



Pulmonary Hypertension in Adult Congenital Heart Disease

The Eisenmenger Syndrome: **Epidemiological and Clinical Aspects**



Eisenmenger – The Story

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 breath-lessness 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

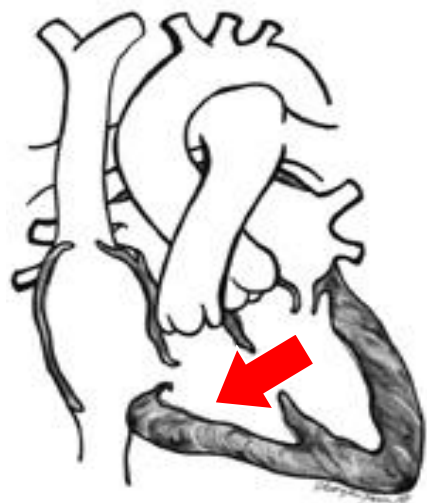
EM Complex

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

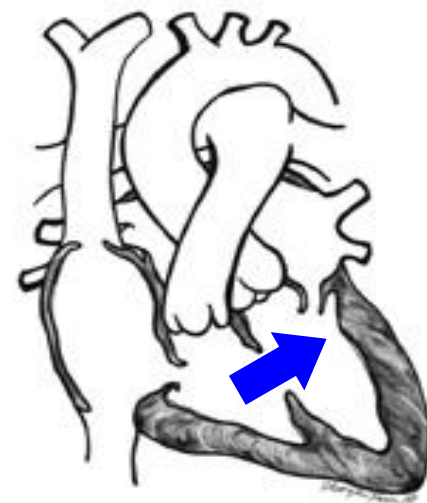
Evolution of the Eisenmenger Syndrome



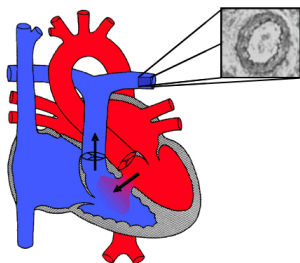
PVR ↑

Shunt Reversal

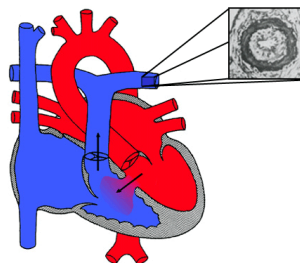
Oxygen Supply ↓



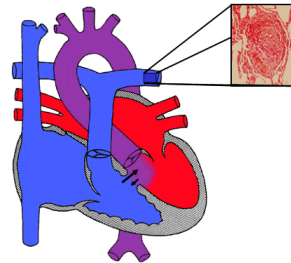
VSD Early



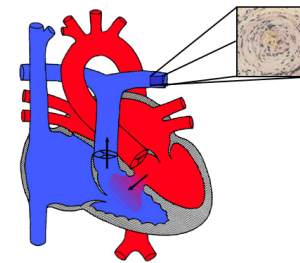
VSD Mid



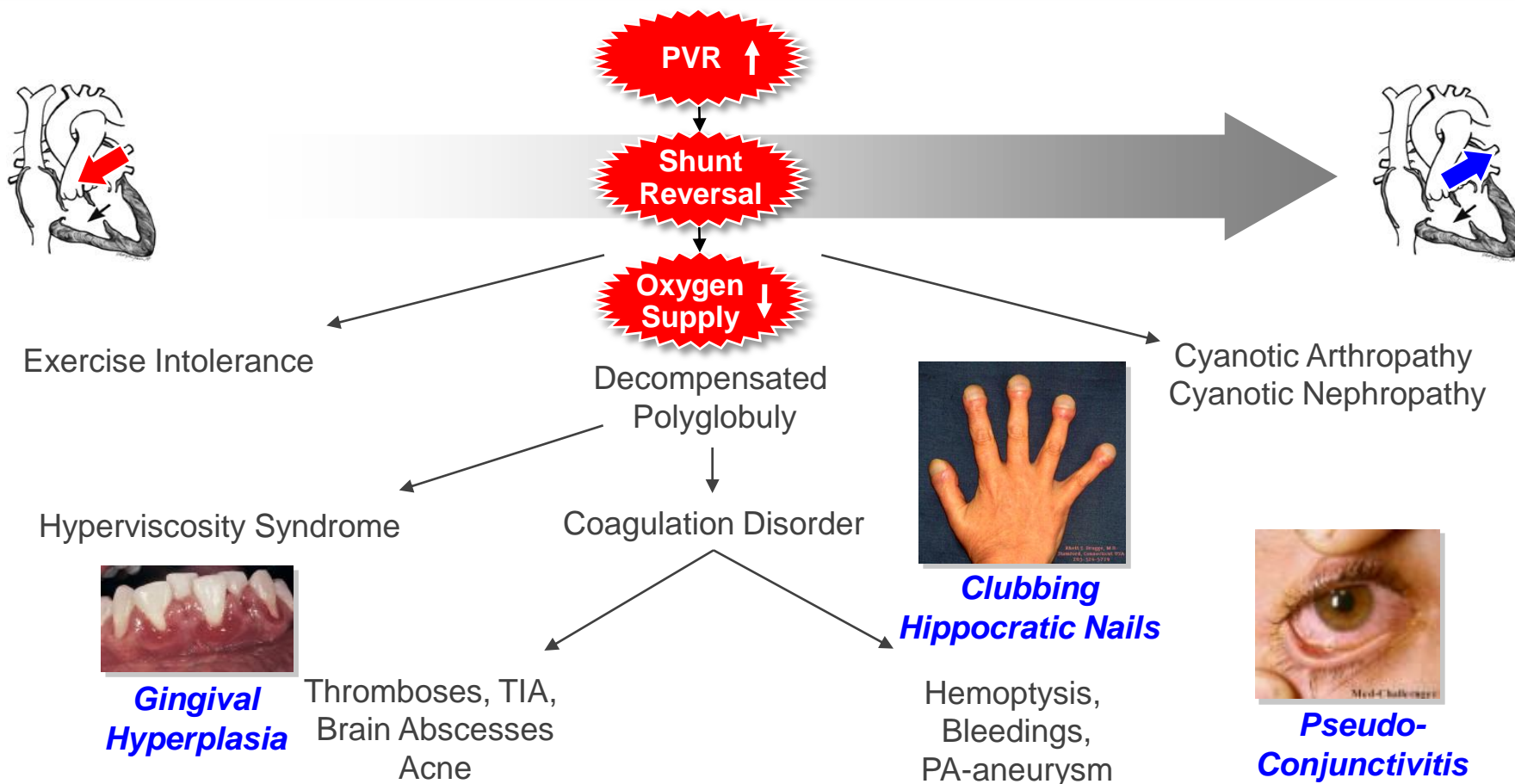
VSD End Stage



VSD Late



Chronic Cyanosis



Symptoms in Eisenmenger Patients

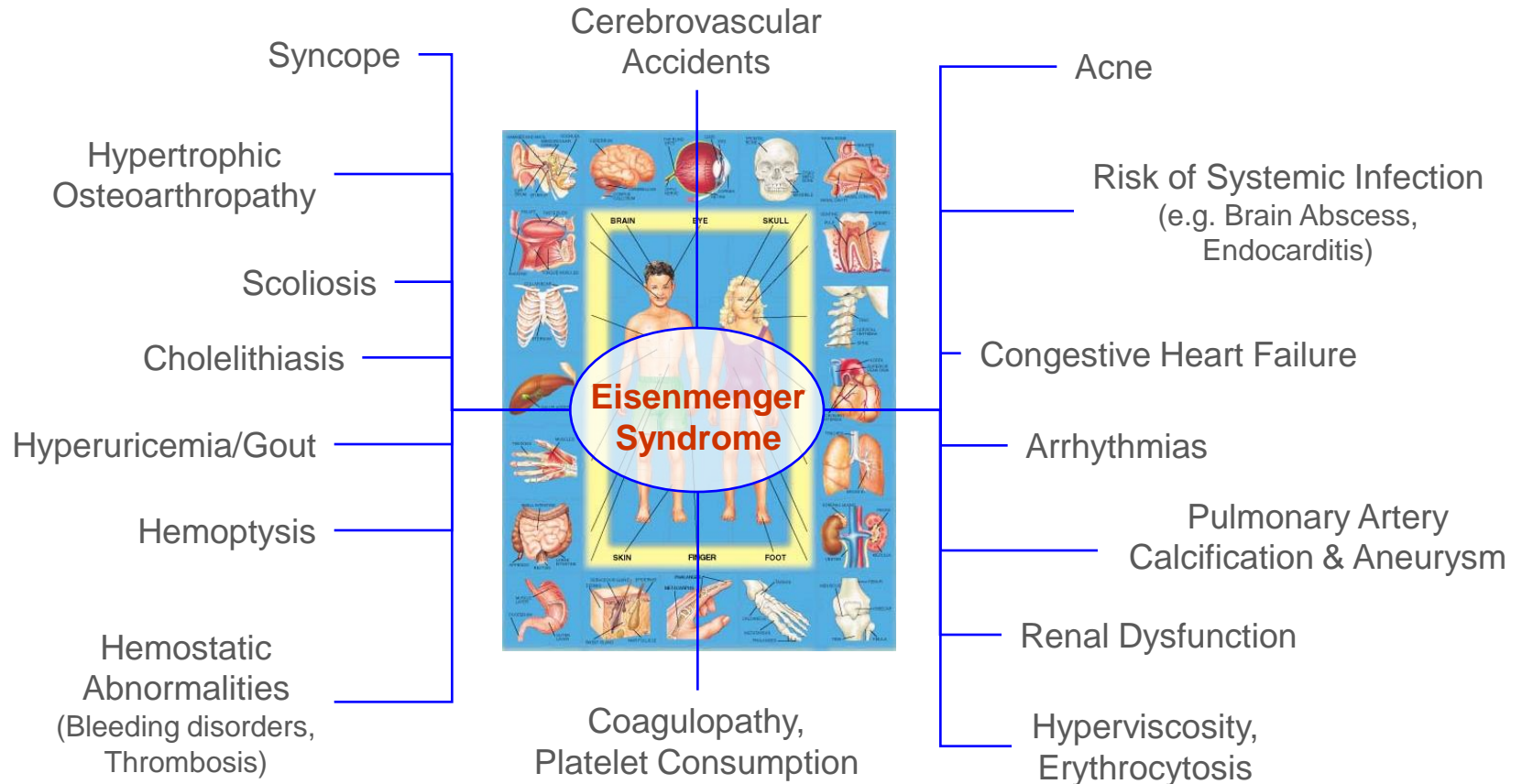
Infants

- 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
 - Dysnpoea, Rapid Breating
 - Fainting/Syncope

Adults

- **All patients are symptomatic !!!**
- **Symptoms:**
 - Cyanosis
 - Dysnpoea
 - Cough
 - Fatigue
 - Chest Pain
 - Hemoptysis
 - Syncope

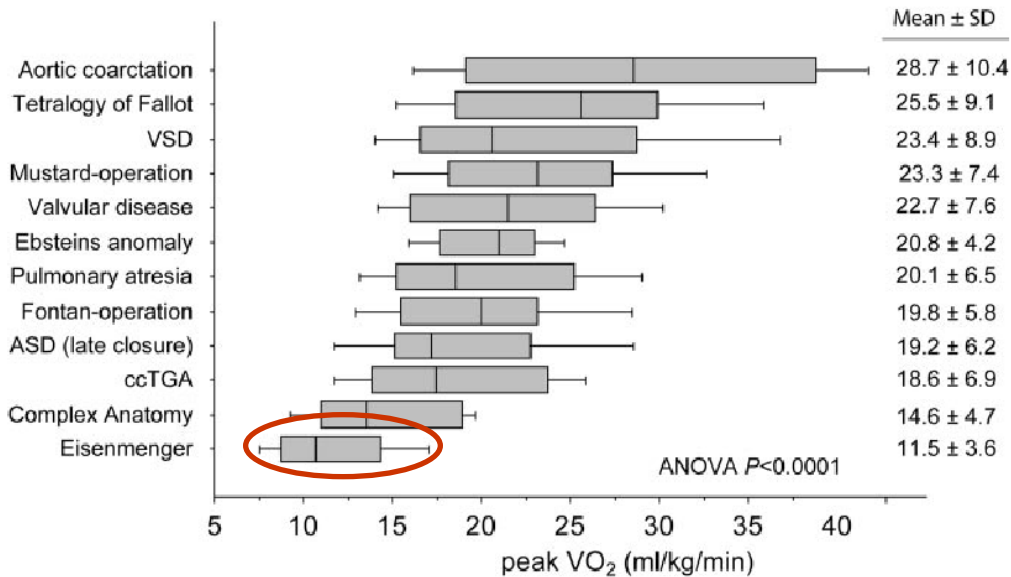
Special Problems in Eisenmenger Patients



Exercise Capacity in Adults with CHD

Distribution of Peak VO₂

in Different Diagnostic Groups



Diller *Circulation* 2005

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

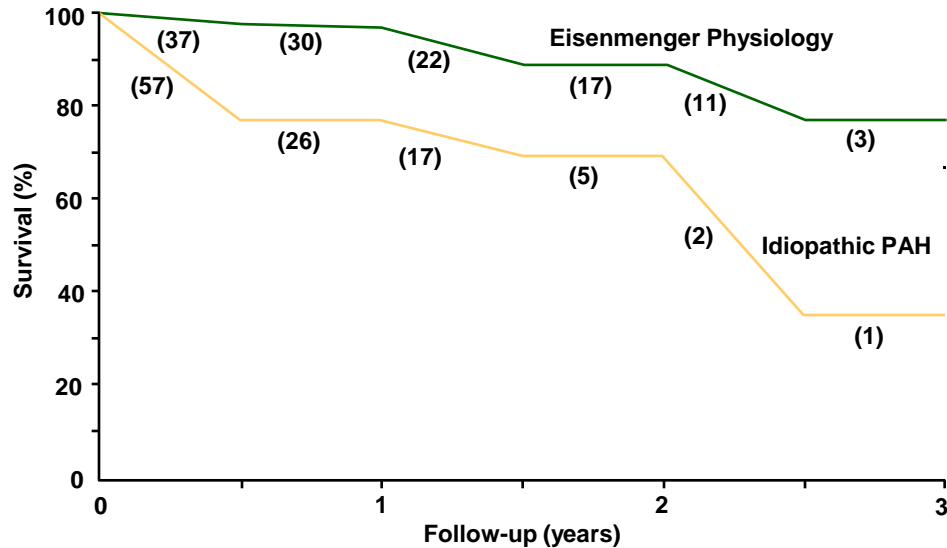
General Aspects

- 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



Hopkins *J Heart Lung Transplant* 1996

Idiopathic PAH

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

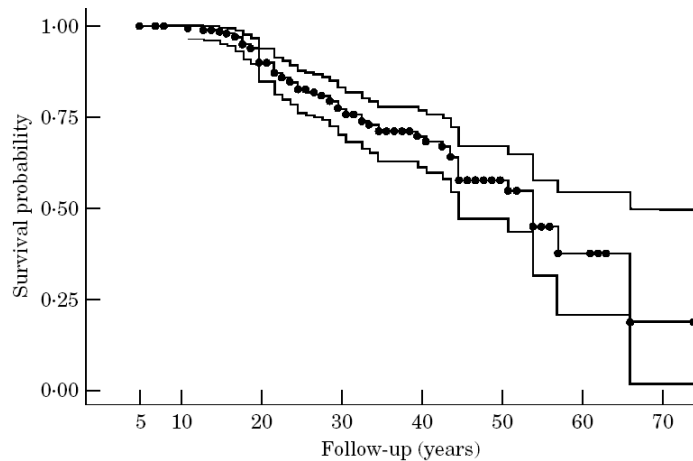
Eisenmenger Physiology

- 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!

D'Alonzo *Ann Intern Med* 1991

Total Population

Kaplan-Meier Survival Curve (Greenwood CI)

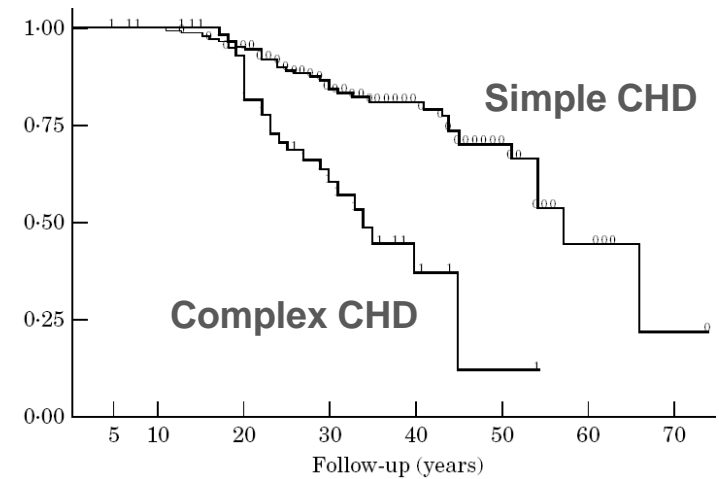


Prognosis is affected by

- RV function
- LV function
- Renal function
- Cyanosis
- Physical function
- UA/Bili/BNP/ET-1/VEGF
- Volume retention
- Age at deterioration

Simple vs. Complex Congenital Heart Defects

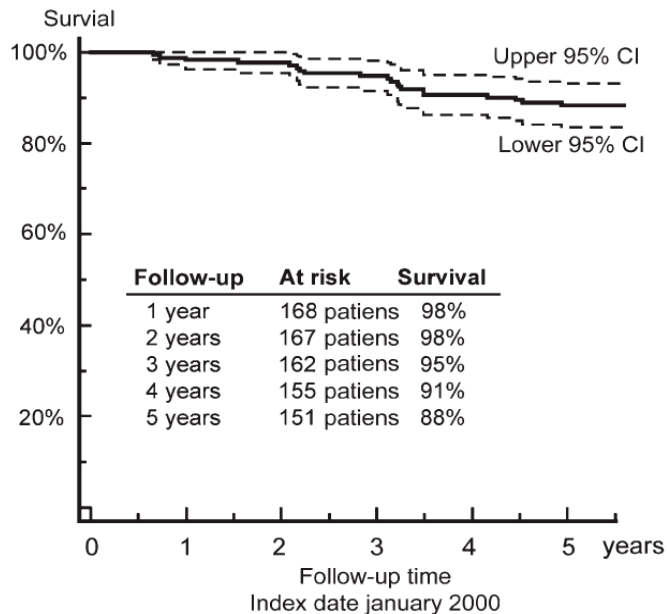
Kaplan-Meier Survival Curve



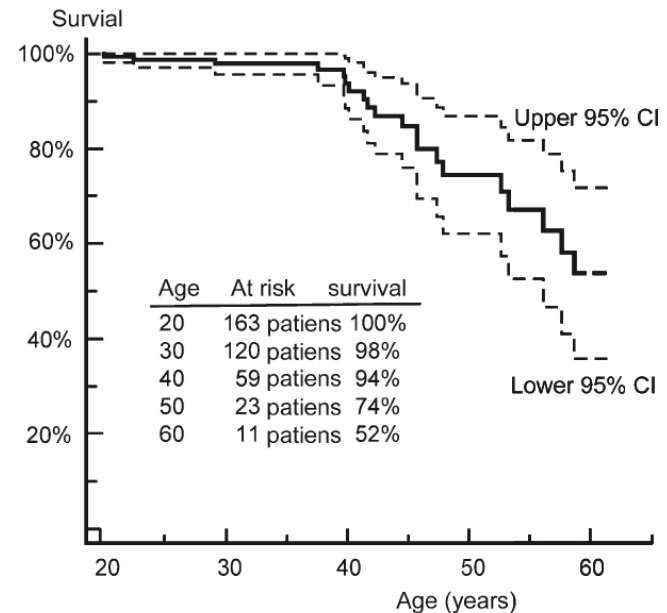
Daliento *Eur Heart J* 2006

Eisenmenger-Survival as function of ...

Follow-up Time



Age



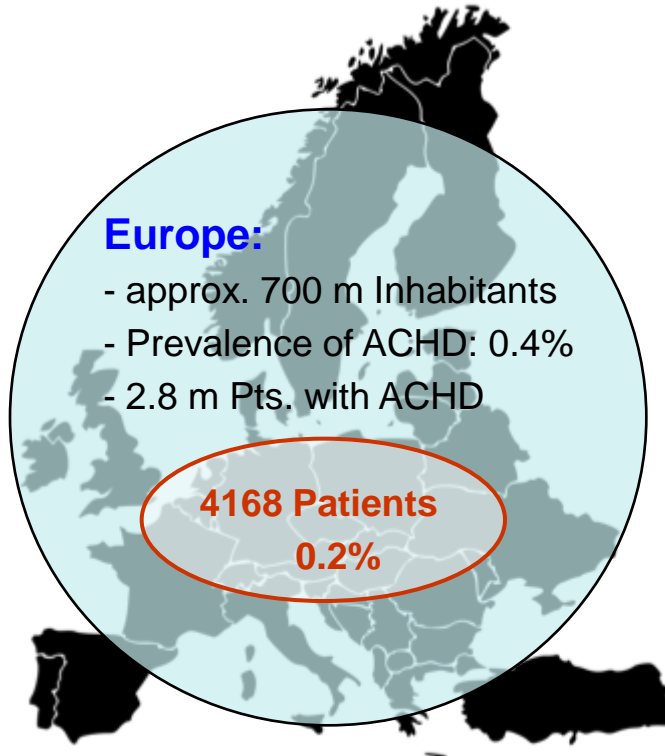
■ Predictors of mortality

- 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



Engelfried *Eur Heart J* 2005
Marelli *Circulation* 2007

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)

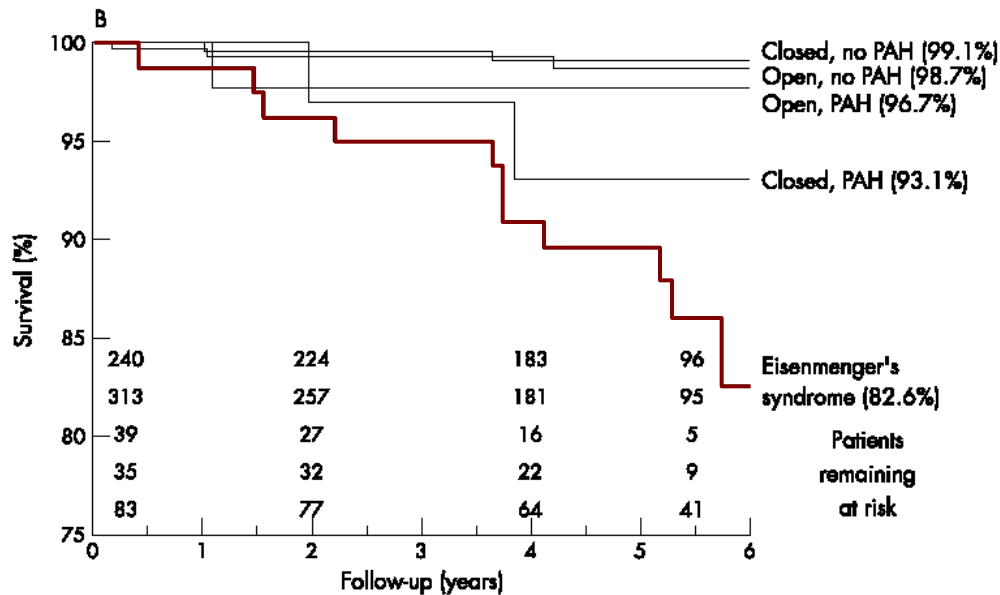
Diagnosis

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

Euro Heart Survey on ACHD

Subgroup of Patients with VSD

Kaplan-Meier Survival Curve



Engelfried *Heart* 2007

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%)

Requirements for Eisenmenger Patients

Interdisciplinary, multilateral Care

Handling in Specialized Centres (Tertiary Care Centres)

Transfer to GUCH-Services

