

Treatment of congenital aortic valve disease:

Neonatal surgical management

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Challenges

- . valvar lesions
- . associated lesions
- . status of left ventricle

Valvar lesions

- . bicuspid or unicuspid valve
- . rarely, tricuspid valve
- . usually, one normal commissure

- . thick dysplastic leaflets
- . excrement fibrous nodular tissue

- . size of annulus : + or - hypoplastic

Associated lesions

- . LVOT lesions
 - subvalvar stenosis
 - supravalvar stenosis
- . mitral stenosis (parachute mitral valve)
- . aortic coarctation

Standard surgical procedure

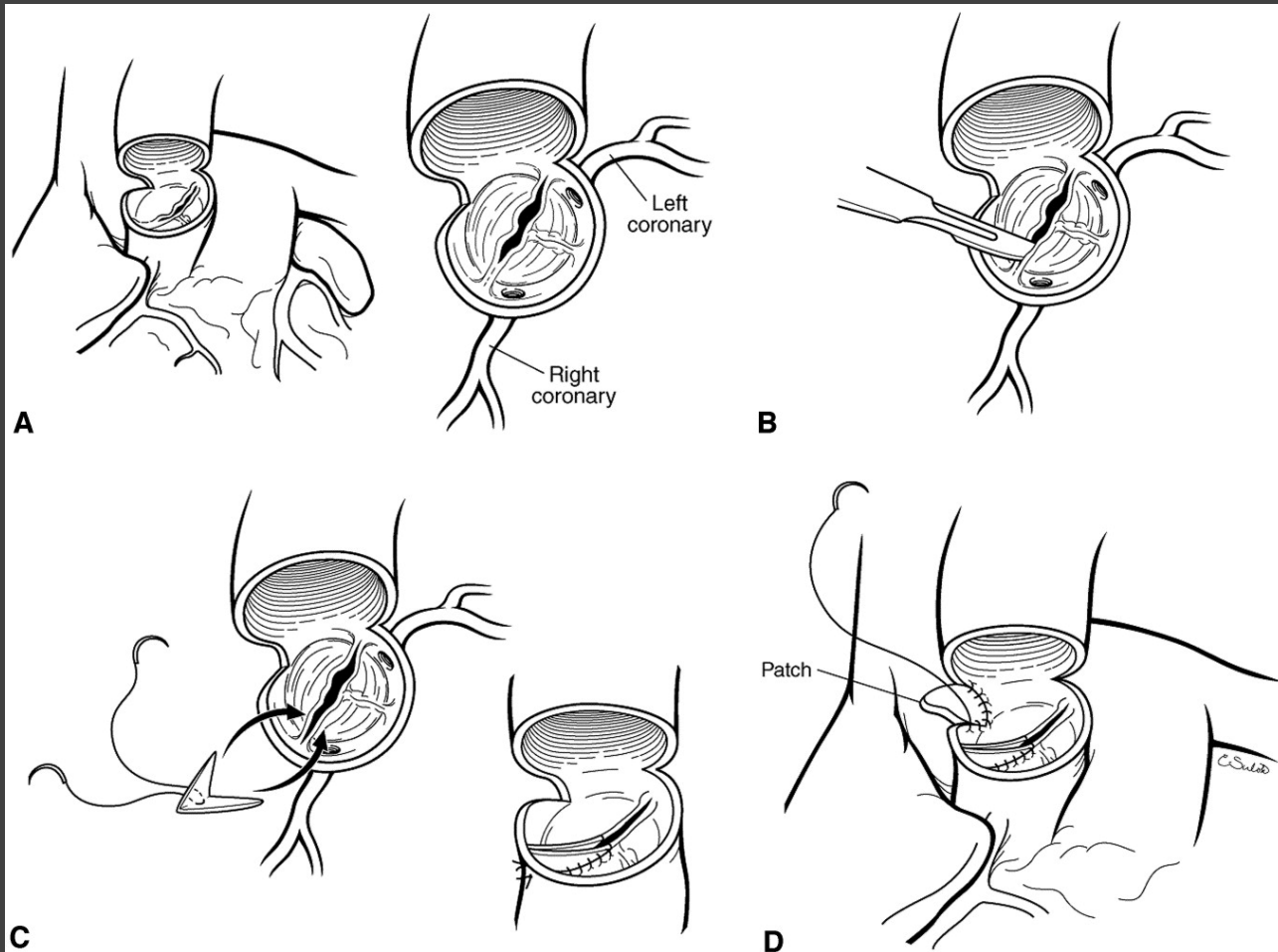
- . extended commissurotomy (bicuspid)
- . aggressive shaving of leaflets
- . excision of fibrous nodules

- . commissural reconstruction
- . supravalvar enlargement

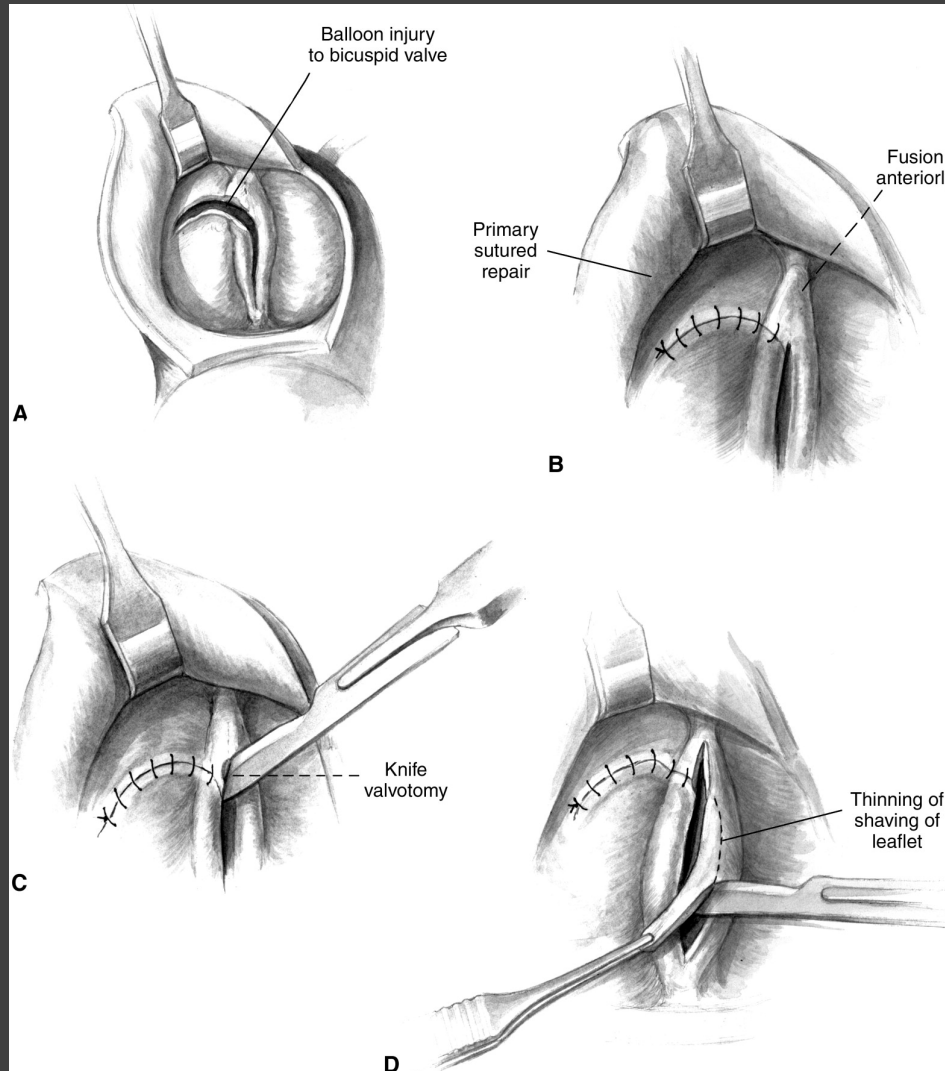
- . ligation of PDA
- . complete / partial closure of ASD
- . coarctation repair if needed

Neonatal aortic valve surgery

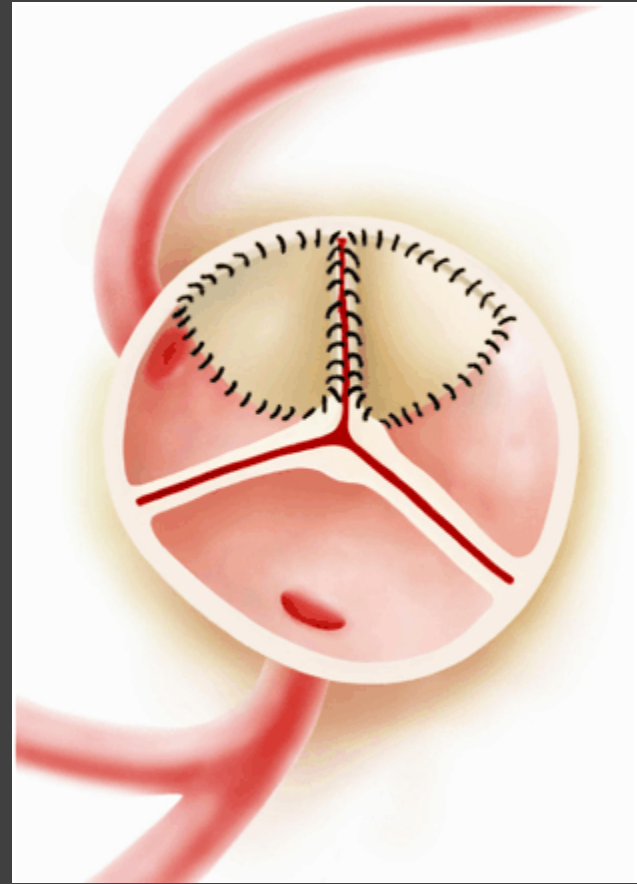
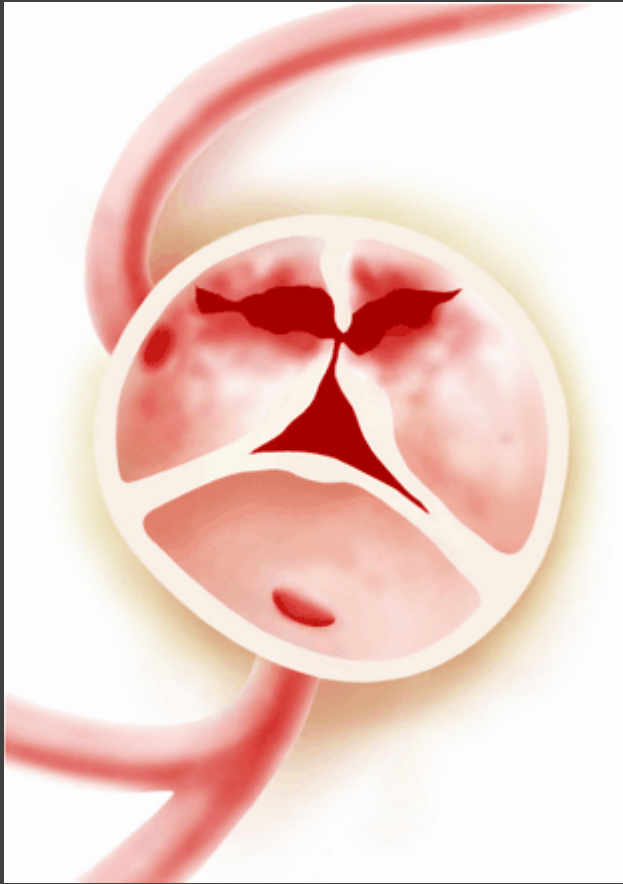
Standard surgical procedure



Standard surgical procedure



Tear repair + leaflet reconstruction



Valvuloplasty in neonates: results

2008 - 2011

early mortality

Necker-Enfants Malades (19 patients)	1/19 (5.3%)
EACTS database (71 patients)	6/71 (8.5%)

Neonatal aortic valve surgery

1016

B. Alsoufi et al. / European Journal of Cardio-thoracic Surgery 31 (2007) 1013–1021

Table 2
Selected results of surgical aortic valvulotomy in neonates and infants

Study	Operative technique	Age (months)	Operative mortality (%)	Survival	Freedom from re-intervention
Koivumäki et al. [36]	TVAV	10	34	9 years, 53%	4 reinterventions
Arora et al. [12]	TVAV	10	55.5	10 years, 52%	10 reinterventions
Cobanovici and Bobbs [34]	TVAV	10	14.3	7 years, 50%	7 reinterventions
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CAV: open aortic valvulotomy; TVAV: transventricular aortic valvulotomy.

3.2.2. Percutaneous balloon aortic valvuloplasty

Since the first report of successful percutaneous balloon aortic valvuloplasty (BAVP) for aortic stenosis in infancy, this minimally invasive technique, in many institutions, has become the favored technique. In many institutions, retrograde access is favored, although an antegrade approach using the umbilical or the femoral

arteries is also used. The normal range within 1–2 years of survival on the LV outflow tract was 48% at 5 years and freedom from aortic valve replacement was 54% at 5 years. Outcomes of selected series of infants undergoing BAVP are detailed in Table 3.

Table 3
Selected results of percutaneous balloon aortic valvuloplasty in neonates and infants

Study	Number of patients	Age (months)	Mortality (%)	Survival	Freedom from re-intervention
Arora et al. [37]	12	10	14	8 years, 88%	8 years, 60%
Arora et al. [42]	142	10	14.1	10 years, 53%	5 years, 50%
Arora et al. [34]	13	10	14.1	10 years, 53%	10 years, 48%
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arteries. However, retrograde approach using the carotid artery has been described [40, 41]. In a report comparing retrograde and antegrade balloon aortic valvuloplasty, antegrade approach was associated with diminished mortality, but compared with retrograde approach [40, 41]. Advantages of a percutaneous intervention include the avoidance of a surgical incision, which may be associated with a number of disadvantages including vascular access complications, inability to determine the degree of stenosis, and the potential for subsequent potential for aortic valve insufficiency and aortic regurgitation. It is important to note that the goal is not complete elimination but improvement of the stenosis so that it can be repeated, and Bissler et al. [42] point out that the procedure can be repeated. Factors were identified including extremely young neonates, older age of intervention and infants with aortic regurgitation. A recent review from Boston of 113 infants < 20 days of age who underwent BAVP showed a mean rate of reintervention (AR) developed in 15% of patients on follow-up. Reintervention with freedom from moderate or severe AR or > 50% reduction in the aortic valve dimension and left ventricular end-diastolic dimension Z-scores increased to

3.2.3. Percutaneous valvuloplasty versus surgical

Both surgical valvotomy and percutaneous balloon valvuloplasty are associated with important mortality and morbidity such as residual or recurrent aortic valve dysfunction, and the need for re-intervention [4, 5, 7, 34, 36, 38]. There are similarities between the two techniques. Numerous reports cited each by the other, but few compared surgical versus percutaneous techniques and those that are linked by the lack of adjustments between the two groups [2, 37, 39]. The largest comparative study by the American Society of Thoracic Surgeons (ASTS) [39] compared surgical aortic valvulotomy (SAV; n = 23) or percutaneous balloon aortic valvuloplasty (BAVP; n = 23). The study demonstrated that while controlling for pre-intervention aortic stenosis, BAVP was more effective for pre-intervention than SAV as evidenced by a greater mean percent reduction in aortic stenosis (34% vs. 24%, p = 0.03) and mean flow (3.9 ml/min/m² vs. 2.0 ml/min/m², p = 0.001). However, BAVP was also associated with a greater need for re-intervention (18% vs. 33%, p = 0.03) and a greater need for re-intervention (18% vs. 33%, p = 0.03). Significant factors for re-intervention included

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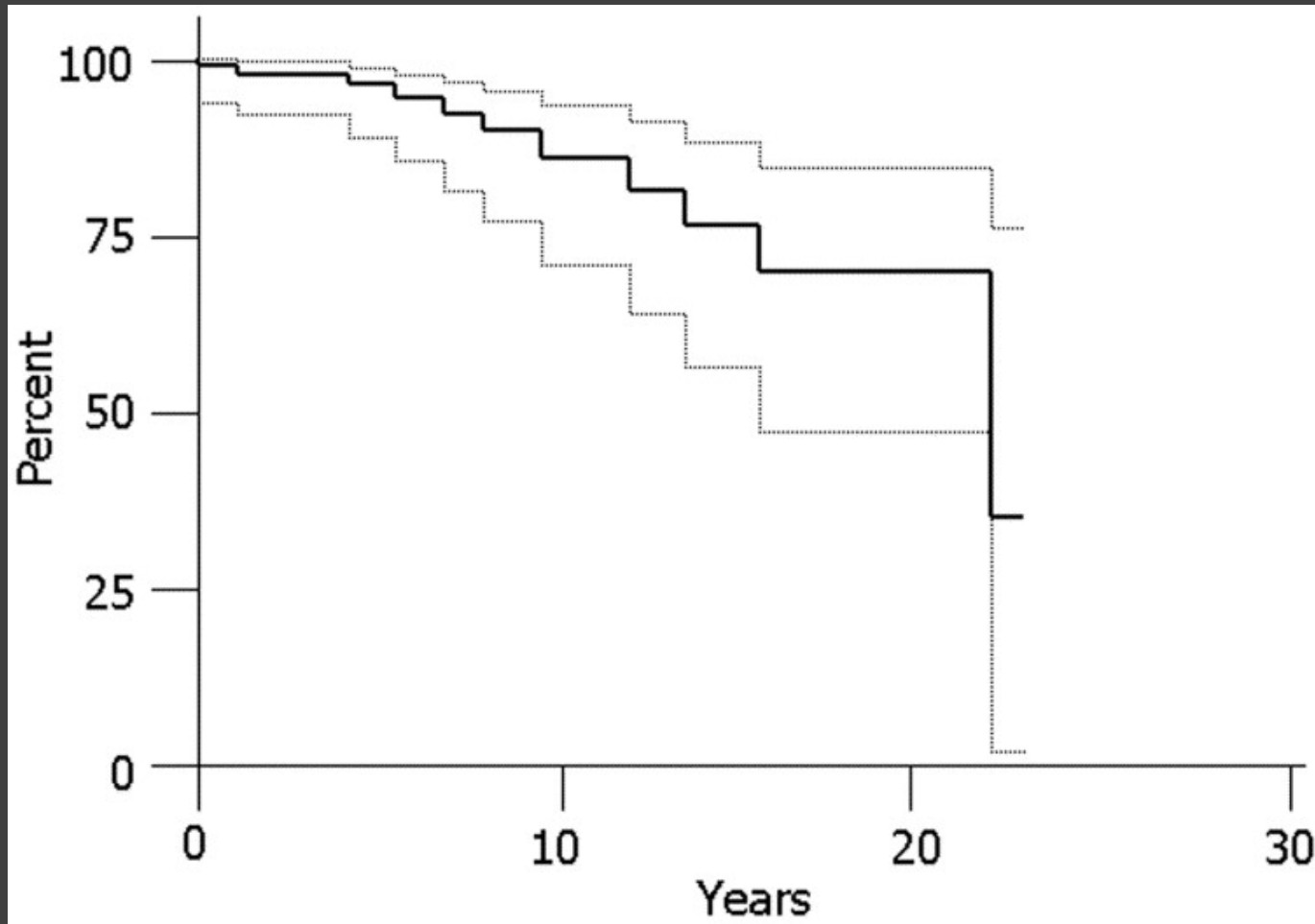
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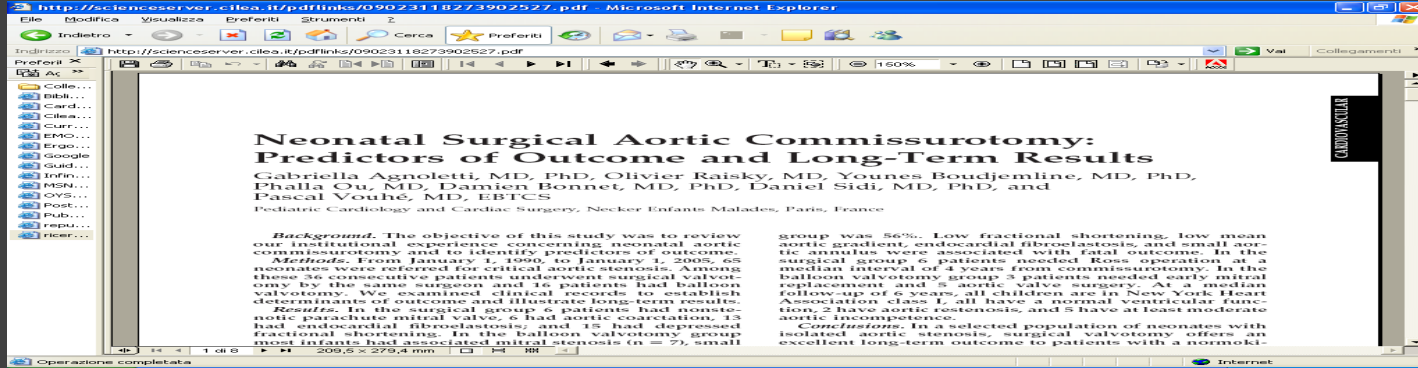
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Simple aortic valve repair : freedom from reintervention

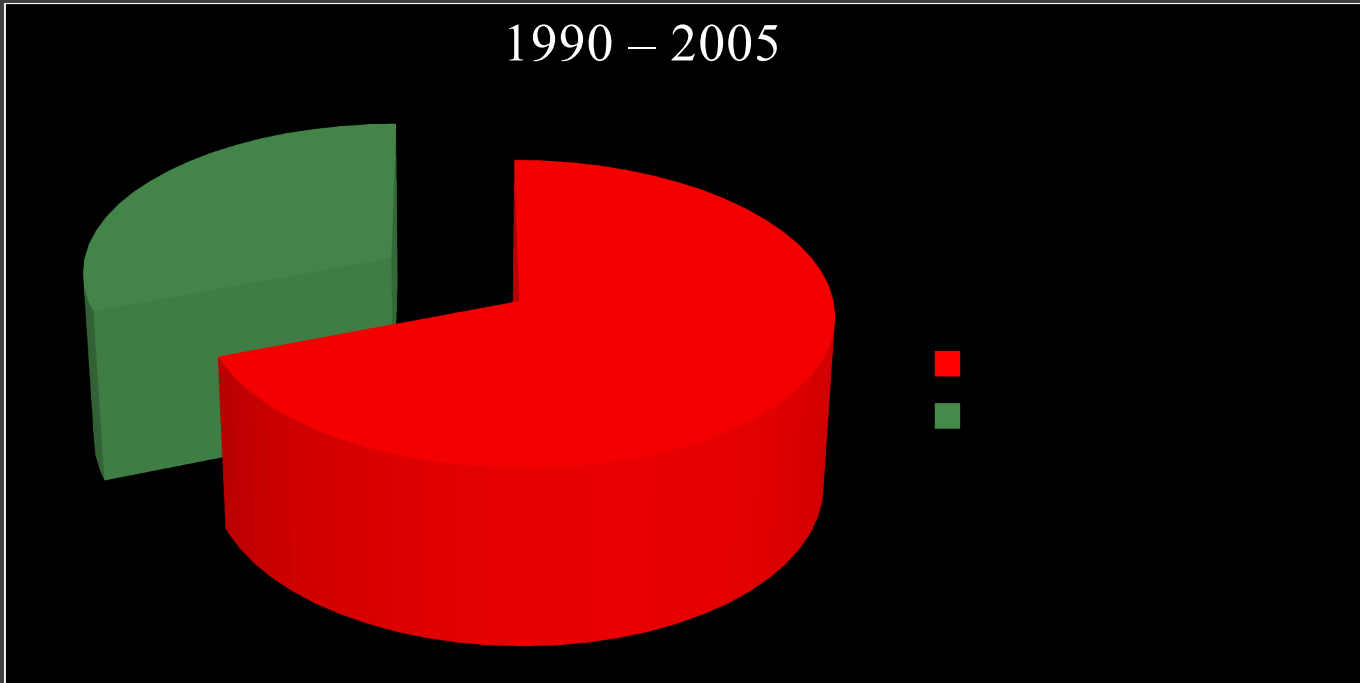


Neonatal aortic valve surgery



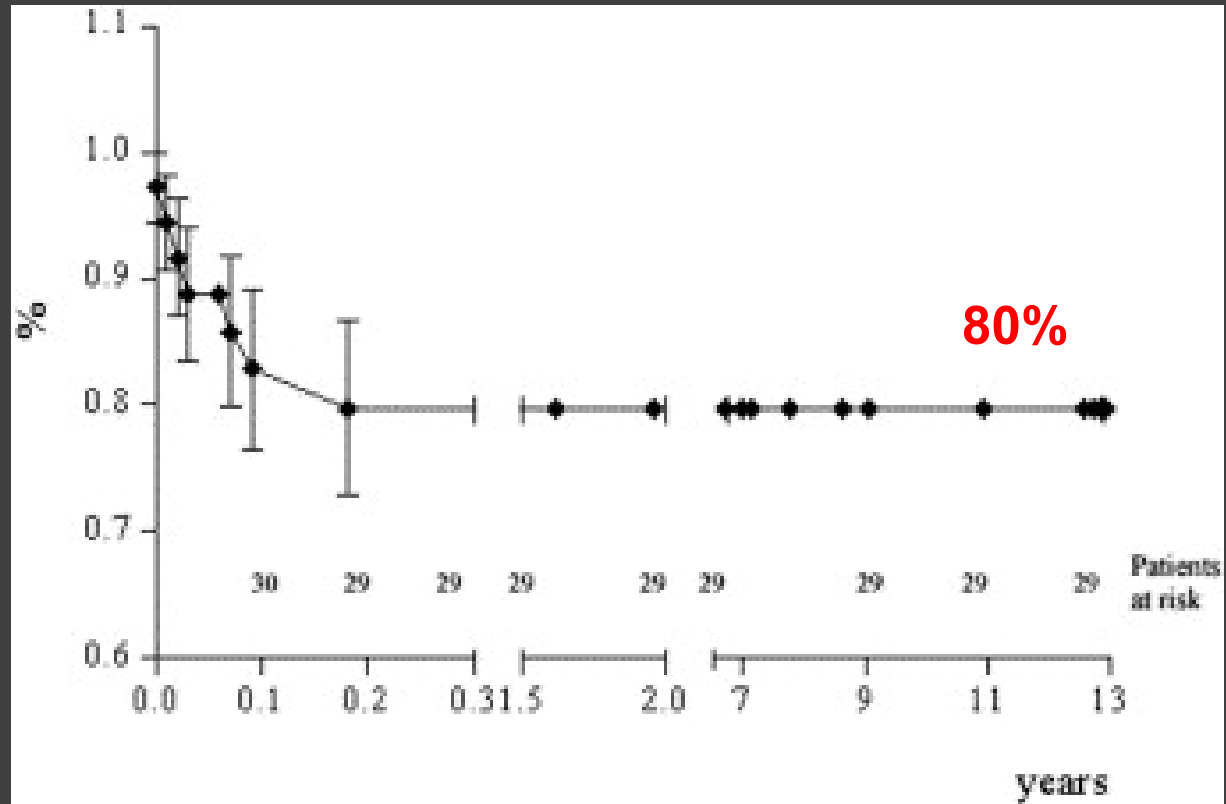
Ann Thorac Surg 2006; 82: 1585-93

1990 – 2005



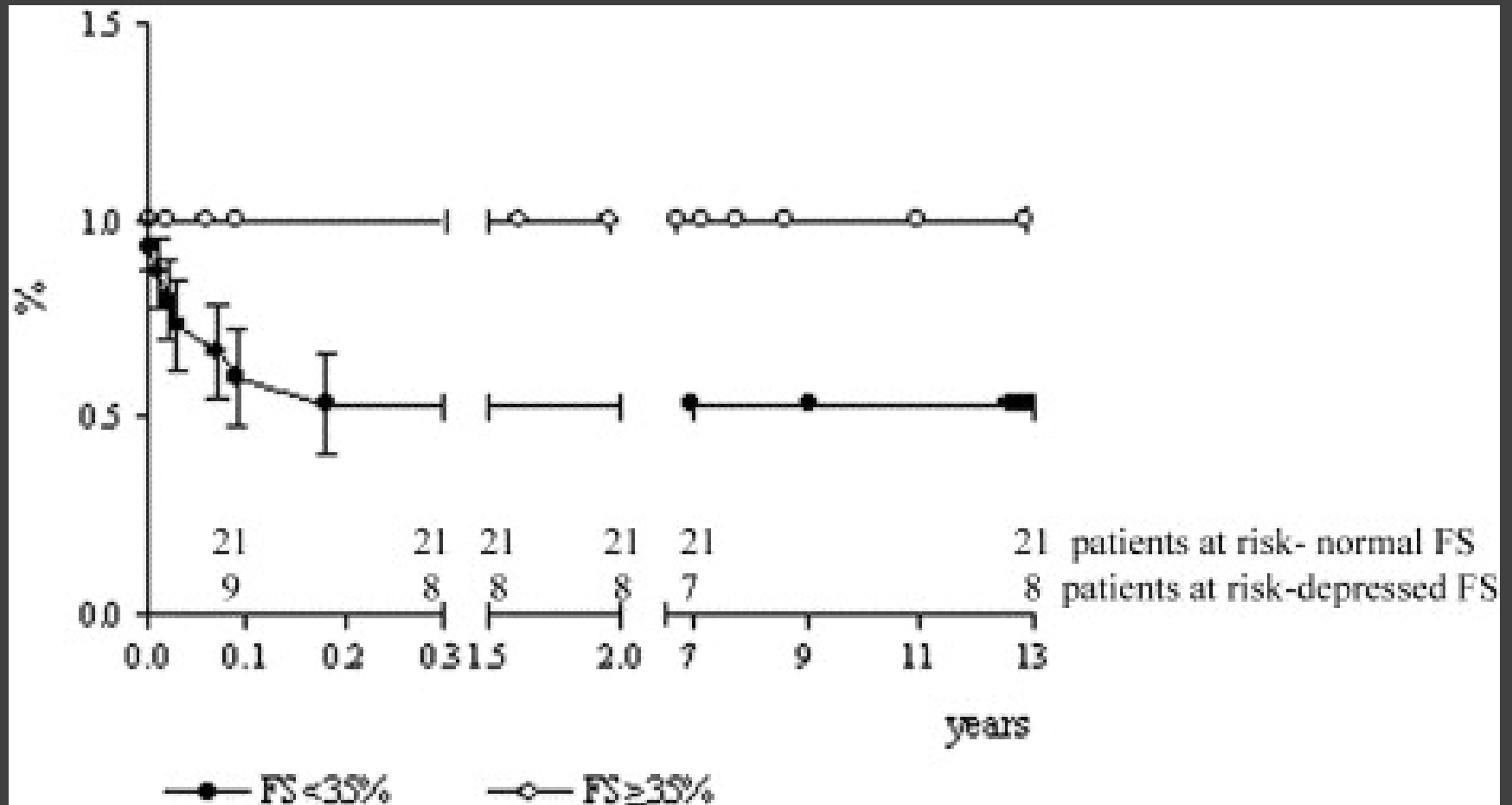
Neonatal aortic valve surgery

* Survival



* Reintervention : 27% of survivors (6 Ross, 2 balloon dilatation)

Neonatal aortic valve surgery



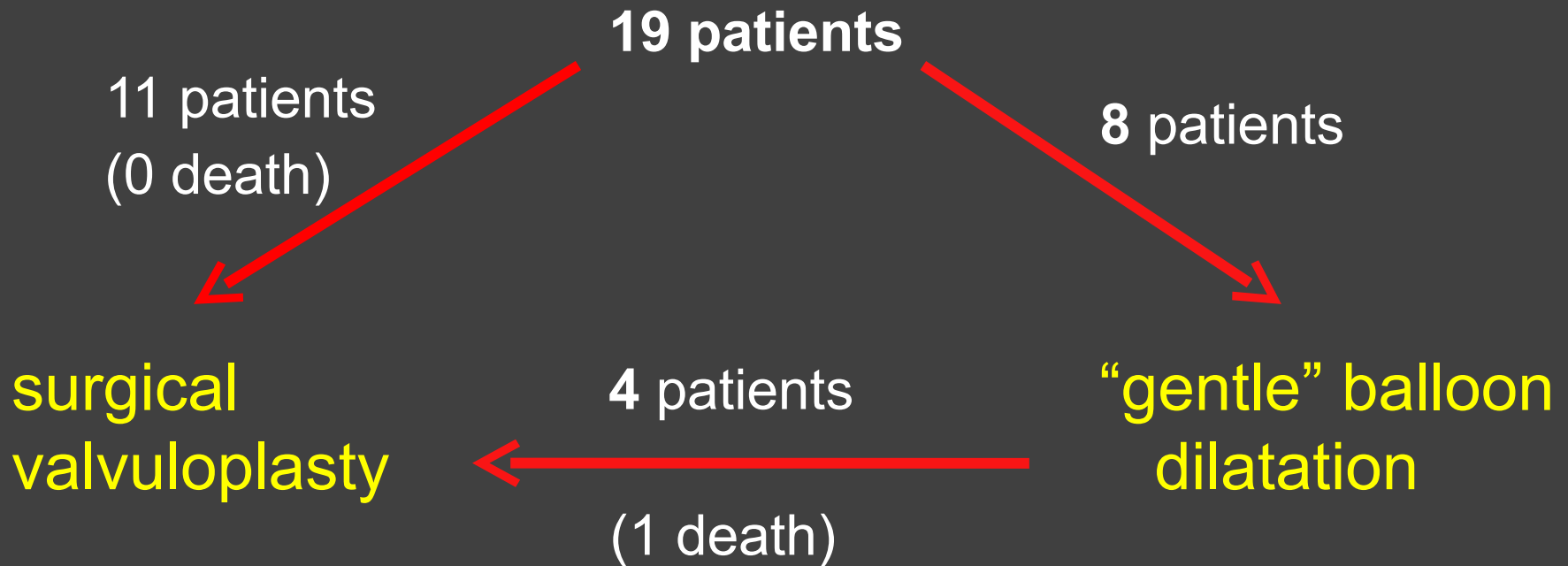
Challenges

- . valvar lesions
- . associated lesions
- . left ventricle: function and size

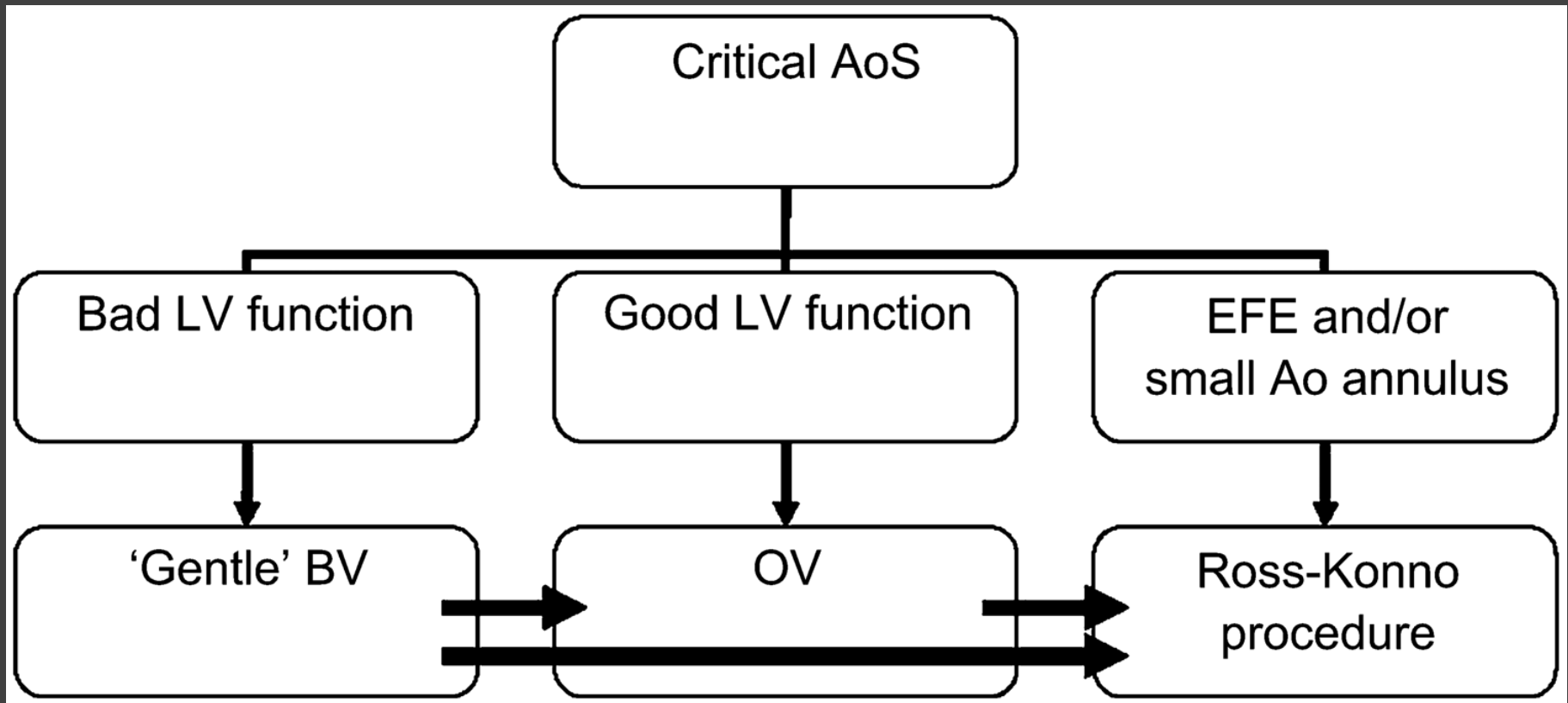
left ventricle

- . left ventricular dysfunction
 - endocardial fibroelastosis
 - impaired systolic / diastolic function
 - potentially reversible
- indication for "partial" balloon dilatation
- . left ventricular hypoplasia

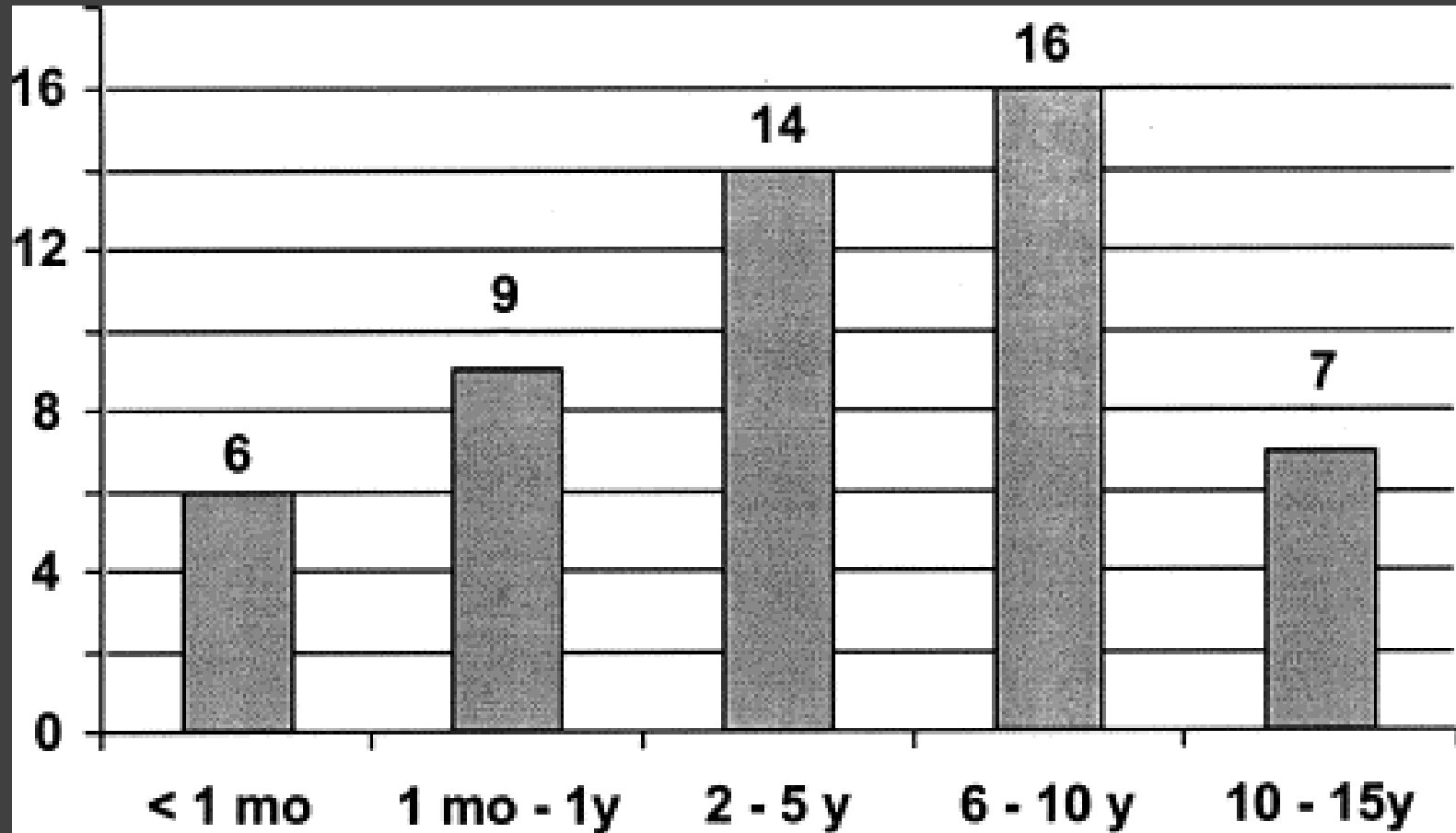
Necker-Enfants Malades 2005 - 2010



left ventricular dysfunction



Ross procedure in children



Ross / Ross-Konno procedures : mortality

	> 1 month	< 1 month
Ross	2/3	1/29
Ross - Konno	2/3	0/17
Total (52 pts)	4/6 (67%) *	1/46 (2.2%)

* all deaths in rescue Ross for failed dilatation

left ventricle

- . left ventricular dysfunction
- . left ventricular hypoplasia
 - absence of absolute criteria for biventricular repair
 - univentricular approach
 - hybrid approach

hybrid approach

- . initial procedure
 - bilateral PA banding
 - duct stenting
 - balloon aortic valvuloplasty
 - ASD size calibration
- . "buys" time for improvement
- . allows delayed decision-making

Conclusions

- 1. subnormal LV function and size** (most patients)
 - surgical valvuloplasty provides excellent results
 - there is no place for balloon dilatation
- 2. severe LV dysfunction**
 - "gentle" balloon dilatation
 - followed by surgical valvuloplasty or Ross-Konno
- 3. LV hypoplasia**
 - individualized management
 - potential place for hybrid approach.