What Can We Learn from an Exercise Test in the Young with Repaired Congenital Heart Disease?

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Disclosures

- Washington University Site Principal Investigator:
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  - Improving Access to Pediatric Cardiac Rehabilitation Through Telemedicine and Remote Patient Monitoring Technology

Exercise Testing Applications

Pulmonary

- Chronic Obstructive Pulmonary Disease (COPD)
- Asthma and Exercise-Induced Bronchoconstriction
- Evaluation of Treatment Efficacy
- Interstitial Lung Disease
- Monitoring Disease Progression and Response to Therapy
- Pulmonary Hypertension
- Assessment of Persistent Dyspnea or Fatigue
- Vocal Cord Dysfunction

Cardiac

Electrophysiological

- Arrhythmias
- Bradycardia
- Bundle Branch Block
- CPVT
- Heart Block
- Long QT
- Orthostatic Intolerance
- Pacemaker Optimization
- PVCs
- Sinus Node Dysfunction
- SVT
- Syncope
- WPW

Congenital Heart Disease (non-repaired/repaired)

Acyanotic

- ASD
- Aortic Valve Regurgitation
- Aortic Valve Stenosis
- Bicuspid Aortic Valve
- Coarctation
- Coronary anomalies/disease
- Mitral Regurgitation
- Mitral Stenosis
- PDA
- Pulmonary Valve Stenosis
- VSD

Cyanotic

- cc-TGA
- D-TGA
- Ebstein Anomaly
- Univentricular Heart
- Tetralogy of Fallot

Cardiomyopathies

- Hypertrophic Cardiomyopathy
- Dilated Cardiomyopathy
- Arrhythmogenic Cardiomyopathy
- LV Noncompaction
- Assessment of Cardiac Reserve
- Family History of Cardiomyopathy

Other Cardiac

- Chest Pain
- Exercise Intolerance
- Preparticipation Screening
- Hypertension

Acquired Heart Disease

- Kawasaki Disease
- MIS-C
- Myocarditis
- Pericarditis

Heart Transplant

- Pre/Post- Heart Transplant
- Cardiac Rehabilitation Planning

Neurological

- Exercise-Induced Seizures
- Cerebral Palsy
- Muscular Dystrophies

Other

- Chemotherapy
- Post-COVID-19 Syndrome
- Exercise-Induced Hypoglycemia
- Obesity
- Pectus
- Peripheral Arterial Disease (PAD)
- Claudication
- Metabolic Disorders

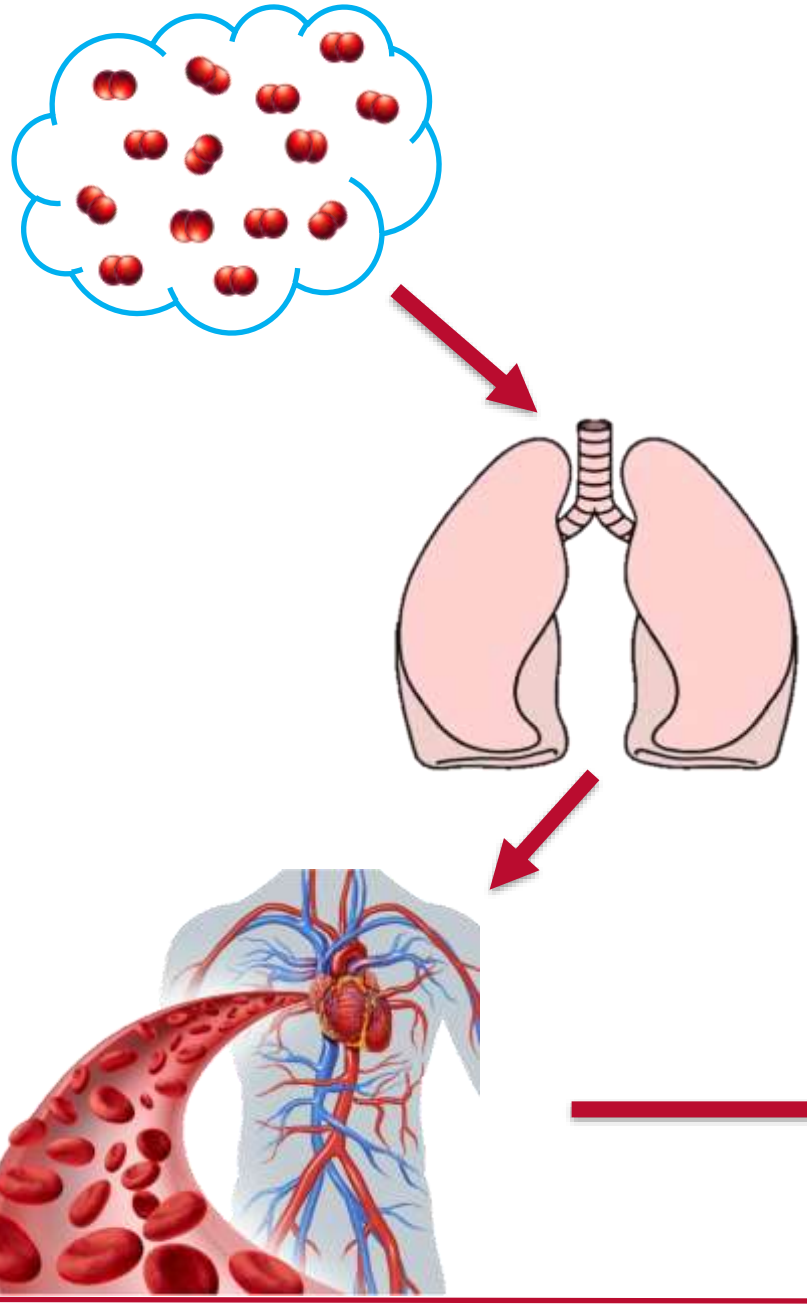
Exercise Test

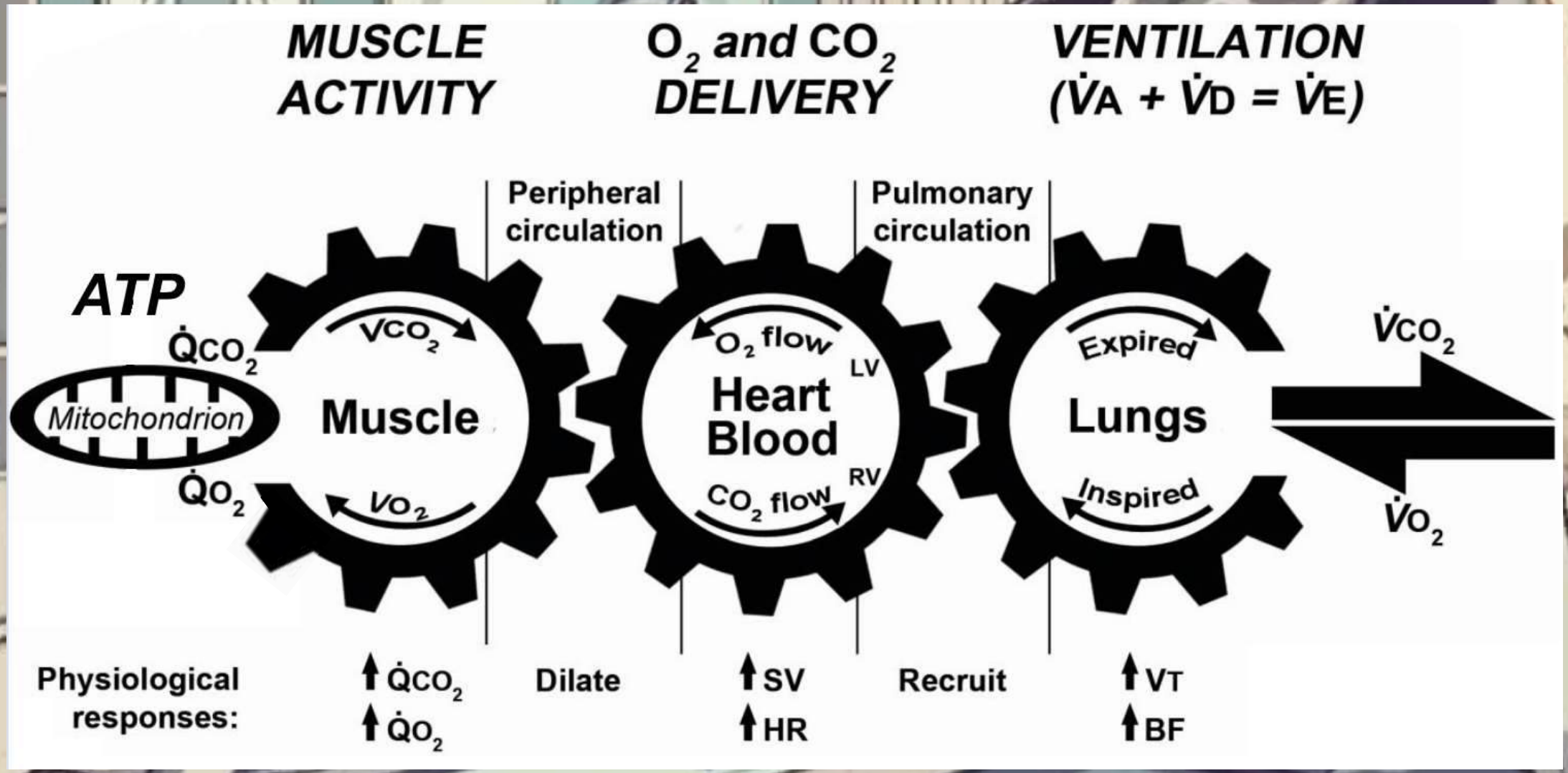
Graded ECG

- HR response
- BP response
- Baseline ECG
- Arrhythmias
- QT intervals
- ST segment changes

Cardiopulmonary Exercise Test (CPET)

- Graded ECG data
- Breath-by-breath data
- Peak oxygen consumption
- Ventilatory anaerobic threshold
- VO_2 and its relationship to: HR, WR, VCO_2
- O_2 Pulse
- Ventilation and its relationship to breathing capacity
- Lung mechanics
- Relationship of VE and CO_2 elimination



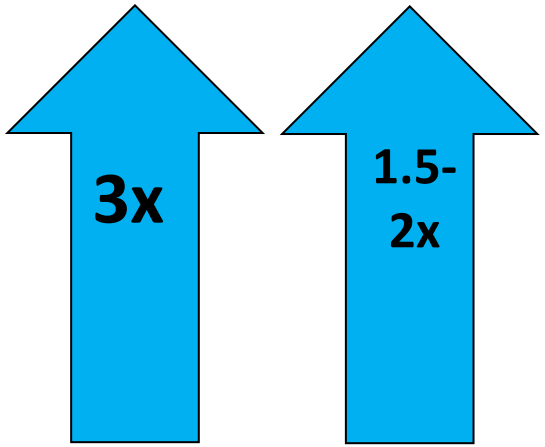

MUSCLE ACTIVITY
O2 and CO2 DELIVERY
VENTILATION (V̇A + V̇D = V̇E)
Peripheral circulation
Pulmonary circulation
ATP
Q̇CO2
Mitochondrion
Q̇O2
VCO2
Muscle
VO2
O2 flow
LV
Heart Blood
RV
CO2 flow
Expired
Lungs
Inspired
V̇CO2
V̇O2
Physiological responses:
↑ Q̇CO2
↑ Q̇O2
Dilate
↑ SV
↑ HR
Recruit
↑ VT
↑ BF

Oxygen Consumption

- VO_2 = [C.O.] x [oxygen extraction]

  = [HR x SV] x [C_aO_2 - C_vO_2]

  = [HR x SV] x [1.36 (Hgb) (S_aO_2 - S_vO_2)]



Sinus node Dysfunction

- ASD (sinus venosus ASD)
- TAPVC
- Fontan Procedure
- Atrial switch (cc-TGA, Mustard, Senning)
- Tetralogy of Fallot
- Ebstein's Anomaly
- Heterotaxy

AV Block

- VSD
- AV canal
- Tetralogy of Fallot
- D-TGA
- DORV
- cc-TGA
- Subaortic stenosis
- Truncus Arteriosus

Pacemaker Optimization

- Rate Response
- AV delay
- Upper Tracking Rate
- Mode Switching
- CRT

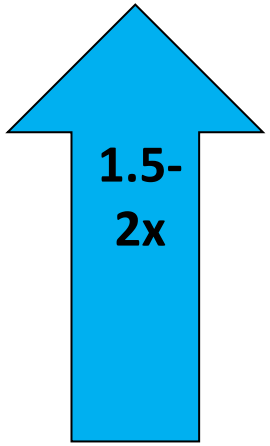
3x

$$=[\mathbf{HR} \times SV] \times [1.36\,(Hgb)\,(S_aO_2 - S_vO_2)]$$

Oxygen Pulse (O_2P)

- $O_2P = VO_2/HR$
  - = Cardiac Output/HR x O_2 extraction
  - = Stroke Volume x O_2 extraction

- ↓ systolic/diastolic function
- Myocardial ischemia
- Cardiomyopathy
- Severe outflow tract obstruction
- Severe valvar regurgitation
- Fontan circulation



$$=[HR \times \mathbf{SV}] \times [1.36\ (Hgb)\ (S_aO_2 - S_vO_2)]$$

Oxygen Extraction

- During exercise mixed venous oxygen saturation typically falls to less than 30%
  - Vasodilation -> larger surface area
  - Accumulation of LA facility release of O2

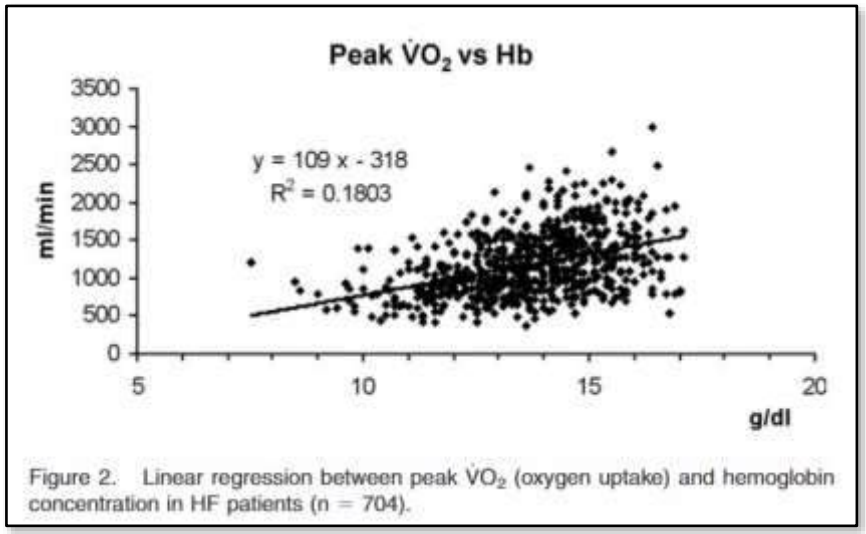
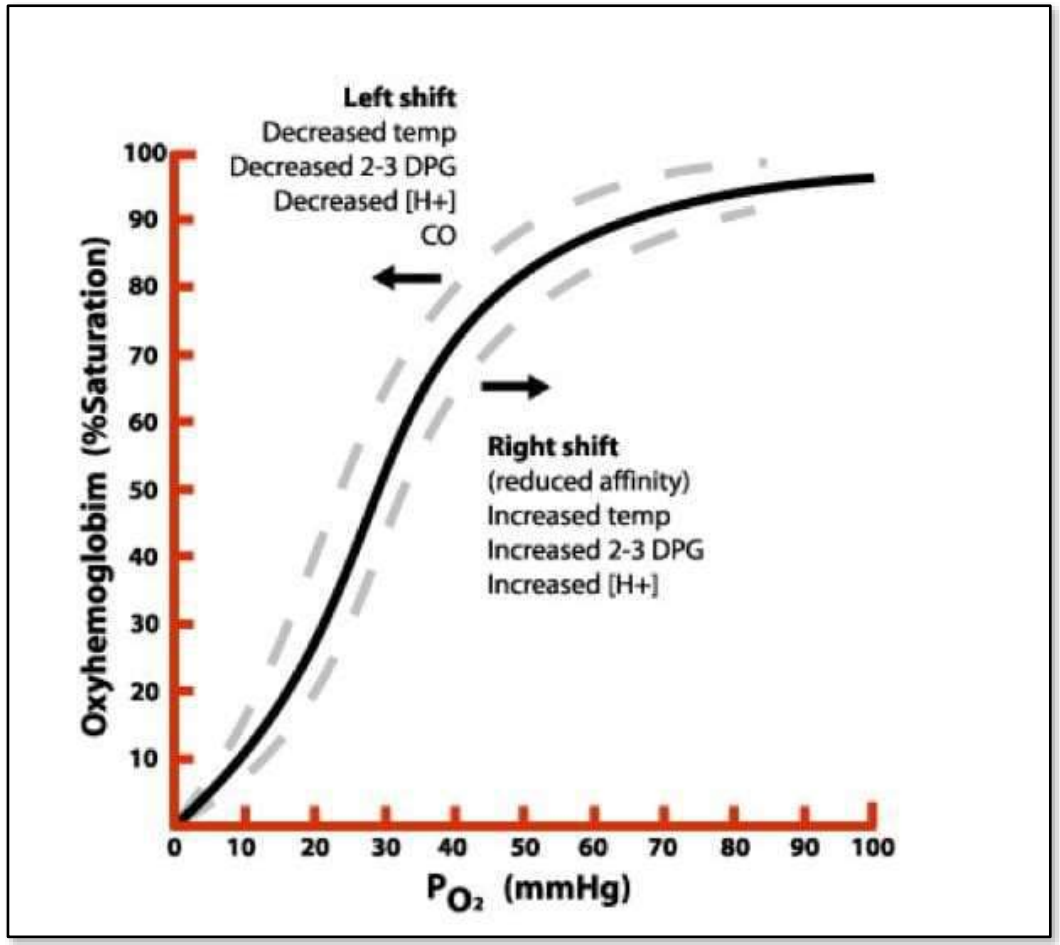
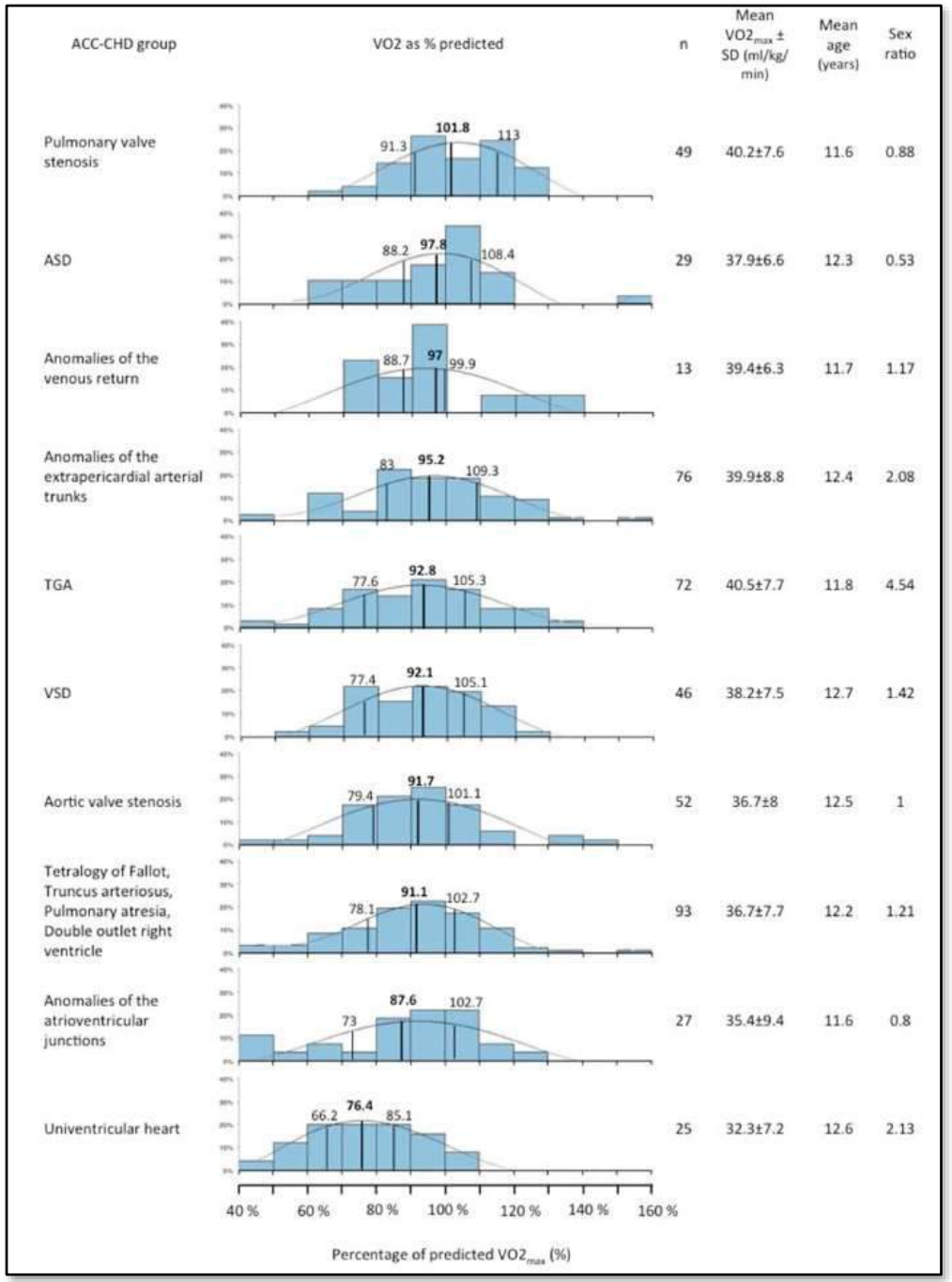


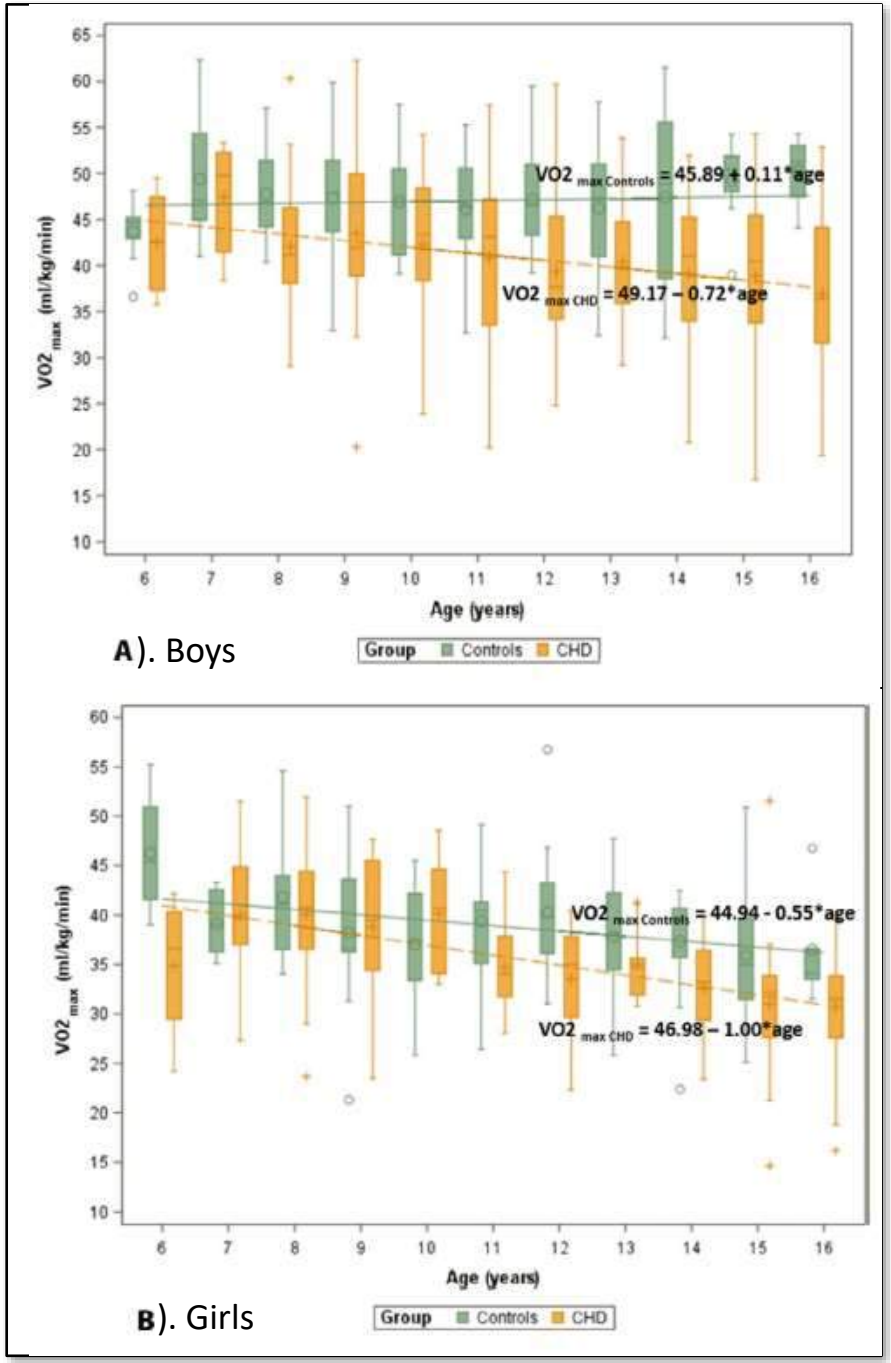
Figure 2. Linear regression between peak $\dot{V}O_2$ (oxygen uptake) and hemoglobin concentration in HF patients (n = 704).

Agostoni, Piergiuseppe et.al. "Relationship of resting hemoglobin concentration to peak oxygen uptake in heart failure patients." *American journal of hematology*



$$=[HR \times SV] \times [1.36\,(\mathbf{Hgb})\,(S_aO_2 - S_vO_2)]$$

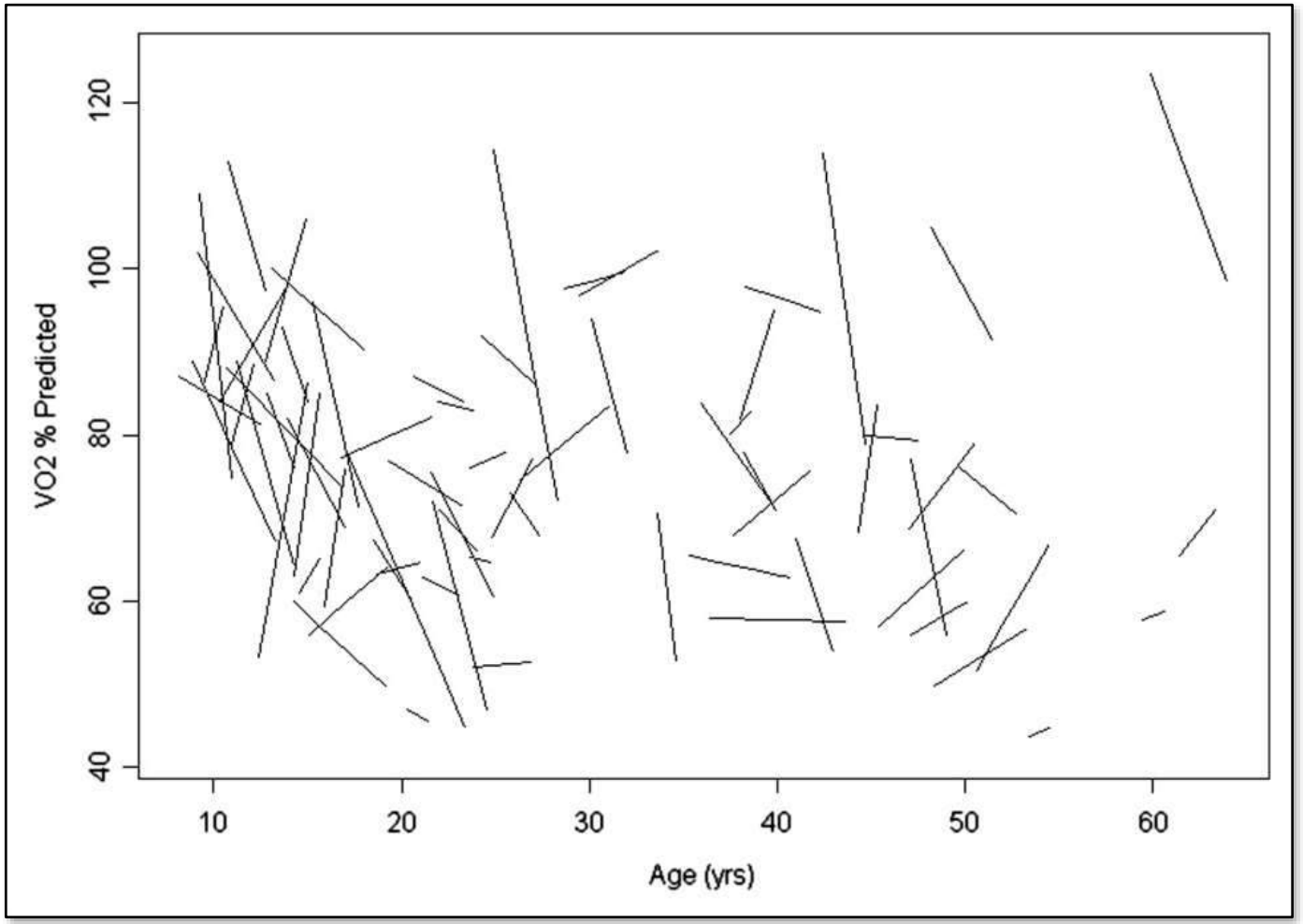




Amedro et. al. “Cardiopulmonary fitness in children with congenital heart diseases versus healthy children.” Congenital Heart Disease

Peak VO2 – Tetralogy of Fallot

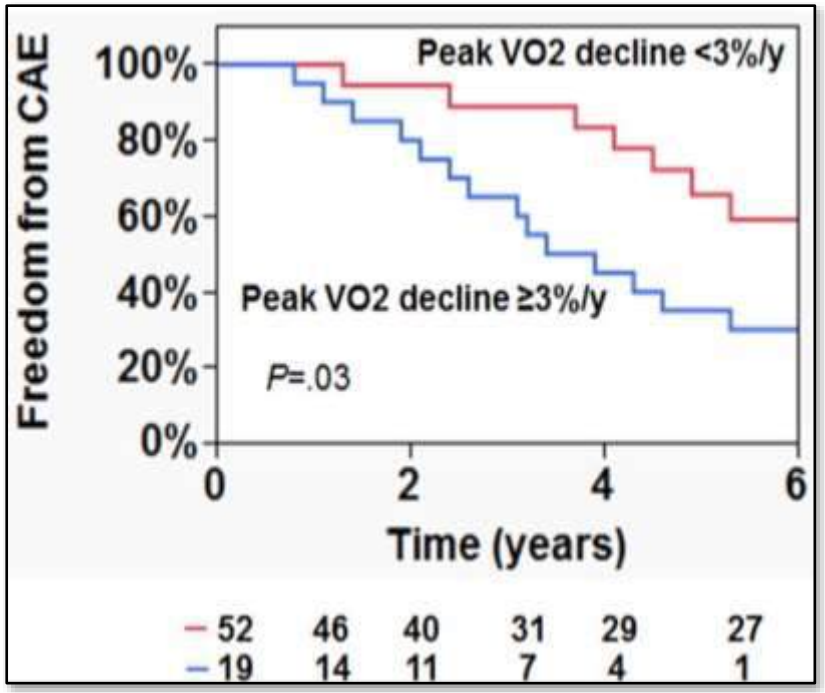
VO2 Observed decrease of 1.4% points/yr



Kipps, Alaina K et.al. "Longitudinal exercise capacity of patients with repaired tetralogy of fallot." *The American journal of cardiology*

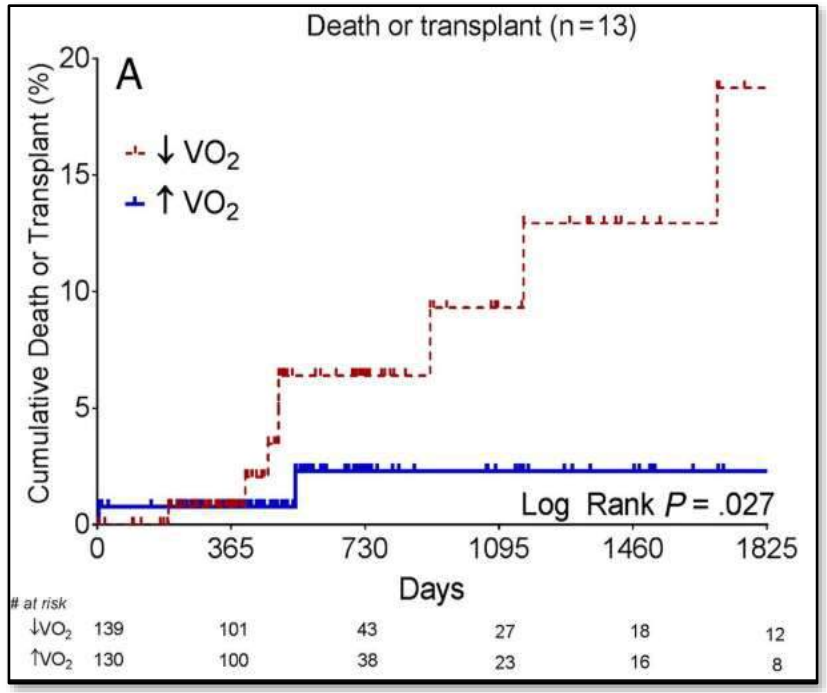
Peak VO2 – Fontan

Adverse cardiovascular events



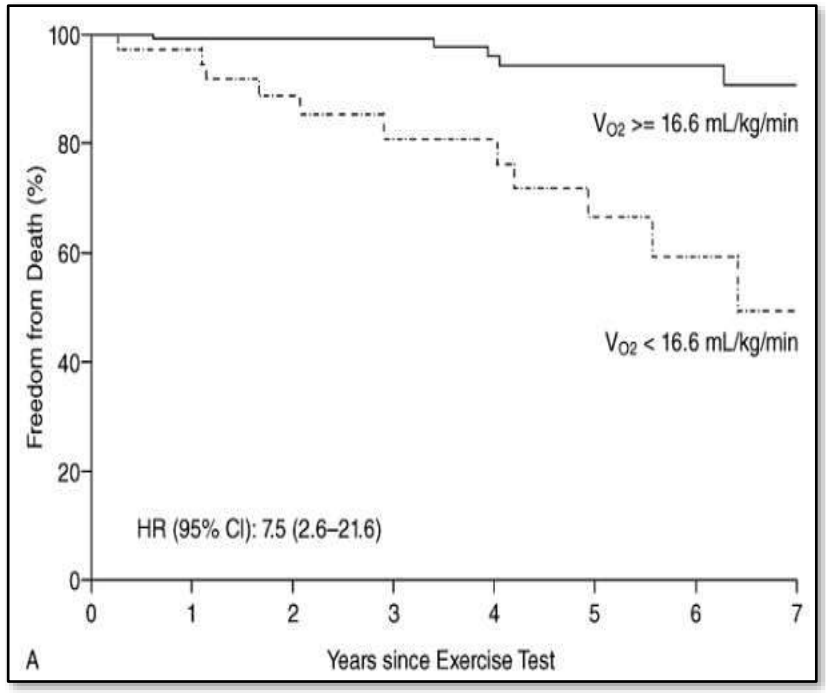
Egbe, Alexander C. et. al. "Cardiopulmonary exercise test in adults with prior Fontan operation: the prognostic value of serial testing." International journal of Cardiology

Death/Cardiac Transplant



Cunningham, Jonathan W. et.al. "Decline in peak oxygen consumption over time predicts death or transplantation in adults with a Fontan circulation." American heart journal

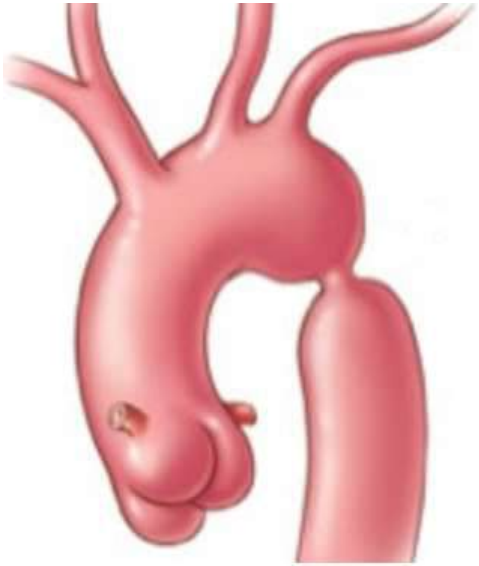
Death

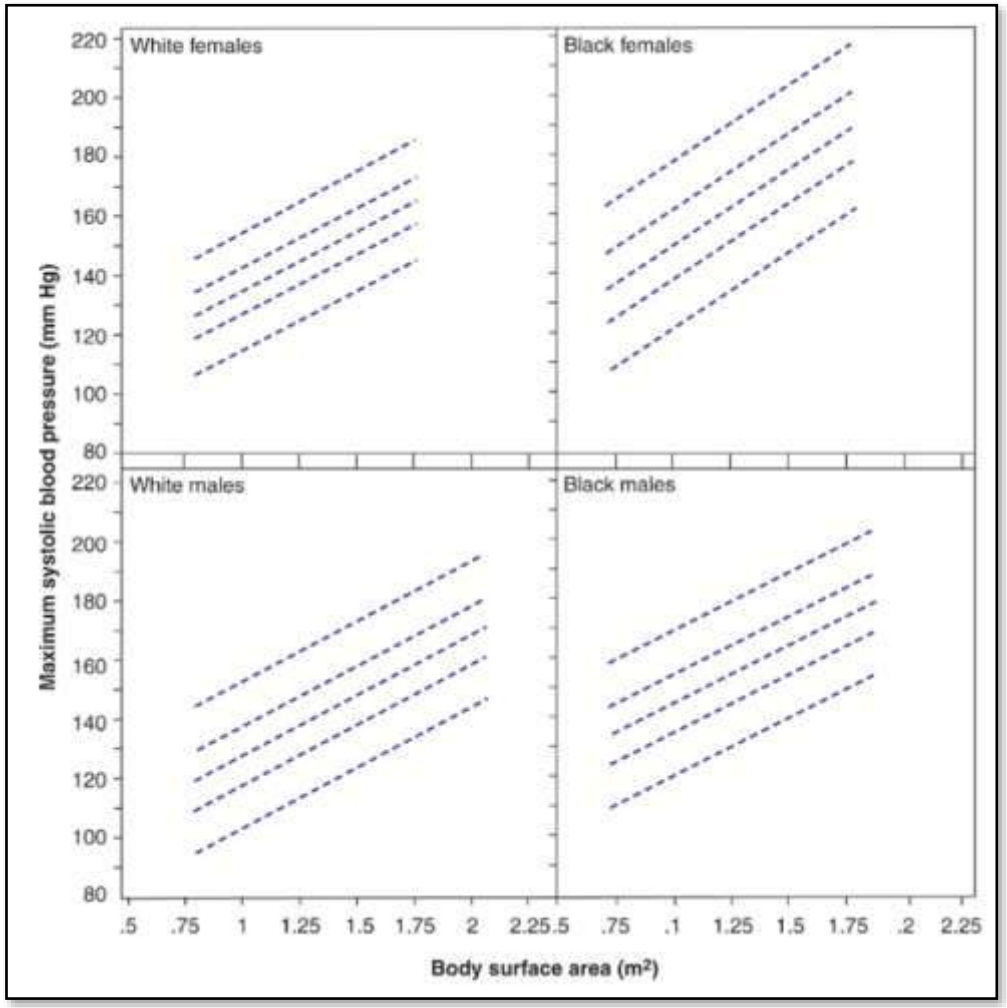


Fernandes, Susan M. et.al. "Exercise testing identifies patients at increased risk for morbidity and mortality following Fontan surgery." Congenital heart disease

Blood Pressure Response to Dynamic Exercise

- SBP increases, DBP small increase (maybe 10 mmHg)
- Systolic blood pressure should exceed resting values by at least 20% or 20 mmHg
- Peak exercise: Rare for systolic blood pressure to exceed 200 in males in 180 in females



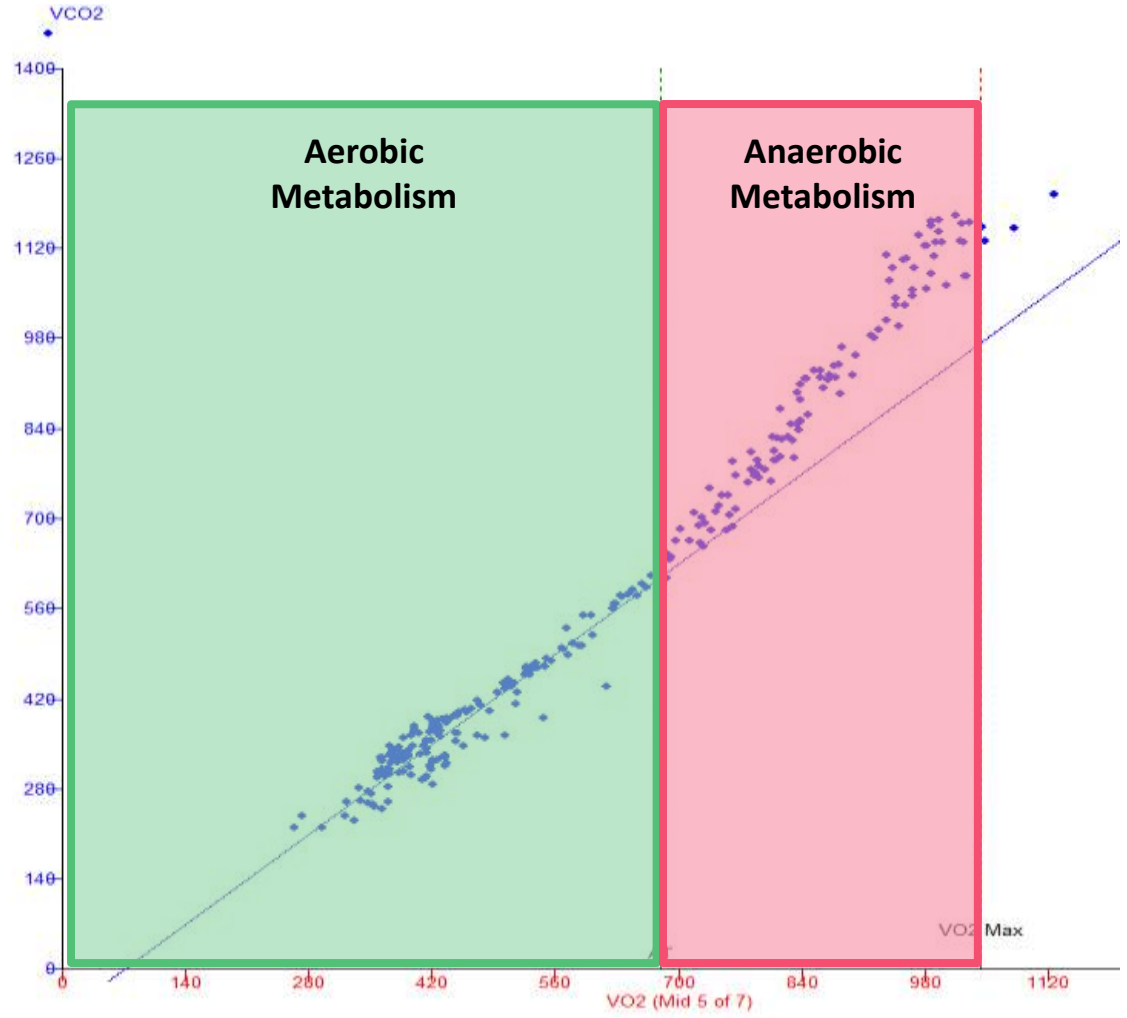


Rhodes, Jonathan et. al. *Exercise physiology for the pediatric and congenital cardiologist*.

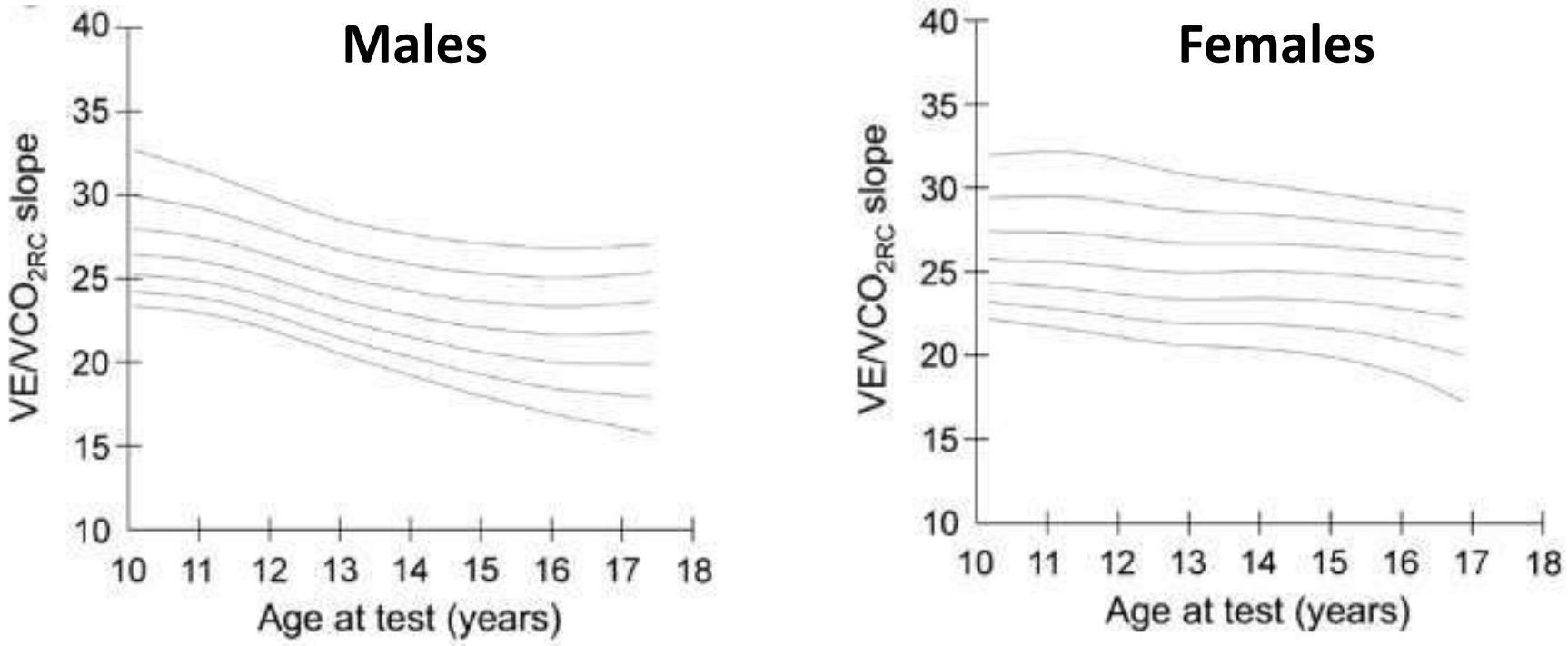
Ventilatory Anaerobic Threshold (VAT)

An excellent index of the cardiovascular system's capacity to support hemodynamic demands of exercise

Any cardiovascular condition that impairs oxygen delivery will lower anaerobic threshold



V_E/VCO_2 Slope



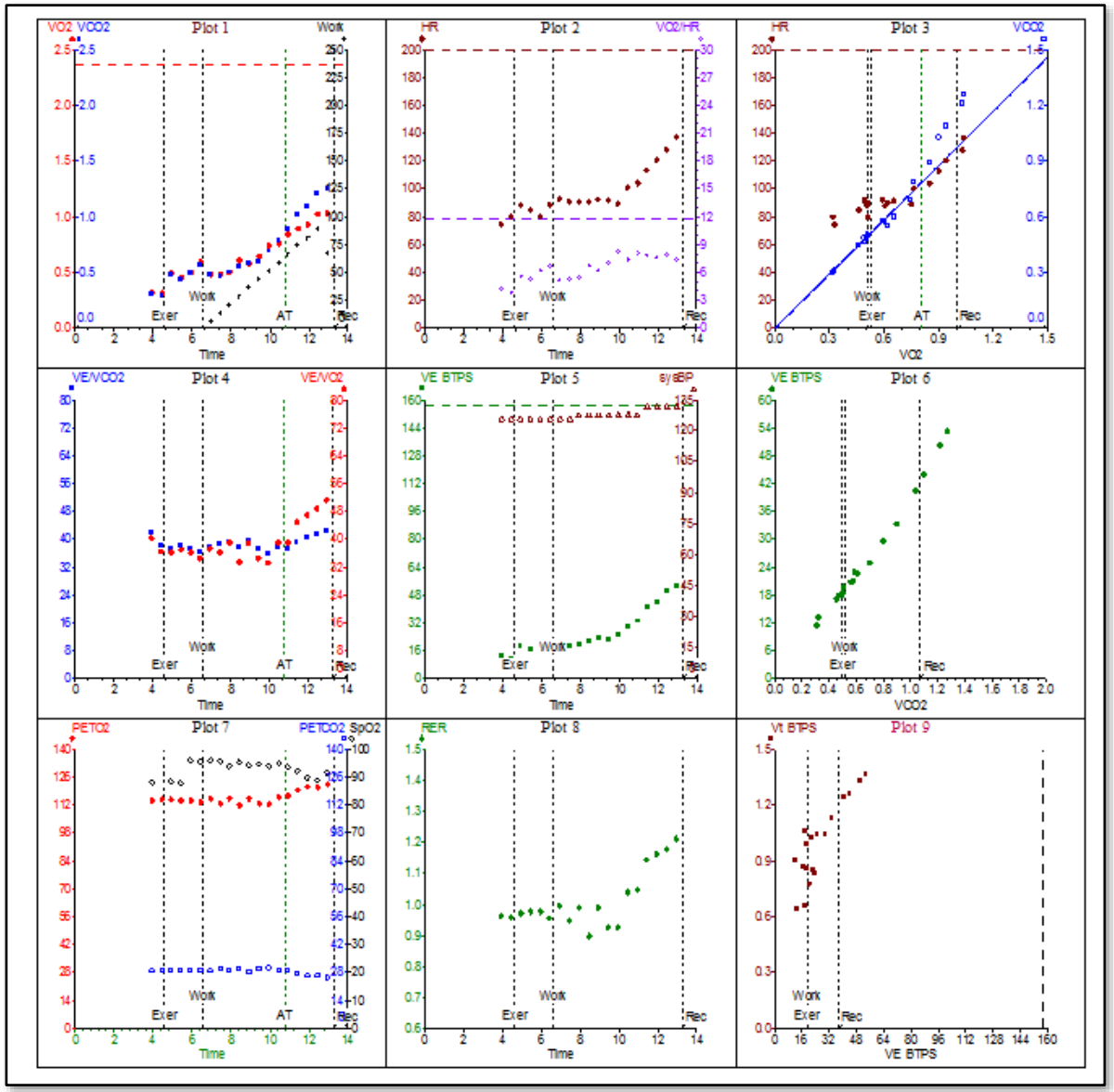
- Impaired transport of gases across the alveolar-capillary membrane
- Fontan Circulation

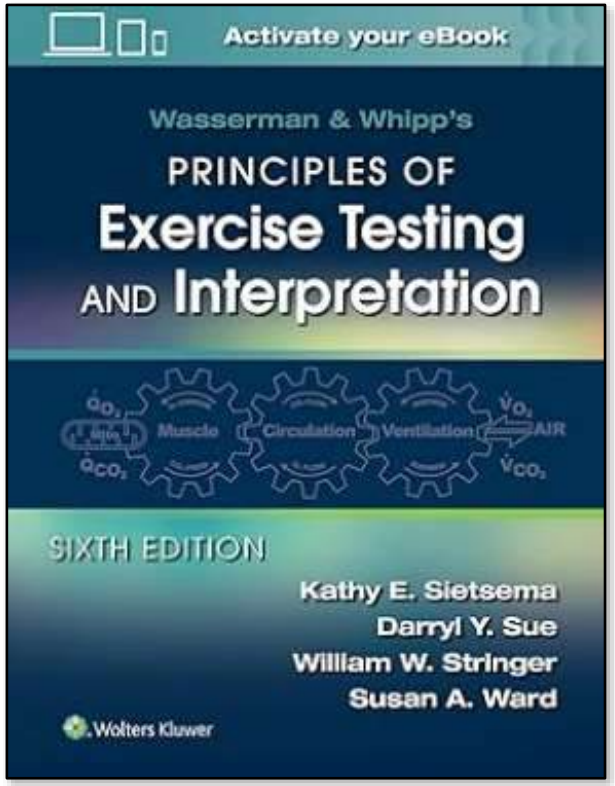
- Pulmonary Stenosis
- Elevated Wedge Pressures

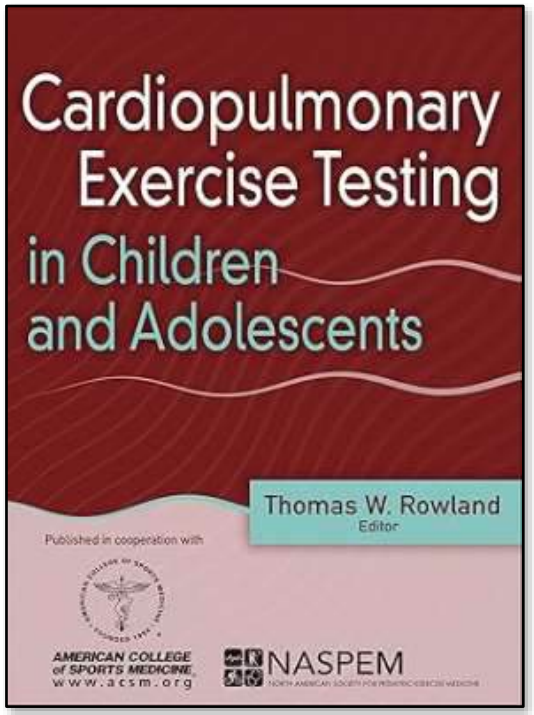
- Right to left shunting
- Pulmonary Hypertension (ASD, PFO)

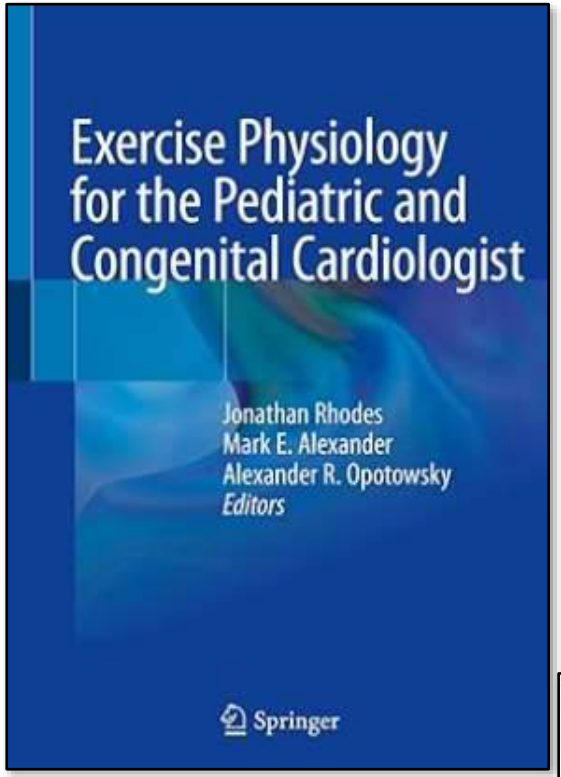
CPET Report

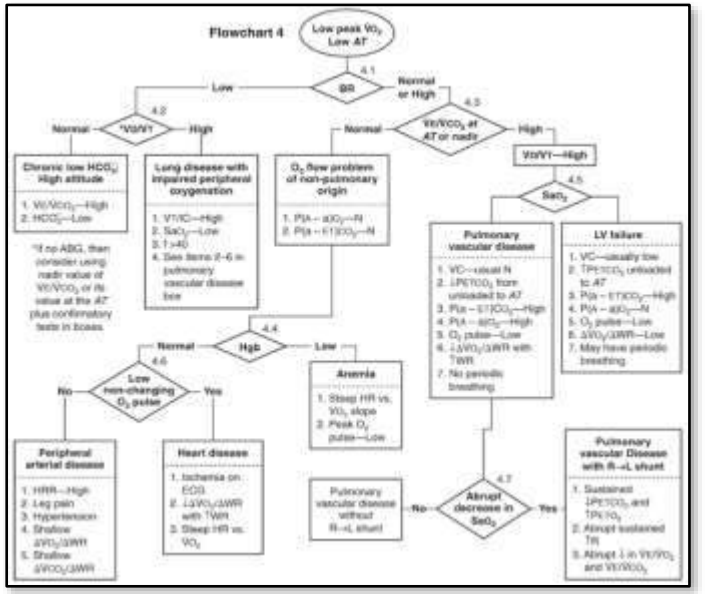
	Rest	AT	V02 Max	Pred	AT / VO2 Max (%)	VO2 Max/Pred (%)
Time (min)	4:21	10:37	11:46			
Ex Time (min)		6:11	7:20			
---- WORK ----						
Work (Watts)	0	63	80	187	79	43
Speed (RPM)	3	53	56		95	
---- VENTILATION ----						
Vt BTPS (L)	0.70	1.02	1.42		72	
RR (br/min)	20	29	30		95	
VE BTPS (L/min)	13.9	29.3	43.1	157.0	68	27
BR (%)	91.1	81.3	72.5		112	
---- O2 CONSUMPTION ----						
VO2 (mL/kg/min)	6.7	14.1	20.7	43.3	68	48
VO2 (mL/min)	365	767	1131	2359	68	48
VCO2 (mL/min)	343	788	1173	2854	67	41
RER	0.94	1.03	1.04		99	
METS	1.9	4.0	5.9	12.4	68	48
---- CARDIAC ----						
HR (BPM)	84	103	113	200	91	56
VO2/HR (mL/beat)	4	7	10	12	74	85
---- V/Q ----						
VE/VCO2	41	37	37	39	101	95
VE/VO2	38	38	38	47	100	82
PETCO2 (mmHg)	28	29	29		102	
PETO2 (mmHg)	115	116	117		99	
----------						
sysBP (mmHg)	126	128	132		97	
diaBP (mmHg)	72	72	70		103	
RatePrsPd SBP*HR/100	106	132	149	380	88	39
Borg PE						
SpO2 (%)		95	92		103	



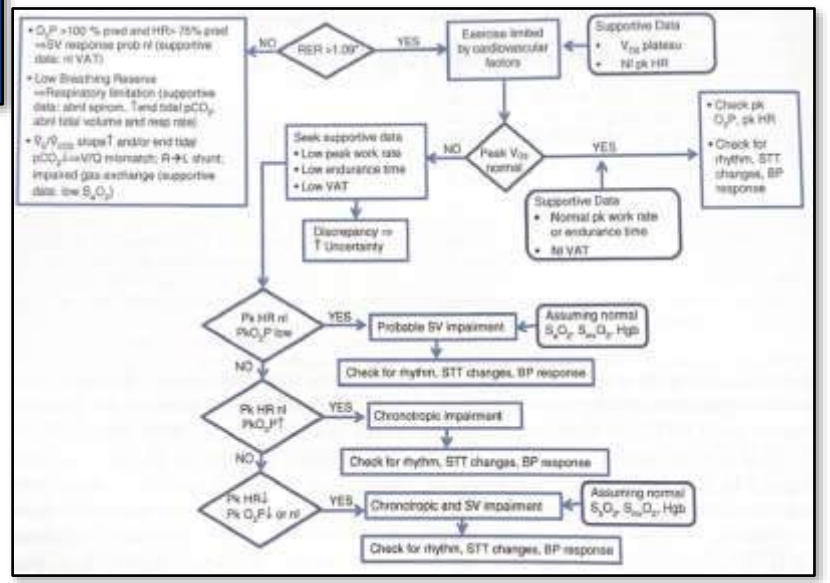








Sietsema, Kathy E., et. al *Wasserman & Whipp's: principles of exercise testing and interpretation.*



Rhodes, Jonathan et. al. *Exercise physiology for the pediatric and congenital cardiologist*.

Prognosis/ Surveillance

Symptom Provocation

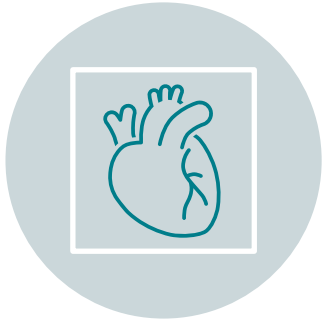
Risk Stratification

Symptom Provocation


DIZZINESS


PALPITATIONS


SYNCOPE

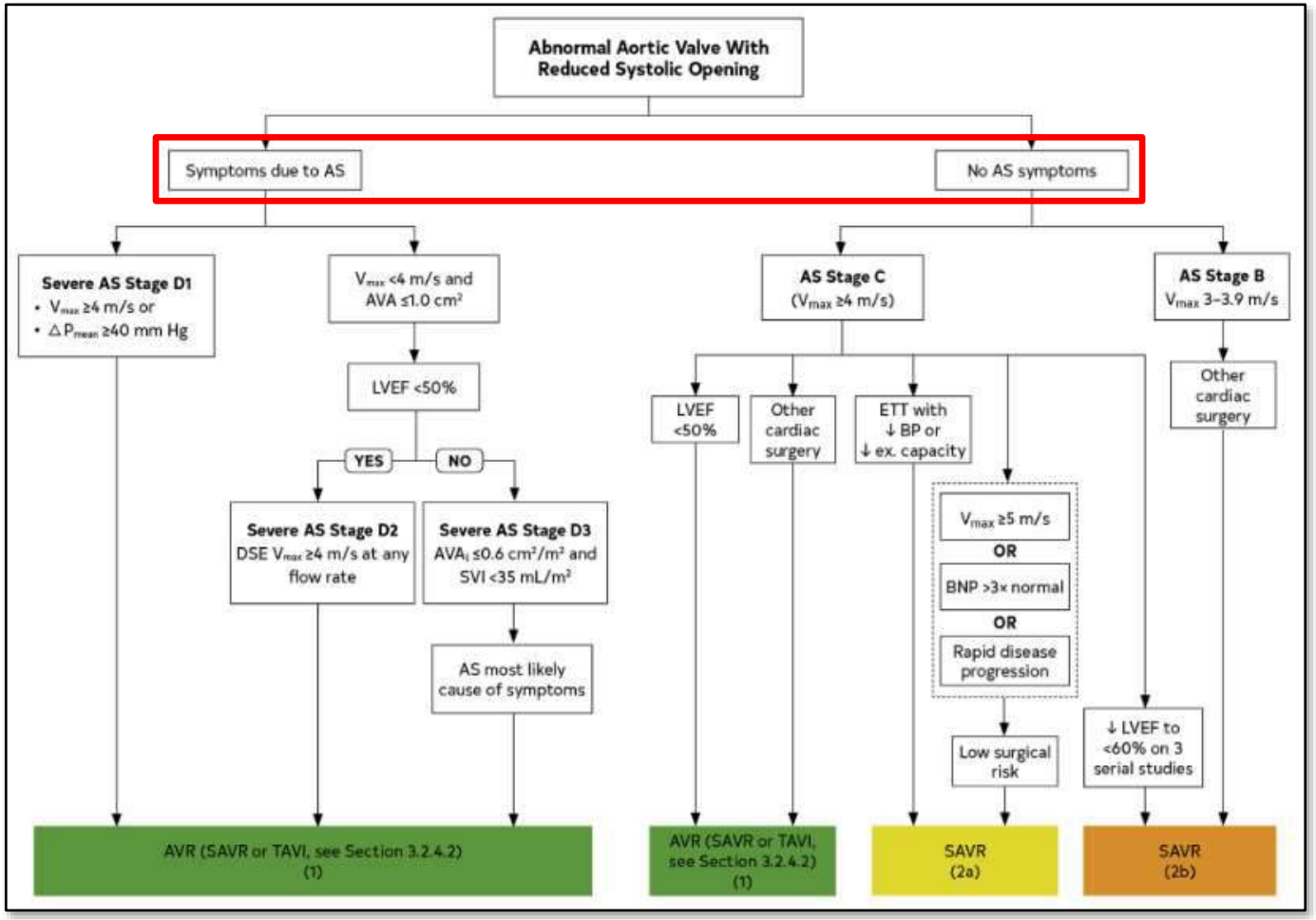

CHEST PAIN


SHORTNESS OF BREATH


EXERCISE INTOLERANCE

- rTOF pulmonary valve replacement

Symptom Provocation



- rTOF pulmonary valve replacement
- Valvular heart disease

Catherine M. Otto et al. "2020 ACC/AHA guideline for the management of patients with valvular heart disease." *JACC*

Risk Assessment

Transposition of the Great Arteries: After Atrial Switch (Mustard or Senning Operation)

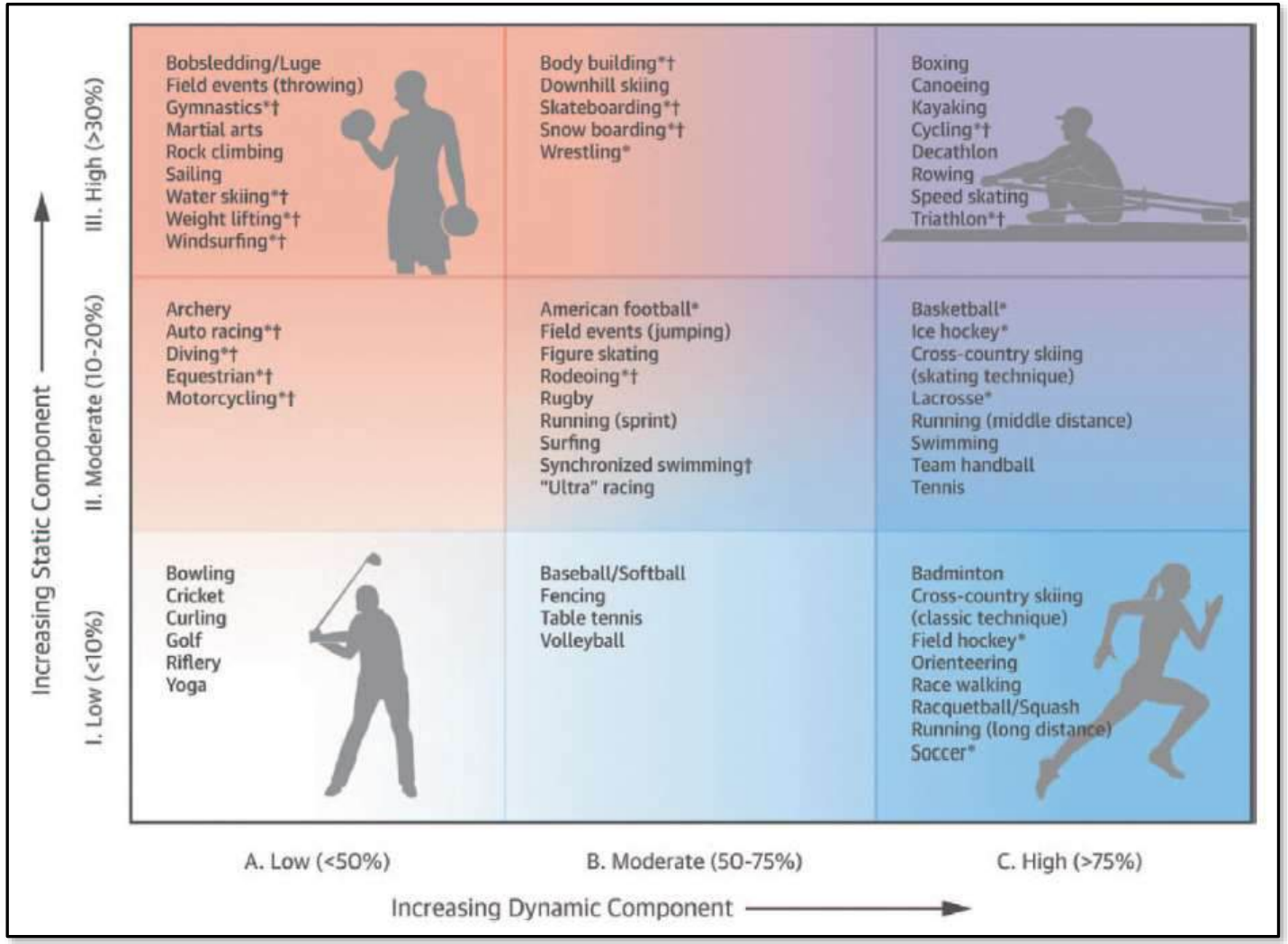
Recommendations

1. It is recommended that before participation in competitive sports, all athletes who have undergone the Senning and Mustard 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. Participation in competitive sports in those athletes with a history of clinically significant arrhythmias or severe ventricular dysfunction may be considered on an individual basis based on clinical stability *(Class IIb; Level of Evidence C)*.
3. Athletes without clinically significant arrhythmias, ventricular dysfunction, exercise intolerance, or exercise-induced ischemia may be considered for participation in low- and moderate-intensity competitive sports (classes IA, IB, IIA, and IIB) *(Class IIb; Level of Evidence C)*.

Ventricular Dysfunction After CHD Surgery

Recommendations

1. Before participation in competitive sports, all athletes with ventricular dysfunction after CHD surgery should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing *(Class I; Level of Evidence B)*.
2. Athletes with normal or near-normal systemic ventricular function (EF ≥50%) can participate in all sports *(Class I; Level of Evidence B)*.
3. It is reasonable for athletes with mildly diminished ventricular function (EF 40%-50%) to participate in low- and medium-intensity static and dynamic sports (classes IA, IB, and IIA and IIB) *(Class IIb; Level of Evidence B)*.



Maron, Barry J. et al. "Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities." *Circulation*

Congenitally Corrected TGA

Recommendations

1. It is recommended that before participation in competitive sports, all CCTGA athletes should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing *(Class I; Level of Evidence B)*.
2. Participation in competitive sports in those CCTGA athletes with a history of clinically significant arrhythmias or severe ventricular dysfunction may be considered on an individual basis based on clinical stability *(Class IIb; Level of Evidence C)*.

Coarctation of the Aorta: Treated by Surgery or Balloon and Stent

Recommendations

1. Athletes who are >3 months past surgical repair or stent placement with <20 mm Hg arm/leg blood pressure gradient at rest, as well as (1) a normal exercise test with no significant dilation of the ascending aorta (z score <3.0), (2) no aneurysm at the site of coarctation intervention, and (3) no significant concomitant aortic valve disease, may be considered for participation in competitive sports, but with the exception of high-intensity static exercise (classes IIIA, IIIB, and IIIC), as well as sports that pose a danger of bodily collision *(Class IIb; Level of Evidence C)*.

Postoperative Tetralogy of Fallot

Recommendations

1. Before participation in competitive sports, it is recommended that all athletes with repaired tetralogy of Fallot should undergo evaluation, including clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing *(Class I; Level of Evidence B)*.
2. Athletes without significant ventricular dysfunction (EF >50%), arrhythmias, or outflow tract obstruction may be considered for participation in moderate- to high-intensity sports (class II to III). To meet these criteria, the athlete must be able to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms *(Class IIb; Level of Evidence B)*.

Martinez, Matthew W. et al. "Sports Participation by Athletes With Cardiovascular Disease." *JACC*

What you learned today

- Another non-invasive tool
- CHD surveillance and progression
- Objective assessment of symptoms
- Keep our patient safe **AND** active
- Consult your local exercise expert
- Become the local expert and build a lab!

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