Sudden cardiac death in children and adolescents: Etiology and treatment

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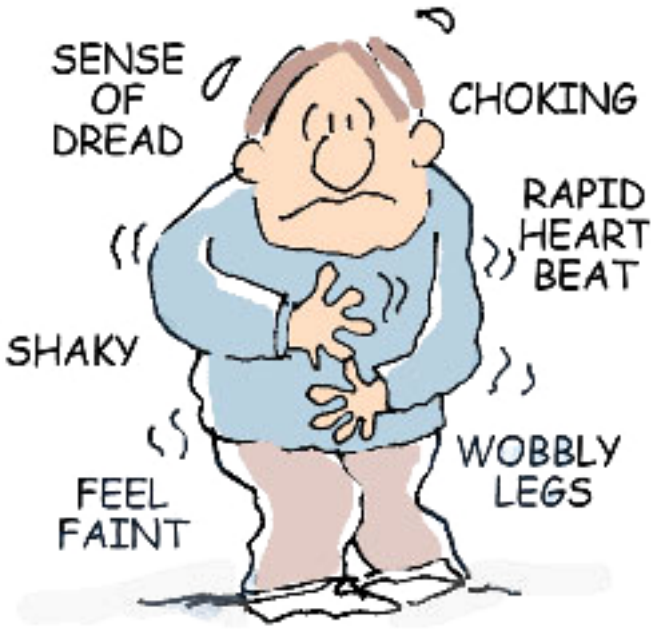


Sudden cardiac death

Children and adolescents

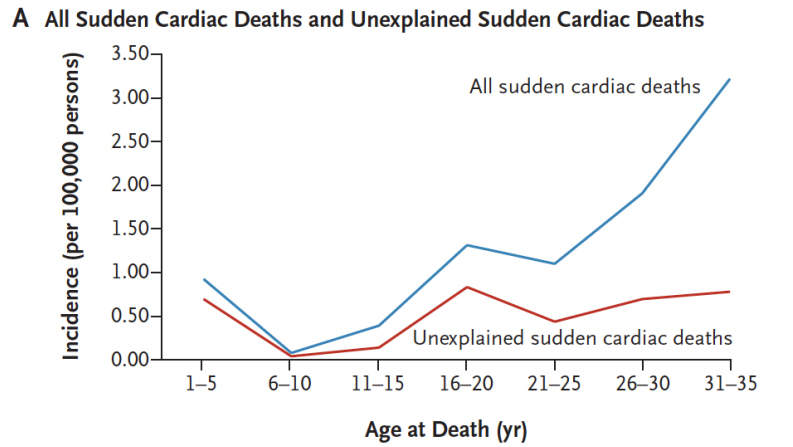
Etiology and treatment

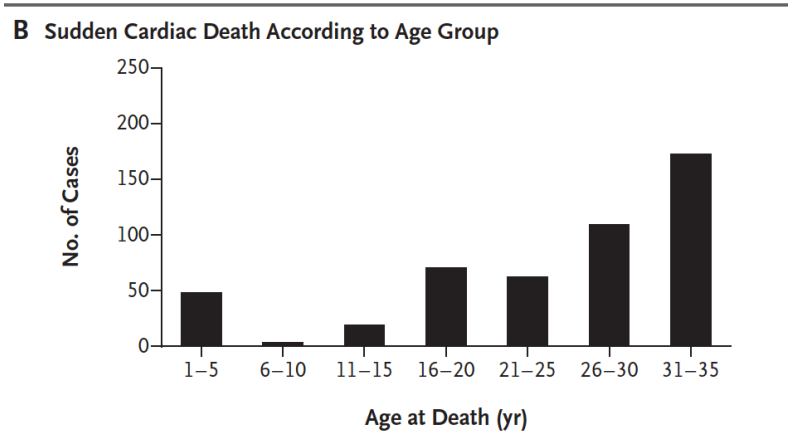
say little about much
or
say much about little

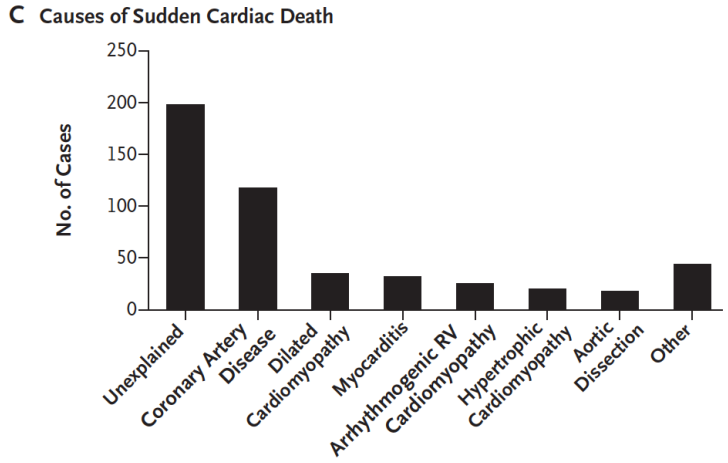
Focus on:

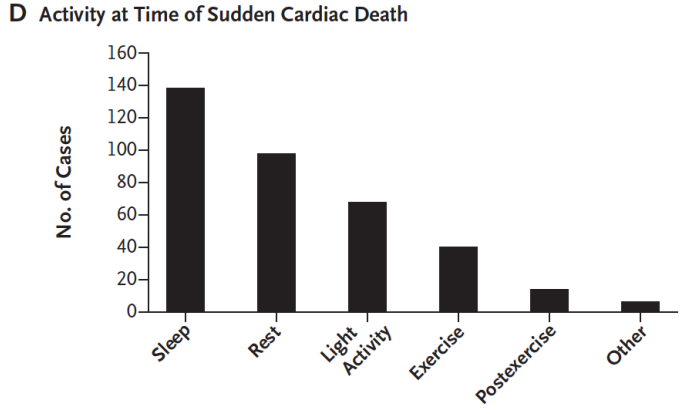
- (Tachy)arrhythmias and sudden cardiac death in *typical* congenital heart disease (CHD)
- No discussion on vaso-vagal syncope, channelopathies, abnormal coronary arteries, hypertrophic cardiomyopathy…
- Relationship between exercise in CHD and arrhythmia/sudden cardiac death

Sudden death: common problem?









Bagnall et al. New Engl J Med 2016; study in New Zealand and Australia; n = 490 (2010 -2012)

Sudden death: from athlete to…

Table 3. Sports-related sudden cardiac death by different age groups in Denmark from 2007 to 2009.

Age group (years)	12-49	12-35	36-49
SrSCD† cases (n)	44	9	35
Of whom were competitive athletes (n)	11	3	8
Incidence rates per 100,000 person-years (denominator)	(95% CI)	(95% CI)	(95% CI)
SCD* (general population)	10.7 (10.0-11.5)	3.15 (2.67-3.69)	21.7 (20.1-23.4)
SrSCD† (general population)	0.54 (0.39-0.72)	0.19 (0.08-0.35)	1.05 (0.73-1.45)
Non-competitive athletes (non-competitive athletes)	1.43 (0.99-2.01)	0.43 (0.16-0.94)	2.95 (1.95-4.30)
Competitive athletes (competitive athletes)	1.51 (0.75-2.71)	0.47 (0.10-1.14)	6.64 (2.86-13.1)
Sensitivity analyses: Incidence rates per 100,000 person-years (denominator)			
Autopsied cases only:			
Non-competitive athletes (non-competitive athletes)	1.08 (0.6-2.5)	0.43 (0.16-0.94)	2.08 (2.3-12.0)
Competitive athletes (competitive athletes)	1.37 (0.7-1.6)	0.47 (0.10-1.14)	5.81 (1.3-3.3)

Risgaard Dan Med J 2016

Sudden death: … to CHD

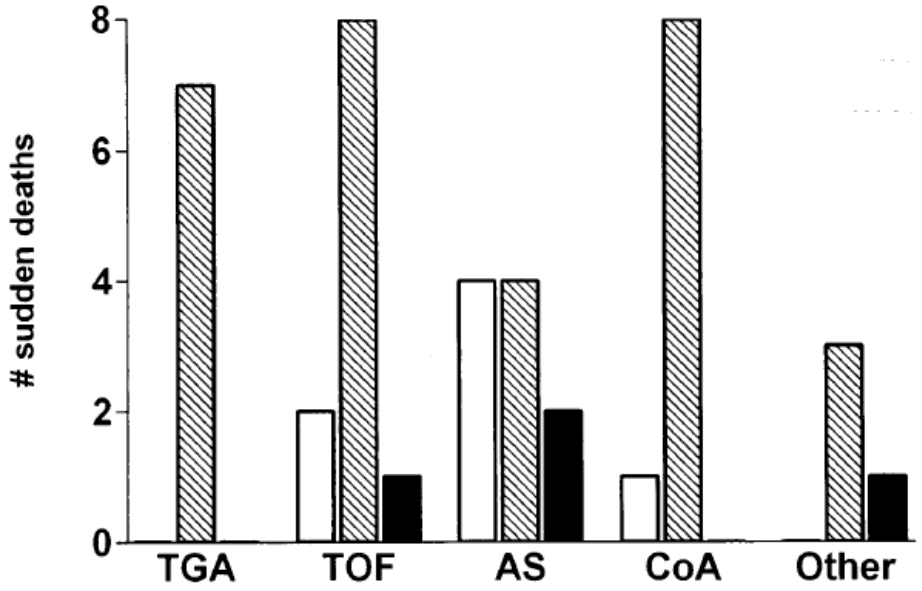
Table 1. Specific Congenital Heart Defects and Incidence of Sudden and Nonsudden Cardiac Death

	No. (%) of Pts With Complete Follow-Up	Total Follow-Up (pt-yr)	Sudden Cardiac Death		Nonsudden Cardiac Death	
			No.	Incidence/ 1,000 Pt-yr	No.	Incidence/ 1,000 Pt-yr
ASD	622 (86%)	7,904	0	0	0	0
VSD	527 (87%)	6,354	1	0.2	8	1.2
AVSD	254 (87%)	2,217	2	0.9	15	6.7
PDA	623 (82%)	8,753	0	0	4	0.4
PS	241 (91%)	3,568	1	0.3	2	0.6
AS	169 (94%)	1,860	10	5.4	9	4.8
CoA	536 (92%)	6,706	9	1.3	17	2.5
TOF	445 (91%)	7,082	11	1.5	9	1.3
D-TGA	172 (95%)	1,413	7	4.9	10	6.9
Total	3,589	45,857	41	0.9	74	1.6

AS = aortic stenosis; ASD = atrial septal defect; AVSD = atrioventricular septal defect; CoA = coarctation of the aorta; PDA = patent ductus arteriosus; PS = pulmonary stenosis; pt-yr = patient-years; Pts = patients; D-TGA = dextro-transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

Silka et al. JACC 1998

Sudden death by arrhythmias?

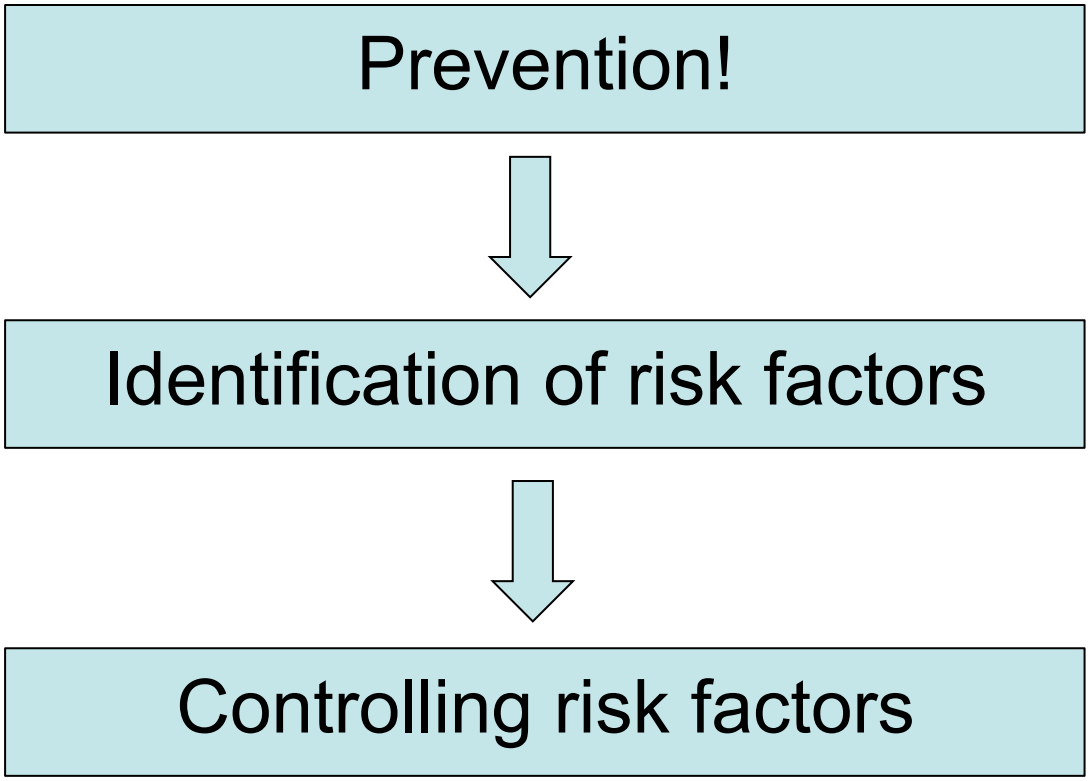


< 5% AV-block

Figure 1. Documented or probable etiologies of death among the 41 patients who died suddenly (arrhythmia **[hatched bars]**, circulatory **[open bars]**, congestive heart failure **[solid bars]**). The anatomic diagnoses of the other patients were pulmonary stenosis in one patient, ventricular septal defect in one and AV septal defect in two.

Silka et al. JACC 1998

Best treatment of sudden death?



What about the underlying defect?

N = 213/25790 (0.83%)

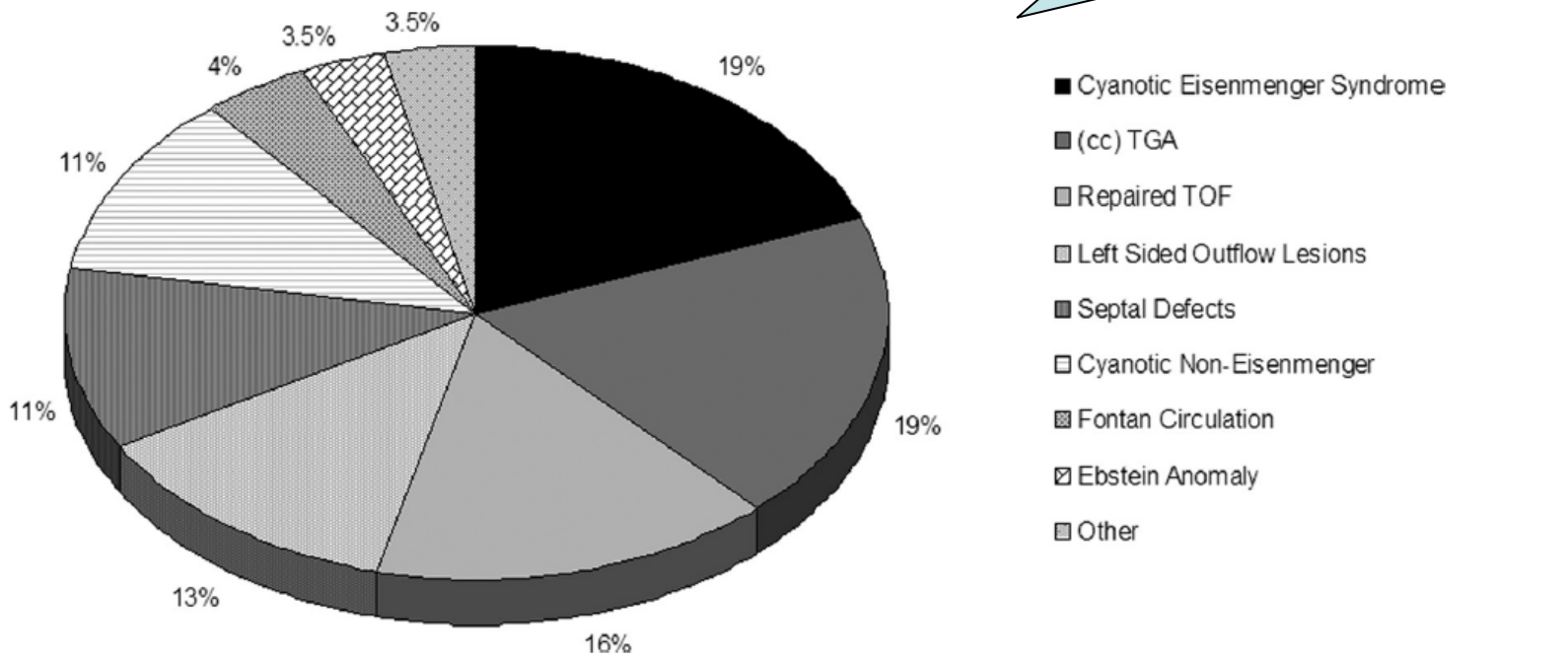


Figure 1. Cardiac defects in 171 sudden cardiac death cases. (cc) TGA indicates (congenitally corrected) transposition of great arteries; TOF, tetralogy of Fallot.

Koyak et al. Circulation 2012

Role of gender and age after childhood?

Table 1. Characteristics of Sudden Cardiac Death Cases and Controls

Variable	Cases (n=165)	Controls (n=310)	*P*
Medical history, n (%)			
Male	106 (64)	195 (63)	0.773
Mean age, y	36±15	37±14	0.480

No predictive value…

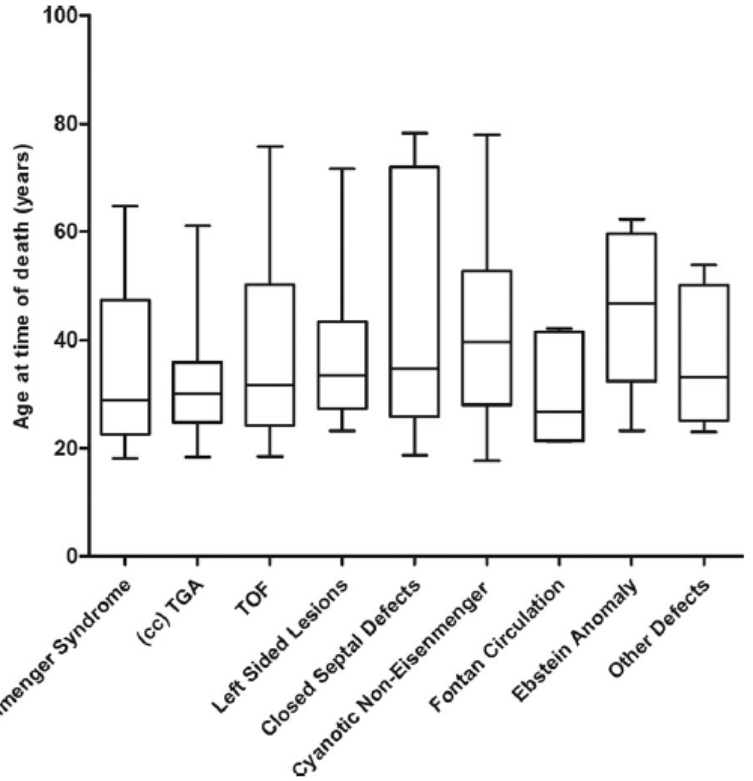


Figure 5. Age at time of death among several cardiac defects. (cc) TGA indicates (congenitally corrected) transposition of great arteries; TOF, tetralogy of Fallot.

Koyak et al. Circulation 2012

Medical history and symptoms?

Table 3. Clinical Variables Associated With SCD in Univariate and Multivariate Analysis in 165 SCD Cases and 310 Matched Controls

	Univariate Analysis			Multivariate Analysis		
Variable	OR	95% CI	*P*	OR	95% CI	*P*
Baseline characteristics						
Documented coronary artery disease	6.4	1.34–30.83	0.020			
Heart failure symptoms	4.2	2.54–6.90	<0.001			
Arrhythmic symptoms	1.6	0.95–2.86	0.075			
Supraventricular tachycardia	2.5	1.50–4.02	<0.001	3.5	1.50–7.95	0.004
Nonsustained ventricular tachycardia	2.0	1.09–3.66	0.025			
Heart failure drugs	5.6	3.24–9.66	<0.001			
Antiarrhythmic drugs	2.5	1.44–4.44	0.001			

Koyak et al. Circulation 2012

Relate to underlying disease!

TABLE 4. Predictors of Inducible Sustained VT

Variable	OR	95% CI	*P*
Univariate analysis			
Age at EP study	1.07	1.04–1.11	<0.0001
Age ≥18 y	6.0	3.0–12.3	<0.0001
Age at corrective surgery	1.07	1.02–1.12	0.0060
Age ≥7 y	3.3	1.9–5.6	<0.0001
Palpitations	2.8	1.6–5.0	0.0004
Syncope	4.9	2.6–9.1	<0.0001
Prior palliative surgery	2.9	1.7–5.1	<0.0001
Documented AF/flutter	2.1	1.0–4.4	0.0522
QRS, ms	1.03	1.02–1.04	<0.0001
QRS ≥180 ms	7.3	3.6–14.6	<0.0001
Modified Lown ≥2			0.0002
RV systolic			0.0574
At		1.3–5.0	0.0074
At	2.4	1.1–4.9	0.0201
Cardiothoracic ratio ≥0.60	3.3	1.8–6.1	0.0001
Multivariate analysis			
Age at EP study ≥18 y	3.3	1.1–10.5	0.0416
Palpitations	2.8	1.2–6.8	0.0234
Prior palliative surgery	3.1	1.2–7.6	0.0163
Modified Lown ≥2	5.6	1.0–30.9	0.0493
Cardiothoracic ratio ≥0.60	3.3	1.2–8.8	0.0200

AF indicates atrial flutter. Other abbreviations as in Table 1.

Tetralogy of Fallot

Table 2. Variables for Which Conditional Logistic Regression Analysis Was Performed, Noted in Mean OR With 95% CI

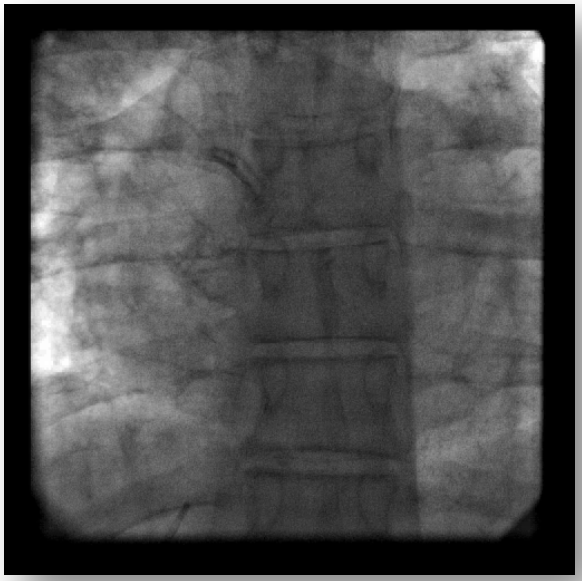
	p Value	OR (95% CI)
Symptoms		
Symptomatic*	<0.0005	6.45 (2.42–17.24)
Arrhythmic symptoms*	0.003	21.60 (2.80–166.79)
Heart failure symptoms*	0.001	4.44 (1.85–10.62)
ECG		
QRS duration	0.723	0.32 (0.001–175.66)
QT interval	0.668	0.16 (0.000–734.27)
QTc interval	0.193	1084.50 (0.029–4.1E + 07)
QRS duration >100 ms	0.251	980 (0.618–6.324)
QT dispersion		988 (0.973–1.003)
Heart rate		017 (1.000–1.035)
Basal heart rhythm nonsinus		1.112 (0.509–2.427)
Chest X-ray		
Enlarged heart size	0.053	2.227 (0.989–5.000)
24-h Holter		
Basal heart rhythm nonsinus	0.037	5.260 (1.10–25.00)
Documented episodes of arrhythmia	0.431	1.770 (0.44–7.25)
Mean heart rate	0.527	0.980 (0.919–1.044)
Minimum heart rate	0.952	1.001 (0.956–1.050)
Maximum heart rate	0.803	0.996 (0.965–1.028)
History of arrhythmia in follow-up		
Documented arrhythmia*	0.005	3.473 (1.451–8.310)
Documented SND in follow-up	0.035	2.405 (1.065–5.432)
Documented AFL/AF in follow-up*	0.001	4.866 (1.900–12.462)
Arrhythmia treatment		
Pacemaker implantation	0.550	0.641 (0.149–2.758)
Medication treatment*	0.002	5.159 (1.863–14.283)

*Statistically significant risk factor ($p < 0.005$).
AFL/AF = atrial flutter/atrial fibrillation; CI = confidence interval; OR = odds ratio; SND = sinus node disease.

Atrial switch operation

Khairy et al. Circulation 2004; Kammeraad et al. JACC 2004

Relate to underlying disease!



Angio IVC > “LA”

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QT dispersion		88 (0.973–1.003)
Heart rate		17 (1.000–1.035)
Basal heart rhythm nonsinus		1.112 (0.509–2.427)
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Information from the resting ECG?

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Variable	OR	95% CI	*P*	OR	95% CI	*P*
ECG						
Basal heart rhythm nonsinus	4.1	1.90–8.98	<0.001			
QRS duration (per 10-ms increase)	1.34	1.22–1.48	<0.001	1.22	1.10–1.34	0.008
QRS duration ≥140 ms	4.1	1.44–11.53	0.008			
QT interval	0.99	0.99–1.00	0.345			
QTc interval	1.00	0.99–1.01	0.108			
QT dispersion (per 10-ms increase)	1.34	1.22–1.63	<0.001	1.22	1.10–1.48	0.008

QRS duration:
- Tetralogy of Fallot: > 180 ms (significant relationship)
- Transposition of the great arteries: no clear relationship
- Eisenmenger syndrome: OR = 1.10/ms, 95%, P = 0.044

Koyak et al. Circulation 2012; Gatzoulis et al. Lancet 2000; Kammeraad et al. JACC 2004; Diller et al. Eur Heart J 2006

Information from the resting ECG?

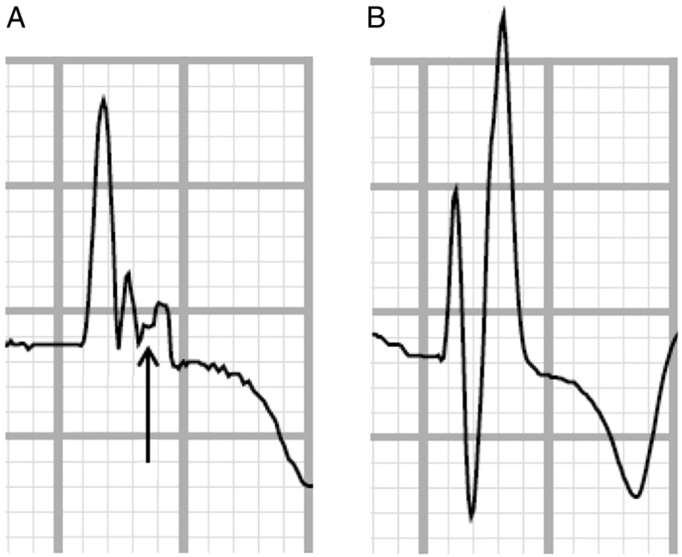


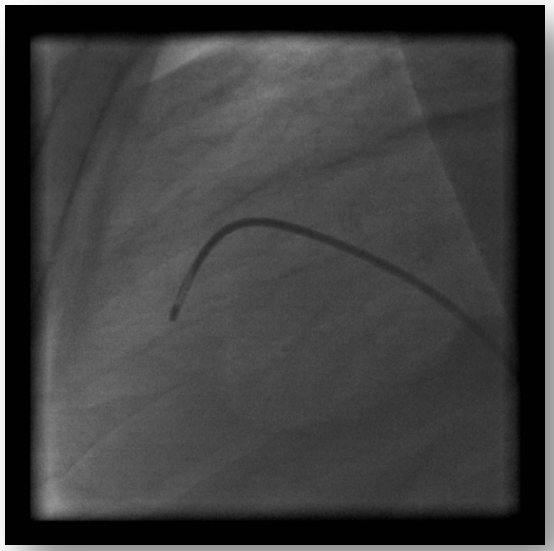
Figure 1 Right bundle branch block (RBBB) with and without QRS fragmentation. Lead V2 of an ECG in a patient with tetralogy of Fallot (TOF) (A) with RBBB and evident fragmentation (arrows) of the prolonged QRS complexes compared with a patient with TOF (B) with a wide RBBB without fragmentation of prolonged QRS complexes. fQRS, fragmented QRS complexes.

Tetralogy of Fallot:
occurrence of ventricular arrhythmia

N = 794	HR	95%CI
Extent of fQRS	2.00	1.26 – 3.16
Atrial fibrillation	4.78	1.45 – 14.8
QRS duration > 180 msec	2.63	1.12 – 6.18

Bokma et al. Heart 2016

Information from echocardiography?



Angio lateral RV TOF

Table 4. Factors Associated With SCD Among Various Cardiac Defects in Univariate Analysis

	Eisenmenger (33 SCD Cases vs 58 Controls) OR (95% CI)	(cc) TGA* (27 SCD Cases vs 52 Controls) OR (95% CI)	TOF (26 SCD Cases vs 52 Controls) OR (95% CI)	Left-Sided Outflow Lesions (21 SCD Cases vs 39 Controls) OR (95% CI)	Septal Defects (19 SCD Cases vs 39 Controls) OR (95% CI)	Cyanotic Non-Eisenmenger† (18 SCD Cases vs 32 Controls) OR (95% CI)
Symptoms						
Heart failure symptoms	2.7 (1.12–6.45)	4.0 (1.22–13.07)	. . .	. . .	. . .	6.6 (1.41–30.97)
Medication						
Heart failure drugs	3.0 (1.20–7.56)	. . .	3.9 (1.20–12.96)	. . .	. . .	18.2 (2.3–141.94)
Antiarrhythmics	3.4 (1.02–11.48)	. . .	. . .	. . .	. . .	. . .
Documented arrhythmias						
Atrial fibrillation/atrial flutter	. . .	3.2 (1.07–9.66)	7.2 (1.51–34.56)	. . .	. . .	. . .
Nonsustained ventricular tachycardia	. . .	. . .	. . .	. . .	. . .	. . .
ECG						
QRS duration (per 10-ms increase)	1.34 (1.03–1.76)	. . .	. . .	. . .	1.52 (1.04–2.22)	1.72 (1.01–2.92)
QT dispersion (per 10-ms increase)	. . .	. . .	1.99 (1.13–3.46)	. . .	. . .	. . .
Echocardiogram						
Impaired systemic ventricular function	8.2 (1.79–37.87)	3.8 (1.18–12.27)	8.5 (0.98–74.91)	. . .	. . .	7.4 (1.58–34.81)
Impaired pulmonary ventricular function	5.0 (1.35–18.73)	. . .	11.7 (1.42–95.92)	. . .	. . .	. . .
Chest x-ray						
Cardiothoracic ratio >0.5	. . .	. . .	8.7 (1.08–70.86)	. . .	. . .	. . .
Pulmonary edema	. . .	. . .	. . .	. . .	. . .	11.1 (1.33–92.54)
Laboratory findings						
Increased creatinine (per 10-μmol/L increase)	1.12 (1.12–2.46)	. . .	. . .	. . .	. . .	. . .

SCD indicates sudden cardiac death; (cc) TGA, (congenitally corrected) transposition of great arteries; TOF, tetralogy of Fallot; OR, odds ratio; and CI, confidence interval.

*Excluding patients with arterial switch and Rastelli procedure.

†Excluding patients with Fontan circulation.

Koyak et al. Circulation 2012

What is the value of 24 Holter?

Symptomatic

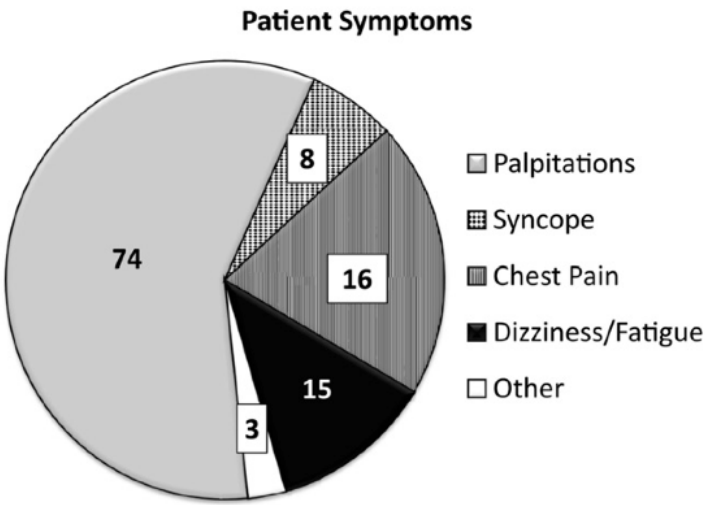


Figure 1. Clinical symptom type: breakdown of clinical patient symptom type as indication for Holter study. Numbers in diagram represent patient numbers.

Table 4
Holter study statistics stratified by age and congenital heart disease (CHD) type

Variable	Individual Holter Study*		Cumulative Data from ≥1 Holter Study†		
	PPV	NPV	Sensitivity	Specificity	NPV
All	0.08	0.97	0.4	0.7	0.96
Age (yrs)					
<18	0.04	0.99			
18–25	0.06	0.97			
18–35	0.17	0.92			
18–<25			0.3	0.6	0.93
>25			0.5	0.6	0.92
Congenital heart disease type					
Tetralogy of Fallot	0.05	0.99	0	0.8	0.98
Senning/Mustard	0.16	0.96	0.5	0.6	0.92
Fontan physiology	0.08	0.95	0.5	0.6	0.94

Sensitivity defined as percentage of patients with Holter study that changed management who then experienced a future clinically significant arrhythmia.

NPV = negative positive value; PPV = positive predictive value.

* Analyzed separately to compare PPV and NPV of Holter monitoring in patients with and without clinical symptoms.

† For each patient analyzed to determine sensitivity, specificity, and NPV of Holter monitoring in detection of clinically significant arrhythmias in individual patients.

Czosek et al. Am J Cardiol 2013

What is the value of 24 Holter?

Asymptomatic

Table 3 Patient characteristics

Characteristic				Overall ($n = 140$)
Mean age: years (range)				26.5 (18–66)
Cardiac diagnosis	Percentage of total ($n = 140$) (%)	Arrhythmias on ECG (%)	Arrhythmias on Holter monitoring (%)	Symptomatic (%)
Tetralogy of Fallot	21	13	43	10
d-TGA	6	22	33	0
Atrial septal defects	7	10	30	30
Single ventricle	5	57	29	29
Ventricular septal defects	4	20	40	0
Other (ASD, pulmonary atresia, mitral valve prolapse/mitral regurgitation, aortic stenosis/subaortic stenosis, Ebstein's anomaly, mitral stenosis, coarctation of aorta, and ccTGA)	57	11	27	10
Total	100	15	31	11

d-TGA dextro-transposition of the great arteries; *ASD* atrioventricular septal defect; *ECG* electrocardiogram; *ccTGA* congenitally corrected transposition of the great arteries

Rodriguez et al. Pediatr Cardiol 2012

What is the value of 24 Holter?

Asymptomatic

Table 3 Patient characteristics

Non sustained ventricular tachycardia in TOF patients found on routine screening: 7 fold risk for sudden cardiac death

	of total ($n = 140$) (%)	on ECG (%)	Holter monitoring (%)	(%)
Tetralogy of Fallot	21	13	43	10
d-TGA	6	22	33	0
Atrial septal defects	7	10	30	30
Single ventricle	5	57	29	29
Ventricular septal defects	4	20	40	0
Other (ASD, pulmonary atresia, mitral valve prolapse/mitral regurgitation, aortic stenosis/subaortic stenosis, Ebstein's anomaly, mitral stenosis, coarctation of aorta, and ccTGA)	57	11	27	10
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Rodriguez et al. Pediatr Cardiol 2012; Czosek et al. Am J Cardiol 2013

Exercise test equals 24 Holter

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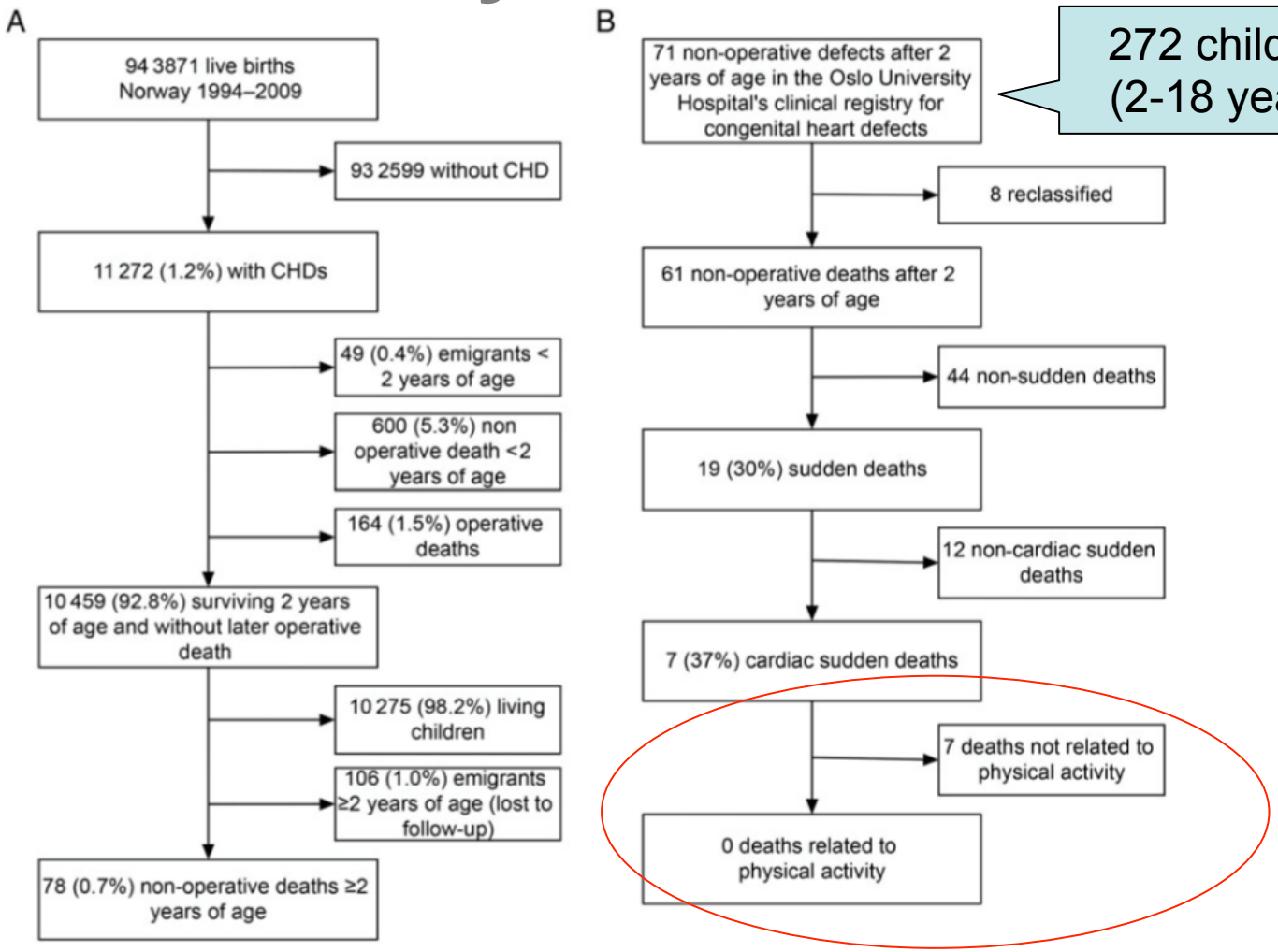
	Univariate Analysis			Multivariate Analysis		
Variable	OR	95% CI	*P*	OR	95% CI	*P*
Exercise test						
Induced arrhythmias	6.6	1.43–30.21	0.016			
Ventricular arrhythmias	4.8	1.00–23.24	0.049			

SCD indicates sudden cardiac death; OR, odds ratio; CI, confidence interval; and LVOT, left ventricular outflow tract.

Sudden death occurred only in 10% of cases during exercise!!

Koyak et al. Circulation 2012

Physical activity and sudden death?



Jortveit et al. Eur Heart J 2015

To EP or not to EP for VT?

Primary prevention

Repaired TOF

TABLE 5. Prognostic Significance of Inducible VT

			Actuarial Freedom From VT and SCD, %			
EP Testing	RR (95% CI)	*P*	1 y	5 y	10 y	15 y
Negative	1.00	...	97.9	92.8	89.3	89.3
SMVT	5.0 (2.1–11.9)	0.0002	79.4	67.1	63.6	63.6
SPVT	12.9 (3.9–43.2)	<0.0001	80.0	53.3	26.7	0.0
SMVT or SPVT	5.8 (2.5–13.2)	<0.0001	79.4	62.6	58.7	50.3

EP indicates electrophysiological; SMVT, sustained monomorphic VT; and SPVT, sustained polymorphic VT.

Khairy et al. Circulation 2004

To EP or not to EP for VT?

Primary prevention

EP study was not predictive for appropriate shocks in primary prevention for dTGA

Table 2. Predictors of Appropriate ICD Shocks

Variable	Hazard Ratio	95% CI	*P* Value
Univariate analysis			
Secondary prevention indication	5.1	1.1,45.5	0.0375
Ventricular septal defect	4.3	0.9,20.8	0.0742
At least moderate tricuspid regurgitation	4.1	0.8,20.5	0.0912
QTc, ms	1.02	1.00,1.05	0.0767
Lack of β-blockers	11.3	1.3,100.1	0.0303
Multivariate analysis			
Secondary prevention indication	18.0	1.2,261.0	0.0341
Lack of β-blockers	16.7	1.3,185.2	0.0301

Khairy et al. Circ Arrhythm Electrophysiol 2008

How to treat VT? Medical therapy?

- No data…

How to treat? VT ablation?

- No convincing data…
- Maybe for the individual patient (TOF)

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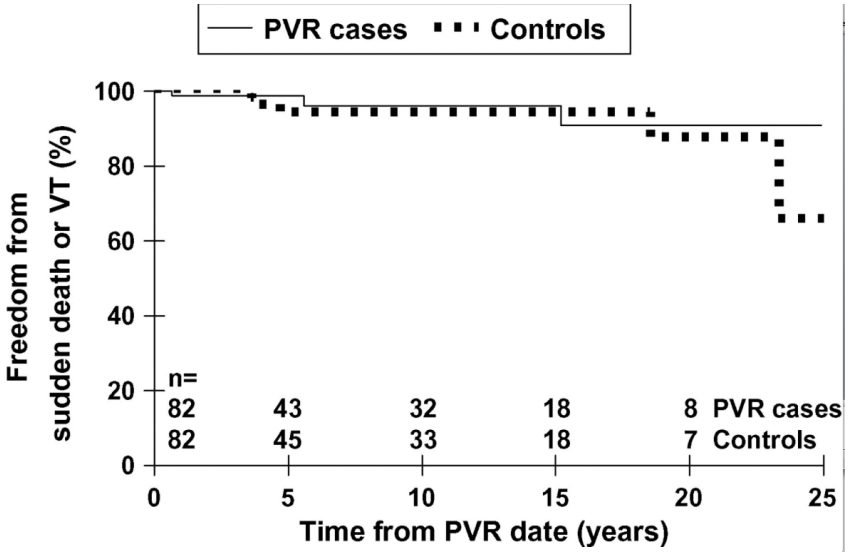
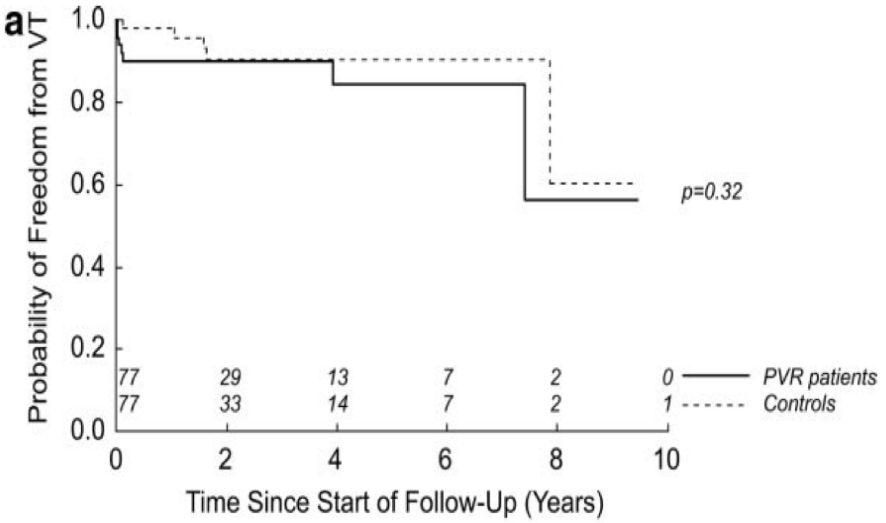
How to treat VT? Structural repair?



Questionable for tetralogy of Fallot



Gengsakul et al. Eur J Cardiothorac Surg 2007; Harrild et al. Circulation 2009

ICD the only solution?

Class I	1.	ICD therapy is indicated in adults with CHD who are survivors of *cardiac arrest* due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia after evaluation to define the cause of the event and exclude any completely reversible etiology *(Level of evidence: B)*. [40, 46, 460, 461 and 462]
	2.	ICD therapy is indicated in adults with CHD and *spontaneous sustained ventricular tachycardia* who have undergone hemodynamic and electrophysiologic evaluation *(Level of evidence: B)*. [40, 46, 97, 426, 460 and 461] Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD therapy in carefully selected patients *(Level of evidence: C)*. [463, 464 and 465]
	3.	ICD therapy is indicated in adults with CHD and a *systemic left ventricular ejection* fraction ≤35%, biventricular physiology, and New York Heart Association (NYHA) class II or III symptoms *(Level of evidence: B)*. [97, 111, 428, 433, 434 and 435]

Khairy et al. Heart Rhythm 2014

ICD implantation

- Risk stratification

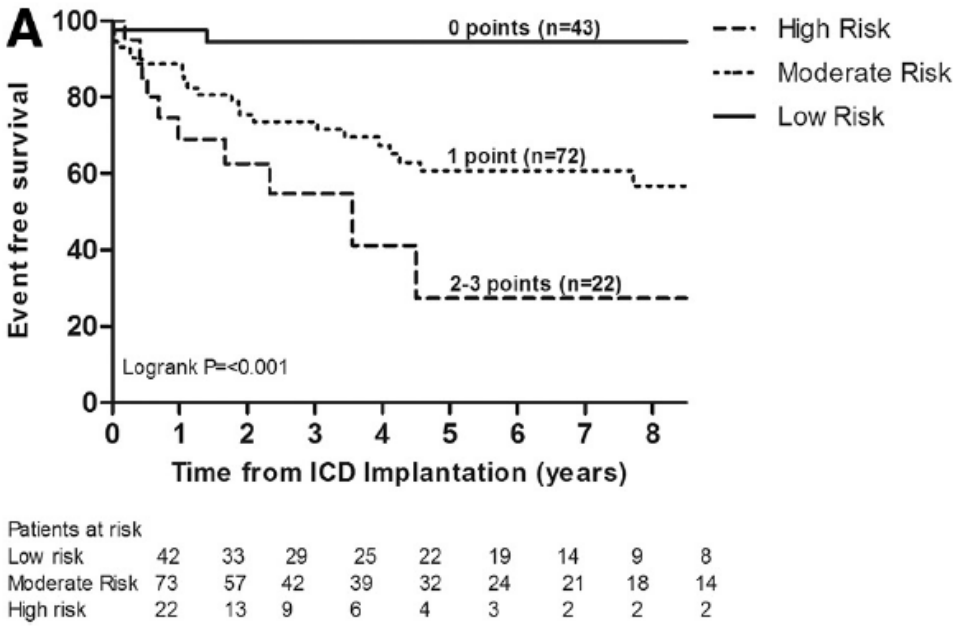
Table 3. Risk Score for Appropriate ICD Shocks in the Total Cohort

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

CAD indicates coronary artery disease.

Subpulmonary ventricular function (primary prevention)

Systemic ventricular function (secondary prevention)



Koyak et al. Circ Arrhythm Electrophysiol 2012

Conclusions

- Arrhythmia and SCD are not uncommon in CHD
- Electrical and structural abnormalities - not specifically exercise - trigger late arrhythmia
- Screening to arrhythmia = screening to risk factors, also in the asymptomatic patient
- The best treatment = avoiding structural changes
- The effect of medical treatment or anatomical repair is unclear to prevent recurrence of arrhythmia
- ICD implantation seems the best solution