



Barth Syndrome: Cardiac Arrhythmia Concerns

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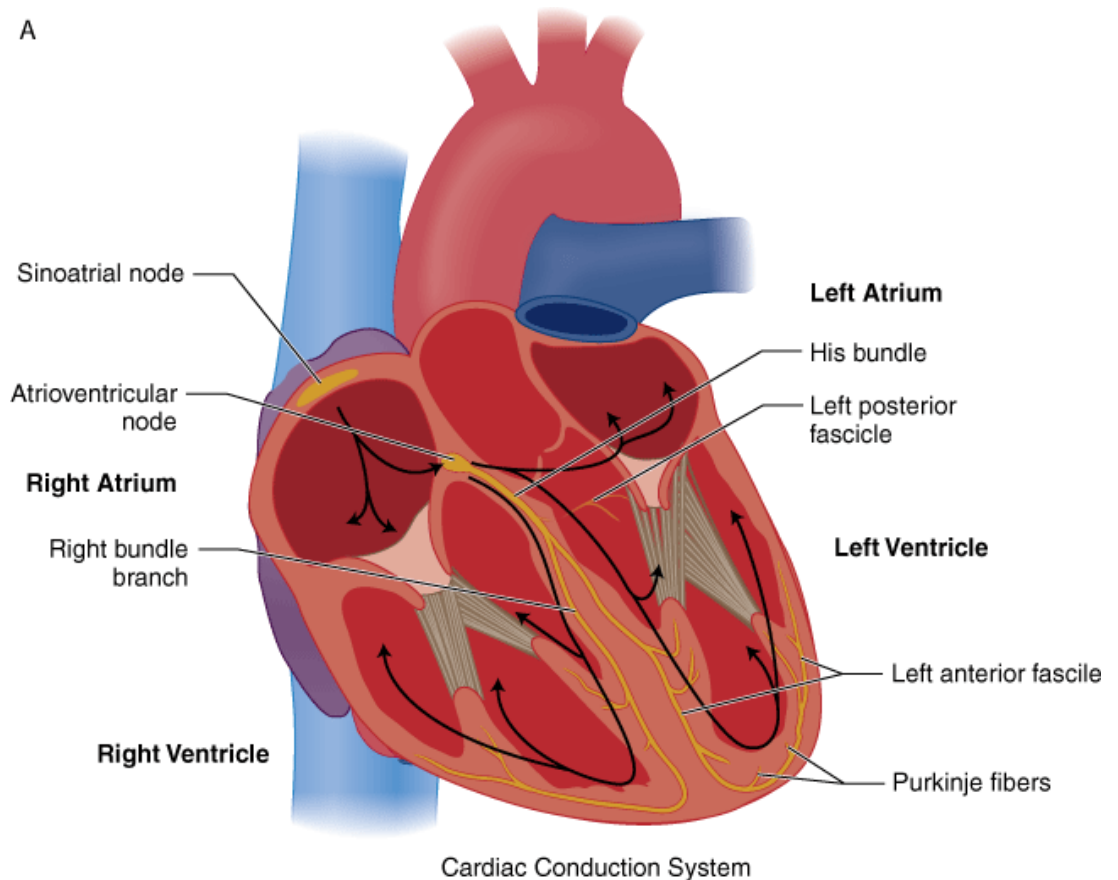


musc
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Changing What's Possible

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



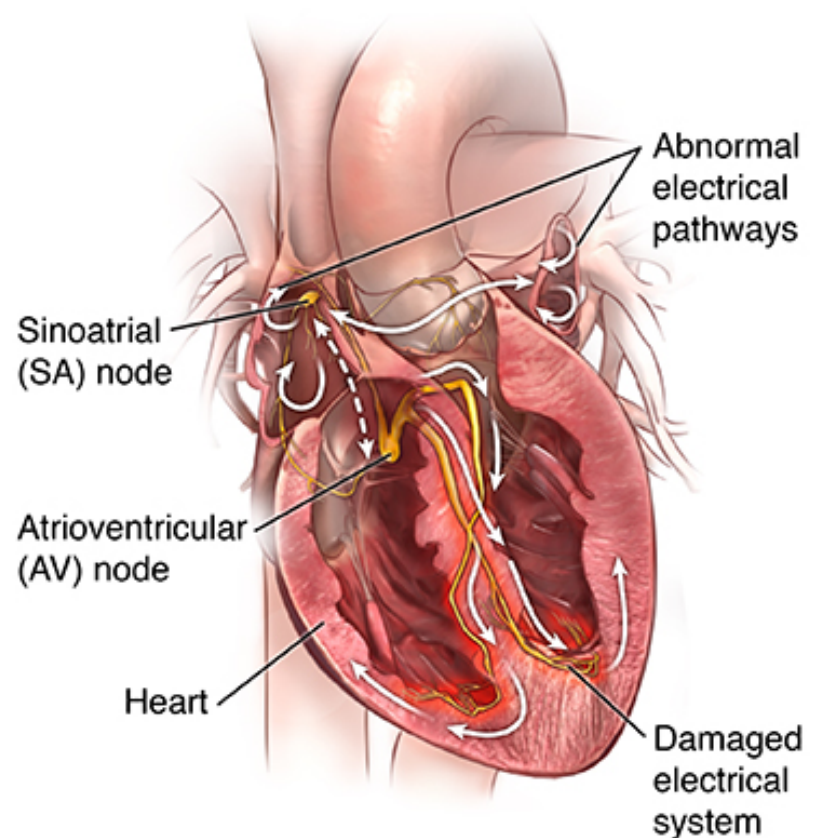
Source: Tintinalli JE, Stapczynski JS, Ma OJ, Cline DM, Cydulka RK, Meckler GD:
Tintinalli's Emergency Medicine: A Comprehensive Study Guide, 7th Edition:
<http://www.accessmedicine.com>
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What are Arrhythmias?

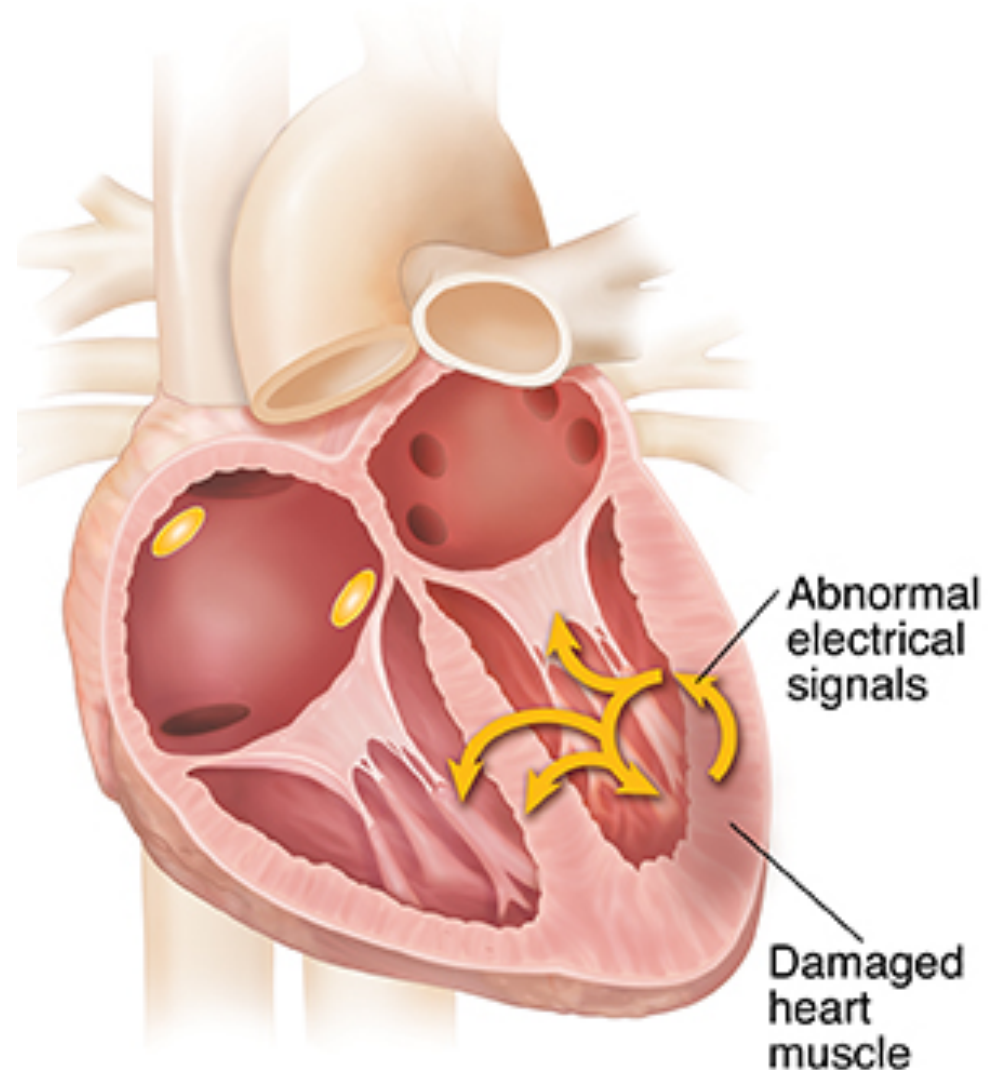
- ▶ Abnormal beating of the heart
 - ▶ Too fast
 - ▶ Too slow
 - ▶ Irregular
- ▶ Can originate from atrium, ventricular, or use both
 - ▶ Atrial Tachycardia
 - ▶ Ventricular Tachycardia
 - ▶ SVT

Damaged electrical system of the heart

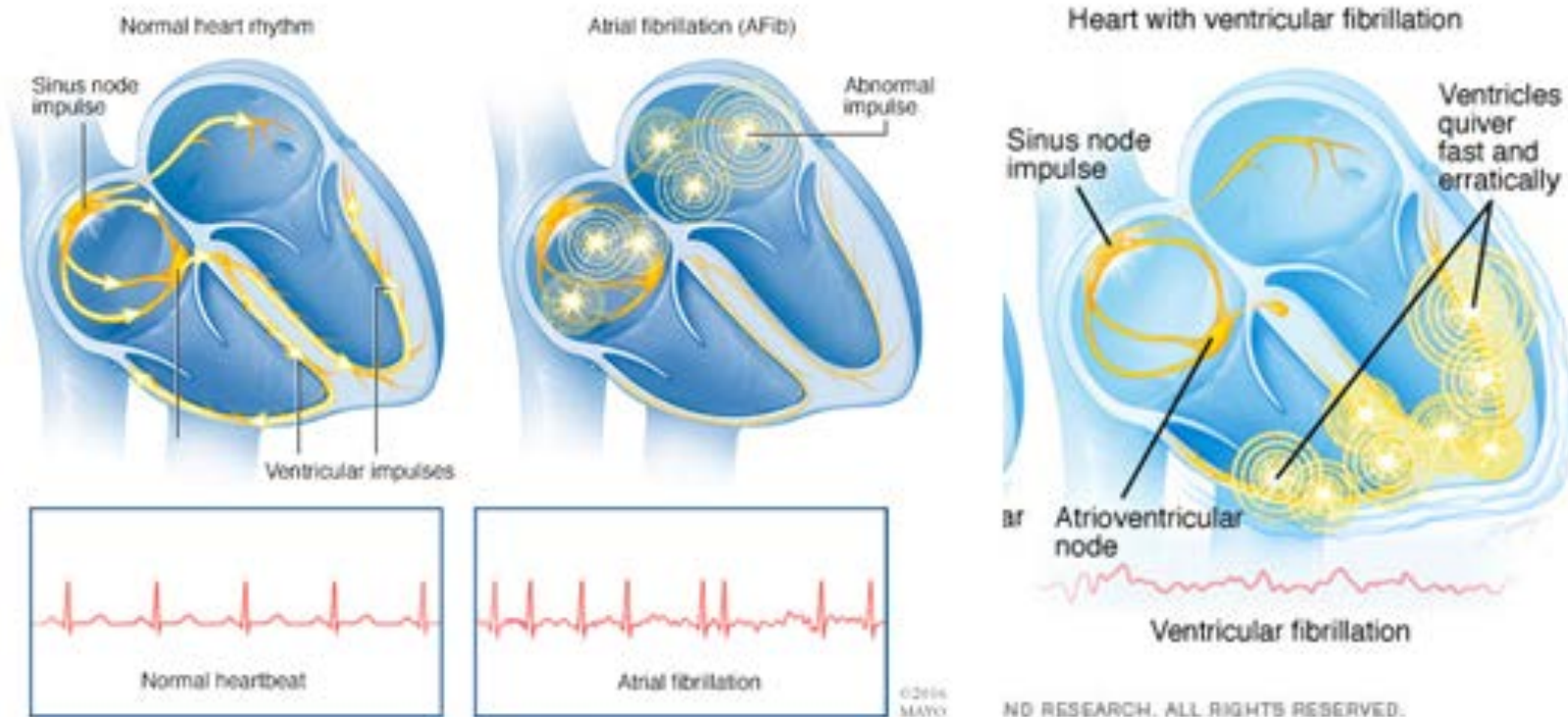


Arrhythmias

- ▶ **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

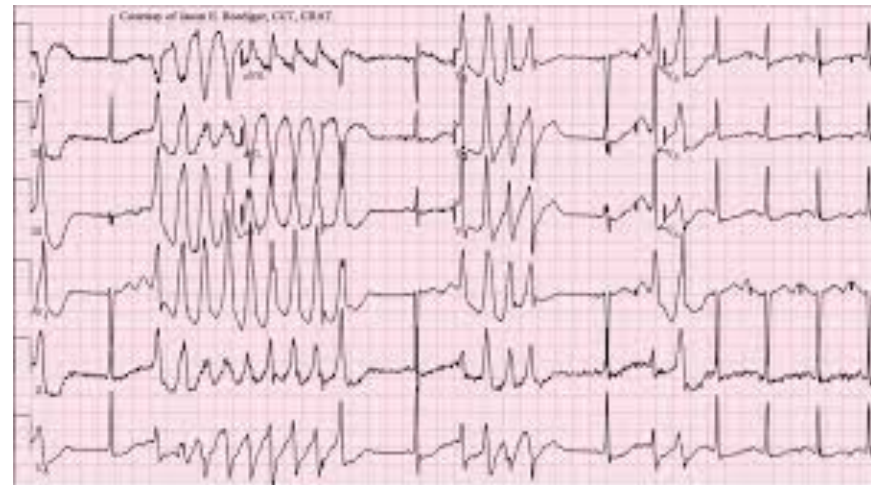
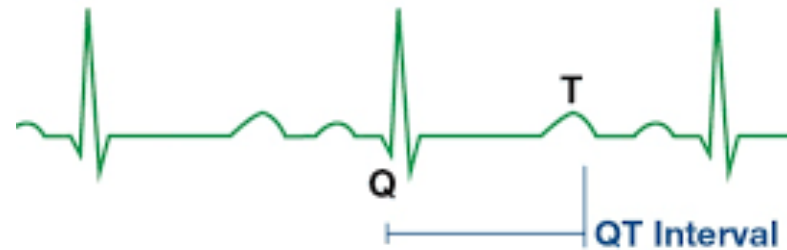


- ▶ Ventricular Tachycardia → Ventricular Fibrillation



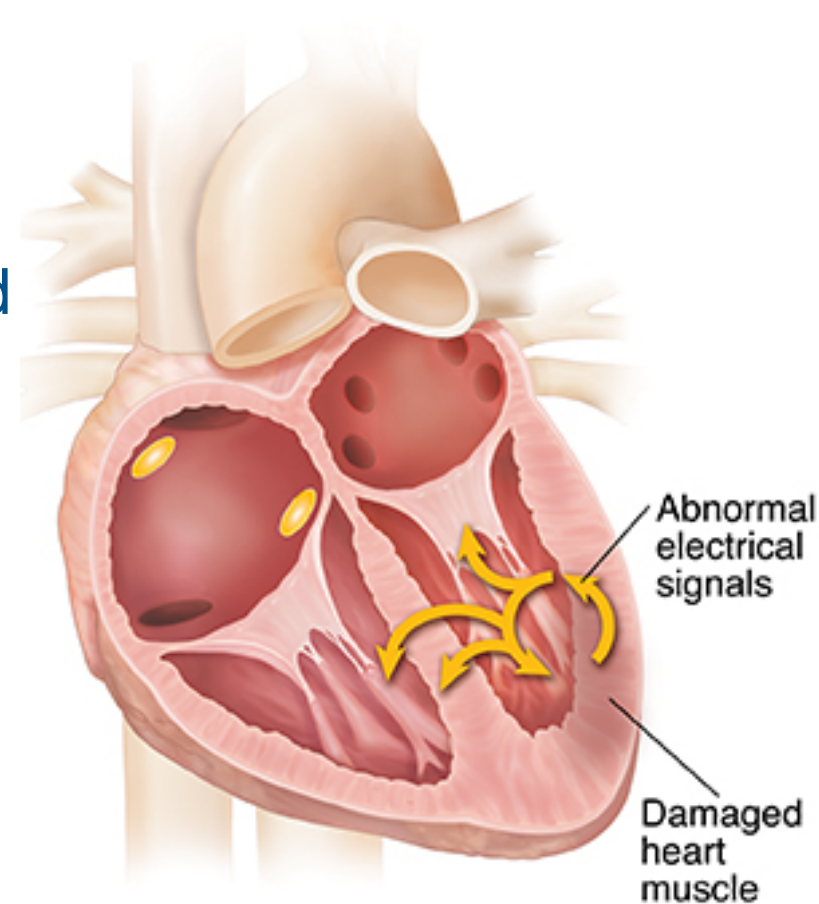
Arrhythmias

- ▶ 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>



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



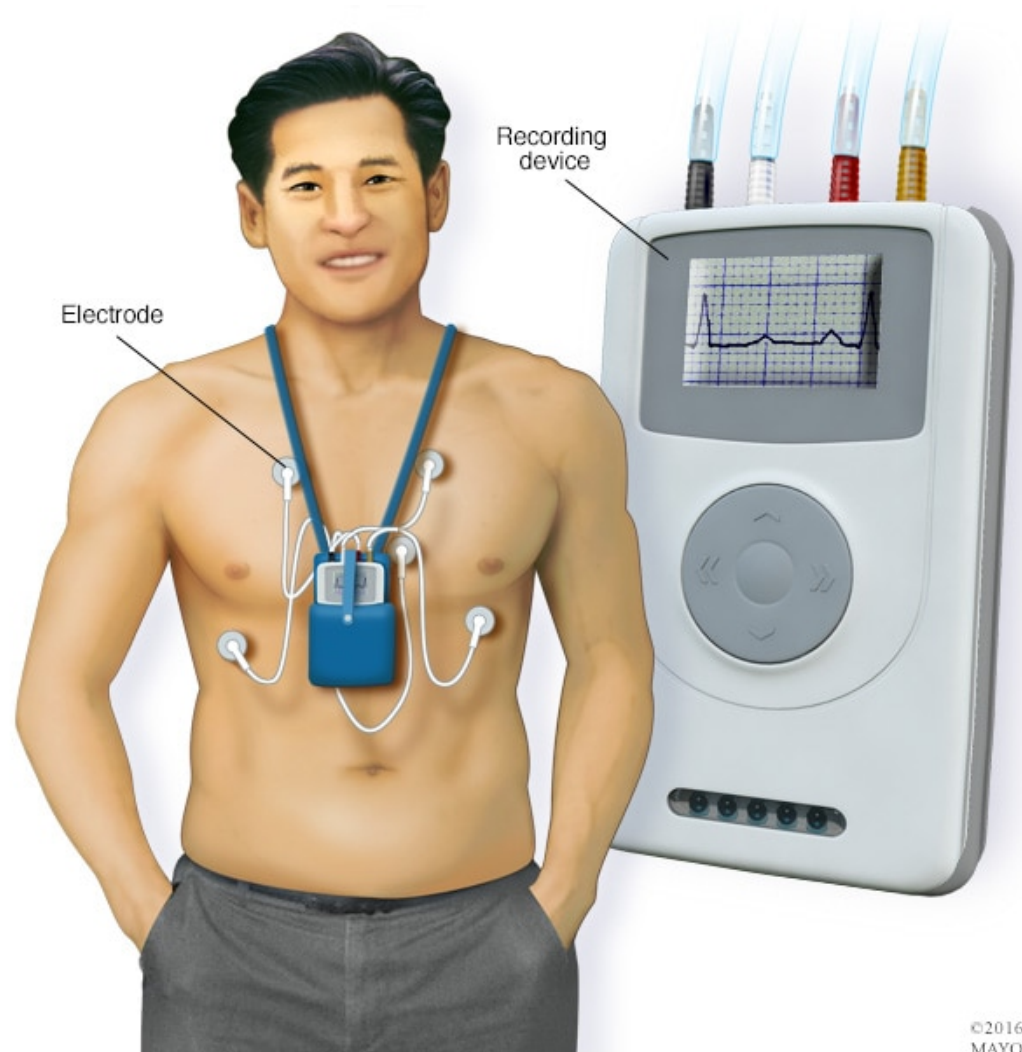
Symptoms of Arrhythmia

- ▶ 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)

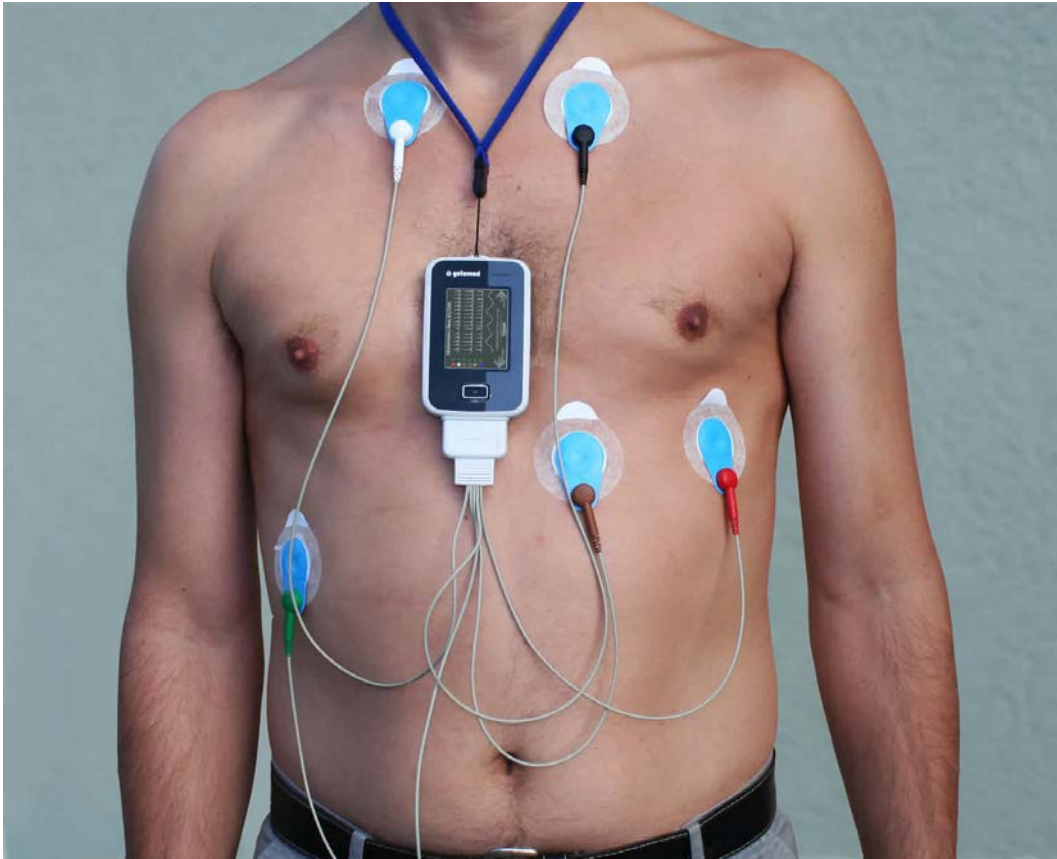


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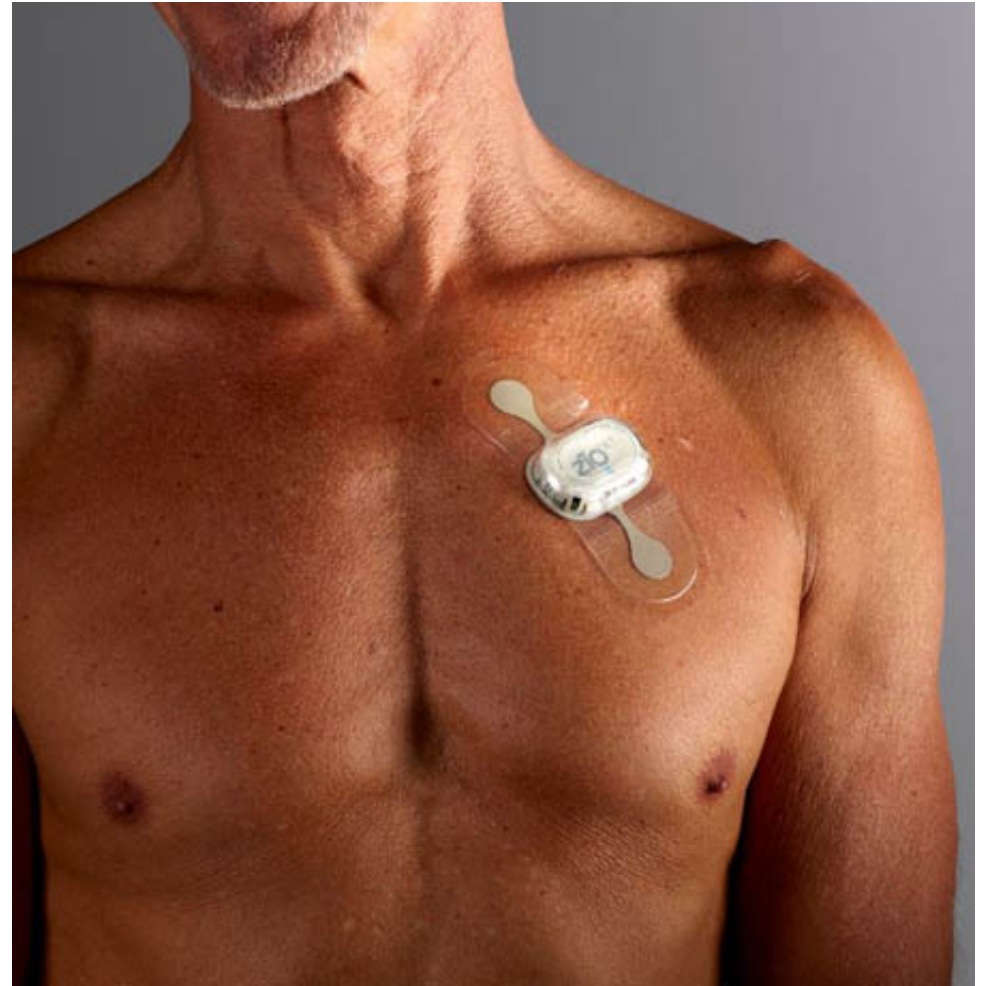
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Newer Ambulatory Monitoring



Newer Ambulatory Monitors

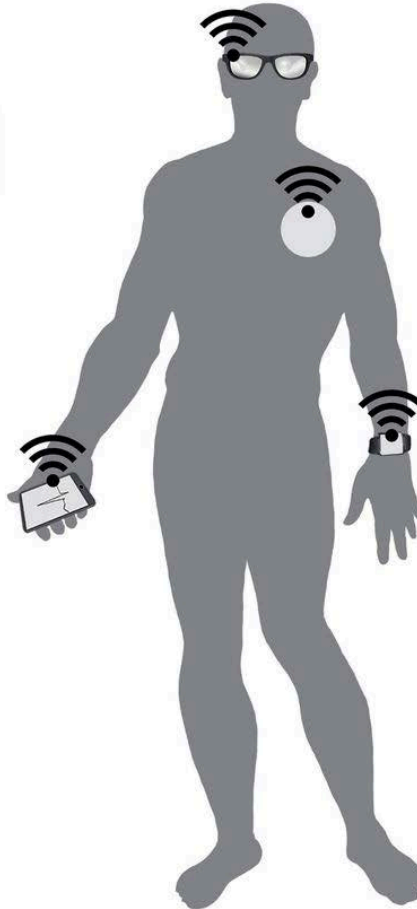


Wearables

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

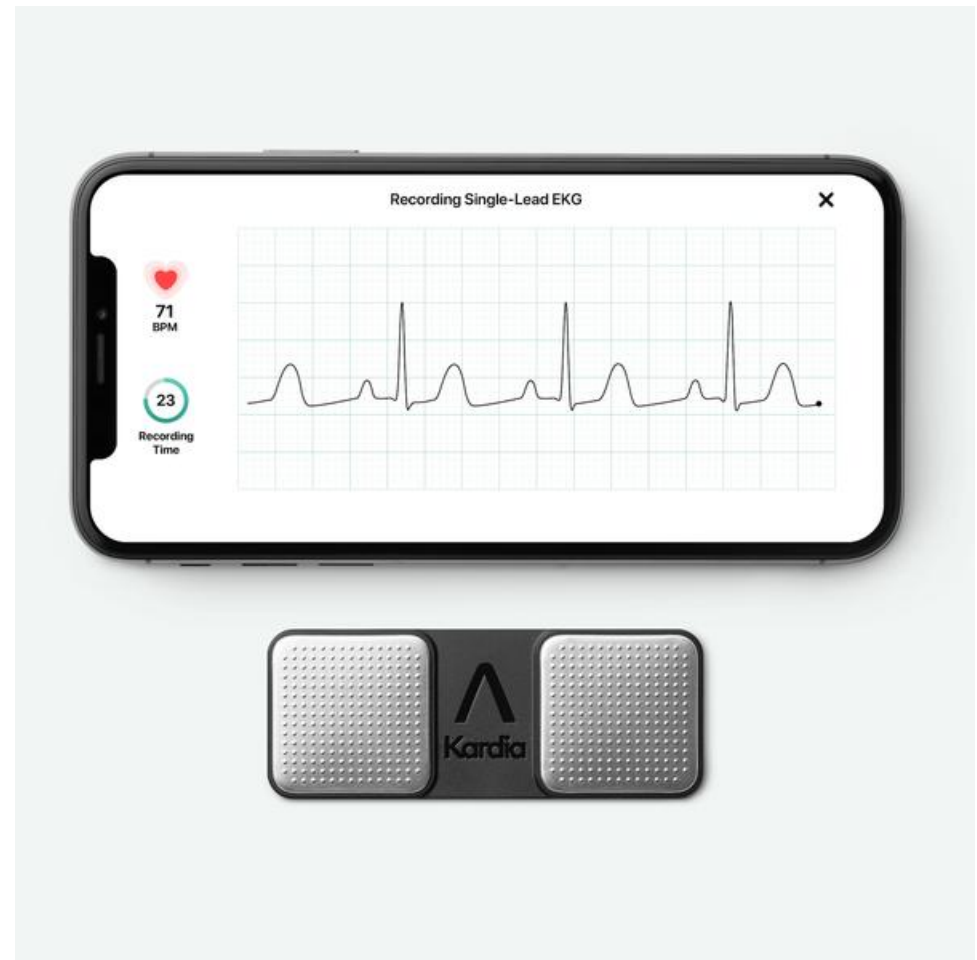
- Wristwatches
- Smartphones
- Patches
- Headbands
- Eye-glasses
- Necklaces

Sana, F. et al. J Am Coll Cardiol. 2020;75(13):1582-92.



Wearables

- ▶ I-Watch
- ▶ Kardia
- ▶ Fitbit
- ▶ Oura Ring

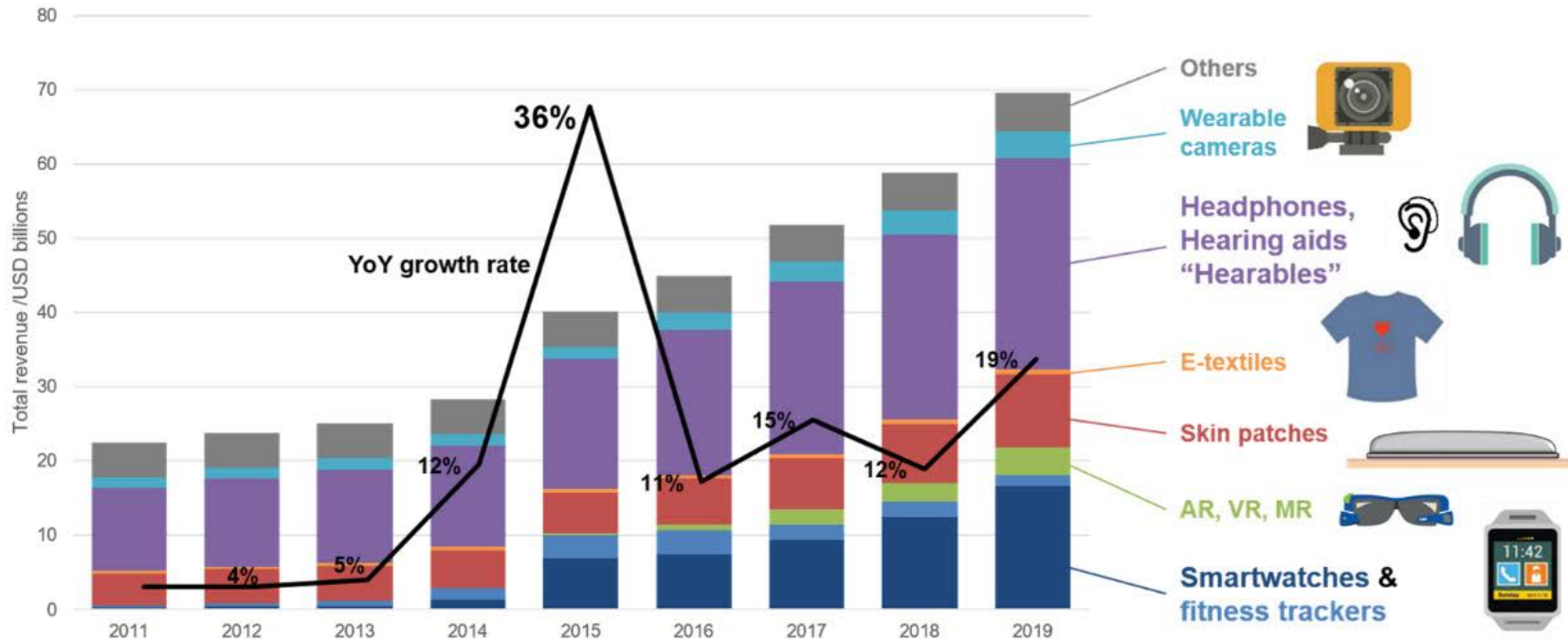


Wearables

Wearables: State of the industry

Annual revenue from wearable technology products

IDTechEx Research



Implantable Loop Recorder



Implantable Loop Recorder

BioMonitor 2 (Biotronik SE & Co, Berlin, Germany)



Reveal LINQ (Medtronic, Minneapolis, USA)



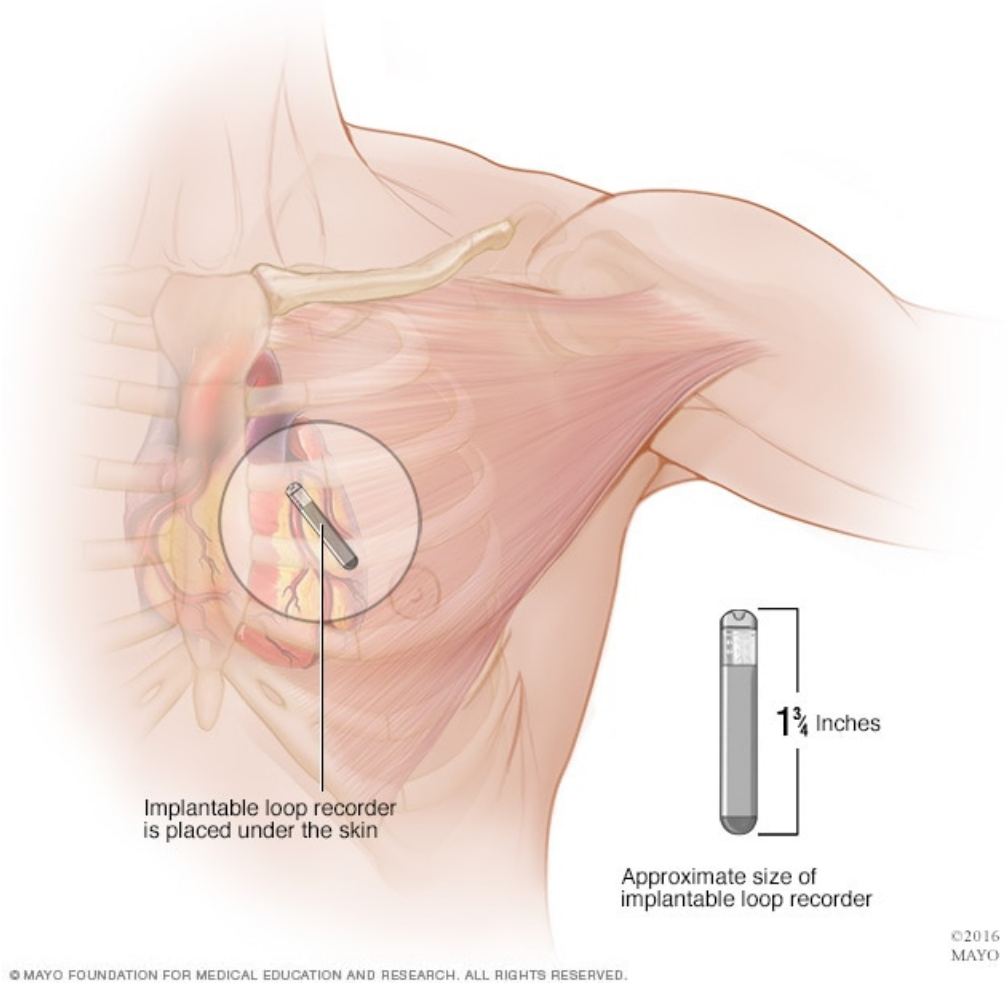
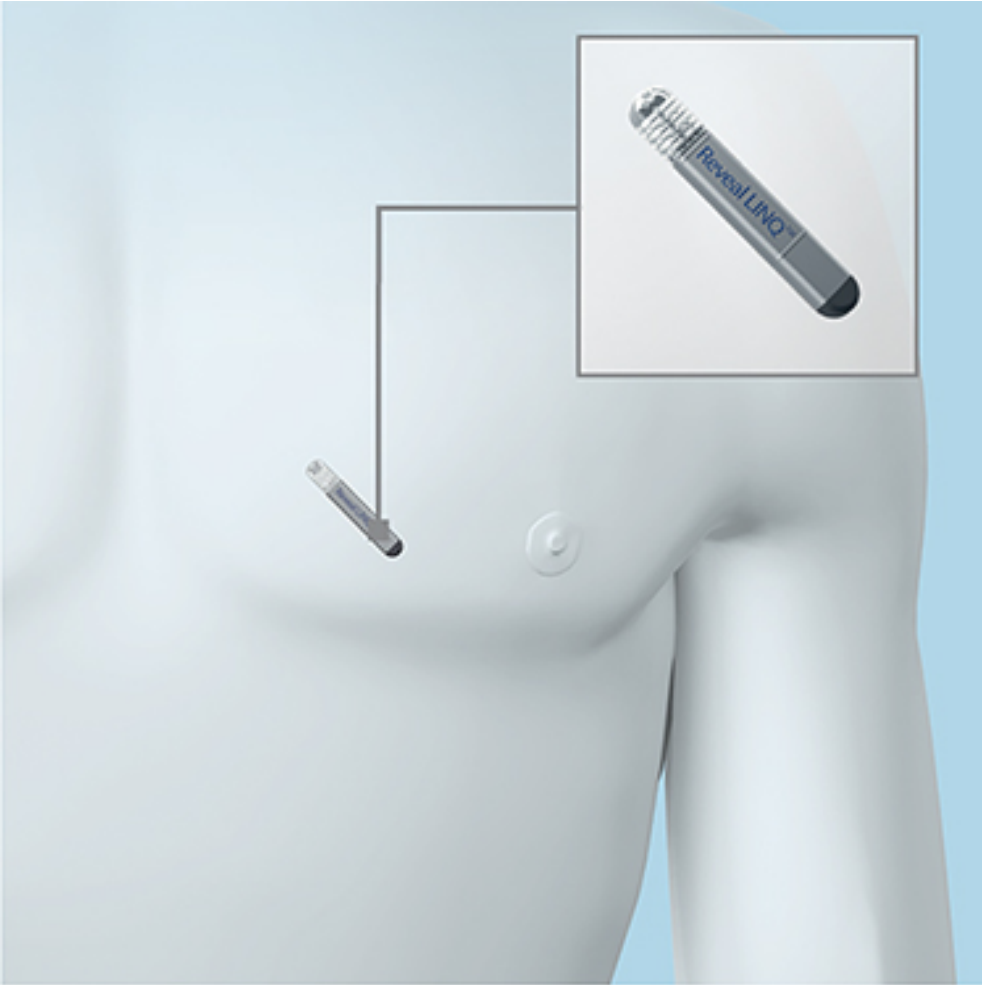
Reveal XT (Medtronic, Minneapolis, USA)



Confirm Rx™ ICM (St Jude Medical, Minnesota, USA)



Implantable Loop Recorder

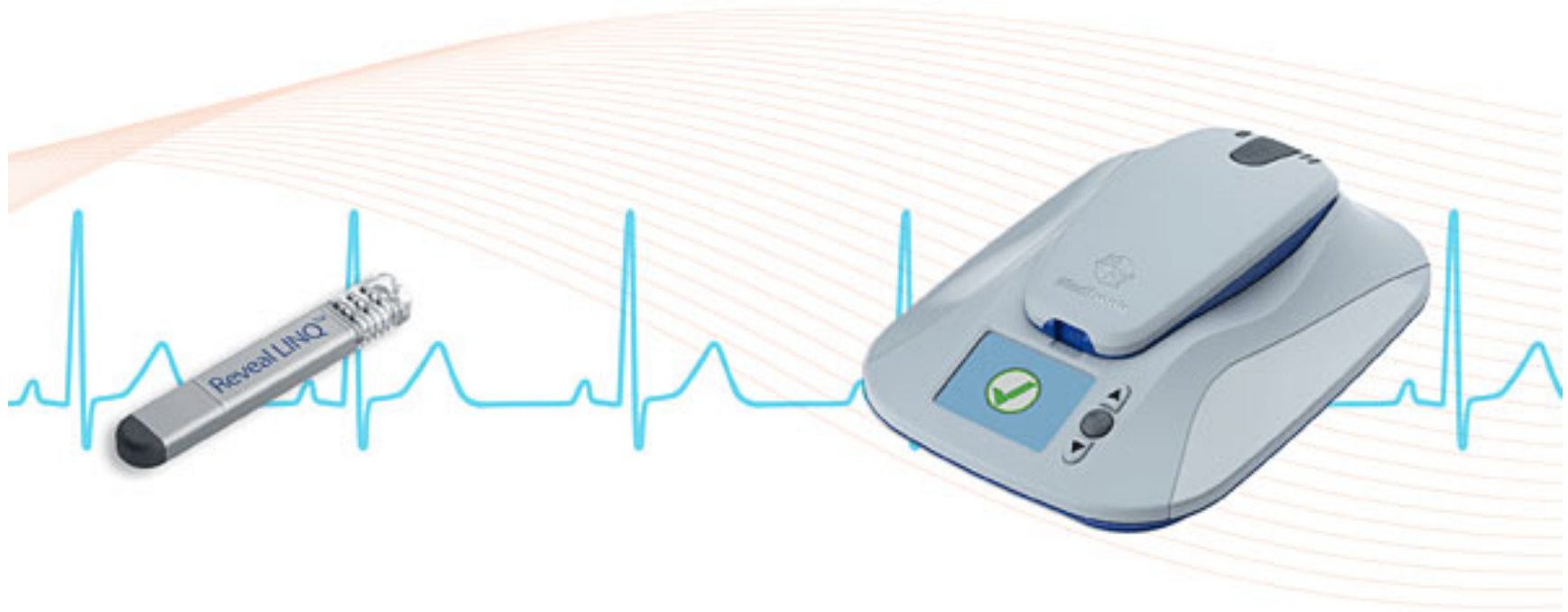


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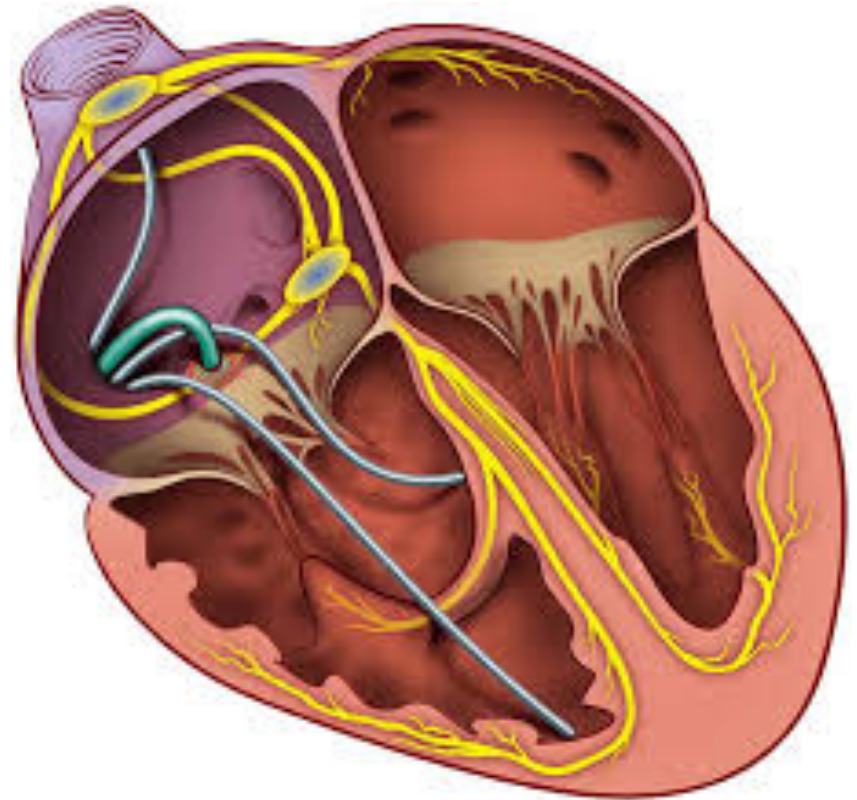


Implantable Loop Recorder



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)

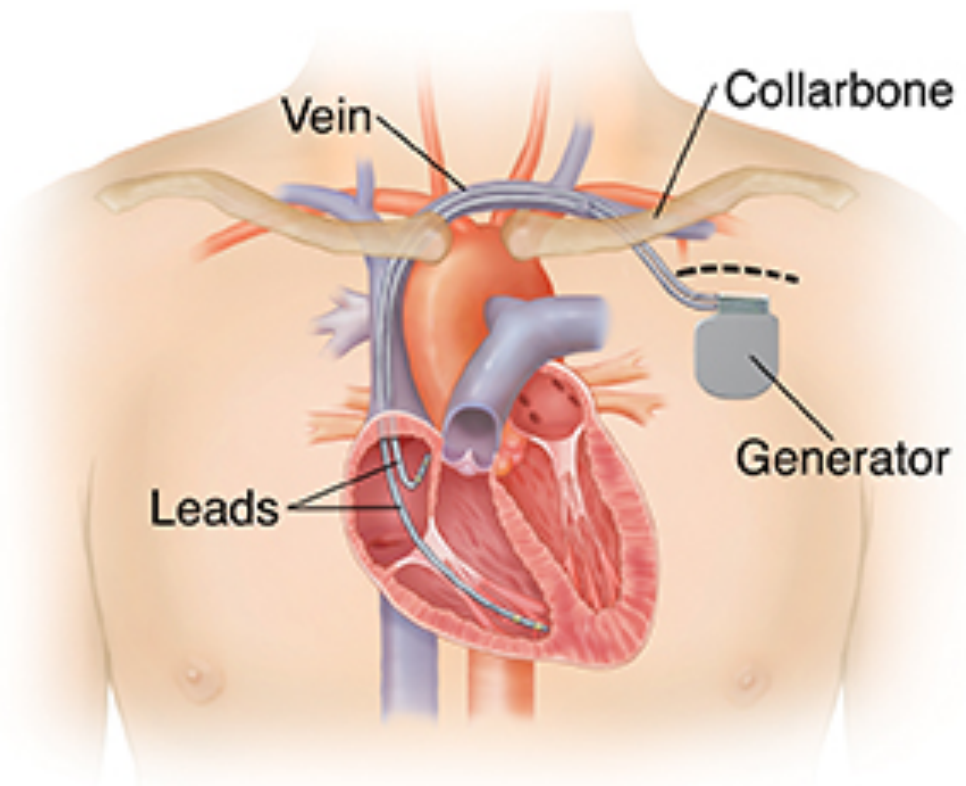
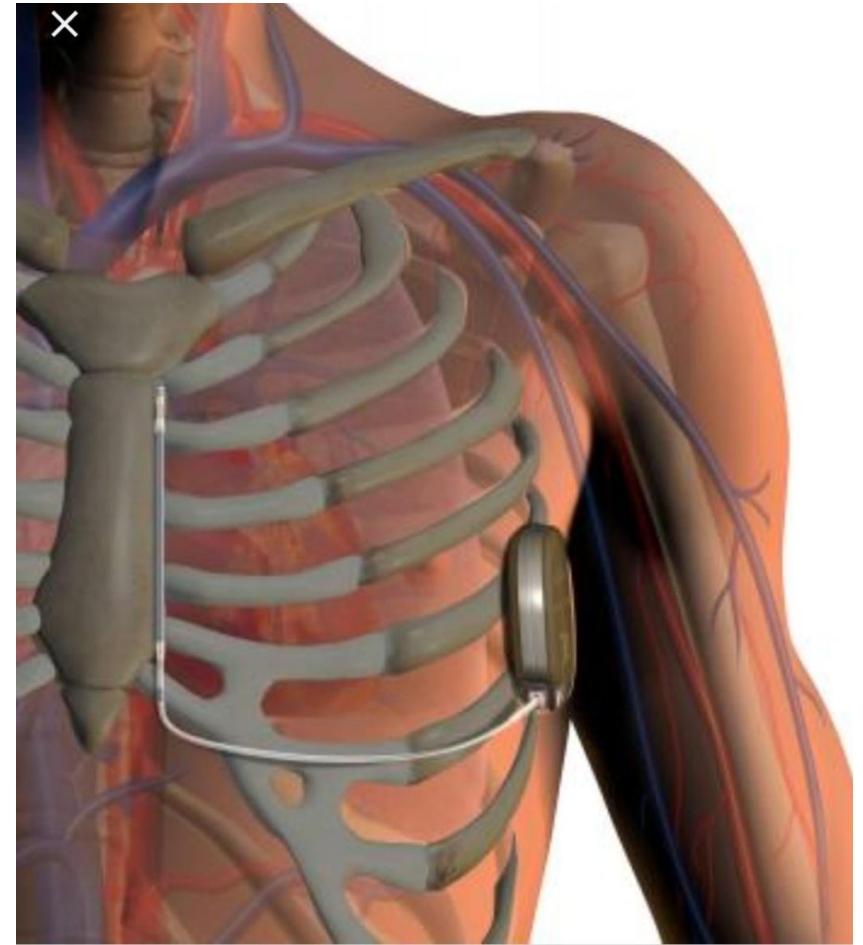
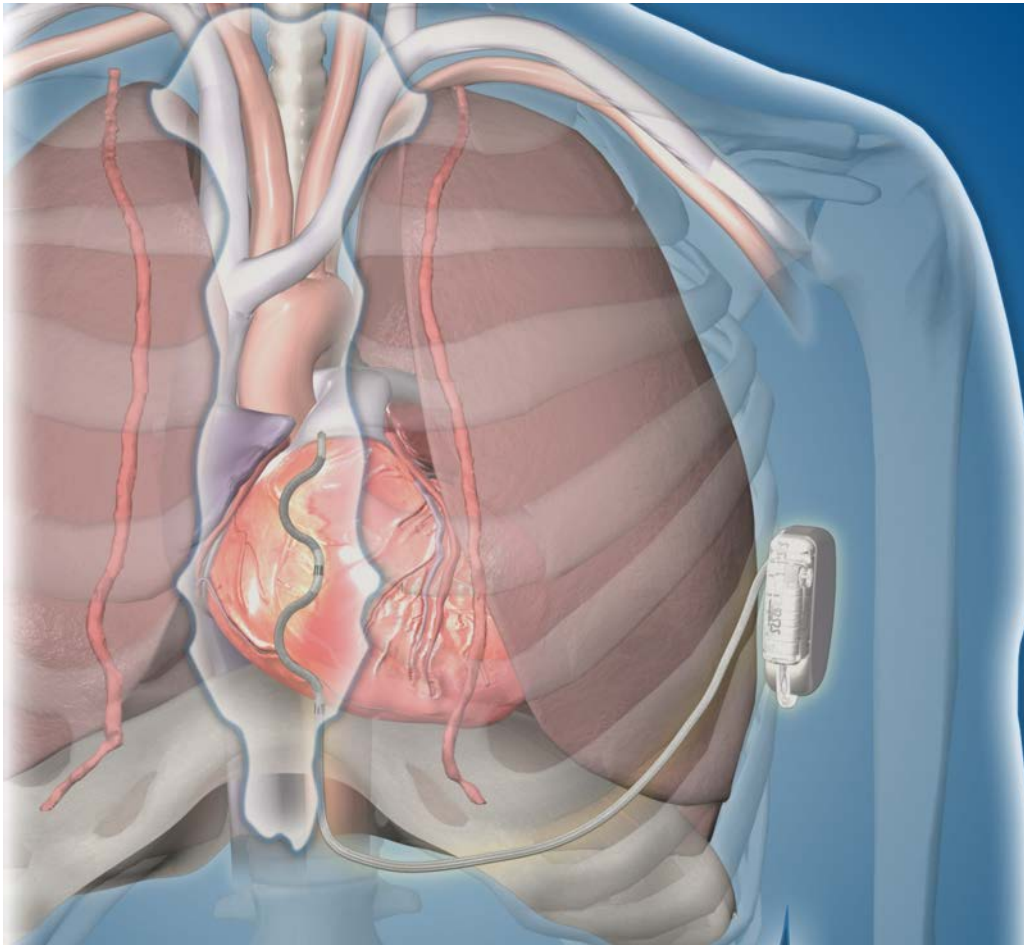


Figure 1. Examples of ICDs (pictures are not to scale).



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?

