Pulmonary Hypertension in Adult Congenital Heart Disease

The Eisenmenger Syndrome: Epidemiological and Clinical Aspects
1897, Victor Eisenmenger:
“Die angeborenen Defecte der Kammerscheidewand des Herzens.“
Zeitschrift für klinische Medizin

1958, Paul H. Wood:
summarized Eisenmenger's accounts

“The patient was a powerfully built man of 32 who gave a history of cyanosis and moderate breathlessness since infancy. He managed well until January of 1894 when dyspnoea increased and oedema set in. Seven months later he was admitted to the hospital in a state of heart failure……
He improved with rest and digitalis, but collapsed and died suddenly on November 13 following a large haemoptysis”

Victor Eisenmenger
(*1864, † 1932)

Paul Hamilton Wood
(*1907; † 1962)
Definition

Victor Eisenmenger (Vienna, *1864, † 1932)
- 1897: 32 y, Dyspnoe, Cyanosis, Haemoptoe
- VSD & PAH

EM Complex

Paul H. Wood (London, *1907, † 1962)
- 1958: syst. PH due to elevated PVR
- PAPVD, ASD, SVD
- AVSD, VSD, TA, AoPW
- PDA, BT-Shunt

EM Syndrome - PVOD
Evolution of the Eisenmenger Syndrome

- PVR (Pulmonary Vascular Resistance) increase
- Shunt Reversal
- Oxygen Supply decrease

Stages:
- VSD Early
- VSD Mid
- VSD End Stage
- VSD Late
Chronic Cyanosis

- PVR
- Shunt Reversal
- Oxygen Supply

Exercise Intolerance
Hyperviscosity Syndrome

Decompensated Polyglobul

Coagulation Disorder

Thromboses, TIA, Brain Abscesses, Acne

Gingival Hyperplasia

Hemoptysis, Bleedings, PA-aneurysm

Cyanotic Arthropyathy
Cyanotic Nephropathy

Clubbing Hippocratic Nails

Pseudo-Conjunctivitis

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Symptoms in Eisenmenger Patients

**Infants**
- Usually healthy childhood
- **Symptoms:**
  - Fatigue
  - especially during crying spells and
  - at feeding time
  - Difficulty eating, breathing or sucking
  - Poor weight gain
  - Slow growth or other physical retardation
  - Low Tolerance for extra Exertion
  - Dyspnorea, Rapid Breathing
  - Fainting/Syncope

**Adults**
- **All patients are symptomatic !!!**
- **Symptoms:**
  - Cyanosis
  - Dyspnoea
  - Cough
  - Fatigue
  - Chest Pain
  - Hemoptyisis
  - Syncope
Special Problems in Eisenmenger Patients

- Syncope
- Hypertrophic Osteoarthropathy
- Scoliosis
- Cholelithiasis
- Hyperuricemia/Gout
- Hemoptysis
- Hemostatic Abnormalities (Bleeding disorders, Thrombosis)

Cerebrovascular Accidents

- Acne
- Risk of Systemic Infection (e.g. Brain Abscess, Endocarditis)
- Congestive Heart Failure
- Arrhythmias
- Pulmonary Artery Calcification & Aneurysm
- Renal Dysfunction
- Hyperviscosity, Erythrocytosis

Eisenmenger Syndrome

Coagulopathy, Platelet Consumption
Exercise Intolerance
- Identifies Patients at Risk of Hospitalization or Death
- Even after accounting for Age, Gender, Functional Class (NYHA), Laboratory Parameters, Underlying cardiac lesions.

→ Patients with a worse exercise capacity: more likely to be admitted to hospital
Special Risks in Eisenmenger Patients

**Eisenmenger Syndrome**
- Pulmonary infection
- Angiography – CARE!
- Altitude exposure
- General anesthesia
- Dehydration
- Venesection, Anemia commonly due to iron deficiency
- Drugs (vasodilator, diuretics, contraception, nonsteroidal anti-inflammatory drugs)
- Hemorrhage
- Intravenous lines (air embolism, infection, brain abscess)
- Cardiac and noncardiac surgery
- Pregnancy (contraindicated)
Natural History in Eisenmenger Patients

General Aspects

- Adult patients limited (Functional Class II-III)
- Survival to 3rd-5th decade common
- **Prognosis:**
  - Simple lesions (ASD, VSD, PDA) better
  - Complex lesions CHD worse
- **Death occurs:**
  - Suddenly (2/3) – Arrhythmias?
  - Heart failure
  - Massive Haemoptysis (e.g. PA rupture)
Natural Course

Survival
Eisenmenger Physiology vs. Idiopathic PAH

- **Idiopathic PAH**
  - Mortality: 50% after 2.8 years

- **Eisenmenger Physiology**
  - Median Age at Diagnoses: 25 (17-34) years
  - Mortality: approx. 3% per year
  - Prognosis appears better than in Idiopathic PAH
  - **But:** Survival is the worst of all of the CHD Patients!

Hopkins *J Heart Lung Transplant* 1996

Eisenmenger’s Physiology and Survival

- **Total Population**
  Kaplan-Meier Survival Curve (Greenwood CI)

- **Simple vs. Complex Congenital Heart Defects**
  Kaplan-Meier Survival Curve

- **Prognosis is affected by**
  - RV function
  - LV function
  - Renal function
  - Cyanosis
  - Physical function
  - UA/Bili/BNP/ET-1/VEGF
  - Volume retention
  - Age at deterioration

Daliento *Eur Heart J* 2006
Eisenmenger-Survival as function of ...

- **Predictors of mortality**
  - Functional class
  - Signs of heart failure
  - Low serum albumin and potassium levels

- History of clinical arrhythmia
- QRS duration and QTc interval

Diller *Eur Heart J* 2006
Euro Heart Survey on ACHD

**Database**
- **79 Centres** (48 specialized), **26 Countries**
- **4168 Patients** (87% in specialized Centres)
- Retrospectively
- Consecutively visiting Centres 1998-04/2004 → median **Follow-up: 5 years** (3.6-5.7 years)

**Diagnosis**
- ASD II
- VSD
- TOF
- Aortic Coarctation
- TGA
- Marfan Syndrome
- Functional Single Ventricle, Fontan Circulation
- **Eisenmenger Syndrome** and other Cyanotic Defects

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Engelfried *Eur Heart J* 2005
Marelli *Circulation* 2007
Euro Heart Survey on ACHD

Subgroup of Patients with VSD
Kaplan-Meier Survival Curve

- **Database**
  - 4168 Patients
  - 12.7% PAH
  - 5.5% Eisenmenger Syndrome

- **Eisenmenger Syndrome**
  - Median Age: 30 years
  - Females: 64%
  - Oxygen saturation at rest: 82%
  - Mortality 20.6% (Range 14.5-26.7%)

Engelfried *Heart* 2007

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Requirements for Eisenmenger Patients

Interdisciplinary, multilateral Care

Handling in Specialized Centres (Tertiary Care Centres)

Transfer to GUCH-Services

- Adult Cardiology
- Pediatric Cardiology
- Surgery

- Rheumatology
- Pneumology
- Haematology
- Others . . .