Unicuspid Aortic Valve Repair

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Aortic Valve Replacement

Reproducible

Low Mortality
(curr. 2-4%)

Late complications:

Thromboembolism
Anticoagulation/Hemorrhage
Structural failure
PV endocarditis

Pediatric AV Replacement

Outcomes and Associated Risk Factors for Aortic Valve Replacement in 160 Children

A Competing-Risks Analysis

Tara Karamlou, MD; Karen Jang, MS; William G. Williams, MD; Christopher A. Caldarone, MD; Glen Van Arsdell, MD; John G. Coles, MD; Brian W. McCrindle, MD, MPH

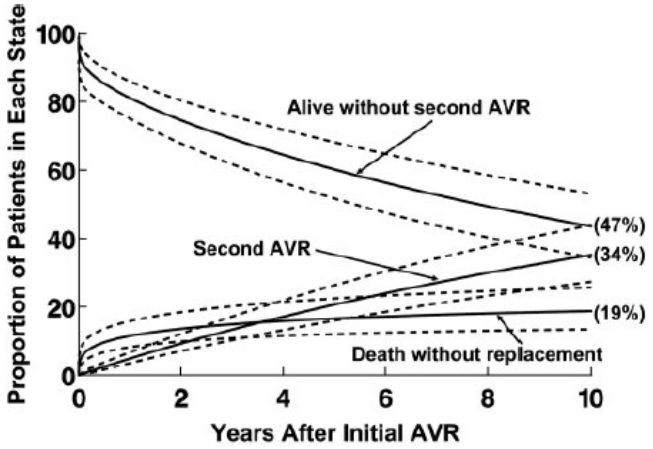
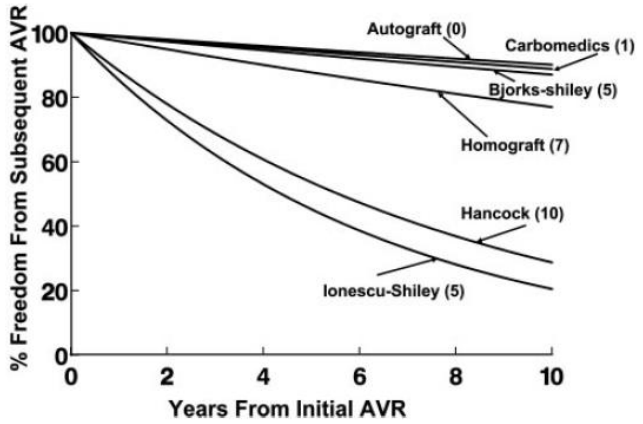
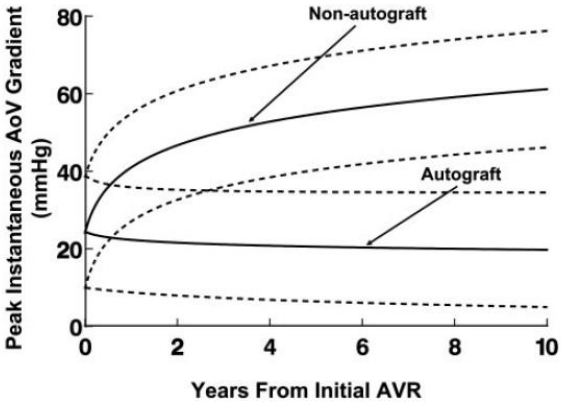


Figure 2. Competing-risks depiction of events after initial AVR in 160 children. All patients are represented in the graph as alive

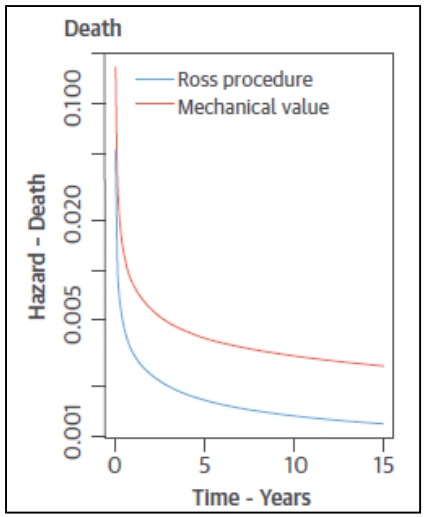


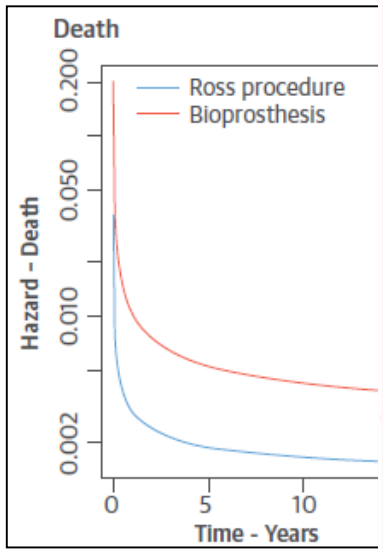


(*Circulation*. 2005;112:3462-3469.)

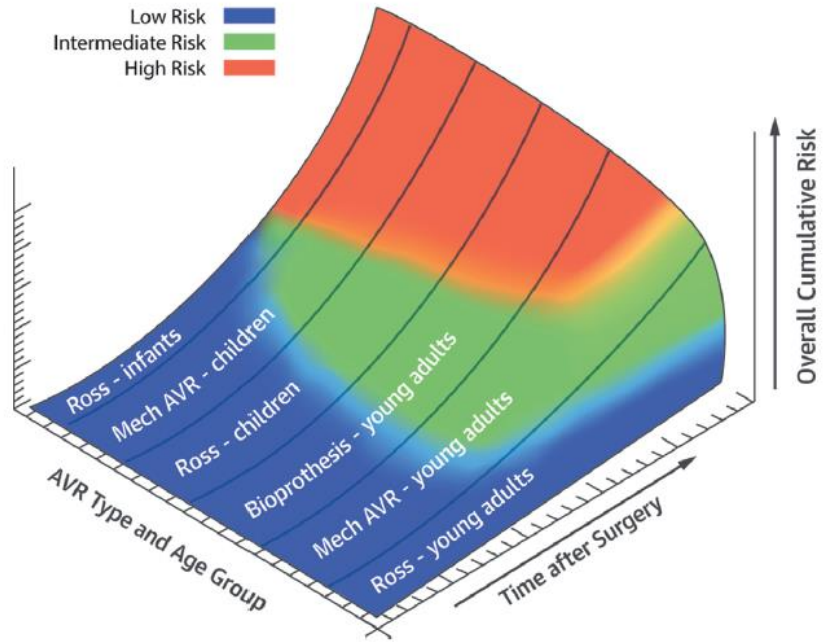
Aortic Valve Replacement and the Ross Operation in Children and Young Adults

Mansour T.A. Sharabiani, PhD,[a] Dan M. Dorobantu, MD,[b,c] Alireza S. Mahani, PhD,[d] Mark Turner, PhD,[b]
Andrew J. Peter Tometzki, MBChB,[b] Gianni D. Angelini, MD,[a,b] Andrew J. Parry, MBChB,[b] Massimo Caputo, MD,[b]
Serban C. Stoica, MD[b]





CENTRAL ILLUSTRATION **AVR and the Ross Operation in Children and Young Adults**



Sharabiani, M.T.A. et al. J Am Coll Cardiol. 2016;67(24):2858-70.

Dynamic changes of risk of mortality or reintervention during follow-up period after aortic valve replacement (AVR) operation by the procedure types and the age groups. Risk of both mortality and reintervention decrease with the patient age and increase during follow-up. The overall increase of risk differs depending on the AVR procedure type and the speed of increase in risk during the various stages of follow-up time is not proportional across the AVR types. Distances and the levels of increase in risk are drawn schematically and, therefore, are not precise.

JACC VOL. 67, NO. 24, 2016
JUNE 21, 2016:2858-70

Options and Disadvantages of AV Replacement in Pediatric Patients

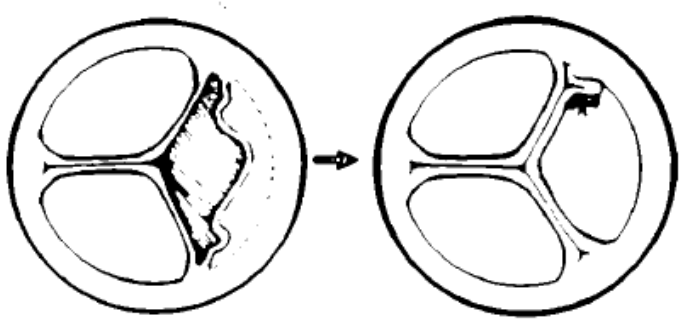
- Absence of growth potential (mechanical/biological AVR)
- Anticoagulation (mechanical AVR)
- Early degeneration (biological AVR)
- Excess mortality (mechanical and biological AVR)

- Extensive surgery (Ross operation)

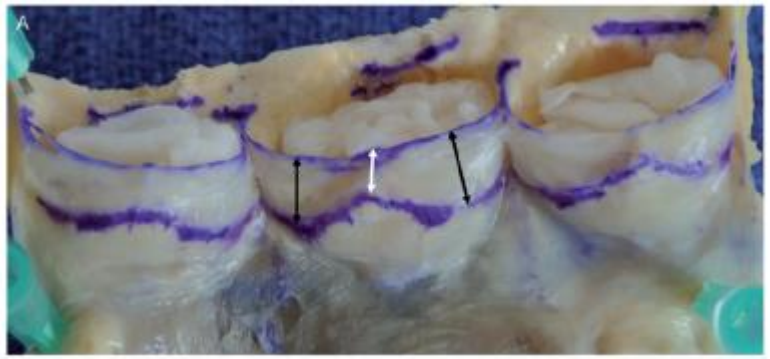
→ Repair as best compromise?

(Postponing the time of replacement to better circumstances e.g. low risk age)

✓All cusp margins should be at equal height for competent valve function.

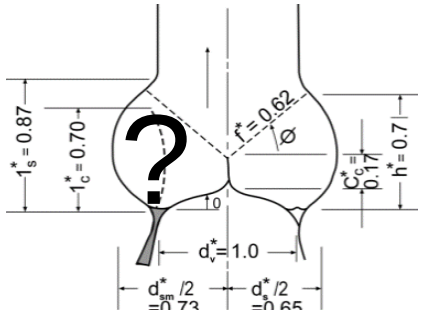


✓Coaptation height should be high for secure diastolic function.

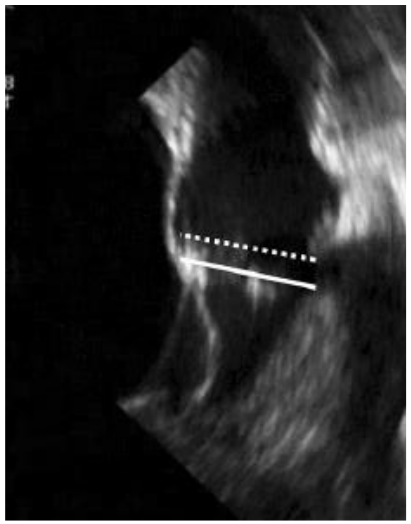


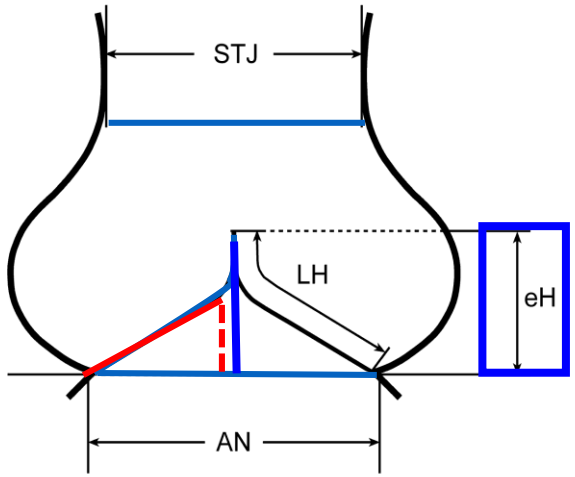
Duran CMG, J Card Surg 1994
De Kerchove L, Eur J Cardiothorac Surg 2018

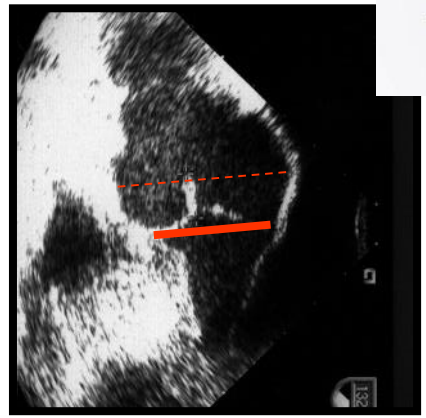
Looks are not enough: Effective and Geometric Height

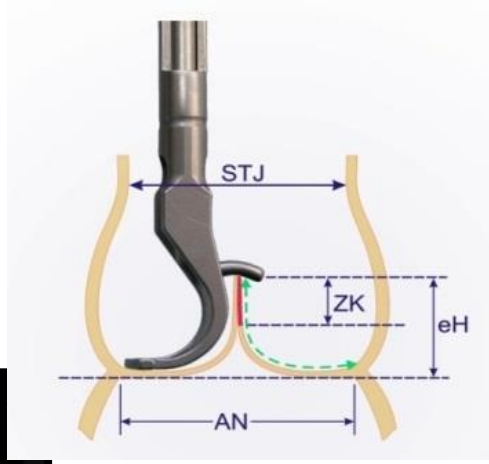


Swanson, Circ Res 1974









A new approach to the assessment of aortic cusp geometry

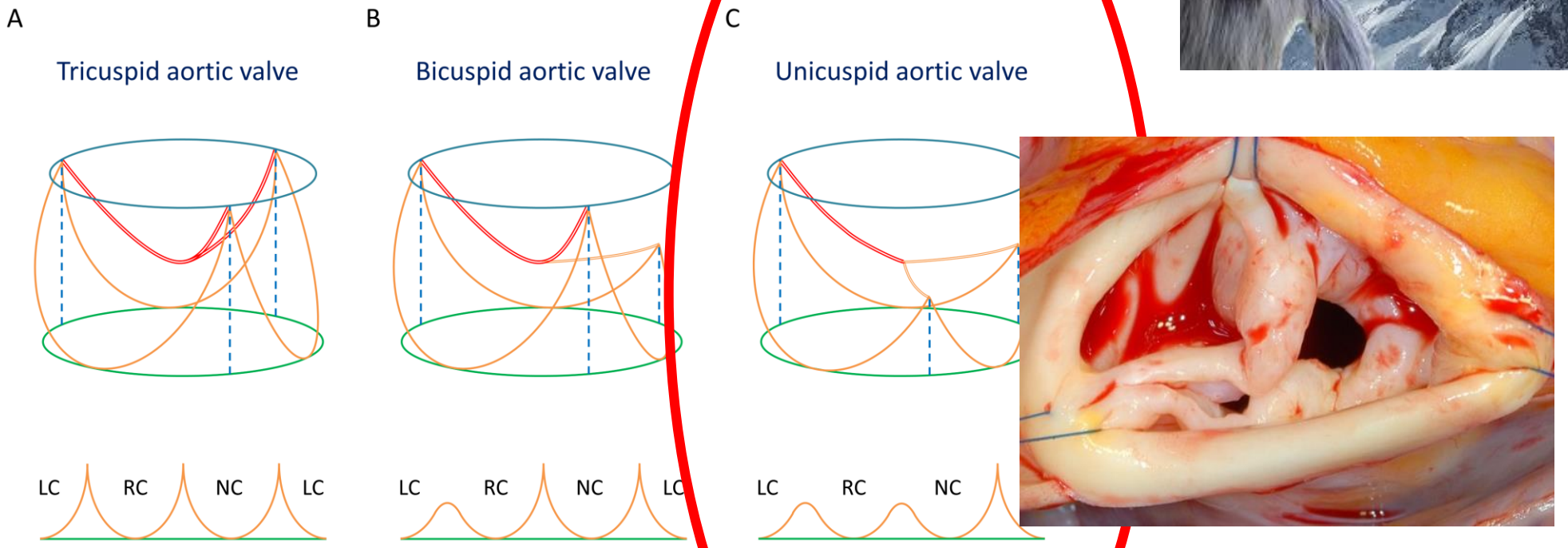
Hans-Joachim Schäfers, MD, PhD, Benjamin Bierbach, MD, and Diana Aicher, MD, Homburg/Saar, Germany

9-10 mm (or 45% of gH) = normal

Schäfers HJ et al, JTCVS 2006

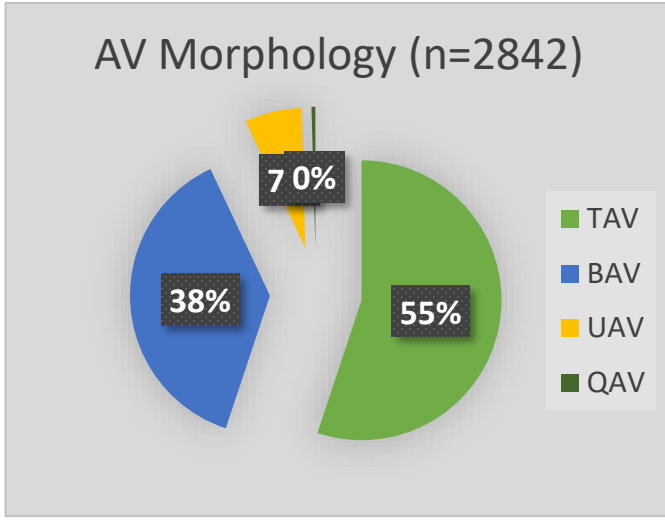
Unicuspid Aortic Valve (Pubmed Hits 161)

Did you mean: bicuspid aortic valve (2929 items)

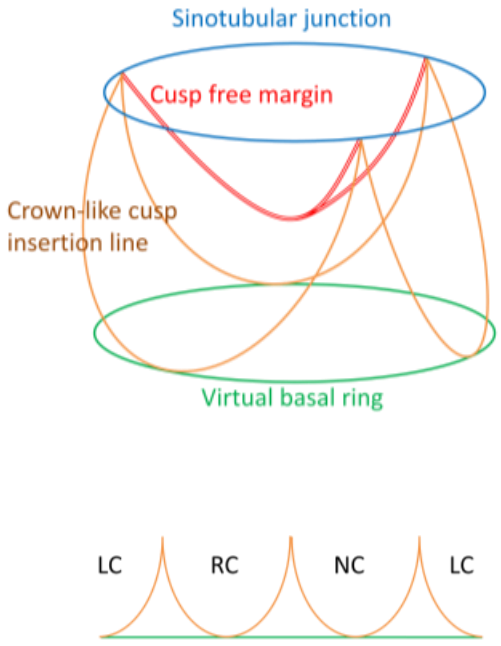


According to Anderson RH

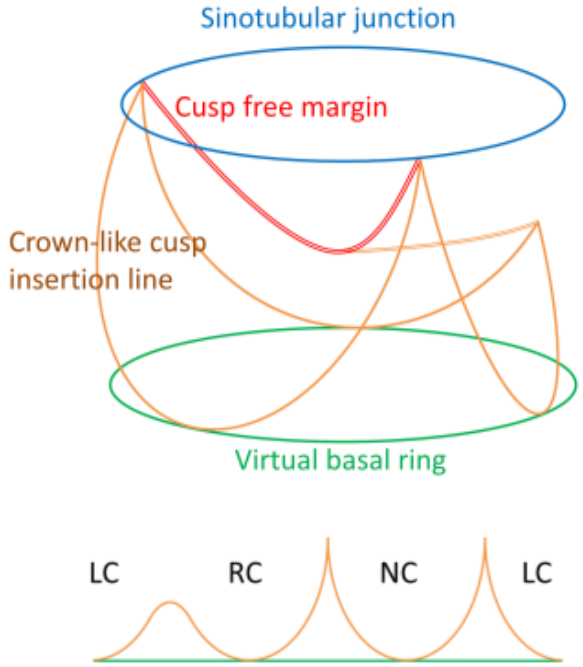
Aortic Valve Morphology



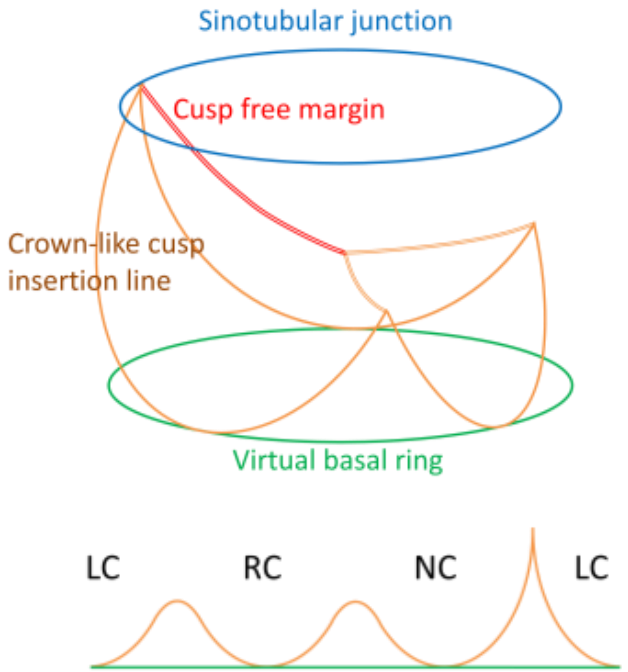
Tricuspid



Bicuspid

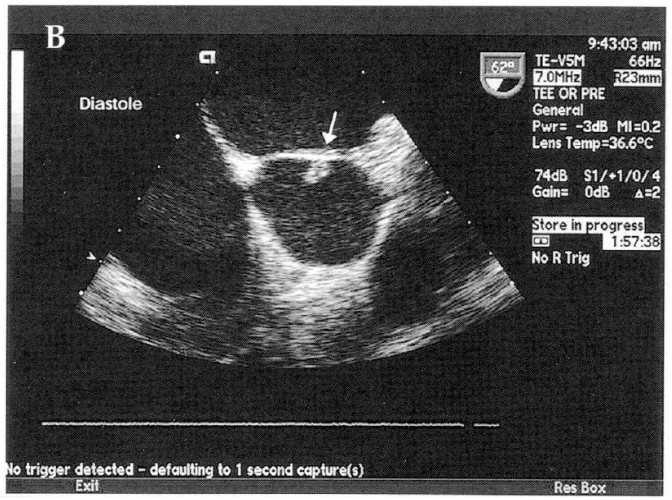


Unicuspid



Incidence and Echocardiographic Features of Congenital Unicuspid Aortic Valve in an Adult Population

Gian M. Novaro[1], Micky Mishra[2], Brian P. Griffin[3]



Patients

All patients who underwent transthoracic echocardiography in the authors' laboratory between January 1, 1990 and May 31, 2002 were screened for the study. Patients aged >18 years and described as having a congenital UAV were identified.

Among 113,552 individual patient echocardiographic examinations, 21 patients (14 males, seven females) with congenital UAVs were identified, resulting in an estimated incidence of 0.019%. The mean patient age was 34 ± 10 years (range: 21 to 60 years), and the mean

The Journal of Heart Valve Disease 2003;12:674-678

But: only adult patients!

Unicuspid Aortic Valve (Pubmed Hits 161)

Did you mean: bicuspid aortic valve (2929 items)

Canadian Journal of Cardiology 32 (2016) 110–116

Clinical Research

New Insights Into Unicuspid Aortic Valve Disease in Adults: Not Just a Subtype of Bicuspid Aortic Valves

Pierre-Emmanuel Noly, MD,[a] Lauren Basmadjian, MD,[a] Ismail Bouhout, MD,[a]
Van Hoai Viet Le, MD,[b] Nancy Poirier, MD,[a] and Ismail El-Hamamsy, MD, PhD[a]

Methods: From 2011 to 2015, all data from adult patients with confirmed UAVs (n = 42) who underwent aortic valve surgery were reviewed.

The prevalence of UAV in our surgical cohort of 2500 AV replacements over the study period was 1.68%. This is comparable with previously published data on the incidence

Table 2. Preoperative echocardiographic characteristics (n = 42 patients)

Characteristic	Value	Median	Range
Type of valvular dysfunction			
Pure stenosis	8 (19)		
Pure regurgitation	5 (12)		
Mixed stenosis and regurgitation	29 (69)		

Accuracy of preoperative echocardiographic diagnosis

Preoperative TTE diagnosis of UAV was made in only 6 patients (14%). The rest of the AVs were described as bicuspid in 33 patients (77%) or undetermined (n = 3; 8%). Intraoperative TEE increased the rate of detection before surgical inspection to 69% (n = 29 patients). To determine the potential for accurate determination of the preoperative diagnosis, all preoperative TTEs were re-examined with full knowledge of the valve phenotype observed during surgery. In

only adult patients!

Aortic Valve Dysfunction in Children and Adolescents
(What are we treating?)

- Congenital aortic stenosis
- AR after thx of aortic [illegible]osis
- Mixed AR / AS

- AR [illegible] BAV/QAV
- [illegible]R due to endocarditis
- Root aneurysm due to connective tissue disease

What is the morphology of the dysfuntional av in childhood and adolescence?

Aortic stenosis of the neonate: A single-center experience

Check for updates

Mathieu Vergnat, MD,[a] Boulos Asfour, MD,[a] Claudia Arenz, MD,[a] Philipp Sachweh, MD,[a] Benjamin [illegible], MD,[a] Viktor H [illegible],

Alain J. Poncelet[a,*], Gébrine El K [illegible],
Stéphane Moniotte[b], Mona [illegible],

ABSTRA

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[a] Department of Cardiovascular and Thoracic Surgery, UCL–Cliniqu
[b] Department of Pediatric Cardiology, UCL–Cliniques Universitaires
[c] Department of An
[d] Department of Pe

* Corresponding au
Bruxelles B-1200,

Received 24 Februar

Abstract

OBJECTIVES: To
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METHODS: A ret
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RESULTS: Sixty-si
36 (55%) and 17
regurgitation the
patients, VSD clo
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All were cardiac
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complication (en

CONCLUSIONS:
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surgery such as th

Keywords: Paedi

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Hans Peter G
James L. Wil
Melbourne, A

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Valve morphologies:
tricuspid
bicuspid
unicuspid ?

Long-term result:
recurrent AS
progressive AR

:84–89
-1243-0

CrossMark

CLE

of Aortic Regurgitation After Different Repair
or Congenital Aortic Valve Stenosis

hannes Kroll[1] · Jan Kiss[1] · Carolin Hess[1] · Brigitte Stiller[2] ·
iedhelm Beyersdorf[1]

Accepted: 3 August 2015 / Published online: 13 August 2015
ness Media New York 2015

t to characterize the incidence of AR
mine risk factors for AR progression
s of infants and children after surgical
tal aortic valvular and supravalvular
ients underwent repair of the aortic
genital aortic valve stenosis ($n = 14$,
ith aortic regurgitation (AR, $n = 16$,
%) had a valvular and $n = 3$ patients
th supravalvular pathology of their
16 patients (53 %) a bicuspid and in
s, a unicuspid valve was present.
was analyzed using the Cox model
ations. Log-rank calculations were
es reaching statistical significance in
erences in survival between groups.

patients had moderate or severe AR grades >2.5+. Patients with a monocuspid aortic valve and patients who had some kind of patch implantation into their cusps or commissures or shaving of thickened cusps were more likely to present with progression of aortic regurgitation. Monocuspid aortic valve and patch implantation, as well as cusp shaving, are probably linked to AR progression. The standard procedure of commissurotomy results in an absolute rate of AR progression of 40 % over a medium-term follow-up period.

Keywords Congenital aortic valve stenosis · Aortic valve repair · Bicuspid aortic valve · Monocuspid aortic valve · Aortic valve regurgitation

f aortic valve repair from a single institution.
c valve repair [median age 13.4 years (range 1 day to 45 years)].
nosis/aortic insufficiency 25 (45%), aortic insufficiency 24 (43%)
ub-commissural plication 36 (64%), commissurotomy 24 (43%),
(14%) and resuspension of commissures 4 (7%). Most patients
additional procedures. Hospital survival was 55/56 [98%; (95%
n for aortic valve pathology. Fifty-three patients [95%; (95% CI
nths; four survivors required aortic valve replacement and two
required repeat aortic valve repair [84%; (95% CI 72–91%)] reintervention-free survival].

Conclusions: **(1) In this study, aortic valve repair for congenital abnormalities avoided reoperation in the majority of patients, avoided anticoagulation and retained growth potential of the valve. (2) Repeat aortic valve repair or replacement was used to treat subsequent valve deterioration.**

(Heart, Lung and Circulation 2006;15:248–255)

Morphologyof Congenital AS

Morphology of the ventriculoaortic junction in critical aortic stenosis

Implications for hemodynamic function and clinical management

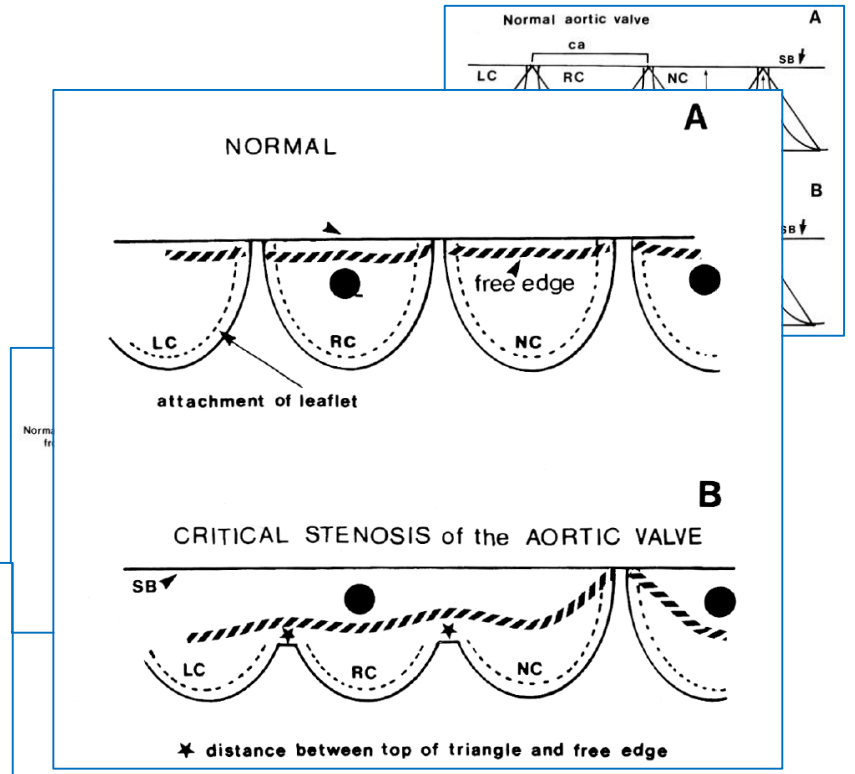
Roxane McKay, MD, FRCS, Audrey Smith, FIMLS, PhD,
Maurice P. Leung, MB, BS, MRCP,* Robert Arnold, MB, ChB, FRCP, and
Robert H. Anderson, BSc, MD, FRCPath,** *Liverpool, England*

The clinical presentation of infants with critical aortic stenosis, as well as the results of surgical treatment, differs from obstruction of the left ventricular outflow tract in older children. To investigate a possible anatomic basis for this situation, we performed a detailed morphometric study of 21 hearts from infants who had critical aortic stenosis and 11 normal hearts from infants less than 3 months of age. In each of the hearts with critical aortic stenosis, only one commissure extended to the sinutubular

Table II. *Height of the interleaflet triangles (*h *on Fig. 1) in millimeters*

	Normal		*Abnormal*		
	N	*Mean ± SD*	*N*	*Mean ± SD*	*p Value*
Left/right triangle	11	6 ± 0.8	20	2 ± 0.8	<0.0006
Noncoronary/right triangle	11	6 ± 0.7	19	2 ± 1.1	<0.001
Noncoronary/left triangle	11	6 ± 0.8	20	5 ± 0.7	<0.05

(J THORAC CARDIOVASC SURG 1992;104:434-42)



Morphology of Congenital AS

MORPHOLOGIC DETERMINANTS FAVORING SURGICAL AORTIC VALVULOPLASTY VERSUS PULMONARY AUTOGRAFT AORTIC VALVE REPLACEMENT IN CHILDREN

Jacques A. M. van Son, MD*
V. Mohan Reddy, MD
Michael D. Black, MD
Hiranya Rajasinghe, MD
Gary S. Haas, MD
Frank L. Hanley, MD

The pulmonary autograft is being used with increasing frequency to replace the diseased aortic valve in the pediatric population. Attempted surgical aortic valvuloplasty with an unacceptable result and return to cardiopulmonary bypass for aortic valve replacement with a pulmonary autograft results in prolonged bypass time and increased potential for morbidity. Therefore, the ability to predict an unsuccessful outcome for valvuloplasty

present study. *Methods:* Twenty-two patients (media weeks to 14 years) with bicuspid ($n = 11$), tricuspid pid ($n = 2$) aortic valves underwent valvuloplasty f 9), aortic regurgitation ($n = 7$), or a combination (n procedures included balloon aortic valvuloplasty (n valvotomy ($n = 1$). Median pressure gradient across

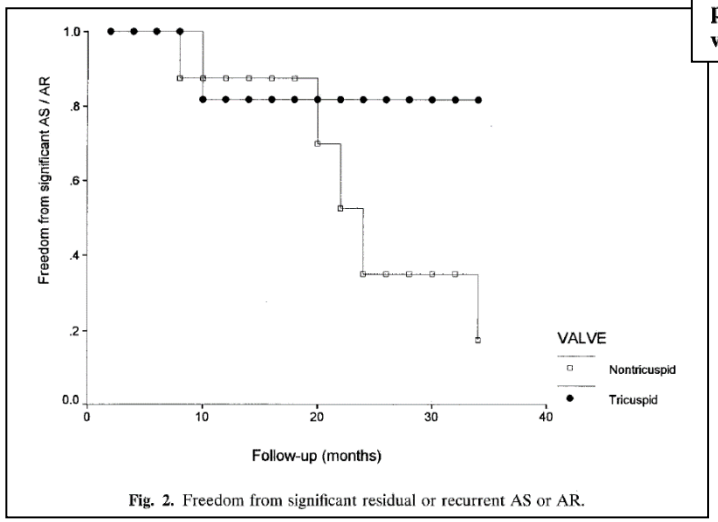


Fig. 2. Freedom from significant residual or recurrent AS or AR.

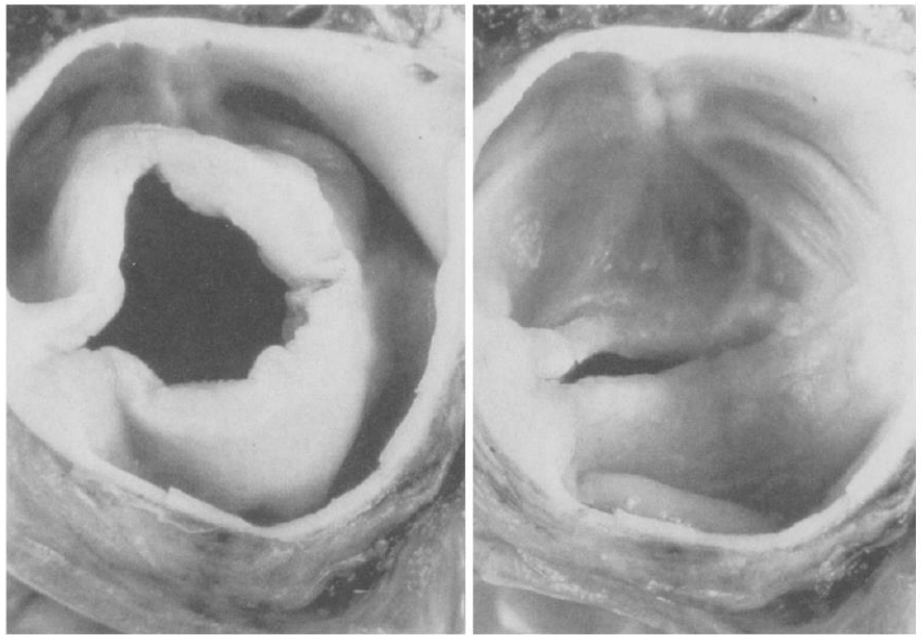


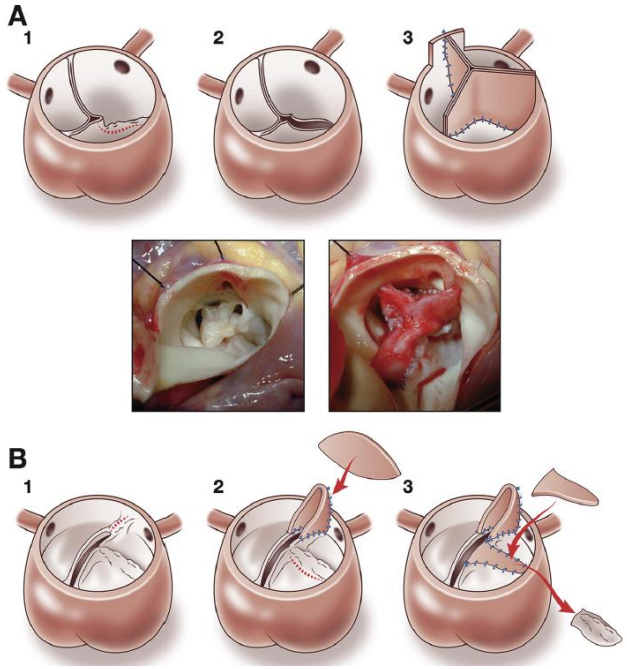
Fig. 1A. Bicuspid aortic valve. *Left,* Valve in open position. *Right,* Valve in closed position. Note commissural fusion and poor coaptation of leaflets caused by relatively annular arrangement of attachment of leaflets with resultant reduced height of interleaflet triangles and reduced depth of cusps.

(J Thorac Cardiovasc Surg 1996;111:1149-57)

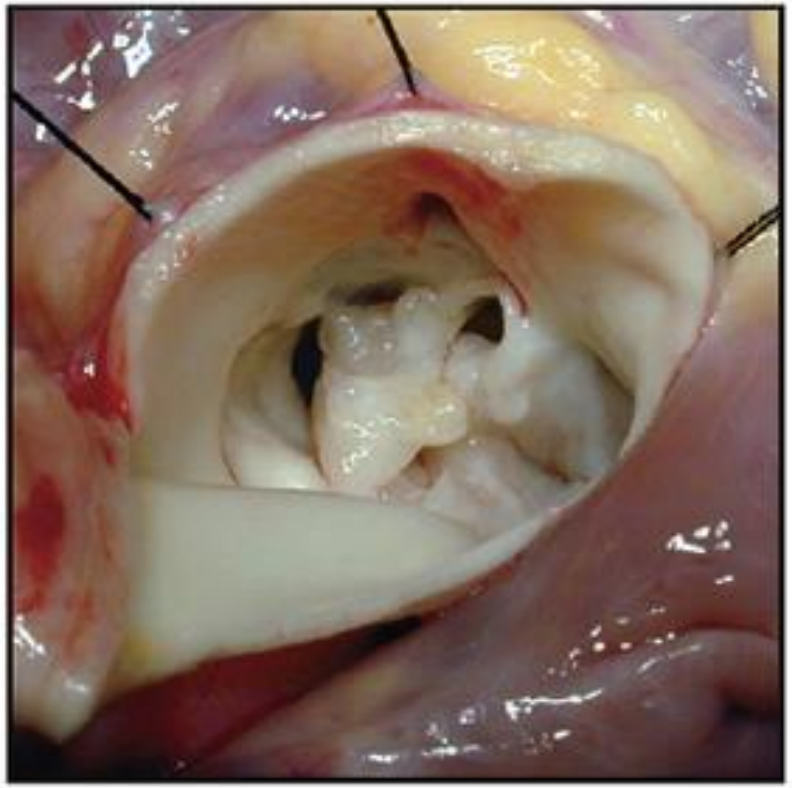
Morphology of Congenital AS

Outcomes After Operations for Bicuspid Aortic Valve Disease in the Pediatric Population

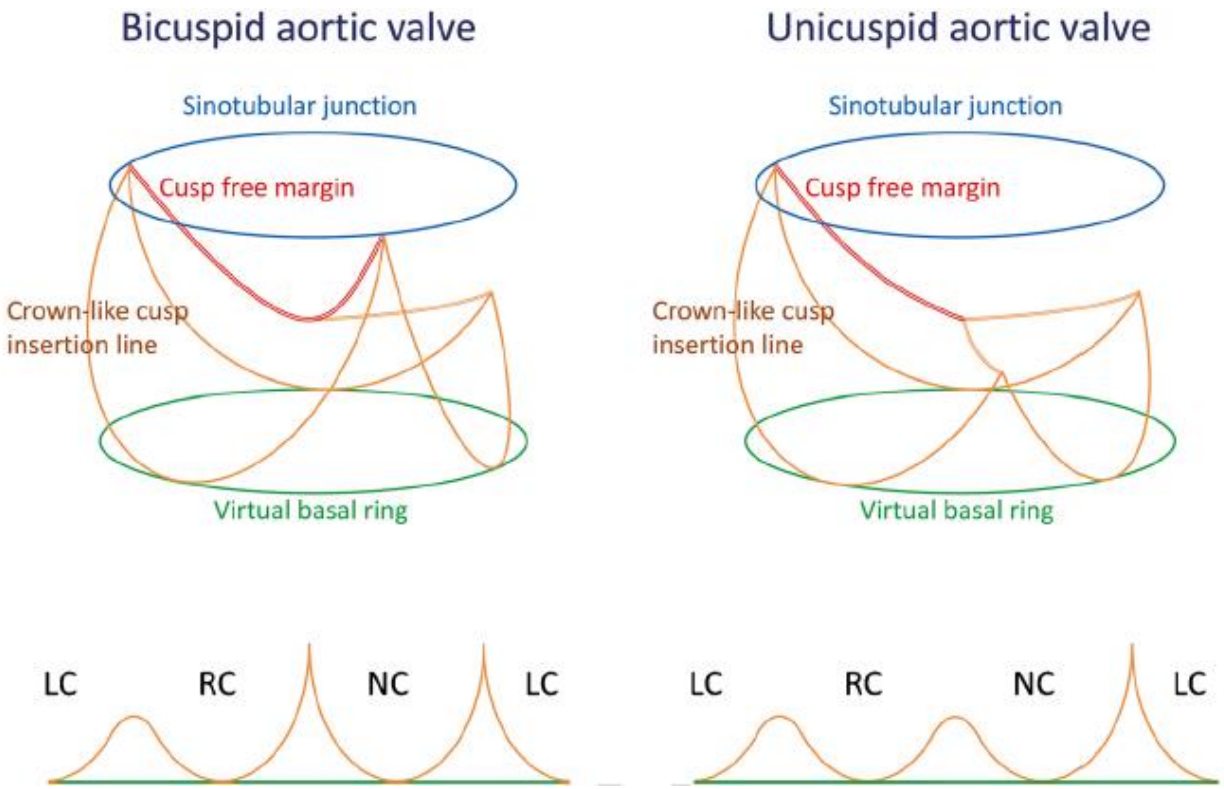
Javariah Siddiqui, MBBS, Christian P. Brizard, MD, Igor E. Konstantinov, MD, PhD, John Galati, BS, PhD, Gavin Wheaton, MD, Michael Cheung, MB ChB, Stephen Horton, PhD, and Yves d'Udekem, MD, PhD

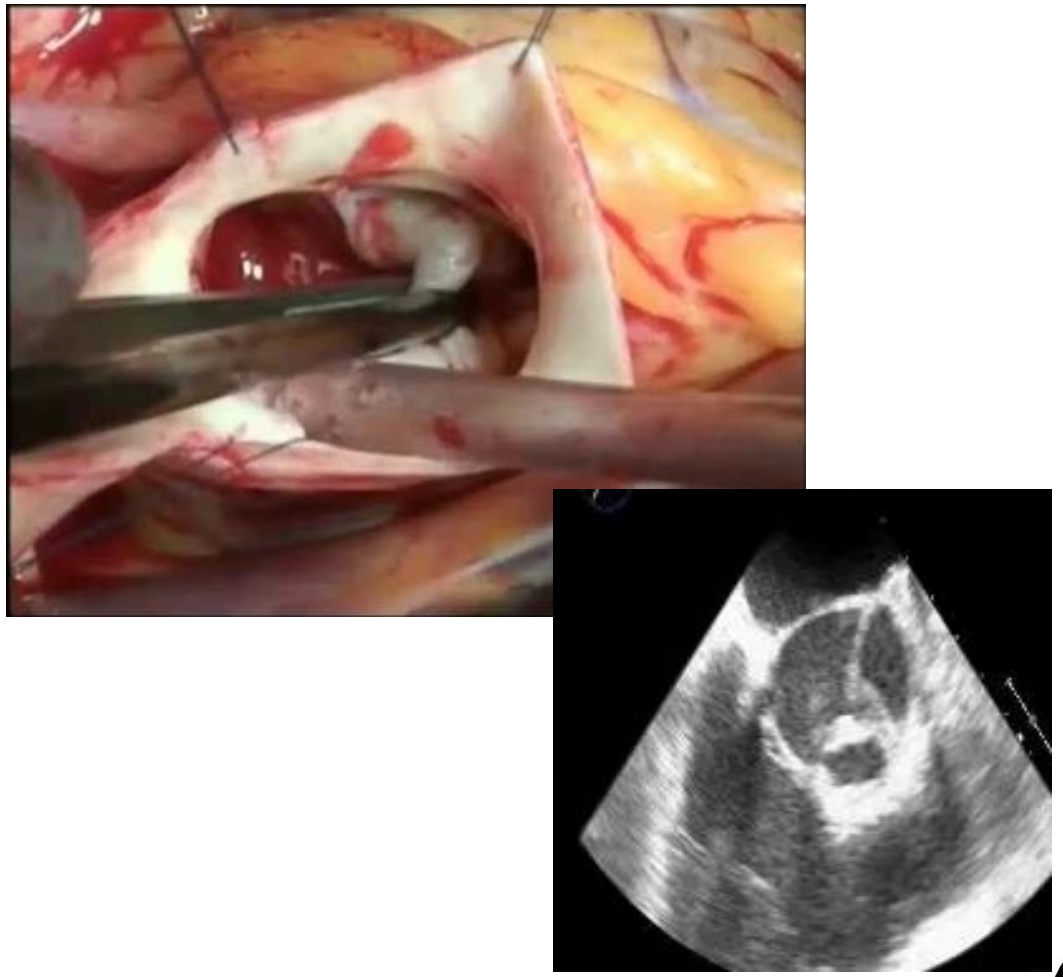


(Ann Thorac Surg 2013;96:2175–83)



Unicuspid Aortic Valve (UAV)





Matsushima S. Indian J Thorac Cardiovasc Surg 2019.

Tricuspidisation of the aortic valve with creation of a crown-like annulus is able to restore a normal valve function in bicuspid aortic valves☆

René Prêtre[a,*], Alexander Kadner[a], Hitendu Dave[a], Dominique Bettex[b], Michele Genoni[a]

Fig. 1. Superior view of a BAV. The fused leaflet is disinserted from annulus to the deepest point of the non-coronary leaflet. The fused leaflet is trimmed to create a 'normal' non-coronary leaflet. The right coronary leaflet is created with a patch of xenopericardium (L: left; R: right; NC: non-coronary).

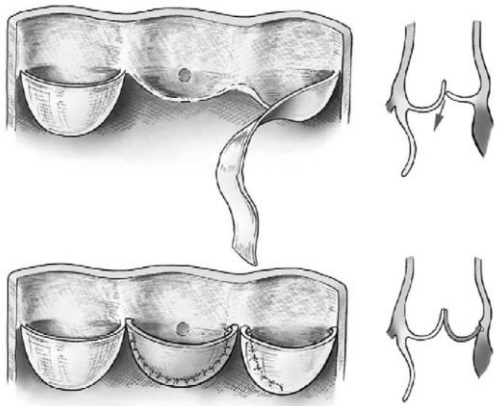


Fig. 2. Deployment of the aortic root showing the detachment of the fused leaflets and the re-attachment of a native and an artificial (dark gray) leaflet over a wider height, thus re-creating the crown-like annulus. Inserts right demonstrate the reason of the eccentric regurgitation (top) and the improved coaptation obtained with the annular remodelling.

Tricuspidisation of the aortic valve could be performed in 12 patients. During the same time period, we performed 46 Ross procedures, 8 other valve repairs and 3 conventional replacements in a similar population (younger than 35 years)

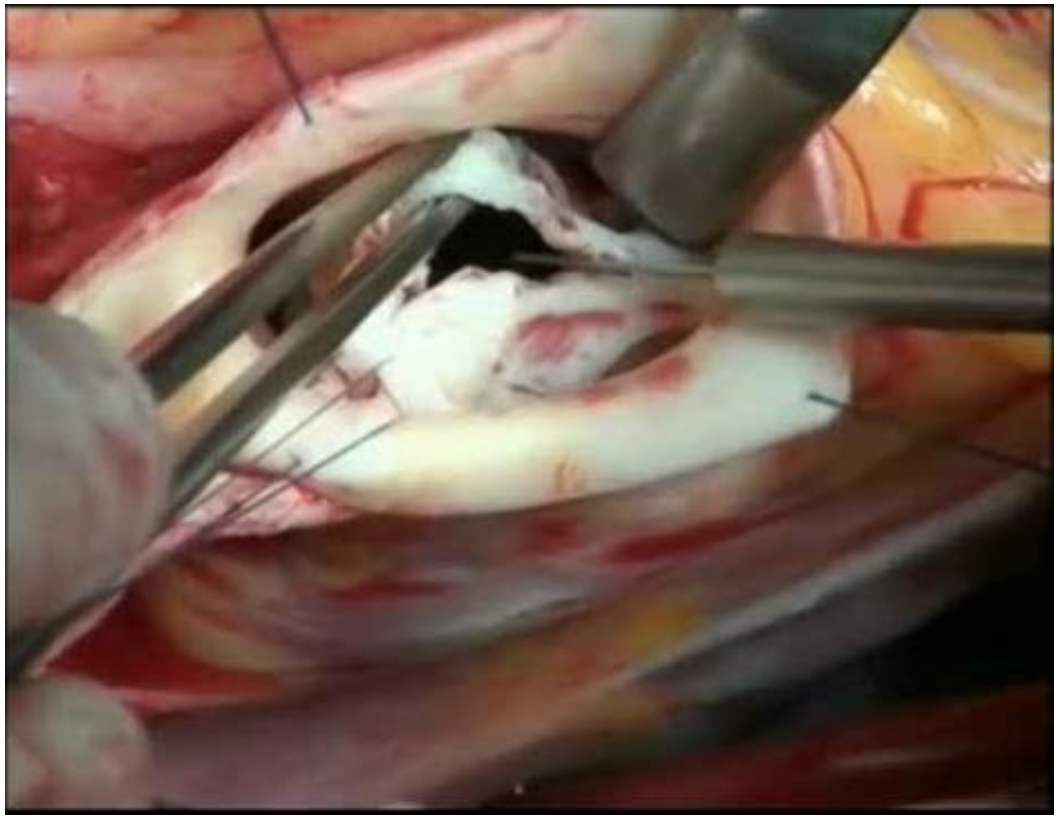
decided after disinsertion and modelling of the fused leaflets. The repair was interrupted in a more advanced phase and converted to a Ross procedure in four patients. In two of them, the conversion occurred after declamping of the aorta.

Tricuspidization (of BAV?) performed in 12 of 69 procedures

Conversion to replacement in 4 instances

European Journal of Cardio-thoracic Surgery 29 (2006) 1001—1007

Bicuspidization – the Next Step



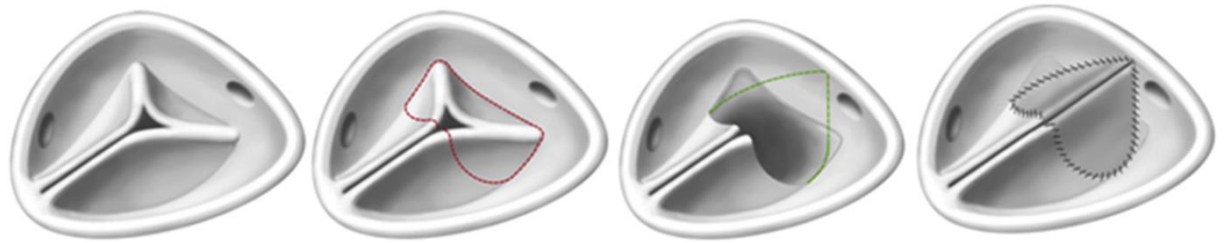
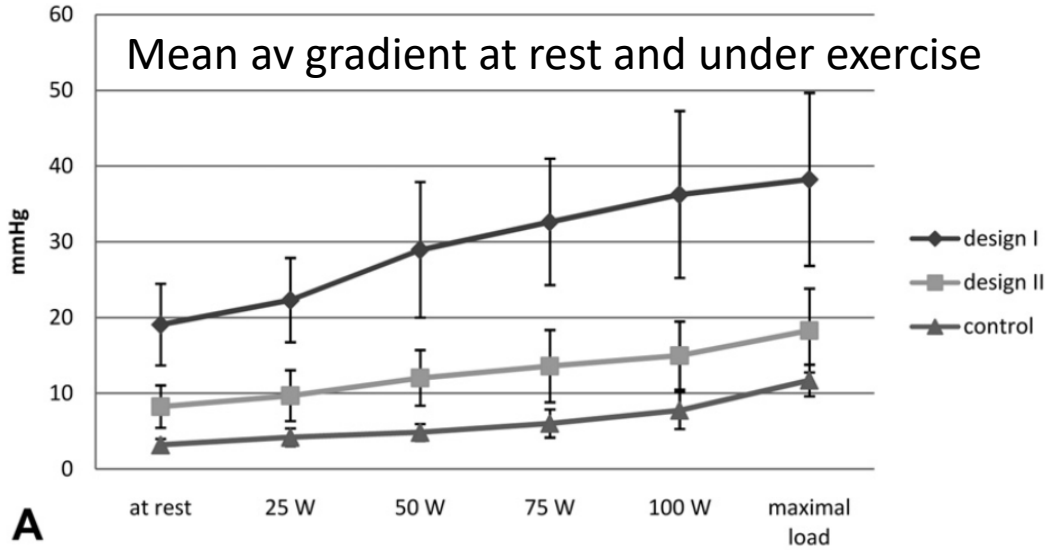


Fig 2. Bicuspidization of the unicuspid aortic valve: Design II.

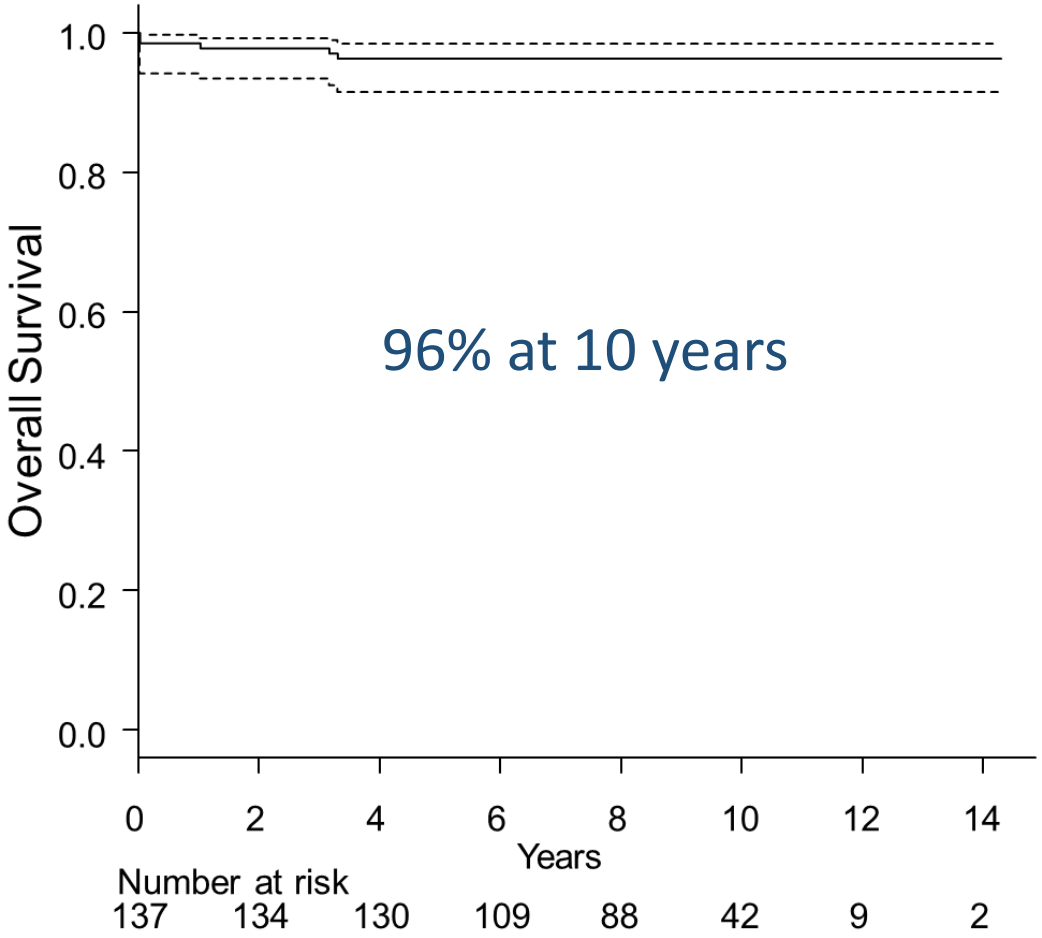


Aicher D. Ann Thorac Surg 2013.

Bicuspidization of the UAV

Variable	n = 137
Perfusion time, mean	80±23 min
Cross-clamp time, mean	60±18 min
Operative technique, n (%)	
Symmetric bicuspidization	112 (82)
External suture annuloplasty (Basal ring ≥ 26mm)	47 (34)
Concomitant procedure, n (%)	
Ascending aortic replacement	30 (22)
Root remodeling	20 (15)
Subaortic stenosis resection	4 (3)
Intraoperative conversion to valve replacement, n (%)	0 (0)

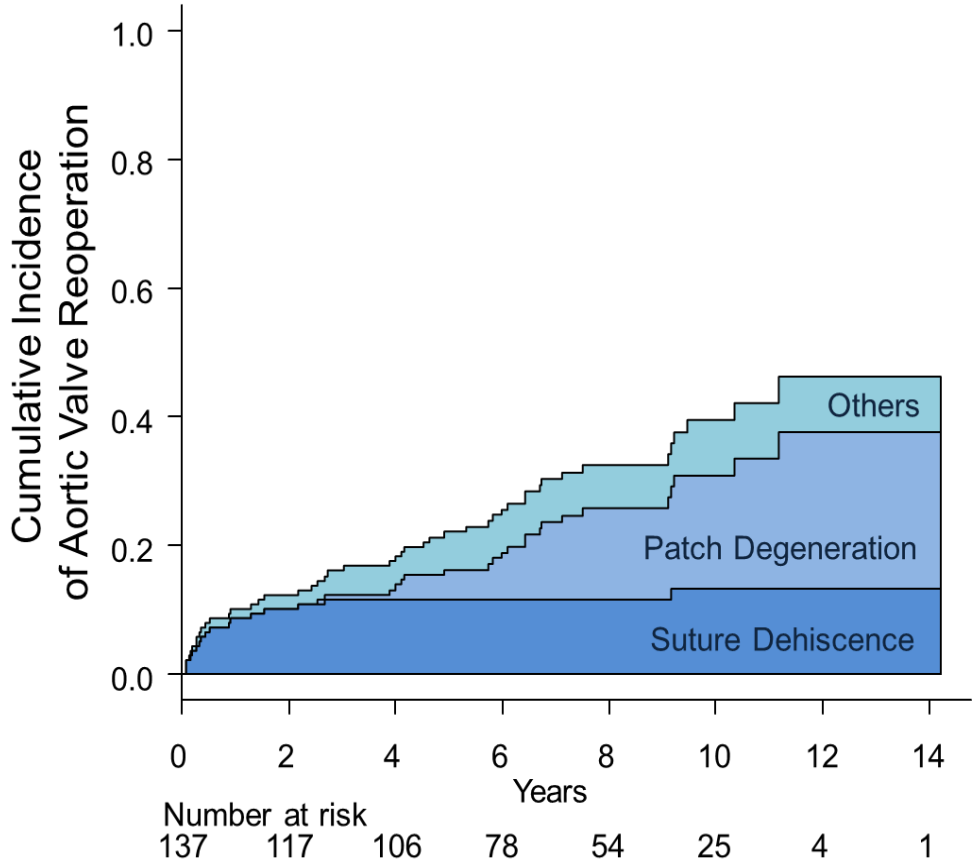
Survival



Causes of deaths

- Infective endocarditis (n=2)
- Ventricular arrhythmia (n=1)
- Traffic accident (n=1)
- Unknown (n=1)

Causes of Aortic Valve Failures (n=47)



<u>Patch degeneration (n=20)</u>

<u>Suture dehiscence (n=17)</u>

Subaortic stenosis (n=3)

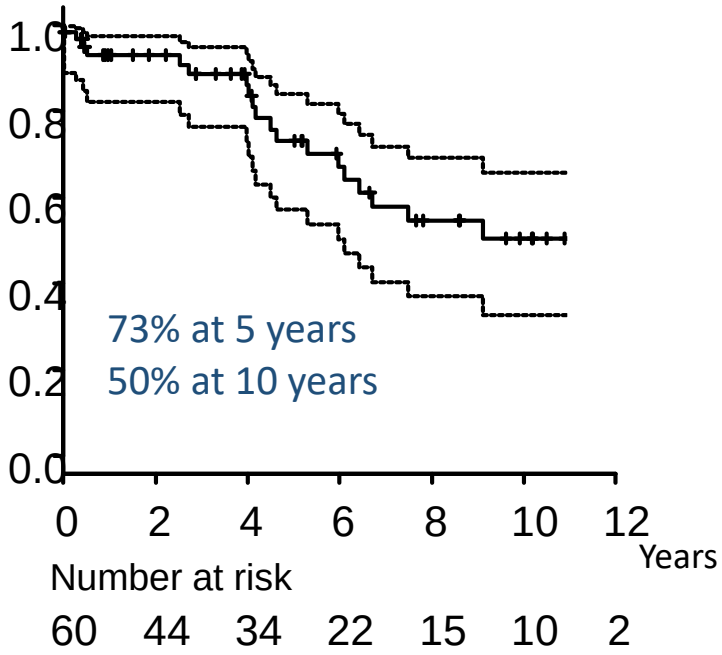
Infective endocarditis (n=3)

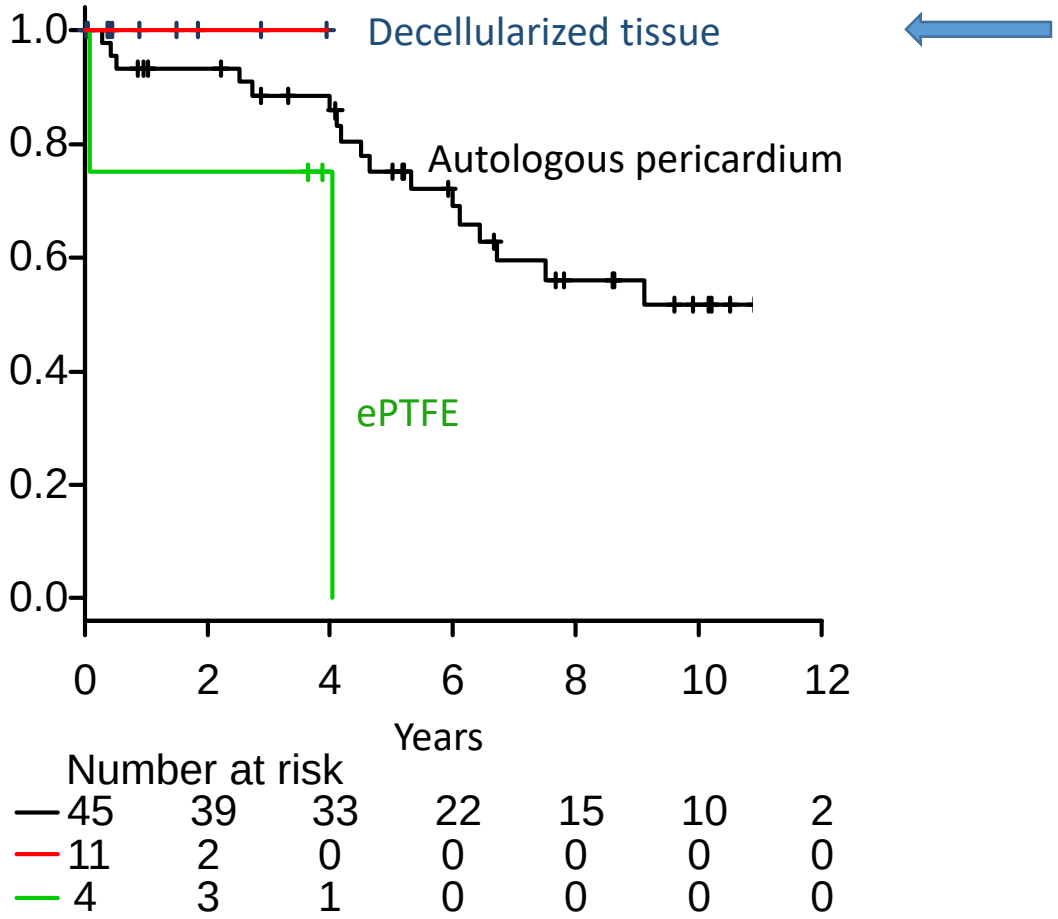
Tissue erosion by braided suture annuloplasty (n=2)

Aortic root dilatation (n=1)

Unknown (n=1)

UAV - Freedom from Reoperation





Conclusions

- The incidence/prevalence of UAV is grossly underestimated
- AV repair is an option for almost all congenital variants of the AV including the UAV
- Repair is associated with excellent survival. It postpones replacement and allows for growth and replacement in a low-risk scenario
- Bicuspidization of the UAV is a safe and reproducible repair approach with excellent survival. The “golden rule” (eH=0.45 x gH) also applies to that scenario.
- Patch degeneration is an unsolved problem with autologous pericardium (decellularized xenopericardial patch?).

Thank you for your attention

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