CARDIOLOGY 2024

Lifting Expectations: Exploring Power Lifting in Fontan Circulation

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Diagnosis List

1. 13 y.o. young man with hypoplastic left heart syndrome
   a) s/p Norwood procedure (11/2008)
   b) s/p Hemi-Fontan procedure with PA augmentation and tricuspid valvuloplasty, (4/2009)
   c) Narrowing of the SVC hemi-Fontan anastomosis by cath (5/2009)
   d) Lateral tunnel Fontan with tricuspid valvuloplasty (4/2011)
2. Tricuspid regurgitation, moderate
   a) Status post tricuspid valvuloplasty x2, (7/2009, 4/2011)
   b) Residual moderate tricuspid regurgitation, unchanged
3. Mildly depressed right ventricular systolic function by echo, stable for several years
4. History of junctional ectopic tachycardia, resolved
5. Pseudo pre-excitation; brief, slow SVT noted on Holter 2018

Diagnostic Data

- Current Medications
- ECG
- Echo
- MRI/CT
- Cath
- Holter
- CPET
- Implanted Devices

Diagnostic Data

- Current Medications
  - aspirin, enalapril, spironolactone
- ECG
  - NSR, RVH
- Echo
  - Unobstructed Fontan conduit
  - Small fenestration (4mmHg)
  - Mild RV dysfunction
  - Moderate TR from 2 jets, unchanged from prior studies
- MRI
  - Severe RV dilation (173 mL/m^2)
  - TR regurgitant fraction 32%
  - Fenestration 20% of caval flow
  - Unobstructed PAs and Ao
- Cath – None recently
- Holter
  - HR: mean 81 bpm (range 56-145)
  - Arrhythmia burden: 2 short runs of SVT (fastest 161 bpm, longest 7 beats)

History

- WK reports he has already been exercising with his father for the past 4 months, **lifting weights under his father's supervision**.
- He typically lifts for about **45-60 minutes most days of the week** and alternates muscle groups each lifting session as well as completes core strengthening exercises.
- He reports he normally will complete **6-10 repetitions in each set** without difficulty.
- He does report **occasional one rep max efforts** to gauge his interval strength improvements.
- He had one **episode of dizziness with bench pressing** prior to starting to lift with his father – he was with a group of **older kids who were challenging him** to bench press 105 pounds, even though he had never lifted any weights before that time.

Baseline CPET

Stage Data

Phase	Stage	Work	HR	BP	SpO2	QTc II	QTc V5	Comment
Pre-Test	Seated		100	102/64	88			
	Warm-up		103	108/72	88			
Exercise	3:00		121	120/78	86			PVC
	6:00		155	120/80	89			RPE 4/10, no symptoms
	Peak (7:27)	108 watts	157	124/70	84			RPE 8-9/10; PVC
Recovery	1:00		139	120/-	85			PVC
	4:00		114	110/68	87			PVC
	8:00		111	110/60	87			

Metabolic Data

	VO2			VE/VCO2		O2 Pulse		OUES	
Phase	ml/kg/min	z-score	RER	slope	z-score	ml/beat	z-score	slope	z-score
Rest	5.5	–	0.81	–	–	2.9	–	–	–
AT	16.7	-2.0	–	31.8	2.7	–	–	1413	-2.6
Peak	26.3	-3.8	1.01	37.9	2.6	9.0	-2.7	1574	-3.3

Reference Dataset: Blanchard

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Rest	5.5	–	0.81	–	–	2.9	–	–	–
AT	16.7	-2.0	–	31.8	2.7	–	–	1413	-2.6
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Reference Dataset: Blanchard

Final Exercise Test Interpretation

1. This was a submaximal exercise test (RER 1.01; HR 76% of age predicted max).
2. Peak oxygen consumption was 1412 ml/min (z-score -3.8). This equates to 26.3 ml/kg/min and 53% of predicted.
3. Ventilatory anaerobic threshold occurred at 63% of peak oxygen consumption and 34% of predicted peak oxygen consumption.
4. Peak O2 pulse was 9 mL/beat (z-score -2.7).
5. Heart rate response was submaximal, though cannot rule out chronotropic incompetence.
6. Blood pressure response to exercise was normal.
7. Oxygen saturation was normal at baseline and unchanged with exercise.
8. Ventilatory efficiency (VE/VCO2) slope from rest to AT was 31.8 (z-score 2.7) and from rest to peak was 37.9 (z-score 2.6).
9. There were no exercise induced symptoms.
10. The ECG was sinus rhythm, RVH with repolarization abnormalities and pseudo-pre excitation pattern at baseline and throughout exercise. PVCs of 2 morphologies were present throughout exercise and recovery. There was 1.5-2.5 mm ST segment depression in the lateral precordial and inferior leads that is most consistent with baseline RVH changes. The specificity for ischemia in this population is very limited.
11. Other comments: Baseline spirometry was normal.
12. There is no prior test for comparison.

ExRx

Select Exercise Prescription Criteria

○ FORCE (Early Intervention ExRx) ◉ Budts (HEART Club)

Physiologic Variables

Ventricles

Anatomy

○ Normal ◉ Single V or Systemic RV

Dysfunction

○ None ◉ Mild ○ Moderate ○ Severe

Hypertrophy

○ Normal ○ Mild ◉ Moderate ○ Severe

Pressure Load

◉ Normal ○ Mild ○ Moderate ○ Severe

Volume Load

○ Normal ○ Mild ◉ Moderate ○ Severe

Pulmonary Artery Pressure

◉ Normal ○ Mildly Elevated ○ Moderate / Severly Elevated

Aortic Dilation

◉ None/Mild ○ Moderate ○ Severe ○ Needs Repair

Coronary Anatomy and Function

◉ Normal ○ Low Concern ○ High Concern

Arrhythmias

○ None ◉ Mild / Non-Malignant ○ Significant / Malignant

Arterial Saturation with Exercise

○ No Cyanosis ◉ Central Cyanosis

Anatomic/Physiologic Definitions

ExRx

Exercise Program Phase

(•) Beginner ( ) Intermediate ( ) Advanced

Intensity Targets

Physiologic Category	A	B	C	D	E
Static Component	High	Moderate	Moderate	Moderate	Low
Relative Intensity	High	High	Moderate	Low	Low
Training HR Range	145 to 150 BPM	145 to 150 BPM	135 to 145 BPM	130 to 135 BPM	130 to 135 BPM

The default intensity targets are based off the selected CPET data and may need to be individualized. If needed, you may override the defalts by filling in the optional fields below.

Alternate Lower HR Target (Optional)

105

Alternate Upper HR Target (Optional)

120

Alternate Talk Test (Optional)

- ( ) breathing too hard to talk
- (•) breathing harder but can still have a conversation
- ( ) breathing easy, can talk and sing

Alternate RPE (Optional)

- ( ) 7-9
- (•) 4-6
- ( ) 2-3

Exercise Prescription

Exercise Program Phase: Beginner

Intensity Goal: Low Intensity

- *How hard you work:* 4-6 on a 0-10 scale
- *Heart rate zone:* 105 to 125 beats per minute
- *Talk test:* breathing harder but can still have a conversation

Physiologic Category: D

Frequency Goal: 6 days per week

- *Aerobic:* 2-3 days per week
- *Strength:* 2-3 days per week
- *Flexibility and Balance:* 1-2 days per week

Prescriber Notes: Heart rate at AT during exercise test was ~125 BPM, will increased heart rate target in the future if doing well and ready for progression.

This exercise prescription is designed to promote regular physical activity for individuals with congenital heart disease. **Using guidelines for competitive sports participation is not recommended for those interested in leisure and recreational physical activity.**[1] This exercise prescription has been created based on your patient's physiologic status and data obtained from their most recent cardiopulmonary exercise test for use during recreational physical activity using consensus recommendations from AHA/ACC/ESC.[2-4]

"We recommend that he modify his current exercise program to includes aerobic and flexibility training in addition to the strength training he is already doing. We had a long discussion regarding the importance of safely lifting weights. We **discouraged WK from trying to attempt "one-rep max" lifting maneuvers** and explained that he should be able to easily breathe without Valsalva while lifting. I think this is going to be one of our primary challenges while working with WK in HEART club. Over time, we will try and help him **build internal metrics** for his physical activity and wellness **instead of comparison to peers** as this is going to become increasingly complicated as he enters high school."

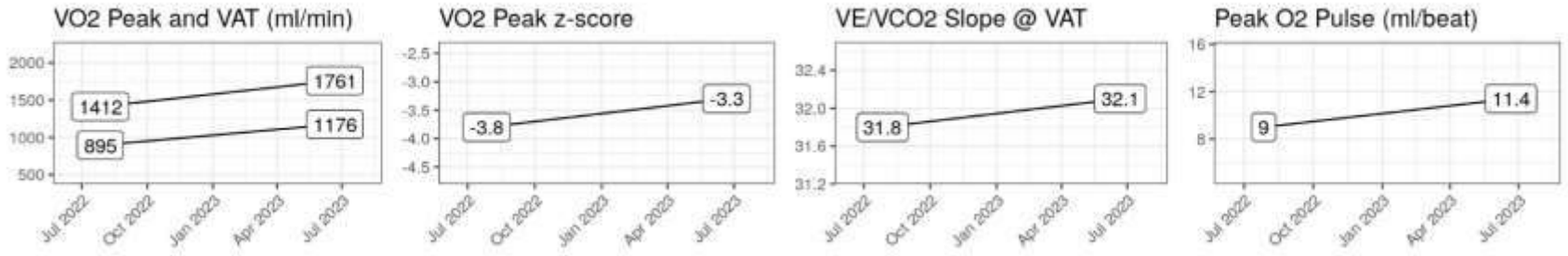
How it’s going…

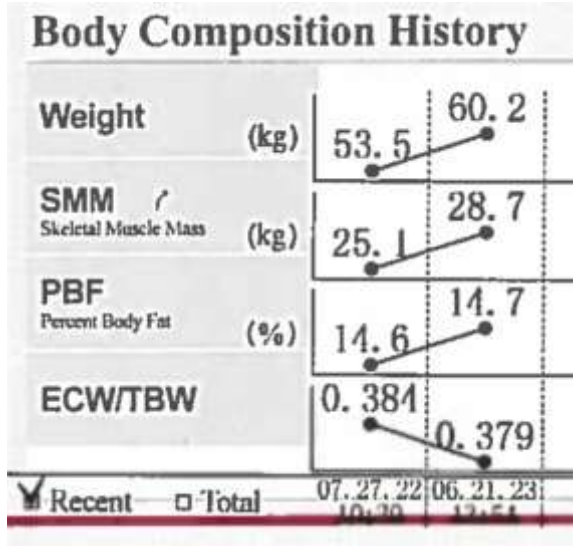
Metabolic Data

	VO2			VE/VCO2		O2 Pulse		OUES	
Phase	ml/kg/min	z-score	RER	slope	z-score	ml/beat	z-score	slope	z-score
Rest	6.1	–	0.82	–	–	3.7	–	–	–
AT	19.5	-1.6	–	32.1	3.4	–	–	1884	-1.8
Peak	29.3	-3.3	1.10	40.1	3.3	11.4	-2.1	1805	-3.0

Reference Dataset: Blanchard

Historical Trends





How it’s going…

1. Hypoplastic left heart syndrome (MA/AA), s/p Norwood-BT (11/04/2008), s/p hemi-Fontan with pulmonary artery augmentation and tricuspid valvuloplasty (03/24/2009), lateral tunnel fenestrated Fontan with additional tricuspid valvuloplasty (04/25/2011).
2. No Fontan pathway obstruction.
3. A small fenestration is seen.
4. Fenestration mean gradient of at least 3 mmHg --likely underestimated due to a suboptimal Doppler angle.
5. Smaller right pulmonary artery than left with unobstructed bilateral antegrade flow.
6. Unrestrictive flow across the atrial septum.
7. Trivial tricuspid stenosis, mean gradient = 4 mmHg at 82 bpm. Mild regurgitation via multiple jets that is mildly improved from the prior study.
8. Moderately dilated RV and mild RV hypertrophy with mildly depressed systolic function of the right ventricle.
9. RV global function is unchanged compared to the prior study.
10. Native to neo-aortic (Damus) connection is widely open.
11. Trivial neo-aortic valve regurgitation.

How it’s going…

12/2023 Cath	Baseline	After fluid challenge, contrast, VV-collateral occlusion
Fontan pressure	12 mmHg	19 - 20 mmHg
RVEDP	8 mmHg	16 mmHg
Ao saturation	85%	91%

How it’s going…

- Dropped high school gym class (peer/coach pressure)
- Not using supplements
- Wants to participate in a natural body building competition
- Continues to challenge his cardiologist with exercise and weightlifting questions
- Both exercise team and primary cardiologist have concerns about body dysmorphia