Congenital Heart Disease: Arrhythmia in Adult Congenital Heart Disease

S-ICD in ACHD

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Disclosure

The COI to declare is as follows

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⑨ Other remuneration such as gifts: none

Background

Sudden cardiac death (SCD) due to life-threatening ventricular tachyarrhythmias is a significant cause of mortality and morbidity in adult patients with congenital heart disease (ACHD).

However, in patients with complex congenital anatomy including complicated venous access, or with right-to-left shunting, the use of a transvenous ICD (TV-ICD) is either impossible or relatively contraindicated due to the increased risk of systemic thromboembolism or venous complications.

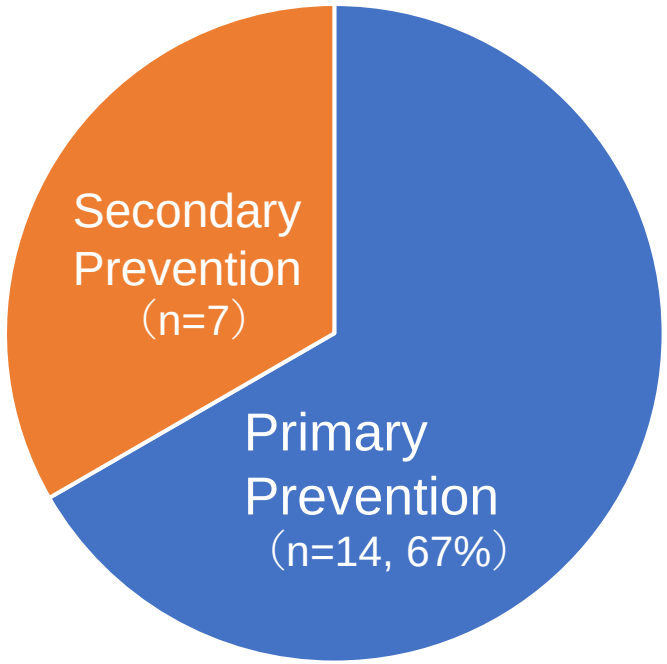
The subcutaneous ICD (S-ICD) is expected as a potential new treatment option for patients with ACHD at high risk for SCD.

Is the S-ICD really beneficial in preventing SCD in patients with ACHD?

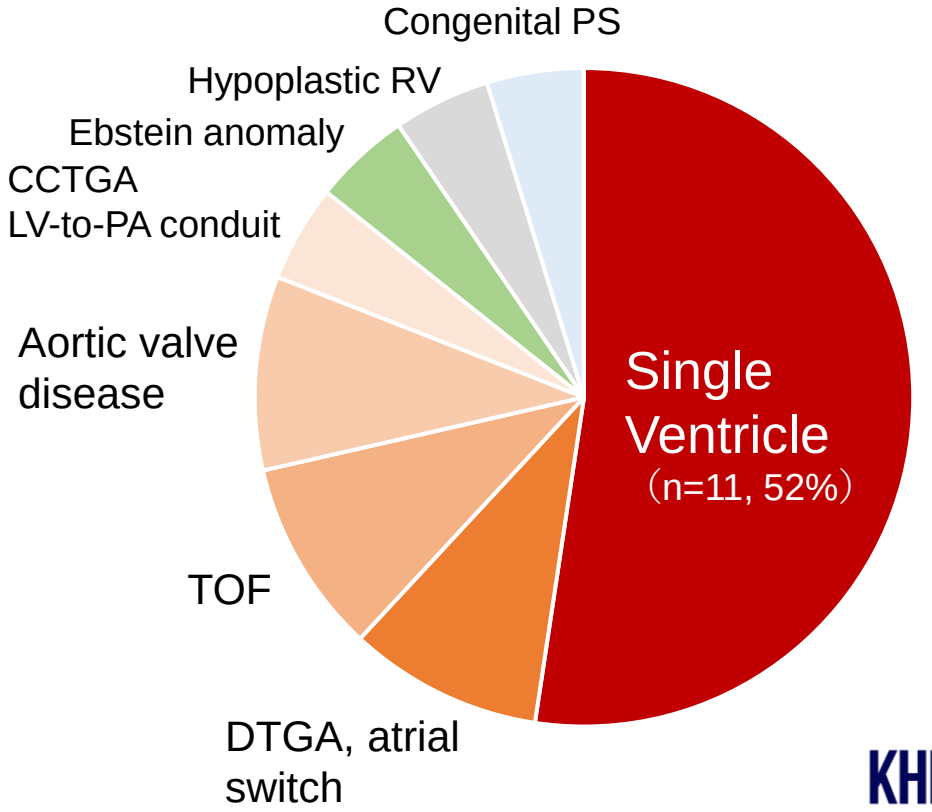
Mid-term experience with the S-ICD in the ACHD population

Twenty-one ACHD patients identified from AARCC (Alliance for Adult Research in Congenital Cardiology) retrospective data were analyzed.

Indication for ICD



Reason for choosing the S-ICD



Moore et al. Circ Arrhythm Electrophysiol. 2016;9:e004338.

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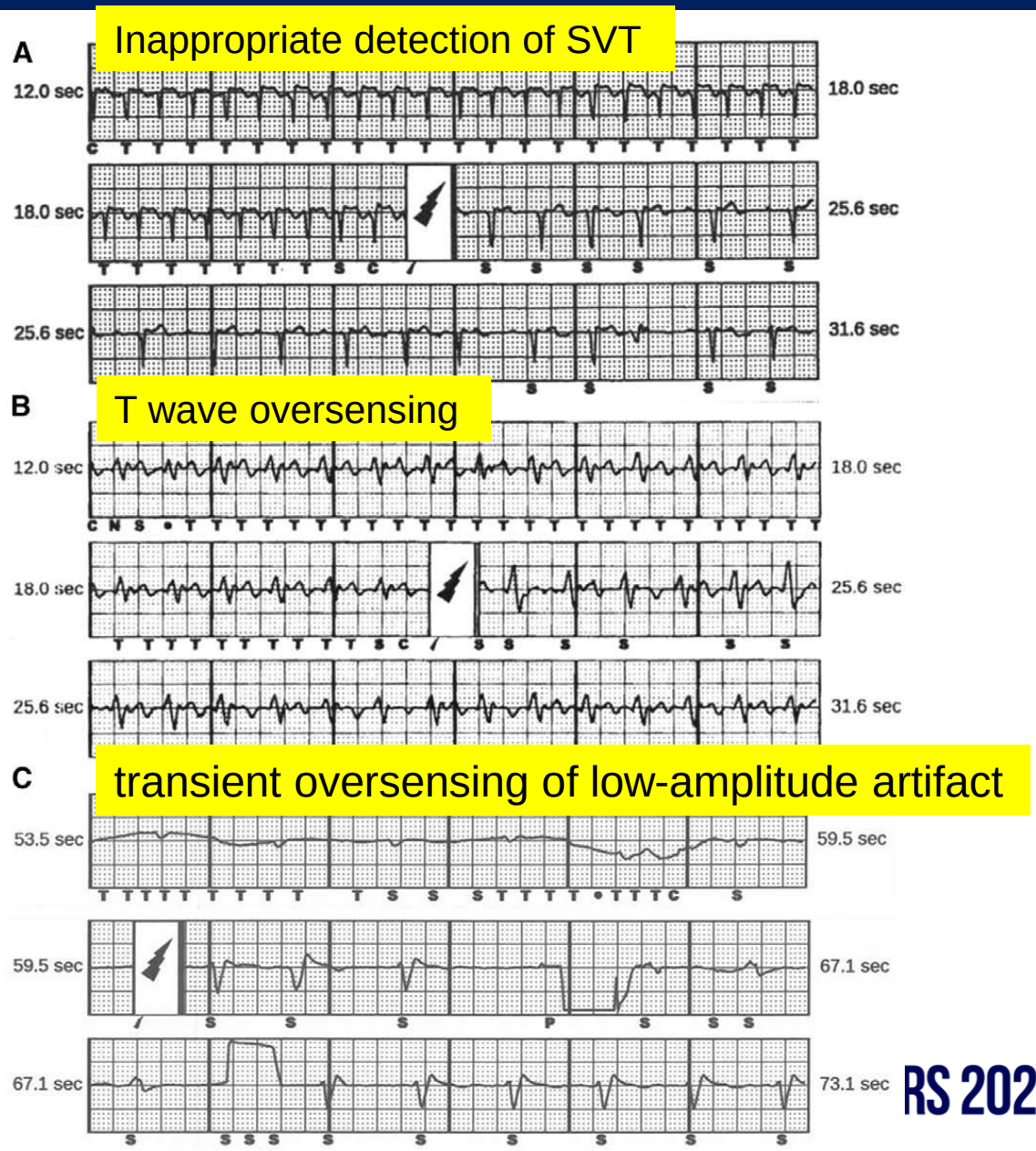
Mid-term outcome

Acute defibrillation test was performed in 18 patients during procedure and resulted in successfully termination at an output of ≤80J for all patients.

During median follow-up period of 14 months (IQR 3-19months), 4 patients (20%) experienced IAS, and 1 patient received appropriate shocks (5%).



Moore et al. Circ Arrhythm Electrophysiol. 2016;9:e004338.

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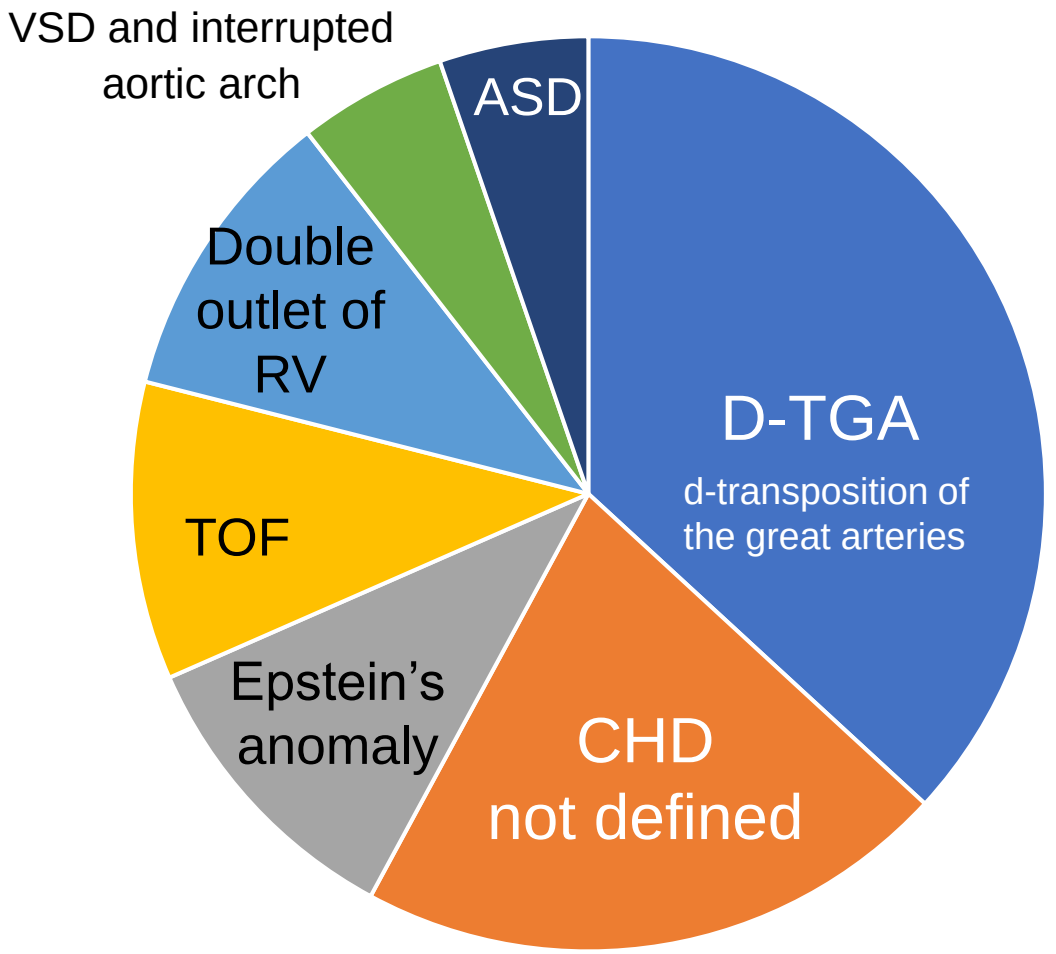
What about the mid-term efficacy of the S-ICD compared to the TV-ICD?

A pooled analysis enrolled 865 patients who registered in the EFFORTLESS registry and the IDE study.

Nineteen CHD patients versus 846 non-CHD patients with a median follow-up of 567 days (18months) and 639 days, respectively, were analyzed.

Details of ACHD (n=19)



D'Souza et al. J Am Coll Cardiol EP 2016;2:615–22.

Comparison of baseline characteristics between 2 groups

TABLE 1 Demographic Data and Medical History for Patients With and Without Congenital Heart Disease

Demographic	Statistic/Category	Congenital Heart Disease Patients	Non-Congenital Heart Disease Patients	p Value
Age (yrs)	Mean ± SD (median)	30.0 ± 13.8 (26.8)	50.7 ± 16.7 (53.0)	<0.0001
	Range	12.0-65.0	7.0-88.0	
Sex, n (%)	Male	10 (52.6)	622 (72.9)	0.0502
	Female	9 (47.4)	231 (27.1)	
BMI (kg/m^2)	Mean ± SD (median)	24.9 ± 6.2 (24.1)	28.3 ± 6.6 (27.1)	0.0284
	Range	16.0-39.0	15.2-69.0	
Indication	Primary prevention	13 (68.4)	602 (70.7)	0.0002
	Secondary prevention	6 (31.6)	250 (29.3)	
Ejection fraction (%)	Mean ± SD (median)	43.9 ± 20.3 (45.0)	39.3 ± 17.6 (34.0)	0.4412
	Range	12.0-70.0	10.0-86.0	
Medical history, n (%)	NYHA functional class II–IV	3 (15.8)	324 (38.0)	0.0477
	Atrial fibrillation	5 (26.3)	136 (16.0)	0.2256
	COPD	0 (0)	56 (6.6)	0.2477
	Diabetes	0 (0)	155 (18.2)	0.0402
	Hypertension	1 (5.3)	330 (38.8)	0.0029
	Myocardial infarction	1 (5.3)	301 (35.4)	0.0064
	Stroke	1 (5.3)	44 (5.2)	0.9856
	Valve disease	4 (21.1)	110 (12.9)	0.2992
	Ablation	5 (26.3)	35 (4.1)	<.0001
	CABG	0 (0)	101 (11.9)	0.1102
	Prior ICD	3 (15.8)	117 (13.7)	0.7969
	Explant due to infection	1 (5.3)	75 (8.8)	0.5887
	Explant due to ICD lead failure	0 (0)	30 (3.5)	0.4052
	Pacemaker	5 (26.3)	17 (2.0)	<.0001
	Percutaneous Revascularization	1 (5.3)	194 (22.8)	0.0702
	Value surgery	5 (26.3)	48 (5.6)	0.0002
NYHA functional class II–IV breakdown, n (%)	Class II	3 (100.0)	216 (66.7)	
	Class III-IV	0 (0)	108 (33.3)	

BMI = body mass index; CABG = coronary artery bypass grafting; COPD = chronic obstructive pulmonary disease; ICD = implantable cardioverter defibrillator; NYHA = New York Heart Association functional class.

Younger (30.0 ± 13.8 vs 50.7 ± 16.7)

More patients with a history of ablation therapy

More patients with pacemaker implants

D'Souza et al. J Am Coll Cardiol EP 2016;2:615–22.

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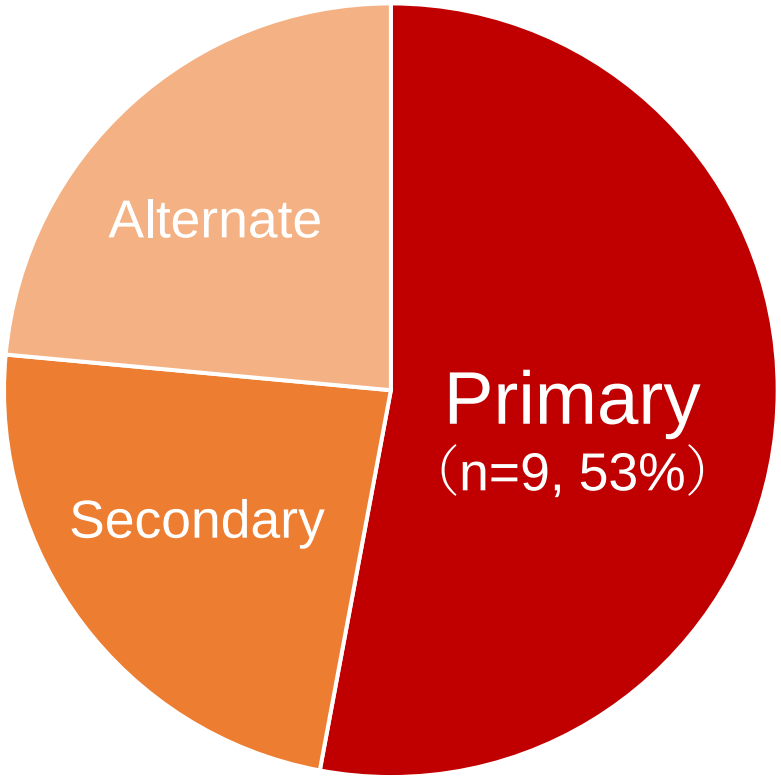
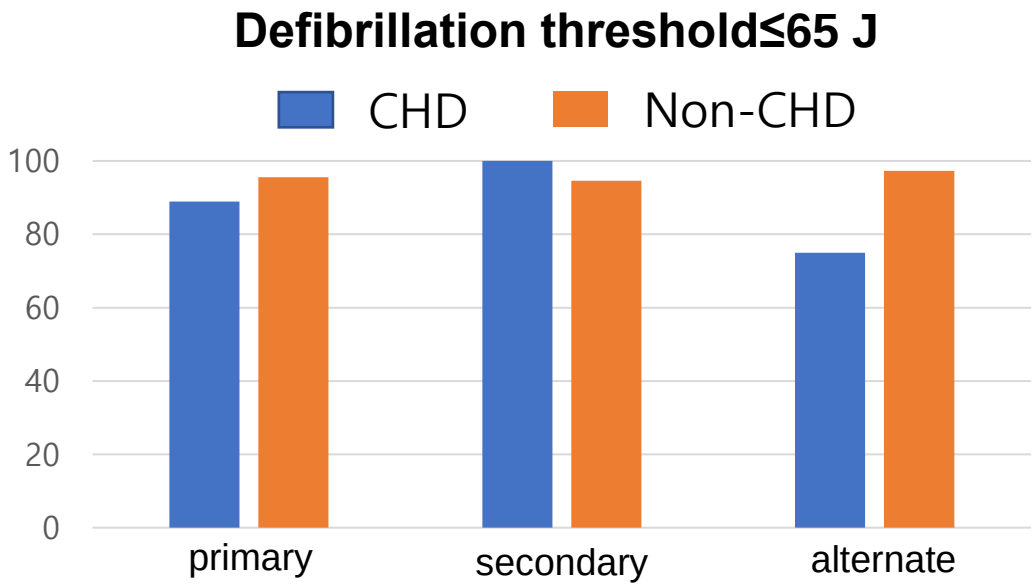
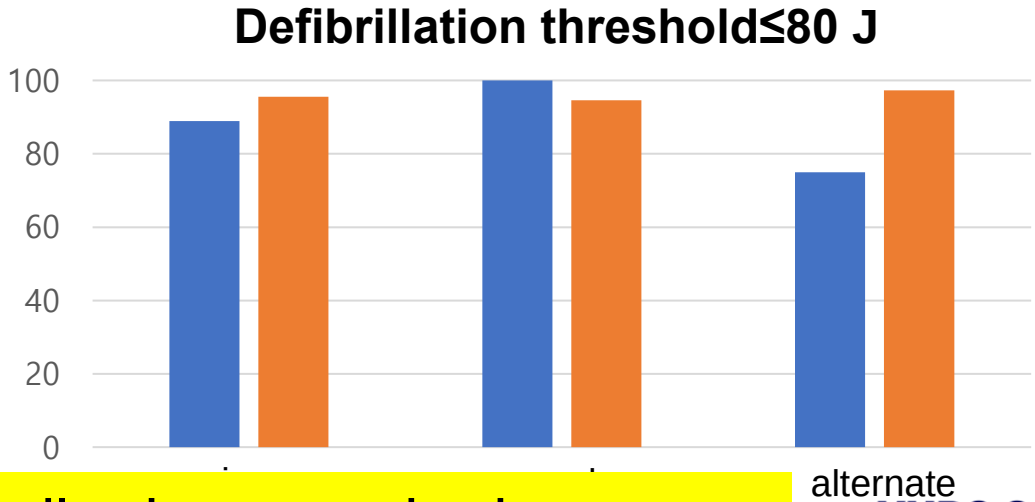
Comparison of efficacy in acute defibrillation test

Selected sensing vector





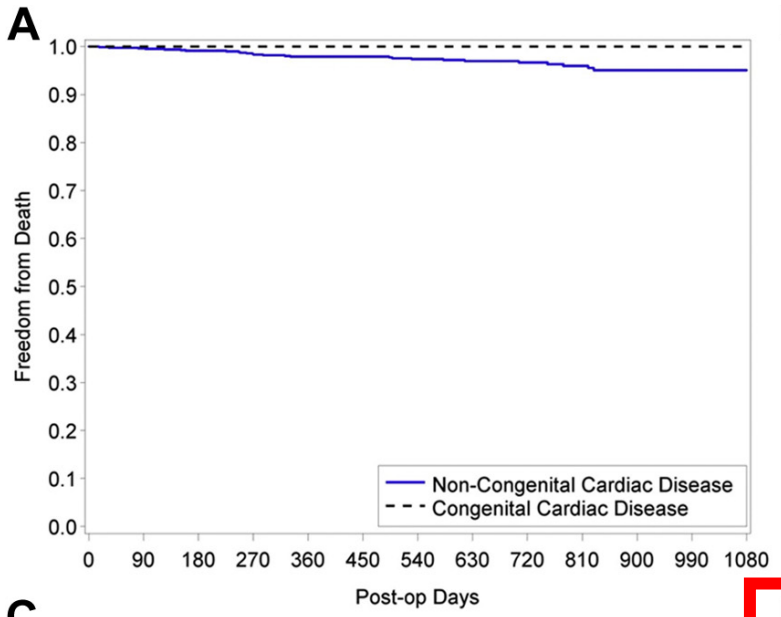


The defibrillation success rate was similar between both groups.

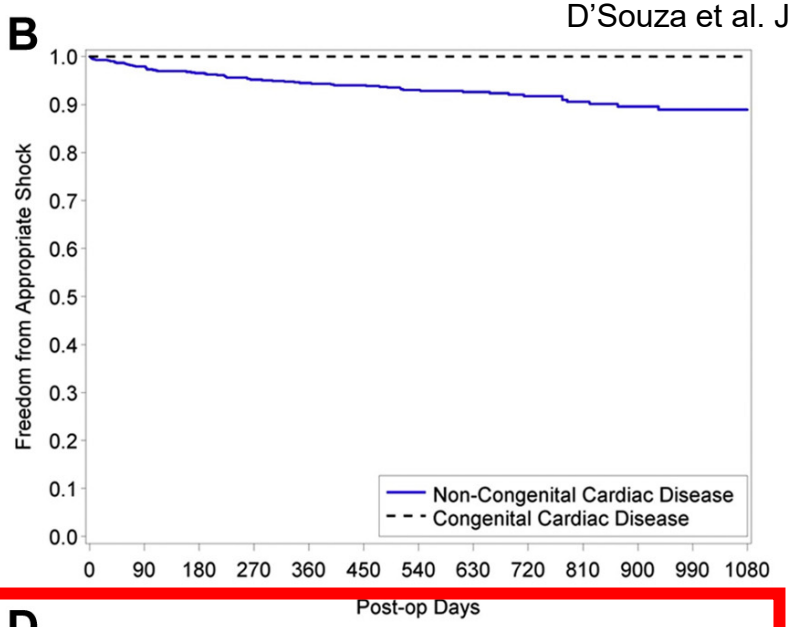
Kaplan-Meier estimates for congenital and non-congenital patients post S-ICD implantation

D'Souza et al. J Am Coll Cardiol EP 2016;2:615–22.

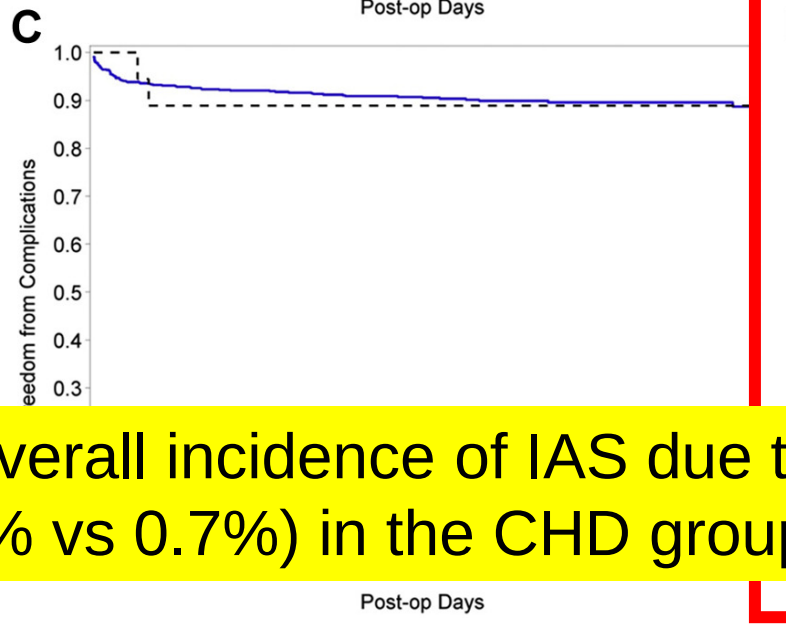
Mortality



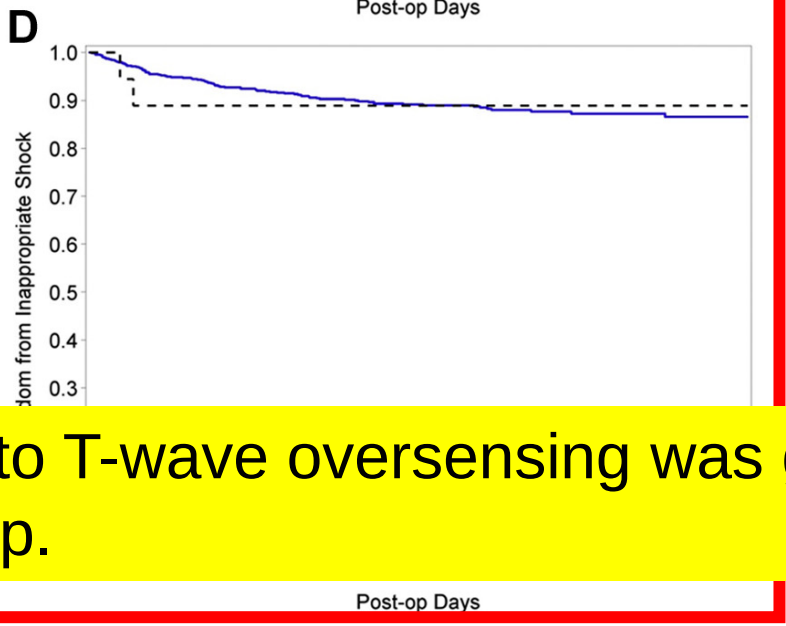
Appropriate shock



All complications



Inappropriate shock (IAS)



The overall incidence of IAS due to T-wave oversensing was greater (10.2% vs 0.7%) in the CHD group.

Long-term experience from a large single-centre analysis

Twenty S-ICD patients with ACHD were investigated during a mean follow-up of 3 years.

Results

Initial sensing vector at time of implantation (*n*)	
Primary	10 (50%)
Secondary	8 (42%)
Alternate	2 (8%)
Operation related S-ICD complications (*n*)	2 (10%)
Pocket haematoma managed conservatively	1 (5%)
Pocket haematoma requiring surgical revision	1 (5%)
Operation time (min)	38.2 ± 7.1
Local anaesthesia preferred due to critical preoperative state	2 (10%)
Successful defibrillation test (*n*)	19 (95%)
Defibrillation test foregone because of perioperative instability	1 (5%)
S-ICD™ explantation due to infectious problems	1 (5%)
Death during follow-up	3 (15%)
Death due to congestive heart failure in palliative patients after deactivation of the S-ICD™	2 (10%)
Death due to pulmonary embolism	1 (5%)

In the acute defibrillation test, VF was successfully terminated by S-ICD shock in all patients.

There were 9 appropriate shock deliveries in 3 patients (15%), all of them terminating VT with the 1st shock.

In 2 patients (10%), an inappropriate shock occurred due to T-wave oversensing.

Willy K, et al. Europace 2019; 21: 1537–1542.

Limitations of evidence proving usefulness of S-ICD for ACHD

There are no RCTs or large-scale studies demonstrating the usefulness of S-ICD for patients with ACHD.

The defibrillation success rate in the acute defibrillation test is close to 100%, but its efficacy against spontaneous VT/VF has not yet been established.

The effect of S-ICD on long-term prognosis is unknown.

What are the remaining challenges in the application of S-ICD to patients with ACHD?

1st issue: Risk of inappropriate sensing

ACHD patients has many structural and functional disturbances…

Cardiac chamber enlargement

Abnormal cardiac position

Mechanical strain

Augmented repolarization

→ Abnormal T wave morphology

↓

IAS due to TWOS

Inappropriate shock rate in ACHD patients

Comparison of the results of several studies

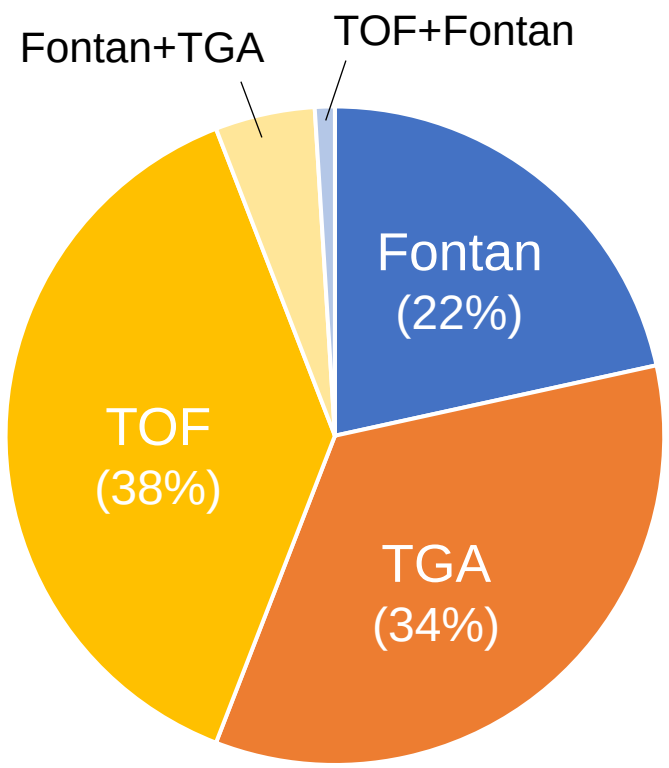
	Moore, et al.	D'Souza, et al.	Ferrero P, et al.	Willy K, et al.
Number of patients	21	19	8	20
Follow-up period	14 months	567 days	874 days	3 years
A				
M				
Systemic ventricle EF, %	41 (35–63)	43.920.3	n.a	46.511.3
IAS, %	20	10.2	12.5	10

Although the observation period is 3 years or less, the IAS rate is over 10%. It is necessary to estimate how much SMART pass technology contributes to prevention for IAS.

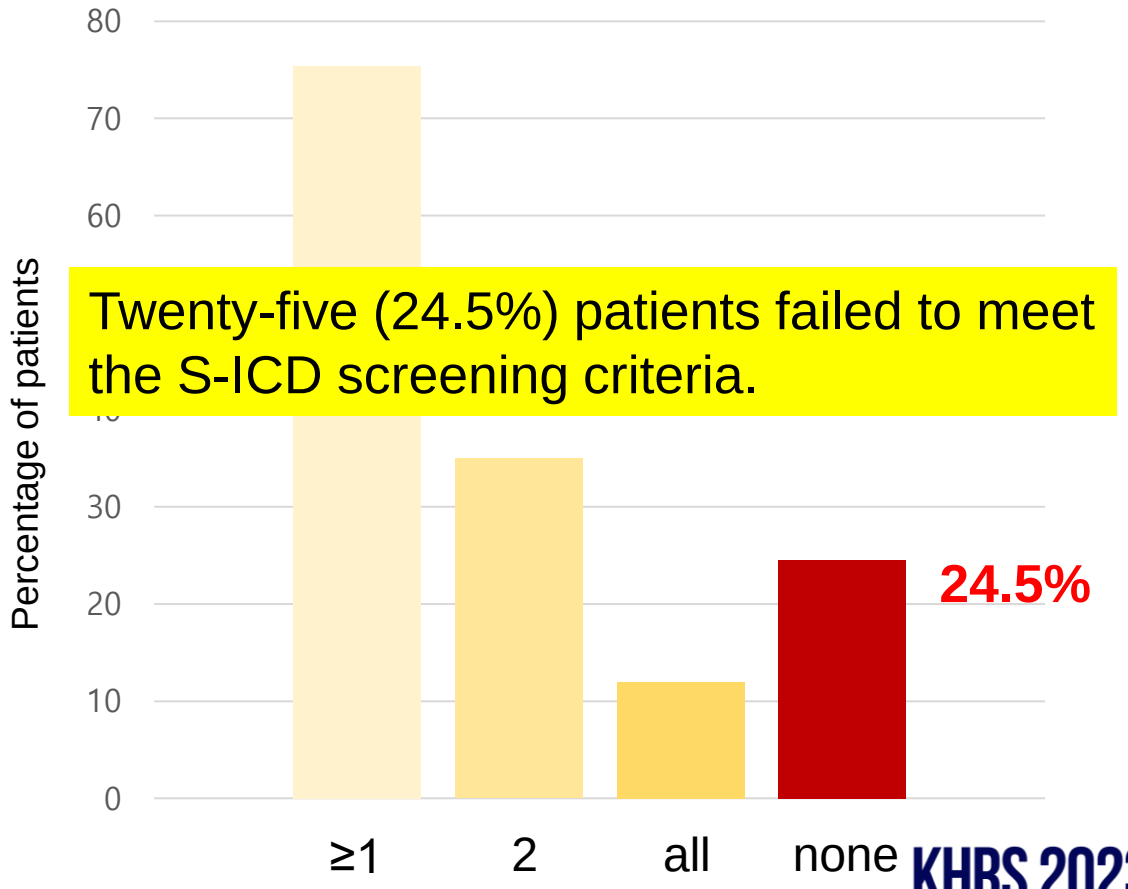
2nd issue: Ineligibility for S-ICD in ECG screening

A standard ECG screening was performed in 102 patients with complex ACHD.

Details of complex ACHD



Number and distribution of suitable vectors



Garside, et al. Pacing Clin Electrophysiol. 2019;42:65–70.

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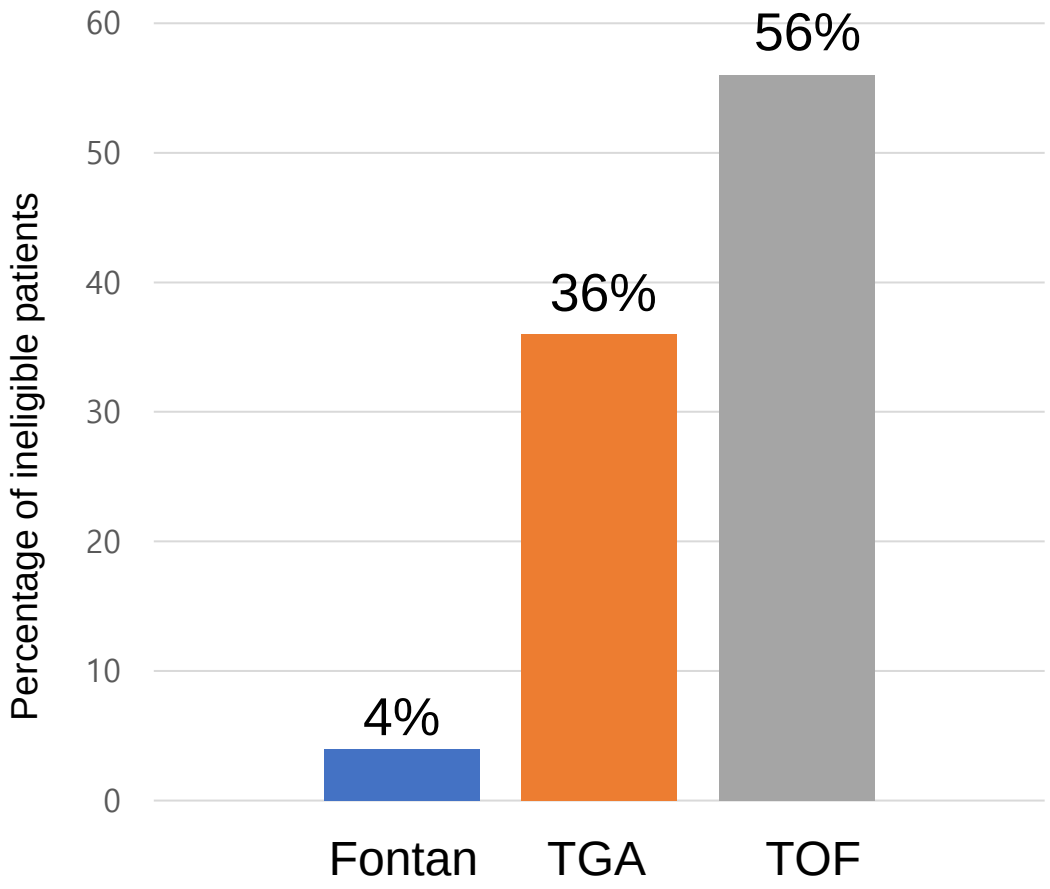
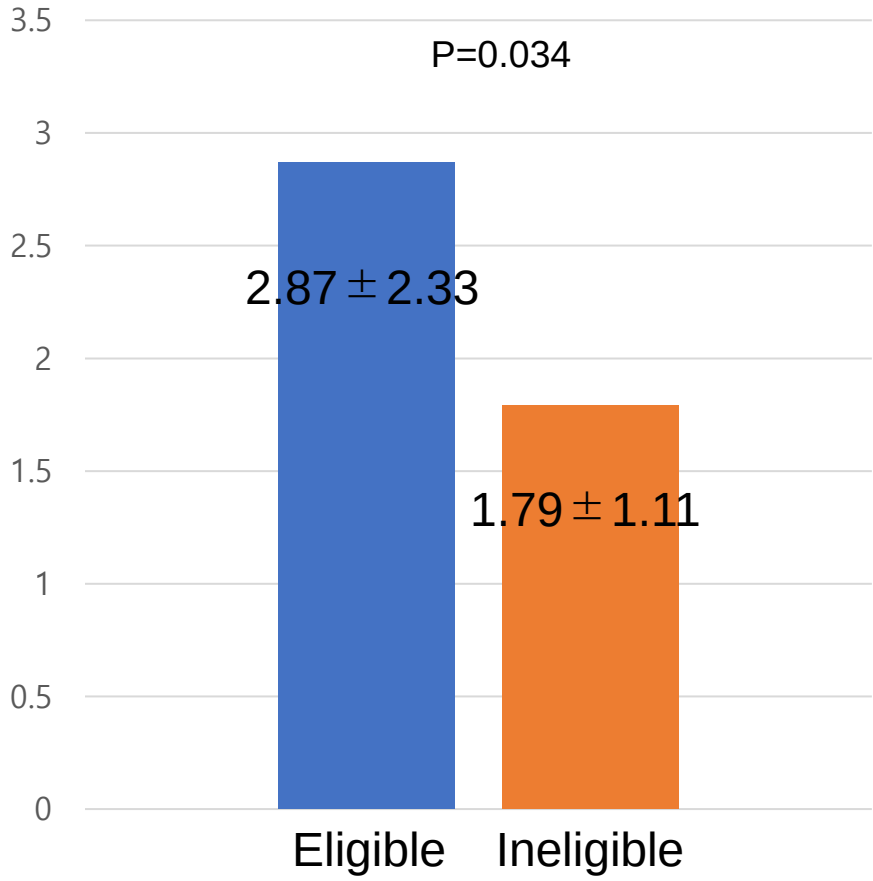
What is the reason for ineligibility for S-ICD in ACHD?

Disease-specific differences in eligibility for ECG screening



Comparison of R:Tmax



Garside, et al. Pacing Clin Electrophysiol. 2019;42:65–70.

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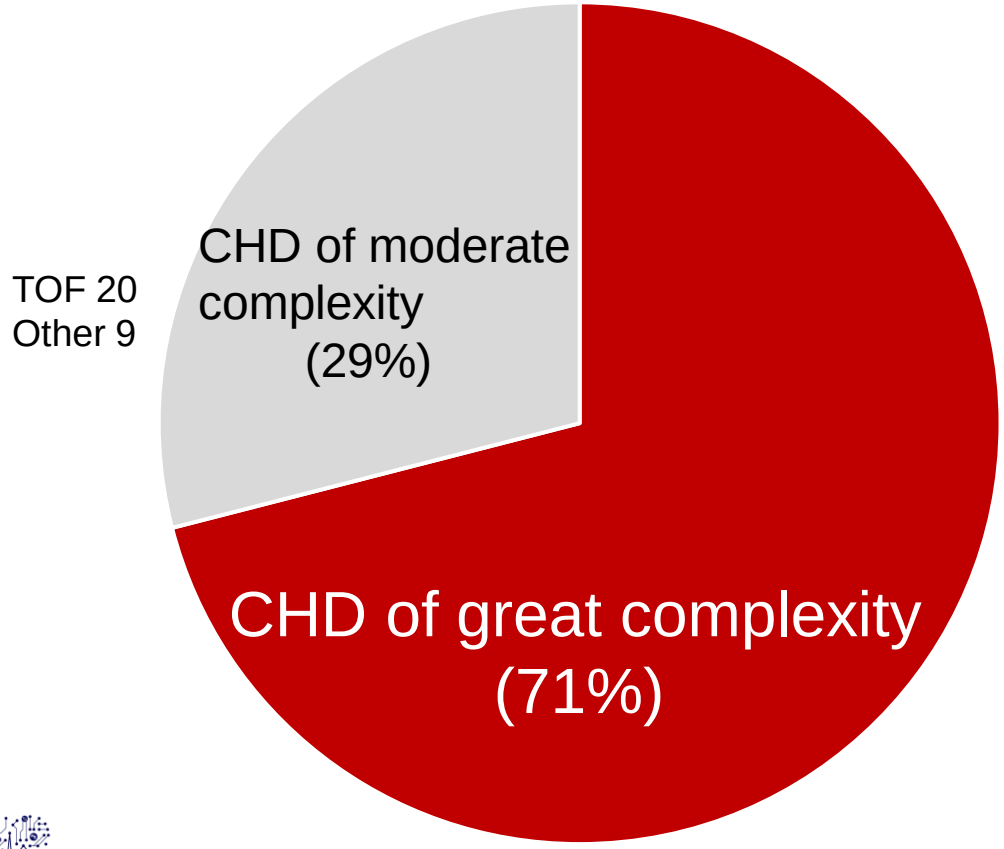
Does utilization of AST improve the eligibility for S-ICD?

One hundred patients with ACHD were screened for S-ICD eligibility with standard ECG-based screening test and automated screening test (AST).

Details of ACHD



Baseline characteristics

Age 38.1 ± 12.2 (years)

Male 66 (%)

BMI 25.8 ± 5.3 (kg/m2)

LVEF 48.0 ± 9.2 (%)

Sinus rhythm 74 (%)

C. Zormpas et al. ESC Heart Failure 2021; 8: 1502–1508.

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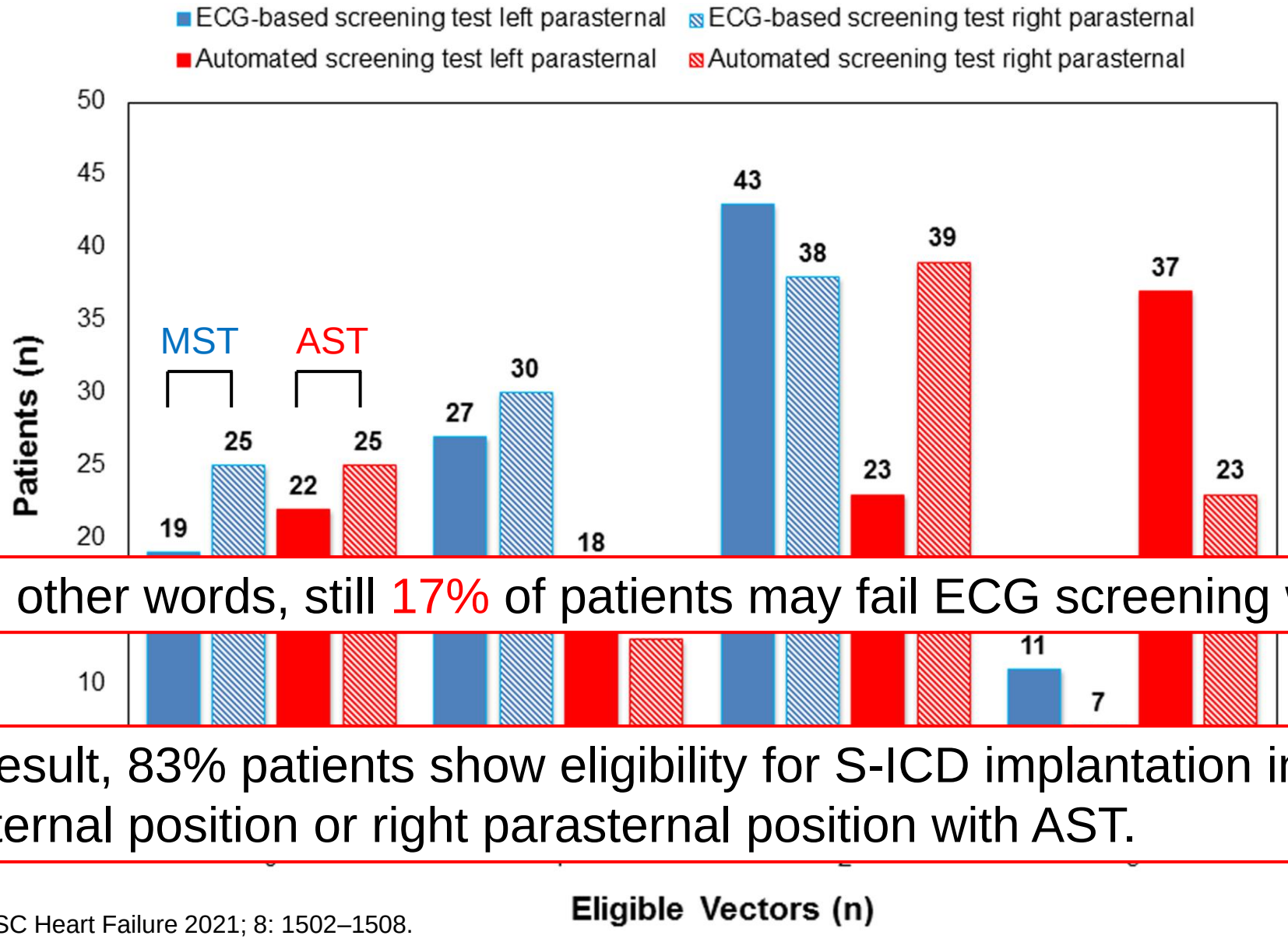
Number of eligible vectors in study population



In other words, still 17% of patients may fail ECG screening with AST.

As a result, 83% patients show eligibility for S-ICD implantation in either left parasternal position or right parasternal position with AST.

C. Zormpas et al. ESC Heart Failure 2021; 8: 1502–1508.

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What are the predictors of failure in AST?

Univariate analysis

Parameter	Eligible $n = 83$	Ineligible $n = 17$	*P*-value
Age (years)	36.5 ± 11.4	34.3 ± 7.5	0.756
Male, *n* (%)	55 (66.3)	11 (64.7)	0.903
Chest circumference (cm)	96.3 ± 12.3	94.5 ± 11.3	0.191
Body mass index (kg/m^2)	25.7 ± 5.7	25.0 ± 3.5	0.223
Sinus rhythm, *n* (%)	65 (78.3)	9 (53.0)	0.030
Cardiac axis (°)	19.9 ± 79.9	34.0 ± 83.5	0.957
QRS duration (ms)	122.8 ± 32.6	170.6 ± 30.1	<0.001
Paced QRS complex, *n* (%)	8 (9.6)	7 (41.2)	<0.001

Sinus rhythm

QRS duration

Paced QRS complex

Multivariate analysis

	OR	95% CI for OR	*P*-value
Sinus rhythm	0.981	0.117–8.228	0.981
QRS duration ≥148 ms	0.102	0.024–0.432	0.002
Paced QRS complex	0.480	0.049–4.732	0.530

A QRS duration ≥148ms was the only independent predictor for failure of the AST.

C. Zormpas et al. ESC Heart Failure 2021; 8: 1502–1508.

Are there any solutions to ineligibility for ECG screening?

100 ACHD patients

Mean age 48 ± 14 years, average QRS duration 134 ± 37 ms
CHD with great complexity 21%, simple or moderate complexity 79%

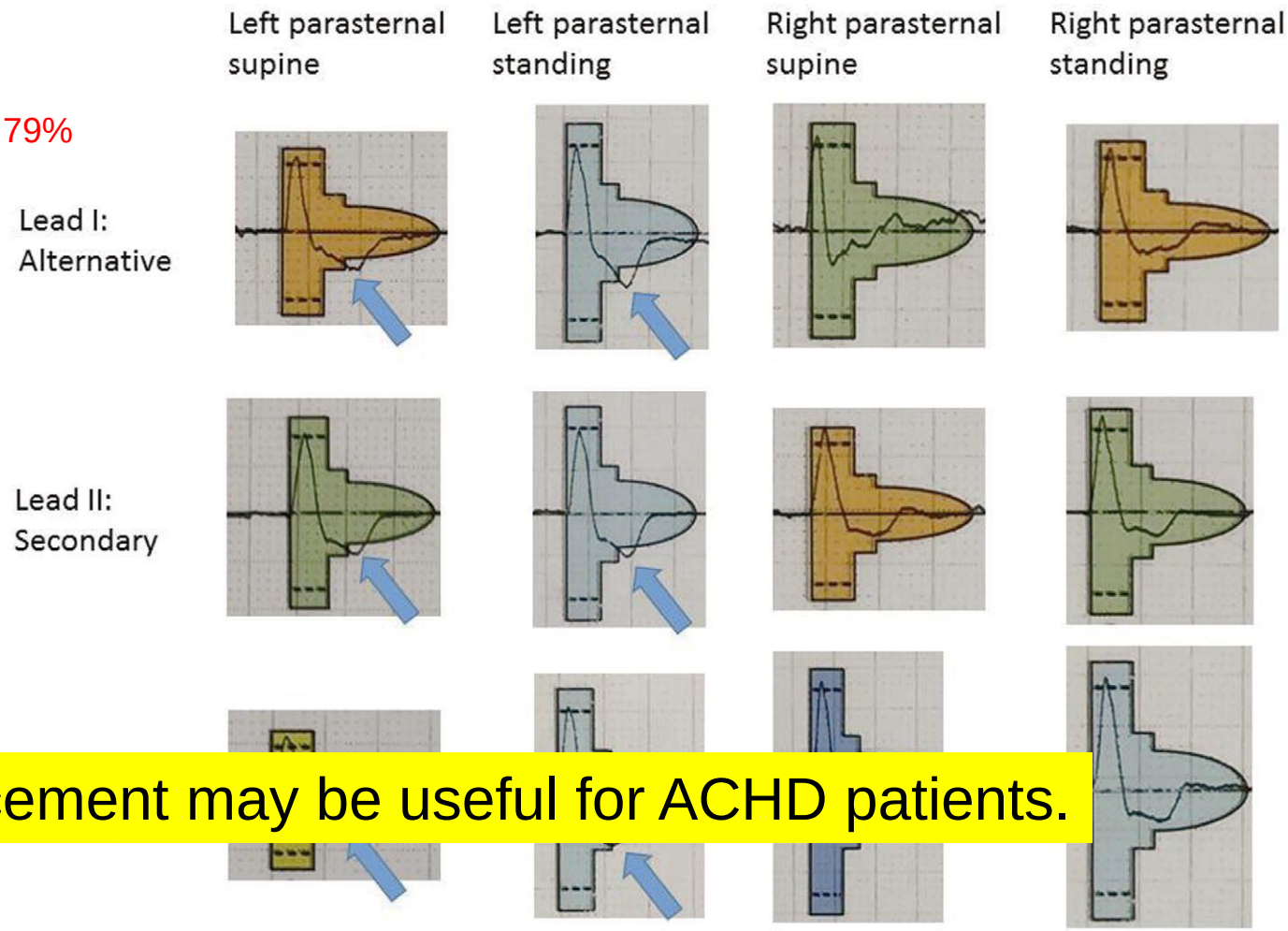
Standard screening at left parasternal position

Passed screening n=79

Failed screening n=21

Right parasternal position

Passed screening n=9



Right parasternal lead placement may be useful for ACHD patients.

Right parasternal position enabled 43% of failed patients to pass screening

Okamura H, et al. Circ J 2016; 80: 1328 – 1335.

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Summary

The S-ICD is a promising option for ACHD patients with limited vascular access, and the post-implantation defibrillation success rate has been reported to be comparable to that of the TV-ICD. On the other hand, much of the evidence is small-group and retrospective studies, lacking RCTs and large-scale studies.

Ineligibility for S-ICD in ECG screening due to abnormal T wave morphology and complication of intraventricular conduction disorders such as right bundle branch block, temporal changes in QRS configuration, remain issues associated with S-ICD selection.