Catecholaminergic Polymorphic VT When do you implant an ICD and how to follow-up?





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#### Lecture Title:

Catecholaminergic Polymorphic VT. When do you implant an ICD and how to follow-up?

Speaker Name: Thomas Kriebel, MD, FEHRA

The following relationships exist related to this presentation: NONE

Off label use of products will not be discussed in this presentation. Will be discussed





### Natural Course of Patients with symptomatic CPVT De Ferrari GM et al., Circulation 2015



- Median age at onset of symptoms 8.5 y
- At age of 15 y 96% major cardiac events (21 syncopes, 33 aborted sudden cardiac deaths or appropriate ICD discharges)





## **Current Indications for ICD-Implantation in CPVT**

urmal of the American College of Cardiology 2013 by the American College of Cardiology Foundation, the American Heart Association, Inc., di the Heart Rhythm Society ublished by Elsevier Inc.

PRACTICE GUIDELINE

#### 2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities

A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society

> Developed in Collaboration With the American Association for Thoracic Surgery and Society of Thoracic Surgeons

2008 WRITING COMMITTEE MEMBERS Andrew E. Epstein, MD, FACC, FAHA, FHRS, *Chair*, John P. DiMarco, MD, PHD, FACC, FHRS; Kenneth A. Ellenbogen. MD, FACC, FAHA, FHRS; N.A. Mark Eates III, MD, FACC, FAHA, FHRS; Roger A. Freedman, MD, FACC, FHRS; Leonard S. Gettes, MD, FACC, FAHA; A. Marc Gillinov, MD, FACC, FAHS; Gabriel Gregoratos, MD, FACC, FAHA; Stephen C. Hammill, MD, FACC, FAHS; David L. Hayes, MD, FACC, FAHA, FHRS; Mark A. Hlatky, MD, FACC, FAHA; L. Kristin Newby, MD, FACC, FAHA;

#### HRS/EHRA/APHRS Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes Heart Rhythm 2013

Silvia G. Priori, MD, PhD, (HRS Chairperson)<sup>1</sup>, Arthur A. Wilde, MD, PhD, (EHRA Chairperson)<sup>2</sup>, Minoru Horie, MD, PhD, (APHRS Chairperson)<sup>3</sup>, Yongkeun Cho, MD, PhD, (APHRS Chairperson)<sup>4</sup>, Elijah R. Behr, MA, MBBS, MD, FRCP<sup>5</sup>, Charles Berul, MD, FHRS, CCDS<sup>6</sup>, Nico Blom, MD, PhD<sup>7,\*</sup>, Josep Brugada, MD, PhD<sup>8</sup>, Chern-En Chiang, MD, PhD<sup>9</sup>, Heikki Huikuri, MD<sup>10</sup>, Prince Kannankeril, MD<sup>11,‡</sup>, Andrew Krahn, MD, FHRS<sup>12</sup>, Antoine Leenhardt, MD<sup>13</sup>, Arthur Moss, MD<sup>14</sup>, Peter J. Schwartz, MD<sup>15</sup>, Wataru Shimizu, MD, PhD<sup>16</sup>, Gordon Tomaselli, MD, FHRS<sup>17,†</sup>, Cynthia Tracy, MD<sup>18,%</sup>



**ESC GUIDELINES** 

#### 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC)



Prioir S et al.



### Class I

• ICD implantation in addition to beta-blockers with or without Flecainide is recommended in patients with CPVT who experienced cardiac arrest, recurrent syncopes (2012 Class IIa) or polymorphic bidirectional VT despite optimal therapy (Level C)

Class III

 ICD as a stand-alone therapy is not indicated in an asymptomatic patient with a diagnosis of CPVT





### Current Indications for ICD-Implantation in CPVT II Additional treatment

### Class IIa

• Flecainide should be considered in addition to betablockers in patients with a diagnosis of CPVT and carriers of an ICD to reduce appropriate ICD shocks

### **Class IIb**

• Left cardiac sympathetic denervation may be considered in patients with a diagnosis of CPVT who experience recurrent time tes, polymorphic/ bidirectional VT or several propriate ICD shocks while on beta-blocker (+Fi cainide) and in patients who are intolerant or have contraindications to betablockers





### **After ICD Implantation**







# **Case Presentation**

### 8-year-old ♂, no family history of SCD, recurrent exercise-induced syncopal episodes 08/04 Holter recordings





### **Treadmill Test** Bidirectional VT, HR 204 bpm



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## **Clinical Course I**

- Diagnosis: CPVT (Genetics negativ)
- Antiarrhythmic therapy with propranolol 5 mg/kg
- ICD implantation due to ongoing polymorphic VT 06/2005
- ICD programming: VT monitor < 330 ms (180 bpm), VF zone < 240 ms (250 bpm) Counter 18/24 for NID and 9/12 for RNID





### Clinical Course II Dec 2005

- Hair loss → pt stopped betablockers
- During exercise recurrent ICD discharges due to non-sustained polymorphic VT
- Back to beta-blockers

ID#	Date/Time	V. Cycle	Duration	Reason
234	Nov 28 19:41:51	290 ms	7 beats	Non-Sustained
233	Nov 28 19:41:43	290 ms	9 beats	Non-Sustained
232	Nov 28 19:41:39	300 ms	9 beats	Non-Sustained
231	Nov 28 19:41:34	310 ms	5 beats	Non-Sustained
230	Nov 19 12:41:53	300 ms	8 beats	Non-Sustained
229	Nov 19 12:41:38	300 ms	5 beats	Non-Sustained
228	Nov 19 12:41:35	280 ms	6 beats	Non-Sustained
227	Nov 19 12:41:24	260 ms	7 beats	Non-Sustained
226	Nov 14 19:55:23	270 ms	7 beats	Non-Sustained
225	Nov 11 09:32:49	260 ms	8 beats	Non-Sustained
224	Nov 05 12:03:00	290 ms	6 beats	Non-Sustained
223	Nov 05 11:59:17	250 ms	19 sec	Wavelet
222	Nov 05 11:59:12	260 ms	5 beats	Non-Sustained
221	Nov 05 11:59:04	240 ms	16 beats	Non-Sustained
220	Nov 05 11:58:55	290 ms	6 beats	Non-Sustained
219	Nov 05 11:58:43	210 ms	18 beats	Non-Sustained
218	Nov 05 11:58:32	250 ms	5 beats	Non-Sustained
217	Nov 05 11:58:30	280 ms	5 beats	Non-Sustained
216	Nov 05 11:58:11	250 ms	5 beats	Non-Sustained
215	Nov 05 11:58:07	280 ms	9 beats	Non-Sustained
214	Nov 05 11:57:54	270 ms	11 beats	Non-Sustained
213	Nov 05 11:57:45	310 ms	7 beats	Non-Sustained
212	Oct 22 11:10:23	260 ms	7 beats	Non-Sustained
211	Oct 22 11:10:21	260 ms	7 beats	Non-Sustained
210	Oct 14 15:18:38	300 ms	9 beats	Non-Sustained
209	Oct 14 15:13:06	230 ms	18 beats	Non-Sustained
208	Oct 14 09:08:39	250 ms	12 beats	Non-Sustained
207	Oct 14 09:08:20	240 ms	5 beats	Non-Sustained
206	Oct 14 08:47:21	240 ms	6 beats	Non-Sustained
205	Oct 14 08:47:13	320 ms	5 beats	Non-Sustained
204	Oct 13 07:59:32	200 ms	10 beats	Non-Sustained
203	Oct 13 07:59:28	290 ms	10 beats	Non-Sustained
202	Oct 13 07:59:07	300 ms	7 beats	Non-Sustained
201	Oct 13 07:59:06	290 ms	5 beats	Non-Sustained
200	Oct 13 07:58:59	300 ms	5 beats	Non-Sustained
199	Oct 13 07:58:58	290 ms	5 beats	Non-Sustained
198	Oct 13 07:58:42	250 ms	15 beats	Non-Sustained
197	Sep 28 14:49:15	280 ms	5 beats	Non-Sustained
196	Sep 26 12:01:03	240 ms	5 beats	Non-Sustained





### Clinical Course III Dec 2005







## **Clinical Course IV**



### 3rd ICD discharge induced self-limited VF Reprogramming of ICD: counter increase to 30/40 for NID and 18/24 for RNID





### Sudden Cardiac Death Despite an ICD in a Young Female with Catecholaminergic VT Mohamed U et al., Heart Rhythm 2006

ID#	Date/Time	Туре	V. Cycle	Last Rx	Success	Duration
7	Jan 11 09:15:58	VF	130 ms	VF Rx 6	No	14 min







### Modification of the ICD Software October 2006; VF zone < 200 ms (300 bpm)

Parameters - Detection									
	Enable	Initial	Redetect	V Interval (Rate)	)				
VF	On	30/40	18/24	200 ms	0				
FVT	Off				0				
VT	Monitor	20	8	330 ms (182 bpm)		330 ms			
						SVT Limit <b>≭ 240 m</b> s			
	Detection	n Enhancem	ents			Sensitivity			
Wave	let		On	]		Ventricular 0.3 mV			
SVT Limit 240 I			240 ms						
Stability Off			Off						
<mark>Onset</mark>			Monitor						
Addit	ional Settings	·							



Herz Zentrum Göttinge

### Clinical Course VI December 2006







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### Clinical Course VI December 2006





Zentrum

Göttir



### Sympathectomy May 2008







## **Clinical Course After Sympathectomy**

Nov 07 – Nov 08







### Outcome of ICD Therapy in CPVT Roses-Noguer F et al., Heart Rhythm 2014

- > 13 pts with CPVT after ICD implantation
- > Median age: 15 y
- Indications: 7/13 Cardiac arrest

6/13 Syncope despite AA treatment

> 10/13 pts. with 96 shocks:







### Outcome of ICD Therapy in CPVT Roses-Noguer F et al., Heart Rhythm 2014

- > 87/96 shocks reviewed:
  63/87 (72%) appropriate
  24/87 (28%) inappropriate (SVT, T-wave oversensing)
- Appropriate shocks:
  20/63 (32%) effective
  43/63 (68%) ineffective
- Shocks to triggered VT 1/40 effective Shocks due to VF 19/23 effective Antitachycardia pacing 2/12 effective





# Optimal ICD Programming in CPVT Patients

- > Target VFib
- Avoid shocks during triggered polymorphic or bidirectional VT
- Detection and redetection durations should be maximized
- Individual reprogramming by the manufacturer may be necessary





### Clinical Management of CPVT - Role of LCSD De Ferrari GM et al., Circulation 2015

- > N=63 pts (n=54 secondary + n=9 primary prevention)
- Pts with major cardiac events despite optimal AA treatment (n=38) reduced from 100% to 32%
- Pts with presurgical ICD: rate of shocks per person per y dropped from 3.6 to 0.6 shocks

Weitplaz-Gradit





# **Conclusions I**

- ICD shocks can be proarrhythmic in CPVT due to the pain and subsequent adrenaline surge
   Effectiveness of appropriate shocks in CPVT
- patients seems to be critically dependent on the underlying rhythm
- Specific algorithm to discriminate VF from polymorphic VT should be developed and incorporated in the ICD software





# **Conclusions II**

> Left cardiac sympathetic denervation should be always considered in patients with CPVT and recurrent ICD shocks despite optimal AA treatment > The indication class of left cardiac sympathetic denervation should be changed to class I in patients with recurrent symptoms despite optimal AA treatment and be considered prior or with ICD implantation

> Westphiz-Gradit



# Thank you very much for Your Attention!





Team of the Department of Pediatrics and Pediatric cardiology 2016/2017



Akademisches Lehrkrankenhaus de Universitäten Mainz und Heidelber