



## ICD Use in CPVT



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# Korean Heart Rhythm Society

## COI Disclosure

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The authors have no financial conflicts of interest to disclose concerning the presentation



# Disclosure

## Relationships with commercial interests:

- Grants/Research Support: No
- Speakers Bureau/Honoraria: No
- Consulting Fees: No
- Other: No



# CASE 1

F 13yr

Frequent syncope

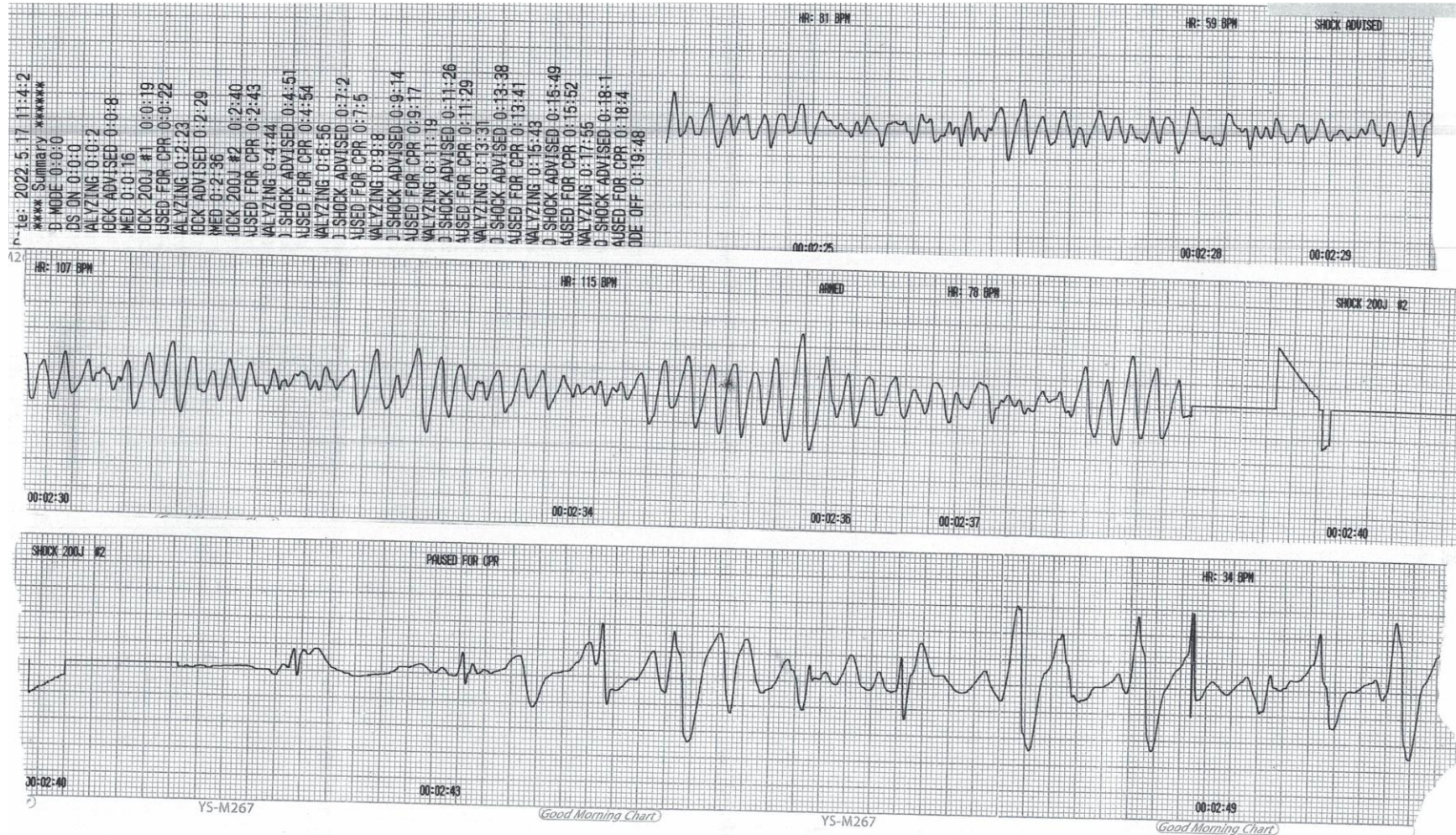
EEG brain MRI at local hospital

Seizure?

Anticonvulsant medication

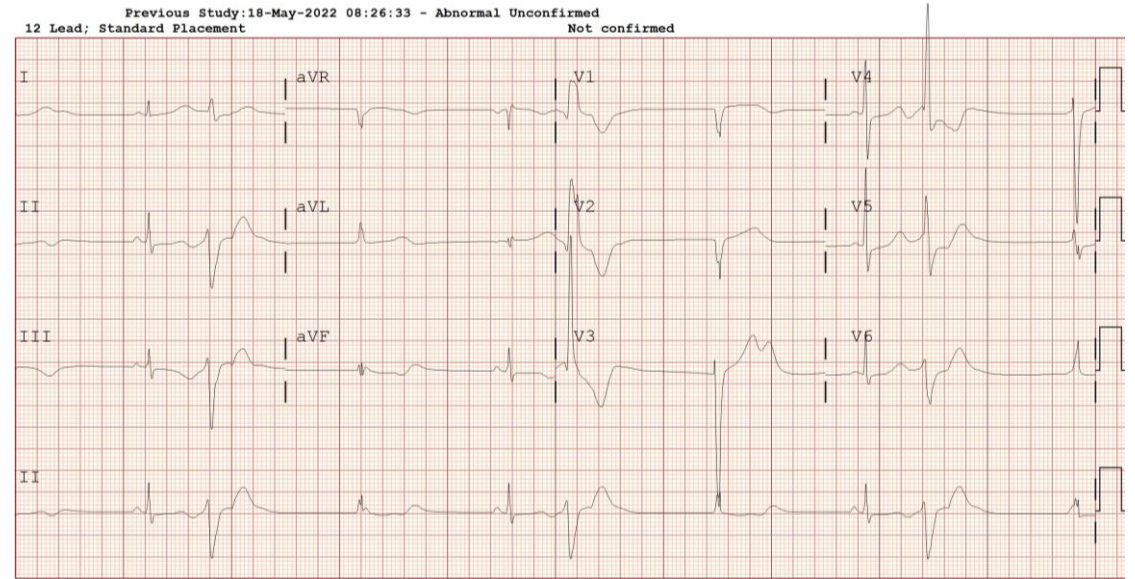


# at School, Seizure, call 119





# Post-shock



# CASE 2

F 12yr

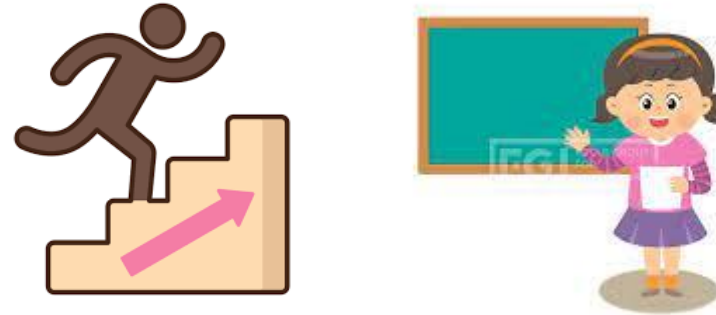
2018 Frequent syncope

HUTT, EEG at KNUCH

Vasovagal syncope? Seizure?

Anticonvulsant medication

2021 Abnormal ECG

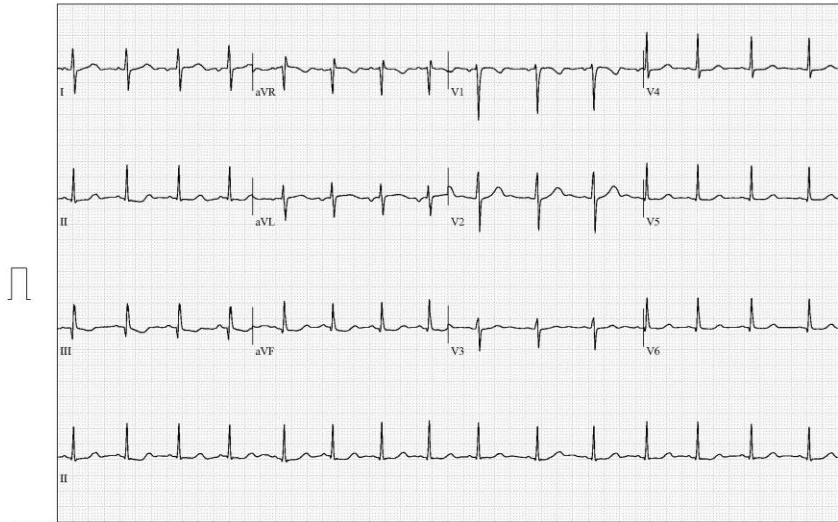




# Exercise test supine

Lee seon a.  
Patient ID: 7156789  
2022/08/09 Female 162 cm 54 kg  
10:34:31 11 yrs Asian

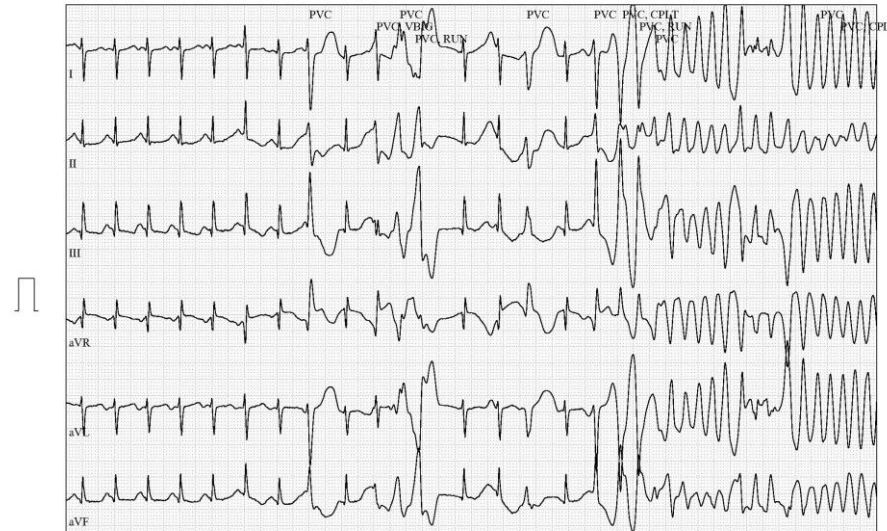
Exercise Test / ECG Strips  
86 bpm  
PRETEST SUPINE  
0:01  
BRUCE  
KNUH



GE CASE V7.0 (10)  
25 mm/s 10 mm/mV 60 Hz 0.04Hz FRF HEART V5.41.1 HR(V2,V1) Unconfirmed  
Attending MD: Kim yeo hyang Page 8

Lee seon a.  
Patient ID: 7156789  
2022/08/09 Female 162 cm 54 kg  
10:37:11 11 yrs Asian

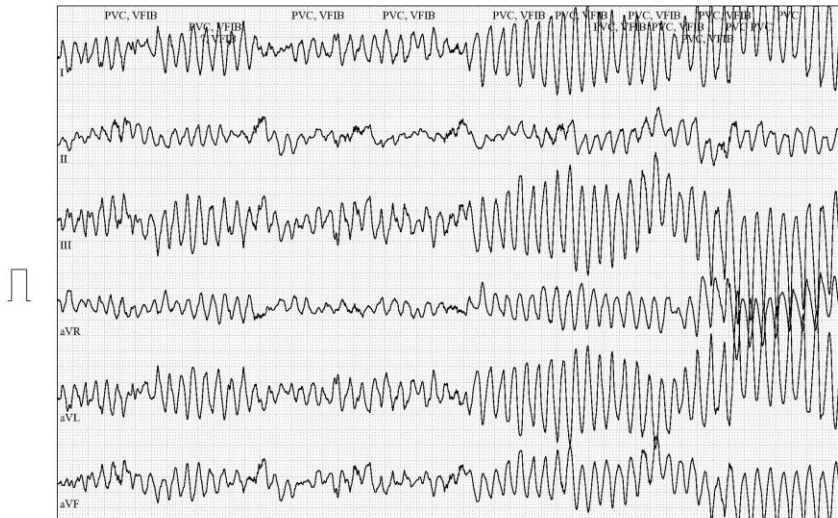
Exercise Test / Arrhythmia Review  
142 bpm  
PRETEST SUPINE  
2:40  
BRUCE  
0.0 km/h  
0.0 %  
KNUH



GE CASE V7.0 (10)  
25 mm/s 10 mm/mV 60 Hz 0.04Hz FRF HEART V5.41.1 HR(V2,V1) Unconfirmed  
Attending MD: Kim yeo hyang Page 1

Lee seon a.  
Patient ID: 7156789  
2022/08/09 Female 162 cm 54 kg  
10:37:39 11 yrs Asian

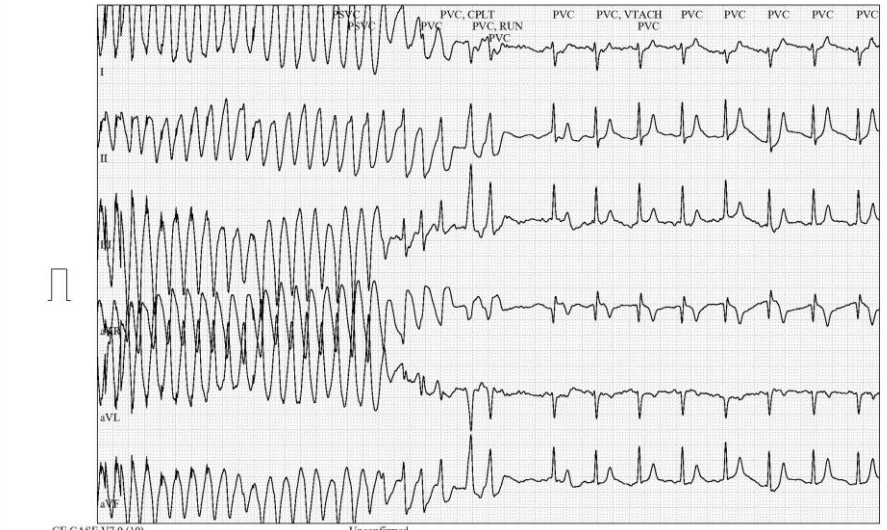
Exercise Test / Arrhythmia Review  
203 bpm  
120/80 mmHg  
PRETEST SUPINE  
3:09  
BRUCE  
0.0 km/h  
0.0 %  
KNUH



GE CASE V7.0 (10)  
25 mm/s 10 mm/mV 60 Hz 0.04Hz FRF HEART V5.41.1 HR(V6,V2) Unconfirmed  
Attending MD: Kim yeo hyang Page 1

Lee seon a.  
Patient ID: 7156789  
2022/08/09 Female 162 cm 54 kg  
10:37:56 11 yrs Asian

Exercise Test / Arrhythmia Review  
157 bpm  
120/80 mmHg  
PRETEST SUPINE  
3:26  
BRUCE  
0.0 km/h  
0.0 %  
KNUH



GE CASE V7.0 (10)  
25 mm/s 10 mm/mV 60 Hz 0.04Hz FRF HEART V5.41.1 HR(V6,V2) Unconfirmed  
Attending MD: Kim yeo hyang Page 1





Lee seon a.  
Patient ID: 7156789  
2022/08/09 10:38:31  
Female 162 cm 54 kg  
11 yrs Asian

Exercise Test / Arrhythmia Review

169 bpm  
120/80 mmHg

PRETEST  
SUPINE  
4:01

BRUCE  
0.0 km/h  
0.0 %

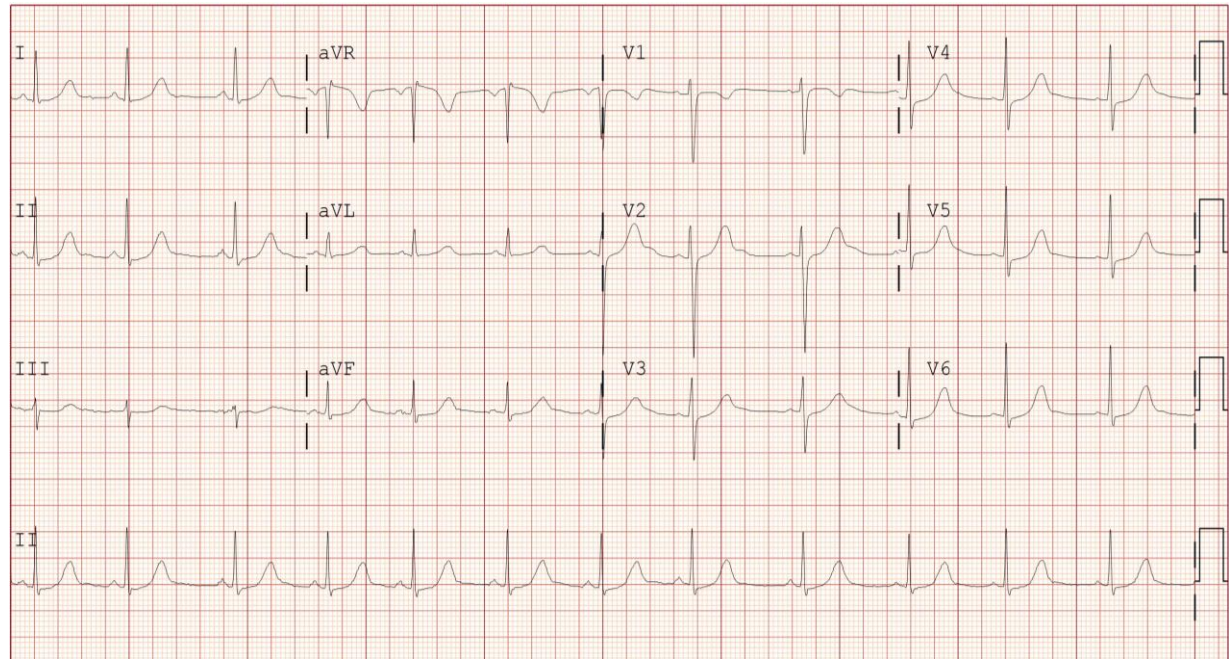
KNUH



GE CASE V7.0 (10)  
25 mm/s 10 mm/mV 60 Hz 0.04Hz FRF HEART V5.41.1 HR(LV2) Unconfirmed

Attending MD: Kim yeo hyang

Page 1



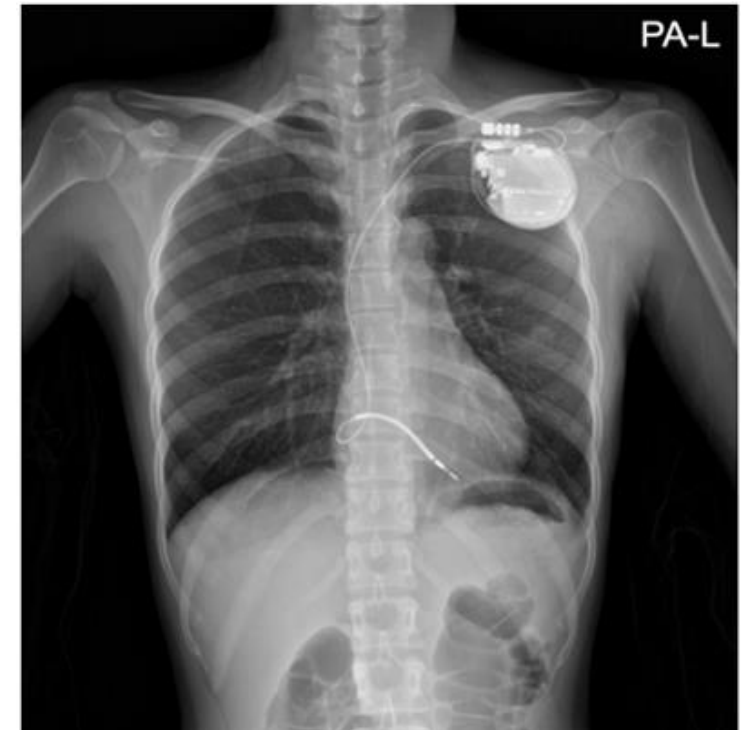
# Diagnosis

- Syncope on exercise
- Seizure
- Cardiac arrest
  
- ECG
- Exercise test
  
- Gene study, RYR2 gene mutation



**CPVT**

# Treatment





# ICD indication

- ESC guidelines and the HRS expert consensus statement
- diagnosis and management of patients with inherited primary arrhythmia syndromes
- ICD implantation in CPVT patients who experienced
  - cardiac arrest,
  - recurrent syncope,
  - polymorphic/bidirectional VT despite optimal medical therapy and/or left cardiac sympathetic denervation (LCSD).
- higher risk of future arrhythmic episodes, marker of adverse outcomes
- ICD as a stand-alone therapy in an asymptomatic patient with CPVT is contraindicated due to the risk of adrenergic storms.



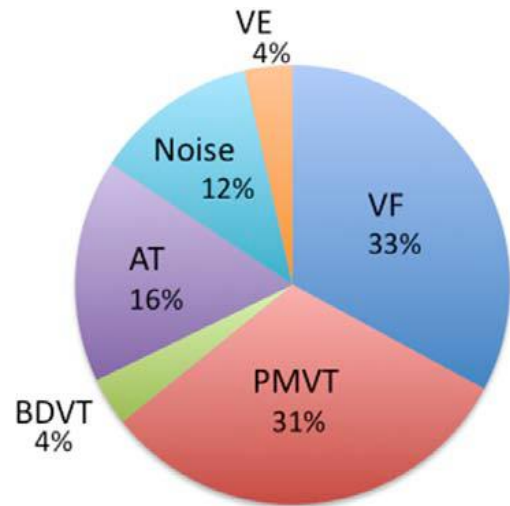
# Real-world data

- the nature of CPVT and its potentially malignant course even with optimal medical therapy
- implanting an ICD seems reasonable
- towards a more liberal use of ICDs in CPVT patients
  
- ICDs are not without their problems.

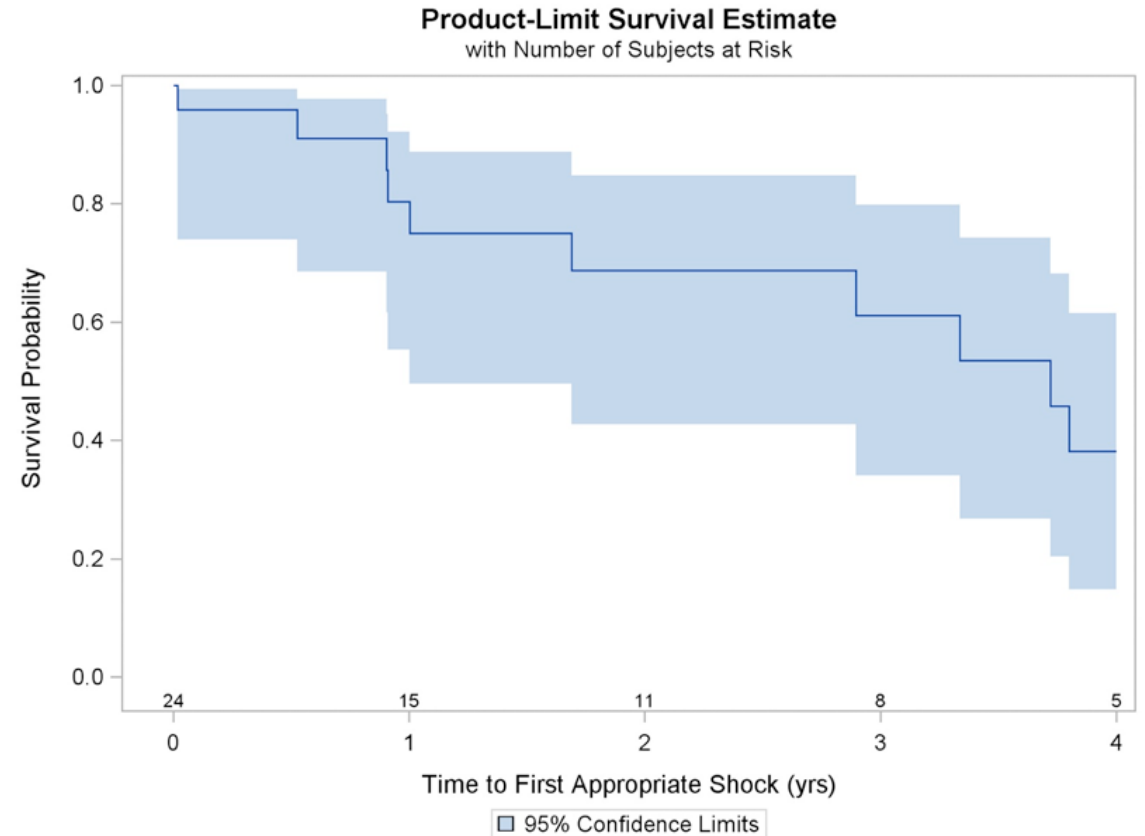


# ICD application for CPVT pros and cons

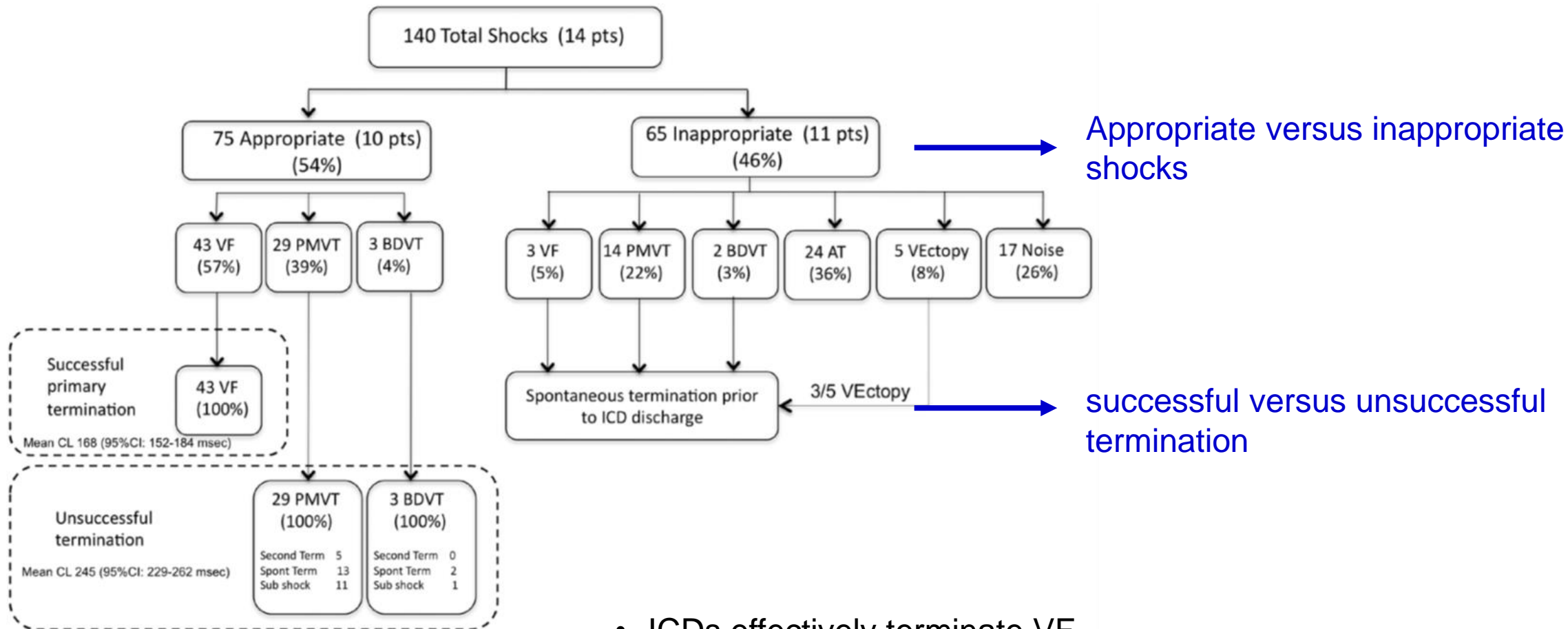
- ICDs might be proarrhythmic in CPVT.
- systematic review and meta-analysis



Arrhythmia mechanisms resulting in ICD discharge



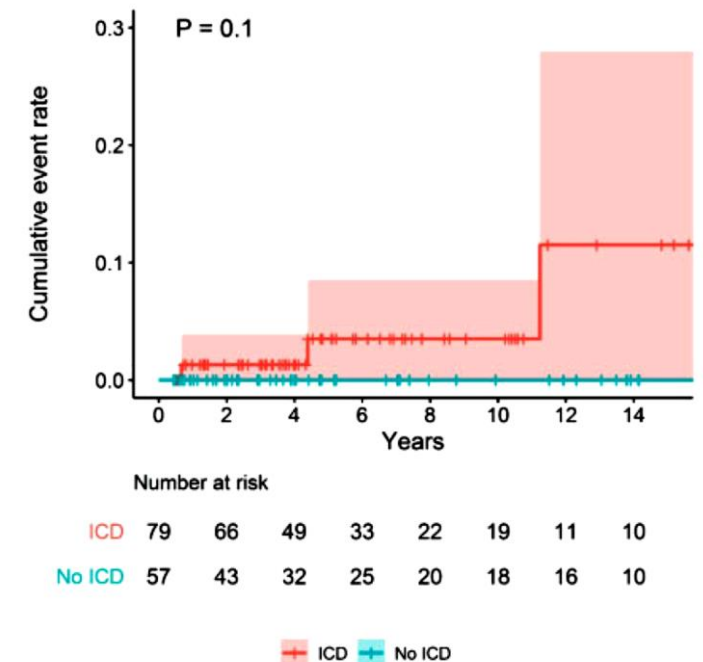




- ICDs effectively terminate VF
- not as effective in terminating VT.
- high rate of ineffective appropriate discharges
- high rate of inappropriate discharges
- Shocks for VT, monomorphic or polymorphic, were ineffective

# ICD application for CPVT pros and cons

- 136 patients who presented with SCD in whom CPVT was diagnosed, observational data
- initiation of guideline-directed therapy with  $\beta$ -blocker and flecainide with or without LCSD
- 58% of the patients had an ICD implanted
- a median follow-up of 4.8 years
- a composite outcome of **SCD, sudden cardiac arrest, syncope**  
**47% with ICD > 15.8% without ICD**
- 24% inappropriate ICD shocks
- 29% other device-related complications
- a **lack of survival benefit in patients with ICDs**



# ICD application for CPVT pros and cons

- prospective cohort study followed up 216 patients with CPVT on  $\beta$ -blockers only
- mean duration of 7.5 years
- 28 (13%) life-threatening arrhythmic event
- 18 had an **ICD already implanted** prior to the event, with subsequent **zero mortality**
- 4 of **the other 10 (40%) died**



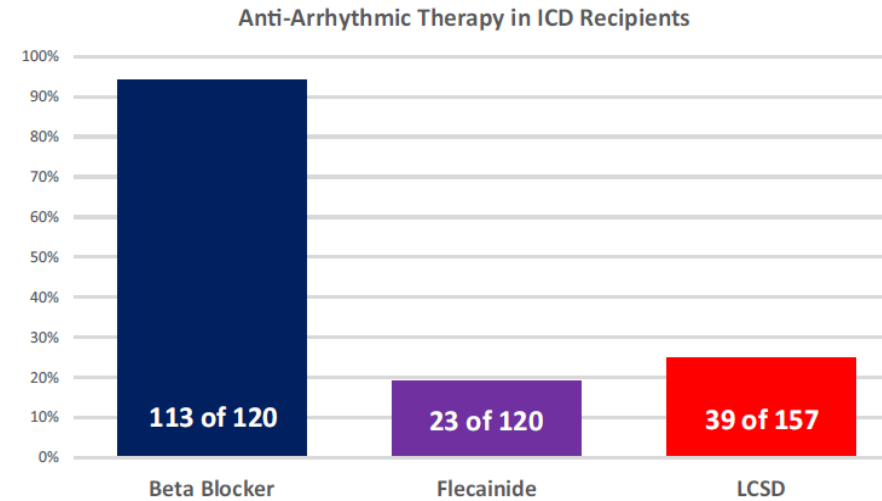


# ICD programming

- the absence of randomised trials
- recommend a conservative approach towards using ICDs in line with the current guidelines
- ICD is implanted following a **shared decision-making process**
- the treating physician should seek to minimise the risk of appropriate and inappropriate therapy by optimising medical treatment with or without LCSD
- Supraventricular tachycardias are common in CPVT patients
- extended monitoring period could also help to minimise these risks, aiming to treat only high-rate arrhythmias, such as VF.



# Shock appropriateness and efficacy






**Table 3** Summary of the 2 studies reporting shock appropriateness and efficacy by inciting arrhythmia

Study	No. of patients	Median duration of follow-up (y)	No. of patients with shocks (% of total patients)	No. of appropriate shocks (% of total shocks)	No. of appropriate shocks for VT (% of successful shocks)	No. of appropriate shocks for VF (% of successful shocks)	No. of inappropriate shocks (% of total shocks)	No. of inappropriate shocks for atrial arrhythmias (% of inappropriate shocks)	No. of inappropriate shocks for self-resolved ventricular arrhythmia (% of inappropriate shocks)	No. of patients with both inappropriate and appropriate shocks	No. of inappropriate shocks for noise/T-wave oversensing (% of inappropriate shocks)
Roses-Noguer et al <sup>18</sup>	13	4.0 (IQR, 1.7–19.9)	10 (77)	63 (72)	40 (3)	23 (79)	24 (28)	16 (67)	1 (4)	NR	7 (29)
Miyake et al <sup>5</sup>	24	3.3 (IQR, 1.1–5.8)	14 (58)	75 (54)	32 (0)	43 (100)	65 (46)	24 (36)	22 (34)	7 (29)	17 (26)
Total	37	4.0 (IQR, 2.4–7.1)	24 (65)	138 (61)	72 (1)	66 (94)	89 (39)	40 (45)	23 (26)		24 (27)

IQR = interquartile range; NR = not reported; VF = ventricular fibrillation; VT = ventricular tachycardia.



# CPVT in KNUCH

sex	Age on diagnosis	Syncope	Neurology f/u	VT/VF	drug	Sympa- thectomy	Re-attack	Exercise PVC
F	12	o	o	o 	Nadolol+flecainide	o	x	x
F	15	o	x	o 	Concor+flecainide	o	o	o
F	12	o	o	o	Nadolol+flecainide	o	x	o
F	13	o	o	o 	Nadolol+flecainide	o	x	o
M	9	o	x	x	flecainide	x	-	x





## Guideline HRS/EHRA/APHRS Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes (2014)




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**“I have only one ICD case for CPVT”**





# Who needs ICD ?

sex	Age on diagnosis	Recurrent Syncope	Neurology f/u	VT/VF	drug	Sympa- thectomy	Re-attack	Exercise PVC
F	12	o	o	o 	Nadolol+flecainide	o	x	x
F	15	o	x	o 	Concor+flecainide	o	o	o
F	12	o	o	o	Nadolol+flecainide	o	x	o
F	13	o	o	o 	Nadolol+flecainide	o	x	o
M	9	o	x	x	flecainide	x	-	x



# Take Home Message

- Recurrent syncope, cardiogenic or neurologic
- Refractory seizure
- ECG / exercise test
- Medical treatment / sympathectomy
- ICD application based on guideline

