

Devices: Indications and Risk

ICD Guidelines in Children and Congenital Heart Patients





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The following relationships exist related to this presentation: NONE

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

ICDs in the YOUNG w/wo CHD Concepts

- 1989 first applied to the young
- ~ 1% of ICD recipients
- Can be effective therapy
- Inappropriate shocks common
- Overall complication rates greater than in adults
- Congenital heart morphologies often limit implant choices



*Pics courtesy of Dr Berul: J Innovations Card Rhythm Mngt 2011

ICDs in the Young Device-related Considerations

Endocardial Single / dual coils

Epicardial

- Patch
- Array
- Subcutaneous
- +/- CRT systems









ICDs in the Young Device-related Considerations

- Medtronic
 - Sprint Fidelis lead fractures and a number of deaths and serious injuries potentially linked to their use
- St. Jude Medical
 - lawsuits over its recalled Riata debrillator leads
- Sorin
 - ICD leads recalled from market after causing inappropriate shocks



Atallah 2013 Circulation

ICDs in the Young Patient-related Problems

- Adult patients
 - **1 5** %
- Pediatric patients
 - **7 36%**









ICDs in the Young Inappropriate Therapy Delivery

N = 76 pts; 90 ICD' S				
Cause	No. of Patients	Median Time to First Shock (months)	No. of Shocks [Mean (max)]	
Lead failure	7	35*	25† (60)	
Sinus tachycardia	8	3	1.6 (4)	
SVT/IARŤ	4	4	2.3 (4)	
T wave oversensing	2	22	1.5 (2)	
Overall (patients)	19 (25%)	16	10 (60)	

Two patients had inappropriate therapy both for lead failure and sinus tachycardia.

*P = 0.028; $\dagger P = 0.038$, Kruskal-Wallis.

IART = intra-atrial reentry tachycardia; SVT = supraventricular tachycardia.

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LEAD PROBLEMS = 36%
SENSING PROBLEMS = 74%
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ICDs in the Young Typical Patient Ages



Alexander 2004 JCVEP



ICDs in the Young Published Clinical Implant Reasons



ICDs in the Young Genetic Arrhythmias HCM Long QT syndrome Dilated cardiomyopathy Other Familial arrhythmia Idiopathic VT/VF ARVC CPVT

Garnreiter. Heart Rhythm 2015

ICDs in the Young Congenital Heart Arrhythmias





There are no published "ICD Guidelines" specific to children or most CHD patients



- End the presentation...Thank you
- Or

Extrapolate

"project into an unexplored (Pediatrics/Congenital heart) situation from observations in an explored (Internal Medicine) field"

ICDs in the Young Important Issues to Remember

- ICDs are designed for adults
- To date, no Pediatric-specific ICD trials
- Pediatric concerns
 - Sensing issues (T wave, heart rates)
 - Growth, activity, lead replacement, QOL (emotional)
 - Vascular, valvular effects
- Patient-specific implant "customization" required
 - Subcutaneous coil arrays
 - Pericardial, pleural implant

ICDs in the Young Important Issues to Consider

Customize therapy delivery based on patient needs

- ICD shocks can be pro-arrhythmic
 - Anti-tachycardia overdrive pacing may be better if feasible
- Arrhythmias can be self-perpetuating (post-op CHD)
 - Ischemic areas, stretch, elevated pressures
 - Correct hemodynamics first
- Concomitant drug therapies can affect ICD function
 - Sodium channel blockade
 - Sensing, defibrillation thresholds

PRE-IMPLANT EP TESTING OF PATIENT- SPECIFIC ARRHYTHMIAS ALLOWS FOR FUTURE PROGRAMMING DECISIONS

ICD GUIDELINES - PUBLICATIONS

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APPROPRIATE USE CRITERIA

ACCF/HRS/AHA/ASE/HFSA/SCAI/SCCT/SCMR 2013 Appropriate Use Criteria for Implantable Cardioverter-Defibrillators and Cardiac Resynchronization Therapy

No specific recommendations for children per se Recommendation for repaired Tetralogy with syncope and inherited Genetic conditions

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ICD GUIDELINES - PUBLICATIONS

EXPERT CONSENSUS STATEMENT

HRS/ACC/AHA Expert Consensus Statement on the Use of Implantable Cardioverter-Defibrillator Therapy in Patients Who Are Not Included or Not Well Represented in Clinical Trials

Developed in partnership with and endorsed by the American College of Cardiology (ACC) and the

No specific recommendations for children or CHD patients

Endorsed by the European Heart Rhythm Association (EHRA), the Asia Pacific Heart Rhythm Society (APHRS) and the Sociedad Latinoamericana de Estimulacion Cardiaca y Electrofisiologia (SOLAECE)-Latin American Society of Cardiac Pacing and Electrophysiology

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ICD GUIDELINES Patient Selection Criteria – 2° Prevention*

- Coronary disease / Acute MI: Unstable VF/VT
 - ? Old Kawasaki
- Coronary disease: VF/VT Exercise
- No Coronary disease: VF/VT
 - Dilated cardiomyopathy
 - Drug abuse
 - Myocarditis, Cardiomyopathy
- Inherited Genetic Disease: VF/VT
 - LQTc, CPVT, ARVD, HCM, Brugada, WPW
- On Waiting List for Transplant: VF/VT



ICD GUIDELINES Patient Selection Criteria – 1° Prevention*

- Coronary disease / Acute MI: non sustained VT
 - LVEF ≤ 30%; 31-40%
 - Chronic / ischemic cardiomyopathy
- Non ischemic cardiomyopathy LVEF ≤ 40%
- Myocardial Disease
 - Giant Cell Myocarditis
 - Amyloid, Sarcoid, Chagas
- Inherited Genetic Disease
 - HCM, ARVD, LQTc, Brugada, DCM
- On Waiting List for Transplant NYHA IV

* Pre-emptive



ICD GUIDELINES

Patient Selection Criteria – <u>Unexplained</u> Syncope

- Non-ischemic Dilated CM
 Esp LVEF ≤ 35%
- Hypertrophic CM*
- LV Non compaction*
- Repaired Tetralology of Fallot*
- ARVD*#
 - * irrespective of Ejection Fraction
 - # regardless of EPS findings



SYNCOPE Etiology

- 16% adolescents bonafide syncope
- 60% adolescents pre-syncope
- 226 children in emergency department with syncope
 - Neurocardiogenic syncope +80%
 - Neurologic disorders 9%
 - Cardiac disorders: 5 cases (2%)

Perform very detailed evaluation of syncope before considering ICD!



Massin et al. Pediatr. 2004



Primary Prevention: Genetic Conditions (Excludes Syncope and Sustained VT)

A Appropriate; CM cardiomyopathy; ECG electrocardiogram; EPS electrophysiological study; GDMT guideline-directed medical therapy; LV left ventricular;

LVEF left ventricular ejection fraction; M May Be Appropriate; MI myocardial infarction; NICM nonischemic cardiomyopathy; NSVT nonsustained ventricular tachycardia;

R Rarely Appropriate; RV right ventricular; SCD sudden cardiac death; VF ventricular fibrillation; VT ventricular tachycardia.

Primary Prevention: Genetic Conditions (Excludes Syncope and Sustained VT)



QTc Risk Factors: >500ms Prior cardiac arrest Events on therapy ≤ 20 years old

GDMT: guideline-mediated medical therapy





Brugada*



Type 3: Saddle-back type "ST-segment elevation"

* Not typically seen in young children

Brugada Syndrome

Typical age 40+/- 22y

Pediatric Data

- N = 91
- 9.7y mean (3m -17y)
- ◆ 65% +FH
- 3% FH SCD
- 1% SCD
- 44% abn ECG at rest
- 27% abn ECG drugs
 - Na+ blockers
 - Ajamaline
 - 1C agents ~80% specificity
- Syncope ~10y age

Brugada Syndrome

V,





Primary Prevention: Genetic Conditions (Excludes Syncope and Sustained VT)



Primary Prevention: Genetic Conditions (Excludes Syncope and Sustained VT)



ICD GUIDELINES Cardiomyopathy

Primary Prevention: non coronary-related cardiomyopathies



ICD GUIDELINES Other Cardiomyopathy

Primary Prevention: non coronary-related cardiomyopathies



ICDs in the Young w/wo CHD Conclusions

- ICDs can be effective in the young
- Current "Guidelines" can be extrapolated somewhat to children and CHD patients
- Strategic planning is mandatory when applying ICD therapy to the young
- Be aware of risks / benefits





THANK YOU



