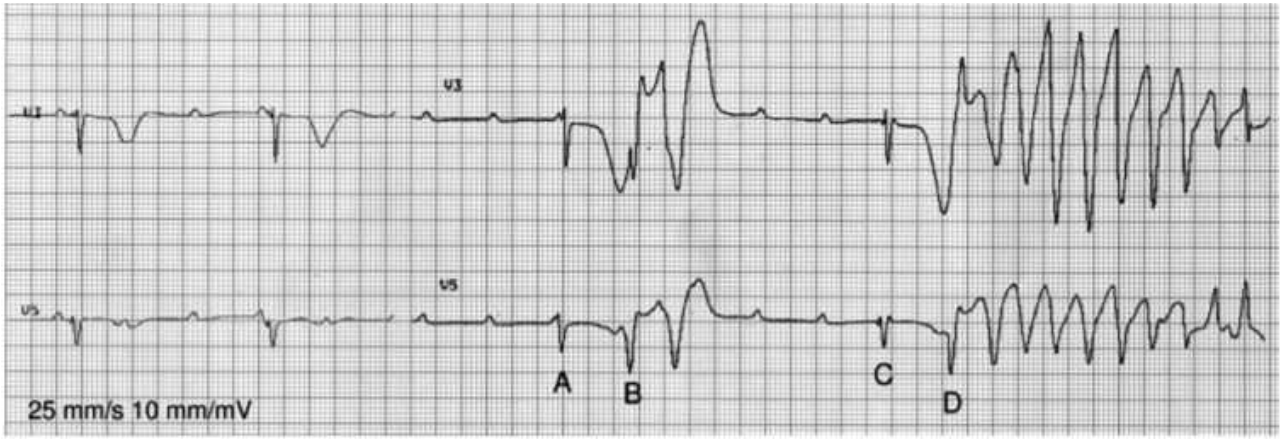
PEDIRHYTHM VII

ICD Programming in Genetic Arrhythmias



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FACULTY DISCLOSURE

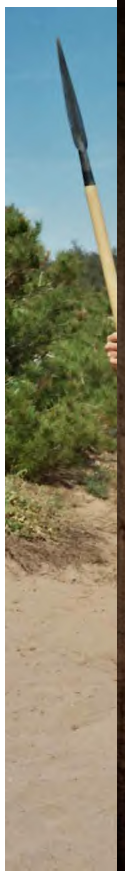
Lecture Title:
ICD Programming in Genetic Arrhythmias

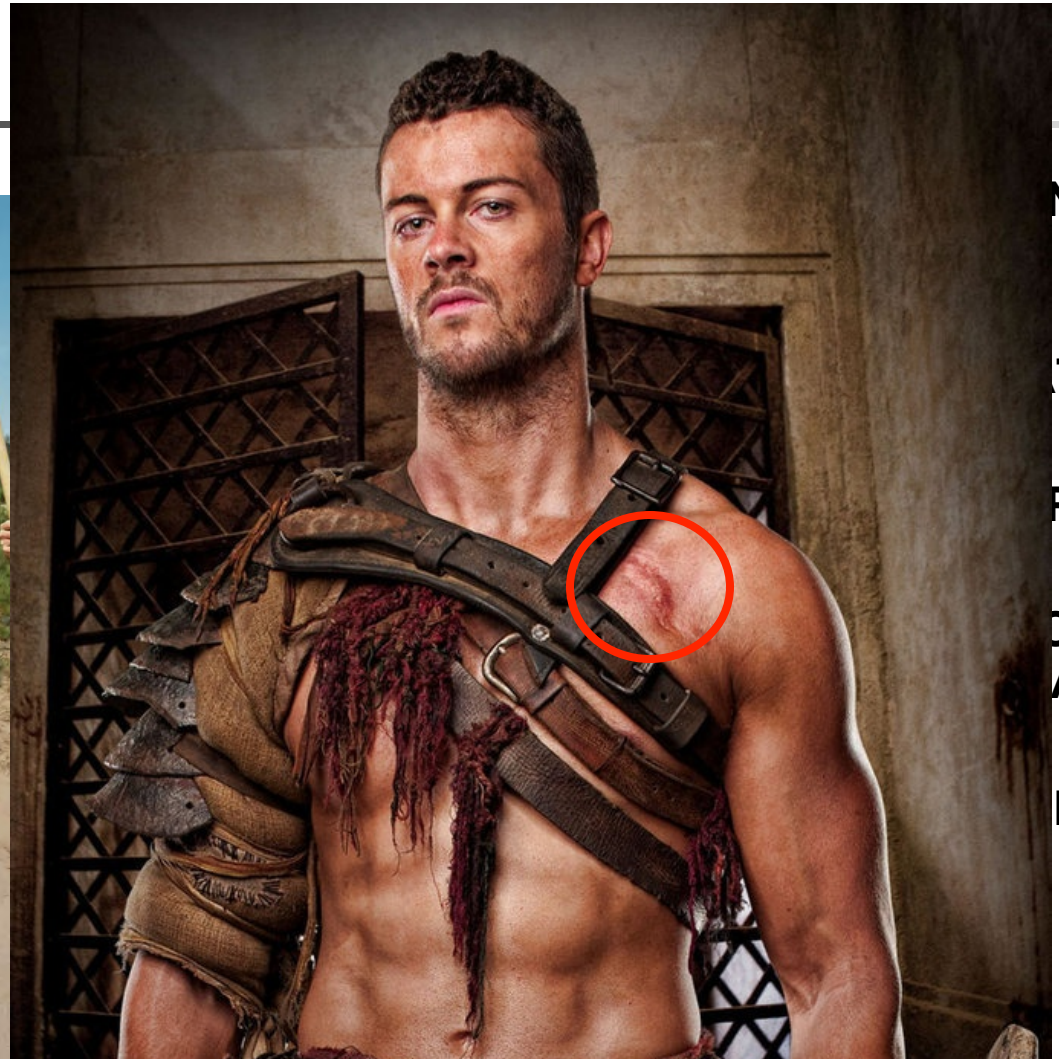
Speaker Name:
Peter P. Karpawich, MD

The following relationships exist related to this presentation:
NONE

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

SUDDEN DEATH IN THE YOUNG ATHLETE





NVASION OF

THON

PHILIPPEDES

OM THE
ATHENS

NQUER…”

But he would have survived if he had an ICD!!

ICD - Genetic Arrhythmias
Overview – Sudden Cardiac Death

- Sudden Cardiac Death < 40yo
  - 3/ 100,000
- 70% Primary arrhythmia
  - "Channelopathies"
- 30% Arrhythmogenic structural heart disease

ICD - Genetic Arrhythmias

SCD Incidence

- ADULT:
  - 350 - 450,000 / YEAR
  - 1 /1,000
  - 20% OF ALL DEATHS
- CHILD-ADOLESCENT:
  - 1 - 6 / 100,000
  - 8% OF ALL DEATHS
- 1:50-100,000 HIGH SCHOOL / COLLEGE ATHLETES
  - 60% ARE HIGH SCHOOL AGES

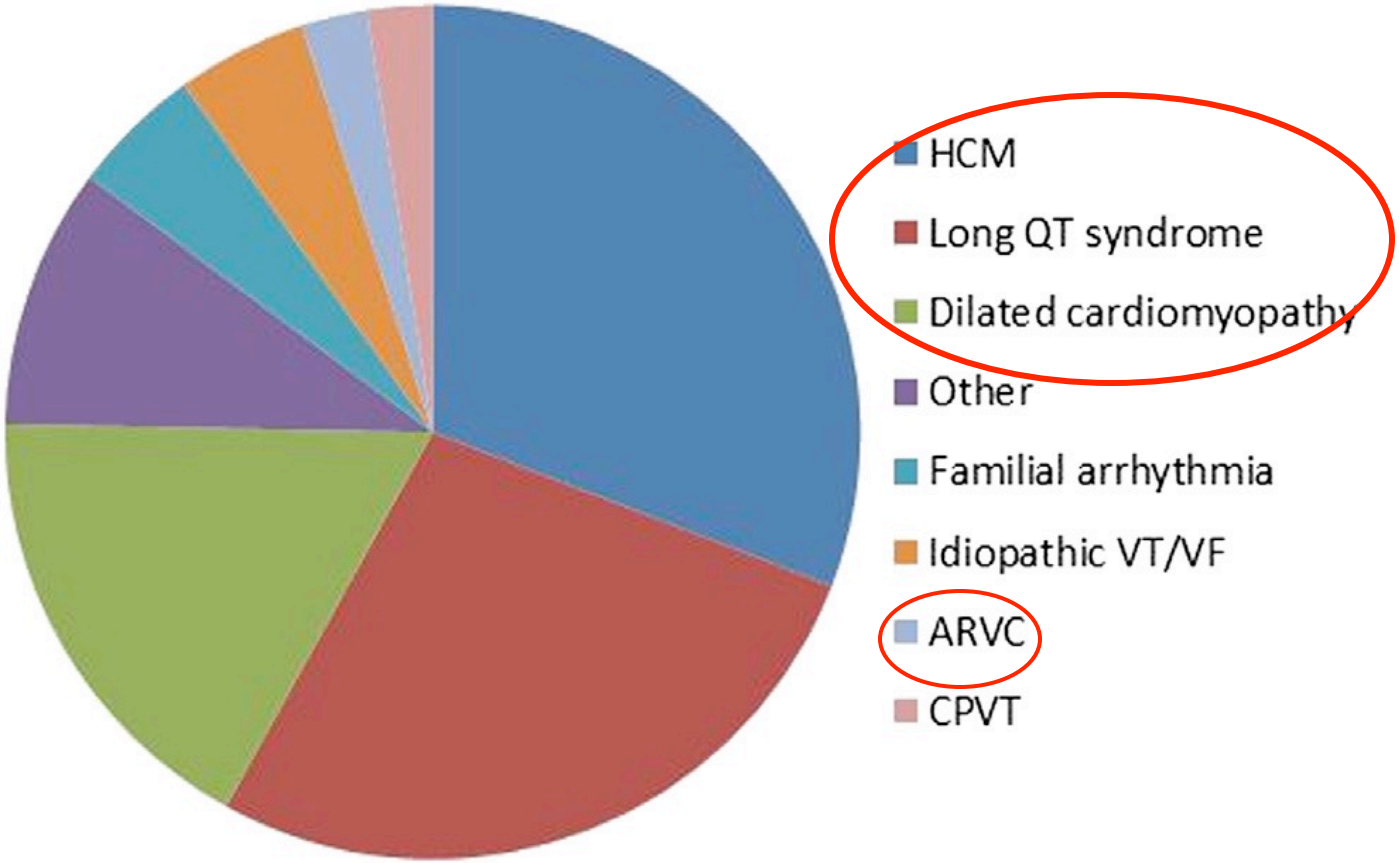
ICD - Genetic Arrhythmias
SCD Incidence

- AGE: 12 – 40y (MEDIAN 17y)
- GENDER: MALE 90%
- RACE: EQUAL
- SPORT (USA):
  - BASKETBALL 35%
  - FOOTBALL 33%
- WHEN:
  - TRAINING 58%
  - COMPETITION 32%

ICD - Genetic Arrhythmias
Syndromes

- **Channelopathies**
  - **Long QTc**
  - **Catecholaminergic Polymorh VT**
  - **Short QTc**
  - **Brugada**
- **Abnormal Structure**
  - **Hypertrophic Cardiomyopathy**
  - **Arrhythmogenic RV Dysplasia**
  - **Dilated Cardiomyopathy**
  - **Noncompaction**

ICD - Genetic Arrhythmias
Syndromes



Garnreiter. Heart Rhythm 2015

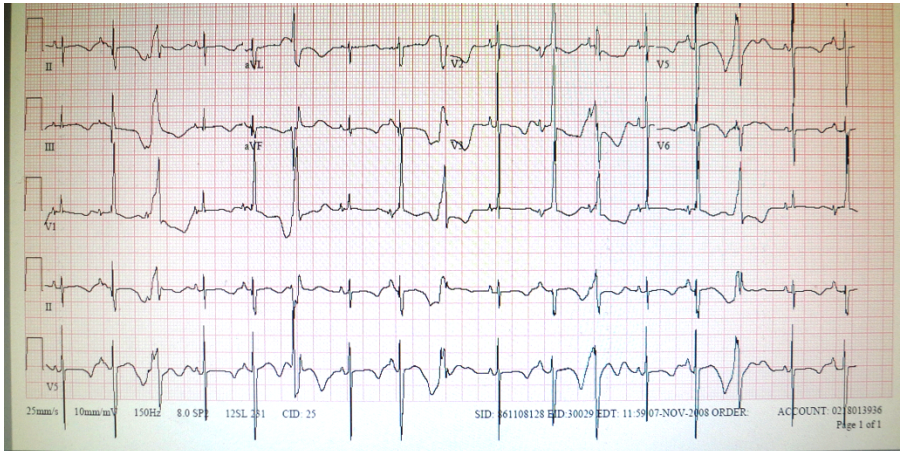
ICD - Genetic Arrhythmias
Long QTc

- Arrhythmogenic Syncope
  - Non-sustained VT, torsade
  - A fib, rapid AV conduction
  - AV block, Sinus arrest
  - Sympathetic surge
    - (exercise, stress, anger)
  - Sudden unexplained awakening at night

Need for pacemaker, ICD or both?

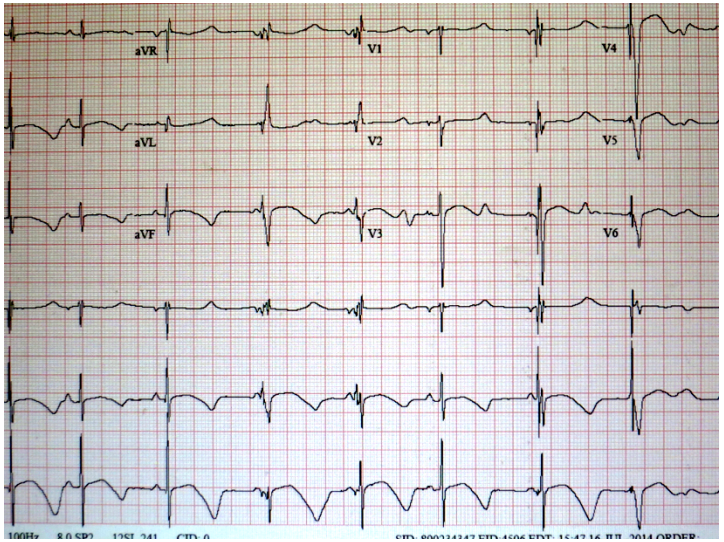
ICD - Genetic Arrhythmias
Long QTc – Case 1

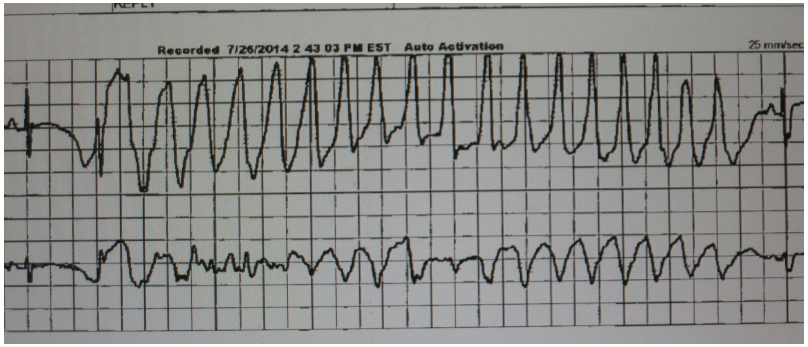
- Male, term NB
- In-utero 2o AVB
- QTc 550ms
- ? Long QT3
- Temp pacing at 1 day
- VT episodes
  - Rx: Propranolol and Mexiletine
- Permanent epicardial VVI pacemaker at 5 day
- Genetics: LQT 2 (HERG)
- Mexiletine discontinued



ICD - Genetic Arrhythmias
Long QTc - Case

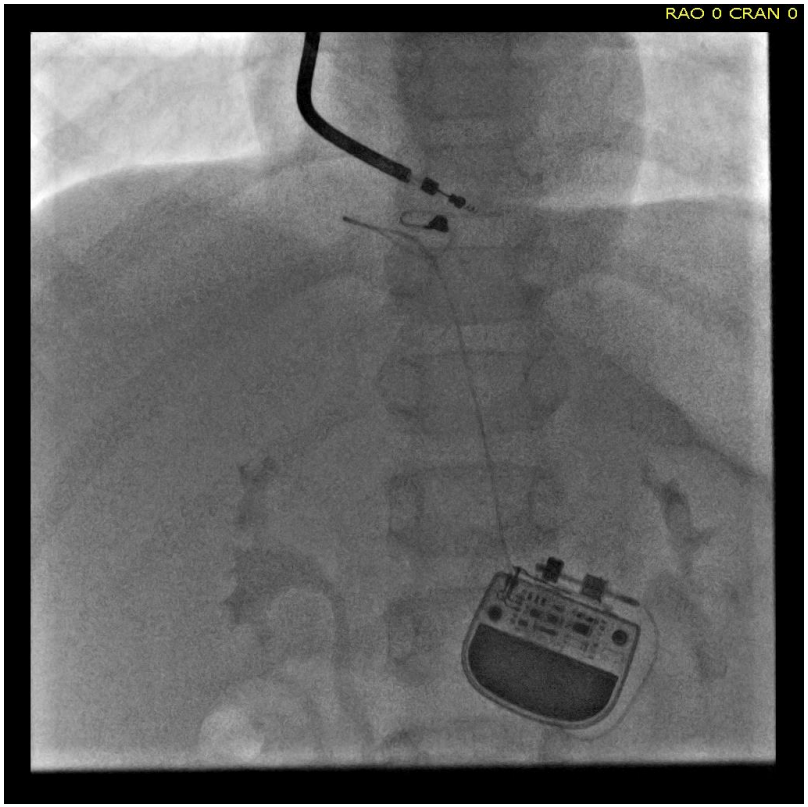
- Did well x 6 y
- Continued on β blocker
- Intermittent V pace
- No VT on stress testing or Holters
- 7yo Holter showed nonsust VT
  - Rate ~ 200bpm
- Switch to ICD

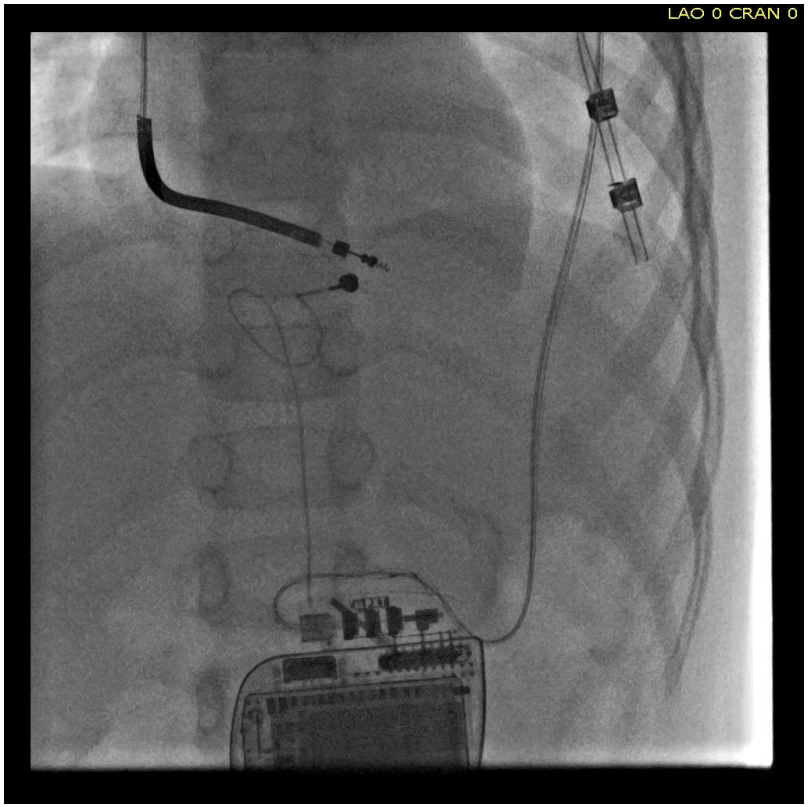




ICD - Genetic Arrhythmias

Long QTc - Case





ICD - Genetic Arrhythmias
Long QTc - Case

- Patient has both intermittent AV block and ventricular tachycardia
- Program both pacing and ICD modalities
- VT is somewhat stable
  - Initial overdrive pace instead of immediate shock

ICD - Genetic Arrhythmias
Long QTc - Case

Pacemaker Programming

Stevens, Jaylen
Device: Evera XT VR DVBB1D4
Serial Number: BWH214836H

Date of Visit: 03-Apr-2015 08:45:49
SW016 Software Version 1.0.1 (5.1)
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Initial Interrogation: Parameters — Page 1

Pacing Summary

Mode	Rates	
Mode VVI	Lower	60 bpm

Pacing Details	RV
Amplitude	3.50 V
Pulse Width	0.40 ms
Capture Management	Adaptive
Amplitude Margin	2.0 X
Min. Adapted Amplitude	2.00 V
Acute Phase Remaining	59 days
Sensitivity	0.30 mV
Pace Polarity	Bipolar
Sense Polarity	Bipolar

Refractory/Blanking	
V. Blank Post VP	200 ms
V. Blank Post VS	120 ms

Arrhythmia Interventions	
V. Rate Stabilization	Off

Stevens, Jaylen
Device: Evera XT VR DVBB1D4
Serial Number: BWH214836H

Date of Visit: 03-Apr-20
SW016 Software Versio
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Initial Interrogation: Parameters

Post Shock Pacing	
Post Shock Pacing*	
V. Amplitude	
V. Pulse Width	
* Settings for first 25 beats post shoc	
Post VT/VF Shock Pacing	

Additional Features	
Rate Hysteresis	Off
Sleep	Off

Device Information

Device	Medtronic	Evera XT VR DVBB1...	BWH214836H	Implanted
RV	Medtronic	6935M	TDL111661V	Implanted

Notes

long qt with bradycardia

ICD - Genetic Arrhythmias

Long QTc - Case

ICD programming

Stevens, Jaylen
Device: Evera XT VR DVBB1D4
Serial Number: BWH214836H

Date of Visit: 03-Apr-2015 08:45:49
SW016 Software Version 1.0.1 (5.1)
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Initial Interrogation: Parameters Page 3

VT/VF Detection

		V. Interval (Rate)	Initial	Redetect
VF	On	270 ms (222 bpm)	18/24	12/16
FVT	OFF			
VT	On	320 ms (188 bpm)	20	12
Monitor	Monitor	400 ms (150 bpm)	32	

[redacted] 270 ms
[redacted] 320 ms
No Rx [redacted] 400 ms

Wavelet		Other Enhancements		Sensitivity
Wavelet	On	Stability	Off	RV 0.30 mV
Template	03-Apr-2015	Onset	Off	
Match Threshold	70 %	High Rate Timeout		
Auto Collection	On	VF Zone Only	0.75 min	
SVT V. Limit	260 ms	All Zones	Off	
		TWave	On	
		RV Lead Noise	On+Timeout	
		Timeout	0.75 min	

Stevens, Jaylen
Device: Evera XT VR DVBB1D4
Serial Number: BWH214836H

Date of Visit: 03-Apr-201
SW016 Software Version
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Initial Interrogation: Parameters

VF Therapies	Rx1	Rx2	Rx3	Rx4	Rx5
VF Therapy Status	On	On	On	On	On
Energy	20 J	35 J	35 J	35 J	35 J
Pathway	B>AX	AX>B	B>AX	AX>B	B>AX

ATP During Charging
Deliver ATP if last 8 R-R >= 240 ms,Burst Pulses = 8, R-S1 = 88 %, Decrement = 10 ms
ChargeSaver = On(1 episodes), SmartMode = On

FVT Therapies	Rx1	Rx2	Rx3	Rx4	Rx5
FVT Therapy Status	Off	Off	Off	Off	Off

VT Therapies	Rx1	Rx2	Rx3	Rx4	Rx5
VT Therapy Status	On	On	On	On	On
Therapy Type	Burst	CV	CV	CV	CV
Energy		20 J	35 J	35 J	35 J
Pathway		B>AX	AX>B	B>AX	AX>B
Initial # Pulses	8				
R-S1 Interval=(%RR)	88 %				
S1S2(Ramp+)=(%RR)					
S2SN(Ramp+)=(%RR)					
Interval Dec	10 ms				
# Sequences	3				
Smart Mode	Off				

ICD - Genetic Arrhythmias
Long QTc - Case

ICD programming

Initial Interrogation: Parameters — Page 5

Shared V. ATP	
V-V Minimum ATP Interval	200 ms
V. Amplitude	8 V
V. Pulse Width	1.5 ms
V. Pace Blanking	240 ms

Shared V. Therapies	
Active Can/SVC Coil	Can+SVC On
Progressive Episode Therapies	Off
Confirmation+	On

Initial Interrogation: Parameters

Data Collection Setup

	Source	Range
LECG	Can to SVC	+/- 2 mV
EGM 1	RVtip to RVring	+/- 8 mV
EGM 2 (Wavelet)	Can to RVcoil	+/- 12 mV
EGM 3	RVtip to RVcoil	+/- 8 mV

Monitored	EGM1 and EGM2
Pre-arrhythmia EGM	Off
Device Date/Time	03-Apr-2015 08:46
Holter Telemetry	Off

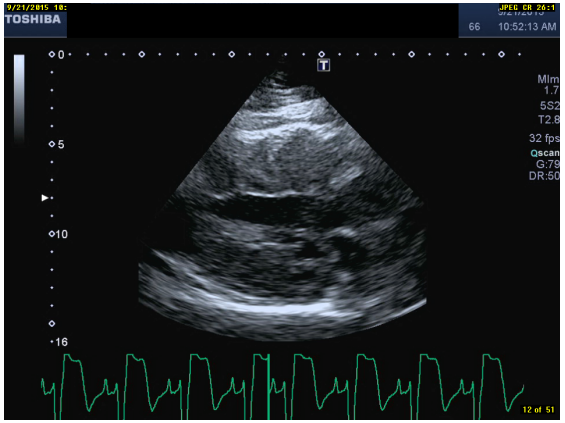
Medtronic CareAlert Setup

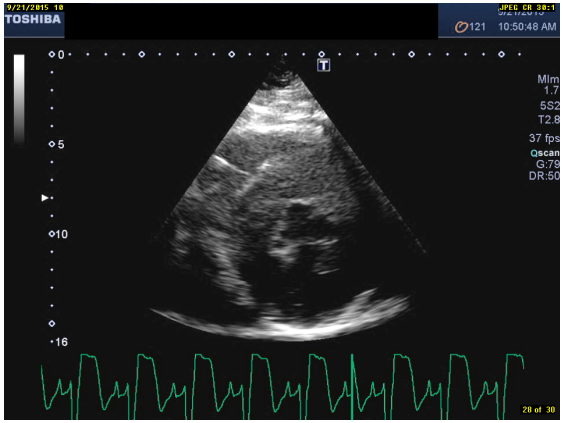
Patient Home Monitor	Yes
Alert Time	09:10

ICD - Genetic Arrhythmias

HCM – Case 2

- 30y, Female
- Hypertrophic cardiomyopathy
- 14yo Severe LVOT obstruction
  - LVOT resection
  - Surgical AV block
  - Post operative epicardial pacemaker (DDD)
- β blocker therapy
- Holter showed short runs of VT
- Switch to ICD
  - Implant ICD testing = immediate Vfib





ICD - Genetic Arrhythmias

HCM – Case 2

IMPLANTED DEVICES

PULSE GENERATOR Virtuoso II DR

Prod #	Serial	Manufacturer	Implant Date	Placement	Status
D274DRG	PZT200426H	Medtronic	03/04/2010	L Pect. SQ	Acute

LEADS AND ADAPTERS

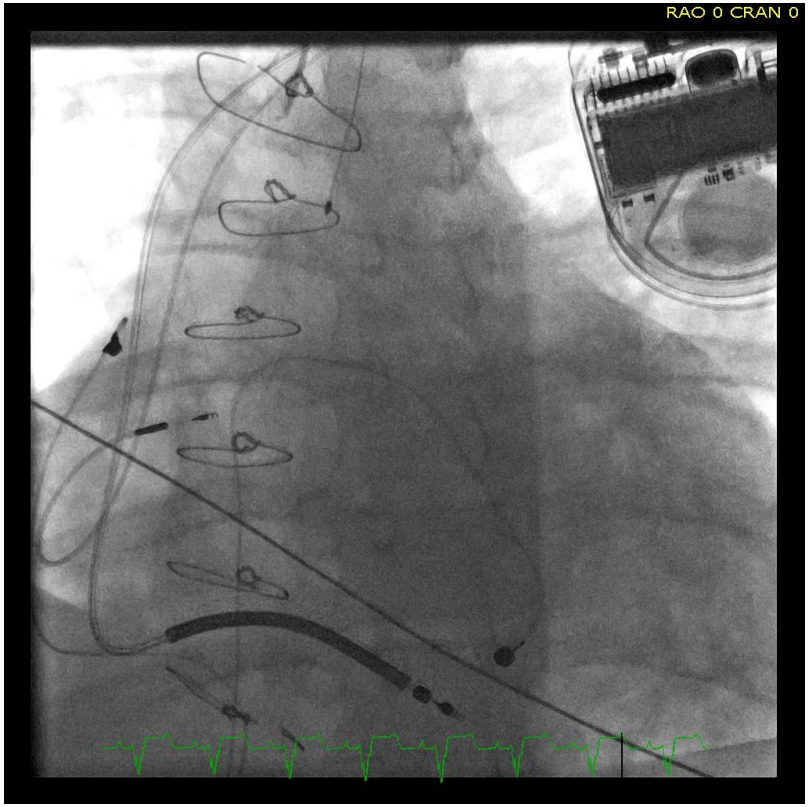
Prod #	Length	Serial Number	Manufacturer	Implant Date	Placement	Status	Chamber
3830	59cm	LFF055303V	Medtronic	03/04/2010	RA-Septum High	Acute	Right Atrium
6935	58cm	TAU010770V	Medtronic	03/04/2010	RV-Apex	Acute	Right Ventricle

BRADY PARAMETER SETTINGS

		Parameter	Atrial	RV	LV		
Pacing Mode:	DDDR	Amplitude (V):	3	3		Safety Pacing:	ON
Mode Switch:	ON	Pulse Width (ms):	0.4	0.4		PVC Response:	ON
Upper Tracking Rate (ppm):	150	Sensitivity (mV):	0.3	0.3		PMT Sensor:	OFF
Upper Activity Rate (ppm):	150	Blanking (mS):	150	120		Rate Response :	7
Lower Rate (ppm):	60	Pacing Config:	NP	NP	N/A	Rate Adaptive AV Interval:	OFF
Paced AV Interval (ms):	180	Sensing Config:	NP	NP		Activity Threshold:	Medium Low
Sensed AV Interval (ms):	150	VV Interval:				Acceleration (min):	30 seconds
PVARP or ARP (mS):	310	IVRP:				Deceleration (min):	Excercise
VRP:							

REPLACED DEVICE INFORMATION

	Prod Number	Prod Name	Serial	Manufacturer	Implant Date
REPLACED ICD/IPG	VEDRO1	VERSA	NWH200018H	Medtronic	12/04/2008
	Prod Number	**Serial**	**Status**	**Manufacturer**	**Implant Date**
LEADS/ ADAPTERS	10295B	LBT000849R	Capped/Reusable	Medtronic	06/21/1994
	10295B	LBT000846R	Capped/Reusable	Medtronic	06/21/1994



ICD - Genetic Arrhythmias

HCM – Case 2

Table 3 Univariate associations with inappropriate shocks

Characteristic	Without inappropriate shocks	With inappropriate shocks	*P*
All patients (N = 144)	130 (90.3)	**14 (9.7)**	
Sex			
Female	46 (35.4)	7 (50)	.28
Male	84 (64.6)	7 (50)	
CHD			
No	64 (49.2)	9 (64.3)	.28
Yes	66 (50.8)	5 (35.7)	
On antiarrhythmic medication			
No	27 (20.8)	3 (21.4)	.95
Yes	103 (79.2)	11 (78.6)	
Type of device			
Single chamber	35 (26.9)	0 (0)	**.02**
Dual chamber	95 (73.1)	14 (100)	
Device manufacturer			
Medtronic	66 (50.8)	6 (42.9)	.37
Boston Scientific	31 (23.8)	6 (42.9)	
St Jude Medical	33 (25.4)	2 (14.3)	
History of SVT	26 (20)	7 (50)	**.02**
Age at implant (y)	17.4 ± 10.1	15 ± 6.6	.40
Weight at implant (kg)	57.3 ± 23.7	52.1 ± 19.9	.45
VF detection rate (beats/min)	226 ± 14	226 ± 23	.97
VF detection duration (beats)	18.7 ± 8.4	16.9 ± 7.3	.46
Shocks programmed in the VT zone	80 (62)	11 (92)*	**.03**

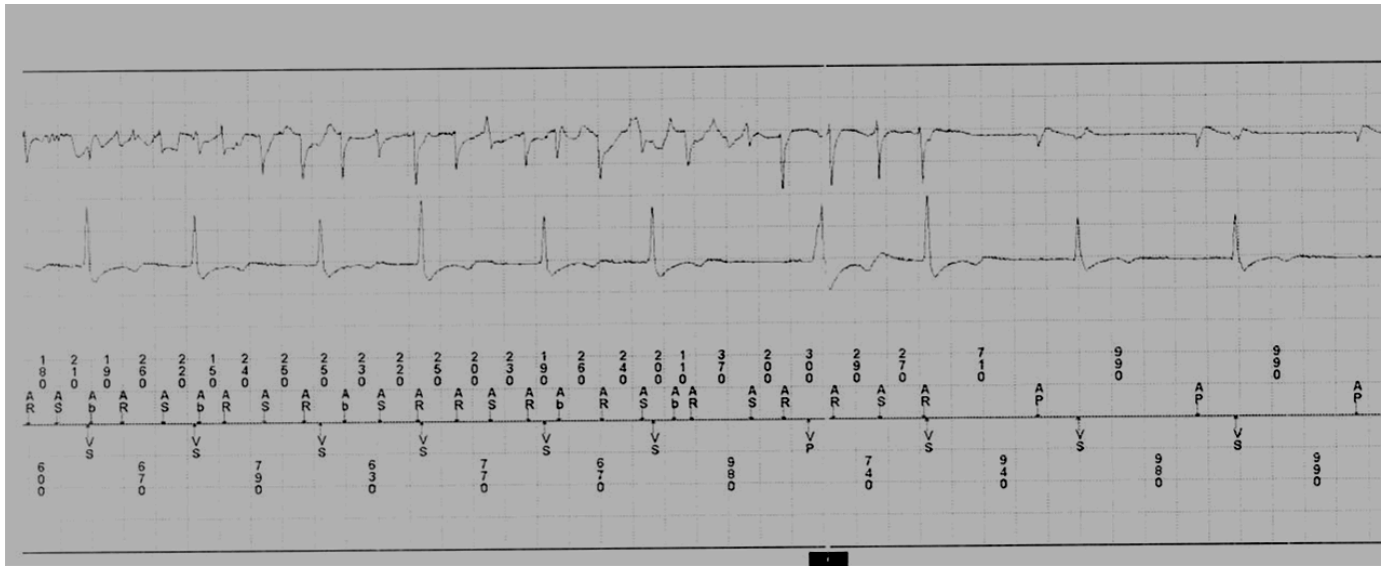
Values are presented as mean ± SD or as n (%). Results in bold indicate statistical significance.
CHD = congenital heart disease; SVT = supraventricular tachycardia; VF = ventricular fibrillation; VT = ventricular tachycardia.
*Excludes 2 patients receiving an inappropriate shock for whom these data are unknown.

Implant atrial lead only if required based on pacing needs
Need to adjust "atrial discriminators"

Garnreiter Heart Rhyhtm 2015

ICD - Genetic Arrhythmias

HCM – Case 2



Atrial rate monitoring

ICD - Genetic Arrhythmias

HCM – Case 2

STIMULATION THRESHOLD

Chamber	Polarity	Pulse Width (ms)	Voltage(V)	Current (mA)	Impedance (Ohms)	P/RWave (mV)	SlewRate (V/Sec)
Right Atrium	Bipolar	0.50	.5	0.7	704	2.2	1.2
Right Ventricle	Bipolar	0.50	.4		562	4	

DEVICE MEASURED DATA

Chamber Pac	P/R Wave (mV)	Pacing Imp	HVB Imp	HVX Imp	Test Shock	Pacing Threshold V	Pacing Threshold ms
Right Atrium	2.0	589				0.5	0.40
Right Ventricle		494	74			0.5	0.40

Note: Impedances given in Ohms

INDUCTIONS

#	Induction	T-SHOCK mSec	T-SHOCK Joule)	Episode	RX	Pre-RX Rhythm	Therapy	Post-RX Rhythm	Imp	Energy	Charge Time	Path	CL	DX	R-Wave VF
1	V 50 Hz			1	5	VF	DEFIB	Paced	71	35	6.5	B > AX	230	1.2	0

DETECTIONS

PARAMETERS	AF	AT	VF		FVT	VT
Interval (ms)			300			
Rate (BPM)						
Initial NID			18	24		
Redetect NID			9	12		

ANTI-TACHY PACING	A	V
Pulse Width (ms):		
Amplitude (V):		
Minimal Interval (ms):		
Blanking After Pace (ms):		

VT/VF Discrimination: VVI/VVOBackup: Tilt (%):

SVT DISCRIMINATION

Afib: ON
Sinus Tach: ON
Other 1:1: OFF
SVT Limit: 280 mS

EGM Width:
Width Threshold (ms):
Slew Threshold (mV):

VT Stability:
VT Onset:

Wavelets
1
2
3

TACHY PARAMETER SETTINGS

VF	Therapy #	Therapy Type	Energy (J/%)	Pathway
	1	Defib	35	B > AX
	2	Defib	35	AX > B
	3	Defib	35	B > AX
	4	Defib	35	AX > B
	5	Defib	35	B > AX
	6	Defib	35	AX > B

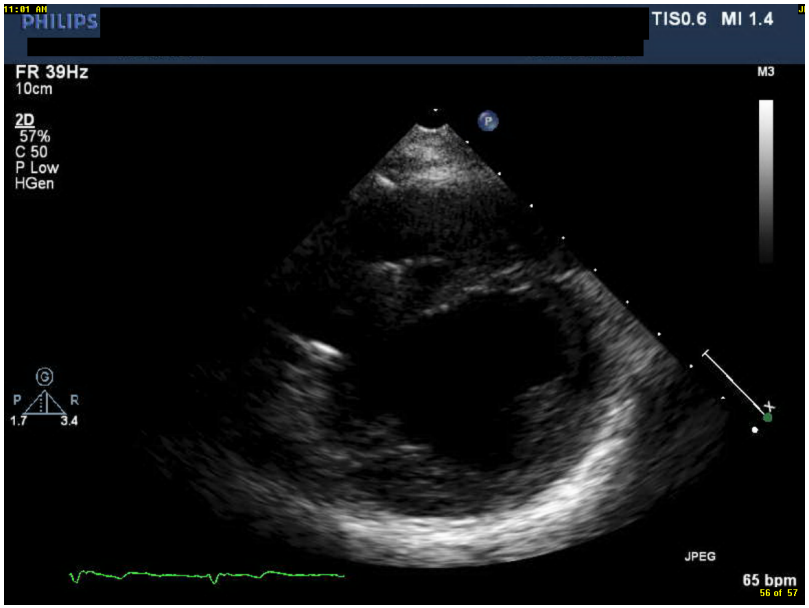
Reconfirm VF: YES
Patient Activated:

Daily Availability: Max A Defibs: Window Start: Window Length: Stop Time:

ICD - Genetic Arrhythmias

IDCM– Case 3

- 10yo Female
- Idiopathic Dilated Cardiomyopathy
- NYHA III
- CRT pace for heart failure
- Onset of non-sustained VT 6y later
  - Variable rates
- Switch to ICD



ICD - Genetic Arrhythmias

IDCM– Case 3

Mode		Rates		CRT	
Mode	DDDR	Lower	60 bpm	AdaptivCRT	Nonadaptive CRT
Mode Switch	171 bpm	Upper Track	150 bpm	V. Pacing	LV->RV
		Upper Sensor	150 bpm	V-V Pace Delay	0 ms
				Paced AV	140 ms
				Sensed AV	130 ms

Pacing Details	Atrial	RV	LV
Amplitude	1.50 V	2.00 V	2.75 V
Pulse Width	0.40 ms	0.40 ms	0.40 ms
Capture Management	Adaptive	Adaptive	Monitor
Amplitude Margin	2.0 X	2.0 X	
Min. Adapted Amplitude	1.50 V	2.00 V	
Max. Adapted Amplitude			6.00 V
Acute Phase Remaining	Off	Off	
Acute Phase Completed	26-Nov-2014	26-Nov-2014	
Sensitivity	2.10 mV	0.30 mV	
Pace Polarity	Bipolar	Bipolar	LVtip to RVcoil
Sense Polarity	Bipolar	Bipolar	

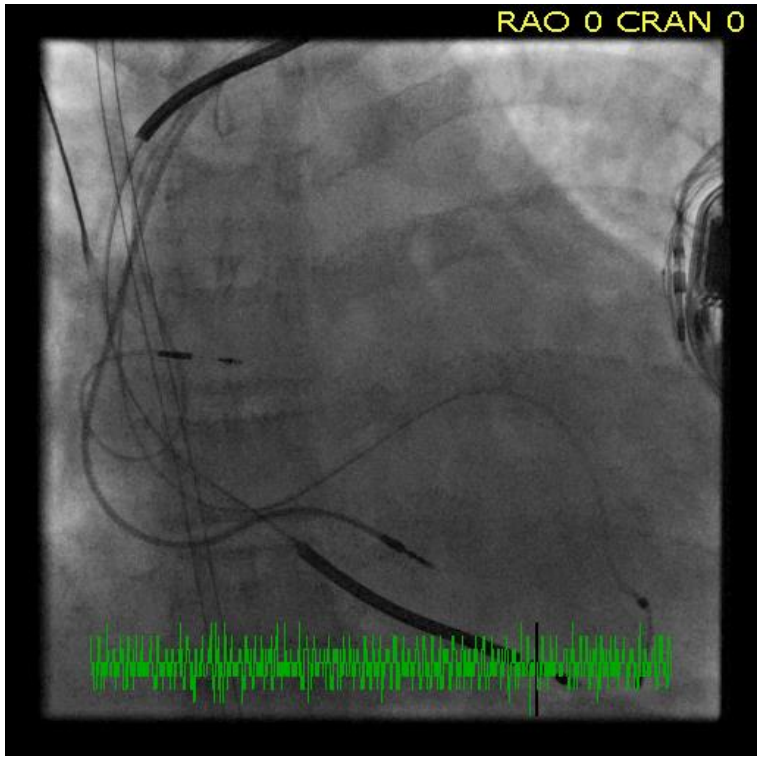
Parameter Summary

Mode	DDDR	Lower Rate	60 bpm	AdaptivCRT	Nonadaptive CRT
Mode Switch	171 bpm	Upper Track	150 bpm	V. Pacing	LV->RV
		Upper Sensor	150 bpm	Paced AV	140 ms
				Sensed AV	130 ms

Detection		Rates	Therapies
AT/AF	Monitor	>171 bpm	All Rx Off
VF	On	>200 bpm	ATP During Charging, 35J x 6
FVT	via VF	200-207 bpm	35J x 6
VT	On	167-200 bpm	35J x 6

Enhancements On: AF/Afl, Sinus Tach, Wavelet, TWave, Noise(Timeout)

Changes This Session	Session Start	Current Value
No parameters have been changed during the current session.		

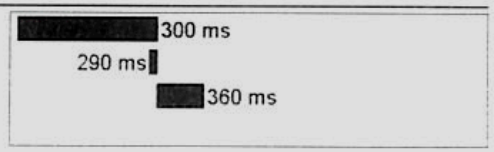


ICD - Genetic Arrhythmias

IDCM– Case 3

VT/VF Detection

		V. Interval (Rate)	Initial	Redetect
VF	On	300 ms (200 bpm)	30/40	12/16
FVT	via VF	290 ms (207 bpm)		
VT	On	360 ms (167 bpm)	32	28
Monitor	Off	450 ms (133 bpm)	32	



PR Logic/Wavelet		Other Enhancements		Sensitivity	
AF/Afl	On	Stability	Off	Atrial	2.10 mV
Sinus Tach	On	Onset	Off	RV	0.30 mV
Other 1:1 SVTs	Off	High Rate Timeout			
Wavelet	On	VF Zone Only	Off		
Template	None	All Zones	Off		
Match Threshold	70 %	TWave	On		
Auto Collection	Off	RV Lead Noise	On+Timeout		
SVT V. Limit	260 ms	Timeout	0.75 min		

ICD - Genetic Arrhythmias
Conclusions

- Genetic arrhythmias are variable
  - Atrial, Ventricular, Both
- There is no one therapeutic approach
- Apply effective preventive management
- Patient-specific device programming is mandatory

Children's Hospital of Michigan
Detroit Medical Center / Wayne State University

PEDIRHYTHM VII

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THANK YOU