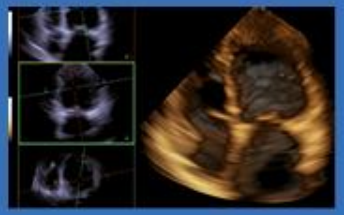
EuroValve
January 26-27, 2017

Pulmonary valve : indication for treatment

Agnès Pasquet, MD, PhD
Pôle de Recherche Cardiovasculaire
Institut de Recherche Expérimentale et Clinique
Université catholique de Louvain



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Faculty disclosure

Agnes Pasquet

I have **no financial relationships** to disclose.

But the hospital where I'm working receive research grant from:
Daichi Sankyo
Astra Zeneca

Guidelines ?

Guidelines on the management of valvular heart disease (version 2012)

The Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS)

Guidelines ?

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These guidelines focus on acquired VHD, are oriented towards management, and do not deal with endocarditis or congenital valve disease, including pulmonary valve disease, since recent guidelines have been produced by the ESC on these topics.[10,11]

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Guidelines ?

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

Guidelines:

ESC Guideli gement of grown-up disease (new version

The Task Force or rown-up Congenital Heart Disease of the Eur ology (ESC)

Endorsed by the Assoc ic Cardiology (AEPC)

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Pulmonary valve disease ?

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ESC Guidelir **gement**

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ngenital Heart

(AEPC)

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Pulmonary valve disease:

Congenital	Pulmonary valve stenosis
	Pulmonary atresia
	Supravalve pulmonary stenosis
	Infundibular pulmonary stenosis
	Idiopathic pulmonary artery dilatation
	Anomalous origin of coronary artery from pulmonary trunk
	Coronary arteriovenous fistula
Acquired	Rheumatic valve disease
	Infective endocarditis
	Carcinoid heart disease
	Tumors
Iatrogenic	Homograft dysfunction following Ross operation
	Homograft reconstruction for total correction of:
	Pulmonary atresia
	Complex form of Tetrology of Fallot
	Common arterial trunk
	Transposition of great arterus with Pulmonary stenosis
	Pulmonary regurgitation following total correction of Tetrology of Fallot or following balloon valvotomy.

Mainly a congenital disease !
7–12% of all congenital heart defects,
80–90% of all Right ventricular outflow tract obstruction.

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Pulmonary stenosis:

Indications	Class[a]	Level[b]
RVOTO at any level should be repaired regardless of symptoms when Doppler peak gradient is >64 mmHg (peak velocity >4m/s), provided that RV function is normal and no valve substitute is required	I	C
In asymptomatic patients in whom balloon valvotomy is ineffective and surgical valve replacement is the only option, surgery should be performed in the presence of a systolic RVP >80 mmHg (TR velocity >4.3 m/s)	I	C
Intervention in patients with gradient <64 mmHg should be considered in the presence of: • symptoms related to PS or, • decreased RV function or, • double-chambered RV (which is usually progressive) or, • important arrhythmias or, • right-to-left shunting via an ASD or VSD.	IIa	C

Ballon valvuloplasty is the preferred treatment method

ESC guidelines GUCH, 2010

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Ballon valvuloplasty is the preferred treatment method

No reference to severe PS !

ESC guidelines GUCH, 2010

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What is a severe pulmonary stenosis ?:

	Mild	Moderate	Severe
Peak velocity m/s	<3	3-4	>4
Peak gradient	<36	36-64	> 64

Baumgartner et al, Eur J Echocardiography (2009) 10, 1–25

Stage	Definition	Valve Anatomy	Valve Hemodynamics	Hemodynamic Consequences	Symptoms
C, D	Severe PS	• Thickened, distorted, possibly calcified leaflets with systolic doming and/or reduced excursion • Other anatomic abnormalities may be present, such as narrowed RVOT	• V_{max} >4 m/s; peak instantaneous gradient >64 mm Hg	• RVH • Possible RV, RA enlargement • Poststenotic enlargement of main PA	• None or variable and dependent on severity of obstruction

AHA ACC guidelines valve disease,2008

Pulmonary stenosis:

Balloon valvotomy is recommended for asymptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg (in association with less than moderate pulmonic valve regurgitation).	I	B
Balloon valvotomy is recommended for symptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg (in association with less than moderate pulmonic regurgitation).	I	C
Surgical therapy is recommended for patients with severe PS and an associated hypoplastic pulmonary annulus, severe pulmonary regurgitation, subvalvular PS, or supravalvular PS. Surgery is also preferred for most dysplastic pulmonary valves and when there is associated severe TR or the need for a surgical Maze procedure.	I	C
Balloon valvotomy may be reasonable in asymptomatic patients with a dysplastic pulmonary valve and a peak instantaneous gradient by Doppler greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg.	IIb	C
Balloon valvotomy may be reasonable in selected symptomatic patients with a dysplastic pulmonary valve and peak instantaneous gradient by Doppler greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg.	IIb	C

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Balloon valvotomy may be reasonable in selected symptomatic patients with a dysplastic pulmonary valve and peak instantaneous gradient by Doppler greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg.	IIb	C

Pulmonary stenosis:

Balloon valvotomy is not recommended for asymptomatic patients with a peak instantaneous gradient by Doppler less than 50 mm Hg in the presence of normal cardiac output.	III	C
Balloon valvotomy is not recommended for symptomatic patients with PS and severe pulmonary regurgitation	III	C
Balloon valvotomy is not recommended for symptomatic patients with a peak instantaneous gradient by Doppler less than 30 mm Hg.	III	C

AHA ACC guidelines GUCH, 2008

Pulmonary stenosis:

Symptomatic patients: intervention if

- PG> 64/60 mmHg Mean: > 40 mmHg
- selected pts 50 mmHg (US)

Asymptomatic patients: intervention if

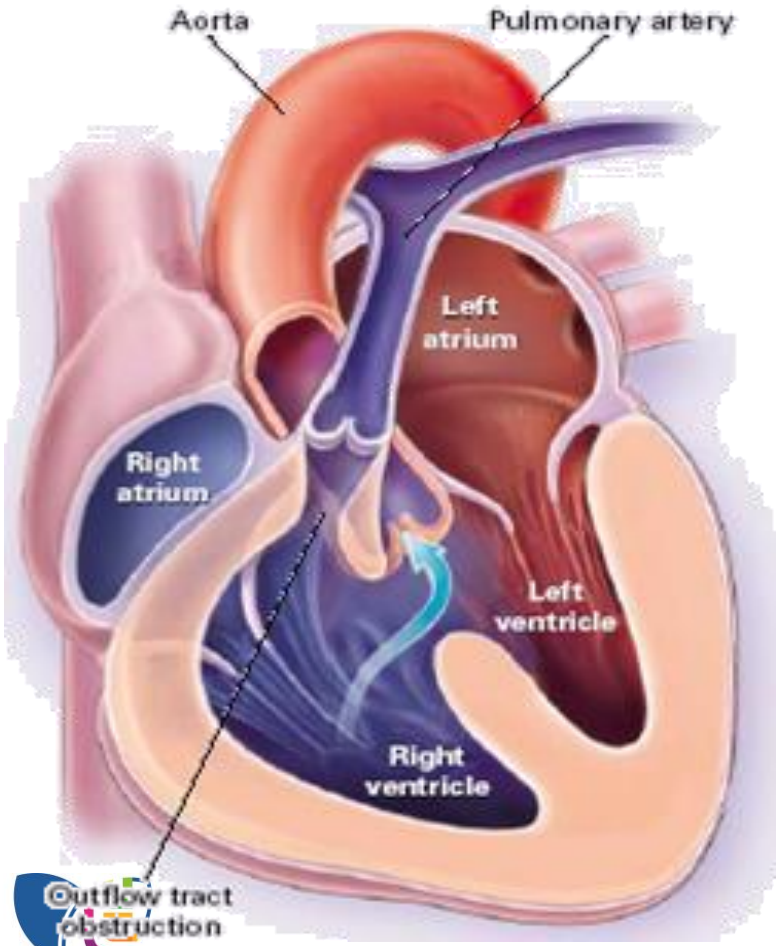
PG > 80 mmHg (TR: >4.3m/s) (EU)

PG > 60 mmHg/40 (US),

no intervention if PG<50 mmHg

EU: consider symptoms RV function, arythmia if lower gradient.

Tetralogy of Fallot:



- *4 features:*
- VSD (non-restrictive)
- Overriding aorta (but ,50%)
- RVOTO : infundibular,valvular, or (usually) a combination of both, with or without supravalvular or branch PA stenosis
- RVH.
- 6-11% all cardiopathies

Tetralogy of Fallot:

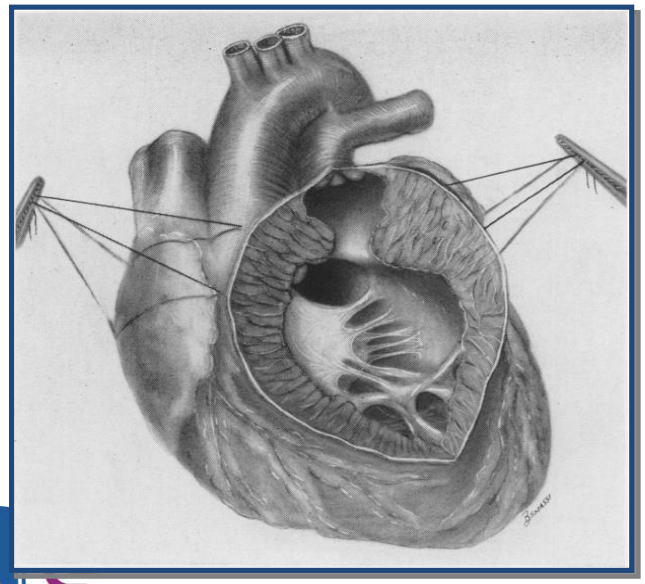
Direct Vision Intracardiac Surgical Correction of the Tetralogy of Fallot, Pentalogy of Fallot, and Pulmonary Atresia Defects

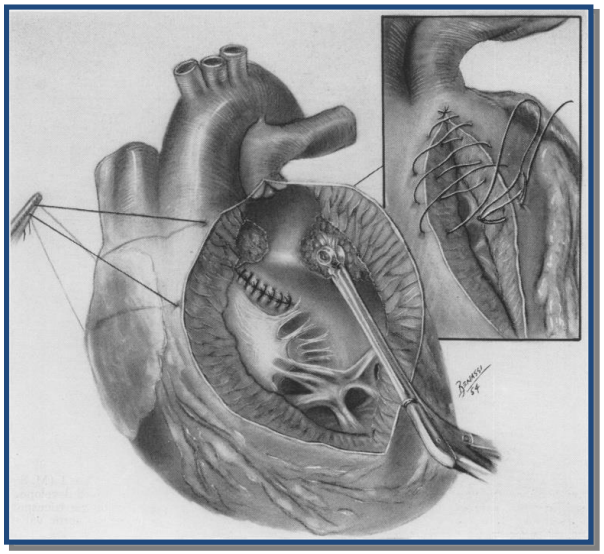
Report of First Ten Cases*

C. Walton Lillehei, M.D., Morley Cohen, M.D., Herbert E. Warden, M.D., Raymond C. Read, M.D., Joseph B. Aust, M.D., Richard A. DeWall, M.D., and Richard L. Varco, M.D.

Minneapolis, Minn.

Annals of surgery 1955, 142:418 - 442





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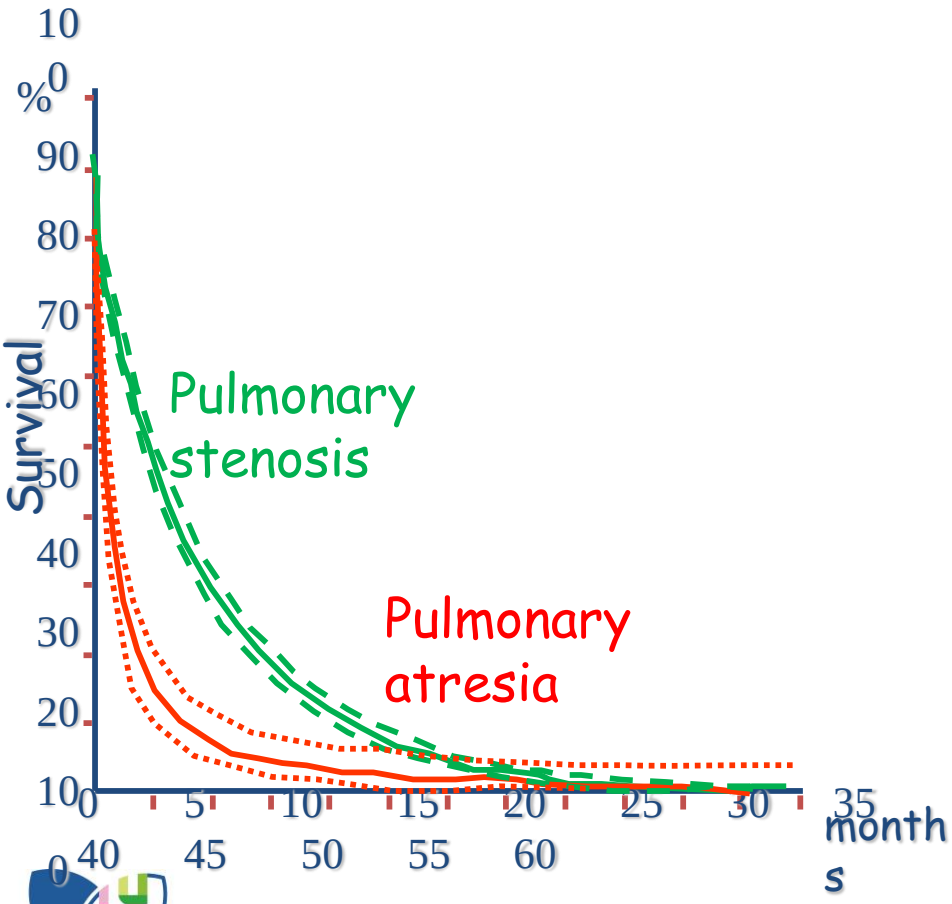

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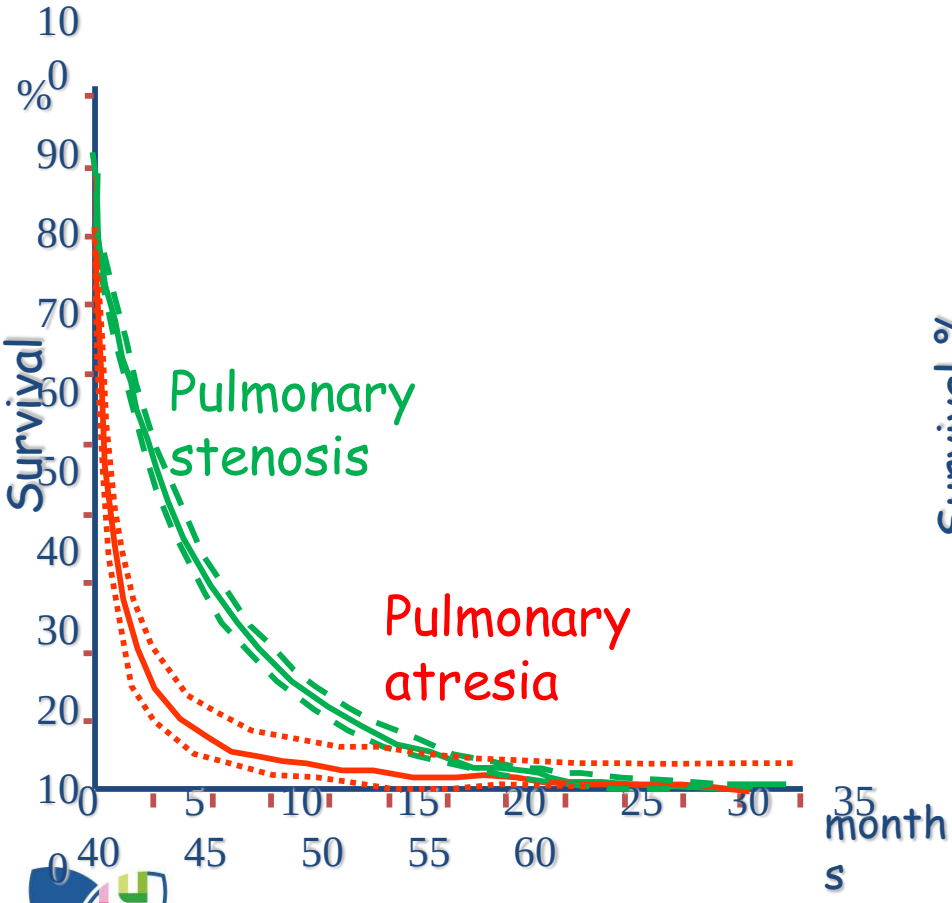
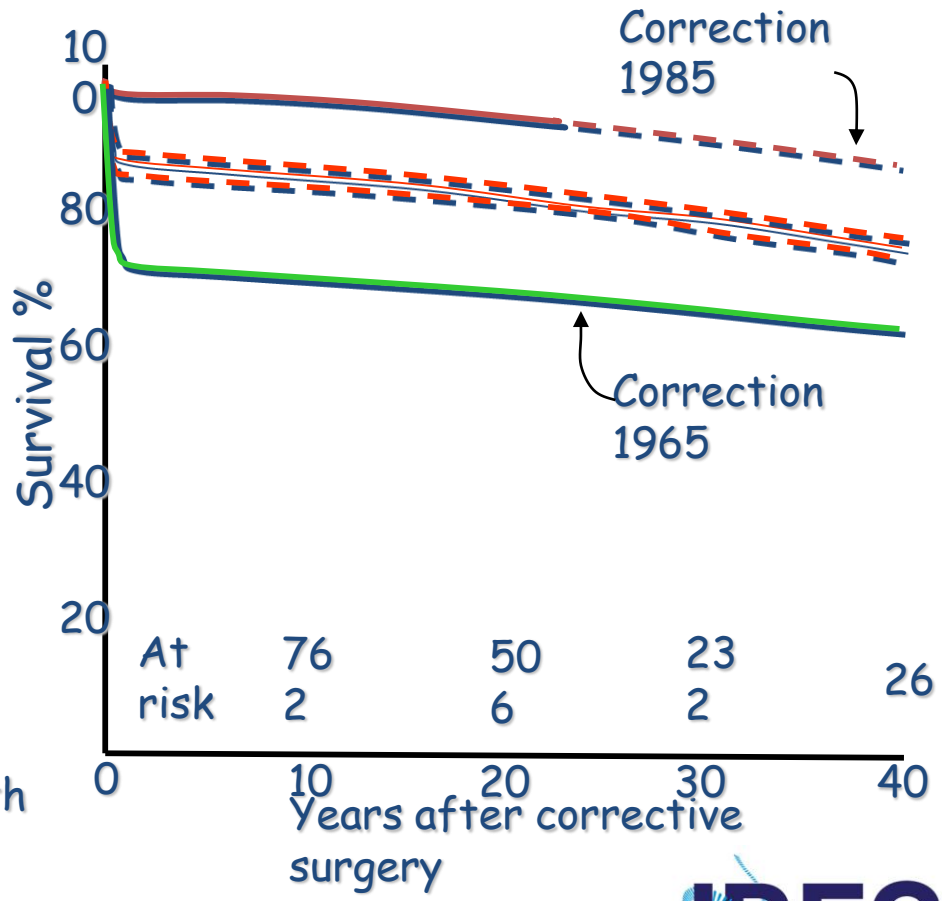
Hickey et al , Eur J Thorac Surg 2009;35: 156-166

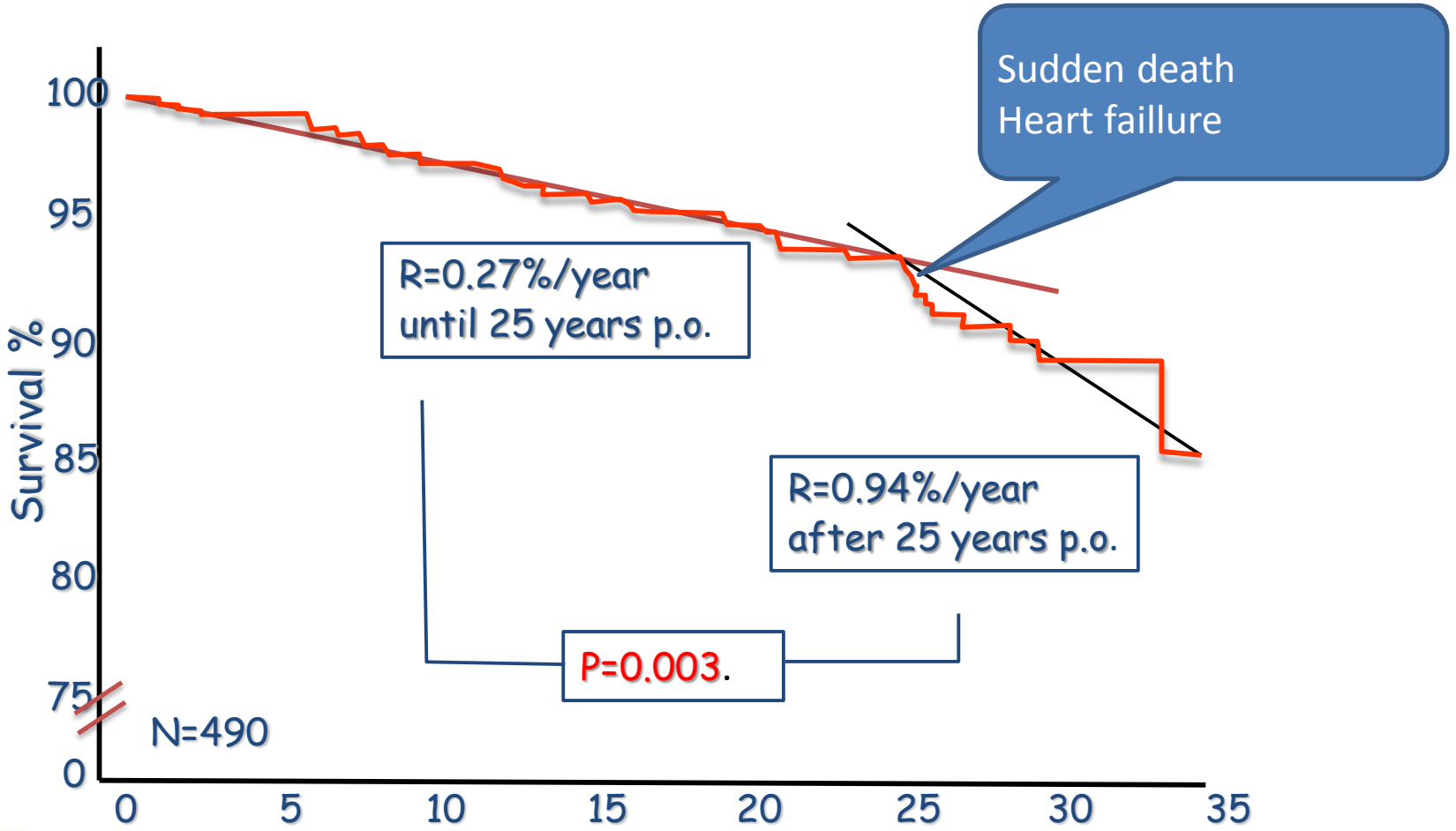
Tetralogy of Fallot:





Hickey et al , Eur J Thorac Surg 2009;35: 156-166

But.....

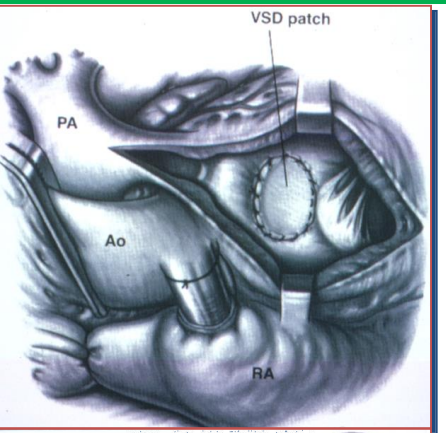


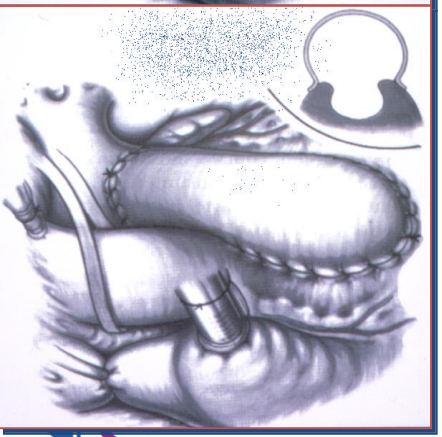
Nollert et al , JACC 1997;30: 1374-1383

Long term FU Tetralogy:

Surgery « patch »

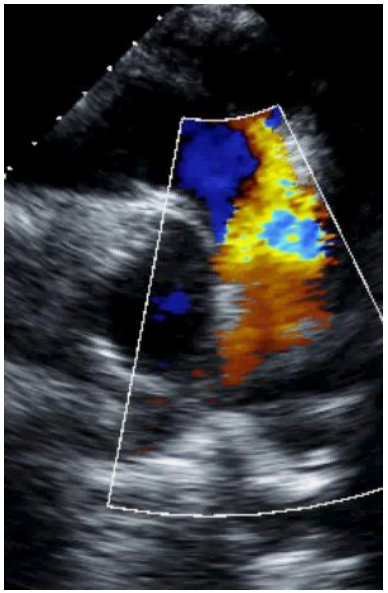
RV outflow tract



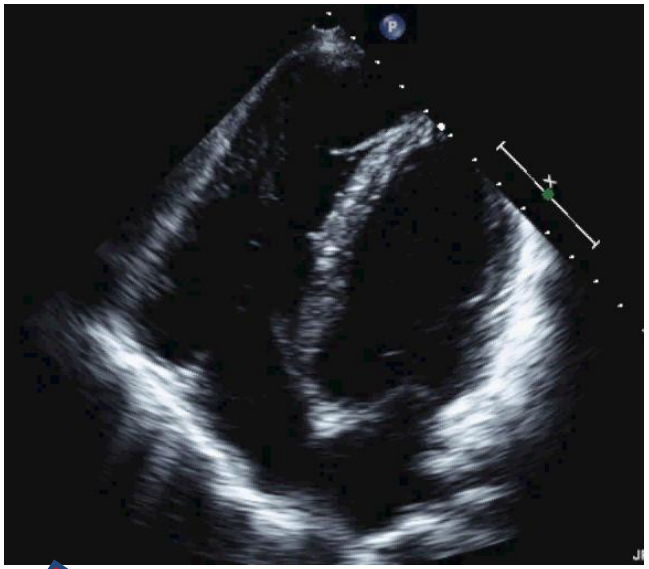


Pulmonary regurgitation

« well tolerated »



RV Dilatation



Reduced exercise capacity

Heart Failure

Arythmia

Interaction RV/LV

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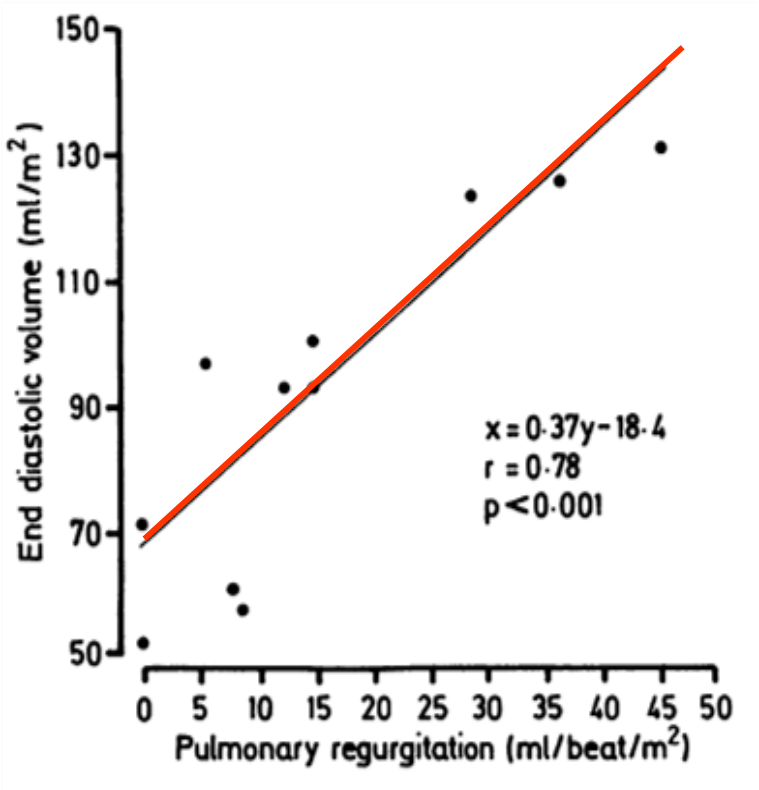
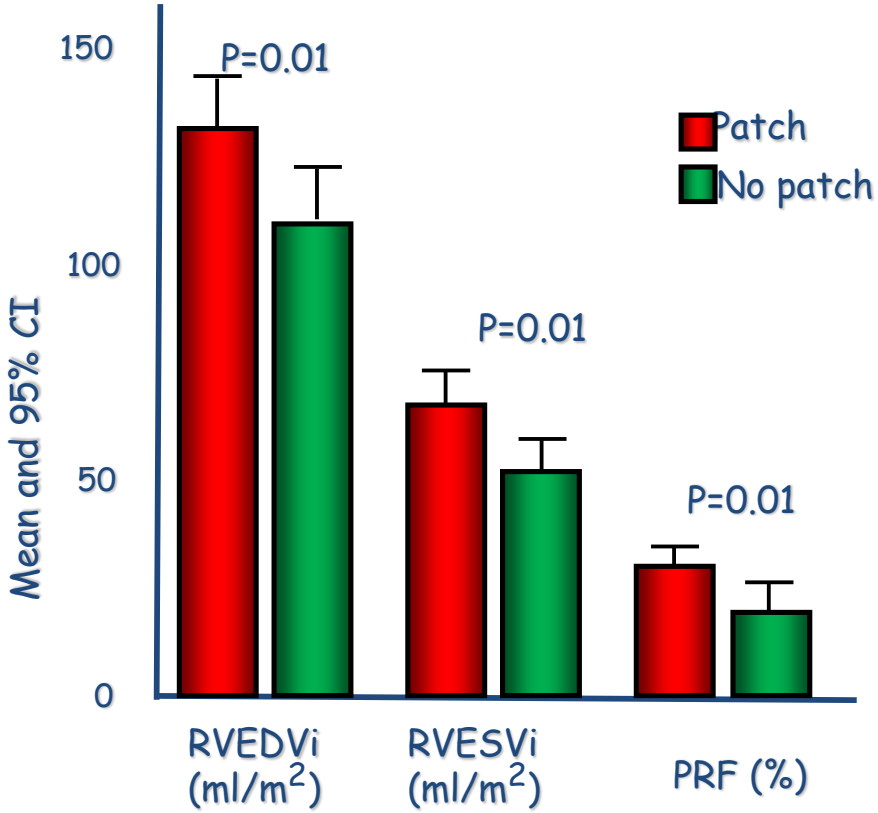
PR and RV dilatation:

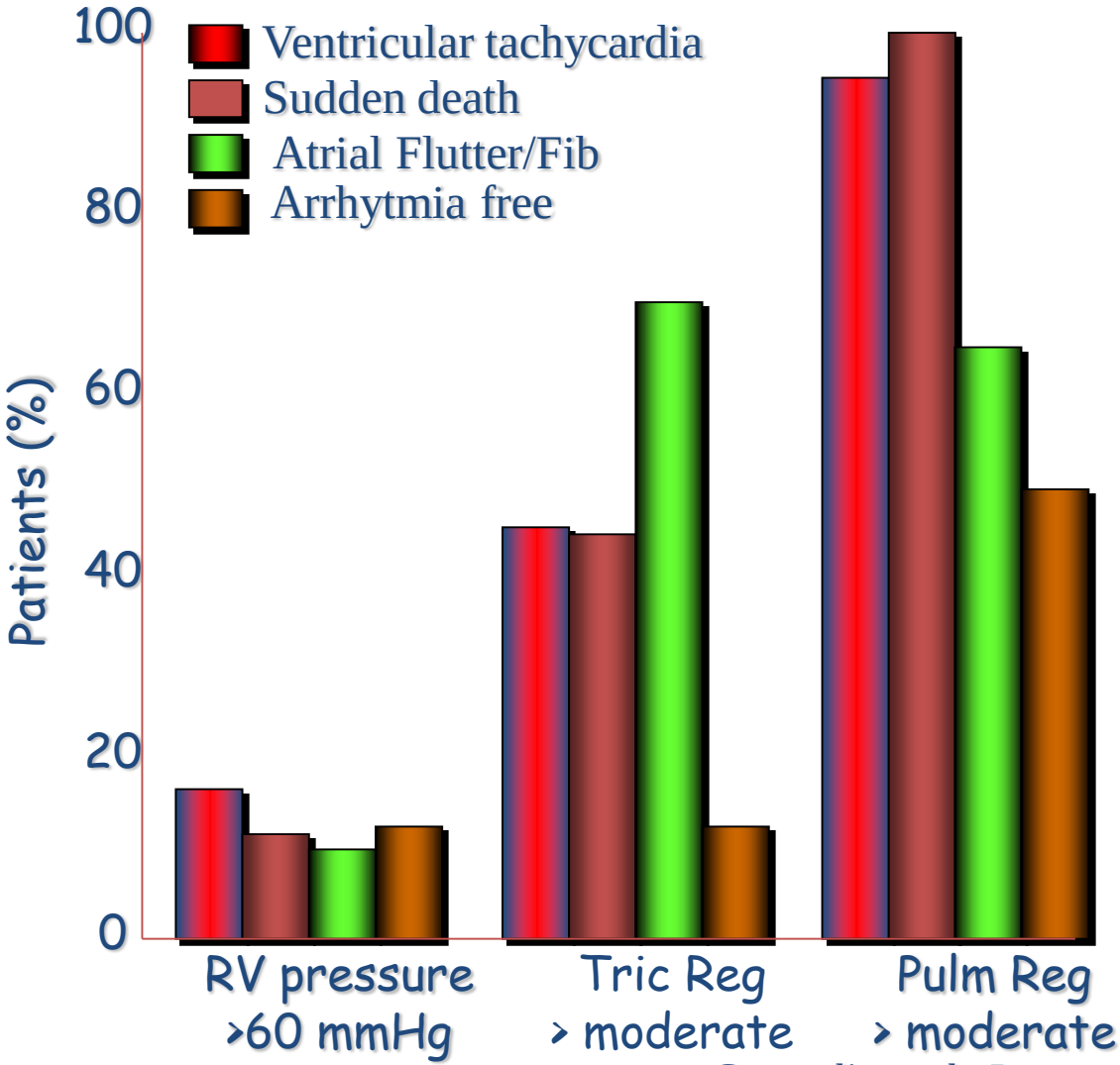




Redington et al , Br Heart J 1988;60: 57-65

Davlouros et al ,JACC2002;40: 2044-2052

PR and risk of arythmias:



Gatzoulis et al , Lancet 2000;356: 975-981

Guidelines:

Pulmonary valve replacement is indicated for severe pulmonary regurgitation and symptoms or decreased exercise tolerance.	I	B
Pulmonary valve replacement is reasonable in adults with previous tetralogy of Fallot, severe pulmonary regurgitation, and any of the following: a. Moderate to severe RV dysfunction. (*Level of Evidence: B*) b. Moderate to severe RV enlargement. (*Level of Evidence: B*) c. Development of symptomatic or sustained atrial and/or ventricular arrhythmias. (*Level of Evidence: C*) d. Moderate to severe TR. (*Level of Evidence: C*)	IIa	C
Surgery is reasonable in adults with prior repair of tetralogy of Fallot and residual RVOT obstruction (valvular or subvalvular) and any of the following indications: a. Residual RVOT obstruction (valvular or subvalvular) with peak instantaneous echocardiography gradient greater than 50 mm Hg. b. Residual RVOT obstruction (valvular or subvalvular) with RV/LV pressure ratio greater than 0.7. b. Residual RVOT obstruction (valvular or subvalvular) with progressive and/or severe dilatation of the right ventricle with dysfunction. c. A combination of multiple residual lesions (eg, VSD and RVOT obstruction) leading to RV enlargement or reduced RV function.	IIa	C

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AHA ACC guidelines GUCH, 2008

IREC Institut de Recherche Expérimentale et Clinique

Guidelines:

Pulmonary valve replacement is indicated for severe pulmonary regurgitation and symptoms or decreased exercise tolerance.	I	B
Pulmonary valve replacement is reasonable in adults with previous tetralogy of Fallot, severe pulmonary regurgitation, and any of the following: a. Moderate to severe RV dysfunction. (*Level of Evidence: B*) b. Moderate to severe RV enlargement. (*Level of Evidence: B*) c. Development of symptomatic or sustained atrial and/or ventricular arrhythmias. (*Level of Evidence: C*) d. Moderate to severe TR. (*Level of Evidence: C*)	IIa	C
Surgery is reasonable in adults with prior repair of tetralogy of Fallot and residual RVOT obstruction (valvular or subvalvular) and any of the following indications: a. Residual RVOT obstruction (valvular or subvalvular) with peak instantaneous echocardiography gradient greater than 50 mm Hg. b. Residual RVOT obstruction (valvular or subvalvular) with RV/LV pressure ratio greater than 0.7. b. Residual RVOT obstruction (valvular or subvalvular) with progressive and/or severe dilatation of the right ventricle with dysfunction. c. A combination of multiple residual lesions (eg, VSD and RVOT obstruction) leading to RV enlargement or reduced RV function.	IIa	C

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Tetralogy of Fallot :

Indications	Class[a]	Level[b]
PVRep should be performed in symptomatic patients with severe PR and/or stenosis (RV systolic pressure >60 mmHg, TR velocity >3.5 m/s)	I	C
PVRep should be considered in asymptomatic patients with severe PR and/or PS when at least one of the following criteria is present: • Decrease in objective exercise capacity • Progressive RV dilation • Progressive RV systolic dysfunction • Progressive TR (at least moderate) • RVOTO with RV systolic pressure >80 mmHg (TR velocity >4.3 m/s) • Sustained atrial/ventricular arrhythmias	IIa	C

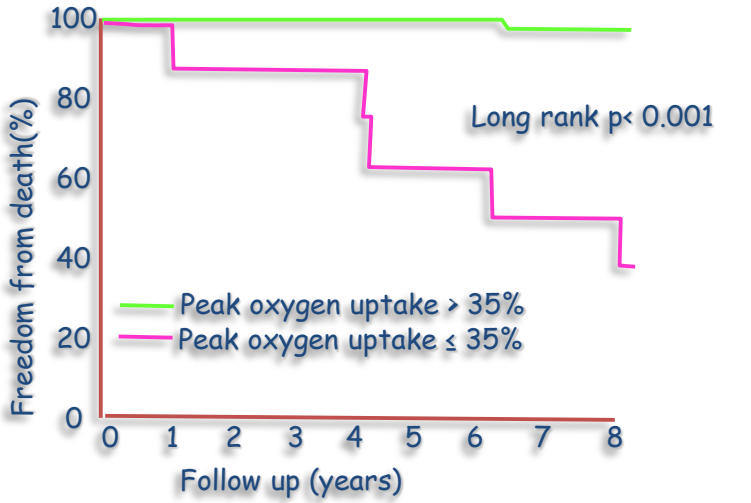
ESC guidelines GUCH, 2010

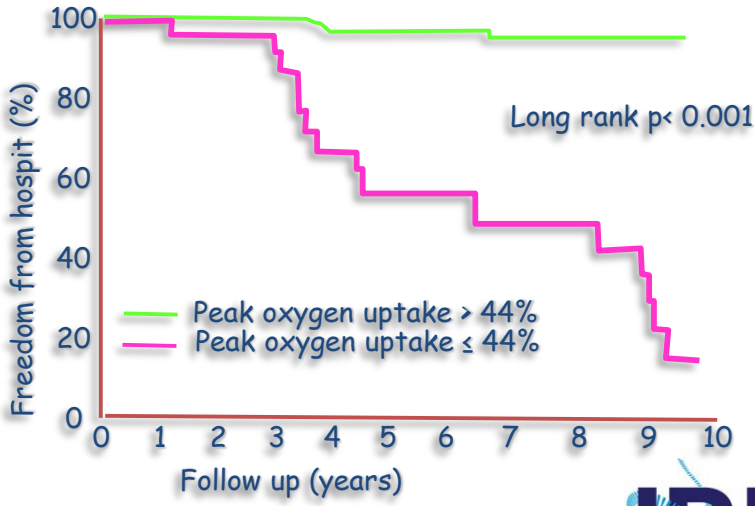
TOF exercise capacity:

Significant univariate and multivariate predictors of death and hospitalization

Variable	p Value	Hazard Ratio	95% CI
Univariate analysis			
NYHA functional class	<0.001	2.286	1.789–3.324
Peak heart rate (beats/min)	<0.001	0.982	0.966–0.998
Right ventricular systolic pressure (mm Hg)	0.002	1.021	1.014–1.036
Peak oxygen uptake (% of predicted)	<0.001	0.962	0.934–0.992
VE/VCO_2 slope	<0.001	1.098	1.046–1.148
Pulmonary regurgitation	<0.001	1.762	1.256–1.987
Right ventricular systolic function	<0.001	1.934	1.623–2.134
Multivariate analysis			
NYHA functional class	0.001	2.118	1.344–3.542
Peak oxygen uptake (% of predicted)	0.01	0.974	0.950–0.994
VE/VCO_2 slope	0.002	1.076	1.038–1.115

Hazard ratios refer to unit increases in NYHA functional class, peak oxygen uptake (percent of predicted), VE/VCO_2 slope, heart rate (beats/min), and right ventricular systolic pressure (mm Hg).





Giardini et al, Am J Cardiol 2007;99: 1462-1467

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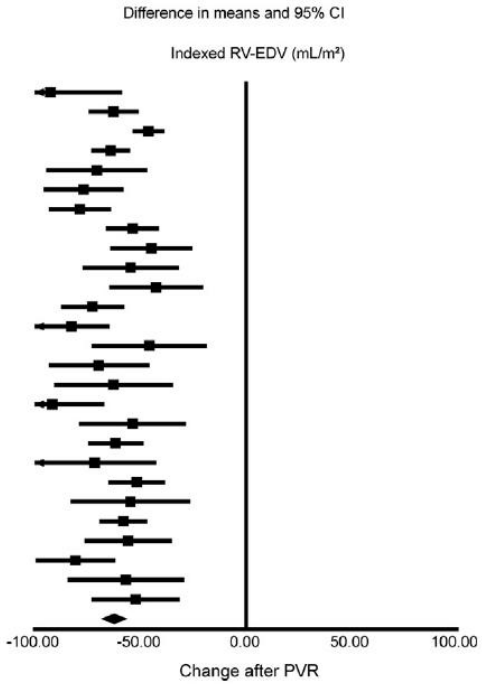
PVR usefull to reduce RV dimensions?

A

Study name	Difference in means	Standard error	P-Value	Relative weight (%)
Chalard 2012	-93.000	17.178	<0.001	1.74
Lee 2012	-63.000	5.867	<0.001	5.37
Quail 2012 (Matched)	-46.400	3.619	<0.001	6.47
Quail 2012 (Unmatched)	-64.400	4.511	<0.001	6.05
Jang 2012 (Matched)	-70.900	12.056	<0.001	2.84
Jang 2012 (Unmatched)	-77.300	9.500	<0.001	3.71
Tobler 2012	-79.000	7.418	<0.001	4.61
Frigiola 2012	-54.000	6.220	<0.001	5.19
Chen 2012	-45.000	9.742	<0.001	3.62
Ovcina 2011 (arm 1)	-54.900	11.437	<0.001	3.03
Ovcina 2011 (arm 2)	-42.800	11.136	<0.001	3.13
Geva 2010 (arm 1)	-73.000	7.485	<0.001	4.58
Geva 2010 (arm 2)	-83.000	8.995	<0.001	3.91
Lindsey 2010	-46.000	13.752	0.001	2.40
Tsang 2010	-70.000	11.982	<0.001	2.87
Harrild 2009	-63.000	14.209	<0.001	2.30
Knirsch 2008	-92.000	12.334	<0.001	2.77
Frigiola 2008	-54.000	12.771	<0.001	2.65
Henkens 2007 (arm 1)	-62.000	6.552	<0.001	5.03
Henkens 2007 (arm 2)	-72.000	14.739	<0.001	2.18
Oosterhof 2007	-52.000	6.646	<0.001	4.98
Ghez 2007	-55.000	14.321	<0.001	2.27
Kleinveld 2006	-58.300	5.613	<0.001	5.50
Therrien 2005	-56.000	10.381	<0.001	3.38
Buechel 2005	-81.100	9.437	<0.001	3.74
Straten 2005	-57.100	13.950	<0.001	2.35
Vliegen 2002	-52.500	10.468	<0.001	3.35
Overall effect	-62.734	2.591	<0.001	

Total (95% CI): 782 (Pre-PVR); 725 (Post-PVR)
Test for heterogeneity: Chi² = 61.96; df = 26 (P < 0.001); I² = 58.3%
Test for overall random effect: Z = -24.20 (P < 0.001)



PVR

- Reduce PR
- ↓ RV diastolic volume
- ↑ Systolic LV function
- ↓ QRS width
- ↓ symptoms

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Cavalcanti et al , JACC 2013;62:2227-2243

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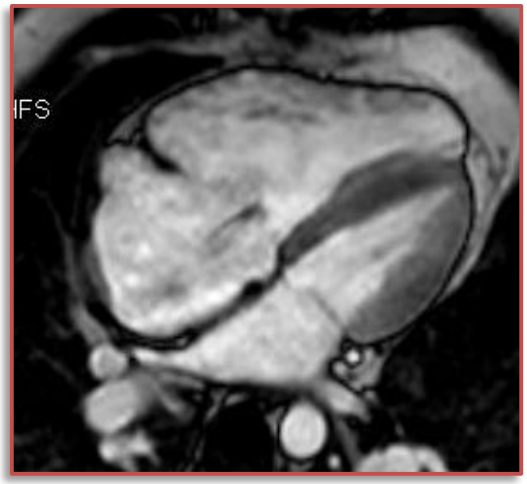
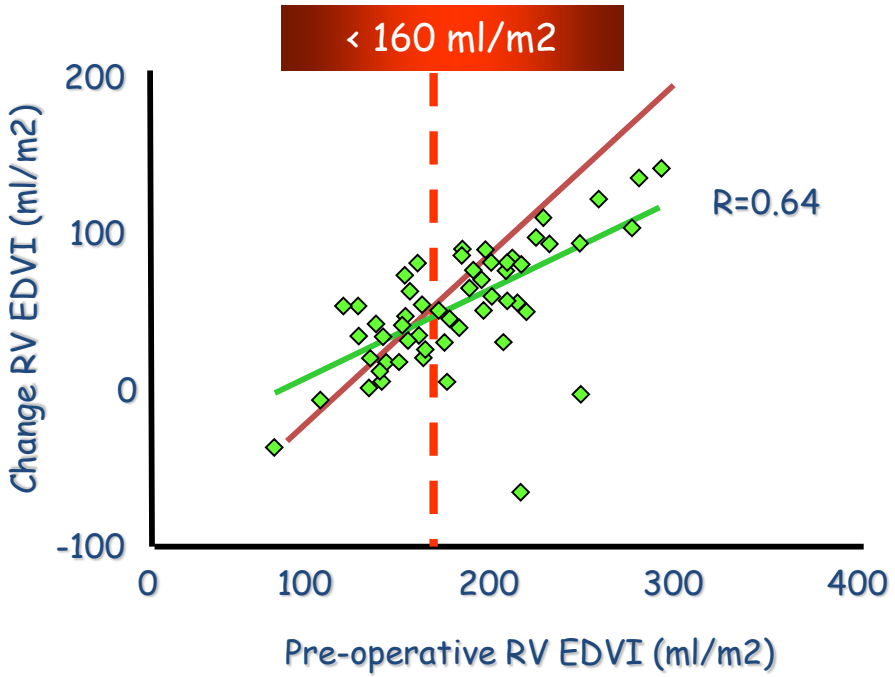
TOF RV dilatation:





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Oosterhol et al , Circulation. 2007;116:545-551

TOF RV dilatation/Function :

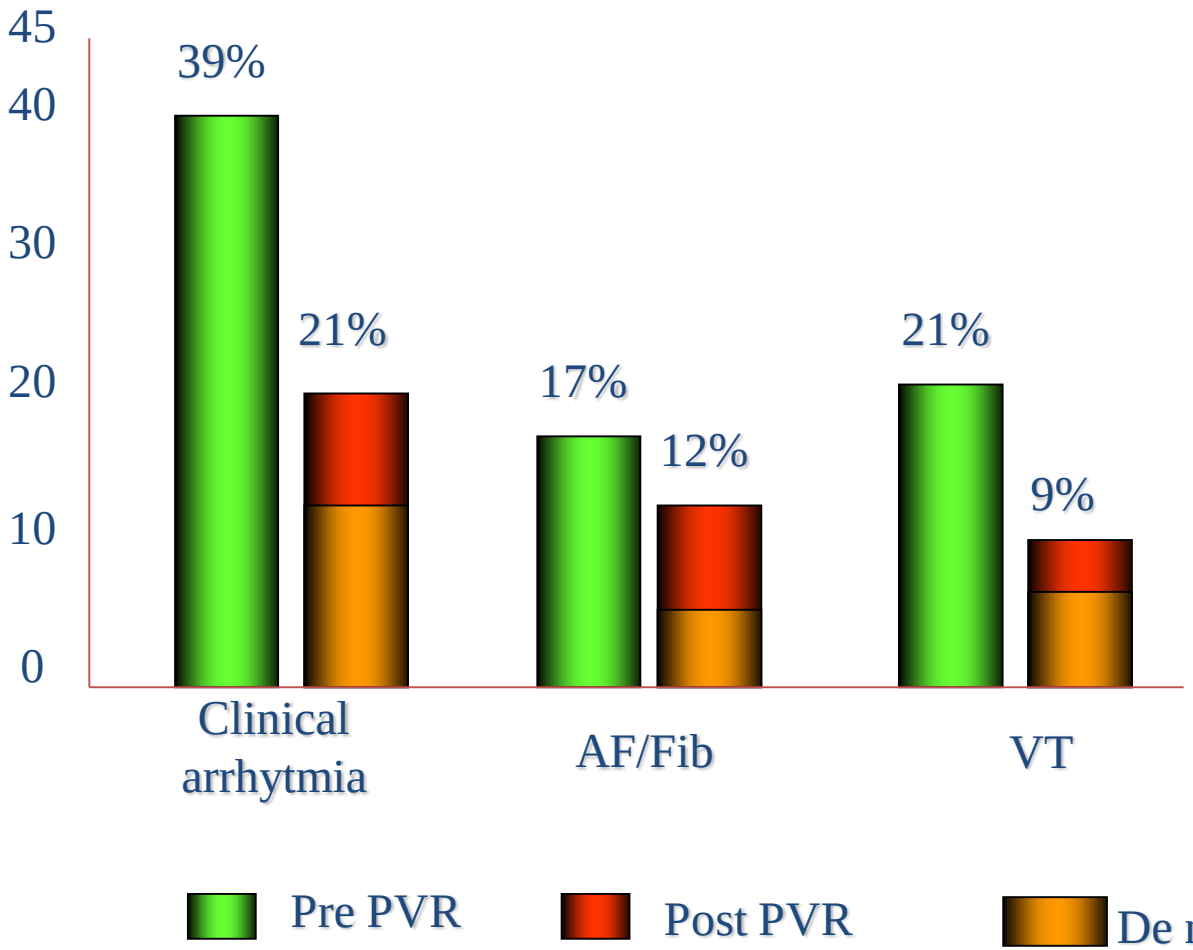
When ?

- Huge dilatation > 160 (150) ml/m2 RV EDVI no recuperation Rv dimension and function

- Lower limit ? 140 ?, 150 ? ???

- Other aspect:
  - Pulmonary pressure
  - Pulmonary branch
  - RV function ?

Effect of PVR on arythmias:



Thierrien et al , Circ 2001;103: 2489-2494

Conclusion:

- Indication for interventions in pulmonary valve disease are less well defined than for other valve disease.

- If there is threshold quite well defined for valve stenosis, the question remain frequently open for pulmonary regurgitation.

- One part of problem is related to assessment of RV dilatation and function and the lack of knowledge of functional recovery of the RV