Eisenmenger Syndrome

Introduction

- One of the most interesting and unique physiology of congenital heart disease.
- Unfortunately, one of the worst prognosis as well. *(Circ 2015;132:2118)*
- From congenital heart disease perspective; Eisenmenger syndrome is a cyanotic heart disease.
- From pulmonary hypertension perspective; Eisenmenger syndrome is within PAH (gr I), subgr. CHD-PAH

Definition

- Defects (could be simple or complex abnormalities) that begin as systemic-to-pulmonic shunts and progress with time to severely elevation of pulmonary vascular resistance (PVR) and to reversal (pulmonary to systemic) or bidirectional shunting, causing cyanosis, erythrocytosis, and multi organ involment *(JACC 2013; 62:25, Suppl D)*
- First described by Victor Eisenmenger in 1897.
- Improved understanding of physiology by Paul Wood in 1951.
- 1-3% of all ACHD. 10% of CHD-PAH which is 10% of PAH. *(REVEAL registry)*
- Uncorrected PDA, VSD and ASD comprises 75% off all Eisenmenger syndrome. (*Int J Cardiol 1994;45(3):199)*
- Other lesions include TAPVR, AVSD, TA, TGA, aortopulmonary communication, and post op systemic to pulmonic shunts surgery such as Waterston or Pott.

Assessment

- Dyspnea; hard to detect since patient have never been "normal".
- Very low mean peak VO2 = 12.5 ml/kg/min = 3.5 METs. *(EHJ 2012;33:1386)*
- Hemoptysis, syncope
- Cyanosis (uniform or differential), clubbing
- Signs of PAH: Prominent A, palpable P2, RV heave, ejection click at pulmonic area
- Signs of RV failure: JVP, V wave, TR, hepatomegaly, ascites, edema

Investigation

- O2 sat (finger and toe)
- Hb, Hct, iron panel, ferritin, Cr, uric acid
- ECG: Signs of RVH
- CXR: Central dilatation of pulmonary trunks, peripheral pruning of pulmonary vasculature
- CV imaging: Sign of RV pressure load. Consider MRI to evaluate for anatomy, function and flow
- Cardiac catheterization +/- O2 and pulmonary vasodilator challenge is essential to evaluate treatment options. PVR is usually more than 10 wU.
- Rule out other causes of reversal shunt (cyanosis but no elevated PVR) e.g. RV outflow obstruction, abnormal RV compliance, extracardiac R → L.
- Rule out other causes of PH e.g. PH due to LHD (systemic ventricular failure, stenosis lesion), pulmonary venous hypertension, venous obstruction, restrictive lungs, etc.

Supportive care and education

- Eisenmenger is a multisystem disease
  - Secondary erythrocytosis *(Int J Cardio 2004;97:109)*
    The same as other cyanosis ACHDs.
    Hyperviscosity syndrome: headache, weakness, fatigue, or poor concentration.
    Avoid phlebotomy unless symptomatic + Hb> 20g/dL or Hct 65%. *(ACC/AHA ACHD 2008)*
    Total of 500 cc of bloodletting with 750-1000 ml of IV volume replacement.
  - Hematologic abnormalities
    Clot - PA thrombosis, careful with INR monitoring
    Bleed - Gingival bleeding, pulmonary bleeding, menorrhagia
    Iron deficiency anemia
  - Neurologic complications, cerebrovascular accidents, brain abscess
  - Renal dysfunction
  - Hyperuricemia: Decreased renal uric acid clearance, gout
  - Cholecystitis (Ca billirunate stone)
  - CV: Arrhythmia, LM compression from enlarged PA, RVH induced RV ischemia
  - Endocarditis risk
  - Emboli: Air filter, avoid endocardial pacing
  - Orthopedic issues such as scoliosis
- Avoid dehydration (hot tubs, excessive alcohol)
- Avoid high altitude but some evidences of safety with commercial air travel
- Strenuous exercise, especially isometric exercise
- Pregnancy: 50% maternal mortality

Conditions in which pregnancy is WHO IV (pregnancy contraindicated)

- PAH of any cause (include Eisenmenger)
- Severe systemic ventricular dysfunction (LVE <30%, NYHA II-IV)
- Previous peripartum cardiomyopathy with any residual impairment of left ventricular function
- Aortic dilatation (> 45 in Marfan, >50 mm in bicuspid AV)
- Native severe coarctation

(ESC CV in pregnancy 2011)

Treatment

- Very limited evidences: Oxygen, digoxin, warfarin?, CCB?
- From a non-randomized, retrospective study, patient with Eisenmenger had better survival rate with PAH specific medication i.e. ERA, PDEinh or prostacyclin. *(Circ 2010;121:20)*
- Bosentan have been shown to improve PVRi, mPA and 6MWD in an RCT of 54 patients *(BREATH-5 Circ 2006).*

Prognosis

- 25% mortality at 5 years. May live until 40-50's yo. Better prognosis than iPAH.
- Pump failure is the most common cause of death. Others causes include SCD, pulmonary hemorrhage, infection, CVA.
- Poor prognosis factors include older age, pre-tricuspid shunt, not sinus rhythm, lower resting O2 sat, presence of pericardial effusion. (*Circ* 2017;135:1432).

Suggested Readings

- The adult patient with Eisenmenger syndrome: Part I-III. (Curr Card Rev 2010, 6, 363-372)
- Improved survival among patients with Eisenmenger syndrome receiving advanced therapy for pulmonary arterial hypertension. (Circ 2010;121:20-25.)