Duchenne Muscular Dystrophy and Sudden Cardiac Death: Does it Happen and How Do We Prevent It?

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Sudden Cardiac Death

- Sudden cardiac death is one of the primary causes of death in adults.

- Sudden cardiac death is due to fast rhythms originating from the bottom chambers of the heart causing a drop in blood pressure.

- These rhythms cause fainting as well as sudden death.
Heart Beat Anatomy

SINUS NODE
• The Heart’s ‘Natural Pacemaker’
  - 60-100 BPM at rest
• Tells the heart to speed up when you run and slow down when you are sleeping
Heart Beat Anatomy

Sinus Node (SA Node)

Atrioventricular Node (AV Node)

AV NODE
• Receives impulse from SA Node
• Delivers impulse to the His-Purkinje System
• 40-60 BPM if SA Node fails to deliver an impulse
Premature Atrial Beats

- Origin: Atrium (outside the Sinus Node)
Monomorphic Ventricular Tachycardia

- Origin: Ventricles (Single Focus)
- Characteristics: Rapid, wide, and regular QRS
Polymorphic Ventricular Tachycardia

- Origin: Ventricles (Wandering Single Focus)
- Characteristics: Wide and irregular QRS
Can We Predict Who is at Risk?

• Patients who had a history of a heart attack were known to have a high risk of sudden cardiac death

• The ability to induce a life threatening rhythm in the catheterization laboratory predicted those at highest risk

• Over time it was found that cardiac catheterizations were not necessary and that the best predictor of sudden cardiac death was poor cardiac function

• Furthermore, the best treatment to prevent sudden cardiac death was found to be the implantable cardiac defibrillator
The Implantable Cardiac Defibrillator (ICD)

ICD System

Device + Lead + Programmer
Implantable Cardiac Defibrillator (ICD)

Device Components Overview

- Battery
- Circuitry
- Active Can
- Capacitor (2 shown)
  200 μF ea.
  400 V ea.
- Leads To Heart
The ICD System – How it Works
The ICD System – How it Works
The ICD System – How it Works

- Atrium & Ventricle
  - Pacing
- Ventricle
  - VT prevention
  - Defibrillation

Atrium & Ventricle
- Pacing
Sensing

- Sensing - what the device "sees"
- Electrical Activity - what the device is looking for
- Lead – contains the ‘eyeball’ of the device
Sensing

The ‘eye’ of the device

• Constantly watches for electrical activity
• Sees or senses all electrical signals that pass
• Signal produced by a passing electrical wave is called an Intracardiac Electrogram (EGM)
Shocking or Defibrillation Circuit

A > B

AX > B
VF Detection and Therapy

<table>
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<th>Type</th>
<th>ATP Seq</th>
<th>Shocks</th>
<th>Success</th>
<th>ID#</th>
<th>Date</th>
<th>Time hh:mm:ss</th>
<th>Duration hh:mm:ss</th>
<th>Avg bpm A/V</th>
<th>Max bpm A/V</th>
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<td>VF</td>
<td>0</td>
<td>20J</td>
<td>Yes</td>
<td>1</td>
<td>06-Oct-2011</td>
<td>10:18:09</td>
<td>---/240</td>
<td>---/-240</td>
<td>---/-240</td>
<td>Rest</td>
</tr>
</tbody>
</table>

- V-V
- A-A

VF = 350 ms

Detection

19.7 J

Term.
VF Detection and Therapy
So Everyone Should Get an ICD

• Imperfections
  • Need anesthesia for implantation
  • Inappropriate discharges
  • Lead complications
  • Psychological impact
  • ICDs do not treat cardiomyopathy

• Indications
  • What is the incidence of sudden cardiac death
  • Can we predict who is at risk
Who Is At Risk – Dilated Cardiomyopathy

Sudden Cardiac Death – Heart Failure Trial

• 2521 patients with left ventricular ejection fraction ≤ 35% followed for 4 years

• Median age of 60 years

• 1211 with cardiomyopathy not due to a heart attack

• ICD therapy reduced overall mortality by 23%

Bardy et al. NEJM 2005
Current Recommendations for ICD Implantation

- Patients who have survived a sudden cardiac arrest (secondary prevention)

- Patients with severely depressed cardiac function (primary prevention)
Who Is At Risk – Dilated Cardiomyopathy

- Dimas et al, 2009 - reviewed all patients with idiopathic dilated cardiomyopathy seen at Texas Childrens Hospital over 14 year period
  - 85 patients with median age of 3.8 yrs
  - Mean ejection fraction was 25% (median 23%)
  - 1 episode of sudden cardiac death (1%)
Inappropriate Discharges in Young Adults

• Berul et al evaluated 443 patients with ICDs with a median age of 16 years at 4 large pediatric institutions

• Inappropriate shocks in 87/409 patients (21%)
  • Mean of 6 per patient (median 4)
  • Mainly due to lead failure or incorrect diagnosis of the arrhythmia

• 24% of patients <18 years experienced at least one inappropriate shock compared with 14% in adults
Kids Are Not Adults

- Incidence of sudden cardiac death in children / adolescents with dilated cardiomyopathy is much lower than in adults

- Role of prophylactic placement of ICD or primary prevention is less clear

- Higher risk of complications both with procedure and following the procedure
What About Sudden Cardiac Death in Patients with Muscular Dystrophy?
Sudden Cardiac Death in DMD

• No data on the incidence of ventricular arrhythmias

• No data on the incidence of sudden cardiac death

• Use of implantable defibrillators based on data obtained from patients with a mean age 60 years with a different reason for their cardiomyopathy
Conclusion

- Implantable cardiac defibrillators prevent sudden cardiac death

- The risk of sudden cardiac death in Duchenne muscular dystrophy is unknown

- Decision to place ICD for primary prevention must take into account not only risk of sudden death but risks of ICD implantation, inappropriate discharges and lead complications
UPCOMING :: NEW RESEARCH STUDY
SUDDEN CARDIAC DEATH IN DUCHENNE MUSCULAR DYSTROPHY:
INCIDENCE AND PREVENTION

This upcoming patient registry will determine the rate at which
these children suffer from sudden cardiac death or irregular
heartbeats and whether current therapy with devices such as
internal cardiac defibrillators is beneficial. This will be a multi-
site study.

Pending approval by the Nationwide Children’s Hospital
Institutional Review Board (IRB), recruitment will begin.

Visit www.parentprojectmd.org or
call 614-722-0494 with questions.