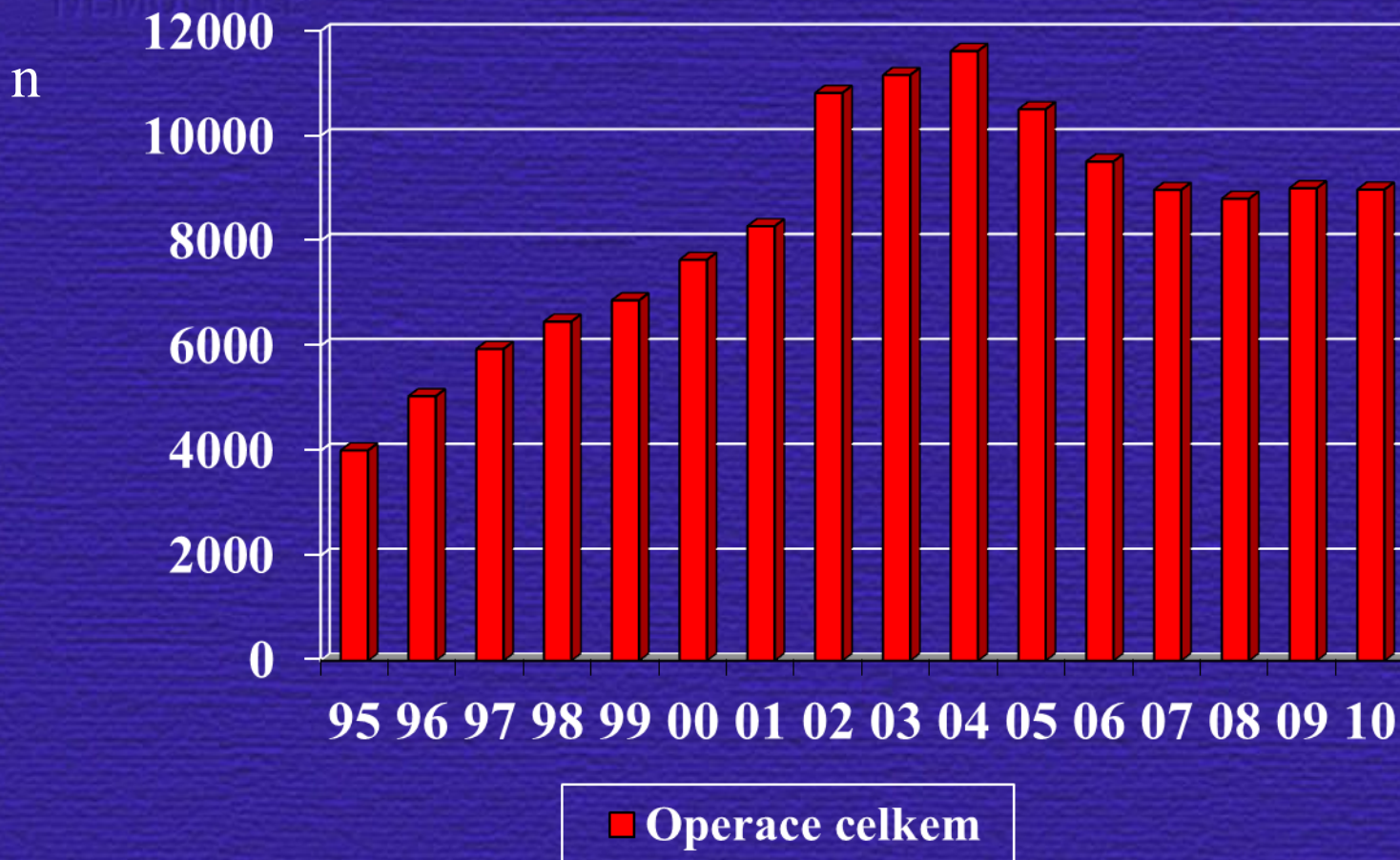


CHIRURGICKÁ LÉČBA VROZENÝCH SRDEČNÍCH VAD V DOSPĚLOSTI

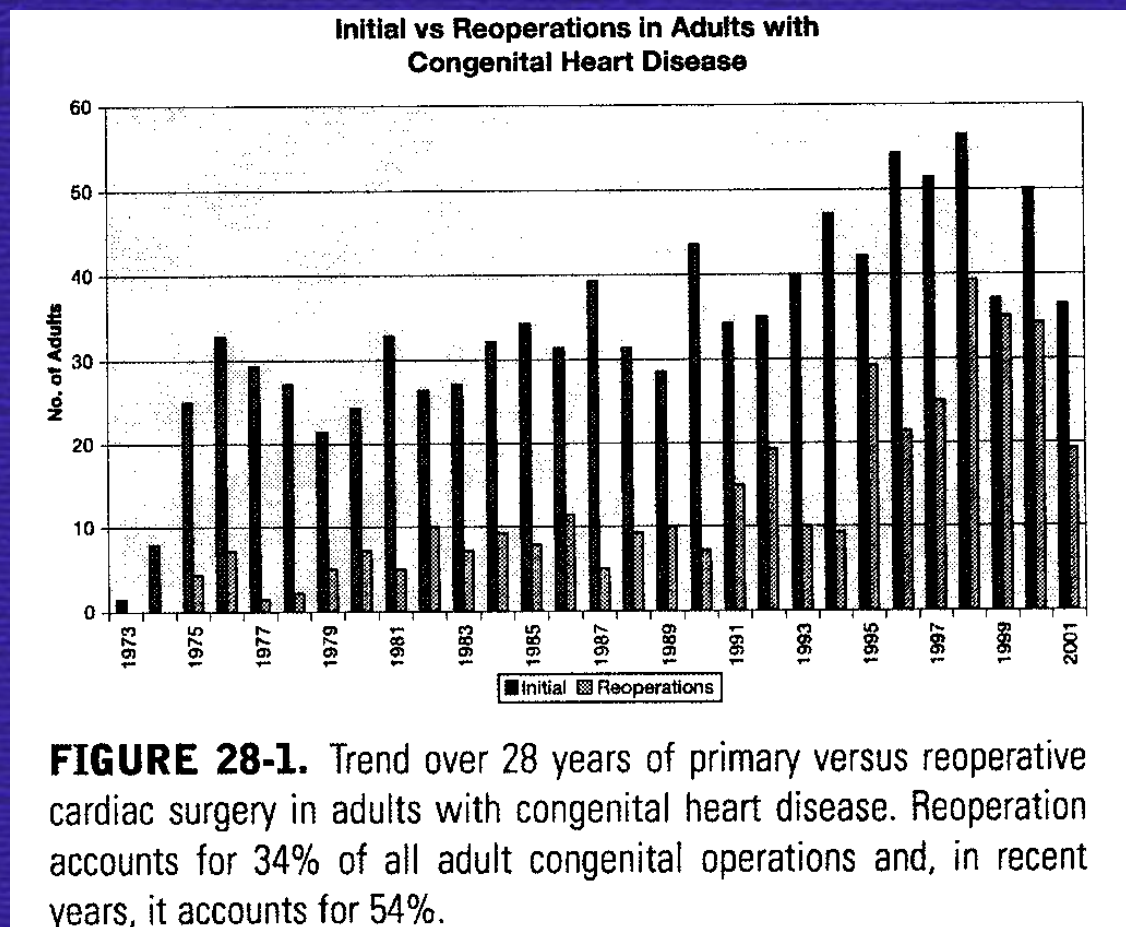


Černý Š., Gebauer R.
*Nemocnice Na Homolce, Praha,
Fakultní nemocnice Motol, Praha*

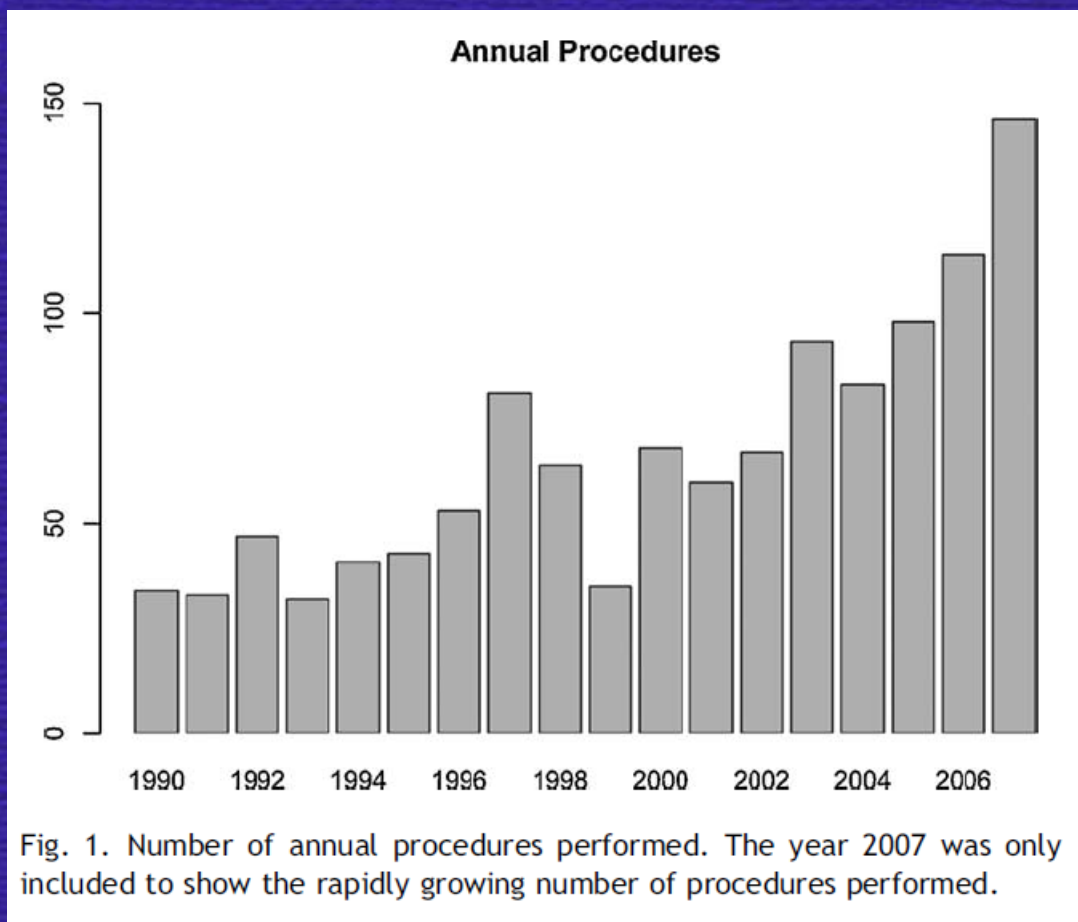
Počty dospělých KCH operací V ČR 1995 - 2010



Počty KCH operací dospělých pacientů s VSV- Toronto General

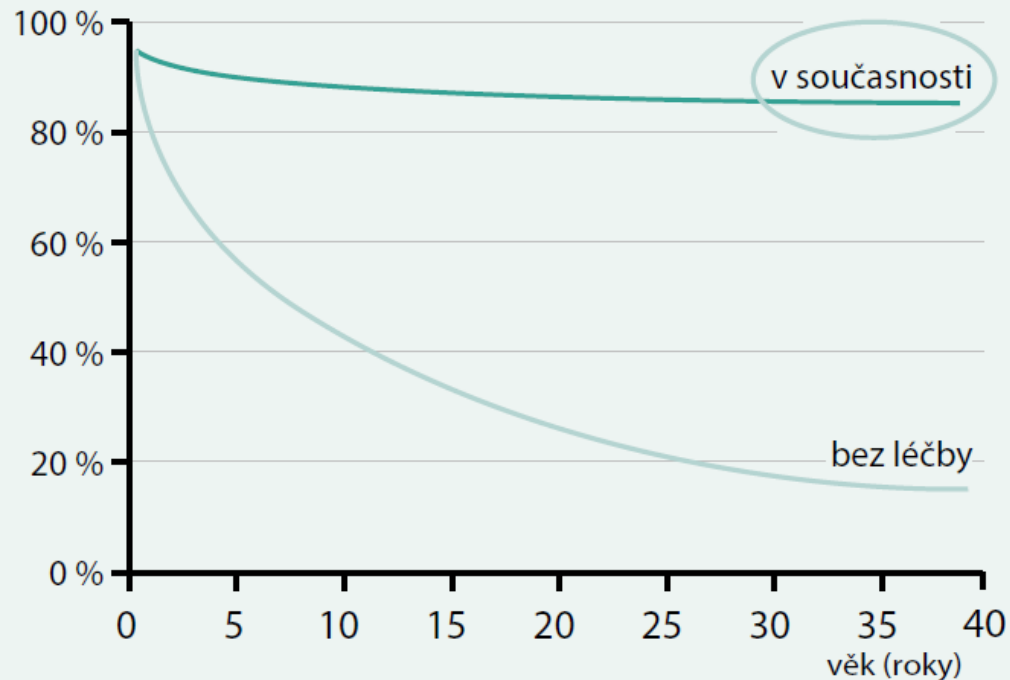


Počty KCH operací dospělých pacientů s VSV- Rotterdam



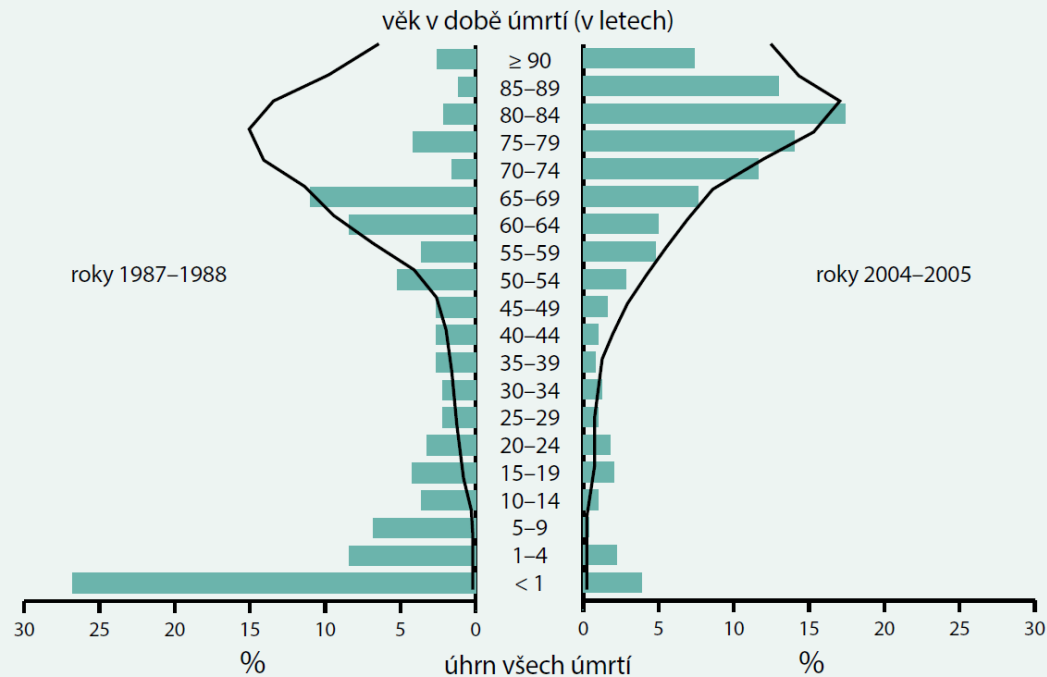
Přežívání nemocných s VSV

Graf 1. Vývoj přežívání pacientů s VSV při komplexní léčbě



Přežívání nemocných s VSV

Graf 2. Vývoj přežívání pacientů s VSV na míře komplexní péče



- V současné době v ČR cca 25.000 pacientů s VSV
- Tento počet bude v následujících letech narůstat a během 20 – 30 let dosáhne cca 60.000 pacientů
- Zhruba ½ těchto pacientů bude v budoucnu potřebovat další specializovanou péči včetně chirurgické

Může chirurgie VSV v dospělosti být řešením pro dospělá kardiochirurgická centra???

- Přes vzrůstající počet výkonů není celkový počet operací tak vysoký (světová velká centra cca 50 – 100 výkonů / rok)
- Superspecializovaná centra pro chirurgii VSV v dospělosti by měla pokrývat populaci cca 3-5 miliónů obyvatel
- Jedná se velmi často o komplexní výkony se složitou anatomií
- Dospělí kardiochirurgové nemají dostatečný trénink v chirurgii VSV
- Stále probíhá diskuze kdo a kde má KCH VSV v dospělosti provádět ...

Kdo a kde by měl provádět KCH léčbu VSV v dospělosti ?

Adult Congenital Heart Surgery: Adult or Pediatric Facility? Adult or Pediatric Surgeon?

Brian E. Kogon, MD, Courtney Plattner, BA, Traci Leong, PhD, Paul M. Kirshbom, MD, Kirk R. Kanter, MD, Mike McConnell, MD, and Wendy Book, MD

Divisions of Cardiothoracic Surgery and Cardiology, Emory University School of Medicine, Rollins School of Public Health, and Sibley Cardiology, Children's Healthcare of Atlanta, Atlanta, Georgia

Background. One of the current controversies in the field of adult congenital heart disease is whether patients should be cared for at an adult or pediatric facility and by an adult or pediatric heart surgeon. After transitioning our program from the children's hospital to the adult hospital, we analyzed our experience with each system.

Methods. Between 2000 and 2007, 303 operations were performed on adults (age ≥ 18 years) with congenital heart disease. One hundred eighty-five operations were performed in an adult hospital and 118 in a pediatric hospital. Forty-six operations were performed by an adult heart surgeon and 257 by a congenital heart surgeon.

Results. Mean age, coexisting medical problems, and preoperative risk factors were higher in both the adult hospital group and adult surgeon group compared with the respective pediatric groups. Mortality was similar at the adult and pediatric hospitals (4.3% versus 5.1%), but

was markedly higher in the adult surgeon group compared with the pediatric surgeon group (15.2% versus 2.7%; $p = 0.0008$). By multivariate analysis, risk factors for mortality included older age at the time of surgery ($p = 0.028$), surgery performed at a children's hospital ($p = 0.013$), and surgery performed by an adult heart surgeon ($p = 0.0004$).

Conclusions. Congenital heart surgery can be performed in adults with reasonable morbidity and mortality. Caring for an anticipated aging adult congenital population with increasingly numerous coexisting medical problems and risk factors is best facilitated in an adult hospital setting. Also, when surgery becomes necessary, these adult patients are best served by a congenital heart surgeon.

(Ann Thorac Surg 2009;87:833-40)

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Kdo a kde by měl provádět KCH léčbu VSV v dospělosti ?

National Practice Patterns for Management of Adult Congenital Heart Disease

Operation By Pediatric Heart Surgeons Decreases In-Hospital Death

Tara Karamlou, MD; Brian S. Diggs, PhD; Thomas Person, MD;
Ross M. Ungerleider, MD, MBA; Karl F. Welke, MD, MS

Background—Surgery for grown-up (age ≥ 18 years) patients with congenital heart disease (GUCH) is frequently performed by surgeons without specialization in pediatric heart surgery. We sought to define national practice patterns and to determine whether outcomes for GUCH patients are improved if they are treated by specialized pediatric heart surgeons (PHSs) compared with non-PHSs.

Methods and Results—We identified index cardiac procedures in patients with 12 congenital heart disease diagnostic groups using the Nationwide Inpatient Sample 1988 to 2003. PHSs were defined as surgeons whose annual practice volumes were made of $>75\%$ annual pediatric heart cases. GUCH operations were defined as operations within these 12 diagnoses occurring in patients ≥ 18 years of age. We identified 30 250 operations, yielding a national estimate of $152\,277 \pm 7875$ operations. Of these, $111\,816 \pm 7456$ (73%) were pediatric operations, and $40\,461 \pm 1365$ (27%) were GUCH operations. PHSs performed 68% of pediatric operations in all diagnostic groups, whereas non-PHSs performed 95% of GUCH operations within the same diagnostic groups ($P < 0.0001$). In-hospital death rates for GUCH patients operated on by PHSs were lower than death rates for GUCH patients operated on by non-PHSs (1.87% [95% CI, 0.62 to 3.13] versus 4.84% [95% CI, 4.30 to 5.38%]; $P < 0.0001$). Survival advantage increased with increasing surgeon annual pediatric volume ($P = 0.0031$).

Conclusions—Pediatric patients within specific diagnostic groups are more likely to undergo operation by PHSs, whereas GUCH patients within the same diagnostic groups are more likely to undergo operation by non-PHSs. In-hospital death rates are lower for GUCH patients operated on by PHSs. GUCH patients should be encouraged to obtain surgical operation by PHS. (Circulation. 2008;118:2345-2352.)

Key Words: epidemiology ■ heart defects, congenital ■ heart septal defects ■ statistics

Kdo a kde by měl provádět KCH léčbu VSV v dospělosti ?

Cardiac surgery in adults performed at children's hospitals: Trends and outcomes

William T. Mahle, MD,^a Paul M. Kirshbom, MD,^b Kirk R. Kanter, MD,^b and Brian M. Kogon, MD^b

The Journal of Thoracic and Cardiovascular Surgery • Volume 136, Number 2 307

Objective: The number of adults with congenital heart disease who require cardiac surgery is projected to increase dramatically. Controversy exists as to whether such procedures should be performed in pediatric centers, which generally have the greatest experience with operations for congenital heart disease. We sought to report the outcomes for cardiac surgery performed in adults (≥ 21 years of age) at children's hospitals and determine how these practices varied among institutions.

Methods: Data from July 2005 to June 2007 from the Child Health Corporation of America, a consortium of 37 free-standing children's hospitals, were analyzed to determine the institutional volume, type of cardiac procedure, outcome, and hospital charges. Individual institutional variables were analyzed to determine which factors might be associated with the practice of performing adult cardiac surgery in children's hospitals.

Results: During the study period, there were 719 admissions for cardiac surgery in adults at Child Health Corporation of America institutions. The median age at the time of operation was 26 years (range, 21–86 years). The most common surgical procedures were implantation or revision of a pacemaker or defibrillator ($n = 207$ [29.2%]), pulmonary valve replacement ($n = 119$ [16.8%]), aortic valve replacement ($n = 59$ [8.3%]), and Fontan revision ($n = 37$ [5.2%]). The median hospital length of stay was 6 days (range, 1–175 days). The hospital mortality was 1.9%. Comorbid conditions likely to require other subspecialty care were present in more than 30% of patients. Among the Child Health Corporation of America centers, adult operations as a proportion of overall cardiac operations varied from 0% to 10.9%. There was no relationship between overall cardiac surgical volume and proportion of adult cases performed in Child Health Corporation of America centers.

Conclusions: A significant number of adult cardiac surgical procedures are being performed at children's hospitals with excellent results. The majority of procedures are not related to complex shunt lesions but rather pacemaker/defibrillator implantation and semilunar valve surgery. Whether adult patients with congenital heart disease should continue to undergo most cardiac surgery in children's hospitals is worthy of discussion.

University Health Network

Toronto General Hospital
Toronto Western Hospital
Princess Margaret Hospital
Toronto Medical Laboratories



STEPAN CERNY
CLINICAL FELLOW
CARDIOVASCULAR SURGERY



TABLE 28-2. Initial Diagnosis in Reoperated Adults with Prior Intracardiac Repair of CHD

<i>Diagnosis at Previous Repair</i>	<i>N</i>	<i>% of Total</i>
Tetralogy repair	139	42
Subaortic stenosis*	31	9
Fontan	30	9
ASD repair†	29	9
VSD repair	20	6
Coarctation repair	5	2
Other	75	23
Total	329	100



Williams WG et al; In: Advanced Therapy in Cardiac Surgery, 2003

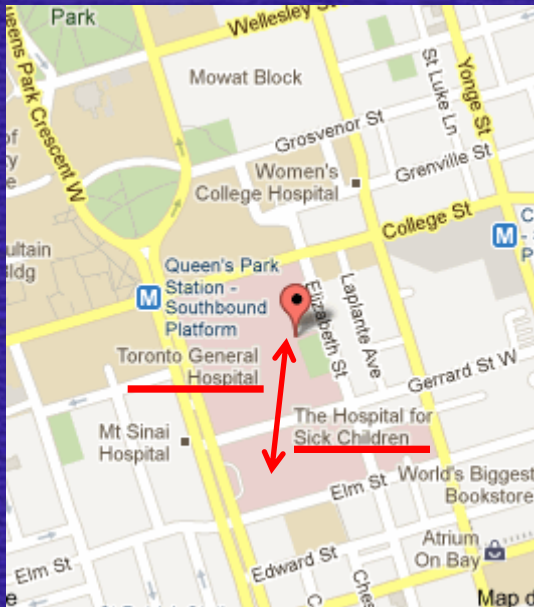
Pulmonary valve replacement in repaired tetralogy of Fallot: Determinants of early postoperative adverse outcomes

Laura Dos, MD,^{a,c} Alexander Dadashev, MD,^a David Tanous, MBBS,^a Ignacio J. Ferreira-González, MD,^d Kim Haberer,^a Samuel C. Siu, MD,^a Glen S. Van Arsdell, MD,^b Erwin N. Oechslin, MD,^a William G. Williams, MD,^b and Candice K. Silversides, MD^a

The Journal of Thoracic and Cardiovascular Surgery • Volume 138, Number 3

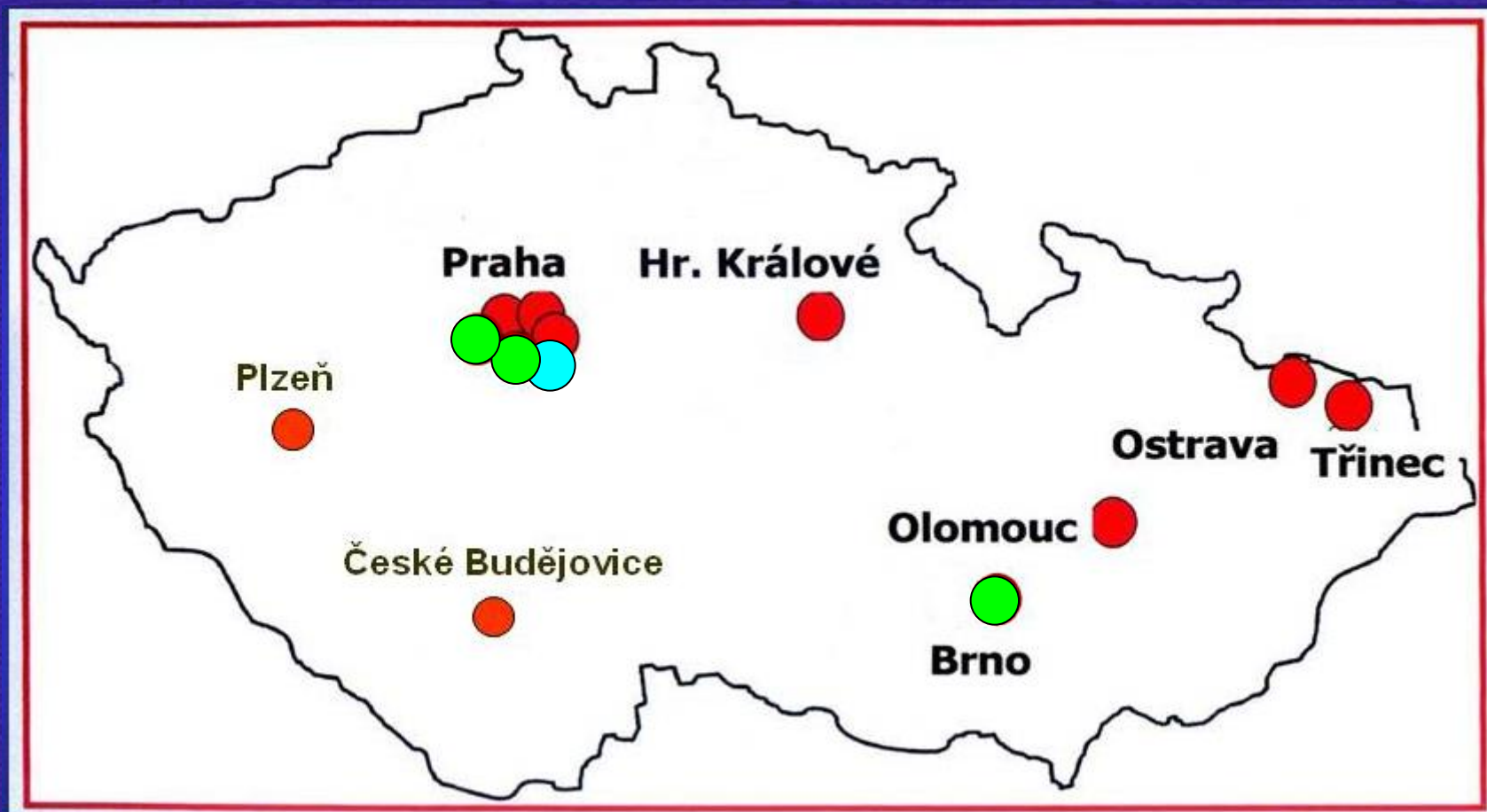
553

Toronto Congenital Cardiac Center for Adults



- Toronto General Hospital + Hospital for Sick Children
- Všichni pacienti jsou sledováni týmem specializovaných kardiologů
- Všichni pacienti jsou operováni dětským (kongenitálním) kardiochirurgiem na oddělení dospělé kardiochirurgie
- Tým intenzivistů na JIP je doplněn lékařem se znalostí problematiky komplexních VSV

KCH VSV v dospělosti v České republice

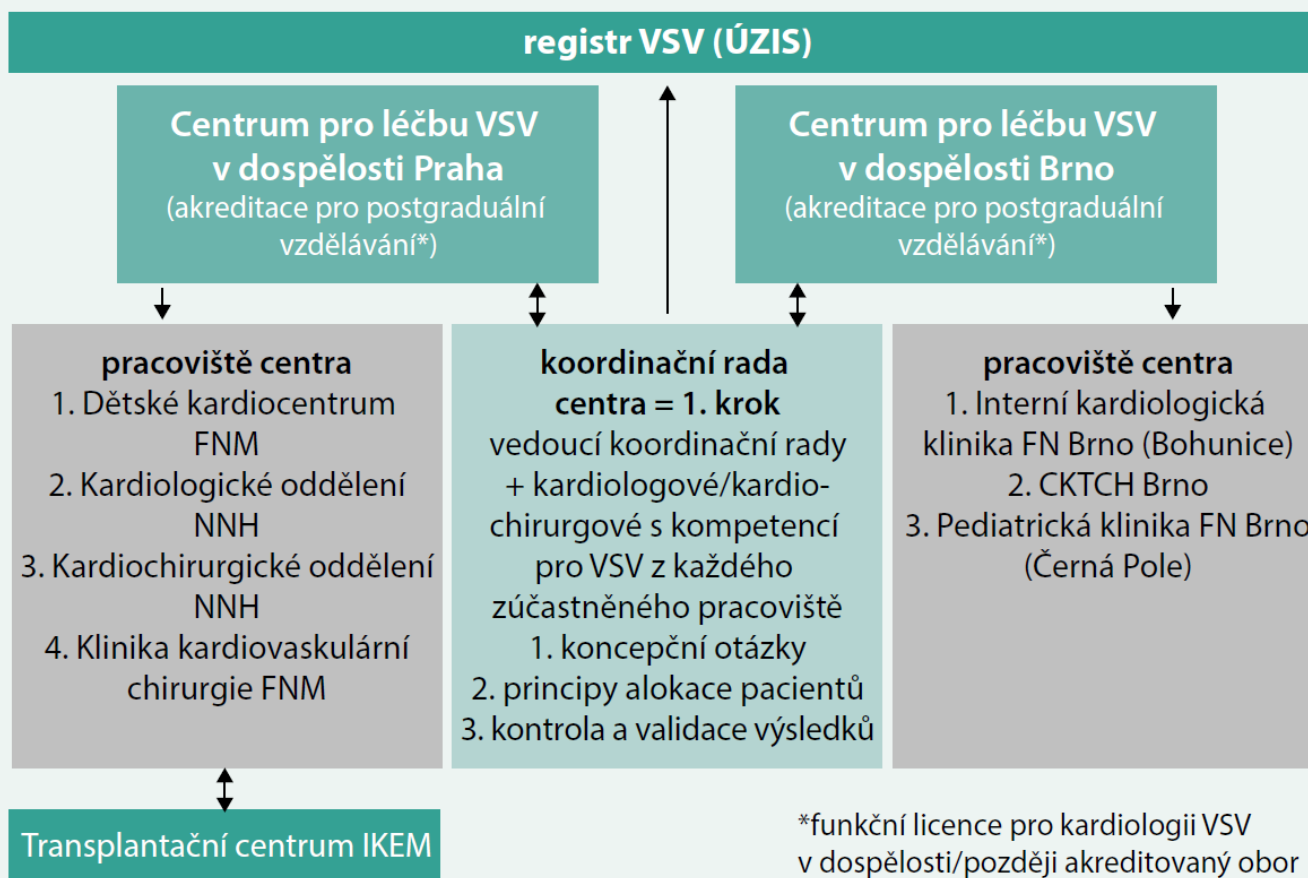


2016:	Dospělá kardiokirurgie	12	●
	Dětská kardiokirurgie	1	●
	KCH VSV v dospělosti (NNH, FNM, CKTCH)	3	●

Česká republika – 10,5 mil. obyvatel

KCH VSV v dospělosti v České republice

Schéma. Návrh organizační struktury center pro léčbu vrozených srdečních vad (VSV) v dospělosti



Chirurgická léčba VSV v dospělosti

Rozdělení vad

- **Dle komplexnosti vad:**
 - Vady jednoduché (běžné)
 - Vady vzácné a komplexní
- **Dle chirurgické léčby:**
 - Primární (nediagnostikované nebo nekorigované) vady
 - Reziduální léze po předchozí korekci

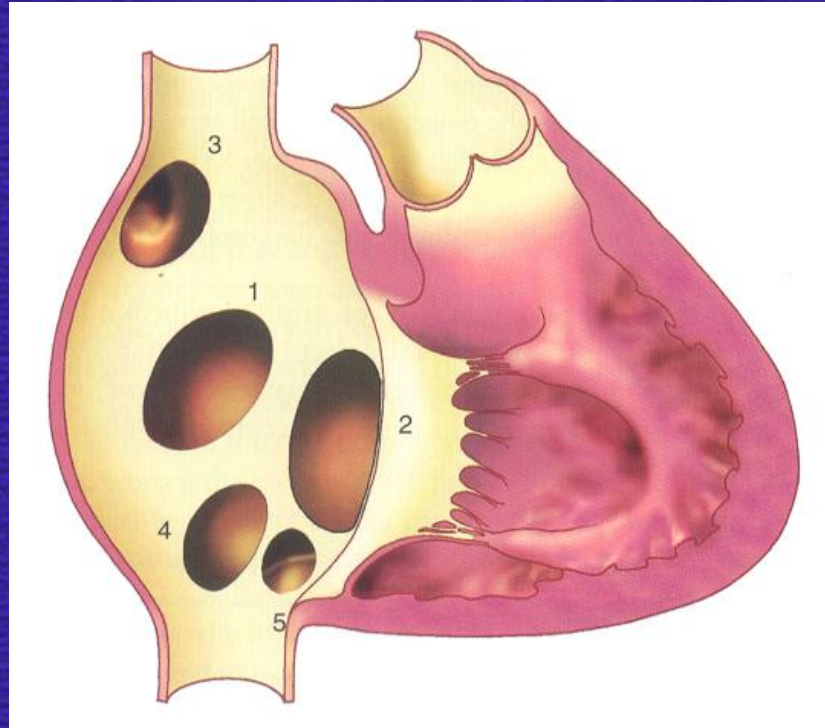
Chirurgická léčba VSV v dospělosti

Rozdělení dle chirurgické léčby

- **Primární (nediagnostikované nebo nekorigované) vady:**
 - Vada nebyla diagnostikována v dětství a projevila se až v dospělosti
 - Vada byla diagnostikována v dětství ale nebyla operována pro malou hemodynamickou závažnost
 - Vada byla dříve považována za inoperabilní, ale v současné době je s vyšším rizikem operabilní

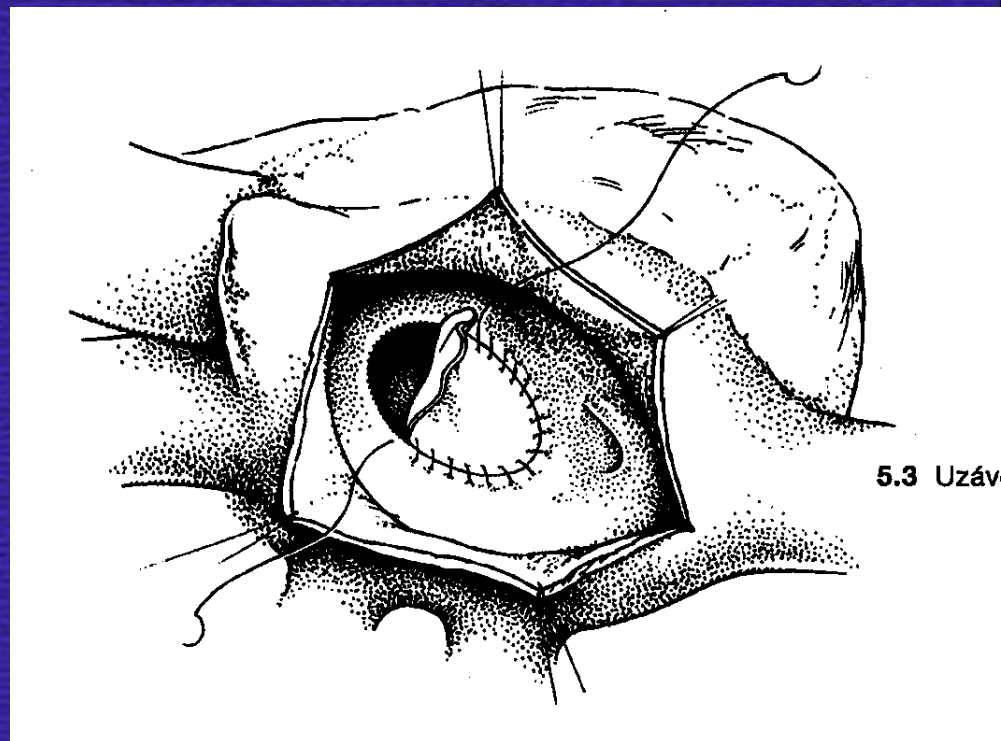
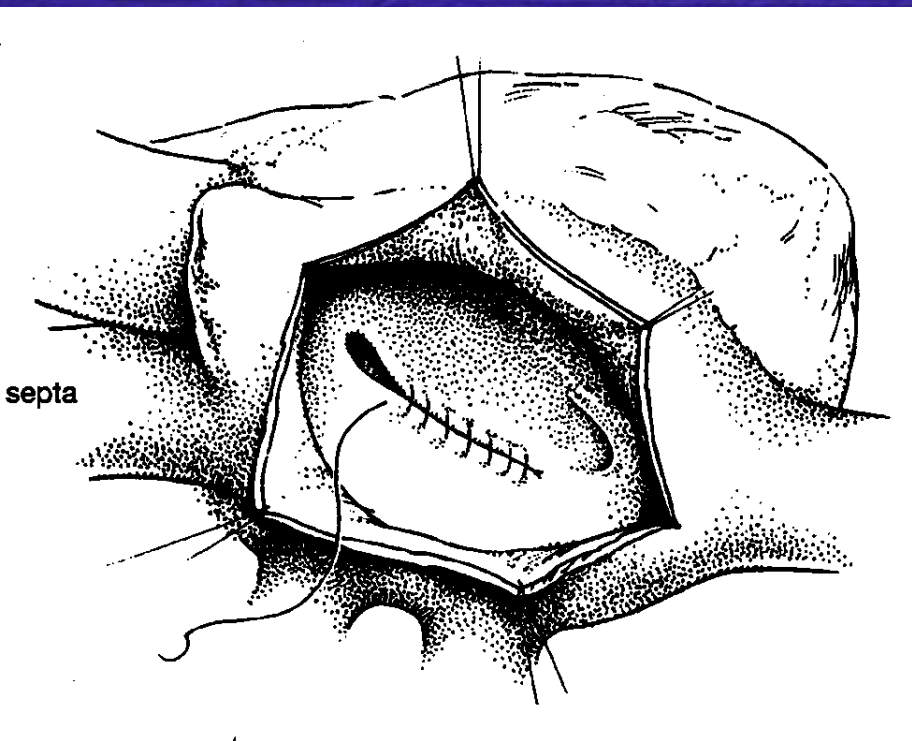
Primární VSV v dospělosti

Defekt septa síňí typu secundum



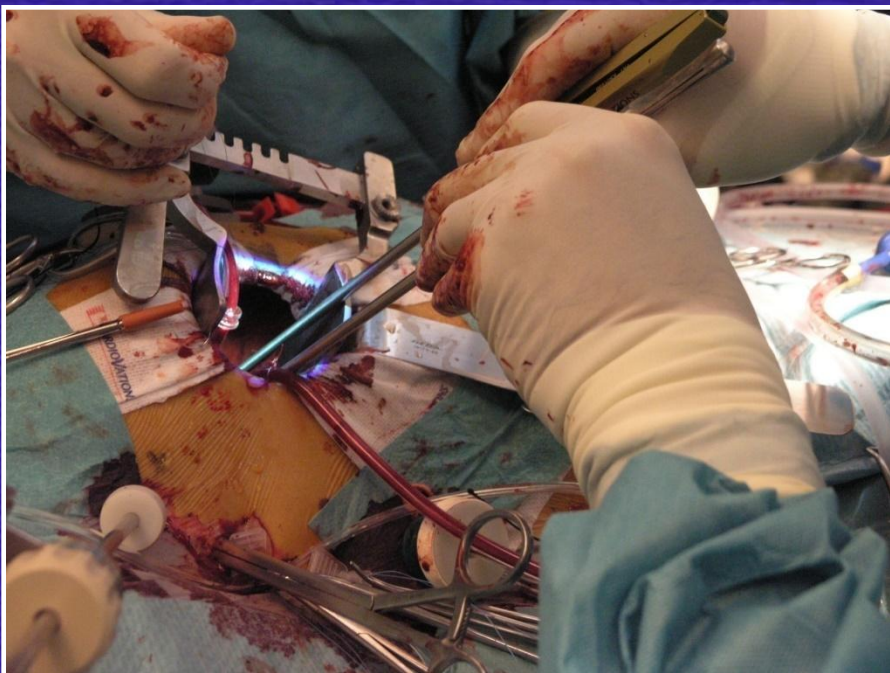
Primární VSV v dospělosti

Defekt septa síní typu secundum



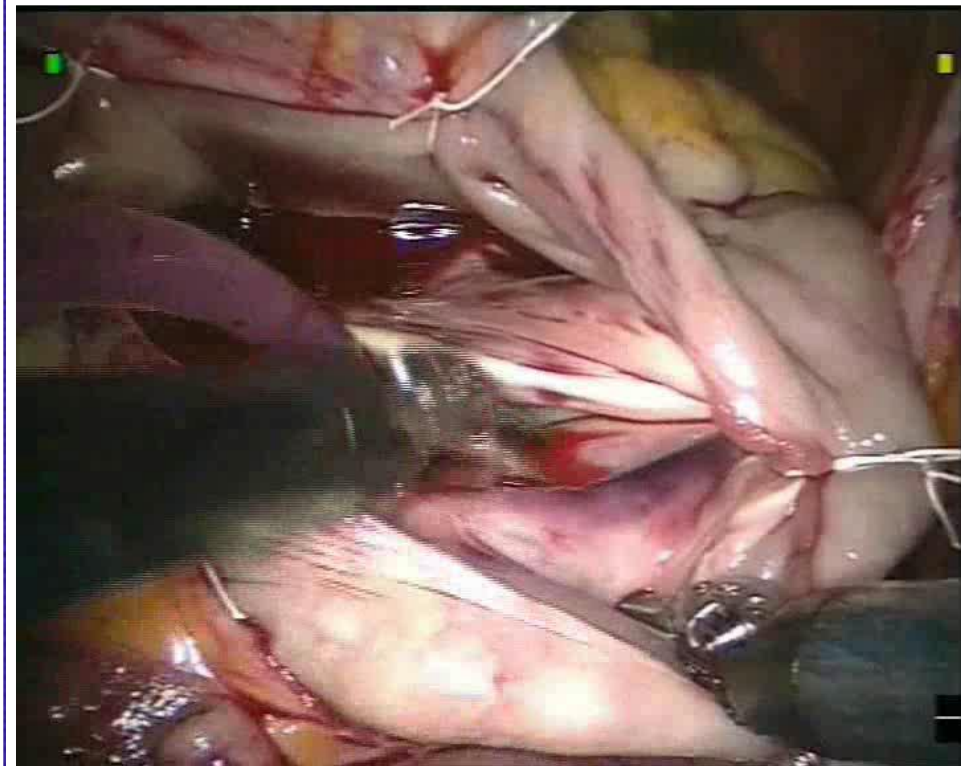
Primární VSV v dospělosti

Defekt septa síňí typu secundum



Primární VSV v dospělosti

Defekt septa síňí typu secundum



Primární VSV v dospělosti

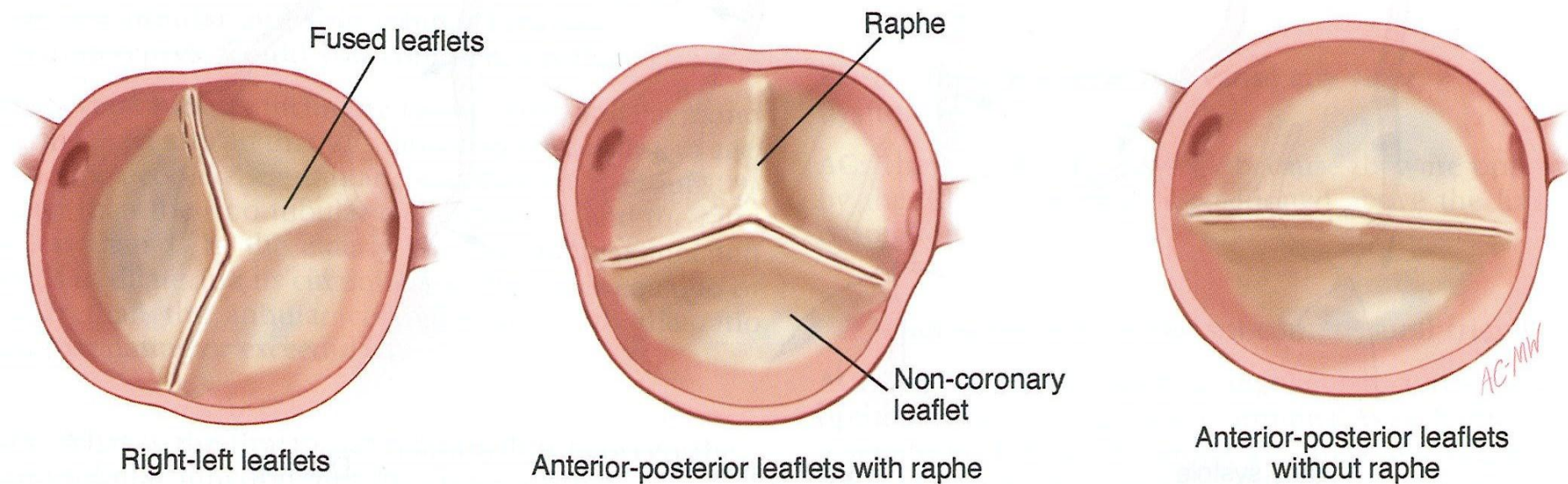
Defekt septa síní typu secundum



Primární VSV v dospělosti

Patologie aortální chlopně a kořene aorty

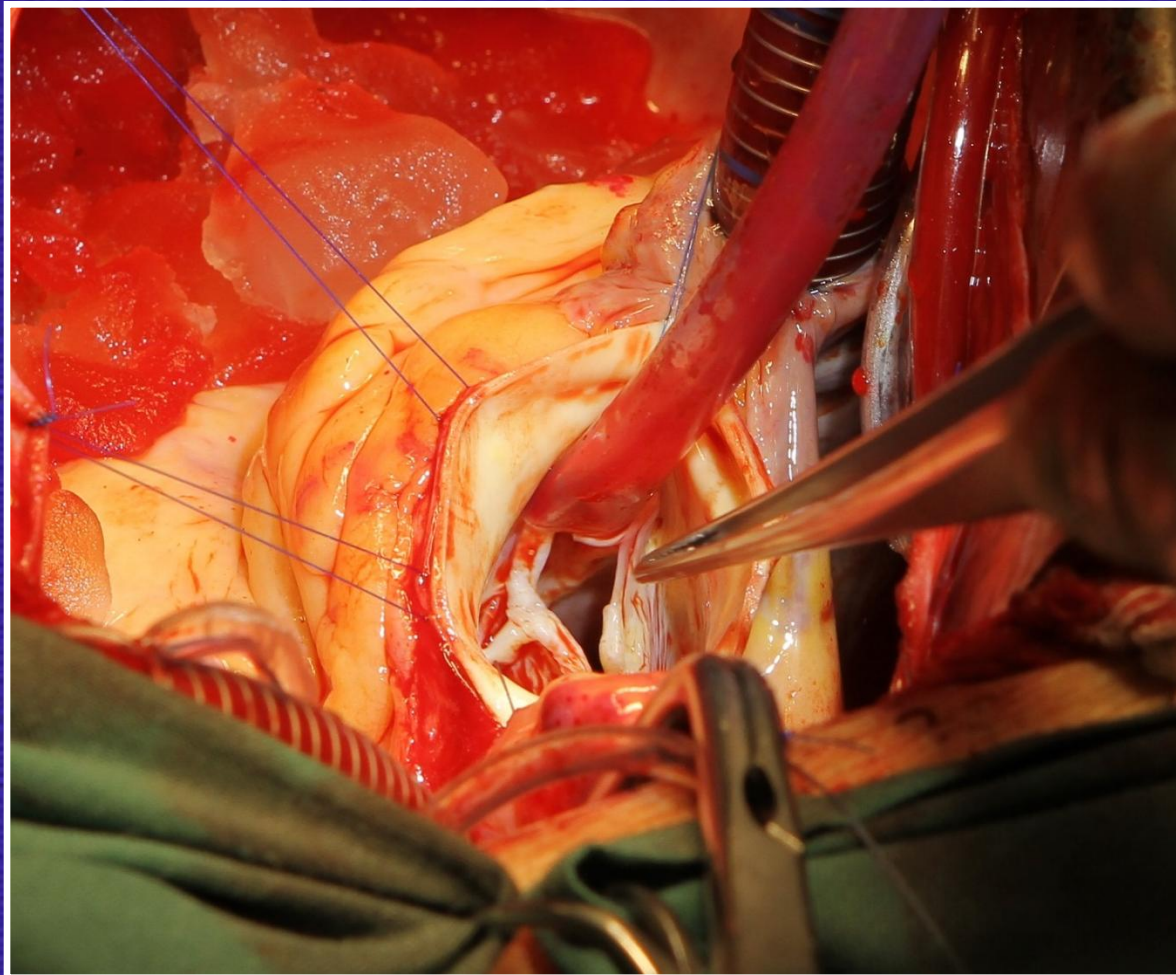
BAV



Primární VSV v dospělosti

Patologie aortální chlopně a kořene aorty

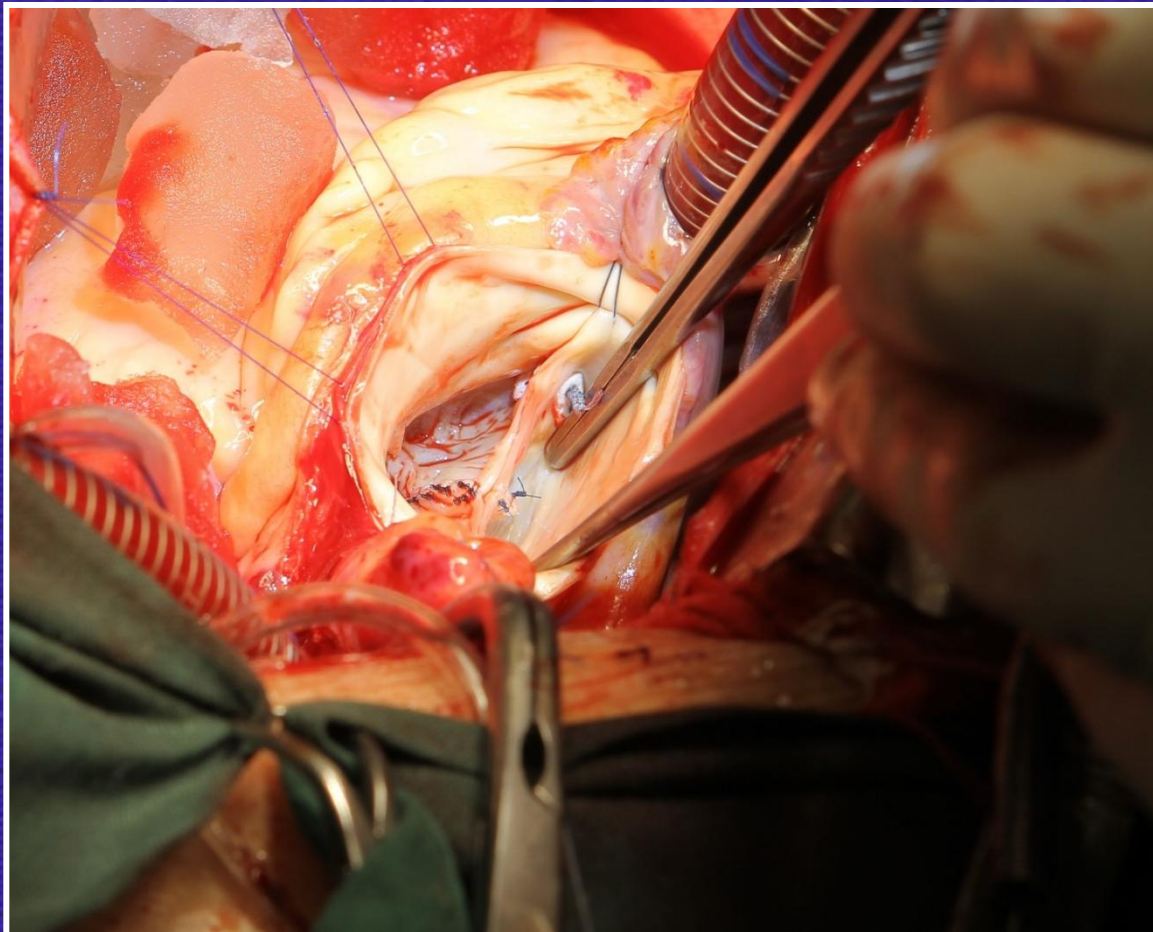
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Primární VSV v dospělosti

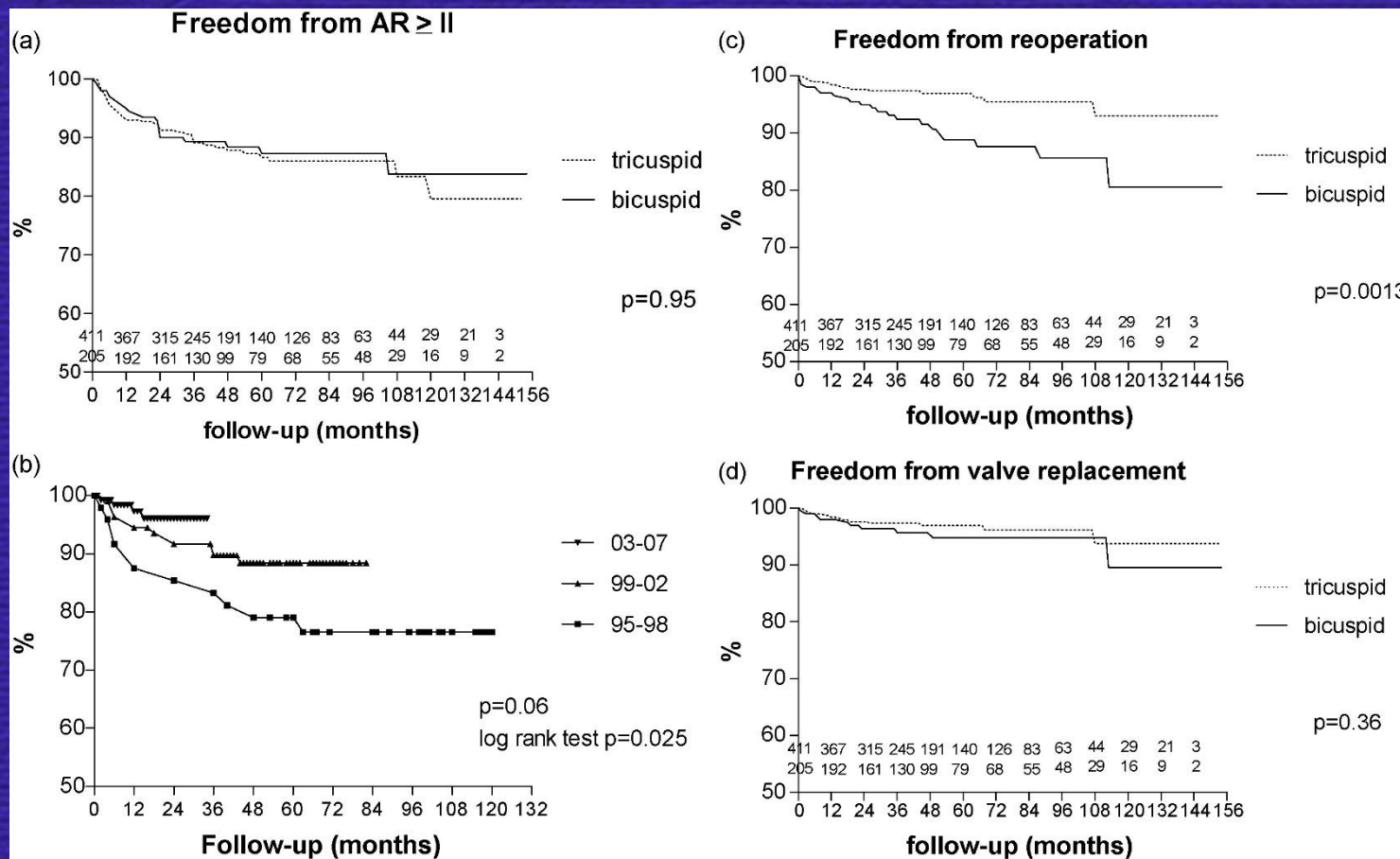
Patologie aortální chlopně a kořene aorty

BAV



Primární VSV v dospělosti

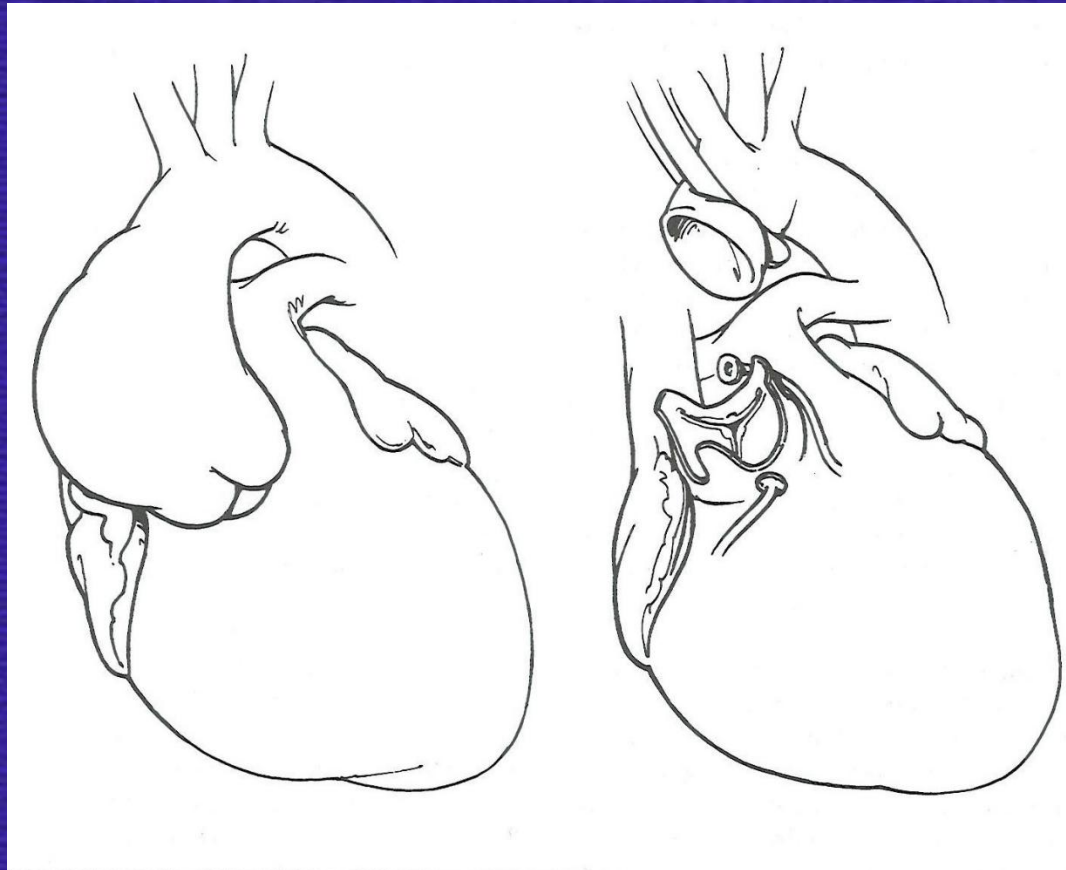
Patologie aortální chlopně a kořene aorty



Primární VSV v dospělosti

Patologie aortální chlopně a kořene aorty

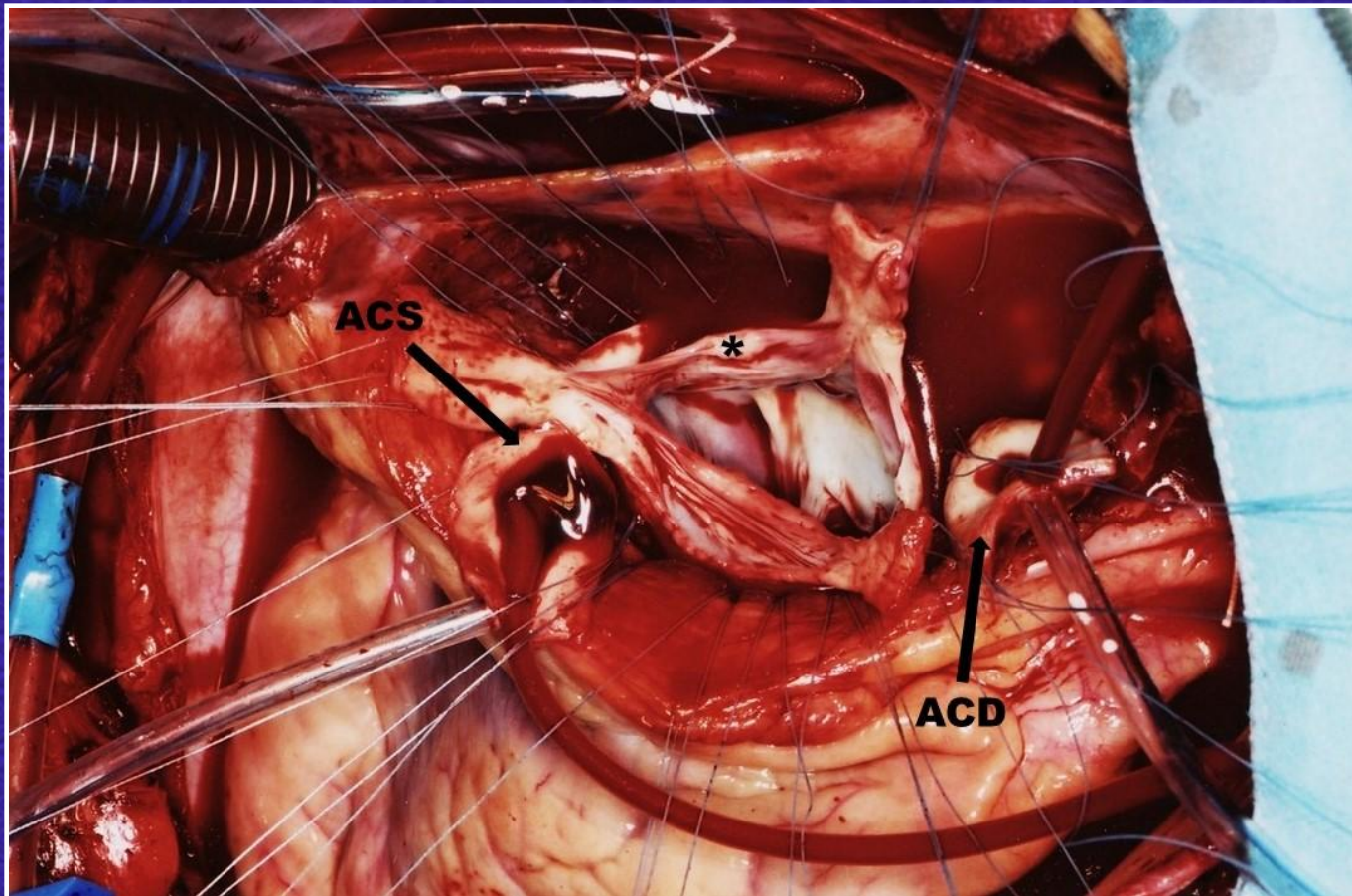
Marfanův syndrom



Primární VSV v dospělosti

Patologie aortální chlopně a kořene aorty

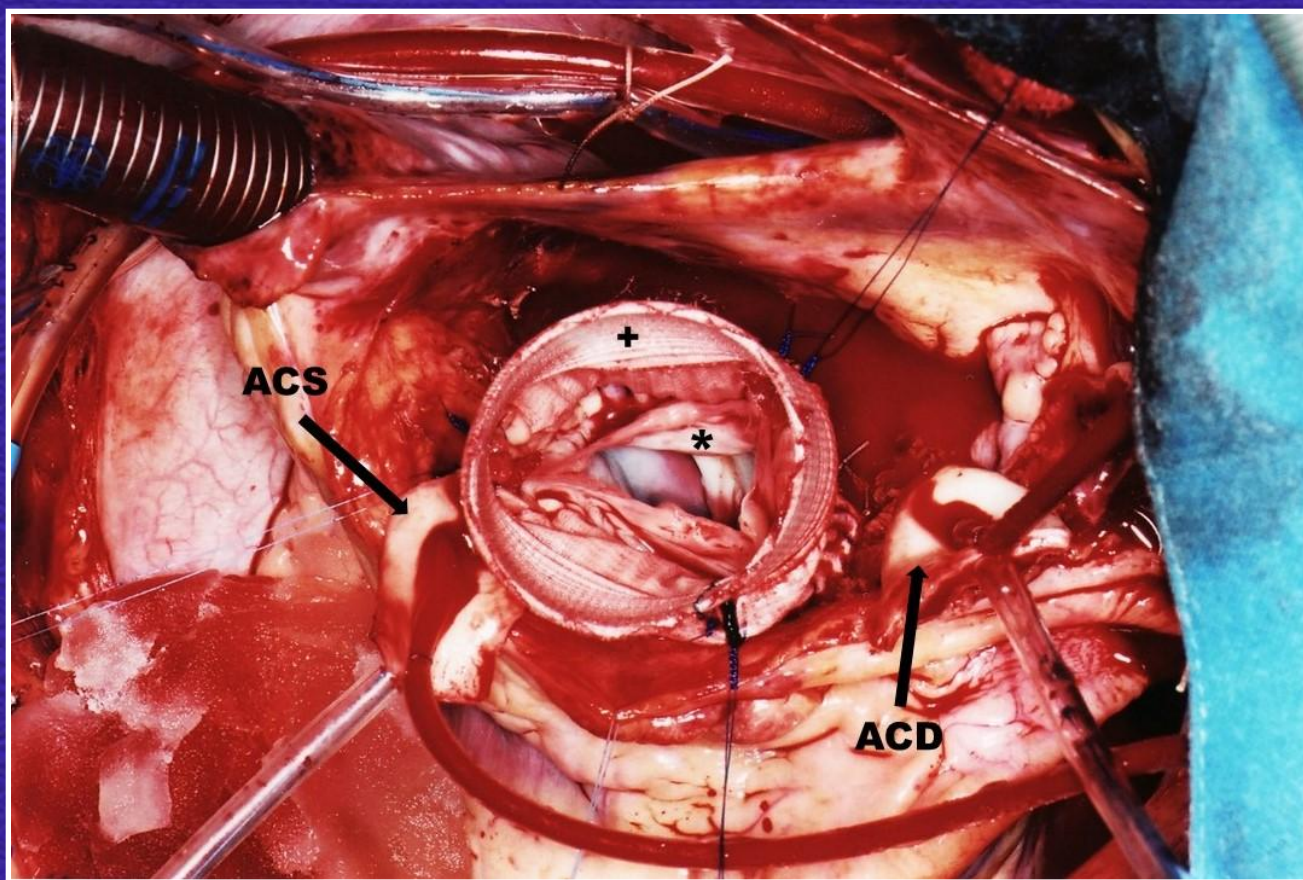
Marfanův syndrom



Primární VSV v dospělosti

Patologie aortální chlopně a kořene aorty

Marfanův syndrom



Primární VSV v dospělosti

Patologie aortální chlopně a kořene aorty

Marfanův syndrom

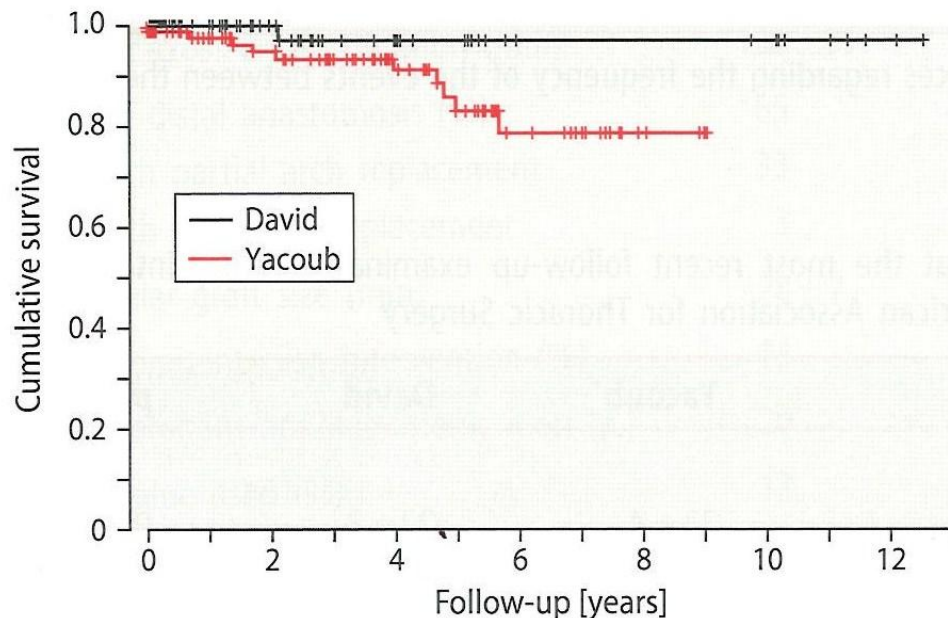
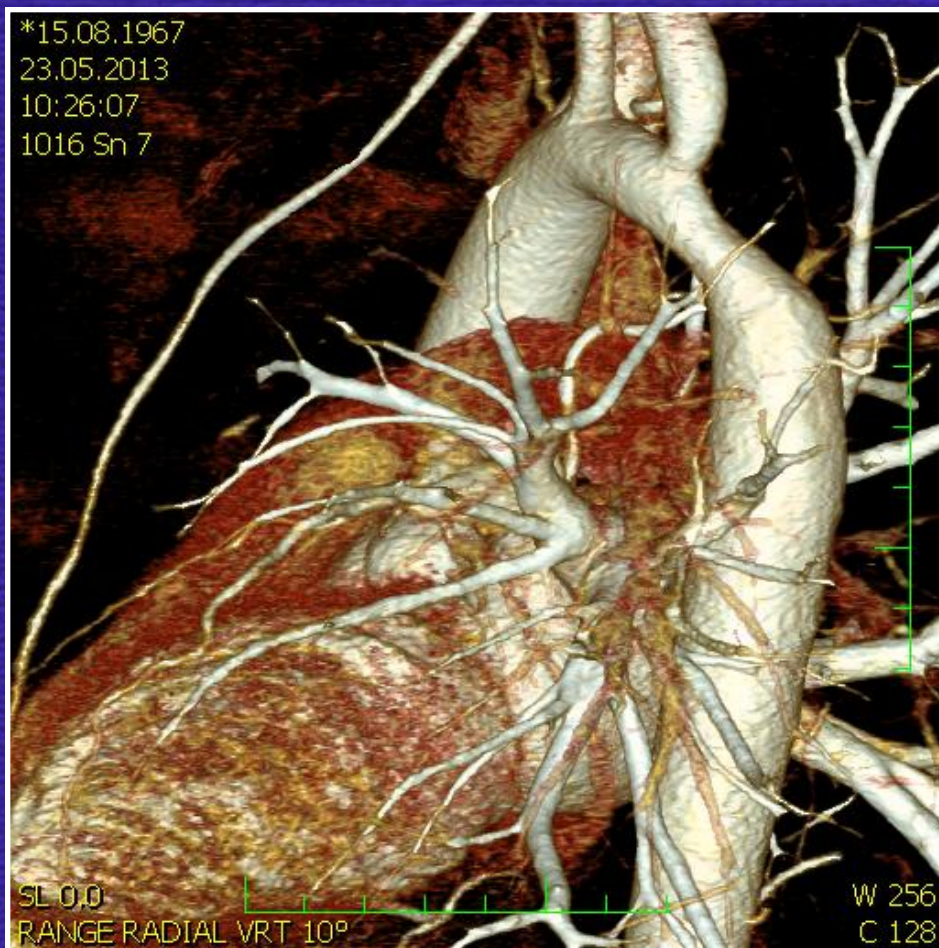


Fig. 4. Valve-related reoperations: actuarial freedom from valve-related reoperations in all patients undergoing valve-sparing aortic root replacement, stratified by technique. Censored patient data are indicated by ticks to the curves. The difference is not quite significant ($p=0.065$)

Primární VSV v dospělosti

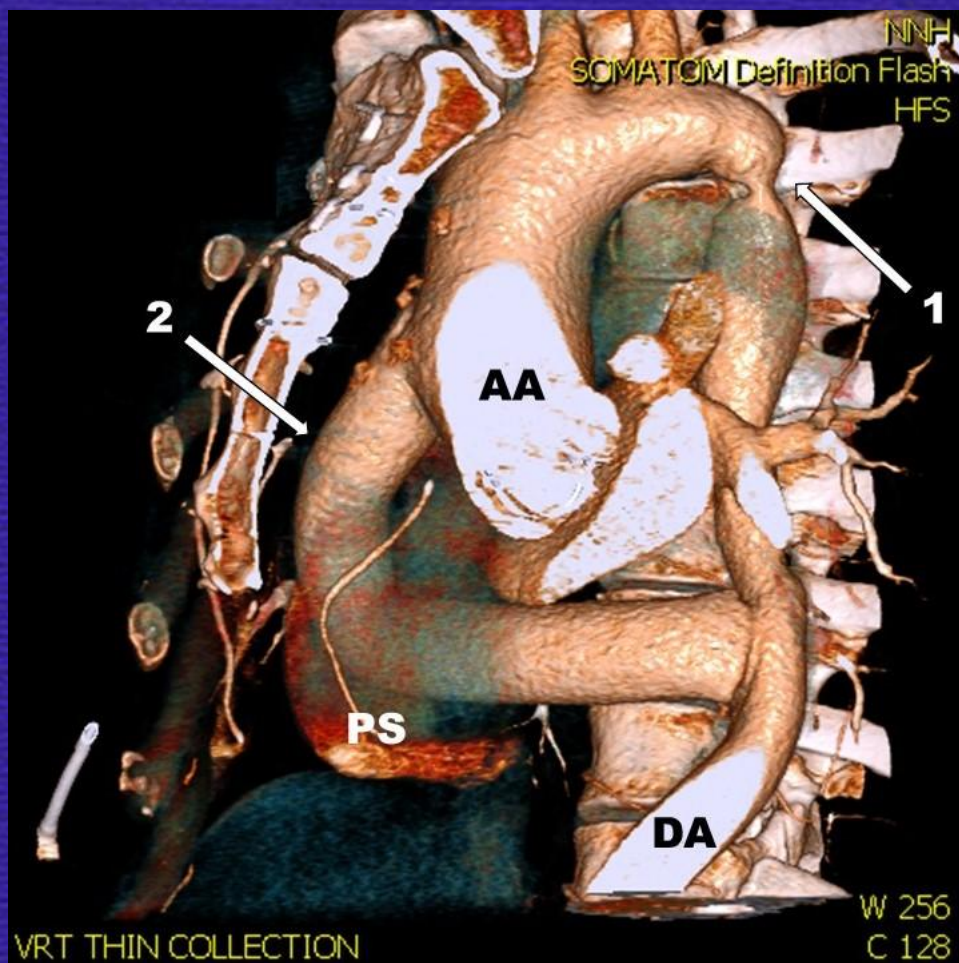
Koarktace aorty



Primární VSV v dospělosti

Koarktace aorty

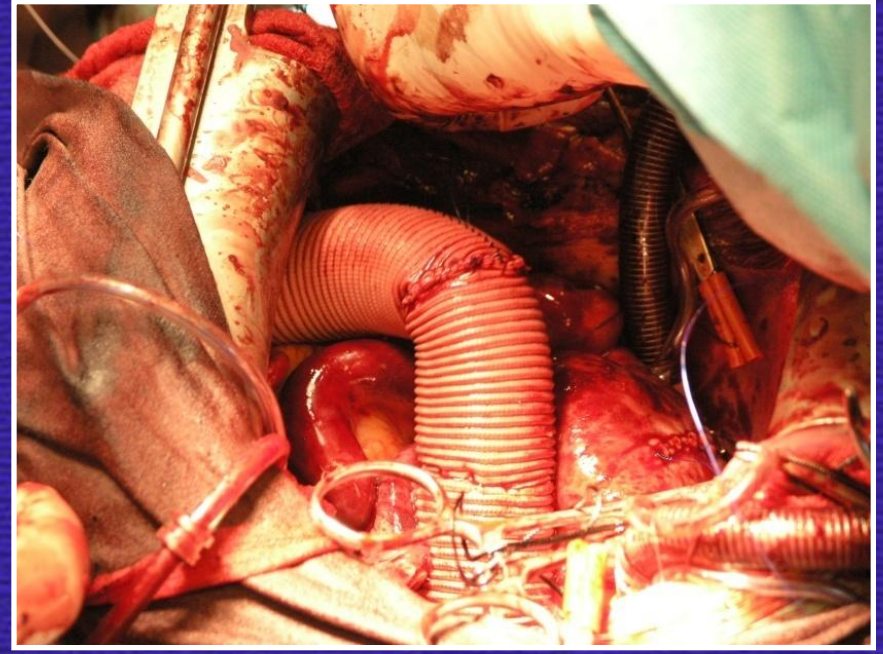
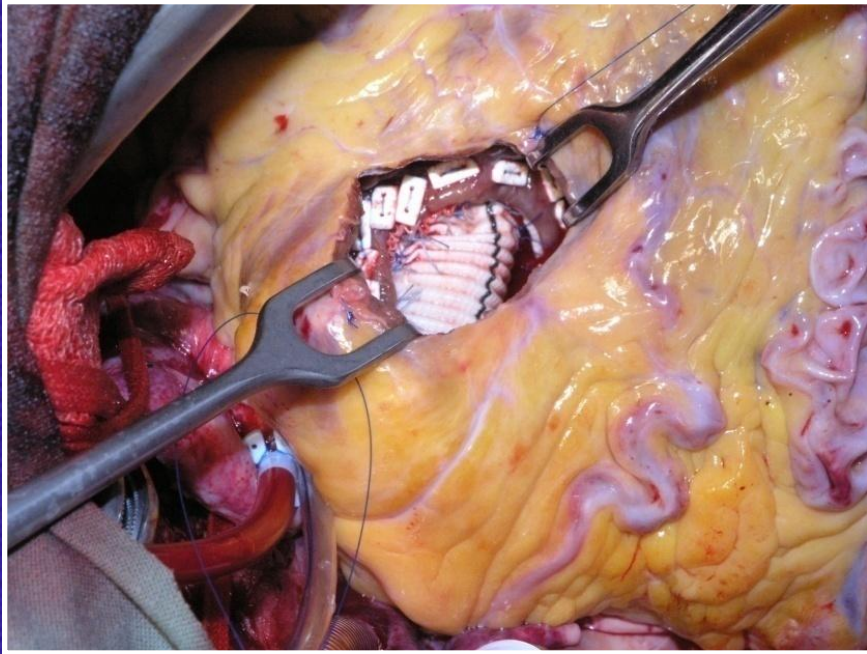
Extraanatomický bypass



Primární VSV v dospělosti

Nekorigovaná TGA + regurgitace AV chlopni

1. fáze - Rastelliho operace



Primární VSV v dospělosti

Nekorigovaná TGA + regurgitace AV chlopní

2. fáze – MVP+TVP+MAZE z MT



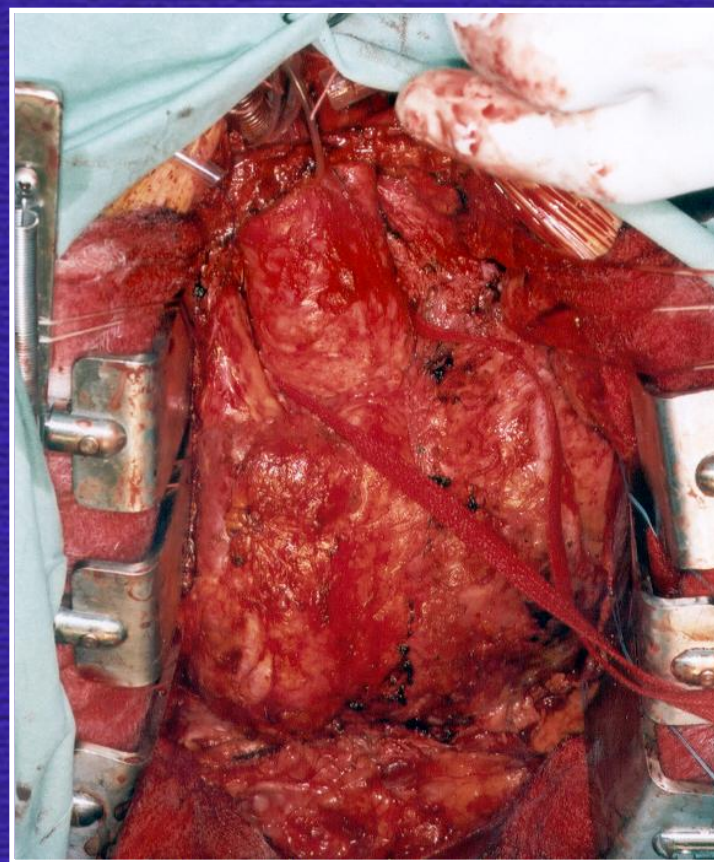
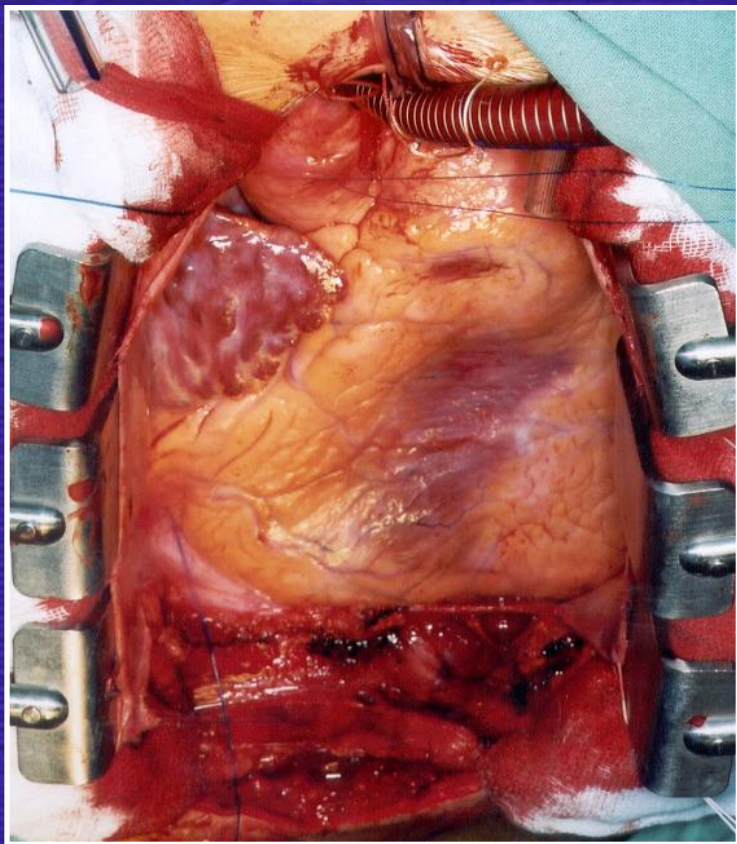
Chirurgická léčba VSV v dospělosti

Rozdělení dle chirurgické léčby

- **Reziduální léze po předchozí chirurgické korekci:**
 - Mohou se vyskytovat s jakýmkoliv odstupem od původní korekce
 - Zahrnují řadu variantních patologií
 - Předvídatelné a předpokládané reziduální nálezy (AS, PS, TOF, degenerace HG, reziduální MR, klasický Fontan atd.)
 - Nepředvídatelné nálezy po korekčních operacích z počátků dětské KCH (60-70. léta 20 století).
 - Dělí se na **PRAVOSTRANNÉ** a **LEVOSTRANNÉ**

Reziduální VSV v dospělosti

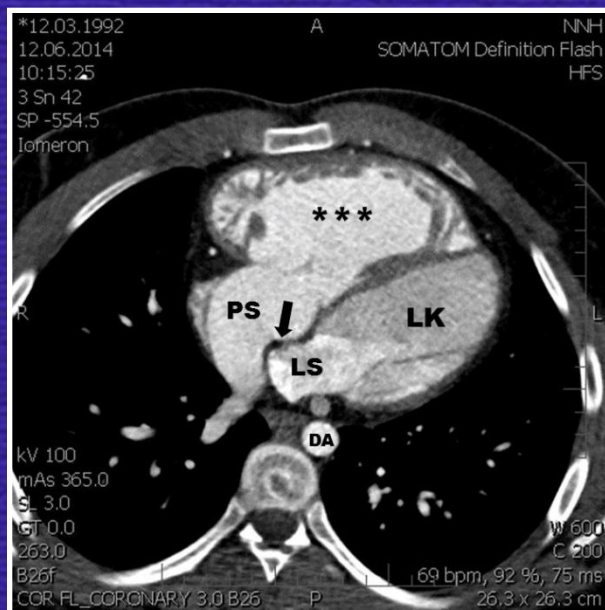
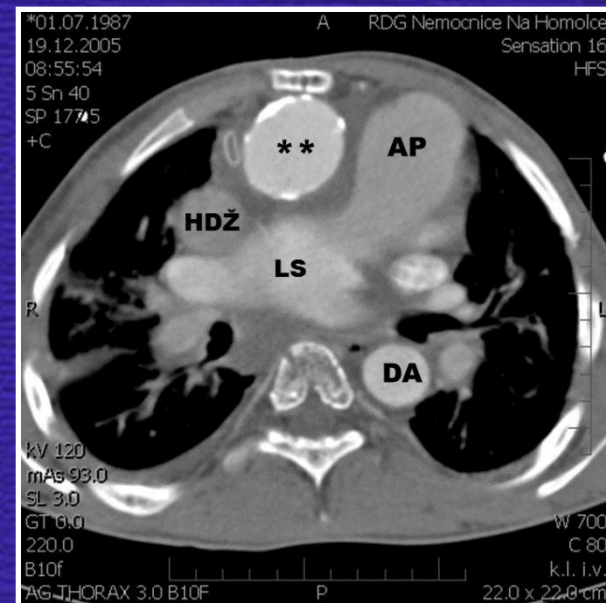
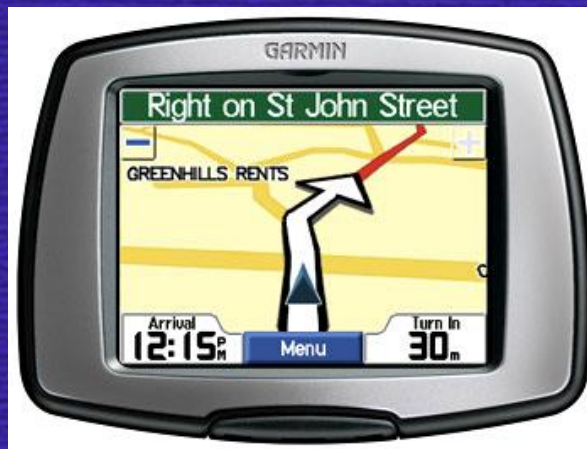
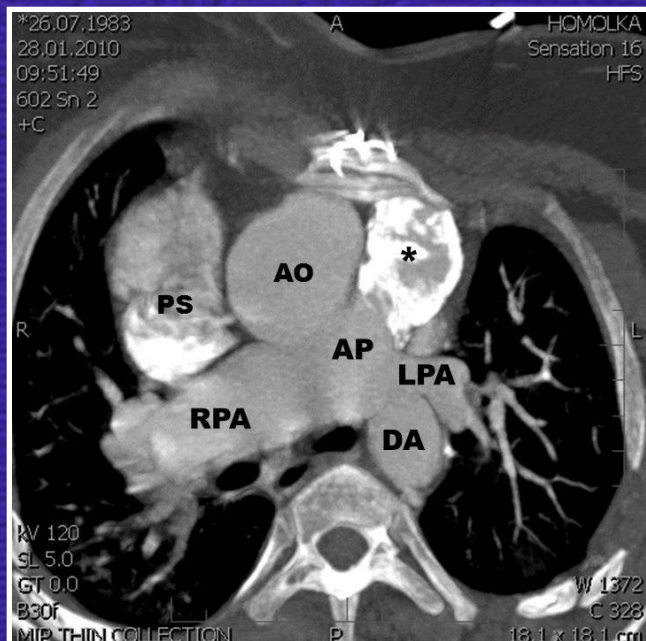
Reoperace



Bezpečně provedená resternotomie je základem úspěšné operace !!!!

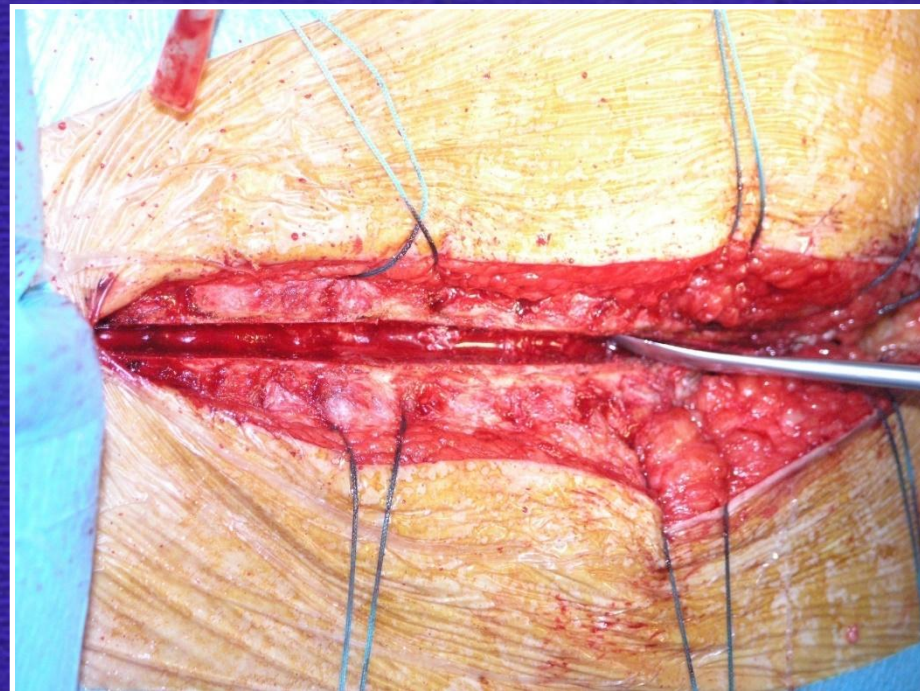
Reziduální VSV v dospělosti

Reoperace



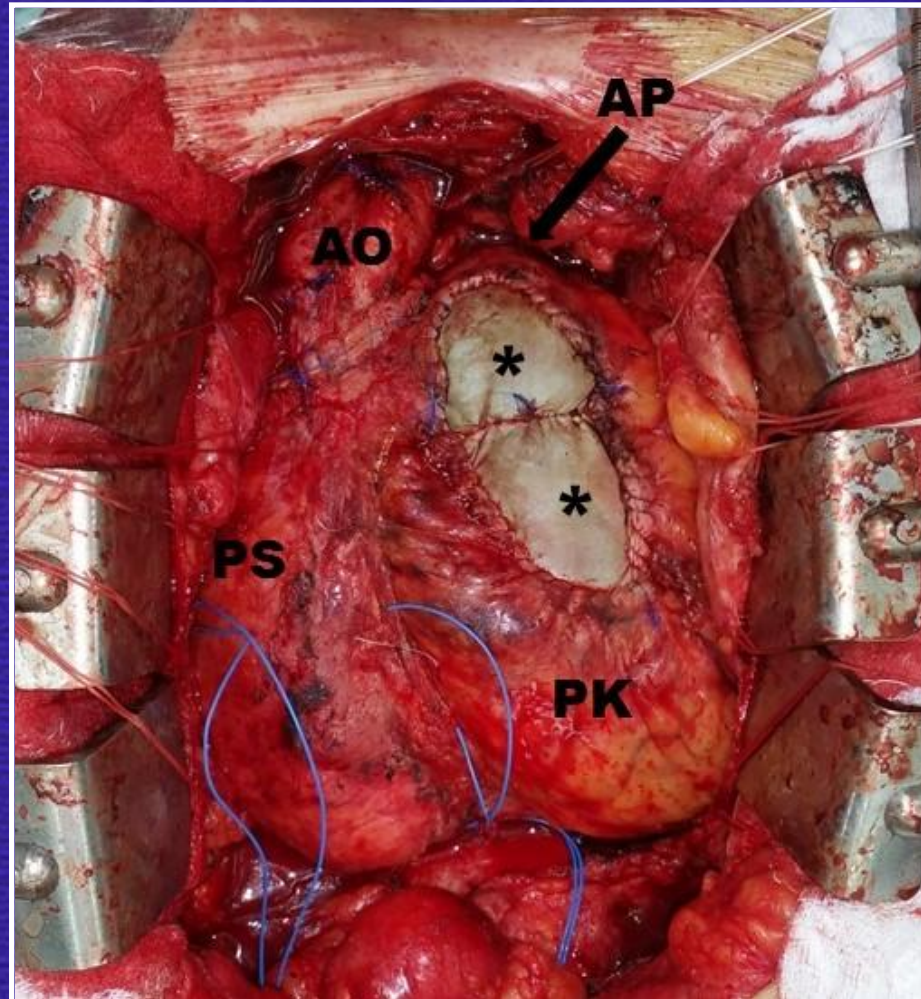
Reziduální VSV v dospělosti

Reoperace



Reziduální VSV v dospělosti

Regurgitace PV po korekci TOF



Reziduální VSV v dospělosti

Regurgitace PV po korekci TOF – PVR bio

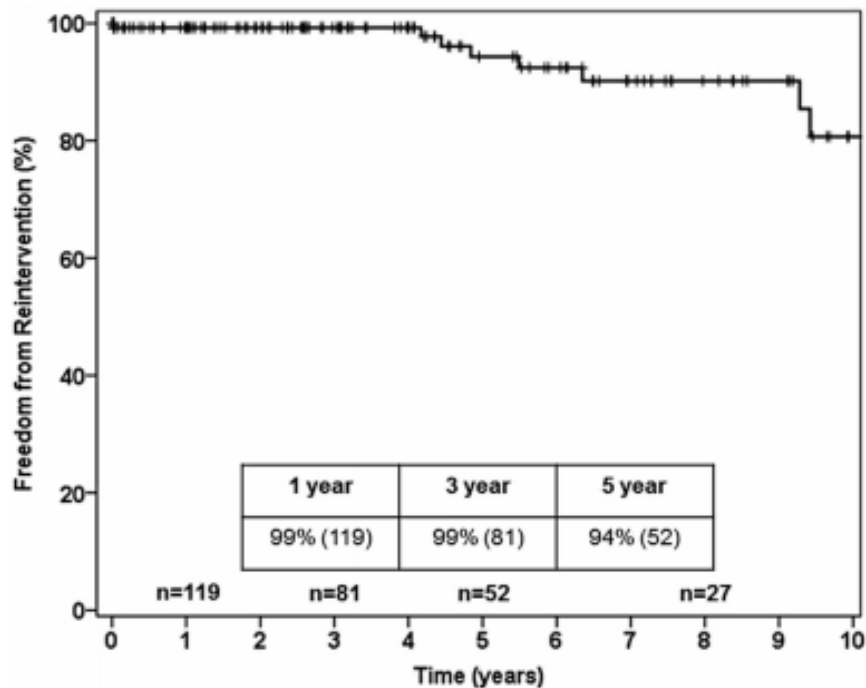


FIGURE 1. Freedom from reintervention for patients eligible for percutaneous intervention who underwent surgical pulmonary valve replacement (PVR).

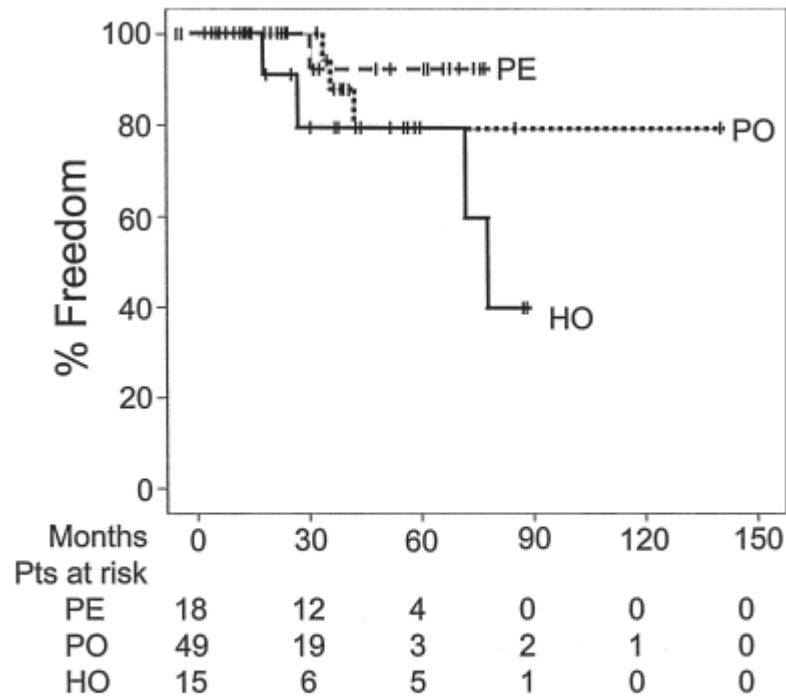


Fig 5. Kaplan-Meier actuarial freedom from valve explantation in patients (Pts) who underwent initial pulmonary valve replacement for chronic pulmonary insufficiency. (HO = homograft; PE = pericardial; PO = porcine.)

ED McKenzie, et al. *J Thorac Cardiovasc Surg*, 2014;148:1450-1453

AC Fiore, et al. *The Annals of Thoracic Surgery*, 2008;85:1712-1718

Reziduální VSV v dospělosti

Regurgitace PV po korekci TOF – PVR mech

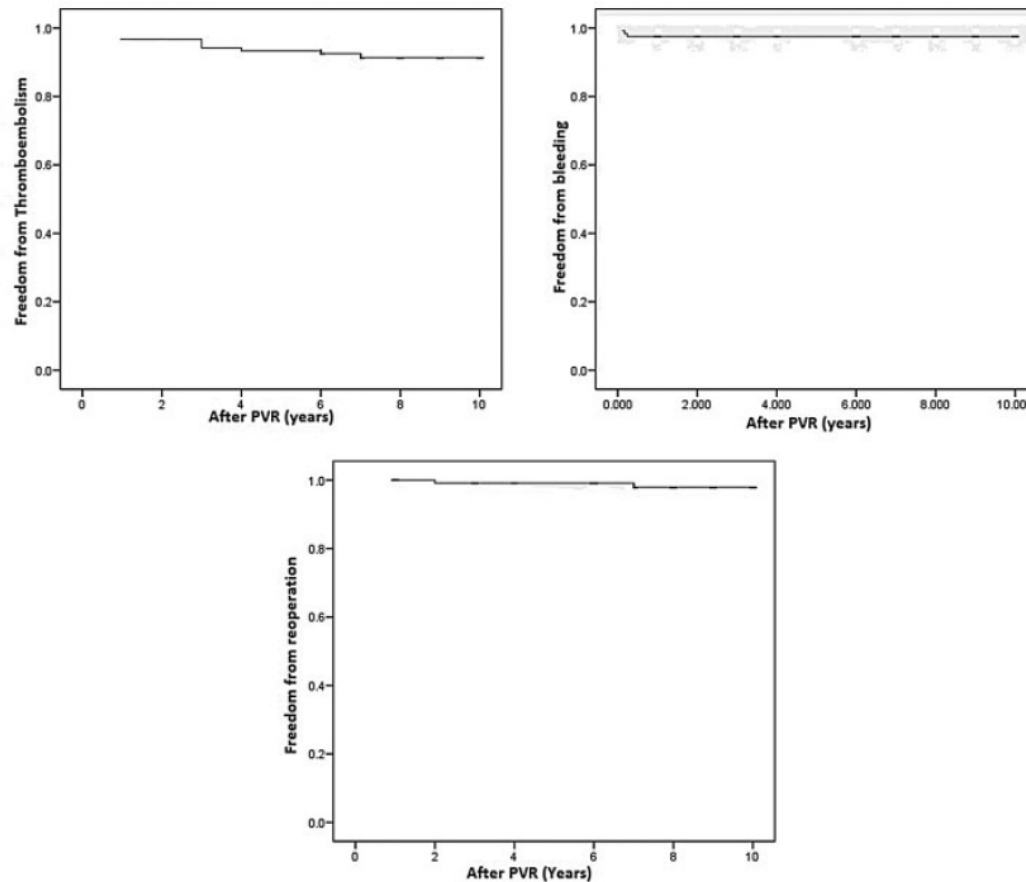
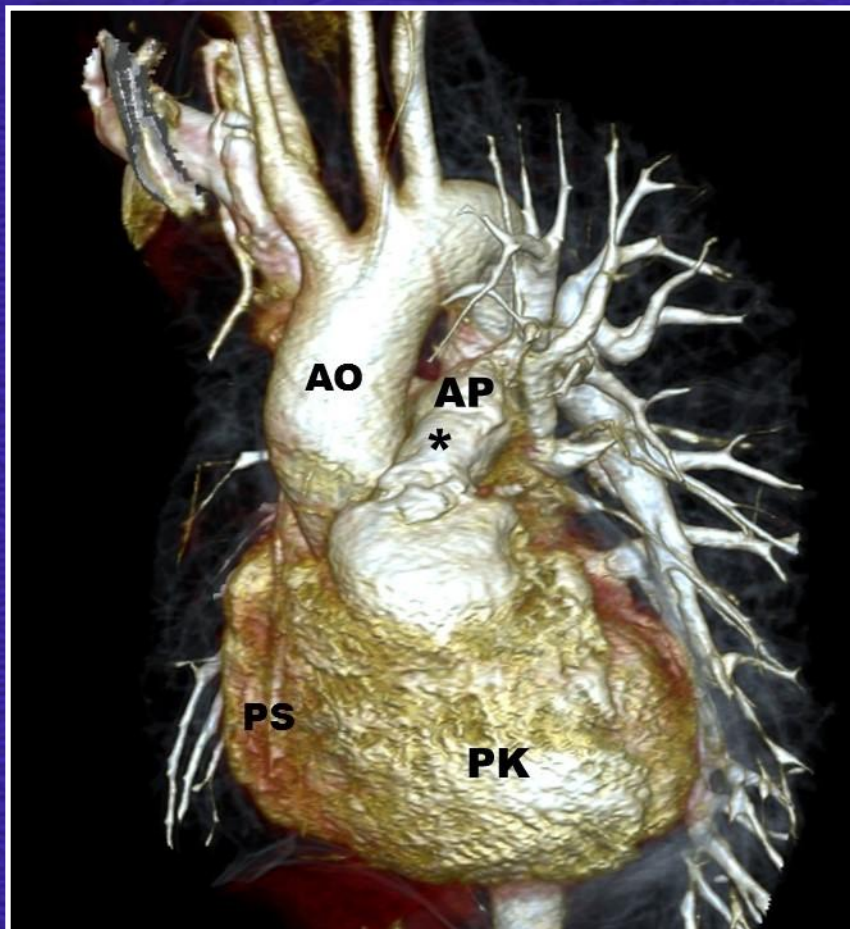


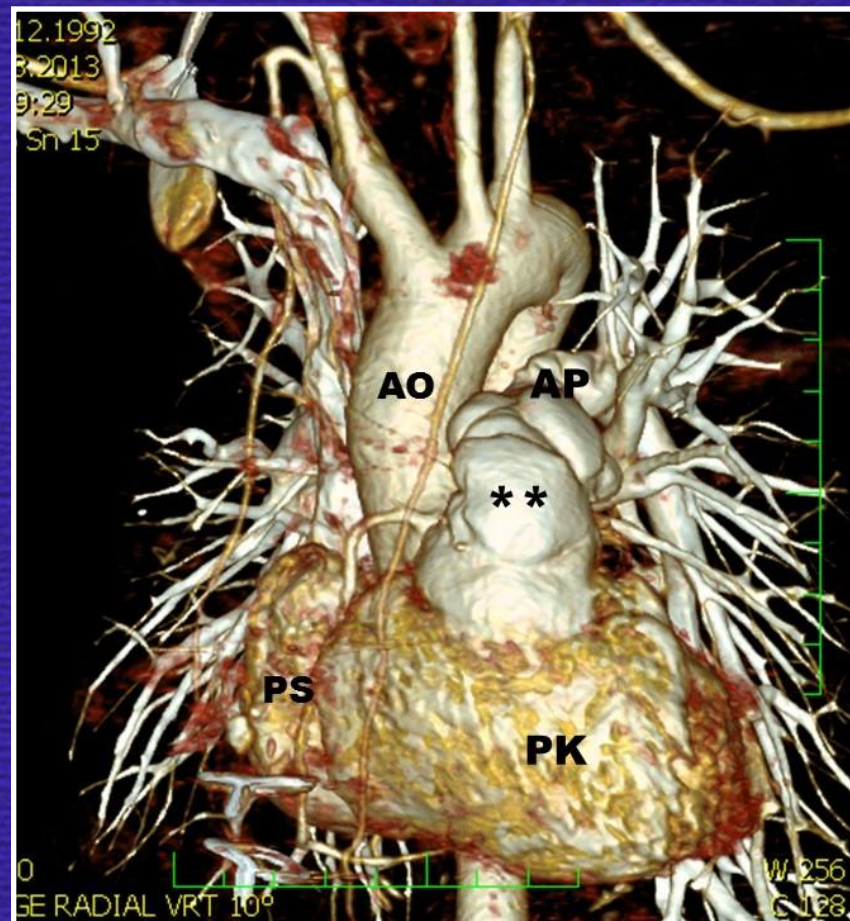
Fig. 1 Freedom from thromboembolism, bleeding, and reoperation after pulmonary valve replacement.

Reziduální VSV v dospělosti

Obstrukce RVOT



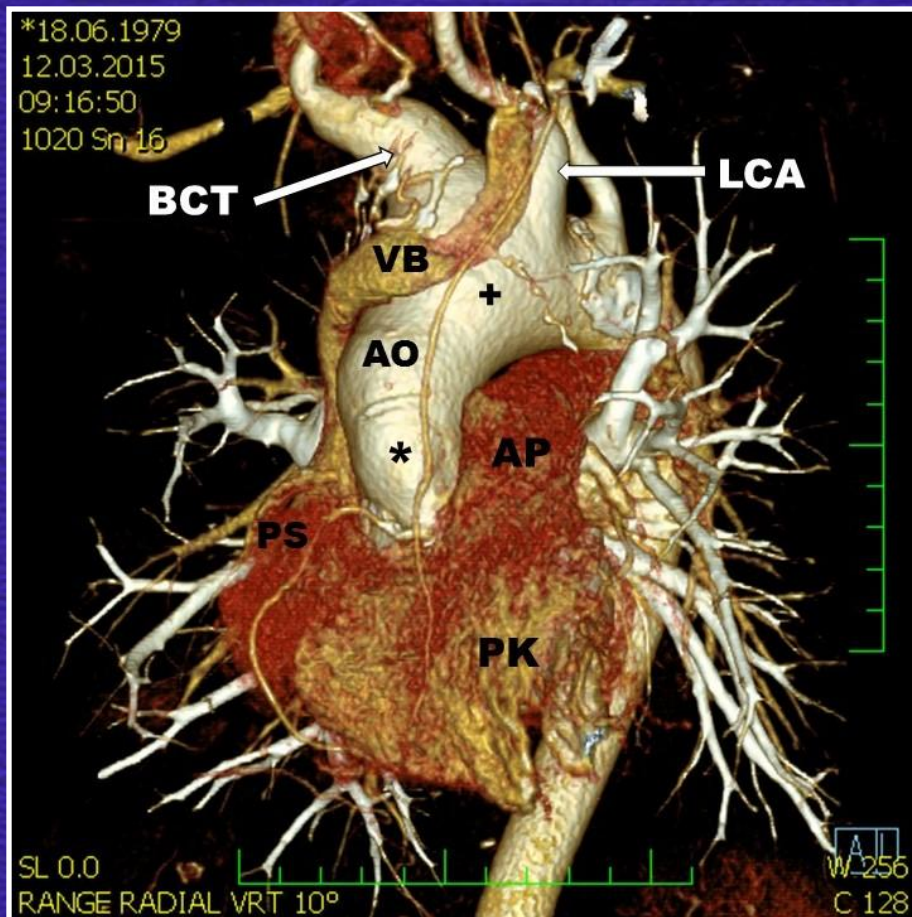
Předoperační



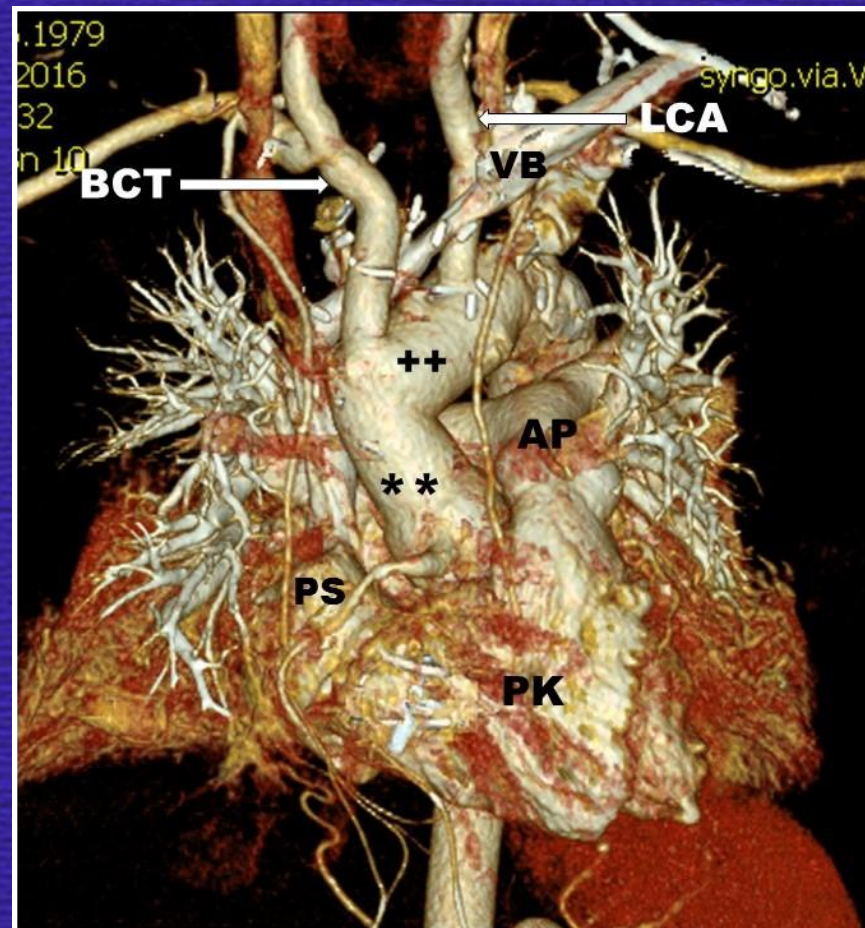
Pooperační

Reziduální VSV v dospělosti

Patologie AV a LVOT



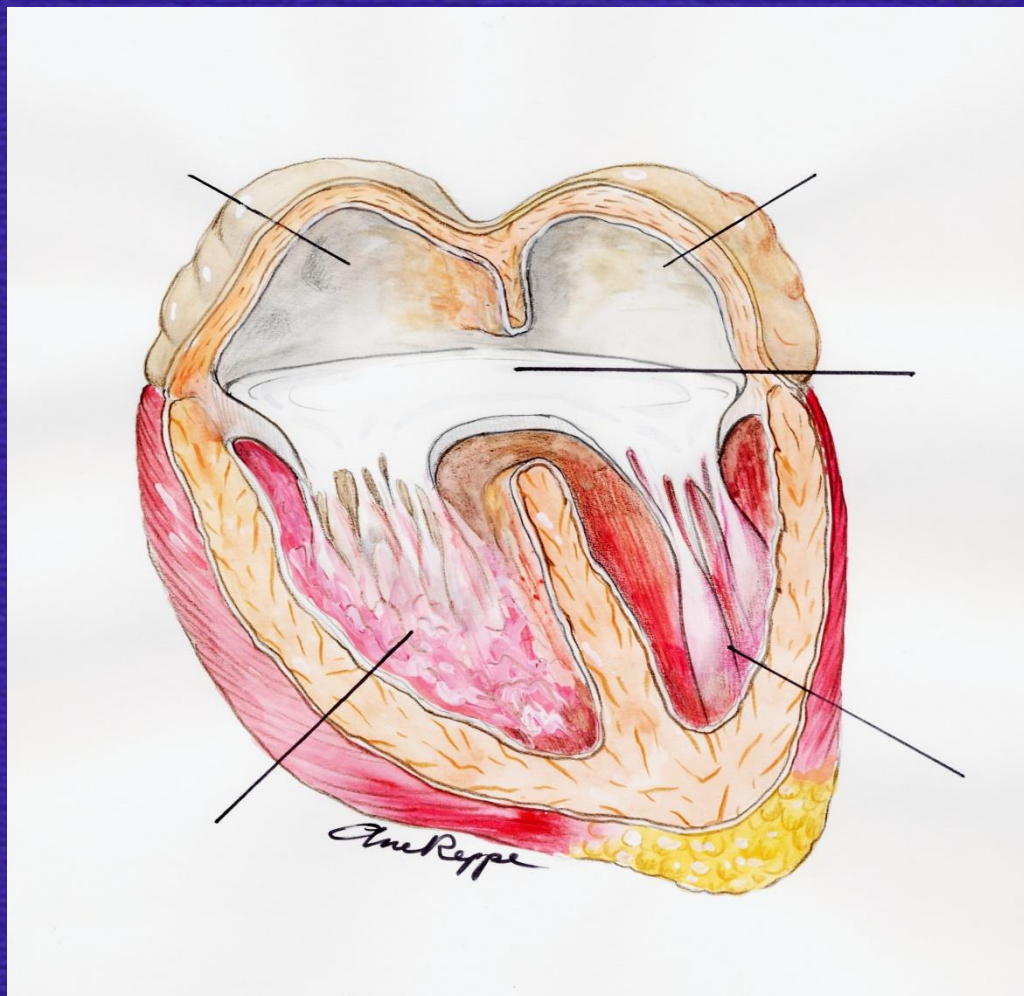
Předoperační



Pooperační

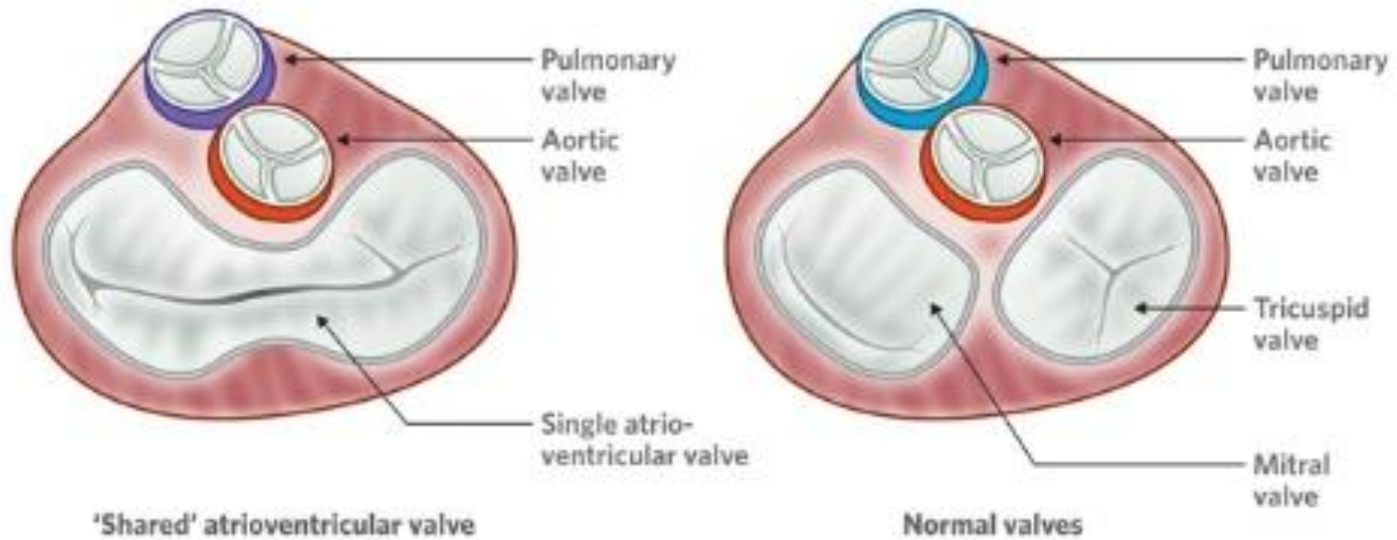
Reziduální VSV v dospělosti

AVSD a regurgitace MV



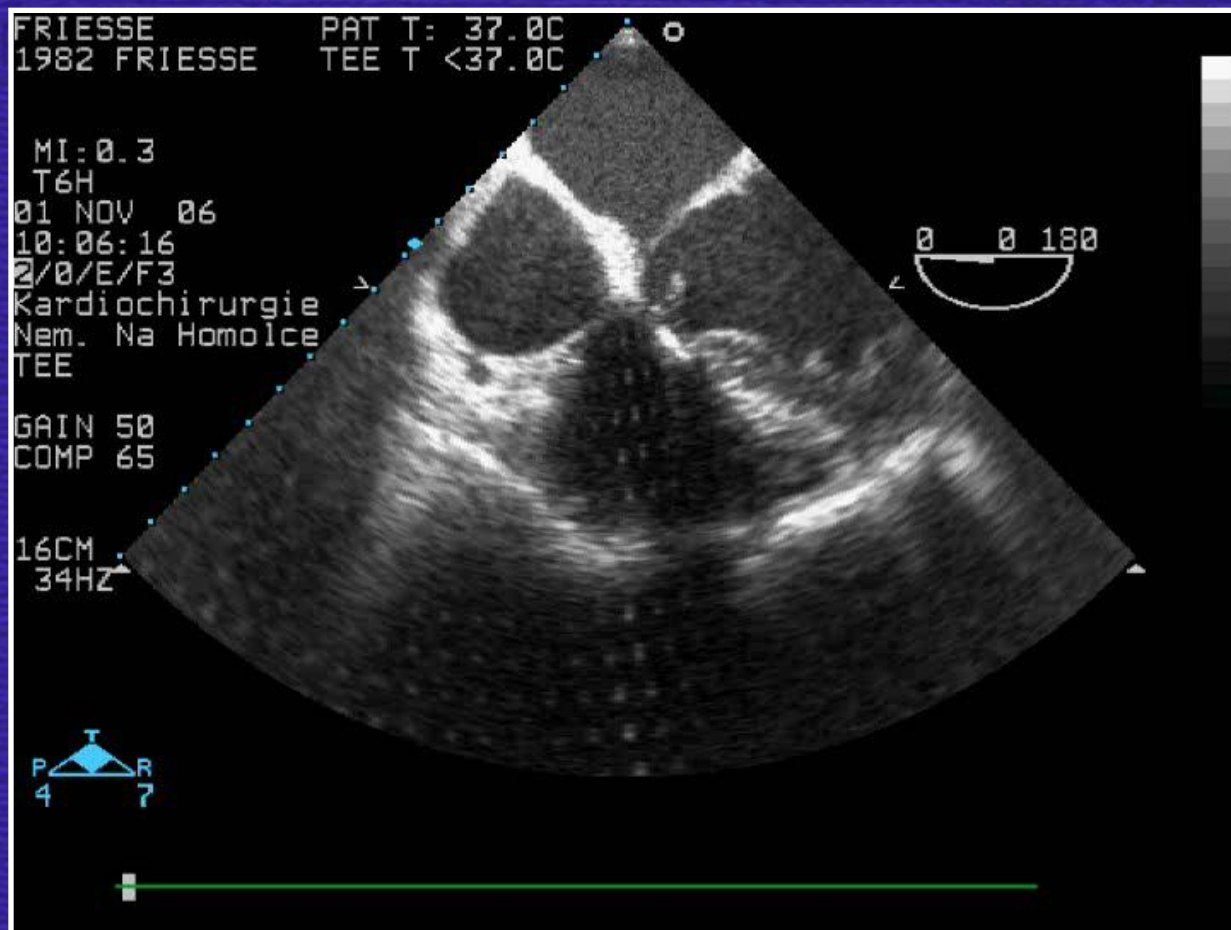
Reziduální VSV v dospělosti

AVSD a regurgitace MV



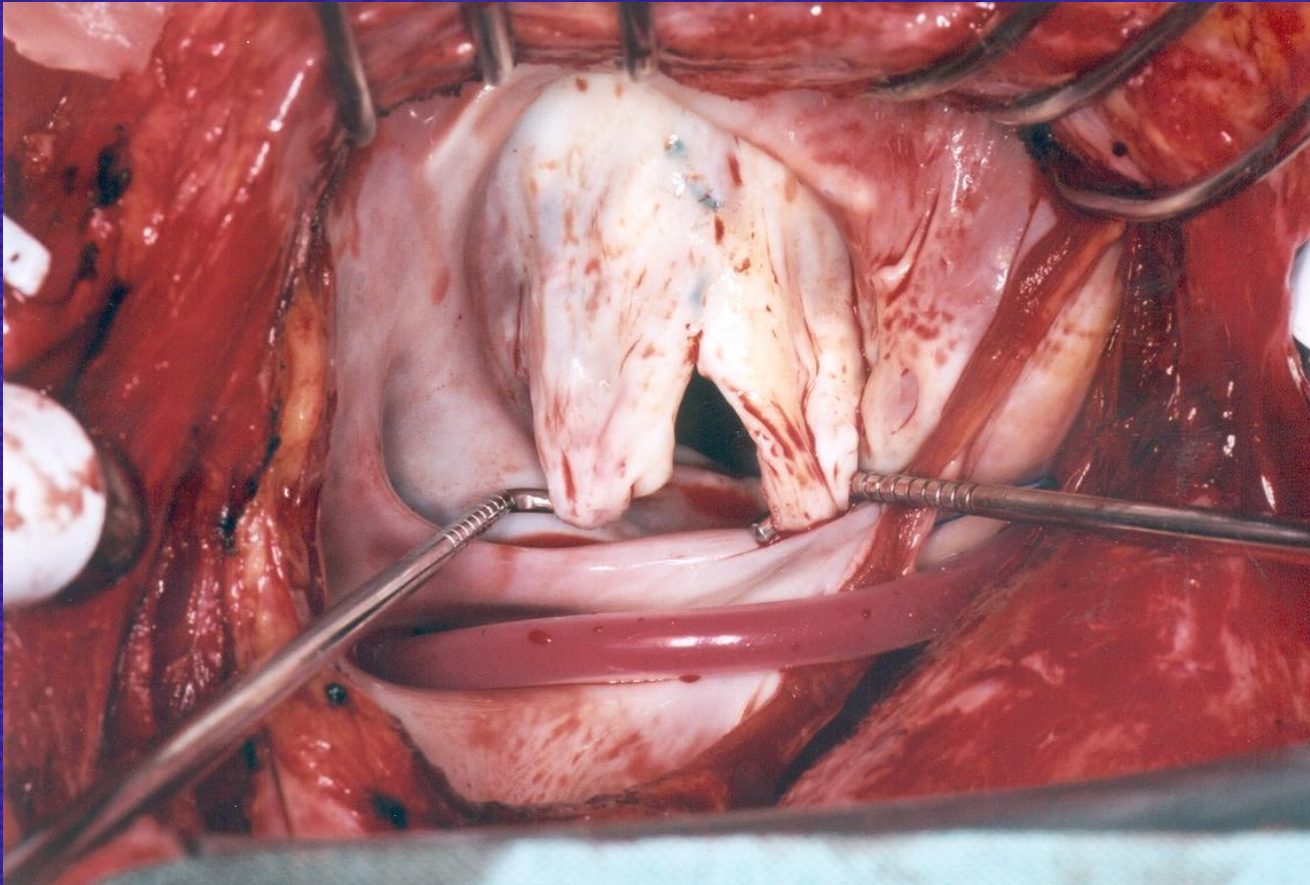
Reziduální VSV v dospělosti

AVSD a regurgitace MV



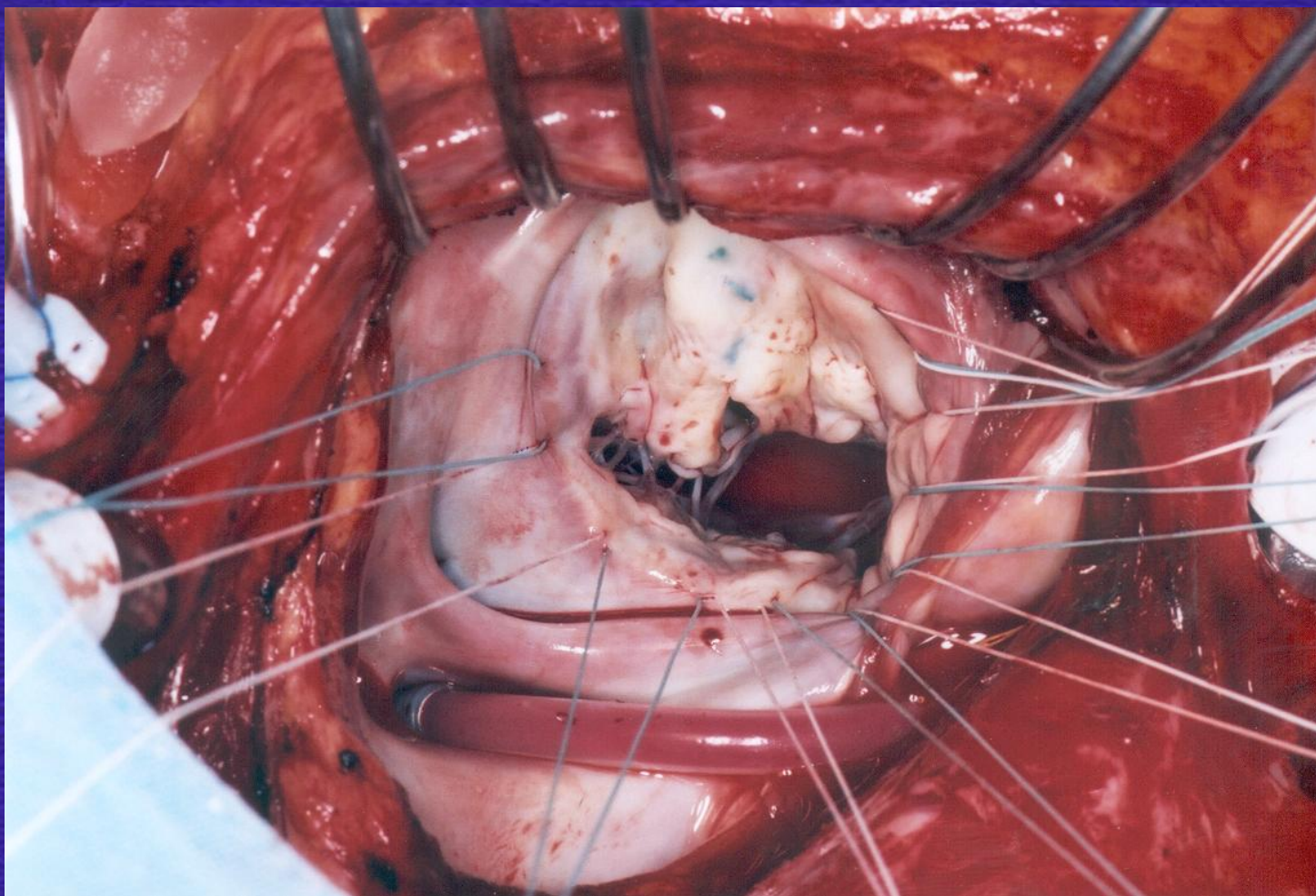
Reziduální VSV v dospělosti

AVSD a regurgitace MV



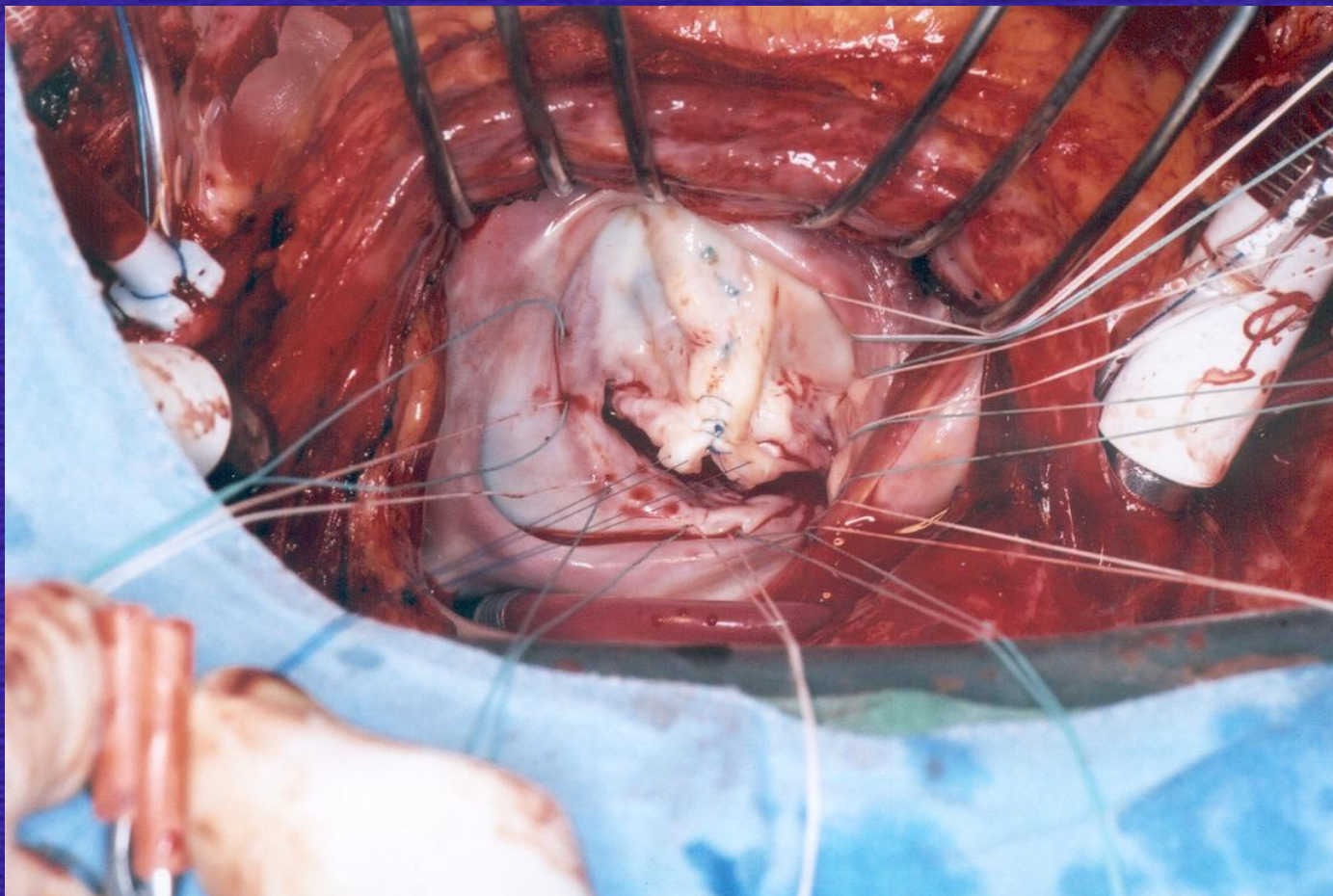
Reziduální VSV v dospělosti

AVSD a regurgitace MV



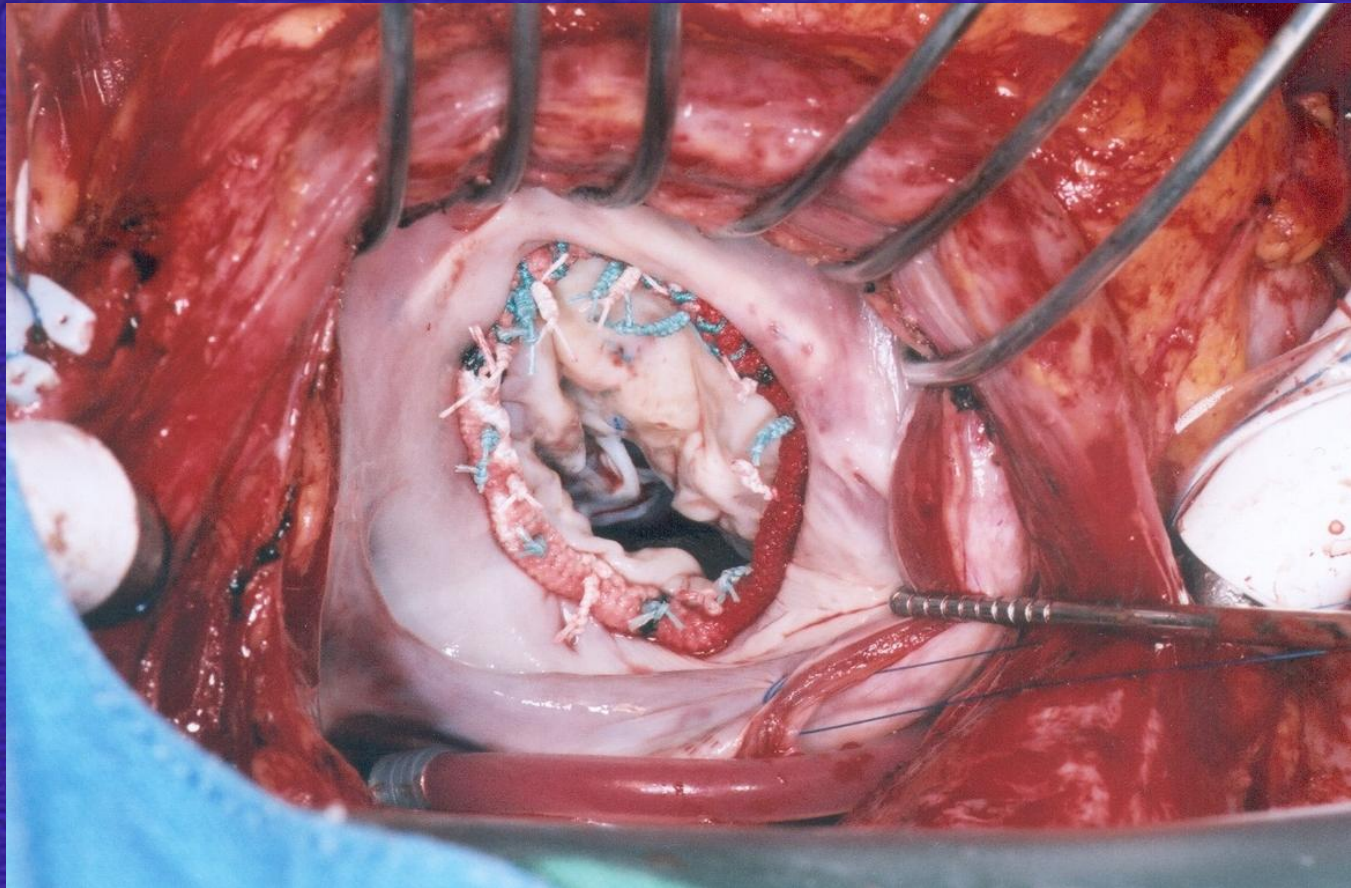
Reziduální VSV v dospělosti

AVSD a regurgitace MV



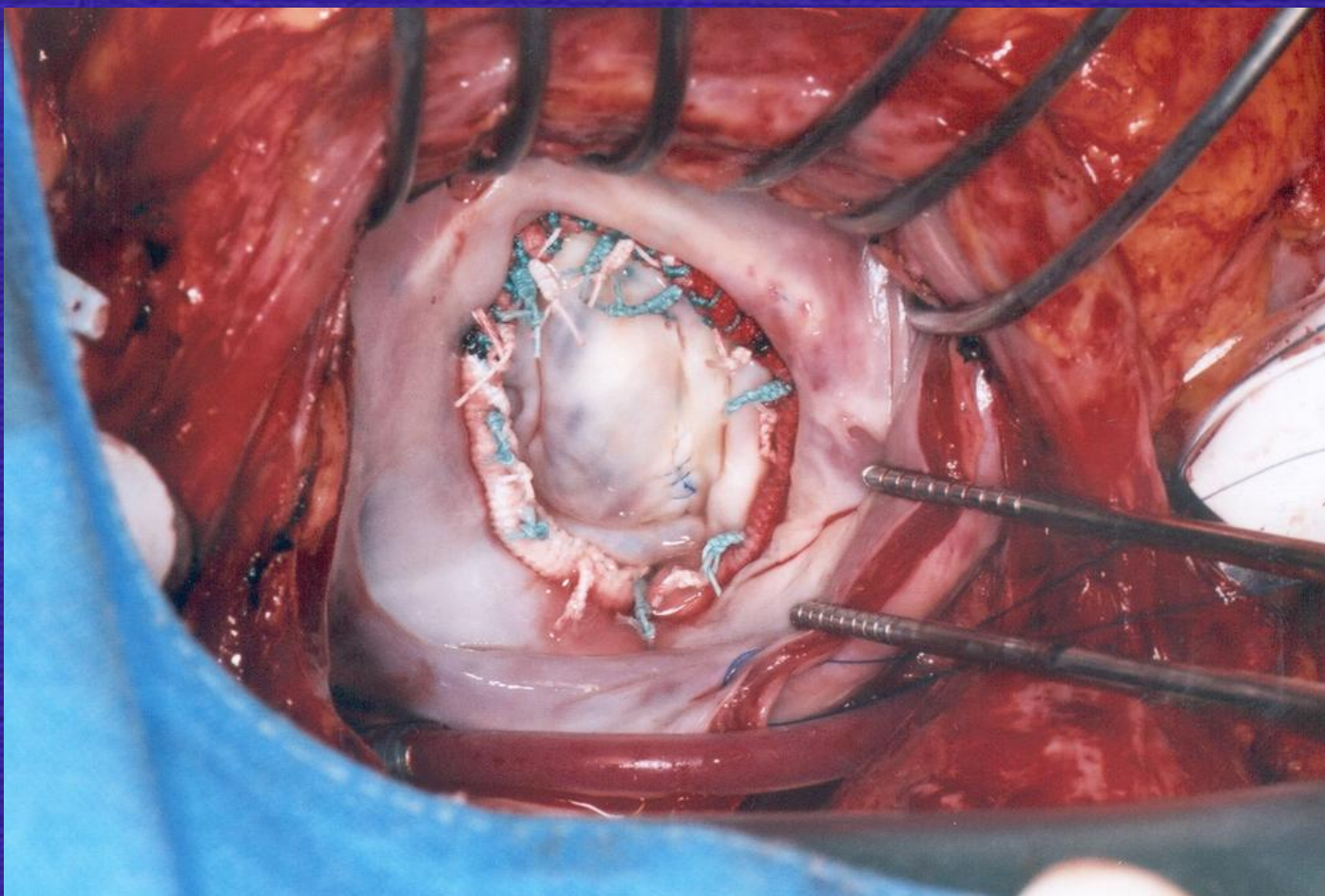
Reziduální VSV v dospělosti

AVSD a regurgitace MV



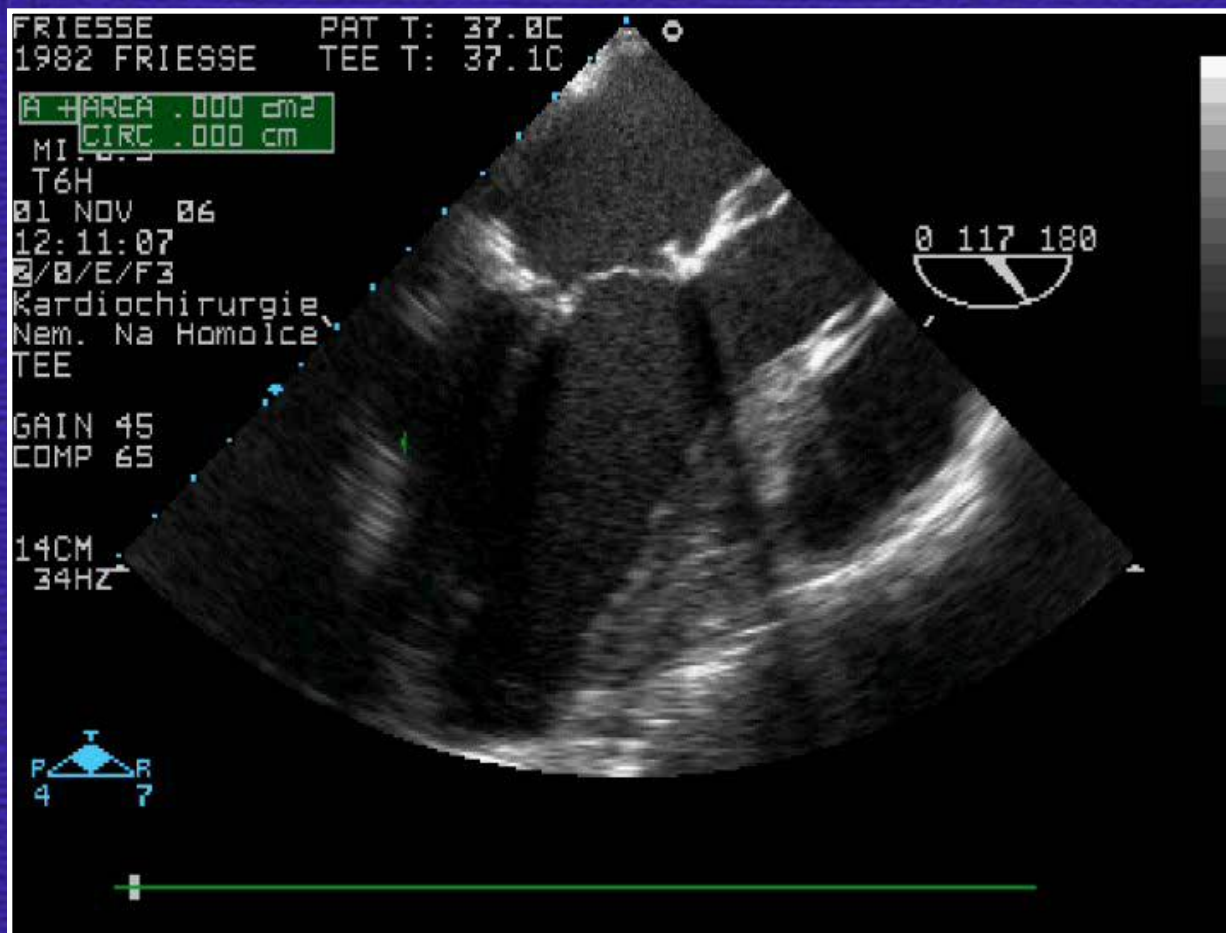
Reziduální VSV v dospělosti

AVSD a regurgitace MV



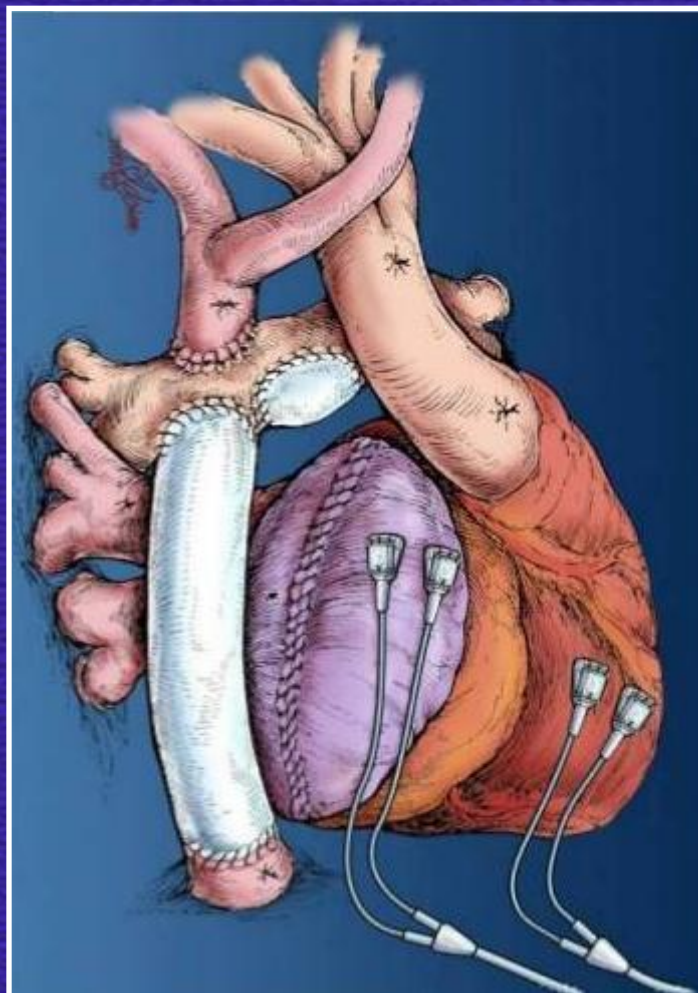
Reziduální VSV v dospělosti

AVSD a regurgitace MV



Reziduální VSV v dospělosti

Fontánovská cirkulace



Chirurgická léčba VSV v dospělosti

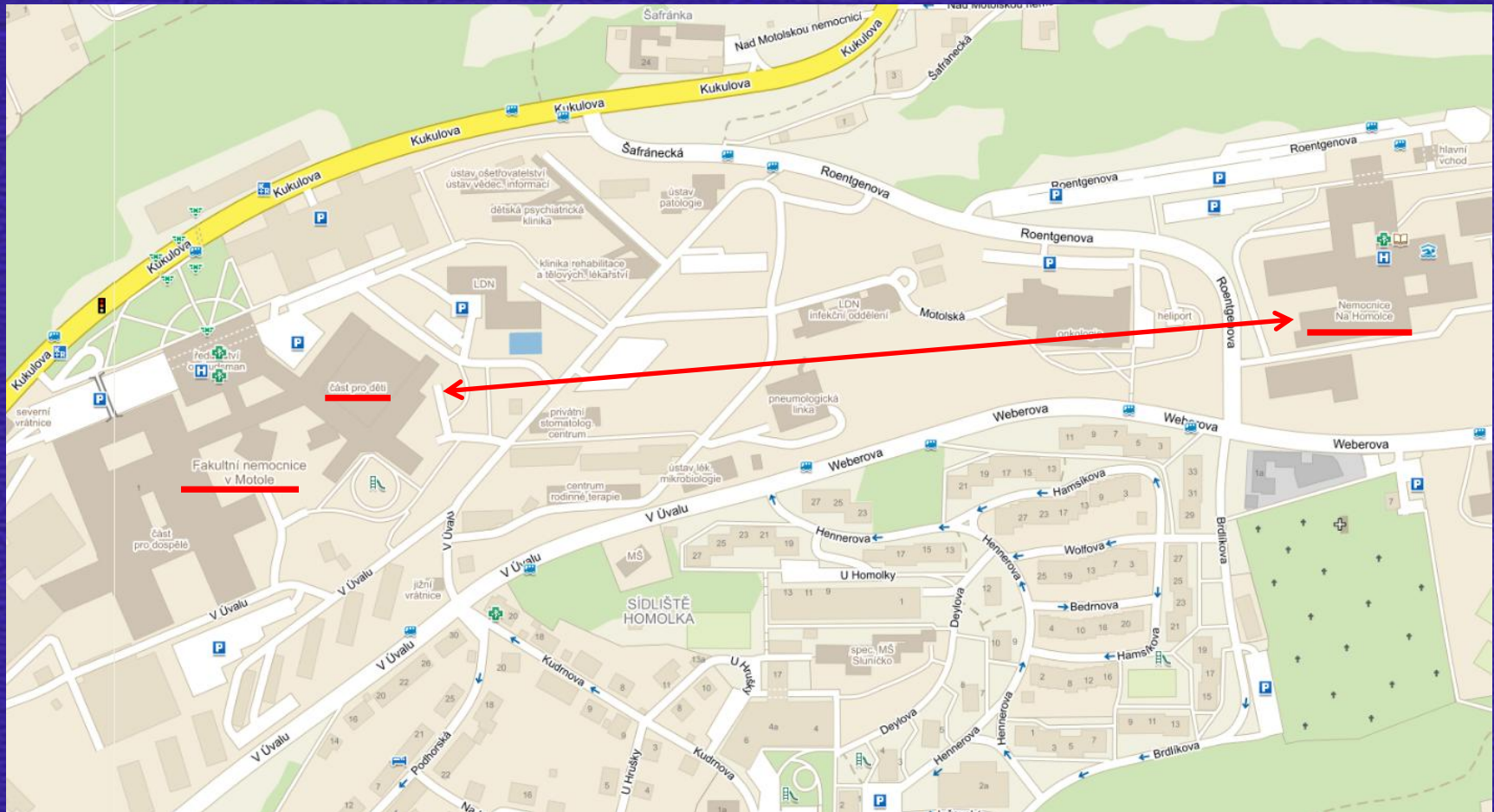
Specifika pooperační péče

- Riziko peri a pooperačního krvácení v terénu reoperací
- Selhání pravé komory srdeční (TOF, PHT, Ebstein)
- Fontánovská cirkulace
- Nemocní se systémovou pravou komorou
- Volba chlopenní náhrady
- Častá extrémní anxiozita pacientů

Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)



Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)

Original research article

Operations of adults with congenital heart disease – Single center experience with 10 years results

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ABSTRACT

During the last 10 years, 844 operations of 805 adults with congenital heart disease (CHD) were performed in Hospital Na Homolce in Prague, Czech Republic. The median age was 37 (interquartile range 25–49, full range 16–81) years. Operations of complex and rare CHD represented 47%. Forty-four percent of patients (354) underwent previous cardiac surgery in childhood or adulthood. Three and more operations were performed in 14% (113 patients). Combined surgical procedures were performed in 70% of operations.

Thirty-day mortality was 1.36%, hospital mortality 1.7% and 5-year survival probability 97%. The risk factors for early and late mortality were NYHA class III and IV symptoms ($p < 0.0001$; OR 30.8), history of heart failure ($p = 0.001$; OR 6.7), cyanosis ($p < 0.0001$; OR 60.5), number of previous operations ($p = 0.00033$), presence of mechanical prosthetic valves ($p = 0.0032$; OR 3.7) and univentricular circulation ($p = 0.0276$; OR 5.4). The difference was not significant for arrhythmias ($p = 0.078$), pulmonary hypertension ($p = 0.072$), age at operation ($p = 0.372$) and gender ($p = 0.48$).

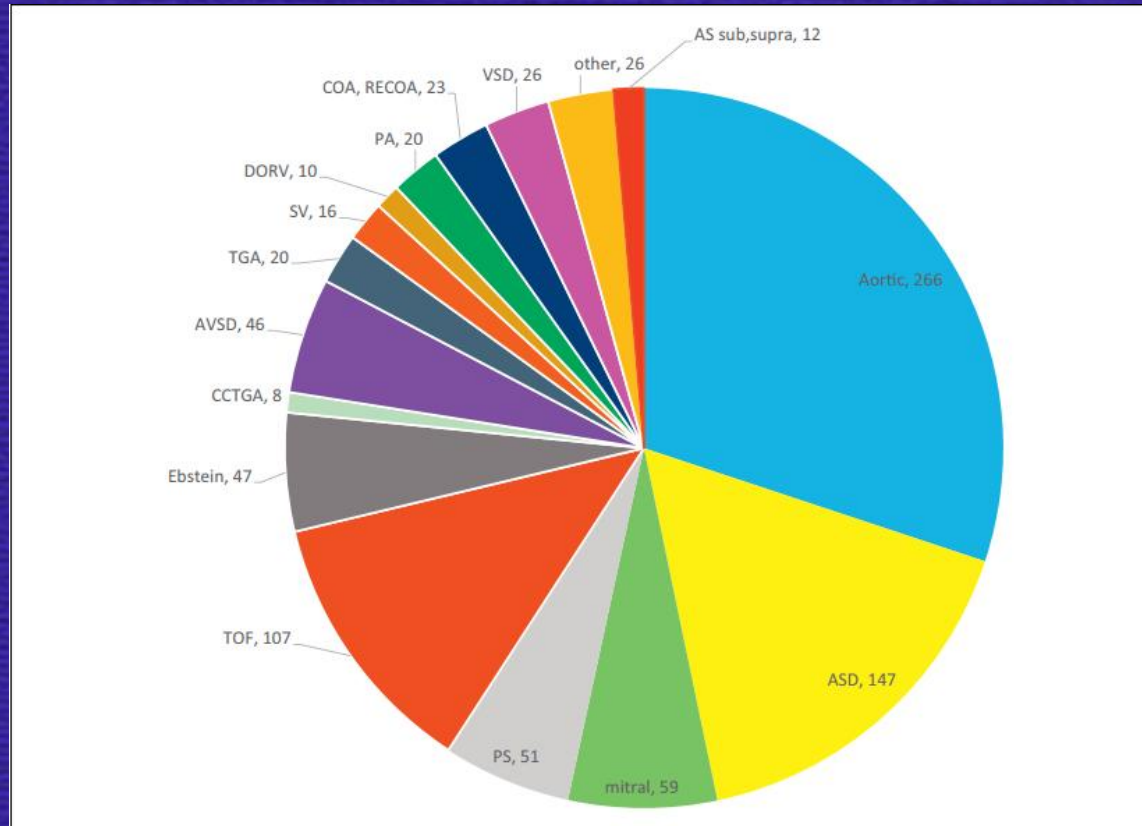
Centralization of adult CHD care in a high volume center carries very good surgical results with low early and late mortality. It is important to perform the operations in time and to eliminate all residual lesions by combined surgical procedure. The presence of pediatric cardiac surgeon is necessary for the operations of complex CHD.

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Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)



Graph 1 - Overview of adults with congenital heart disease operated in the years 2005–2015 according diagnoses.

Abbreviations - ASD: atrial septal defect, AS: aortic stenosis, AVSD: atrio-ventricular septal defect, COA: coarctation of the aorta, CCTGA: congenitally corrected transposition of the great arteries, DORV: double outlet right ventricle, PA: pulmonary atresia, PS: pulmonary stenosis, RECOA: recoarctation of the aorta, SV: functionally single ventricle, TGA: transposition of the great arteries, TOF: tetralogy of Fallot.

Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)

Jednoduché vady

Table 1a – Summary of the operations of common (simple) ACHD.

Diagnosis	Age (years) median (IQR)	N of operations (% of all ACHD operations)	N (%) of reoperations after operation in childhood	Hospital mortality (N)	Hospital mortality for the diagnosis (%)
Congenital aortic valve and aorta disease	35 (26–64)	266 (30%)	81 (31%)	0	0%
Atrial septal defect type II	50 (37–63)	147 (16%)	13 (9%)	2	1.36%
Congenital mitral valve disease	38 (27–49)	59 (7%)	16 (27%)	0	0%
Altogether		472 (53%)	110 (23%)	2	0.4%

N: number; ACHD: adult congenital heart diseases; IQR: interquartile range.

Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)

Komplexní vady

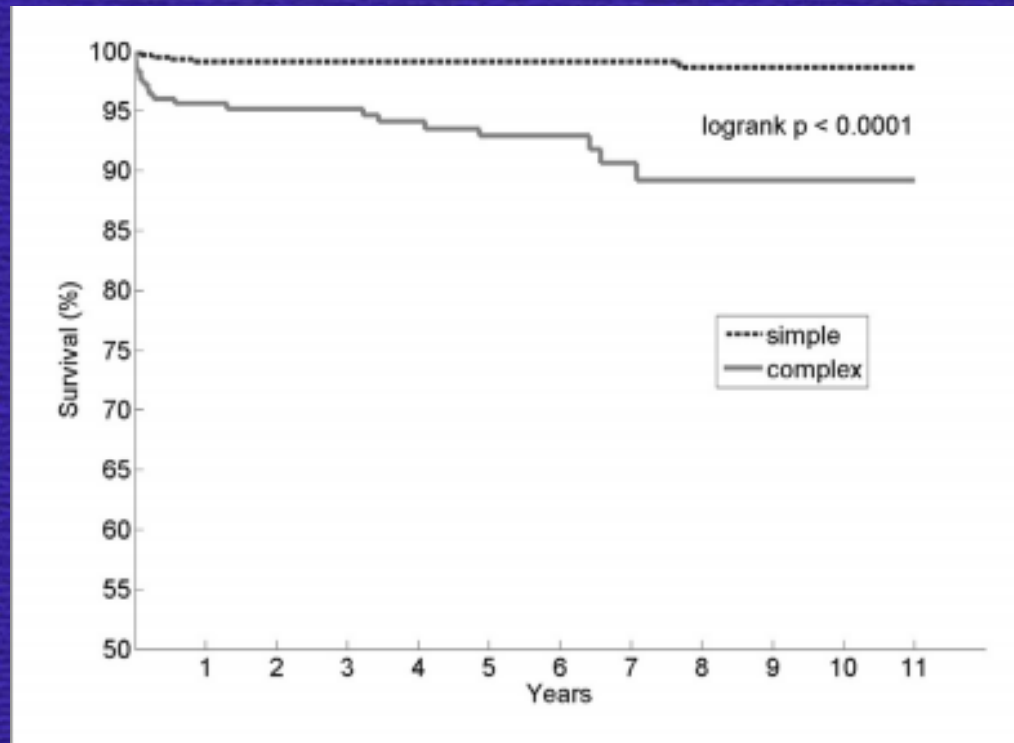
Table 1b – Summary of the operations of rare and complex ACHD.

Diagnosis	Age (years) median (IQR)	N of operations (% of all ACHD operations)	N (%) of reoperations after operation in childhood	Hospital mortality (N)	Hospital mortality for the diagnosis (%)
Tetralogy of Fallot	30 (23–37)	107 (12%)	107 (100%)	1 ^a	0.9%
Pulmonary stenosis	42 (33–51)	51 (6%)	39 (76%)	1	1.9%
Ebstein anomaly	48 (32–64)	47 (5.5%)	13 (28%)	2	4.2%
CCTGA	41.5(32–51)	8 (1%)	2 (25%)	1	12.5%
AVSD	42 (28–56)	46 (5.2%)	27 (59%)	2	4.3%
TGA					
Mustard/Senning	25 (20–30)	20 (2.4%)	19 (95%)	1	5%
Rastelli	32 (26–38)				
Arterial switch	19.5 (NA)				
SV	32 (24–39)	16 (1.8%)	13 (81%)	1	6%
DORV	26 (18–35)	10 (1.1%)	9 (90%)	1	10%
Pulmonary atresia	23 (20–26)	20 (2.3%)	20 (100%)	1*	5%
Coarctation of the aorta	47 (34–59)	23 (2.6%)	17 (74%)	1	4%
VSD	41 (28–53)	26 (3%)	17 (65%)	0	0%
AS sub-, supra-, aorto-LV tunnel	32 (16–48)	12 (1.4%)	6 (50%)	0	0%
Truncus arteriosus	21 (20–23)	4 (0.5%)	4 (100%)	0	0%
Other (PAPVC, scimitar sy, ALCAPA, another shunt)	47 (32–62)	19 (2.2%)	5 (11%)	1	5.2%
Tricuspid valve disease	36 (30–43)	8 (1%)	6 (75%)	0	0%
Altogether		412 (47%)	302 (73%)	12	2.9%

Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)



Graph 2 - Kaplan-Meier survival analysis of simple (dashed line) and complex (solid line) congenital heart disease after operation in adulthood.

Chirurgická léčba VSV v dospělosti

Vlastní zkušenosti

(2005-2015, n=844)

Rizikové faktory

Table 4 – Comparison of the frequency of risk factors among survivors and deceased.

Risk factor	Survivors N = 780	Deceased N = 25	p	OR
NYHA III–IV before operation	150 (19%)	22 (88%)	<0.0001	30.8
Cyanosis before operation	6 (0.77%)	8 (32%)	<0.0001	60.5
History of congestive heart failure	35 (4.5%)	6 (24%)	0.001	6.7
Presence of mechanical valve prosthesis	120 (15.4%)	10 (40%)	0.0032	3.7
Univentricular circulation	19 (2.4%)	3 (12%)	0.0276	5.4
Pulmonary hypertension	106 (13.6%)	7 (28%)	0.072	2.4
Arrhythmias	238 (30%)	12 (48%)	0.078	2.1
Men	419 (54%)	9 (36%)	0.103	0.48
Age at operation	39	40	0.636	NA
Number of previous operations	538	35		
Previous operations per patient	0.63	1.4	0.00033	NA

OR: odds ratio.

Závěry

- Chirurgie VSV v dospělosti je rychle se vyvíjející subspecializace na pomezí dětské a dospělé kardiochirurgie
- Těsná spolupráce mezi kongenitálními a dospělými chirurgy je zásadní pro dosažení dobrých výsledků
- Spektrum pacientů se skládá jak z primárně neoperovaných vad tak z reziduálních nálezů
- Důležitá je včasná indikace k chirurgické léčbě
- Při koncentraci tohoto typu kardiochirurgické péče do superspecializovaných center lze dosáhnout nízké peroperační mortality a výborných dlouhodobých výsledků



Děkuji za pozornost !!!!!