



DGPK Guideline Pulmonary Regurgitation Version 2016

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General Information

Which patients are affected?

- Congenital isolated pulmonary regurgitation (rare)
- Residual pulmonary regurgitation
 - after repair of tetralogy of Fallot
 - after treatment of pulmonary stenosis
- Dysfunction of a RV to PA-conduit

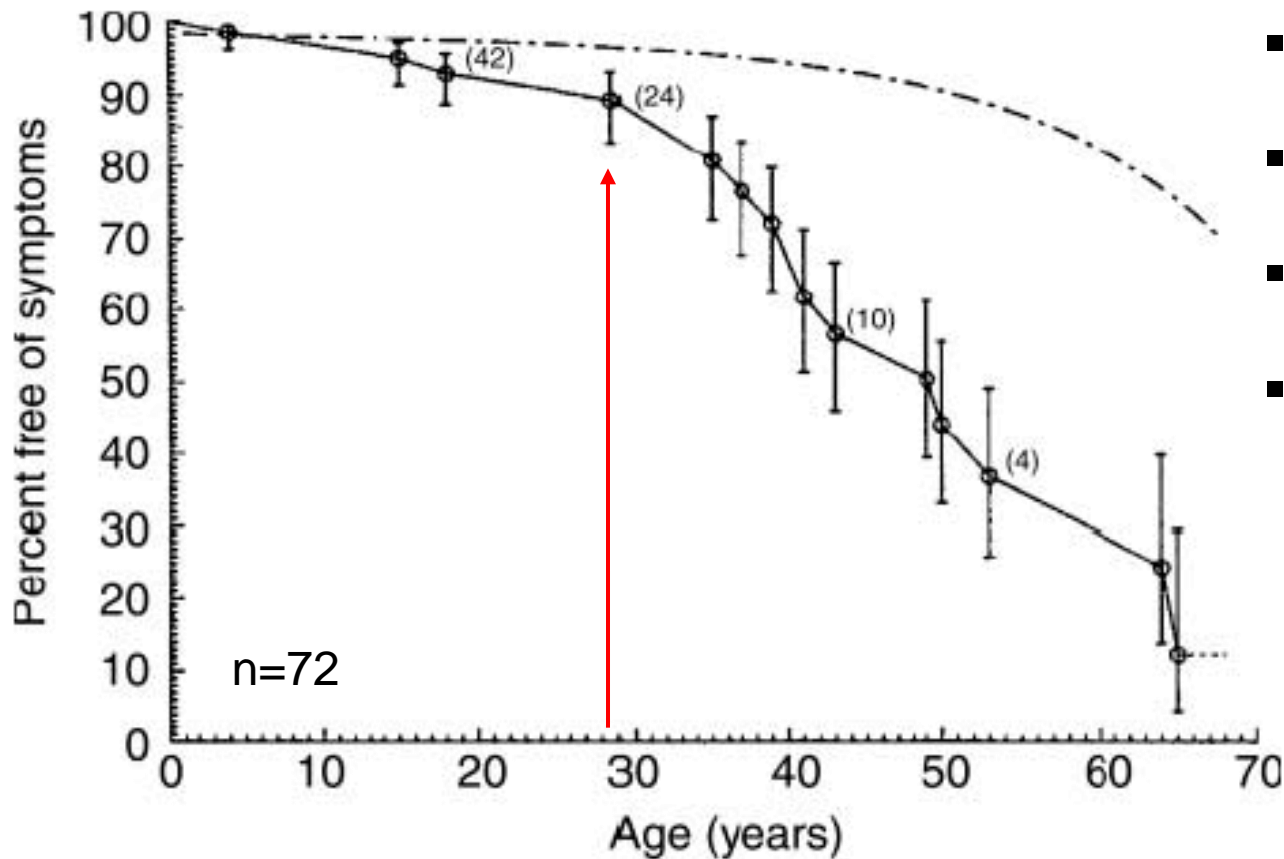


Sequelae

- Symptoms
- Impaired long-term survival
- Exercise intolerance
- Right ventricular enlargement
- Right (and left) ventricular dysfunction
- Electrical disturbance



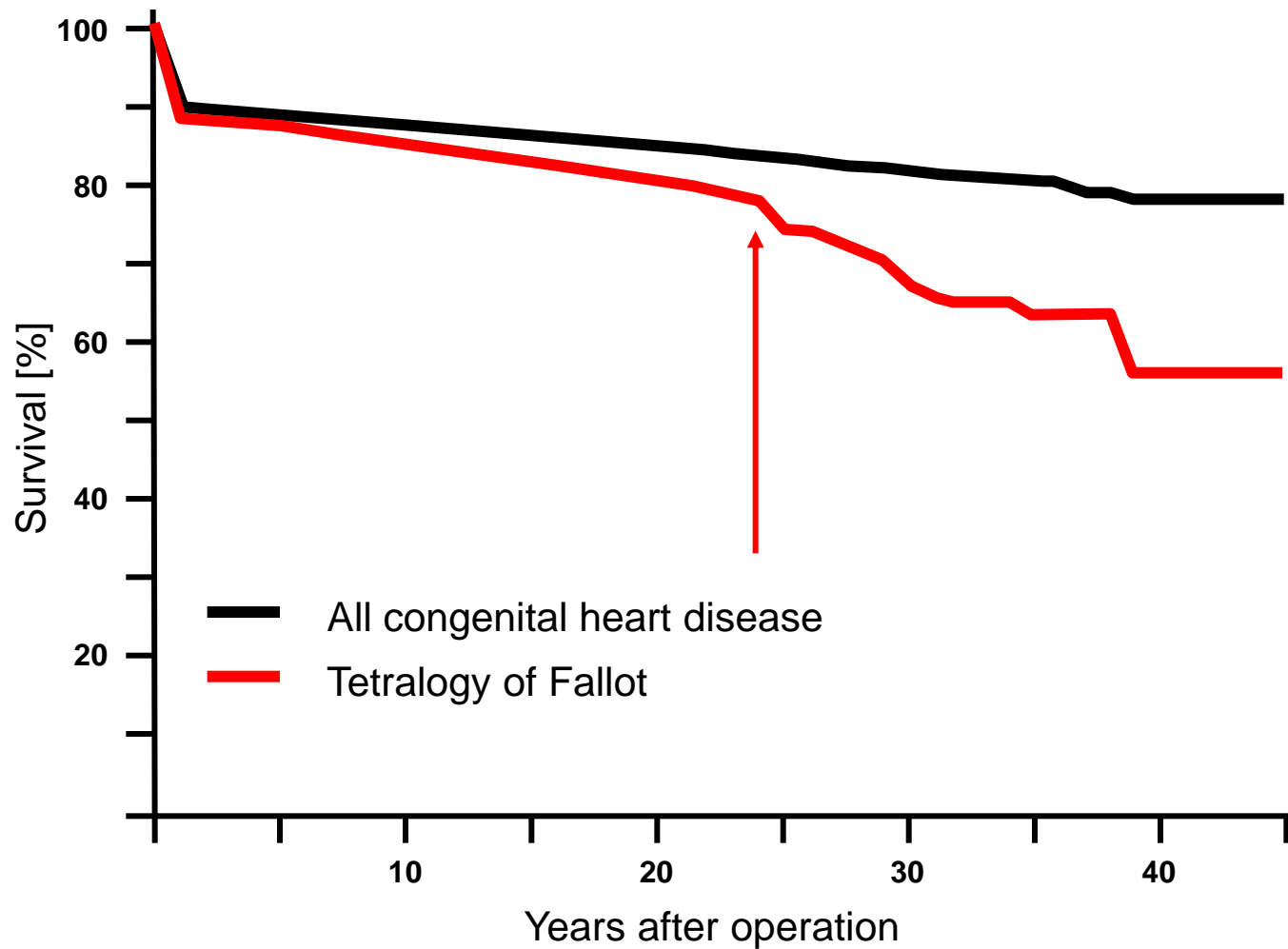
Symptoms



- Exercise intolerance
- Dyspnoea
- Oedema
- Sudden death

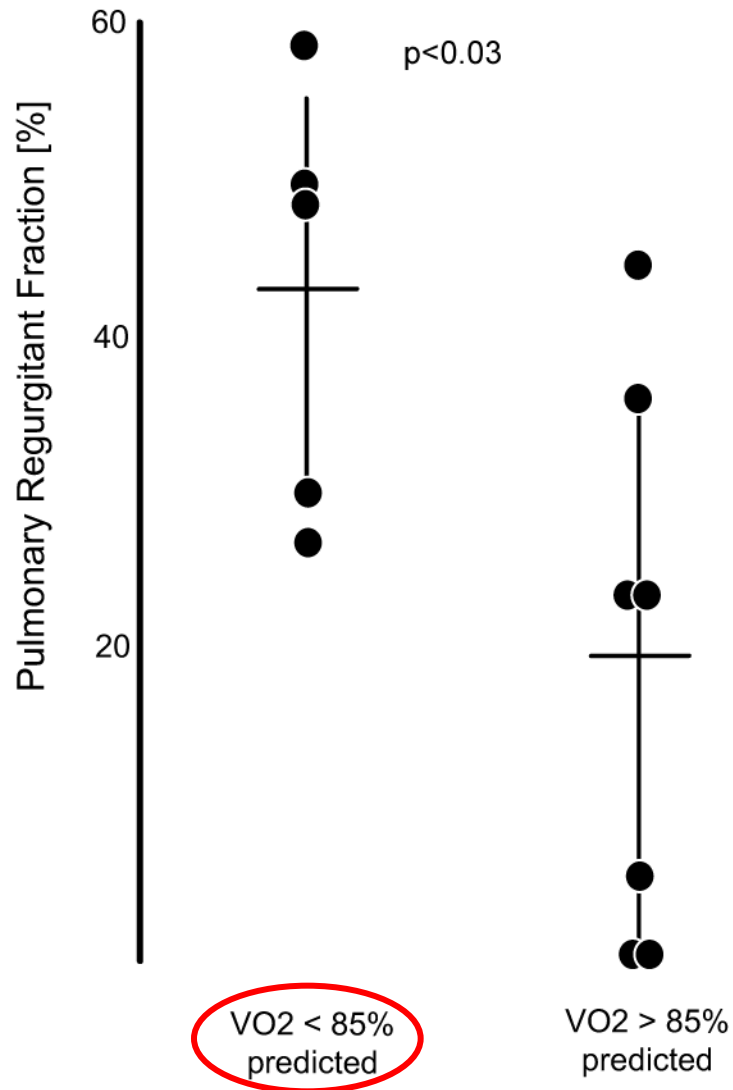


Impaired Long-term Survival



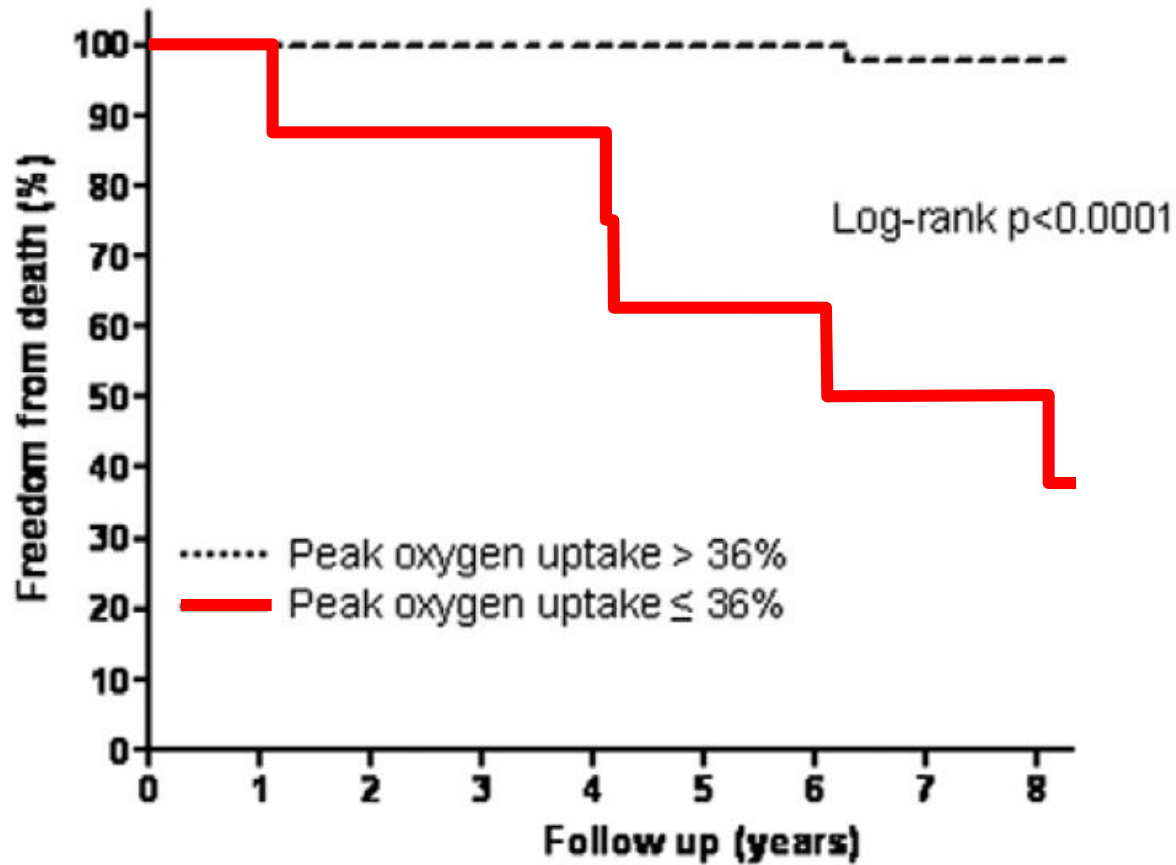


Exercise Intolerance

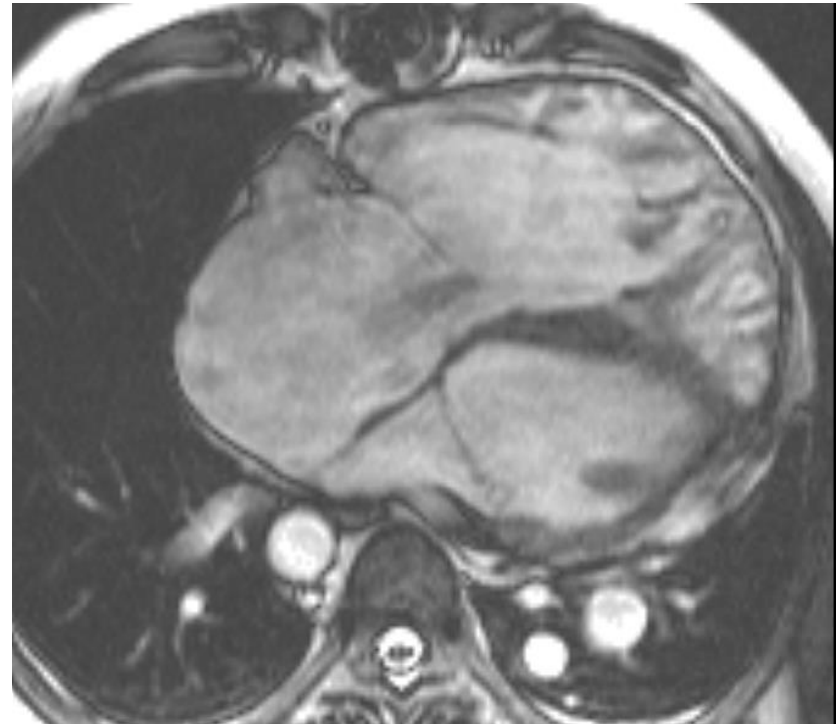
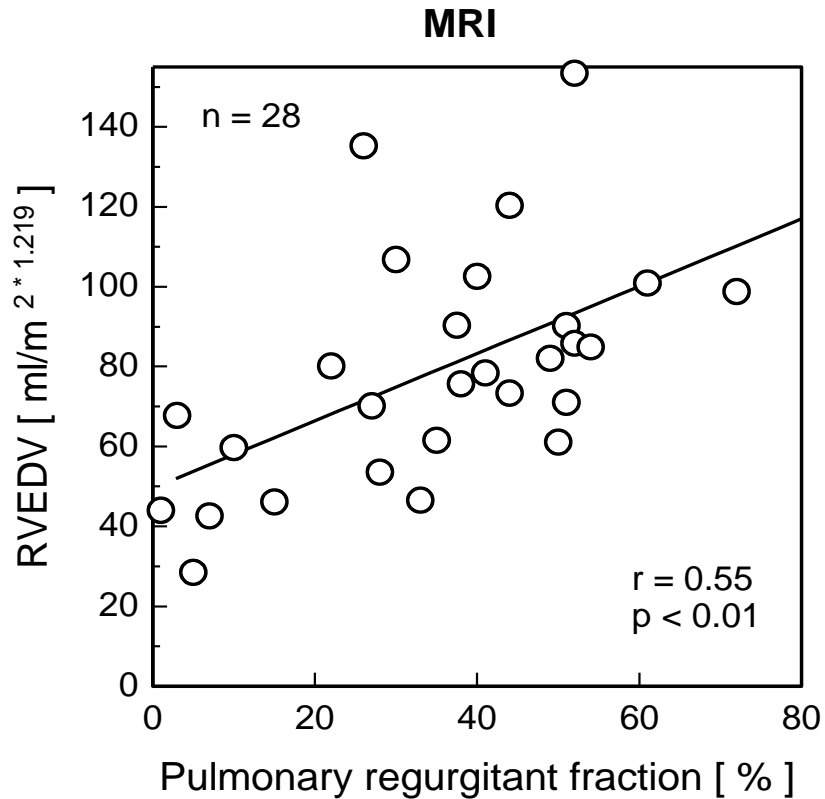




Exercise Intolerance

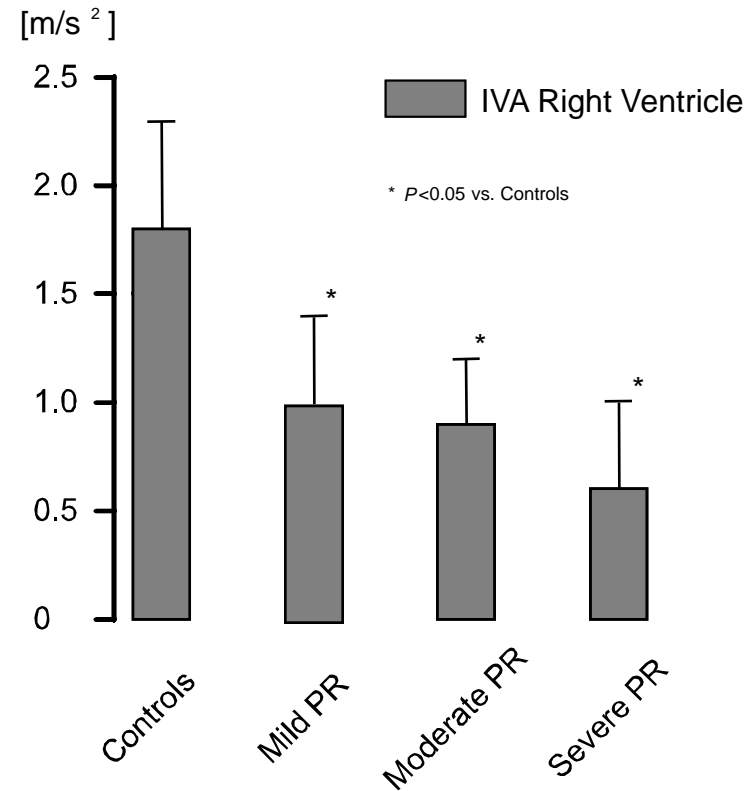
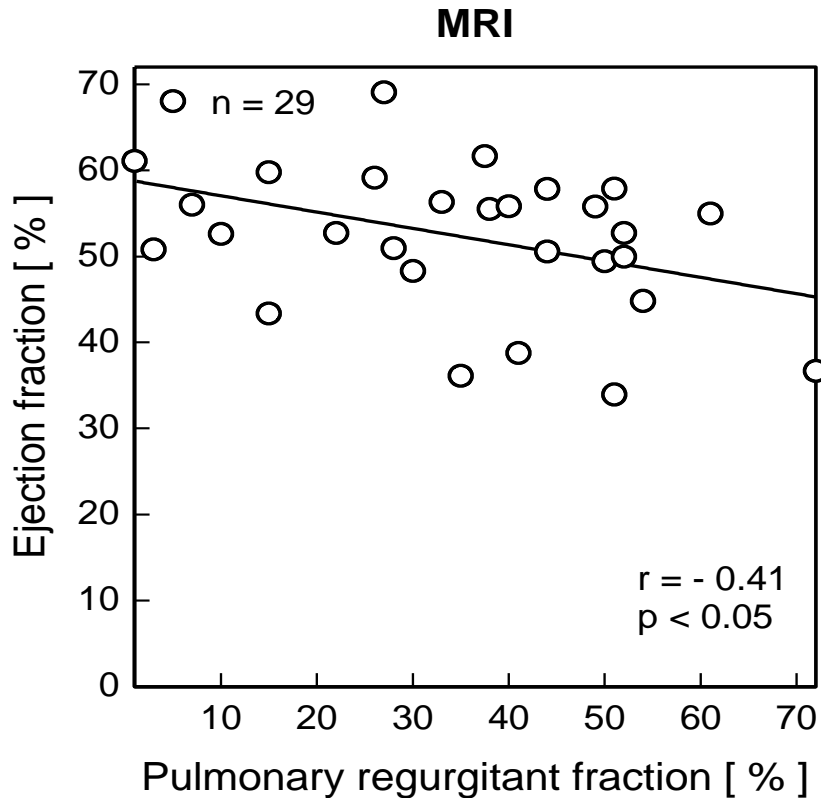


Right Ventricular Enlargement



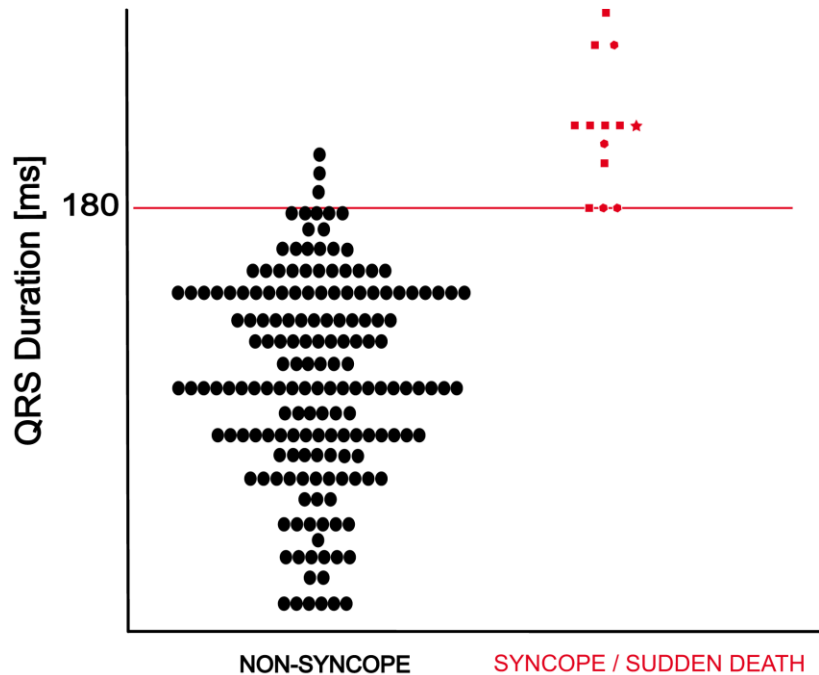
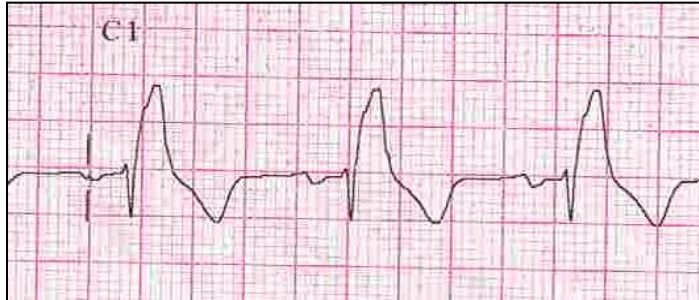


Right Ventricular Dysfunction





Electrical Disturbance



- RV enlargement correlates with QRS duration
- **QRS duration >180 ms** and **increase of QRS duration > 3.5 ms/year** are highly sensitive **predictors of sustained VT and sudden cardiac death**



Diagnostic Aims

- Determination of PR severity
- Disclosure of potential sequelae of PR
 - RV dysfunction
 - RV enlargement
 - Risk stratification for malignant arrhythmias
- Disclosure of associated or additional lesions



Diagnostic Modalities

ECG:	QRS duration (progression over time?) Present arrhythmia
Holter:	Ventricular ectopy Intermittent arrhythmia
Chest X-ray:	Heart size (CTR) Size of the pulmonary arteries Asymmetric lung perfusion
Echocardiography:	Estimation of PR severity Estimation of RV size and RV function Detection of associated lesions (VSD, RVOT obstruction etc.)
MRI:	Quantification of PR (RGF) RV function/ volume RV fibrosis („late enhancement“)
Exercise testing:	Quantification of exercise tolerance
Cardiac catheter:	Exclusion of pulmonary hypertension (previous shunt procedures) Interventional procedures (dilatation of PA stenoses)



Therapy - Remarks

- The treatment of pulmonary regurgitation is pulmonary valve replacement (or reconstruction in selected cases)
- Indications for pulmonary valve replacement are in evolution
- Balancing the risk of late RV dysfunction, arrhythmia and sudden cardiac death against the finite lifespan of a valve prosthesis and risks of surgery is the major challenge for the timing of pulmonary valve replacement



Pulmonary Valve Replacement is reasonable...

in the **presence of significant PR** (RGF >30% on MRI)

and at least one of the following findings:

- Symptoms
- Development of symptomatic of sustained atrial or ventricular arrhythmias
- Moderate to severe RV dysfunction (or deterioration during follow-up)
- Moderate to severe RV dilatation (or deterioration during follow-up)
- Development of tricuspid regurgitation in the presence of RV enlargement
- QRS duration > 180 ms or increase in QRS duration $\geq 3,5$ ms/year
- Additional lesions needing bypass surgery



Pregnancy and Pulmonary Regurgitation

Womens desire to become pregnant should be included in the decision making for pulmonary valve replacement

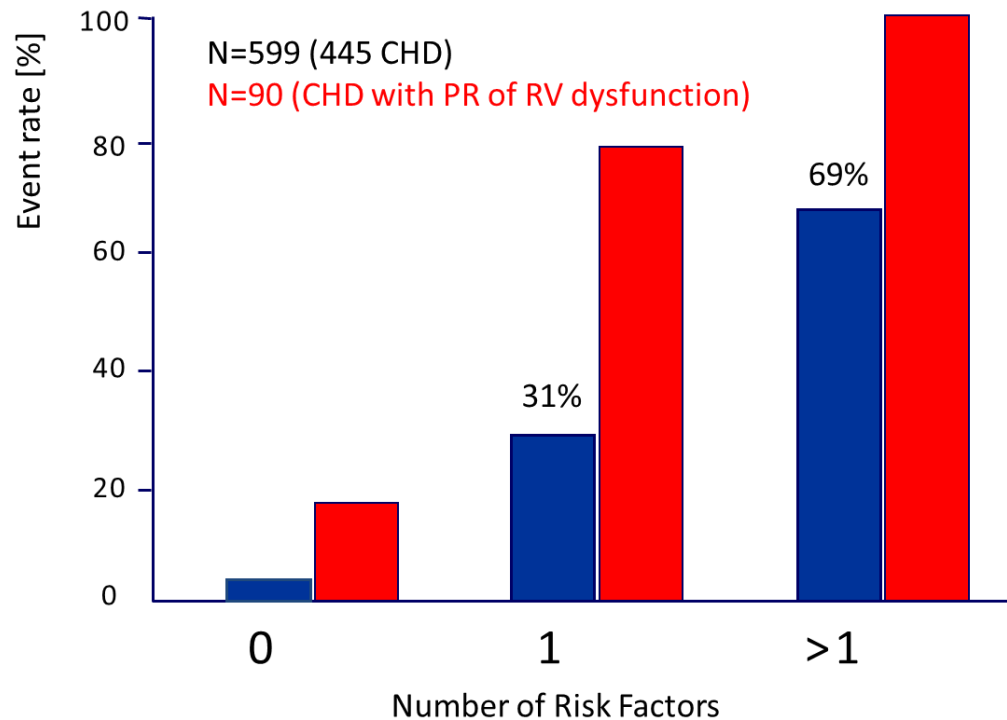
- **Pulmonary regurgitation increases the risk for maternal complications associated with pregnancy in the presence of additional risk factors**



Pregnancy and Pulmonary Regurgitation

CAPREG Risk Factors

- Prior cardiac events (heart failure/ stroke/ arrhythmia)
- NYHA III of IV or cyanosis
- Significant left heart obstruction
- Systemic ventricular dysfunction





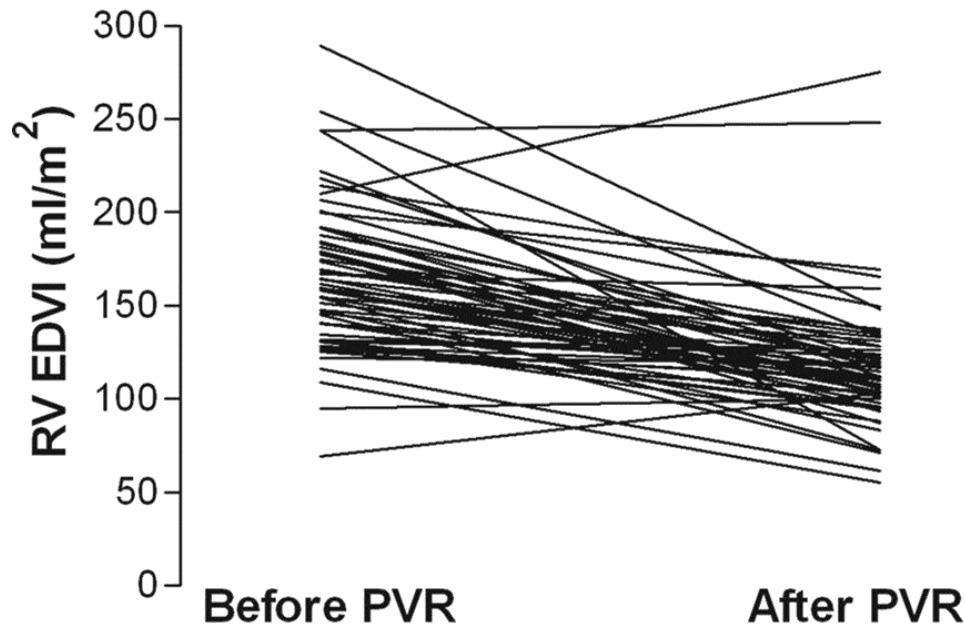
Symptoms

- Symptoms can be mild
- Patients are often unaware of their limitations
- **Serial exercise testing should be used to assess functional capacity objectively**



Right Ventricular Enlargement

- Pulmonary valve replacement should be carried out before the **end diastolic volume index exceeds 150-160 ml/m²** or the **end systolic volume index exceeds 80-90 ml/m²** on MRI assessment.



'Cut off values' for normalisation:

RV EDVi: 160 ml/m²

RV ESVi: 82 ml/m²

n=57
prospective
all ≥ moderate PR



Right Ventricular Enlargement

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Journal of the American College of Cardiology
© 2012 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Vol. 60, No. 11, 2012
ISSN 0735-1097/\$36.00
<http://dx.doi.org/10.1016/j.jacc.2012.03.077>

Congenital Heart Disease

Outcomes of Pulmonary Valve Replacement in 170 Patients With Chronic Pulmonary Regurgitation After Relief of Right Ventricular Outflow Tract Obstruction

Implications for Optimal Timing of Pulmonary Valve Replacement

Cheul Lee, MD,* Yang Min Kim, MD,† Chang-Ha Lee, MD,* Jae Gun Kwak, MD,*
Chun Soo Park, MD,* Jin Young Song, MD,‡ Woo-Sup Shim, MD,‡ Eun Young Choi, MD,‡
Sang Yun Lee, MD,‡ Jae Suk Baek, MD‡

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Midterm outcomes of PVR in patients with chronic PR were acceptable. PVR should be considered before RV EDVI exceeds **163 ml/m²** or RV ESVI exceeds **80 ml/m²**, with more attention to RV ESVI. (J Am Coll Cardiol 2012;60:1005–14) © 2012 by the American College of Cardiology Foundation

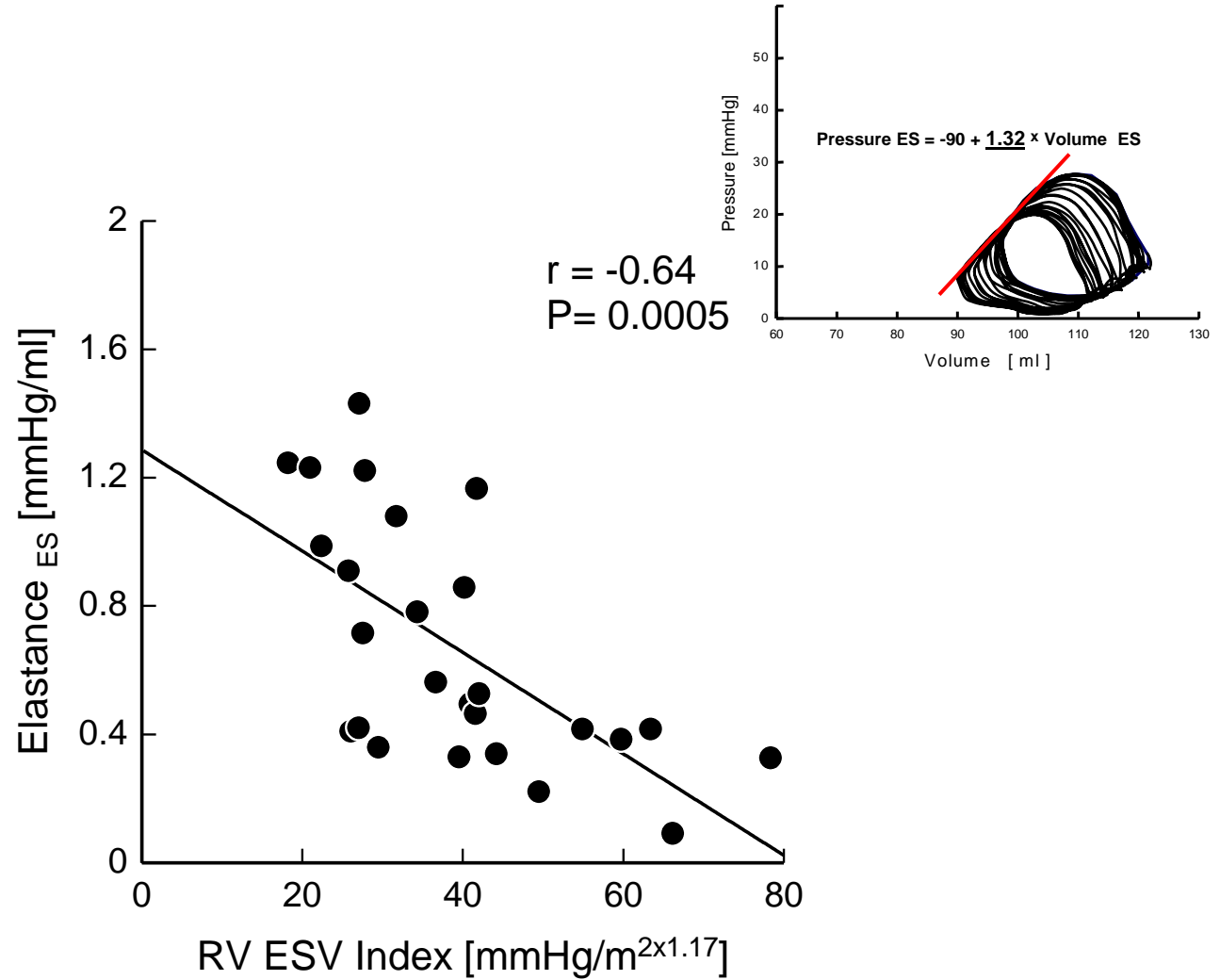
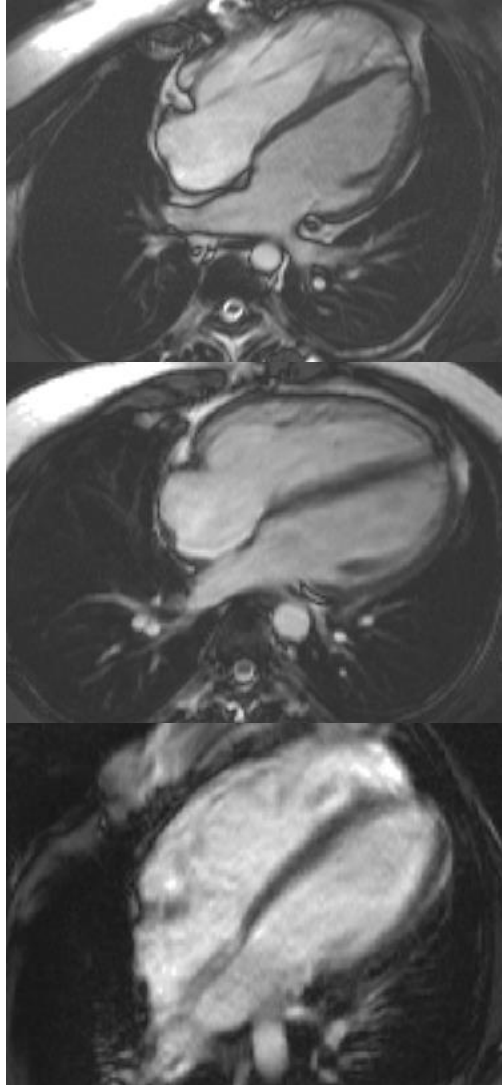


Right Ventricular Dysfunction

- RV function is difficult to quantify.
- The diagnosis of RV dysfunction should be based on MRI and echocardiographic data obtained by experienced investigators.
- The end systolic volume index is a good estimate of RV contractility.
- Progressive RV enlargement on serial MRI assessment suggests deterioration of RV contractility.



Right Ventricular Dysfunction





Right Ventricular Enlargement

- Pulmonary valve replacement should be carried out before the **end diastolic volume index exceeds 150-160 ml/m²** or the **end systolic volume index exceeds 80-90 ml/m²** on MRI assessment.



European Heart Journal
doi:10.1093/eurheartj/ehv550

CLINICAL RESEARCH

Congenital heart disease

Preoperative thresholds for mid-to-late haemodynamic and clinical outcomes after pulmonary valve replacement in tetralogy of Fallot

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Received 25 March 2015; revised 4 August 2015; accepted 25 September 2015

Patients with preoperative **RV ESV 95 ml/m²** were at increased risk for unfavourable mid-to-late haemodynamic outcome



Associated Lesions

- RVOT obstruction with an RV/LV pressure ratio $>0,7$ or progressive RV dilatation and/or RV dysfunction.
- Residual VSD with a left-to-right shunt greater than 1,5 : 1,0.
- Severe aortic regurgitation with associated symptoms or more than mild LV dysfunction.
- A combination of multiple residual lesions.



Effects of Pulmonary Valve Replacement

- Reverse remodelling of the right ventricle (reduction of RV size).
- Improvement of functional and exercise capacity.
- Reduction of the incidence of arrhythmias
(esp. when combined with cryoablation).

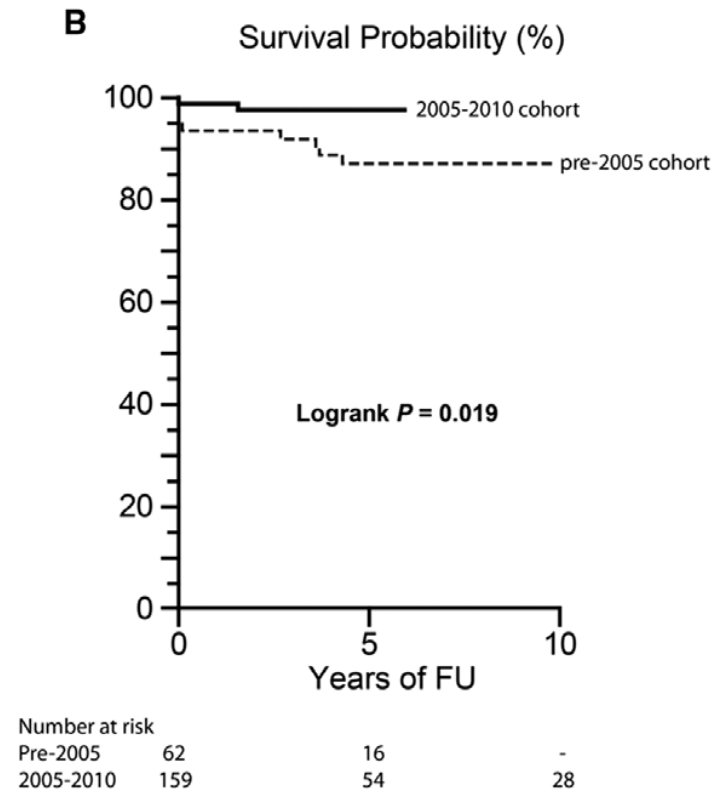
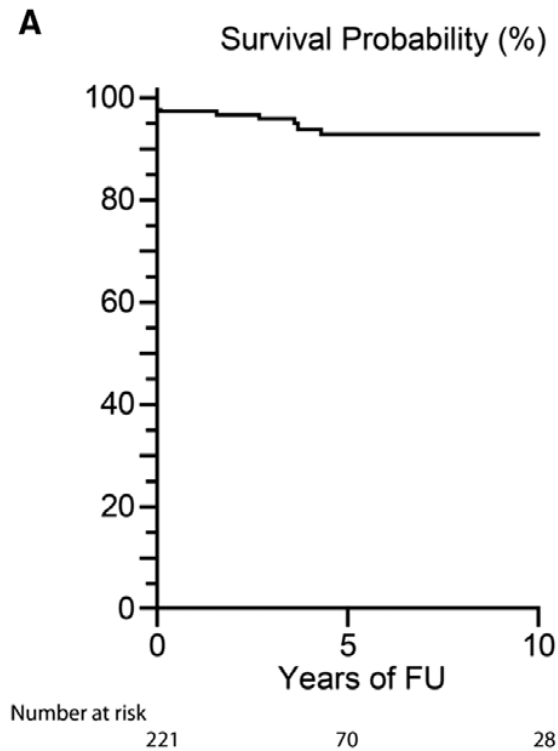


Technique of Pulmonary Valve Replacement

- There is no general consensus as to which type of biological valve prosthesis should be used for pulmonary valve replacement.
- When technically possible percutaneous pulmonary valve replacement is an alternative to surgical pulmonary valve replacement.
- The risk of infective endocarditis is potentially higher following percutaneous valve replacement when compared with homograft pulmonary valve replacement

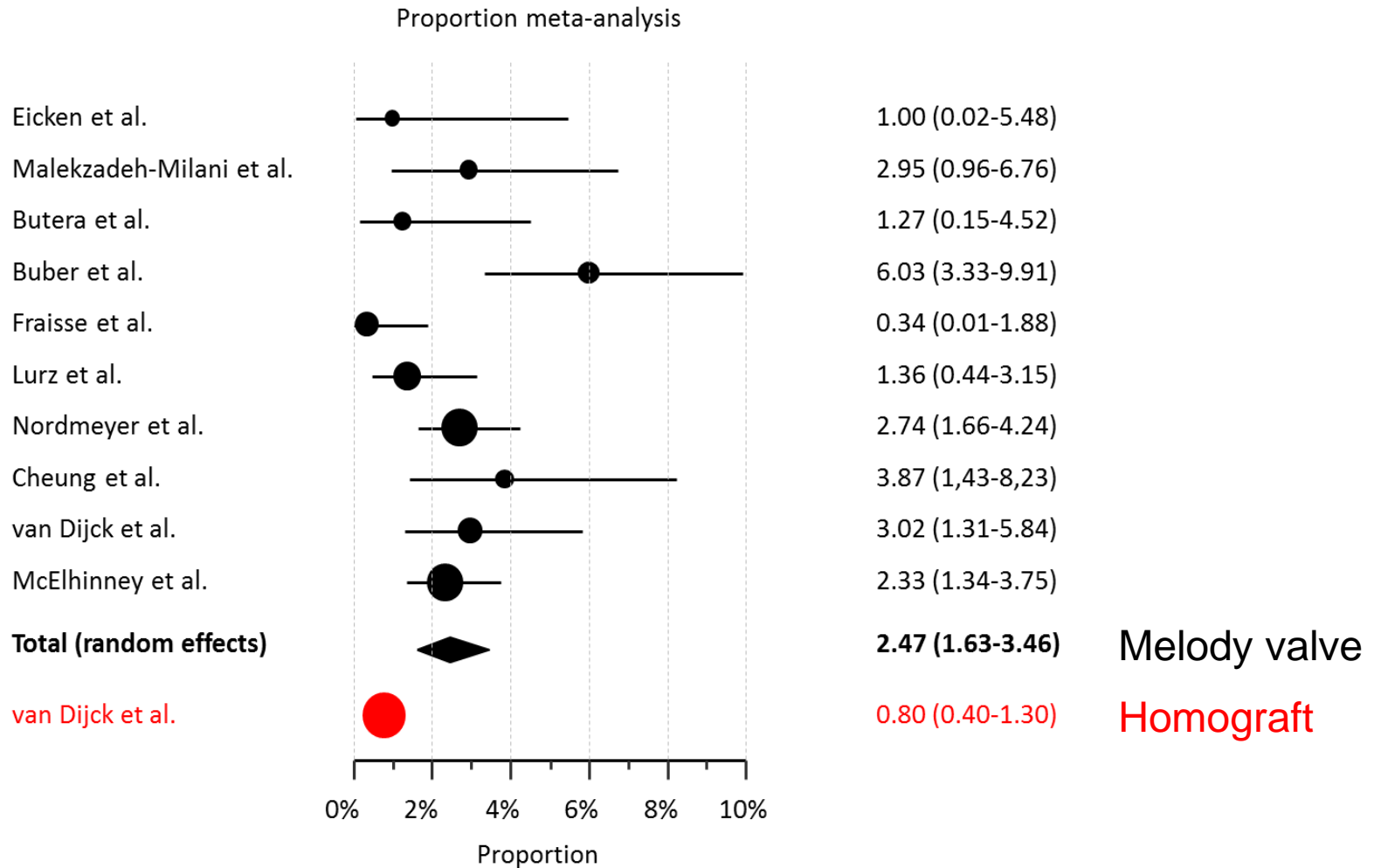


Technique of Pulmonary Valve Replacement





Technique of Pulmonary Valve Replacement





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Level of evidence

- Ref 1, 12-14 are review article (level of evidence IV)
- Ref 11 and 18 are consensus guidelines (level of evidence IV)
- Ref 2-10, 15-17 and 19-25 are descriptive studies (level of evidence III)
- Ref 26 is a scientific comment containing a meta-analysis (evidence II)