Pediatric Patients with Inherited Arrhythmia Syndromes

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Slides courtesy of Dr. Ronn Tanel
What’s so different about pediatric patients?

- Diagnosis
- Treatment
- Lifestyle
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Familial Screening

- Identification of children or siblings after an affected family member is diagnosed
- Genotype + Phenotype + or -
Age at Presentation for Inherited Arrhythmia Syndromes

LQTS
BrS
ARVC
CPVT

0 10 20 30 40

Age
For some inherited arrhythmias, an early age at onset of symptoms is a predictor of future sudden cardiac events.
Cardiac Events in LQTS

Goldenberg and Moss, JACC 2008
Sudden Infant Death Syndrome (SIDS)

SIDS is the sudden death of an infant under one year of age which remains unexplained after a thorough case investigation, including performance of a complete autopsy, examination of the death scene, and review of the clinical history.

*Willinger et al, 1991*
Sudden Infant Death Syndrome (SIDS)
Does LQTS cause SIDS?

- Genetic testing suggests that LQTS and other inherited arrhythmias may cause 5% - 10% of SIDS deaths.
- ECG screening of parents of SIDS victims suggests that the incidence may be higher.
- Some data regarding SIDS speaks against LQTS playing a significant role, such as the “Back to Sleep” program.
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Follow-up for the Pediatric Patient at Risk for an Inherited Arrhythmia

- Clinical follow-up throughout childhood
- Repeat serial ECGs
- Additional monitoring/testing in those with symptoms
- Interpretation of studies is dependent on age
- Consideration of special studies based on specific diagnosis, as well as other factors (eg. MRI for ARVC)
What’s so different about pediatric patients?

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ICD Therapy
Pacemaker and ICD in Pediatrics

- Children and teens are involved in more dynamic activity than adult patients, potentially resulting in added stress and strain on leads and generators.

- Device implantation must take into consideration continued body growth.

- Studies have demonstrated a higher incidence of lead complications in pediatric device patients.

- Cardiac devices must be implanted and cared for with the idea that pediatric patients have the potential for many decades of active lives.
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Impact of Cardiac Devices on the Quality of Life in Pediatric Patients
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Lifestyle
36th Bethesda Conference: Eligibility Recommendations for Competitive Athletes with Cardiovascular Abnormalities

<table>
<thead>
<tr>
<th>Increasing Static Component</th>
<th>III. High (&lt;50% MVC)</th>
<th>II. Moderate (20-50% MVC)</th>
<th>I. Low (&lt;20% MVC)</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Low (&lt;40% Max O₂)</td>
<td>B. Moderate (40-70% Max O₂)</td>
<td>C. High (&gt;70% Max O₂)</td>
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<tr>
<td>Bobsledding/Luge†, Field events (throwing), Gymnastics*†, Martial arts*, Sailing, Sport climbing, Water skiing†, Weight lifting†, Windsurfing†</td>
<td>Body building*†, Downhill skiing†, Skateboarding*†, Snowboarding*†, Wrestling*</td>
<td>Boxing*, Canoeing/Kayaking, Cycling†, Decathlon, Rowing, Speed-skating*†, Triathlon†</td>
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<tr>
<td>Archery, Auto racing†, Diving†, Equestrian*†, Motorcycling*†</td>
<td>American football*, Field events (jumping), Figure skating*, Rodeoing†, Rugby*, Running (sprint), Surfing††, Synchronized swimming†</td>
<td>Basketball*, Ice hockey*, Cross-country skiing (skiing technique), Lacrosse*, Running (middle distance), Swimming, Team handball</td>
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<tr>
<td>Billiards, Bowling, Cricket, Curling, Golf, Riflery</td>
<td>Baseball/Softball*, Fencing, Table tennis, Volleyball</td>
<td>Badminton, Cross-country skiing (classic technique), Field hockey*, Orienteering, Race walking, Racquetball/Squash, Running (long distance), Soccer*, Tennis</td>
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Increasing Dynamic Component
36th Bethesda Conference: Eligibility Recommendations for Competitive Athletes with Cardiovascular Abnormalities

![Graph showing eligibility recommendations for competitive athletes with varying degrees of cardiovascular abnormalities. The graph categorizes activities based on the static and dynamic components of exercise intensity. Activities such as billiards, bowling, cricket, curling, golf, and riflery are recommended for athletes in the moderate category (40-70% Max O₂).]
Comprehensive consultation and counseling to enable an informed decision making regarding sports participation

- 353 patients; 130 (37%) remained active in sports (11 y)
- 25% high school athletes; 6% college athletes
- 1 sport-related cardiac event (0.003 events/athlete year)
## Sports Played by LQTS patients

| Static Component, Peak % Maximal Voluntary Left Ventricular Contraction | No. of Patients |
|---|---|---|
| III High >50 | 25 | 4 | 0 |
| II Moderate 20-50 | 0 | 14 | 34 |
| I Low <20 | 3 | 22 | 28 |

| Dynamic Component, Peak % Maximal Oxygen Uptake |
|---|---|---|
| A Low <40 | B Moderate 40-70 | C High >70 |

Activity Guidelines

- Children and teens need to be involved in meaningful and rewarding activities that provide a sense of fulfillment and accomplishment.

- Choosing an activity, hobby, or sport should begin early, be modified as necessary, and involve the patient.

- The ideal activity is one that is acceptable to the patient and fits the medical treatment plan.
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Activity Guidelines

Long QT Syndrome and Sports Participation
Oil and Water or an Acceptable and Manageable Combination?*
Protective Devices
Adolescence

- Physical activity
- Driving
- Alcohol and recreational drug use
- Sexual activity
- Education and vocational training
- Career planning
Adolescence

- Will he take his medication?
- Will she tell me if she has symptoms?
- How will he respond to “peer pressure”?
- Can she get her driver’s license?
- Who will be watching out for my child when I’m not there?
Adolescence

- Teens should be involved in personal health care decisions
- Teens should develop an understanding and responsibility for their diagnosis and treatment
- Teens should develop support networks within their social framework, including teachers, coaches, clergy, club leaders, guidance counselors, health care providers, and other mentors
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The ultimate goal of pediatric health care is to allow the child/adolescent to continue with as normal activity as is reasonably possible in order to promote healthy development, while minimizing significant risk.
Acknowledgement

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