Results of operations in Adults with Congenital Heart Disease – Na Homolce Hospital experience

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CARDIAC SURGERY
CARDIAC SURGERY

Why should we perform adult congenital cardiac surgery ?

- In period 1977-2010, total of 14483 procedures was done

35 Výsledky chirurgické léčby vrozených srdečních vad u dětí v Dětském kardiocentru v Praze-Motole

V průběhu 34leté činnosti Dětského kardiocentra v Praze-Motole bylo na tomto pracovišti provedeno 14 483 operací dětí s vrozenou srdeční vadou (tab. 1).

Tab. 1 Přehled operací vrozených srdečních vad v Dětském kardiocentru Fakultní nemocnice v Motole, Praha, v letech 1977–2010 (n = 14 483)

typ vady	operace			
	na zavřeném srdci	mimotělní oběh	invazivní katétr	celkem
defekt komorového septa	116	2105	6	2227
defekt síňového septa	0	1567	215	1782
otevřená tepenná dučej	952	0	606	1558
koarktace aorty	1286	28	66	1380
transpozice velkých arterií	177	1079	0	1256
fallotova tetralogie	192	1011	0	1203
funkčně jediná komora	220	673	0	893
stenóza/regurgitace aorty	0	503	350	853
defekt atrioventrikulárního septa	16	681	0	697
pulmonální atrézie	376	205	0	581
pulmonální stenóza	1	129	384	514
dvojvýtoková pravá komora	148	261	1	410
úplný anomální návrat plicních žil	0	195	0	195
vady mitrální chlopně	0	154	0	154
cévní prstenec	153	0	0	153
společný arteriální trunkus	0	148	1	149
interrupce oblouku aorty	23	65	0	88
anomálie koronárních arterií	10	70	0	80
anomálie trikuspidální chlopně	0	56	0	56
srdeční tumor	0	42	0	42
aortopulmonální defekt	2	29	0	31
cor triatriatum	0	19	0	19
aplazie chlopně plicnice	0	10	0	10
aortoventrikulární tunel	0	8	0	8
ektopie srdce	7	0	0	7
transplantace srdce	0	2	0	2
ostatní operace	135	0	0	135
celkem	**3814**	**9040**	**1629**	**14 483**

Podpořeno MZČR – RVO, Nemocnice Na Homolce – NNH-00023884, IG140201

Who and where should perform adult congenital cardiac surgery ?

NA HOMOLCE HOSPITAL

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

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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)

Kogon BE et al, Ann Thorac Surg 2009;87:833-40

Adult congenital cardiac surgery

Adult cardiac surgeon perspective – what to learn from paediatric cardiac surgeons?

- Tolerance of young hearts to ischemia and tolerance of young patients to very long pump runs (second, third…)
- Precise anatomical insight and understanding of CHD
- Hemodynamical understanding of CHD (shunts, single ventricle circulation, PH…)
- Complete repair of heart defect is often feasible despite very complex procedures
- Prevention of heart block
- Using autologus and homograft tissue for repairs

Adult congenital cardiac surgery

Paediatric surgeon perspective – what to learn from adult cardiac surgeons?

- Behaviour of old hearts (over 40) and their myocardial protection
- Alternative incisions and cannulation techniques (multiple redos, peripherial cannulation, MICS)
- Major aortic surgery
- Valve repair techniques
- Valve replacement techniques esp. biological valves
- Arrhythmia surgery (esp. right heart)
- Coronary surgery (CABG etc…)
- Adult ICU care

Adult congenital cardiac surgery

„Ideal“ adult CHD center design

NA HOMOLCE HOSPITAL

- High volume adult cardiac surgical unit
- Surgical experience in:
  - Aortic surgery, valve reconstructive surgery
  - Arrhythmia surgery
  - MICS of AV valves and atrial septum
- ICU experience in:
  - PH treatment
  - VAD therapy
- Close cooperation with paediatric cardiac surgeon
- Access to HTx and LTx .....
- The team!!

Adult congenital cardiac surgery

„Ideal“ adult CHD center design

European Heart Journal (2014) **35**, 686–690
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CURRENT OPINION

Recommendations for organization of care for adults with congenital heart disease and for training in the subspecialty of ‘Grown-up Congenital Heart Disease’ in Europe: a position paper of the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology

**Helmut Baumgartner[1]*, Werner Budts[2], Massimo Chessa[3], John
Andreas Eicken[5], Johan Holm[6], Laurence Iserin[7], Folkert Meijbo
Andras Szatmari[10], Pedro T. Trindade[11], and Fiona Walker[12], fo
on Grown-up Congenital Heart Disease of the European Society**

Table 1 Staff requirements of a specialist grown-up congenital heart centre

Adult/paediatric cardiologist with GUCH certification	At least 2
GUCH imaging specialist (echo, CMR, CT)	At least 2
Congenital invasive cardiologist	At least 2
CHD surgeon	At least 2
Anaesthesiologist with CHD experience and expertise	At least 2
Invasive electrophysiologist with GUCH experience	At least 1
Psychologist	At least 1
Social worker	At least 1
Cardiovascular pathologist	At least 1

Na Homolce Hospital experience 2005 - 2015

Total No of operations	**884**
- Common Congenital Heart Defects	**472**
- Rare /complex Congenital Heart Defects	**412**

-Redo cases	**361 (41%)**
-Recurring/redo cases	**117 (14%)**
-Combined procedures	**590 (67%)**

30-day mortality 1,36%

hospital mortality 1,70%

Podpořeno MZČR – RVO, Nemocnice Na Homolce – NNH-00023884, IG140201

Overview of operations of common congenital heart defects in adulthood by diagnoses, including hospital mortality (NNH 2005-2015)

Diagnoses *(age in years in date of operation); age of the oldest patient*	**Number of procedures**	**Share from all operations**	**Redo after surgery in childhood % (N)**	**Hospital/in house mortality (N)**	**Hospital mortality for a given diagnosis (%)**
Congenital defects of aortic valve and aorta (37±13); 69 years	**266**	**30 %**	**31 % (81)**	**0**	**0 %**
Atrial septal defect Type septum secundum (49±16); 81 years	**147**	**16 %**	**9 % (13)**	**2**	**1,36 %**
Congenital defets of mitral valve (38±14); 58 years	**59**	**7 %**	**27% (16)**	**0**	**0 %**
Total	**472**	**53 %**	**23 % (110)**	**2**	**0,4 %**

Podpořeno MZČR – RVO, Nemocnice Na Homolce – NNH-00023884, IG140201

Summary of operations of rare / complex congenital heart defects in adulthood, according to diagnoses, including hospital mortality (NNH 2005-2015) 1/3

Diagnoses *(age in years in date of operation); age of the oldest patient*	**Number of procedures**	**Share from all operations**	**Redo after surgery in childhood % (N)**	**Hospital/in house mortality (N)**	**Hospital mortality for a given diagnosis (%)**
Tetralogy of Fallot (33±11); 61 years	**107**	**12 %**	**100% (107)**	**1**	**0,9 %**
Pulmonary stenosis (43±12); 67 years	**51**	**6 %**	**76% (39)**	**1**	**1,9 %**
Ebstein anomaly (33±15); 70 years	**47**	**5,5 %**	**28% (13)**	**2**	**4,2 %**
CCTGA (43±15); 71 years	**8**	**1 %**	**25% (2)**	**1**	**12,5 %**
AVSD (40±13,5); 70 years	**46**	**5,2 %**	**59% (27)**	**2**	**4,3 %**
TGA Must/Senn (27±5,6); 33 Rastelli (34±7); 48 arter.switch (19,5±2); 21	**20**	**2,4 %**	**95% (19)**	**1**	**5 %**

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Summary of operations of rare / complex congenital heart defects in adulthood, according to diagnoses, including hospital mortality (NNH 2005-2015) 2/3

Diagnoses *(age in years in date of operation); age of the oldest patient*	**Number of procedures**	**Share from all operations**	**Redo after surgery in childhood % (N)**	**Hospital/in house mortality (N)**	**Hospital mortality for a given diagnosis (%)**
SV (32,5±11); 54 years	**16**	**1,8 %**	**81% (13)**	**1**	**6 %**
DORV (28,5±9); 41 years	**10**	**1,1 %**	**90 % (9)**	**1**	**10 %**
Pulmonary atresia (24,5±5); 37 years	**20**	**2,3 %**	**100% (20)**	**0**	**0 %**
Coarctation of the aorta (45±14); 65 years	**23**	**2,6 %**	**74% (17)**	**0**	**0 %**
VSD (42±13); 63 years	**26**	**3 %**	**65% (17)**	**0**	**0 %**
AS sub-, supra-, aorto-LK tunnel (40±19); 72 years	**12**	**1,4 %**	**50% (6)**	**0**	**0 %**
Truncus arteriosus (21±2,5); 24 years	**4**	**0,5 %**	**100% (4)**	**0**	**0 %**

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Summary of operations of rare / complex congenital heart defects in adulthood, according to diagnoses, including hospital mortality (NNH 2005-2015) 3/3

Diagnoses *(age in years in date of operation); age of the oldest patient*	**Number of procedures**	**Share from all operations**	**Redo after surgery in childhood % (N)**	**Hospital/in house mortality (N)**	**Hospital mortality for a given diagnosis (%)**
PAPVD, scimitar sy (45±16); 70 years	**6**	**0,7 %**	**17% (1)**	**1**	**16,7 %**
ALCAPA (35±19); 57 years	**3**	**0,3 %**	**67% (2)**	**0**	**0 %**
Other shunt defects (50±14); 63 years	**3**	**0,3 %**	**0% (0)**	**0**	**0 %**
Tricuspid valve defects (37±11); 55 years	**8**	**1 %**	**75% (6)**	**0**	**0 %**
Other	**2**	**0,2 %**	**0% (0)**	**0**	**0 %**
Total	**412**	**47 %**	**73% (302)**	**11**	**2,67 %**

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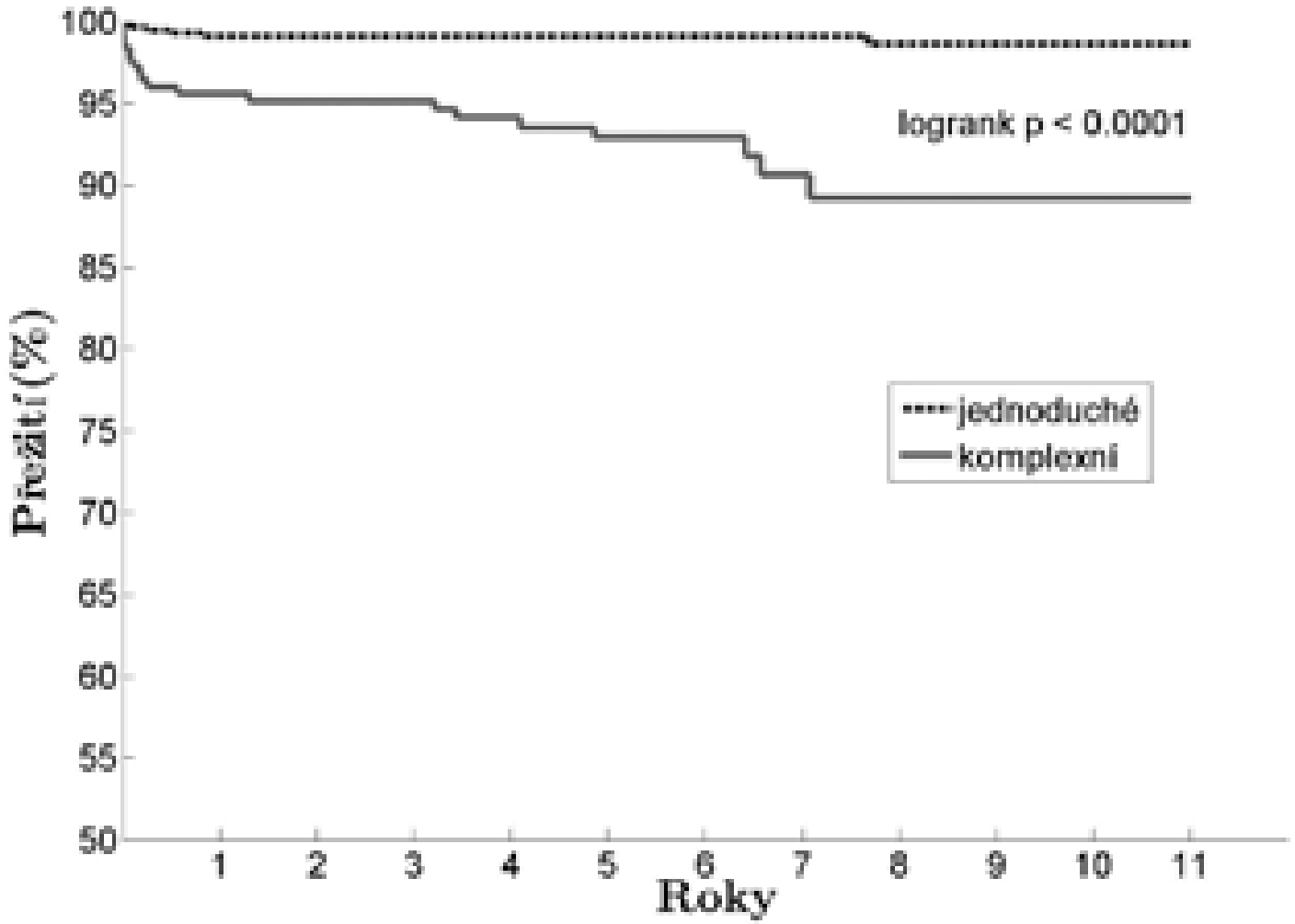
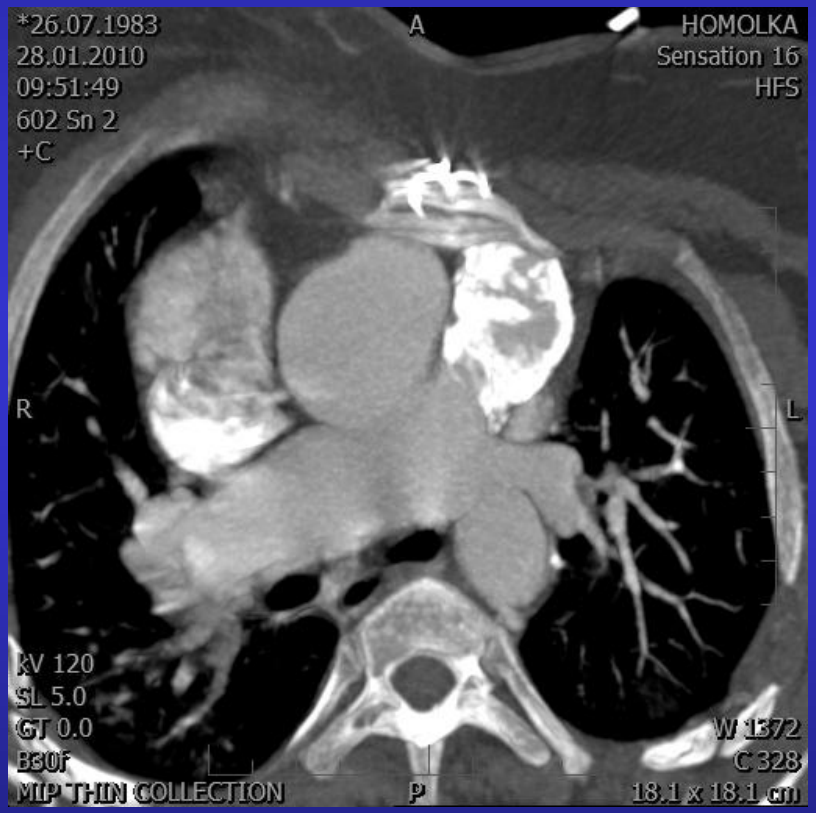


Popelova et al: Operations of adults with congenital heart disease, Cor et Vasa, 2016

Pulmonary valve replacement

surgical experience

Pulmonary valve procedures...



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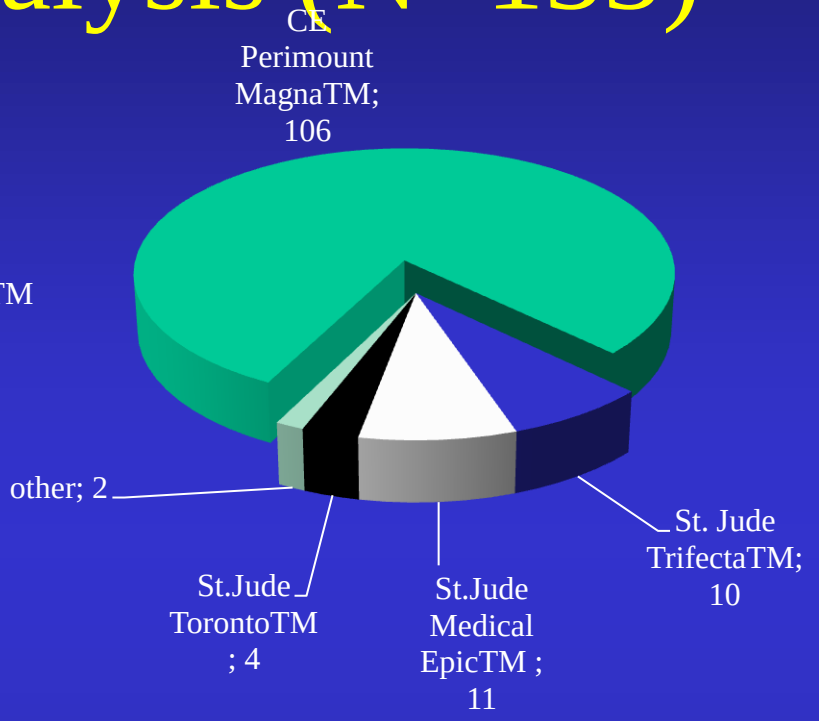
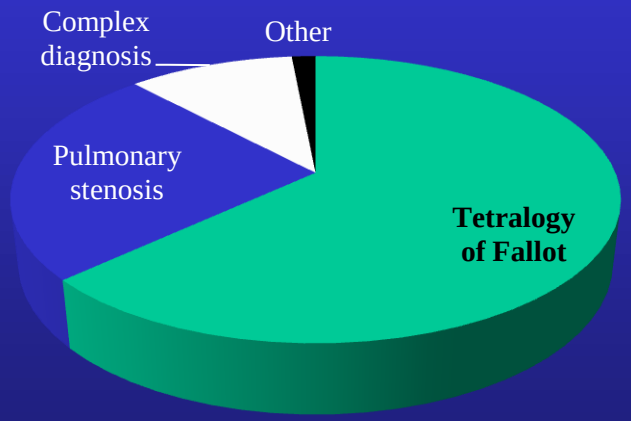
Procedure data analysis (N=133)

- Valves used
  - Carpentier-Edwards Perimount Magna™
  - St. Jude Trifecta™
  - St. Jude Medical Epic™
  - St. Jude Toronto™



- Procedure diagnosis
  - Tetralogy of Fallot (n=84)
  - Pulmonary stenosis (n=33)
  - Complex diagnosis (n=14)
  - Other (n=2)



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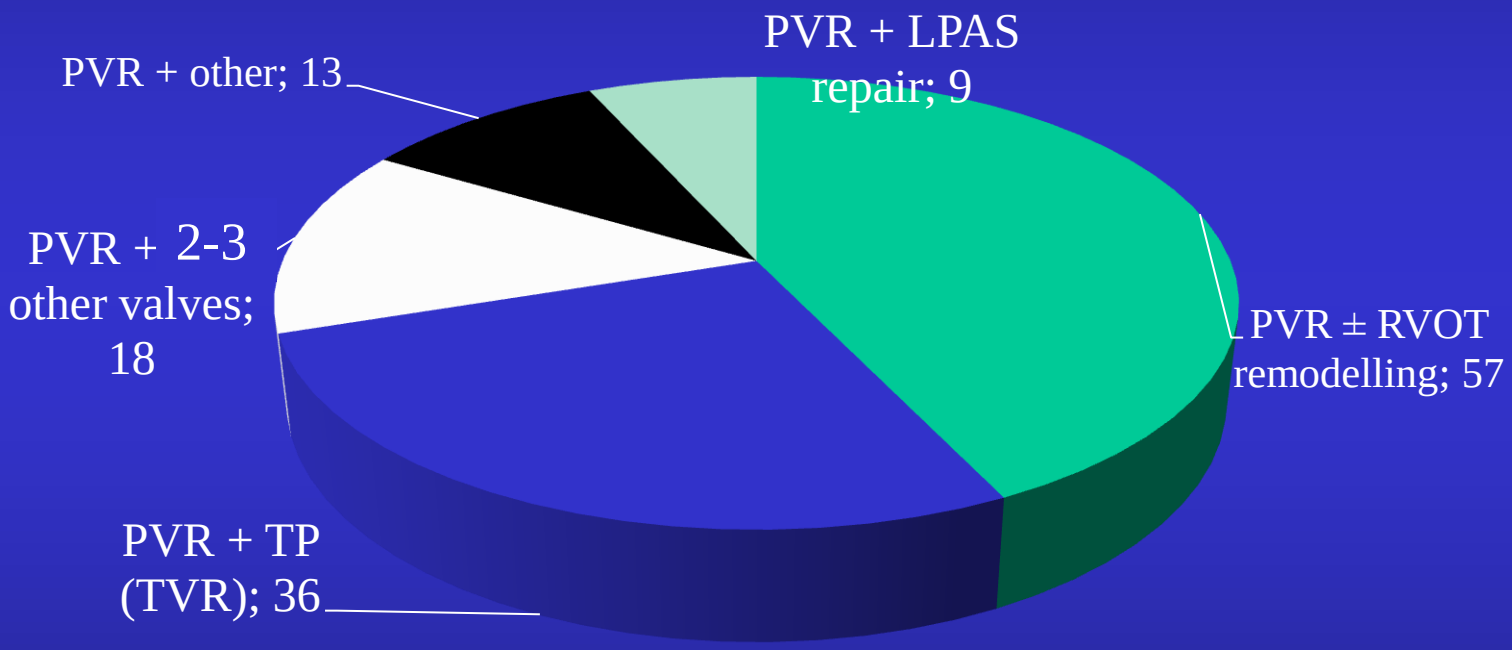
Types of surgical procedures

- 182 previous procedures in 129 patients



- Size of implanted valves 21-29 (mean 27)

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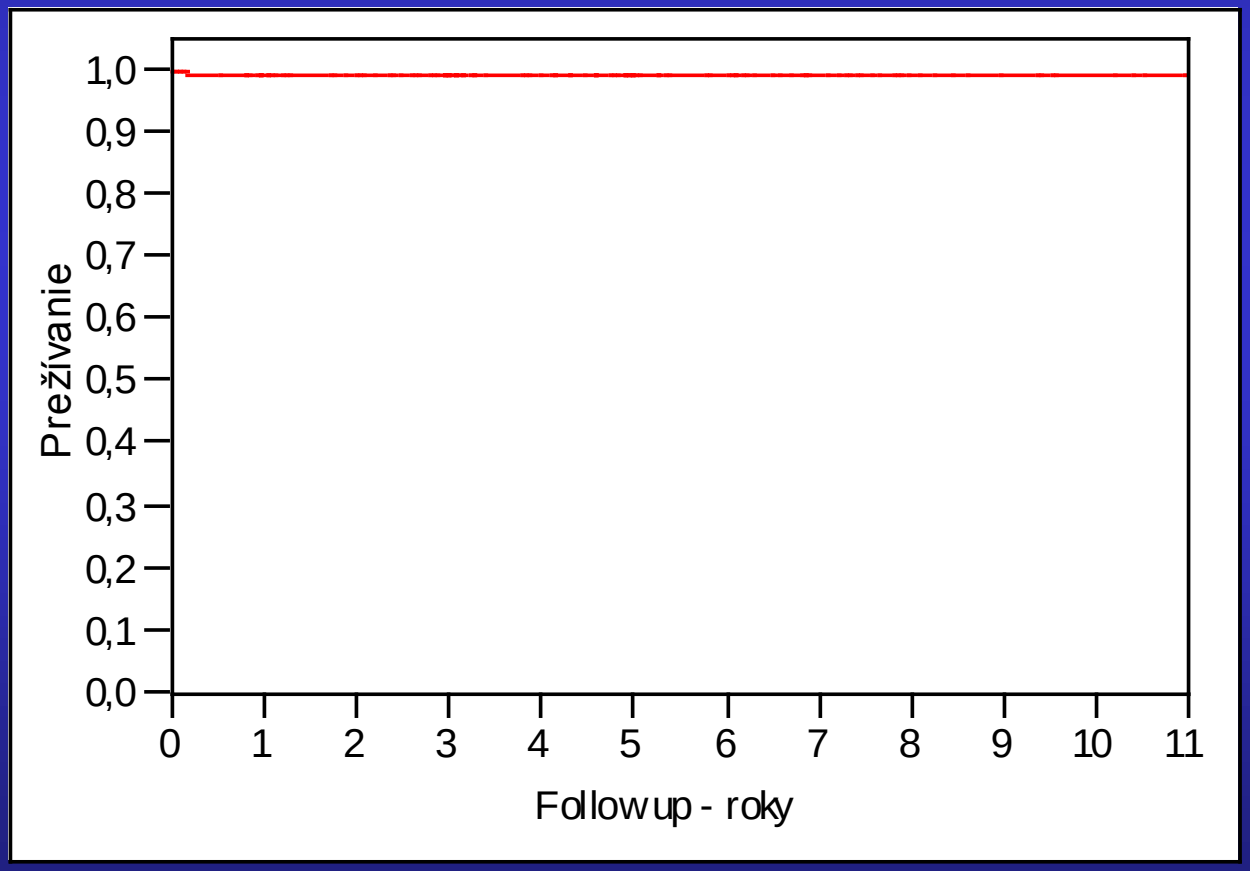
Results

- Late mortality - 1 patient



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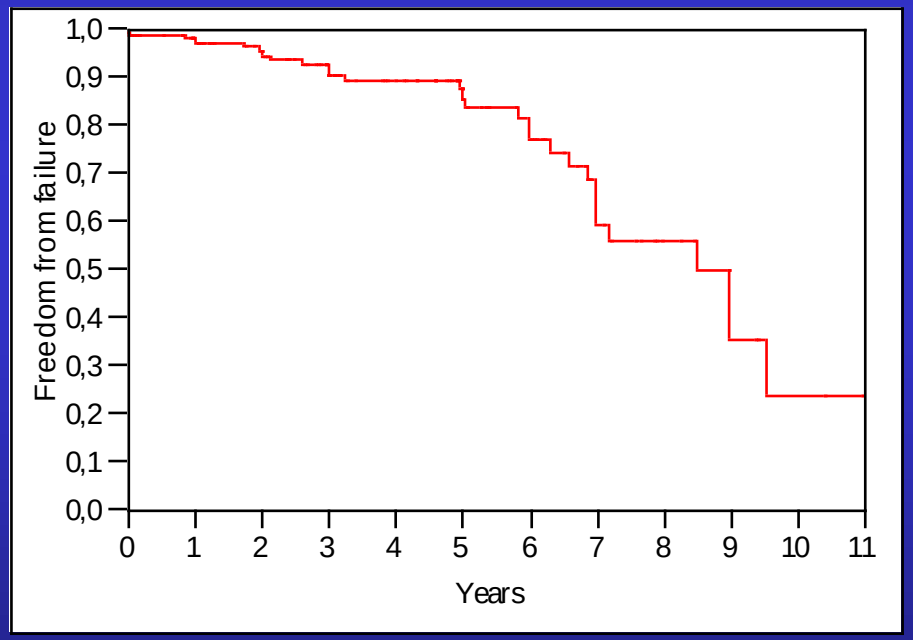
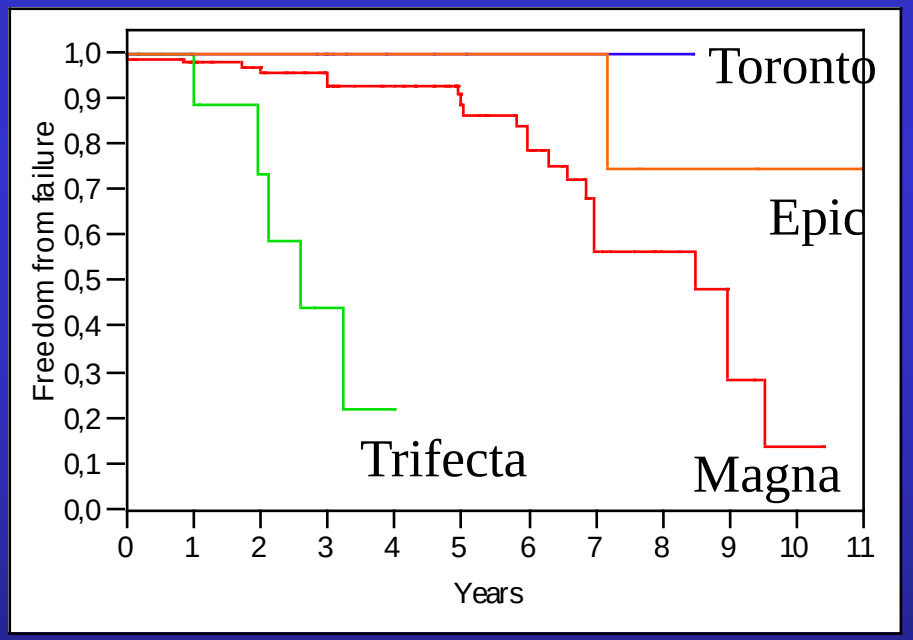
Analysis of the failure of implanted bioprostetic valves

- Systolic gradient ≥ 40 mmHg
- Insufficency of implanted bioprosthetic valve ≥ 2,5 score





- 1, 4 and 8 years – 97%, 90% a 56%
- Difference in various types of implanted valves functions
  - Trifecta p<0,0001

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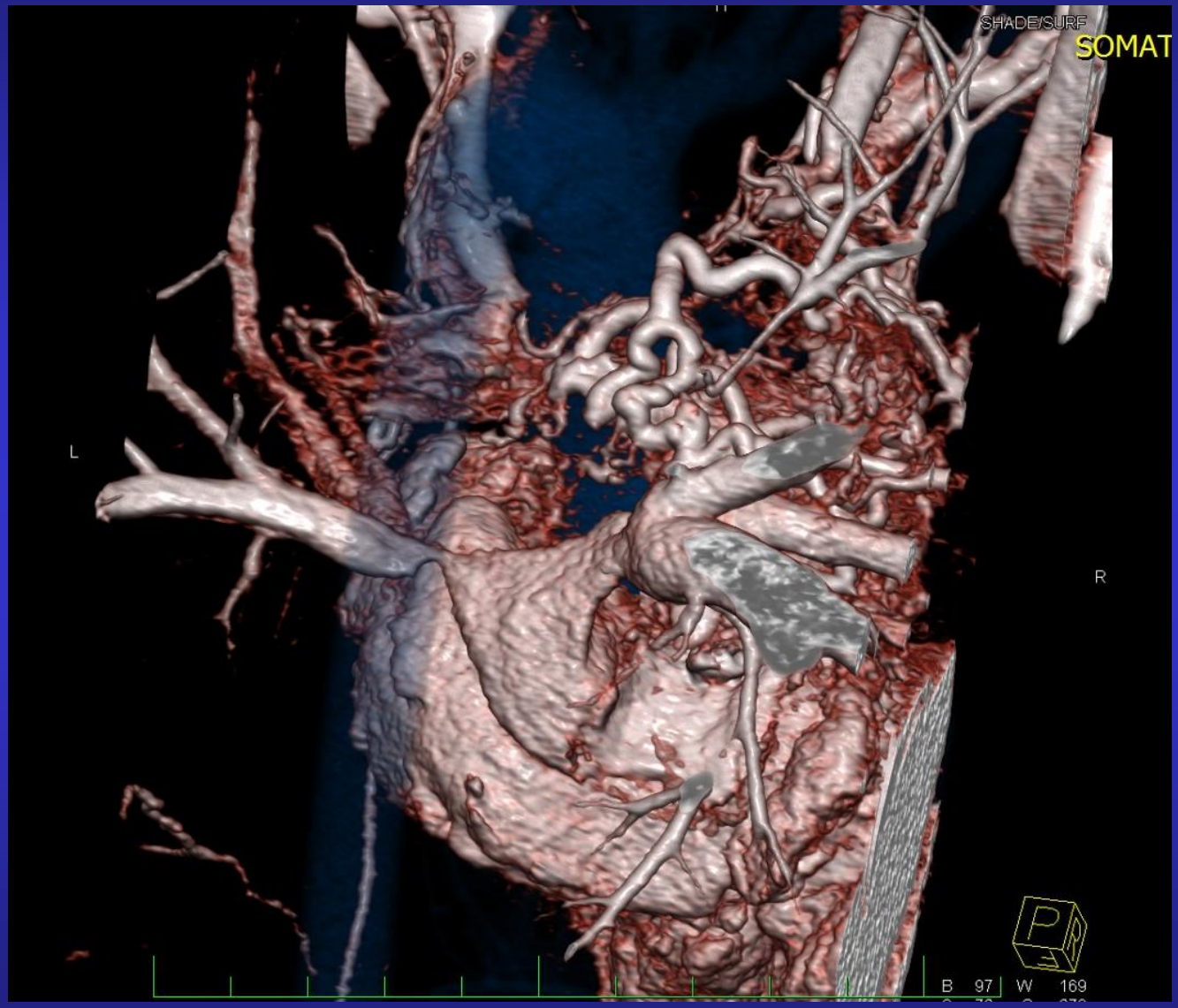
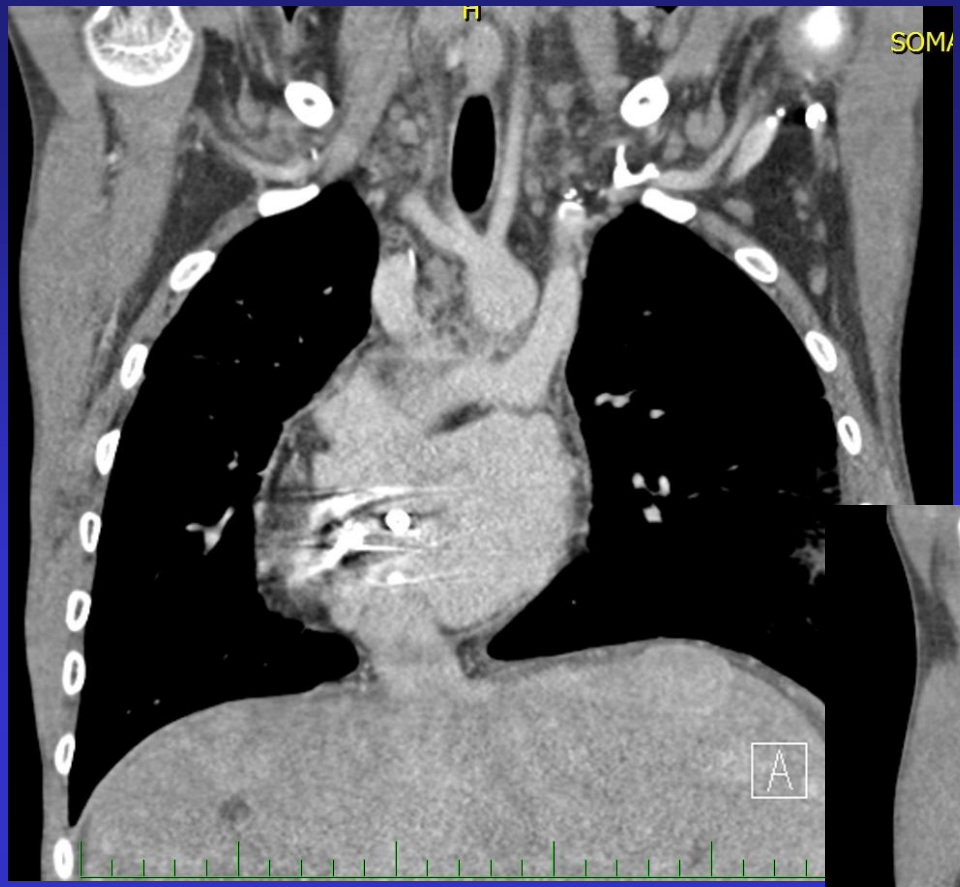
Single ventricle

teamwork

Case report – woman 25 years
Dg: f-SV(DORV, AVSD, ISOM, DXC)

- 1993 MBT
- 1995 TCPC – bilat BCPA, intraatrial goretex tunnel
- 2006 1st redo – closure of a lateral baffle leak, RPA plasty
- 2015 2nd redo – AV valve replacement –SJM 33
- 2015, 2016 cardioversion - SVT intraatrial reentry
- 2016 RFA SVT (multiorgan failure, NYHA IV)


SHADE/SURF
SOMAT
L
R
B 97
W 169


H
SOMA
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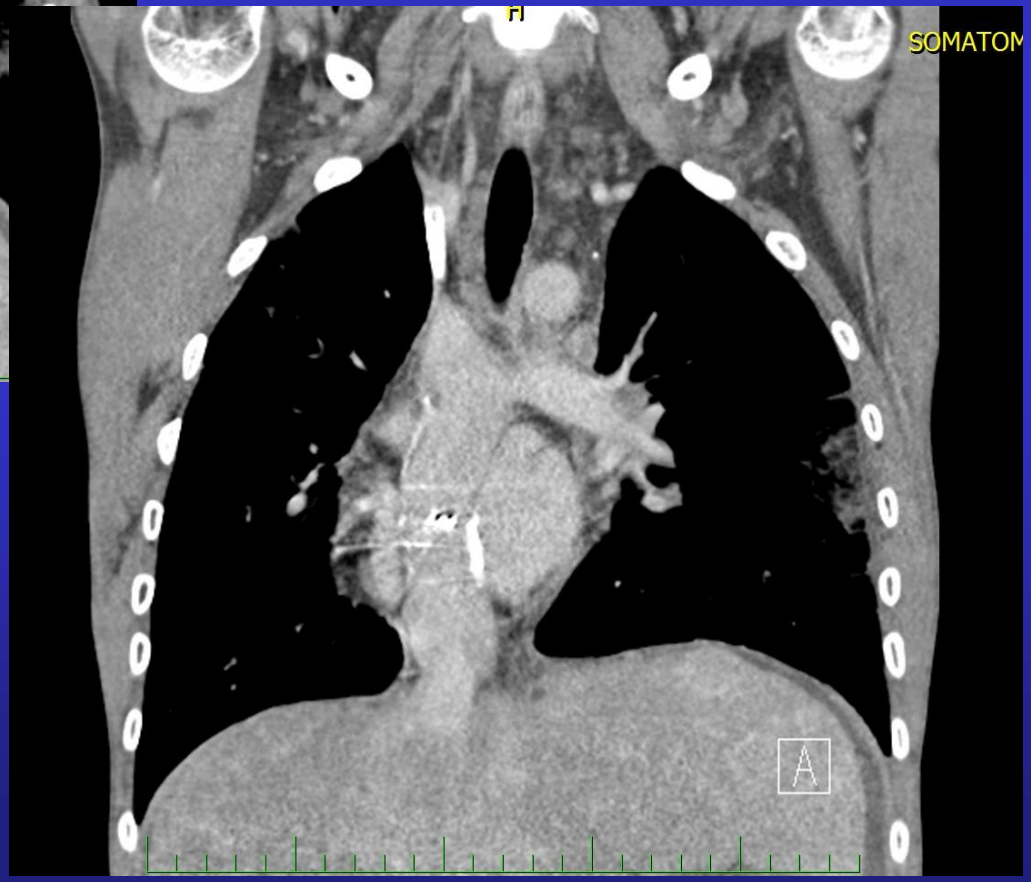
H
SOMATOM
A

Bilateral SVC and femoral percutaneus venous cannulation, femoral arterial cannulation for ECC

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Case report – woman 25 years
Dg: f-SV(DORV, AVSD, ISOM, DXC)

- 1993 MBT
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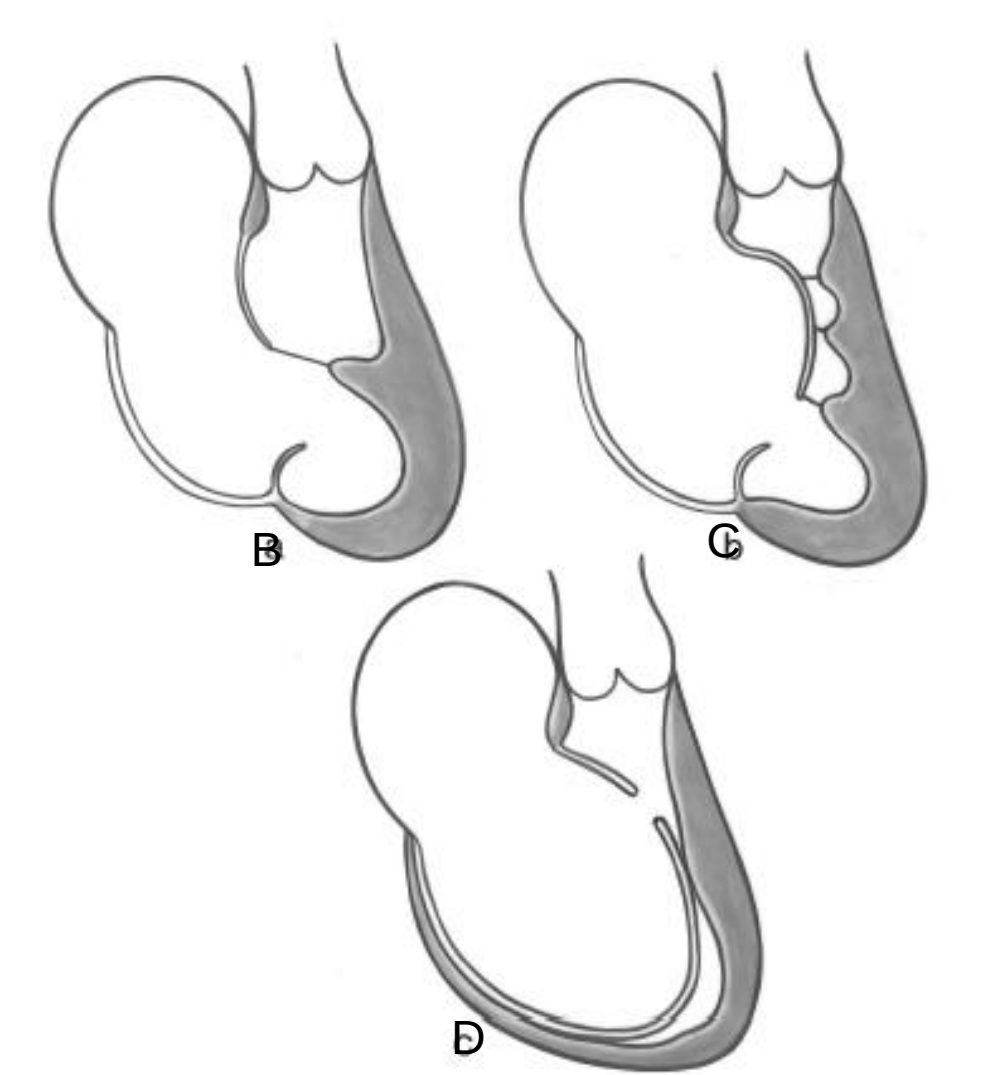
- 2006 1st redo – closure of a lateral baffle leak, RPA plasty
- 2015 2nd redo – AV valve replacement –SJM 33
- 2016 RFA SVT intratrial reentry (multiorgan failure), NYHA IV
- 2016 3rd redo – extracardiac TCPC
  left pulmonary veins plasty
  NYHA II

Ebstein anomaly

centralization

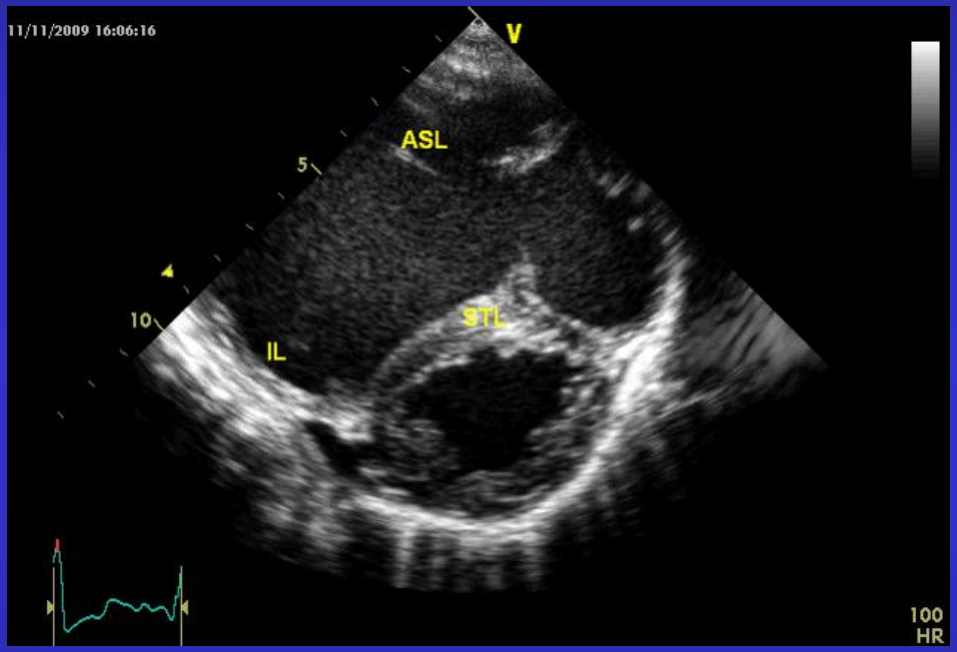
Ebstein's Anomaly Classification

Carpentier 1988

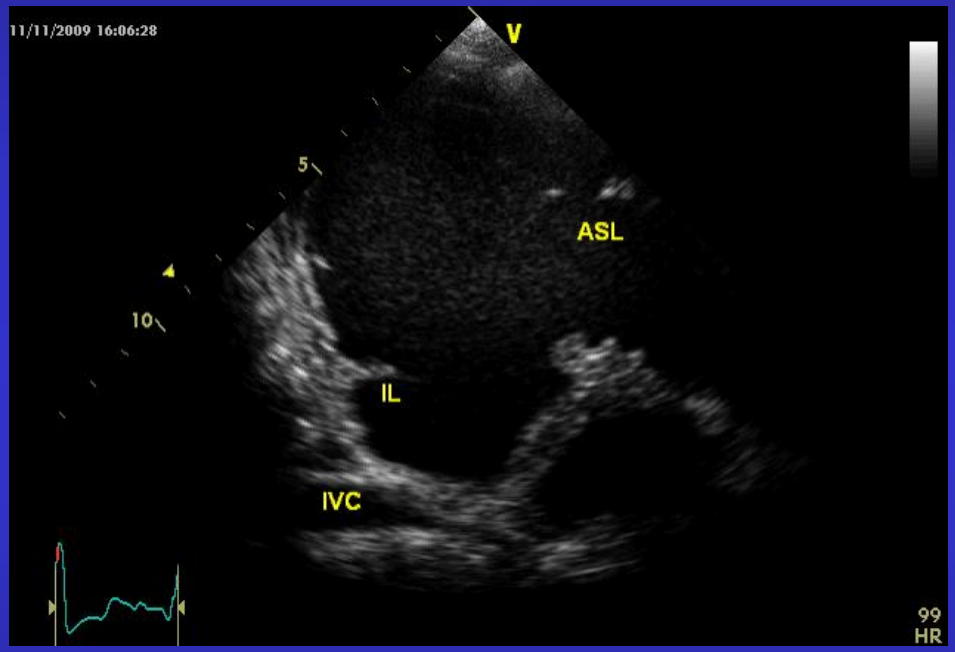


- Type A: adequate RV volume
- Type B: Large atrialised component of RV with mobile anterior leaflet
- Type C: Restricted anterior leaflet, with RVOTO obstruction
- Type D: Complete atrialisation of RV with small infundibular component

Ebstein anomaly: 2D Echocardiography

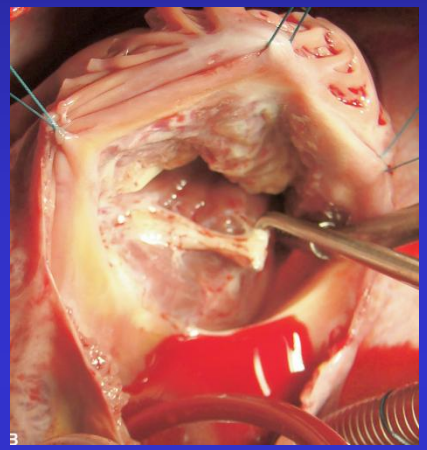


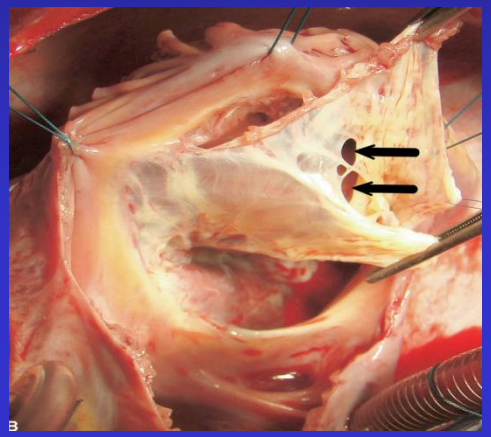
Parasternal short axis view

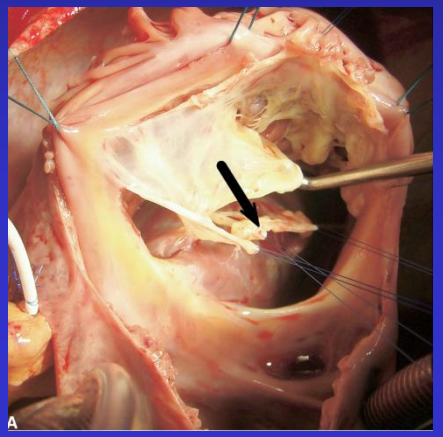


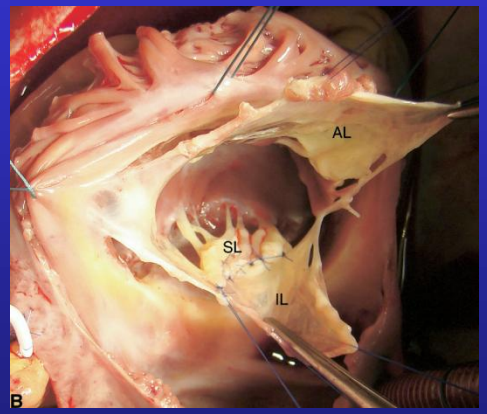
Parasternal right inflow view

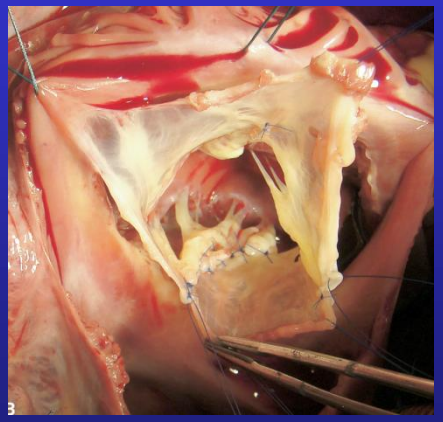
Da Silva technique/Cone repair/anatomic rotation…

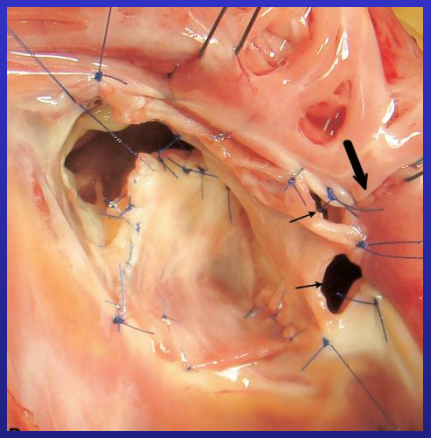












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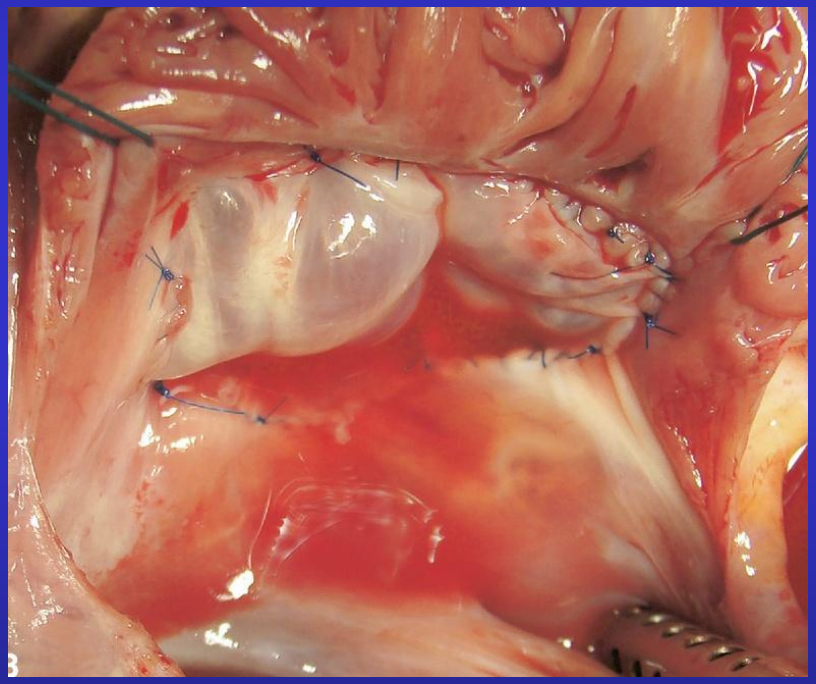
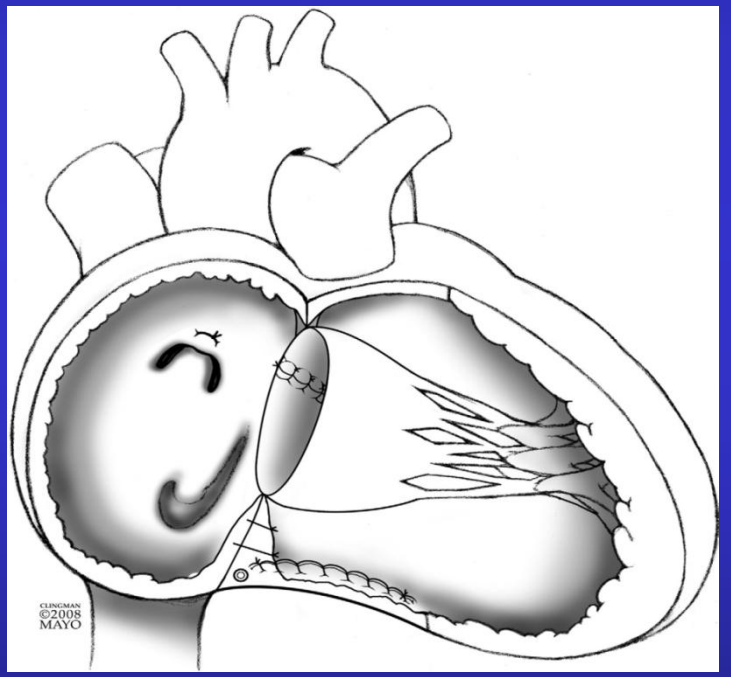
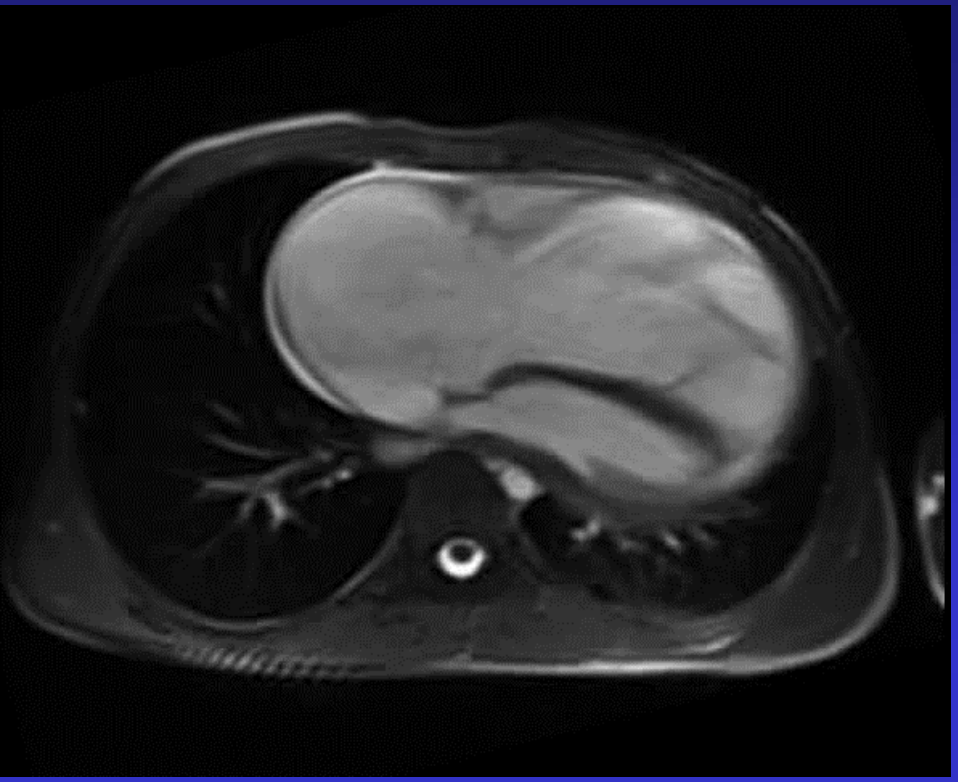
Cone repair – final result

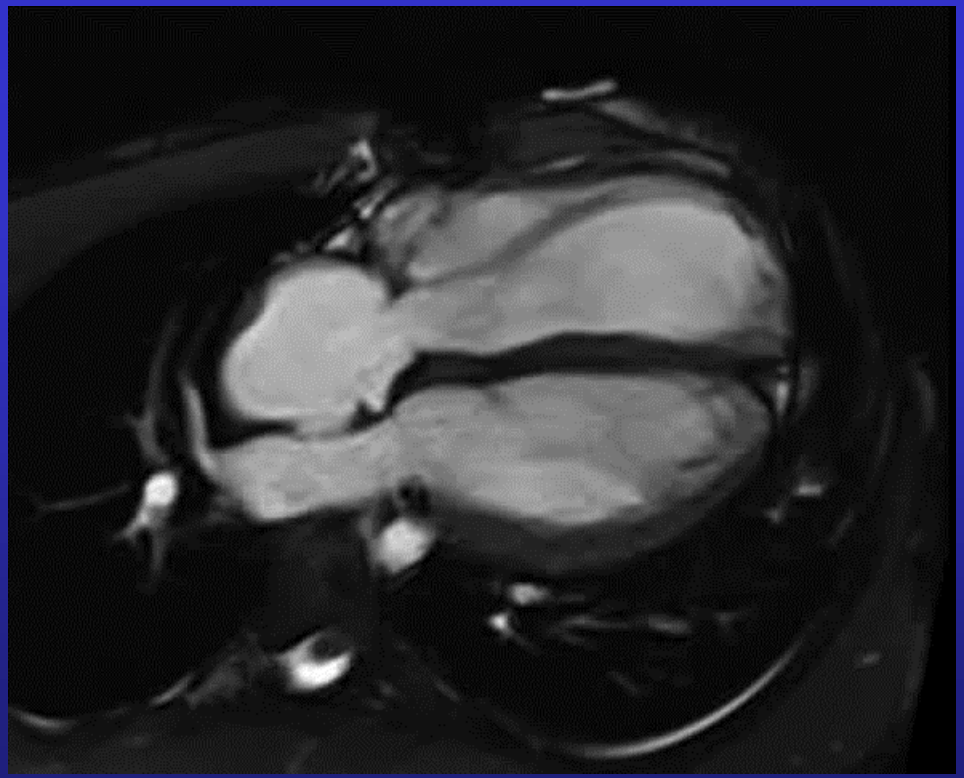




Cone Reconstruction of the Tricuspid Valve for Ebstein's Anomaly: Anatomic Repair. Joseph A. Dearani, MD, Emile Bacha, MD, and José Pedro da Silva, MD



4 chamber view
before and after repair



Preop data (N=47)

• Age	median 33 (18-70) years
• Tricuspid insufficiency	3° 2 pts
	4° 43 pts
• Mitral insufficiency	3° 4 pts
• Ischemic heart disease	1 pt
• Cyanosis (sat. 71-90%)	5 pts
• Arrhythmia	
(WPW,AF, flutter, palpitations)	19 pts (2x RF ablation)
• Previous surgery	10 pts

Surgery (N=51 in 47 pts)

Tricuspid valve procedure **TVP 25**
TVR (bio) 25

- BCPA: 8
- MAZE - dx /sin: 19/2
- Pacemaker: 3
- ASD / FOA closure: 12
- MV repair: 4
- CABG: 1

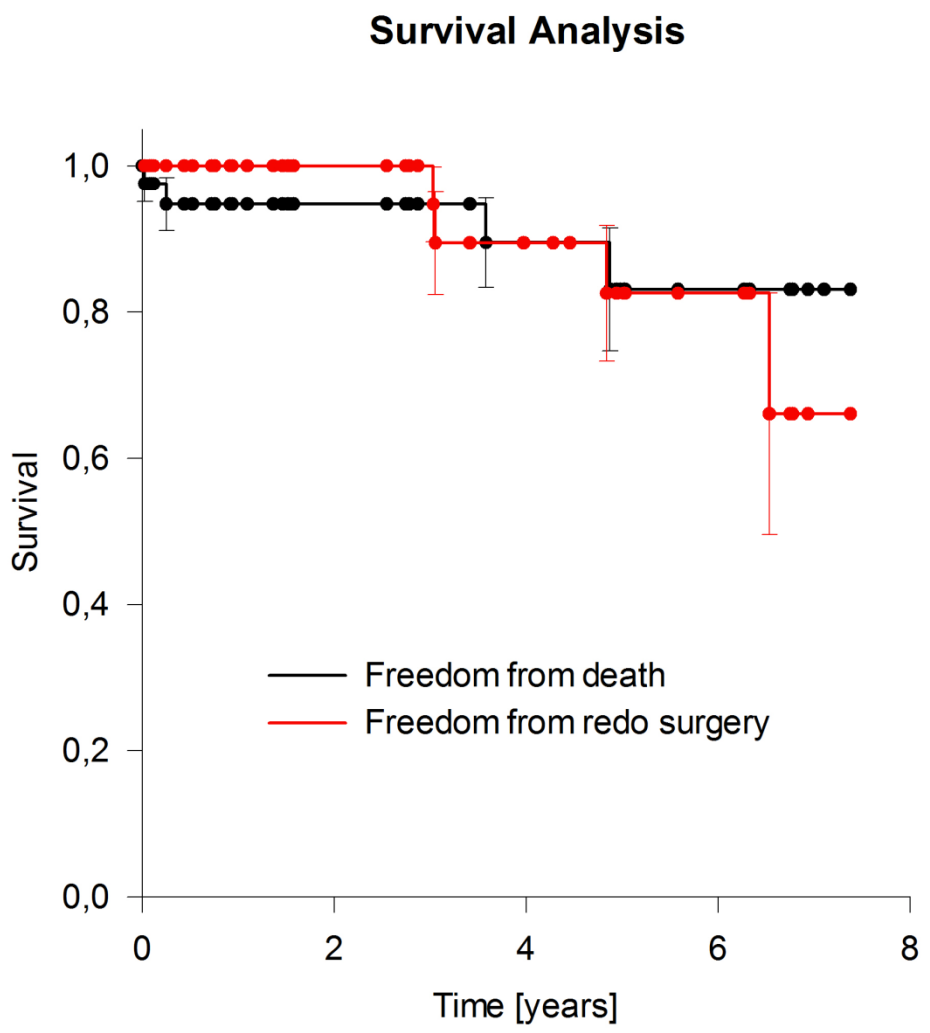
EC 95-255 (152) min, crossclamp 42-150 (105) min
Cardioplegia blood, cold, intermittent

Results

mortality 4/47 (early 2 pts 4,25%, late 2 pts 4,25%)

	before surgery	**after suregy**	**p**
NYHA	2,6 ± 0,7	1,6 ± 0,6	< 0,001
Tricuspid regurgitation 1st-4th degree in TVP patients	3,8 ± 0,5	1,4 ± 0,9	< 0,001
RV EF %	35 ± 10,3 %	40 ± 11,3%	0,01

Survival


Survival Analysis
1,0
0,8
0,6
0,4
0,2
0,0
Survival
0
2
4
6
8
Time [years]
Freedom from death
Freedom from redo surgery

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