My Heart is not Alone: Lung Disease in the Adult with Congenital Heart Disease

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Physical Activity
Gas Exchange
Blood Oxygenation
Oxygen Transport to Muscle

Shortness of breath
Shortness of Breath

- Most common complaint in ACHD patients
- Exercise intolerance is common in ACHD pts
- Exercise capacity measured by peak VO2
  - Predictive of hospitalization
  - Predictive of mortality
- Often attributed to cardiac etiology
  - ↓ HR response (HR reserve and chronotoricopic index)
  - Cardiac dysfunction (systolic and diastolic)

Fredrickson et al Am J Card 2001
Inuzuka et al Circulation 2012
Diller et al Circulation 2005
Restrictive Lung Disease

- Powerful predictor ↓ exercise capacity in ACHD

Ginde et al 2013

- 44% ACHD pts had spirometry suggestive of RLD
- Markedly ↑ compared to general population of 9.2%
- Diagnosis most common with restrictive lung defect
  - TOF, Fontan, and TGA S/P atrial switch
- Independent predictor for exercise capacity

Alonso-Gonzalez et al 2013

- 47% of pts had restrictive lung disease

Presence of Moderate Restrictive Lung Disease Predictive of Mortality in ACHD

N=1188 pts
Follow-up 6.7 yrs

1.6 ↑ mortality

Alonso-Gonzales et al Circulation 2013
Restrictive Lung Disease and Freedom from Hospitalization

Log Rank p<0.001

Cohen et al 2014 ACHA Scientific Session
Etiology of Restrictive Lung Disease

- Group of conditions characterized by ↓ lung volume
- Spirometry good screen but confirmed by lung volumes

**Major causes**

- **Extrinsic**
  - Disease of pleura, chest wall, neuromuscular apparatus
- **Intrinsic**
  - Alteration lung parenchyma

Cohen et al Congenital Heart Disease 2013
Extrinsic Causes

- **Diaphragmatic weakness**
  - Estimated prevalence 10%

- **Restrictive thoracic cage**
  - Multiple surgeries
  - Previous thoracotomies

- **Scoliosis**
  - 16% of pts mod-severe scoliosis
  - 5 fold ↑risk for RLD

- **Respiratory muscle weakness**

Cohen et al Congenital Heart Disease 2013
Obesity in Adults with CHD

- 1 in 3 adults in world are overweight or obese
  - ↑ 23% since 1980

- Prevalence in ACHD population even higher

- 40-54% of ACHD are overweight or obese

Pemberton, *et al* Circulation 2010
Stevens *et al* Population Health Metrics 2012
Intrinsic Causes

- CHD with ↓ PBF leads to ↓ growth and development of lung parenchyma leading to hypoplasia

- Cardiopulmonary bypass at young age induces inflammatory response
  - Damage to alveoli
  - Lung growth

- Parenchymal lung disease
  - Toxicity from amiodarone

Relationship of Restrictive Lung Disease to Exercise Intolerance

♦ Combination:
  – ↓ Ventilatory capacity and ↑ ventilatory demand

Normal Exercise
♦ As CO2 production ↑
  Minute ventilation ↑
  (TV X RR)
♦ Constant relationship
  (VE/VCO2 slope)

Restrictive Lung Disease
♦ Ability to ↑ minute ventilation dependent on RR
♦ Abnormal ventilatory response
  • For any given in ↑ CO2 exaggerated ↑ in minute ventilation

Mascolo et al Curr Opin Pulm Med 2003
Dimoupolos et al Circulation 2006
Now What?

- Limit other insults
  - Smoking
  - Weight loss
  - Amiodarone
  - Obstructive sleep apnea

- Risk stratify and optimize
  - Time of surgery
  - Time of cath

- Increase activity/Cardiopulmonary rehab
Types of Activity and Intensity Levels

♦ Physical activity
  – Includes all types of physical movement not solely exercise

♦ Exercise
  – Planned repetitive physical activity designed to increase fitness

♦ Competitive athletics
  – High intensity

Longmuir  P. et al Circulation 2013
Benefits of Physical Activity

Physiologic Benefits

• Improves overall functional capacity
• Weight management
• Strength

Psychological Benefits

• Improves feelings of well-being
• Self-confidence & self-esteem
• Fairly active patients positively correlate daily activity & exercise capacity
Thank you for your attention!