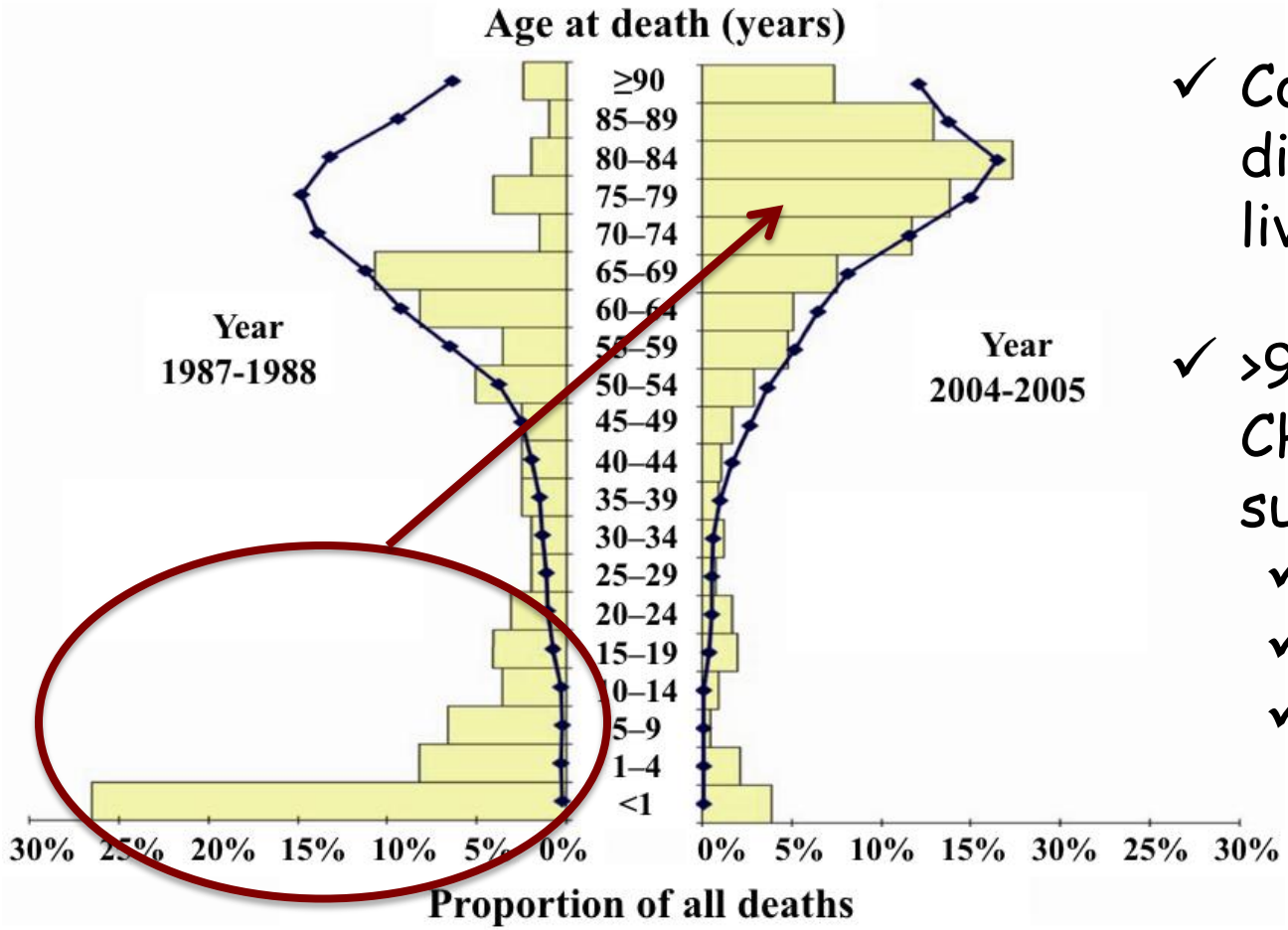
Primární a sekundární prevence náhlé srdeční smrti u vrozených srdečních vad

Roman A. Gebauer

HERZZENTRUM
LEIPZIG

ADULTS WITH CHD: A GROWING POPULATION

71,686 pts with CHD followed for 982,363 patient-years.



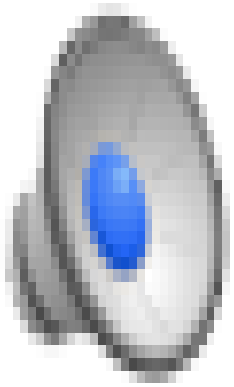
- ✓ Congenital heart disease: ~9 per 1000 live births
- ✓ >90% of children with CHD are expected to survive to adulthood:
  - ✓ USA: >1,000,000
  - ✓ Canada: >100,000
  - ✓ Europe: 1,800,000

Khairy P et al. JACC 2010

Sudden cardiac death

- death due to a cardiovascular cause within one hour of the onset or significant worsening of symptoms, or unwitnessed death in the absence of a known non-cardiac condition as the proximate cause of death.

✓ Arrhythmic SCD encompasses death due to documented or presumed arrhythmias



Mortality rates after surgical repair of CHD

Authors	Years	Patients	Deaths	SCD	CHF	Other CV	Non-cardiac
Oeschlin et al	1981-1996	2609	197	26%	21%	34%	18%
Silka et al	1958-1996	3589	176	23%	13%	35%	12%
Nieminen et al	1953-1998	5919	582	15%	27%	31%	8%
Verheugt et al	2001-2009	6933	197	19%	26%	32%	23%
Zomer et al	2001-2010	8595	231	22%	26%	29%	24%
Total		*27,645*	*1,383*	*19%*	*24%*	*36%*	*15%*

Sudden cardiac death ≈ 20 - 25 % of all late deaths in GUCH

Annual incidence of SCD < 0.1% in GUCH

Journal of the American College of Cardiology
© 2008 by the American College of Cardiology Foundation and American Heart Association, Inc.
Published by Elsevier Inc.

Vol. 52, No. 23, 2008
ISSN 0735-1097/08/$34.00
doi:10.1016/j.jacc.2008.10.001

PRACTICE GUIDELINE: FULL TEXT

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

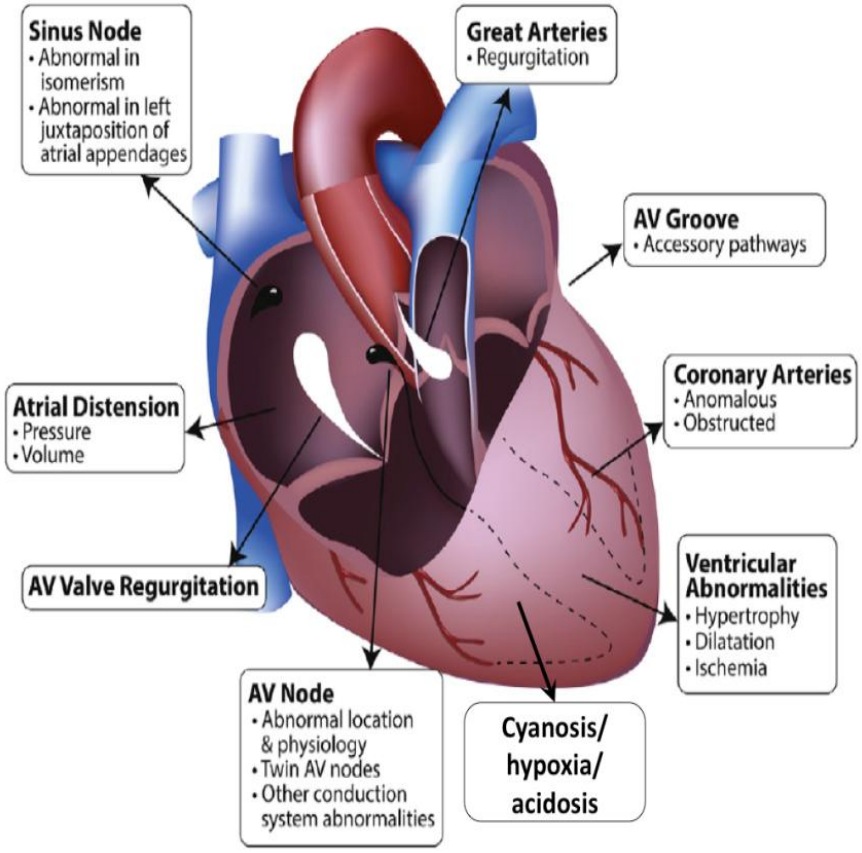
PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD).

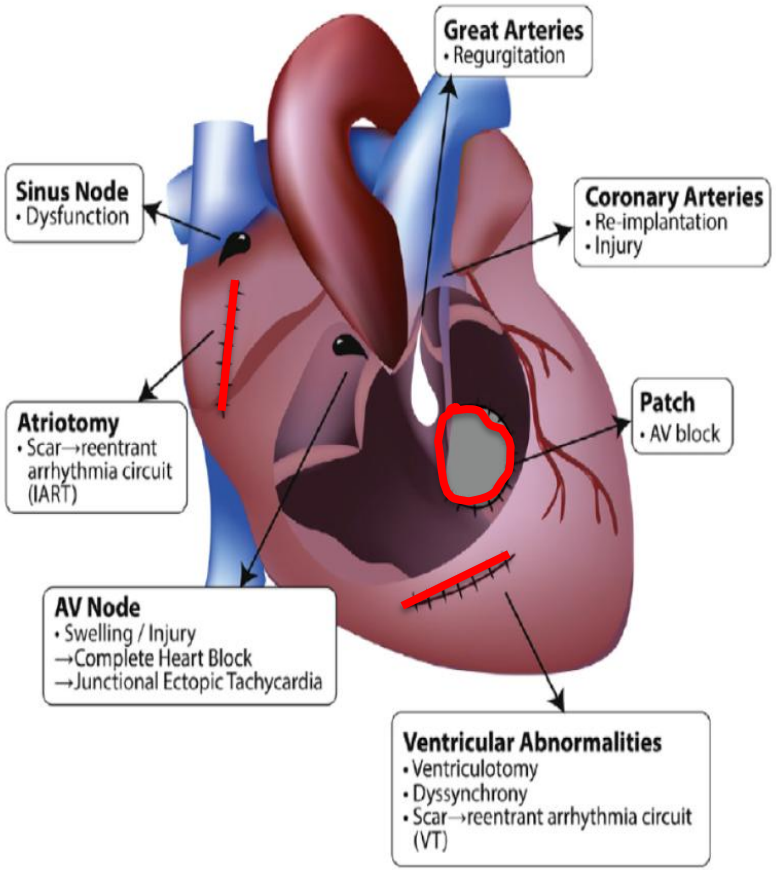
Khairy P et al. Heart Rhythm 2014

FACTORS LEADING TO (LATE) ARRHYTHMIAS IN CHD

Pre-operative



Post-operative



HERZZENTRUM LEIPZIG

Adapted from Escudero C et al. Can J Cardiol 2013

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease, HeartRhythm, 2014

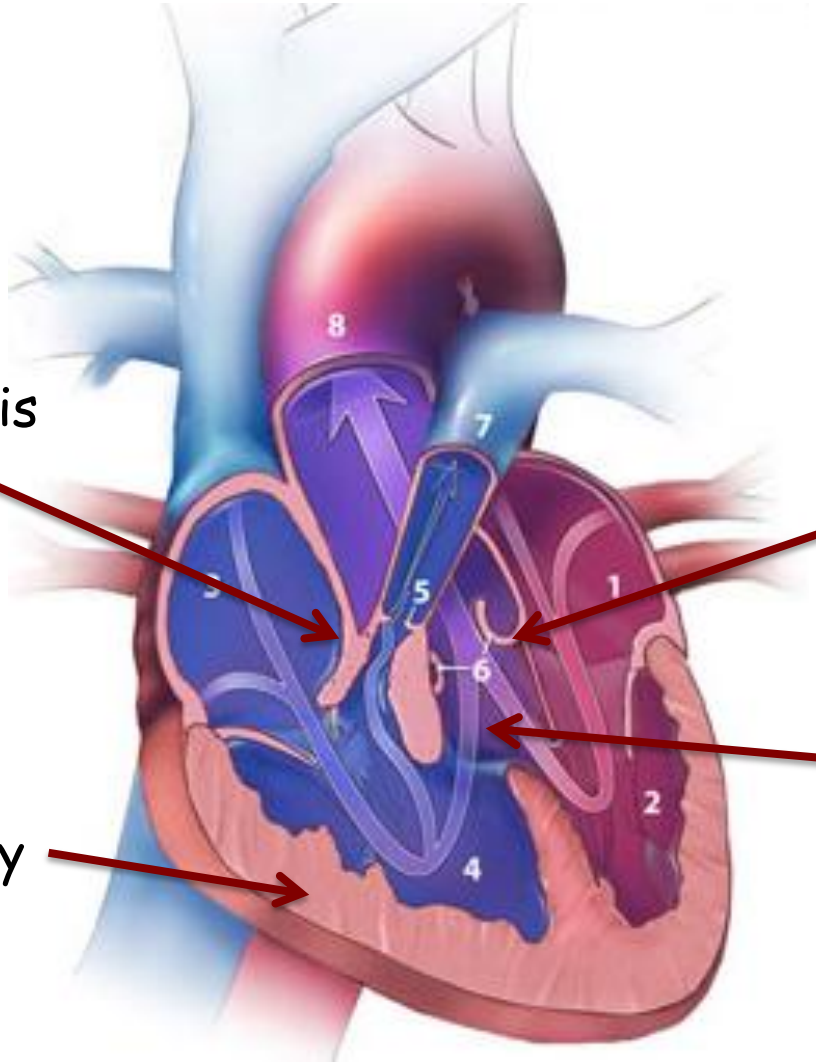
APPROXIMATE RISK ESTIMATES of ARRHYTHMIAS

Complexity of CHD	Type of CHD	Prevalence (in CHD population)	Atrial Arrhythmia			Ventricular Arrhythmia	Other Pacing Needs		
			AT	AF	Other		SND	AV block	Dyssynchrony, heart failure
Simple	Patent ductus arteriosus	6-8%							
	Pulmonary stenosis	6-8%							
	Ventricular septal defect	30-32%							
	Secundum atrial septal defect	8-10%							
Moderate	Aortic coarctation	5-7%							
	Anomalous pulmonary venous return	0.5-2.5%							
	Atrioventricular septal defect	3-5%							
	Aortic stenosis	3-5%							
	Ebstein's anomaly	0.5-1.5%							
	Tetralogy of Fallot	8-10%							
	[illegible] atrial septal defect	2-3%							
Severe	Truncus arteriosus	1.5-2%							
	Pulmonary atresia	2-2.5%							
	DORV	1.5-2%							
	D-Transposition of GA	6-7%							
	L-Transposition of GA	1-2%							
	Hypoplastic left heart syndrome	3-4%							
	Other (heterotaxy, other single ventricles)	7-10%							

Tetralogy of Fallot

Congenital Heart Defect of Moderate Complexity

Warnes CA et al., ACC/AHA 2008 Guidelines, Circulation 2008



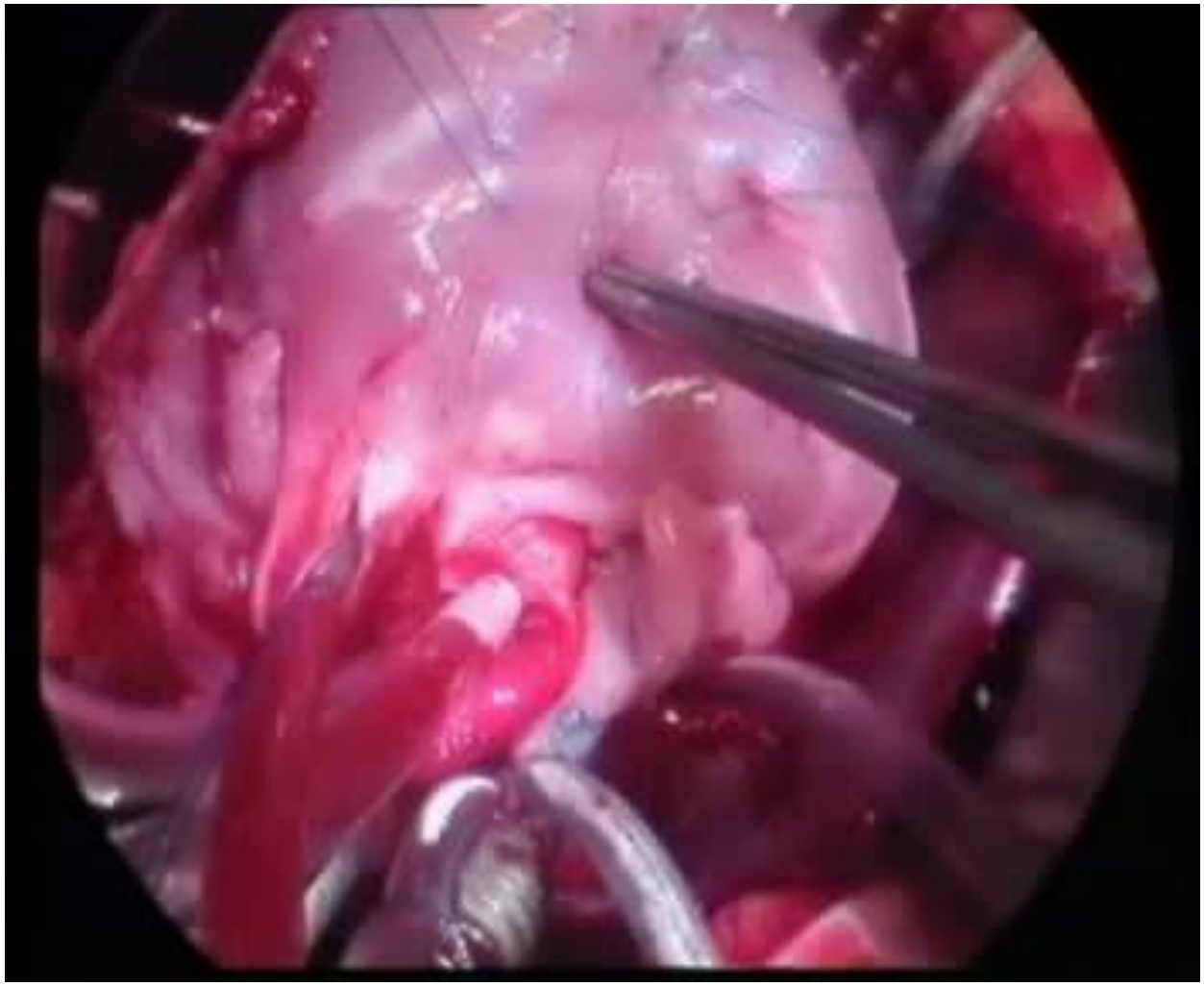
Pulmonary stenosis

Overriding aorta

Ventricular septal defect

RV hypertrophy

ToF repair in infancy



Courtesy prof. Martin Kostelka

Update on Interventional Electrophysiology in Congenital Heart Disease

Evolving Solutions for Complex Hearts

Elizabeth D. Sherwin, MD; John K. Triedman, MD; Edward P. Walsh, MD

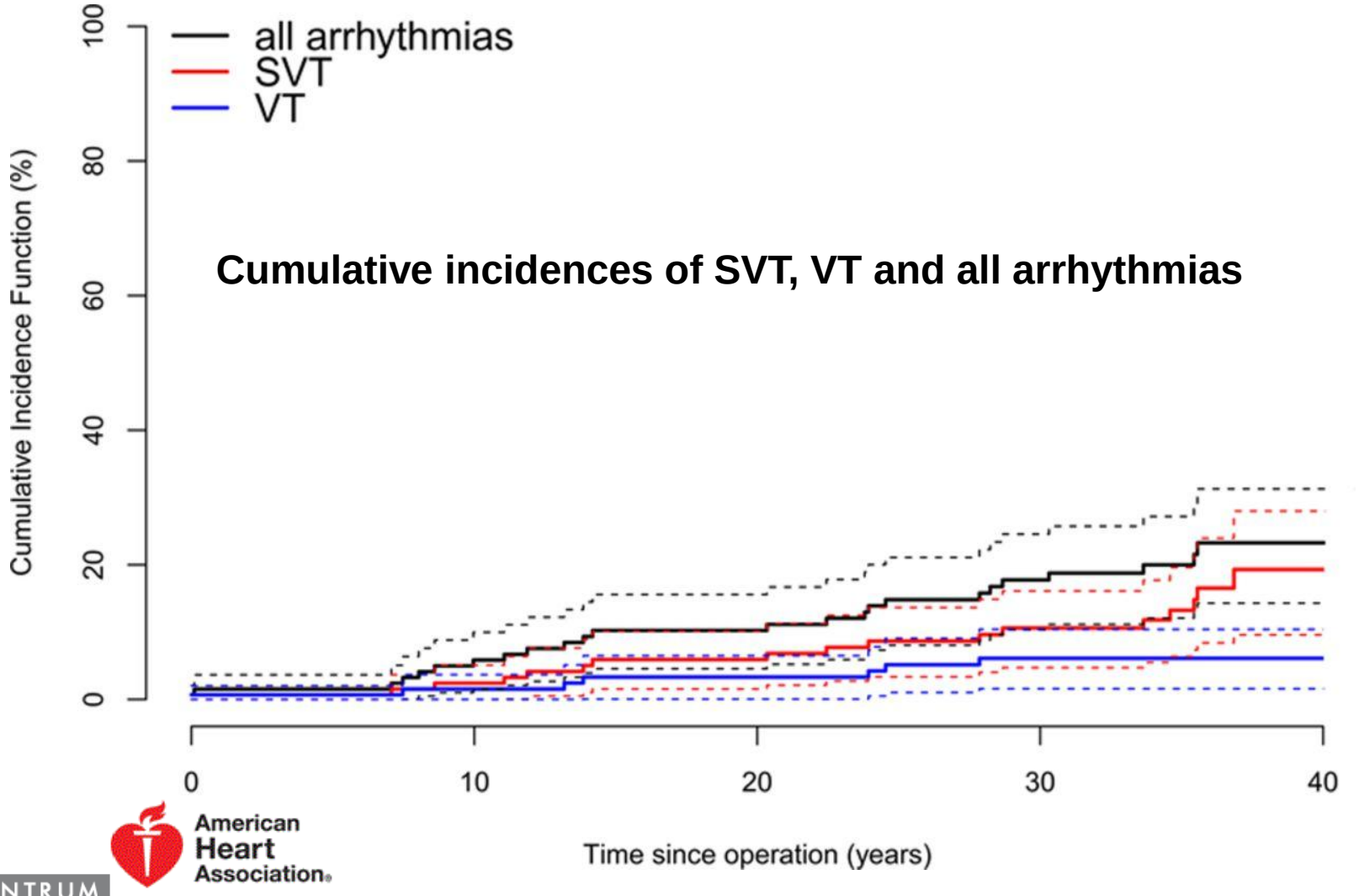
VT/VF substrates in repaired ToF

- Discrete corridors of slow conduction supporting ≥1 monomorphic reentrant VT circuits
  - Consequence of surgery
- Diffusely abnormal myocardium leading to disorganized polymorphic VT and VF
  - Consequence of pressure/volume overload and cyanosis

Circ Arrhythm Electrophysiol, 2013

Consequencies of ToF repair

Arrhythmias



Cumulative incidences of SVT, VT and all arrhythmias

American Heart Association.

HERZZENTRUM LEIPZIG

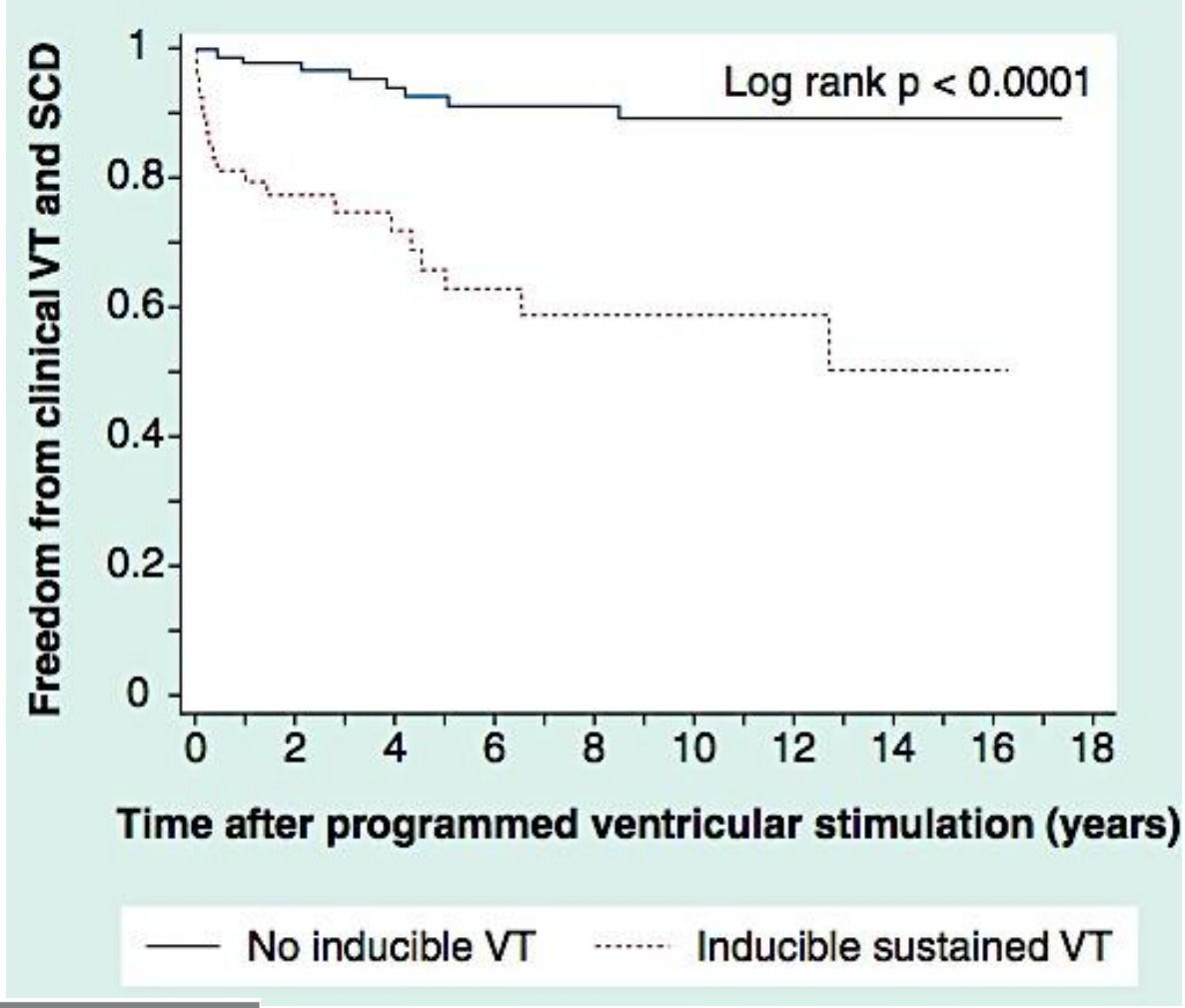
Cuypers J A et al. Circulation. 2014;130:1944-1953

Copyright © American Heart Association, Inc. All rights reserved.

How to identify the patients at risk of SCD???

Value of Programmed Ventricular Stimulation After TOF Repair

A Multicenter Study

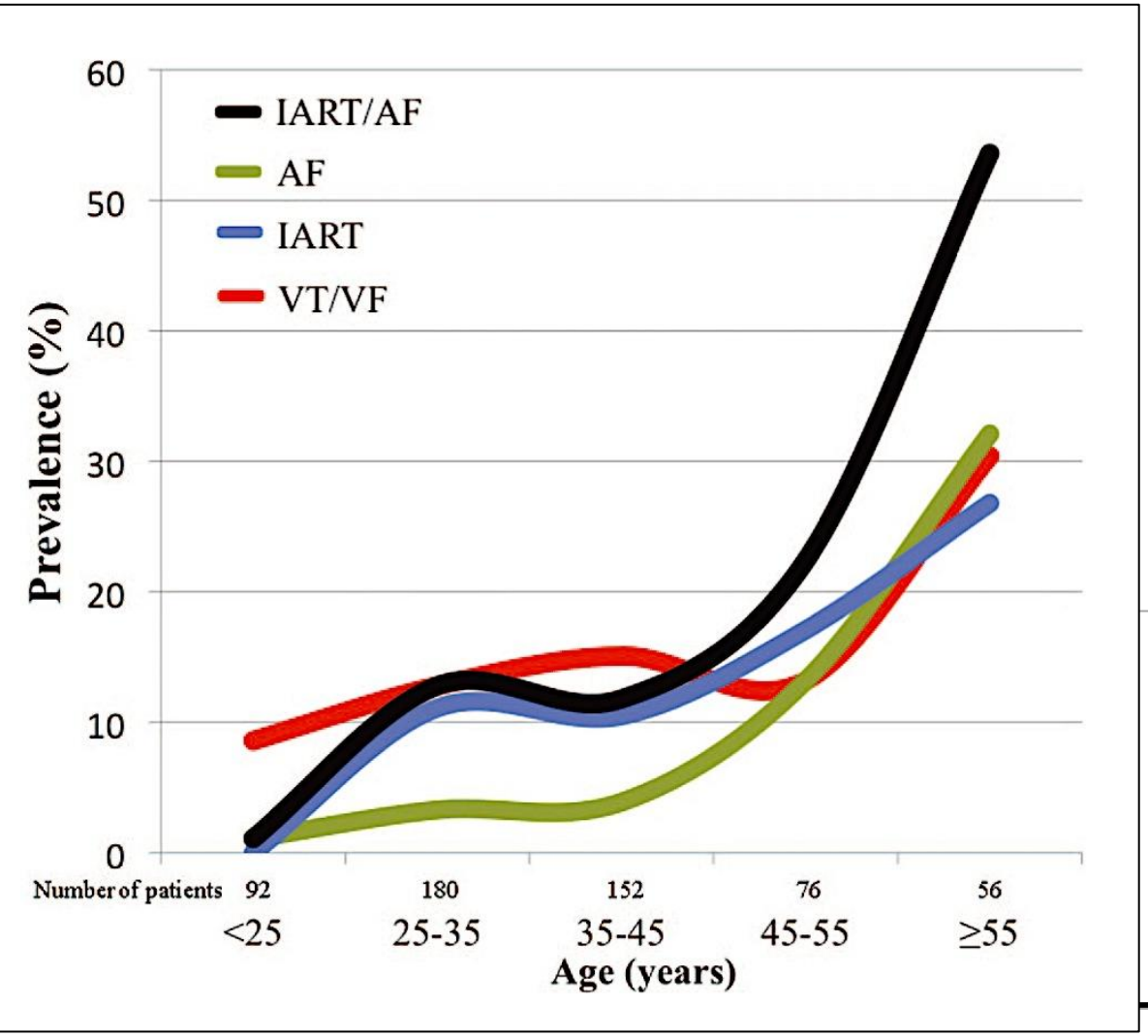


- ✓n=252 pts.
- ✓Δt after surgery 18.5 yrs
- ✓Δt post EPS 6.5 yrs

HERZZENTRUM LEIPZIG

Khairy P et al., Circulation 2004

Arrhythmia Burden in Adults With Surgically Repaired ToF



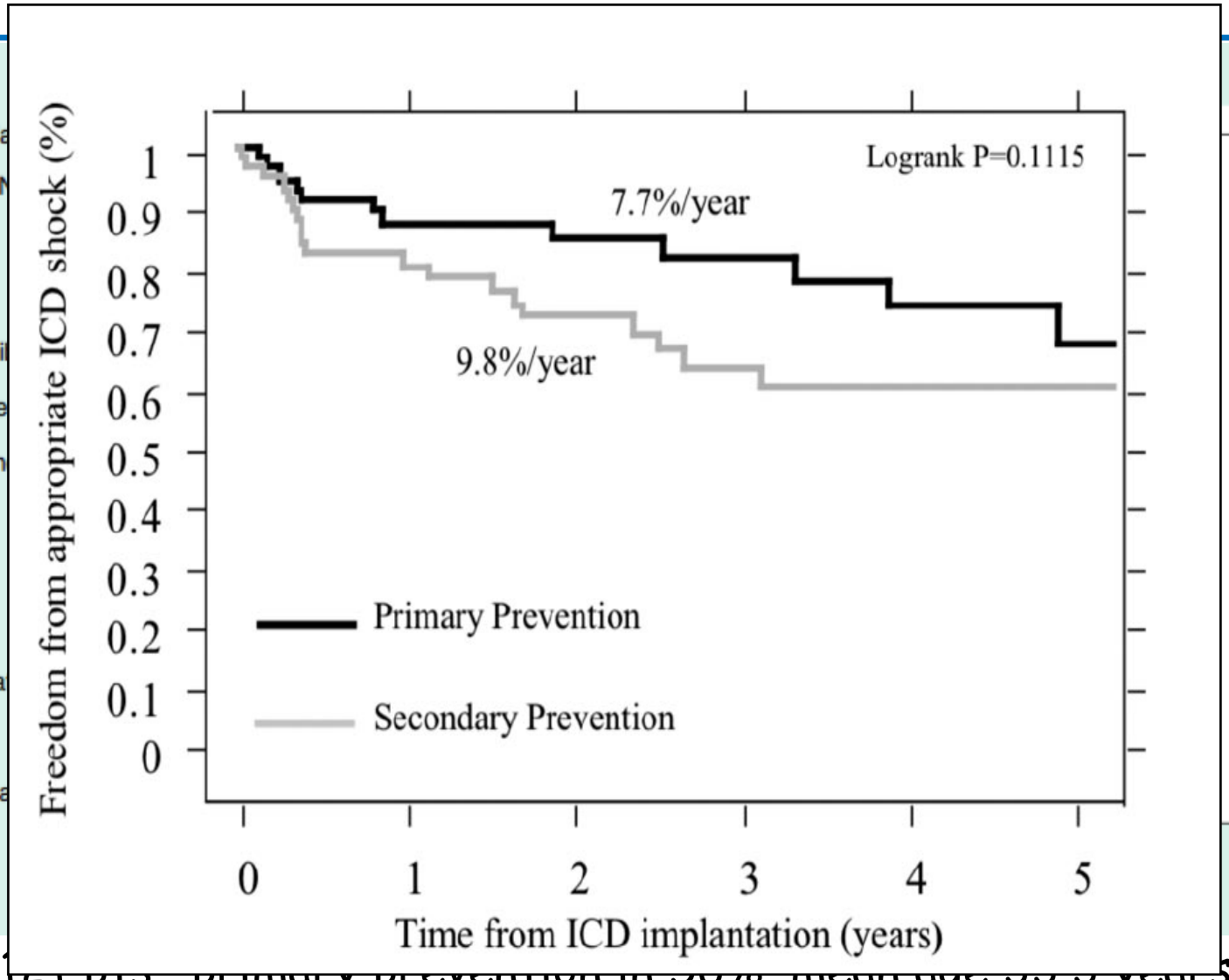
Khairy P et al., Circulation 2010

- ✓ N=556,
- ✓ 11 centers
- ✓ mean age 36.8+12 years
- ✓ sustained VT/VF in 14,6 %

1.3	1.1–1.6	0.005
1.02	1.01–1.03	0.014
3.3	1.5–7.1	0.002

ICD in Tetralogy of Fallot

Khairy P et al., Circulation 2008



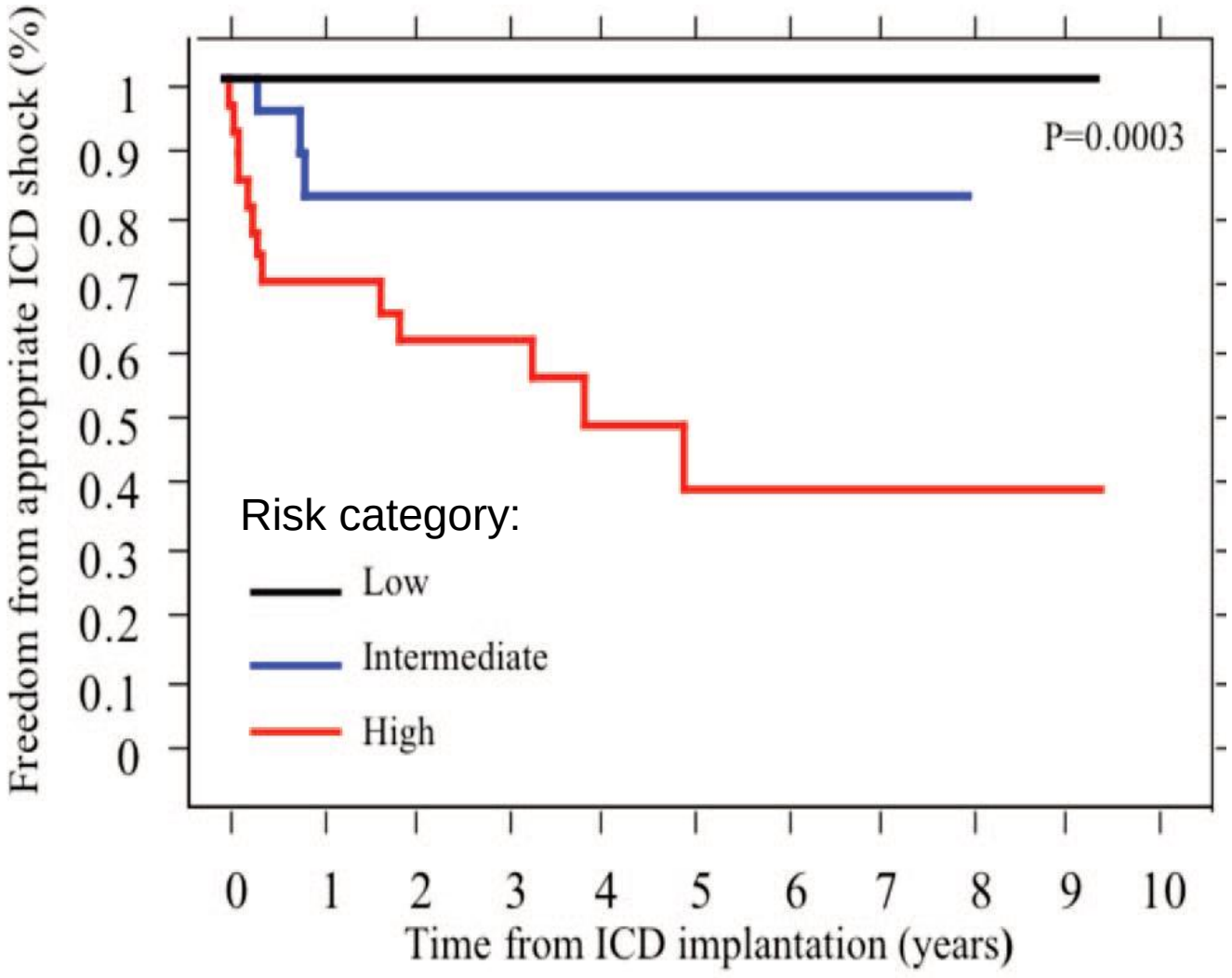
n=121 pts., primary prevention in 56%, mean age 35.5 years,

Δt post ICD 3.7 years, discharge in 9.8% pts/year (SP)

Risk stratification for malignant ventricular arrhythmia after ToF repair

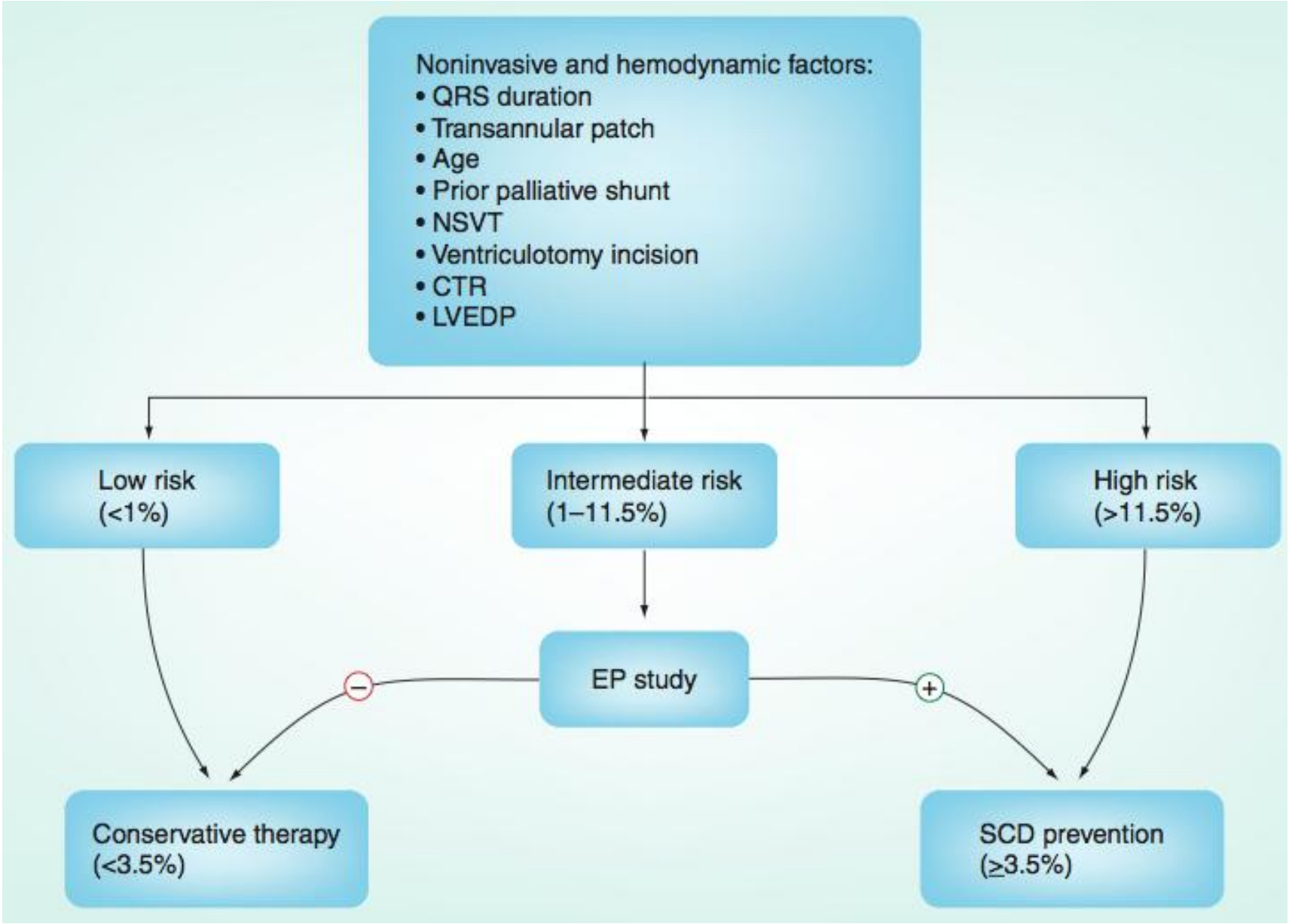
Table 3. Ris
Prevention

Variable
Prior palliative s
Inducible sustai
QRS duration ≥
Ventriculotomy
Nonsustained v
LVEDP ≥12 mr
Total points



Khairy P et al., Circulation 2008

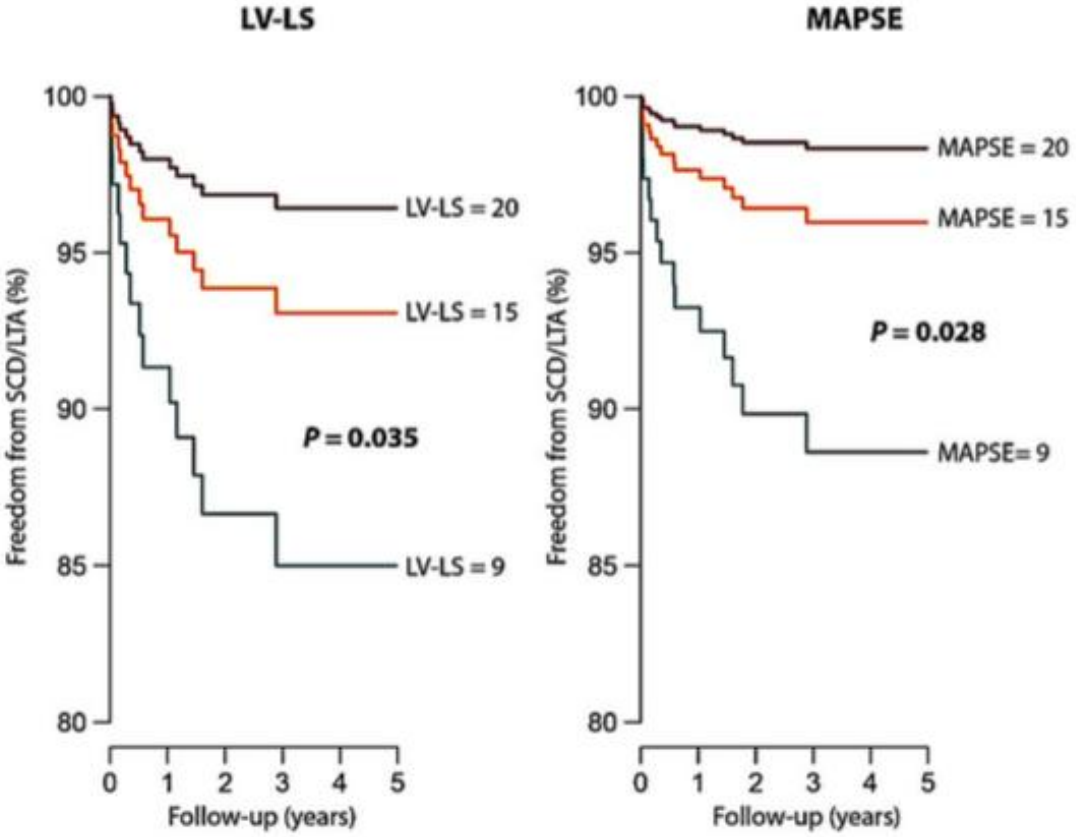
Risk Stratification Approach in TOF



Khairy P, Expert Rev Cardiovasc Ther 2009

Left Ventricular Longitudinal Function Predicts Life-Threatening Ventricular Arrhythmia and Death in Adults with Repaired Tetralogy of Fallot

Diller GP et al., Circulation 2012



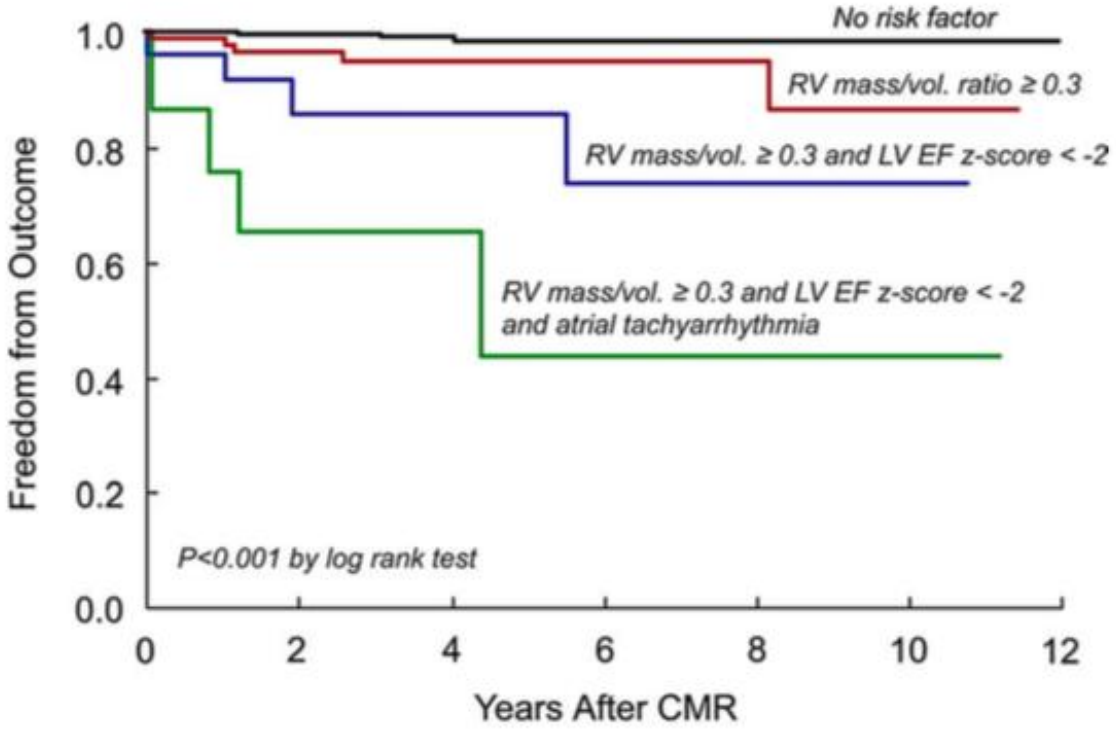
N=413, mean age 36.8+13 years, QRS duration 148+27 ms, LVEF 55+10%

Mean follow-up 2.9 years: SD n=5, sustained VT n=9, appropriate ICD shock n=5

Annual probability of VT/SD 2.4%

Contemporary Predictors of Death and Sustained VT in Repaired TOF Patients Enrolled in the INDICATOR Cohort

Valente AM et al., Heart 2014



No. at Risk:						
No risk factors	435	249	113	22	2	1
RV mass/vol. ≥ 0.3	96	69	43	29	13	7
RV mass/vol. ≥ 0.3 + LVEF z < -2	26	16	11	7	6	3
All three factors	15	6	5	2	1	1

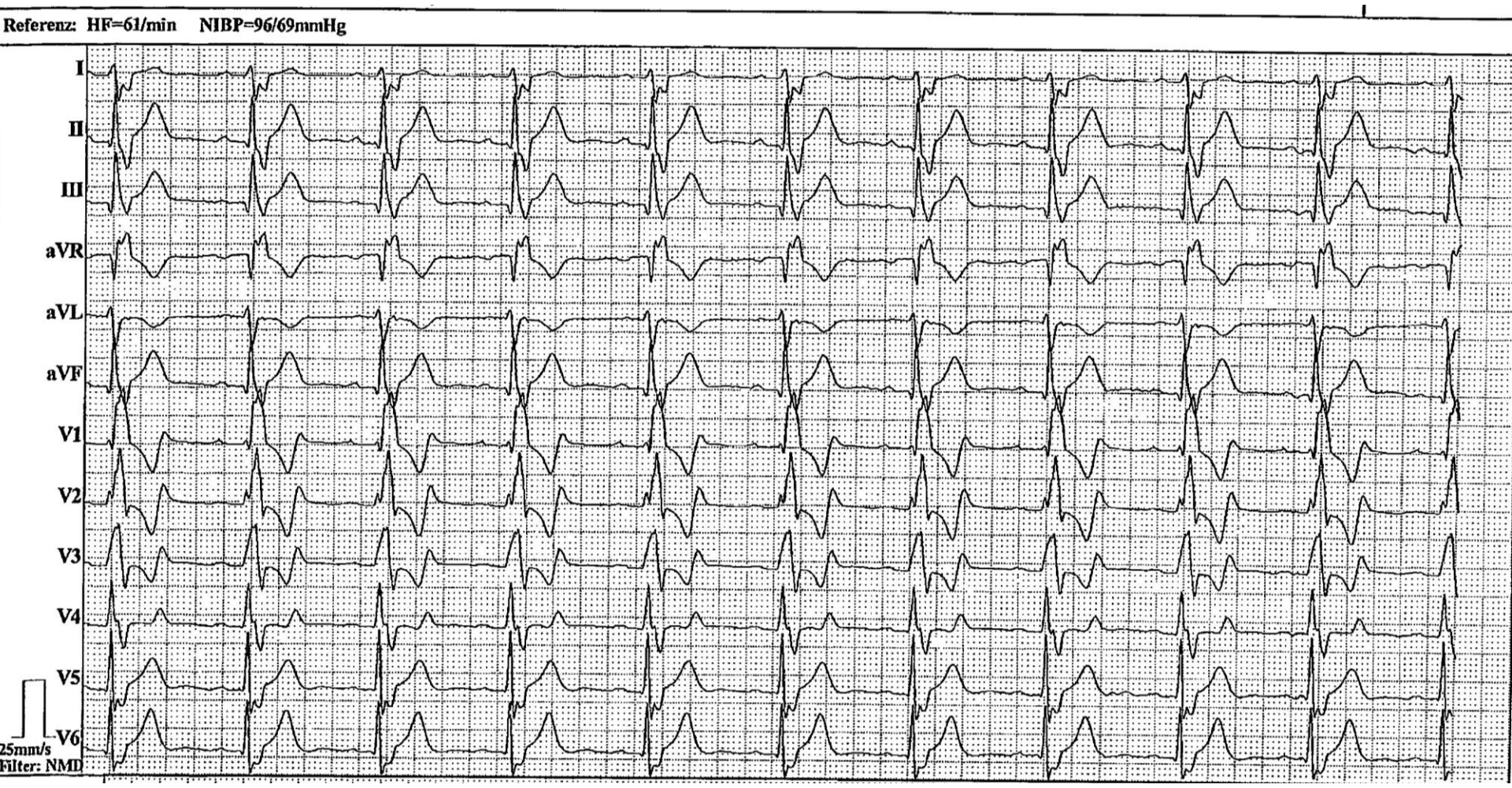
N=873, median age 24.4 years, ECG, exercise, CMR

Median follow-up 4.2 years: death n=28, sustained VT n=4 (3.7%)

Kazuistika 1

- TOF, 33 let, muž
- korekce vady (3 r.)
  - ✓ VSD/ptch, TAN
- Transvenózní implantace DDD pace pro bradykardii (14 let)
- Výměna pace (21 let)
- Ve 23 letech presynkopa, na Holteru středně četné izol. mmVES
  - ✓ PSK – bez indukce VT/VF
- Ve 31 letech implantace konduitu do pulmonální pozice pro PI, rekonstrukce mitrální chlopně
  - ✓ Negativní PSK po operaci

Amb. kontrola ve 33 letech



Amb. kontrola ve 33 letech

O 3 měs. později pac. umírá náhlou smrtí, v paměti pace VF

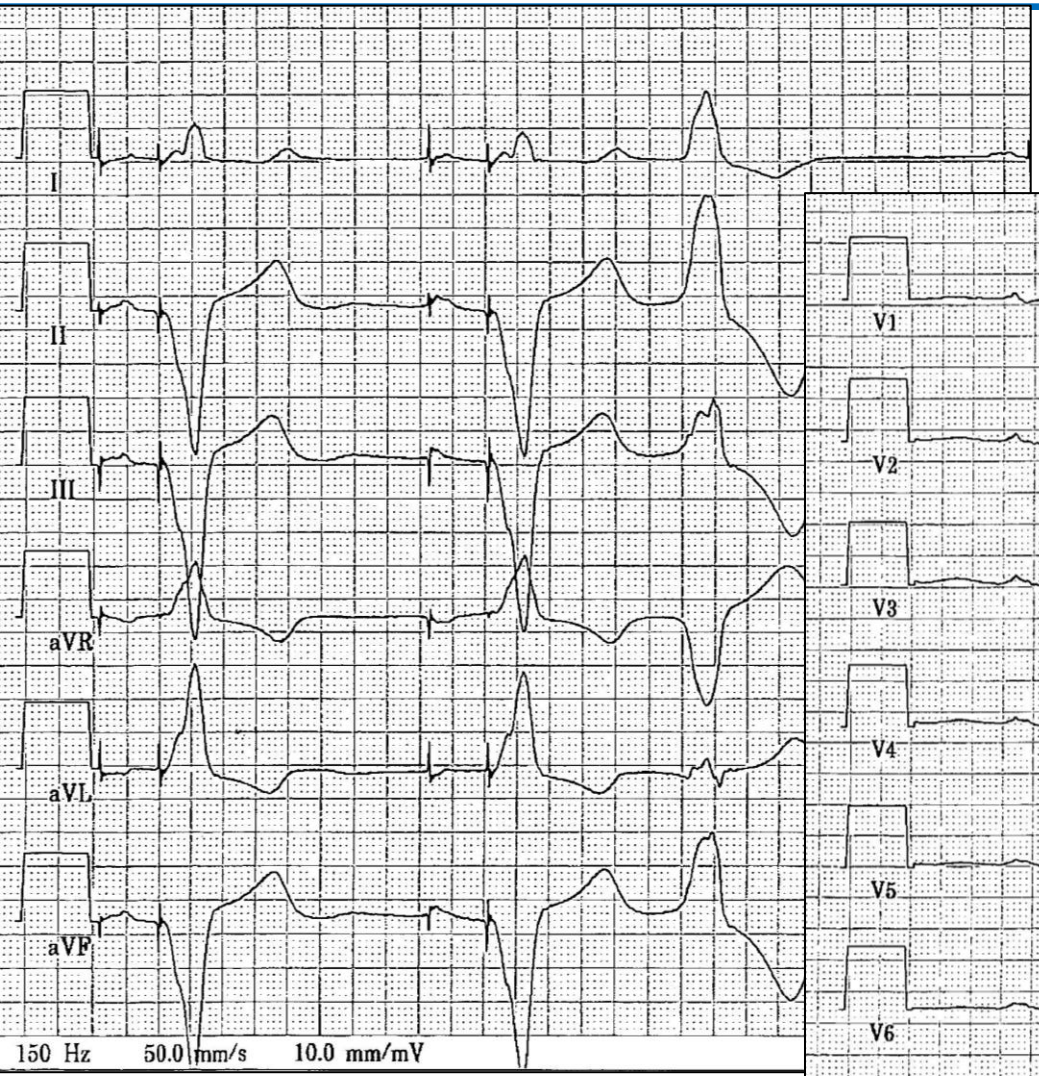
ICD INDICATIONS: CLASS I/IIa

COR	LOE	Recommendation
I	B	ICD therapy is indicated in adults with CHD who are survivors of **cardiac arrest** due to VF or hemodynamically unstable VT after evaluation to define the cause of the event and exclude any completely reversible etiology
I	B C	ICD therapy is indicated in adults with CHD and **spontaneous sustained VT** who have undergone hemodynamic and EP evaluation. Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD therapy in carefully selected patients.
I	B	ICD therapy is indicated in adults with CHD and a **systemic LVEF ≤35%**, biventricular physiology, and NYHA class II or III symptoms
IIa	B	ICD therapy is reasonable in selected adults with **tetralogy of Fallot** and multiple risk factors for sudden cardiac death such as LV systolic or diastolic dysfunction, non-sustained VT, QRS duration ≥180 ms, extensive RV scarring, or inducible sustained VT at EP study

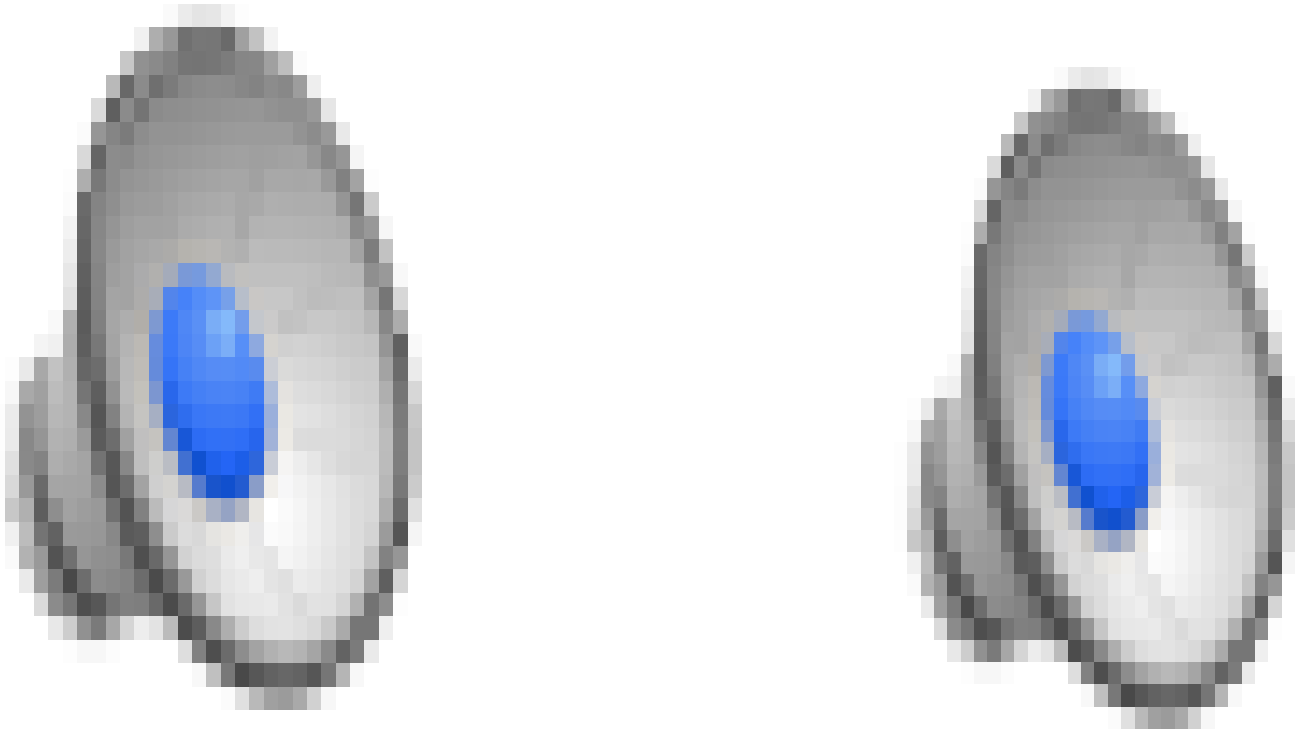
Kazuistika 2

- L-TGA (ccTGA), 34 let, muž
- Implantace DDD pace pro bradykardii při pokroč. AVB II° (17 let)
- Výměna pace (25 let)
- Pravidelné kontroly v "GUCH" ambulanci
- Dobrý klinický stav, stabilní nálezy
- Administrativní práce na plný úvazek

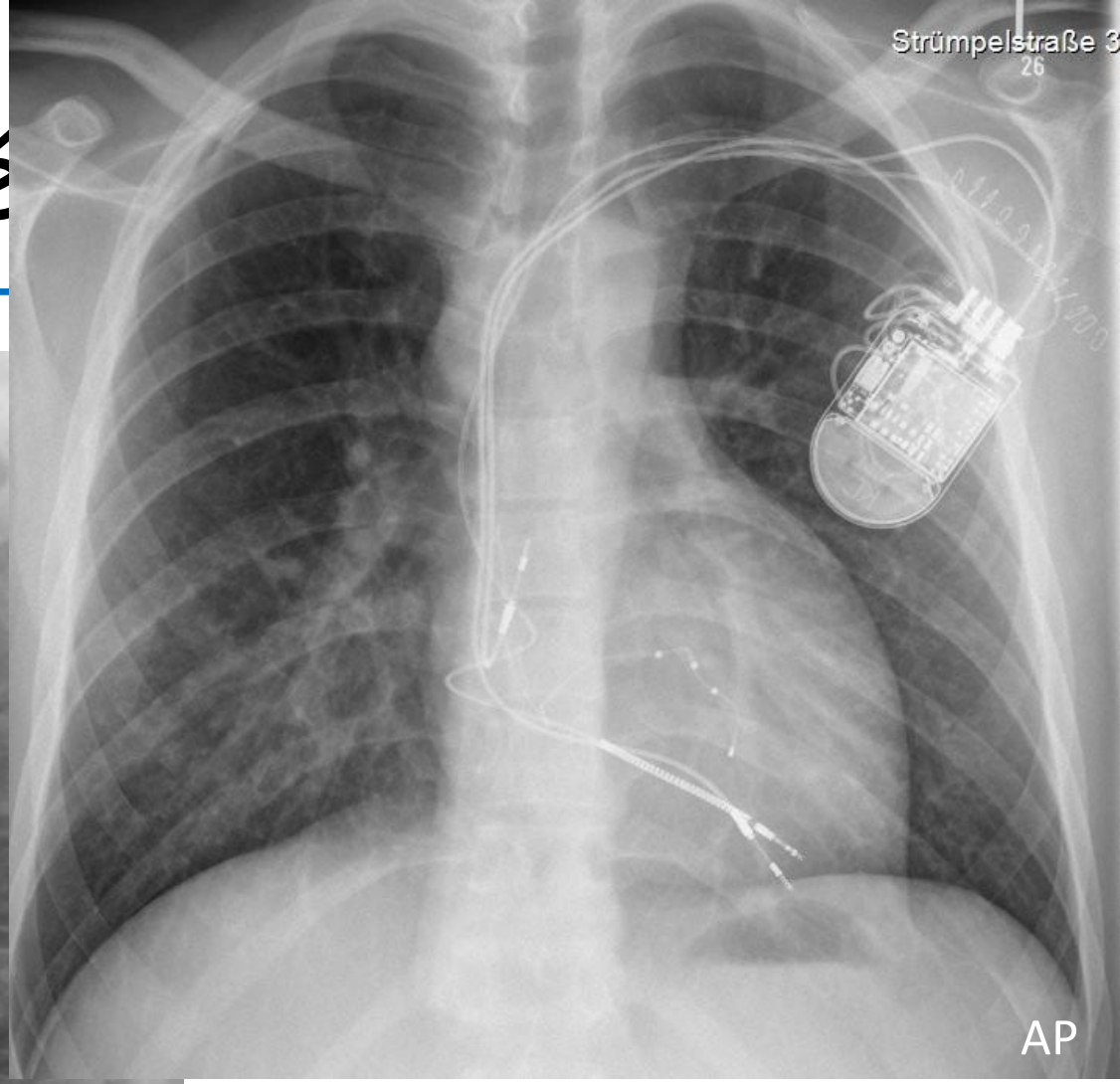
Amb. kontrola ve 31 letech



Echo



o 5 mě



ICD INDICATIONS: CLASS IIb

COR	LOE	Recommendation
IIb	C	ICD therapy may be reasonable in adults with a **single or systemic right ventricular ejection fraction <35%**, particularly in the presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA functional class II or III symptoms, QRS duration ≥140 ms, or severe systemic AV valve regurgitation
IIb	C	ICD therapy may be considered in adults with CHD and a **systemic ventricular ejection fraction <35%** in the absence of overt symptoms (NYHA class I) or other known risk factors
IIb	B	ICD therapy may be considered in adults with CHD and **syncope of unknown origin** with hemodynamically significant sustained ventricular tachycardia or fibrillation inducible at EP study
IIb	C	ICD therapy may be considered for non-hospitalized adults with CHD **awaiting heart transplantation**
IIb	C	ICD therapy may be considered for adults with **syncope** and **moderate or complex CHD** in whom there is a high clinical suspicion of ventricular arrhythmia and in whom thorough invasive and non-invasive investigations have failed to define a cause

Summary – prevention of SCD in GUCH

- Check the hemodynamic status and correct residua first!
- Secondary prevention is "easy" ☺
- Primary prevention - "individual decision" ☹
  - ✓ limited data for ToF/TGA after atrial switch only
  - ✓ no data for single ventricle physiology
- Knowledge of anatomy/surgery is essential – lack of venous access to the heart
  - ✓ S-ICD
  - ✓ alternative implantation techniques

Děkuji za pozornost!