Regular Exercise for the Fontan Circulation: How important is it?

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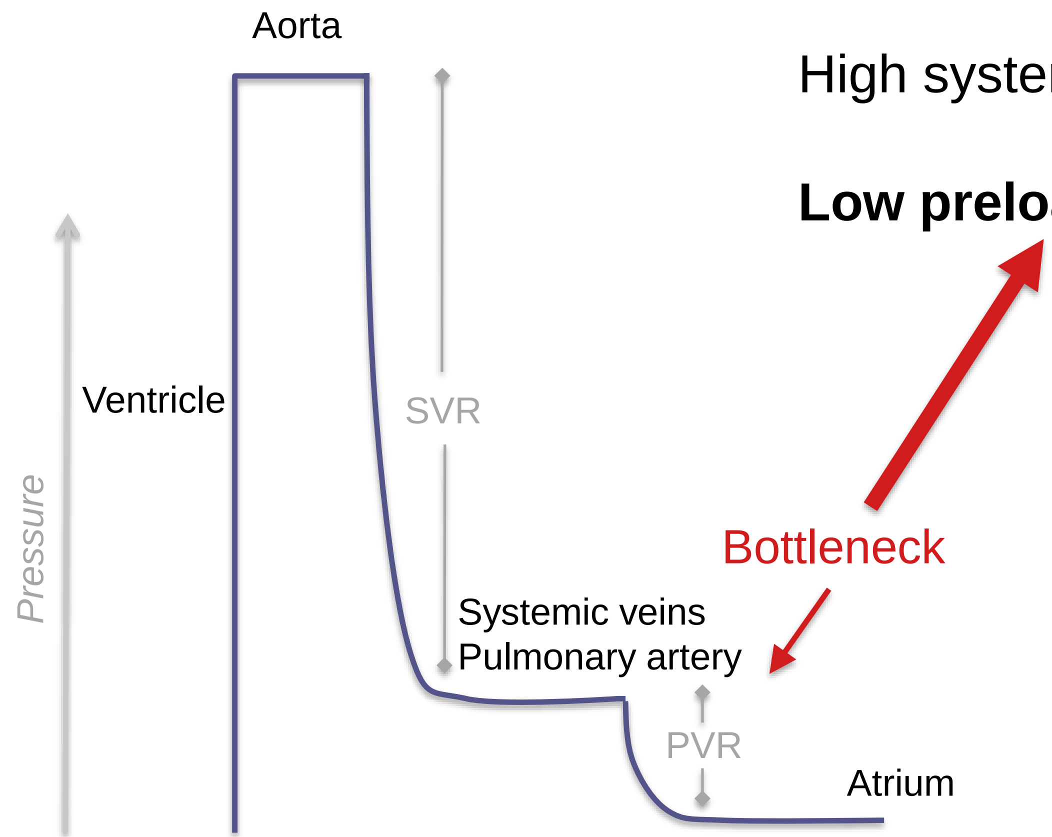
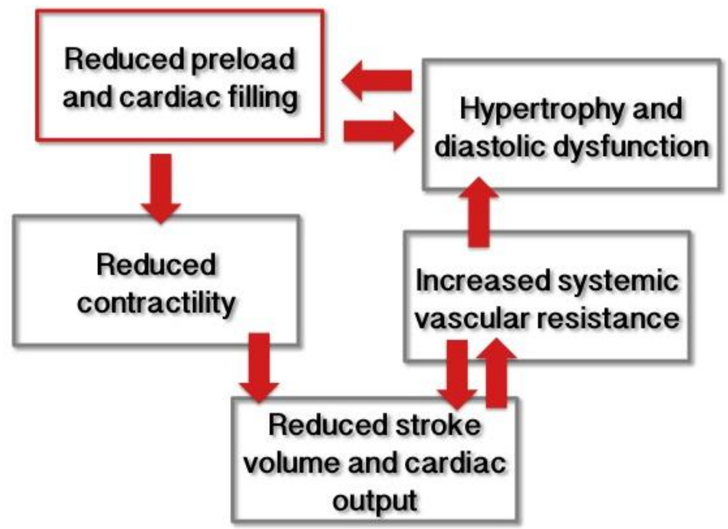
The Fontan Circulation

High systemic venous pressure

Low preload = ↓SV rise during exercise

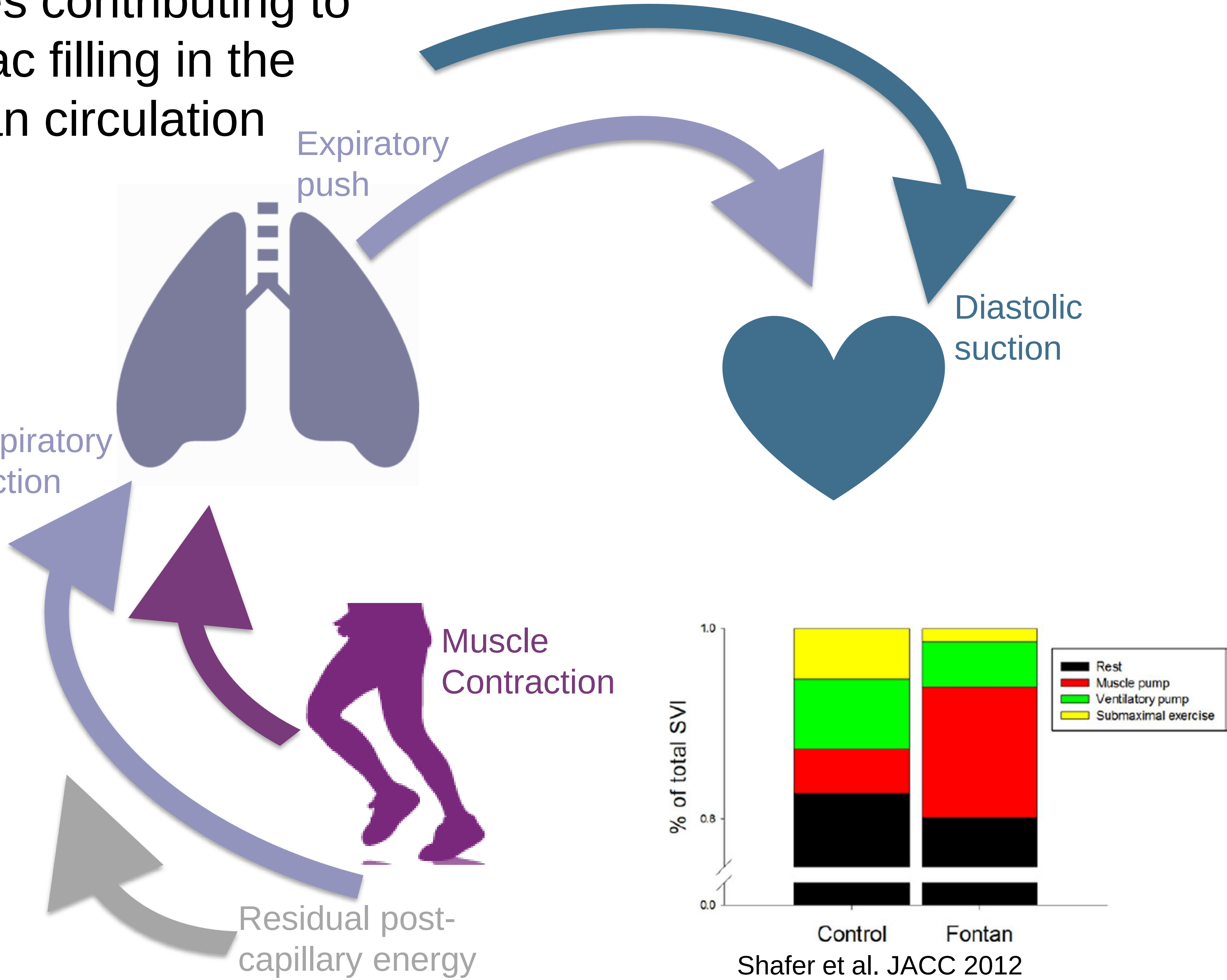




Systemic vascular resistance=SVR
Pulmonary vascular resistance=PVR

Courtesy of Marc Gewillig

Forces contributing to cardiac filling in the Fontan circulation



Shafer et al. JACC 2012

A Fontan Myopathy?

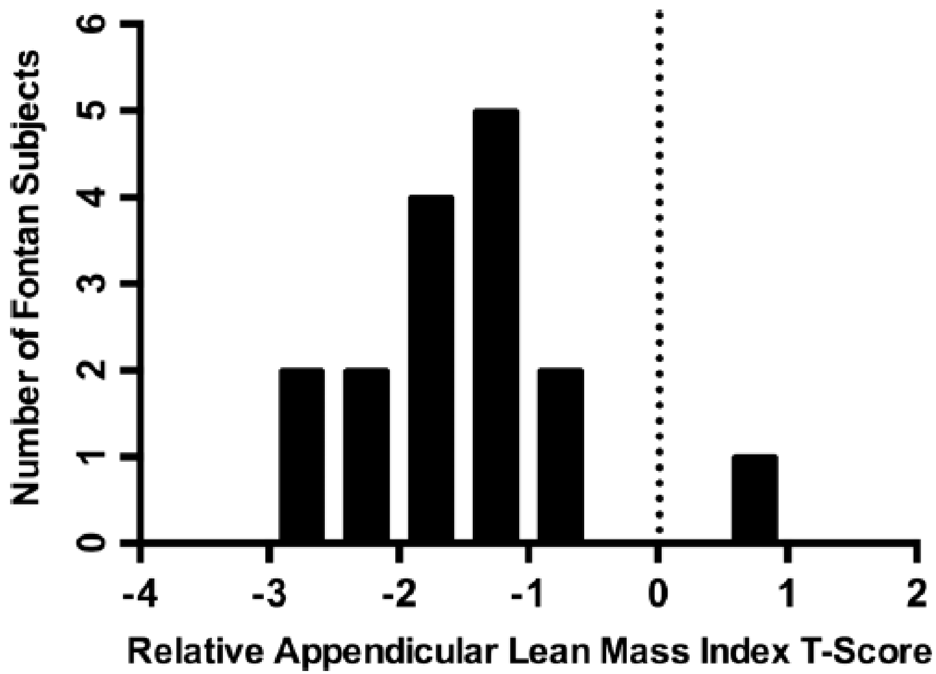


Figure 1 A T-score represents the number of SDs from the young normal reference mean. A value <−2.0 represents marked muscle wasting, defined as in the sarcopenic range.

Mean T-score was -1.47 ± 0.21

Mean Z-score was markedly abnormal −1.46 ± 0.22

(p<0.0001)

Cordina et al. Heart 2013

A Fontan Myopathy?

Congenital heart disease

ORIGINAL ARTICLE

Lean mass deficits, vitamin D status and exercise capacity in children and young adults after Fontan palliation

Catherine M Avitabile,[1] Mary B Leonard,[2,3,4] Babette S Zemel,[4] Jill L Brodsky,[5] Dale Lee,[6] Kathryn Dodds,[1] Christina Hayden-Rush,[1] Kevin K Whitehead,[1,4] Elizabeth Goldmuntz,[1,4] Stephen M Paridon,[1,4] Jack Rychik,[1,4] David J Goldberg[1,4]

Heart 2014

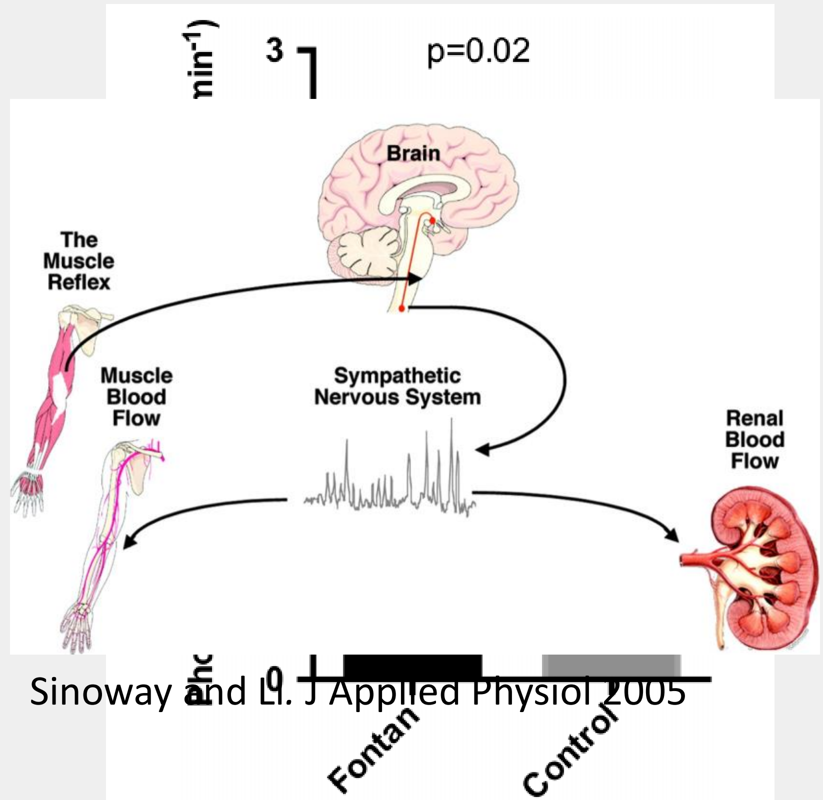
A Fontan Myopathy?

The ability of skeletal muscle to extract oxygen during exercise and post-exercise reoxygenation is abnormal.

Inai *et al.* American Journal of Cardiology 2004

Important skeletal muscle afferent nerves that control blood flow and other autonomic responses are impaired in Fontan.

Brassard *et al.* International Journal of Cardiology 2006



Sinoway and Li. J Applied Physiol 2005

Cordina et al. Heart 2013

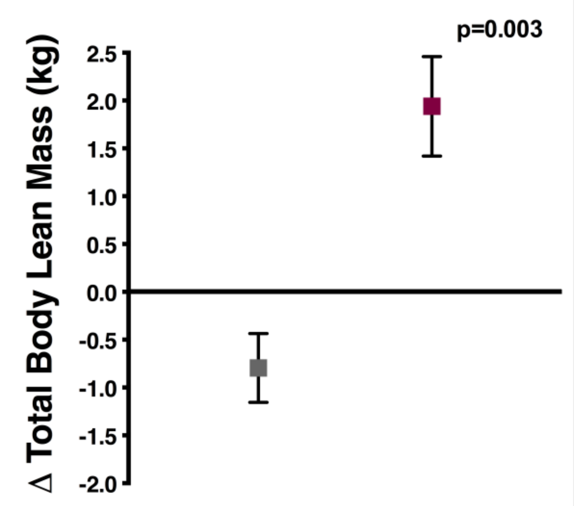
↓

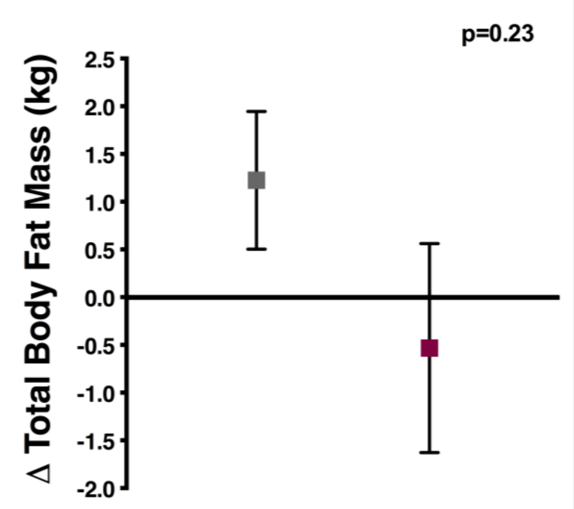
After 8 weeks of aerobic and light resistance training, ergoreceptor function normalised.

Can the muscle pump be augmented?

- We hypothesised that resistance training to augment the peripheral muscle pump in subjects with a Fontan circulation might improve:
  - Cardiac filling
  - Stroke volume and
  - Exercise capacity
- 11 adults (mean 31 years, 2 females, 6 trainers and 5 controls) were recruited
- 3 days/week of high-intensity total body resistance training for 20 weeks. Carefully trained and supervised

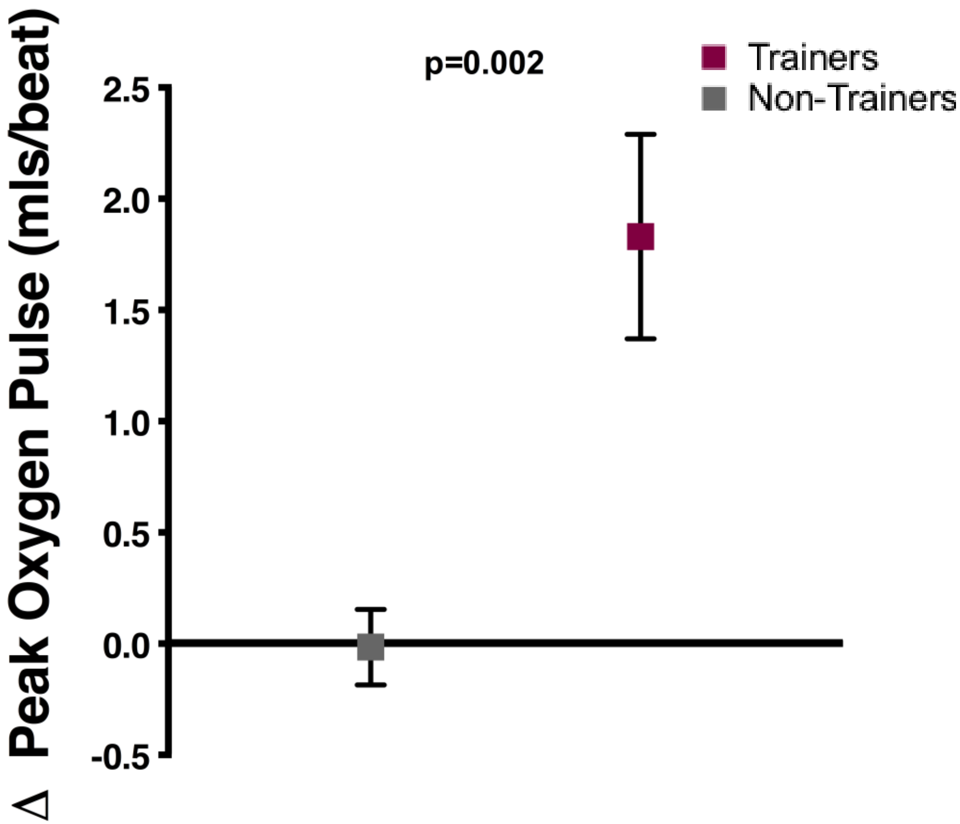
Strength increased by 43 ± 7 % (p=0.002)

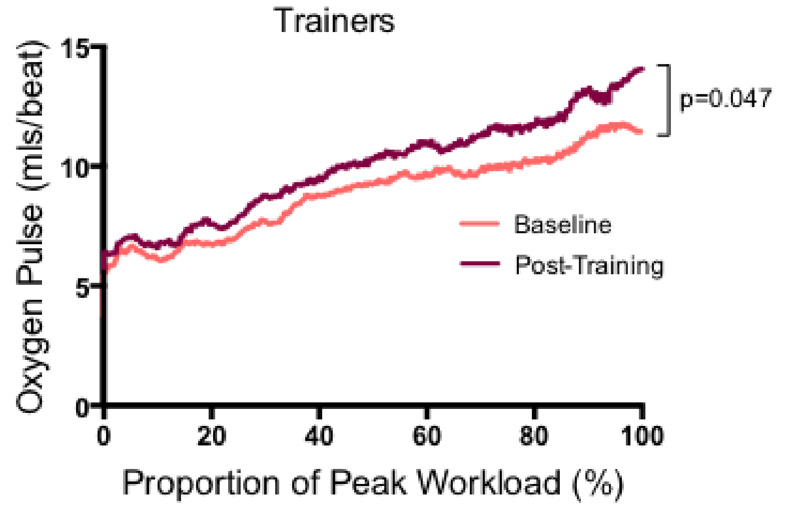


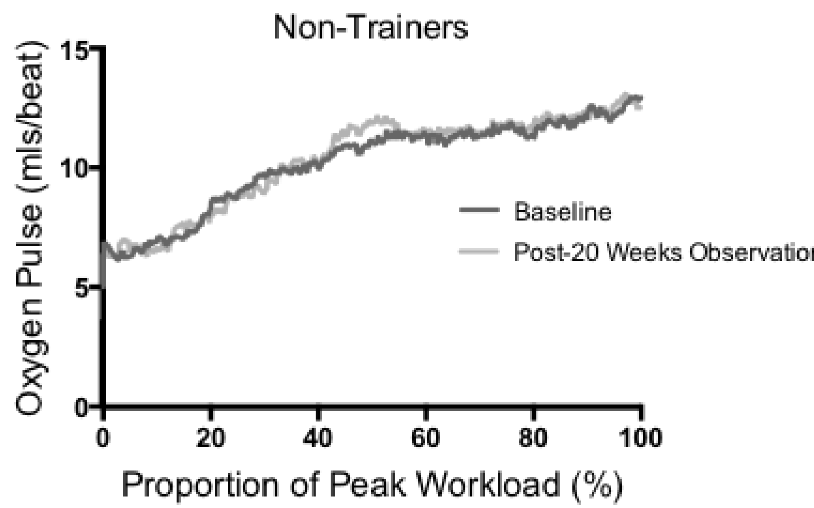


Cordina...Celermajer, International Journal of Cardiology 2013

Peak VO_2 increased by 10% (p=0.03)



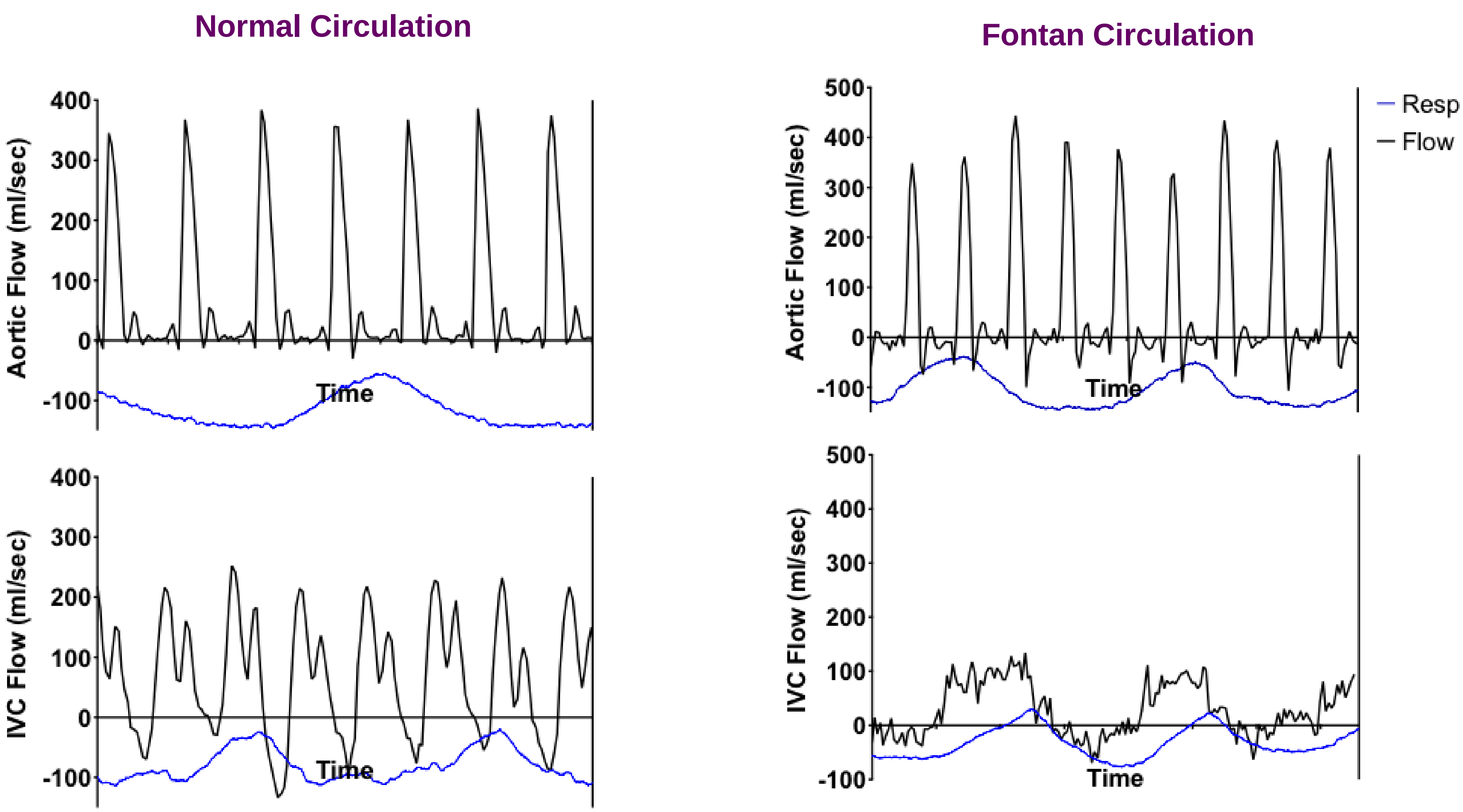




Cordina et al. International Journal of Cardiology 2013

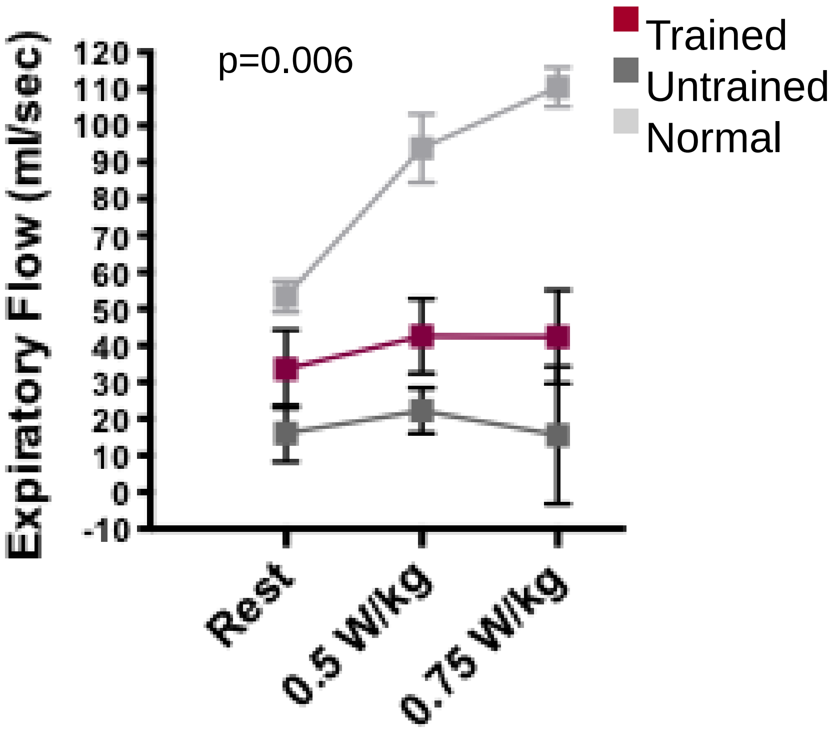
Fontan REGISTRY rpa

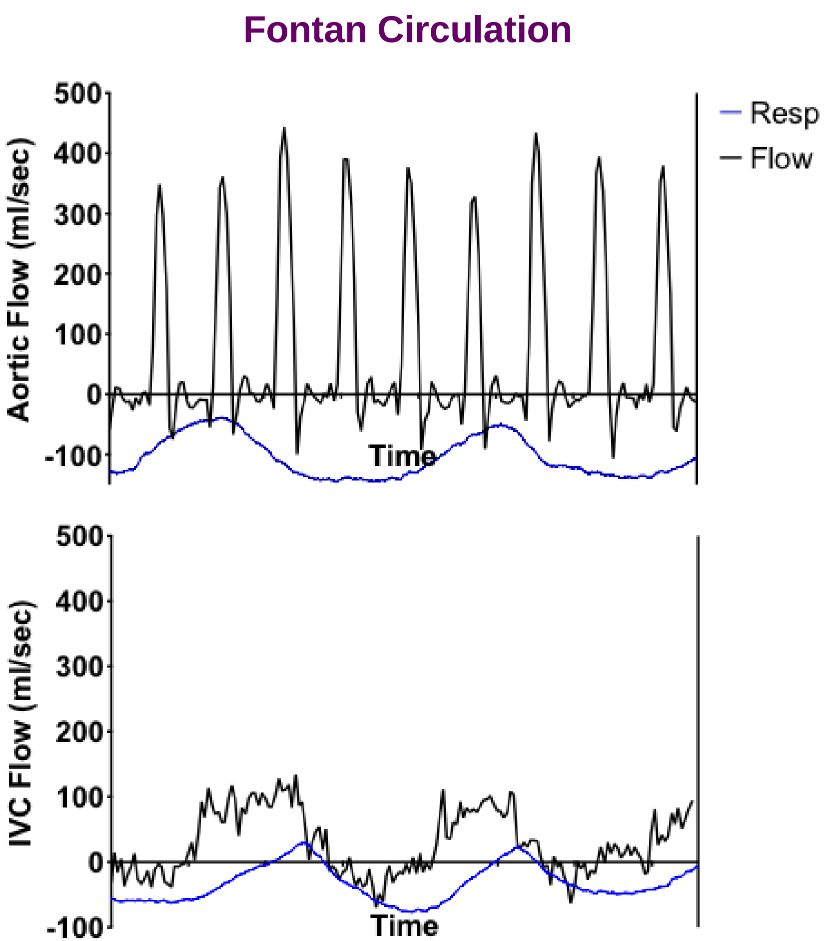
Respiratory Dependence in Fontan



Cordina et al. International Journal of Cardiology 2013

Respiratory Dependence in Fontan





Cordina et al. International Journal of Cardiology 2013

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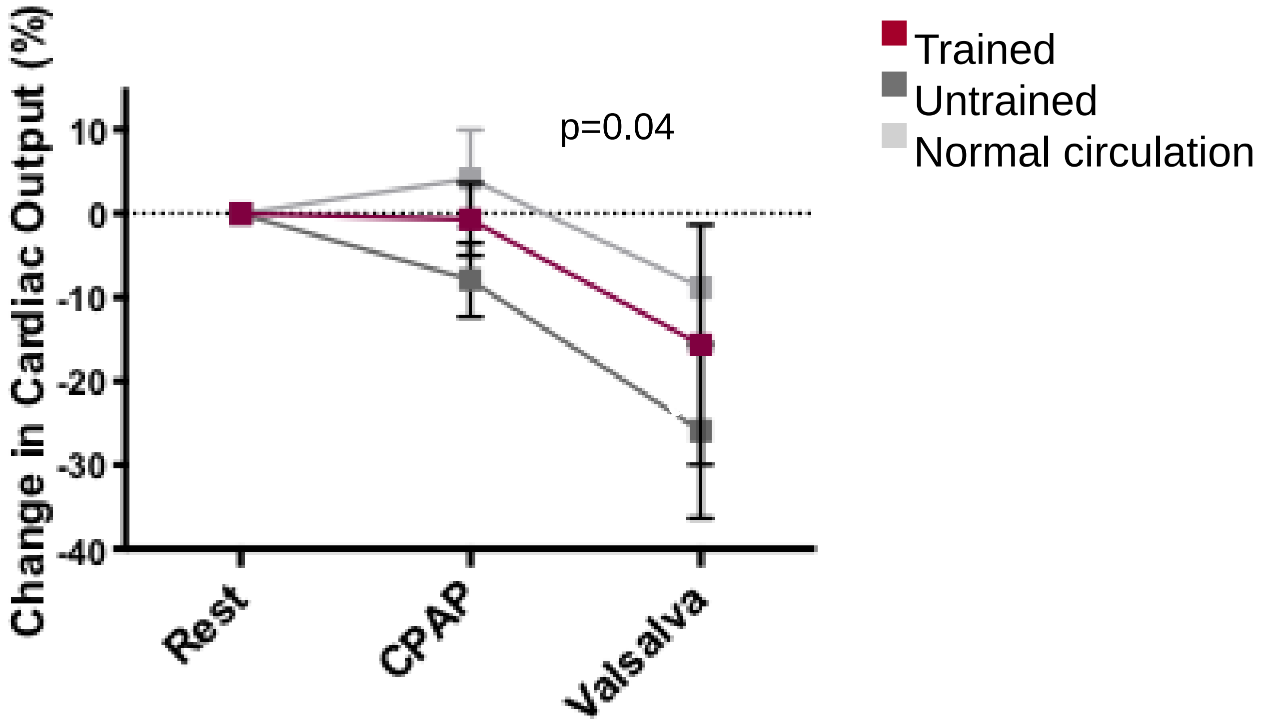
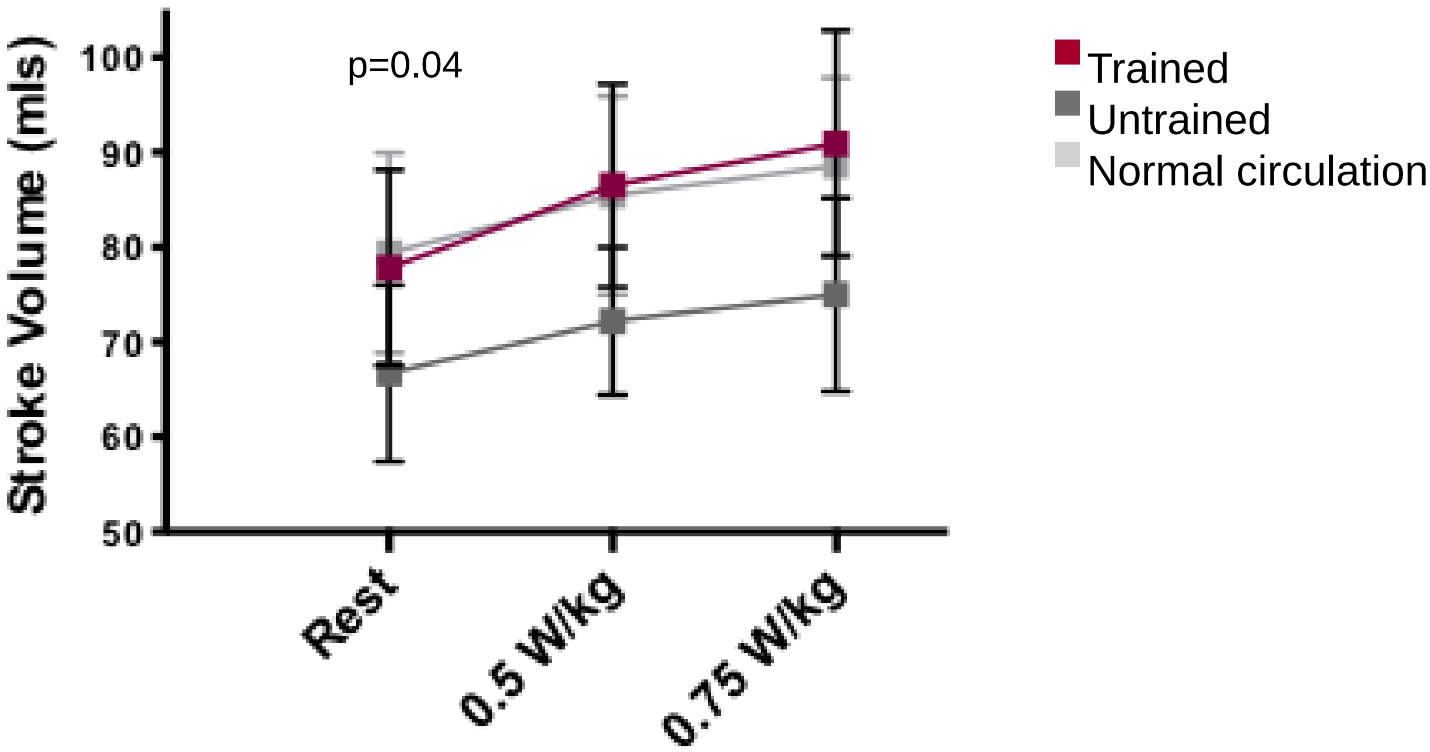
Effects of Training on Tolerance to Inspiratory Stress Assessed at Free Breathing Real-Time MRI



Cordina et al. International Journal of Cardiology 2013

Real time free-breathing MRI



Cordina et al. International Journal of Cardiology 2013

Aerobic training can also improve exercise capacity

	Mean Age (yr ± SD)	n	Training Design	Results
Minamisawa *et al.* 2001	19 ± 4	16	2-3 months home-based aerobic exercise (60-80% peak HR) Uncontrolled	□VO_2 (7%) O_2 pulse tended to improve (5%)
Rhodes *et al*. 2005	12 ± 2	11/16 Fontan	12 weeks aerobic (game-based) + light resistance Uncontrolled	□VO_2 (22%) and O_2 pulse (18%)
Opocher *et al*. 2005	9 ± 1	10	8 months of aerobic home-based training (up to 70% VO_{2peak}) Uncontrolled	□VO_2 (20%) and O_2 pulse (16%)
Longmuir *et al.* 2013	9 ± 2	61	12 month physical activity prescription vs. education and game-based intervention	Both □VO_2 (5%) and motor skills Maintained □ in physical activity at 2y

200 patients, no adverse events

What are the mechanisms that drive improvement?

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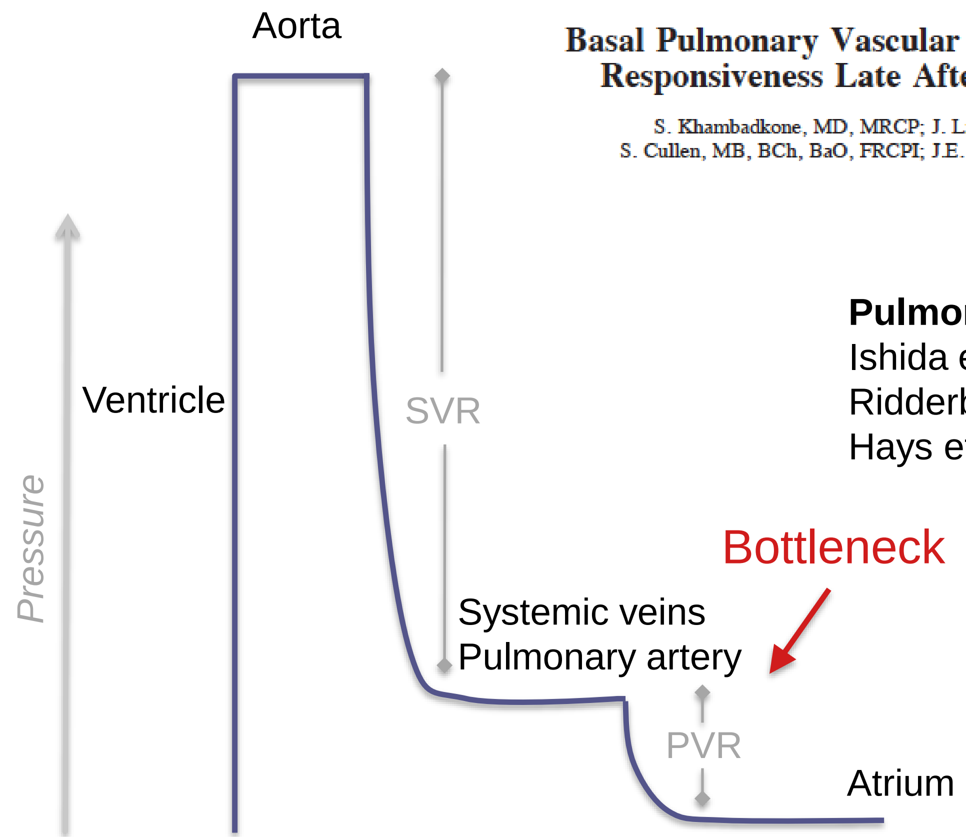
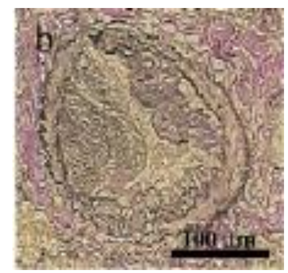
The Fontan Circulation

Basal Pulmonary Vascular Resistance and Nitric Oxide Responsiveness Late After Fontan-Type Operation

S. Khambadkone, MD, MRCP; J. Li, MBBS; M.R. de Leval, MD, FRCS;
S. Cullen, MB, BCh, BaO, FRCPI; J.E. Deanfield, FRCP; A.N. Redington, FRCP



Pulmonary vascular remodelling
Ishida et al. Int J Cardiol 2012
Ridderbos et al. JHLT 2015
Hays et al. Heart 2017



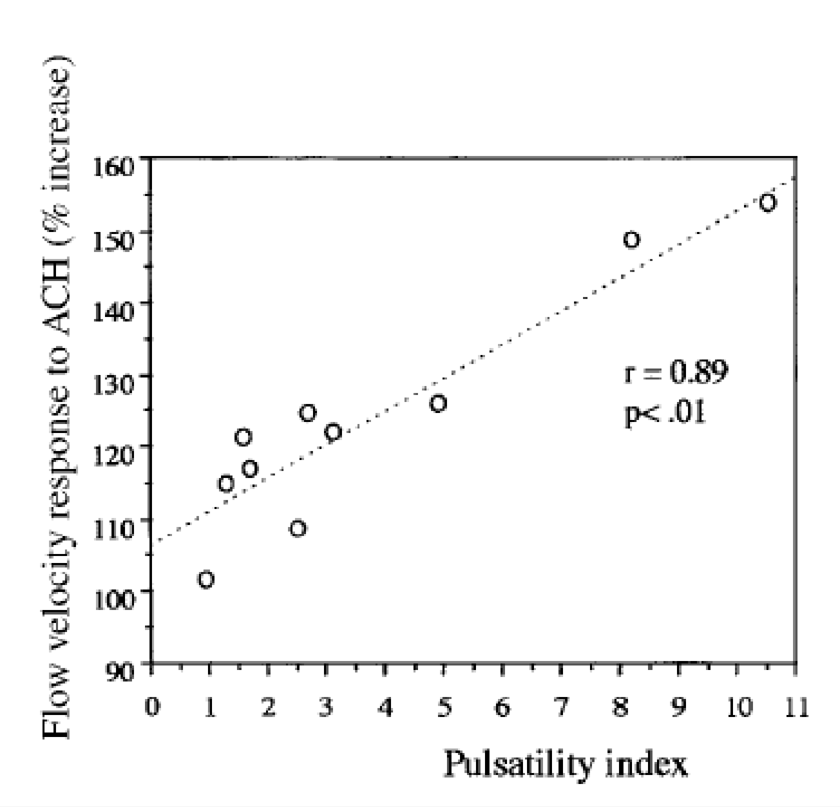
Systemic vascular resistance=SVR
Pulmonary vascular resistance=PVR

Courtesy of Marc Gewillig

Fontan - Pulmonary Endothelial Function

- There is evidence for pulmonary endothelial dysfunction that probably relates to reduced flow and pulsatility in the vascular bed.

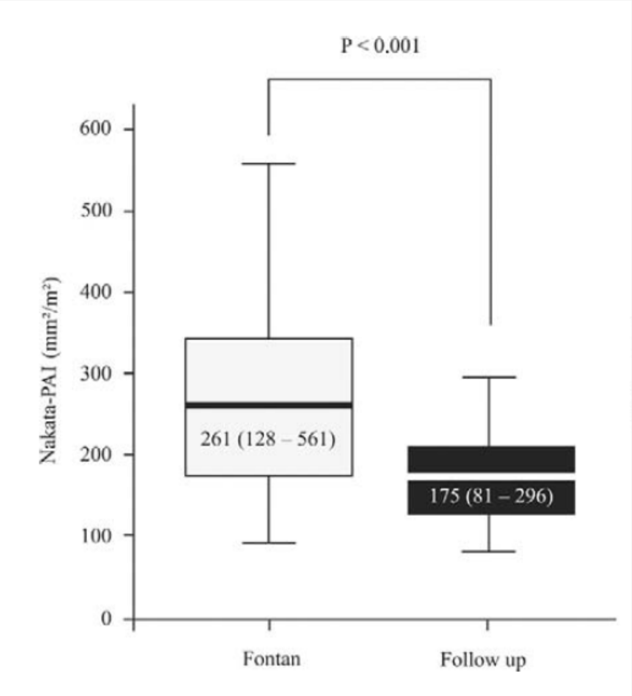
  Kurotobi et al., JTCVS 2001
  Khambadkone et al., Circ 2003.



Kurotobi et al. JTCVS 2001

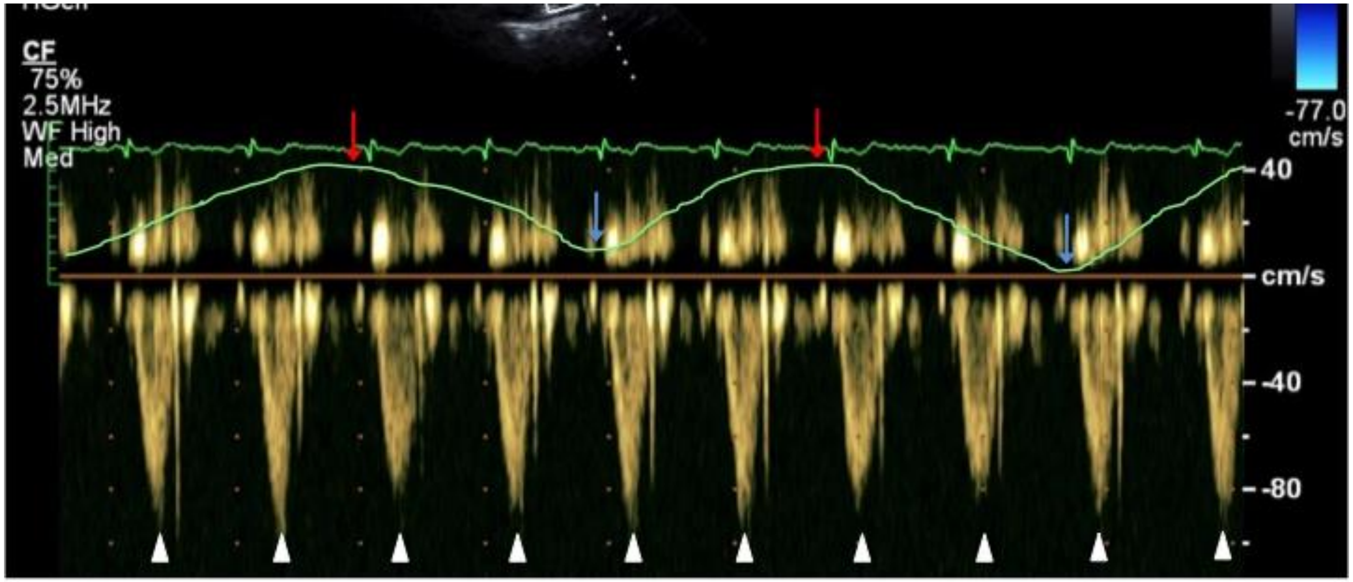
Fontan - Pulmonary Artery Growth

- Pulmonary artery growth is probably attenuated or even ceases at Fontan completion

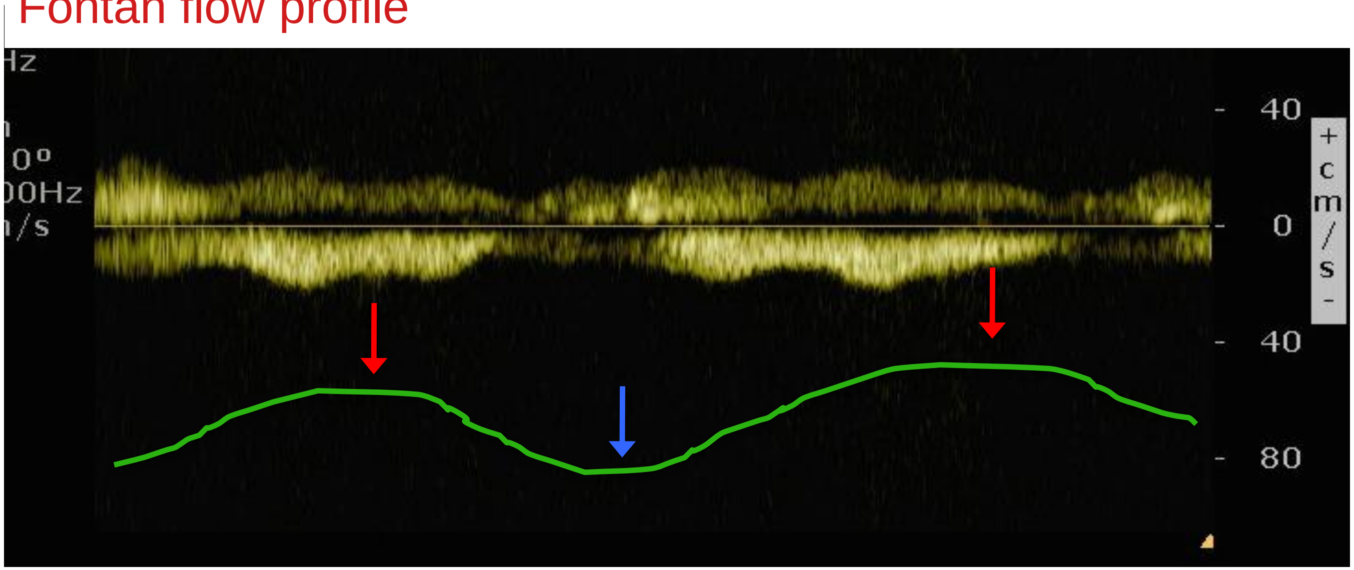


Ouvroutski et al. Annals of Thoracic Surg 2009

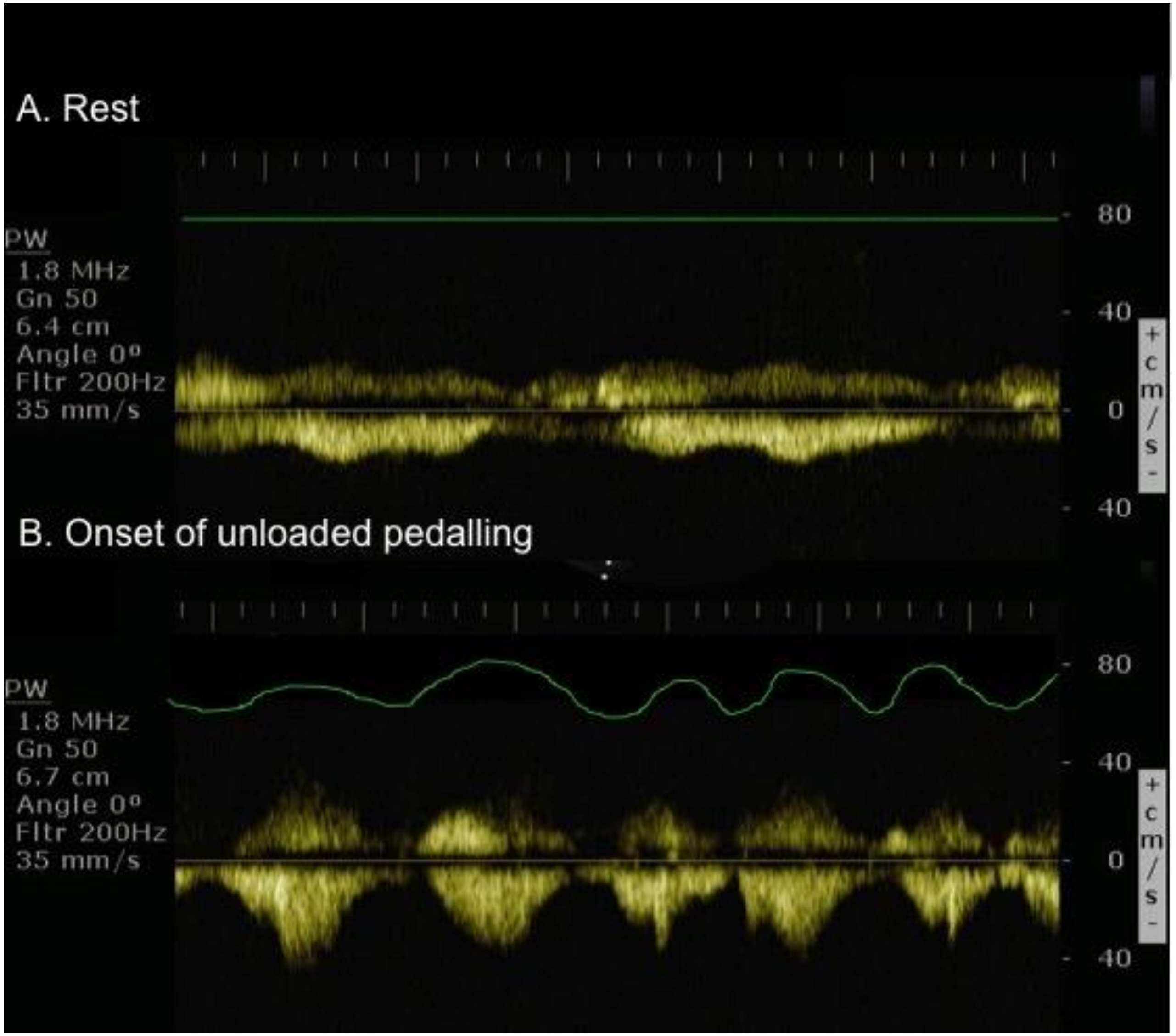
Normal flow profile in pulmonary artery



Fontan flow profile



Fontan – The Peripheral Muscle Pump



Cordina et al. Under Review

The Journal of Thoracic and Cardiovasc[...]

Available online 27 October 2017

In Press, Accepted Manuscript

Super-Fontan: Is it possible?

Rachael Cordina MBBS PhD FRACP [a, b], Karin du Plessis PhD [c, d], Derek d'Udekem MD PhD FRACS [c, d, e]

TABLE 1. Fontan subject characteristics

	All (N = 14)
Sex (female:male)	7:7
Age (y)	24 ± 5 (16-34)
BMI (kg/m^2)	23.0 ± 3.0 (16.7-27.8)
Predominant ventricular morphology	
Left	12 (86)
Right	1 (7)
Biventricular	1 (7)
Dominant cardiac defect	
Tricuspid atresia	10 (71)
Double-inlet left ventricle	2 (14)
Complex double-outlet right ventricle	2 (14)
Type of TCPC repair	
Atriopulmonary connection	3 (21)
Intracardiac lateral tunnel	7 (50)
Extracardiac conduit	3 (21)
Extracardiac conduit post-APC conversion*	1 (7)
Patent fenestration	5 (36%)
Pacemaker	
AAI (not paced during exercise)	1 (7)
DDDR (100% ventricular pacing)	1 (7)
DDDR (atrial pacing during exercise)*	1 (7)
Age at Fontan completion (y)	4 ± 2 (2-11)
Surgical procedures pre-Fontan completion	1.5 ± 1 (0-3)
Surgical or percutaneous interventions post-Fontan	
Electrophysiologic procedure and ablation	3 (21)
APC conversion to extracardiac conduit and epicardial pacemaker implantation	1 (7)
Epicardial pacemaker implantation	2 (14)
Fenestration post-Fontan and subsequent closure	1 (7)
Sustained arrhythmia	3 (21)
Other comorbidities	
Type I diabetes	1 (7)
Medications	
Warfarin/NOAC	7 (50)
Aspirin	7 (50)
ACEI/ARB	2 (14)
Sotalol	1 (7)
Echocardiography	
Ventricular systolic function	
Normal	12 (86)
Mild to moderate impairment	2 (14)
Atrioventricular valve regurgitation	
None-trivial	8 (57)
Mild	6 (43)

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Systemic endothelial function

Systolic function

Confidence

Body image

Cardiovascular risk

Concentration ……

Diastolic function

Mood and anxiety levels

Quality of life

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Regular Exercise
for the Fontan Circulation:

How important is it?

Regular Exercise
for the Fontan Circulation:

How?

Fontan REGISTRY

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Conclusions

- A Fontan myopathy exists that has important implications for the peripheral muscle pump and venous return
- Regular exercise to maintain peripheral muscle bulk improves cardiac preload and exercise capacity
- ? Periodically increasing pulmonary blood flow with exercise may have beneficial effects on pulmonary vascular physiology
- Frequent vigorous exercise is central to a well-functioning Fontan circulation

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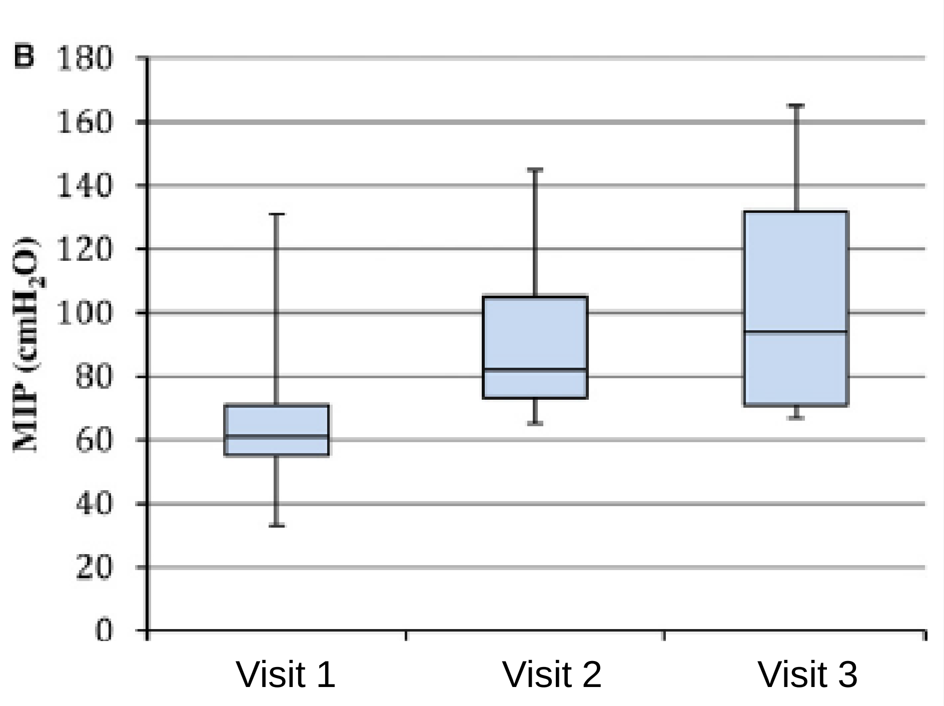
Impact of Inspiratory Muscle Training in Fontan



Laohachai..Ayer, JAHA 2017

Impact of Inspiratory Muscle Training in Fontan

Table 8. Change in Stroke Volume and Ejection Fraction Between Rest and Peak Exercise

Parameter	Rest, Mean±SD	Peak Exercise, Mean±SD	*P* Value
Pre-IMT stroke volume, mL	64.0±12.7	72.4±12.6	0.003
Pre-IMT ejection fraction, %	50.1±3.9	56.7±6.1	0.001
Post-IMT stroke volume, mL	63.7±10.4	68.2±9.0	0.03
Post-IMT ejection fraction, %	52.8±6.1	55.3±2.1	0.02

IMT indicates inspiratory muscle training.

Laohachai..Ayer, JAHA 2017

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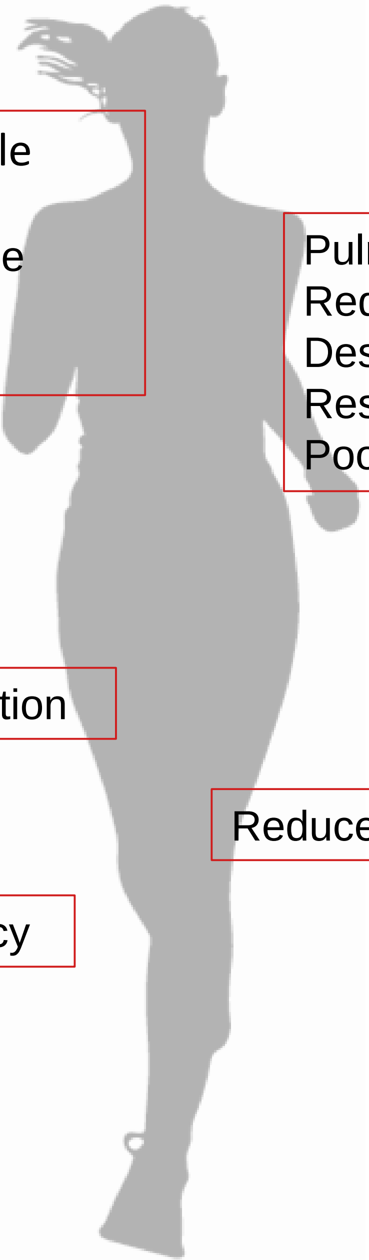
Exercise limitation in the Fontan circulation



No subpulmonary ventricle
Structural issues
Chronotropic incompetence
Diastolic dysfunction
(Systolic dysfunction)

Pulmonary endothelial dysfunction
Reduced lung volumes
Desaturation
Respiratory muscle dysfunction
Poor pulmonary vascular development

Systemic endothelial dysfunction

Reduced skeletal muscle mass and function

Iron deficiency