The Electrocardiogram in Adult Congenital Heart Disease

I shall focus on five aspects of this topic:

1) History of the electrocardiogram.
2) Vectorcardiography
3) Unusual or unfamiliar forms of common arrhythmias.
4) The Long QT Interval
5) The Signal Averaged Electrocardiogram (SAECG)
The History of Electrocardiography

Many brilliant minds contributed to the development of electrocardiography as a clinical science. The early history (1900-1945) was dominated by Professor Willem Einthoven in the Netherlands, Sir Thomas Lewis in England and Frank N. Wilson in the United States. These pioneers laid the foundation for modern electrocardiography.
Professor Willem Einthoven

The 1924 Nobel Prize in Physiology or Medicine. Awarded to Willem Einthoven "for his discovery of the mechanism of the electrocardiogram".
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Sir Thomas Lewis

Cardiologist and Clinical Scientist
Established atrial fibrillation as a clinical
Frank N. Wilson

Introduced the modern era of electrocardiography. He demonstrated negativity of the ventricular cavity and activation from endocardium to epicardium.
Vectorcardiography

Spatial Vector Electrocardiography: Clinical Electrocardiographic Interpretation.
Robert Purves Grant & E. Harvey Estes
Unfamiliar or Unusual Forms of Common Arrhythmias

Sinus Arrhythmia
(Heart Rate Variability)
Sinus arrhythmia can only occur if the atrial septum is intact. It is minimal or absent with an atrial septal defect.
Atrial Septal Defect
No Sinus Arrhythmia

Normal Sinus Rhythm
Atrial Septal Defect Closed
Sinus Arrhythmia
Long QT Interval
Lange-Nielsen Syndrome

An autosomal recessive disorder characterized by congenital deafness, QT interval prolongation, ventricular tachycardia, syncope, and sudden death.
Dalmatian Coach Hounds May Look Alike

But some dogs drop dead on the hunt because of canine Long QT Lange-Nielson Syndrome.
Coupled Babies
Coupled Rhythms
The Signal Averaged Electrocardiogram

In the 1970’s, Michael B. Simson, a cardiology fellow at the University of Pennsylvania, developed the signal averaged electrocardiogram to detect the slow conduction substrate of reentry.
Signal Averaged Electrocardiogram

The SAECG identifies low-amplitude potentials (curved arrow below) at the end of the QRS complex. Late potentials represent delayed ventricular activation and increased risk of reentrant monomorphomorphic ventricular tachycardia.
The Signal Averaged Electrocardiogram

Ventricular Tachycardia

Electrophysiologic mechanisms of ventricular tachycardia:

_reentry, automaticity, triggered activity._

Inducible sustained ventricular tachycardia is typically _reentry & monomorphic:_

![Ventricular Tachycardia ECG](image)
Reentrant substrates remain dormant unless activated (triggered). The overt expression of reentrant MVT requires both a substrate and a trigger. The signal averaged ECG detects the substrate. Severe pulmonary regurgitation serves as a trigger.
In patients with a positive SAECG, slow conduction reentrant substrates are usually along ventriculotomy scars.
Reentrant substrates can be eliminated by radiofrequency ablation or surgical revision of the ventriculotomy scar.
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Thank You