수술은 안 했으면 좋겠는데……

6 year old female

VSD, SA type, small

mild RCC prolapsed with trivial AR
(no progression for several years)

subarterial VSD

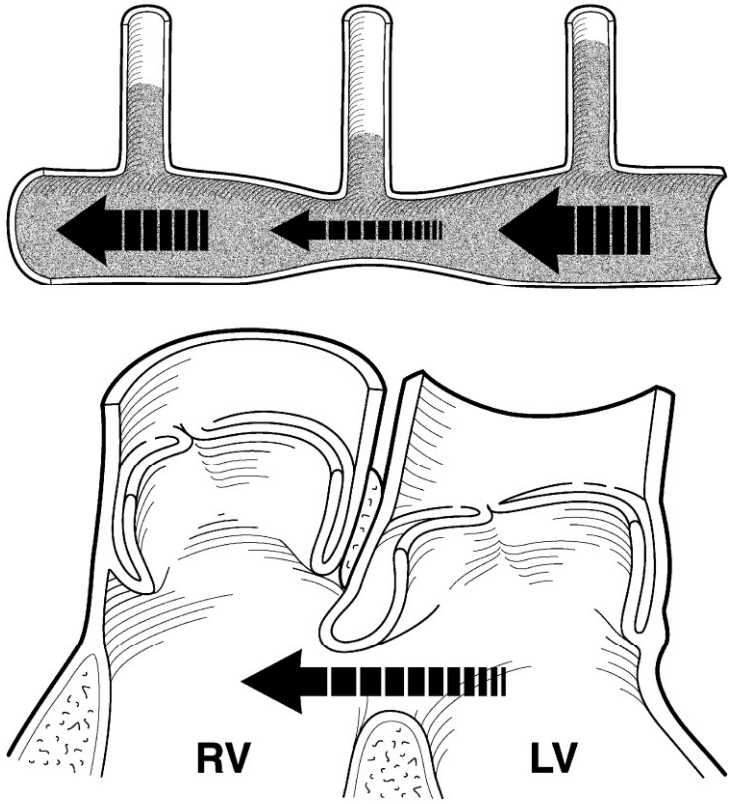
AV prolapse → AR

Aneurysm of Sinus Valsalva

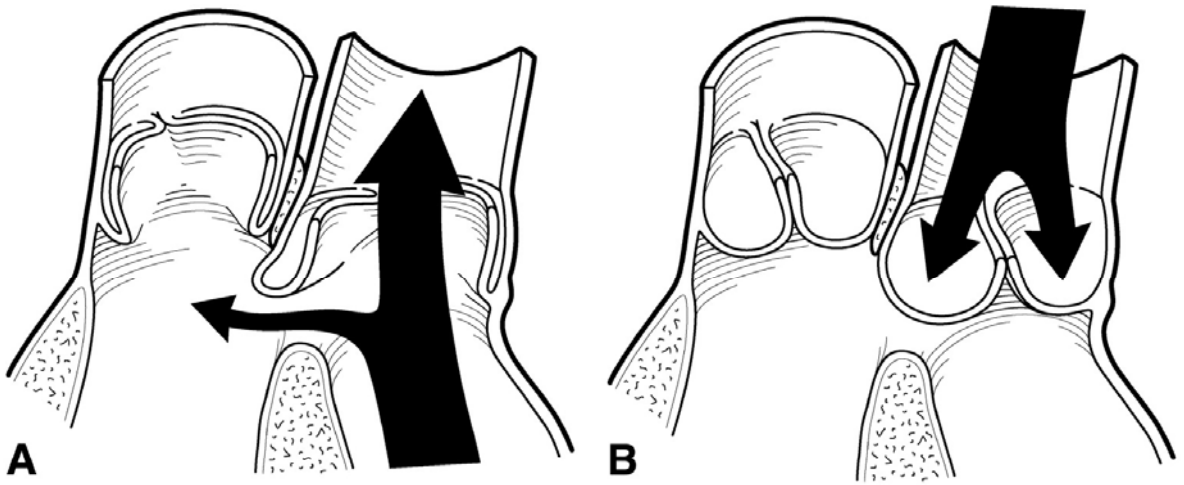
Spontaneous closure rarely occurs.

Progression of AR?

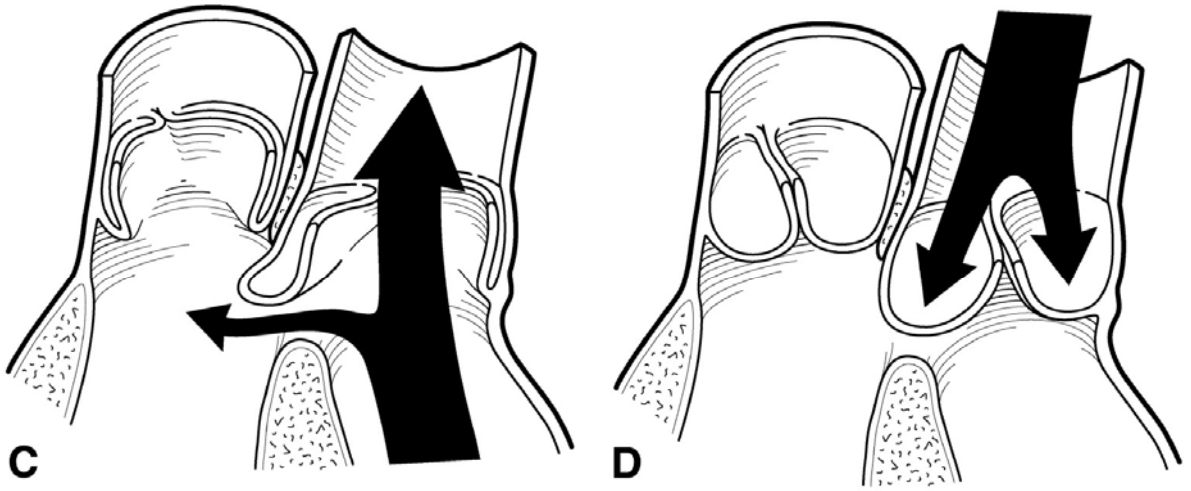
Proposed Mechanisms of AR



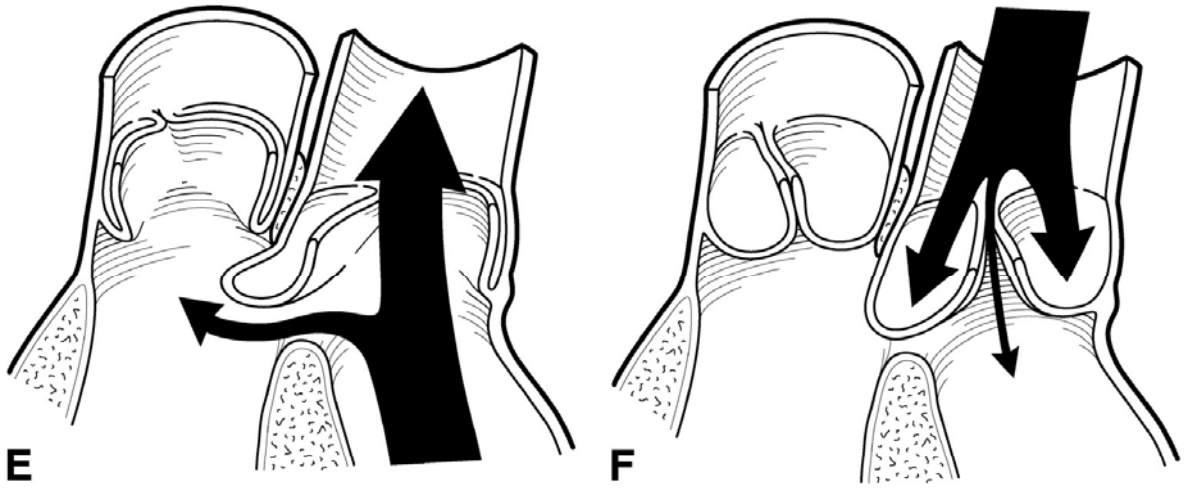
Pathophysiology of AR



Pathophysiology of AR



Pathophysiology of AR



Natural History of Subarterial Infundibular Ventricular Septal Defect

Momma K, Toyma K, Takao A, Ando M, Nakazawa M, Hirosawa K, et al.
Am Heart J 1984;108:1312

Group	Prolapse	AR	Pulm HiBP	Patients	%
1	-	-	-	76	19
2	+	-	-	77	20
3	+	+	-	95	24
4	-		+	111	28
5	Aneurysm of sinus of Valsalva			36	9
Total				345	100

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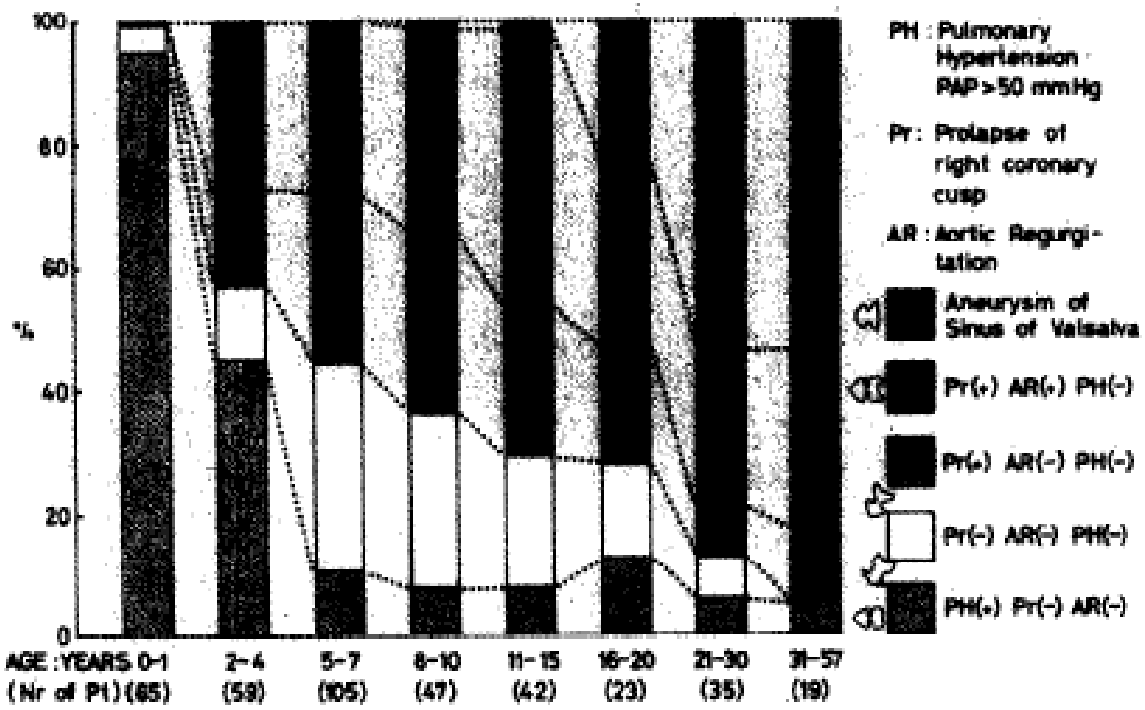

Frequency of Complications in Age Groups

Natural History of Subarterial Infundibular Ventricular Septal Defect

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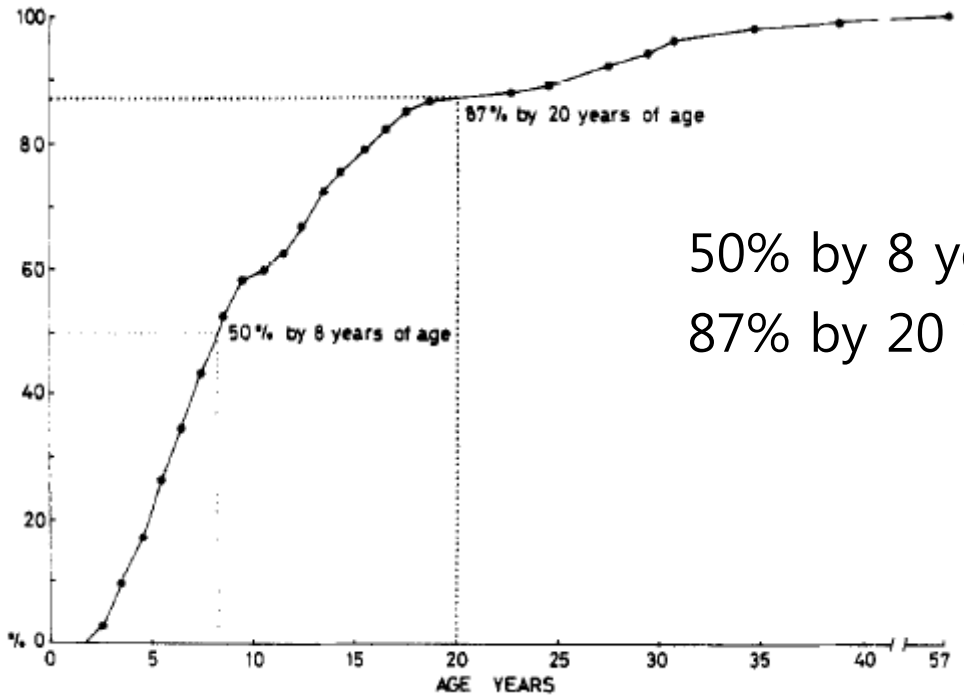


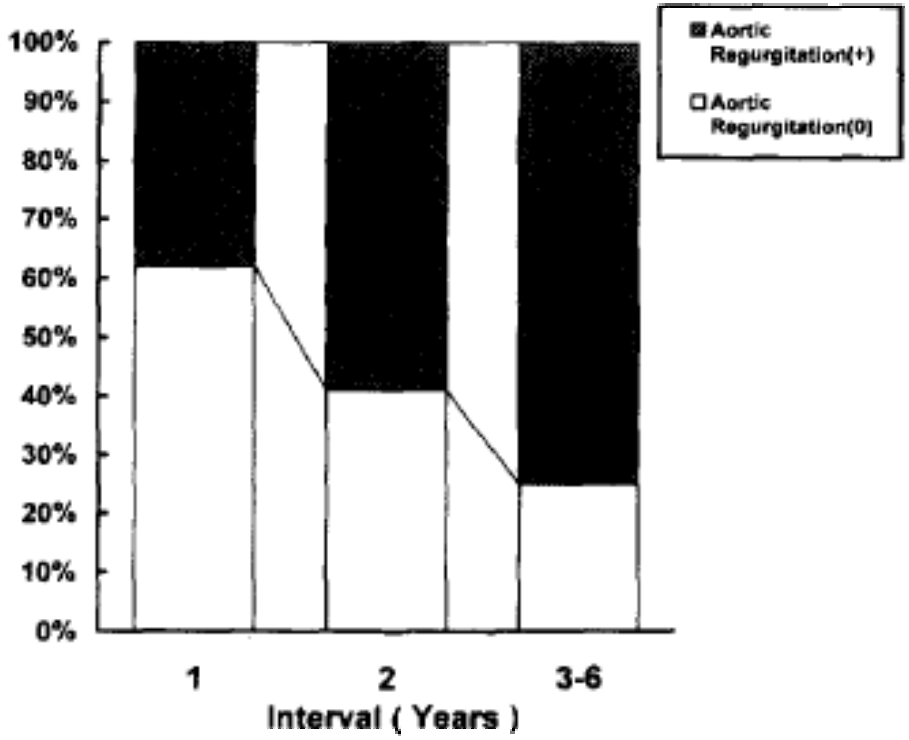
50% by 8 years of age
87% by 20 years of age

Cumulative Age Distribution of AV prolapse and AR

Aortic Valve Prolapse and Aortic Regurgitation associated with Subpulmonic VSD

Tohyama K, Satomi G, Momma K

Am J Cardiol 1997;79:1285

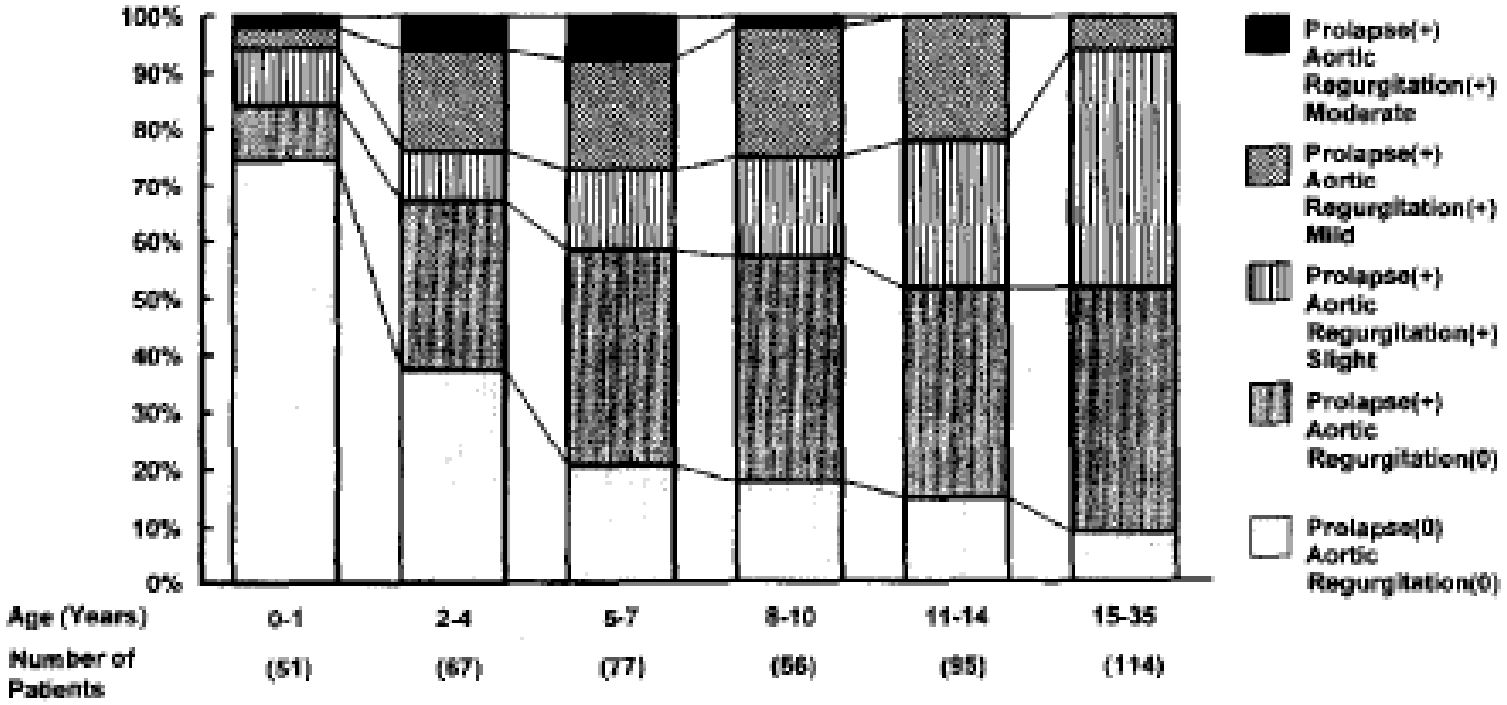


Incidence of AR after the development of AV Prolapse

Aortic Valve Prolapse and Aortic Regurgitation associated with Subpulmonic VSD

Tohyama K, Satomi G, Momma K

Am J Cardiol 1997;79:1285

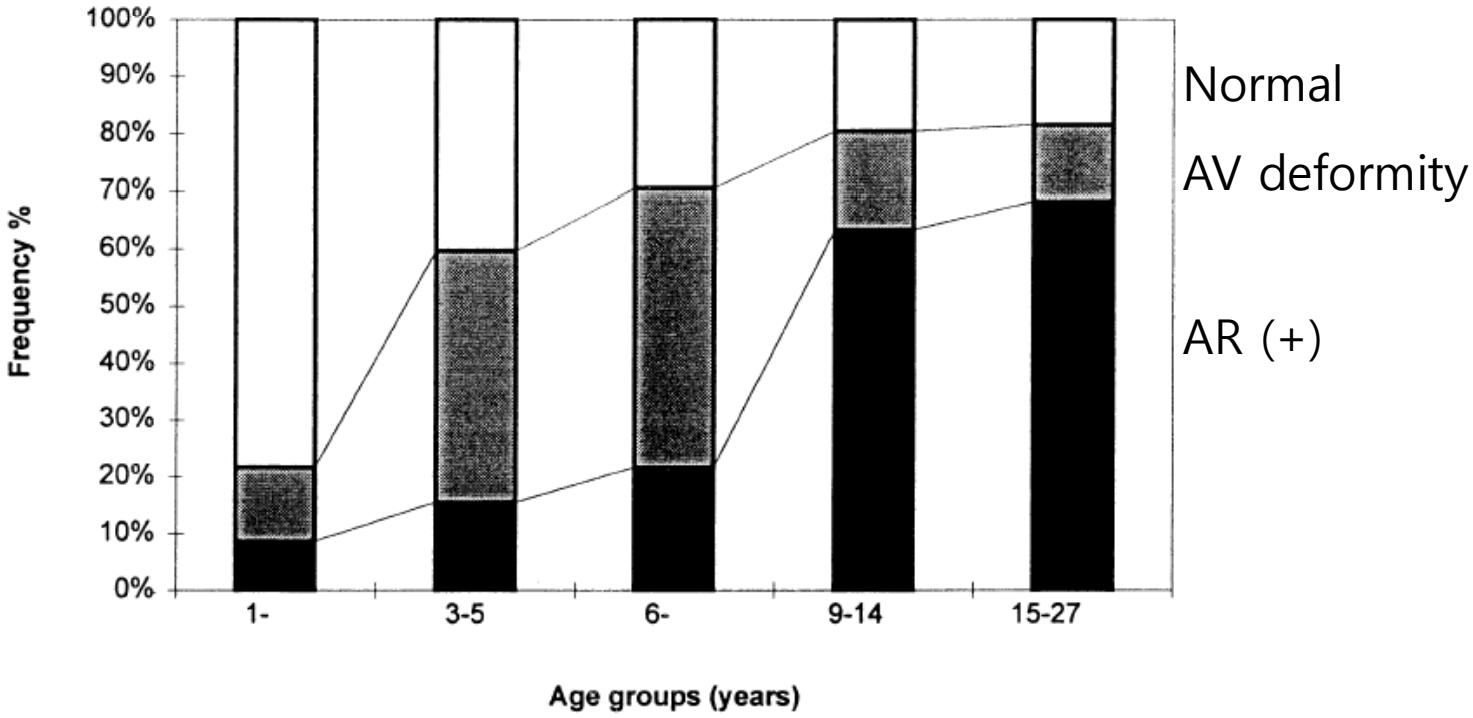


Complications in Patients with subpulmonic VSD

Influence of Surgery on Aortic Valve Prolapse and Aortic Regurgitation in Doubly Committed Subarterial VSD

Sim E, Grignani RT, Wong ML, Quek SC, Wong JCL, Yip WCL, Lee CN

Am J Cardiol 1999;84:1445

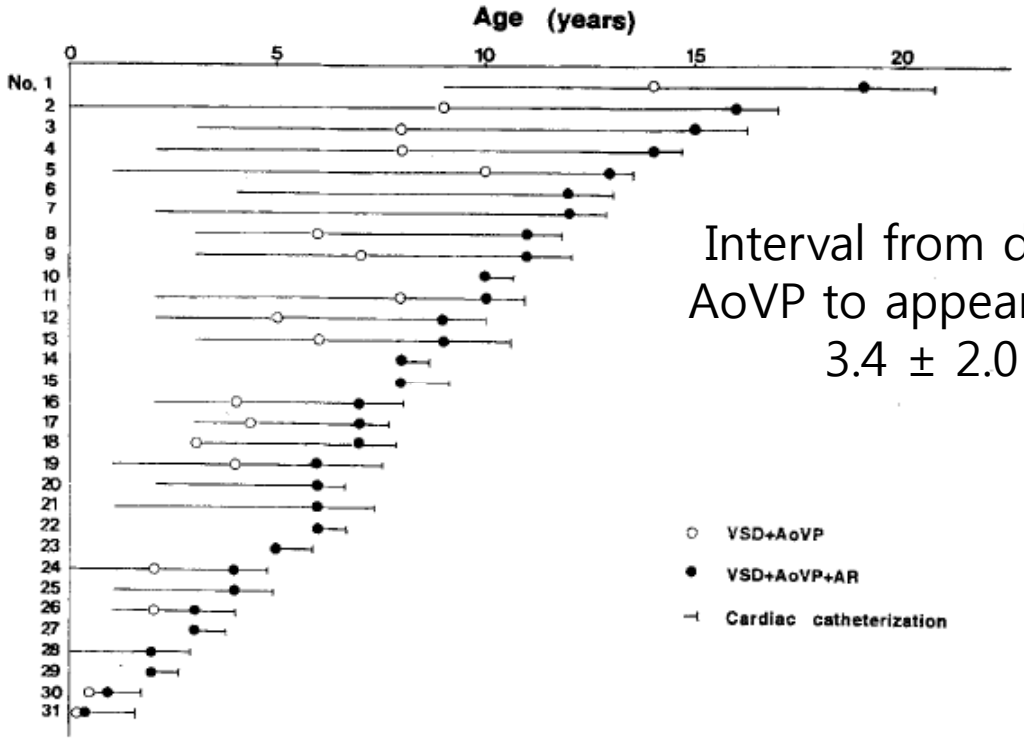


Frequency of AV Deformity and AR in 5 Age Groups

Echocardiographic Evaluation of the development of Aortic Valve Prolapse in Supracristal VSD

Mori K, Matuoka S, Tatara K, Hayabuchi Y, Nii M, Kuroda Y

Eur J Pediatr 1995;154:176



Interval from detection of AoVP to appearance of AR: 3.4 ± 2.0 years

Time course of AoVP and AR

Progression of AR?

highly probable
unavoidable

Predictor of progression?

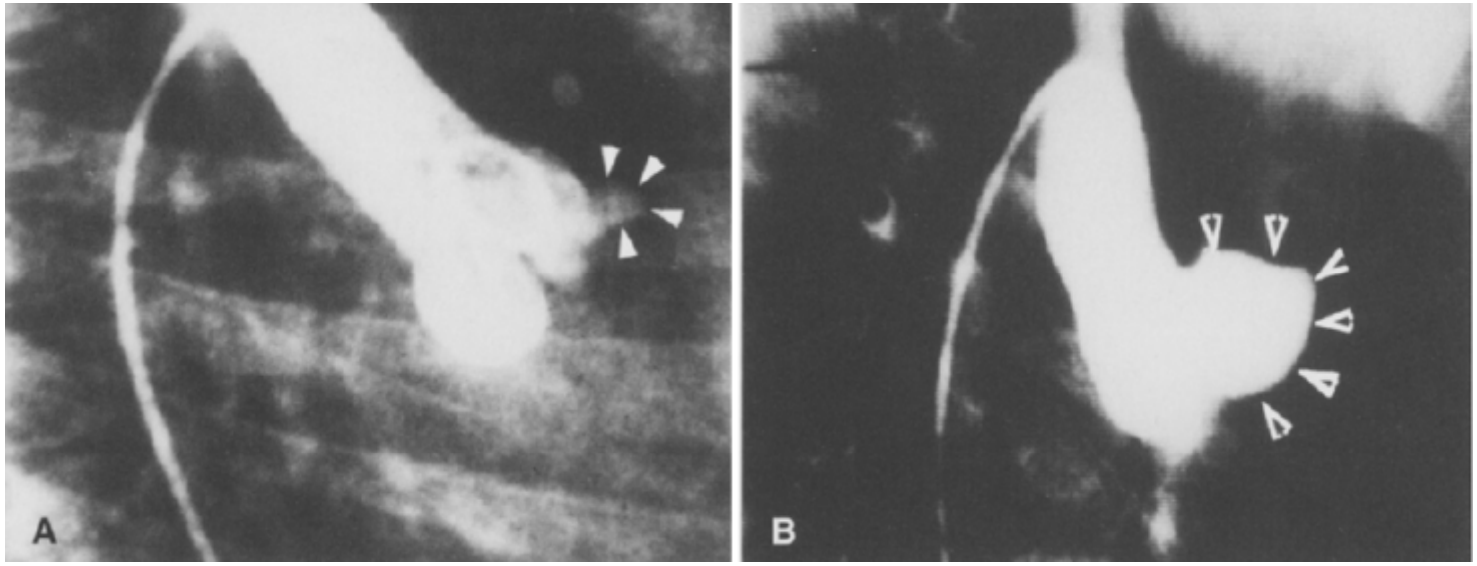
Echocardiographic Evaluation of the development of Aortic Valve Prolapse in Supracristal VSD

Mori K, Matuoka S, Tatara K, Hayabuchi Y, Nii M, Kuroda Y

Eur J Pediatr 1995;154:176

Tear drop (small) prolapse

Box type (large) prolapse



No frequency difference between AVP & AVP+AR group

Even minor AoVP can result in AR

Predictor of progression?

….

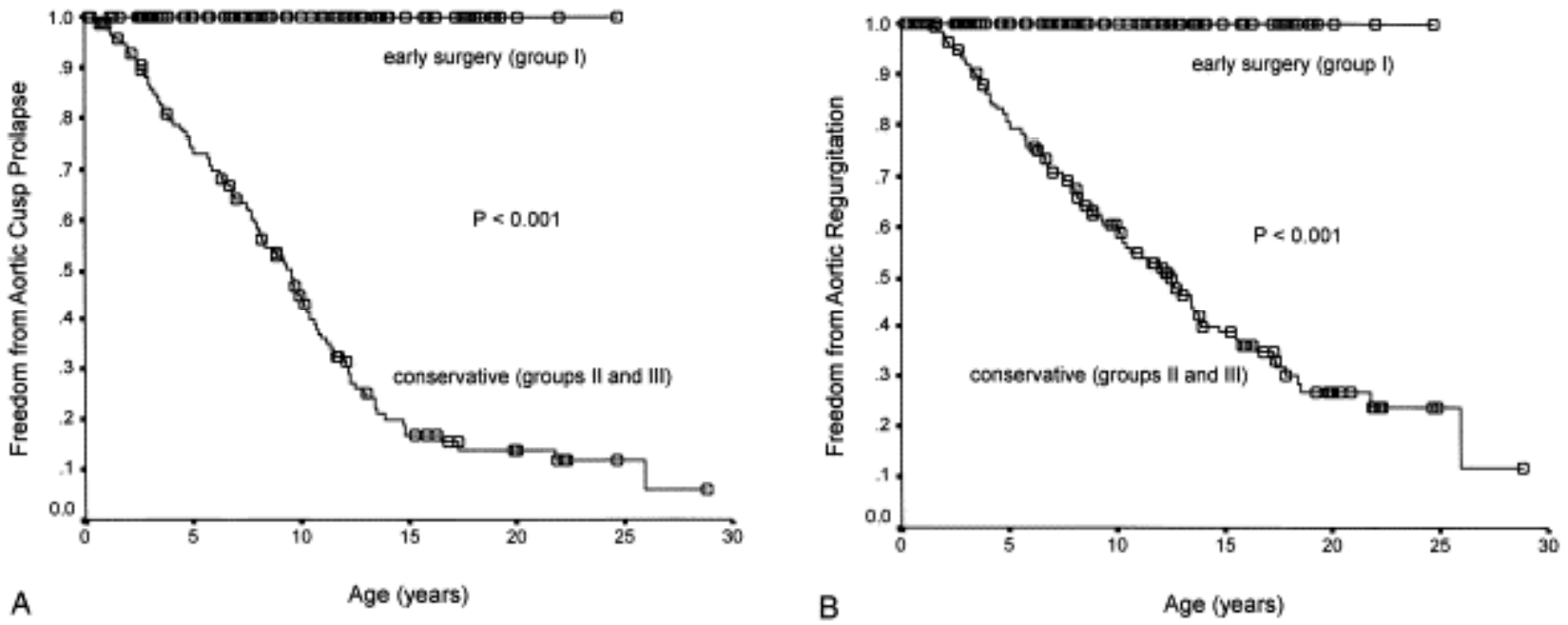
Prevention of AR

Surgical Repair

Analysis of Indications for Surgical Lcosure of Subarterial VSD without associated Aortic Cusp Prolapse and Aortic Regurgitation

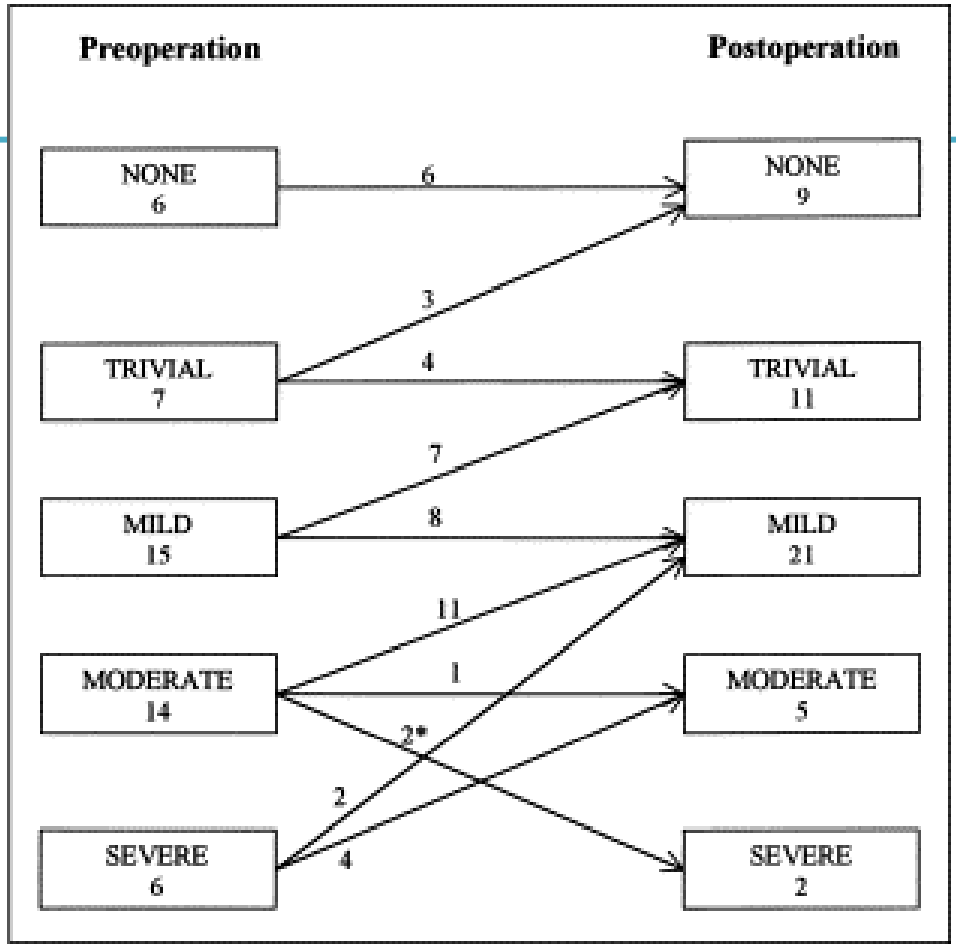
Lun K, Li H, Leung P, Chau AK et al

Am J Cardiol 1997;79:1285



Freedom from Aoritic Cusp Prolapse and AR

Analysis of Indications for Surgical Lcosure of Subarterial VSD without associated Aortic Cusp Prolapse and Aortic Regurgitation



Impact of Surgery on AR

Influence of Surgery on Aortic Valve Prolapse and Aortic Regurgitation in Doubly Committed Subarterial VSD

Sim E, Grignani RT, Wong ML, Quek SC, Wong JCL, Yip WCL, Lee CN

Am J Cardiol 1999;84:1445

Surgical Results

Aortic Valve	No	Fate of AR
AV deformity (+) AR (+)	26	4: AV repair → improved 22: remained same
AV deformity (+) AR (-)	10	5: AR remained unchanged 5: AR progress
AV deformity (-)	9	No progression of AR

Surgery should be offered once AV deformity is present, and, even before the onset of any AV deformity.

Progression of AR even after VSD repair in some patients with trivial or mild AR.

- AV prolapse progresses

  → ↓ intra-ventricular shunting

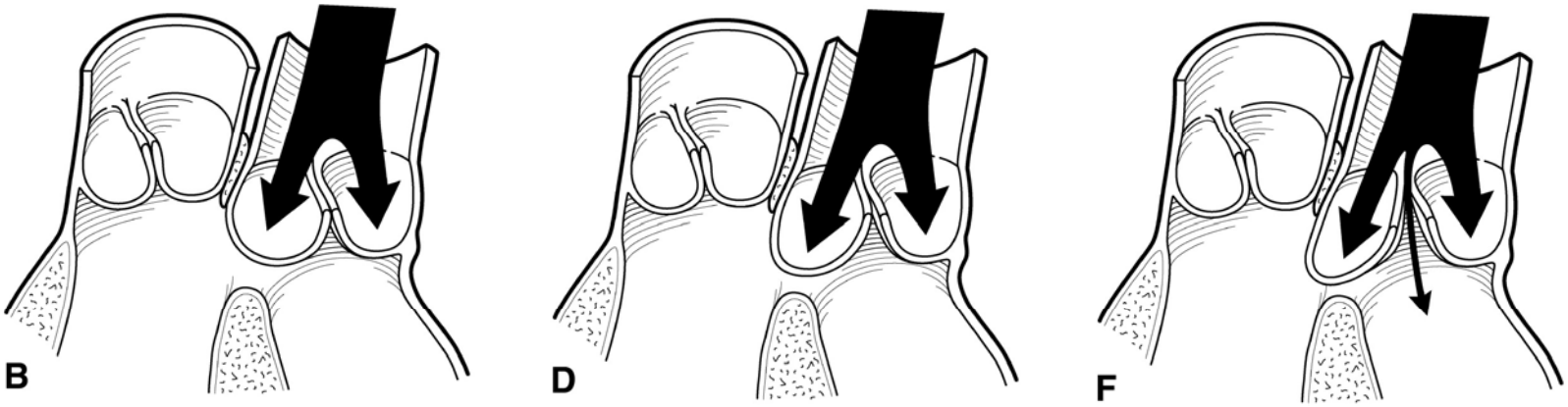
  ↑ AV distortion, AR

  → ↑ risk of endocarditis

  LV volume overload

  sinus Valsalva aneurysm

Optimal timing of Surgery?

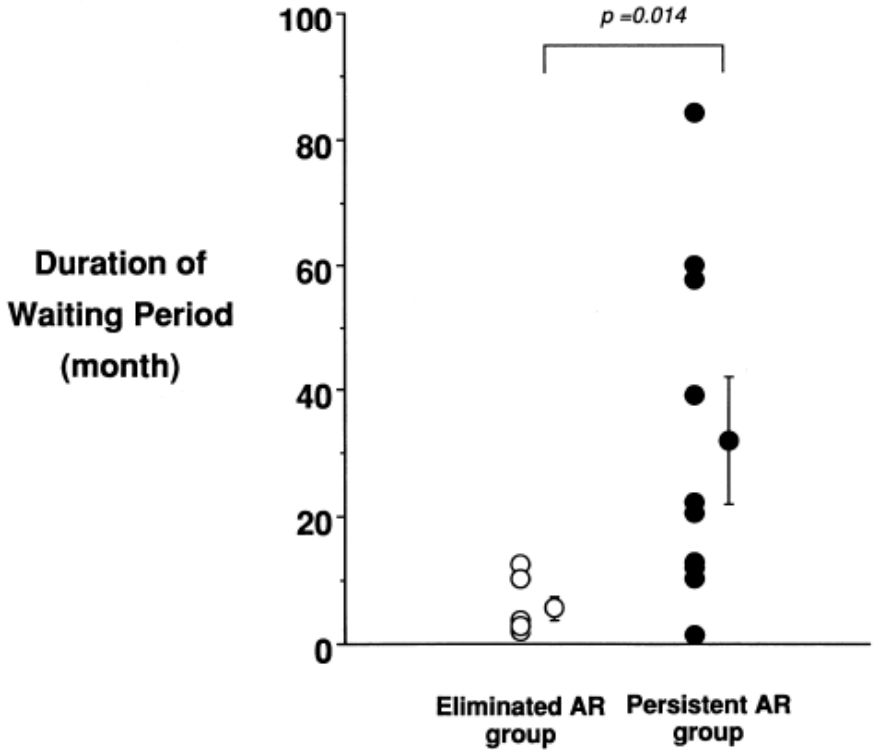


Optimal timing of Surgery?

Before progression of AV deformity

Surgical Strategy for Doubly Committted Subarterial VSD with Aortic Cusp Prolapse

Komai H, Naito Y, Fujiwara K et al.
Ann Thorac Surg 1997;64:1146



Period from the first diagnosis of AR and operation

Risk factor of Progression of AR

- Duration of waiting period (first diagnosis - Operation)

  Komai et al. Ann Thorac Surg 1997;64:1146

- Old age at VSD repair

  Chiu et al. Am Heart J 2007;153:336

Optimal timing of Surgery?

early

Natural Course of small VSD

‘small’ VSD

- Definition:
  - L- R shunt < 50%
  - Normal PA pressure
  - Absence of symptoms do not require surgical repair
- Clinical outcome: good
- Treatment: prophylaxis for endocarditis

Complications of ‘small’ VSDs

- Endocarditis
- Congestive heart failure
- Significant AR
- Occurrence of arrhythmia
- Sudden death

Small ventricular septal defects in adults

U. Neumayer, S. Stone and J. Somerville

The Jane Somerville Grown-Up Congenital Heart Unit, Royal Brompton Hospital, London, U.K.

Aims To establish the frequency of complications in adults with small ventricular septal defects, which have not undergone surgery.

Methods and Results One hundred and eighty-eight adults aged 17–72 (mean, 29·2) years with a small ventricular septal defect were studied. They were referred to a national cardiac centre (National Heart Hospital) and specialized grown-up congenital heart unit. One hundred and thirty-eight were examined in 1994–95. Fifty patients (26·6%) had additional cardiovascular lesions, most commonly a bicuspid aortic valve and/or coarctation. Spontaneous closure occurred in 19 (10%) between the age of 17 and 45 (mean, 27) years. Twenty-one (11·2%) had infective endocarditis. Aortic regurgitation developed in 37 (19·7%) patients; it was severe in nine. Atrial arrhythmias (supraventricular tachycardia or atrial fibrillation) occurred in 12 patients. In four patients, atrial fibrillation produced severe right-sided congestion with a left ventricular to right atrial shunt and haemodynamic features suggesting 'restrictive cardiomyopathy'. Four patients had ventricular arrhythmia. Disproportionate left ventricular enlargement on echocardiography and/or chest radiography was present in 26 (13·8%) without lesions to account for it.

Conclusions Eighty nine patients (47%) aged 17–44 (mean, 26·8) years had no complications through many years, while spontaneous closure occurred in 19 (10%) during adulthood. Forty-six (25%) had serious complications: infective endocarditis (11%), progressive aortic regurgitation (5%), age-related symptomatic arrhythmias (8·5%) and atrial fibrillation the commonest. Accepting that there may be a referral bias for those with complications, the course of a small ventricular septal defect is not necessarily benign during adult life.

(Eur Heart J 1998; 19: 1573–1583)

Key words: Small ventricular septal defects, adult congenital, complications.

Small ventricular septal defects in adults

U. Neumayer, S. Stone and J. Somerville

The Jane Somerville Grown-Up Congenital Heart Unit, Royal Brompton Hospital, London, U.K.

- 188 adults (age: 17-45, mean 29.2 years)
- 46 (25%) patients had serious complications
  - Endocarditis 11%
  - AR 37%
  - Symptomatic arrhythmia 6.9%
  - Cardiomyopathy 2.1%
  - LV abnormality 14%
- The course of a small VSD is <u>not entirely benign</u>.

Behavior of Unrepaired Perimembranous Ventricular Septal Defect in Young Adults

Soufflet V, Van de Bruaene A, Troost E, et al.

Am J Cardiol 2010;105:404

- 220 patients, Median follow-up: 6 years

- Death (1%)
- Endocarditis (4%)
- Pacemaker (0.5%), ICD (0.5%)
- VSD repair (7%), Pulmonary arterial hypertension (9%)

- Mid-term follow-up of adolescents and young adults with a small and unrepaired pmVSD was not uneventful.

Long-term follow-up in isolated ventricular septal defect considered too small to warrant operation

Otterstad JE, Erikssen J, Michelsen S, Nitter-Hauge S

J Intern Med 1990;228:305

- 70 patients
- follow-up duration: 21 (6-29) years
- Cardiac death (8.7%)
- Major complication (22%)
  - Cardiac surgery indicated (12%)
  - Significant dyspnea (22%), chest pain (19%),
  - Cardioactive drug (19%)
- The un-operated adults with a small VSD should be monitored closely since this condition is far from benign.

Infective Endocarditis

- Incidence
  - 1-15%
  - 1.5 - 21/1,000 patient year

    Rhodes et al. Am J Cardiol 1990
  - 3.7% (2.4 /1,000 patient-year)

    Corone et al. Circulation 1977
  - 15% (5.7/1,000 patient-years)

    Otterstad et al. Br Heart J 1983

Incidence of Bacterial Endocarditis in Ventricular Septal Defects

Shah P, Singh WS, Rose V, Keith JD
Circulation 1966

Estimated risk of bacterial endocarditis for 5 year-old patient up to age of 70 years: 13.6%

Risk of Late Endocarditis following Congenital Heart Repair: 30 year follow-up in 610 patients

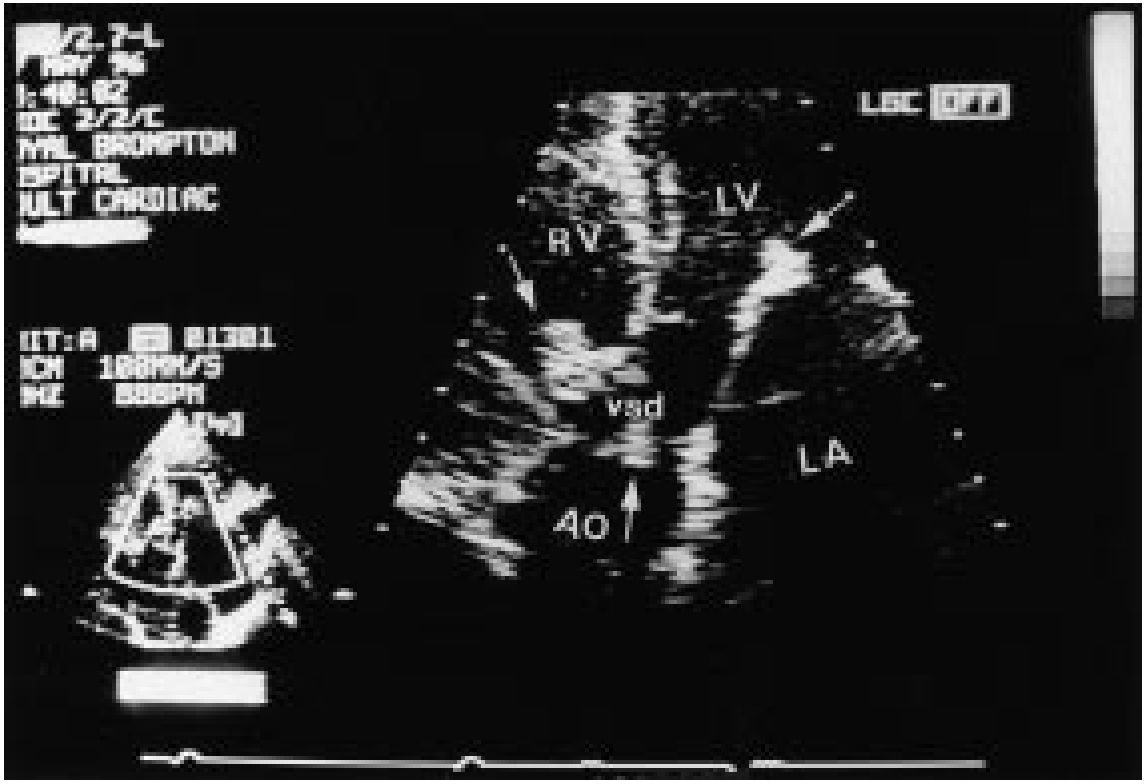
Murphy JG et al.

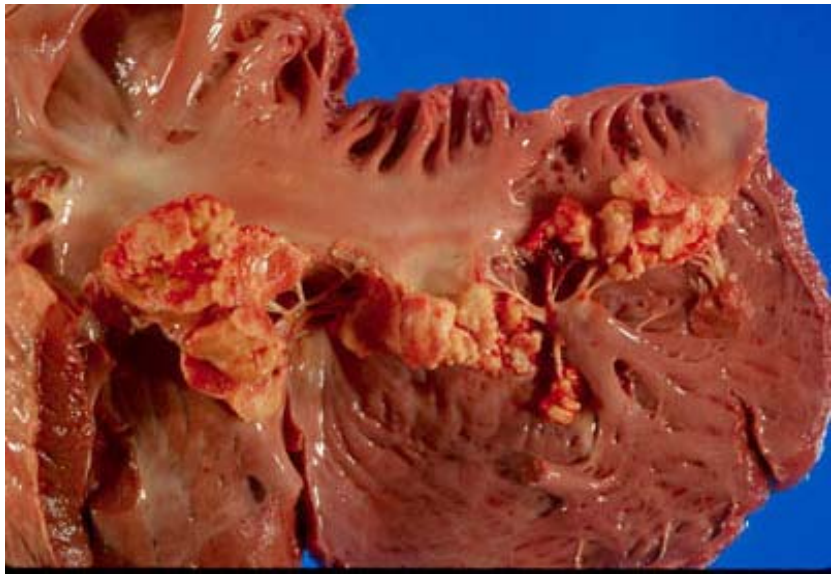
J Am Coll Cardiol 1991;17:135A

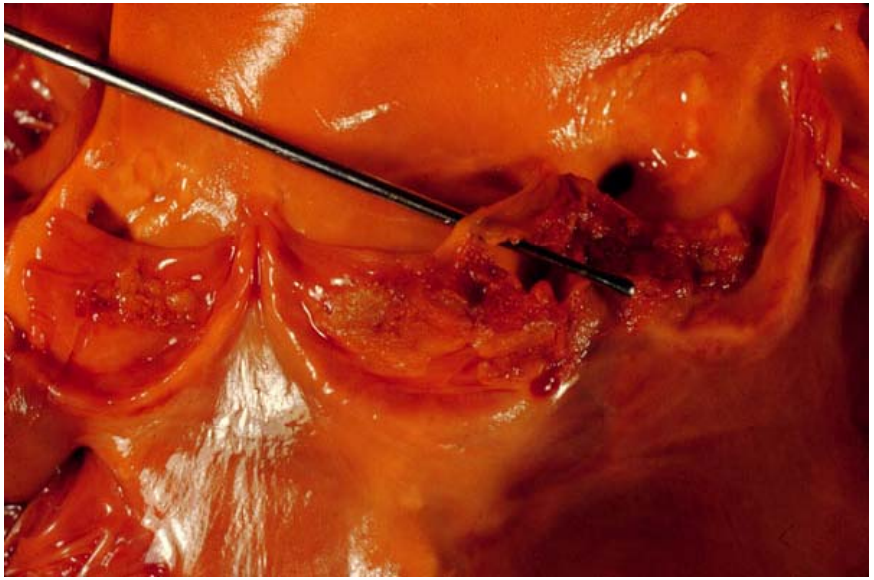
Table 2 Basic cardiac abnormalities in 185 patients (214) episodes of infective endocarditis

Lesions	Group I (episodes)	Group II (episodes)
Left ventricular outflow tract	22 (24)	20 (21)*
Ventricular septal defect	31 (37)	6 (6)*
Fallot (shunt 6, valvotomy 1)	12 (13)	11 (11)
Corrected transposition	11 (18)	2 (2)
Mitral valve prolapse	17 (18)	(1)†
Pulmonary atresia (shunt 7)	10 (13)	2 (2)
One ventricle (shunt 7, PA banding 1)	12 (15)	—
Classic transposition (shunt 2)	5 (9)	3 (3)
Atrioventricular defect	2 (2)	8 (8)
Coarctation	1 (1)	3 (3)
Common trunk	2 (2)	1 (1)
Infundibular pulmonary stenosis	2 (2)	—
Duct	1 (1)	—
Ebstein	—	1 (1)‡
Total	128 (155)	57 (59)

Infective Endocarditis







Risk of Late Endocarditis following Congenital Heart Repair: 30 year follow-up in 610 patients

Murphy JG et al.

J Am Coll Cardiol 1991;17:135A

Outcome of 185 patients with Endocarditis

Outcomes	No	%
Cured	156	84.3
Recurrent	21	11.3
Death	8	4.3
Total	185	100

Prevention of Endocarditis

Antibiotic prophylaxis

Risk of Late Endocarditis following Congenital Heart Repair: 30 year follow-up in 610 patients

Murphy JG et al.

J Am Coll Cardiol 1991;17:135A

Table 3 Pre-disposing events recorded in 87 episodes of infective endocarditis*

Pre-disposing events	Group I	Group II
Dental instrumentation	33	9
Open heart surgery	—	15
Skin infection	8	2
Cardiac catheterization	2	1 (stent)
Others†	14	3
Total	57	30

127/214 (59%) no definite predisposing event

Risk of Late Endocarditis following Congenital Heart Repair: 30 year follow-up in 610 patients

Murphy JG et al.

J Am Coll Cardiol 1991;17:135A

No occurrence of endocarditis on

- ASD secundum
- Pulmonary valve stenosis
- Aorta-pulmonary defect
- Anomalous pulmonary venous drainage
- Unoperated Ebstein
- Closed VSD or PDA

Prevention of Endocarditis

Antibiotic prophylaxis

Definite prevention: Closure of VSD

- Complete closure of a VSD appeared to abolish the risk of infective endocarditis unless aortic or mitral valves were abnormal.

Gersony WM et al.Circulation 1993;87(Suppl 1):121

Moller JH et al. Am J Cardiol 1992;70:661

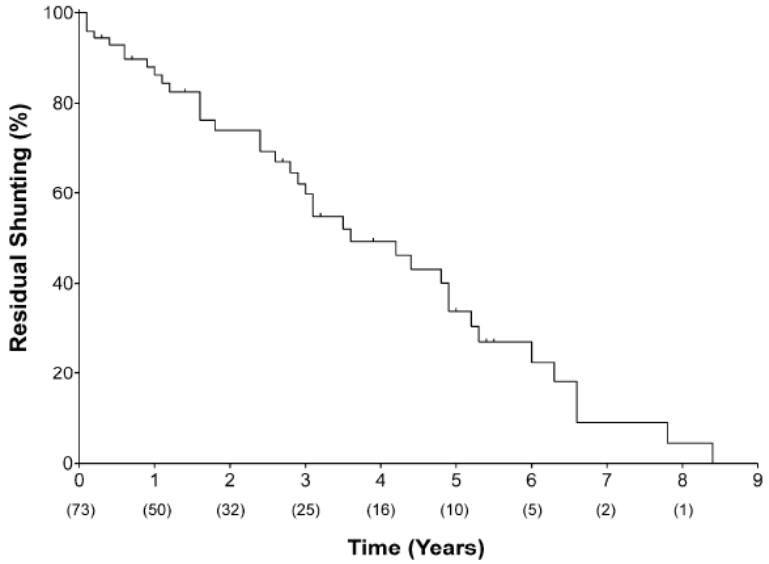
Concerns of Surgical Repair

Risk of surgery

- The operative mortality rate for repair of uncomplicated VSD < 2%
- Increased risks
  - More than one defect
  - Moderate pulmonary hypertension
  - AR

Concerns of Surgical Repair

- Residual VSD:
  - often small, hemodynamically insignificant
  - disappear spontaneously



Bol Raap et al. EJCTS 2003

Concerns of Surgical Repair

- Residual VSD:
  - often small, hemodynamically insignificant
  - disappear spontaneously
- Conduction defects such as RBBB
- Complete heart block
- Late sinus node dysfunction
- Late arrhythmia
- Scar

Socioeconomic Aspect

Socioeconomic stigma of unrepaired CHD

- Employability
- Starting a family
- Pregnancy
- Life or Health Insurance

benefit of medical observation?

혹시...

지금 수술 해 주는 것이 좋겠네