Heart Failure in Adults with Congenital Heart Disease

Webinar ACHD

19th of May 2020

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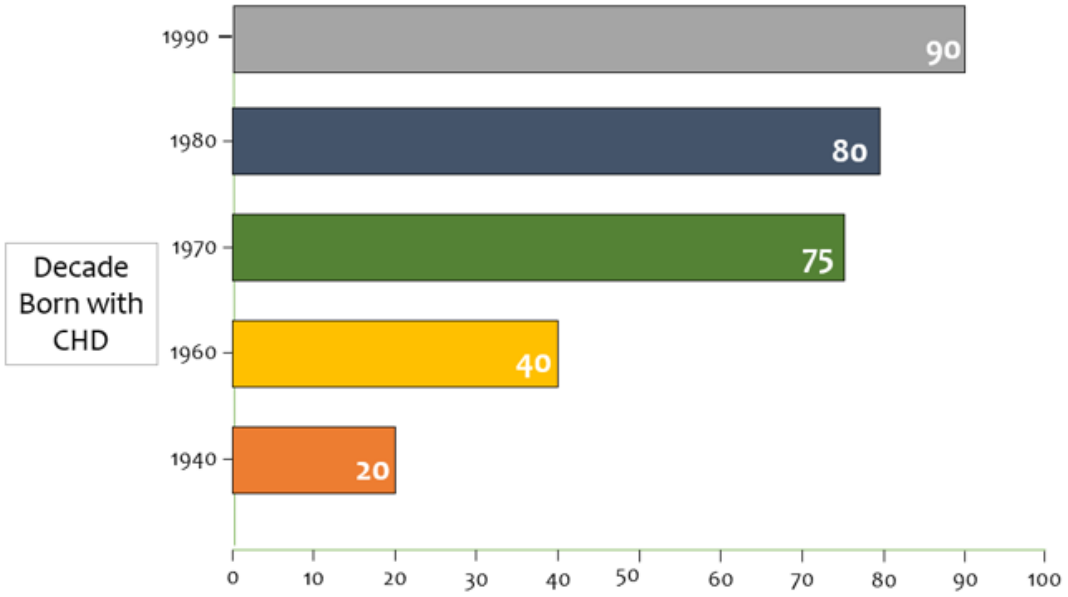
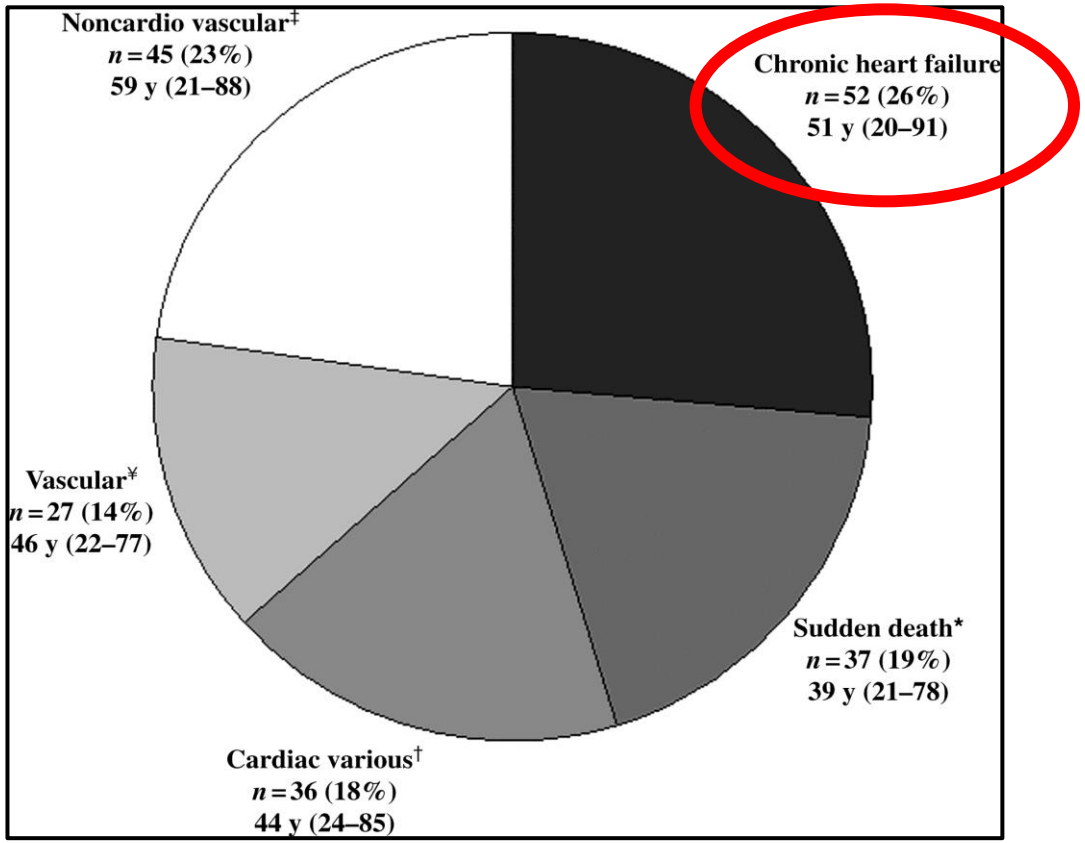
What's the size of the problem?



Warnes CA et al. *J Am Coll Cardiol*. 2001
Moons P et al. *Circulation* 2010



Verheugt et al *Eur Heart J 2010*
Diller et al *Circulation 2015*

Table 3.1 Definition of heart failure with preserved (HFpEF), mid-range (HFmrEF) and reduced ejection fraction (HFrEF)

Type of HF		HFrEF	HFmrEF	HFpEF
CRITERIA	1	Symptoms ± Signs[a]	Symptoms ± Signs[a]	Symptoms ± Signs[a]
	2	LVEF <40%	LVEF 40–49%	LVEF ≥50%
	3	–	1. Elevated levels of natriuretic peptides[b]; 2. At least one additional criterion: a. relevant structural heart disease (LVH and/or LAE), b. diastolic dysfunction (for details see Section 4.3.2).	1. Elevated levels of natriuretic peptides[b]; 2. At least one additional criterion: a. relevant structural heart disease (LVH and/or LAE), b. diastolic dysfunction (for details see Section 4.3.2).

BNP = B-type natriuretic peptide; HF = heart failure; HFmrEF = heart failure with mid-range ejection fraction; HFpEF = heart failure with preserved ejection fraction; HFrEF = heart failure with reduced ejection fraction; LAE = left atrial enlargement; LVEF = left ventricular ejection fraction; LVH = left ventricular hypertrophy; NT-proBNP = N-terminal pro-B type natriuretic peptide.

[a]Signs may not be present in the early stages of HF (especially in HFpEF) and in patients treated with diuretics.

[b]BNP >35 pg/ml and/or NT-proBNP >125 pg/mL.

2016 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC)

Table 3.4 Aetiologies of heart failure

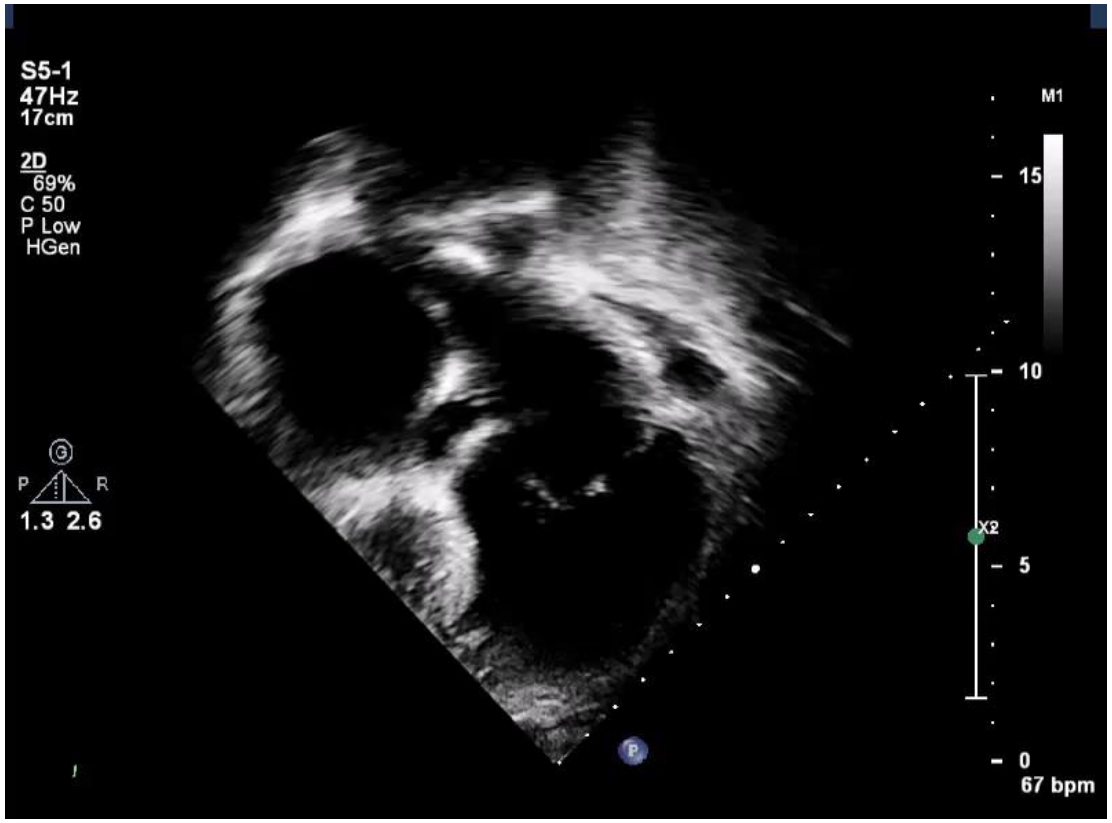
DISEASED MYOCARDIUM		
Ischaemic heart disease	Myocardial scar	
	Myocardial stunning/hibernation	
	Epicardial coronary artery disease	
	Abnormal coronary microcirculation	
	Endothelial dysfunction	
Toxic damage	Recreational substance abuse	Alcohol, cocaine, amphetamine, anabolic steroids.
	Heavy metals	Copper, iron, lead, cobalt.
	Medications	Cytostatic drugs (e.g. anthracyclines), immunomodul antibodies such as trastuzumab, cetuximab), antidep anti-inflammatory drugs, anaesthetics.
	Radiation	
Immune-mediated and inflammatory damage	Related to infection	Bacteria, spirochaetes, fungi, protozoa, parasites (Ch
	Not related to infection	Lymphocytic/giant cell myocarditis, autoimmune dise arthritis, connective tissue disorders, mainly systemi eosinophilic myocarditis (Churg–Strauss).
Infiltration	Related to malignancy	Direct infiltrations and metastases.
	Not related to malignancy	Amyloidosis, sarcoidosis, haemochromatosis (iron), g lysosomal storage diseases (e.g. Fabry disease).
Metabolic derangements	Hormonal	Thyroid diseases, parathyroid diseases, acromegaly, C disease, Addison disease, diabetes, metabolic syndro to pregnancy and peripartum.
	Nutritional	Deficiencies in thiamine, L-carnitine, selenium, iron, p (e.g. malignancy, AIDS, anorexia nervosa), obesity.
Genetic abnormalities	Diverse forms	HCM, DCM, LV non-compaction, ARVC, restrictive c expert documents), muscular dystrophies and lamin
ABNORMAL LOADING CONDITIONS		
Hypertension		
Valve and myocardium structural d	Acquired	Mitral, aortic, tricuspid and pulmonary valve disease
	Congenital	Atrial and ventricular septum defects and others (fo
Pericardial and endomyocardial pathologies	Pericardial	Constrictive pericarditis Pericardial effusion
	Endomyocardial	HES, EMF, endocardial fibroelastosis.
High output states		Severe anaemia, sepsis, thyrotoxicosis, Paget's disease, arteriovenous fistula, pregnancy.
Volume overload		Renal failure, iatrogenic fluid overload.
ARRHYTHMIAS		
Tachyarrhythmias		Atrial, ventricular arrhythmias.
Bradyarrhythmias		Sinus node dysfunctions, conduction disorders.

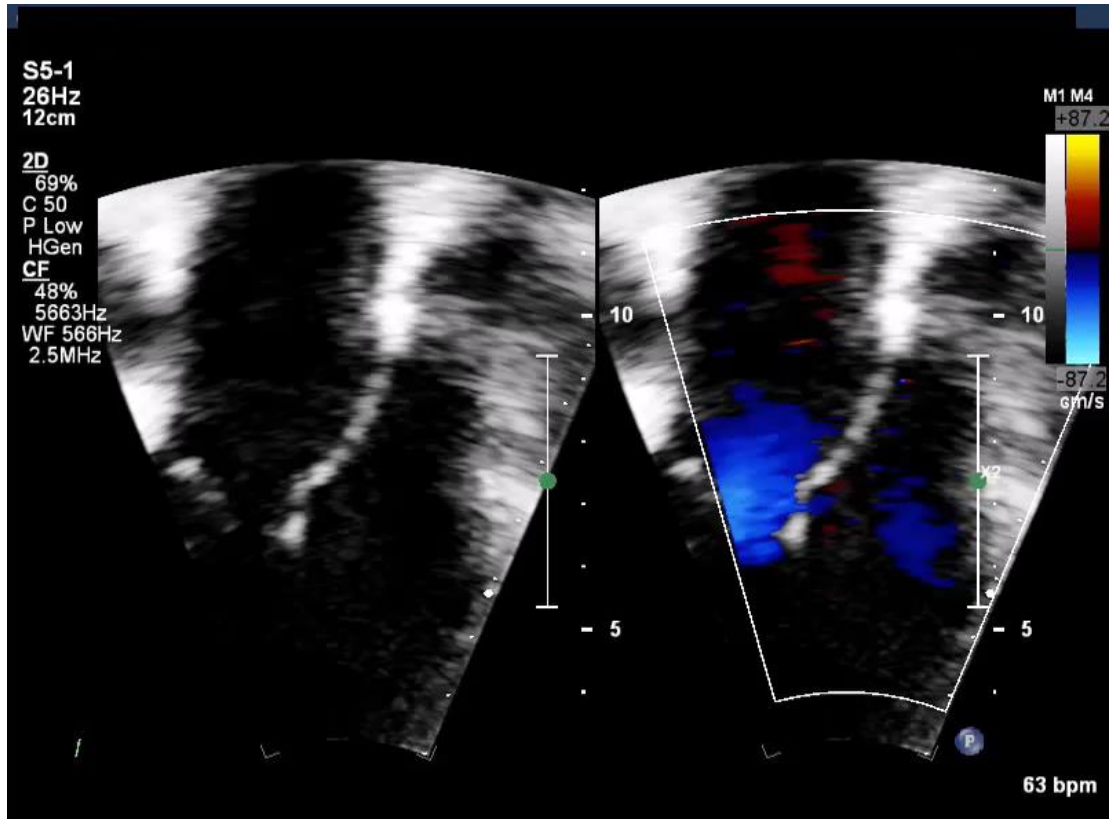
Criteria in the 2018 position statement[2]

1. Severe and persistent symptoms of heart failure (NYHA class III [advanced] or IV)
2. Severe cardiac dysfunction, defined by:
   - reduced LVEF ≤30%
   - isolated RV failure (e.g. ARVC)
   - non-operable severe valve abnormalities
   - congenital abnormalities

   persistently high (or increasing) BNP or NT-proBNP values and data showing severe diastolic dysfunction or LV structural abnormalities, according to the ESC definition of HFpEF and HFmrEF
3. Episodes of pulmonary or systemic congestion requiring high-dose intravenous diuretics (or diuretic combinations) or episodes of low output requiring inotropes or vasoactive drugs or malignant arrhythmias causing >1 unplanned visit or hospitalisation in the past 12 months
4. Severe impairment of exercise capacity with inability to exercise or low 6MWTD (<300 m) or pVO_2 (<12–14 ml/kg/min), estimated to be of cardiac origin

Heart failure?





32y old Fontan (tricuspid atresia), NYHA II, 6 MWT 310m, normal NT pro BNP, normal LV EDP, no peripheral edema, no HF admission , no arrhythmias

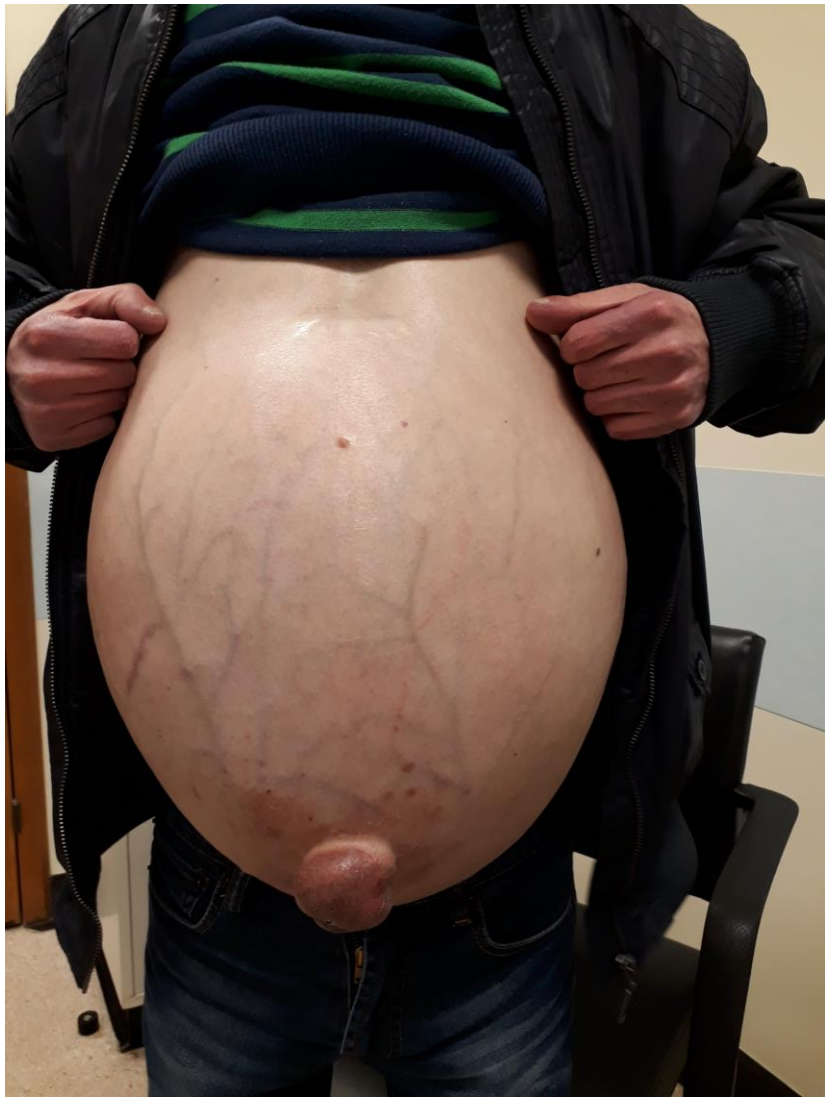
Yes, this is a failing Fontan

High Fontan pressures

Severe ascites

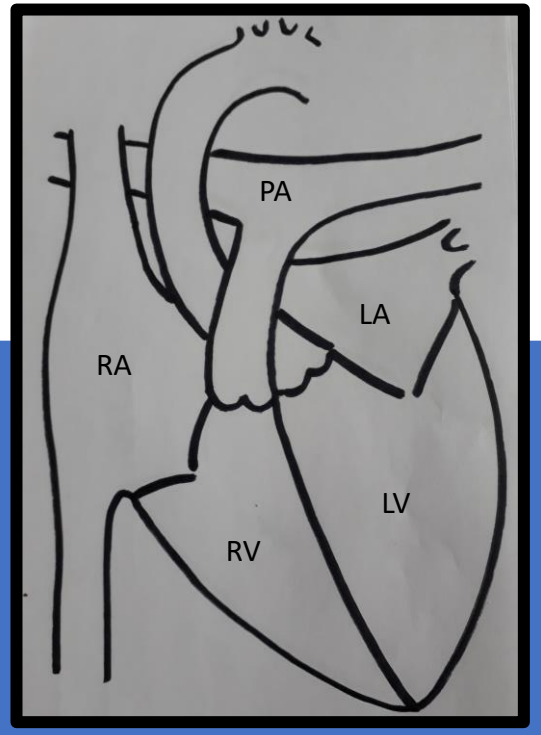
Liver cirrhosis

Major portal hypertension

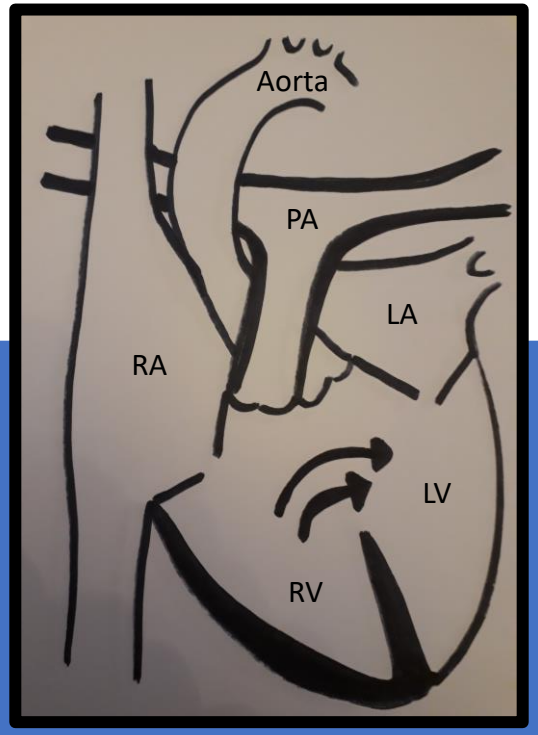


Challenging heterogenous population

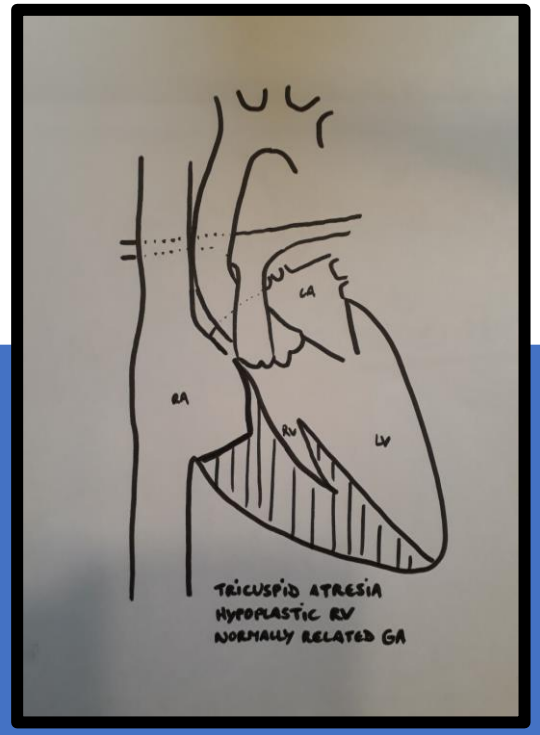
- Cardiac anatomy
- Previous surgical/interventional procedures
- Residual hemodynamic lesions/natural course



Biventricular Physiology



Cyanotic Heart Disease

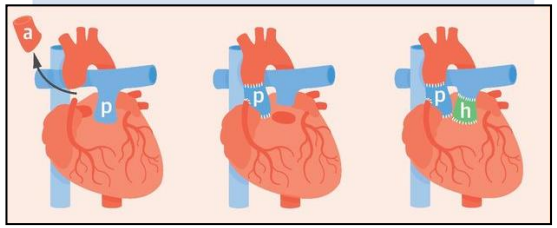


Univentricular Physiology

Biventricular Physiology – Systemic LV

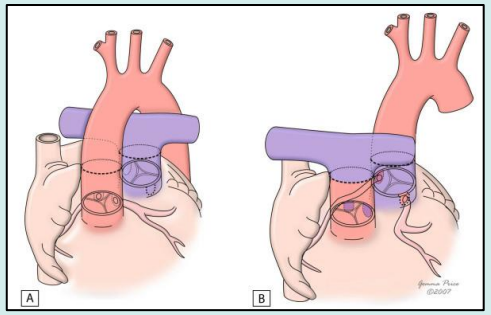
(Supra/Sub) Valvular problems

- Bicuspid aortic valve
- Dysplastic valves
- Pulmonary stenosis
- Ebstein
- .....
- Valve repair/replacement
- Ross procedure

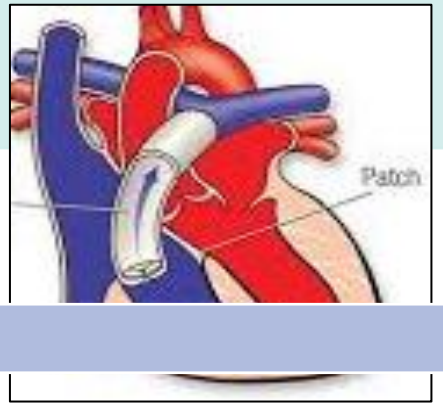


Transposition of the Great Arteries

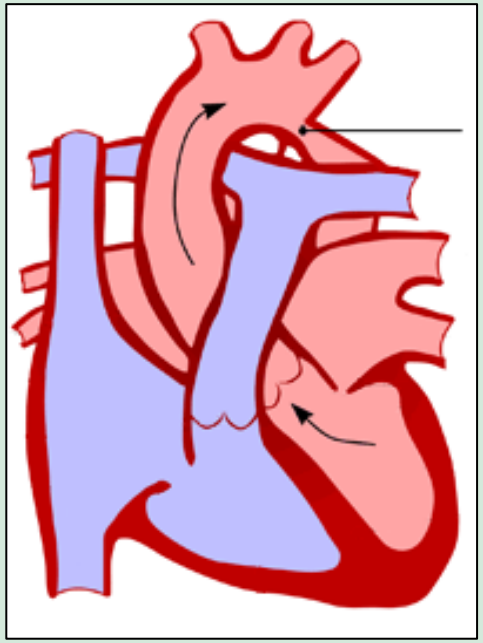
- Post arterial switch (Jatene) procedure



- TGA/VSD post Rastelli repair



Coarctation of the aorta



Intracardiac shunt lesions

- Atrial septal defect
- Ventricular septal defect
- AVSD
- Patent ductus arteriosus
- Anomalous pulmonary veins
- Tetralogy of Fallot
- Truncus arteriosus

Systemic LV – potential complications

Congenital (supra/sub) valvular problems

Valvular dysfunction

Patient prosthesis mismatch

Obstruction of blood flow

TGA

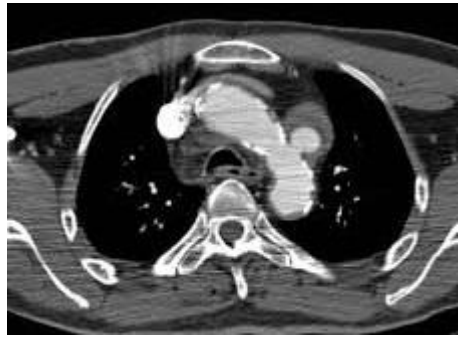
Arterial switch:
Branch PA stenosis
Valvular dysfunction
Coronary problems
(coronary re-implantation)

Rastelli:
RV to PA conduit stenosis/regurgitation
LVOT obstruction

Coarctation of the aorta

Re-coarctation

Pseudo-aneurysm at prior repair site



Arterial hypertension

Intracardiac shunt lesions

(Un)repaired ASD/VSD/AVSD/PDA:
Residual L>R shunt
(Reversal of shunt/cyanosis)

Repaired ToF:
Pulmonary regurgitation/stenosis

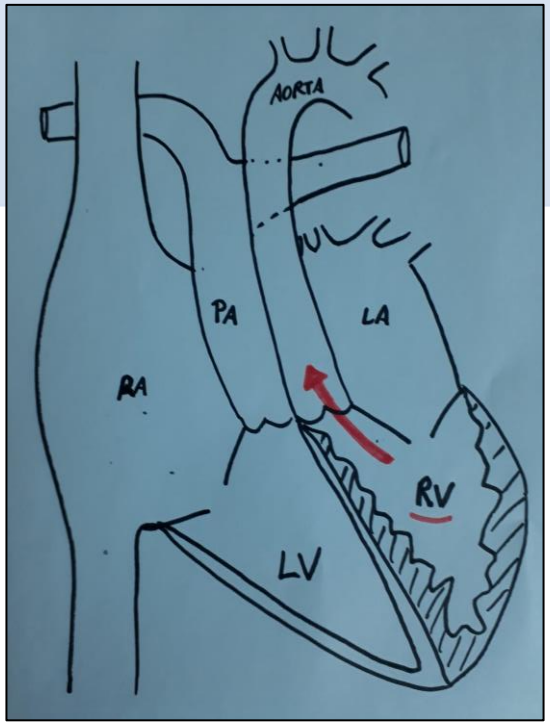
Repaired Truncus:
RV to PA conduit stenosis/regurgitation
Truncal/aortic valve dysfunction

Valvular dysfunction, arrhythmias, arterial hypertension, pulmonary hypertension, volume overload, ventricular impairment

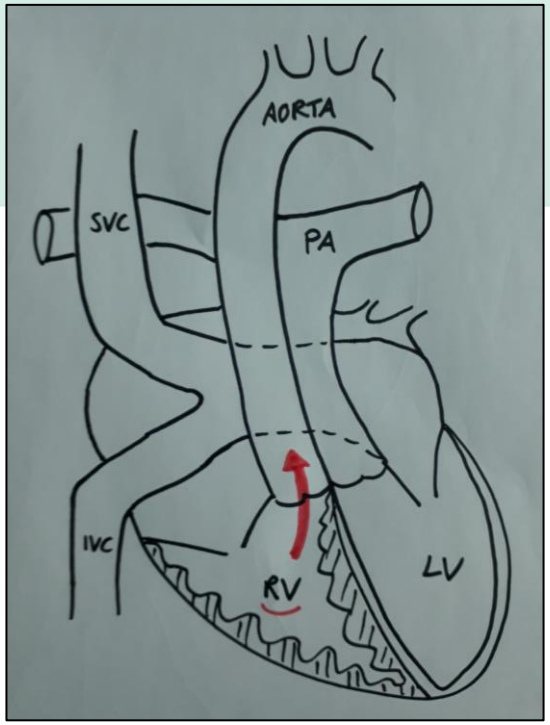
Heart failure

Biventricular Physiology – Systemic RV

Congenitally corrected TGA



TGA post atrial switch (Mustard and Senning procedure)



Systemic RV – potential complications

Congenitally corrected TGA

TGA post atrial switch (Mustard and Senning procedure)

Valvular dysfunction
Conduction abnormalities, brady/tachyarrhythmias, SCD
Pulmonary hypertension
Baffle obstruction/leakage
Ventricular impairment

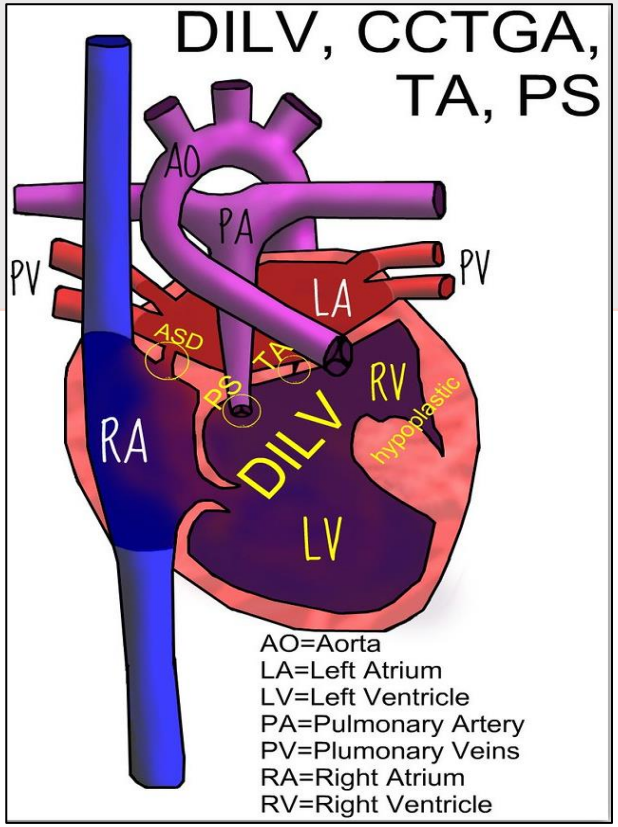
Heart failure

Graham et al. Outcome in ccTGA JACC 2000

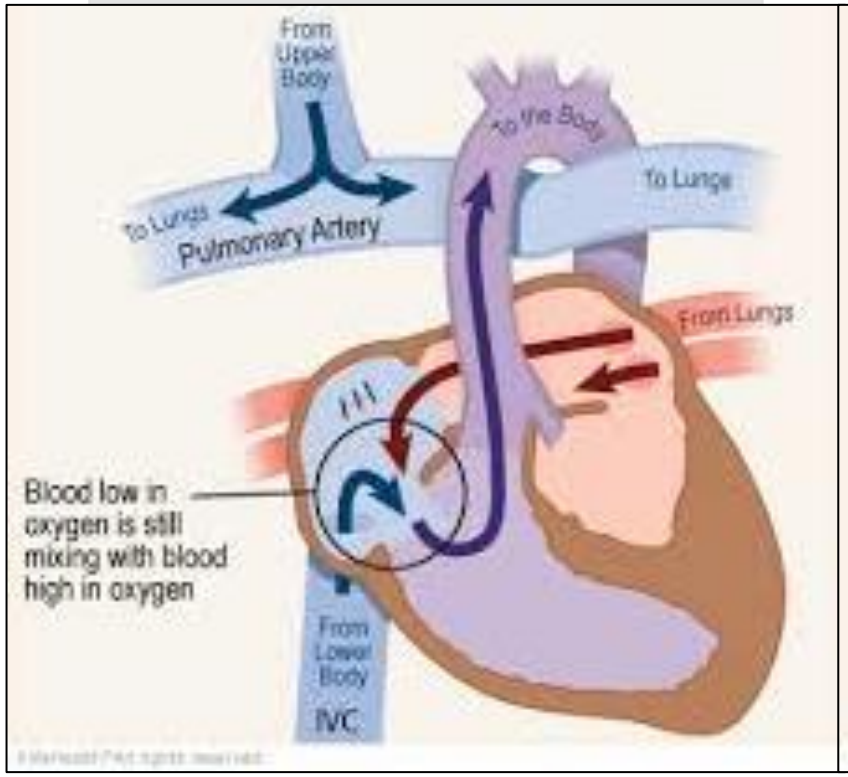
Roos-Hesselink et al. Eur Heart 2004

Univentricular Physiology

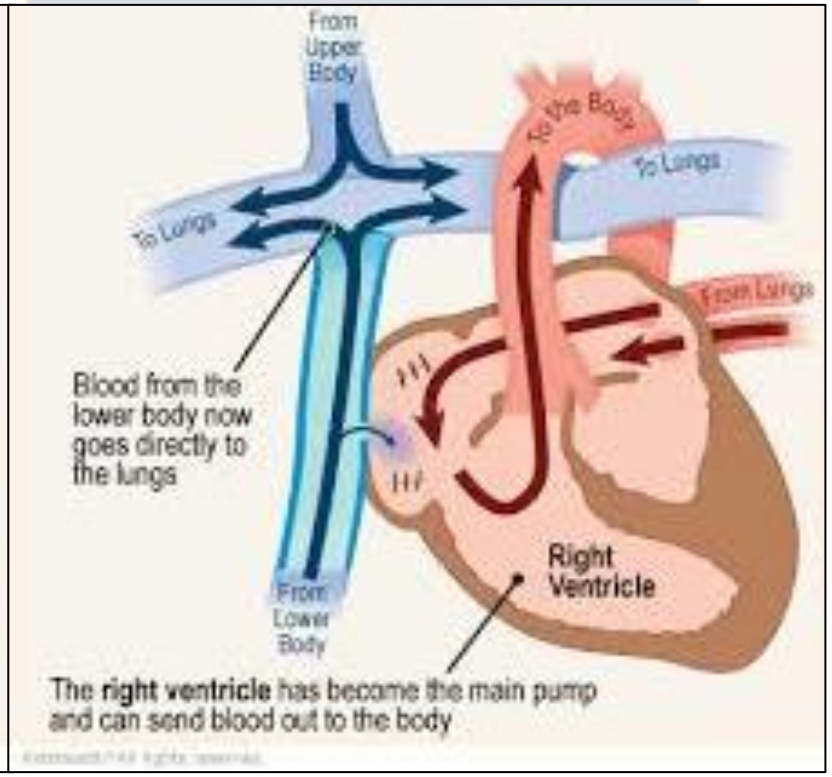
Unrepaired SV



Glenn palliation

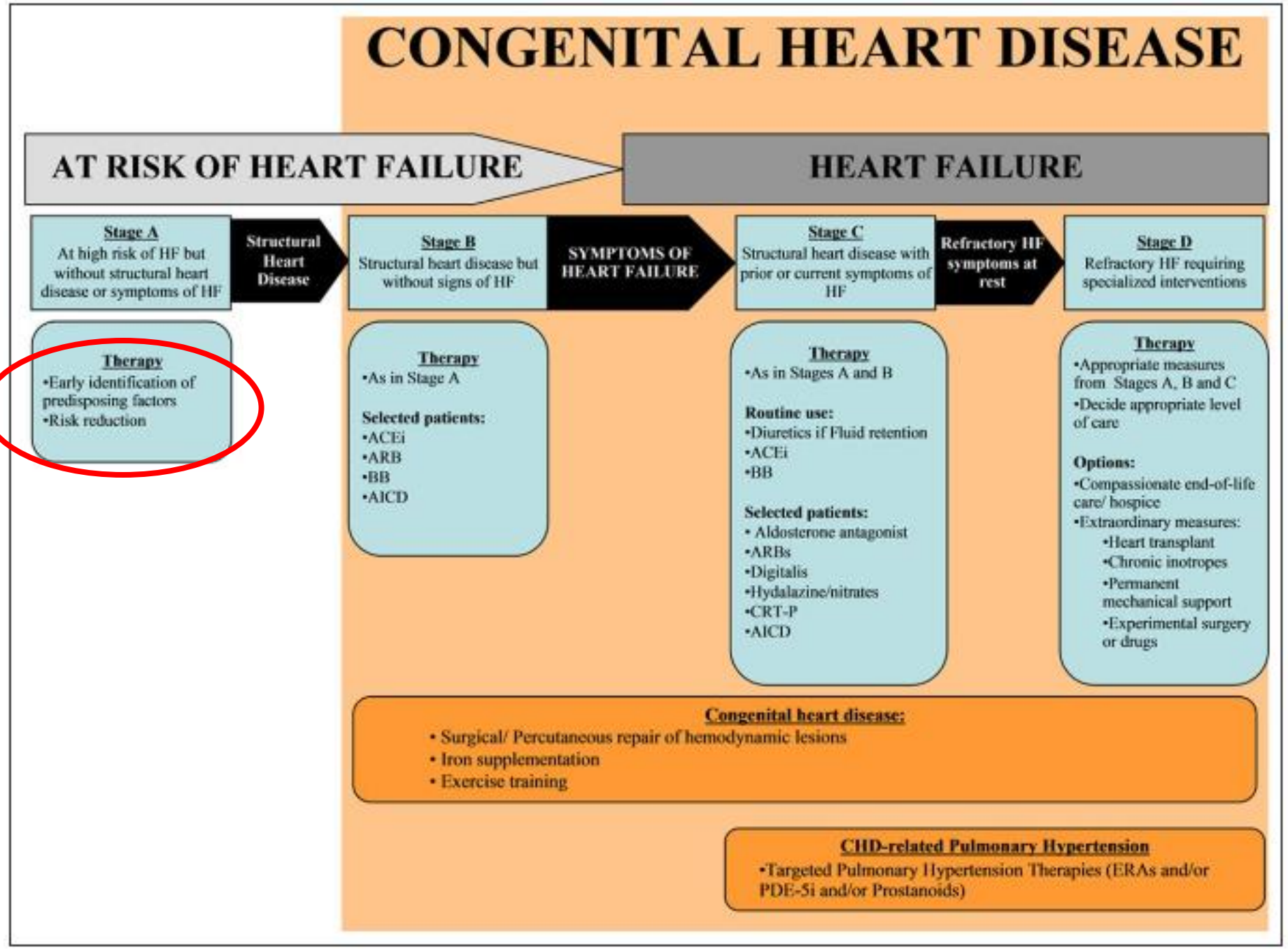


Fontan palliation



Univentricular Physiology – complications

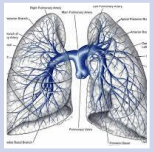
Unrepaired SV	Glenn palliation	Fontan palliation
Valvular dysfunction, ventricular impairment, arrhythmias, thrombo-embolic events Cyanosis, collaterals (venovenous/aortopulmonary)		
Heart failure		
	Lack of subpulmonary ventricle, systemic venous hypertension, SV preload deficiency	
	Fontan failure	



Stout et al Circulation 2016

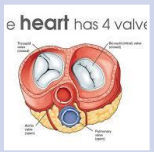
Determinants of well-functioning cardiac/fontan physiology

Sinus rhythm



CVP and PVR

Ventricular function



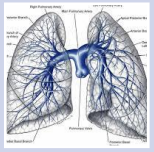
Valves

Anticipation & management of premature heart/fontan failure

Treat arrhythmias

Antiarrhythmic drugs, DCCV, EPS/ablation

?TCPC conversion



Avoid drugs/factors that increase PVR

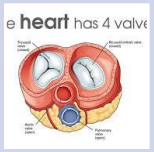
Consider pulmonary vasodilators

? Fontan fenestration

Avoid drugs with negative inotropic effects

Consider classical heart failure drugs

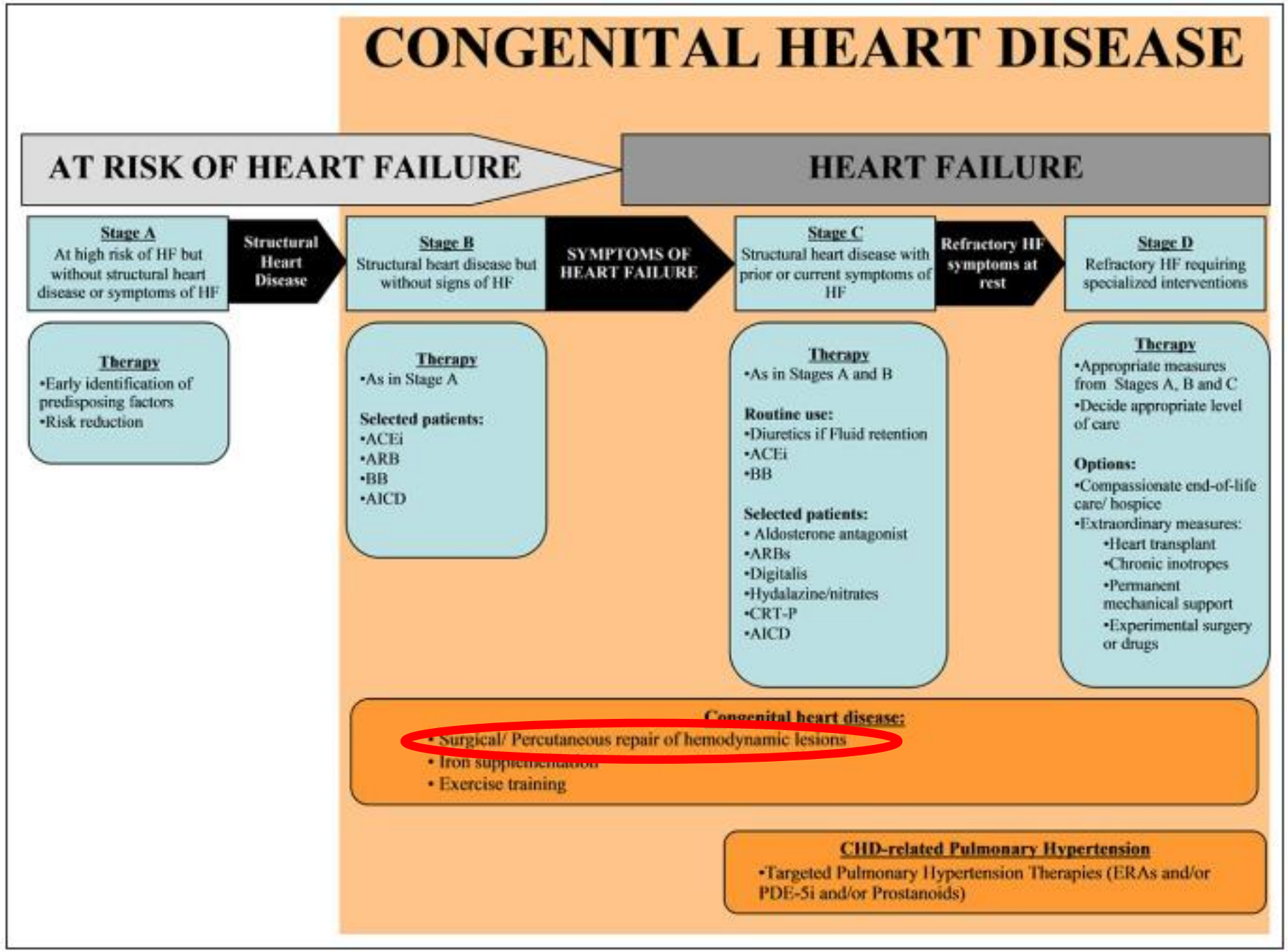
Consider coil embolisation AP collaterals



Surgical management – careful patient selection!

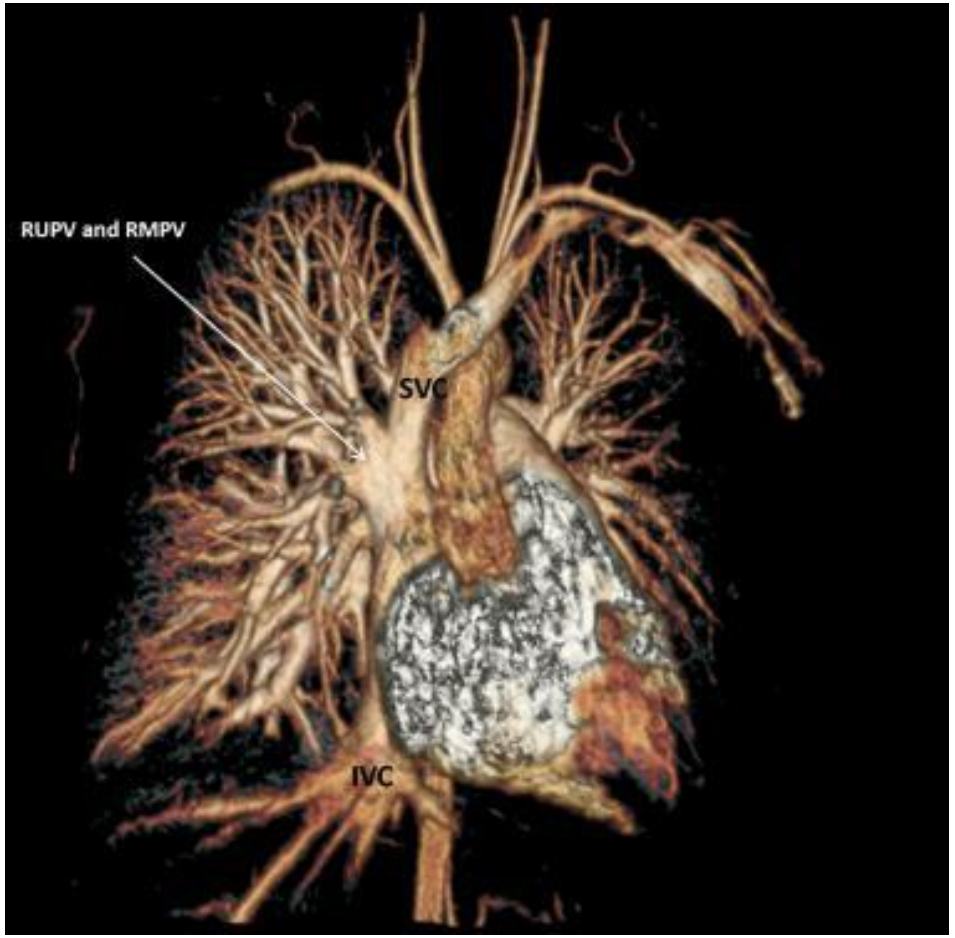
? Medical management

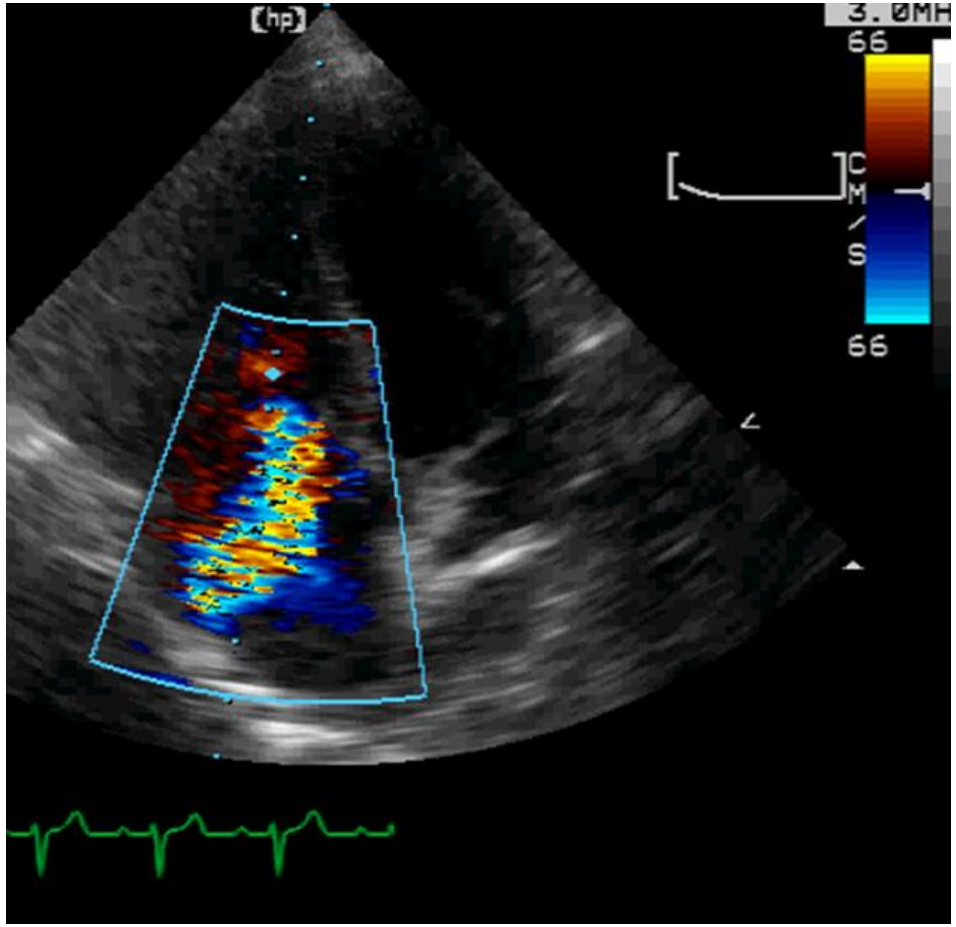
? Treatment of AP collaterals



Stout et al Circulation 2016

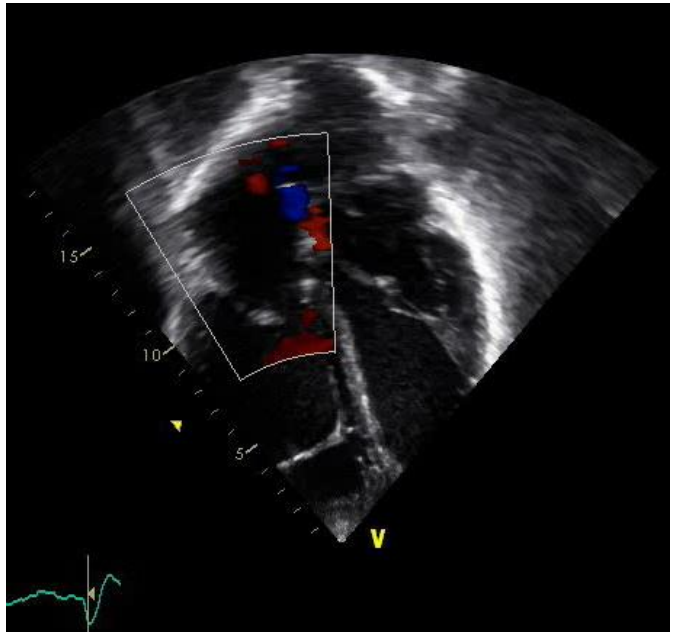
Late diagnosis – ? late repair



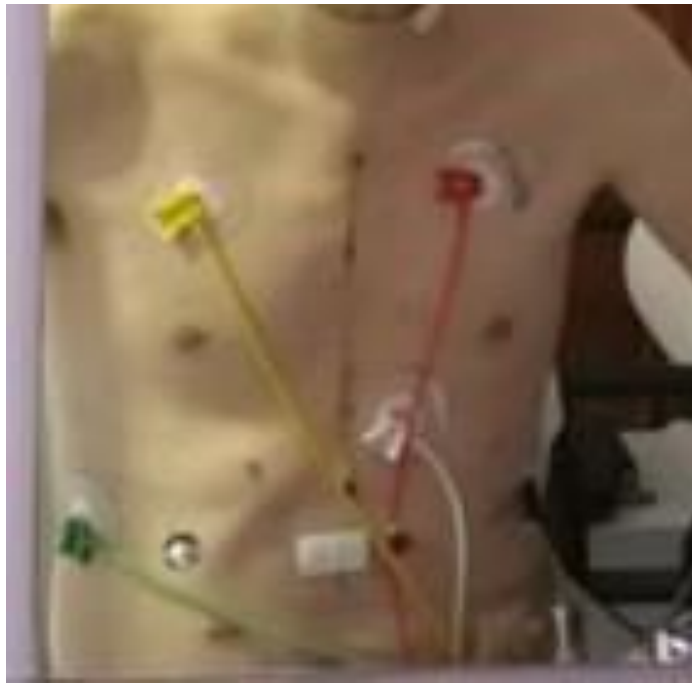


Other challenges in congenital heart failure

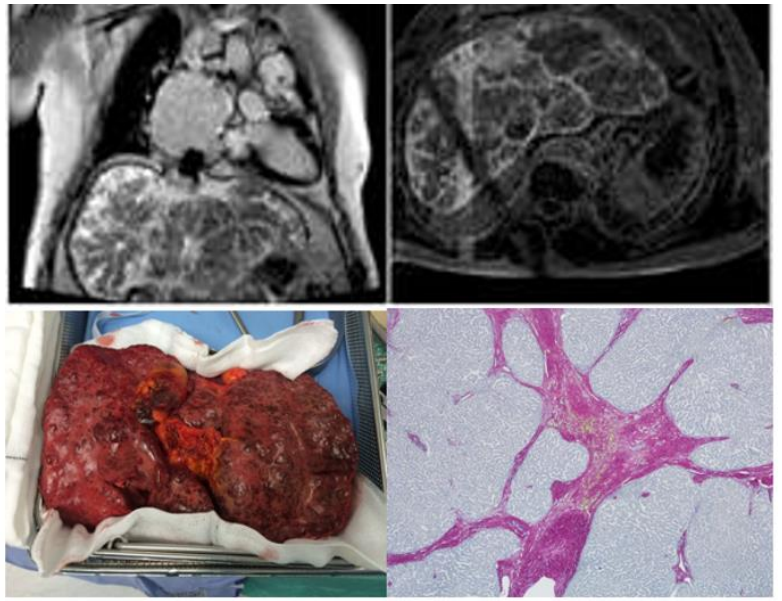
Functional/asymptomatic



Clinical/symptomatic



Fontan failure



Risk factors for death

- CONCOR registry
  - 1st HF admission 1.2 per 100 patient years
  - Admission with HF 5x risk of mortality
  - 1 year mortality after 1st HF admission 24%
  - 3 year mortality after 1st HF admission 35%

- Toronto ACHD-HF clinic - risk death/VAD/Tx
  - Lower NYHA
  - BNP >164pg/ml (BiV patients)
  - Serum sodium <136

- Other risk factors
  - Anaemia
  - Low albumin
  - Impaired VO2
  - Renal failure

Zomer et al International J Cardiology 2013

Roche et al Int J Cardiol 2018

The role of BNP in Fontan?

- BNP normal in majority of patients up to 15 years after Fontan completion
- Elevated and increasing BNP levels associated with increased morbidity and late mortality

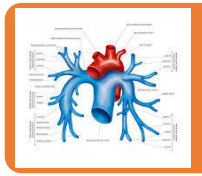
The majority of patients with cavopulmonary connection had normal BNP levels more than 15 years after surgery. In fact, the highest BNP level was less than 300 pg/ml. Comparable BNP levels have been reported in smaller groups composed of both Glenn and Fontan patients [13,14].

BNP is secreted mainly by the ventricles in response to volume expansion and pressure load [1,2]. Therefore, one possible explanation for the surprisingly low BNP levels is the reduced ventricular preload in patients with Fontan circulation together with a limited preload reserve [15,16].

Unfortunately, we were not able to correlate BNP levels with systolic function in our patients, because the broad spectrum of underlying cardiac malformation made it impossible to obtain reliable ejection fraction data by standard echocardiographic procedures. Nevertheless, typical complications in the long-term follow-up after the Fontan procedure were associated with higher BNP levels. A strong positive correlation was found between the severity of atrioventricular valve regurgitation and BNP. Moderate to severe atrioventricular valve regurgitation is a well known major risk factor influencing late mortality after total cavopulmonary connection [17]. In clinical practice, however, the predictive value of a

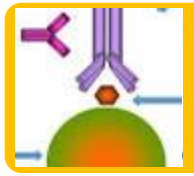
Koch et al. *Eur J Heart Failure 2008*

Transplant challenges

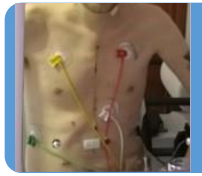


Technical aspects – complex anatomy

Bleeding risk (previous sternotomies, adhesions, FALD…)



HLA antibodies



Cardiac cachexia

Psychological aspects, social/family context

Increased demand for heart transplant - Shortage of donors

In summary

Growing population at risk of heart failure

Understanding of anatomy/physiology to understand mode of heart failure

Management:

- Anticipation of heart failure
- Early referral for advanced heart failure therapies

Two very important ‘tools’ when dealing with adults with CHD

Thank you!