Risk stratification to avoid unnecessary ICD in patients with structural CHD

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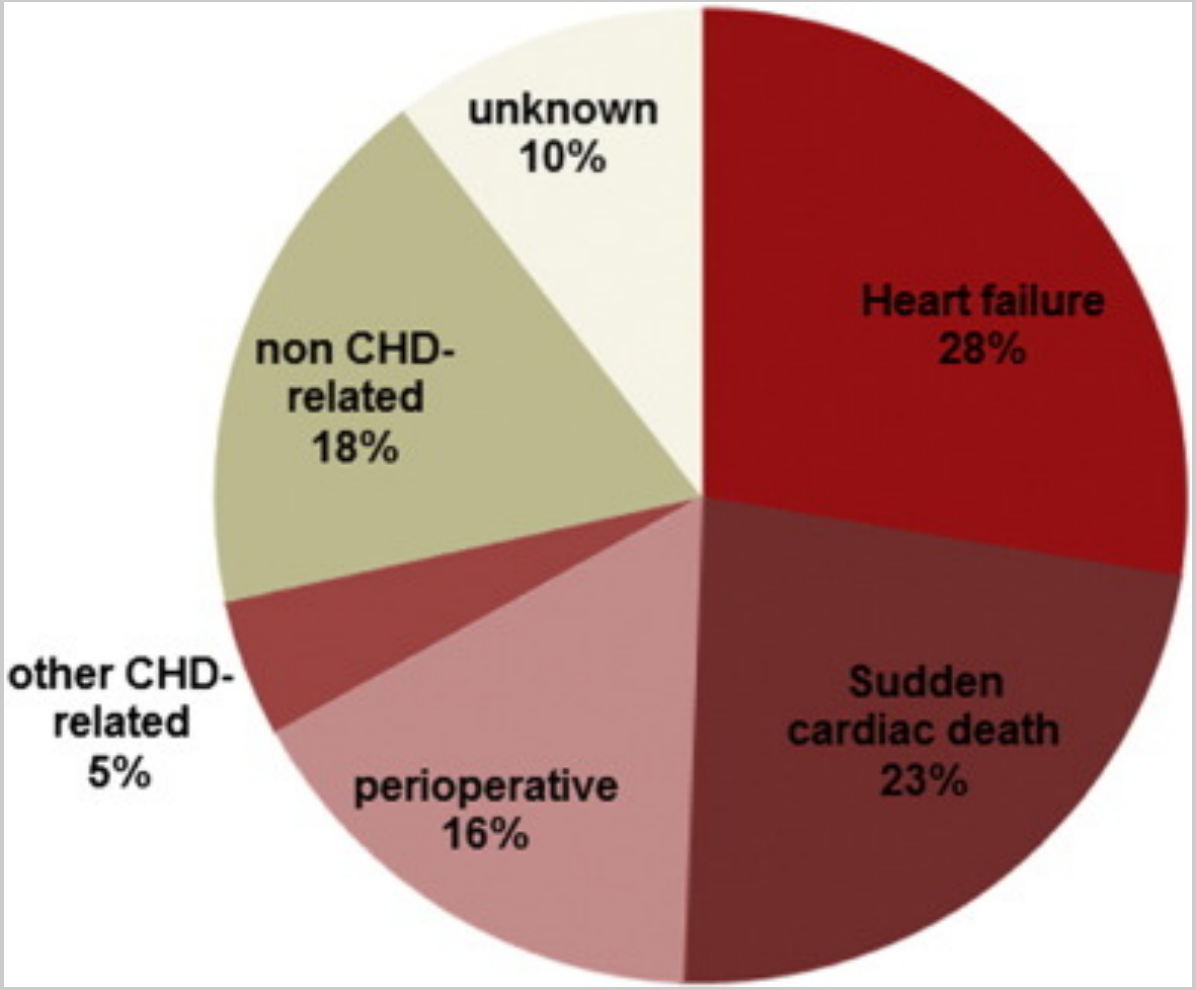
Pedirhythm VII, Thessaloniki, Greece, February 2017

Cause of death in adults with congenital heart disease — An analysis of the German National Register for Congenital Heart Defects

Engelings et al, Int J Cardiol Vol 211, 2016: 31–36



Implantable defibrillators in CHD

- ICD therapy has proven its efficacy as secondary prevention in patients with CHD and sustained ventricular tachycardia or resuscitated cardiac arrest in the absence of a reversible cause

- Risk–benefit assessment for primary prevention ICDs is a major challenge, due to the small number of cases, multiple different types of CHD and potential complications related to these devices

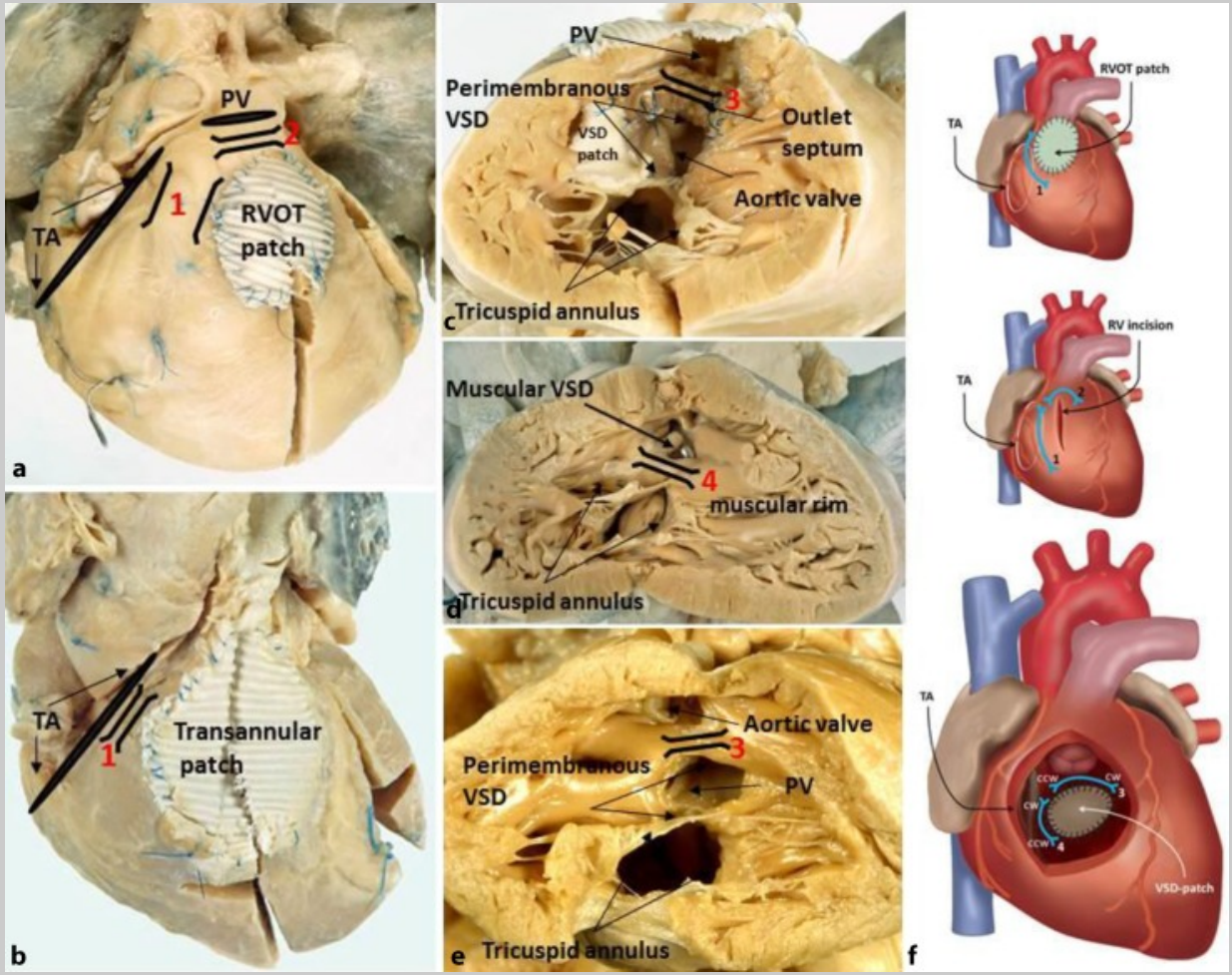
Key factors in weighing risks and benefits of implantable cardioverter-defibrillator (ICD) therapy

- ▸ Estimated risk of sudden cardiac death due to malignant ventricular arrhythmias
- ▸ Competing risks for mortality
- ▸ Effectiveness of ICD in preventing sudden cardiac death
- ▸ Morbidity and mortality associated with ICD therapy
- ▸ Cost considerations

VT/SCD: Most common culprits

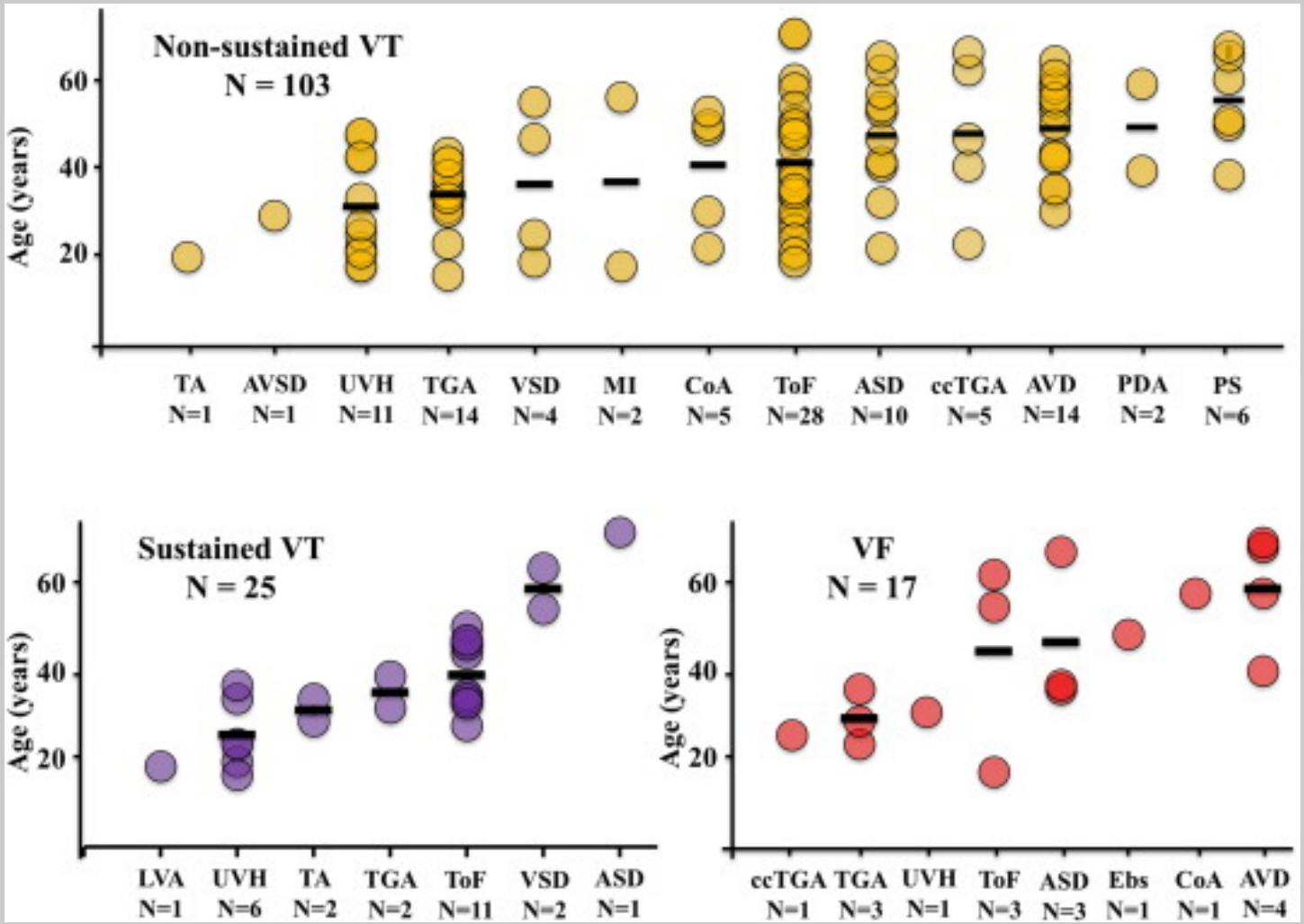
- Tetralogy of Fallot and related lesions
- LV obstructive lesions
- Systemic RV (D-TGA s/p Mustard/Senning, L-TGA)
- Single ventricle
- Eisenmenger syndrome
- Other, e.g. Ebstein's anomaly

Anatomical isthmuses and VT in repaired CHD

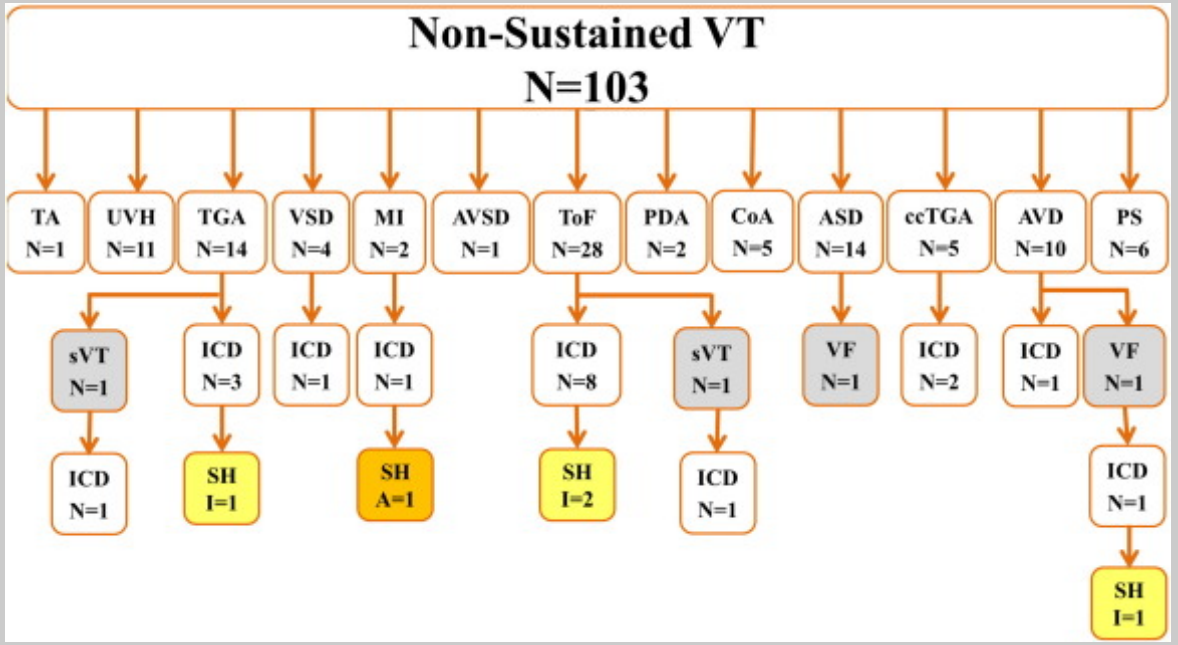


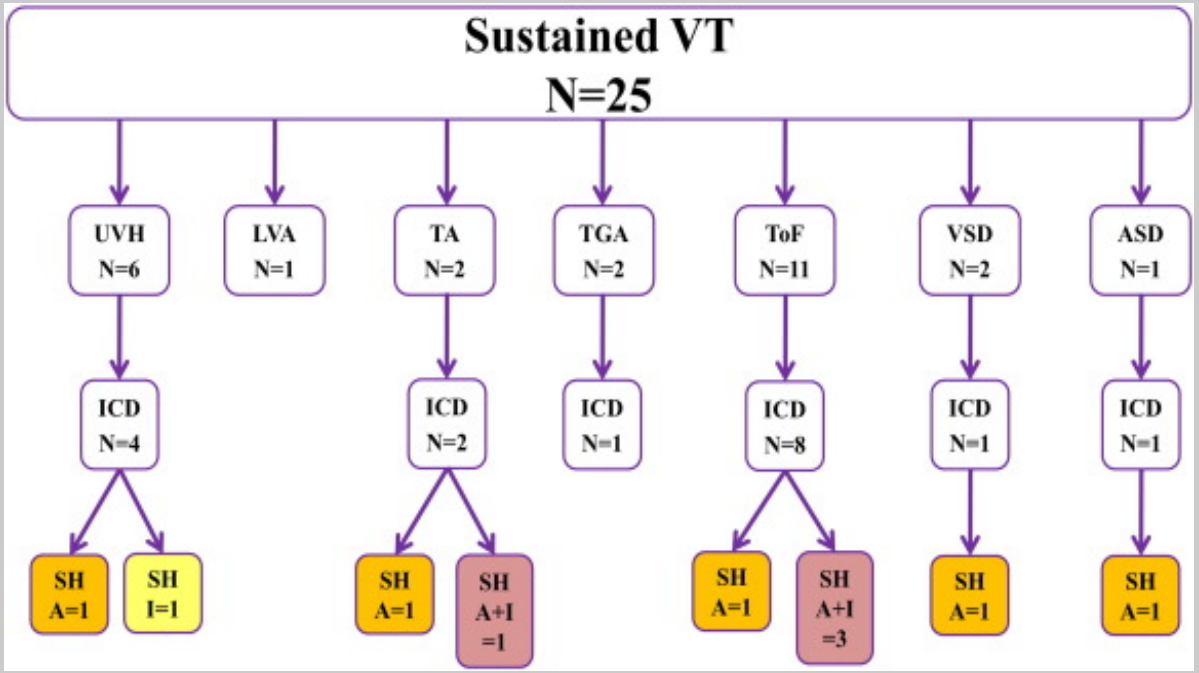
Zeppenfeld K, Herzschrittmacherther Elektrophysiol 2016; 27: 131–136.

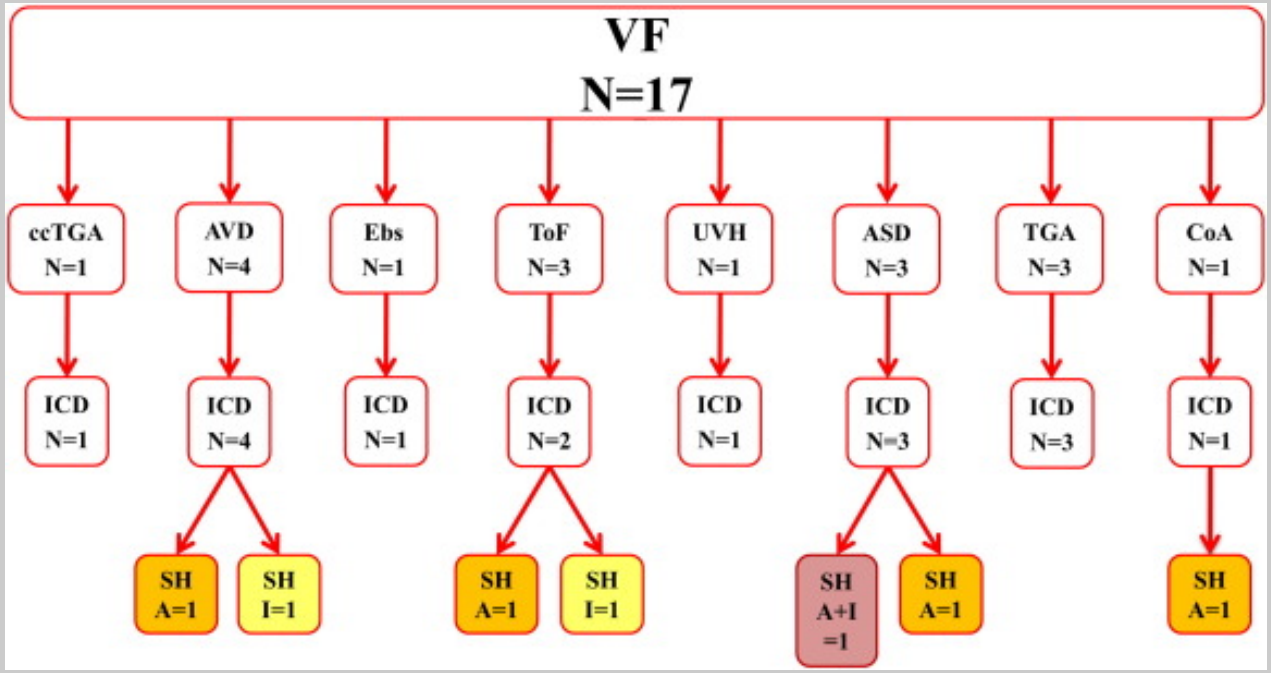
Is non-sustained VT important?



Tewen CP et al, International Journal of Cardiology, Vol 206, 2016:158–163


Non-Sustained VT
N=103
TA N=1
UVH N=11
TGA N=14
VSD N=4
MI N=2
AVSD N=1
ToF N=28
PDA N=2
CoA N=5
ASD N=14
ccTGA N=5
AVD N=10
PS N=6
sVT N=1
ICD N=3
ICD N=1
ICD N=1
ICD N=8
sVT N=1
VF N=1
ICD N=2
ICD N=1
VF N=1
ICD N=1
SH I=1
SH A=1
SH I=2
ICD N=1
ICD N=1
SH I=1


Sustained VT
N=25
UVH N=6
LVA N=1
TA N=2
TGA N=2
ToF N=11
VSD N=2
ASD N=1
ICD N=4
ICD N=2
ICD N=1
ICD N=8
ICD N=1
ICD N=1
SH A=1
SH I=1
SH A=1
SH A+I =1
SH A=1
SH A+I =3
SH A=1
SH A=1


VF
N=17
ccTGA N=1
AVD N=4
Ebs N=1
ToF N=3
UVH N=1
ASD N=3
TGA N=3
CoA N=1
ICD N=1
ICD N=4
ICD N=1
ICD N=2
ICD N=1
ICD N=3
ICD N=3
ICD N=1
SH A=1
SH I=1
SH A=1
SH I=1
SH A+I =1
SH A=1
SH A=1

Asymptomatic NS-VT is not a risk factor for SCD/VT in patients with TOF

S. Cullen, D.S. Celermajer, R.C. Franklin, K.A. Hallidie-Smith, J.E. Deanfield

Prognostic significance of ventricular arrhythmia after repair of tetralogy of Fallot: a 12-year prospective study

J. Am. Coll. Cardiol., 23 (1994), pp. 1151–1155

M.A. Gatzoulis, S. Balaji, S.A. Webber, *et al.*

Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study

Lancet, 356 (2000), pp. 975–981

Symptomatic NS-VT in CHD patients is a predictor for appropriate shocks in ICD for 1^{o} prevention

P. Khairy, L. Harris, M.J. Landzberg, *et al.*

Implantable cardioverter-defibrillators in tetralogy of Fallot

Circulation, 117 (2008), pp. 363–370

Z. Koyak, J.R. de Groot, I.C. Van Gelder, *et al.*

Implantable cardioverter defibrillator therapy in adults with congenital heart disease: who is at risk of shocks?

Circ. Arrhythm. Electrophysiol., 5 (2012), pp. 101–110

ICDs in postoperative TOF

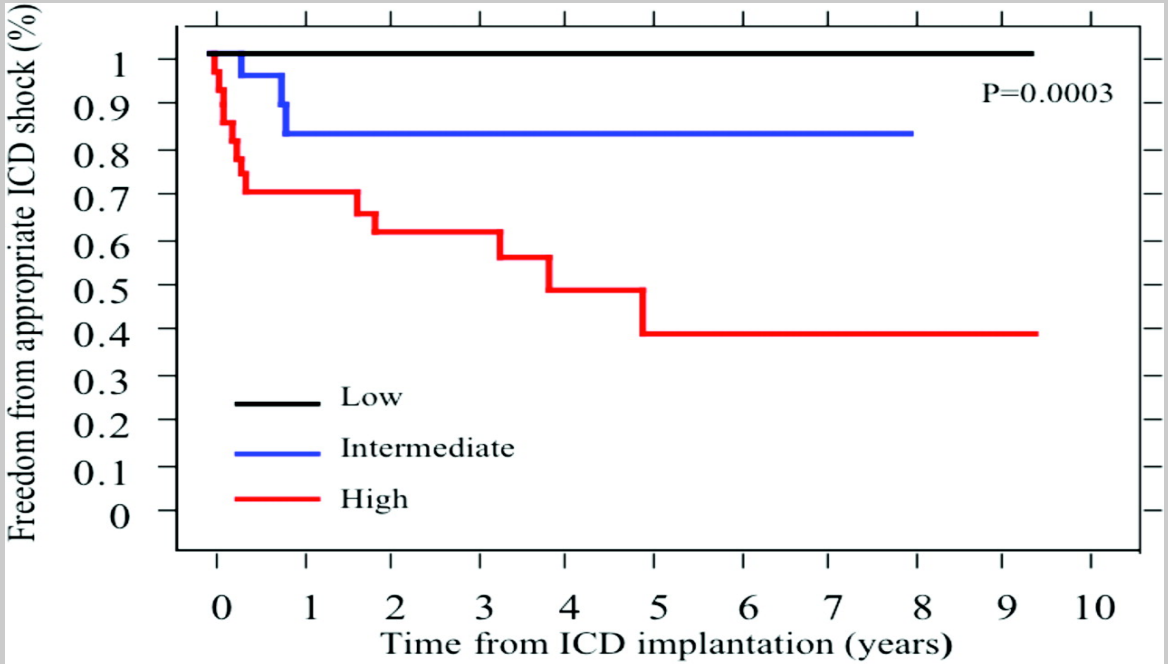
- 121 pts (median 33 yrs, 3,7 yrs F/U)
- 56% primary, 44% secondary prevention (sustained VT or resuscitated SCD)
- 30% at least 1 appropriate shock (actuarial annual rate of 8%)
- Predictors of appropriate shock in primary prevention: Higher LVEDP and non-sustained VT
- Complications: 30% (including 5.8% per year inappropriate shocks)
- Mortality: 2.2% per year

Khairy P et al, Circulation 2008;117:363-70

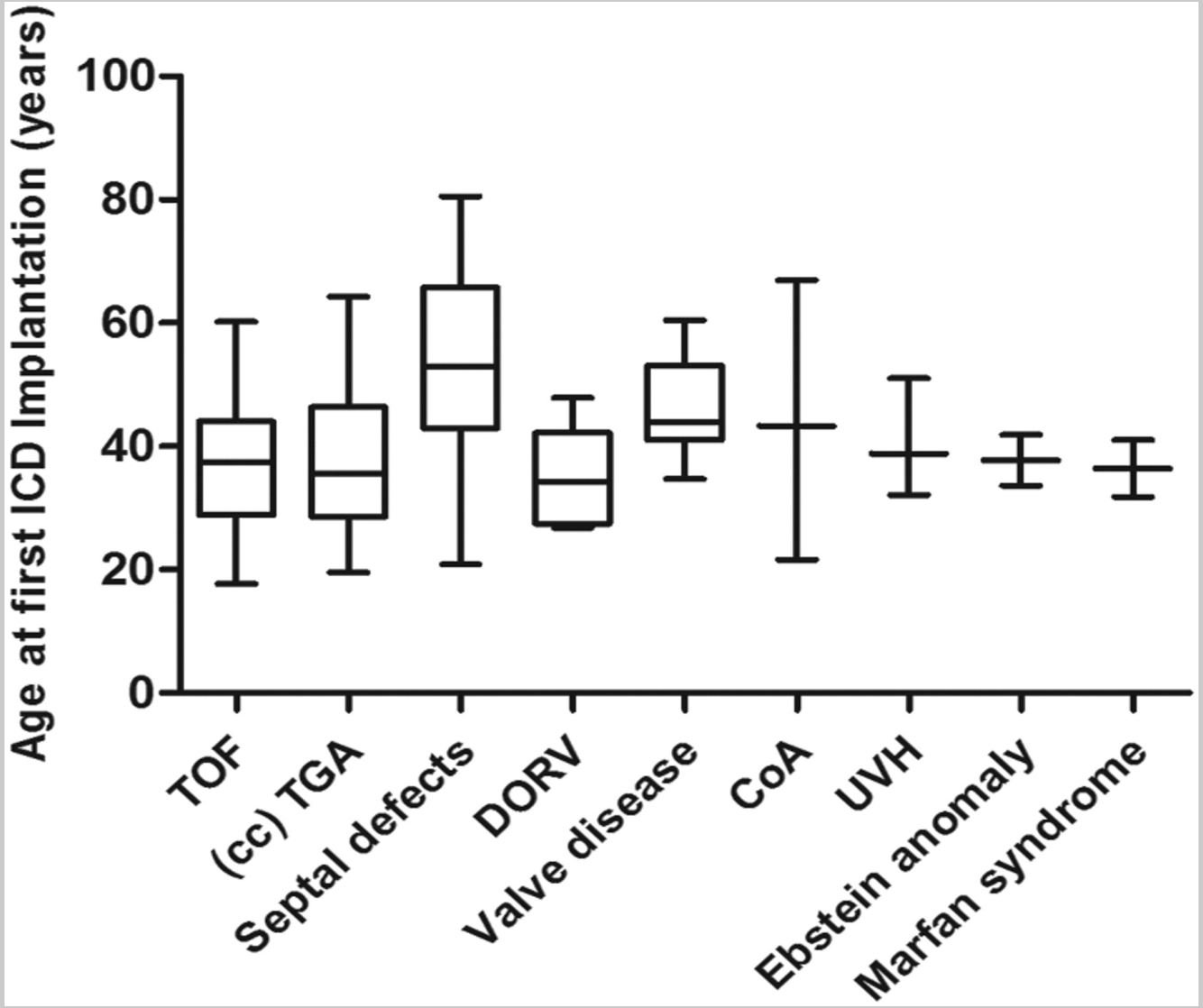
ICDs in postoperative TOF

Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention

Variable	Exp(β)	Points Attributed
Prior palliative shunt	3.2	2
Inducible sustained ventricular tachycardia	2.6	2
QRS duration ≥180 ms	1.4	1
Ventriculotomy incision	3.4	2
Nonsustained ventricular tachycardia	3.7	2
LVEDP ≥12 mm Hg	4.9	3
Total points	...	0–12

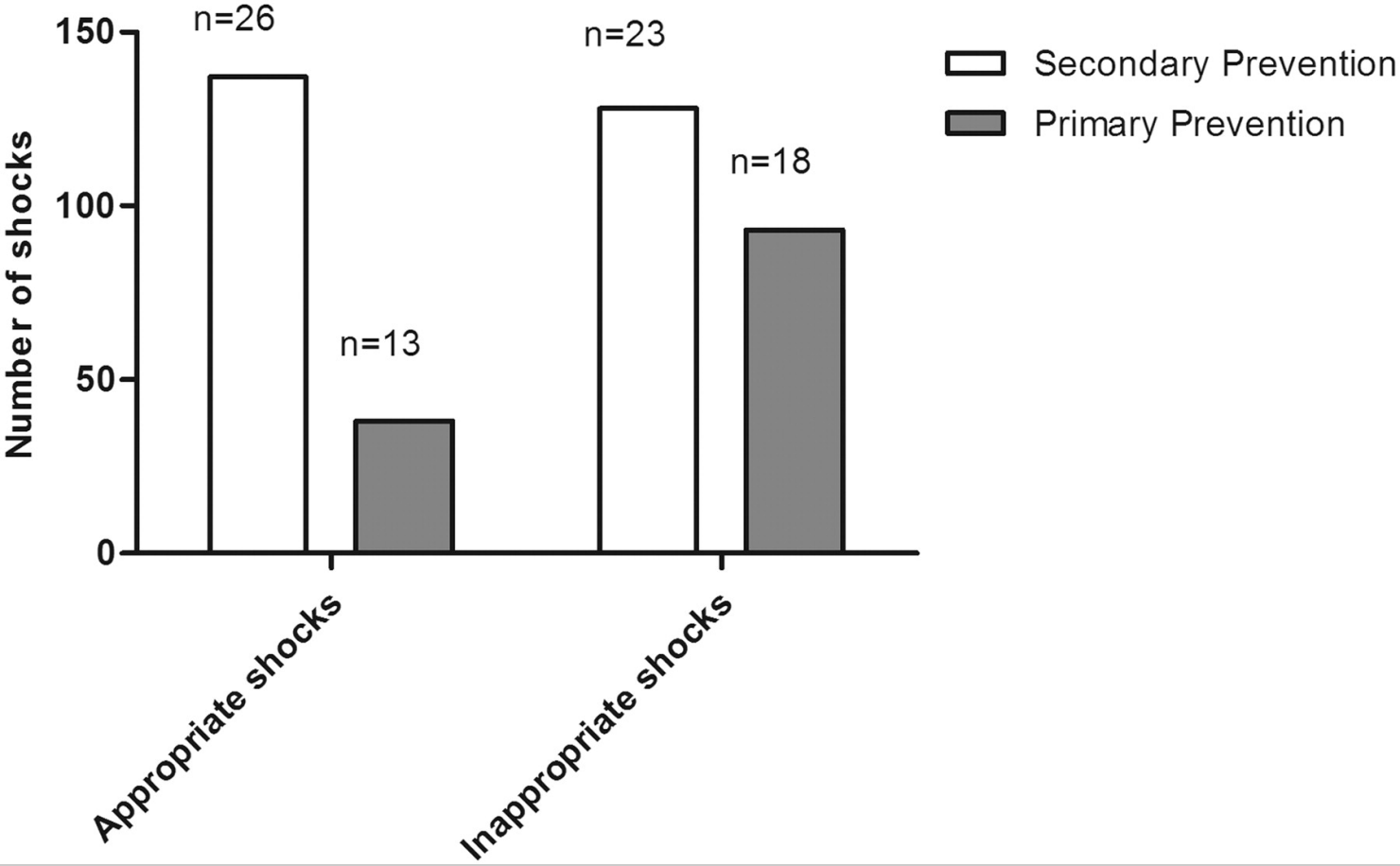


Risk score	Risk category	N	Annualized rate of appropriate shocks
0-2	Low	18	0%
3-5	Intermediate	24	3.8%
6-12	High	26	17.5%



Zeliha Koyak et al. Circ Arrhythm Electrophysiol. 2012;5:101-110

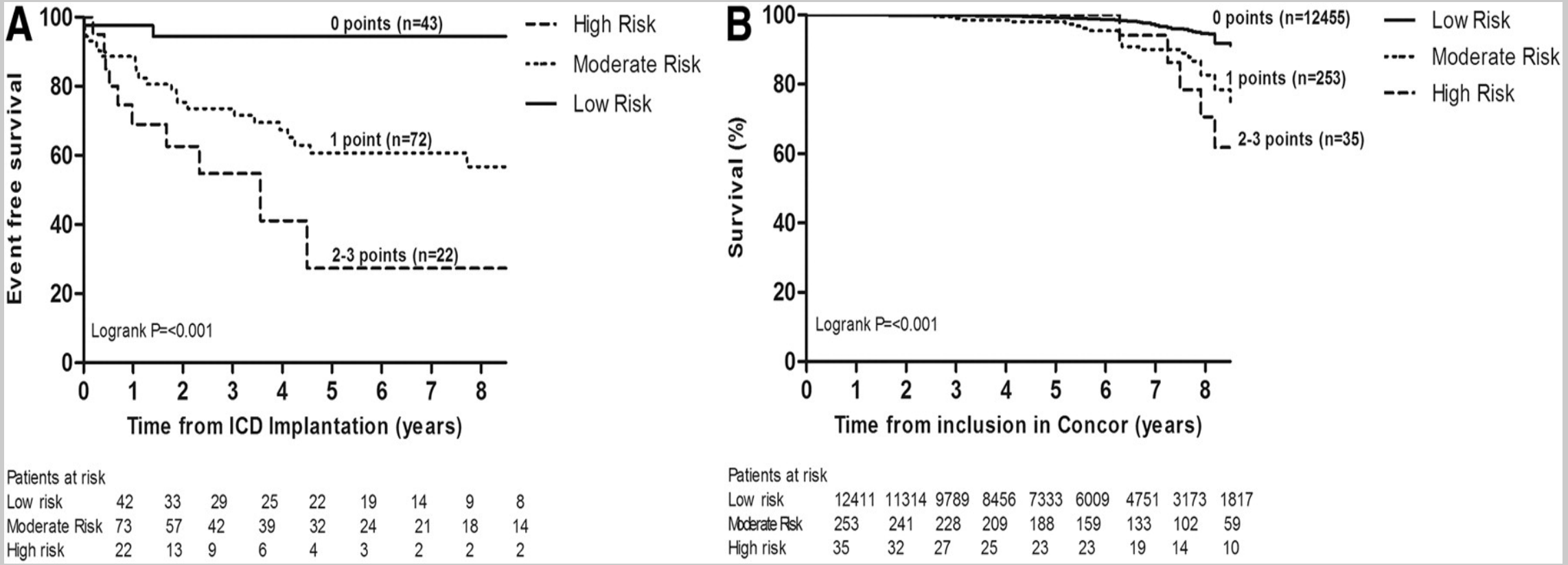
Appropriate vs inappropriate shocks in 1º and 2º prevention



Risk Score for Appropriate ICD Shocks

Variable	HR	Points Attributed
Secondary prevention indication	3.6	1
Documented CAD	2.7	1
Symptomatic NSVT	9.1	2

A. Event-free survival for a first appropriate implantable cardioverter defibrillator (ICD) shock according to an increasing risk score in all patients. **B**, Survival in relation to the presence of risk factors in patients without an ICD in the Concor database.



Zeliha Koyak et al. Circ Arrhythm Electrophysiol. 2012;5:101-110

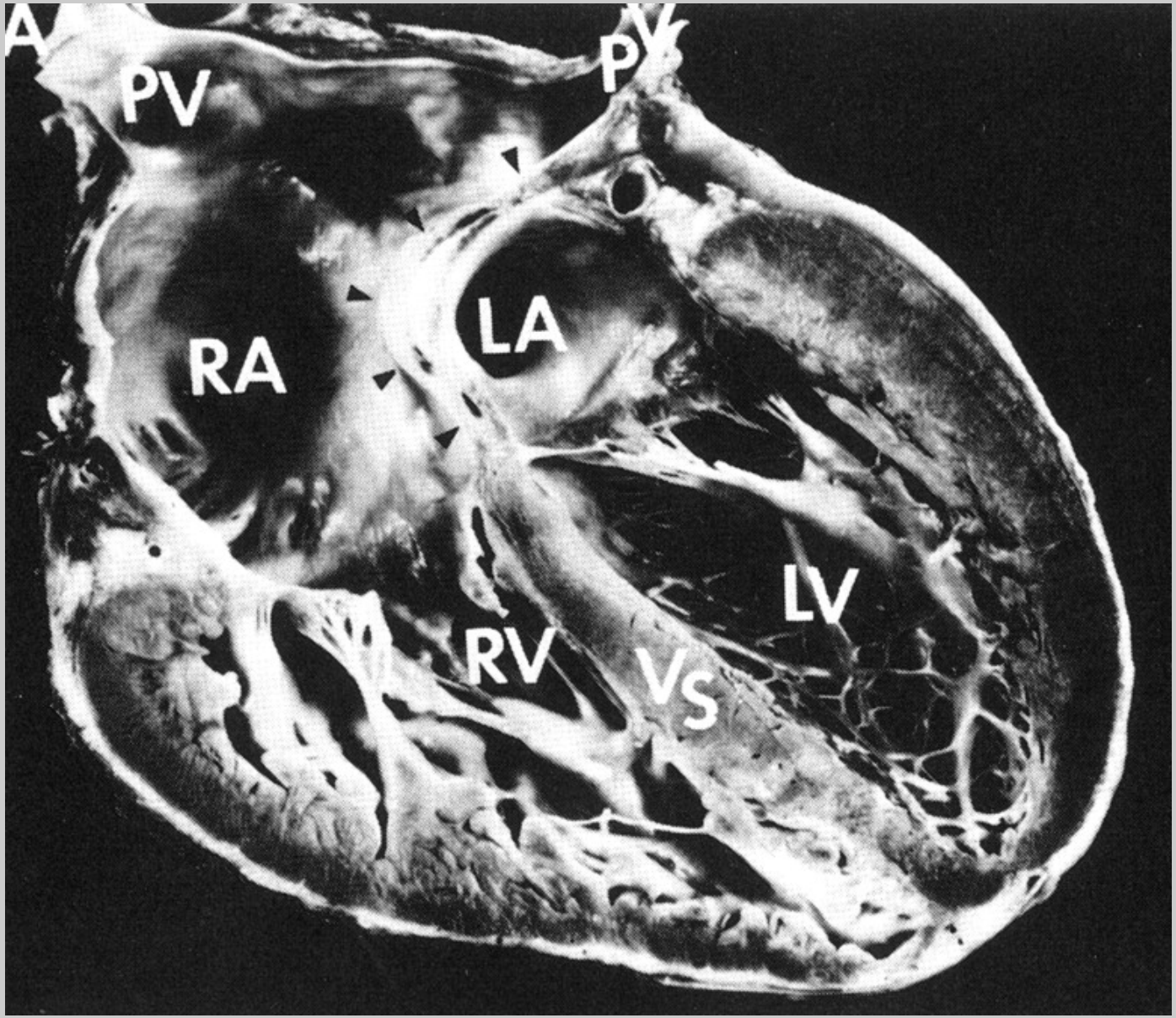
American Heart Association

Primary prevention ICD indications for TOF

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

Khairy et al, Heart Rhythm, 2014-10-01, Volume 11, Issue 10, Pages e102-e165

- Class IIa (Level of evidence: B)
- ICD therapy is reasonable in selected adults with tetralogy of Fallot and multiple risk factors for sudden cardiac death
  - left ventricular systolic or diastolic dysfunction
  - nonsustained ventricular tachycardia
  - QRS duration 180 ms
  - extensive right ventricular scarring
  - inducible sustained ventricular tachycardia at electrophysiologic study

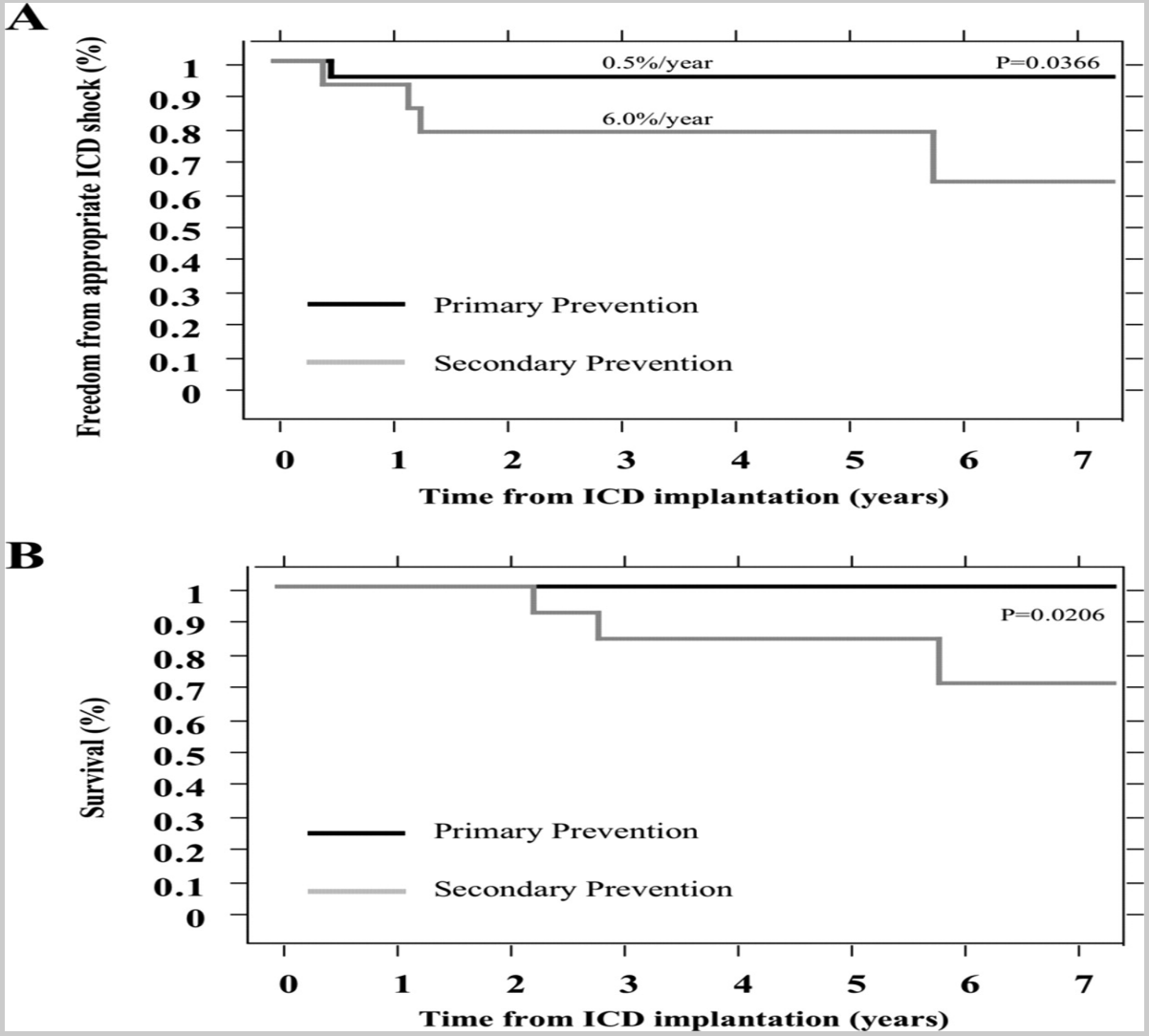

A
PV
PV
RA
LA
LV
RV
Vs

Sudden Death and Defibrillators in Transposition of the Great Arteries With Intra-atrial Baffles

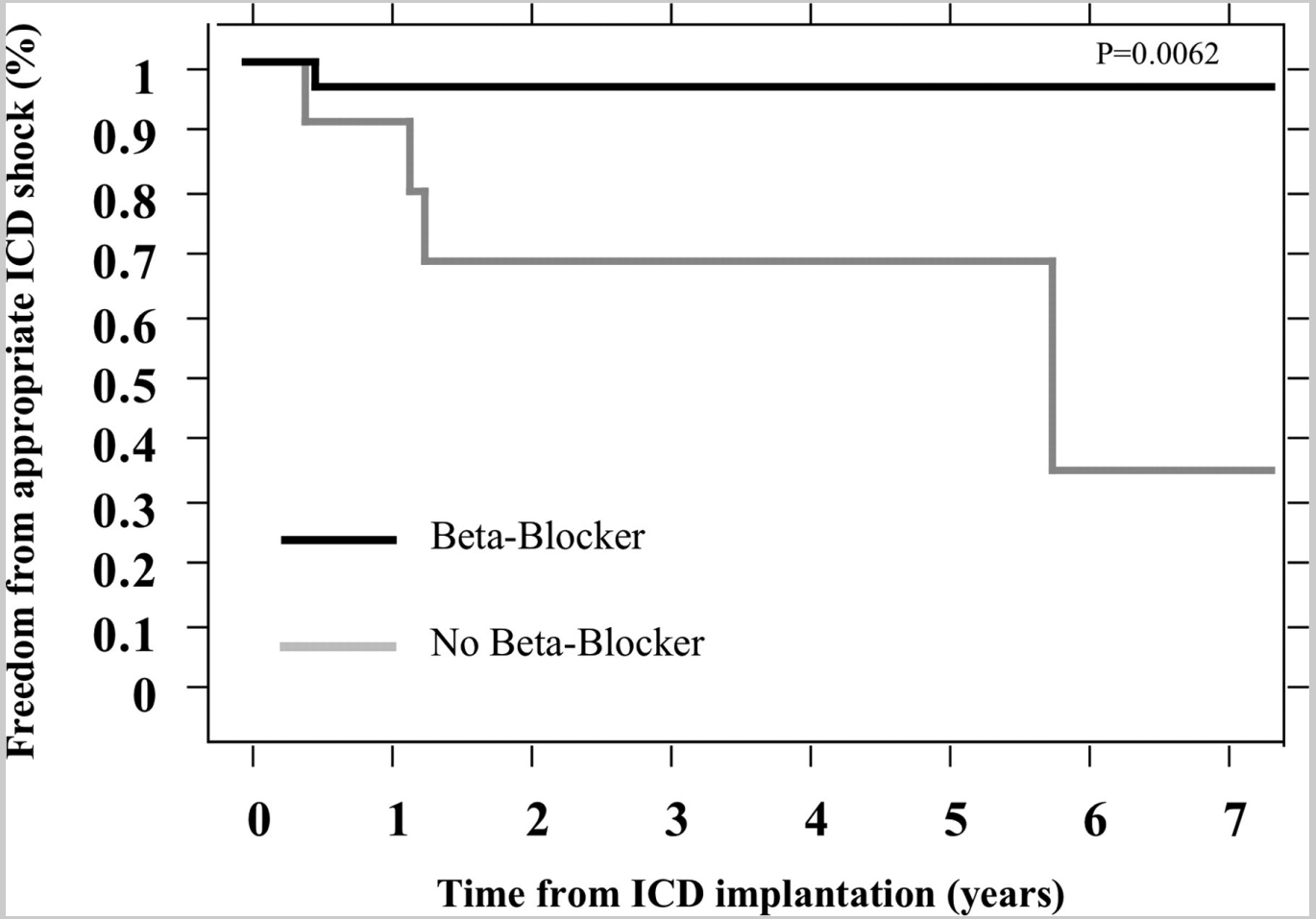
- 37 pts (age, 28.0±7.6 yr) from 7 centers
- ICDs implanted for primary prevention in 23 (62.1%) patients and secondary prevention in 14 patients (37.8%).
- Annual rates of appropriate shocks were 0.5% and 6.0% in primary and secondary prevention, respectively (*P*=0.0366).
- Independent predictors: secondary prevention indication (hazard ratio, 18.0; *P*=0.0341) and lack of β-blockers (hazard ratio, 16.7; *P*=0.0301). ***Inducible VT during EPS not a predictor***
- Intracardiac electrograms documented SVT preceding or coexisting with VT in 50%.
- Inappropriate shocks occurred in 6.6% per year
- 14 patients (37.8%) experienced complications

- Khairy et al, Circ Arrhythmia Electrophysiol *2008; 1: 250-257*

Appropriate ICD shocks (A) and survival (B) in 1º and 2º prevention



Appropriate ICD shocks in patients with and without β-blockers



Predictors of sudden cardiac death after Mustard or Senning repair for transposition of the great arteries

- Retrospective, multicenter, case-controlled study
- 47 patients after Mustard or Senning operation who experienced an SD event
- Symptoms of arrhythmia or CHF and history of documented A Fib or A Flutter were found to increase the risk of SD.
- Neither medication nor pacing was found to be protective. Most SD events (81%) occurred during exercise.
- VT/VF recorded during SD in 21 of 47 patients.

Kammeraad et al, JACC;44(5):1095-102.

Sudden death after Fontan operation

- 261 patients, first Fontan at 7.9 years of age
- Over a median of 12.2 years, 76 (29.1%) died
- Actuarial freedom from death or transplantation was 93.7 and 82.6% at 5 and 20 years respectively with no significant difference between types of Fontan (AP, AV or TCPC)
- **Late deaths** were classified as **sudden** in 7 patients **(9.2%)**, with no predictors identified
- Two patients with sudden death had previously documented intra-atrial reentrant tachycardia
- Absence of aspirin or warfarin therapy shown to be a powerful independent predictor of mortality from thromboembolism

Khairy et al, Circulation. 2008 Jan 1;117(1):85-92

ICD recommendations for single or systemic RV

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

Khairy et al, Heart Rhythm, 2014-10-01, Volume 11, Issue 10, Pages e102-e165

- Class IIB
- ICD therapy may be reasonable in adults with a *single or systemic right ventricular ejection fraction* <35%, particularly in the presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA functional class II or III symptoms, QRS duration ≥140 ms, or severe systemic AV valve regurgitation

Sudden death in other CHD

- Ebstein's anomaly
- Wolff-Parkinson-White syndrome
- Atrial tachycardia
- Ventricular tachycardia

- Aortic or subaortic stenosis
- Related to degree of stenosis and usually of ischemic etiology
- Ventricular arrhythmias in neglected cases

- *Treatment of underlying anatomic problems with tricuspid and aortic valve and ablation of accessory pathways largely prevents sudden cardiac death*
- *Assess risk of VT*

Thank you for coming to Thessaloniki!