Electricity Makes the Heart Beat

- Cells in the heart send out electrical impulses
- These impulses move through the heart
- Electrical impulses tell the heart to contract and pump blood
- Electricity
- Plumbing
What are Arrhythmias?

- Abnormal beating of the heart
  - Too fast
  - Too slow
  - Irregular
- Can originate from atrium, ventricular, or use both
  - Atrial Tachycardia
  - Ventricular Tachycardia
  - SVT
Arrhythmias

- **Atrial Tachycardia**
  - Originate from atrium
  - Faster than normal

- **Ventricular Tachycardia**
  - Abnormal impulses from ventricle
  - Faster than normal
  - Associated with abnormal heart muscle

- **Ventricular Fibrillation**
  - Disorganized electrical impulses
  - Heart stops beating
Ventricular Fibrillation

- Ventricular Tachycardia → Ventricular Fibrillation
Arrhythmias

- **Long QT Syndrome**
  - QT interval on ECG is long
  - Represents delayed relaxation of the heart
  - Can lead to arrhythmias
    - Ventricular Tachycardia
      - Torsades de pointes
    - Ventricular Fibrillation
  - Can be exacerbated by QT prolonging medications
- [https://www.crediblemeds.org](https://www.crediblemeds.org)
Arrhythmia in Barth Syndrome

- Patients with Barth Syndrome:
  - Increased incidence of arrhythmias
  - Abnormal heart muscle
- Ventricular arrhythmias are associated with sudden cardiac death
- Sudden cardiac death is a known risk in Barth Syndrome
  - Caused by malignant ventricular arrhythmias
- Unclear how to clearly risk stratify for sudden cardiac events
Arrhythmia in Barth Syndrome

- Patients with cardiomyopathy have an increased incidence of ventricular arrhythmia

- Cardiomyopathies associated with Barth Syndrome
  - Left Ventricular Non-Compaction
  - Dilated Cardiomyopathy
  - Hypertrophic Cardiomyopathy

- Mechanisms of arrhythmia
  - May be related to mitochondrial disease
    - mitochondrial activities that are important for cardioprotection
  - May be related to type of cardiomyopathy
Arrhythmia in Barth Syndrome

- The Barth Syndrome Registry: distinguishing disease characteristics and growth data from a longitudinal study.
  - Spencer et al.
  - 70 patients with Barth
  - 9 had documented ventricular arrhythmias
  - Most older than 11 years
- Malignant arrhythmias described in younger children
- Unclear if there is clear relationship between prolonged QT interval and increased risk of death
Surveillance

- Regular Screening
  - ECG
    - Signal-Averaged ECGs
    - T wave alternans testing
  - Ambulatory Monitors
- Symptom Review

- Increased Frequency with Higher Risk Patients
  - Left Ventricular Non-Compaction
  - Poor Ventricular Function
  - Syncope
Symptoms of Arrhythmia

- Palpitations
  - Fluttering sensation
  - Pounding sensation
- Syncope (Fainting)
- Lightheadedness
- Shortness of Breath
- Anxious feeling
- Chest Pain
- Asymptomatic
Traditional Ambulatory Monitors

- Holter Monitors
- 30 Day Looping and Non-looping Monitors
- Mobile Cardiac Telemetry (MCT)
Newer Ambulatory Monitoring
Newer Ambulatory Monitors
Wearables

CENTRAL ILLUSTRATION: Wearable Monitoring Devices

Ambulatory Monitoring Capabilities
- ECG
- Heart Rate
- Arrhythmia
- Blood Pressure
- Cardio-Respiratory Fitness
- Stress
- Respiratory Rate
- Temperature
- Oxygen Saturation
- Ischemia
- Apnea

Devices for Ambulatory Monitoring
- Wristwatches
- Smartphones
- Patches
- Headbands
- Eye-glasses
- Necklaces

Wearables

- I-Watch
- Kardia
- Fitbit
- Oura Ring
Wearables: State of the industry

Annual revenue from wearable technology products

IDTechEx Research

Total revenue (USD billions)


YoY growth rate

Wearable cameras
Headphones, Hearing aids “Hearables”
E-textiles
Skin patches
AR, VR, MR
Smartwatches & fitness trackers

Others

Changing What’s Possible | MUSCkids.org
Implantable Loop Recorder
Implantable Loop Recorder

**BioMonitor 2** (Biotronik SE & Co, Berlin, Germany)
- 88 mm
- 15 mm

**Reveal LINQ** (Medtronic, Minneapolis, USA)
- 44.8 mm
- 7.2 mm

**Reveal XT** (Medtronic, Minneapolis, USA)
- 62 mm
- 19 mm

**Confirm Rx™ ICM** (St Jude Medical, Minnesota, USA)
- 49 mm
- 9.4 mm
Implantable Loop Recorder
Implantable Loop Recorder
Electrophysiology Study

- Catheters placed in heart
- Look at electrical properties of heart
- Can evaluate for arrhythmias
- A way to risk stratify
Automatic Implantable Cardioverter Defibrillator (AICD)

Figure 1. Examples of ICDs (pictures are not to scale).
Subcutaneous AICD
AICD placement

- Want to place before event occurs
- Sometime clear indications
  - Secondary prevention
  - Ejection Fraction <35%
  - Syncope consistent with arrhythmia
- Other times may not be clear cut
  - Multidisciplinary approach
  - Cardiologist, Electrophysiologist, Family
Electrophysiologist

- When to refer?
  - Syncope
  - Abnormal Heart Monitors
  - Significant Ventricular Dysfunction
  - Never Too Early
- Can help guide timing of AICD
  - And place it
Questions?