



PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

**ROMA**

Centro Congressi  
di Confindustria

**Auditorium  
della Tecnica**

**9ª Edizione**

**30 Settembre**

**1 Ottobre**

**2022**

**Cardiopatie Congenite**

**PACING E SICD NEI CONGENITI**

**Berardo Sarubbi**

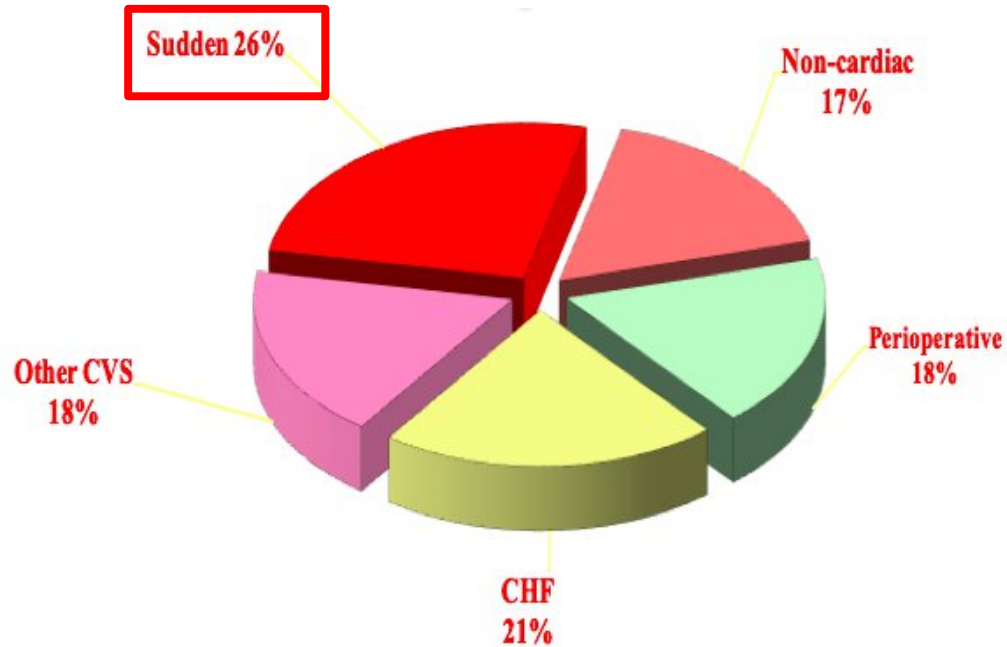
*UOSD Cardiopatie Congenite dell'Adulto*

*A.O.R.N. dei Colli - Ospedale Monaldi - Napoli*

*[www.berardosarubbi.it](http://www.berardosarubbi.it)*

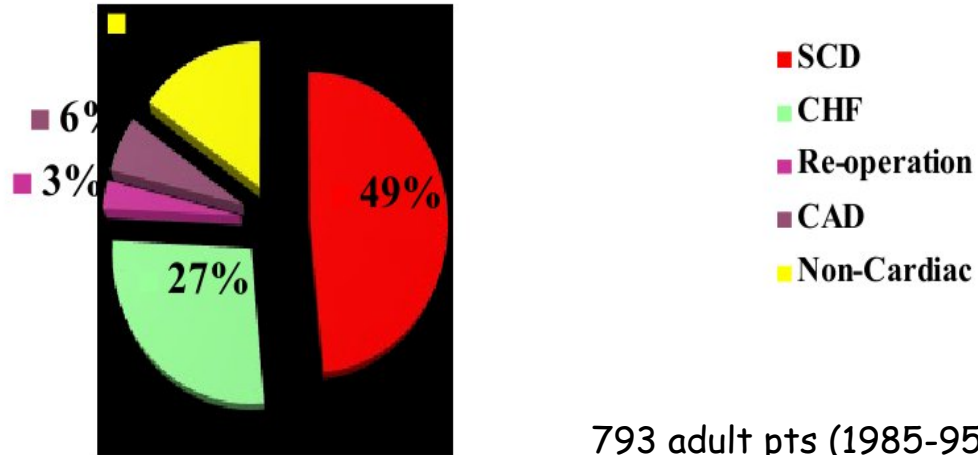


# Causes of Death in GUCH



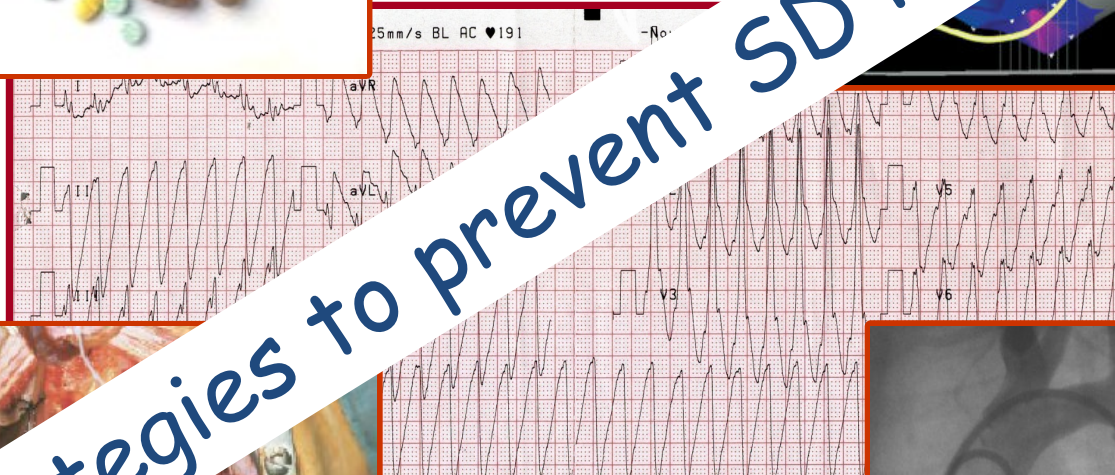
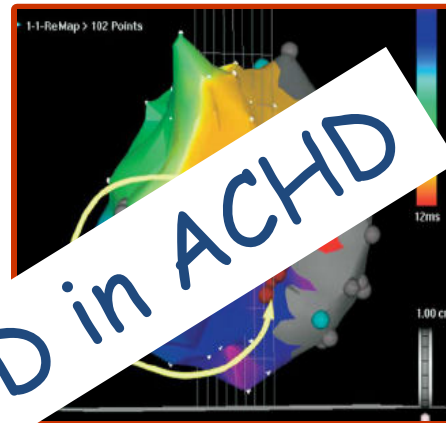
*Oechsling et al Am J Cardiol 2000*

# Late Death in Repaired Tetralogy



793 adult pts (1985-95)  
33 pts died (4.2% mortality)

*Gatzoulis et al Lancet 2000*



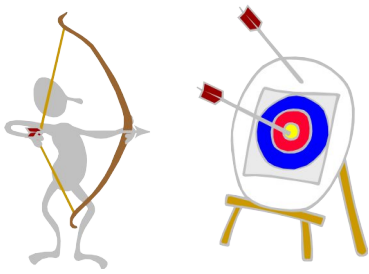
Strategies to prevent SD in ACHD



## Recommendations for treatment of arrhythmias ACHD (3)

Recommendations	Class	Level
<b>Implantable Cardiac Defibrillator</b>		
ICD implantation is indicated in adults with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically intolerated VT after evaluation to define the cause of the event and exclusion of reversible causes.	<b>I</b>	<b>C</b>
ICD implantation is indicated in adults with CHD and sustained VT after haemodynamic evaluation and repair when indicated. EP evaluation is required to identify patients in whom catheter ablation or surgical ablation may be beneficial as adjunctive treatment or in whom it may offer a reasonable alternative.	<b>I</b>	<b>C</b>

©ESC



# Issues for the use of ICD in ACHD

- ✓ Indications
- ✓ Inappropriate shocks and lead failure
- ✓ Unique anatomical situations in CHD
- ✓ Technical difficulties

# Morbidity associated with ICDs in TOF - inappropriate therapies and lead fracture

## Implantable cardioverter-defibrillator therapy in adult patients with tetralogy of Fallot

Klaus K. Witte\*, Christopher B. Pepper, J. Campbell Cowan, John D. Thomson, Kate M. English, and Michael E. Blackburn

Department

Received 1 De



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

Clinical research  
Arrhythmia/electrophysiology

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

Sing-Chien Yap<sup>1\*</sup>,  
Hubert W. Vliege  
and Willem Drent

Journal of the American College of Cardiology  
© 2008 by the American College of Cardiology Foundation  
Published by Elsevier Inc.

Vol. 51, No. 17, 2008  
ISSN 0735-1097/08/\$14.00  
doi:10.1016/j.jacc.2008.01.033

Heart Rhythm Disorders

### Results of a Multicenter Retrospective Implantable Cardioverter-Defibrillator Registry of Pediatric and Congenital Heart Disease Patients

Charles I. Berul, MD,\* George F. Van Hare, MD,†† Naomi J. Kertesz, MD,§ Anne M. Dubin, MD,† Frank Cecchin, MD,\* Kathryn K. Collins, MD,† Bryan C. Cannon, MD,§ Mark E. Alexander, MD,\* John K. Friedman, MD,\* Edward P. Walsh, MD,\* Richard A. Friedman, MD§  
Boston, Massachusetts; Palo Alto and San Francisco, California; and Houston, Texas

## Inappropriate shock:

10 % SCD-HEFT  
21 % Paed ICD Registry  
25 % rTOF Leeds  
30-40 % rTOF Euroheart survey

## 14% lead failure

Berul CI et al. JACC 2008

## Congenital Heart Disease

## 9.1% lead fracture in TOF

Khairy P et al. Circulation 2008

## Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;  
Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD;  
Susan M. Fernandes, MHP, PA-C; Luc Beauchesne, MD; Judith Therrien, MD; Philippe Chetaille, MD;  
Elaine Gordon, MD; Isabelle Vorder Muhll, MD; Frank Cecchin, MD

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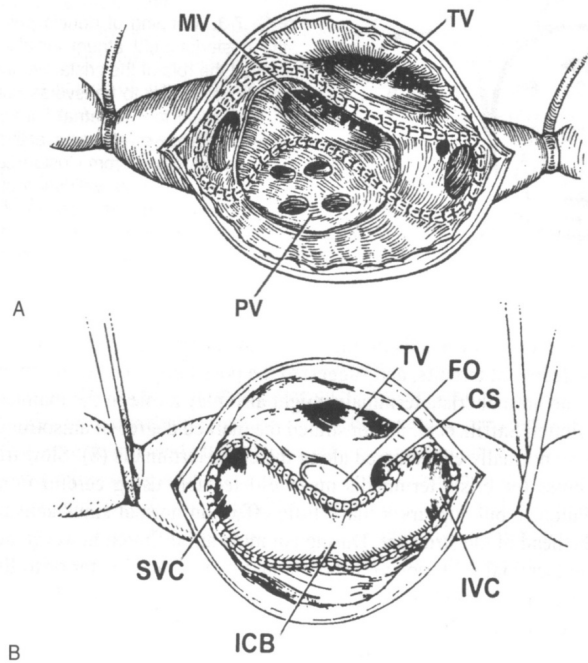
## **High Failure Rate for an Epicardial Implantable Cardioverter-Defibrillator Lead: Implications for Long-Term Follow-Up of Patients With an Implantable Cardioverter-Defibrillator**

PETER A. BRADY, MB, MRCP, PAUL A. FRIEDMAN, MD, JANE M. TRUSTY, RN,  
SUELLEN GRICE, RN, STEPHEN C. HAMMILL, MD, FACC, MARSHALL S. STANTON, MD, FACC

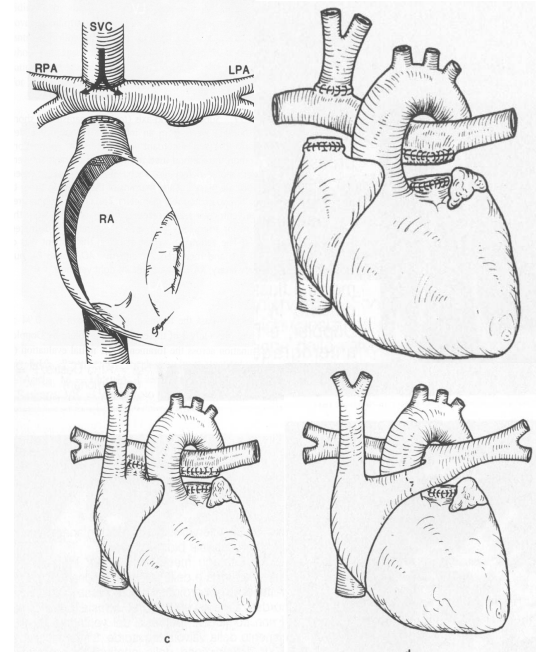
*Rochester, Minnesota*

- ✓ **Epicardial lead malfunction is common on long -term follow-up.**
- ✓ **Some leads have a failure of 28% at 4yrs**

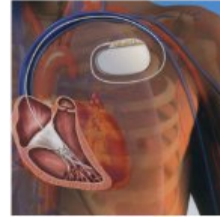
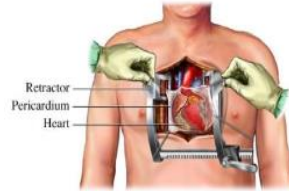
# TGA s/p Mustard/Senning



# s/p Fontan



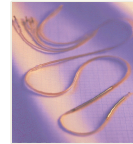
# Cardiac Defibrillator Therapy



195  
Paul Zoll  
1<sup>st</sup> human  
external  
defibrillation



198  
1<sup>st</sup> human  
implant of  
AICD



198  
8  
1<sup>st</sup> human implant of  
endocardial shocking  
lead  
*Eliminated need  
for thoracotomy*



2008  
S-ICD



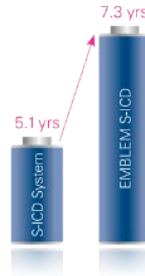
# EMBLEM™ S-ICD System

20% reduction  
in Device Profile



20% reduction in device profile

Longevity > 7,3 yrs



2 year improvement in projected  
longevity with Boston Scientific  
battery technology<sup>2</sup>

Compatible Latitude



LATITUDE Remote Patient  
Management Enabled<sup>2</sup>

2015



**Table 2. Characterization of Patient Groups for S-ICD Implantation**



**Who  
Impl:**

**The Su  
(ICD)**

**Who D**

*Jeanne E. J*

S-ICD is preferred device

No venous access (occluded veins or congenital anomalies)

High risk of complications for transvenous systems have (dialysis, pediatric, and immunocompromised)



Channelopathies (long-QT syndrome, Brugada, hypertrophic cardiomyopathy)

Previous device infections or lead failures

History of endocarditis

S-ICD should be strongly considered

Young patients

*or*

Life expectancy >10 y

Primary prevention indicated patients with ischemic/nonischemic heart failure

Prosthetic valves

Women (preferred generator placement lateral wall)

Selected secondary prevention indicated patients (survivors of out-of-hospital VF, no evidence of monomorphic VT)

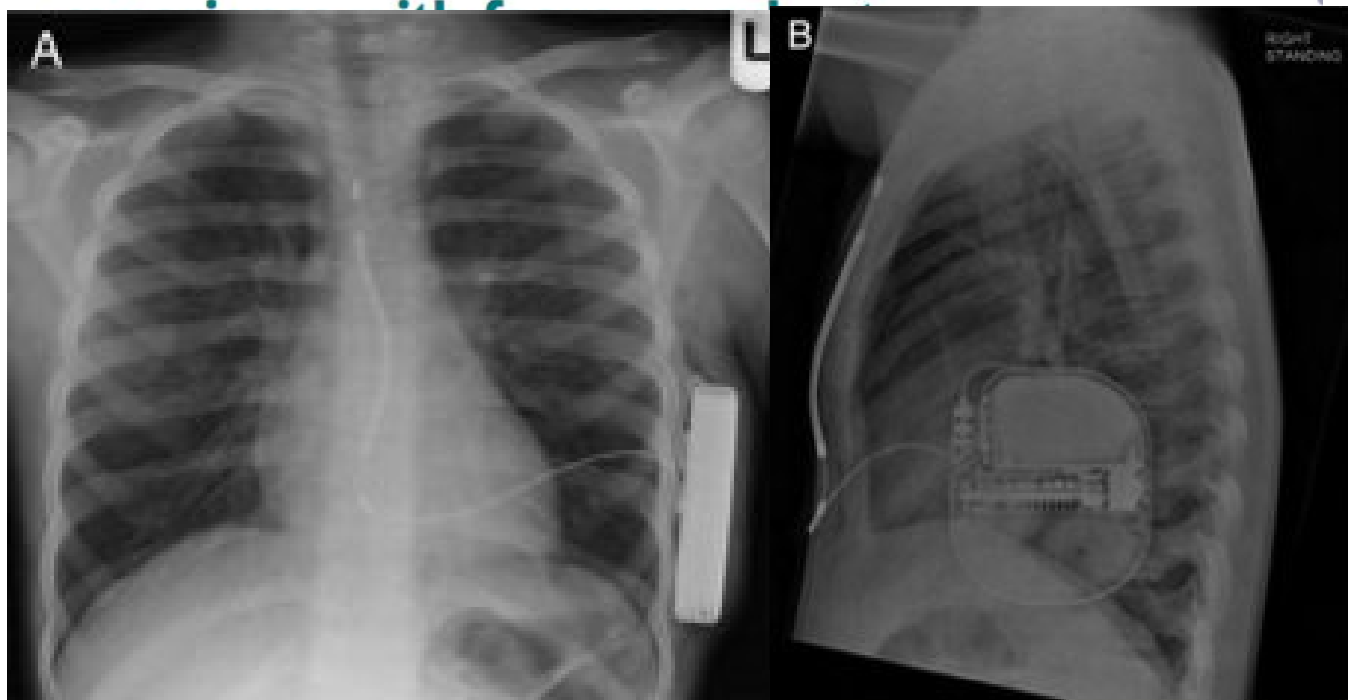
S-ICD should be avoided

Systolic heart failure and LBBB who are indicated for CRT

Symptomatic bradycardia requiring pacemaker

Recurrent sustained monomorphic VT for whom ATP is deemed appropriate

## Implantable cardioverter defibrillator therapy in paediatric practice: a single-centre UK





Le  
Alt

Le  
Se

Le  
Pri



**ESC**

European Society  
of Cardiology

Europace (2018) 0, 1–8

doi:10.1093/europace/euy139

**CLINICAL RESEARCH**

# **Subcutaneous implantable cardioverter-defibrillator: is it ready for use in children and young adults? A single-centre study**

**Massimo Stefano Silvetti<sup>1</sup>, Vincenzo Pazzano<sup>1</sup>, Letizia Verticelli<sup>1</sup>, Irma Battipaglia<sup>1</sup>, Fabio Anselmo Saputo<sup>1</sup>, Sonia Albanese<sup>2</sup>, Mariolina Lovecchio<sup>3</sup>, Sergio Valsecchi<sup>2</sup>, and Fabrizio Drago<sup>1</sup>**

**Table 1** Baseline patient characteristics and therapy

No.	Gender	Arrhythmia symptom	Age (years)	Height (cm)	Weight (kg)	BSA (m <sup>2</sup> )	BMI	Heart disease	LVEF (%)	Prevention	Drugs
1	Male	SND, AFI, NSVT	30	175	83	2.03	27.1	TGA s/p Mustard	32	Primary	Carvedilol, ASA
2	Male	SND, AFI, NSVT	29	180	70	1.86	21.6	TGA s/p Mustard	49	Primary	Sotalol, ASA, ACEI
3	Female	NSVT	17	174	82	2.01	27.1	ARVC	50	Primary	Sotalol
4	Male	NSVT	10	139	38	1.23	19.7	Familial HOCM	60	Primary	Metoprolol, ASA
5	Female	PVC	15	153	43	1.36	18.4	AT, TGA s/p Glenn	20	Primary	Carvedilol, OAT, Digoxin
6	Female	NSVT, syncope	15	142	49	1.43	24.3	ARVC	45	Primary	Carvedilol, ACEI
7	Female	NSVT	14	169	52	1.55	18.2	Familial HNOCM	68	Primary	Metoprolol
8	Female	No	11	156	52	1.52	21.4	Familial HNOCM	65	Primary	Metoprolol
9	Female	VT, CA	15	164	75	1.88	27.9	Familial ARVC	40	Secondary	Amiodarone, ASA
10	Female	SND, VT, syncope	28	160	65	1.73	25.4	UVH s/p Fontan	38	Primary	Carvedilol, OAT
11	Male	LQT >500 ms	15	170	55	1.60	19.0	LQTS2	68	Primary	Propranolol Mexiletin
12	Male	NSVT	31	170	60	1.69	20.8	PA, VSD, MAPCA s/p repair, PH	35	Primary	Metoprolol, Sildenafil Diuretics
13	Female	NSVT, syncope	20	171	62	1.72	21.2	Familial ARVC	50	Primary	Nadolol
14	Female	VF inducible	10	151	61	1.64	26.8	ARVC	47	Primary	Carvedilol
15	Female	PVC	15	166	58	1.63	20.7	ALCAPA, DCM	29	Primary	Carvedilol, ACEI, ASA, Diuretics, Ivabradin



## Subcutaneous implantable cardioverter defibrillator in children and adolescents: results from the S-ICD “Monaldi care” registry

Berardo Sarubbi<sup>1</sup>  • Diego Colonna<sup>1</sup> • Anna Correra<sup>1</sup> • Emanuele Romeo<sup>1</sup> • Michele D’Alto<sup>1</sup> • Maria Teresa Palladino<sup>2</sup> • Salvatore Virno<sup>1</sup> • Antonio D’Onofrio<sup>3</sup> • Maria Giovanna Russo<sup>2</sup>

Received: 10 May 2020 / Accepted: 15 February 2021  
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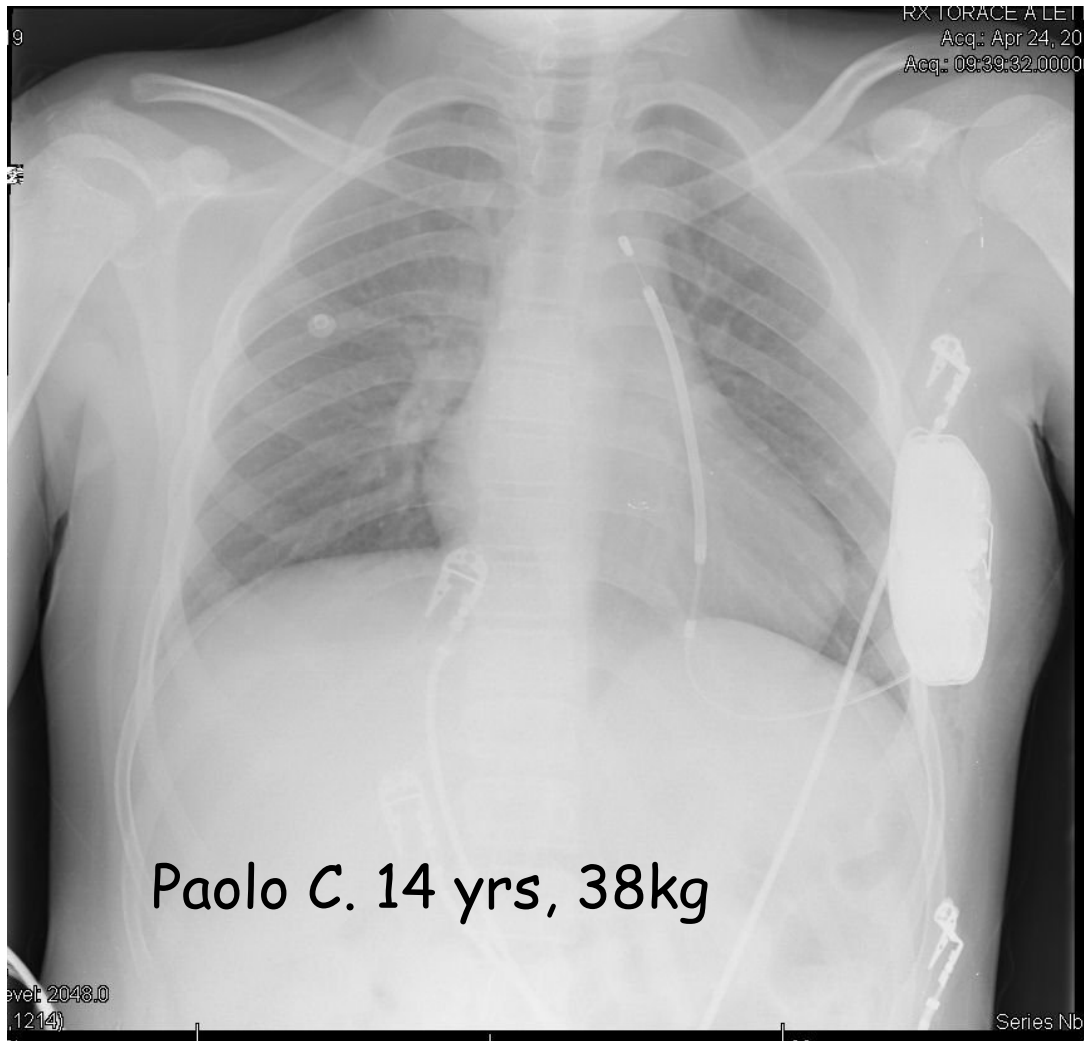
- 297 patients enrolled in the S-ICD “Monaldi care” April 2014 to June 2020
- 21 consecutive children and adolescents
- mean age 13.9 years, range 8-18 years
- mean body weight 59.3 kg, range 38-100 kg
- Mean follow-up 41.9±21.9 months.

**Table 2** Procedural data and outcome

Pt.	Previous device	Years of implant	S-ICD type	Sensing vectors	Incision type	Lead position	SICD position	Conditional shock zone/ shock zone	Intra-op. def. test	Complications	AS	IAS	F-U months	Outcome
#1	No	2018/emblem A219	3	2	Left	Left/Intermusc	220/250 bpm	Yes	None	No	No	No	24	Good
#2	No	2015/emblem A209	1	3	Left	Left/Intermusc	220/250 bpm	Yes	None	No	No	No	64	Good
#3	No	2014/emblem A209	3	3	Left	Left/Intermusc	220/250 bpm	No (no induc.)	None	No	No	No	78	Good SICD replaced (2020) Emblem A219
#4	No	2017/emblem A219	3	2	Left	Left/Intermusc	220/250 bpm	Yes	None	No	No	No	35	Good
#5	No	2016/emblem A209	3	2	Right	Left/Intermusc	220/250 bpm	Yes	None	No	Yes (double count)	Yes	55	Good
#6	No	2016/emblem A209	2	2	Left	Left/Subcut	220/250 bpm	Yes	None	No	No	No	55	Good
#7	No	2017/emblem A219	3	2	Left	Left/Intermusc	200/250 bpm	Yes	None	Yes	No	No	41	Good
#8	No	2019/emblem A219	2	2	Left	Left/Intermusc	200/250 bpm	No (no induc.)	None	No	No	No	18	Good
#9	No	2019/emblem A219	3	2	Left	Left/Intermusc	200/250 bpm	No (low EF)	None	No	No	No	16	Good
#10	No	2019/emblem A219	2	2	Right	Left/Intermusc	220/250 bpm	Yes	None	No	No	No	16	Good
#11	No	2019/emblem A219	3	2	Left	Left/Intermusc	200/250 bpm	Yes	None	No	No	No	20	Good
#12	No	2014/emblem A219	1	3	Left	Left/Subcut	220/250 bpm	Yes	None	No	No	No	70	Good
#13	No	2019/emblem A219	2	2	Left	Left/Intermusc	200/250 bpm	Yes	Skin erosion/-infection	No	No	No	15	Good
#14	No	2016/emblem A209	2	2	Left	Left/Intermusc	200/250 bpm	Yes	None	Yes	Yes (double count)	Yes	36	Reposition lead implant ICD
#15	No	2017/emblem A219	1	2	Left	Left/Intermusc	200/250 bpm	Yes	None	No	Yes (atrial arrhythm)	Yes	40	Good
#16	No	2016/emblem A209	3	2	Left	Left/Intermusc	220/250 bpm	Yes	None	No	No	No	55	Good
#17	No	2014/emblem A209	3	3	Left	Left/Subcut	200/220 bpm	Yes	None	No	No	No	75	Good
#18	No	2020/emblem A219	3	2	Left	Left/Intermusc	200/250 bpm	Yes	None	No	No	No	6	Good
#19	Yes	2020/emblem A219	2	2	Right	Left/Intermusc	200/250 bpm	No (no induc.)	None	No	No	No	7	Good
#20	No	2020/emblem A219	2	2	Left	Left/Intermusc	230/250 bpm	No (no induc.)	None	No	No	No	4	Good
#21	No	2020/emblem A219	2	2	Left	Left/Intermusc	230/250 bpm	Yes	None	No	Yes (double count)	Yes	4	Good

Intermusc, intermuscular; Subcut, subcutaneous; EF, ejection fraction; AS, appropriate shock; IAS, inappropriate shock



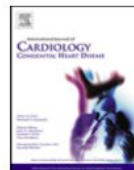




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International Journal of Cardiology Congenital Heart Disease

journal homepage: [www.journals.elsevier.com/international-journal-of-cardiology-congenital-heart-disease](http://www.journals.elsevier.com/international-journal-of-cardiology-congenital-heart-disease)



## Subcutaneous implantable cardioverter defibrillator in complex adult congenital heart disease. Results from the S-ICD “Monaldi Care” registry



Berardo Sarubbi<sup>a,\*</sup>, Anna Correra<sup>a</sup>, Diego Colonna<sup>a</sup>, Emanuele Romeo<sup>a</sup>, Michela Palma<sup>a</sup>, Assunta Merola<sup>a</sup>, Michele D'Alto<sup>b</sup>, Giancarlo Scognamiglio<sup>a</sup>, Flavia Fusco<sup>a</sup>, Rosaria Barracano<sup>a</sup>, Nunzia Borrelli<sup>a</sup>, Nicola Grimaldi<sup>a</sup>, Antonio D'Onofrio<sup>c</sup>, Maria Giovanna Russo<sup>d</sup>

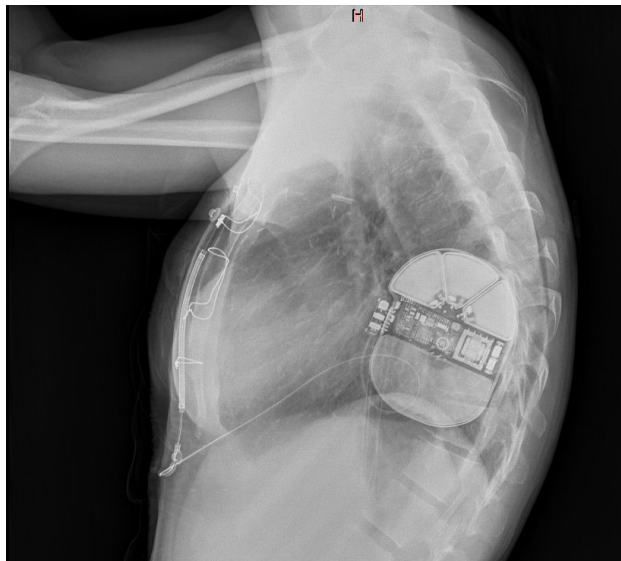
<sup>a</sup> Adult Congenital Heart Disease Unit, Monaldi Hospital, Naples, Italy

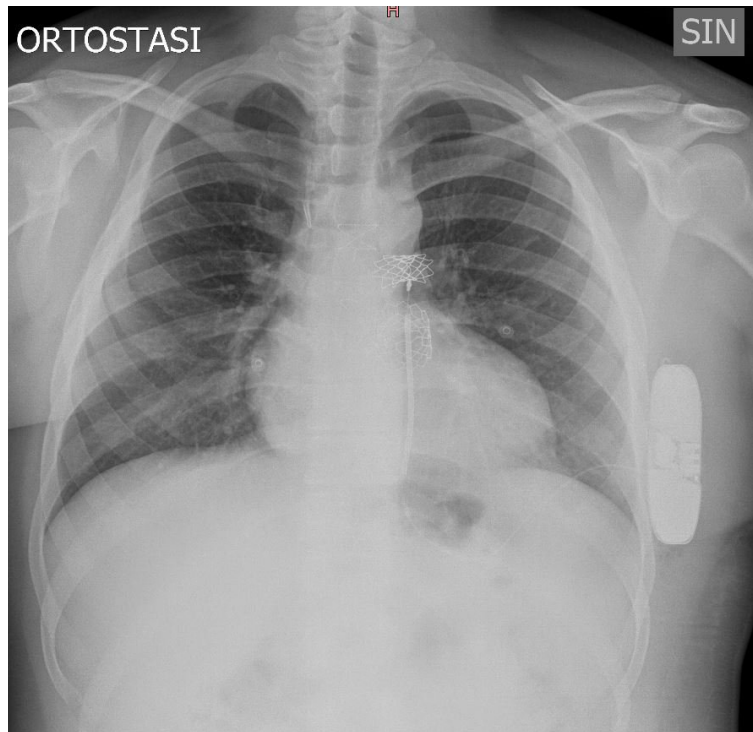
<sup>b</sup> Cardiology Unit. “L. Vanvitelli” University, Monaldi Hospital, Naples, Italy

<sup>c</sup> Electrophysiology and Cardiac Pacing Unit, Monaldi Hospital, Naples, Italy

<sup>d</sup> Paediatric Cardiology Unit. “L. Vanvitelli” University, Monaldi Hospital, Naples, Italy

- From a cohort of 297 pts enrolled in the S-ICD “Monaldi care” registry
- 14 consecutive complex ACHD patients (aged 35.9 +/- 16.7 years).
- All the patients showed a good compliance to the device system with no complications (infections or skin erosions).
- Four patients were listed for heart transplantation (HTX).





Impact of S-ICD vs ICD  
in ACHD patients  
*Data from "Monaldi Care Registry"*  
*"ACHD population subset"*

No significant Differences in terms of:

- Hospital care
- Time for procedure
- Acute complications
- Early/Late complications
- .....



6 hours after implant



# Good Cosmetic Outcome





**Table 2. Characterization of Patient Groups for S-ICD Implantation**



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**The Su (ICD)**

**Who D**

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High risk of complications for transvenous systems have (dialysis, pediatric, and immunocompromised)



Channelopathies (long-QT syndrome, Brugada, hypertrophic cardiomyopathy)

Previous device infections or lead failures

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Young patients

*or*

Life expectancy >10 y

Primary prevention indicated patients with ischemic/nonischemic heart failure

Prosthetic valves

Women (preferred generator placement lateral wall)

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Systolic heart failure and LBBB who are indicated for CRT

Symptomatic bradycardia requiring pacemaker

Recurrent sustained monomorphic VT for whom ATP is deemed appropriate

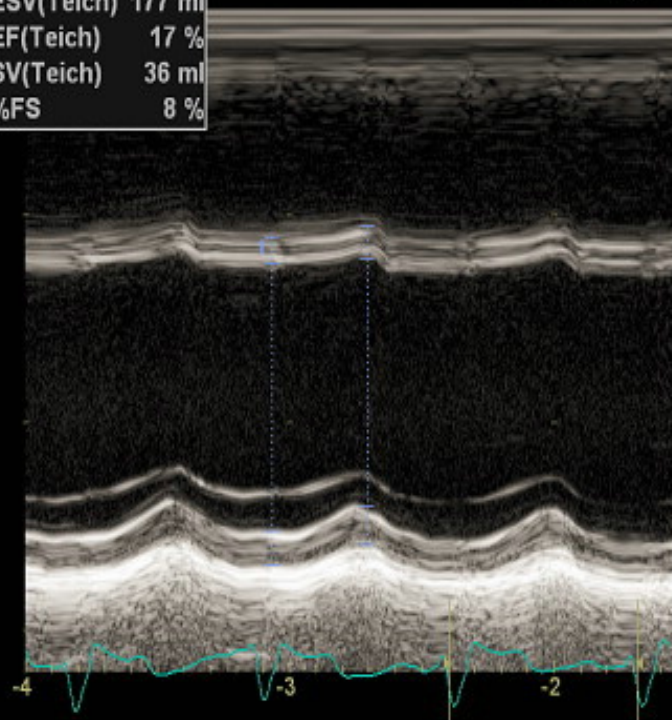
# A. L. 27 yrs

## Diagnosis of TGA + VSD+ PS

- Aged 9 mths: Blalock-Hanlon atrial septectomy
- Aged 3 yrs: Rastelli procedure
- Aged 16 yrs: RV-PA conduit replacement
- Aged 21yrs: Diagnosis of early conduit stenosis
- Aged 25 yrs: Bicameral Pacemaker implant
- Aged 26yrs: Repeated hosp.admission for CHF.  
LV EF:10%
- Aged 27 yrs: in the waiting list for HTx.  
ECG 24h Holter: repeated n.s. VT

10:50:16

1	IVSd	0.6 cm
	IVSs	0.8 cm
	LVIDd	6.5 cm
	LVIDs	6.0 cm
	LVPWd	0.8 cm
	LVPWs	0.9 cm
	EDV(Teich)	213 ml
	ESV(Teich)	177 ml
	EF(Teich)	17 %
	SV(Teich)	36 ml
	%FS	8 %



-[cm]

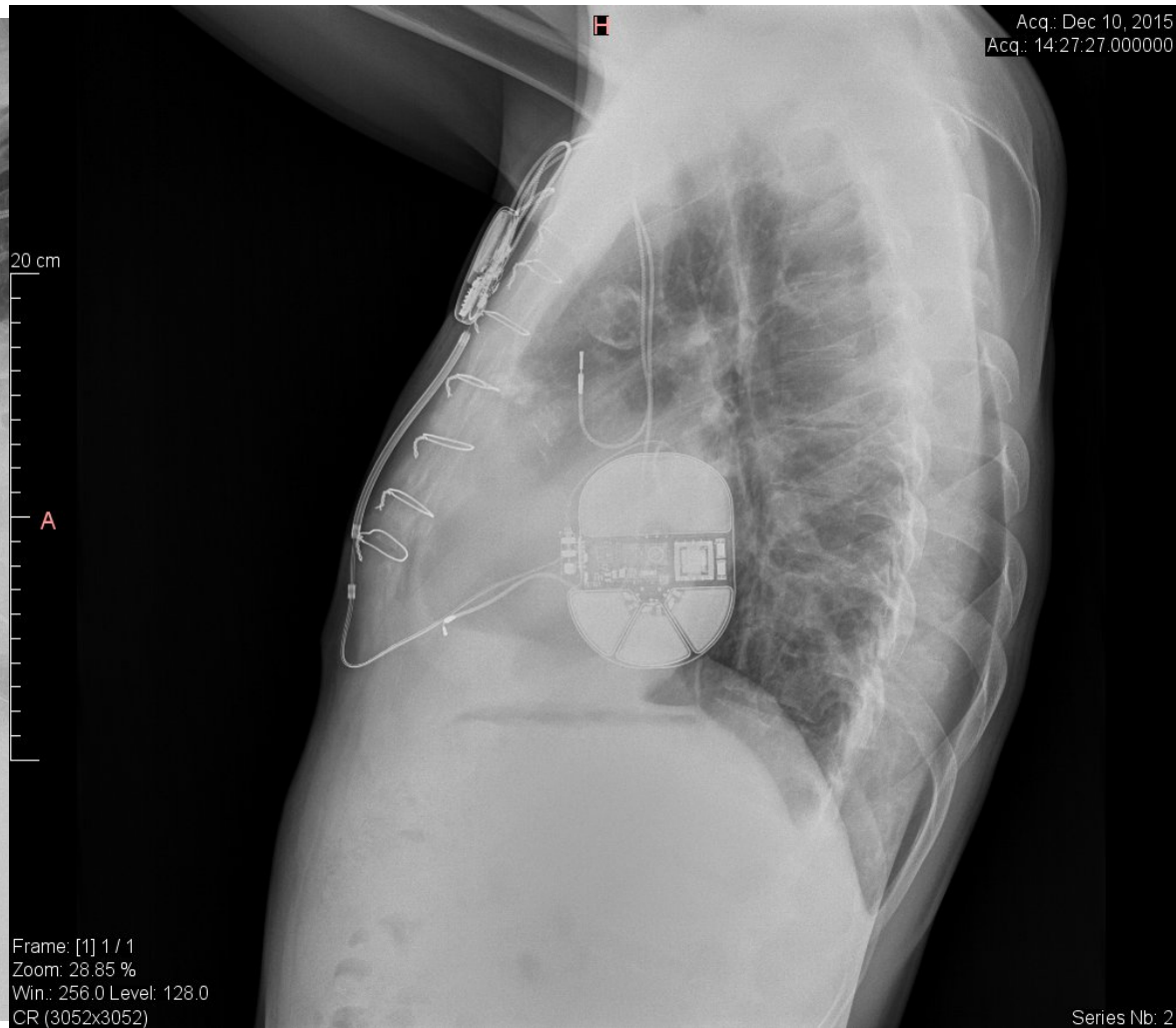
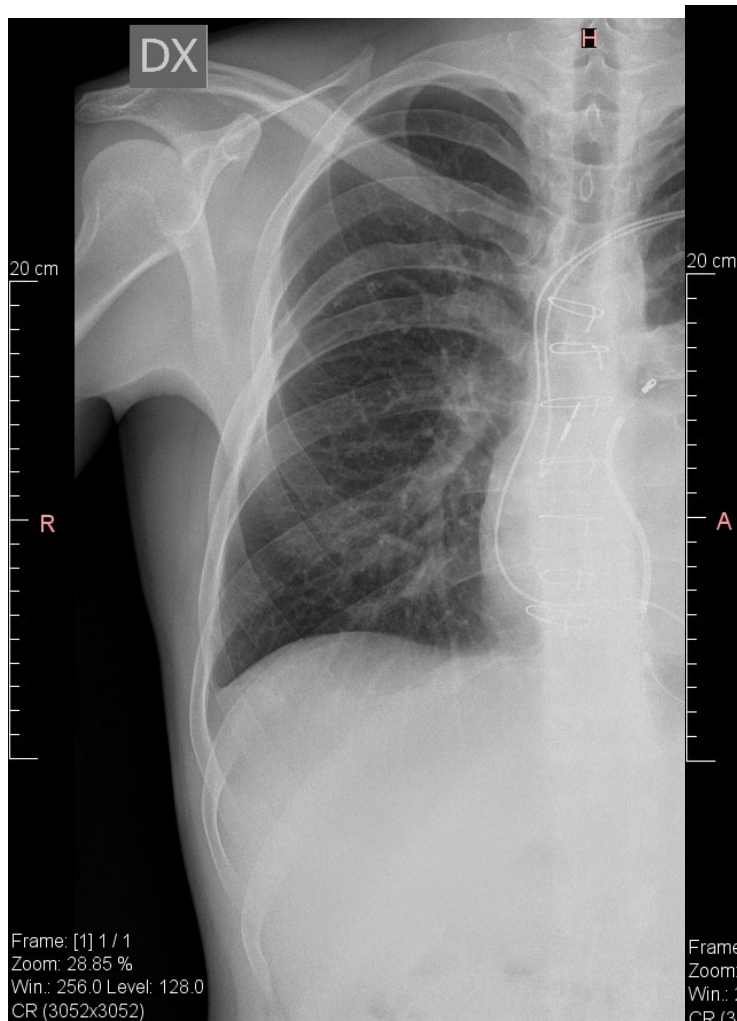
-5

-10

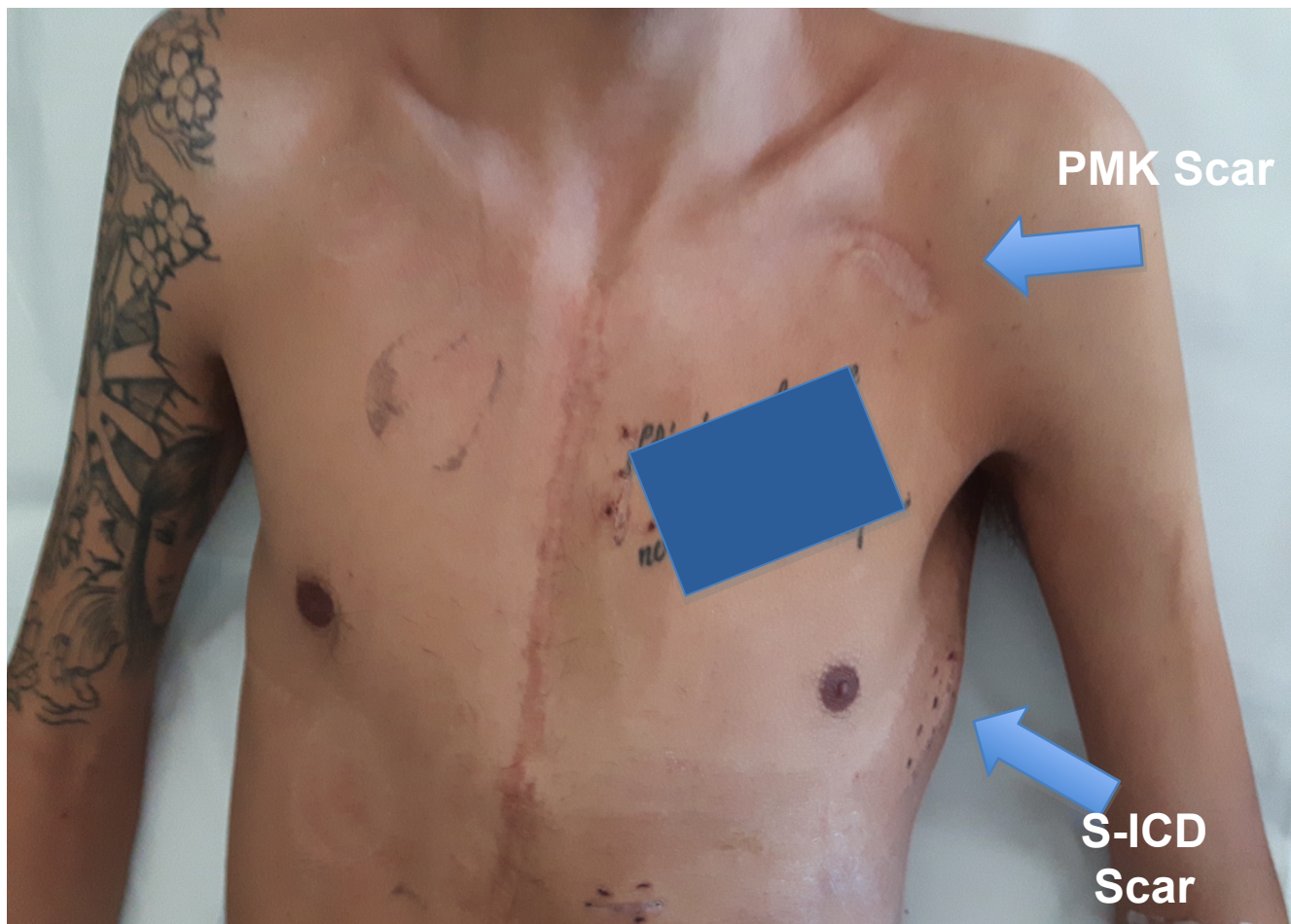
-15

50 mm/s

83  
HR







**CASE REPORT**WILEY *Journal of Arrhythmia*

# Combination of a leadless pacemaker and subcutaneous implantable cardioverter defibrillator therapy for a Japanese patient with prosthetic valve endocarditis

Ryo Ito MD<sup>1</sup> | Yusuke Kondo MD, PhD<sup>2</sup>  | Joachim Winter MD, PhD<sup>3</sup> |  
Tomohiko Hayashi MD<sup>1</sup> | Miyo Nakano MD<sup>1</sup> | Takatsugu Kajiyama MD, PhD<sup>1</sup>  |  
Masahiro Nakano MD, PhD<sup>2</sup> | Yoshio Kobayashi MD, PhD<sup>1</sup>

biventricular pacing met criteria compared to during RV pacing alone (80% vs. 46%,  $P < 0.01$ ). Patients that were paced from the RV septum were more likely to qualify compared to those paced from the RV apex (67% vs. 37%, respectively,  $P < 0.01$ ).

**Conclusion:** While S-ICD implantation may be considered as supplemental therapy in select patients with preexisting transvenous devices, relatively fewer candidates who are paced from the RV apex qualify. QRS morphologies generated from biventricular pacing as well as from septal RV pacing are more likely to screen in based on the recommended S-ICD template. (*J Cardiovasc Electrophysiol*, Vol. 28, pp. 544-548, May 2017)

## **Inappropriate shock due to quadruple counting in a patient with subcutaneous implantable cardioverter-defibrillator and a dual-chamber pacemaker**

**Halim Marzak\*, Olivier Morel, and Laurence Jesel**

Pôle d'activité médico-chirurgicale cardio-vasculaire, Nouvel Hôpital Civil, CHU de Strasbourg, 1, place de l'Hôpital, F-67091 Strasbourg, France

\* Corresponding author. Tel: +33 (0)3 69 551467; fax: +33 (0)3 69 550970. E-mail address: halim.marzak@chru-strasbourg.fr

## **Adverse device-device interaction between pacemaker and subcutaneous implantable cardiac defibrillator**

Nicholas Abbott MD<sup>1</sup> | Aron Bender MD<sup>1</sup> | Charles Henrikson MD<sup>1</sup> |  
Jared Miller MD<sup>1</sup> | Babak Nazer MD<sup>1</sup> | Seshadri Balaji MBBS, MRCP, PhD<sup>2</sup>



Adult Congenital Heart Disease Unit. Monaldi Hospital - Naples, Italy

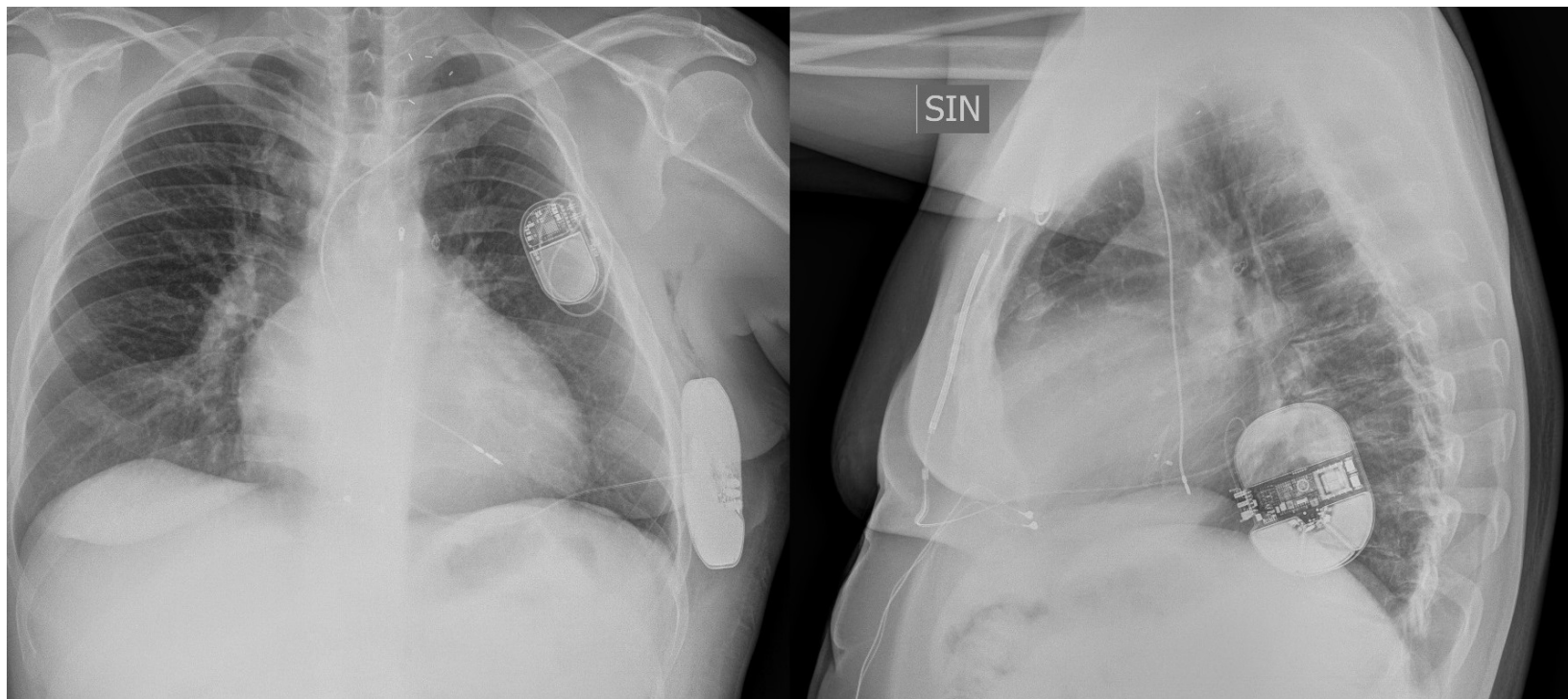
**Combined subcutaneous implantable cardioverter defibrillator and pacemaker devices in complex adult congenital heart disease.**

**A single-center experienced based study.**

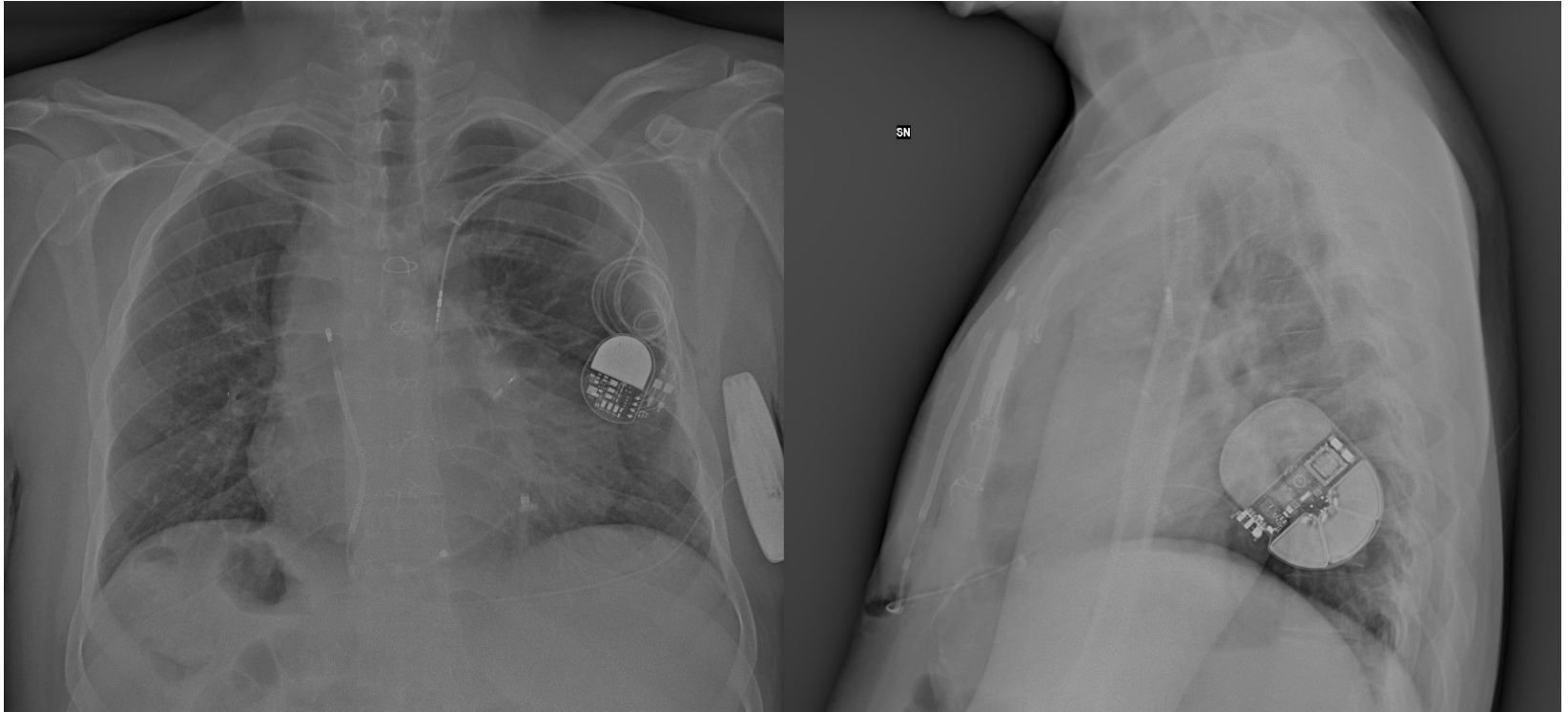
Sarubbi B et al. 2022

*Unpublished data*

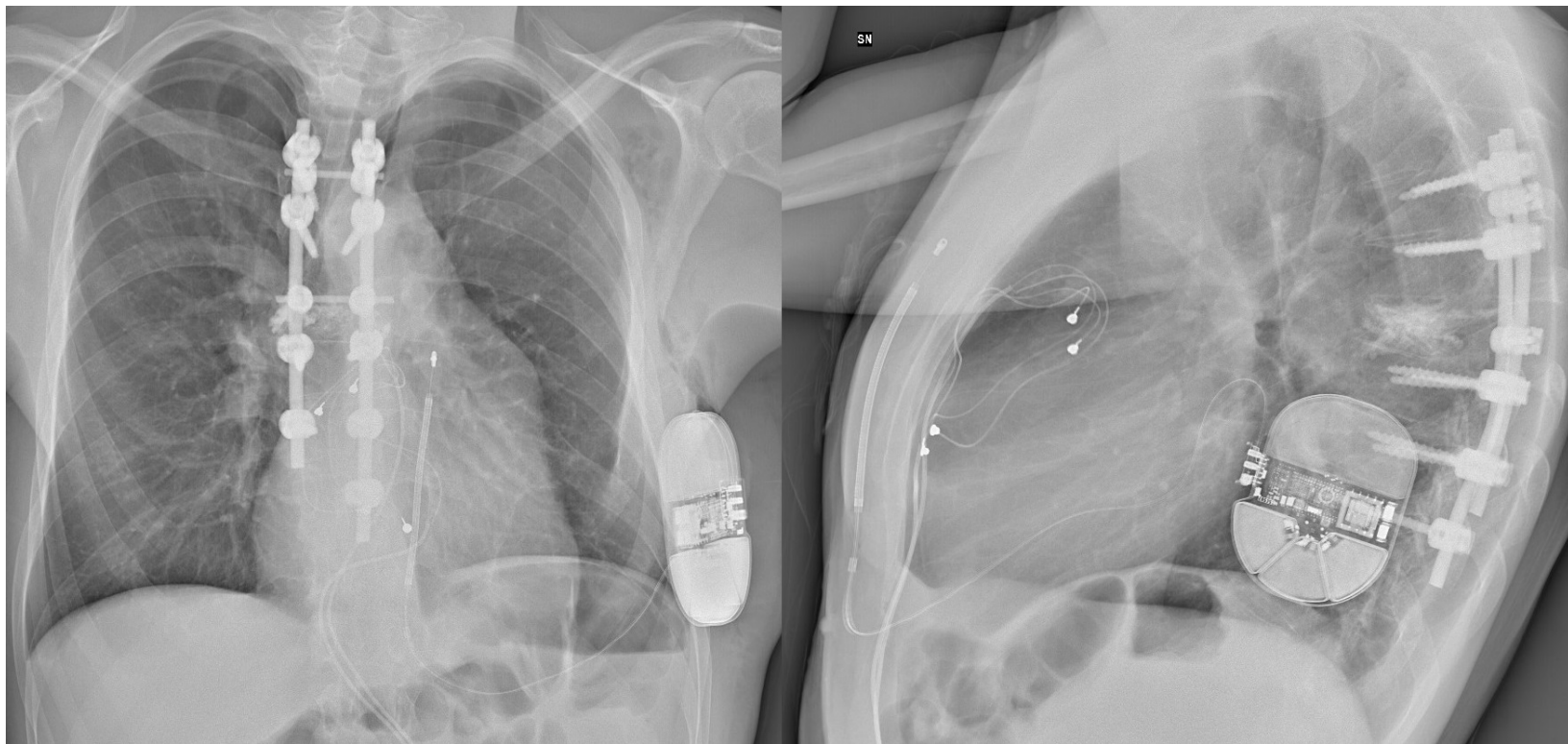
Pt.	Sex	Age	BSA (m²)	Heart disease	Indication to PMK implant	Indication to S-ICD implant
#1	M	27	1.75	TGA + VSD + PS s/p Rastelli procedure	SND	1° prevention
#2	M	73	1.84	Mitral valve stenosis s/p MV replacement	AF with SVR	1° prevention
#3	M	41	2.07	TGA + VSD + PS s/p Senning procedure	III° AVB	1° prevention
#4	M	29	2.12	ccTGA + VSD + PS + situs inversus s/p VSD closure + pulmonary valve replacement	III° AVB	2° prevention (VT/VF storm)
#5	M	43	1.82	DILV + TGA s/p pulmonary banding	III° AVB	1° prevention
#6	M	18	2.01	ccTGA s/p tricuspid valve replacement	III° AVB	1° prevention
#7	F	13	1.38	TOF + LQTS type II s/p radical correction	Postop III° AVB	1° prevention
#8	M	61	2.35	Tricuspid valve endocarditis + atrial flutter s/p tricuspid valve replacement + Maze procedure	Postop III° AVB	1° prevention
#9	M	51	1.74	ccTGA	III° AVB	1° prevention
#10	M	35	1.94	TGA s/p Mustard + SVC baffle stenting	SND	1° prevention



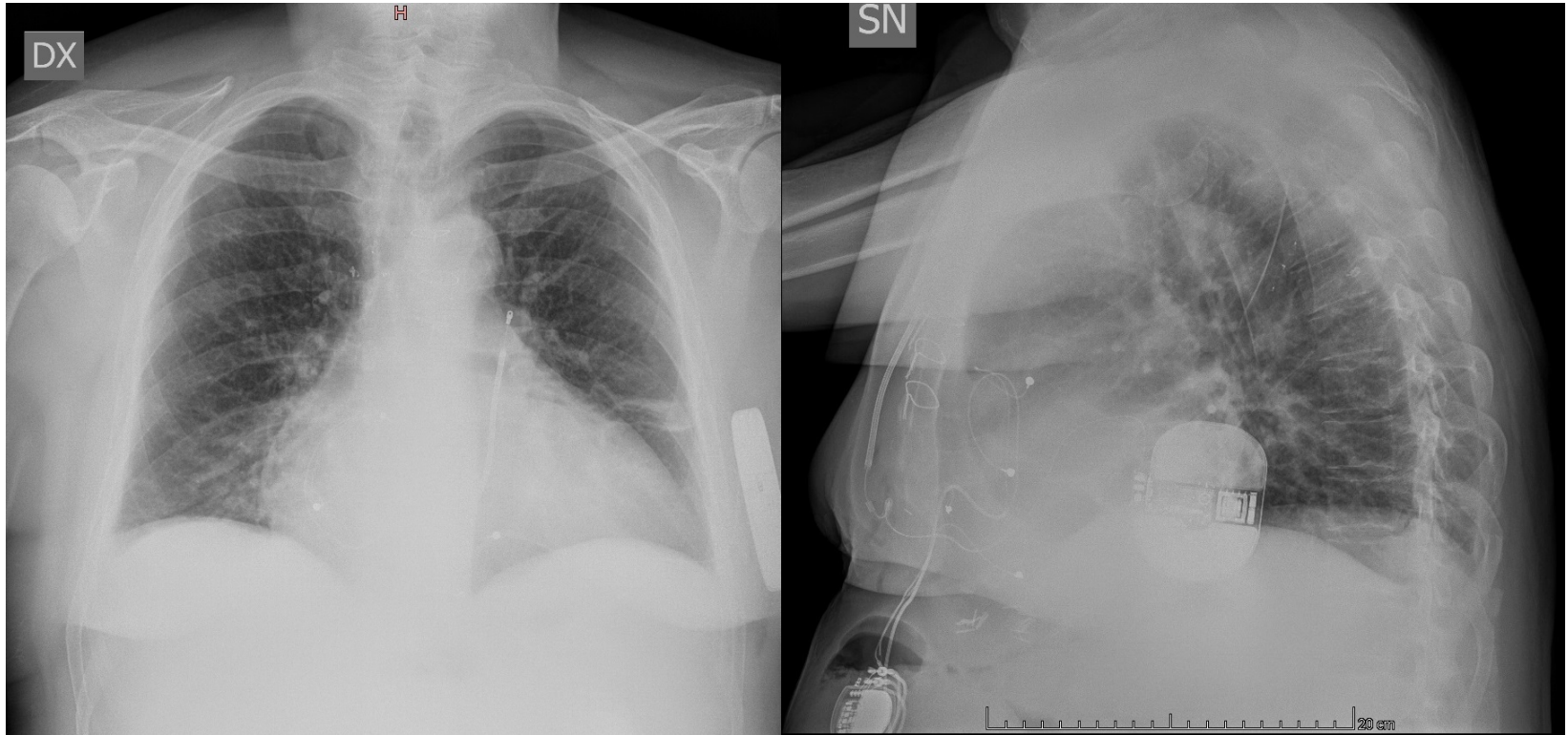
TGA + VSD + PS s/p Senning procedure



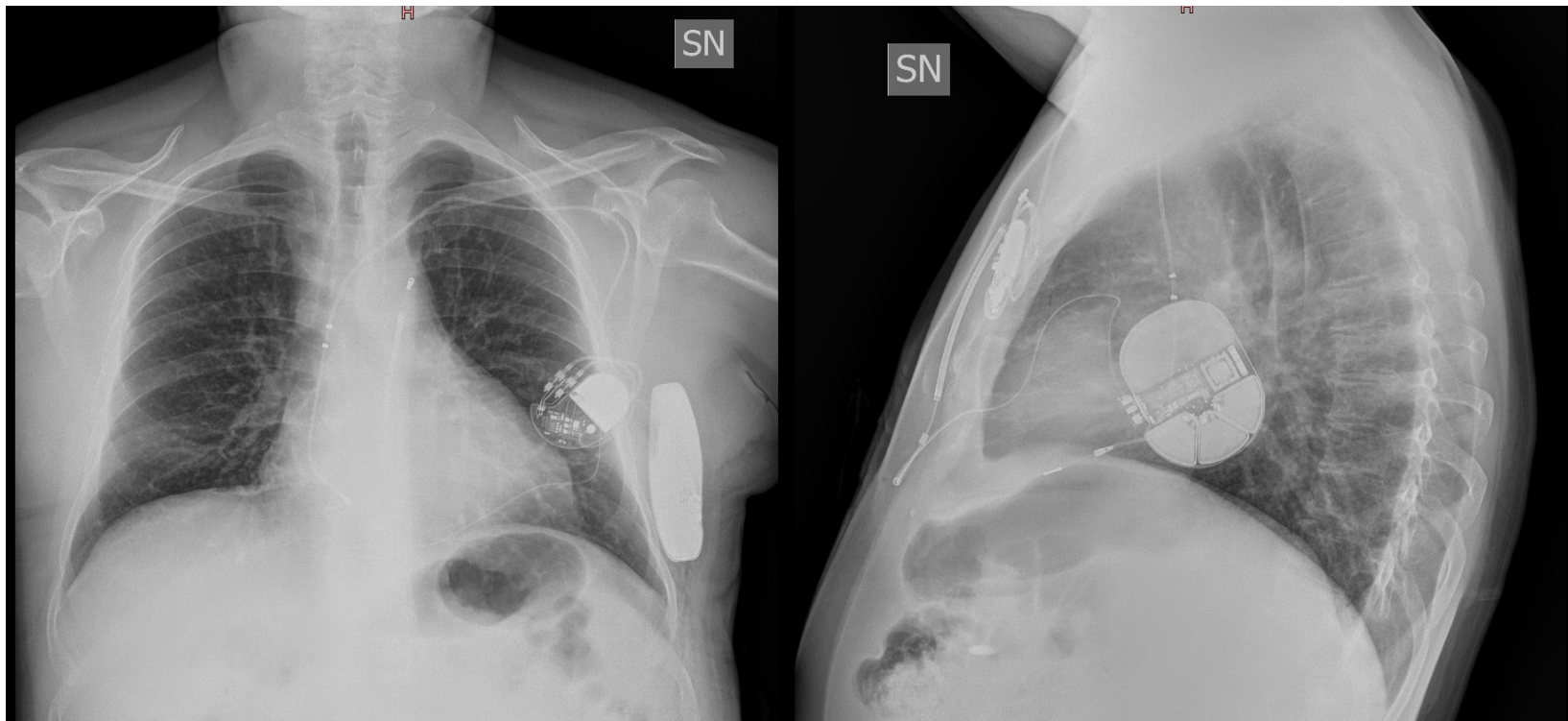
ccTGA + VSD + PS + situs inversus s/p VSD closure + pulmonary valve replacement



DILV + malposition of great arteries s/p pulmonary banding

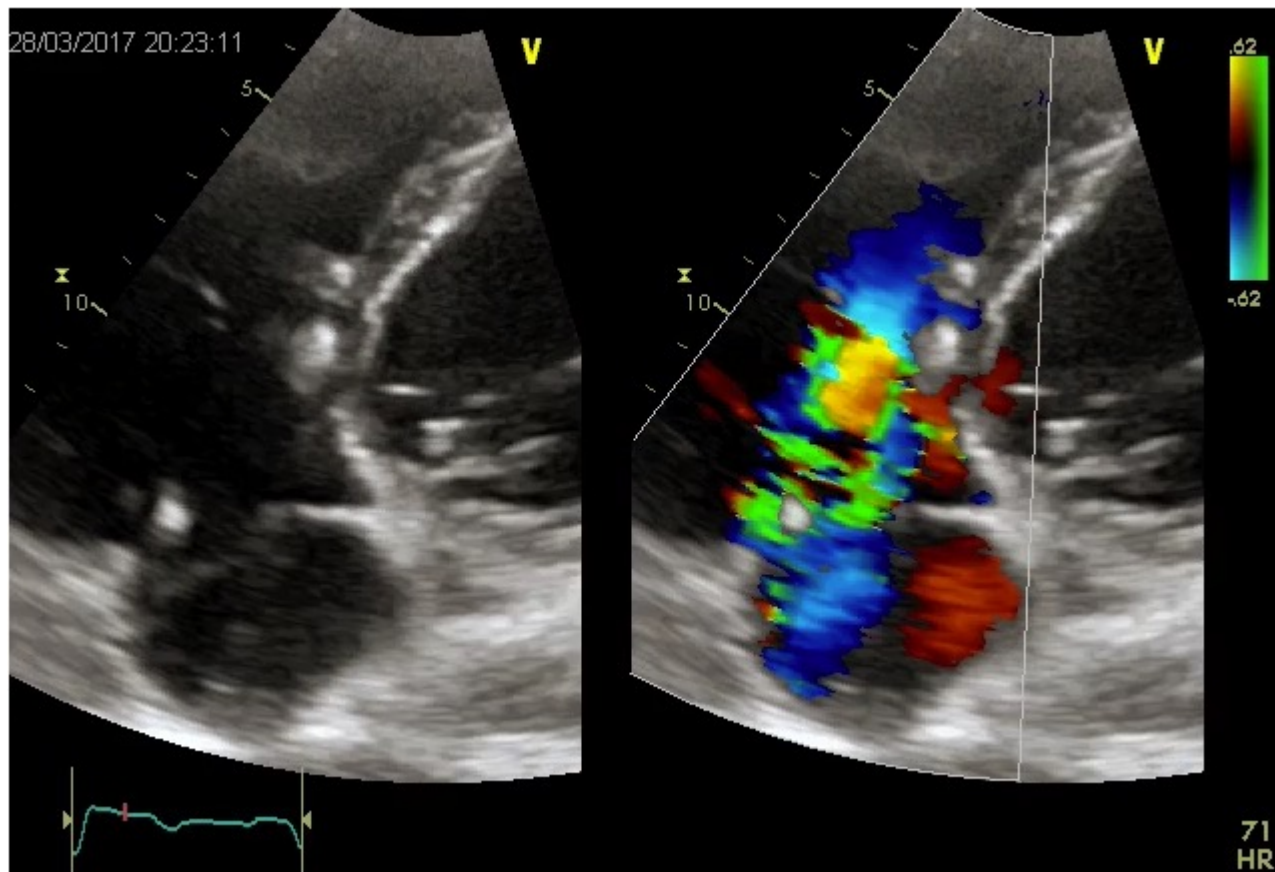


Tricuspid valve endocarditis + atrial flutter s/p tricuspid valve replacement + Maze procedure



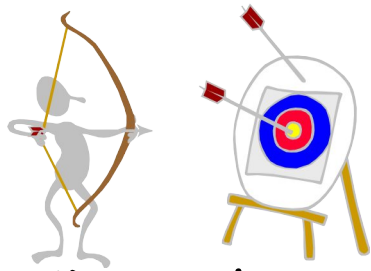
ccTGA





F.R. 34 yrs

Valvular and subvalvular Stenosis s/p Surgery



# Issues for the use of PMK and SICD in ACHD

SICD in the presence of unipolar pacing has been relatively contraindicated.

- ✓ Artefacts from unipolar pacing could interfere with appropriate detection of VT/VF by the S-ICD
- ✓ Risk of the S-ICD under-sensing true VF due to inappropriate pacing
- ✓ Inappropriate shock from double counting
- ✓ Test SICD during ventricular pacing with maximal output

# Take-home message

## PMK and S-ICD in ACHD

- S-ICD can be used safely in ACHD with a permanent PMK.
- There are important issues with regards to testing and programming that need to be addressed at the time of implantation.
- Patients should undergo the same screening as non-paced patients.
- Interference between the devices should be evaluated.
- Pacing spikes could be counted independently from the R waves by the S-ICD.
- Post-shock pacing from the S-ICD could inhibit pacing from the pacemaker and should be turned off.

## The Italian subcutaneous implantable cardioverter-defibrillator why not?

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Cristiano G. Boriani<sup>16</sup>, Mariolina Lovecchio<sup>17</sup>, Sergio Valsecchi<sup>17</sup>,  
Antonio G. Di Biase<sup>18</sup>; on behalf of 'AIAC S-ICD Why Not' Survey Investigators

S-ICD in ACHD with a PMK,  
why not?

# Take-home message

## PMK and SICD in ACHD

- For PMK-dependent patients, the conditional zone for SVT discrimination is not necessary.
- For pts who have tachy-brady synd., screening should be done during pacing and native rhythm and both should pass in at least 1 vector.
- Some PMKs have safety features that convert pacing to unipolar in the event of abnormal lead impedance or reversion to a back-up safety mode. This should be turned off, if possible, or the device tested with the leads programmed to unipolar pacing.