



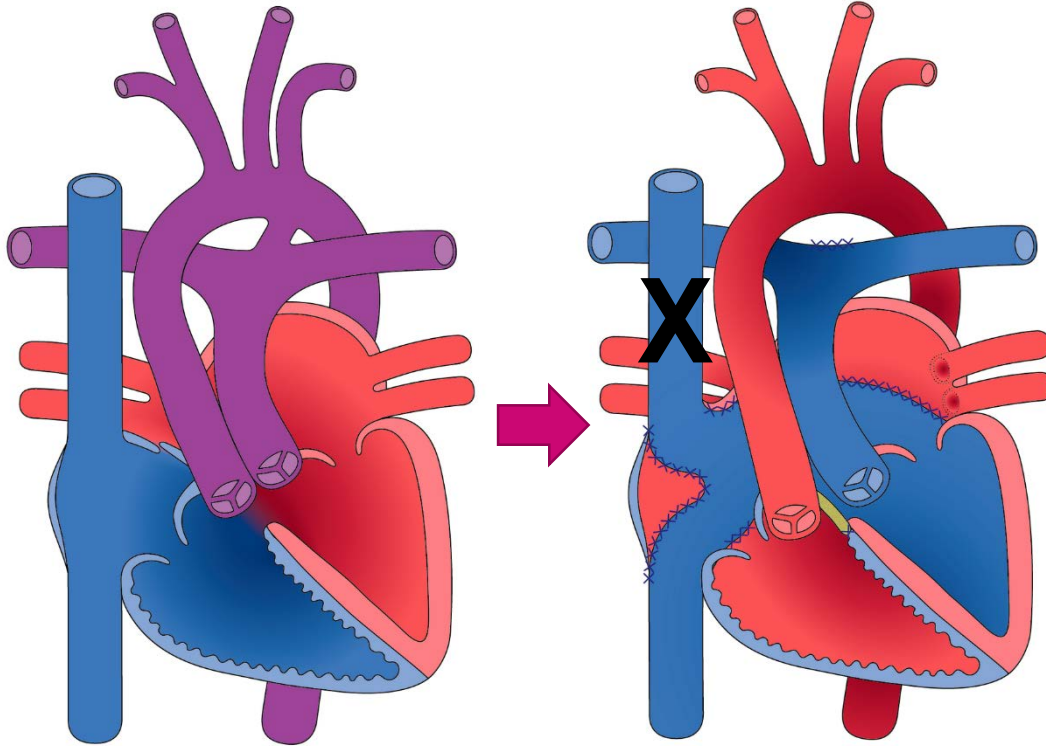
Heart failure medication in (A)CHD

Prof. Markus Schwerzmann





Case presentation - Anatomy



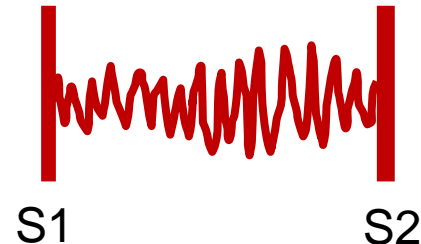
- Male, born 1967
- D-TGA with large VSD
- Septectomy and PA-banding
- Obstructed SVC
- “Modified” Mustard by Prof. Senning at age 8



Case presentation - 2013

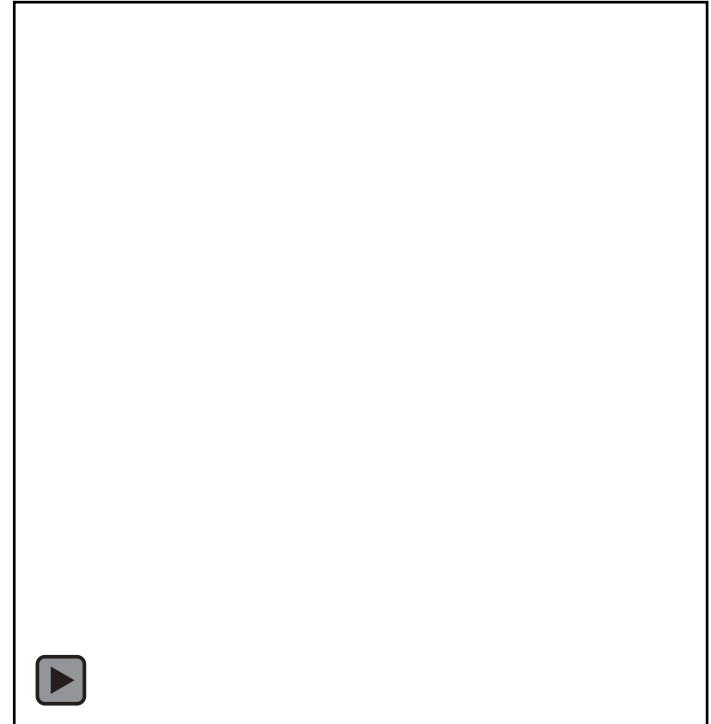


- referred for cardiac evaluation
- married, 2 kids, engineer
- very sportive, healthy lifestyle
- athletic guy, BMI: 22 kg/m²
- BP 116/68 mmHg, HR 62 bpm
- no signs of left or right heart failure





Case presentation - 2013





Case presentation - 2013

- Cardiac MRI 2013: RVEDVi 212 ml/m²; RV-EF 35%; RF of TI 51%; aortic sinus 50 mm; Qp:Qs 1.2
- Cardiopulmonary exercise testing: 240 Watts; peak VO₂ 26 ml/kg/min; normal heart rate response with peak HR 172 bpm

A) Is this heart failure?

B) Does this patient need heart failure medication?



HF according to guidelines...

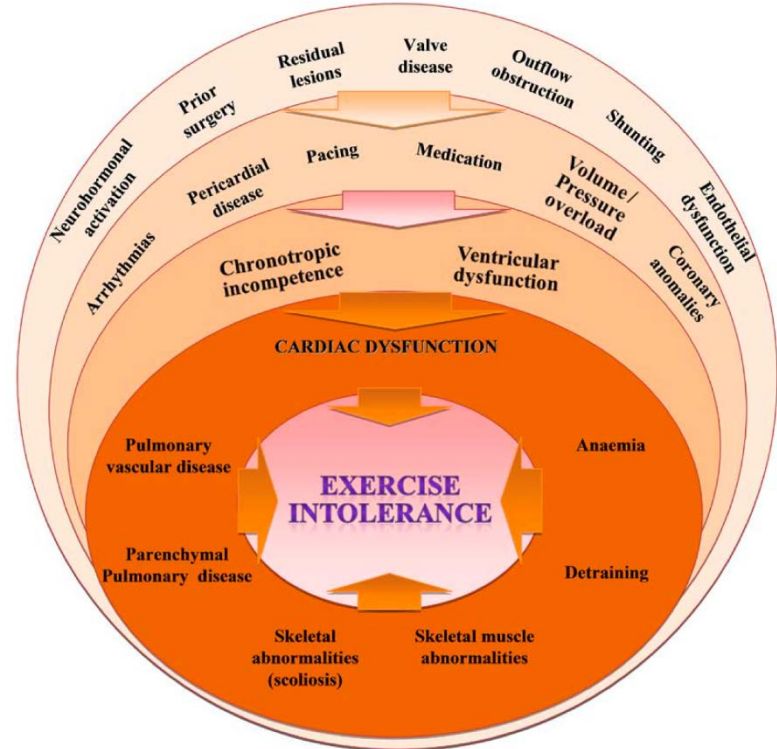
- HF is a **clinical syndrome** characterized by typical symptoms and signs
- **Pathophysiology:** Heart is unable to pump blood at a sufficient rate to meet metabolic demands, or only if the filling pressures are abnormally high
- Asymptomatic cardiac abnormalities (systolic or diastolic ventricular dysfunction): precursors of HF.

Eur Heart J 2016;**37**:2129-200.



HF in ACHD – not defined, because...

- most patients claim to be asymptomatic
- nearly all have structural heart disease
- Objective measures are less reliable
 - reduced exercise capacity is frequent
 - correlation between neurohumoral activation and outcome is lesion specific
- Confounders: cyanosis, syndromic defects





Guidelines

- ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure. Eur Heart J 2016;**37**(27):2129-200
- Chronic Heart Failure in Congenital Heart Disease: A Scientific Statement From the American Heart Association. Circulation 2016;**133**(8):770-801
- Treatment of heart failure in adult congenital heart disease: a position paper of the Working Group of Grown-Up Congenital Heart Disease and the Heart Failure Association of the European Society of Cardiology. Eur Heart J 2016;**37**(18):1419-27



Acquired left heart failure

- Heart failure with preserved ejection fraction (HFpEF): diastolic heart failure
- Heart failure with reduced ejection fraction (HFrEF): systolic heart failure

LV-EF > 50% or < 40%; elevated natriuretic peptides; symptoms

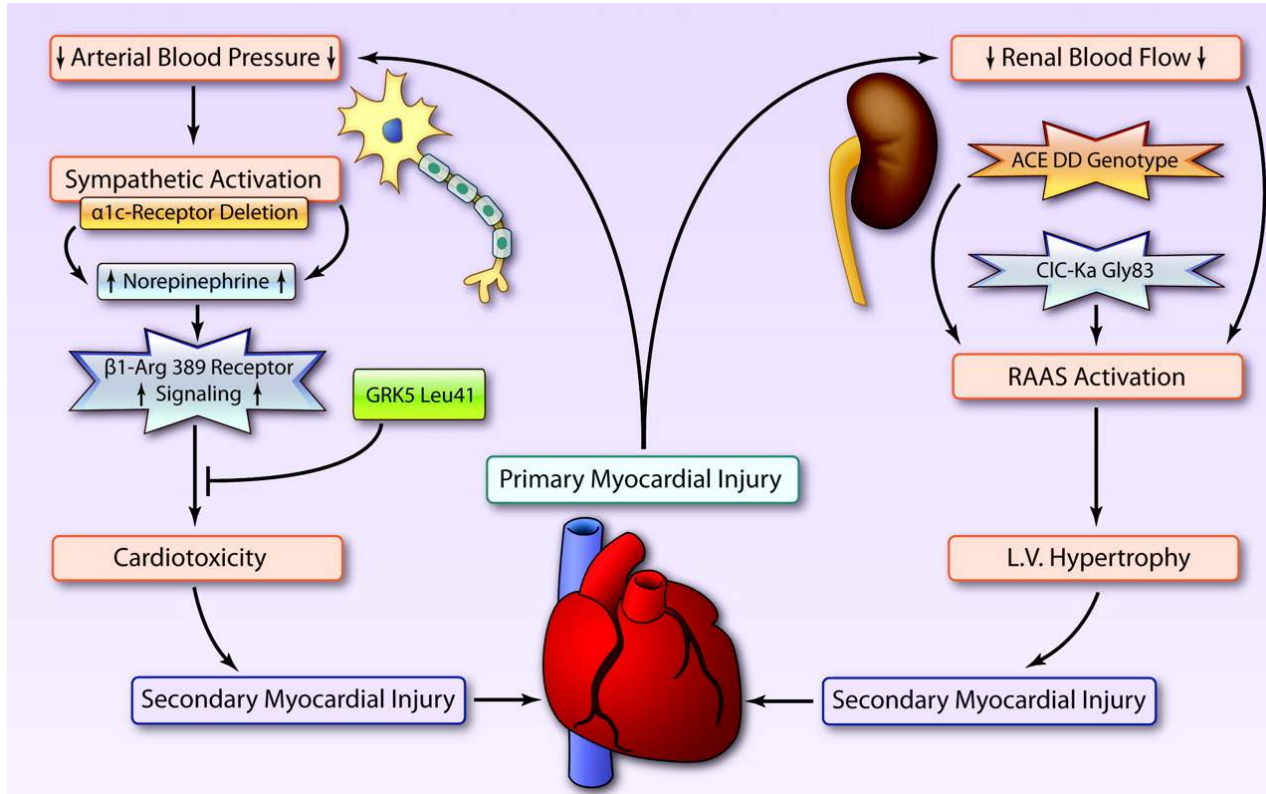
Distinctive medical therapies for HFrEF with impact on prognosis:

- Betablockers, ACE-inhibitor, Mineralocorticoid receptor antagonist, Ivabradine (Procoralan), Angiotensin Rezeptor Neprilysin Inhibitor (Entresto); cardiac resynchronization therapy

Medical therapies for HFpEF with impact on prognosis: ...

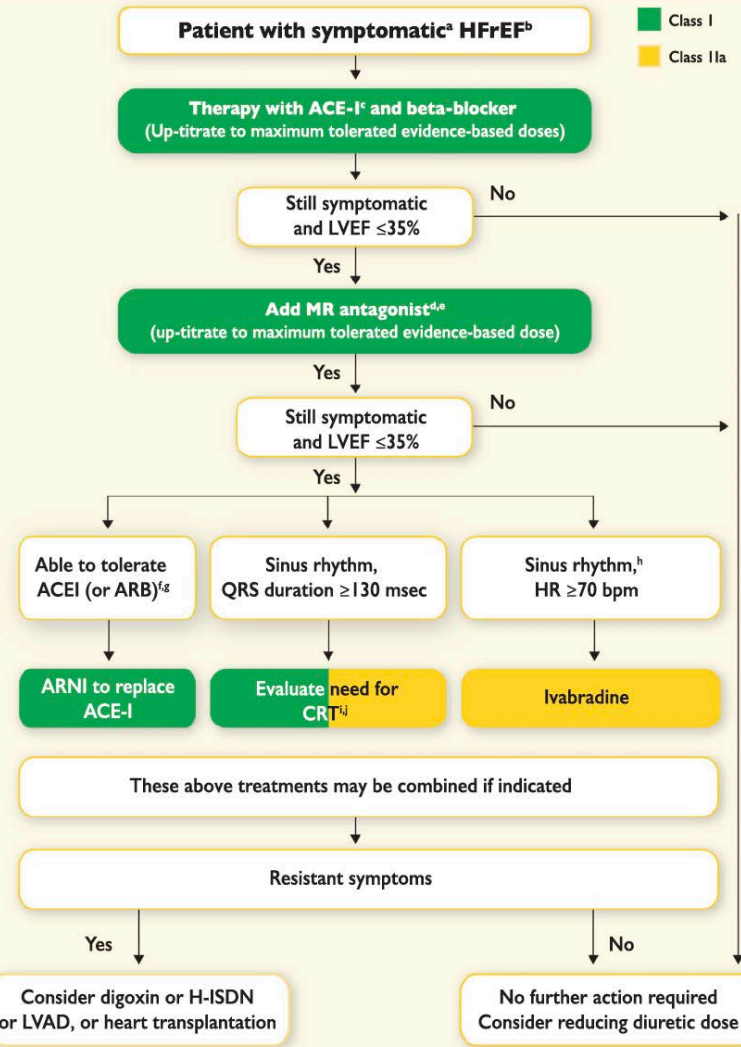


Neurohumoral activation



Diuretics to relieve symptoms and signs of congestion

If LVEF \leq 35% despite OMT
or a history of symptomatic VT/VF, implant ICD



ESC Guidelines 2016

	Starting dose (mg)	Target dose (mg)
ACE-I		
Captopril ^a	6.25 t.i.d.	50 t.i.d.
Enalapril	2.5 b.i.d.	10–20 b.i.d.
Lisinopril ^b	2.5–5.0 o.d.	20–35 o.d.
Ramipril	2.5 o.d.	10 o.d.
Trandolapril ^a	0.5 o.d.	4 o.d.
Beta-blockers		
Bisoprolol	1.25 o.d.	10 o.d.
Carvedilol	3.125 b.i.d.	25 b.i.d. ^d
Metoprolol succinate (CR/XL)	12.5–25 o.d.	200 o.d.
Nebivolol ^f	1.25 o.d.	10 o.d.
ARBs		
Candesartan	4–8 o.d.	32 o.d.
Valsartan	40 b.i.d.	160 b.i.d.
Losartan ^{b,c}	50 o.d.	150 o.d.
MRA s		
Eplerenone	25 o.d.	50 o.d.
Spironolactone	25 o.d.	50 o.d.
ARNI		
Sacubitril/valsartan	49/51 b.i.d.	97/103 b.i.d.
If-channel blocker		
Ivabradine	5 b.i.d.	7.5 b.i.d.



HFpEF

Recommendations for treatment of patients with heart failure with preserved ejection fraction and heart failure with mid-range ejection fraction

Recommendations	Class ^a	Level ^b	Ref ^c
it is recommended to screen patients with HFpEF or HFmrEF for both cardiovascular and non-cardiovascular comorbidities, which, if present, should be treated provided safe and effective interventions exist to improve symptoms, well-being and/or prognosis.	I	C	
Diuretics are recommended in congested patients with HFpEF or HFmrEF in order to alleviate symptoms and signs.	I	B	178, 179

HFmrEF = heart failure with mid-range ejection fraction; HFpEF = heart failure with preserved ejection fraction.

^aClass of recommendation.

^bLevel of evidence.

^cReference(s) supporting recommendations.





HFpEF

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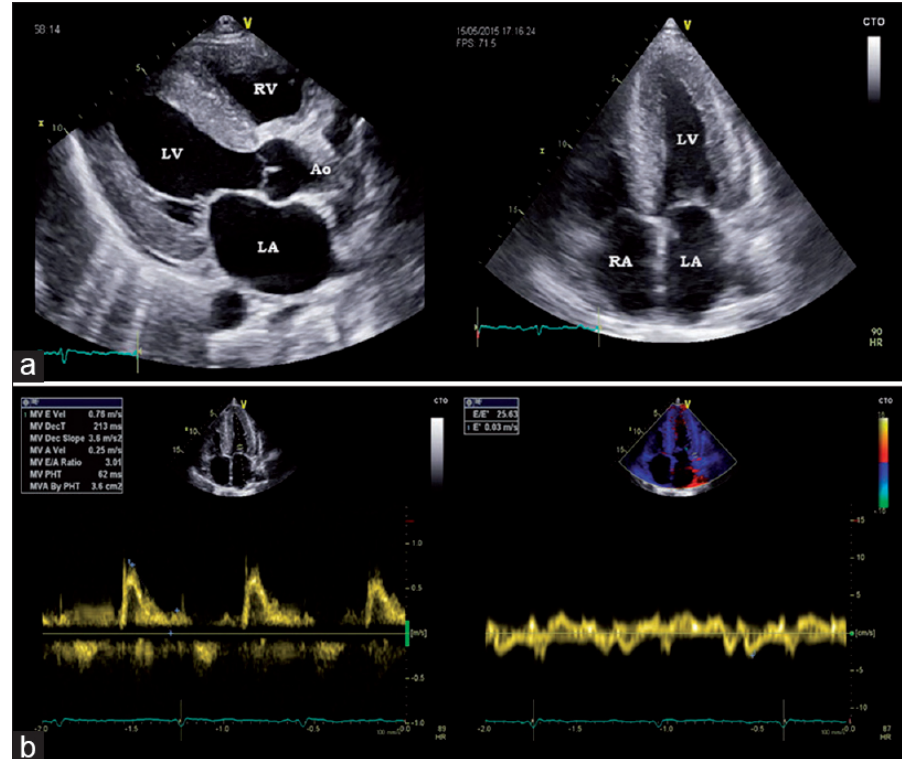
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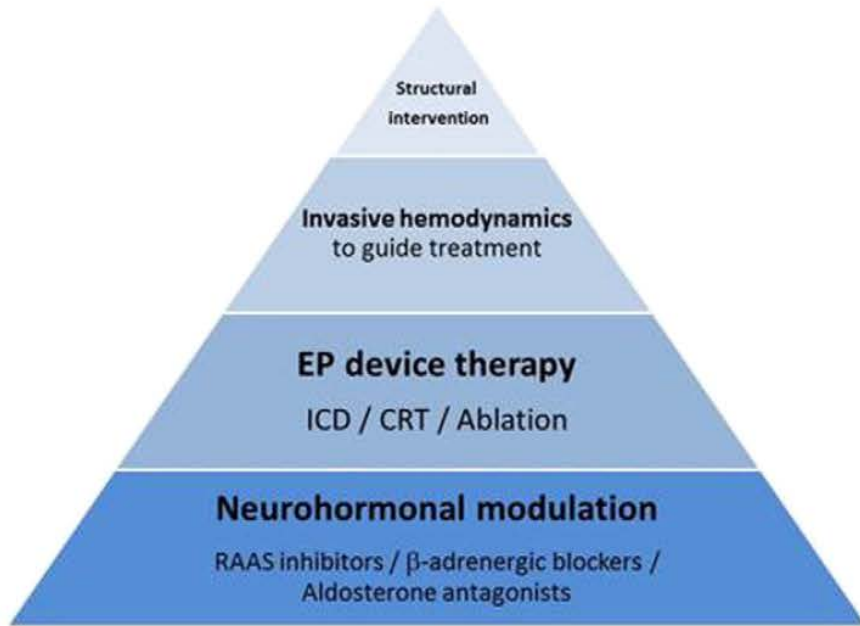
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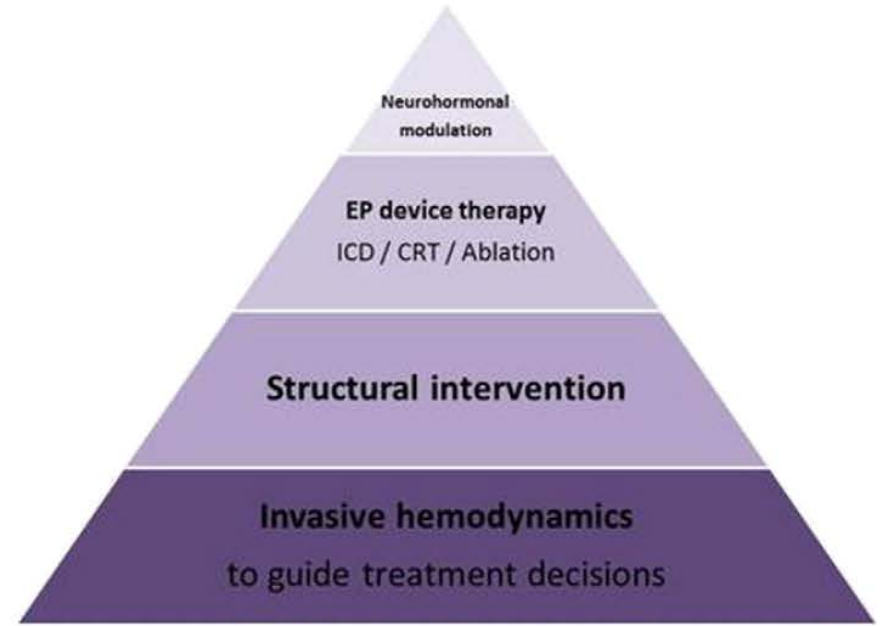




The heart failure concept in ACHD



A) Heart failure treatment in acquired heart failure



B) Heart failure treatment in ACHD



Heart failure in ACHD

