PROGRESS
FROM A CLINICIANS PERSPECTIVE

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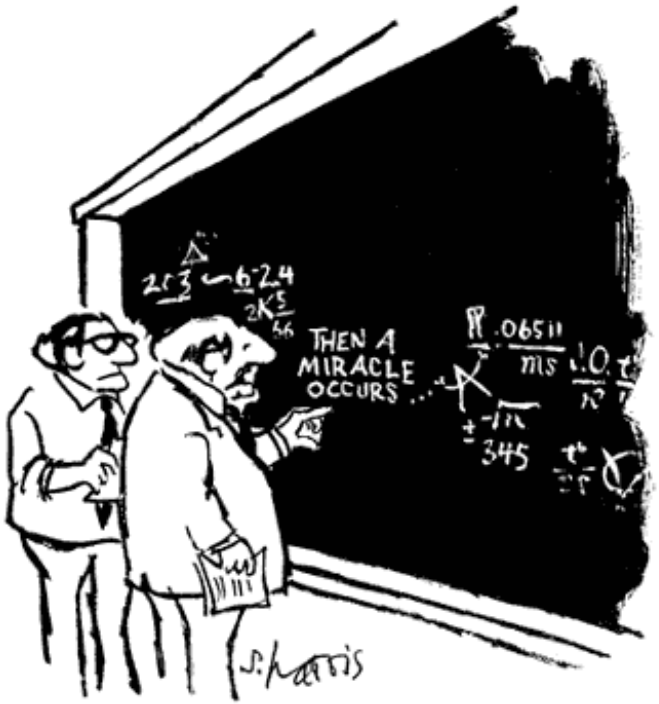
What's changed?

How we train and are organized

The data and guidelines

When I started

You are what you say you are



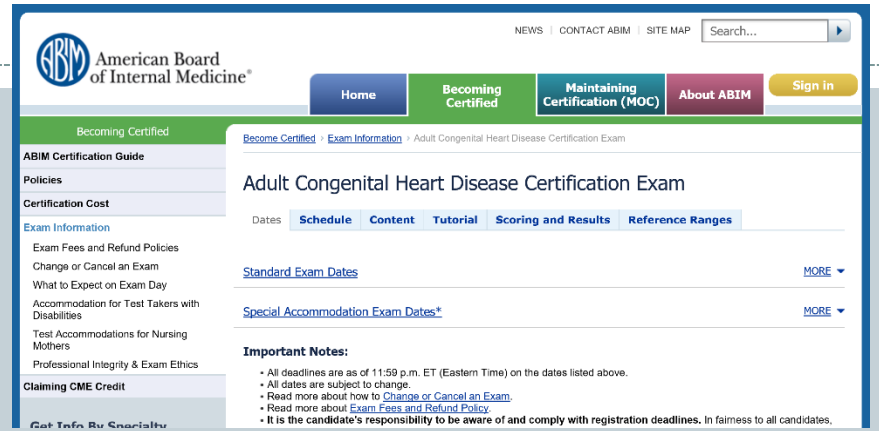
"I think you should be more explicit here in step two."

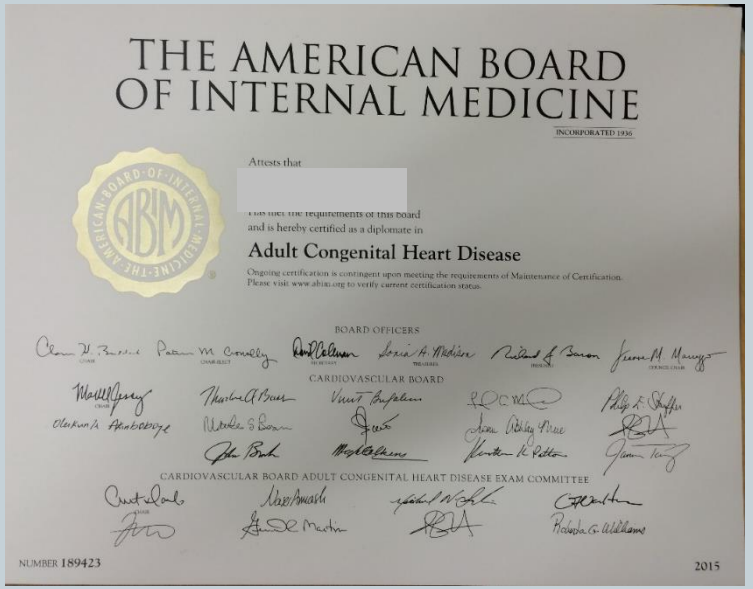
I am an ACHD cardiologist

Accreditation Council for Graduate Medical Education

ACGME

ACGME Program Requirements for Graduate Medical Education in Adult Congenital Heart Disease (Internal Medicine)






VOTED BEST
BURGER
IN TOWN BY
SOME GUY
WHO LIKES
BURGERS

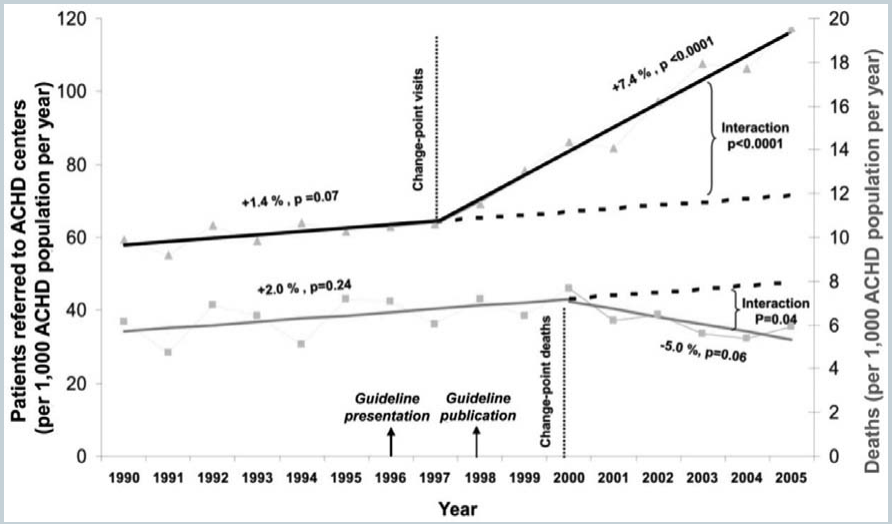
vs.

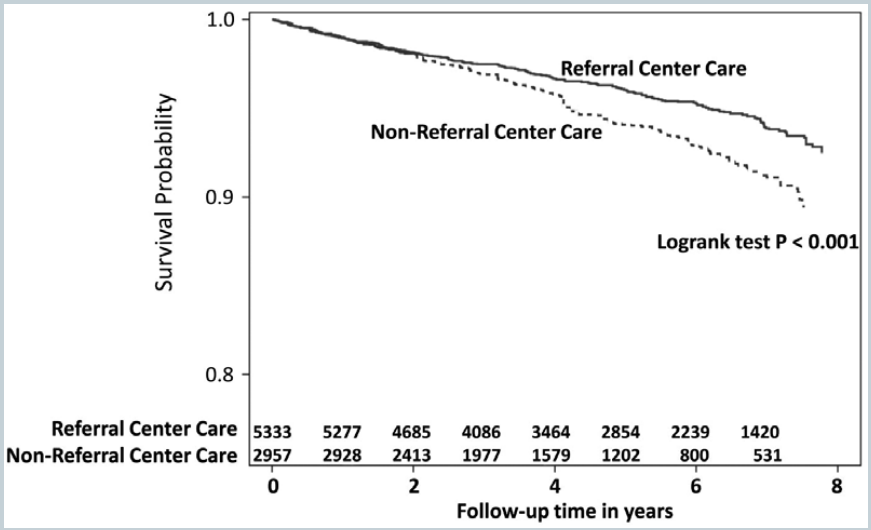

MICHELIN STARS

Impact of ACHD Referral on Mortality

71,467 patients in Quebec

Referral patterns and mortality before and after GL publication





Mylotte *Circulation* 2014


Adult
Congenital
Heart
Association
ACHD ACCREDITED
CARE CENTER


Adult
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COMPREHENSIVE CARE CENTER

Life's hard. It's harder when you're stupid.

~ *John Wayne*

1982-1999

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2001 Nov 15 [updated 2016 Feb 25].
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Int J Cardiol. 2017 Jul 15;239:20. doi: 10.1016/j.ijcard.2017.02.133. No abstract available.
PMID: 28560967

2. Challenge of Timing Redo Aortic Valve Replacement: Is There a Potential Role for Left Ventricular Global Longitudinal Strain?
Baumgartner H.
Circ Cardiovasc Imaging. 2017 Jun;10(6). pii: e006556. doi: 10.1161/CIRCIMAGING.117.006556. No abstract available.
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ACC/AHA Guideline

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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2017 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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Anatomic complexity in ACHD

Table 3. Types of Adult Congenital Heart Disease of Great Complexity*

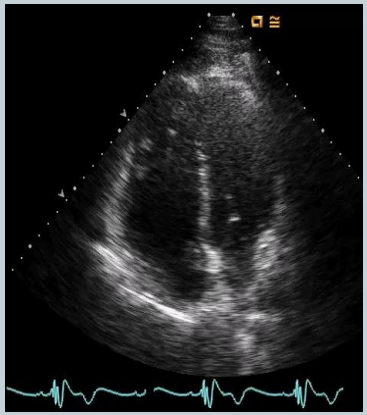
Conduits, valved or nonvalved
Cyanotic congenital heart (all forms)
Double-outlet ventricle
Eisenmenger syndrome
Fontan procedure
Mitral atresia
Single ventricle (also called double inlet or outlet, common, or primitive)
Pulmonary atresia (all forms)
Pulmonary vascular obstructive disease
Transposition of the great arteries
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of atrioventricular or ventriculoarterial connection not included above (ie, crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Table 4. Diagnoses in Adult Patients With Congenital Heart Disease of Moderate Complexity*

Aorto–left ventricular fistulas
Anomalous pulmonary venous drainage, partial or total
Atrioventricular septal defects (partial or complete)
Coarctation of the aorta
Ebstein's anomaly
Infundibular right ventricular outflow obstruction of significance
Ostium primum atrial septal defect
Patent ductus arteriosus (not closed)
Pulmonary valve regurgitation (moderate to severe)
Pulmonary valve stenosis (moderate to severe)
Sinus of Valsalva fistula/aneurysm
Sinus venosus atrial septal defect
Subvalvular AS or SupraAS (except HOCM)
Tetralogy of Fallot
Ventricular septal defect with:
Absent valve or valves
Aortic regurgitation
Coarctation of the aorta
Mitral disease
Right ventricular outflow tract obstruction
Straddling tricuspid/mitral valve
Subaortic stenosis

The adult with repaired tetralogy of fallot

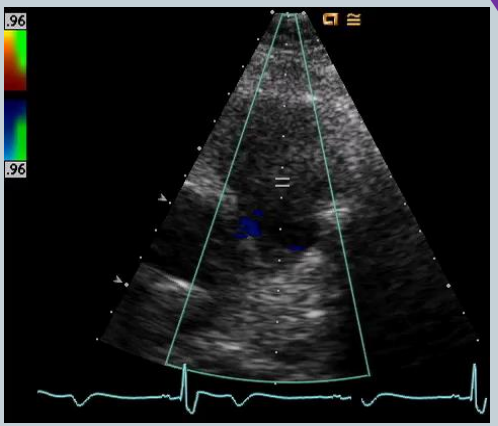
Spectrum of severity



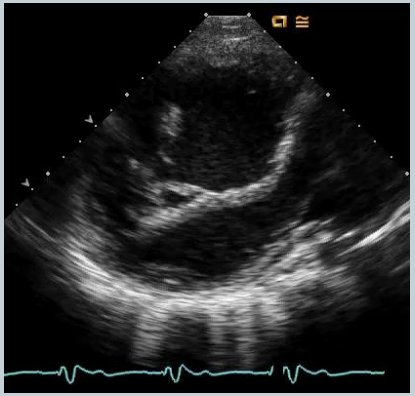
PRETTY GREAT

22 year old asymptomatic, normal right ventricular function, no significant PR

FINE



40 year old with objective exercise intolerance, significant PR, PS, RVE and RV dysfunction



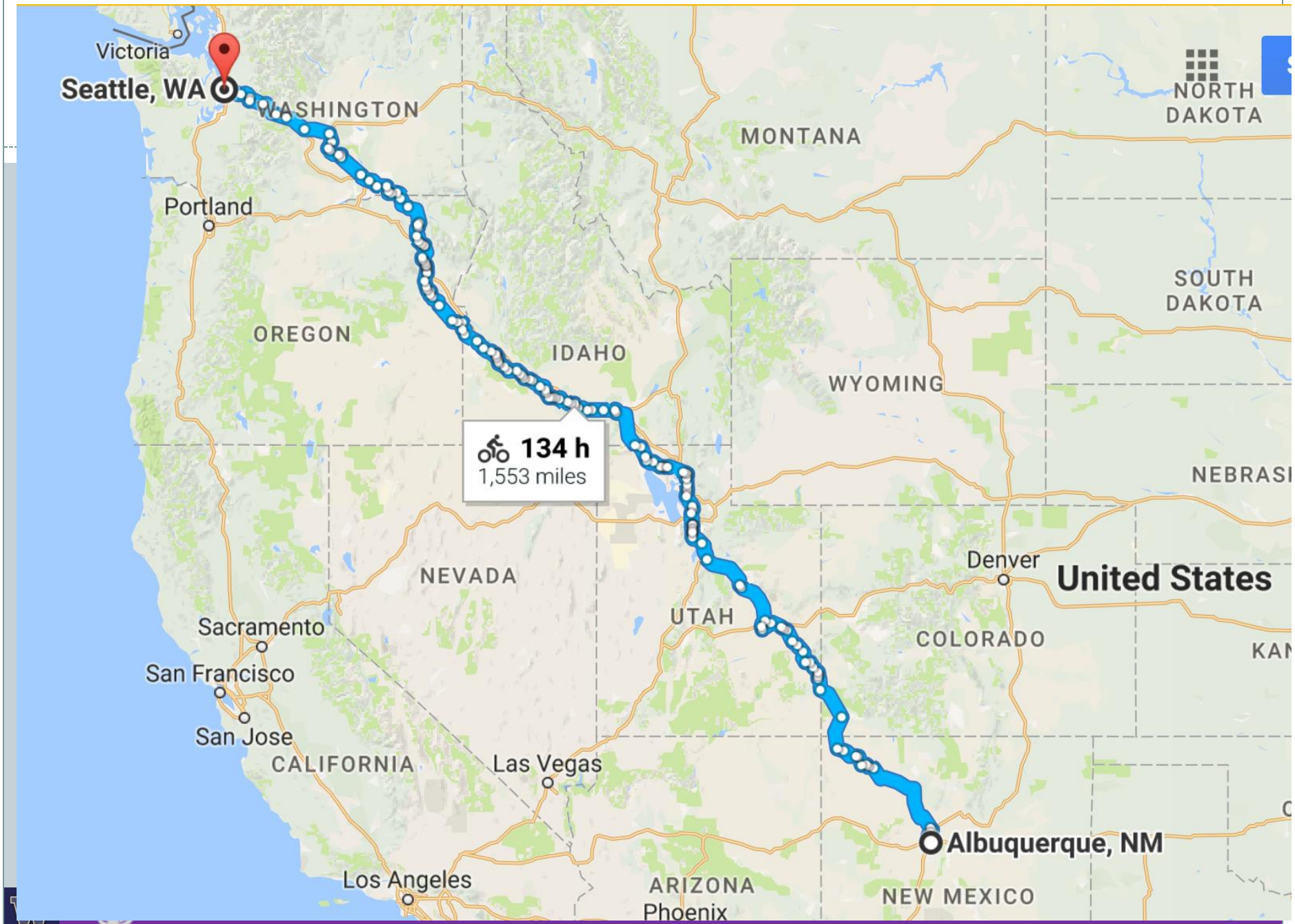
NOT AS GREAT

20 year old NYHA class III with pulmonary atresia and branch pulmonary stenosis

An ACHD patient

Subjectively normal exercise capacity

Rode his bike from home to his annual ACHD clinic evaluations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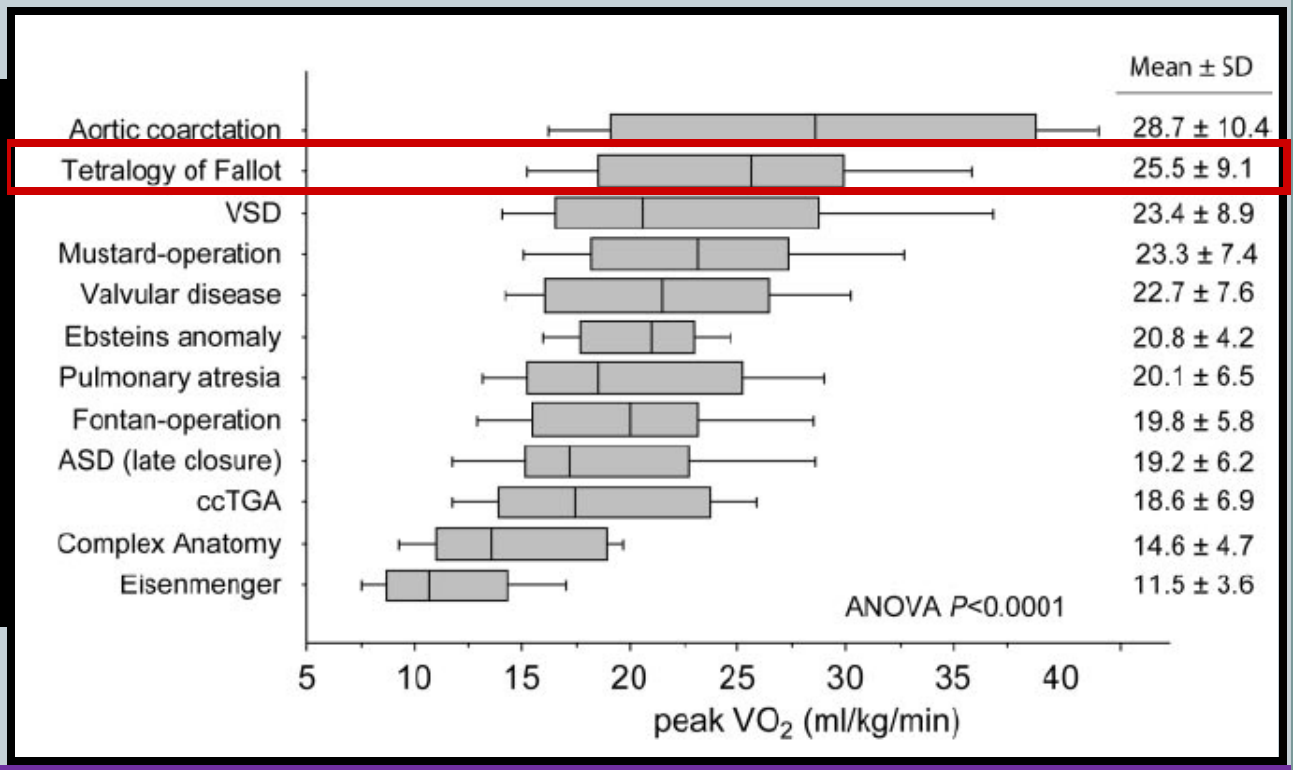
335 consecutive ACHD patients
mean age 33

Exercise capacity predicts morbidity and mortality

Diminished exercise capacity despite lack of symptoms



Diller, et al *Circulation* 2005

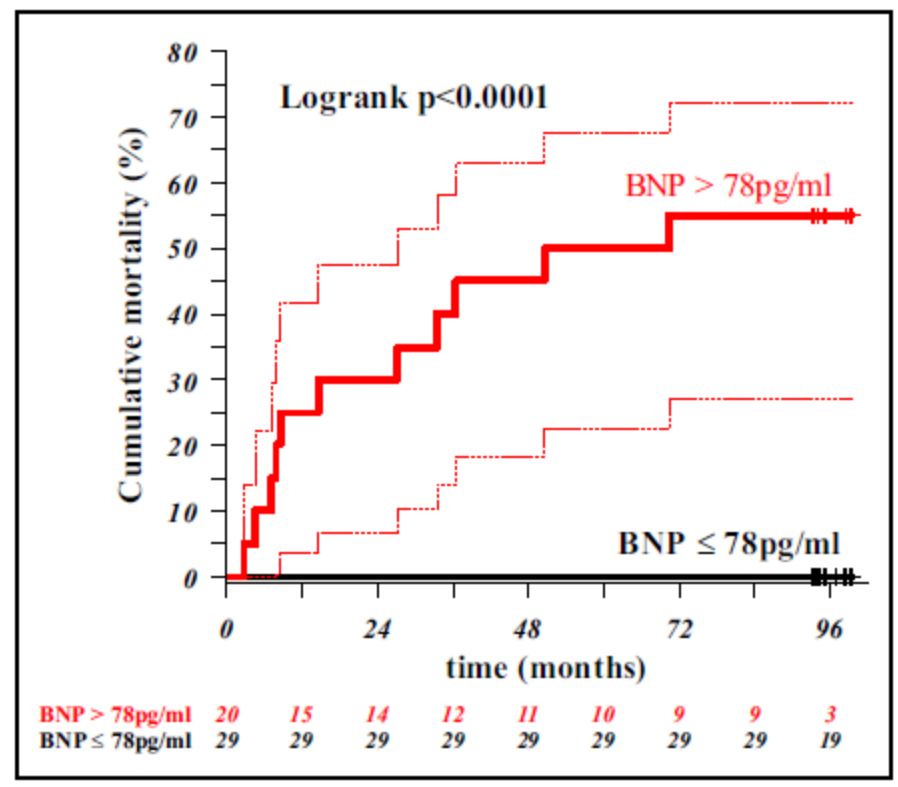
BNP as a prognostic marker in ACHD

49 consecutive ACHD patients from clinic

>1/2 were NYHA class II

Median f/u ~8 years

36% TOF, 30% single ventricle, 15% systemic RV



Giannakoulas Amer J Cardiol 2010

PVR after operative repair of TOF
Meta-analysis of 3118 patients from 48 studies

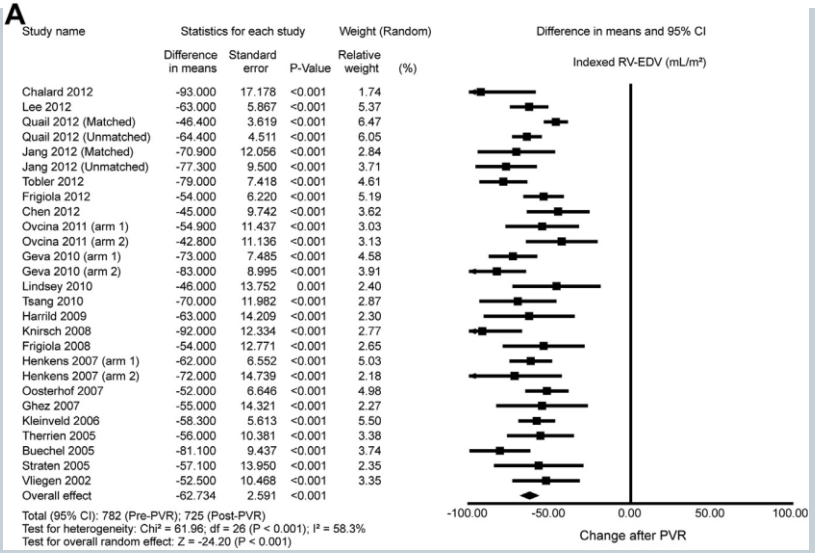
PVR improves

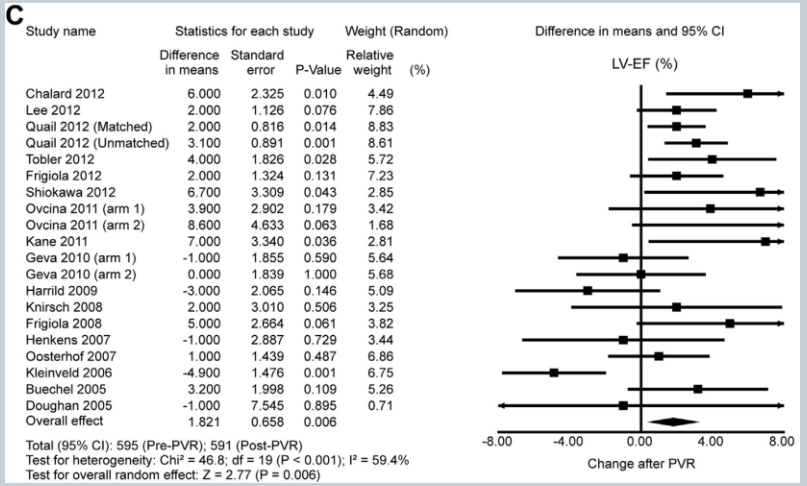
- **PR**
- **RVEDV**
- **RVESV**
- **NYHA class**

Less impressive

- **LVEDV**
- **LVESV**
- **LVEF**

Ferraz *J Am Coll Card* 2014





High quality data ⟷ Limited data/expert opinion

What our field has learned from our patients

A pessimist is one who makes difficulties of his opportunities and an optimist is one who makes opportunities of his difficulties.

~Harry Truman