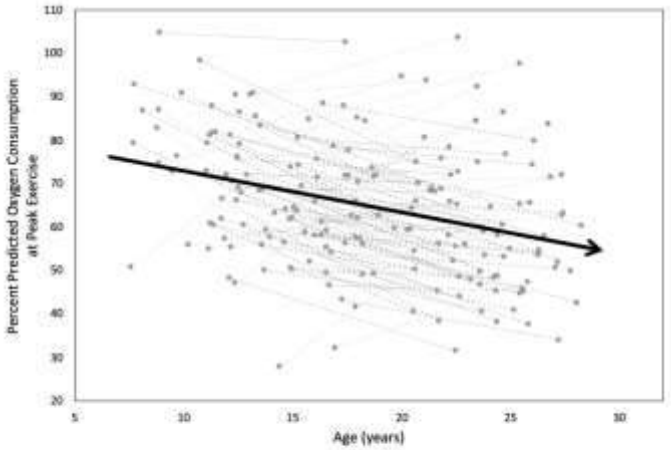
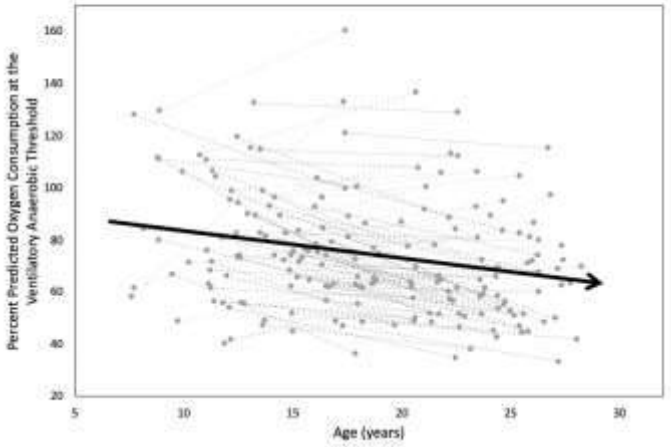
THE PROBLEM

- 95 cases with paired tests from Fontan 1 to Fontan 3 study with RER> 1.1 at both time points
- Percent (%) predicted VO2 at peak exercise and at the Ventilatory Anaerobic Threshold (VAT) declines over time





Goldberg et al., Ped Card 2021

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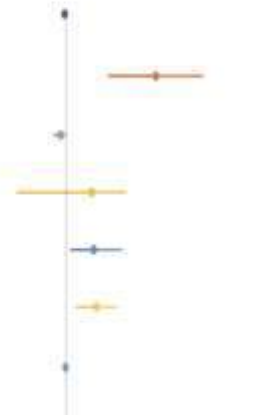
THE PROBLEM

- Systematic review: 7 eligible articles with ≥ 30 subjects and ≥16 yo
- Peak VO2 (percent predicted VO2) was diminished in all studies
- Peak VO2 (percent predicted VO2) as predictor of mortality/transplant was inconsistent between studies
- Decline in VO2 in serial studies associated with mortality/transplant

Table 3A Exercise capacity as a determinant of mortality

Study	Variable	Late outcome	HR (95% CI)
VO_2			
Diller *et al*[22]	Peak VO_2 (mL/kg/min).	Death/transplantation.	0.959 (0.905 to 1.015)
Fernandes *et al*[23]	Peak VO_2 (cut-off value <16.6).	All-cause mortality.	7.5 (2.6 to 21.6)
Ohuchi *et al*[24]	Peak VO_2 (% of predicted value).	All-cause mortality.	0.88 (0.76 to 0.98)
Egbe *et al*[25]	Peak VO_2 (% of predicted value).	Death and cardiac surgery (CAE).	1.77 (0.33 to 3.76)
Egbe *et al*[25]	Peak VO_2 (−3 percentage points/year).	Predictors of 5-year risk of CAE.	1.86 (1.11 to 3.48)
Cunningham *et al*[26]	% Change in peak VO_2, /−10%.	Death/transplantation.	1.96 (1.24 to 3.11)
Atz *et al*[27]	Per cent predicted VO_2 at anaerobic threshold.	Death/transplantation.	0.98 (0.96 to 1.0)



Udholm et al., Open Heart 2018

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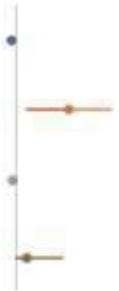
THE PROBLEM

- Systematic review: 7 eligible articles with ≥ 30 subjects and ≥16 yo
- Peak VO2 (percent predicted VO2) was consistently associated with morbidity, particularly cardiac hospitalizations

Table 3B Exercise capacity as a determinant of unscheduled hospitalisation

Study	Variable	Late outcome	HR (95% CI)
VO_2			
Diller *et al*[22]	Peak VO_2 (mL/kg/min).	Hospitalisation.	0.938 (0.912 to 0.965)
Fernandes *et al*[23]	Peak VO_2 (cut-off value <18.9).	Combined end point.	1.95 (1.14 to 3.36)
Ohuchi *et al*[24]	Peak VO_2 (% of predicted value).	Hospitalisation.	0.95 (0.91 to 0.99)
Cunningham *et al*[28]	% Change in peak VO_2, /−10%.	Combined end point.	1.14 (1.0 to 1.8)



Udholm et al., Open Heart 2018

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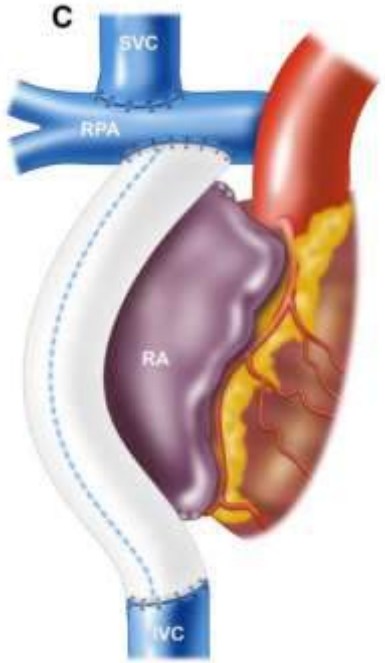
THE REASONS

- Absence of a sub-pulmonary pump
  - “Ceiling” of central venous pressure
  - Limited ability to augment stroke volume in response to demand
  - No pulsatility with diminished recruitment of pulmonary bed
  - Low cardiac output
- Chronotropic incompetence
- Cyanosis
- Restrictive lung function
- Reduced skeletal muscle mass



Rychik et al Circ 2019

THE PAST: 1994

Recommendation. Athletes can participate in low intensity competitive sports (class IA). Selected athletes can engage in sports of either moderate or low static demand if they have normal or near-normal ventricular function and oxygen saturation and near-normal exercise tolerance on formal exercise testing.

Graham et al., JACC 1994

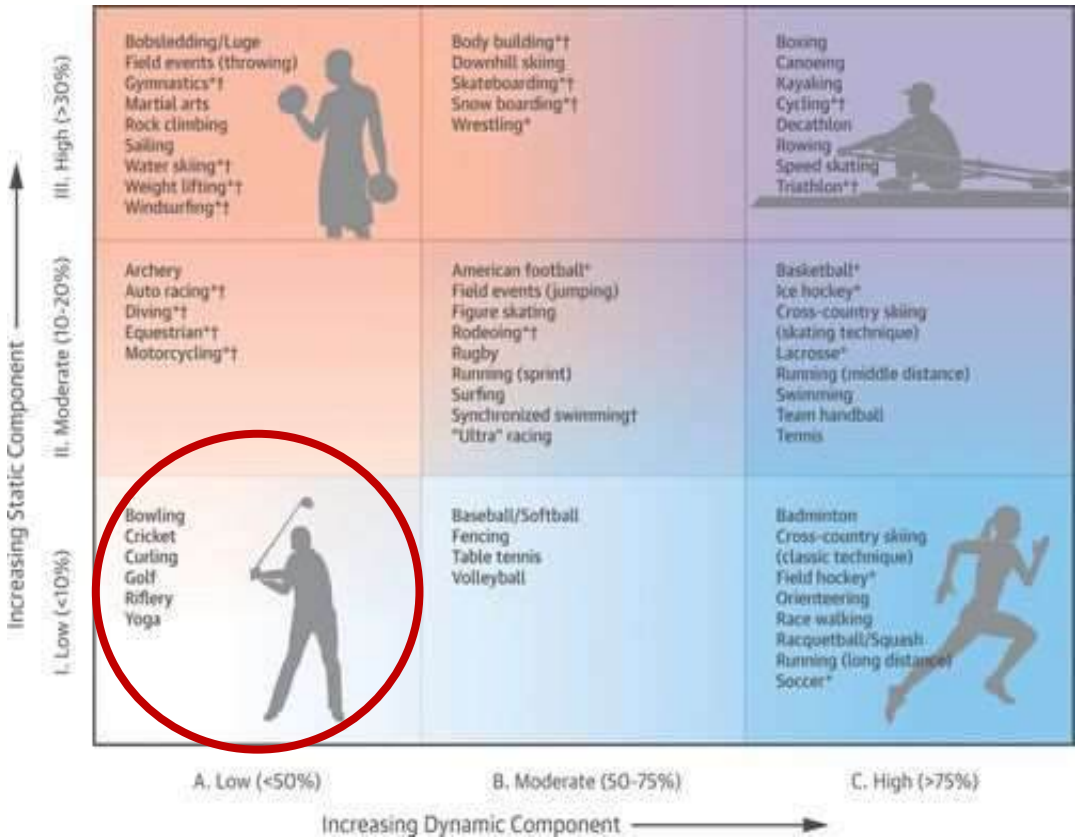
THE PAST: 2015

- ACC/AHA Scientific Statement 2015
- Eligibility and Disqualification Recommendations for Competitive Athletes with CVS Abnormalities: Task Force 4: Congenital Heart Disease

Recommendations

1. **It is recommended that before participation in competitive sports, all athletes who have undergone the Fontan procedure should undergo an evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing *(Class I; Level of Evidence B)*.**
2. **Athletes who have undergone the Fontan procedure and who have no symptomatic heart failure or significantly abnormal intravascular hemodynamics can participate only in low-intensity class IA sports *(Class I; Level of Evidence C)*.**
3. **Participation in other sports may be considered on an individual basis with regard for the athlete's ability to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms *(Class IIb; Level of Evidence C)*.**

THE PAST



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CONSEQUENCES: INACTIVITY

- 147 patients ages 7-18 yrs
- Measured time spent in moderate to vigorous physical activity by accelerometry
- Not related to self-reported activity levels

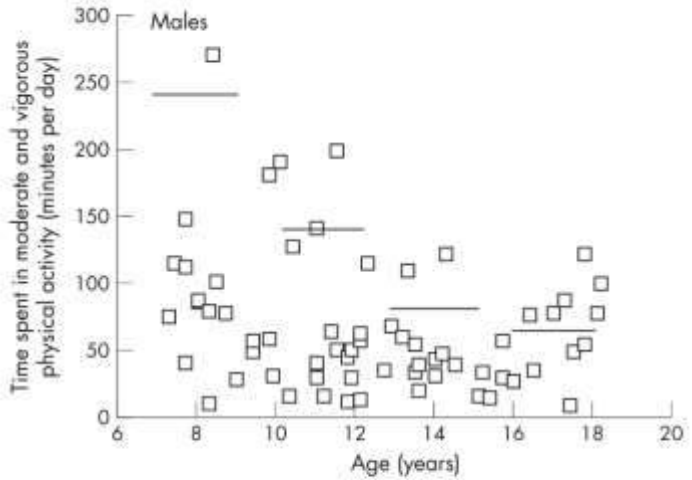

Figure 1 Daily time spent in moderate and vigorous physical activity for each Fontan patient by patient age for males. Reference lines are the age group 50th percentiles for normal, healthy children.[27]

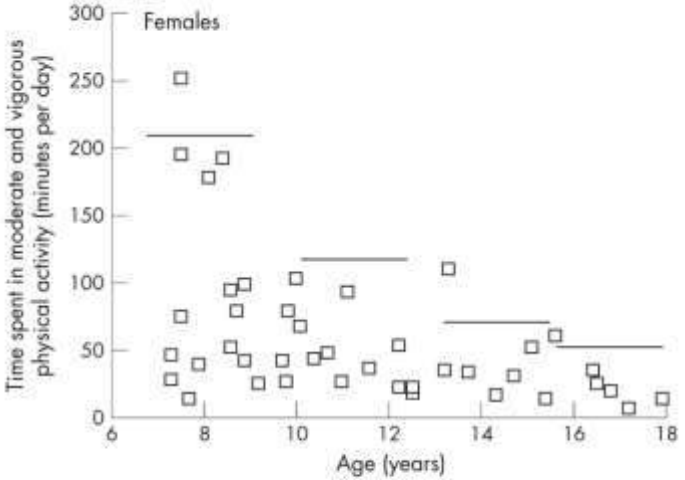

Figure 2 Daily time spent in moderate and vigorous physical activity for each Fontan patient by patient age for females. Reference lines are the age group 50th percentiles for normal, healthy children.[27]

McCrindle et al Arch Dis Child 2007

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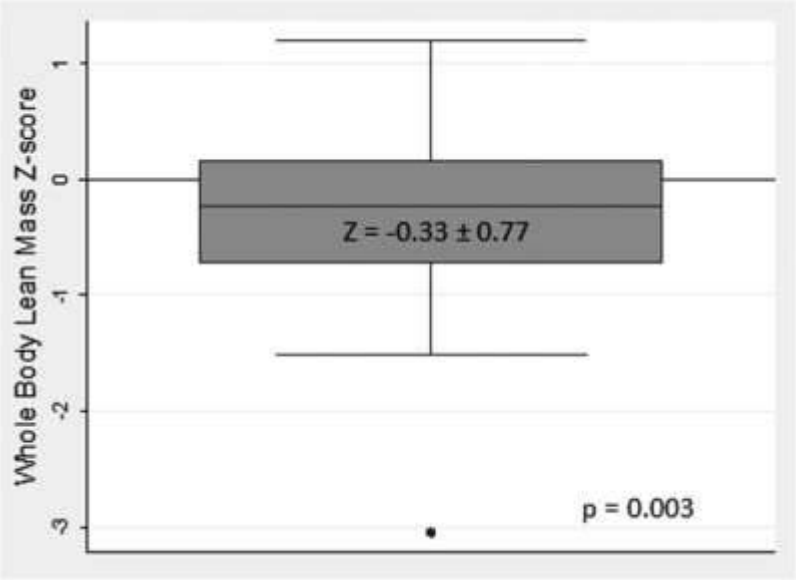
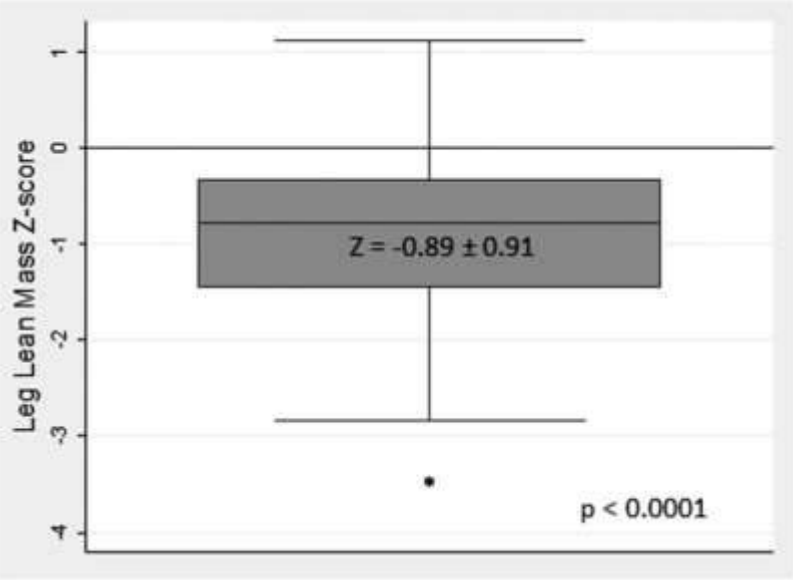
CONSEQUENCES: MYOPENIA

- Leg lean mass in 50 Fontan patients ≥ 5yrs
- Peak VO2 by CPET in 28 cases





Avitabile et al, Congen Heart Dis 2014

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WHAT WERE WE WORRIED ABOUT?

- SUDDEN DEATH
- Long term effects

CARDIAC REHABILITATION: IS IT SAFE?

- Literature does not report sudden death associated with exercise
  - Meta-analysis of 16 prospective intervention studies
  - NO significant exercise-induced adverse events.

Scheffers et al., Eur J of Preventive Cardiology 2020

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ACTIVE PATIENTS HAVE BETTER PEAK VO2

- 112 Fontan with max effort on CPET
- 22 had peak VO2 ≥ 80% predicted (High Capacity Fontan, HCF)
- Multivariable analysis associated with peak VO2:
  - Self-reported active life-style
  - Ventricular systolic function
- Physically active = regular attendance at fitness classes or moderate to vigorous exercise ≥ 4 days/week for ≥ 30 min

Powell et al., Canadian J of Cardiol 2020

Table 5. Results of the univariate and multivariable analyses for percent predicted of VO_2peak

Univariate analysis	R value	*P* value
Active lifestyle	0.45	< 0.0001
Pre-CPET systolic function	0.29	0.004
Postoperative complications	−0.25	0.030
BMI at hospital discharge	−0.25	0.030
Fontan LOS	−0.22	0.040
Cardiopulmonary bypass time	−0.16	0.190
Weight at hospital discharge	−0.13	0.300
Age at CPET	−0.05	0.600
Time from Glenn to Fontan	−0.01	0.900
Height at hospital discharge	0.003	0.900
Multivariable analysis	**Standardized β coefficient (parameter estimates)**	***P* value**
Active lifestyle	0.24 (9.5 ± 4.6)	0.040
Pre-CPET systolic function	−0.20 (−7.4 ± 3.6)	0.040
BMI at hospital discharge	−0.20 (−1.7 ± 0.9)	0.070
Postoperative complications	−0.03 (−1.1 ± 4.9)	0.800
Fontan LOS	−0.02 (−0.03 ± 0.2)	0.900

BMI, body mass index; CPET, cardiopulmonary exercise test; LOS, length of stay; VO_2, oxygen uptake.

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ACTIVE PATIENTS HAVE BETTER PEAK VO2

- 115 patients with complete extracardiac conduit Fontan had CPET
- 6-20 years, average 12.8 years
- Participation in sports clubs in middle (junior high school) and high school “not highly recommended” but 23% reported participating anyway.

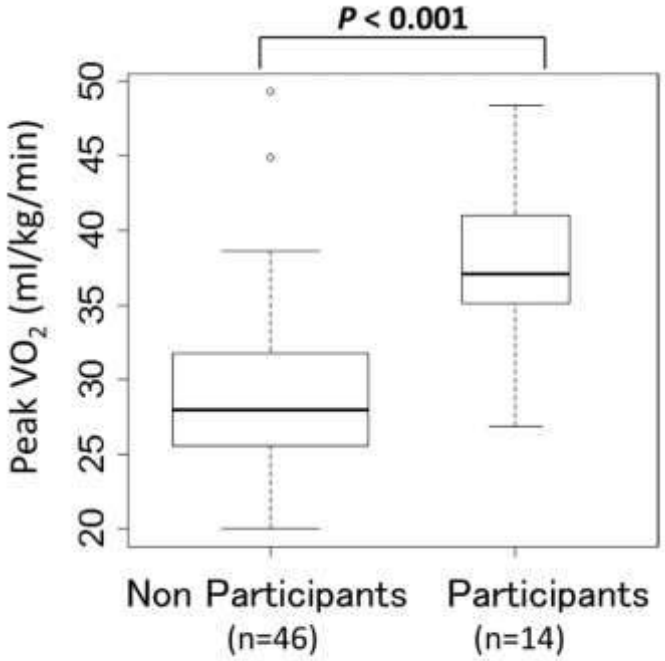
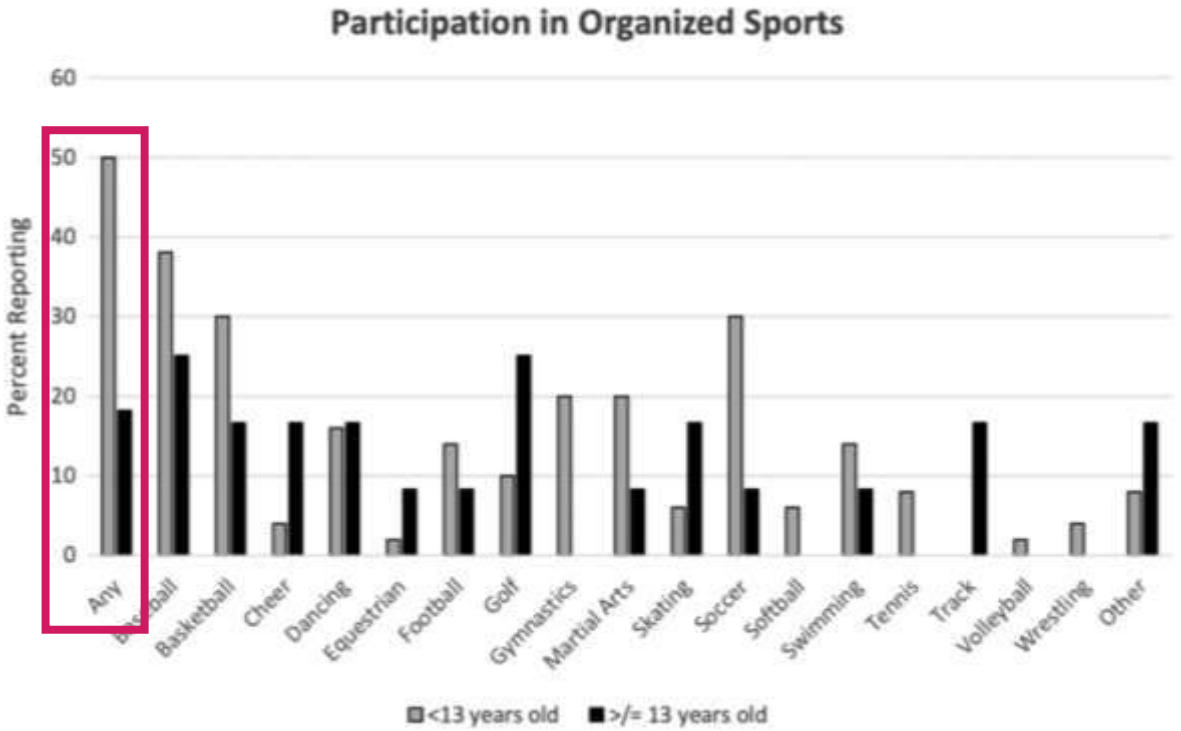


Fig. 2 The difference in peak VO2 between non-participants and participants in sports clubs

Kodama et al Ped Cardiol 2018

WHAT ARE OUR PATIENTS DOING?



Pierick et al., Cardiol Young 2024

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WHAT ARE OUR PATIENTS DOING?

Table 2. Individual participation in gym class at school

	Full participation	Limited participation	No participation
All	36/103 (35%)	62/103 (60.2%)	5/103 (4.9%)
< 13 years old	33/81 (40.7%)	46/81 (56.8%)	2/81 (2.5%)
≥ 13 years old	3/22 (13.6%)	16/22 (72.7%)	3/22 (13.6%)

Pierick et al., Cardiol Young 2024

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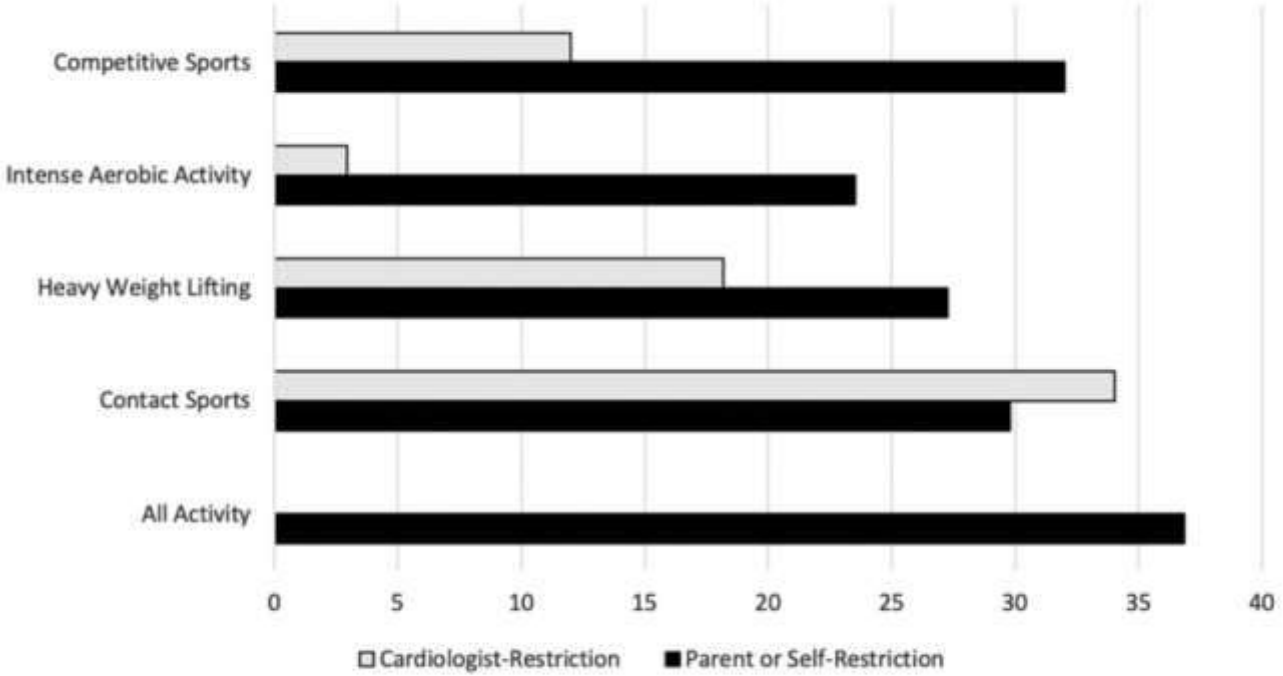
WHY AREN'T THEY MORE ACTIVE?



Pierick et al., Cardiol Young 2024

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EXERCISE: YES! BUT HOW?

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EXERCISE: YES! BUT HOW?

- We have to believe in it!

EXERCISE: YES! BUT HOW?

- TALK ABOUT IT!

FONTAN OUTCOME NETWORK (FON): PHYSICAL ACTIVITY AND IMPROVEMENT PROJECT

- Assess and Support Physical Activity
  - Assess current physical activity levels using the PROMIS physical activity assessment tool
  - Review and discuss the PROMIS score with patients and families

FONTAN OUTCOMES NETWORK

	In the past 7 days…	No days	1 day	2-3 days	4-5 days	6-7 days
PAC_M_009R1	How many days did you exercise or play so hard that your body got tired?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_105R1	How many days did you exercise really hard for 10 minutes or more?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_002R1	How many days did you exercise so much that you breathed hard?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_008R1	How many days were you so physically active that you sweated?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_010R1	How many days did you exercise or play so hard that your muscles burned?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_011R1	How many days did you exercise or play so hard that you felt tired?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_114R1	How many days were you physically active for 10 minutes or more?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
PAC_M_134R1	How many days did you run for 10 minutes or more?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

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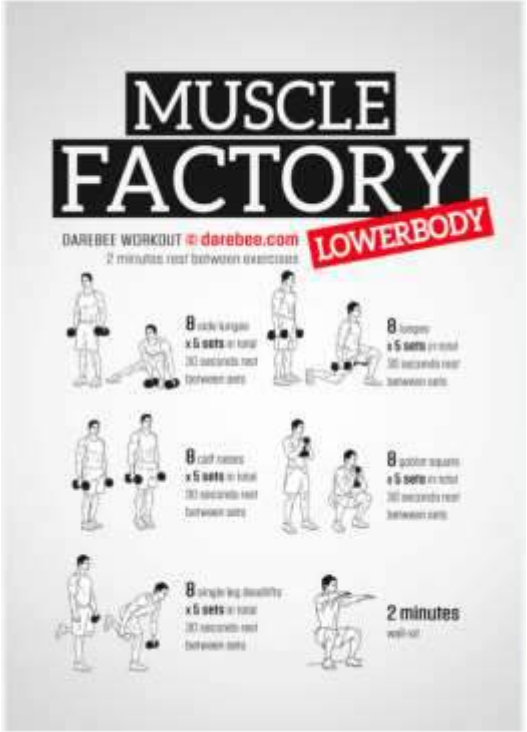
FONTAN CARDIAC REHABILITATION

Aerobic Training



Lower Extremity Resistance Training

Inspiratory Muscle Training

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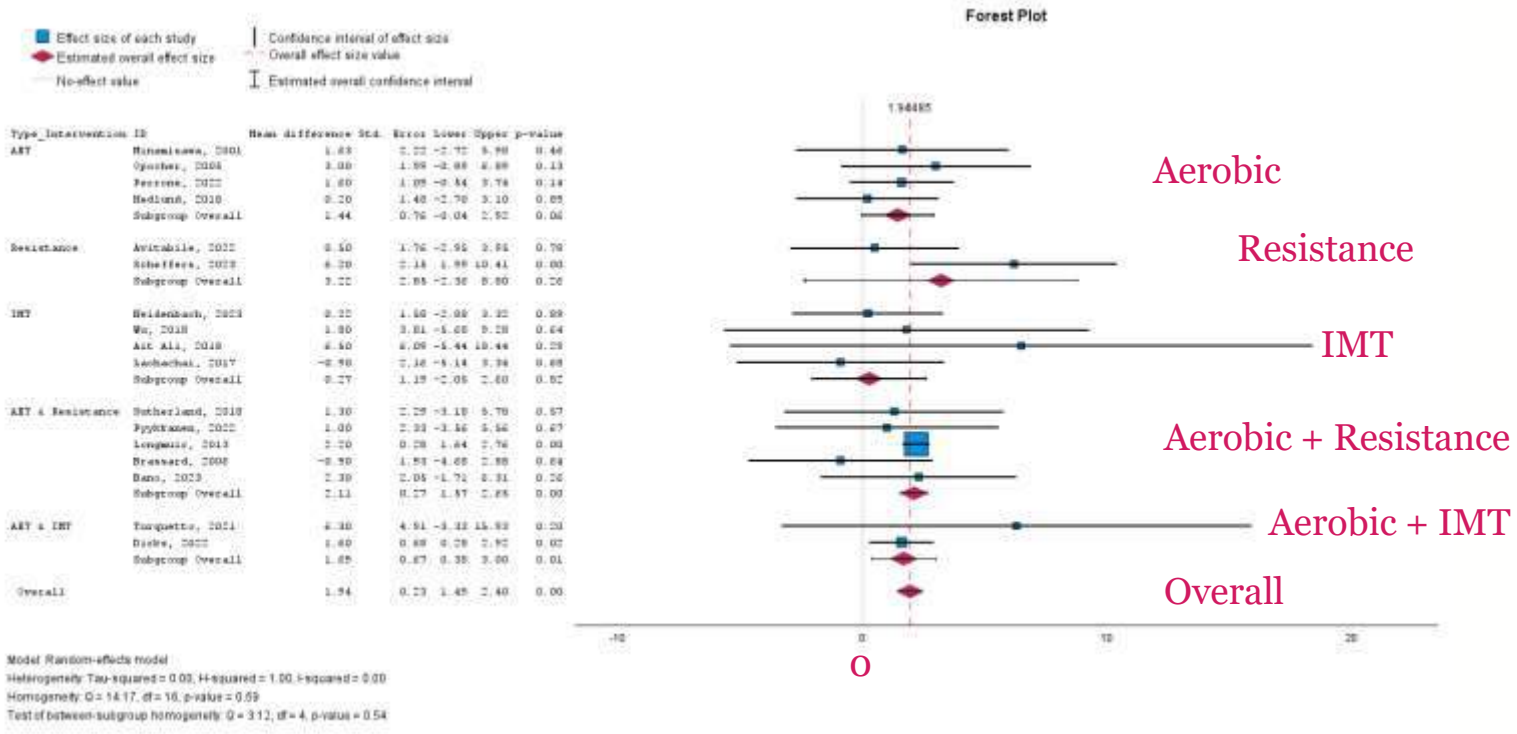
CARDIAC REHAB: TYPE OF INTERVENTION
(OUTCOME = PEAK VO_2)



Luna van de Ven et al., Medicina 2024

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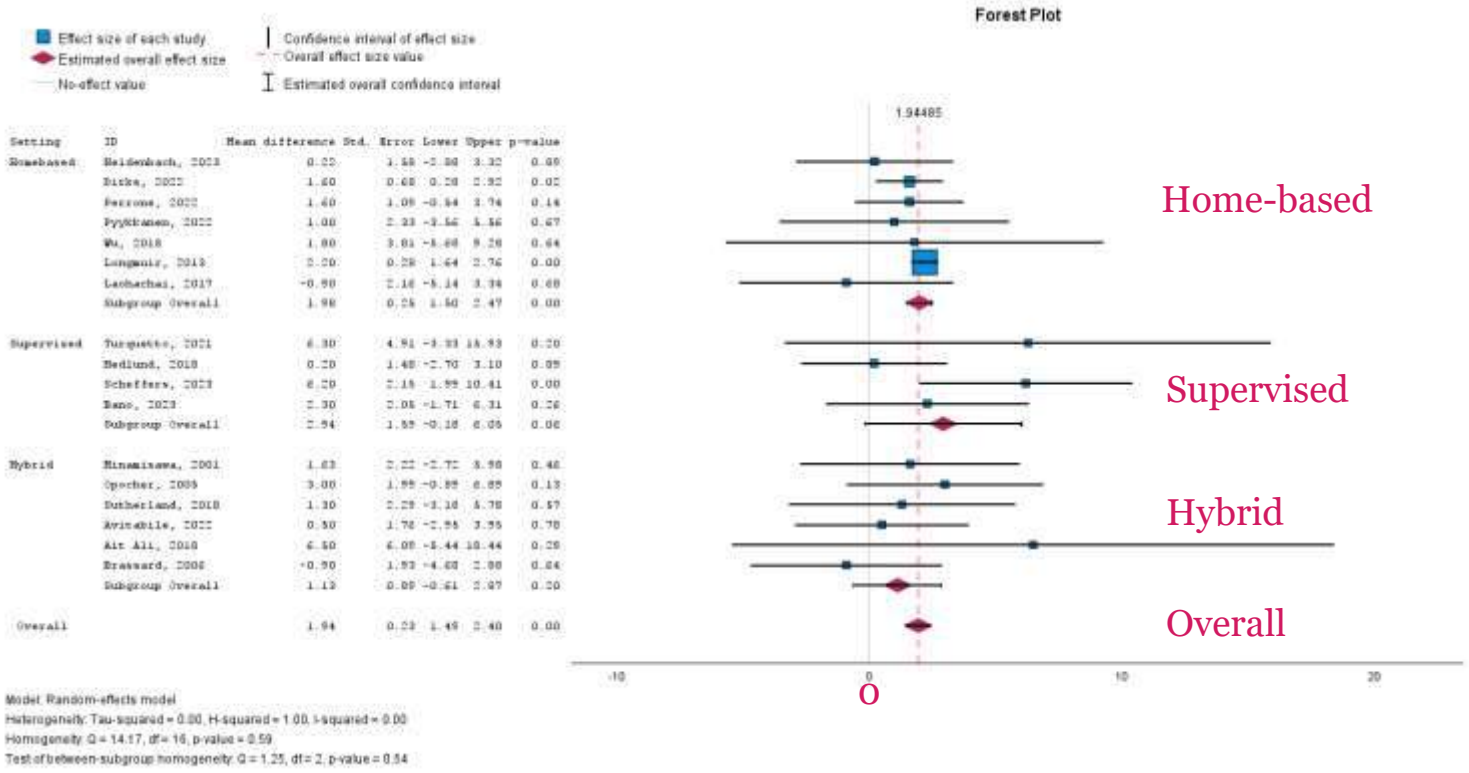
CARDIAC REHAB: BEST LOCATION?
(OUTCOME = PEAK VO_2)



Setting	ID	Mean difference	Std. Error	Lower	Upper	p-value
Homebased	Heidenbach, 2023	0.22	1.58	-2.88	3.32	0.89
	Dirks, 2022	1.60	0.68	0.26	2.92	0.02
	Perrone, 2022	1.40	1.09	-0.84	3.74	0.14
	Pyykkonen, 2022	1.00	2.33	-3.56	5.56	0.67
	Wu, 2018	1.80	3.81	-5.68	9.28	0.64
	Longmuir, 2013	2.20	0.28	1.64	2.76	0.00
	Lachachai, 2017	-0.90	2.16	-5.14	3.34	0.68
	Subgroup Overall	1.98	0.26	1.50	2.47	0.00
Supervised	Turquetto, 2021	6.30	4.91	-3.33	15.93	0.20
	Hedlund, 2018	0.20	1.48	-2.70	3.10	0.89
	Scheffers, 2023	6.20	2.15	1.99	10.41	0.00
	Bano, 2023	2.30	2.06	-1.71	6.31	0.26
	Subgroup Overall	2.54	1.55	-0.18	6.06	0.06
Hybrid	Hinamizawa, 2001	1.03	2.22	-2.72	5.98	0.46
	Opocher, 2005	3.00	1.99	-0.89	6.89	0.13
	Sutherland, 2018	1.30	2.29	-3.18	5.78	0.57
	Avitabile, 2022	0.50	1.76	-2.95	3.95	0.78
	Ait Ali, 2018	6.50	6.09	-5.44	18.44	0.28
	Brassard, 2008	-0.90	1.93	-4.68	2.88	0.64
	Subgroup Overall	1.19	0.89	-0.61	2.97	0.20
Overall		1.94	0.23	1.49	2.40	0.00

Model: Random-effects model
Heterogeneity: Tau-squared = 0.00, H-squared = 1.00, I-squared = 0.00
Homogeneity: Q = 14.17, df = 16, p-value = 0.59
Test of between-subgroup homogeneity: Q = 1.25, df = 2, p-value = 0.54

Luna van de Ven et al., Medicina 2024

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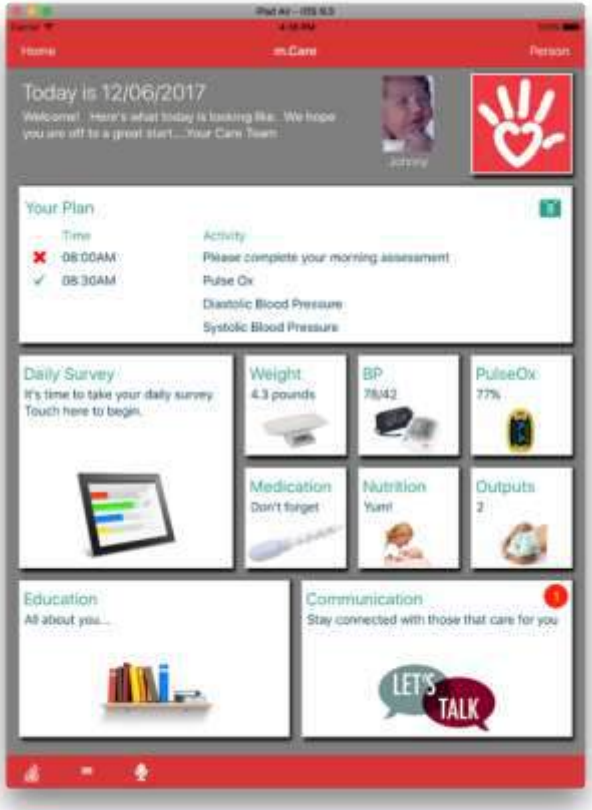
TOOLS TO AUGMENT PARTICIPATION

- Commercial wearables
  - Immediate feedback to patient
  - Proprietary algorithms
- Research grade wearables
  - 3 axial acceleration data to measure physical activity
  - Collect data 10-200x/second
  - Raw data collection allows application of different (and known) algorithms to quantify physical activity

TOOLS TO AUGMENT PARTICIPATION

- Apps
  - Setting goals/schedules
  - Tracking activity
  - Communication
  - Instructional videos
  - Customizable to programs



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TOOLS TO AUGMENT PARTICIPATION

- Incentives
  - Gain-framed: awards accumulated if behavior change goals achieved
  - Loss-framed: upfront endowment with deductions if goals not achieved
- Competition
  - Individual or group
  - Gamification

NEW TRIALS

Exercise Intolerance, Benefits, and Prescription for People Living With a Fontan Circulation: The Fontan Fitness Intervention Trial (F-FIT)—Rationale and Design

Derek L. Tran[1,2,3,4†], Hannah Gibson[3†], Andrew J. Maiorana[5,6], Charlotte E. Verrall[7,8], David W. Baker[1,2], Melanie Clode[9], David R. Lubans[10], Diana Zannino[9], Andrew Bullock[11], Suzie Ferrie[12], Julie Briody[13], Peter Simm[9], Vishva Wijesekera[14], Michelle D'Almeida[3], Sally E. Gosbell[2,3,8], Glen M. Davis[4], Robert Weintraub[9,15], Anthony C. Keech[1,2,16], Rajesh Puranik[1,2], Martin Ugander[17], Robert Justo[18], Dominica Zentner[19,20], Avik Majumdar[2,21], Leeanne Grigg[19,20], Jeff S. Coombes[22], Yves d'Udekem[23], Norman R. Morris[24,25], Julian Ayer[7,8], David S. Celermajer[1,2,3] and Rachael Cordina[1,2,3,9]*

Frontiers in Pediatrics 2022

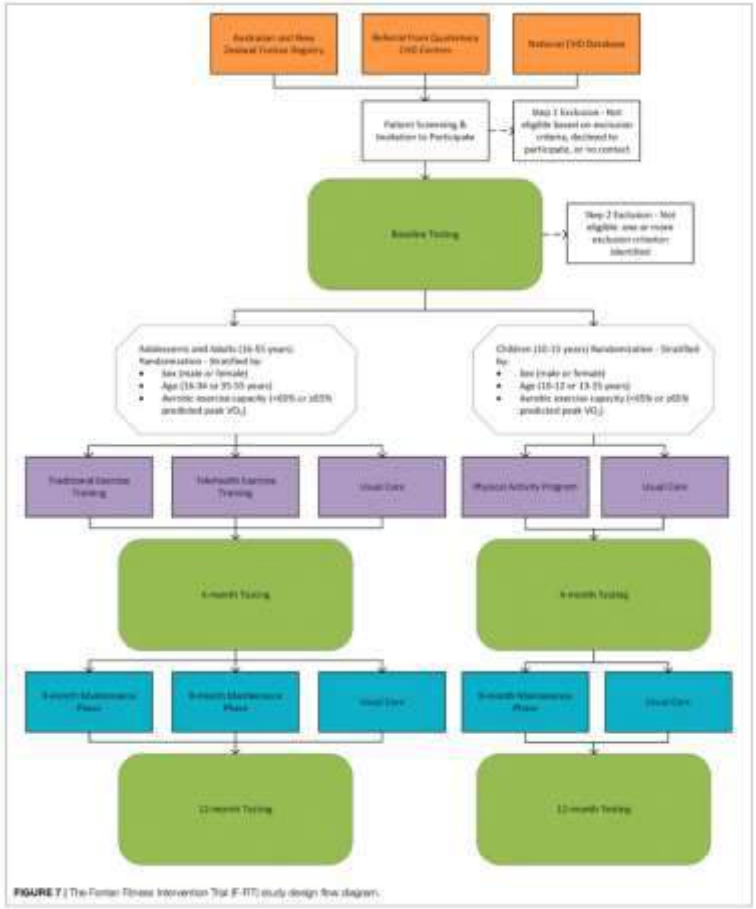

FIGURE 1 | The Fontan Fitness Intervention Trial (F-FIT) study design flow diagram.

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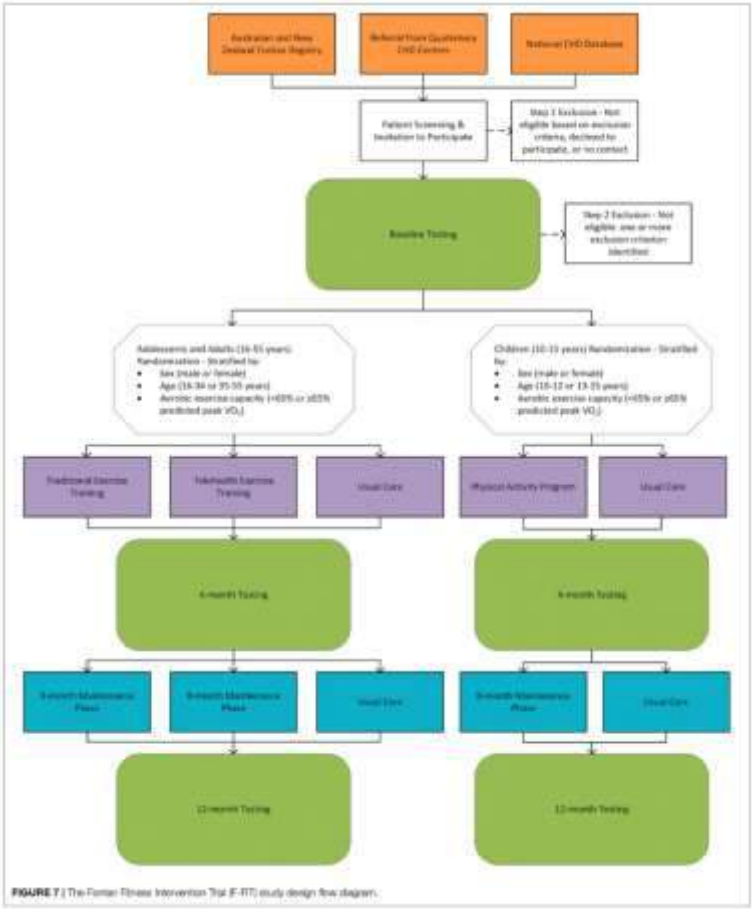
NEW TRIALS

- 110 Adolescents and Adults (16-55 yrs)
  - Gym-based: MVPA 3x/wk, 4 mos, supervised groups in local facility
  - Telehealth: MVPA 3x/wk, 4 mos partially supervised
  - Usual care
- 70 children (10-15)
  - Exercise training program: 1x/wk, 4 mos in person, small local groups, and tasks to complete
  - Usual care
- Detailed evaluation at enrollment
  - 4 month intervention
  - 8 month maintenance phase
- Repeat evaluations at 4 and 12 months



Tran et al., Frontiers in Pediatrics 2022

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NEW TRIALS

TABLE 2 | Assessments and testing.

	Outcomes measures	
Cardiopulmonary exercise testing	Aerobic exercise capacity (peak VO_2)[‡], V_E/VCO_2 ratio and slope, RER, HR, OUES, VO_2 at AT, work rate, oxygen pulse, VO_2/work rate slope, and peripheral venous pressure	
Respiratory muscle and lung function tests	FEV_1, FVC, FEV_1/FVC ratio, TLC, DL_{CO}, PImax, and PEmax	
Dual-energy x-ray absorptiometry	Lean mass, fat mass, bone mineral content, and bone mineral density[†]	
Liver elastography	Liver stiffness	
Near-infrared spectroscopy[†]	HHb, HbO_2, and skeletal muscle oxidative capacity	
Neurocognitive function assessment (Cogstate)	Psychomotor function, attention, visual learning and memory, verbal learning and memory, processing speed, social-emotional cognition, working memory, and executive function scores	
Habitual physical activity (accelerometers; Actigraph GT9X Link)	Counts per minute, steps per day; and time spent in sedentary, light, moderate, vigorous, and moderate-to-vigorous activity	
Nutrition and dietary assessments (ASA24, SGA[†] or SGNA[†], GSRS[†], and indirect calorimetry[†])	SGA (in adults)/SGNA (in children) classification of nutritional status; GSRS (reflux, abdominal pain, indigestion, diarrhea, constipation scores, and total score); dietary macronutrient and micronutrient intake and composition, and REE	
Flow-mediated dilation (FMD)[†]	FMD% (Δ diameter), baseline diameter, peak diameter, and time to peak	
Laboratory and biochemical investigations	NT-proBNP and metabolomic analysis	
Transthoracic echocardiography	AVV S/D ratio, valvular function, VTI, annulus size, aortic flow, and ventricular function	
Resting and exercise cardiac MRI[†]	Ventricular volumes (end-diastolic, end-systolic, stroke volume), ejection fraction, flows (aortic, vena caval), diastolic function (feature tracking, T1 mapping E'), pulmonary artery size (Nakata index), lung water density, hepatic T1 mapping, and AV valve function	
Anthropometry and BIA	Height, weight, waist circumference, BMR, total body water, %BF, and skeletal muscle index	
Quality of life (PedsQL core and cardiac modules)	Physical functioning, emotional functioning, social functioning, school/work functioning, psychosocial functioning, heart problems and treatment, perceived physical appearance, treatment anxiety, cognitive problems, communication and total scores	
	Adolescents and adults	**Children**
Musculoskeletal fitness testing	Chest press 1RM, leg press 1RM, number of leg press repetitions at 70% 1RM (muscular endurance), and handgrip strength	Number of sit-ups, number of push-ups, standing long jump distance, and handgrip strength
Health economic analysis (EQ-5D-5L, CHU-9D, patient cost, and health care expenditure data linkage)	Health state in EQ-5D dimensions, patient cost, and health care utilization	CHU-9D scores, patient cost, and health care utilization

Tran et al., Frontiers in Pediatrics 2022

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NEW TRIALS

Study Protocol of the Exercise Study: Unraveling Limitations for Physical Activity in Children With Chronic Diseases in Order to Target Them With Tailored Interventions—A Randomized Cross Over Trial

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Goals:

1. Limitations to PA in children with different chronic diseases
2. Effects of tailored exercise regimen, possibly with dietary advice and/or psychological counseling

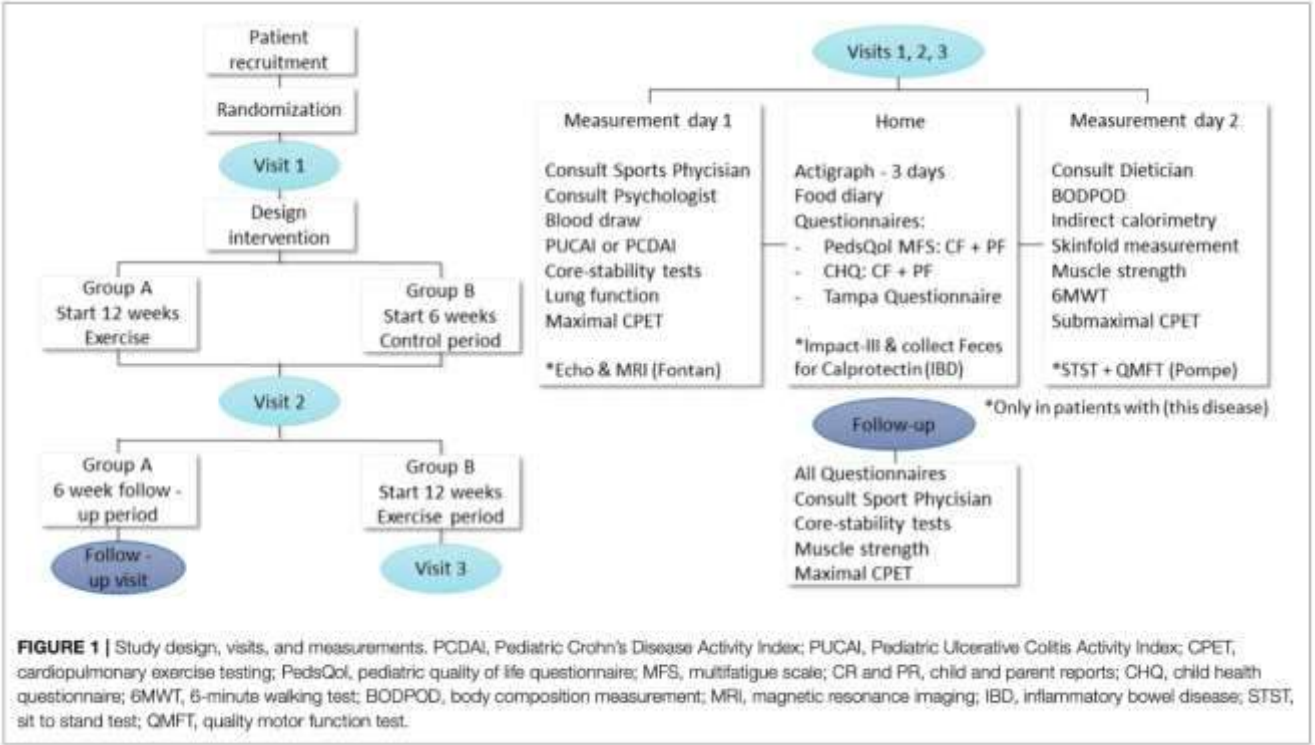

FIGURE 1 | Study design, visits, and measurements. PCDAI, Pediatric Crohn's Disease Activity Index; PUCAI, Pediatric Ulcerative Colitis Activity Index; CPET, cardiopulmonary exercise testing; PedsQol, pediatric quality of life questionnaire; MFS, multifatigue scale; CR and PR, child and parent reports; CHQ, child health questionnaire; 6MWT, 6-minute walking test; BODPOD, body composition measurement; MRI, magnetic resonance imaging; IBD, inflammatory bowel disease; STST, sit to stand test; QMFT, quality motor function test.

Scheffers et al Frontiers in Peds 2022

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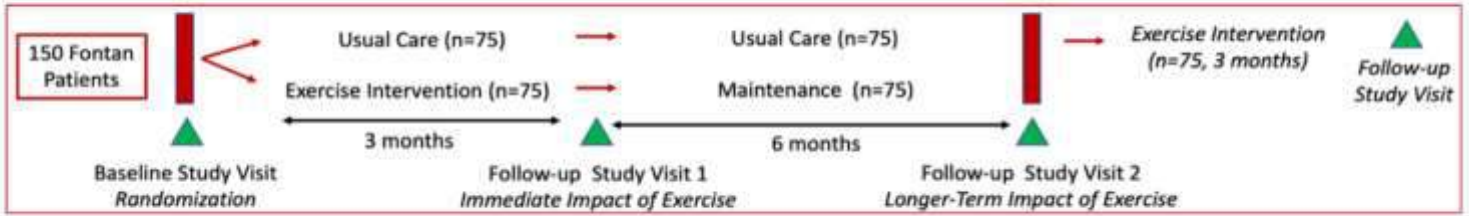
NEW TRIALS

- 150 Fontan patients (9-19 yrs): RCT into intervention or usual care
- Intervention:
  - Live-video-supervised exercise sessions in small peer groups: 3x/wk, 3 mos
  - Maintenance: live-video 1x/wk, independently 2x/wk
- Exercise includes aerobic and resistance training

Figure 5

Supervised-live-video exercise.

Figure 1



Study design.

Tierney et al., Am Heart J 2023

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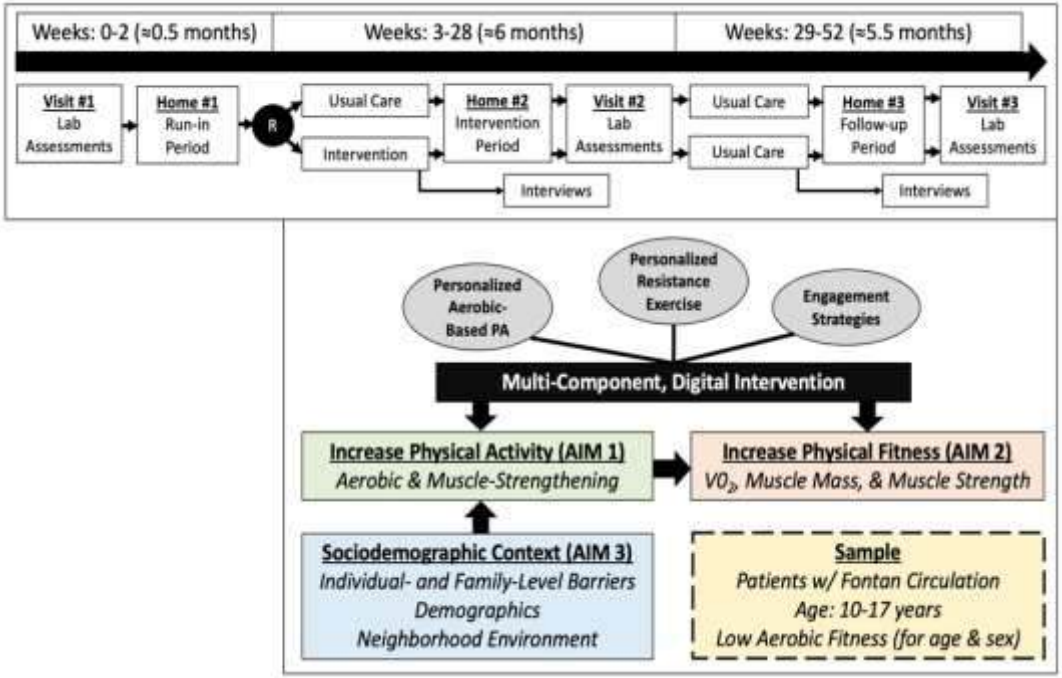
NEWEST TRIAL: FONTAN FITNESS AT CHOP

- Randomized case-control (n=120)
- Home-based, wearable device
- App for schedule, instructions & videos.
- Personalized physical activity designed with exercise physiologist.
- Aerobic and resistance
- 6 mos intervention, 6 mos f/u
- Engage family members
- Incentives included
- Goals: sustained ↑ PA and peak VO2
- Identify facilitators and impediments
- Identify why some "succeed" and some don't to modify design
- Work towards an actual "prescription"
- Generalizable
- Accessible to all



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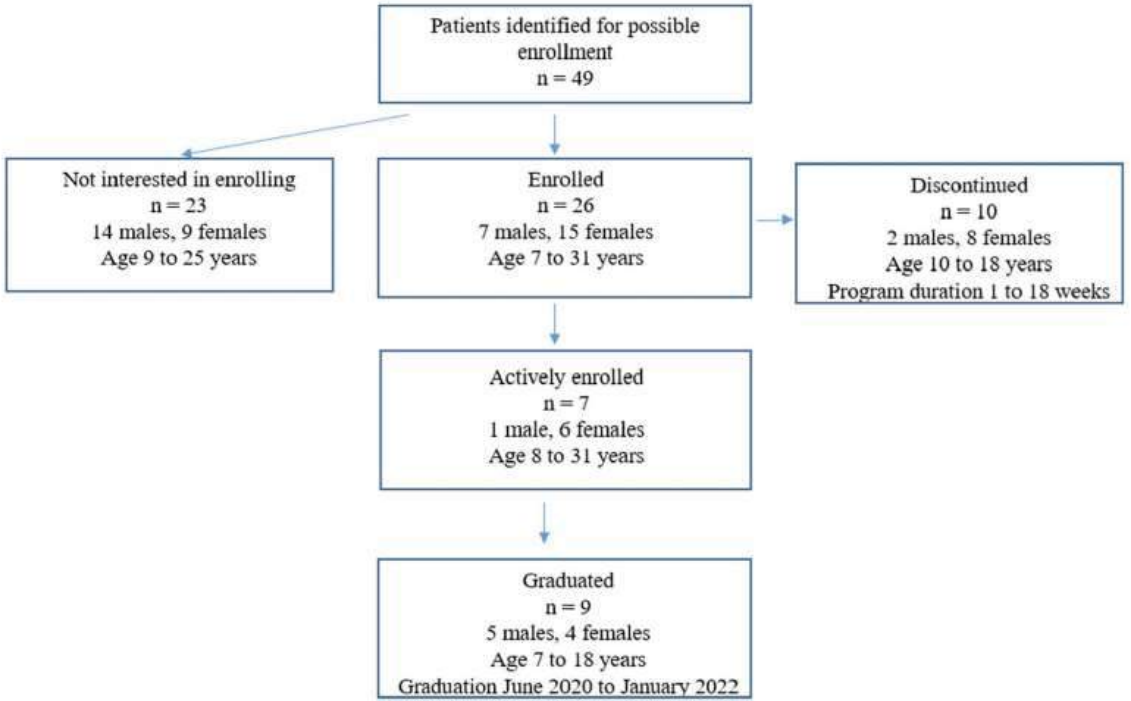
TRIALS AND TRIBULATIONS

- The PREVAIL study in Norway for complex CHD
  - Multicenter, RCT using e-Health, individually tailored encouragements to be physically active for one year (design published in 2012)
  - Klausen et al. Internat'l J of Cardiol vol 221, 2016 publish results:
    - 158 cases, 13-16 yrs with complex CHD rec'd intervention for 52 weeks.
    - Adding tailored e-Health intervention had NO IMPACT on fitness, physical activity, or HRQoL
    - Challenges: interest and retention
- McKillop et al. Pediatr Phys Ther vol 30, 2018
  - Pilot study in 36 patients to assess effect of motivational interviewing on physical activity (MVPA) in adolescents with CHD
  - Both cases and controls given individualized exercise intervention but cases received 12 weeks of motivational interviews
  - Interviews were acceptable but showed no effect on self-efficacy or exercise
  - Possible that a small study not well powered to detect changes

TRIALS AND TRIBULATIONS: HEART CHARGERS

- 12-month partially reimbursable, home-base, personalized exercise prescription based on EST
- Given commercial monitoring device
- Aerobic activities, skeletal and inspiratory muscle training
- Support team communicating by phone, email and telemed check-ins



Fernie et al., Pediatr Cardiol 2023

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HOW DO WE GET ANYONE TO EXERCISE?

- In hospital versus at home?
- What kind of monitoring?
- What kind of communication?
- Prescribed or personalized?
- Family engagement?
- Incentives?
- Generalizability?
- Incorporating into real life?
- Sustain the effort?
- Something we can prescribe?

HOW DO WE HELP FONTAN PATIENTS EXERCISE?

- Safety and efficacy
- Personalized intervention
- Understand/change attitudes
- Facilitators/impediments
- As compared to or in combination with what other intervention?

FUNDAMENTAL QUESTIONS FOR FONTAN PATIENTS

- Which came first:
  - Are they “Super Fontan’s” and therefore they CAN exercise? OR,
  - Are they “Super Fontan’s” BECAUSE they exercise?

- If they are able to increase their peakVO2, will that translate into better long-term outcomes?

- How can the adolescent participate in organized sports or physical activity given competitive nature high-school teams?

EXERCISE, YES! BUT HOW (NOW)?

- First, let's talk about it! Let's measure it!
- Second, combination of aerobic and resistance workouts but details TBD.
- Third, location based on availability and need but should not be an impediment.
- **For now: just do something!**

IMAGINE THE FUTURE: A NEW PRESCRIPTION

- Create the mindset: Promote physical activity from the very beginning
- Develop age- and physiologically appropriate standards from which we can set goals
- Measure reported activity at every visit
- Measure actual activity periodically with wearable device
- If goals not met on measurement, prescribe intervention beyond encouragement
  - Consider current medical status and level of physical activity
  - Use wearable device and app to implement plan, track progress
  - Personalized, home and family based.
  - Goal: modify behavior
- Challenges:
  - Same as any other child/person without Fontan
  - Specific fears/medical and physical limitations of the Fontan
  - Integration into a competitive sports environment

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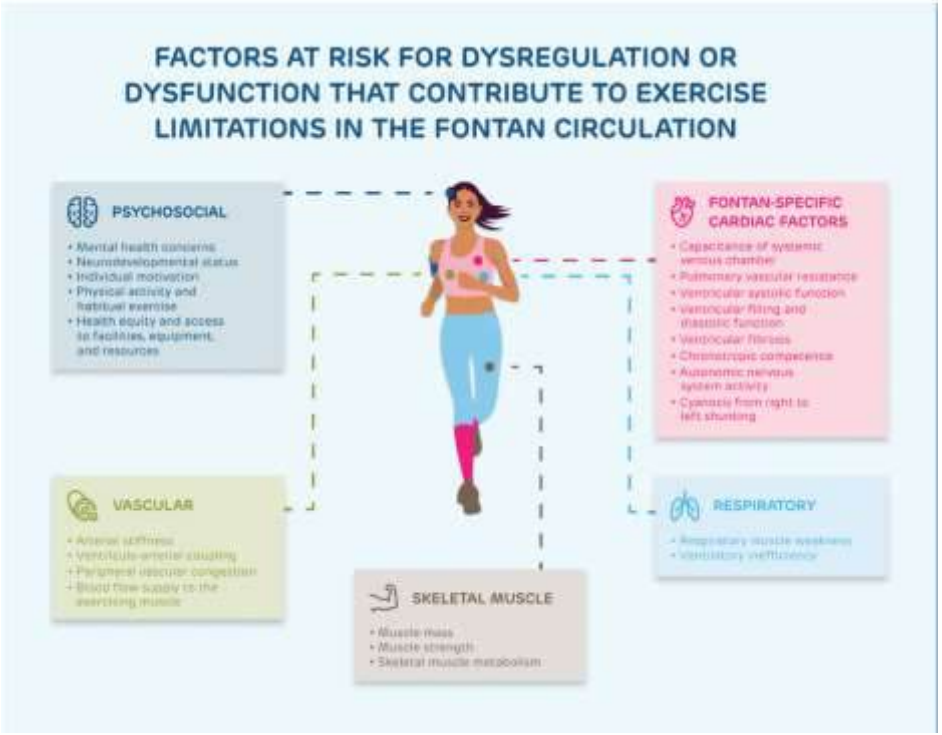
THE HOPE AND PROMISE?

- Improve medical outcomes and longevity
- Obviate the need for (some) medications
- Improve psychosocial aspects of chronic disease
- Likely many other benefits…



Avitabile et al., Int J of Cardiol 2021

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