Arrhythmias and congenital heart disease

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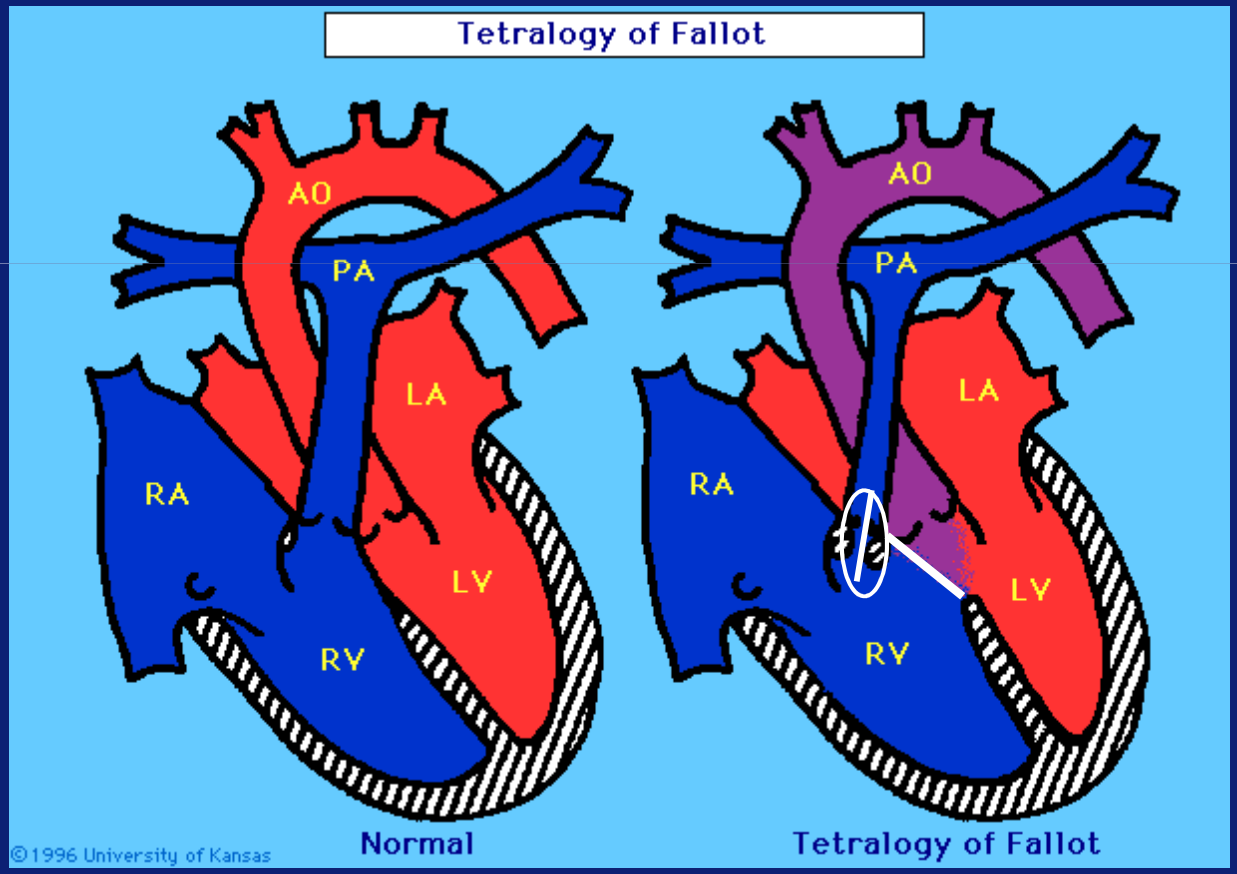
Patient W, born in 1969

Tetralogy of Fallot

1975 at the age of 6 years surgical correction



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2002 Patient W: complaints

Symptoms of diminished exercise tolerance

Physical examination:

L:	1.75 m
W:	75 Kg
RR:	130/70 mmHg
O2 sat:	98%
CVD:	normal

Systolic thrill palpable at 2L

Grade 4/6 systolic ejection murmur and 1/6 diastolic murmur

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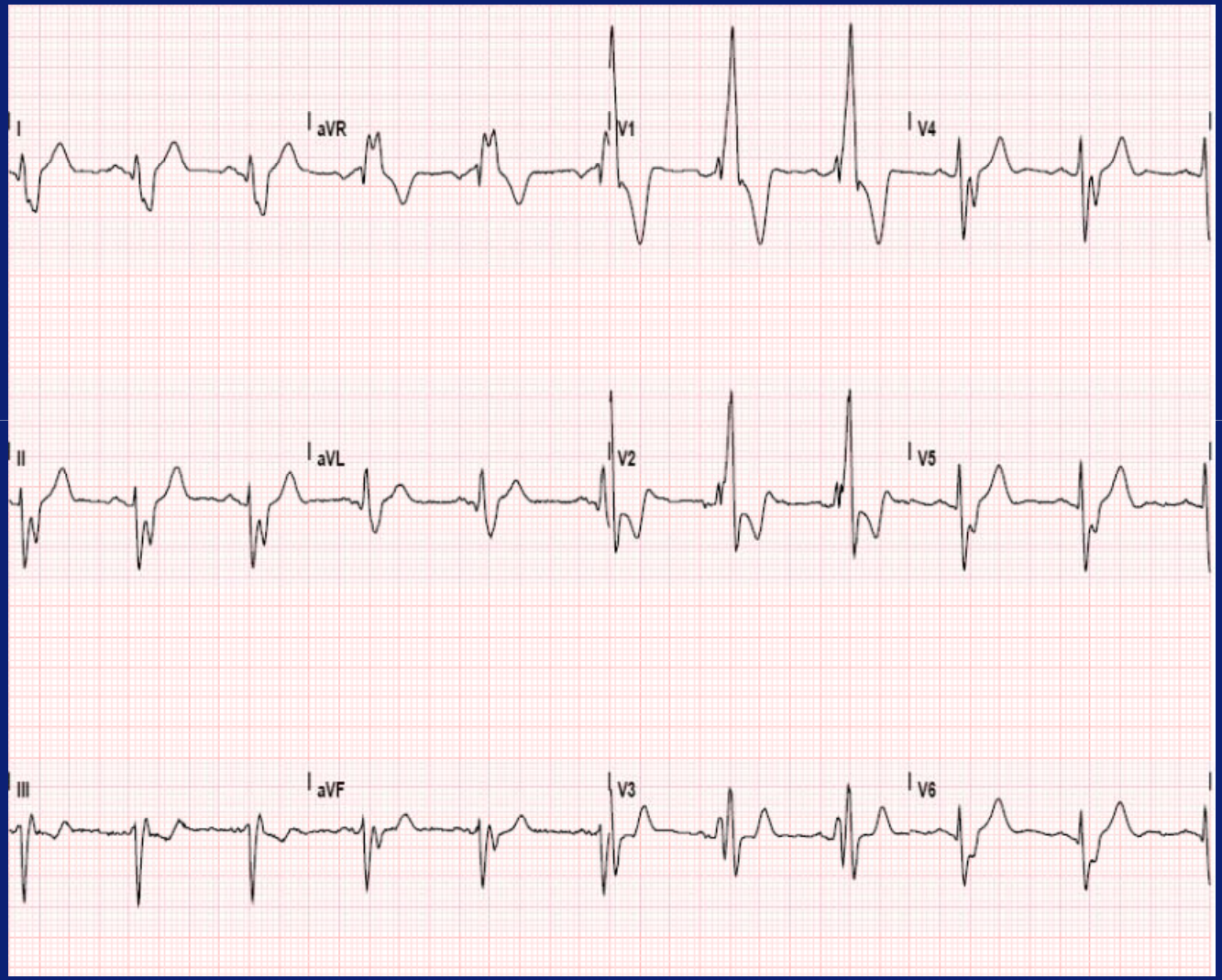
ECG



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What is the most important diagnosis in this patient with corrected Fallot?

1) severe pulmonary stenosis

2) severe pulmonary regurgitation

3) severe aortic stenosis

4) severe aortic regurgitation

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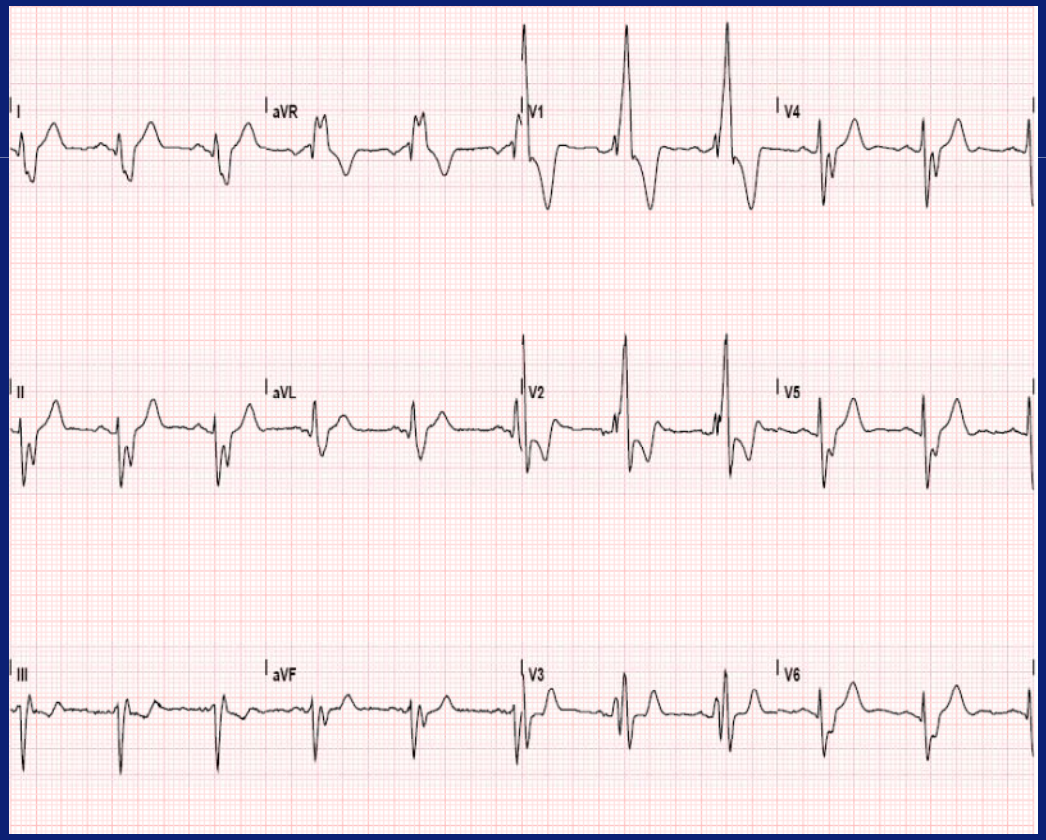
Answer: predominantly severe PS

The most common residual lesion after corrected Fallot is PI

But in this patient a systolic thrill and loud systolic murmer are present.

On ECG RBTB and RVH



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Treatment: surgery

Implantation pulmonary homograft (nr 24) and desobstruction RVOT

Rethoracotomy for bleeding

On the 3rd postop day:

- Arrhythmia
- Blood pressure 80 systolic
- Complaints of dizziness

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ECG

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What is your diagnosis?

1) intra-atrial re-entry tachycardia

2) atrial flutter

3) atrial fibrillation

4) ventricular tachycardia

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ECG

Wide complex tachycardia 200 beats/min

right axis, right bundle brunch block configuration

QRS duration 200 ms

RS pattern in V1,

R/S < 1 in V6

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Diagnosis: VT

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Causes of arrhythmias

‘Natural history’ (abnormal anatomy)

Older age at operation: more fibrosis

Surgical scars

Allways see the surgical report!

Hemodynamic residual lesions: PS, PI, TI, VSD

Long duration of FU / additional pathology (hypertension, coronaries)

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Risk factors for sudden death in Fallot

In older days:

- 3 PVCs on routine ECG
- Trifascicular block
- Elevated pressures in RV

Now:

rotten right ventricle

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Relation QRS-width and sudden death

- Broad QRS: predictor for syncope/ sudden death

→ indicator of RRV = rotten right ventricle

↓

strong correlation with transannular patch

- Increase in QRS width:

predictor for syncope / sudden death

→ indicator of RV worsening

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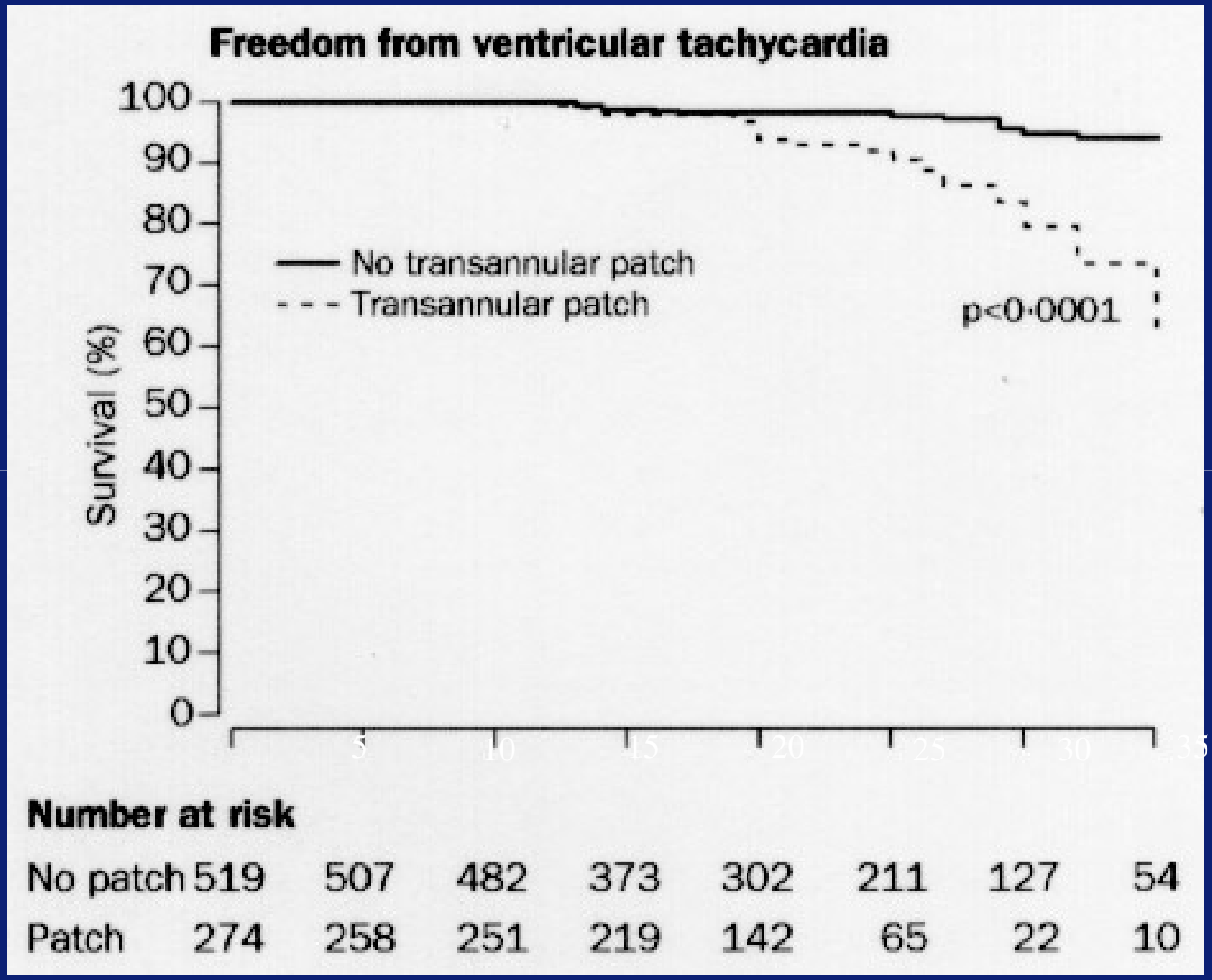
Event-free survival



Gatzoulis et al, Lancet 2000:356:975-81

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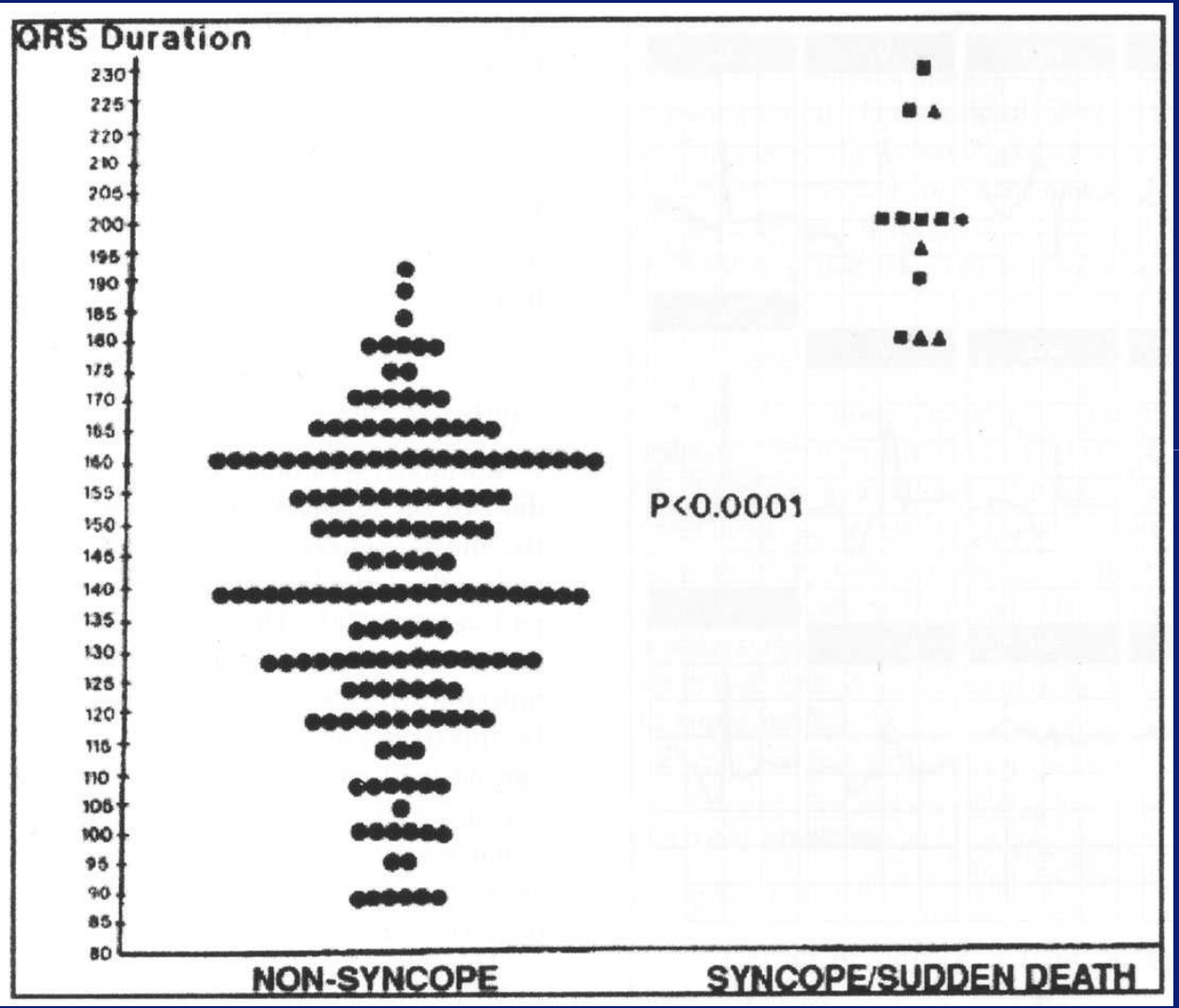
QRS-width and syncope/sudden death



Gatzoulis et al, Circulation 1995;92(20):231-237

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Arrhthmias in older Fallot patients

-34% develop symptomatic atrial or supraventricular tachycardias

-8.5% develop sustained ventricular tachycardia (VT)

-increasing number of implantable defibrillators due to a sudden-death estimate of 2% per decade.

-Thus, an estimated 50,000 adults with repaired Fallot will require electrophysiology follow-up with 100 sudden deaths per year in the USA.

Roos-Hesselink et al. Circulation 1995;91:2214-2219
Lam et al. Electrophysiology issues in adult congenital heart disease Methodist Debakey Cardiovasc J 2011;7:13-7

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Treatment

Electrical cardioversion, start betablockade.

1 day later again VT: ECV

DDD-ICD implantation

After that: no arrhytmias, good clinical condition for 6 years

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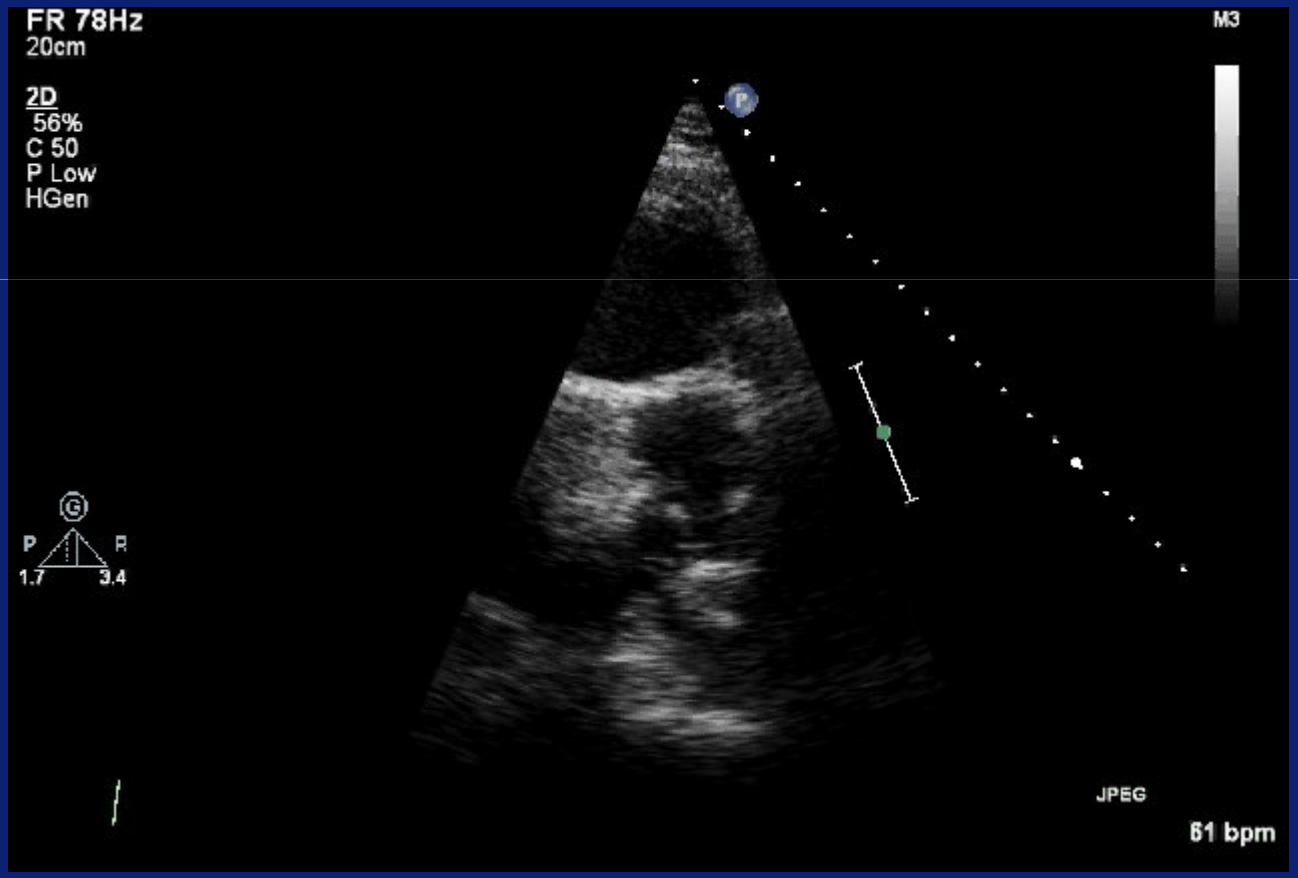
2008 new problem

Endocarditis with oral streptococci.

Antibiotic treatment was started.



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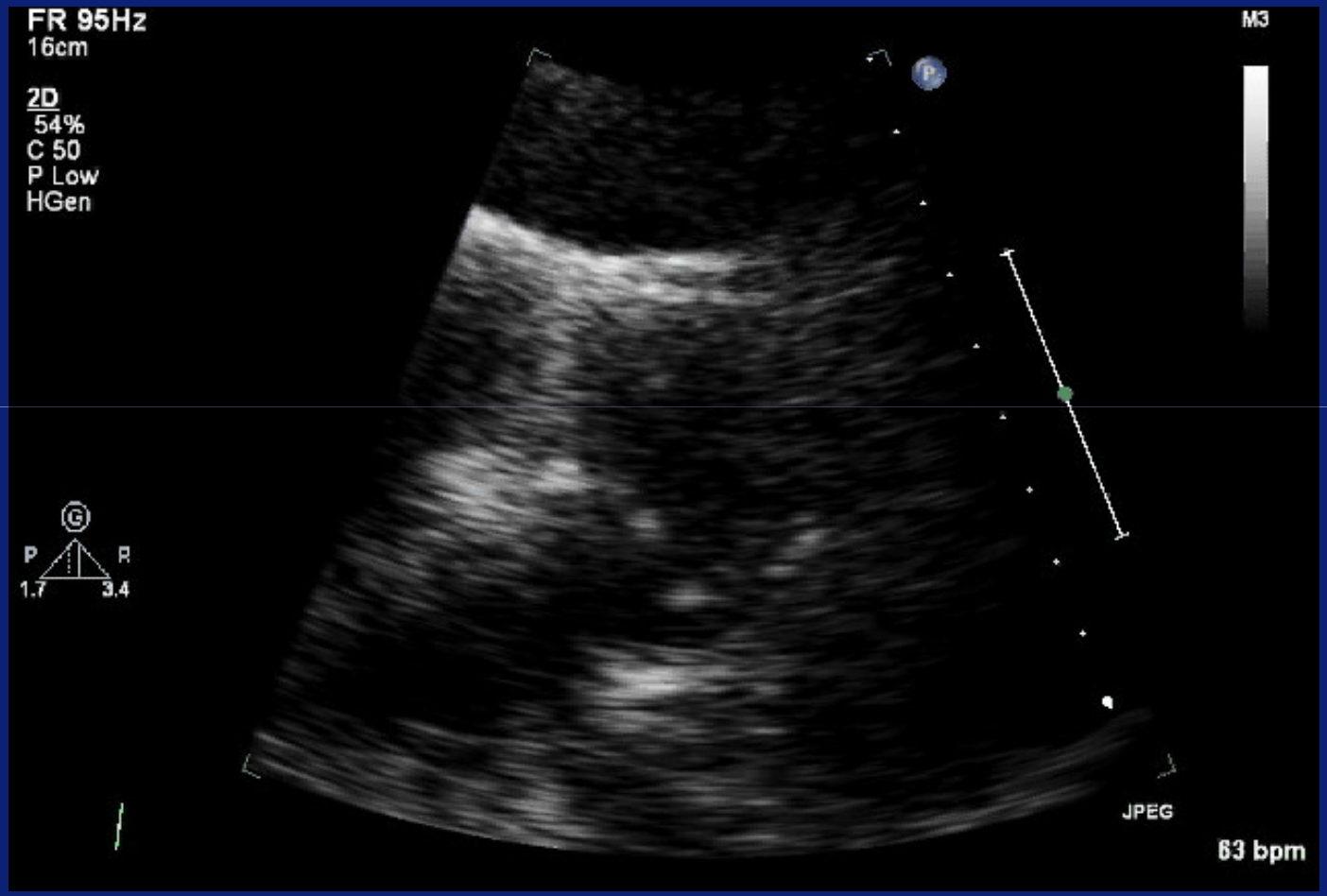
Pulmonary valve



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Should the ICD be removed?

1) no, first 6 weeks of AB treatment, removal may not be necessary

2) yes, immediately, as it is a foreign object

3) remove ICD battery only, leave wires in situ (wires may be re-used for new ICD implantation)

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Our decision

We first tried 6 weeks of antibiotics. The fever disappeared and the patient felt better.

However, 1 month later he developed fever again and we removed the ICD system in total, including the wires. He was treated with AB for 6 weeks afterwards in hospital

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What is your next step?

1) Implant new ICD

2) Implant subcutaneous ICD

3) Stop AB, wait 4 additional weeks in hospital before implantation of new system

4) Send patient home without ICD

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Q4

Our decision

Because he had had no VTs for 6 years, it was decided to let him go home without a new ICD system.

1 year later: VTs on Holter monitoring: a new ICD was implanted

But now a VVI-ICD because of the remaining vegetations on the pulmonary valve

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2012 complaints

Repeated ICD shocks.

ICD rhythm: intra-atrial tachyarrhythmia

Shocks inappropriate!

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European Heart Journal (2007) 28, 1854–1861
doi:10.1093/eurheartj/ehl306

EUROPEAN SOCIETY OF CARDIOLOGY®

Clinical research
Arrhythmia/electrophysiology

Outcome of implantable cardioverter defibrillators in adults with congenital heart disease: a multi-centre study

Sing-Chien Yap[1]*, Jolien W. Roos-Hesselink[1], Elke S. Hoendermis[2], Werner Budts[3], Hubert W. Vliegen[4], Barbara J.M. Mulder[5,6], Arie P.J. van Dijk[7], Martin J. Schalij[4], and Willem Drenthen[2]

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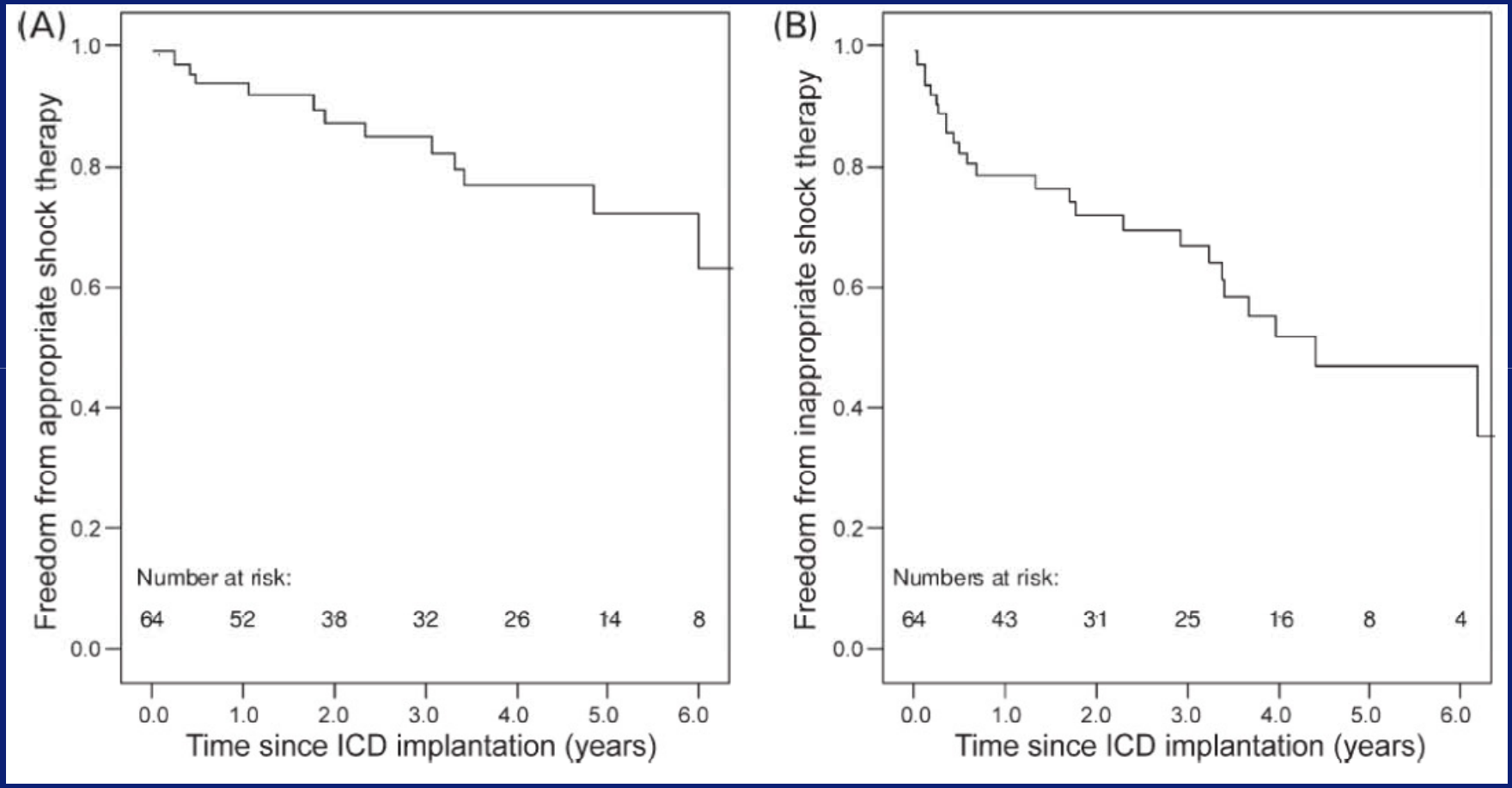
Freedom from appropriate inappropriate shocks



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Is ablation of SVT a good option?

1) yes, it is a safe and effective therapy

2) no, results of ablation are disappointing

3) no, medication should be optimized first

4) no, upgrade the VVI-ICD to DDD-ICD

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Prevalence of arrhythmias (CONCOR)

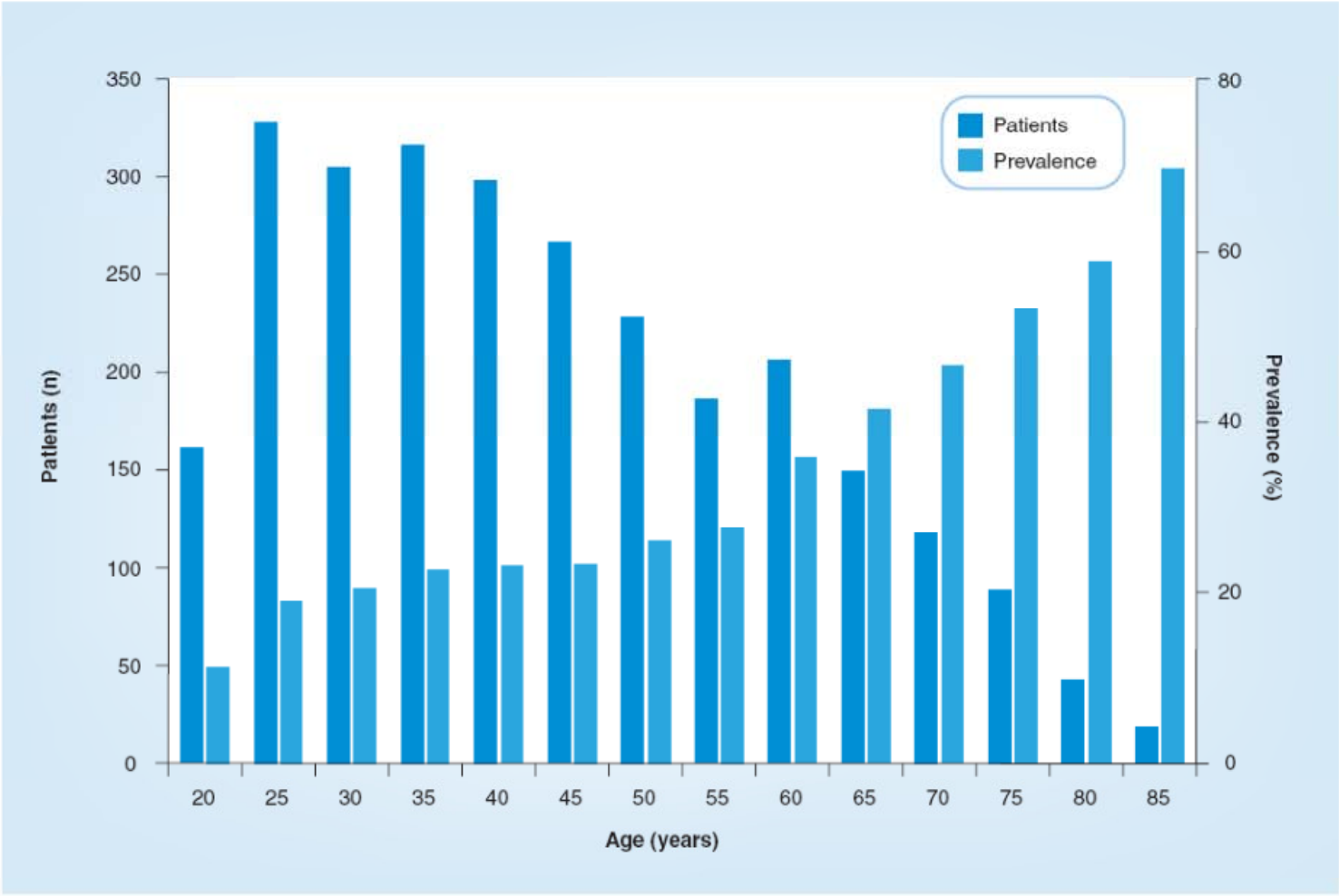

Figure 1. Age distribution of CONCOR® patients with arrhythmia.

Koyal et al. Expert Rev. Cardiovasc. Ther. 8(12), 1753–1766 (2010)

Current literature

The Electroanatomic Mechanisms of Atrial Tachycardia in Patients with Tetralogy of Fallot and Double Outlet Right Ventricle

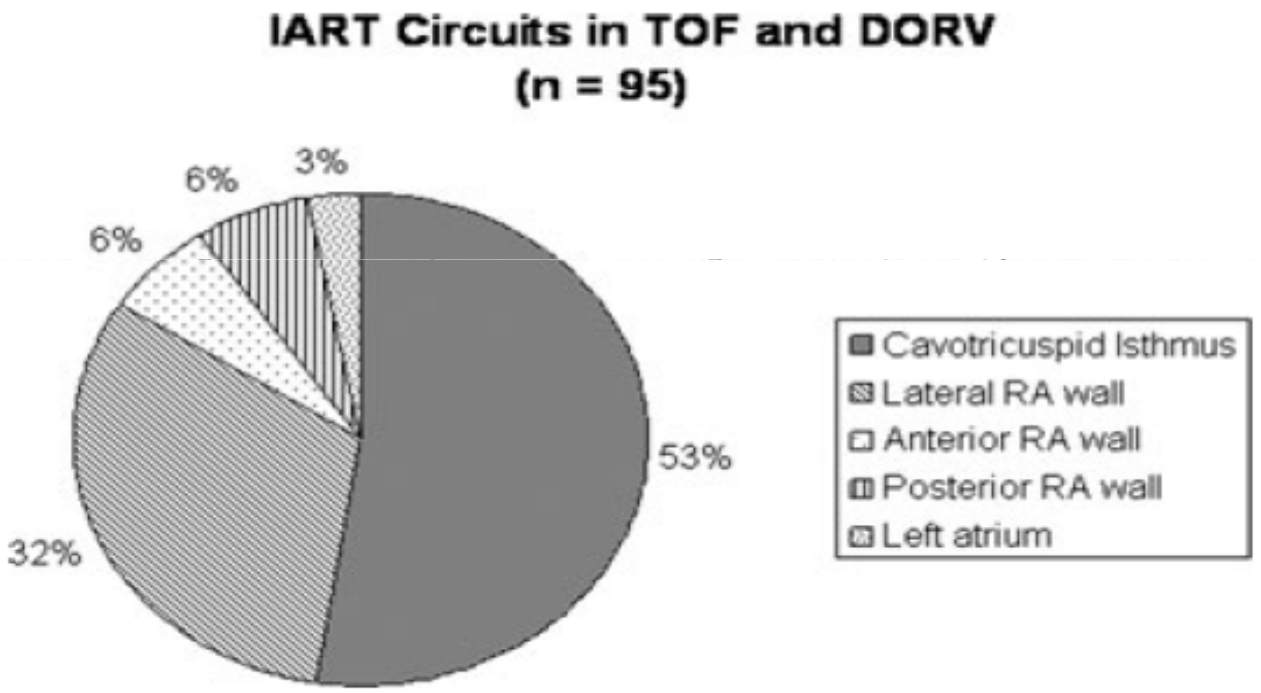

Figure 1. *Location of critical regions of ablation for IART circuits in patients with TOF or DORV (n = 95).*

Mah et al. J Cardiovasc Electrophysiol, Vol. 22, pp. 1013-1017, September 2011

Results

Acute Success in Ablating Atrial Tachycardias

	Acute Success	Unable to Ablate
IART		
Cavotricuspid isthmus	47	3
Lateral RA	30	0
Anterior RA	2	4
Posterior RA	6	0
Left atrium	0	3
Focal atrial tachycardia	12	1
Ectopic atrial tachycardia	4	0
Atrial fibrillation	0	15
Unmapped + mapped with unavailable documentation	4	18
Total	105	44

Mah et al. J Cardiovasc Electrophysiol, Vol. 22, pp. 1013-1017, September 2011

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Acute success for RF ablation

Author (year)	Patients (n)	IART circuits (n)	Acute success (%)	Mapping technique
De Groot *et al.* (2009)	19	30	73	3D electroanatomic
Tanner *et al.* (2004)	36	52	87	Entrainment and electroanatomic
De Groot *et al.* (2001)	17		85	3D electroanatomic
Zrenner *et al.* (2003)	12	14	86	Entrainment and electroanatomic
Kannankeril *et al.* (2003)	40	47	87	
Triedman *et al.* (2002)	134	369	66	Entrainment and electroanatomic
Delacretaz *et al.* (2001)	20	47	81	Entrainment and 3D electroanatomic
Zrenner *et al.* (2001)	25	43	91	Entrainment
Collins *et al.* (2000)	88	110	71–80	Entrainment
Chan *et al.* (2000)	19	21	90	Entrainment
Kanter *et al.* (2000)	11	13	73	Entrainment
Triedman *et al.* (1997)	45		73	Entrainment
Kalman *et al.* (1996)	18	26	81	Entrainment
Van Hare *et al.* (1996)	10	13	77	Entrainment
Triedman *et al.* (1995)	10	30	77	Entrainment

IART: Intra-atrial re-entrant tachycardia.

Koyak et al. Expert Rev. Cardiovasc. Ther. 8(12), 1753–1766 (2010)

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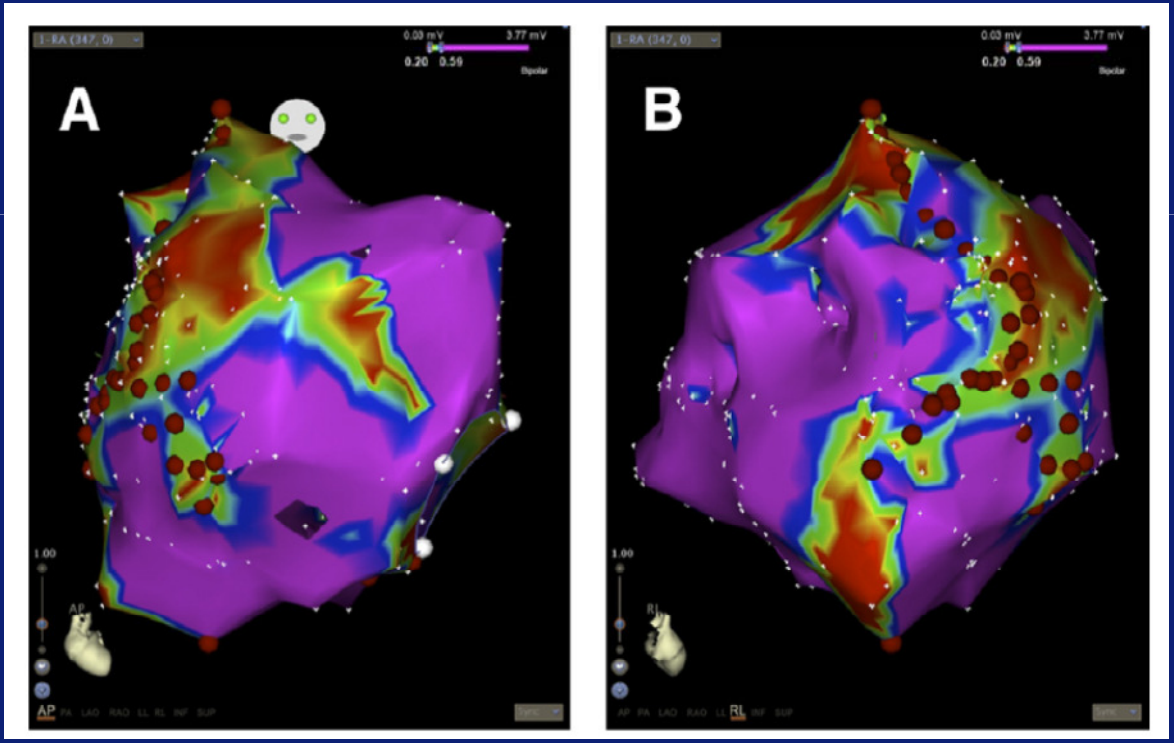
Acute and Long-Term Outcomes of Catheter Ablation Using Remote Magnetic Navigation in Patients With Congenital Heart Disease

Ferdi Akca, Tamas Bauernfeind, MD, Maarten Witsenburg, MD, Lara Dabiri Abkenari, MD, Judith A. Cuypers, MD, Jolien W. Roos-Hesselink, MD, PhD, Natasja M.S. de Groot, MD, PhD, Luc Jordaens, MD, PhD, and Tamas Szili-Torok, MD, PhD*



(Am J Cardiol 2012;xx:xxx)

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Baseline characteristics

Age (years)	
Mean ± SD	35 ± 19
Range	2–77
Gender	
Male	21
Female	15
Congenital heart disease complexity	
Grade I	4 (11%)
Grade II	15 (42%)
Grade III	17 (47%)
Amiodarone prescription	8 (22%)
Sotalol prescription	14 (39%)
β Blocker prescription	8 (22%)
Digoxin prescription	1 (3%)
Verapamil prescription	3 (8%)
Heart surgery (number of operations)	1.7 ± 0.8

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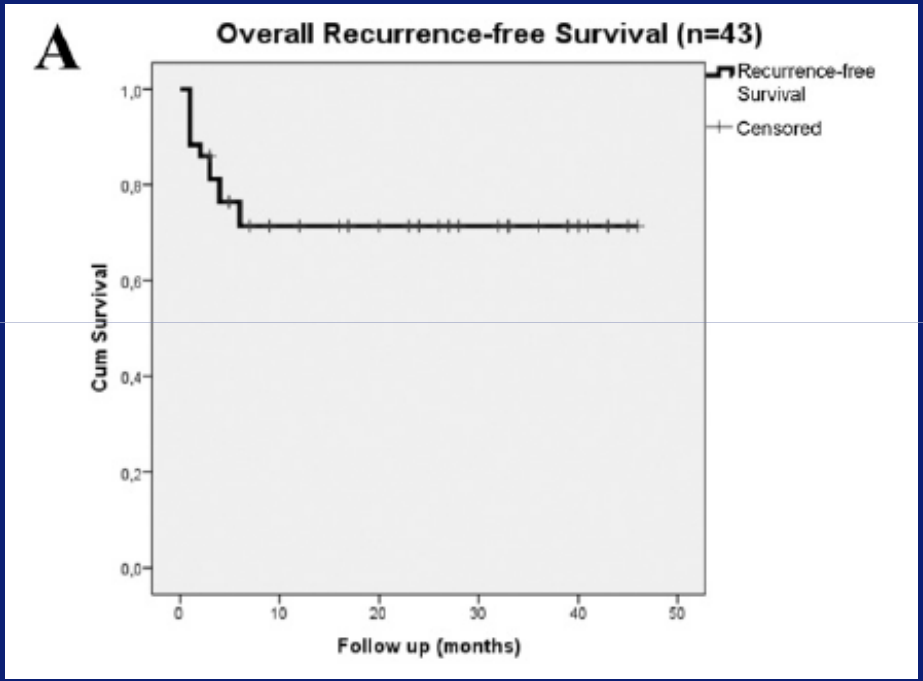
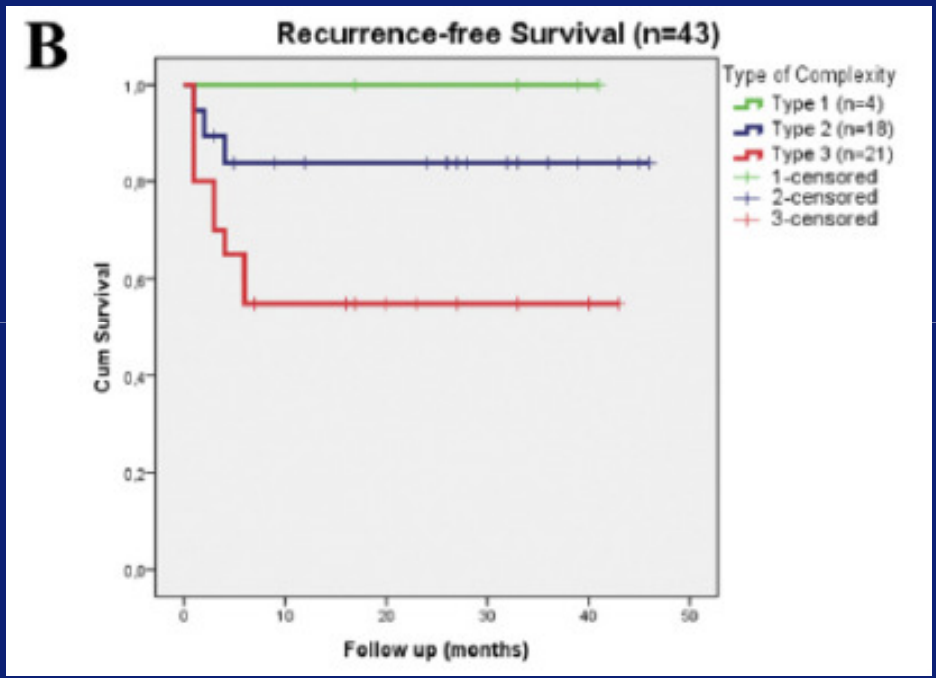
Am J Cardiol 2012;xx:xxx

Results: efficacy

Overall



per diagnosis



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Results: Safety

Fluoroscopy time	40 min
Procedure time	216 min
Mean numbers of RF applications	42

No major complications

3 minor complications: all groin hematomas

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Our patient:

successful RF ablation in 2012

Needs now psychological support

Psychosocial impact of implantable cardioverter defibrillators (ICD) in young adults with Tetralogy of Fallot. *Opic et al in press*

“In patients with Fallot, ICD implantation had a major impact on psychosocial functioning.”

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Conclusions

1. Arrhythmias are common in patients with congenital heart disease
2. Atrial arrhythmias are most frequent and the major cause of morbidity and mortality
3. Ventricular arrhythmias are potentially life threatening, ICD implant may be necessary
4. Beware of the high rate of inappropriate shocks and psychological impact in these young patients
5. Ablation is safe and effective

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Arrhythmias and congenital heart disease

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