What to Recommend: Fontan Circulation

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Disclosures

- Consultancy fees:

  Actelion
  MSD

Disclosures

Dr Rachael Cordina

Dr Julian Ayer

Pr David Celermajer

Melbourne Children's
Excellence in clinical care, research and education

The Royal Children's Hospital Melbourne

Murdoch Childrens Research Institute

The University of Melbourne

- **Current recommendations**
- **Physiology**
- **What we know**
- **Future recommendations**

Fontan REGISTRY

AHA/ACC Scientific Statement

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 4: Congenital Heart Disease

A Scientific Statement From the American Heart Association and American College of Cardiology

George F. Van Hare, MD, FACC, Chair; Michael J. Ackerman, MD, PhD, FACC; Juli-anne K. Evangelista, DNP, APRN, CPNP-AC, FACC; Richard J. Kovacs, MD, FAHA, FACC; Robert J. Myerburg, MD, FACC; Keri M. Shafer, MD; Carole A. Warnes, MD, FACC; Reginald L. Washington, MD, FAHA; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology

Fontan REGISTRY

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)*.**

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Fontan REGISTRY

Recommendations

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Table 1. Classification of Sports (based on peak dynan

	A. Low Dynamic
I. Low Static	Billiards
	Bowling
	Cricket
	Curling
	Golf
	Riflery

ity to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms *(Class IIb; Level of Evidence C)*.

Recommendations

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Fontan REGISTRY

- Current recommendations
- Physiology
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Fontan REGISTRY

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ISSN 0735-1097/08/$34.00
doi:10.1016/j.jacc.2008.02.081

MINI-FOCUS ISSUE: THE FONTAN PROCEDURE

A Cross-Sectional Study of Exercise Performance During the First 2 Decades of Life After the Fontan Operation

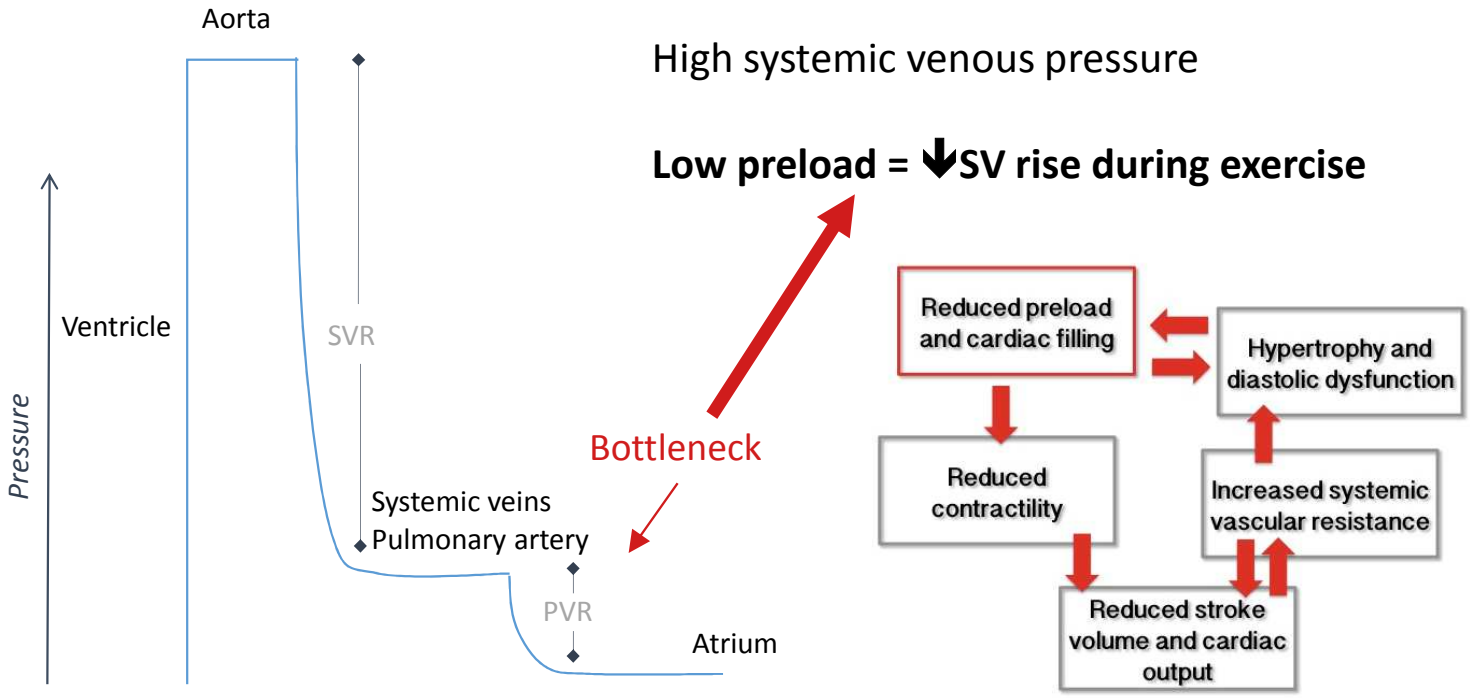
Stephen M. Paridon, MD,* Paul D. Mitchell, MS,† Steven D. Colan, MD,** Richard V. Williams, MD,‡
Andrew Blaufox, MD,§ Jennifer S. Li, MD,‖ Renee Margossian, MD,** Seema Mital, MD,¶
Jennifer Russell, MD,# Jonathan Rhodes, MD,** for the Pediatric Heart Network Investigators

Philadelphia, Pennsylvania; Watertown and Boston, Massachusetts; Salt Lake City, Utah; Charleston, South Carolina; Durham, North Carolina; New York, New York; and Toronto, Ontario, Canada

*Of 411 subjects tested (12.43.2 years of age), 166 achieved a maximal aerobic capacity. Peak oxygenconsumption (VO2) was 26.3 ml/kg/min (**65% of predicted for age and gender [% predicted]**) for the entirepopulation*

Fontan REGISTRY

The Fontan Circulation

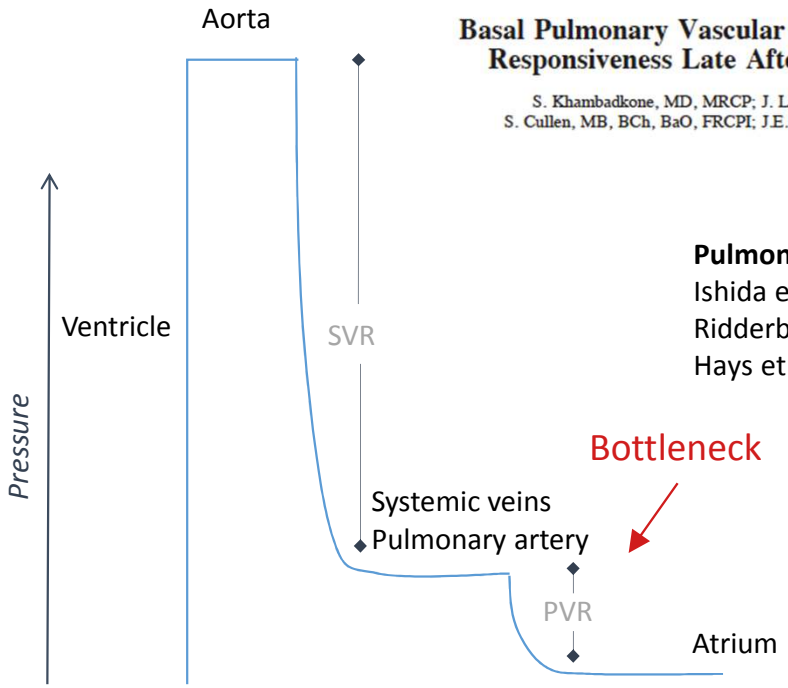


Systemic vascular resistance=SVR
Pulmonary vascular resistance=PVR

Courtesy of Marc Gewillig

Fontan REGISTRY

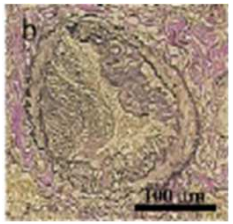
The Fontan Circulation



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
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Fontan REGISTRY

A Fontan Myopathy?

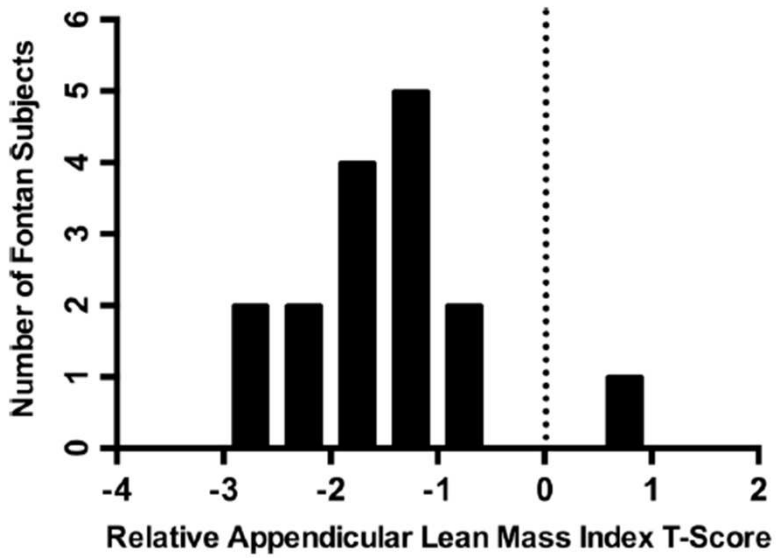


Figure 1 A T-score represents the number of SDs from the young normal reference mean. A value <−2.0 represents marked muscle wasting, defined as in the sarcopenic range.

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

Skeletal muscle abnormalities and exercise capacity in adults with a Fontan circulation

Rachael Cordina,[1,2] Shamus O'Meagher,[1,2] Haslinda Gould,[3,4] Caroline Rae,[5] Graham Kemp,[6] Julie A Pasco,[3,4] David S Celermajer[1,2]

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]

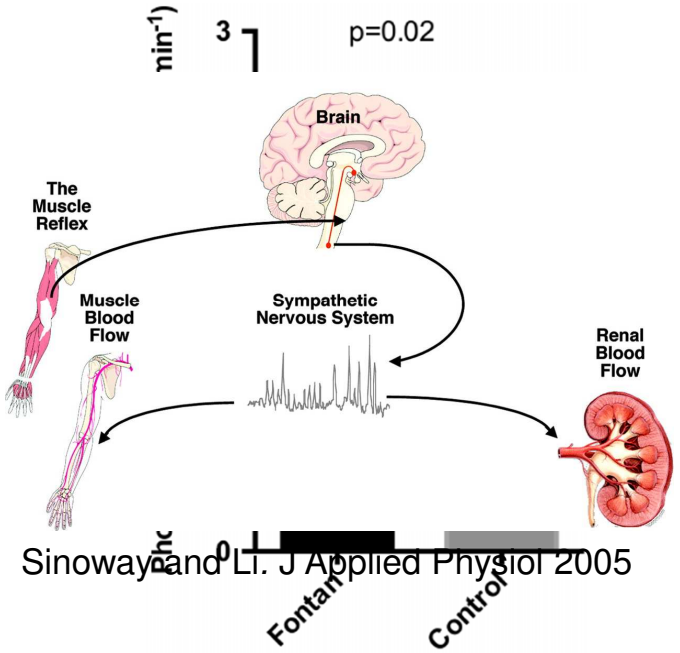
Heart 2014

Fontan REGISTRY

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

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

Cordina et al. Heart 2013

Fontan REGISTRY

International Journal of Cardiology 167 (2013) 1333–1338

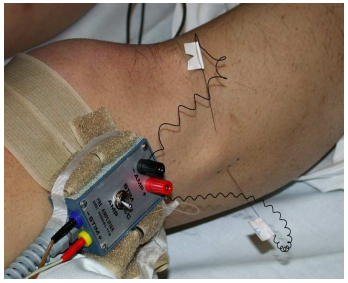
Contents lists available at SciVerse ScienceDirect

International Journal of Cardiology

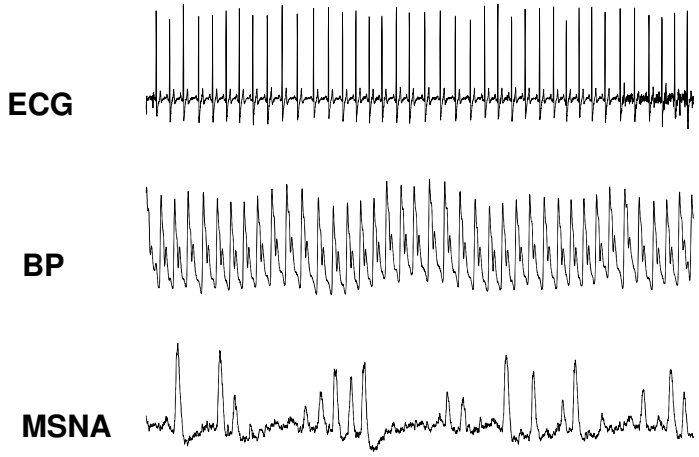
journal homepage: www.elsevier.com/locate/ijcard

Sympathetic and vascular dysfunction in adult patients with Fontan circulation

Elisabeth Lambert [a,e,*,1], Yves d'Udekem [f,g,1], Michael Cheung [g,h], Carolina Ika Sari [a], Julia Inman [a], Anna Ahimastos [c], Nina Eikelis [a], Atul Pathak [j], Ingrid King [f,g], Leanne Grigg [i], Markus Schlaich [b,d], Gavin Lambert [a,d]



Clinical microneurography



Fontan REGISTRY

- Current recommendations
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Fontan REGISTRY

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?

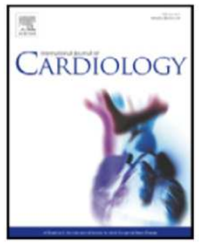
Fontan REGISTRY

International Journal of Cardiology 168 (2013) 780–788

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard



Resistance training improves cardiac output, exercise capacity and tolerance to positive airway pressure in Fontan physiology ☆

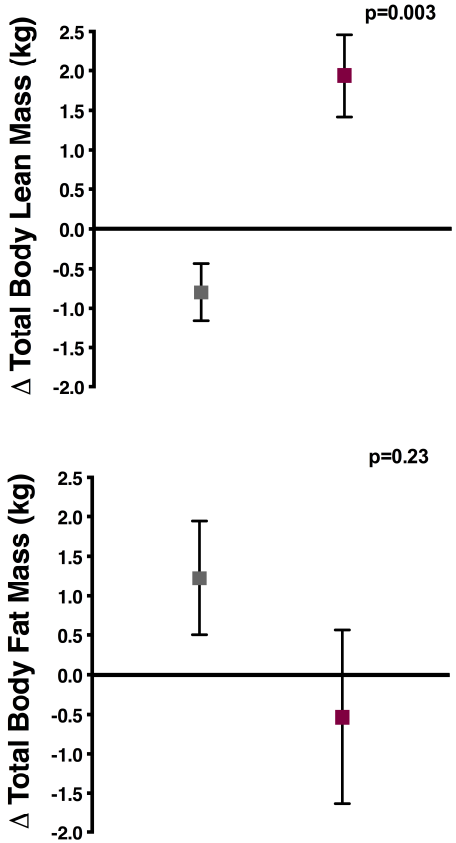
Rachael L. Cordina [a,b], Shamus O'Meagher [a,b], Alia Karmali [a], Caroline L. Rae [c,d], Carsten Liess [e], Graham J. Kemp [f], Raj Puranik [a,b], Nalin Singh [g,h], David S. Celermajer [a,b,*]

Fontan REGISTRY

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

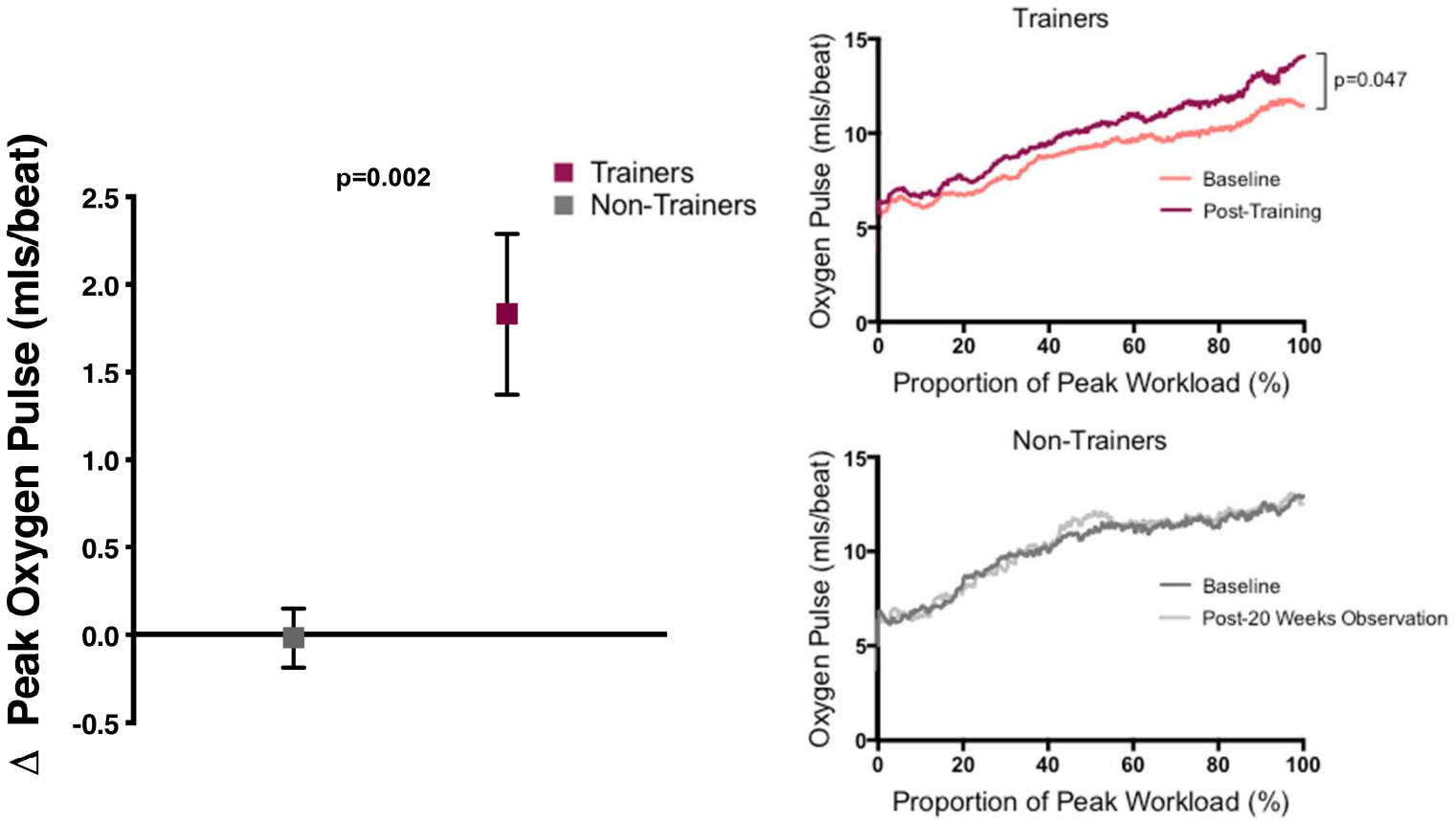
Strength increased by 43 ± 7 %
(p=0.002)



Cordina...Celermajer, International Journal of Cardiology 2013

Fontan REGISTRY

Peak VO_2 increased by 10% (p=0.03)

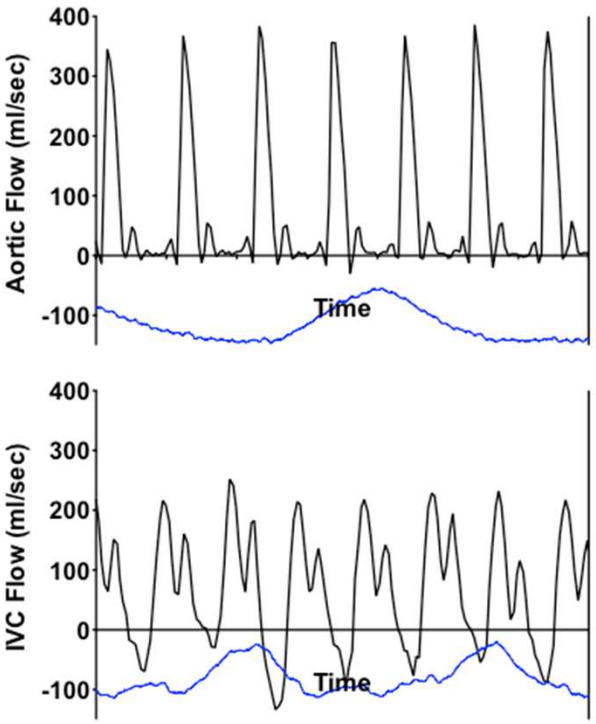


Cordina et al. International Journal of Cardiology 2013

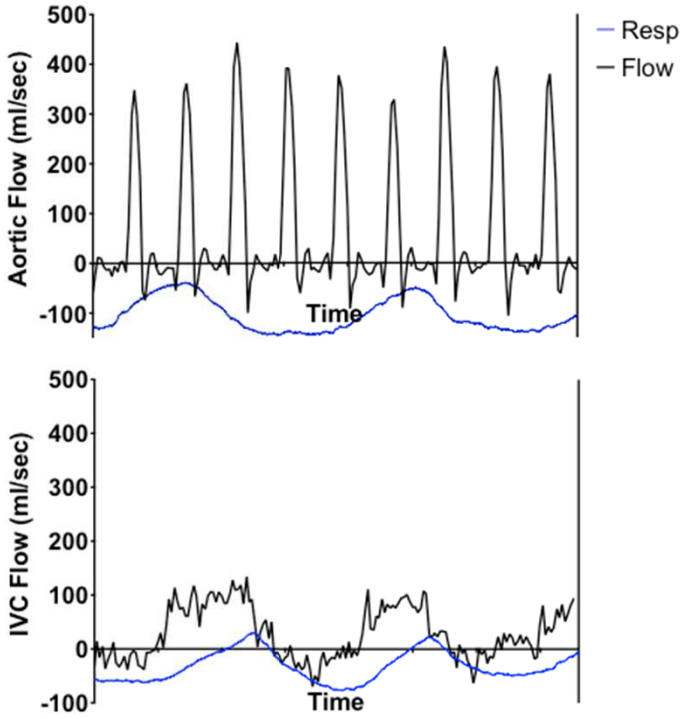
Fontan REGISTRY

Respiratory Dependence in Fontan

Normal Circulation



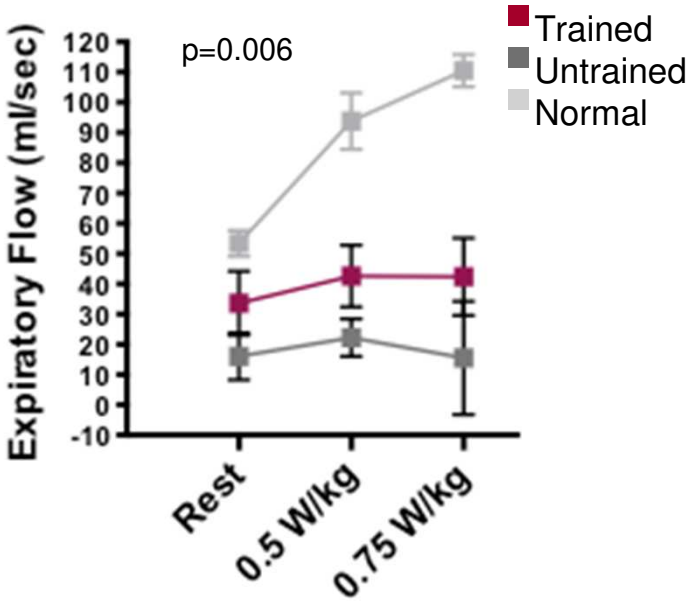
Fontan Circulation

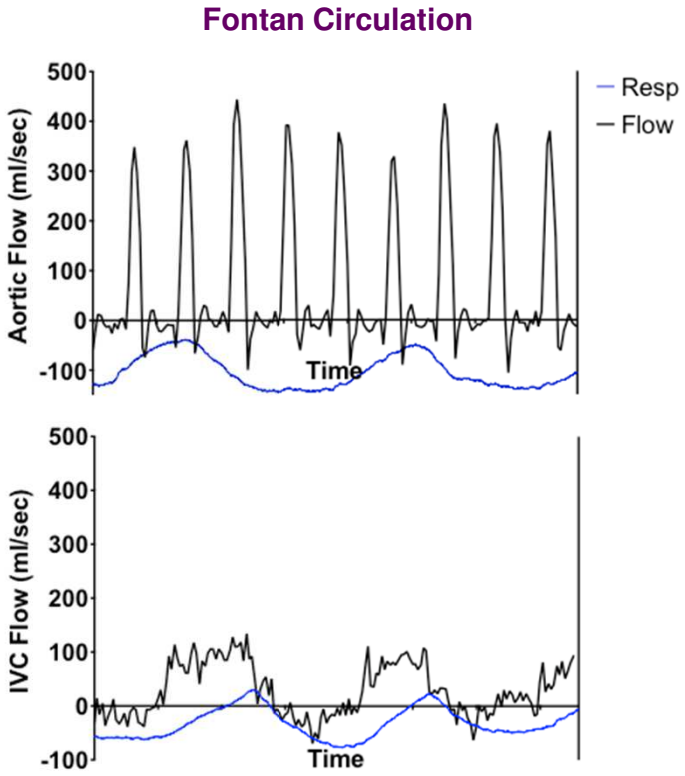


Cordina et al. International Journal of Cardiology 2013

Fontan REGISTRY

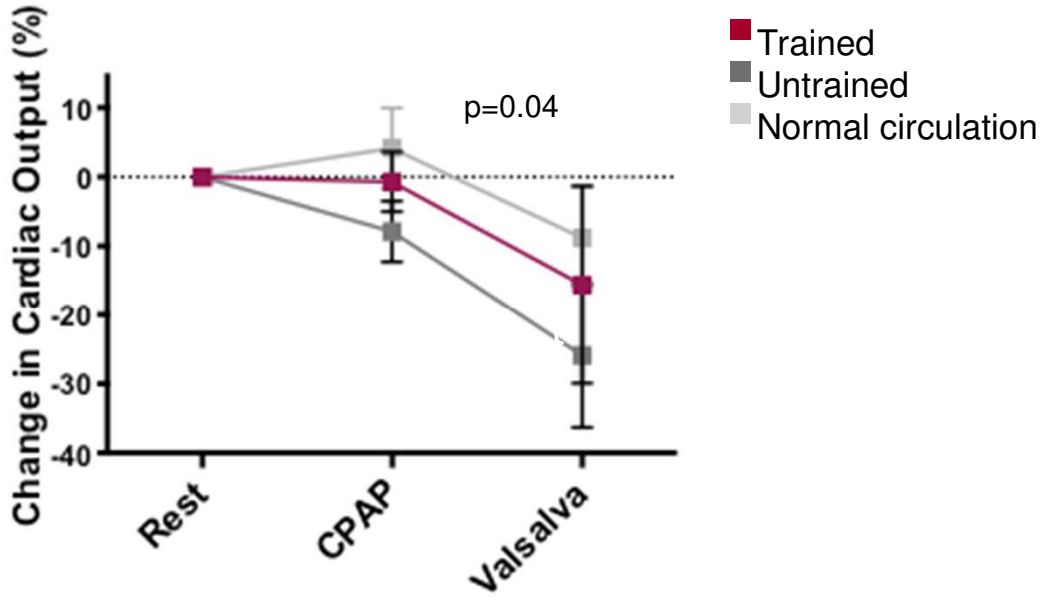
Respiratory Dependence in Fontan





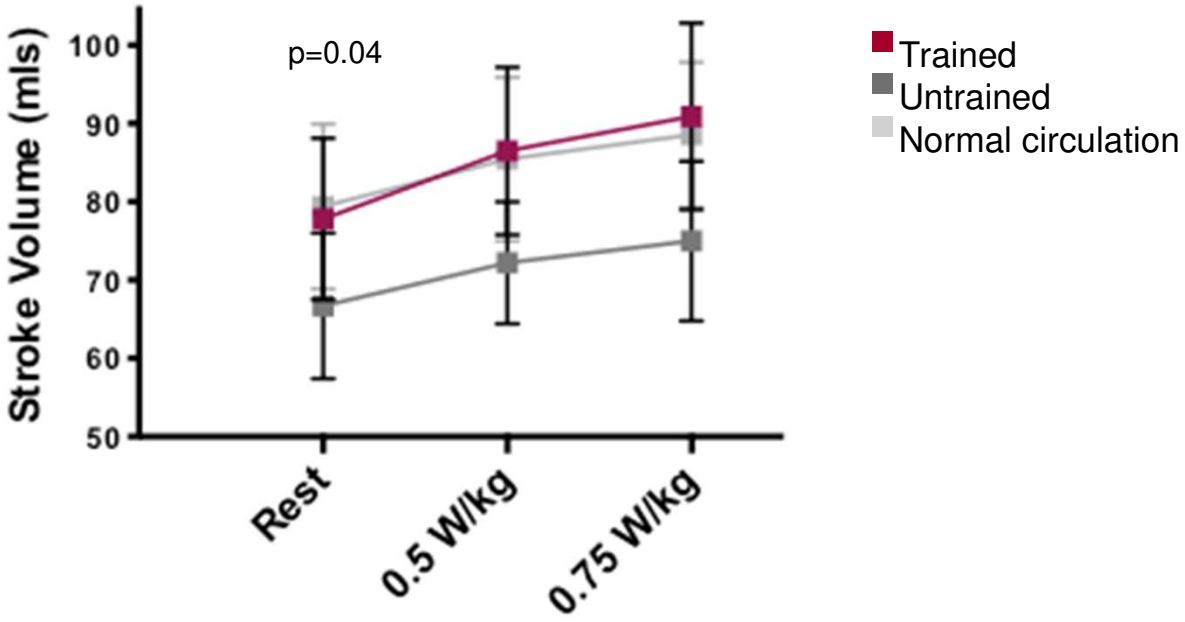
Cordina et al. International Journal of Cardiology 2013

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

Impact of Inspiratory Muscle Training in Fontan

ORIGINAL RESEARCH

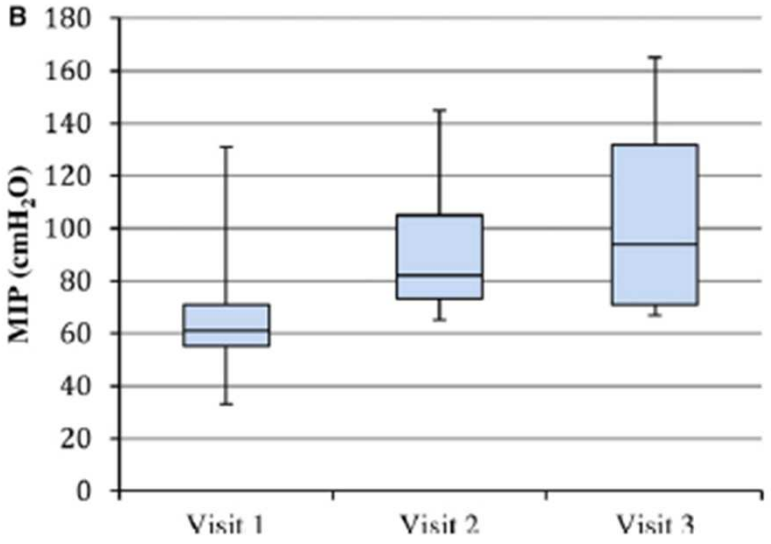
American Heart Association | American Stroke Association

Inspiratory Muscle Training Is Associated With Improved Inspiratory Muscle Strength, Resting Cardiac Output, and the Ventilatory Efficiency of Exercise in Patients With a Fontan Circulation

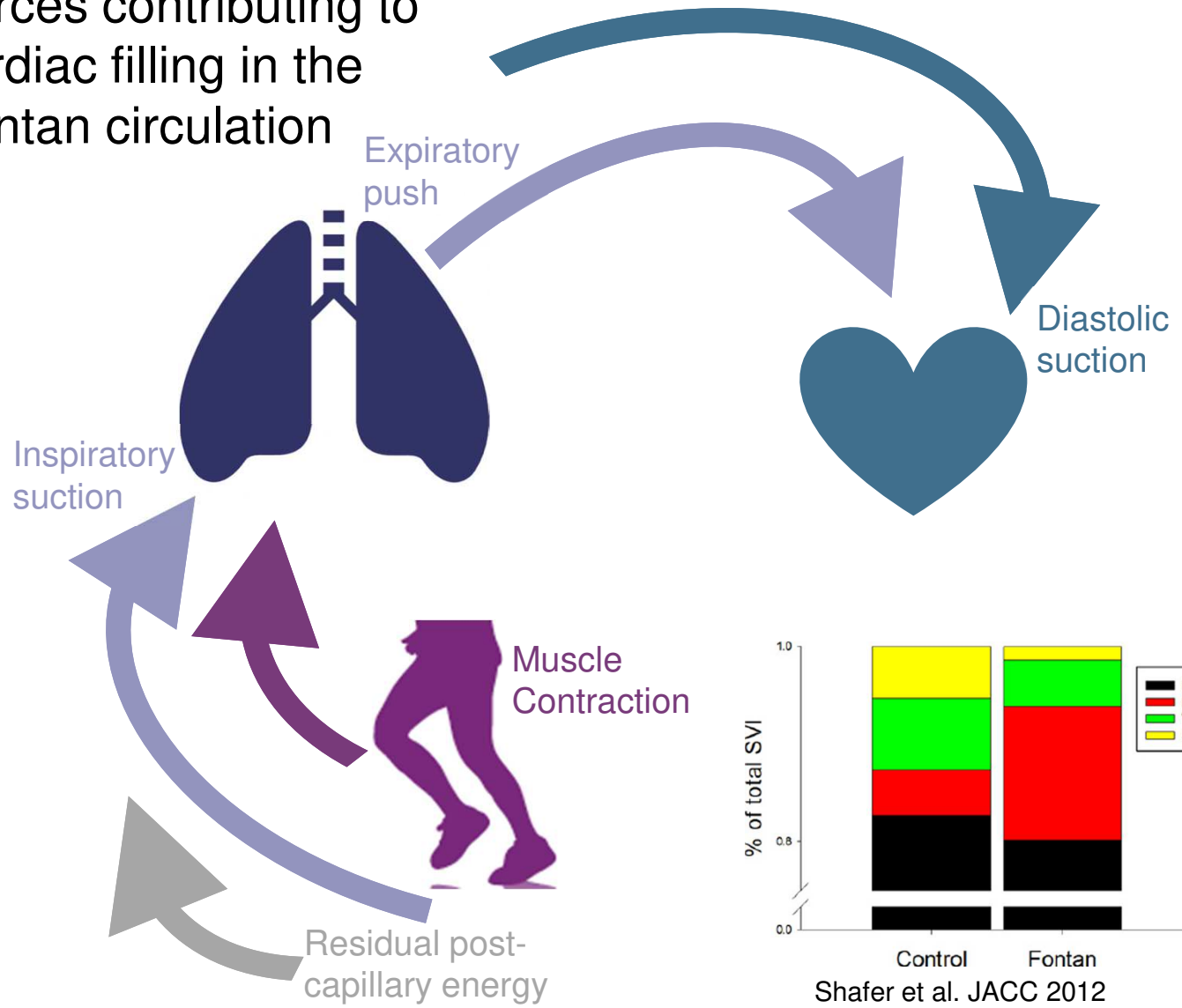
Karina Laohachai, MBBS, FRACP; David Winlaw, MBBS(Hons), MD, FRACS; Hiran Selvadurai, MBBS, FRACP, PhD; Ganesh Kumar Gnanappa, MBBS, MD; Yves d'Udekem, MD, PhD, FRACS; David Celermajer, MBBS, MSc, FRACP, PhD, DSc; Julian Ayer, BSc(Med), MBBS, FRACP, PhD

(*J Am Heart Assoc*. 2017;6:e005750. DOI: 10.1161/JAHA.117.005750.)

Laohachai..Ayer, JAHA 2017



Forces contributing to cardiac filling in the Fontan circulation



Shafer et al. JACC 2012

The Journal of Thoracic and Cardiovascular Surgery

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 Tran BAppSc(ExPhys) [a], Yves d'Udekem MD PhD FRACS [c, d, e]

133 pts who had a recent CPET study

14 (11%) had VO_2 max > 80% of predicted value

All of them participating in moderate to vigorous sporting activities at least 3 times a week.

All employed or studying full time for graduate or post-graduate education

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)

Fontan REGISTRY

Cardiology in the Young

cambridge.org/cty

Images in Congenital Cardiac Disease

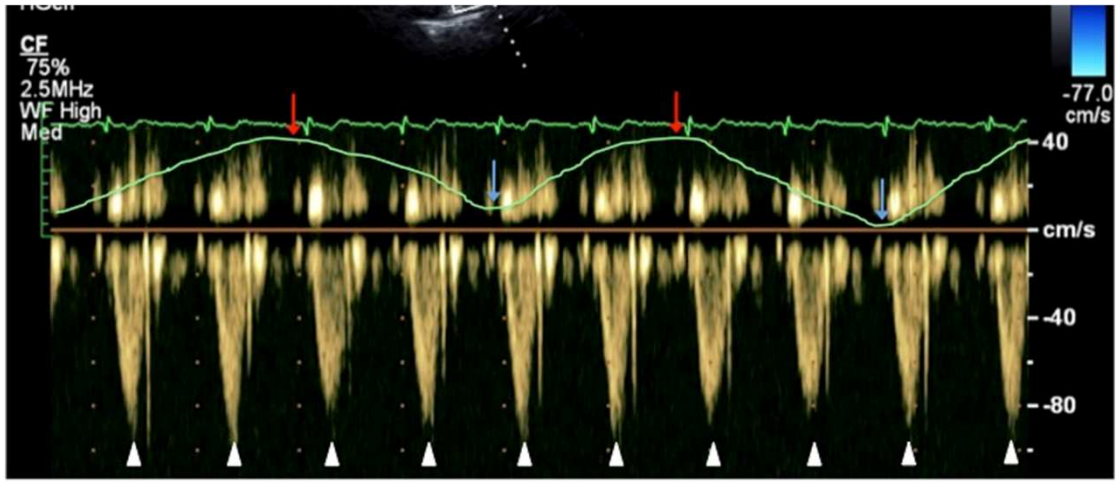
Lower limb exercise generates pulsatile flow into the pulmonary vascular bed in the setting of the Fontan circulation

Rachael Cordina[1,2], David S. Celermajer[1,2] and Yves d'Udekem[3,4,5]

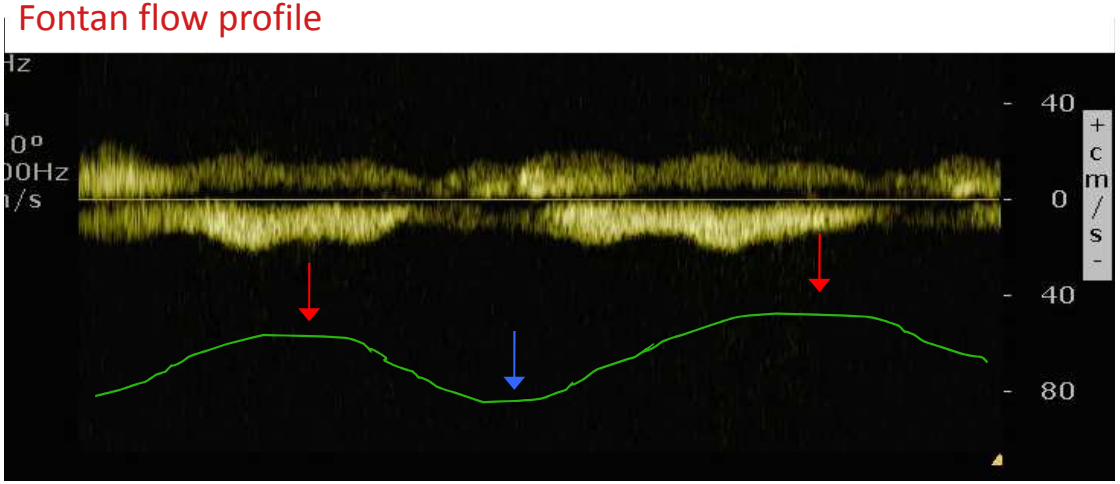
[1]Sydney Medical School, University of Sydney, Sydney, NSW, Australia, [2]Department of Cardiology, Royal Prince Alfred Hospital, Sydney, NSW, Australia, [3]Heart Research Group, Murdoch Children's Research Institute,

Fontan REGISTRY

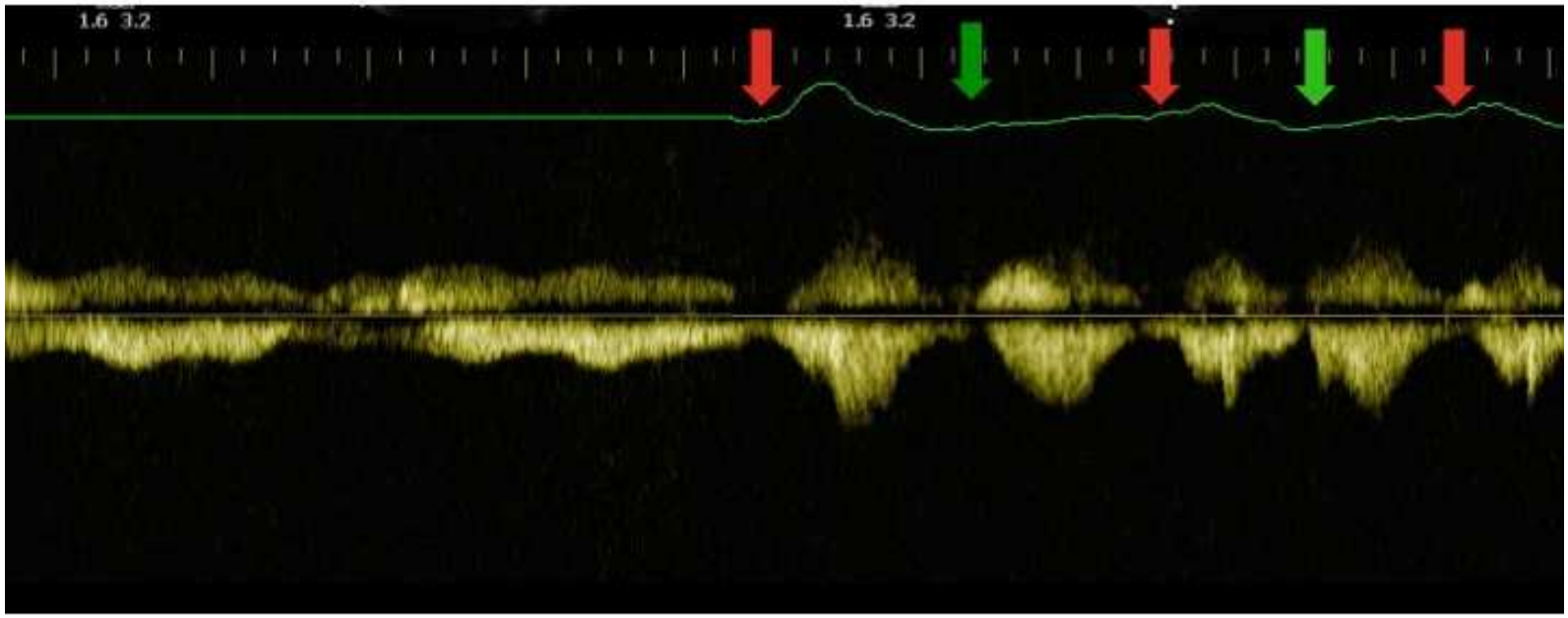
Normal flow profile in pulmonary artery



Fontan flow profile



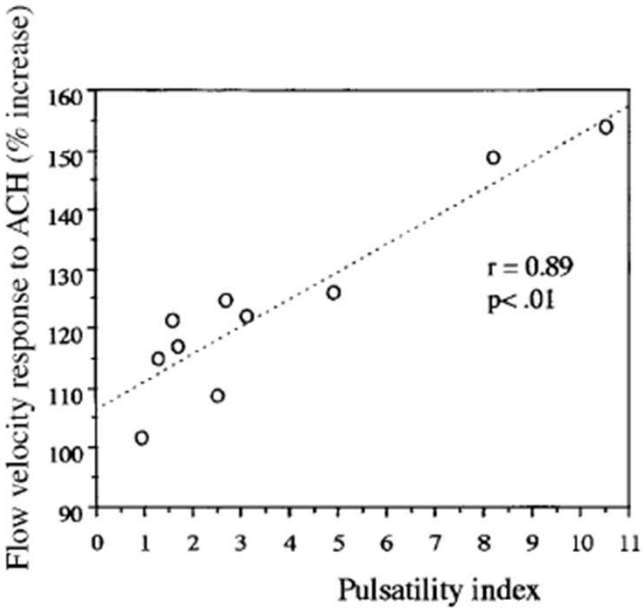
Fontan REGISTRY



Fontan
REGISTRY

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.

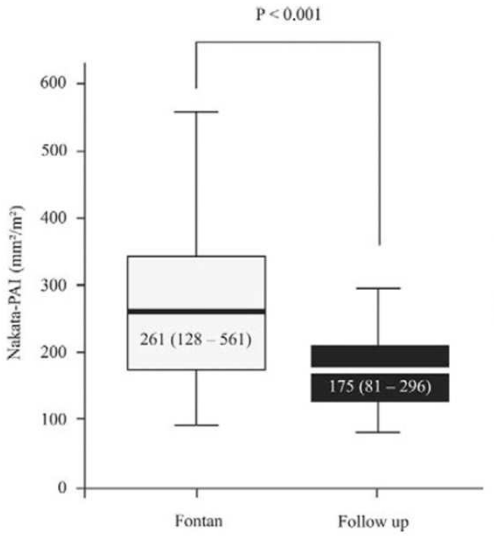


Kurotobi et al. JTCVS 2001

Fontan REGISTRY

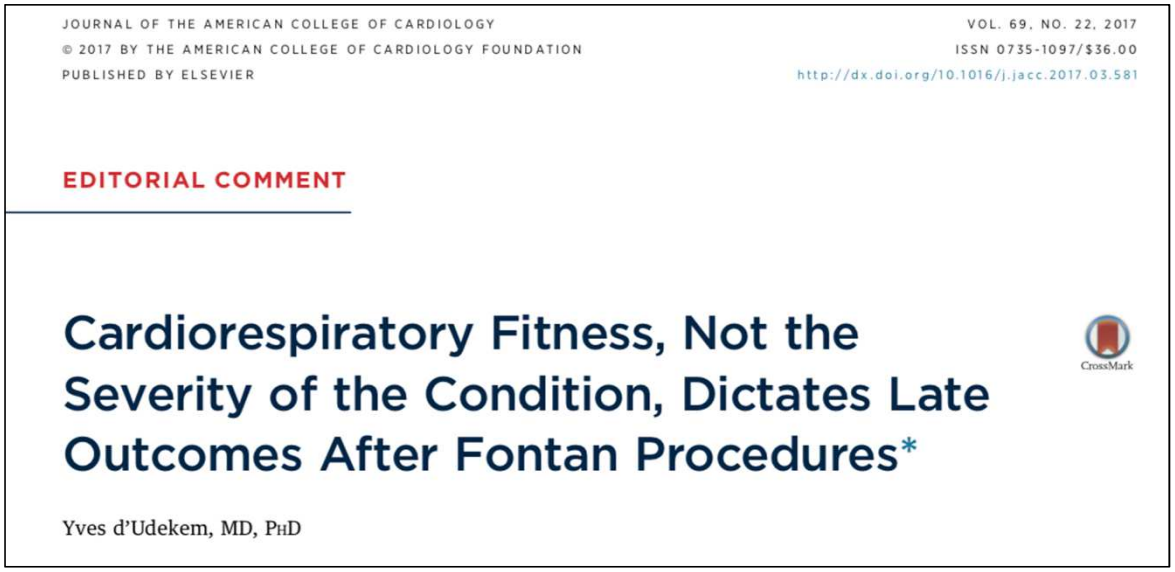
Fontan - Pulmonary Artery Growth

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



Ouvroutski et al. Annals of Thoracic Surg 2009

Fontan REGISTRY



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EDITORIAL COMMENT

Cardiorespiratory Fitness, Not the Severity of the Condition, Dictates Late Outcomes After Fontan Procedures*

Yves d'Udekem, MD, PhD

Lower physical activity score at first testing was predictive of death and transplantation !!!

In the meantime, the only advice that I will keep giving to the young patients who are coming to see me after Fontan procedures is "Exercise, exercise, exercise!"

Fontan REGISTRY

Systemic endothelial function

Systolic function

Confidence

Body image

Cardiovascular risk

Diastolic function

Mood and anxiety levels

Quality of life

Fontan REGISTRY

Increased Body Mass Index Is Associated with Congestive Heart Failure and Mortality in Adult Fontan Patients

Sara C. Martinez, MD, PhD,* Mirnela Byku, MD, PhD,* Eric L. Novak, MS,* Ari M. Cedars, MD,* Pirooz Eghtesady, MD, PhD,† Philip A. Ludbrook, MD,* and Joseph J. Billadello, MD*

*Cardiovascular Division, Department of Internal Medicine, Washington University School of Medicine in St. Louis, St. Louis, Mo, and †Section of Pediatric Cardiothoracic Surgery, Department of Surgery, Washington University School of Medicine in St. Louis, St. Louis, Mo, USA

Congenit Heart Dis. 2016;11:71–79

Fontan REGISTRY

Heart, Lung and Circulation (2015) **24,** 753–768
1443-9506/04/$36.00
http://dx.doi.org/10.1016/j.hlc.2015.03.005

REVIEW

Should We Recommend Exercise after the Fontan Procedure?

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Conclusions Exercise training is safe and beneficial in patients with a Fontan circulation. Exercise training should become a standard of care within this population. Physiological adaptation following exercise training needs to be investigated more extensively.

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ORIGINAL ARTICLE

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CONGENITAL

Long-term outcomes of single-ventricle palliation for pulmonary atresia with intact ventricular septum: Fontan survivors remain at risk of late myocardial ischaemia and death†

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Fontan REGISTRY

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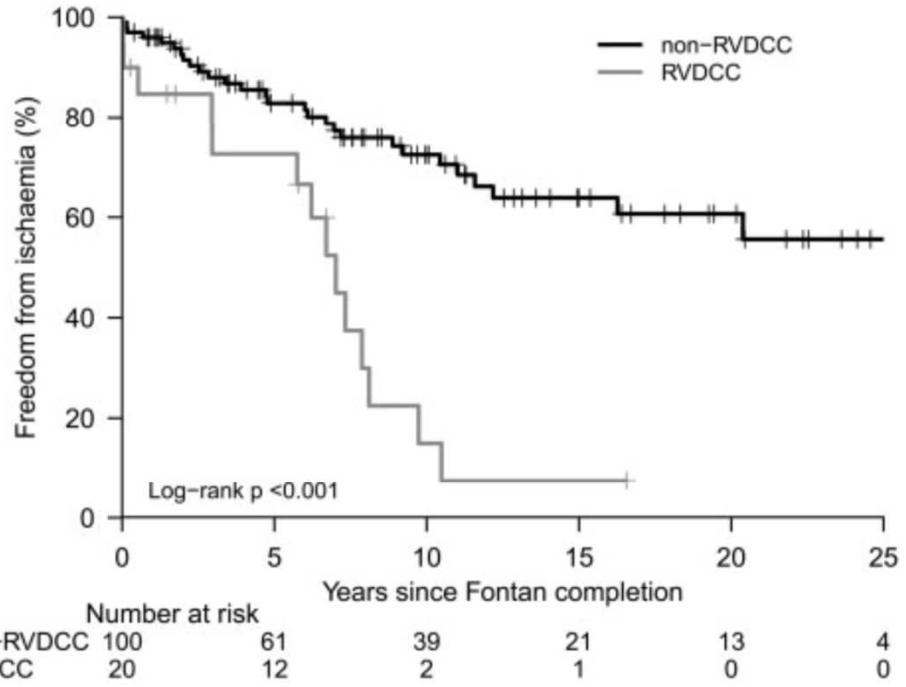

Figure 3: Kaplan–Meier survival curves of freedom from new onset myocardial ischaemia following Fontan completion. Log-rank test, *P* <0.001. RVDCC: right ventricle-dependent coronary circulation.

Regular Exercise
for the Fontan Circulation:

How?

Fontan
REGISTRY

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

Fontan REGISTRY

- Current recommendations
- Physiology
- What we know
- Future recommendations

Fontan REGISTRY

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)*.**

Fontan REGISTRY


JUST DO IT.