Adults with Congenital Heart Disease:

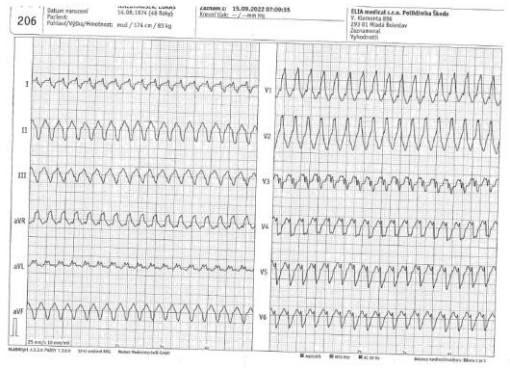
Happy-end or growing problems?

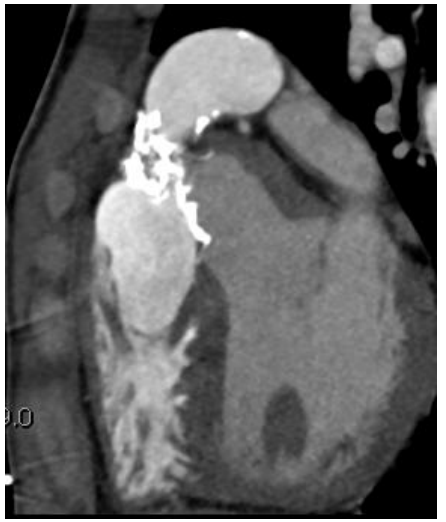
Jana Rubáčková Popelová

Center for Adults with Congenital Heart Disease

Cardiac Surgery, Na Homolce Hospital, Prague 5

Prague Symposium on CHD, 5.11.2022

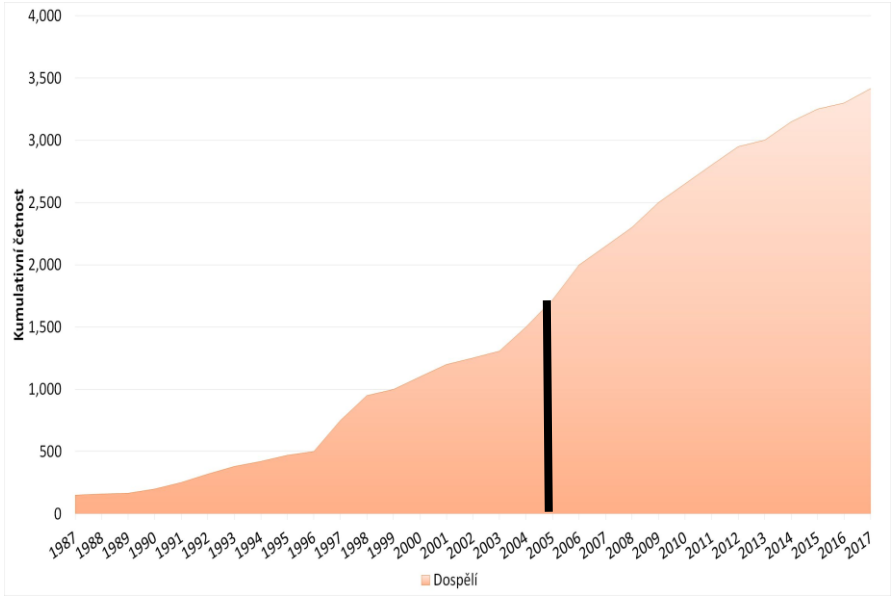




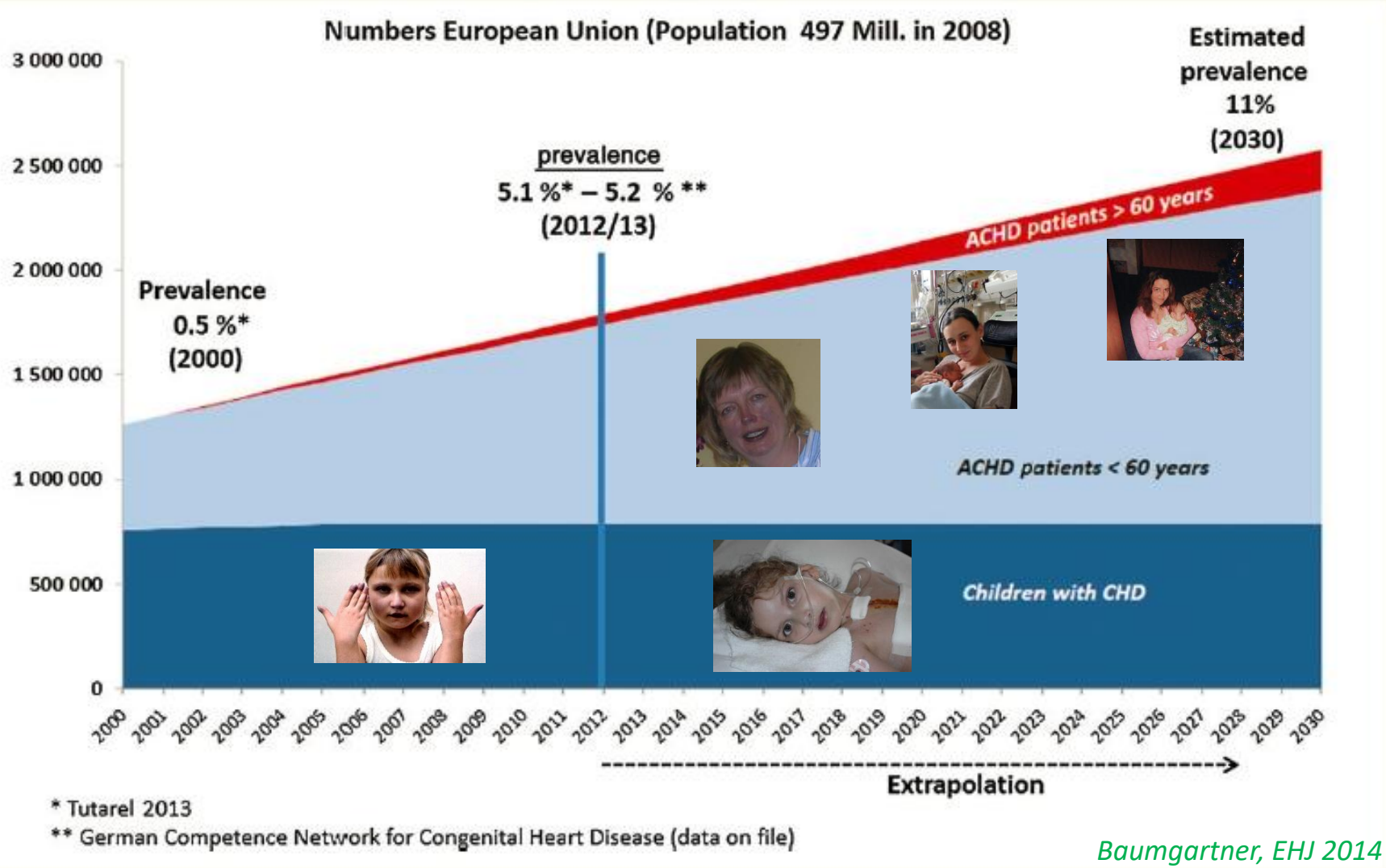
Center for Adults with Congenital Heart Disease

Na Homolce Hospital, Prague

- Since the year 2005
- Adult patients not only from Children Heart Center, but also from cardiologists and hospitals from the whole Czech Republic
- Database: **3759** adult patients with CHD
- Mean age **47 ± 15** years
- New patients **80-100** per year
- Cardiac surgery in adulthood was performed in **1245** patients with CHD



Increasing number of adults with CHD



Baumgartner, EHJ 2014

ACHD issues:

Survival and mortality

Arrhythmias

Heart failure

Operations and interventions in adults

Infective endocarditis

Pregnancy

Quality of life

Survival and mortality

How many children with CHD reach adulthood?

1956 – 1980: 67 %

1980 – 1990: 77 %

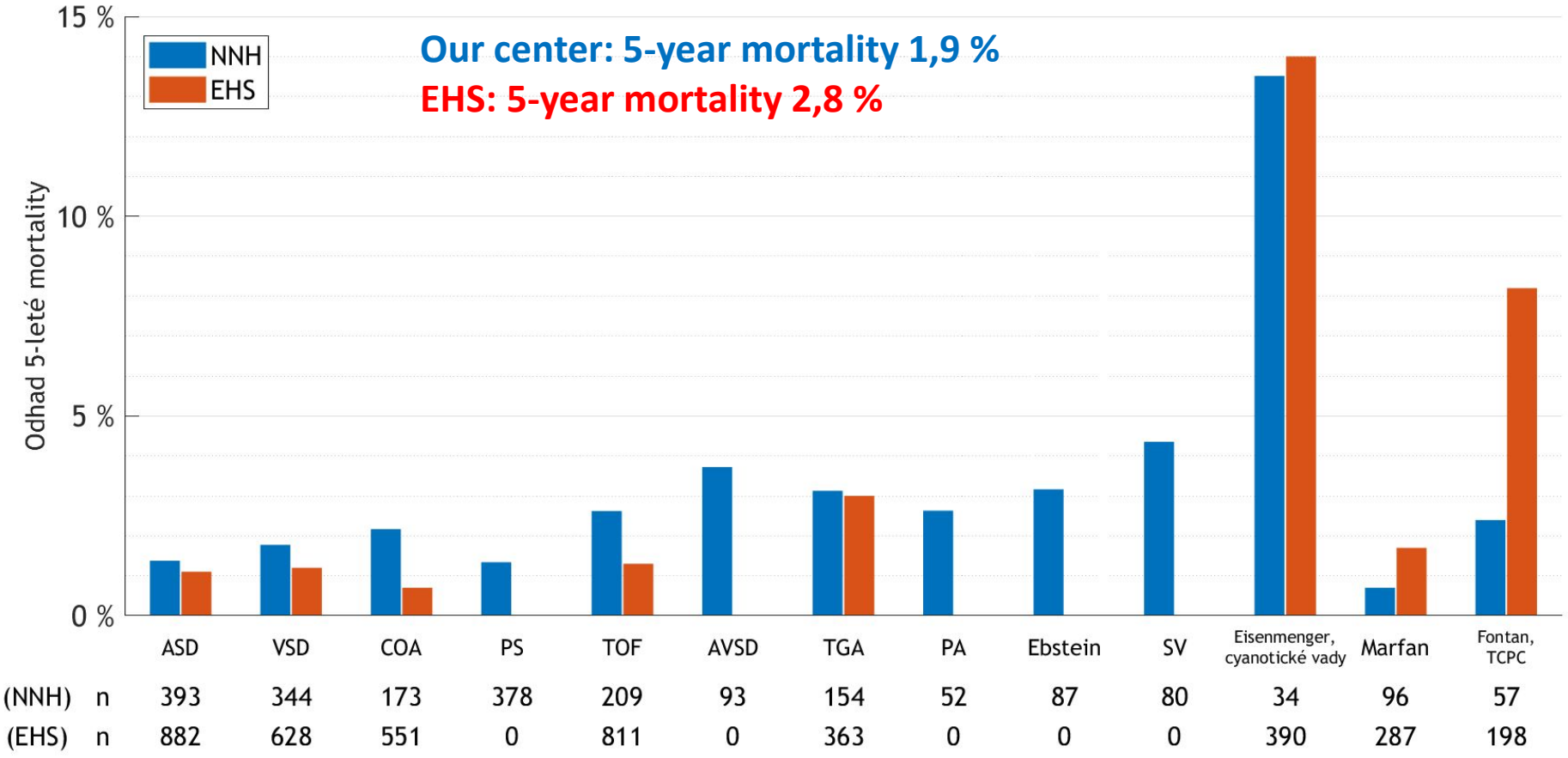
nowadays: > 90 % *(Moons 2010; Stout 2018)*

- Children with simple CHD: 98 %
- Moderately complex CHD: 90 %
- Very complex CHD: 56,4 %
- Univentricular heart: 49 %
- Hypoplastic left heart 7,5 % *(Moons 2010)*

1-year mortality in ACHD: 0,9%

(Diller 2015; Engelings 2016; Greutmann 2014)

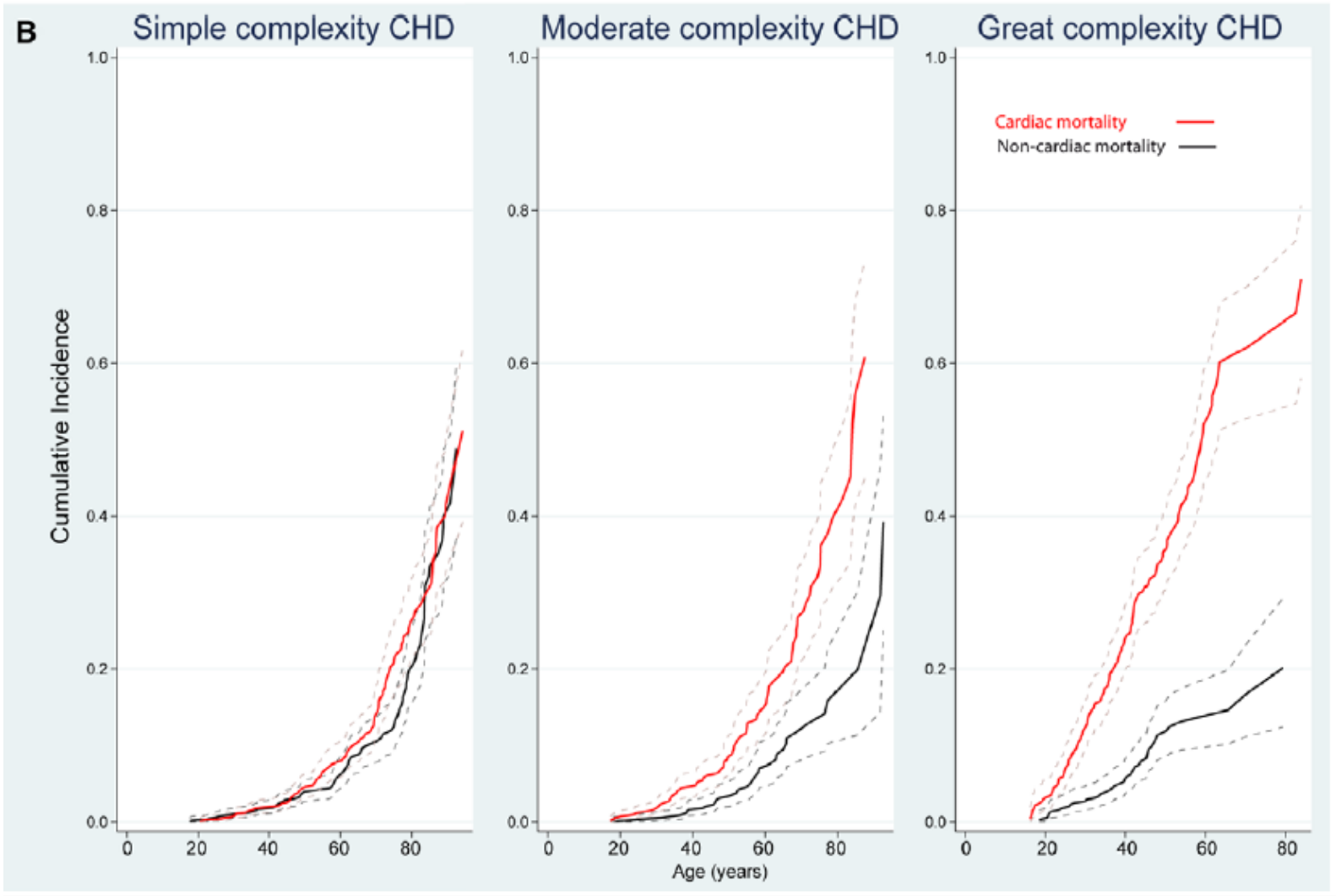
5-year mortality of adults with CHD



EHS: Euro Heart Survey on ACHD / GUCH 2003-2004, retrospective study, 5-years mortality

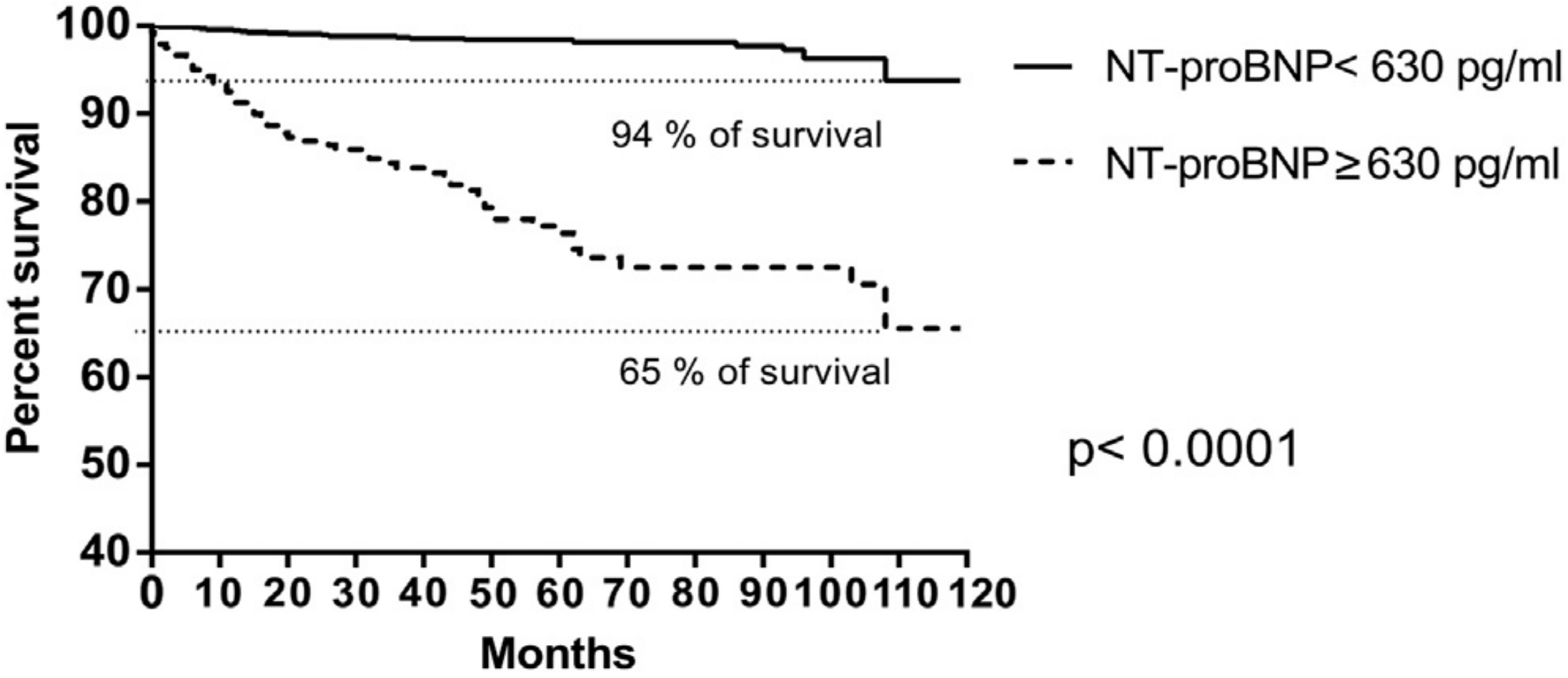
Our center: databasis of adults with CHD, Center in Hospital Na Homolce, 5-years mortality

Cardiac and non-cardiac mortality of adults with CHD



Diller, Circulation 2015

Risk stratification and mortality prediction by NT-proBNP in adults with stable CHD



Popelová, Am J Card, 2015

Cause of death of adults with CHD

Heart failure	**42 %**
Sudden death, arrhythmias	23 %
Pneumonia	10 %
Tumor	6 %
Bleeding	5 %
Perioperative death	13 – 16 %

Diller 2015; Engelings 2016; Greutmann 2014)

Heart failure in ACHD

European Heart Journal (2016) **37**, 1419–1427
doi:10.1093/eurheartj/ehv741

CURRENT OPINION

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

Werner Budts[1]*, Jolien Roos-Hesselink[2], Tanja Rädle-Hurst[3], Andreas Eicken[4], Theresa A. McDonagh[5], Ekaterini Lambrinou[6], Maria G. Crespo-Leiro[7], Fiona Walker[8], and Alexandra A. Frogoudaki[9]

[1]Congenital and Structural Cardiology, University Hospitals Leuven, Herestraat 49, B-3000 Leuven, Belgium; [2]Department of Cardiology, Erasmus Medical Center Rotterdam, Rotterdam, The Netherlands; [3]Department of Pediatric Cardiology, Saarland University Medical Center, Homburg, Germany; [4]Deutsches Herzzentrum München, Munich, Germany;

Heart failure in ACHD

Systemic RV:

- Pharmacological options are not well defined
- The effect of ACEI/ARB is questionable
- Valsartan did not improve survival, had less events in symptomatic pts, 88 pts *(van Dissel, 2018)*
- no evidence of ACEi/ ARB benefit on morbidity and mortality, 359 pts. *(Ladouceur, Heart, 2021)*
- ARNI? Sacubitril/valsartan – 20 pts.,well tolerated, decrease of NT-proBNP
- SGLT2i ??

LV systolic dysfunction

- Treatment of HF as in non-CHD (BB, ACEI/ARB, ARNI, MRA, SGLTi??),
- Diuretics, some pts have Digoxin since childhood
- CRT

RV systolic dysfunction

- Diuretics, pulmonary vasodilatation in PH

Fontan circulation

- Decrease of PAP, diuretics, MRA, pulmonary vasodilatation

Diastolic heart failure in CHD !!!


Congenital heart disease

OPEN ACCESS

Original research

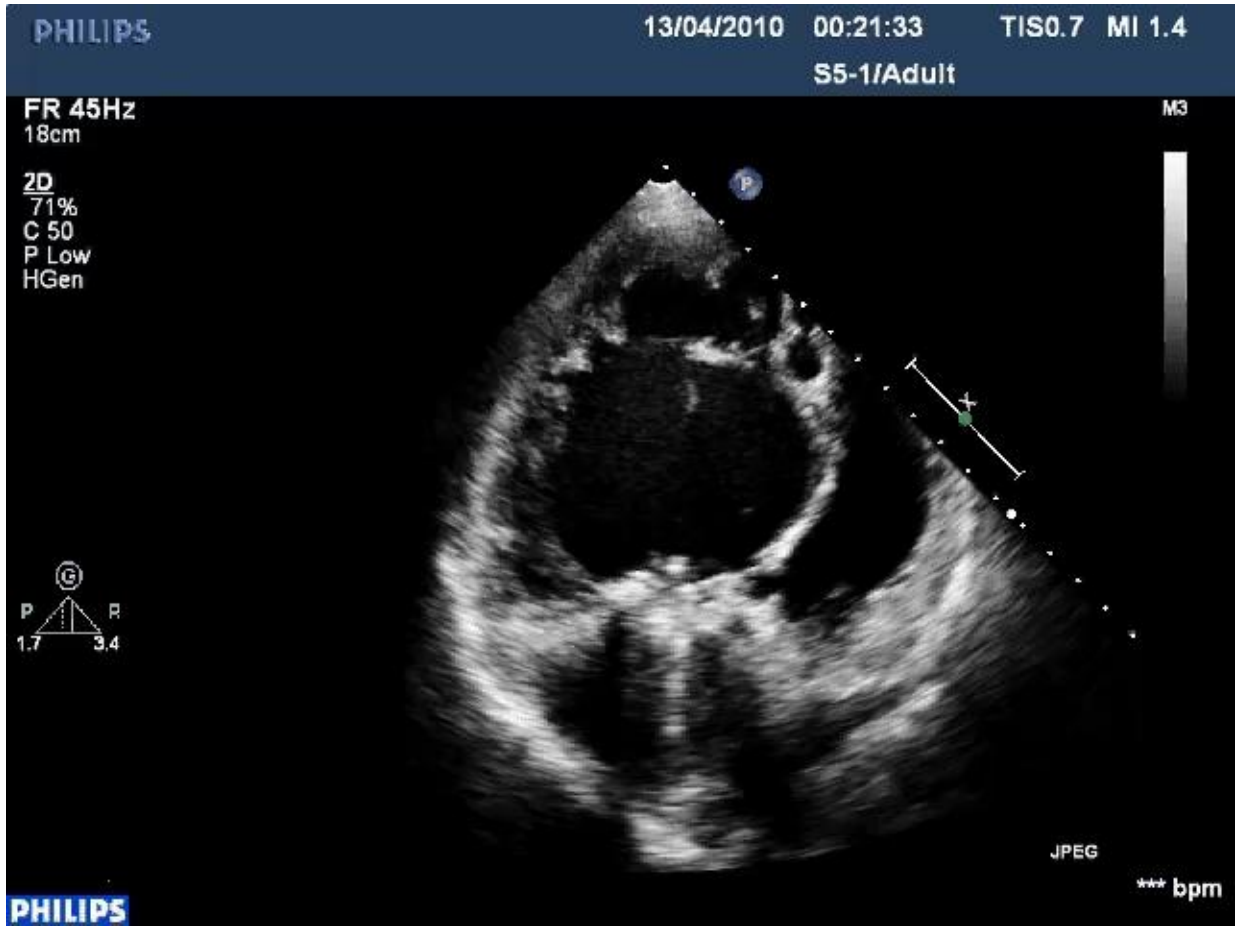
Sacubitril/valsartan in the treatment of systemic right ventricular failure

Tjitske E Zandstra ,[1] Marieke Nederend,[1] Monique R M Jongbloed ,[1,2] Philippine Kiès,[1] Hubert W Vliegen,[1] Berto J Bouma,[3] Laurens F Tops,[4] Martin J Schalij,[4] Anastasia D Egorova [1]

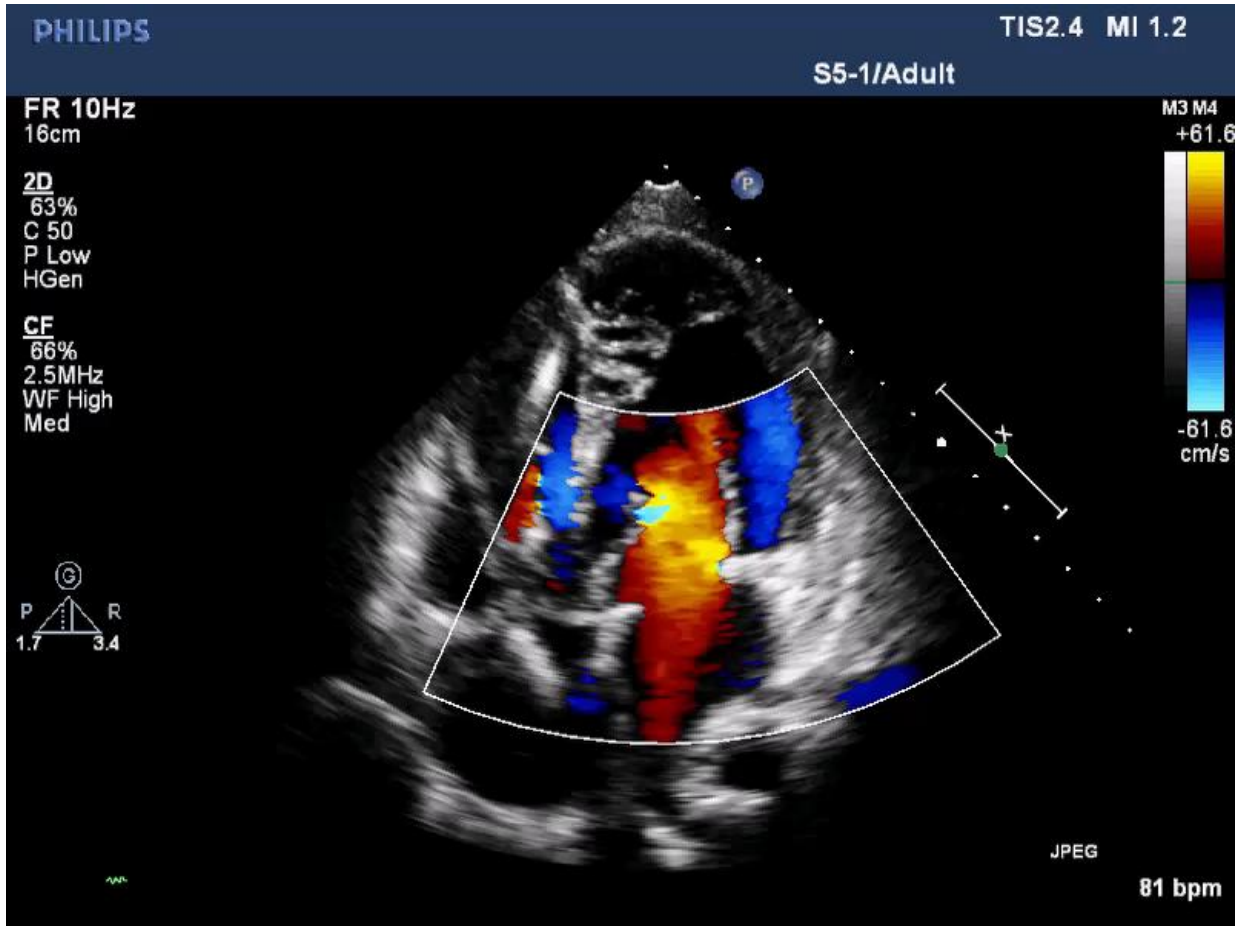
Valve disease? Cardiac surgery?

Systemic right ventricular dysfunction after operation for systemic tricuspid regurgitation

d-TGA, Mustard correction, mechanical AV valve

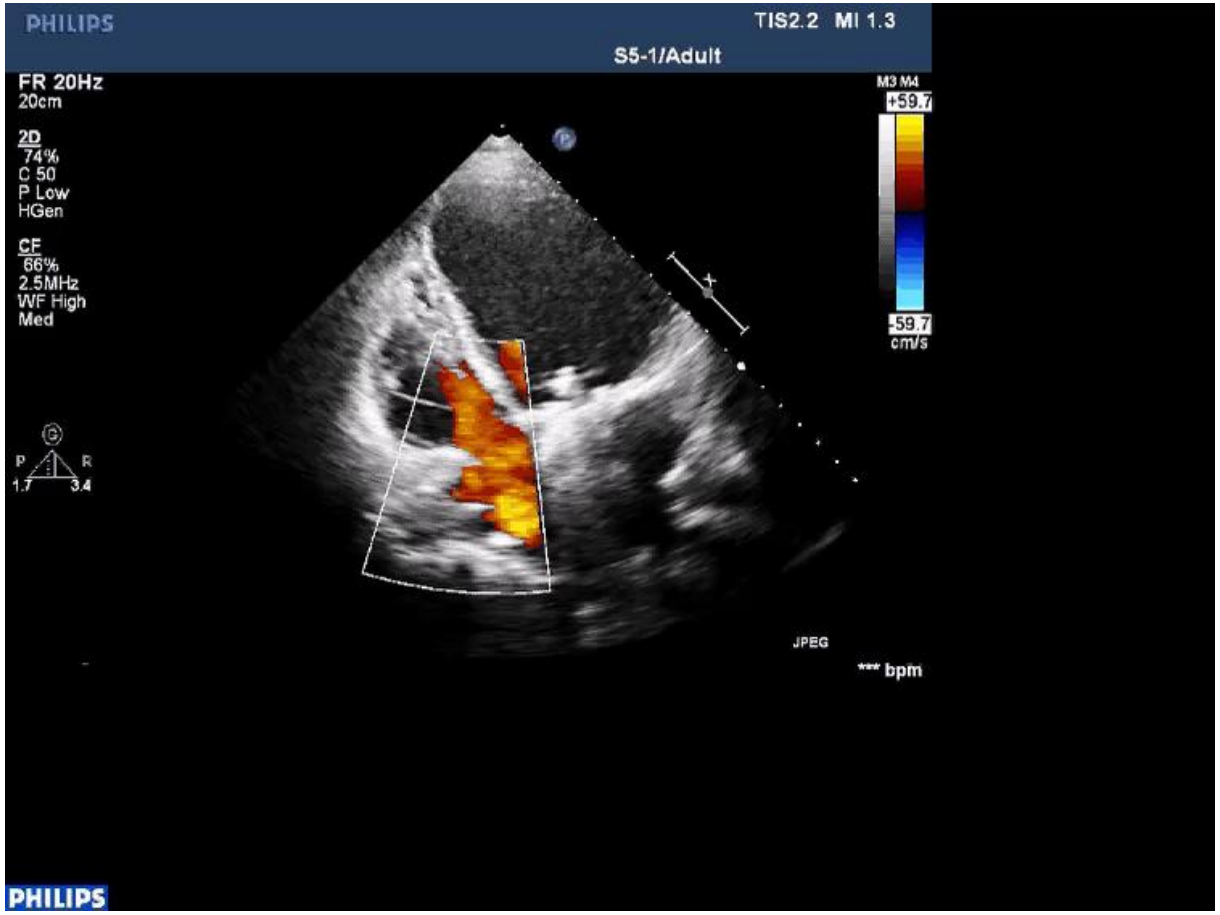


CCTGA after repair of systemic tricuspid valve

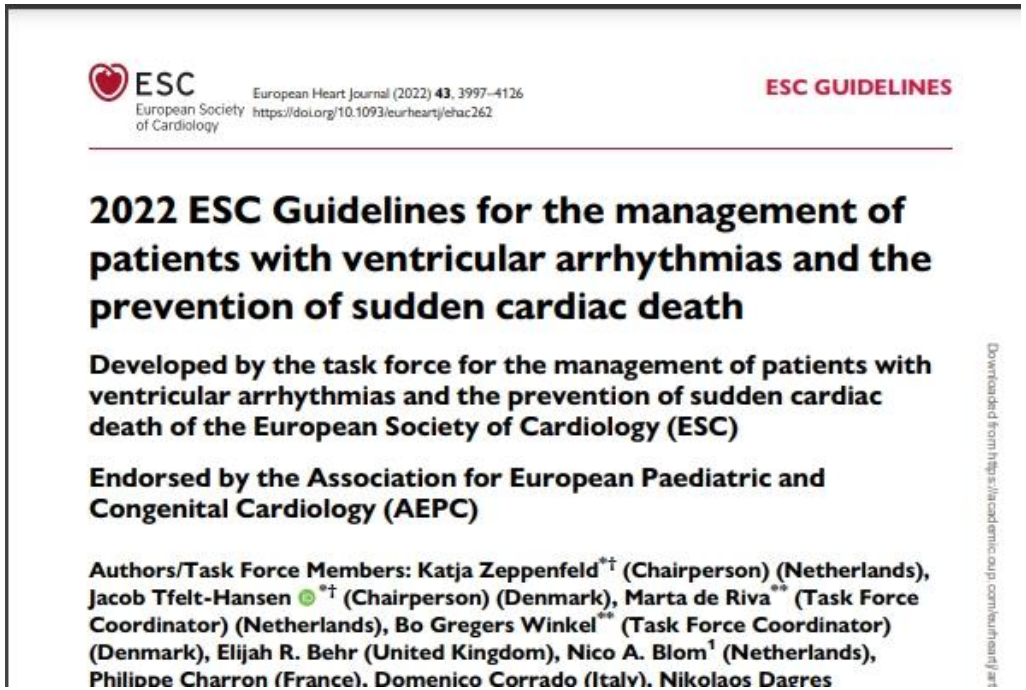


Left ventricular dysfunction

AVSD with late mitral valve replacement due to severe MR
mechanical valve, both mitral leaflets and chordae were removed



Arrhythmias in ACHD



ESC European Society of Cardiology

European Heart Journal (2022) 43, 3997–4126
https://doi.org/10.1093/eurheartj/ehac262

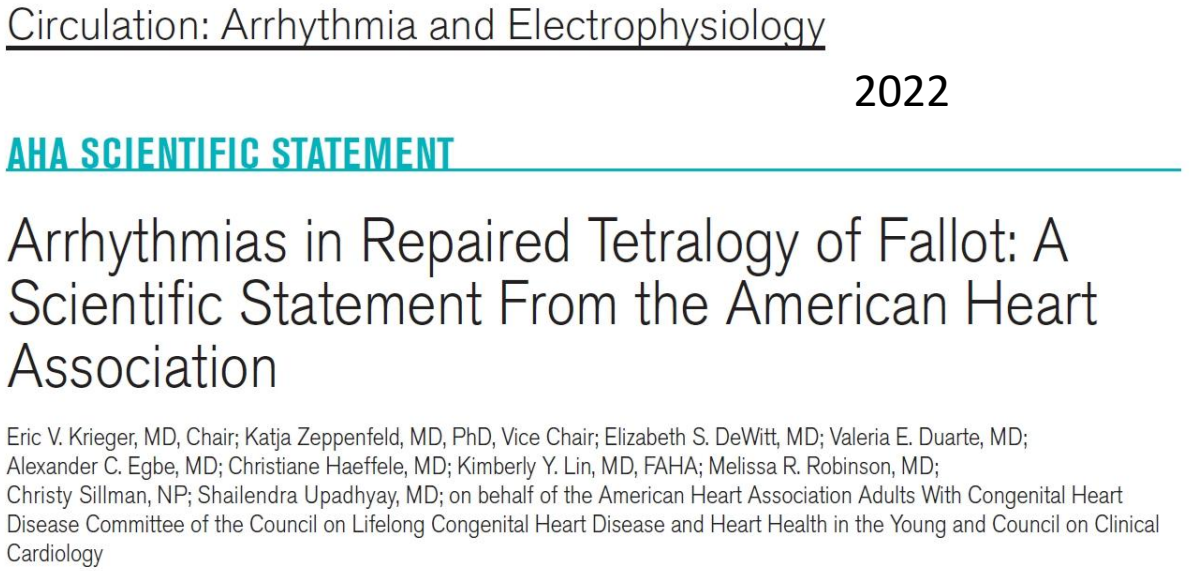
ESC GUIDELINES

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC)

Authors/Task Force Members: Katja Zeppenfeld[*†] (Chairperson) (Netherlands), Jacob Tfelt-Hansen[*†] (Chairperson) (Denmark), Marta de Riva[**] (Task Force Coordinator) (Netherlands), Bo Gregers Winkel[**] (Task Force Coordinator) (Denmark), Elijah R. Behr (United Kingdom), Nico A. Blom[1] (Netherlands), Philippe Charron (France), Domenico Corrado (Italy), Nikolaos Dagres



Circulation: Arrhythmia and Electrophysiology

2022

AHA SCIENTIFIC STATEMENT

Arrhythmias in Repaired Tetralogy of Fallot: A Scientific Statement From the American Heart Association

Eric V. Krieger, MD, Chair; Katja Zeppenfeld, MD, PhD, Vice Chair; Elizabeth S. DeWitt, MD; Valeria E. Duarte, MD; Alexander C. Egbe, MD; Christiane Haeffele, MD; Kimberly Y. Lin, MD, FAHA; Melissa R. Robinson, MD; Christy Sillman, NP; Shailendra Upadhyay, MD; on behalf of the American Heart Association Adults With Congenital Heart Disease Committee of the Council on Lifelong Congenital Heart Disease and Heart Health in the Young and Council on Clinical Cardiology

Risk estimates for arrhythmic events and bradycardias in ACHD

	Supraventricular arrhythmias			Ventricular arrythmias and SCD		Bradycardia			
						SND		AV block	
Type of CHD	AVRT	IART/ EAT	AF	Sustained VT	SCD	Congenital	Acquired	Congenital	Acquired
Secundum ASD		++	++			(+)	+		(+)
Superior sinus venosus defect		++	+				+		
AVSD/primum ASD		++	++	(+)		(+)		(+)	++
VSD		+	(+)	+	(+)[a]				+
Ebstein anomaly	+++	++	+	(+)	++[b]		++		
TOF		++	++	++	++		+		+
TGA									
Atrial switch		+++	+	++[c]	+++[b]		+++		+
Arterial switch		+		+[c]	(+)		(+)		
ccTGA	++	+	+	(+)	++[b]			+	++
Fontan operation									
Atriopulmonary connection		+++	++		+[b]		++		
Intracardiac lateral tunnel		++	+		+[b]		++		
Extracardiac conduit		+	+		+[b]		+		
Eisenmenger physiology Incompletely palliated CHD		++	++		++[d]				

Empty cells indicate that although not specifically indicated, arrhythmic events may occur (no symbol).

(+) = minimal risk + = mild risk ++ = moderate risk +++ = high risk

ESC European Society of Cardiology
European Heart Journal (2020) **00**, 1–83
doi:10.1093/eurheartj/ehaa554

ESC GUIDELINE

2020 ESC Guidelines for the management of adult congenital heart disease

The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Adult Congenital Heart Disease (ISACHD)

Authors/Task Force Members: Helmut Baumgartner* (Chairperson) (Germany),

Antiarrhythmic transcatheter interventions in adults with CHD

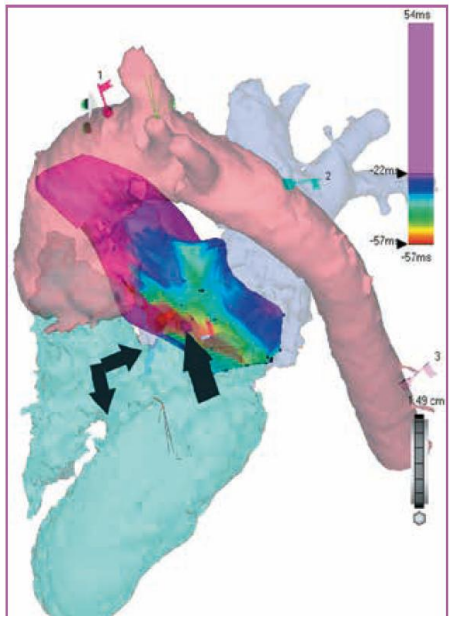
in Na Homolce Hospital

- **182 transcatheter ablations in ACHD**

MUDr. Jan Škoda, MUDr. Petrů, prof. Neužil

- **50 high complexity ablations**

- **39 with limited venous approach: 27x retrograde transaortic approach with remote magnetic navigation**
  9x baffle punction
  1x transhepatic approach in IVC agenesis + BCPA
  2x azygos vein

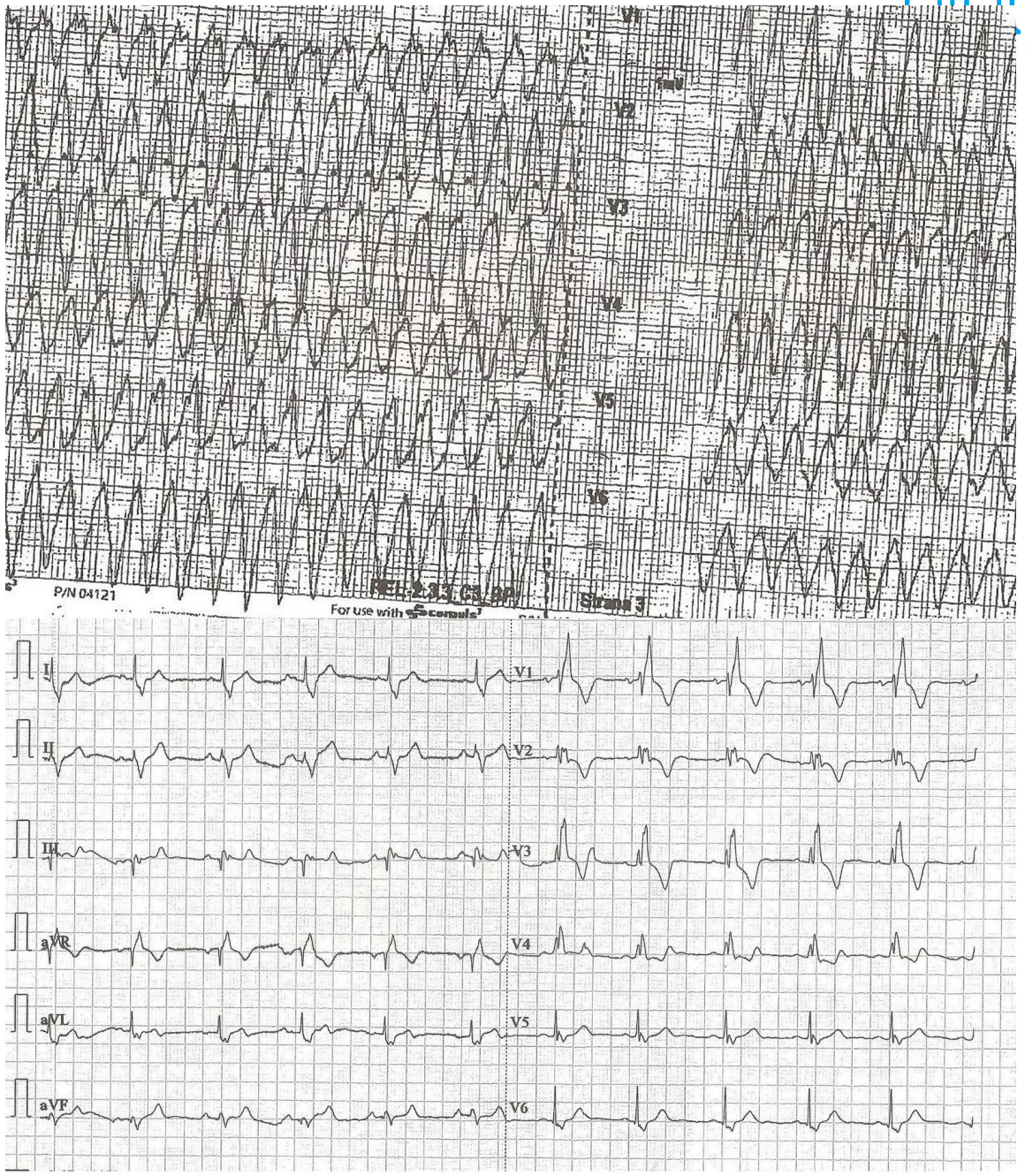


EA mapping with CT integration, focal atrial tachycardia, from right paraseptal region
d-TGA, SV, TCPC, transaortic approach with remote magnetic navigation

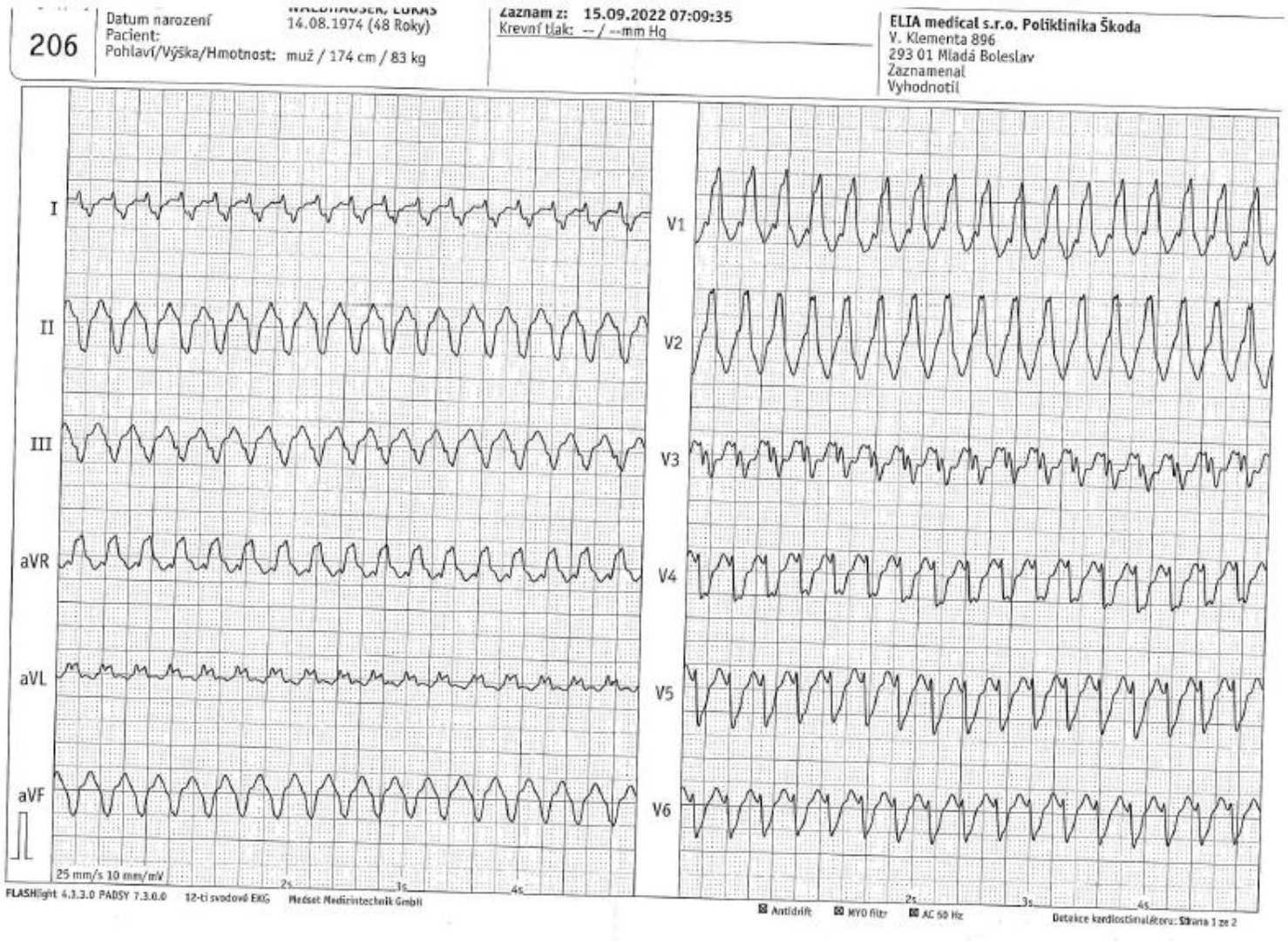
Monomorphic ventricular tachycardia

in repaired TOF

- 36-years old lady with repaired TOF 32 years ago
- No symptoms, NYHA I,
- ECG: RBBB with QRS 160 msec
- After physical effort she had syncope and resuscitation, on ECG **sustained monomorphic ventricular tachycardia with 300 beats per minute**
- After electric cardioversion sinus rhythm 66/min, RBBB
- Echocardiography: mild RV dilatation, good RV systolic function.
- Severe pulmonary regurgitation
- According to the Guidelines she had no indication for pulmonary valve replacement.



IART in repaired Tetralogy of Fallot



- *48-years old man with repaired TOF*
- *Good function of pulmonary bioprosthesis*
- *Syncope when going on the bicycle*
- *ECG: Postincisional flutter with rapid ventricular response (200 beats per minute (1:1 conduction),*
- *Electrical cardioversion*
- *Transcatheter ablation is planned soon*

Operations and interventions in adulthood

Our experience with surgery of adults with CHD

- Surgery was performed in **1245** adult patients with CHD *(up to 30.6.2022)*
- Average age at surgery **40 ± 14 years** (16 - 79 years)
- History of previous operation: **46 %** (573) of our operated patients
- History of previous 3-6 operations: **16 %** (202 pts)
- Most reoperations: truncus arteriosus and pulmonary atresia

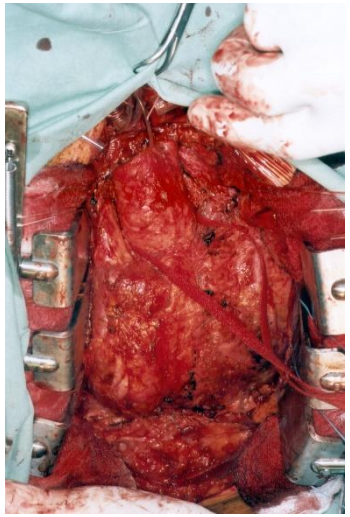
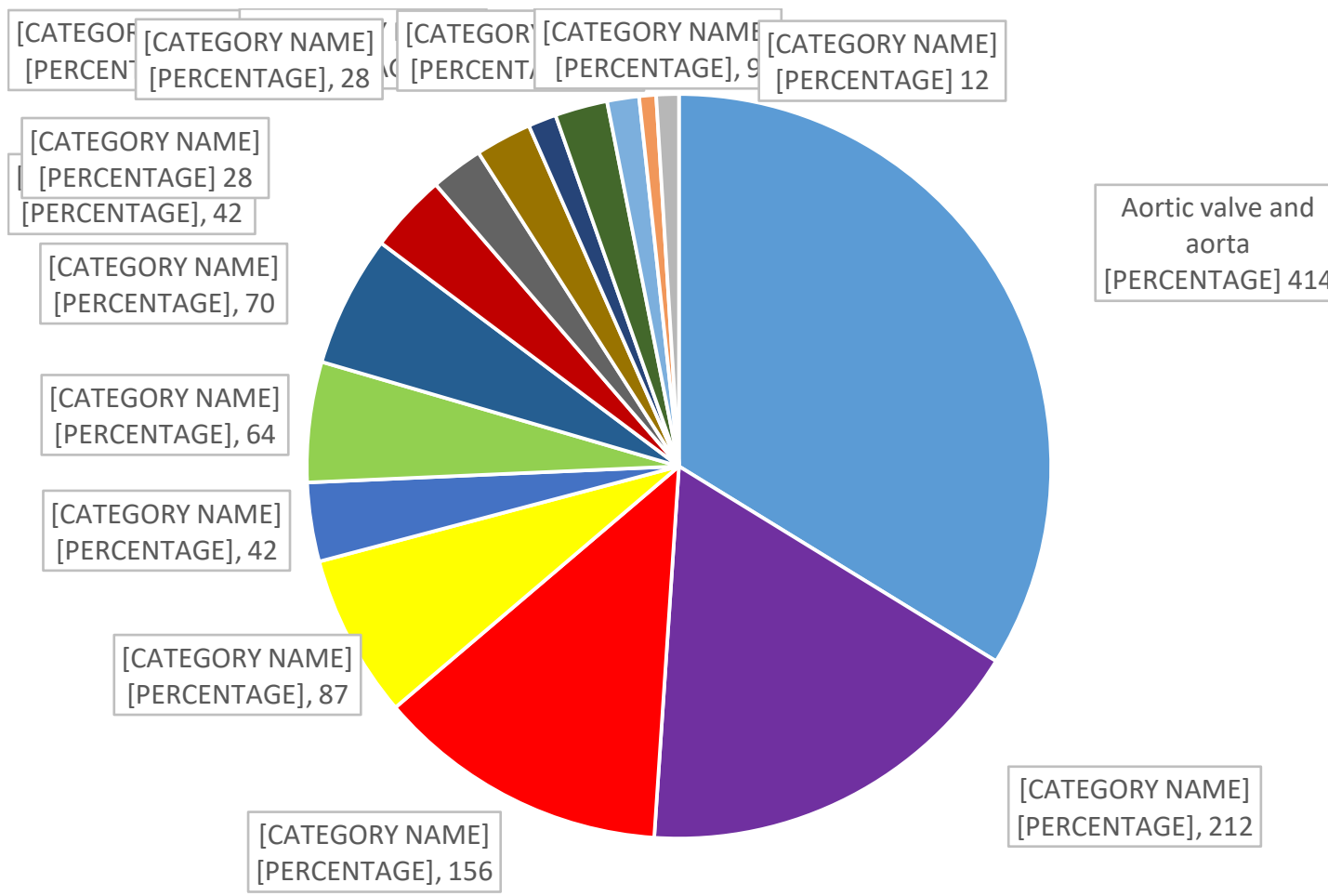


Photo: MUDr. Štěpán Černý

Complex lesions: 35 %



Our experience with surgery of adults with CHD

30-days mortality: **1,3 %**

in-hospital mortality: **1,7 %**

In-hospital mortality:

- Pulmonary atresia **7 %** (+2/28; sepsis, IE)
- Ebstein anomaly **4,5 %** (+4/87)
- COA: **0 %** (+0/42)
- TOF after radical repair: **0 %** (+0/156)
- Pulmonary stenosis: **0 %** (+0/70)
- Truncus arteriosus: **0 %** (+0/9) (one death on WL OTS)

Our surgeons:

MUDr. Roman Gebauer
MUDr. Štěpán Černý
MUDr. Petr Pavel
MUDr. Ivo Skalský
MUDr. Aleš Klváček

Risk factors of mortality after operation of adults with CHD

- **permanent cyanosis before surgery** (OR 60,5), in-hospital mortality **41%** (+7/17)
- **NYHA class III-IV** (OR 30,8), in-hospital mortalty **12%** (+ 22/170)

Tabulka 4 – Rizikové faktory časných i pozdních úmrtí po operaci vrozené srdeční vady v dospělosti

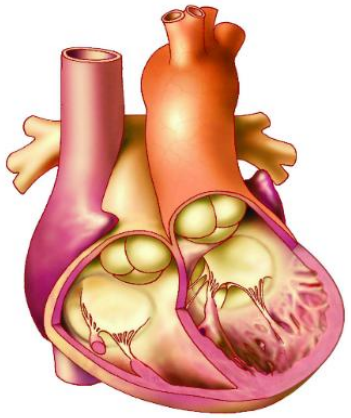
Rizikový faktor	Přeživší (N = 780)	Zemřelí (N = 25)	*p*	OR
NYHA třída III–IV před operací	150 (19 %)	22 (88 %)	< 0,0001	30,8
Cyanóza před operací	6 (0,77 %)	8 (32 %)	< 0,0001	60,5
Anamnéza městnavého srdečního selhání	35 (4,5 %)	6 (24 %)	0,001	6,7
Přítomnost mechanické chlopenní náhrady	120 (15,4 %)	10 (40 %)	0,0032	3,7
Univentrikulární cirkulace	19 (2,4 %)	3 (12 %)	0,0276	5,4
Plicní hypertenze	106 (13,6 %)	7 (28 %)	0,072	2,4
Arytmie	238 (30 %)	12 (48 %)	0,078	2,1
Muži	419 (54 %)	9 (36 %)	0,103	0,48
Věk v době operace	39	40	0,636	NA
Počet předchozích operací Předchozí operace na pacienta	538 0,63	35 1,4	0,00033	NA

NA – nelze určit, OR – odds ratio.

(Rubáčková Popelová, Cor Vasa, 2016)

Operations of systemic right ventricle

N = **26** operations; **2,3%** of all CHD operations, in-hospital mortality **7,6%**

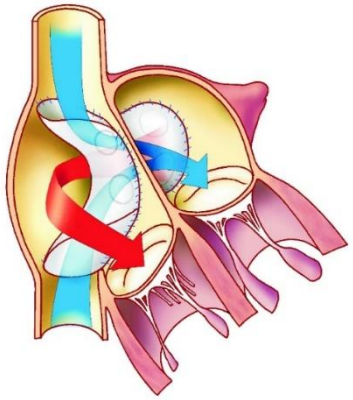


CCTGA, systemic RV is left-side

Operations – systemic tricuspid valve:
12x mechanical prosthesis
2x bioprosthesis
6x tricuspid valve repair

Aortic valve:
1x bioprosthesis

Resynchronisation therapy in TGA/Must/Senn
5x epicardial electrodes implantation
1x extraction of epicardial electrodes due to infection



TGA, Mustard, Senning, systemic RV is right-side

CCTGA: 15 operations, **mortality 13%** (+2/15)

TGA after atrial switch: 11 reoperations, **mortality 0%** (0/11)

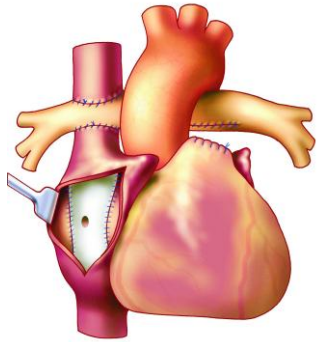
Operations of adults with univentricular circulation

N = **36** operations; **3%** of all CHD operations, **47%** of all our 76 followed-up Fontans

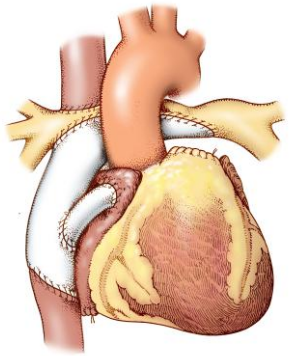
in-hospital mortality: **16,7%** (+6/36)

- **Types of operations:**

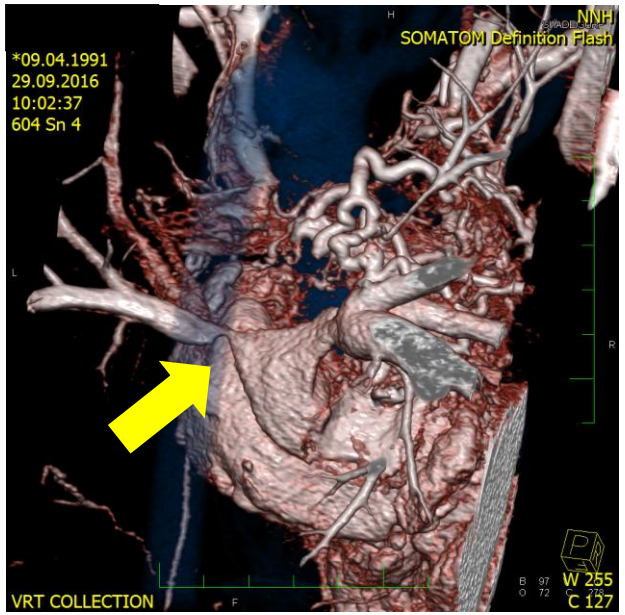
- TCPC in adulthood, Kawashima **(6x)**
- conversion of classical Fontan operation to TCPC **(4x)**
- mitral valve repair (5x)
- closure of tricuspid valve (4x)
- mechanical prosthesis (5x)
- excision of ventricular septum to remove of subaortic stenosis (2x)
- repair of pulmonary veins stenosis (1x)
- excision of mediastinal tumor (1x)
- completion of TCPC by hepatic veins connection (3x)
- epicardial electrodes (5x)



Intra-atrial lateral tunnel with fenestration



extracardial conduit

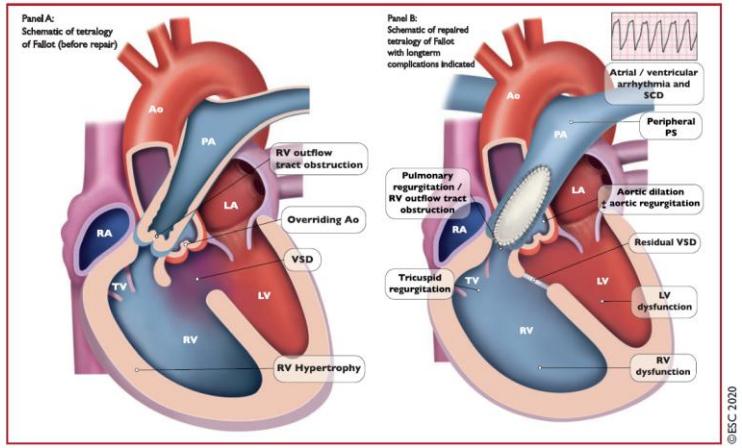


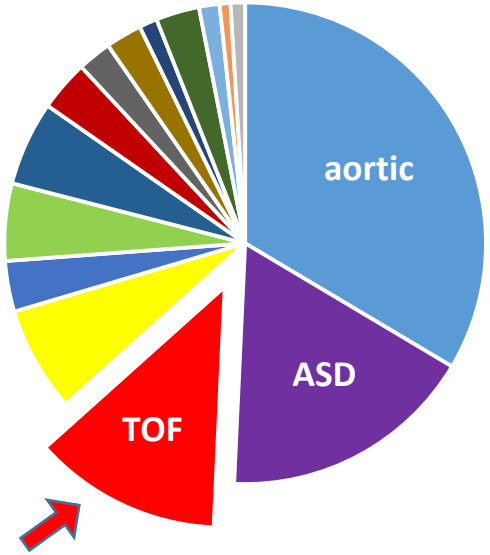
Stenosis of left-sided pulmonary veins after RFA

Tetralogy of Fallot

N = **156** operations; **13%** of all CHD operations

mortality **0 %** (+0/156)

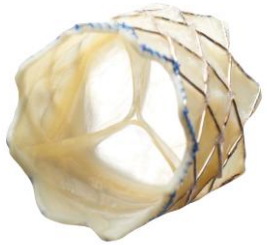




Reoperations TOF, DORV, PA and other complex CHD:

193 pulmonary bioprosthesis
60 pulmonary homografts
17 mechanical pulmonary prosthesis
8 pulmonary repair and Ozaki autopericardial valve

42 TPVI: transcatheter pulmonary valve implantation in homograft or bioprosthesis

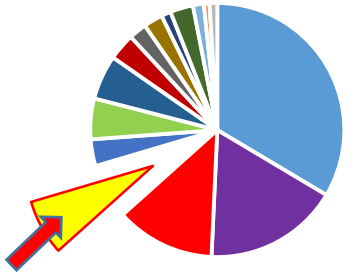


TPVI Melody

TPVI Edwards Sapien

Ebstein anomaly of tricuspid valve

(N = **87** operations; **7 %** of all CHD operations, in-hospital mortality **4,5%** (+4/87), daSilva mortality **0%**

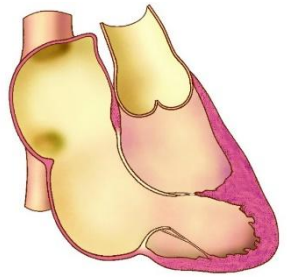


Ebstein anomaly
of tricuspid valve

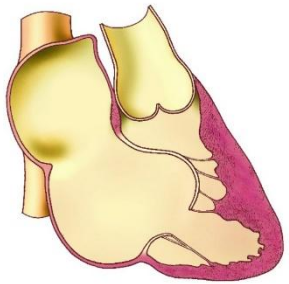
Our oldest operated patient with Ebstein anomaly:
***In 68 years**: tricuspid bioprosthesis*
***In 72 years**: TS (15/6mmHg) a TR, valve-in-valve implantation*

In 76 Years.....

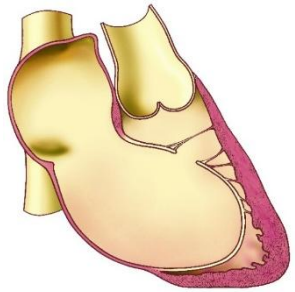
type A-B



type C

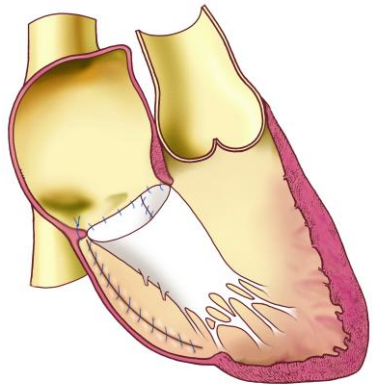


type D



Cone-repair (daSilva)

In our center: **since 2010**
Operations performed by Roman Gebauer, MD



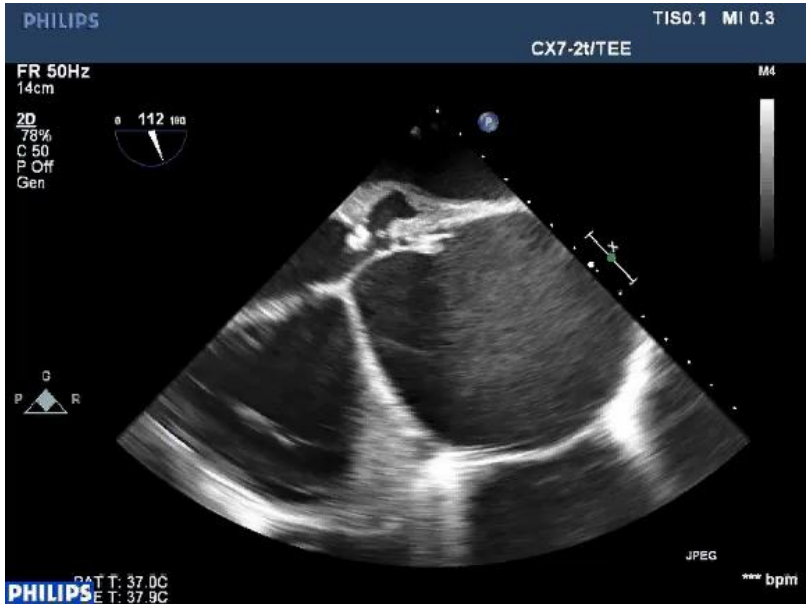
Infective endocarditis

IE in adults with CHD in our registry

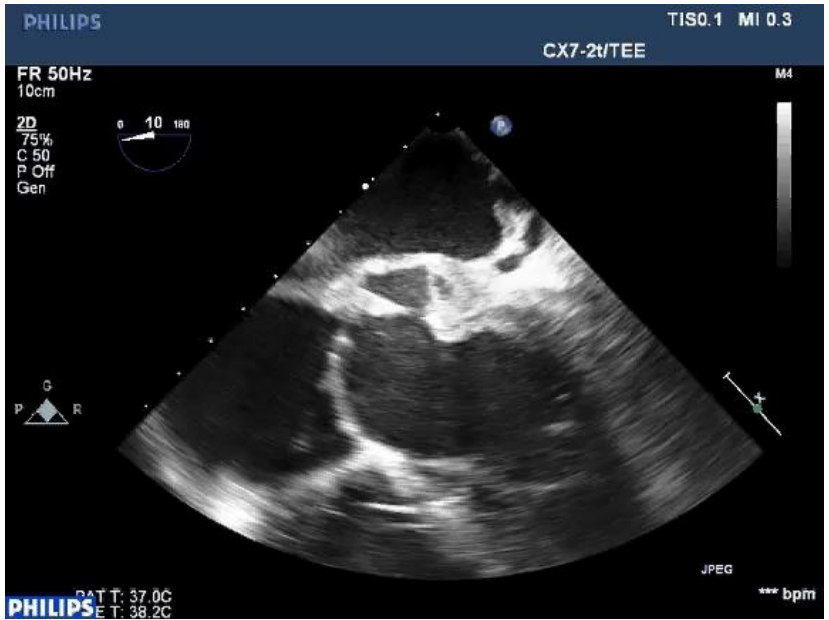
- **BAV** **9,5 %** **(60/627)**
- **VSD** **5%** **(33/660)**
- **ASD** **2%** **(16/804)**

- **IE on pulmonary valve**:
- PVR HMGR**:** **22%** (23/106) – HMGR, also from childhood
- PVR bio: **12 %** (25/208)
- PVR mech: **16,7%** (3/18)

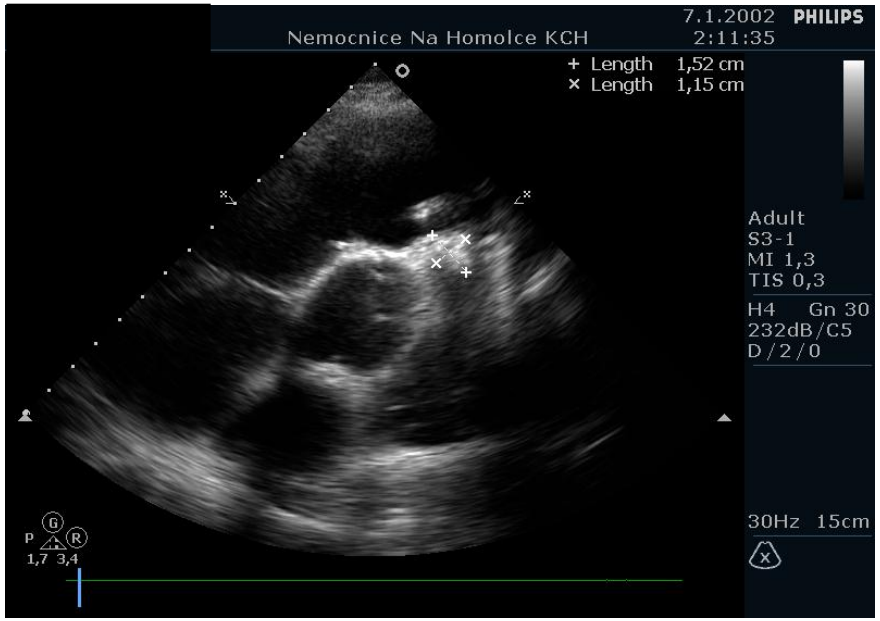
- TPVI: **12%** (5/42 all TPVI)
- **TPVI Melody** **26 % (5/19 TPVI Melody only)**

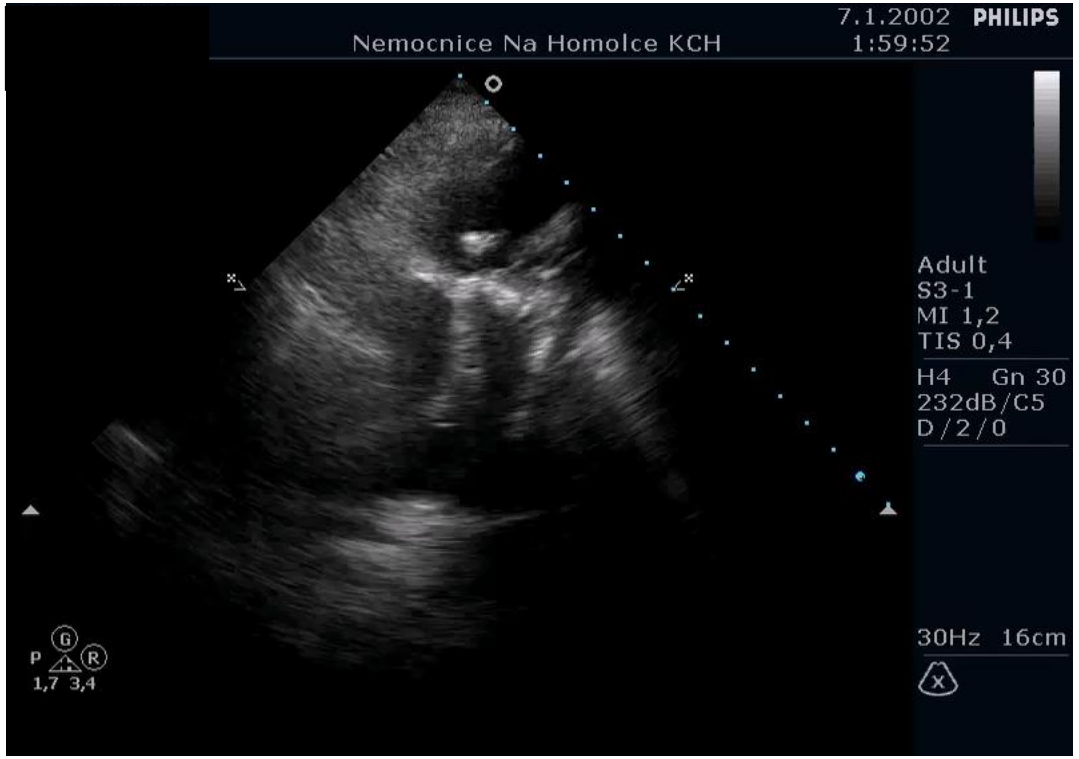


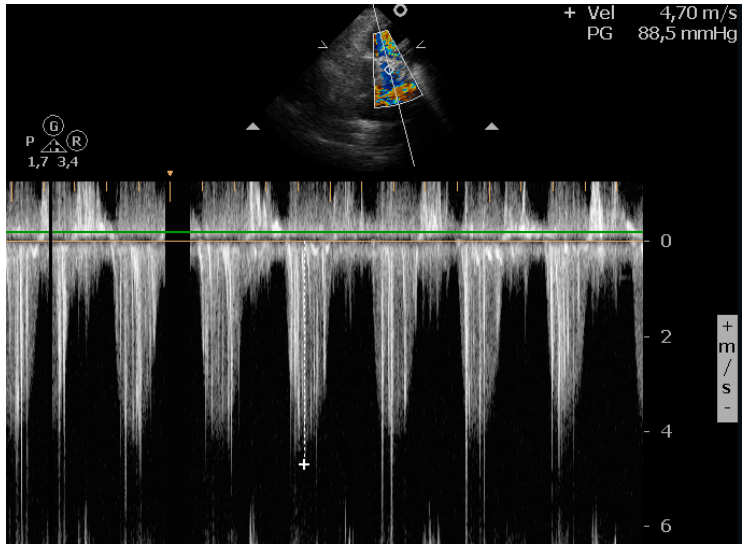
Unoperated BAV with IE, abscess emptying to LVOT, AS, AR, severe dilatation of aortic root,



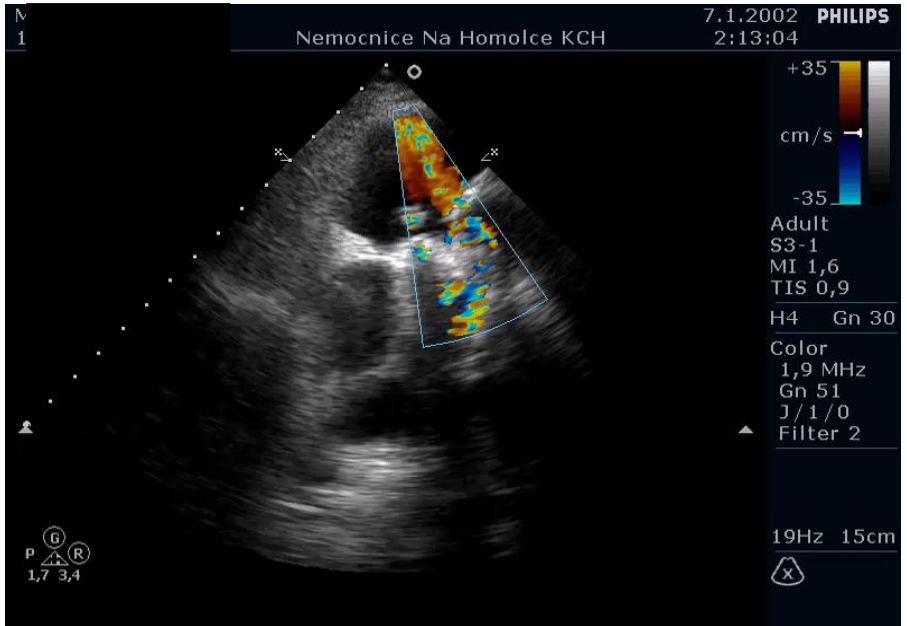
Staphylococcus IE on older pulmonary HMGR with severe pulmonary stenosis







Grad. 89mmHg



Pregnancy

Out of 1293 women with CHD in our database
389 were pregnant during our follow-up = 30 %

ESC European Society of Cardiology
European Heart Journal (2018) **00**, 1–83
doi:10.1093/eurheartj/ehy340

ESC GUIDELINES

2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

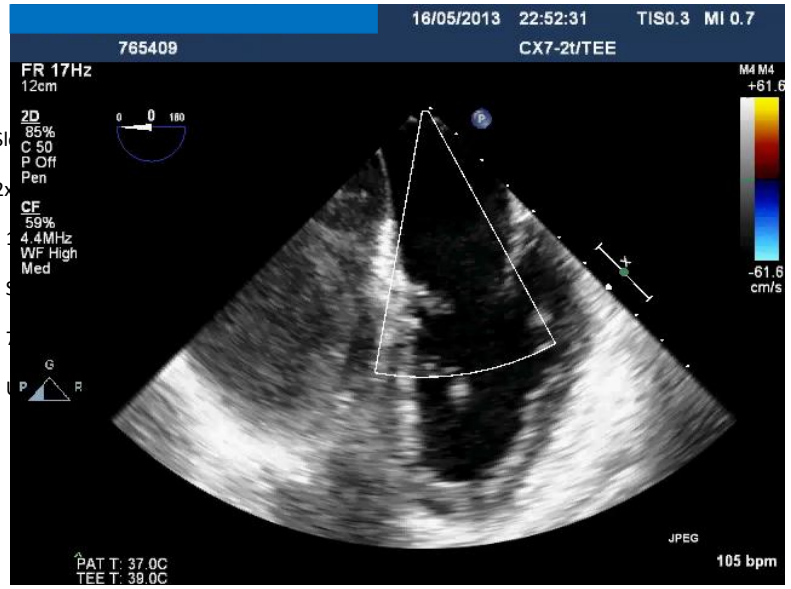
The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC)

Endorsed by: the International Society of Gender Medicine (IGM), the German Institute of Gender in Medicine (DGesGM), the European Society of Anaesthesiology (ESA), and the European Society of Gynecology (ESG)

Authors/Task Force Members: Vera Regitz-Zagrosek* (Chairperson) (Germany), Jolien W. Roos-Hesselink* (Co-Chairperson) (The Netherlands), Johann Bauersachs

Fontan circulation and pregnancy – difficult decision

- Pregnancy with high risk is possible only in the „best“ Fontans,
- NYHA I, no residual findings, VO2max ≥ 25 ml/kg/min
- 50 % abortions, preliminary delivery and neonatal death, frequent infertility



Fontan – Doty

TCPC

Our experience: 34 women with Fontan circulation in fertile age

13x pregnancies in 9 women (26 %), 1 death after delivery

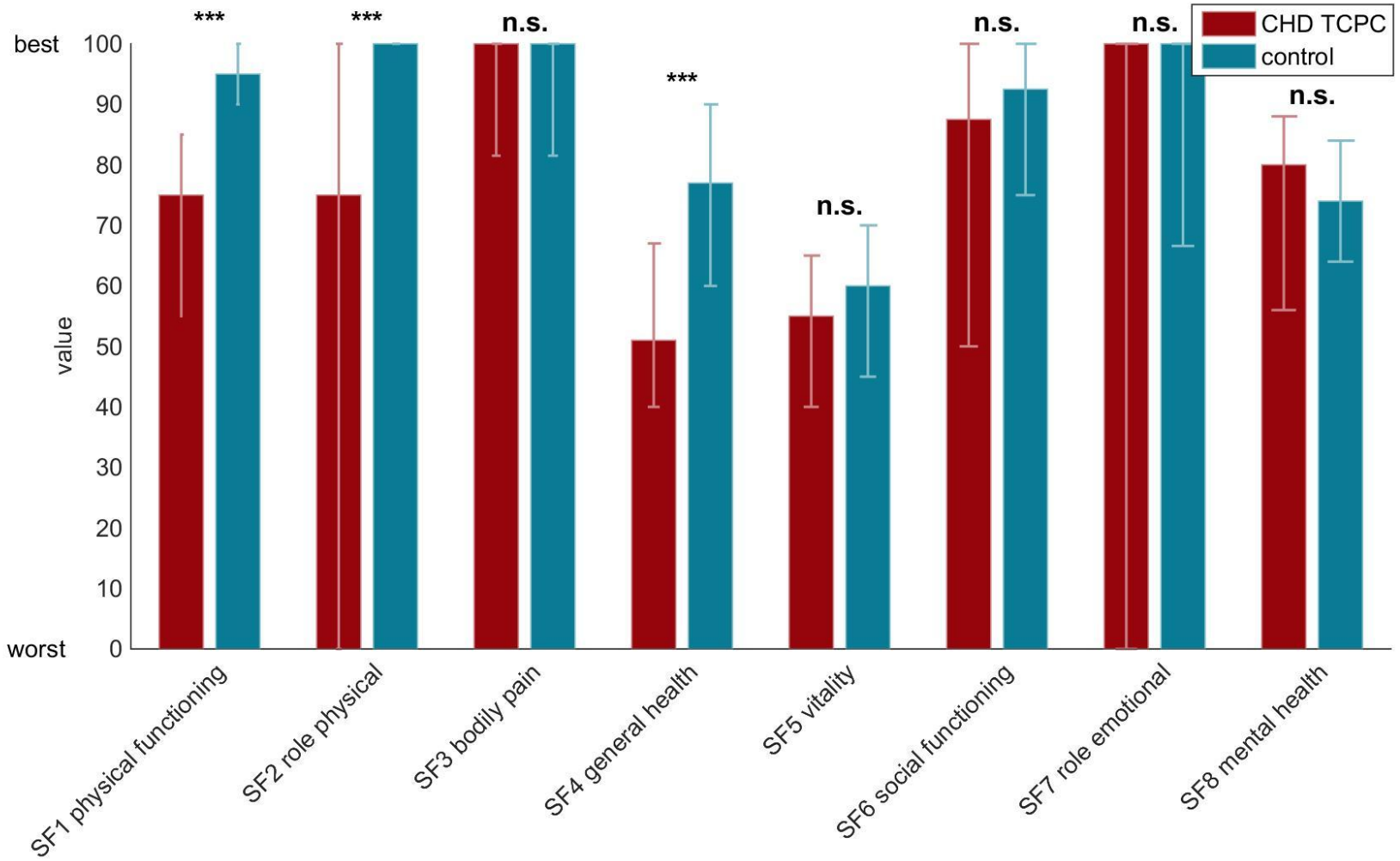
7x healthy children (= 54 %)

6x spontaneous abortion

Quality of life

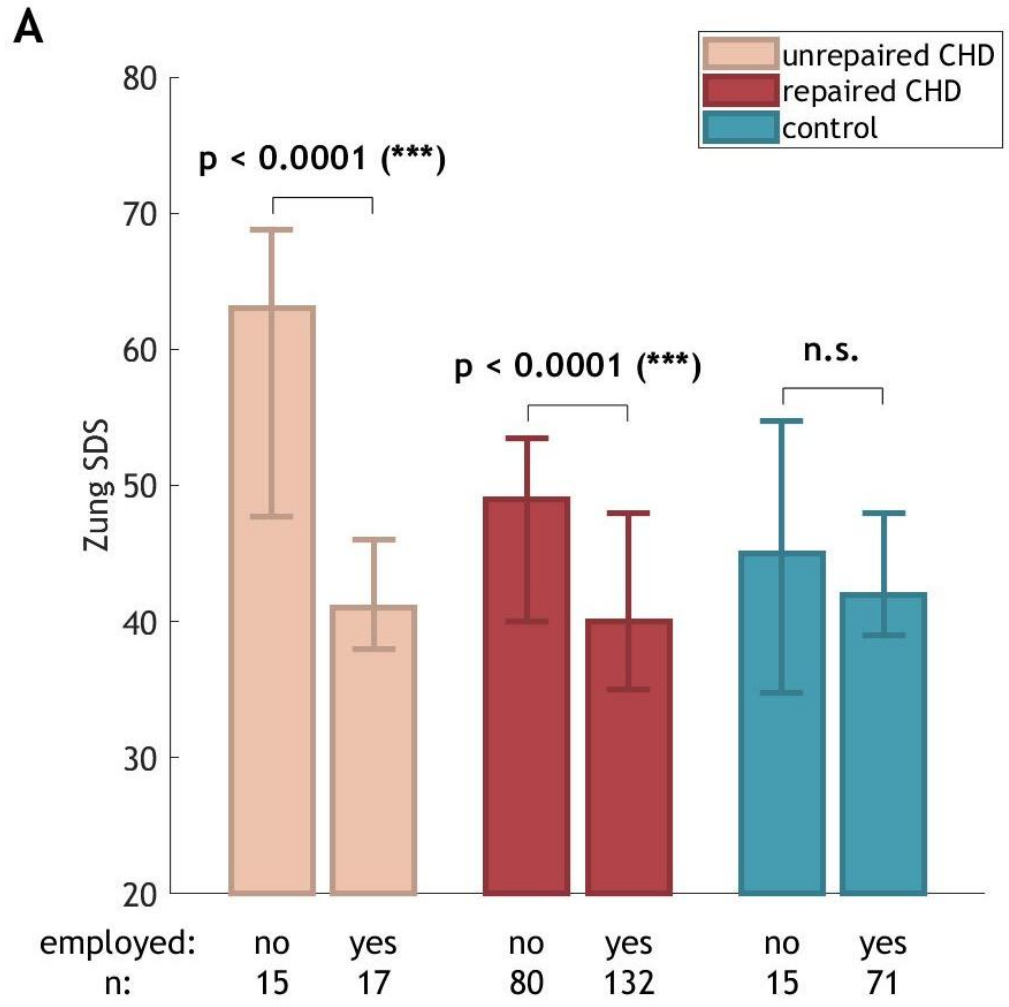
Quality of life in patients with TCPC and healthy population

SF-36 questionnary



Popelová, Cor Vasa, 2018

The role of employment for unrepaired and repaired complex cyanotic CHD



Patients with unrepaired cyanotic CHD without employment had the highest depression score
Also patients with repaired cyanotic CHD, (but not the controls) had higher depression score if unemployed.

Quality of life in repaired complex cyanotic CHD

223 adults with repaired complex cyanotic CHD

mean age 36 ± 10 years

Diagnoses: PA, TGA, TOF, SV (TCPC)

- **57 %** had higher education (secondary school or university)
- **46 %** had full-time job
- **96 %** were in NYHA class 1 or 2

- **40 %** had regular physical activity including recreational sports

- **31 %** women delivered a baby (39/95)

Živná, Rubáčková Popelová, 2017

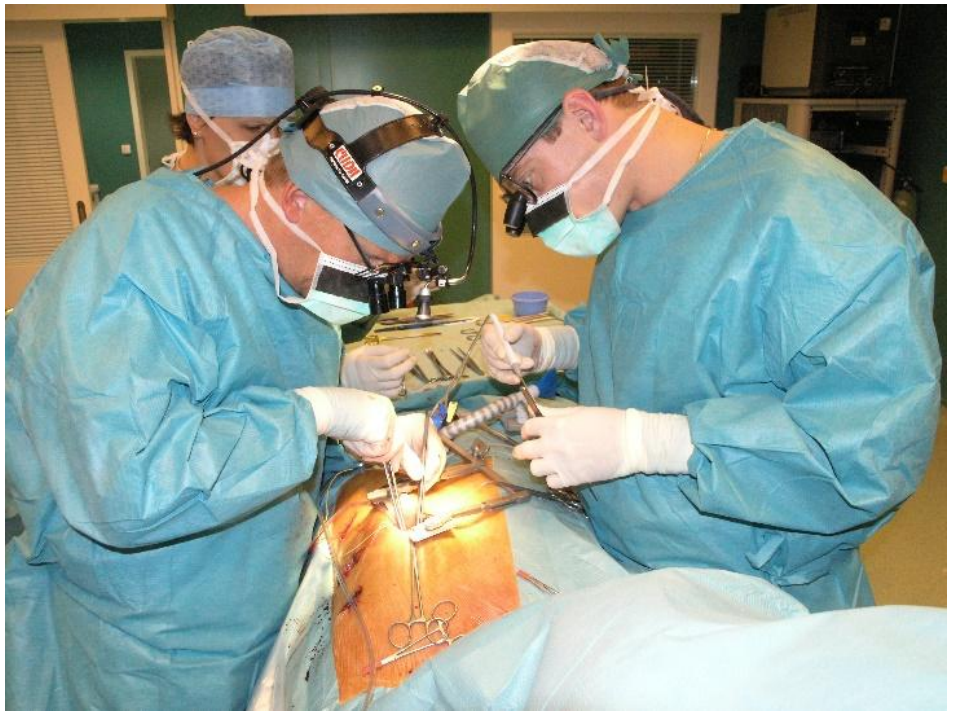
Do adults with CHD represent happy- end or growing problems…… ??

Lady with Ebstein anomaly type C,
born in 1946, operated in 68 yrs

Lady with TCPC

Lady with TOF
after reoperation

Lady with truncus
arteriosus and
6 cardiac operations



…. adults with CHD represent growing success….

….. based on the excellent results of Childern´s Heart Center together with

the effort of the Centers for adults and their co-operation….

Congratulations to 45 birthday !!!!

A lot of success !!!

Many thanks for the excellently operated patiens, for the co-operation and friendship….

Longitudinal follow-up with repeated NT-proBNP measurments

NT-proBNP	Number of pts with at least one sample exceeding this value	mortality
220 pg/ml (median of all patients)	388 pts with at least one NT-proBNP value ≤ 220 pg/ml	**1 %**
1548 pg/ml (median of deceased patients)	54 pts with at least one NT-proBNP value > 1548 pg/ml	**41 %**

Popelová, Am J Card, 2015