Exercise Capacity in Children and Young Adults after Repair of Congenital Heart Disease

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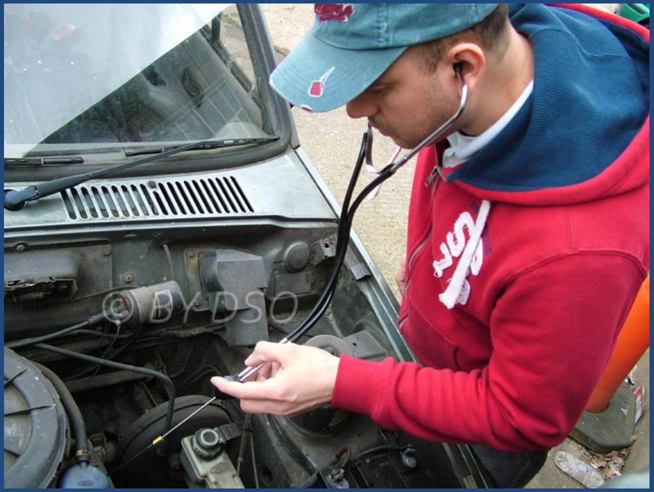
I have no disclosure

Introduction

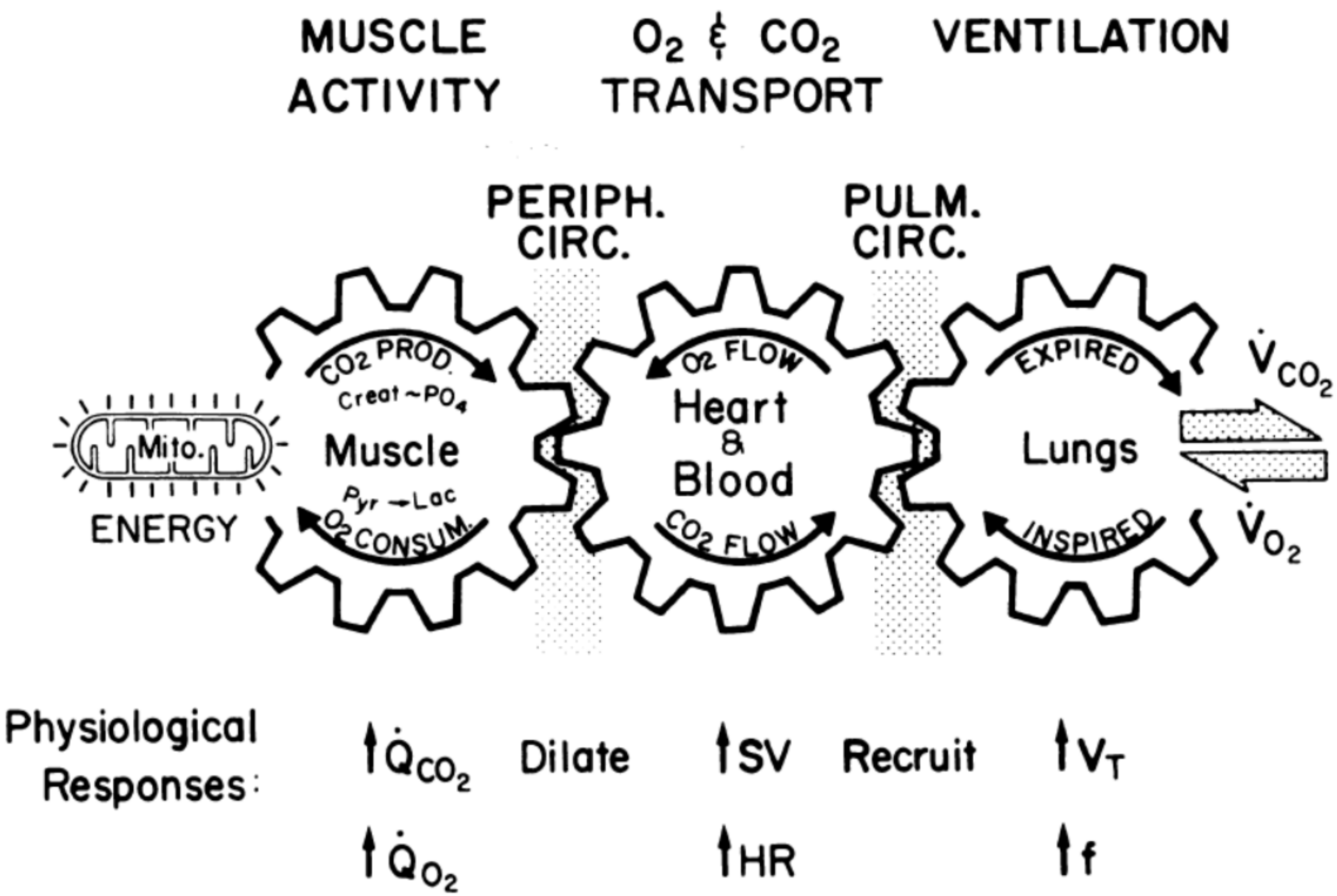
- CHD patients are usually followed-up using resting echocardiography/MRI, and no *functional* tests are performed

- Cardiopulmonary exercise testing (CPET) enables evaluation of maximal cardiac capacity, providing important information on functional outcome.

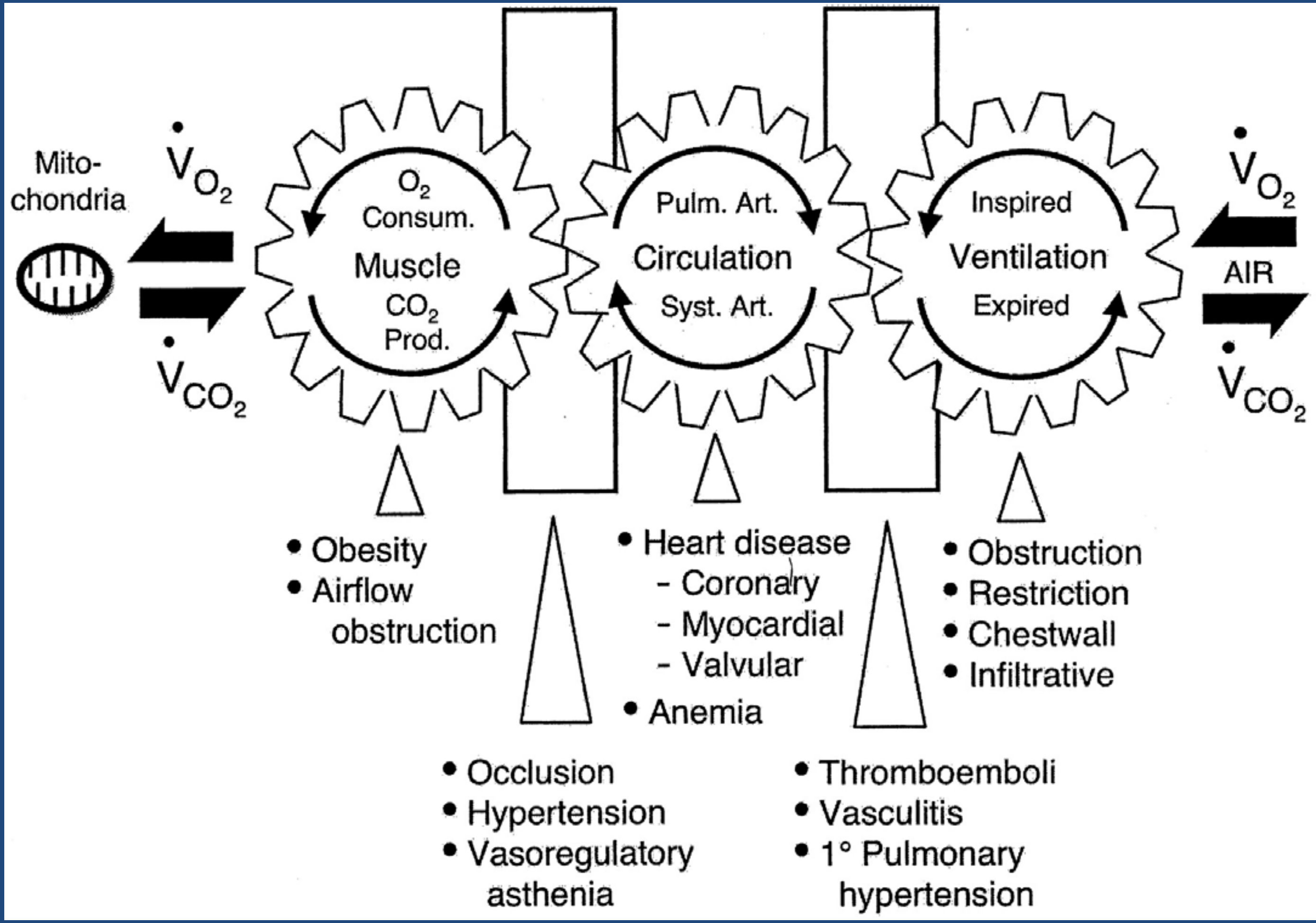
Introduction



Introduction- Gas exchange



Derangements of gas exchange in disease



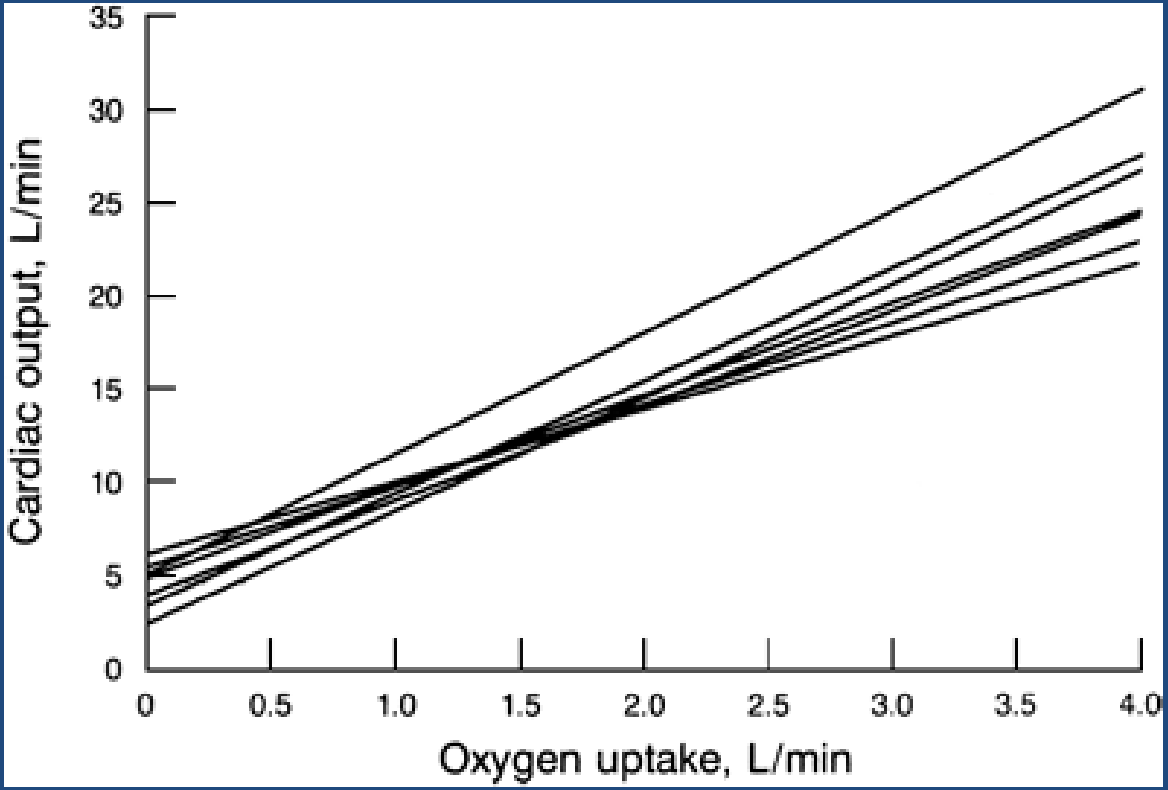
Milani, R. V. et al. Circulation 2004;110:e27-e31

Circulation

VO_2

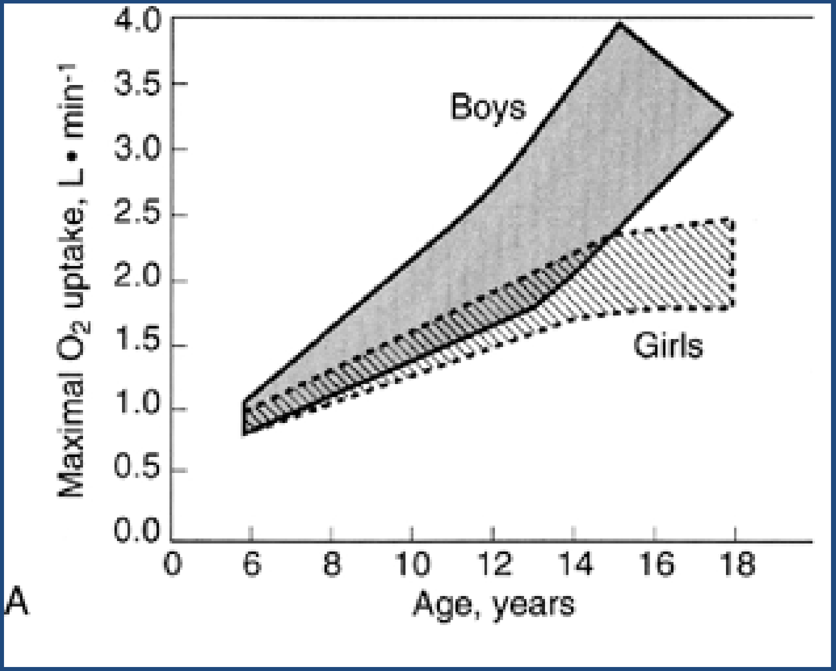
- Oxygen uptake
  - determined by cellular demand
  - level of maximal O_2 transport
- Normal VO_2
  - Age
  - Sex
  - Body size
  - Training
  - Motivation

Cardiac Output vs. O_2 uptake

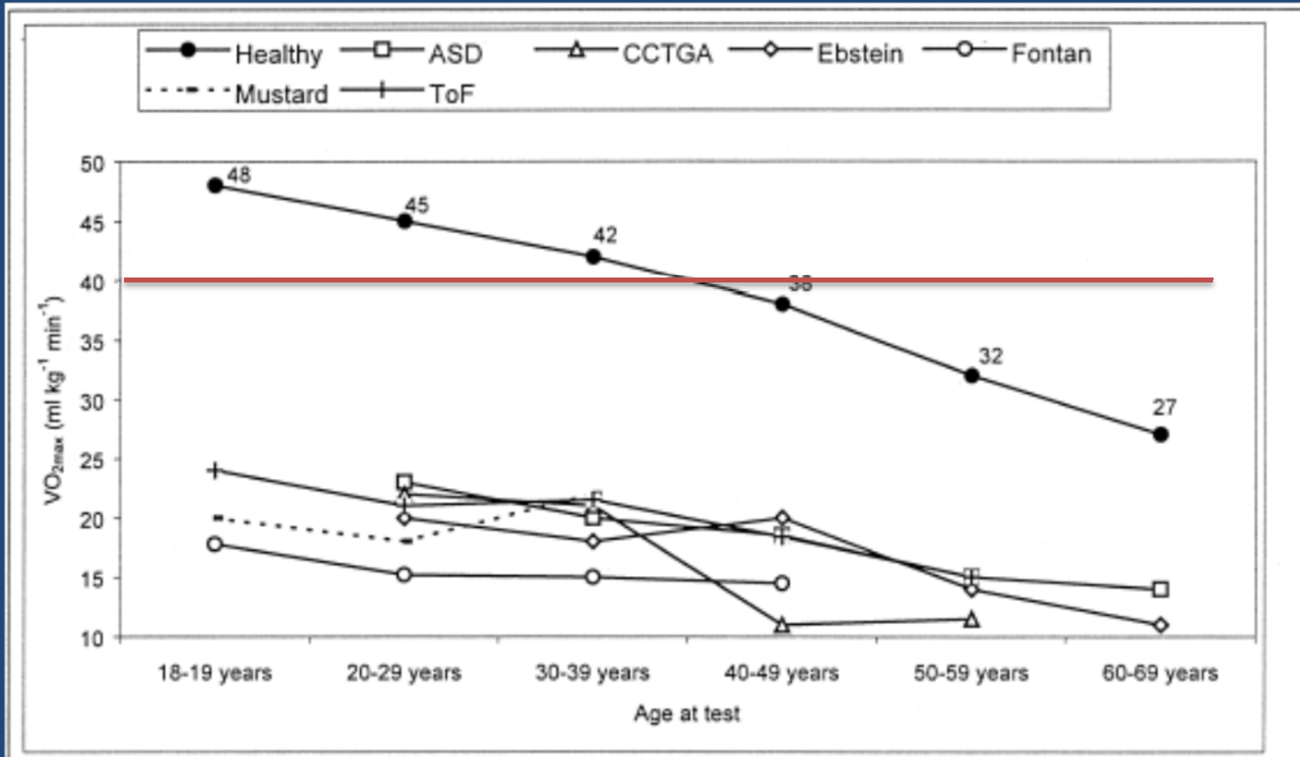


VO_2max –normalized to age and gender

- Boys 42 ml/kg/min
- Girls 38 ml/kg/min



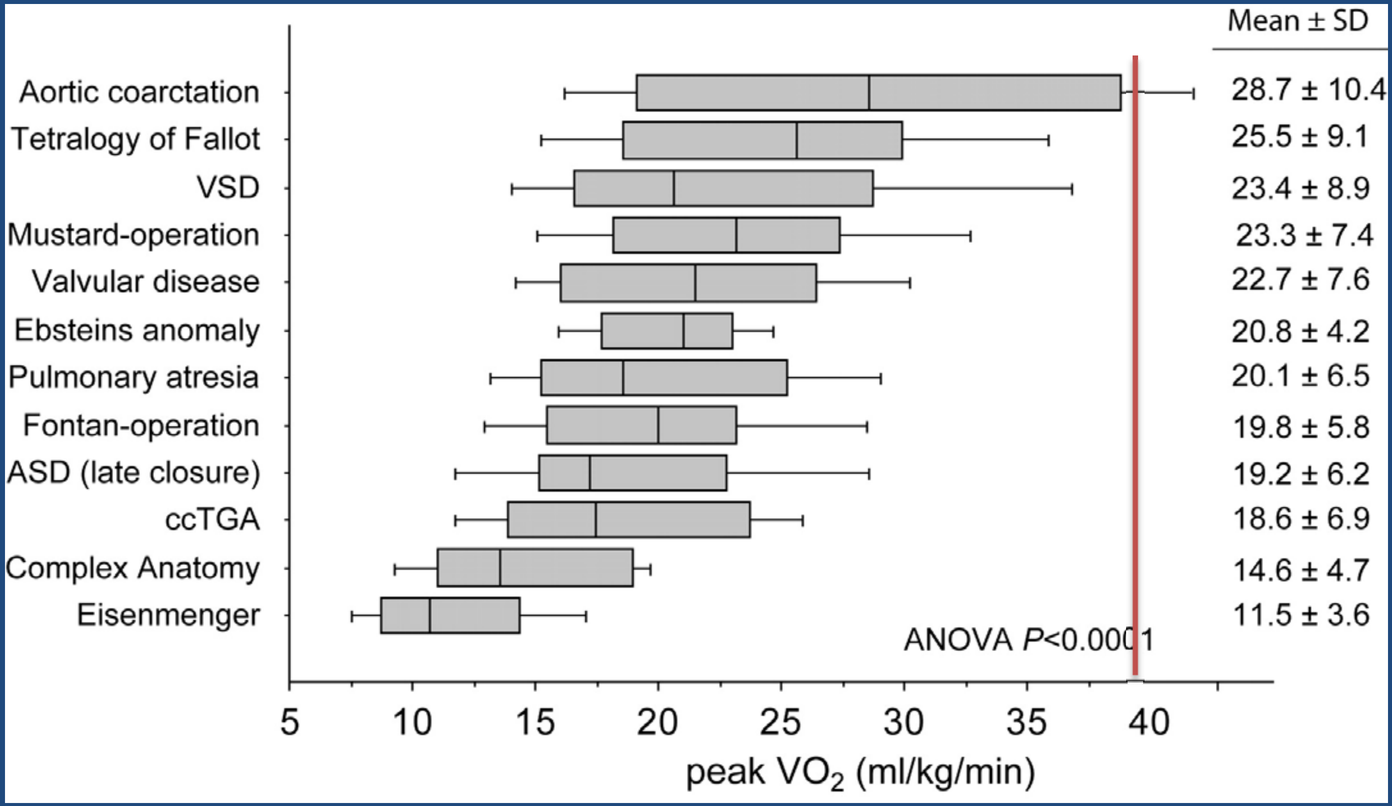
Aerobic Capacity in Adults With Various Congenital Heart Diseases



ASD		18	27	18	16	14
CCTGA		15	8	4	2	
Ebstein		9	11	9	5	3
Fontan	5	30	12	4		
Mustard	16	57	8			
ToF	7	63	50	30	17	

Am J Cardiol 2001;87:310–314

Distribution of peak VO2 in different diagnostic groups



Diller, G.-P. et al. Circulation 2005;112:828-835

Circulation

American Heart Association
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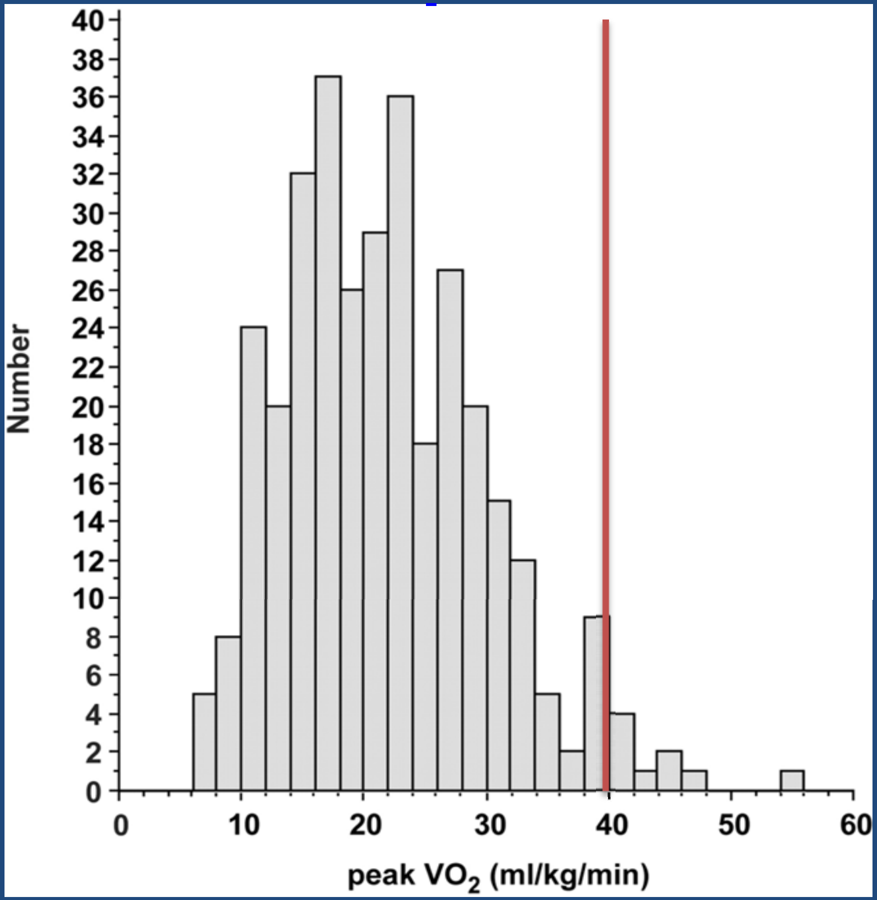
Distribution of peak VO_2 in asymptomatic patients with ACHD (NYHA class I)



Exercise intolerance in ACHD: comparative severity, correlates, and prognostic implication

Diller, G.-P. et al. Circulation 2005;112:828-835

Circulation

Self estimated functioning vs. VO_2peak

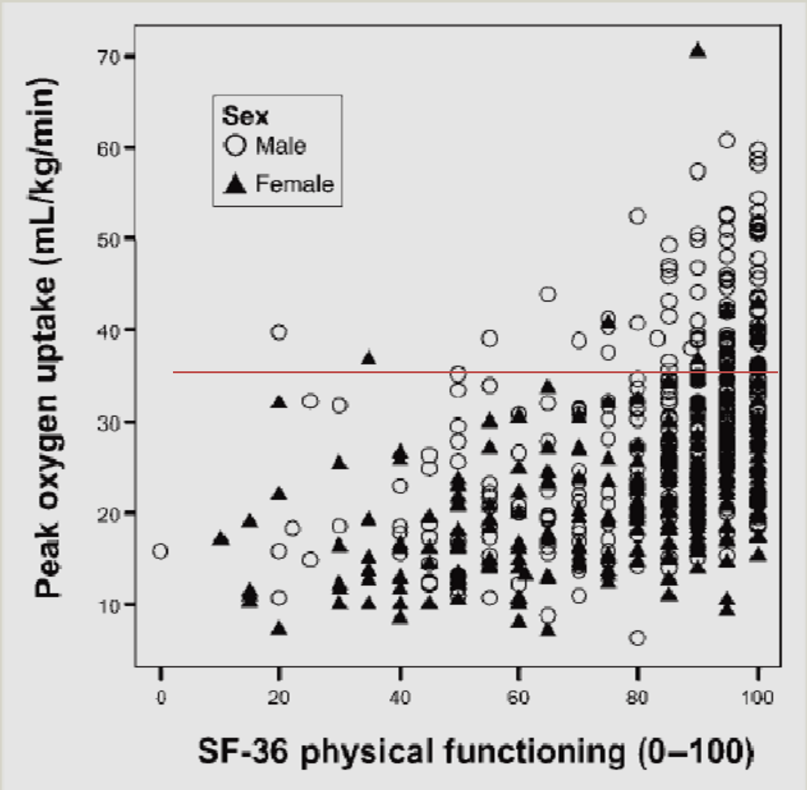


Figure 3 Correlation of objectively measured aerobic physical capacity and self-estimated physical functioning in 564 adolescents and adults with congenital heart disease ($r = 0.435$, $P = 1.72 \times 10^{-27}$); additionally depicting that many patients overestimate their physical capabilities.

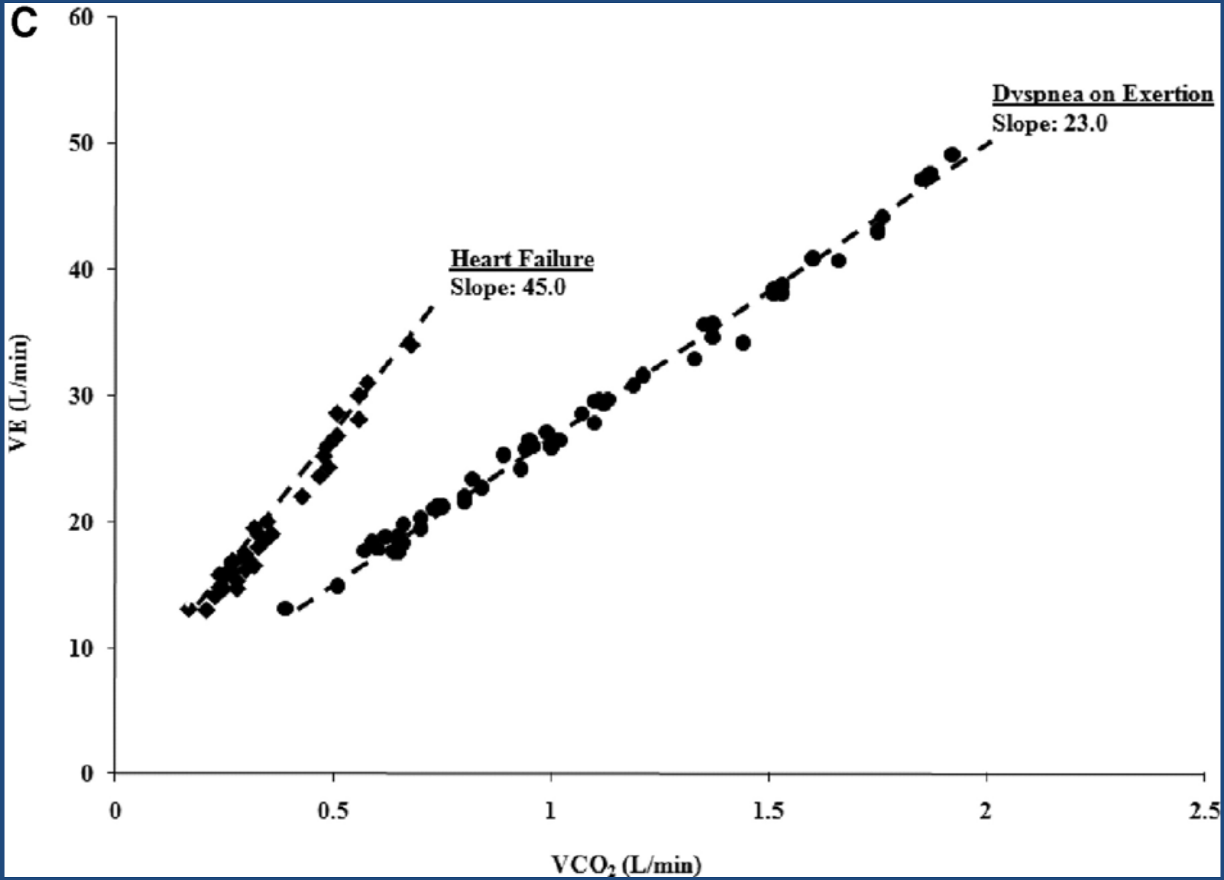
European Heart Journal (2009) **30**, 497–504

Self-estimated physical functioning in CHD patients

V_E/V_{CO2} slope

- Minute ventilation /CO_2 production
- Index of gas exchange efficiency during exercise
  - Liters of air exhaled /1 Liter of CO2 eliminated
- In children < 28

V_E/V_{CO2} in HF vs. respiratory dyspnea



Balady, G. J. et al. Circulation 2010;122:191-225
Clinician's Guide to cardiopulmonary exercise testing in adults: a scientific statement from the American Heart Association

Circulation

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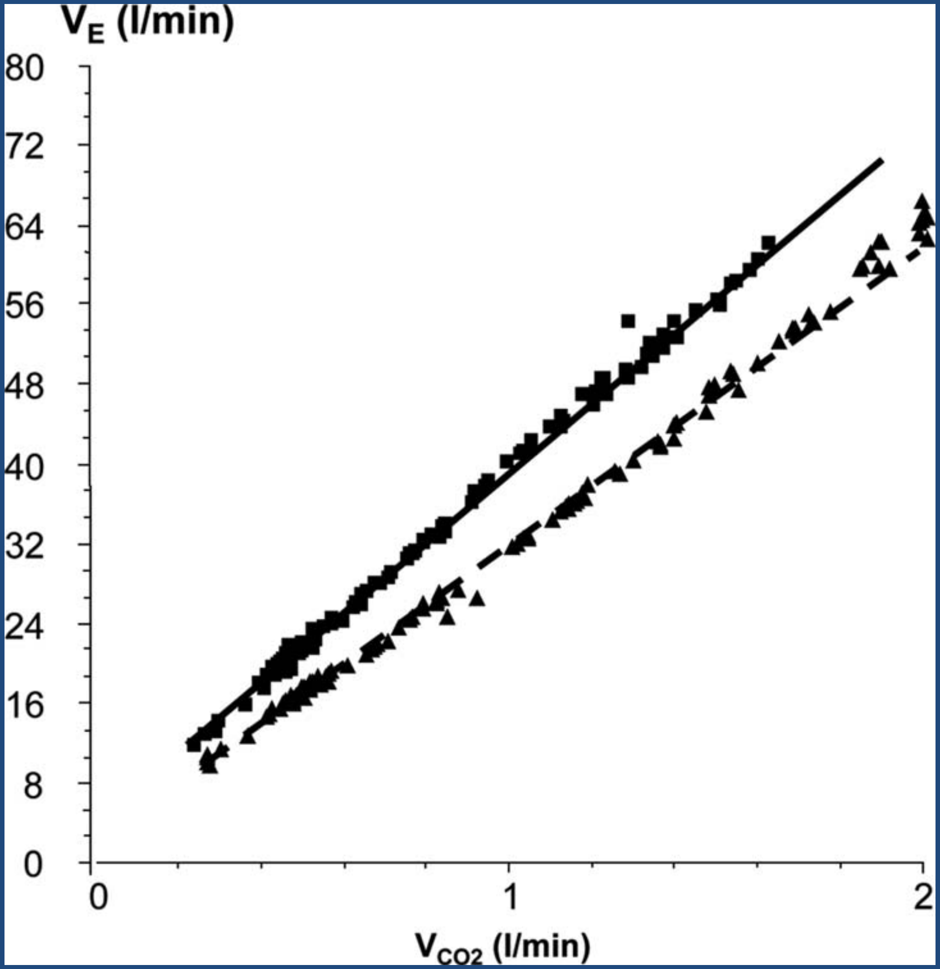
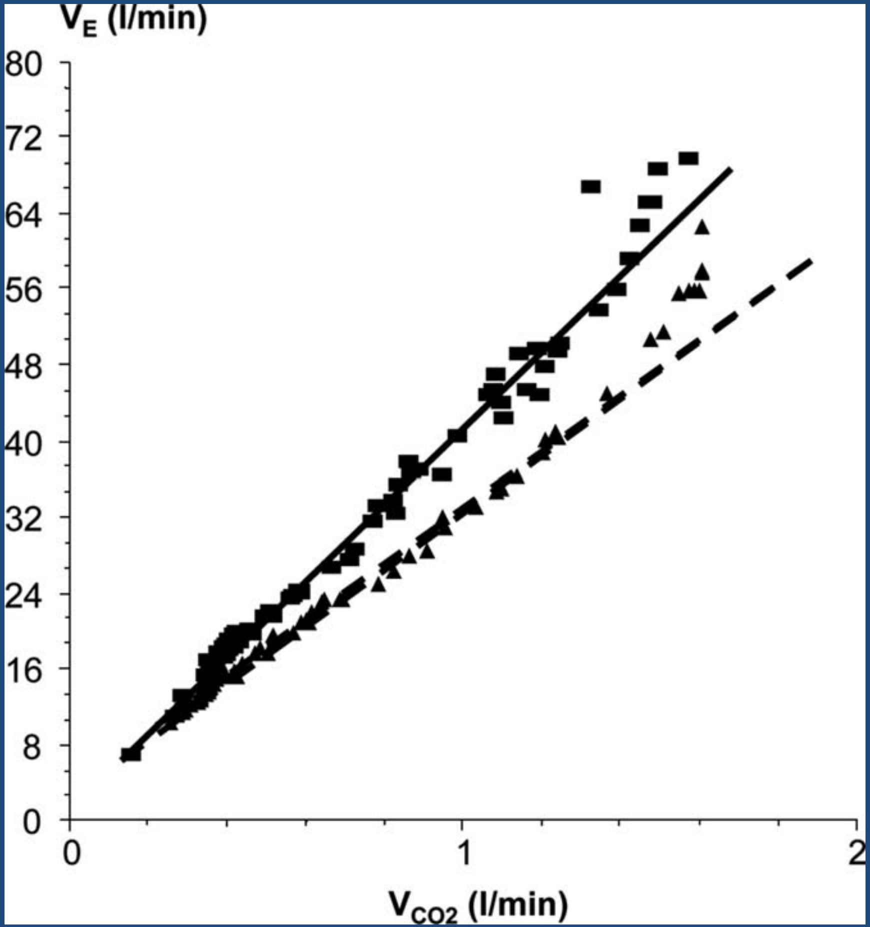
V_E /V_{CO2} in a TOF patient before and after successful LPA balloon angioplasty



Rhodes, J. et al. Circulation 2010;122:1957-1967

Circulation

V_E/V_{CO2} in a patient with fenestrated Fontan before and after fenestration closure



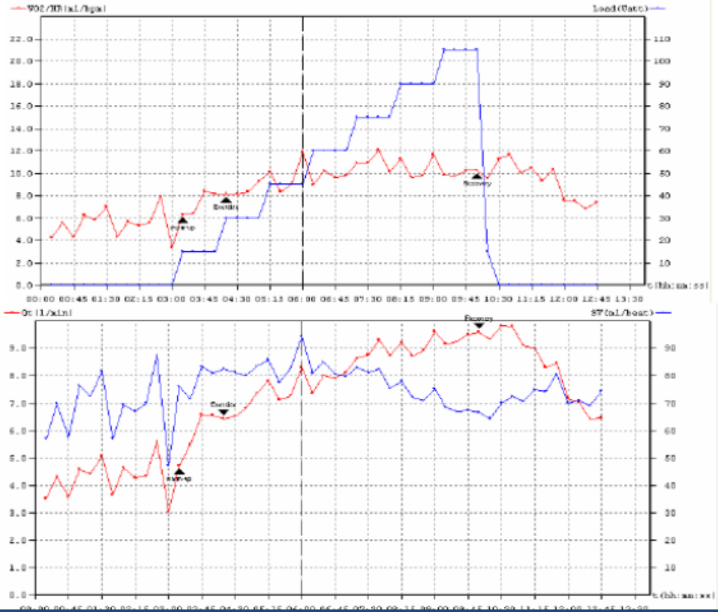
Rhodes, J. et al. Circulation 2010;122:1957-1967

Circulation

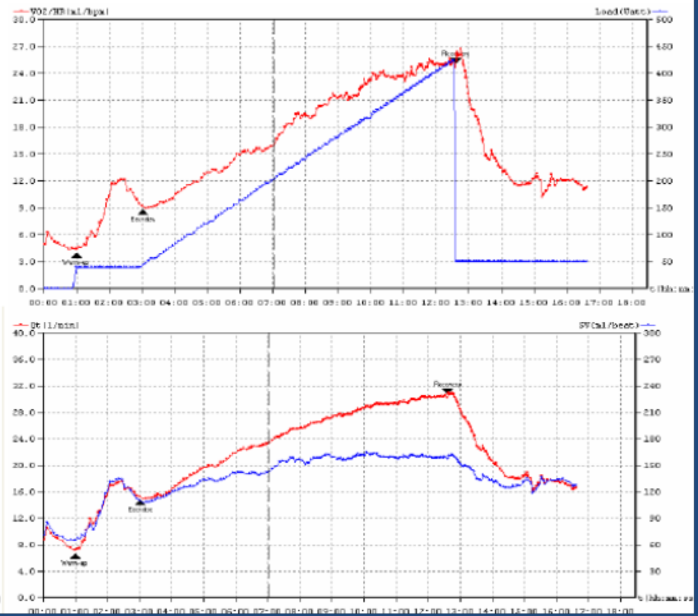
VO_2/HR = Oxygen pulse (O_2P)

- Indirect measurement of SV (stroke volume)
- Increases throughout a ramping exercise
- Falling values during increasing workloads indicate an abnormal SV
- Should be > 80% of predicted at max exercise

Cardiac Patient



Elite Athlete



ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

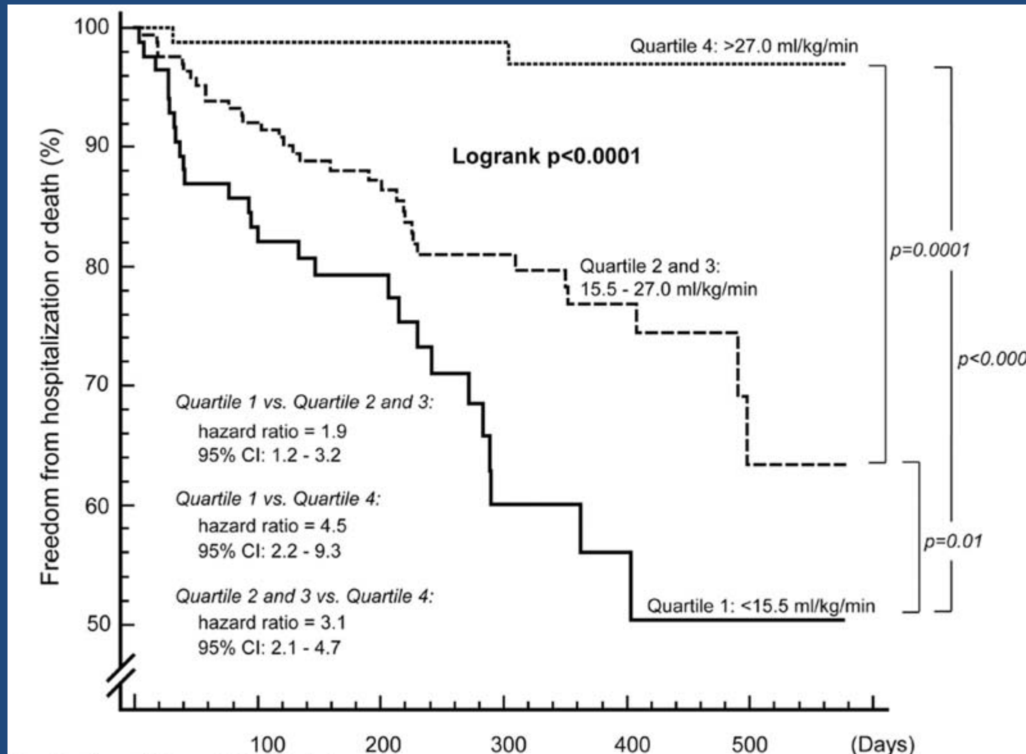
Endorsed by the Association for European Paediatric Cardiology (AEPC)

3.2.4 Cardiopulmonary exercise testing

Formal exercise testing has an important role in the GUCH population, in which quality of life and functional capacity are key measures of the success of intervention. Traditional exercise testing uses protocols that are largely designed for risk stratification of ischaemic heart disease and are often not appropriate in GUCH patients. CPET, including assessment of objective exercise capacity (time, maximum oxygen uptake), ventilation efficiency (VE/VCO_2 slope), chronotropic and blood pressure response, as well as exercise-induced arrhythmia, gives a broader evaluation of function and fitness, and has endpoints which correlate well with morbidity and mortality in GUCH patients.[4] Serial exercise testing should therefore be a part of long-term follow-up protocols and interventional trials. It plays an important role in the timing of interventions and re-interventions.

European Heart Journal (2010) **31**, 2915–2957

Combined end point of hospitalization or death (event-free survival)



Quartile 1: peak VO_2 <15.5 ml/kg/min

At risk	84	68	41	20	10	3
Event-free survival	100%	82.1%	79.3%	60.1%	56.1%	50.5%

Quartile 2 and 3: peak VO_2 15.5 - 27.0 ml/kg/min

At risk	164	147	104	63	33	11
Event-free survival	100%	92.1%	87.2%	80.9%	76.8%	63.4%

Quartile 4: peak VO_2 >27.0 ml/kg/min

At risk	81	79	69	55	32	10
Event-free survival	100%	98.8%	98.8%	97.0%	97.0%	97.0%

Diller, G.-P. et al. Circulation 2005;112:828-835

Circulation

American Heart Association

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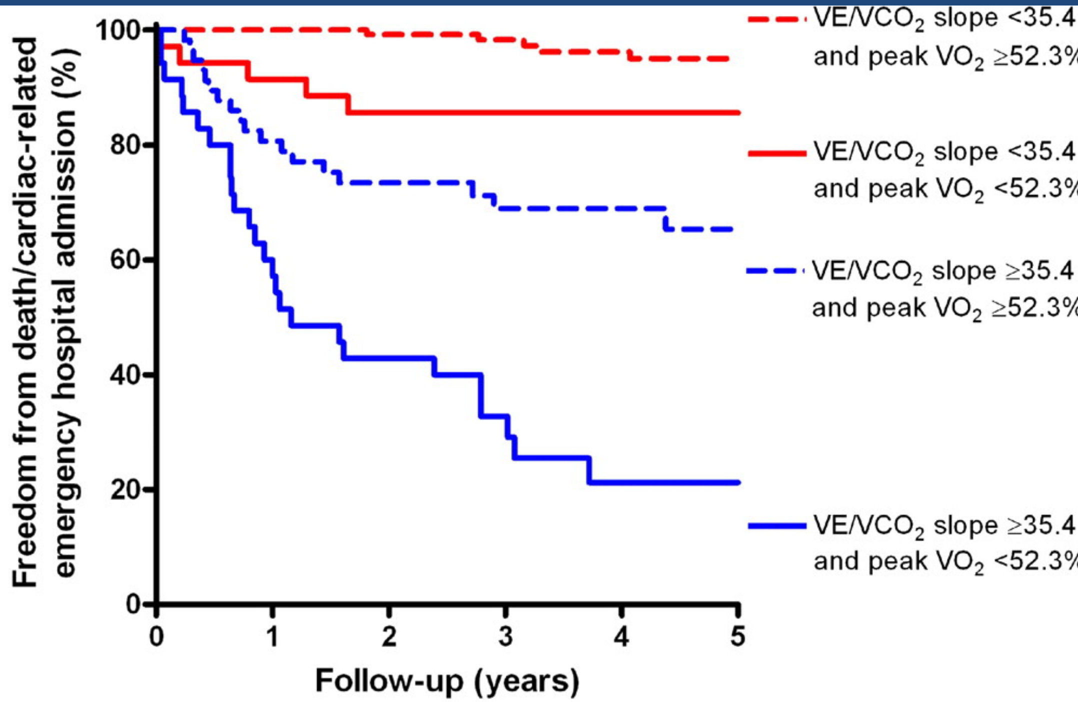
Freedom From Death/Emergency Cardiac-Related Hospital Admission Stratified by Combination of VE/VCO_2 Slope and Peak VO_2



Patients at risk

VE/VCO_2 slope <35.4 and peak VO_2 ≥52.3%:	147	145	126	104	79
VE/VCO_2 slope <35.4 and peak VO_2 <52.3%:	35	33	29	26	21
VE/VCO_2 slope ≥35.4 and peak VO_2 ≥52.3%:	57	46	38	30	20
VE/VCO_2 slope ≥35.4 and peak VO_2 <52.3%:	35	21	16	11	6

Giardini, A. et al. J Am Coll Cardiol 2009;53:1548-1555

Ventilatory efficiency and aerobic capacity predict event-free survival in adults with atrial repair for TGA

Clinical Practice and Education Paper

Physical performance and physical activity in grown-up congenital heart disease

Tony Reybrouck[a,c] and Luc Mertens[b]

Departments of [a]Cardiovascular Rehabilitation, [b]Pediatric Cardiology, University Hospital Gasthuisberg, Leuven, Belgium and [c]Department of Rehabilitation Sciences, University of Leuven (KU Leuven), 3000 Leuven, Belgium.

European Journal of Cardiovascular Prevention and Rehabilitation 2005, 12:498–502

Evaluation of children with congenital heart defects

To get an objective assessment of the functional capacity of children and adolescents with congenital heart defects, formal exercise testing should be performed with continuous measurement of gas exchange. Other methods such as history taking and questionnaires are inaccurate and not sensitive.

Guidelines for the Outpatient Management of Complex Congenital Heart Disease

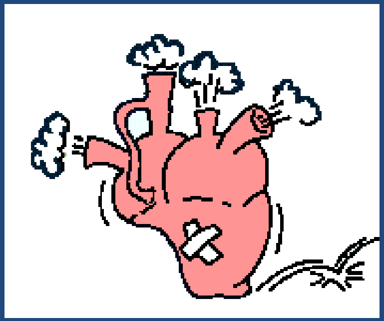
Gil Wernovsky, MD*‡, Jonathan J. Rome, MD*‡, Sarah Tabbutt, MD, PhD*‡, Jack Rychik, MD*‡, Meryl S. Cohen, MD*‡, Stephen M. Paridon, MD*‡, Gary Webb, MD*‡, Kathryn M. Dodds, RN, MSN, CPNP[II], Maureen A. Gallagher, RN, MSN[II], Desiree A. Fleck, RN, MSN, CRNP[II], Thomas L. Spray, MD†§, Victoria L. Vetter, MD*‡, and Marie M. Gleason, MD*‡

we believe that individual assessment of the child's cardiopulmonary function during exercise combined with the routine resting cardiovascular evaluations is essential to tailor appropriate activity level recommendations for these children and adolescents.

Congenit Heart Dis. 2006;1:10–26

Multidisciplinary lab

Multidisciplinary team



Pediatric Cardiologist

Sport Medicine

Physiologist

Objectives

- To determine exercise capacity and cardiac function of patients with repaired CHD compared with normal controls.

- To compare measures of fitness, cardiac and pulmonary functions between CHD patients with complete or incomplete repair, as determined by resting echocardiography.

Methods

- Design: Retrospective analysis of prospectively-collected data

- Population:
- All CHD patients <40 yrs old, with no significant additional co-morbidities,
- After biventricular corrective interventions (surgery or catheterization),
- CHD subgroups divided by the presence of significant anatomical residua on a resting echocardiogram

Methods

- Controls
- otherwise healthy children and adolescents referred to our lab for evaluation of chest pain, palpitations, arrhythmias, conduction disorders etc, and were determined to have normal cardiac function

- CPET on a cycle ergometer in our institution.

Methods- CHD subgroups

Complete repair (n=49)

- TOF – 13
- TGA (all s/p ASO)- 8
- VSD/ DCRV- 8
- COA – 4
- Ross- 3
- PS- 2
- AVC, MS, DORV, PAPVR, IHSS

Methods -CHD subgroups

Incomplete repair (n=24)

- TOF with residual PI -8
- PS/PPS (TOF, VSD+PS, DORV, Rastelli) -5
- TGA s/p Mustard -3
- LV dysfunction (TGA, TOF, ASO)- 4
- PS (PS, Ross)- 2
- AS -1
- AI -1

Methods

- Measures of cardiac function were compared between CHD (n=73) and control (n=76) groups using multiple linear regression techniques and ANCOVA, adjusting for age and sex.

- Similar comparisons were made between CHD patients with complete (n=49) vs. incomplete (n=24) repair
- Values also expressed as % predicted for age and sex

Methods

- CPET data analyzed for this study were:
  - Peak VO_2: aerobic capacity
  - Peak O_2pulse: relates to stroke volume at peak exercise
  - V_E/VCO_2 slope: gas exchange efficiency ≈ cardiac function

Results- peak VO_2 (ml/kg/min)

<u>Age adjusted results</u>:

		p value	
• Complete	32.7 +/-1.2		
		0.59	
• Incomplete	28.8 +/-1.9		0.03
		0.05	
• Control	36.4 +/-1.1		

Results- peak VO_2 (ml/kg/min)

% of predicted value:

		p value	
• Complete	73.8 +/-17.9		
		0.96	
• Incomplete	66.3 +/-20.1		0.02
		0.01	
• Control	92.9 +/-22.0		

Results- O_2 pulse

Age adjusted results:

			p value	
• Complete	9.9	+/-0.47		
			0.059	
• Incomplete	8.0	+/-0.70		0.28
			0.00	
• Control	11.0	+/-0.40		

Results- O_2 pulse

% of predicted value:

		p value	
• Complete	90.5 +/-2.9		
		0.047	
• Incomplete	78.4 +/-4.3		0.26
		0.001	
• Control	94.4 +/-2.4		

Results- V_E/VCO_2 slope

Age adjusted results:

p value

- Complete 27.7 +/-0.6

0.55

- Incomplete 30.2 +/-0.9

0.04

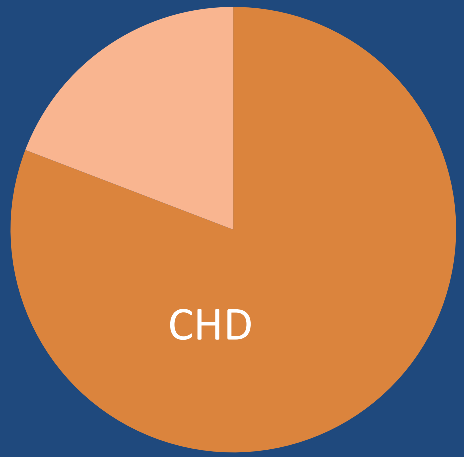
0.001

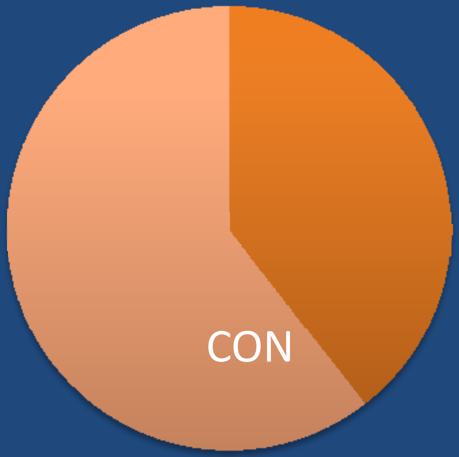
- Controls 25.8 +/-0.5

Results- Peak VO_2

- CHD patients had 25% lower <u>aerobic fitness</u> compared with controls:
  - Peak VO_2 29±8 vs. 38±10 ml/kg/min, p=0.001

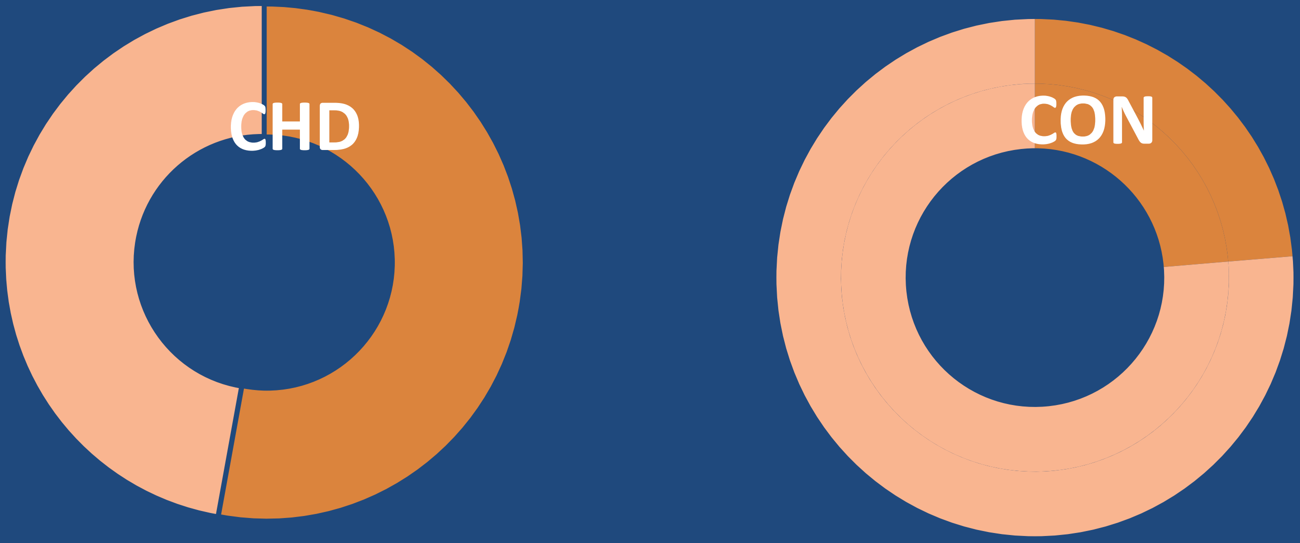
- 19% of CHD patients had normal fitness (peak VO_2 >85% predicted) vs.
- 62% of controls (p<0.001):





Results- Peak O_2pulse

- Peak O_2pulse was abnormal in
- 53% of CHD patients vs.
- 24% of controls ($p<0.001$)



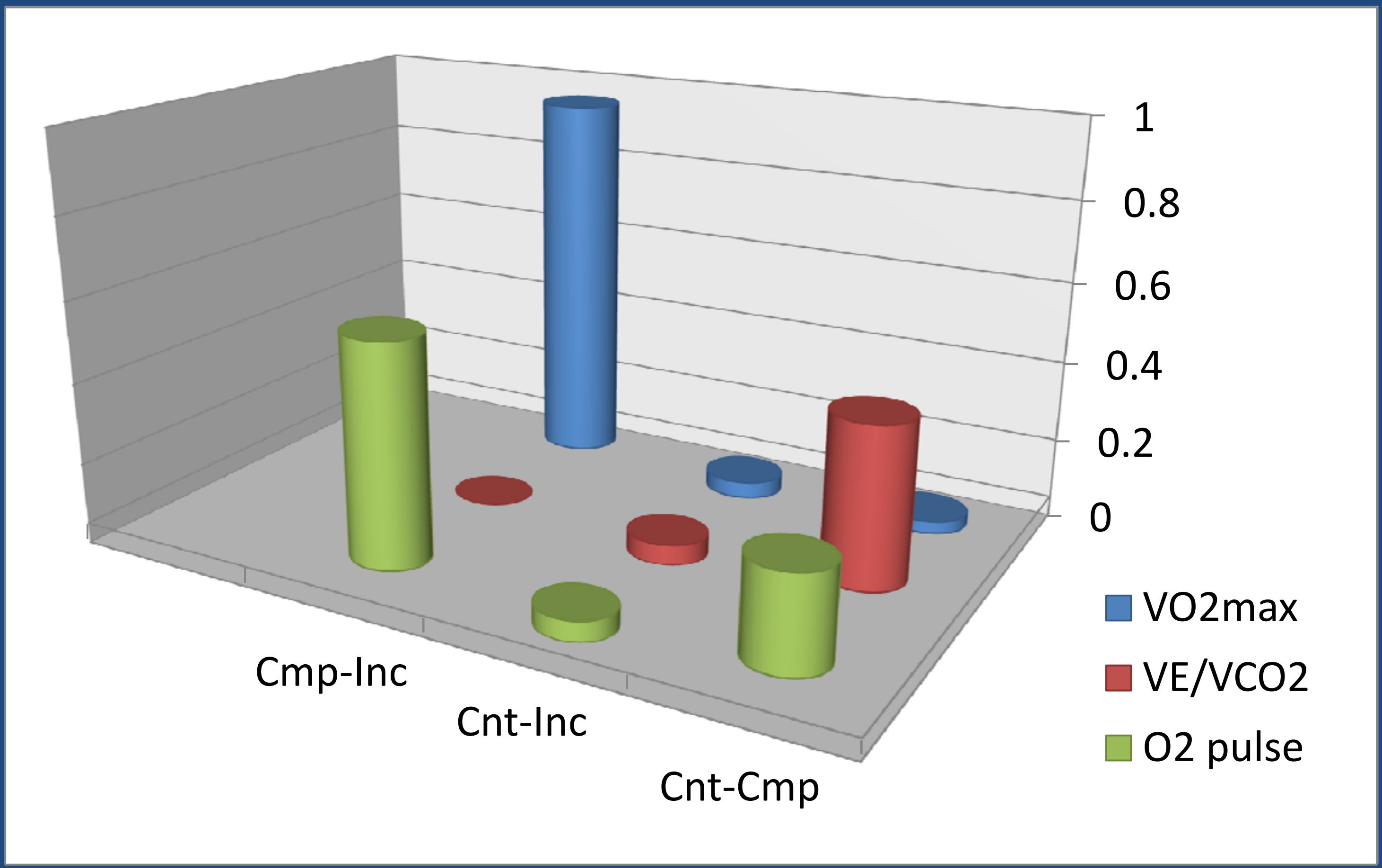
Results- VE/VCO_2 slope

- A significantly higher VE/VCO_2 slope was seen in CHD group compared to controls - 28±5 vs. 26±3 ($p=0.019$)

- 30% of CHD patients had an abnormal slope (>30), compared with 14% of controls ($p=0.021$)

- None of the measured parameters differed between CHD subgroups, except VE/VCO2 slope:
  - An abnormal slope was found in 22% with complete repair, but 46% with incomplete repair.

Study groups vs. variables



Conclusions

- Patients with biventricular CHD repair have a significantly decreased exercise capacity, due to abnormal cardiac function and deconditioning

- The measured parameters were low in all CHD patients, indicating the limited ability of resting echocardiography in assessing cardiac *capacity*.

- Patients with incomplete repair have a significantly higher VE/VCO_2 slope

Conclusions

- Functional cardiopulmonary capacity should be determined, in order to assign our patient a safe level of activity

- Peak VO2 and VE/VCO2 are important prognostic factors

  – CPET should be performed routinely to help plan interventions

  – Patients should learn to view physical activity as an important component of their medical care

Thank you:

- Ronen Reuveny, PhD
- Omer Rosenblum, medical student
- Avshalom Koren, PhD
- Gal Dubnov-Raz, MD

QUESTIONS?