

42yr Old Male with Severe AR

Mild LV dysfunction s/p TOF

-AV Replacement(tissue valve) or AoV plasty-

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Current Guideline for AR s/p TOF

✓ Surgery is reasonable in adults with prior repair of TOF

Recommendations	COR	LOE
Residual RVOT obstruction (valvular or subvalvular) and Severe AR with associated symptoms or more than mild LV dysfunction - ACC/AHA 2008 Guidelines -	Ila	C
Significant aortic regurgitation associated with symptoms and/or progressive LV systolic dysfunction - Canadian Cardiovascular Society 2009 Consensus -	Ila	C

However, there was **no recommendation** related to the most appropriate type of surgery.

AORTIC VALVE REPLACEMENT AFTER REPAIR OF PULMONARY ATRESIA AND VENTRICULAR SEPTAL DEFECT OR TETRALOGY OF FALLOT

G. Alfred Dodds III, MD^a
Carole A. Warnes, MD, MRCP^a
Gordon K. Danielson, MD^b

Objective: Little attention has been paid to the occurrence of aortic regurgitation after complete repair in patients with pulmonary atresia and ventricular septal defect or tetralogy of Fallot. To highlight the development of aortic regurgitation or aortic root dilation severe enough to necessitate aortic valve replacement with or without aortic aneurysmorrhaphy or aortic root replacement, we retrospectively reviewed the records of patients who underwent aortic valve operation at our institution subsequent to repair of pulmonary atresia and ventricular septal defect or tetralogy of Fallot. **Methods:** We searched the Mayo Clinic database for patients with pulmonary atresia and ventricular septal defect or tetralogy of Fallot who subsequently had aortic valve or aortic root operations. The degree of aortic regurgitation before operation was noted. Aortic sinus and root dimensions were measured. **Results:** Sixteen patients underwent complete repair at a median age of 17 years, followed by an aortic operation a median of 13.5 years later. All 16 patients had dilated aortic sinuses at the time of the aortic valve operation. These 16 patients had aortic valve replacement: 11 with mechanical prostheses and 5 with bioprostheses. Five

The first series – progressive AR and aortic root dilatation after repair TOF or PA/VSD

16 pts – 10 mechanical AVR, 5 tissue AVR, 1 Bentall

Median age at the time of repair : 13.5yr

Conclusions : Progressive aortic regurgitation and aortic root dilation can occur despite complete repair of pulmonary atresia and ventricular septal defect or tetralogy of Fallot.



Prevalence and optimal management strategy for aortic regurgitation in tetralogy of Fallot

Toru Ishizaka^{a,*}, Hajime Ichikawa^a, Yoshiki Sawa^a, Norihide Fukushima^a, Koji Kagisaki^a, Haruhiko Kondo^a, Shigetoyo Kogaki^b, Hikaru Matsuda^a

Abstract

Objective: Aortic regurgitation (AR) in the tetralogy of Fallot (TOF) is not frequent, but when present it impacts significantly on surgical management. Furthermore, the incidence of late AR development has been increasing, along with surgical interest in current practices. **Methods:** Pre- and post-operative studies on 427 patients (TOF, 374; TOF/PA (TOF with pulmonary atresia), 53) who survived corrective operation were reviewed. AR (\geq mild) was detected in 28. **Results:** Nine had AR preoperatively, while 25 (including six with preoperative AR) exhibited AR post-operatively. In the 19 who developed AR post-operatively, the aortic root diameter (AoRoD) and indexed AoRoD (%AoRoD) were 42 ± 11 mm and $166 \pm 36\%$, increased from the preoperative values of 30 ± 10 mm and $149 \pm 24\%$. AR-free rate at 20 years

1968 – 2003, 427pts – surgical correction of TOF

25pts (5.85%) – (6pts - \geq mid AR before surgery, 19 pts – developed AR post-operatively)

The average interval between corrective surgery and the detection of AR : 12 ± 9 yrs

Absolute values for aortic root size : 42 ± 11 mm

Management : medical f/u (21pts), Operation (mechanical AVR 3pts, AVP 1pt)

Conclusions : After repair of TOF, careful observation for a late progression of AR is needed for the optimal timing of surgical intervention.....

Mechanism for Occurrence of AR

Progressive Aortic Root Dilatation in Adults Late After Repair of Tetralogy of Fallot

Koichiro Niwa, MD; Samuel C. Siu, MD, SM; Gary D. Webb, MD; Michael A. Gatzoulis, MD, PhD

216 pts, previously undergone reparative surgery for TOF

Group A : aortic root(AoRt) dilatation (n = 32) Group B : controls (n = 54)

Patients from group A had a longer shunt-to-repair interval ($P = 0.048$)

Conclusions : A subset of adult TOF exhibits ongoing dilatation of AoRt late after repair. This dilatation relates to **previous long-standing volume overload** of AoRt and possibly to intrinsic properties of AoRt and may lead to AR.

Mechanism for Occurrence of AR



Intrinsic Histological Abnormalities of Aortic Root and Ascending Aorta in Tetralogy of Fallot Evidence of Causative Mechanism for Aortic Dilatation and Aortopathy

J.L. Tan, MBBS, MRCP; P.A. Davlouros, MD; K.P. McCarthy, BSc;
M.A. Gatzoulis, MD, PhD; S.Y. Ho, PhD, FRCPath

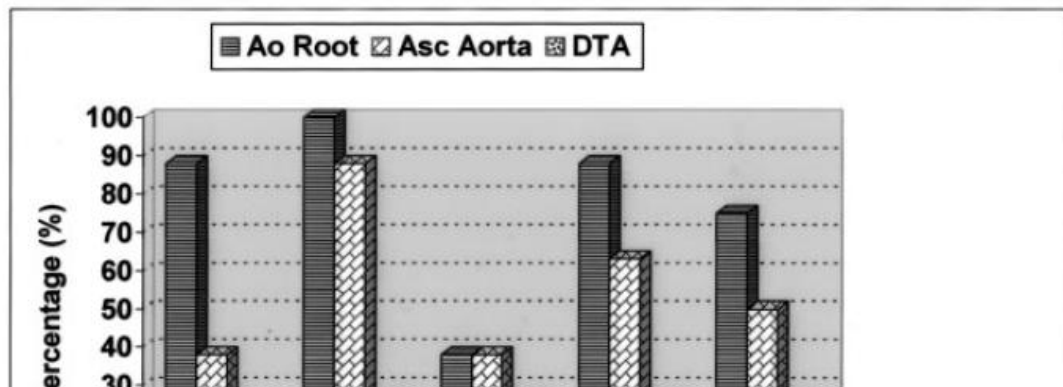


Figure 3. Abnormal histological features of grade 2 or 3 in the aortic root (at least 1 of the aortic sinuses), in the ascending aorta, and in the descending thoracic aortic wall of the adult TOF group (n=8). Asc A indicates ascending

Conclusions : There are marked histological abnormalities in the aortic root and ascending aortic wall of patients with TOF present from infancy, suggesting a causative mechanism for subsequent aortic root dilatation.

Mechanism for Occurrence of AR

- Previous volume overload of Aortic root +
Intrinsic properties of Aortic root (histologic abnormality)
- Aortic root dilatation (prevalence 15-87%)
- Progressive aortic valve regurgitation

Considering Factors for Valve Choice

- Aortic root dilatation
- History of cardiac operations
- State of the valve in the pulmonic position
- Life expectancy of TOF patients
- Possibility of pregnancy

Aortic root dilatation

- Valve-sparing operations
- The ideal operation for young patients



Aortic valve–sparing reimplantation for dilatation of the ascending aorta and aortic regurgitation late after repair of congenital heart disease

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J Thorac Cardiovasc Surg 2007;133:876-9

TABLE 1. Summary of data before valve-sparing operation

Conclusions : Aortic valve–sparing reimplantation is an effective treatment for **aortic root dilatation and aortic regurgitation after corrective repair of complex congenital heart disease**. This procedure leads to correction of aortic regurgitation, favorable durability of the native aortic valve, and excellent hemodynamics, **avoiding a prosthetic valve and anticoagulation**.

Aortic root dilatation

Valve-Sparing Replacement of the Aortic Root after Repair of Tetralogy of Fallot

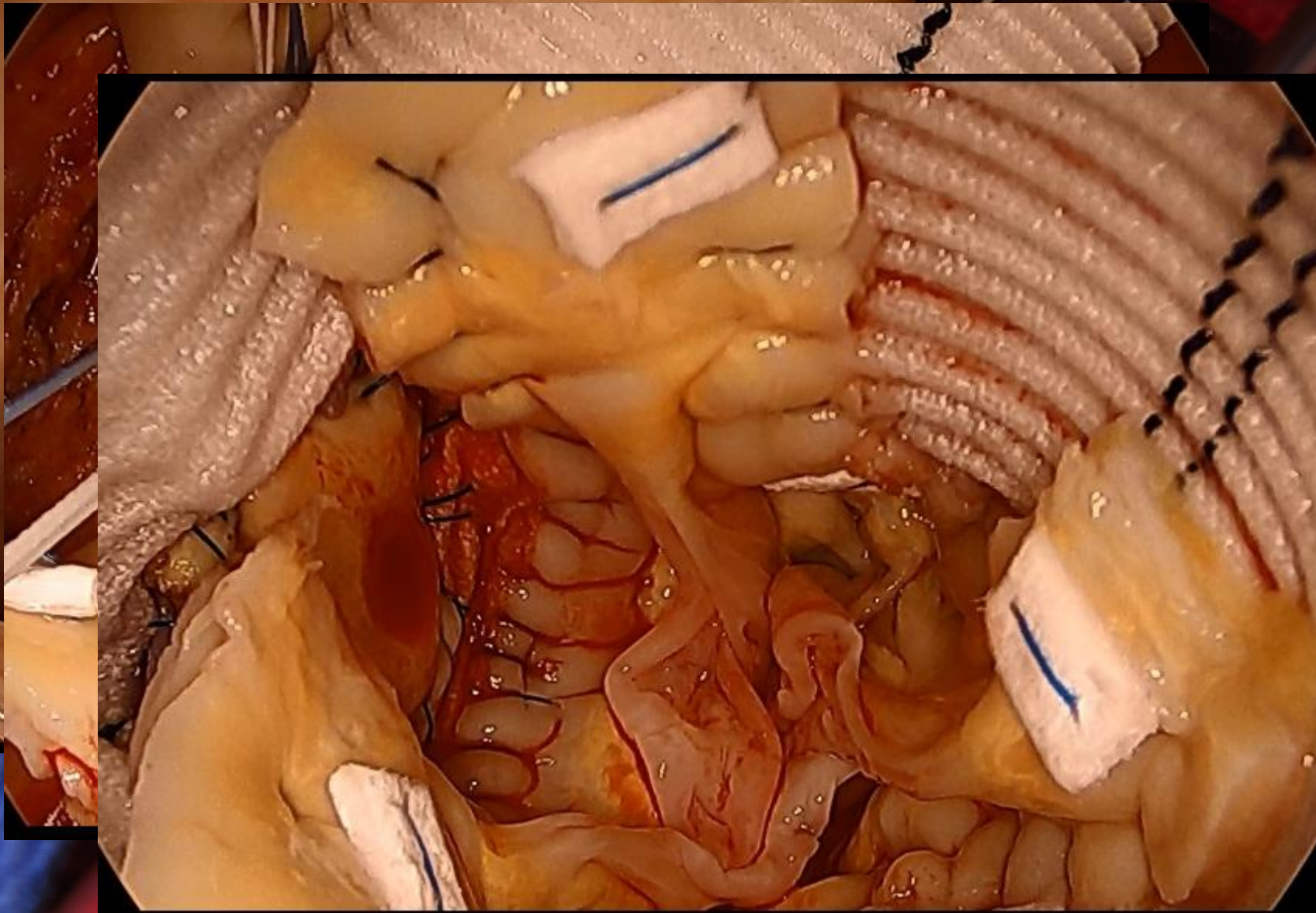
Yoshio Ootaki, MD, PhD,¹ Ryuichi Kuromaru, MD,² and Ross M. Ungerleider, MD¹

Ann Thorac Cardiovasc Surg 2013;19:170-2

Progressive aortic root dilatation is a common feature after surgical repair of tetralogy of Fallot. This report describes a successful valve-sparing replacement of the aortic root in a patient with significant dilated aortic root and aortic regurgitation after repair of tetralogy of Fallot.

16-year-old male, aortic root =40mm, moderate AR

However, it is not easy to perform valve sparing operation for severe AR. Because reoperation due to remnant or recurrent AR would be very risky.



History of Cardiac operation

- ✓ B-T shunt, total repair, pulmonary valve replacement, Permanent pacemaker insertion, etc.
- ✓ Risk of redo-cardiac surgery
 - **Sutureless AVR** : Shortening the op time (CPB and ACC time), Edwards INTUITY > Sorin Perceval S
 - TAVI : Inappropriate approach considering the aortic root dilatation as the first treatment → However, good alternative methods for NSVD or SVD in replaced aortic valve

State of valve in Pulmonic Position

- Native valve or bioprosthesis
 - Consider the risk of anticoagulation and the other factors (RV function, possibility of PV intervention in the future)
- Mechanical valve in pulmonic position
 - Mechanical valve > Bioprosthesis
 - Valve click sounds: uncomfortable and annoying

Life Expectancy

- Current guideline for mechanical valve

Recommendations	COR	LOE
A mechanical prosthesis is reasonable for AVR or MVR in patients <60 y of age who do not have a contraindication to anticoagulation - 2014 AHA guideline -	IIa	B

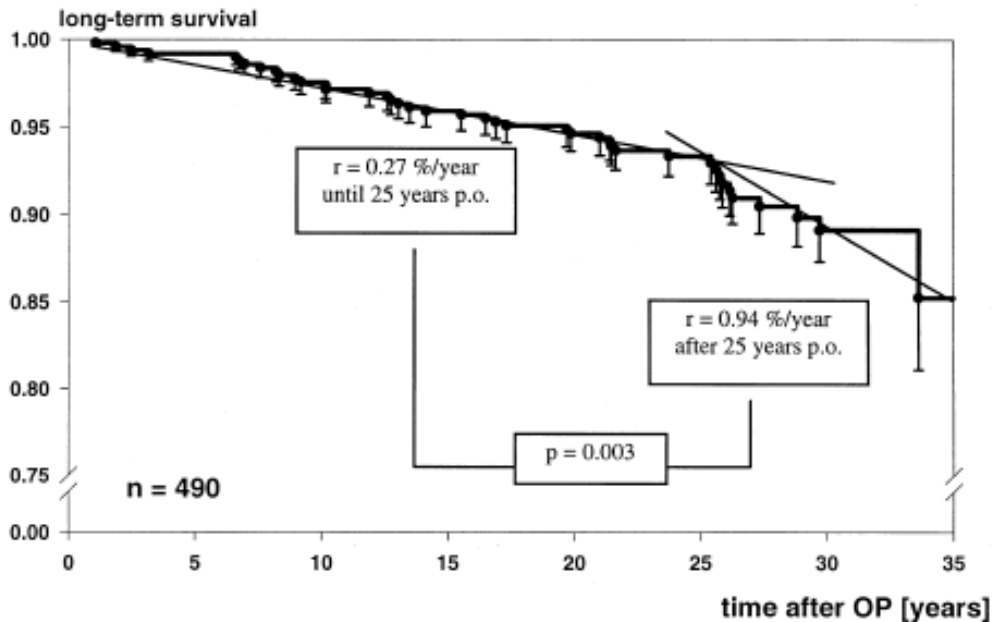
- life expectancy in 60yr : 25yrs (Korea)

<http://kosis.kr>

- ✓ What is the life expectancy of 42 year-old patient
underwent repair of TOF?

Life Expectancy

- Overall survival of TOF patients



Repair of Tetralogy of Fallot: of the First Year After

STEFAN BOUTERWEK, DMD,
RUNO REICHART, MD

89% and 85%

Figure 2. Long-term survival after correction of TOF. All patients who died within the first year after correction were excluded for calculation of long-term survival. The curve shows two different phases that are distinct. The early, low risk phase lasts 25 years; thereafter, the risk increases significantly. Mortality risk (r) per year, as a linearized number, is calculated for each phase. Note the break in the y axis. OP = operation; p.o. = postoperatively.

Mortality increased 25 years postoperatively from **0.27%/year** to **0.94%/year** ($p = 0.003$)

Life Expectancy

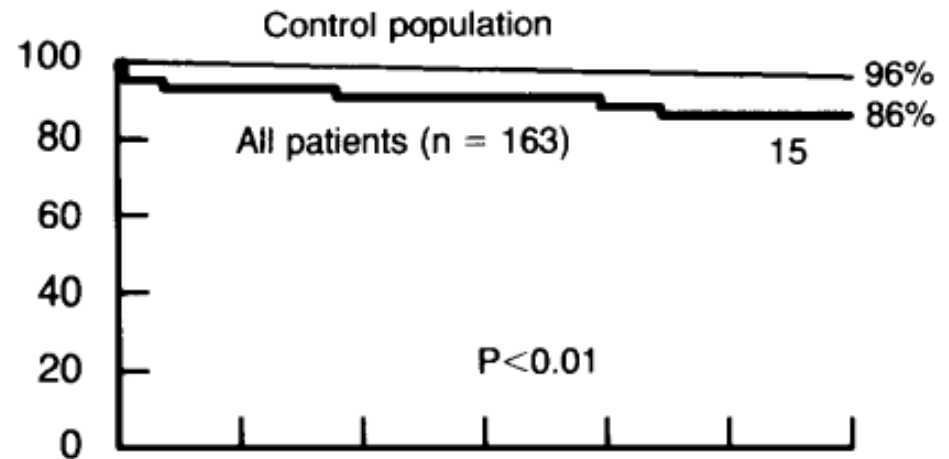
LONG-TERM OUTCOME IN PATIENTS UNDERGOING SURGICAL REPAIR OF TETRALOGY OF FALLOT

JOSEPH G. MURPHY, M.D., M.R.C.P.I., BERNARD J. GERSH, M.B., CH.B., D.PHIL., DOUGLAS D. MAIR, M.D.,
VALENTIN FUSTER, M.D., MICHAEL D. MCGOON, M.D., DUANE M. ILSTRUP, M.S.,
DWIGHT C. MCGOON, M.D., JOHN W. KIRKLIN, M.D., AND GORDON K. DANIELSON, M.D.



Overall 30year survival : 86%

Conclusions : Among patients with surgically repaired tetralogy of Fallot, the long-term survival after the postoperative period is excellent but **remains lower than that in the general population.**



Possibility of Pregnancy

✓ Anticoagulation (Warfarin : Category X)

Category X

Studies in animals or humans have demonstrated fetal abnormalities and/or there is positive evidence of human fetal risk based on adverse reaction data from investigational or marketing experience, and the risks involved in use of the drug in pregnant women clearly outweigh potential benefits.

Example drugs: atorvastatin, simvastatin, warfarin, methotrexate, finasteride

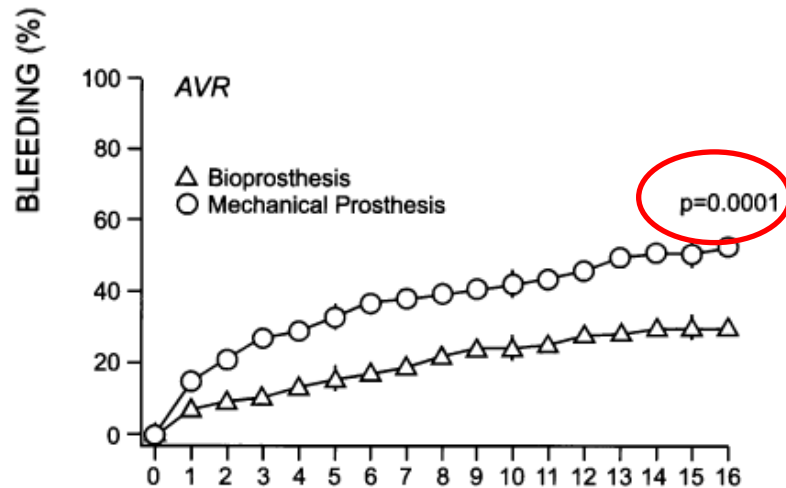
- Risk of fetal abnormalities
- Risk of thromboembolism for changing to LMWH

Risk of Anticoagulation



Outcomes 15 Years After Valve Replacement With a Mechanical Versus a Bioprosthetic Valve: Final Report of the Veterans Affairs Randomized Trial

1977 – 1982, 575pts, randomized trial
Bjork-Shiley valve vs Hancock valve



Risk of Anticoagulation

Twenty year comparison of a Bjork-Shiley mechanical heart valve with porcine bioprostheses

H Oxenham, P Bloomfield, D J Wheatley, R J Lee, J Cunningham, R J Prescott, H C Miller

1975 – 1979

Bjork-Shiley valve vs porcine prostheses

	Aortic valve replacement		
	10 Years	20 Years	p Value*
Bleeding: all episodes			
Bjork-Shiley valve	16.3 (4.2)	61.1 (7.6)	0.001
Porcine valve	5.9 (2.9)	42.4 (12.1)	
Bleeding: major episodes			
Bjork-Shiley valve	12.2 (3.7)	37.8 (7.1)	0.021
Porcine valve	4.2 (2.4)	32.0 (12.6)	

Risk of Anticoagulation



Aortic Valve Replacement

A Prospective Randomized Evaluation of Mechanical Versus Biological Valves in Patients Ages 55 to 70 Years

1995 – 2003, randomized trial, 310pts

Bioprosthetic aortic valve vs Mechanical aortic valve

Table 4 Linearized Rate of Valve-Related Events

MP (n = 149) BP (n = 147)

Thromboembolism
Bleeding
Endocarditis
Valve dysfunction
Valve thrombosis
Nonstructural valve dysfunction
Reoperation

Conclusions : a similar survival rate as well the same rate of occurrence of thromboembolism, bleeding.....

→ However, they should calculate the **composite occurrence rate of thromboembolism and bleeding**

CI = confidence interval, valve dysfunction as in Table 1

Risk of Anticoagulation

Patient outcome after aortic valve replacement with a mechanical or biological prosthesis: Weighing lifetime anticoagulant-related event risk against reoperation risk



1982 – 2003, 3934pts

Bioprosthesis (73%) vs Mechanical prosthesis (27%)

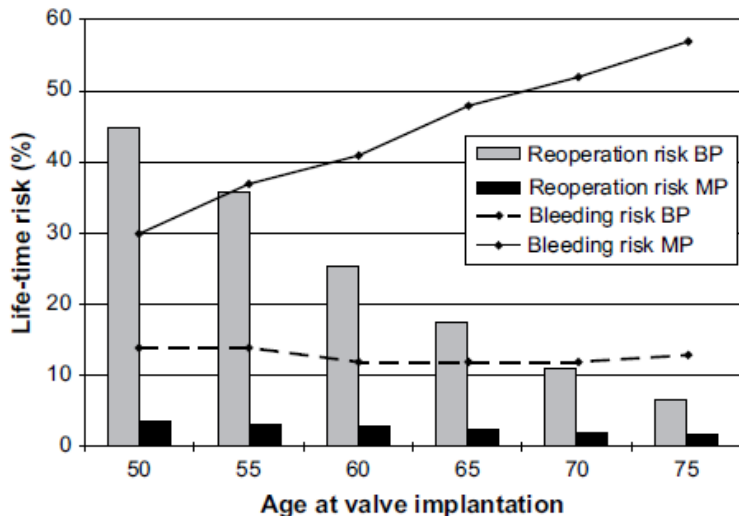


FIGURE 2. Lifetime risks of reoperation and bleeding after AVR with mechanical and bioprostheses. BP, Bioprosthesis; MP, mechanical prosthesis.

Rapid Deployment Valve

EDWARDS INTUITY Elite

VALVE SYSTEM



EDWARDS INTUITY Elite Valve System



EDWARDS INTUITY Elite Valve System

Proven Pericardial
Technology

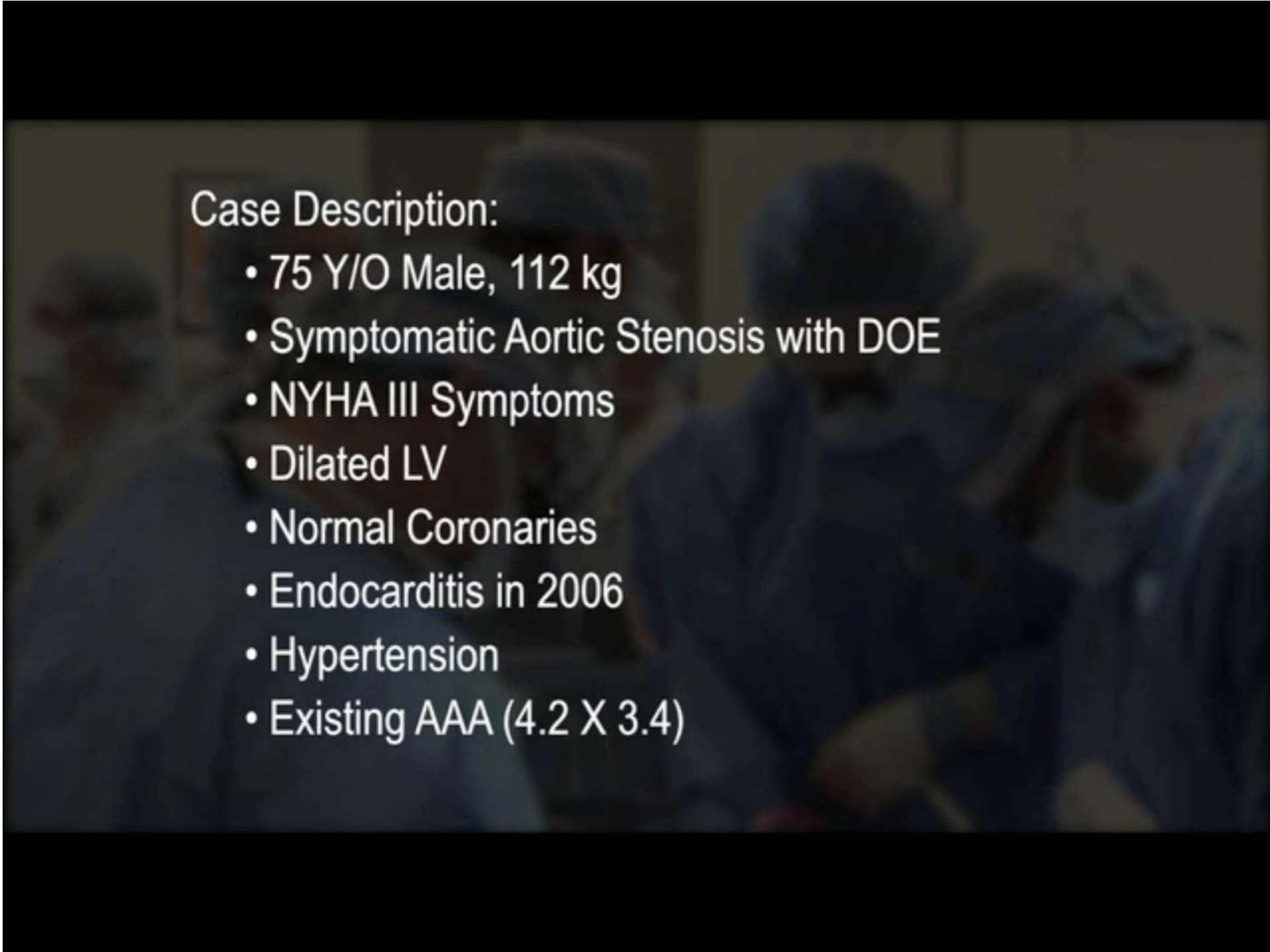
New
Innovations



EDWARDS INTUITY Elite Valve System combines..

- Proven Pericardial valve technology (Perimount Magna Ease valve)
- Innovation in transcatheter heart valve (Sapien valve)

***Rapid Deployment System
with 3 guiding sutures &
Balloon expandable system.***



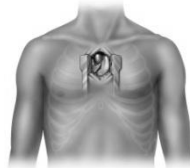
Case Description:

- 75 Y/O Male, 112 kg
- Symptomatic Aortic Stenosis with DOE
- NYHA III Symptoms
- Dilated LV
- Normal Coronaries
- Endocarditis in 2006
- Hypertension
- Existing AAA (4.2 X 3.4)

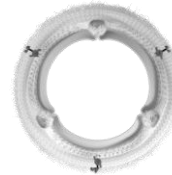
EDWARDS INTUITY Elite Valve System Clinical Data



Procedure Times



Smaller Incision



Hemodynamics



PVLs / Pacemaker

CADENCE-MIS

41.3 min XCT MIS RDAVR,
p<0.0001 vs. FS AVR

Bochum

26 min XCT MIS RDAVR,
9 min implantation time

Leipzig

39 min XCT MIS RDAVR;
42 min XCT in all pts

CADENCE-MIS

Proven time benefit in MIS,
comparable outcomes

Bochum

100% MIS AVR, very short
XCT, excellent outcomes

Leipzig

72% MIS AVR, short XCT,
excellent outcomes

TRITON 3-Yr

8.7 mm Hg @ 3 years,
n=287

CADENCE-MIS

Statistically better gradients
than conventional valves

Leipzig

8.8 mm Hg @ discharge,
n=69

TRITON 3-Yr

0.7% early PVLs (>2+);
1.2% late PVLs

CADENCE-MIS

0% early PVLs (>2+);
4.3% (2) pacemaker

Bochum

1.7% (1) early PVLs (>1+)
and pacemaker; 0% late

Conclusions

The choice of valve for AR with the previous repaired TOF should be **individualized** according to the aortic root dilatation, history of cardiac operations, state of the valve in pulmonic position, age and possibility of pregnancy

Even if the root replacement do not perform, the clinical course might be less malignant compared to Marfan's Syndrome

Conclusions

The aortic valve sparing operations could be considered in 42-year old patient. Of course, comprehensive information including complications of valve sparing operations, the problems of prosthetic valve, risk of anticoagulation and possibility of TAVI should be provided for the patients.

If the patient decided to undergo ascending aorta replacement with supracoronary AVR using tissue valve, rapid deployment valve could be a good option in terms of decreasing of surgical risk.