Debate in Congenital Heart disease

- 6세 여아,
  VSD SA type, small,
  mild RCC prolapsed with trivial AR
  (no progression for several years)

- Observation

Points of issue

- Natural course of SA VSD
- Post-operative course
  - In patients with < slight AR
  - In patients with ≥ moderate AR
- Risk factors for progression of aortic valve prolapse / aortic regurgitation
  - Size of defect?
  - Morphology / position of defect?

Natural course

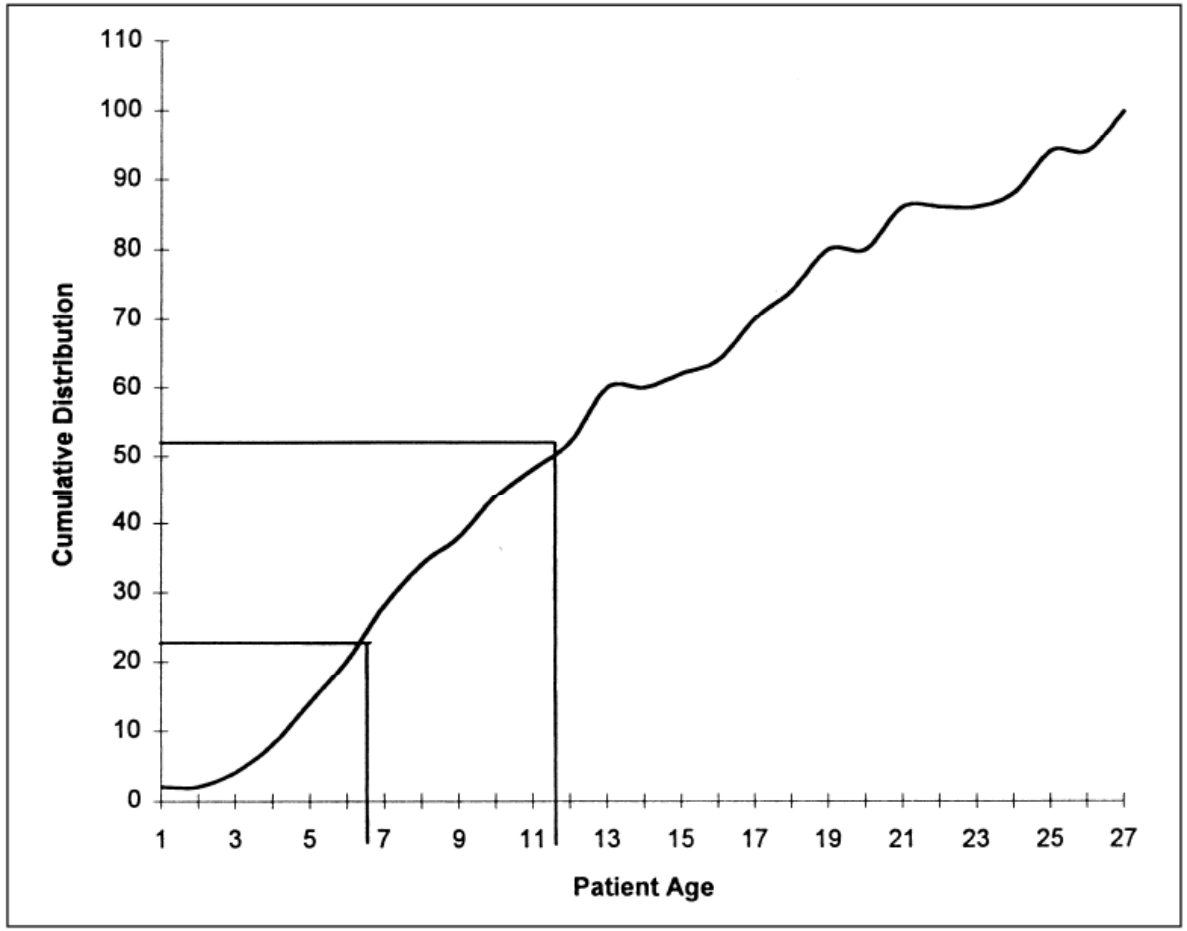

FIGURE 1. Cumulative age distribution at time of study with AR and AV deformity.

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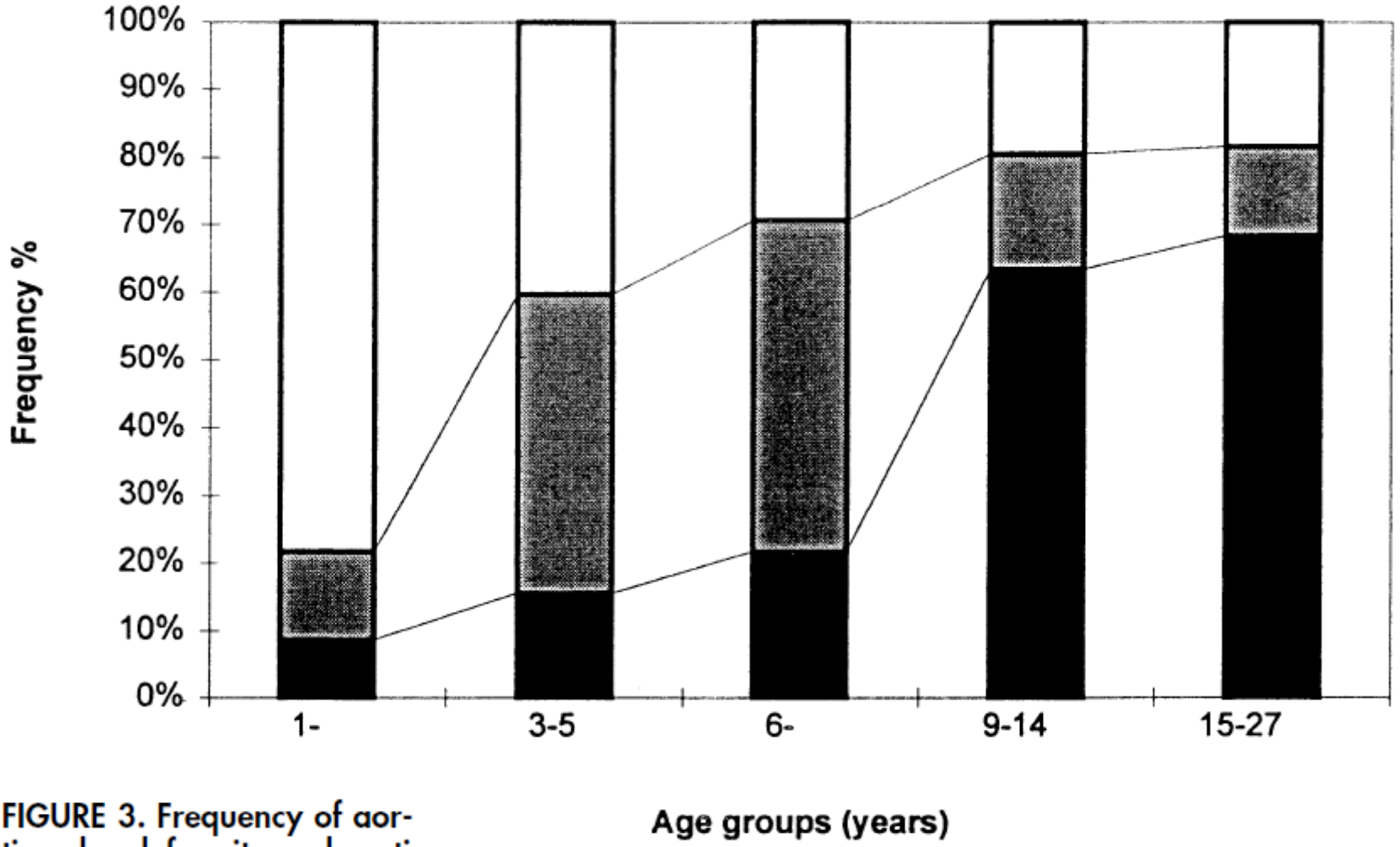

FIGURE 3. Frequency of aortic valve deformity and aortic regurgitation in 5 age groups. *White,* normal; *shaded,* AV; *black,* AV + AR.

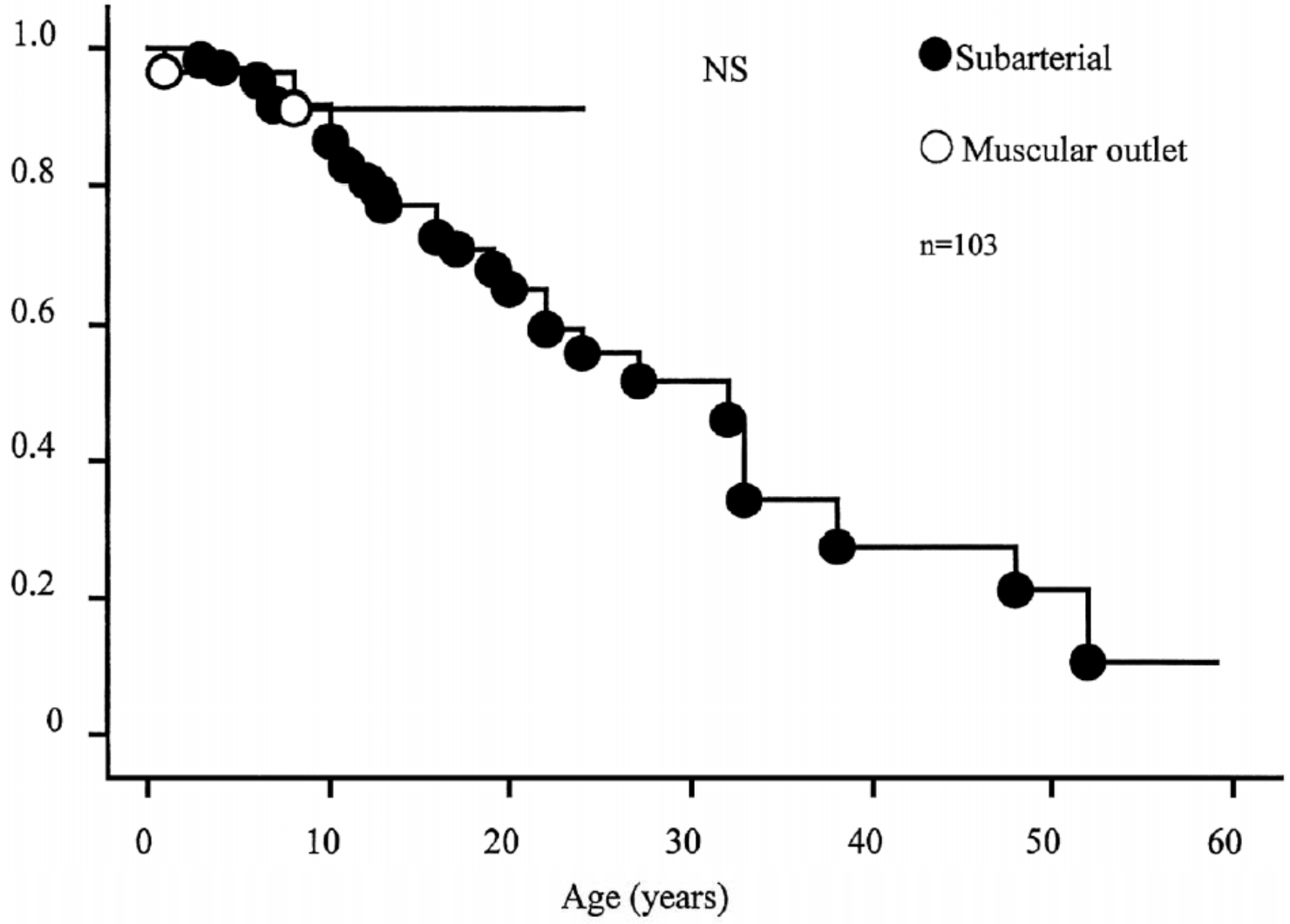

Fig 2. Freedom from aortic regurgitation ≥slight.

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Table 1 Background Data for Each Group of Ventricular Septal Defect (VSD) Patients

	I		II		III		IV		V		Total
	Subarterial	Muscular outlet	Subarterial	Muscular outlet	Subarterial	Muscular outlet	Subarterial	Muscular outlet	Subarterial	Muscular outlet	
n	64	39	48	45	7	3	48	18	33	11	
RCCD											
Range			0.14–0.29	0.11–0.29	0.19–0.29	0.22–0.27	0.30–1.00	0.31–0.46	0.30–0.89	0.32–0.95	
Mean±SD			0.24±0.04	0.22±0.05	0.24±0.04	0.25±0.03	0.39±0.11	0.37±0.05	0.47±0.15	0.47±0.17	
R/L											
Range			0.97–1.29	0.95–1.27	1.31–1.61	1.33–1.42	1.03–1.29	0.82–1.25	1.30–1.64	1.30–1.55	
Mean⊥SD			1.15⊥0.08	1.15⊥0.08	1.38⊥0.11	1.37⊥0.05	1.17±0.07	1.13±0.10	1.39±0.08	1.39±0.09	
Age at initial exam (years)											
Range	0–49	0–42	0–17	0–14	0–4	0–12	0–57	0–9	0–59	0–61	
Mean±SD	7±12	7±9	5±5	4±4	1±1	5±5	10±12	3±3	19±20*	8±18	
Age at last exam (years)											
Range	0–49	0–42	0–32	0–28	3–16	5–19	1–59	4–29	0–60	1–72	
Mean±SD	9±14	12±10	14±8	13±7	8±4	10±7	18±12	13±6	24±18*	15±19	
Age when RCCP first detected (years)											
Range			0–18	0–23	2–6	5–14	0–57	0–28	0–60	1–63	
Mean±SD			7±5	6±5	3±1	8±5	11±12	7±7	20±20*	10±18	
F/U interval after RCCP first detected (years)											
Range			0–16	0–16	0–8	0–5	0–17	0–15	0–17	0–10	
Mean±SD			7±4	6±5	4±3	2±2	7±5	5±4	3±4	3±4	
AR											
None	64	39	30	37	2	1	25	9	3*	4	214
Trivial	0	0	12	7	1	1	12	6	7	4	50
Slight	0	0	6	1	1(1)	1(1)	9(1)	3(2)	6(1)	2	29(6)
Moderate	0	0	0	0	3(3)	0	2(1)	0	7(1)*	1	23(5)
Valsalva	0	0	0	0	0	0	1	0	7	1	

RCCD, right coronary cusp deformity index; R/L, right coronary cusp imbalance index; F/U, follow-up; RCCP, right coronary cusp prolapse; AR, aortic regurgitation; Valsalva, rupture of the sinus of Valsalva aneurysm; ( ), obvious progression of AR.
**p<0.01.*

Table 2 Original Diameter of the Ventricular Septal Defect (VSD) in Each Group

	I		II		III		IV		V	
	Subarterial	Muscular outlet	Subarterial	Muscular outlet	Subarterial	Muscular outlet	Subarterial	Muscular outlet	Subarterial	Muscular outlet
Diameter <5 mm	0	2	2	7	0	1#	0	0	0	1
Diameter ≥5 mm	34	8	22	12	6	1	29	10	33	7
Actual diameter (mm)										
Range	6.0–23.0	3.3–11.0	3.9–15.9	1.5–13.0	6.0–9.6	4.3–7.0	2.0–30.0	2.0–15.0	5.5–2.50	4.6–10.0
Mean±SD	11.1±3.3*	7.3±2.5	8.1±2.9	6.2±2.9	7.7±1.3	5.7±1.9	9.6±4.8	7.8±4.0	12.8±4.8*	7.9±2.1

*#Trivial AR; *p<0.01.*

Post-operative course

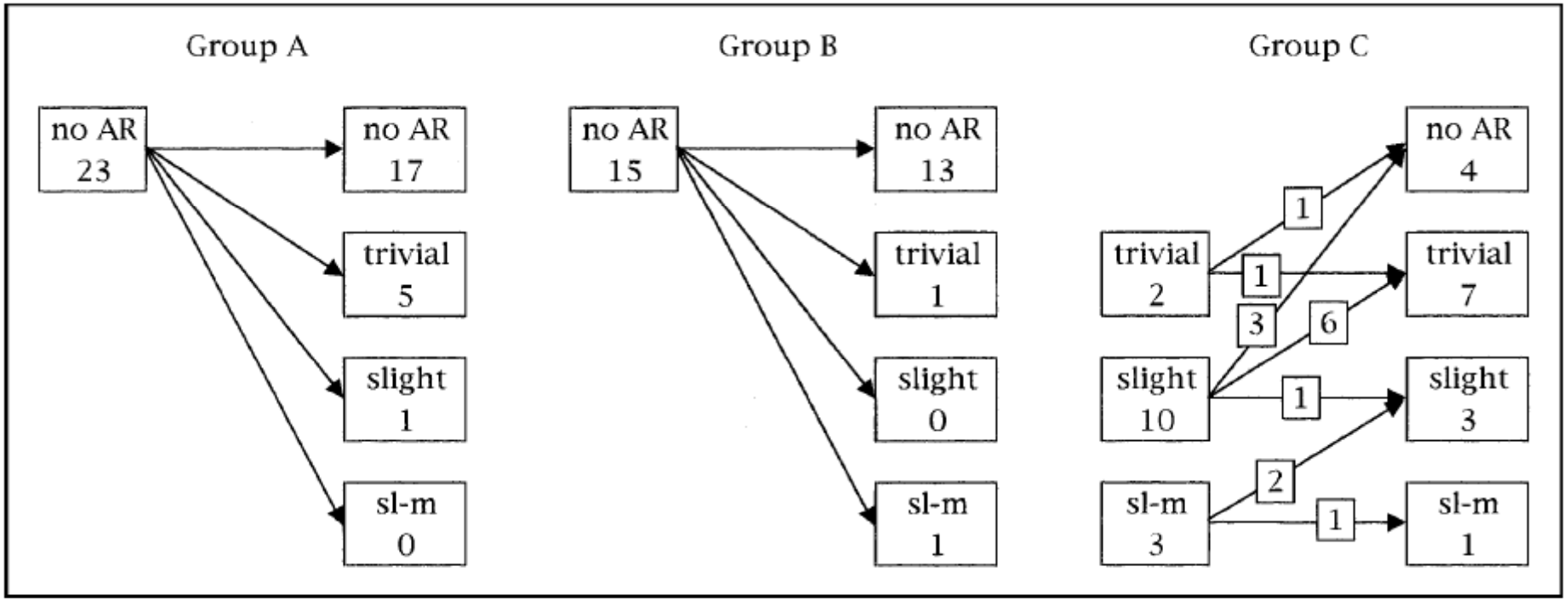

FIGURE 1. Fate of AR before and after patch closure of VSD in each group. sl-m = slight-moderate.

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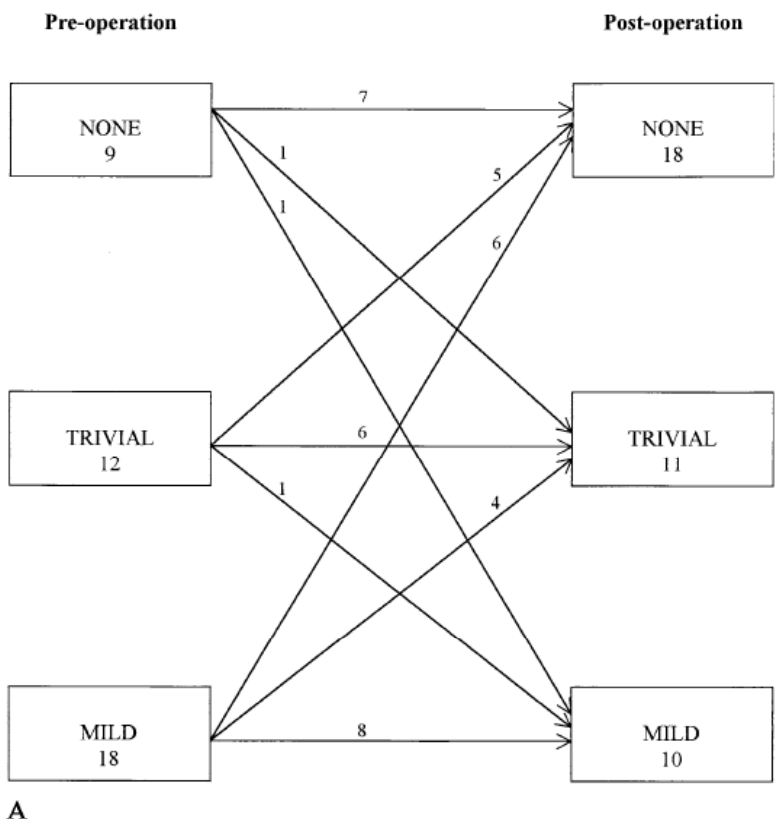

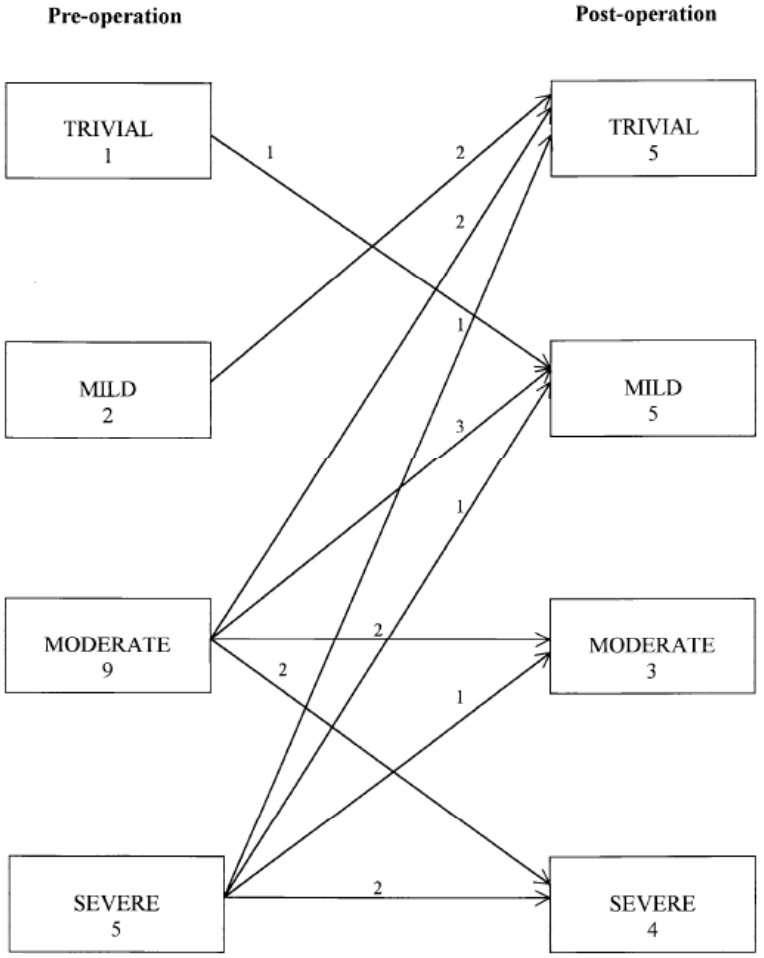

Fig 1. (A) Impact of simple closure of the ventricular septal defect on aortic regurgitation in group II patients who had mild to moderate aortic cusp prolapse. (B) Impact of aortic valvoplasty in addition to patch closure of ventricular septal defect in group III patients who had moderate to severe aortic cusp prolapse.

Cheung Y et al. Ann Thorac Surg 2002

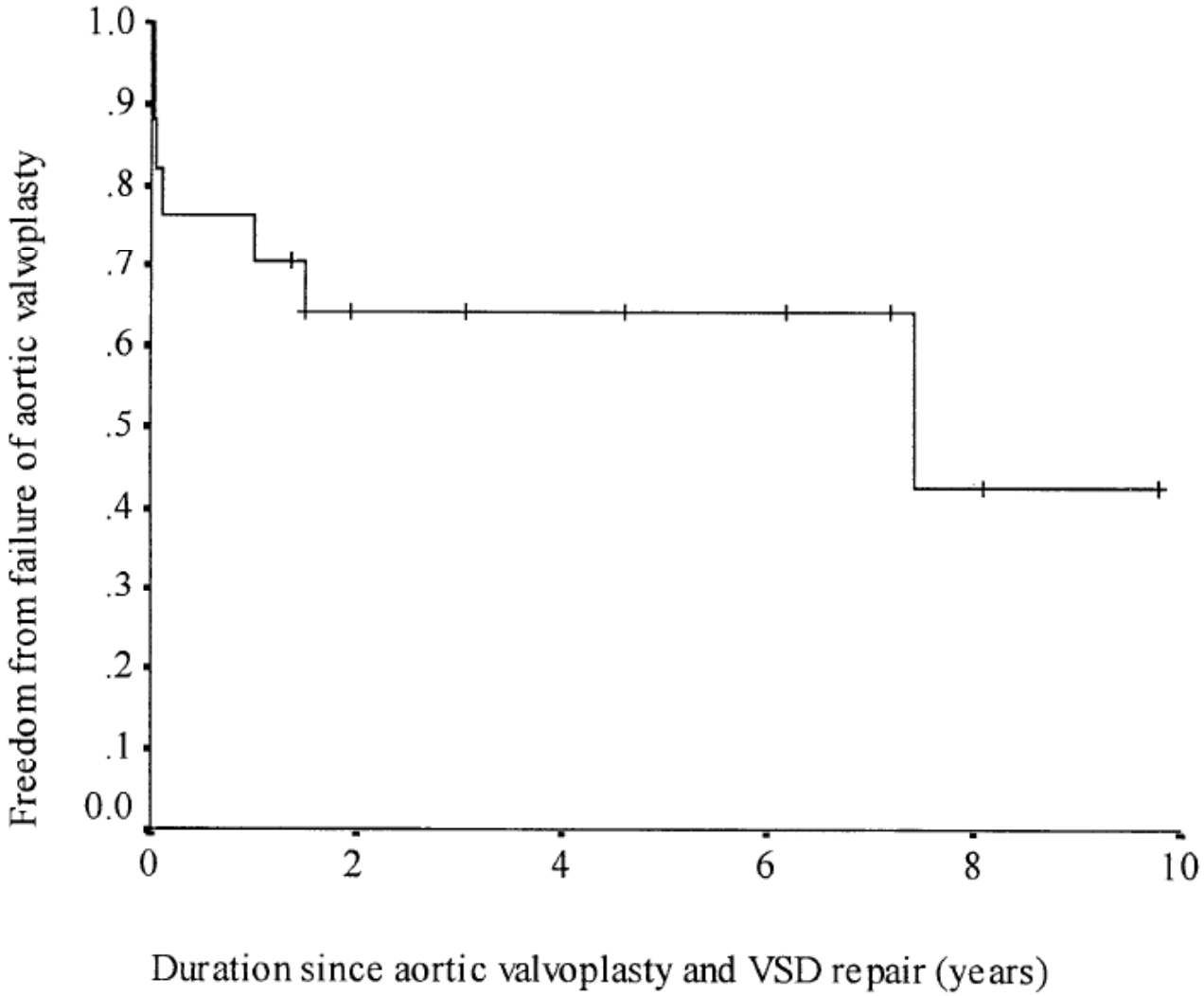

Fig 2. Kaplan-Meier actuarial survival curve showing freedom from failure of aortic valvoplasty in group III patients after operation. (VSD = *ventricular septal defect.*)

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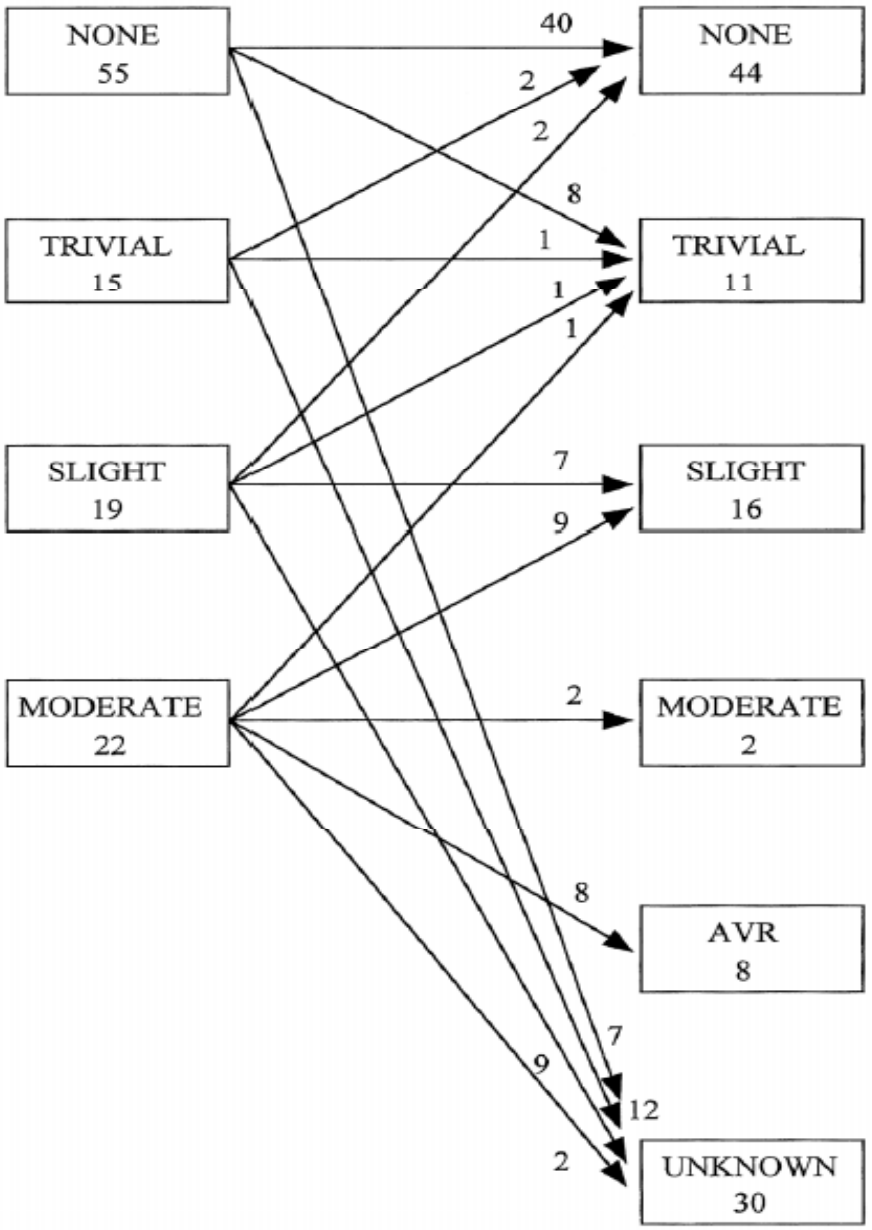

Fig 3. Impact of surgery on aortic regurgitation.

Risk factors for progression of aortic valve prolapse / aortic regurgitation

Small sized defect is risky?

Table 1. Comparison of Ventricular Septal Defect Size and Shunt Size in Patients With Doubly Committed Subarterial Defects

Group	I	II	III	Total
Qp:Qs				
Mean	1.5	1.8	2.9	1.7
Range	1.1–2.6	1.2–2.5	2.3–>4.0	1.1–>4.0
VSD size				
Mean	4.8	4.2	8.2	5.8
Range	2.1–10.2	2.0–4.7	4.3–11.0	2.0–11.0

Qp:Qs = mean pulmonary-to-systemic blood flow ratio; VSD = ventricular septal defect.

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TABLE I Ratio of Pulmonary to Systemic Blood Flow and Ventricular Septal Defect Size in Different Subgroups of Patients With Doubly Committed Subarterial Ventricular Septal Defects

Associated Complications	Aortic Valve Deformity + AR	Aortic Valve Deformity	None	Total
Pulmonary/systemic flow ratio				
Mean	1.4	1.5	1.8	1.6
Range	1.1–2.6	1.0–2.5	1.2–>4	1.0–>4
Ventricular septal defect size				
Mean	4.5	3.6	5.3	4.3
Range	1.8–10.2	2.0–6.5	2.5–12.2	1.8–12.2

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TABLE III Summary of Characteristics of the Three Groups

	Group A (n = 23)	Group B (n = 15)	Group C (n = 15)	p Value
Age at VSD closure (mo)	5–79 (23 ± 16)	31–198 (100 ± 54)	23–146 (59 ± 38)	<0.01
Follow-up interval (yrs)	5–13 (10 ± 3)	5–14 (10 ± 3)	5–14 (7 ± 3)	<0.05
Age at last follow-up (yrs)	7–18 (11 ± 3)	10–28 (18 ± 5)	16–26 (19 ± 3)	<0.01
Diameter of VSD (mm)	7–17 (10 ± 2)	7–18 (11 ± 3)	8–16 (11 ± 3)	NS

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Large sized defect is risky?

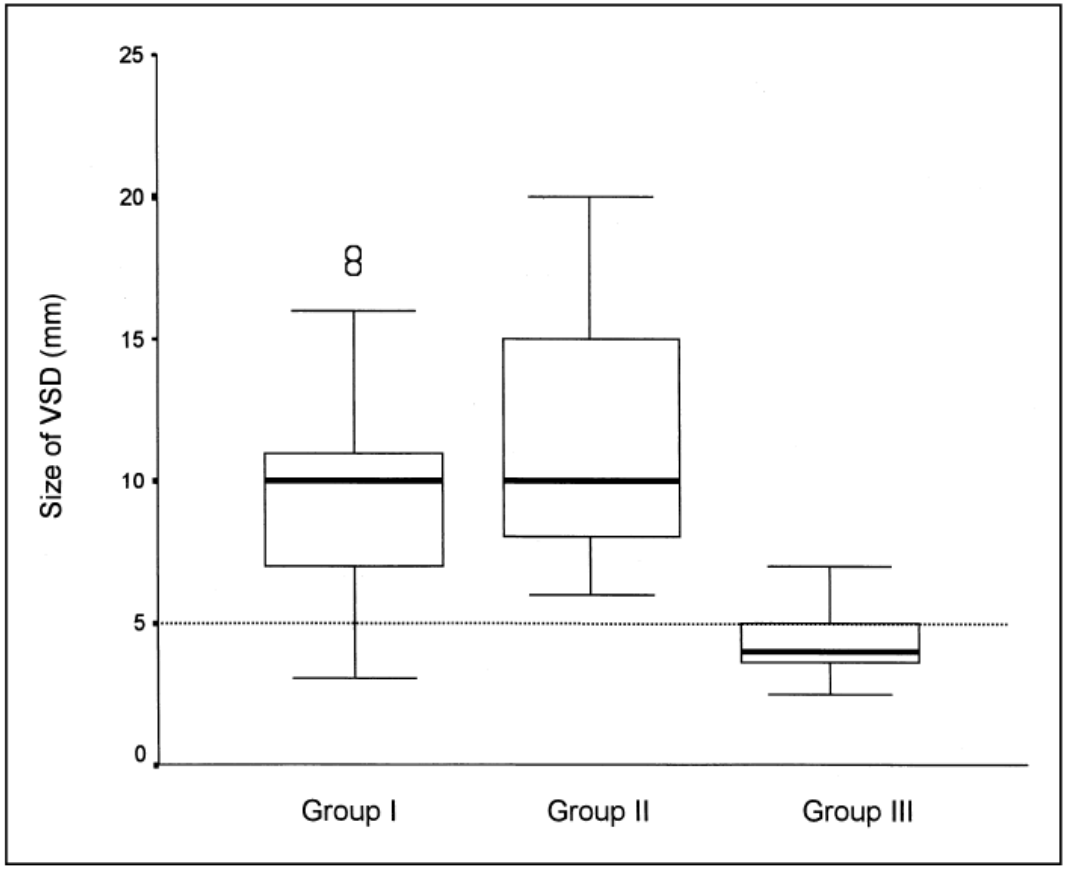

FIGURE 1. Box plot of size of VSD by patient groups. *Bold lines*, medians in each group. All patients in group II and all but 1 patient in group I had a VSD size ≥5 mm.

- Group I 75 patients. CHF (+), prolapse (-), AR (-)
  Group II 102 patients. CHF (-), prolapse (+), AR (±)
  Group III 37 patients. CHF (-), prolapse (-), AR (-)

(Lun K et al. Am J Cardiol 2001)

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Mean±SD			0.24±0.04	0.22±0.05	0.24±0.04	0.25±0.03	0.39±0.11	0.37±0.05	0.47±0.15	0.47±0.17	
R/L											
Range			0.97–1.29	0.95–1.27	1.31–1.61	1.33–1.42	1.03–1.29	0.82–1.25	1.30–1.64	1.30–1.55	
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RCCD, right coronary cusp deformity index; R/L, right coronary cusp imbalance index; F/U, follow-up; RCCP, right coronary cusp prolapse; AR, aortic regurgitation; Valsalva, rupture of the sinus of Valsalva aneurysm; ( ), obvious progression of AR.
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Diameter ≥5 mm	34	8	22	12	6	1	29	10	33	7
Actual diameter (mm)										
Range	6.0–23.0	3.3–11.0	3.9–15.9	1.5–13.0	6.0–9.6	4.3–7.0	2.0–30.0	2.0–15.0	5.5–2.50	4.6–10.0
Mean±SD	11.1±3.3*	7.3±2.5	8.1±2.9	6.2±2.9	7.7±1.3	5.7±1.9	9.6±4.8	7.8±4.0	12.8±4.8*	7.9±2.1

*#Trivial AR; *p<0.01.*

Subclassification of SA VSD

- Synonyms

  Conal, infundibular, subpulmonary, subpulmonic, supracrystal, subarterial, doubly committed, etc

① Absence of conal septum
② High muscular VSD with a muscular rim
③ Circular defect underneath part of the RCC
④ Crescentic defect underneath the RCC

(Brizard C. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2006)

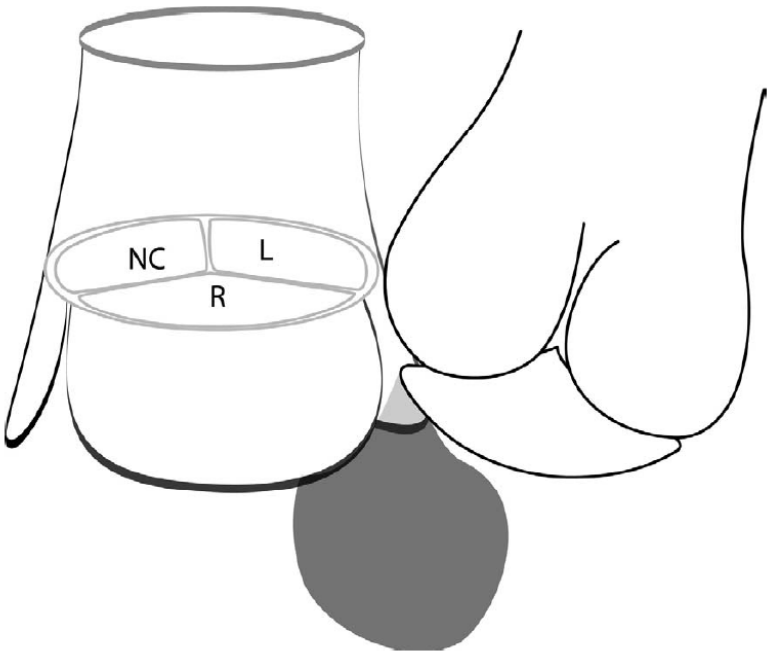

Figure 1 Schematic representation of a large defect in the conal septum with little relationship with the right coronary cusp.

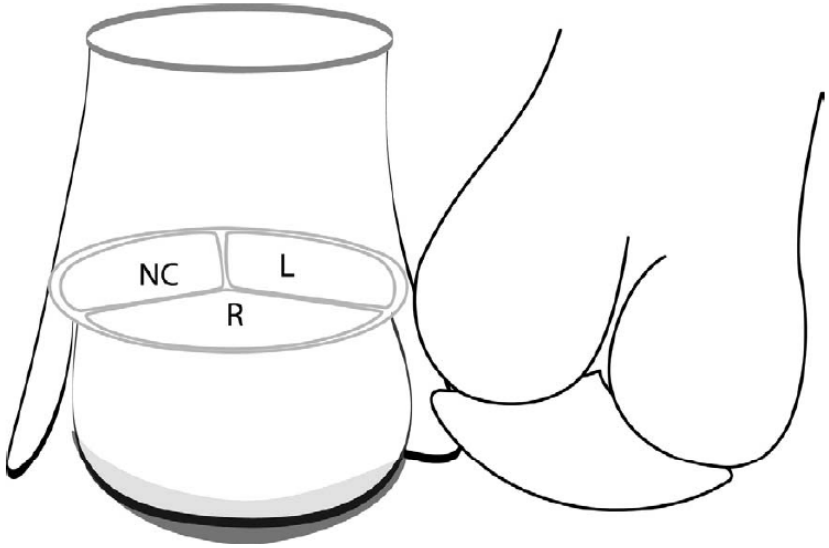

Figure 2 Schematic representation of an "infundibular VSD" as we see it.

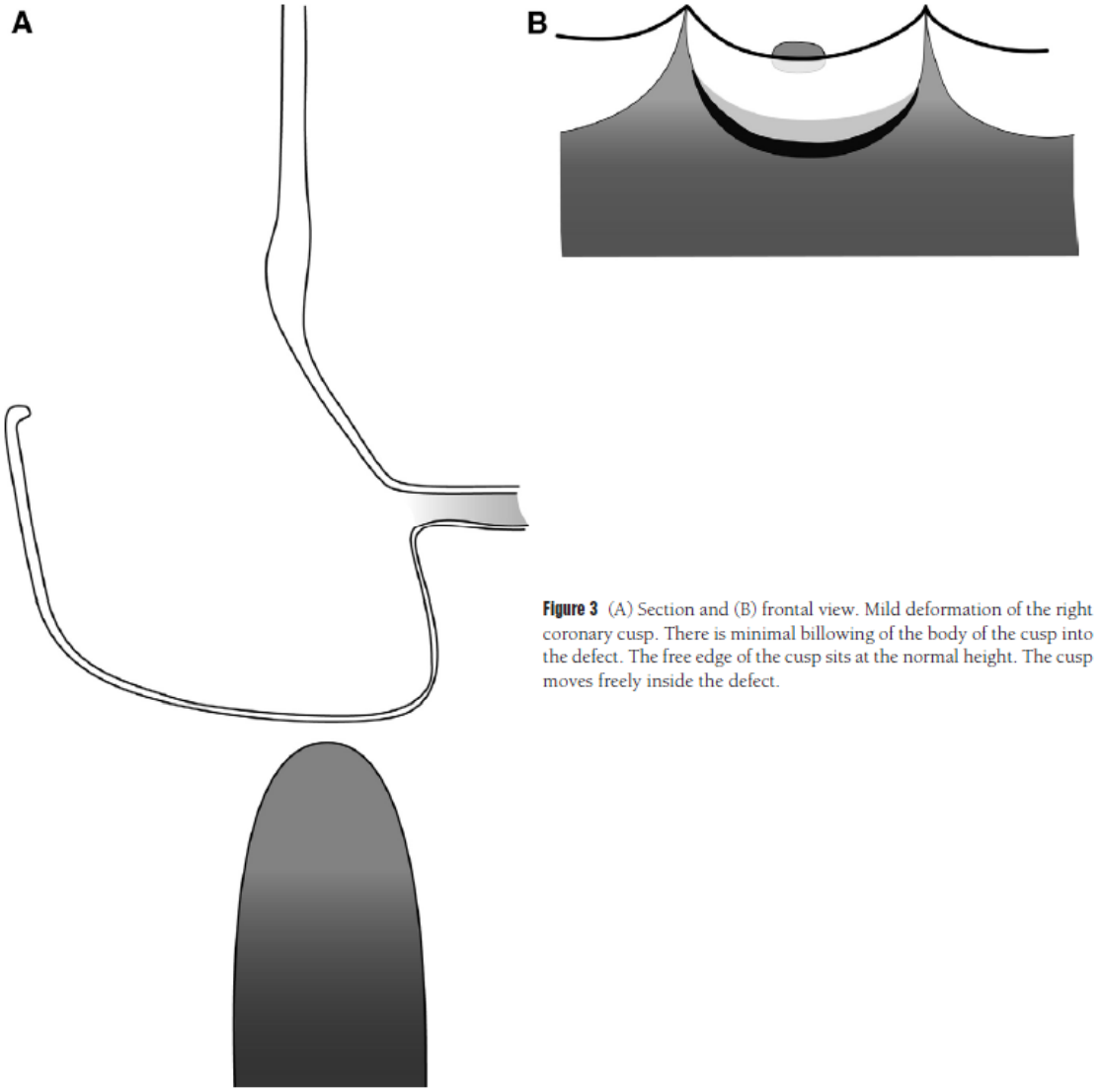

Figure 3 (A) Section and (B) frontal view. Mild deformation of the right coronary cusp. There is minimal billowing of the body of the cusp into the defect. The free edge of the cusp sits at the normal height. The cusp moves freely inside the defect.

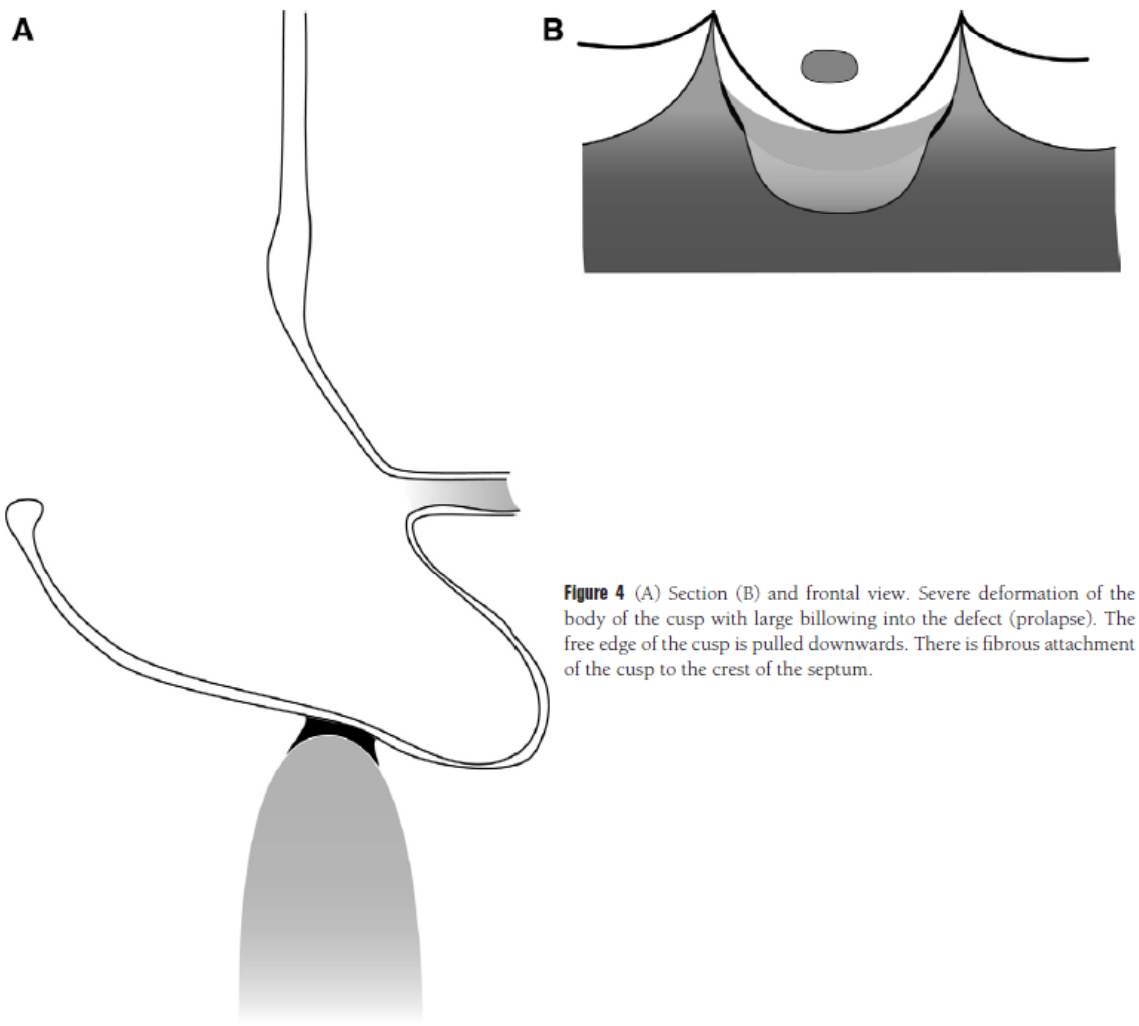

Figure 4 (A) Section (B) and frontal view. Severe deformation of the body of the cusp with large billowing into the defect (prolapse). The free edge of the cusp is pulled downwards. There is fibrous attachment of the cusp to the crest of the septum.

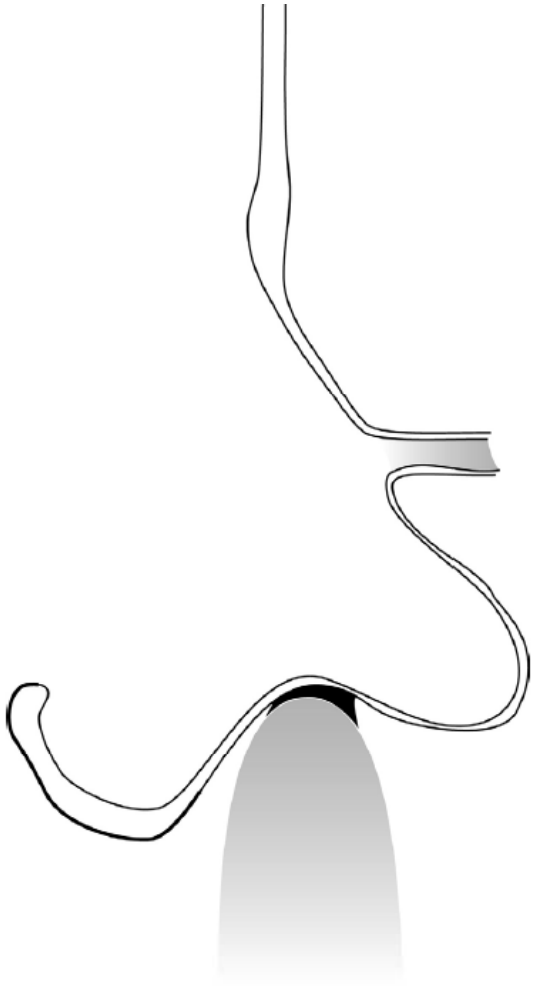

Figure 5 The height of the cusp is limited while the free edge is elongated. There are significant alterations of the cusp tissue.

Severity indices of RCC prolapse

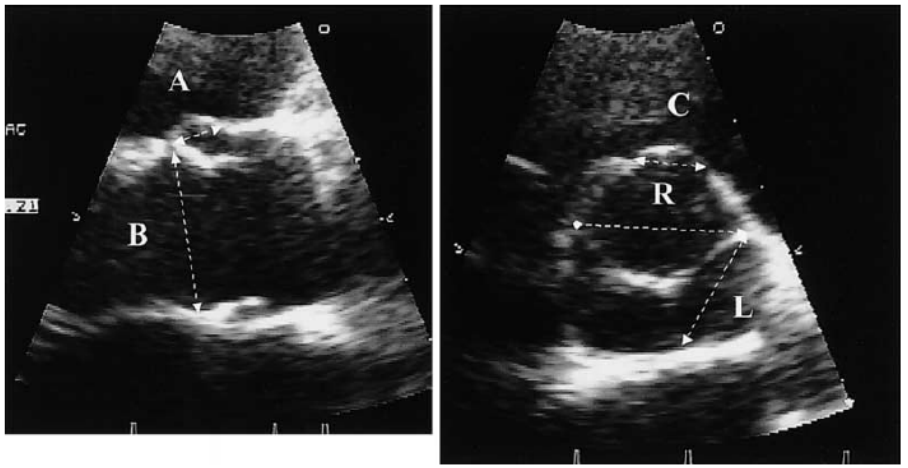

Fig 1. Method of measuring the right coronary cusp deformity index (Left), right coronary cusp imbalance index, and the maximum diameter of the original defect (Right). Right coronary cusp deformity index was obtained by dividing the prolapsed part of the right coronary cusp (A) by the ring diameter of the aortic valve (B) at end-systole, and the right coronary cusp imbalance index was calculated as the width of the right coronary cusp (R) divided by the width of the left coronary cusp (L). The maximum diameter of the original defect (C) was measured with the assumption that the right coronary cusp did not cover the defect.

- RCC deformity index = A/B (< 0.3)
- RCC imbalance index = R/L (< 1.3)

Tomita H et al. Circ J 2004

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Mean±SD			0.24±0.04	0.22±0.05	0.24±0.04	0.25±0.03	0.39±0.11	0.37±0.05	0.47±0.15	0.47±0.17	
R/L											
Range			0.97–1.29	0.95–1.27	1.31–1.61	1.33–1.42	1.03–1.29	0.82–1.25	1.30–1.64	1.30–1.55	
Mean⊥SD			1.15⊥0.08	1.15⊥0.08	1.38⊥0.11	1.37⊥0.05	1.17±0.07	1.13±0.10	1.39±0.08	1.39±0.09	
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RCCD, right coronary cusp deformity index; R/L, right coronary cusp imbalance index; F/U, follow-up; RCCP, right coronary cusp prolapse; AR, aortic regurgitation; Valsalva, rupture of the sinus of Valsalva aneurysm; ( ), obvious progression of AR.
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Mean±SD	11.1±3.3*	7.3±2.5	8.1±2.9	6.2±2.9	7.7±1.3	5.7±1.9	9.6±4.8	7.8±4.0	12.8±4.8*	7.9±2.1

*#Trivial AR; *p<0.01.*