# Long term outcome after AVSD repair

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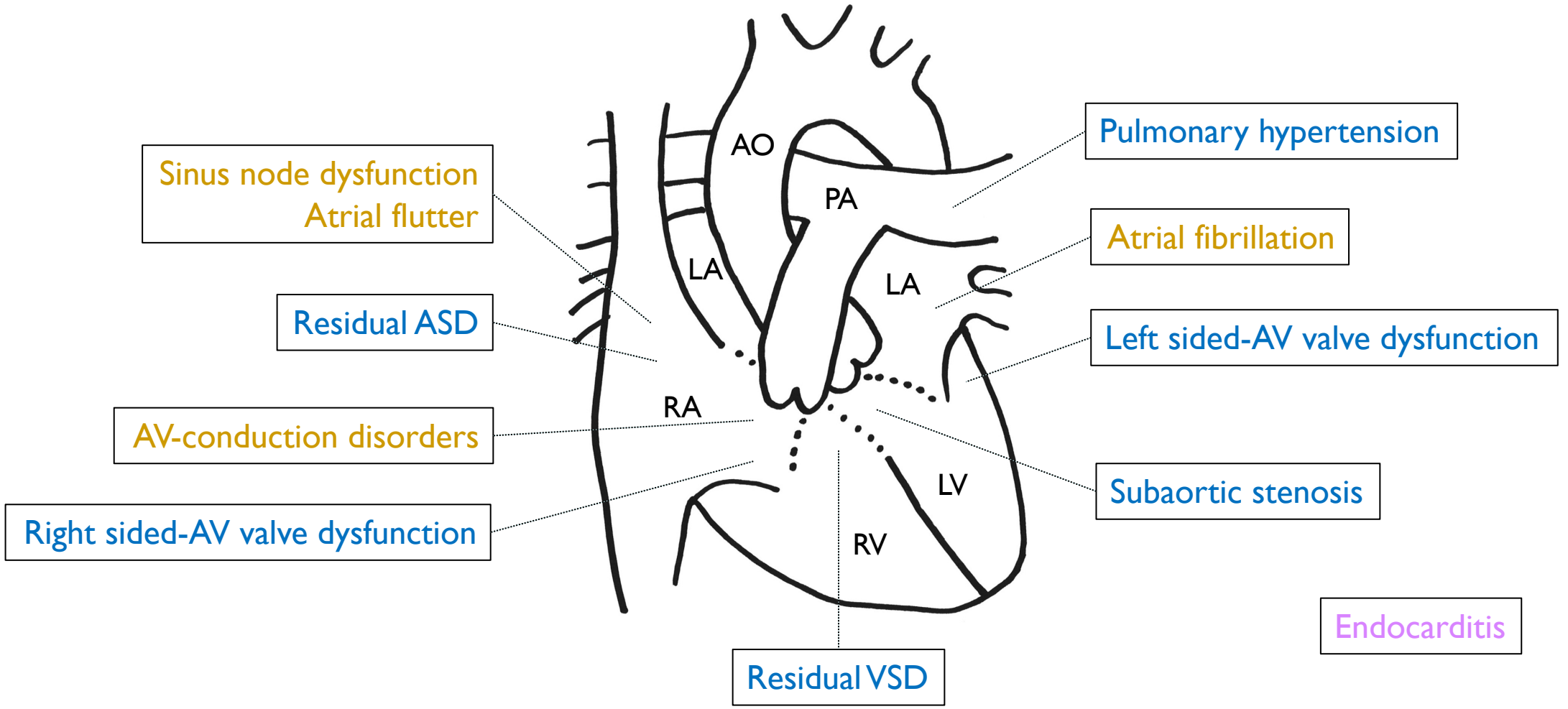
# Conflict of interest

- Adult congenital cardiologist
  - Facing the results of the therapeutic decisions by pediatric cardiologists and congenital cardiac surgeons

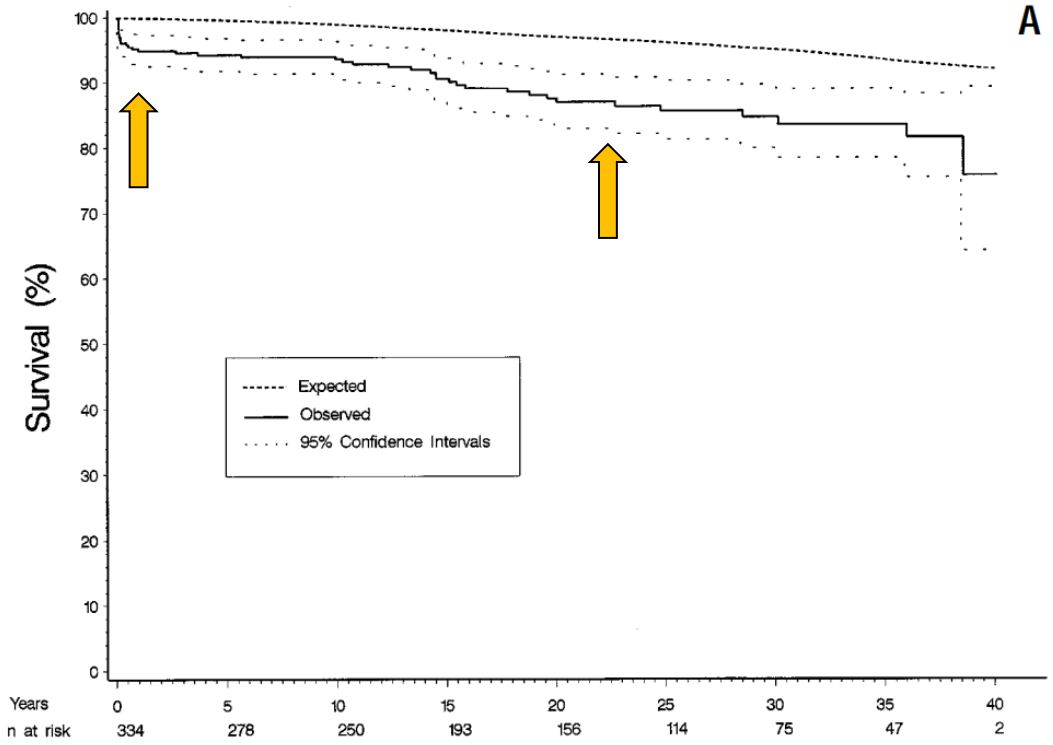
# Sequellae late after atrioventricular septal defect repair



# Survival and decades of surgery

90% at 15 years

85% at 40 years




95% at 15 years

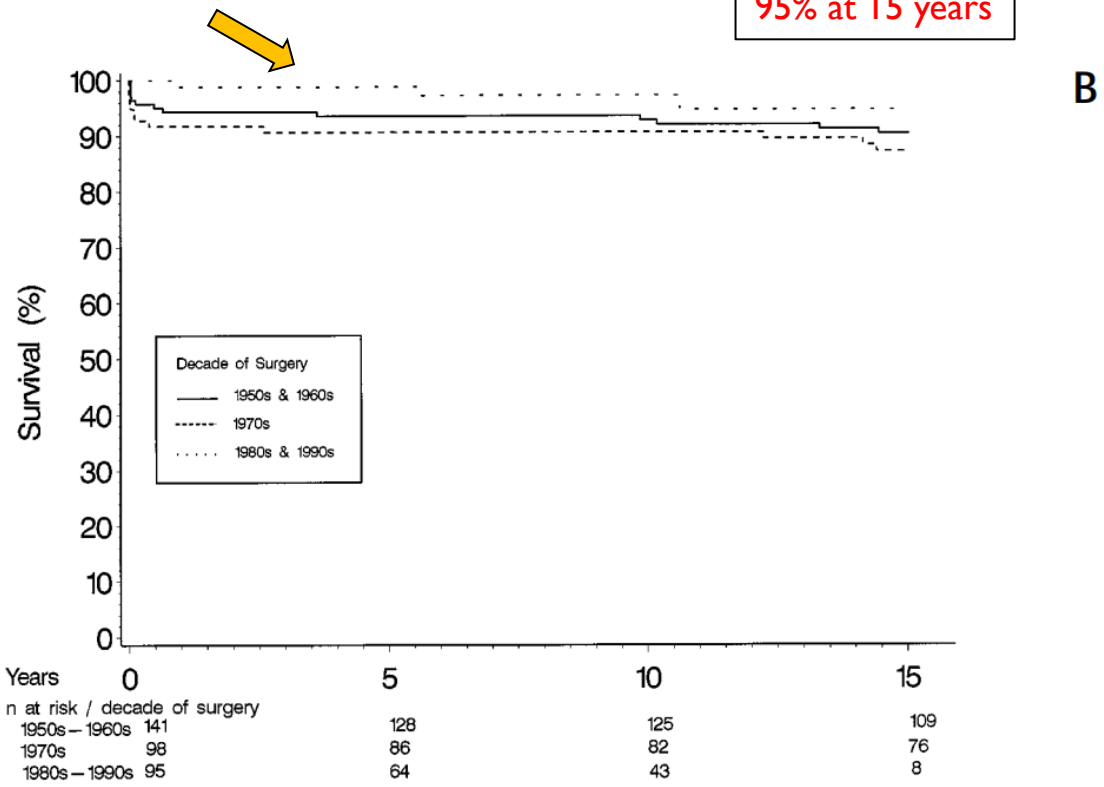
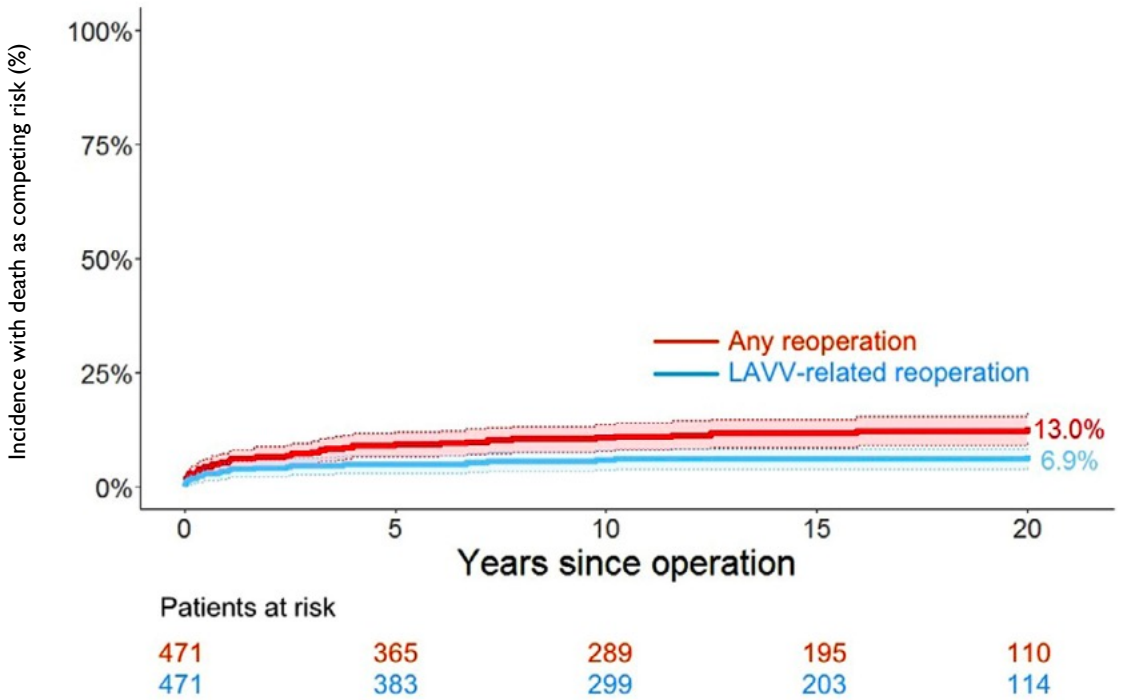



**Fig 1.** Kaplan-Meier curve represents overall survival **(A)** and by the decade of the operation **(B)** in this cohort of patients.

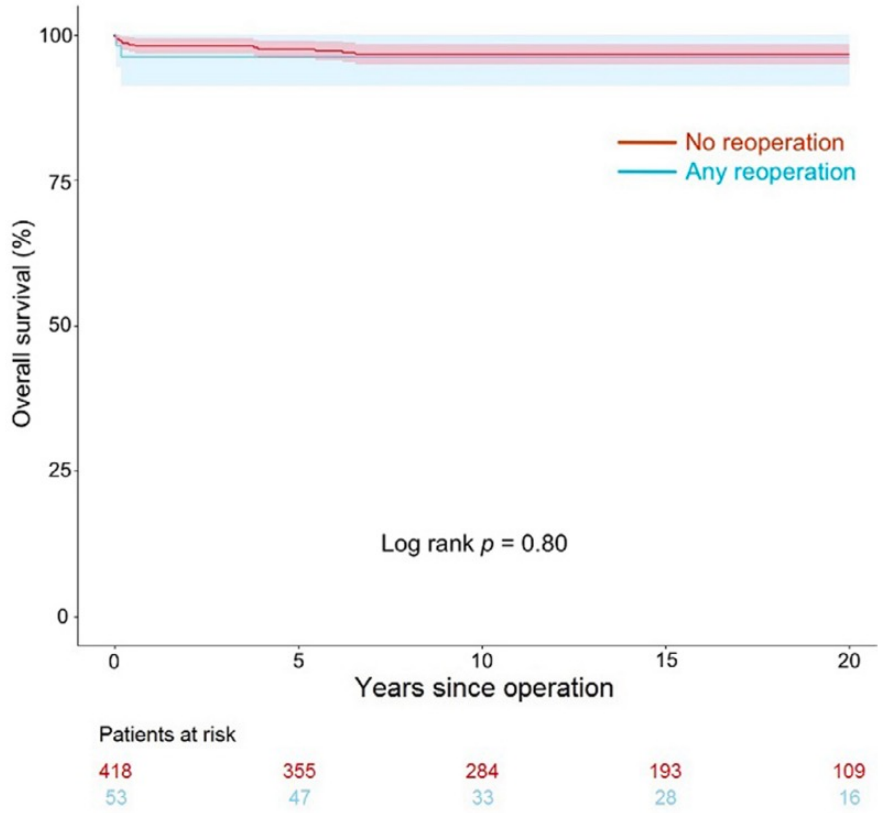
El-Najdawi et al. J Thorac Cardiovasc Surg 2000

# Re-operation and impact on survival

Overall re-operation rate: 10 -15%




No significant impact on mortality (perioperative ✝ 3.8%, late ✝ 0%)




Ramgren et al. Semin Thoracic Surg 2022

# Risk factors of overall re-operation

**Table 3.** Univariable and Multivariable Analysis of Risk Factors for Any Reoperation After Primary Repair of AVSD

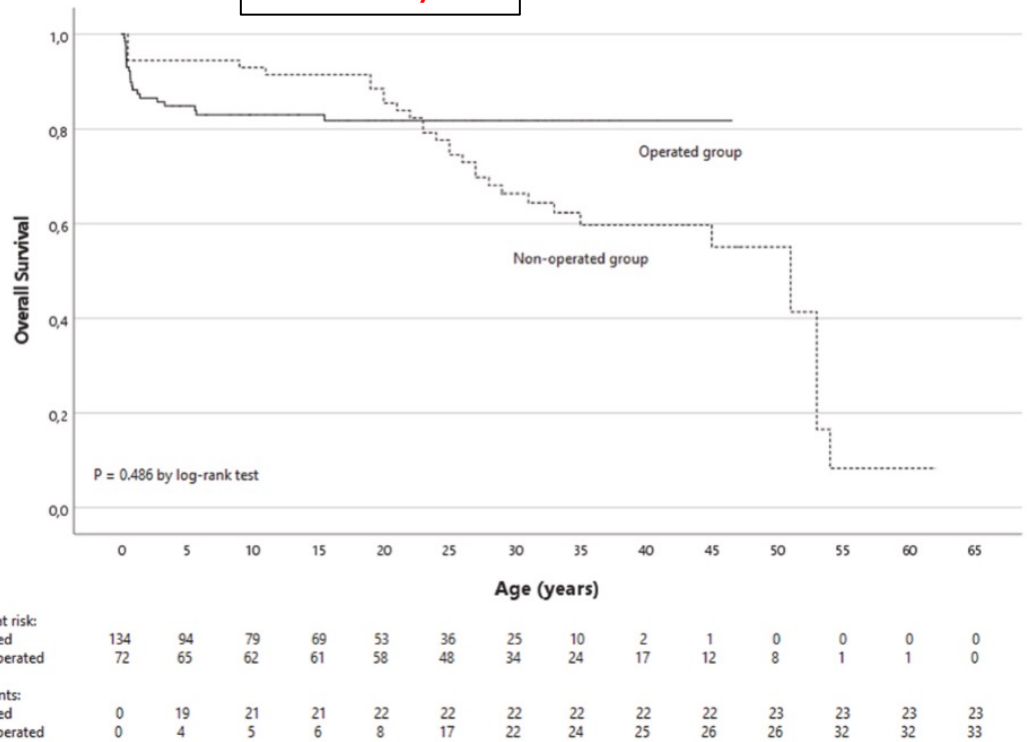
| | Characteristic | UnivariableHR (95% CI) | *P* value | MultivariableHR (95% CI) | *P* value |
|---|---|---|---|---|---|
| | AVSD subtypes | | | | |
| | pAVSD | 1 | | | |
| | tAVSD | 5.15 (1.72-15.4) | 0.003 | | |
| | cAVSD | 1.96 (0.69-5.55) | 0.206 | | |
| | Age at primary repair (y) | 0.99 (0.98-1.01) | 0.276 | | |
| | Weight at primary repair (kg) | 0.98 (0.94-1.02) | 0.367 | | |
| | Female gender | 0.93 (0.54-1.60) | 0.797 | | |
| - | Trisomy 21 | 0.39 (0.22-0.67) | 0.001 | 0.51 (0.29-0.90) | 0.019 |
| | Associated cardiac malformations | 1.78 (1.00-3.14) | 0.049 | | |
| | Coarctation | 1.79 (0.44-7.36) | 0.421 | | |
| + | Tetralogy of Fallot | 5.15 (1.59-16.7) | 0.006 | 6.13 (1.86-20.2) | 0.003 |
| | Prior cardiac/palliative surgery | 2.06 (0.74-5.70) | 0.166 | | |
| | Double orifice LAVV | 1.05 (0.25-4.31) | 0.950 | | |
| | Incomplete commissures | 3.00 (1.56-5.77) | 0.001 | | |
| | Extra cleft | 1.63 (0.73-3.62) | 0.230 | | |
| | LV single papillary muscle head | 3.29 (1.40-7.69) | 0.006 | | |
| + | Unbalanced AV-valves/ventricles | 5.24 (2.36-11.6) | <0.001 | 5.24 (2.30-11.9) | <0.001 |
| | Incomplete cleft closure | 2.43 (1.28-4.63) | 0.007 | | |
| | Residual mitral regurgitation | | | | |
| | None/mild | 1 | | | |
| + | Moderate | 3.65 (1.76-7.59) | 0.001 | 2.88 (1.37-6.05) | 0.005 |
| | Severe | 69.4 (24.5-197) | <0.001 | 40.7 (14.9-111) | <0.001 |

AVSD, atrioventricular septal defect; AV, atrioventricular; CI, confidence interval; HR, hazard ratio; LAVV, left atrioventricular valve; LV, left ventricle; p/t/cAVSD, partial/transitional/complete atrioventricular septal defect.
Values were expressed as number and percentage (%) or as mean ± standard deviation.

Ramgren et al. Semin Thoracic Surg 2022

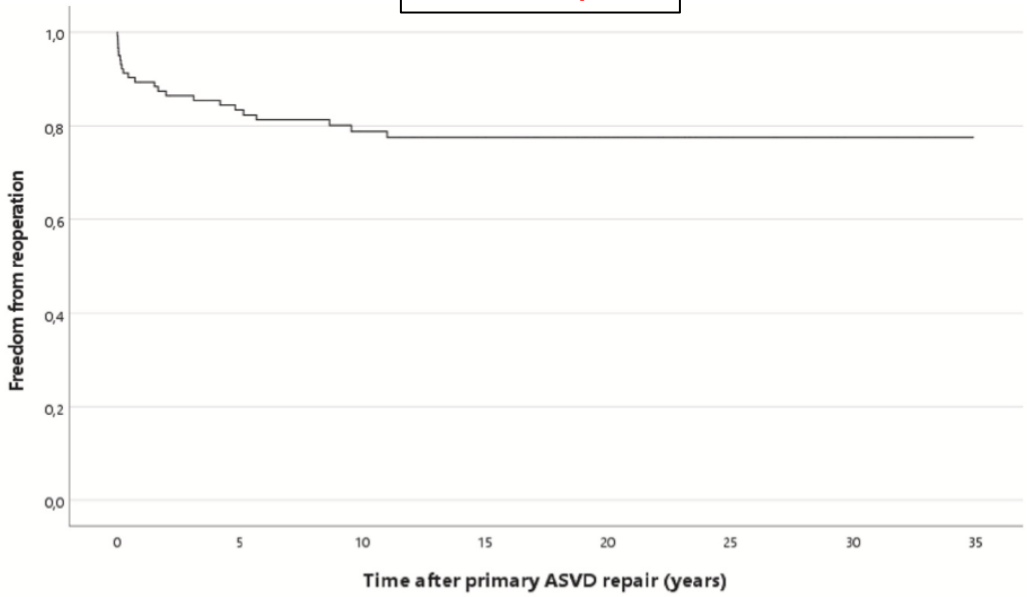
# Survival and re-operation in Down syndrome

80% at 15 years




| Patients at risk: | | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| operated | 134 | 94 | 79 | 69 | 53 | 36 | 25 | 10 | 2 | 1 | 0 | 0 | 0 | 0 |
| non-operated | 72 | 65 | 62 | 61 | 58 | 48 | 34 | 24 | 17 | 12 | 8 | 1 | 1 | 0 |
| **Total events:** | | | | | | | | | | | | | | |
| operated | 0 | 19 | 21 | 21 | 22 | 22 | 22 | 22 | 22 | 22 | 23 | 23 | 23 | 23 |
| non-operated | 0 | 4 | 5 | 6 | 8 | 17 | 22 | 24 | 25 | 26 | 26 | 32 | 32 | 33 |

80% at 15 years




| Patients at risk: | 134 | 80 | 60 | 53 | 35 | 13 | 6 | 0 |
|---|---|---|---|---|---|---|---|---|
| Total events: | 0 | 18 | 22 | 23 | 23 | 23 | 23 | 23 |

**Fig. 2.** Kaplan Meier redo surgery free curve in repaired patients.



Daene et al. Int J Cardiol CHD 2023

# Re-operations and findings on the left-sided AV-valve

50% of re-operation at left sided AV-valve

**Table 2.** Indications for First Reoperation After Primary Repair for AVSD

| Indication for Reoperation | pAVSD | tAVSD | cAVSD | Total |
|---|---|---|---|---|
| Number of primary repairs | 78 (16.6) | 88 (18.7) | 305 (64.8) | 471 (100)* |
| LAVV pathology | 2 (2.6) | 9 (10.2) | 17 (5.6) | 28 (5.9) |
| LVOTO | 1 (1.3) | 6 (6.8) | 9 (3.0) | 16 (3.4) |
| Residual shunt | 1 (1.3) | 2 (2.3) | 4 (1.3) | 7 (1.5) |
| Pacemaker implantation | 0 (0) | 0 (0) | 2 (0.7) | 2 (0.4) |
| Total | 4 (5.1) | 17 (19.3) | 32 (10.5) | 53 (11.1) |

LAVV, left atrioventricular valve; LVOTO, left ventricle outflow tract obstruction; p/t/cAVSD, partial/transitional/complete atrioventricular septal defect.
Values presented as number and (%).
*Six patients were lost to follow-up (2 with pAVSD and 4 with cAVSD).

Findings on left AV-valve

**Table 4.** Perioperative Findings in LAVV at Re-repair

| Pathology* | *n* | % |
|---|---|---|
| Repair dehiscence | 15 | 58 |
| Residual cleft | 7 | 27 |
| Dysplastic leaflet tissue | 7 | 27 |
| Stenosis after prior repair | 4 | 15 |
| Abnormal leaflet pliability | 3 | 12 |
| Leaflet perforation | 3 | 12 |
| Leaflet prolapse-tethering | 2 | 7 |
| Double orifice | 2 | 7 |
| Deficient mural leaflet | 1 | 4 |
| Additional cleft (previously unrepaired) | 1 | 4 |
| Septation patch dehiscence | 1 | 4 |
| Myxomatous leaflet tissue | 1 | 4 |
| Chordal rupture | 1 | 4 |

LAVV, left atrioventricular valve.
*More than one pathologic finding may have been present in any given patient.

Ramgren et al. Semin Thoracic Surg 2022

# Re-operation for left or right atrioventricular valve

Re-operation at median 7 years; range 1 month - 71 years

Table 3
First reoperations performed in the total study population

| Reoperation | PAVSD $n=31$ (%) | CAVSD $n=28$ (%) | Total $n=59$ (%) | *P*-value |
|---|---|---|---|---|
| Repair of the LAVV | 22 (71) | 23 (82) | 45 (76) | 0.31 |
| Cleft closure | 21 (68) | 23 (82) | 44 (75) | 0.21 |
| Commissuroplasty | 9 (29) | 10 (36) | 19 (32) | 0.58 |
| Annuloplasty | 2 (6) | 0 (0) | 2 (3) | 0.49 |
| Mechanical valve replacement of the LAVV | 6 (19) | 2 (7) | 8 (14) | 0.26 |
| Residual ASD repair | 3 (10) | 0 (0) | 3 (5) | 0.24 |
| Enucleation of LVOT obstruction | 0 (0) | 3 (11) | 3 (5) | 0.10 |
| Additional corrections | | | | |
| Repair of the RAVV | 5 (16) | 5 (18) | 10 (17) | 0.86 |
| Cleft closure | 0 (0) | 3 (11) | 3 (5) | 0.10 |
| Commissuroplasty | 4 (13) | 3 (11) | 7 (12) | 1.0 |
| Annuloplasty | 1 (3) | 0 (0) | 1 (2) | 1.0 |
| Residual VSD repair | 2 (6) | 5 (18) | 7 (12) | 0.24 |
| Residual ASD repair | 9 (29) | 3 (11) | 12 (20) | 0.10 |
| Enucleation of LVOT obstruction | 0 (0) | 1 (4) | 1 (2) | 0.22 |
| Mechanical aortic valve replacement | 1 (3) | 0 (0) | 1 (2) | 1.0 |

PAVSD, partial atrioventricular septal defect; CAVSD, complete atrioventricular septal defect; LAVV, left atrioventricular valve; RAVV, right atrioventricular valve; VSD, ventricular septal defect; ASD, atrial septal defect; LVOT, left ventricular outflow tract.

Birim et al. Interact Cardiovasc Thorac Surg 2009

# Replacement or repair of the left AV-valve?

Risk for re-re-operation

Table 1
Incidence of left AV valve anomalies and results of valve reconstruction

| Anomalies | Left AV valve incompetence after reoperation (grade) | | | |
|---|---|---|---|---|
| | 0 | I | II | III |
| (No. of patients) | ($n = 13$) | ($n = 5$) | ($n = 4$) | ($n = 6$) |
| Dysplastic valvular tissue | 1 | 1 (1) | | 3[a] |
| Fibrotic deformity of septal leaflet | 1 | 2 | 2 | 3 |
| Posterior leaflet prolapse | | 1 (1) | | 1[a] |
| Parachute valve | | | 2 | |
| Severe deformity of the valve | | | | 1[a] |
| Double orifice valve | 1 | | 1 | 1 |
| Total | 3 | 4 | 5 | 9 |

( ), numbers in parenthesis indicate deceased patients.
[a] Subsequent left AV valve replacement performed.

Risk for pacemaker implantation

*Prosthetic Valve Complications*

Overall, 61.2% of patients (19 of 31) were free of a prosthetic valve. There were no major bleeding or thromboembolic complications for patients who underwent prosthetic mitral valve replacement. Severe left ventricular systolic dysfunction developed in 3 patients with a prosthetic valve. One of these patients required heart transplantation owing to progression of cardiomyopathy.

*Need for Pacemaker Implantation*

Pacemaker placement was required in 8 patients (25%). In this series, patients who underwent valve replacement had a significantly higher incidence of pacemaker insertion than did patients who underwent valve repair (62.5% versus 13.0%, $p = 0.005$).

*Durability of Left Atrioventricular Valve Repair*

At follow-up, 71.4% of operative survivors of valve repair (15 of 21) were free of prosthetic valve replacement. The median time to valve replacement after recurrent LAVVR was 3.3 years. Those with a durable repair had mild or less LAVVR in 93.3% (14 of 15) at recent echocardiography. The remaining patient had moderate LAVVR, was stable on serial echocardiograms, and was clinically asymptomatic.

Analysis of factors impacting durability of valve repair

Alexi-Meskishvili et al. Eur J Cardiothorac Surg 1997; Malhotra et al. Ann Thorac Surg 2008

# Mechanical valve or repair of the left AV-valve?

Long-term survival

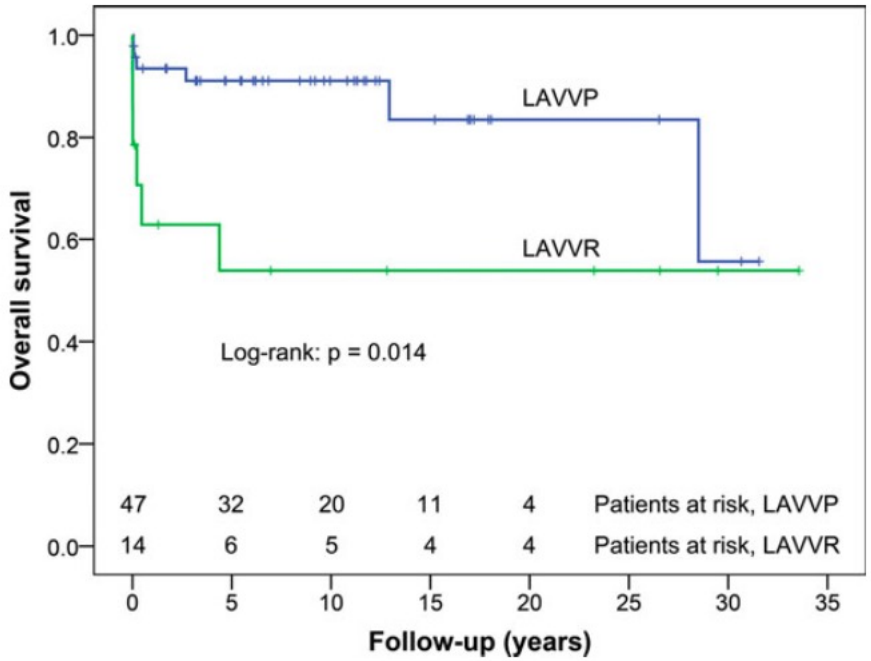



**Figure 1:** Overall survival after reoperation for LAVV pathology at the first reoperation ($n = 61$). LAVV: left atrioventricular valve; LAVVP: left atrioventricular valvuloplasty; LAVVR: left atrioventricular valve replacement.

Long-term data on reoperation

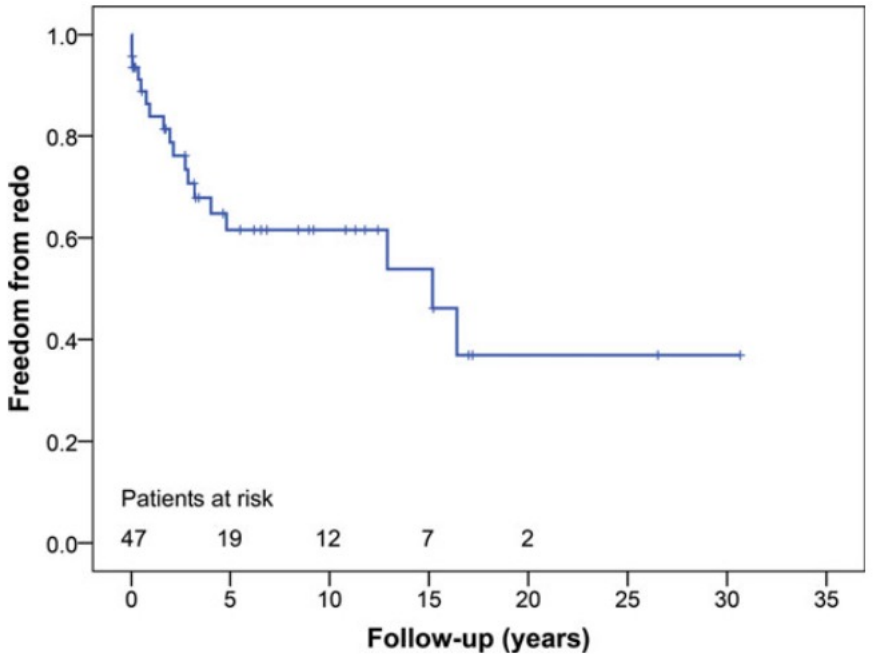



**Figure 4:** Durability of LAVVP at the first reoperation ($n = 47$). LAVVP: left atrioventricular valvuloplasty.

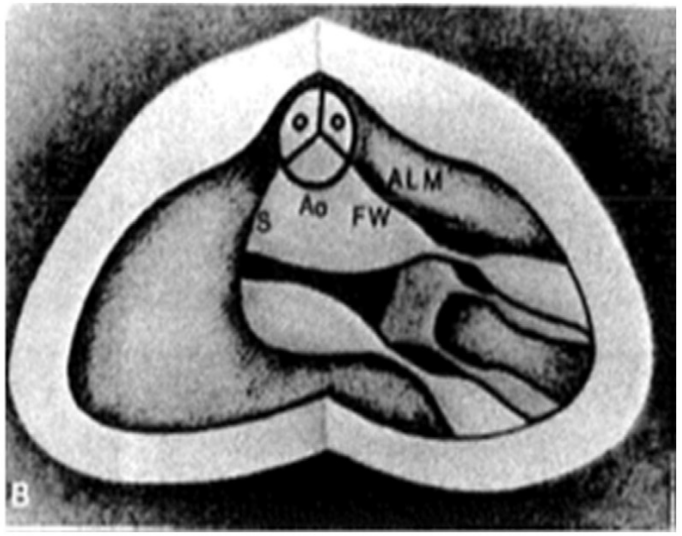
Sojak et al. Eur J Cardiothorac Surg 2016

# When to intervene on the left AV-valve?

| AV valve regurgitation | | |
|---|---|---|
| Valve surgery, preferably AV valve repair, is recommended in symptomatic patients with moderate to severe AV valve regurgitation and should be performed by a congenital cardiac surgeon. | I | C |
| In asymptomatic patients with severe left-sided AV valve regurgitation, valve surgery is recommended when LVESD ≥45 mm[d] and/or LVEF ≤60% provided other causes of LV dysfunction are excluded. | I | C |
| In asymptomatic patients with severe left-sided AV valve regurgitation, preserved LV function (LVESD <45 mm[d] and/or LVEF >60%), high likelihood of successful valve repair, and low surgical risk, intervention should be considered when atrial fibrillation or systolic PAP >50 mmHg is present. | IIa | C |

ESC ACHD guidelines 2020

# Re-operation for left ventricle outflow tract obstruction




Reoperation occurs at 5 – 7 years
Reoperation free after 10 years: 95%

*Predictors of LVOTO*

- Displacement of the AV valve leaflet tips into the LV
- Presence of fixed or thick chords in the LVOT
- An acute aortoseptal angle
- High insertion of the anterolateral papillary muscle
- An abnormal aorta to subaortic ratio

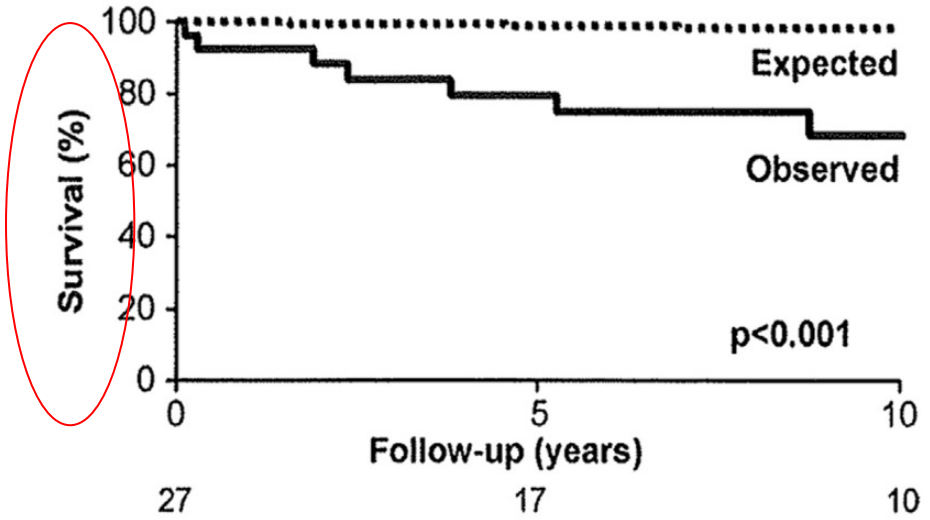



**Figure 1** Actuarial survival after reoperation for LVOTO after repair of AVSD. A significant difference ($P < .001$) is observed as compare with age and gender matched population.

Overman. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2014; Najm et al. Circulation 1997

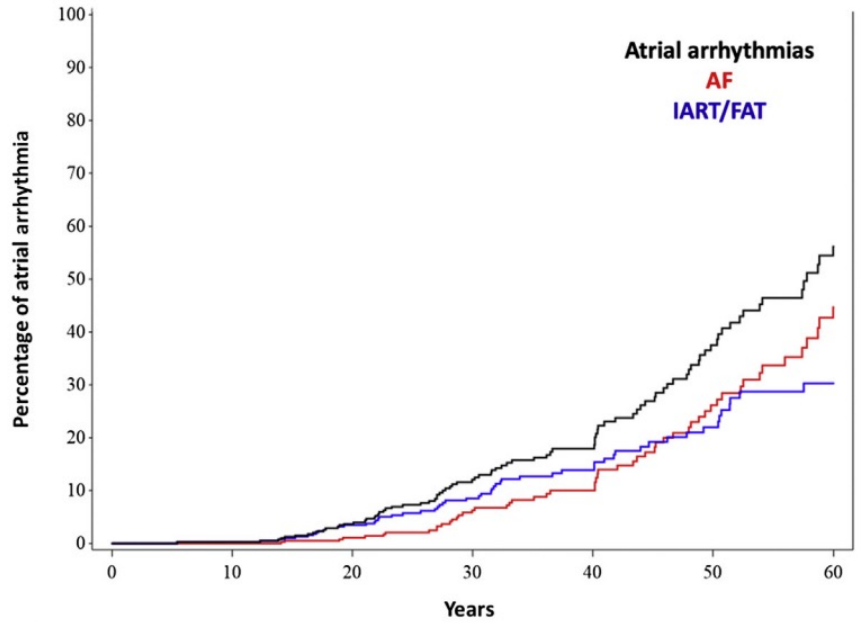
# When to intervene on the LVOT?

| Recommendations | Class[a] | Level[b] |
|---|---|---|
| In symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥40 mmHg[c] or severe AR, surgery is recommended. | I | C |
| Asymptomatic patients should be considered for surgery when one or more of the following findings are present:<br>• Mean gradient <40 mmHg but LVEF <50%.<br>• AR is severe and LVESD >50 mm (or 25 mm/m$^2$ BSA) and/or EF <50%[d].<br>• Mean Doppler gradient is ≥40 mmHg[c] and marked LVH present.<br>• Mean Doppler gradient is ≥40 mmHg[c] and there is a fall in blood pressure below baseline on exercise. | IIa | C |
| Asymptomatic patients may be considered for surgery when one or more of the following findings are present:<br>• Mean Doppler gradient is ≥40 mmHg,[c] LV is normal (EF >50% and no LVH), exercise testing is normal, and surgical risk is low.<br>• Progression of AR is documented and AR becomes more than mild (to prevent further progression). | IIb | C |

ESC ACHD guidelines 2020

# Supraventricular arrhythmia

**FIGURE 1** Lifetime Cumulative Incidences

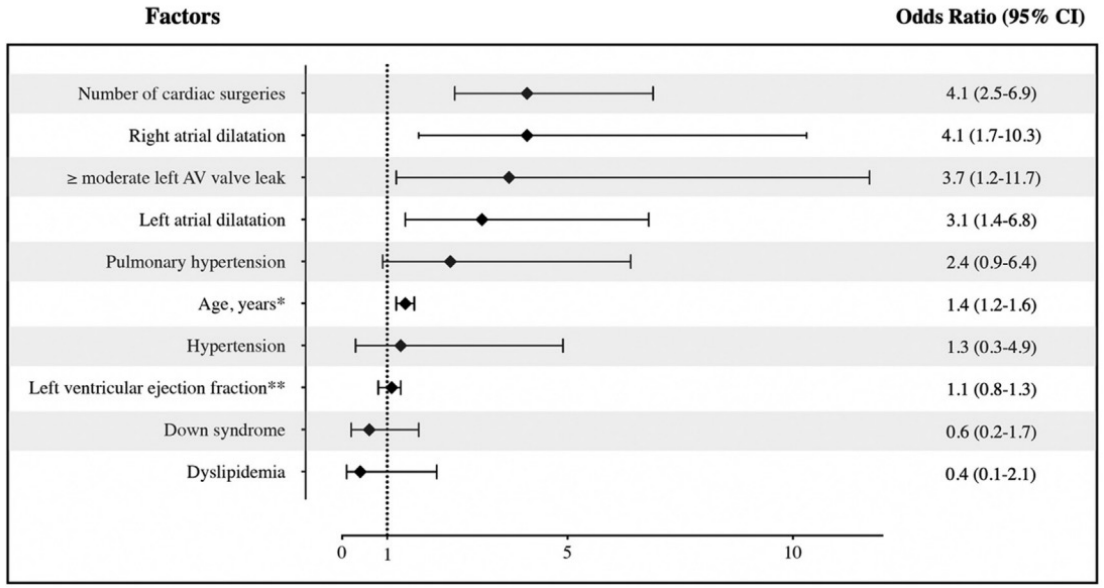



No. at risk

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Atrial arrhythmias | 391 | 390 | 327 | 203 | 115 | 63 | 26 |
| AF | 391 | 391 | 333 | 207 | 117 | 67 | 28 |
| IART/FAT | 391 | 390 | 329 | 209 | 121 | 75 | 37 |

Lifetime cumulative incidences of atrial arrhythmias, atrial fibrillation (AF), and intra-atrial re-entrant tachycardia/focal atrial tachycardia (IART/FAT)

**FIGURE 2** Forest Plot



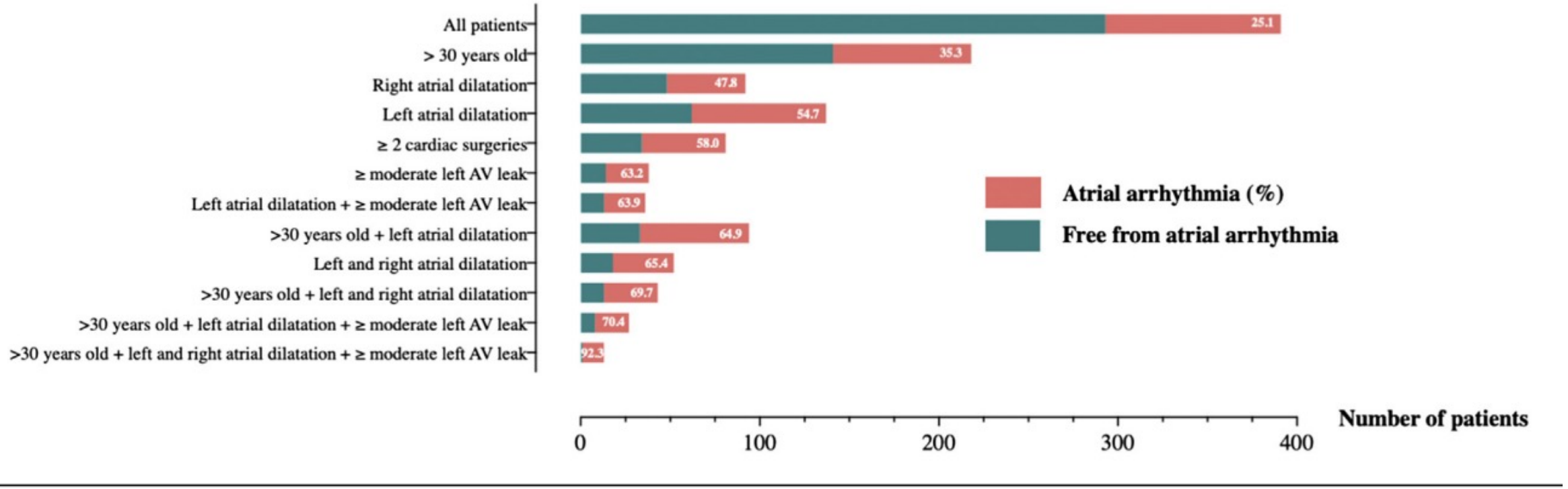

Forest plot of factors associated with atrial arrhythmias in multivariate analysis. *By 5-year increments. **By 5% increment. AV = atrioventricular; CI = confidence interval.

Jacquemart et al. J Am Coll Cardiol EP 2022

# Supraventricular arrhythmia and mechanical/functional data

391 patients (61.6% women)
mean age of 36.3 ± 16.3 years

**FIGURE 3** Prevalence of Atrial Arrhythmia




Prevalence of atrial arrhythmia in different subgroups of patients. Abbreviation as in Figure 2.

Jacquemart et al. J Am Coll Cardiol EP 2022

# Ventricular arrhythmia

c AVSD: N = 238
pAVSD: N =177

mean follow-up duration of 9 years
(range: <30 days - 47 years)

**Table 2**
Early and late post-operative arrhythmias.

| | | cAVSD (n = 23) | | pAVSD (*n* = 25) | |
|---|---|---|---|---|---|
| | | DS (*n* = 18) | NS (*n* = 5) | DS (n = 3) | NS (*n* = 22) |
| Overall post-operative arrhythmias | SVT (*n* = 48) | 18 | 5 | 3 | 22 |
| | VT/VF (*n* = 6) < 1,0% | 2 | 1 | 0 | 3 |
| Early post-operative arrhythmias | SVT (n = 33) | 16 (88,9%) | 4 (80%) | 3 (100%) | 10 (45,5%) |
| Late post-operative arrhythmias | SVT (*n* = 15) | 2 (11,1%) | 1 (20%) | 0 (0%) | 12 (54,5%) |

cAVSD = Complete atrioventricular septal defect, DS = Down syndrome, NS = Non-syndromic, pAVSD = Partial atrioventricular septal defect, SVT = Supraventricular tachycardia, VF = Ventricular fibrillation, VT = Ventricular tachycardia.

Karbanda et al. Int J Cardiol 2018

# Sinus node dysfunction and atrio-ventricular block

**Table 1** Characteristics of patients with partial and complete AVSD undergoing PM implantation

| | Partial AVSD ($n=17$) | Complete AVSD ($n=21$) | $p$ |
|---|---|---|---|
| Male gender | 10 (59) | 9 (43) | 0.32[a] |
| Age at first surgery (years) | 4.1 ± 7.9 | 2.7 ± 3.9 | 0.5[b] |
| Down syndrome | 7 (41) | 13 (62) | 0.2[a] |
| Age at PM implantation (years) | 7.0 ± 8.6 | 8.9 ± 8.3 | 0.5[b] |
| Early AVB | 7 (41) | 7 (33) 2,4% | 0.62[a] |
| Late AVB | 8 (47) | 4 (19) 1,4% | 0.06[a] |
| Late SND | 1 (6) | 9 (43) 3,1% | 0.01[a] |
| Timing of AVB onset (years) | 4.3 ± 3.5 | 6.6 ± 7.2 | 0.45[b] |
| Timing of SND onset (years) | 2 ± 0 | 10.6 ± 6 | 0.22[b] |
| Mortality | 2 (12) | 2 (9.5) | 0.55[a] |

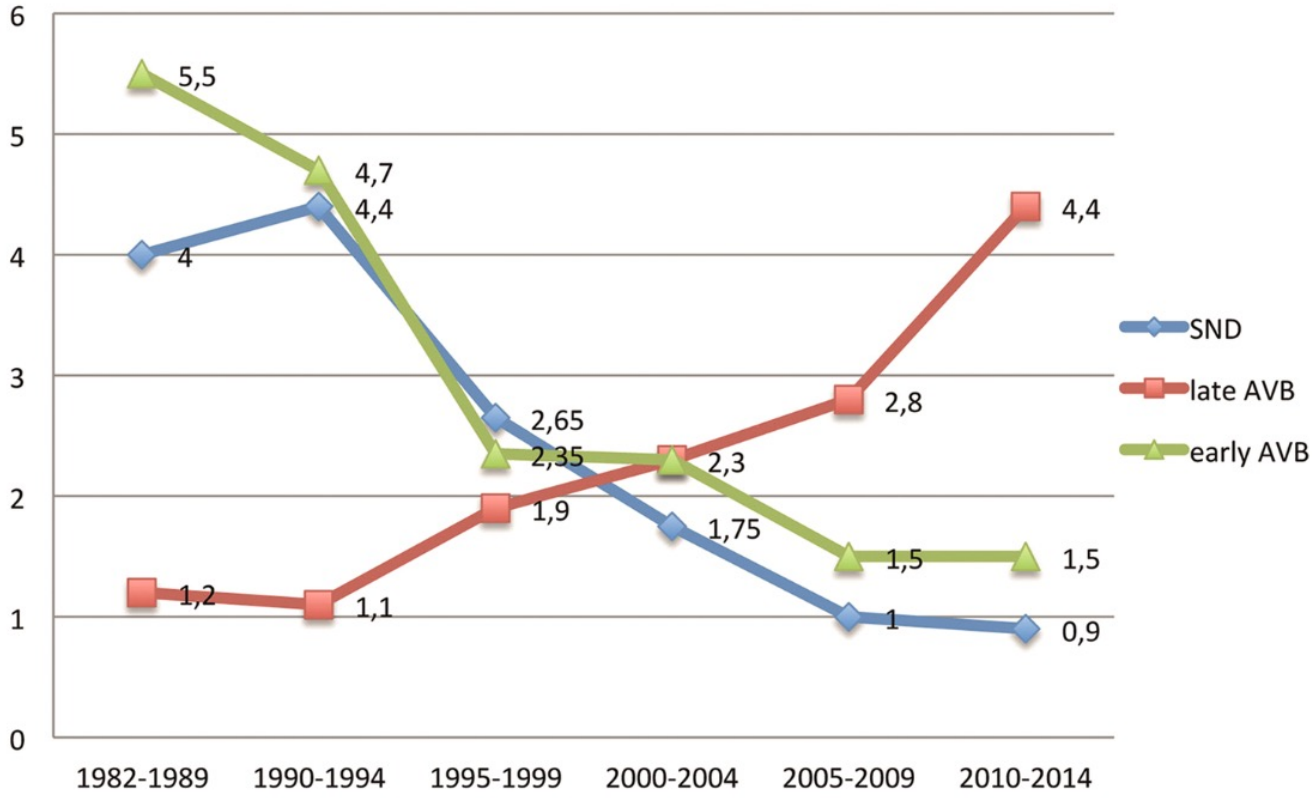
Values are expressed as number (%) or mean ± SD

*AVB* atrioventricular block, *AVSD* atrioventricular septal defect, *SND* sinus node dysfunction, *PM* pacemaker

[a]Chi-square test

[b]Independent *t* test

Changing trends




Di Mambro et al. Pediatric Cardiology 2018

# Residual pulmonary hypertension

| Variable | No. of Patients | All, $n = 88$ | Non-Residual PH after Surgical Correction, $n = 61$ (69.3%) | Residual PH after Surgical Correction, $n = 27$ (30.7%) | $p$-Value |
|---|---|---|---|---|---|
| Female sex, $n$ (%) | 88 | 44 (50.0) | 33 (54.1) | 11 (40.7) | 0.248 |
| Age at diagnosis (RHC) (years) | 88 | 0.84 (0.61–3.03) | 0.84 (0.60–3.60) | 0.84 (0.62–2.00) | 0.469 |
| Diagnosis, $n$ (%) | 88 | | | | |
| ASD | | 3 (3.4) | 3 (4.9) | 0 (0) | 0.550 |
| APVD | | 1 (1.1) | 1 (1.6) | 0 (0) | 1.000 |
| PDA | | 6 (6.8) | 5 (8.2) | 1 (3.7) | 0.662 |
| VSD | | 26 (29.5) | 17 (27.9) | 9 (33.3) | 0.605 |
| Combined | | 24 (27.2) | 12 (19.6) | 12 (44.4) | 0.016 |
| TF | | 3 (3.4) | 3 (4.9) | 0 (0) | 0.550 |
| CAVSD | | 14 (15.9) | 11 (18.0) | 3 (11.1) | 0.536 |
| TGA | | 3 (3.4) | 3 (4.9) | 0 (0) | 0.550 |
| TA | | 1 (1.1) | 1 (1.6) | 0 (0) | 1.000 |
| Other complex defects | | 7 (8.0) | 5 (8.2) | 2 (7.4) | 1.000 |

*All preop PH*

*Redo cath after 10 years*

*AVSD: 3/14 21%*

Jancauskaite et al. Medicina 2020

# Residual pulmonary hypertension

*Pulmonary arterial hypertension (shunt)*
*Postcapillary pulmonary hypertension (left heart)*

*Prevalence in AVSD population at late follow-up: ? %*

*Prevalence in Down patients: 11.7%*
*- Echocardiographic diagnosis*
*- Median follow-up time: 19 years*

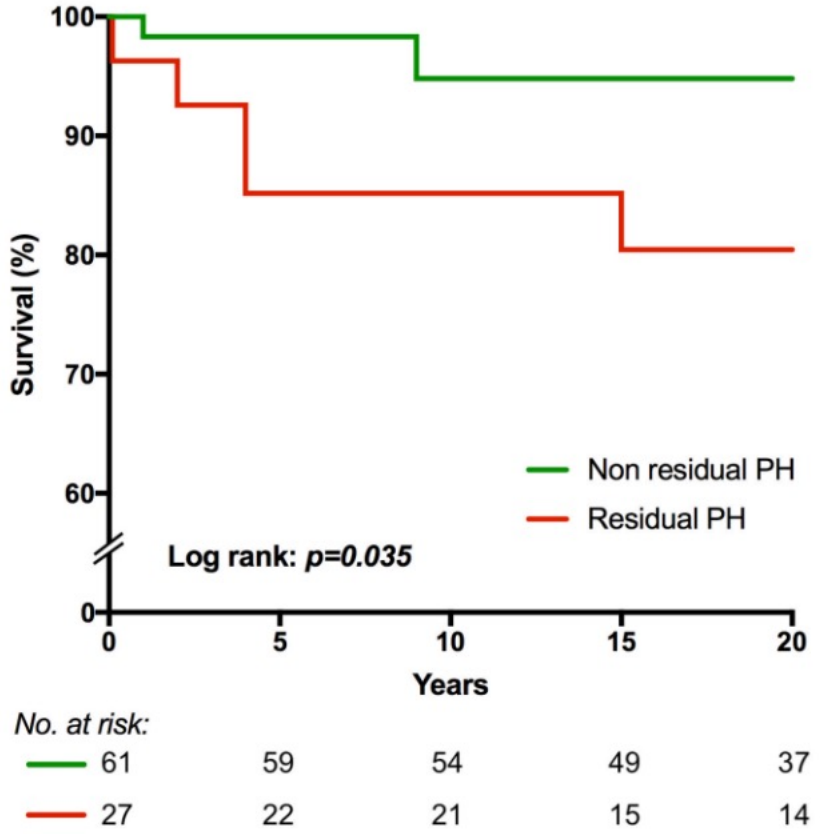



**Figure 2.** Kaplan–Meier survival analysis. PH—pulmonary hypertension.

Jancauskaite et al. Medicina 2020; Daene et al. In J Cardiol CHD 2023

# Endocarditis

Table 2.—Annualized Risk of Endocarditis Within This Population

| Risk for Endocarditis | No. of Cases per 1000 Patient-Years |
|---|---|
| High | |
| Pulmonary atresia with ventricular septal defect | 11.5 |
| Tetralogy of Fallot with palliative systemic-to-pulmonary shunt | 8.2 |
| Aortic valve stenosis* | 7.2 |
| Pulmonary atresia* | 6.4 |
| Unoperated ventricular septal defect | 3.8 |
| Moderate to low | |
| Primum atrial septal defect with cleft mitral valve* | 1.8 |
| Coarctation of the aorta* | 1.2 |
| Complete atrioventricular septal defect* | 1.0 |
| Tetralogy of Fallot* | 0.7 |
| Dextrotransposition of the great arteries* | 0.7 |
| Ventricular septal defect*† | 0.6 |
| No documented risk | |
| Atrial septal defect* | 0 |
| Patent ductus arteriosus* | 0 |
| Pulmonic stenosis* | 0 |

*After definitive surgical repair. For pulmonary atresia, this represents establishment of right ventricle to pulmonary artery continuity.

†All cases of endocarditis occurred either with a residual ventricular septal defect or with associated aortic valve anomalies including bicuspid aortic valve and aortic insufficiency. No cases of endocarditis occurred with closed ventricular septal defect in the absence of other anomalies.

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# Endocarditis

**Table 2.** Primary CHD lesion in patients with infective endocarditis

| Diagnosis | n | % | Adult | Child | Infant |
|---|---|---|---|---|---|
| Tetralogy of Fallot* | 150 | 22.8% | 84 | 56 | 10 |
| VSD | 129 | 19.6% | 76 | 46 | 7 |
| Bicuspid aortic valve | 70 | 10.7% | 59 | 11 | 0 |
| Aortic valve disease (AS / AR) | 57 | 8.7% | 46 | 11 | 0 |
| Discordant VA connections (TGA) | 42 | 6.4% | 20 | 16 | 6 |
| Coarctation of the aorta | 33 | 5.2% | 26 | 6 | 1 |
| Mitral valve anomaly | 31 | 4.7% | 22 | 8 | 1 |
| Common arterial trunk | 31 | 4.7% | 7 | 21 | 3 |
| AVSD | 25 | 3.8% | 6 | 11 | 8 |
| ASD** | 18 | 2.7% | 8 | 5 | 5 |
| Hypoplastic left heart syndrome | 14 | 2.1% | 0 | 3 | 11 |
| Pulmonary valve anomaly | 14 | 2.1% | 13 | 0 | 1 |
| Congenitally corrected TGA | 9 | 1.4% | 7 | 2 | 0 |
| Tricuspid valve anomaly | 7 | 1.1% | 6 | 1 | 0 |
| PDA | 7 | 1.1% | 7 | 1 | 0 |
| Functionally UVH*** | 6 | 0.9% | 5 | 1 | 0 |
| Other **** | 14 | 2.1% | 7 | 5 | 2 |
| Unknown | 79 | 10.7% | 36 | 31 | 10 |

**Table 5.** Risk factors for inpatient mortality on univariate and multivariable analysis (Cox-regression analysis)

| Factor | Univariate analysis | | Multivariable analysis | |
|---|---|---|---|---|
| | HR (95% CI) | *P* | HR (95% CI) | *P* |
| **CHD diagnosis** | | | | |
| Tetralogy of Fallot | 0.58 (0.26–1.3) | .19 | | |
| VSD | 0.96 (0.45–2.1) | .92 | | |
| Bicuspid aortic valve | 0.19 (0.026–1.4) | .1 | | |
| TGA | 1.1 (0.33–3.4) | .91 | | |
| Coarctation of the aorta | 0.45 (0.061–3.2) | .42 | | |
| Mitral valve anomaly | 0.85 (0.21–3.5) | .82 | | |
| Common arterial trunk | 1.9 (0.58–6) | .3 | | |
| AVSD | 3.2 (1.4–7.7) | .0083** | 3.0 (1.2–7.6) | 0.017* |
| ASD | 0.98 (0.13–7.1) | .98 | | |
| HLHS | 3.9 (1.2–13) | .024* | 3.2 (0.81–12.6) | 0.096 |
| Pulmonary valve anomaly | 1.3 (0.17–9.2) | .81 | | |
| Congenitally corrected TGA | 2 (0.28–15) | .48 | | |
| Tricuspid valve anomaly | 2.3 (0.32–17) | .4 | | |
| Functionally UVH | 0.56 (0.24–13) | .58 | | |
| PDA | 3.6 (0.5–27) | .2 | | |

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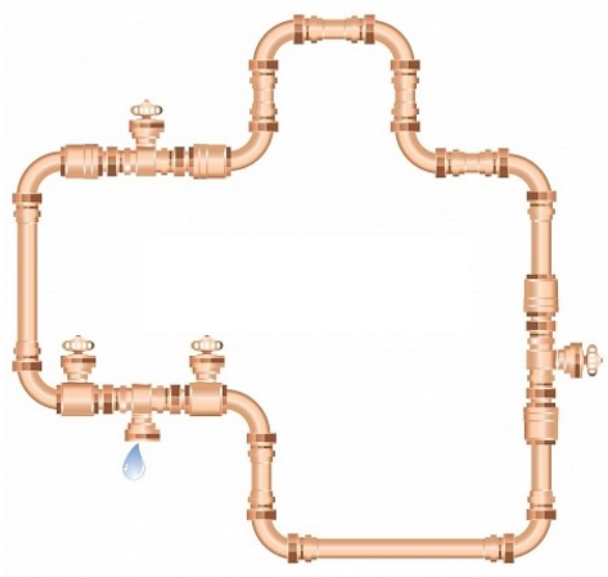
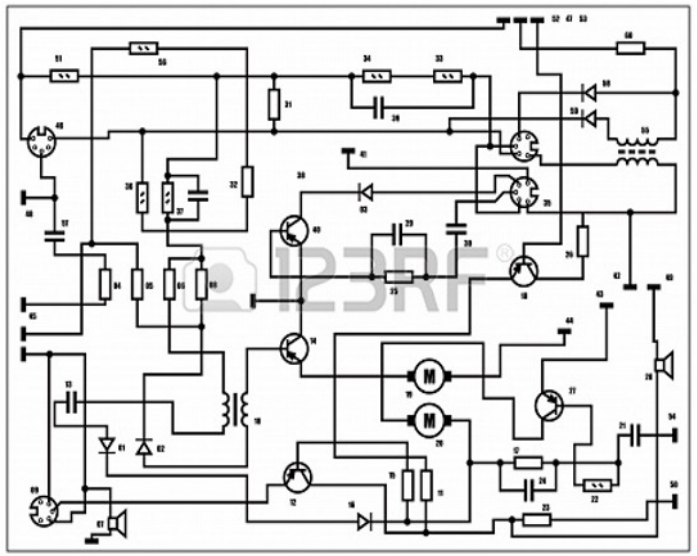
# Other issues addressed by individual approach





- Sequellae
- Residual lesions

- Heart failure
- Arrhythmias

- Contraception
- Heredity
- Pregnancy

- Sportmedical issues
- Sociolegal issues
- Vocational issues

- ...

# Conclusions

- Overall long term outcome is good after AVSD repair.
- Re-interventions mainly focus on the left-sided AV-valve.
- There is an increased prevalence of supraventricular arrhythmias, especially among older individuals.
- Long-term considerations include the need for pacemaker implantation due to atrioventricular block
- Although patients with Down syndrome may display some differences, the overall prognosis is favorable.