

# *The Electrocardiogram in Adult Congenital Heart Disease*

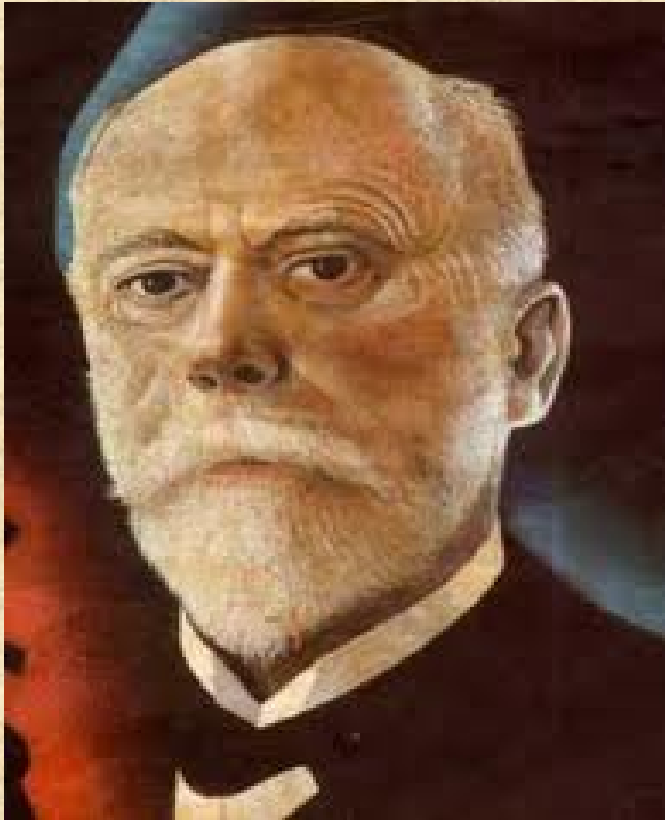
I shall focus on five aspects of this topic:

- 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*

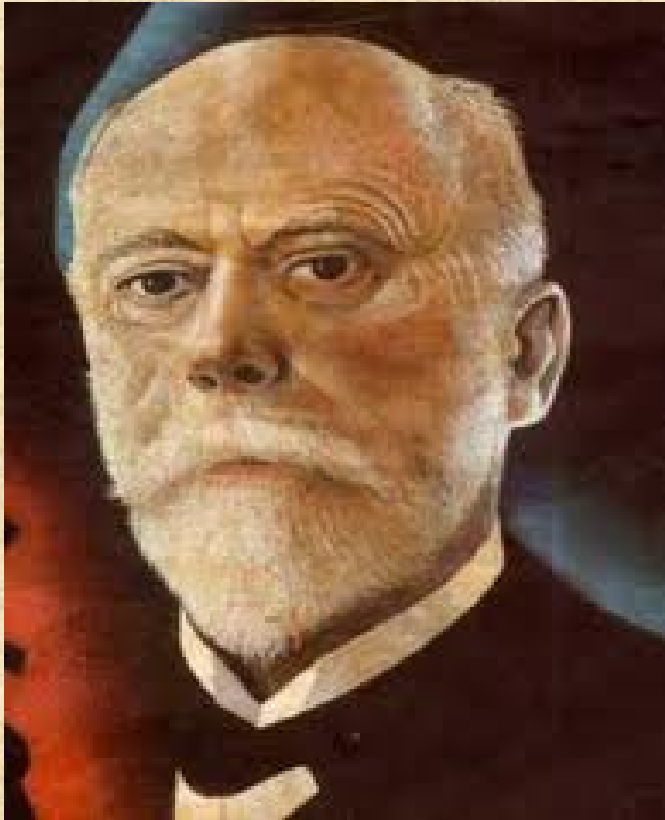
Many brilliant minds contributed to the development of electrocardiography as a clinical science. The early history (1900-1945) was dominated by Professor Willem Einthoven in the Netherlands, Sir Thomas Lewis in England and Frank N. Wilson in the United States. These pioneers laid the foundation for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

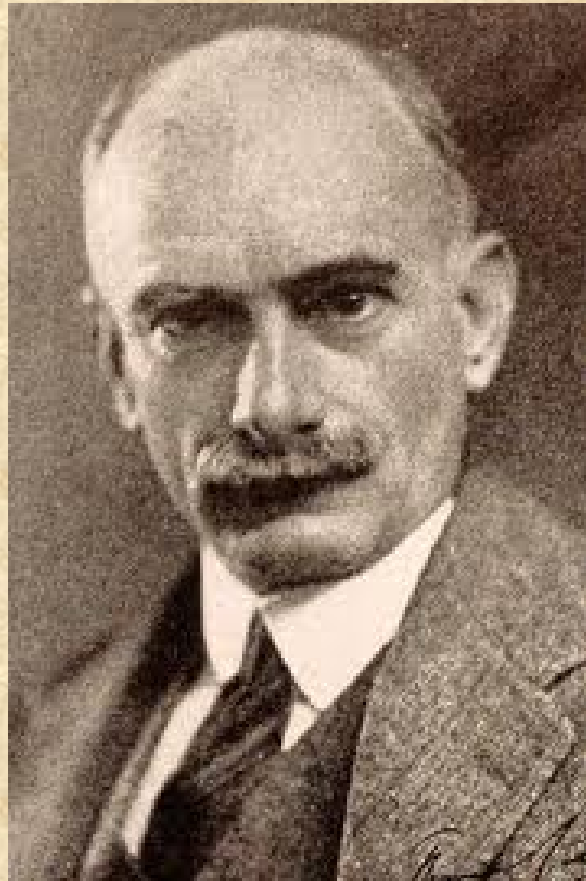
# Professor Willem Einthoven



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



# *Sir Thomas Lewis*



*Cardiologist and Clinical Scientist*

Established atrial fibrillation as a clinical

# *Frank N. Wilson*

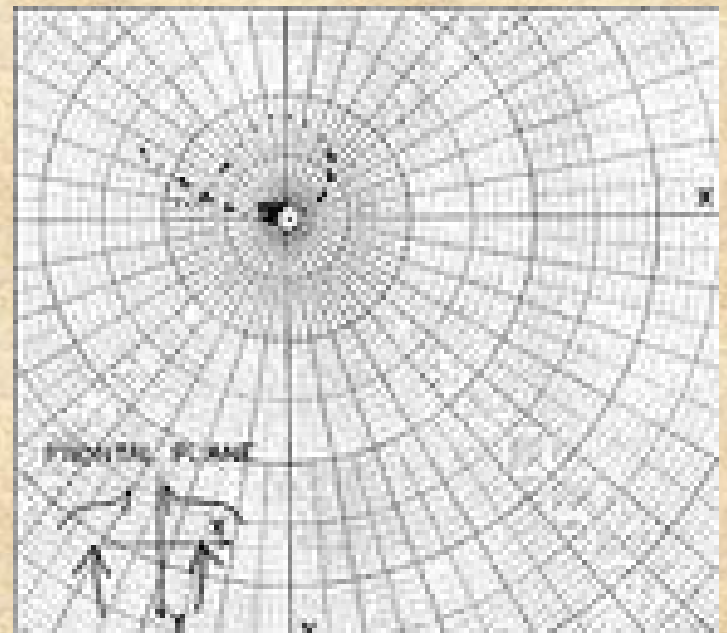
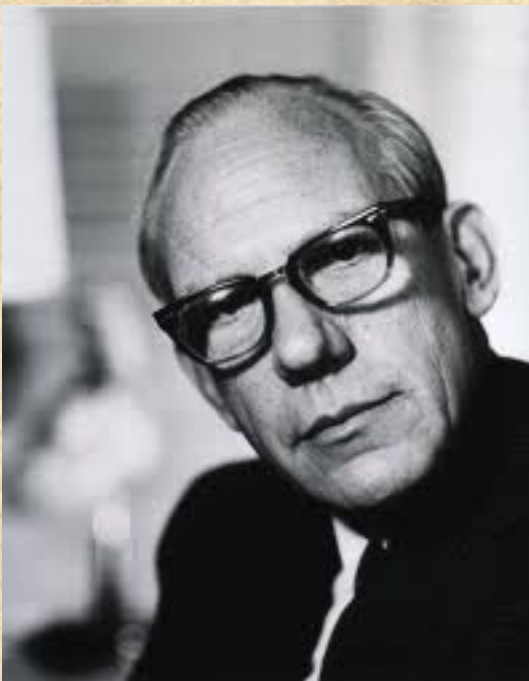


Introduced the modern era of electrocardiography.  
He demonstrated negativity of the ventricular cavity  
and activation from endocardium to epicardium.

# *Vectorcardiography*

Spatial Vector Electrocardiography: Clinical  
Electrocardiographic Interpretation.

Robert Purves Grant & E. Harvey Estes



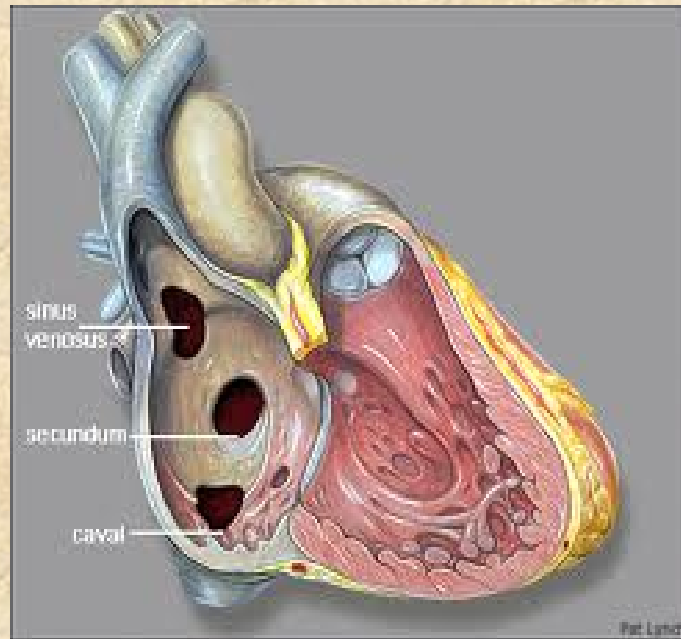
# *Unfamiliar or Unusual Forms of Common Arrhythmias*

*Sinus Arrhythmia  
(Heart Rate Variability)*



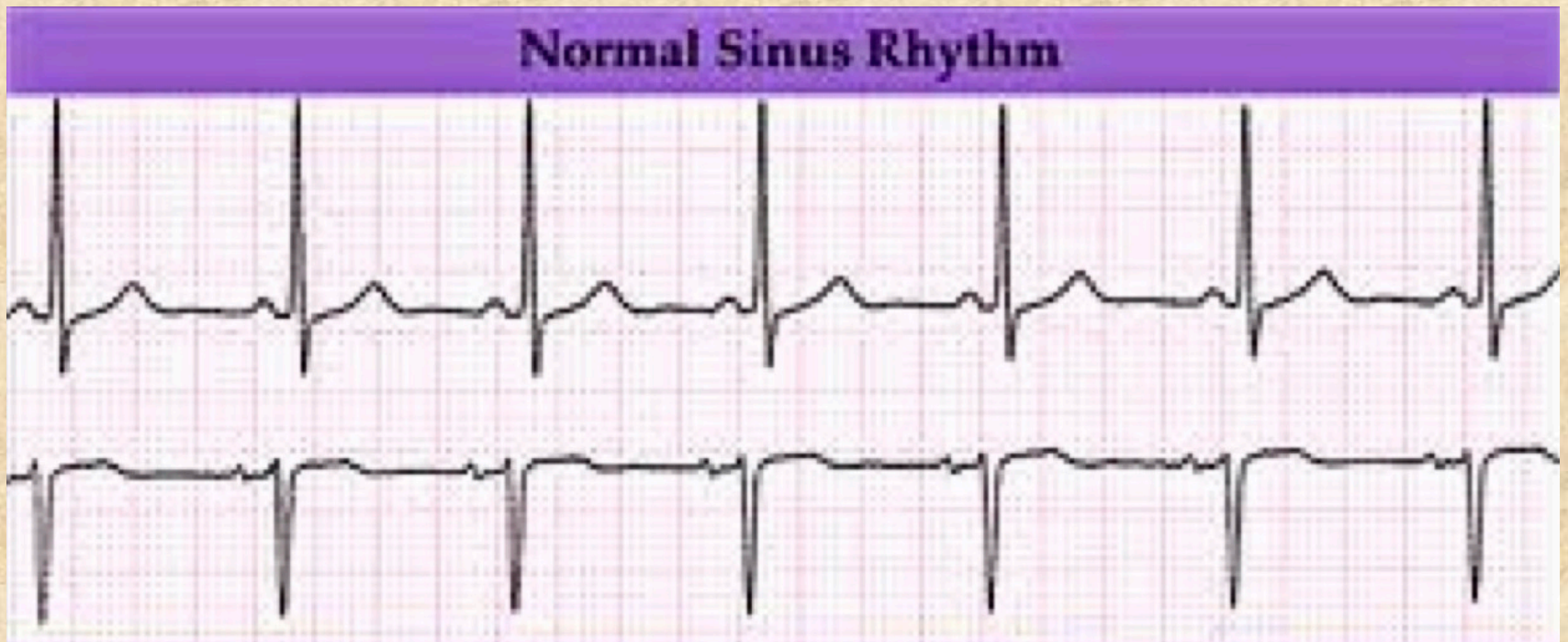
# *Sinus Arrhythmia*

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*

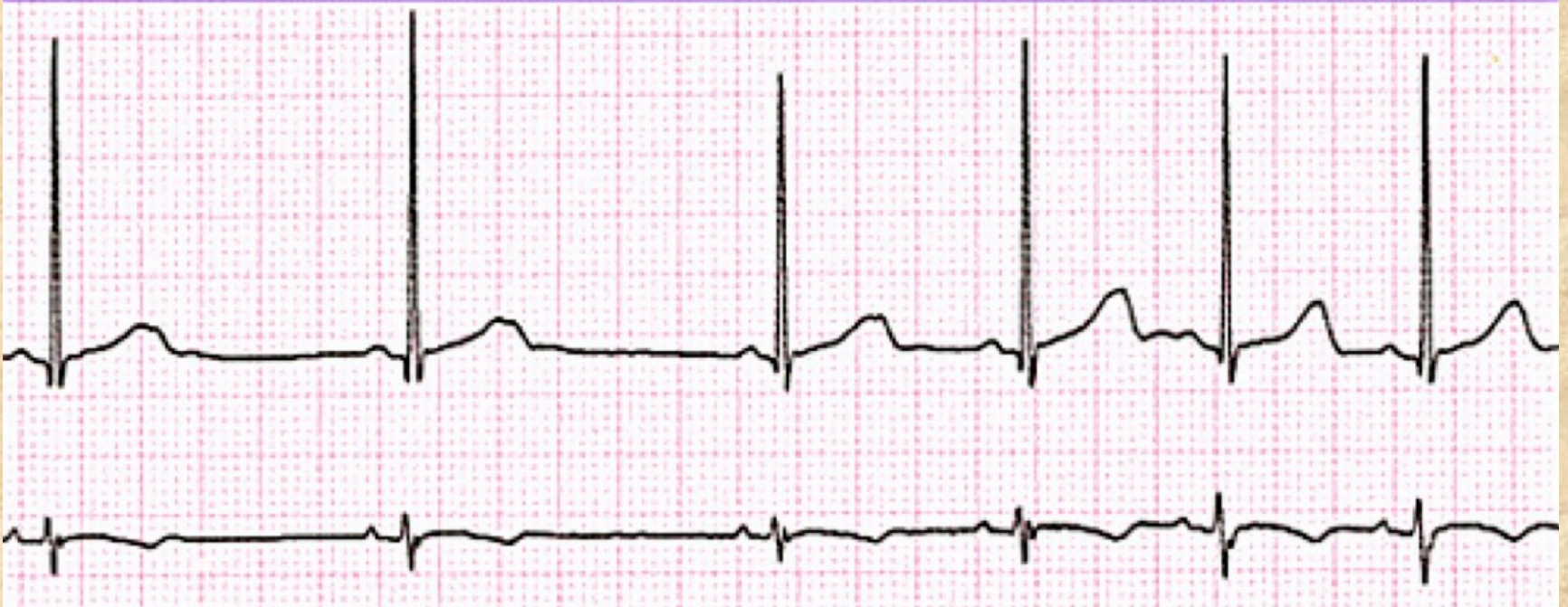




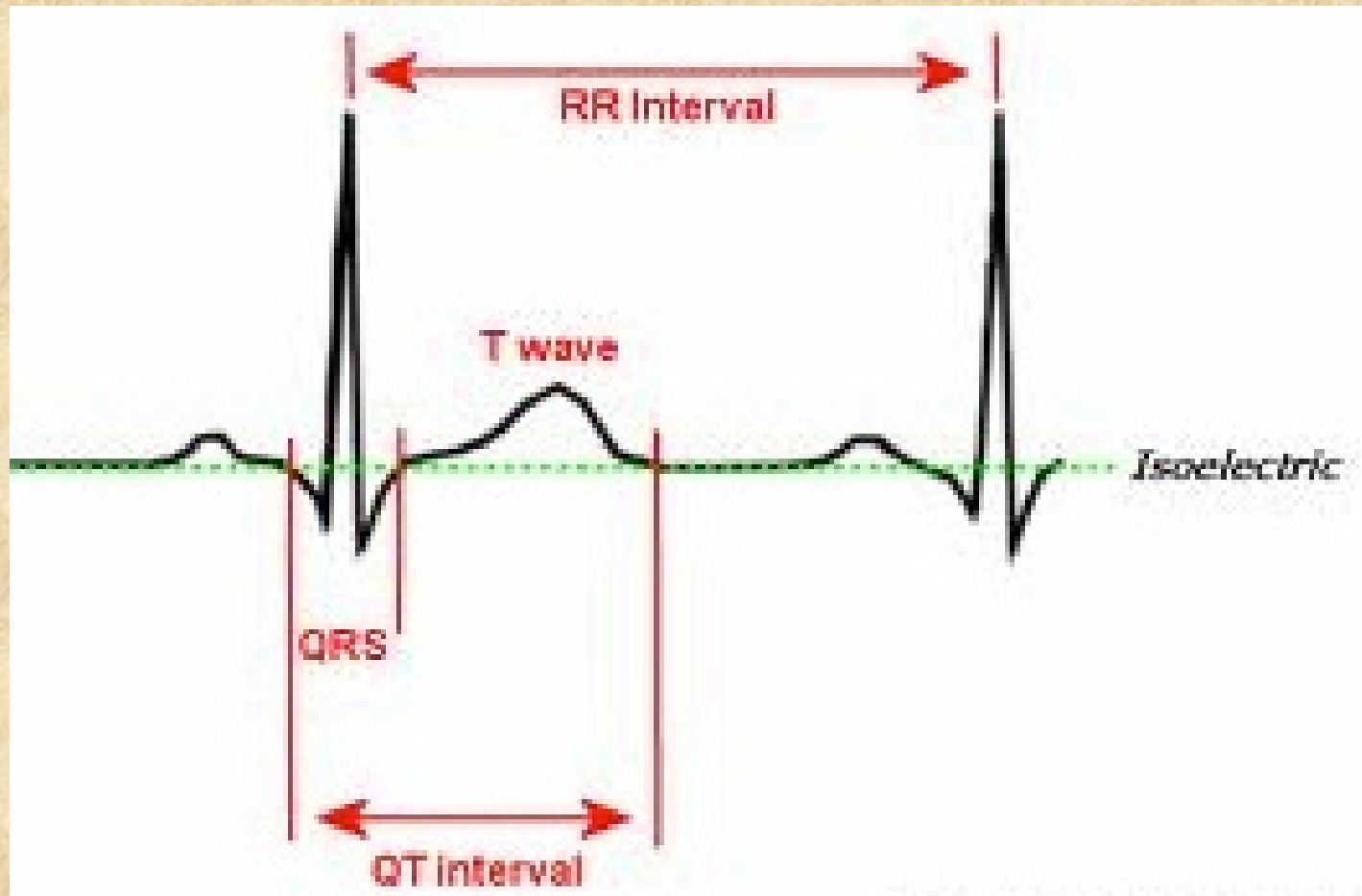
# *Atrial Septal Defect Closed*

## *Sinus Arrhythmia*

**Sinus Arrhythmia**



# *Long QT Interval*





## *Lange-Nielson Syndrome*

An autosomal recessive disorder characterized by congenital deafness, QT interval prolongation, ventricular tachycardia, syncope, and sudden death.

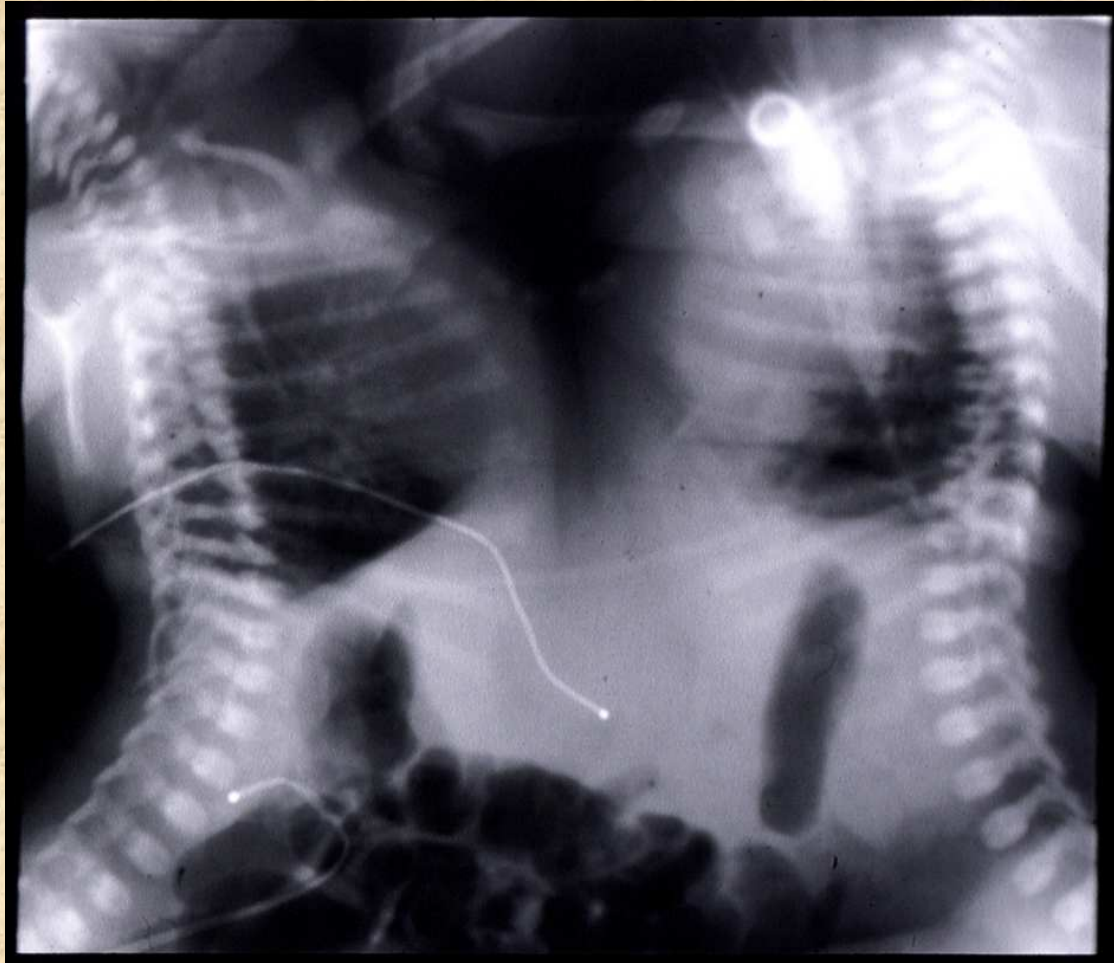


# *Dalmatian Coach Hounds May Look Alike*



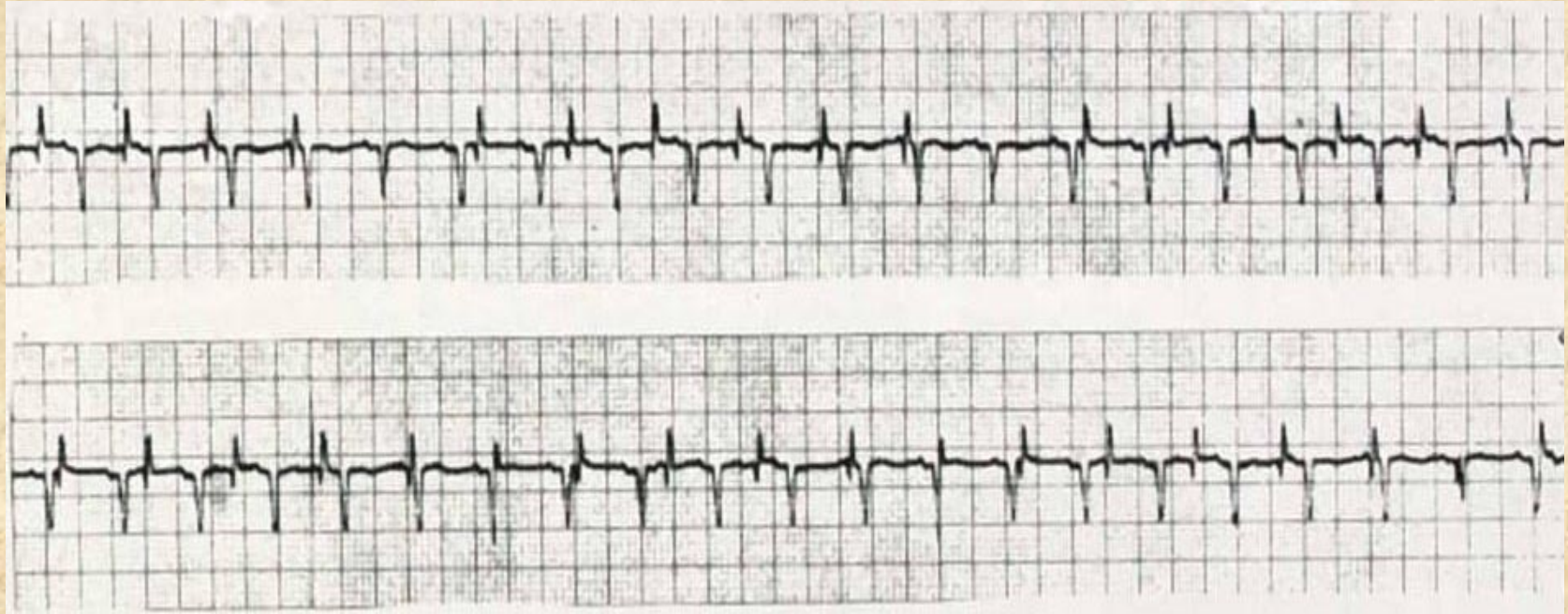
But some dogs drop dead on the hunt because  
of canine *Long QT Lange-Nielson Syndrome*.

# *Coupled Babies*





# *Coupled Rhythms*

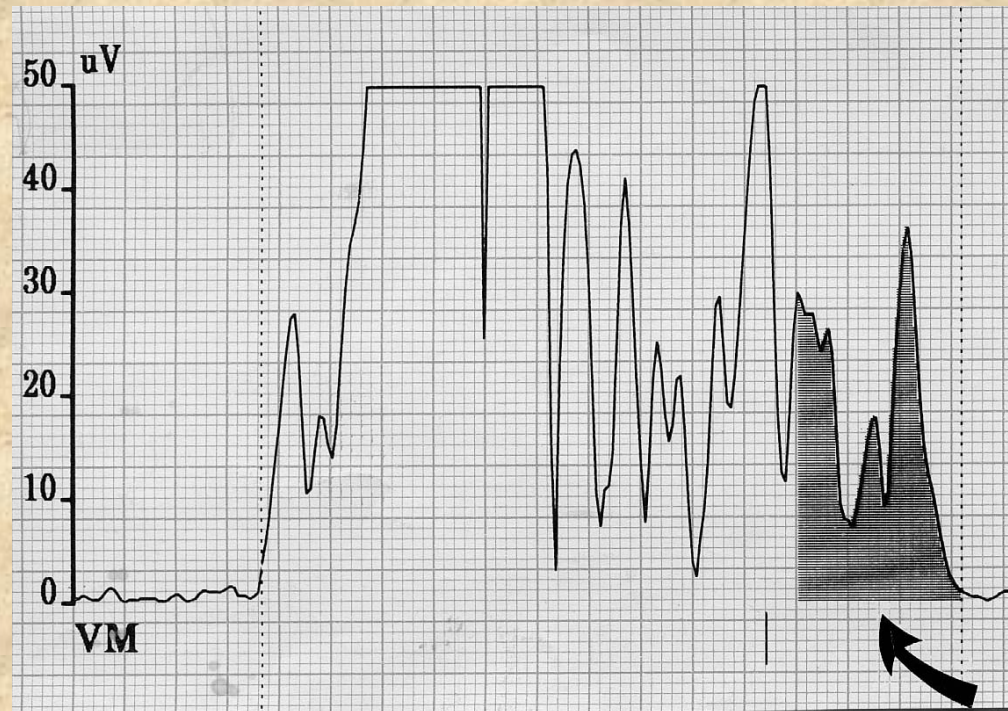


# *The Signal Averaged Electrocardiogram*

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 monomorphic ventricular tachycardia.





# *The Signal Averaged Electrocardiogram*

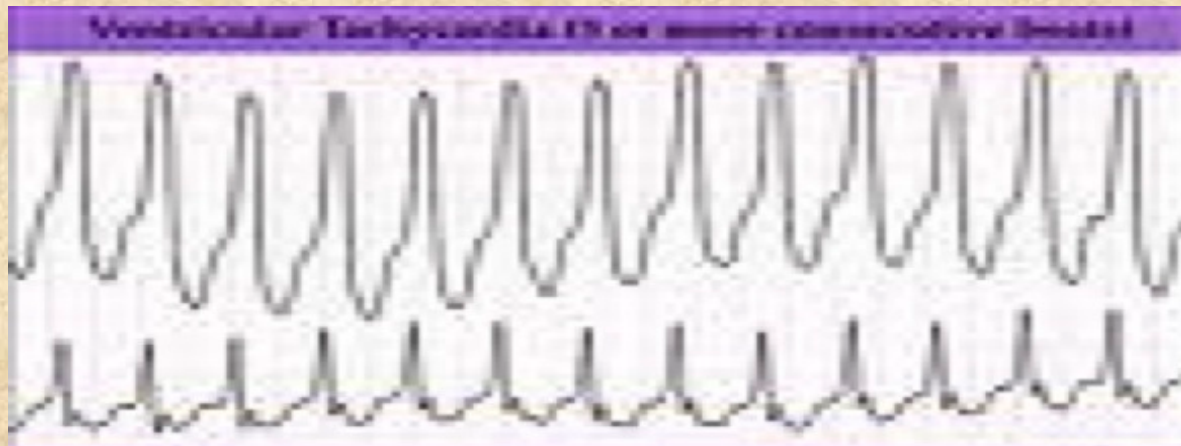
**Perloff JK, Middlekauf HR, Stevenson WG.  
The Post-ventriculotomy Signal Averaged  
Electrocardiogram in Congenital Heart Disease.  
Am J Cardiol 2006;98:1646-1651.**

# *Ventricular Tachycardia*

Electrophysiologic mechanisms of  
ventricular tachycardia:

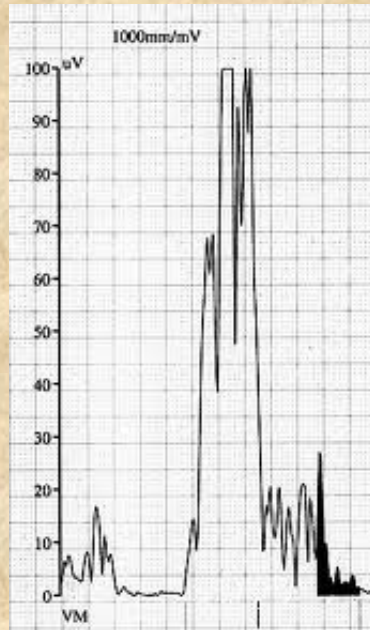
*reentry, automaticity, triggered activity.*

Inducible sustained ventricular tachycardia is  
typically *reentry* & *monomorphic*:

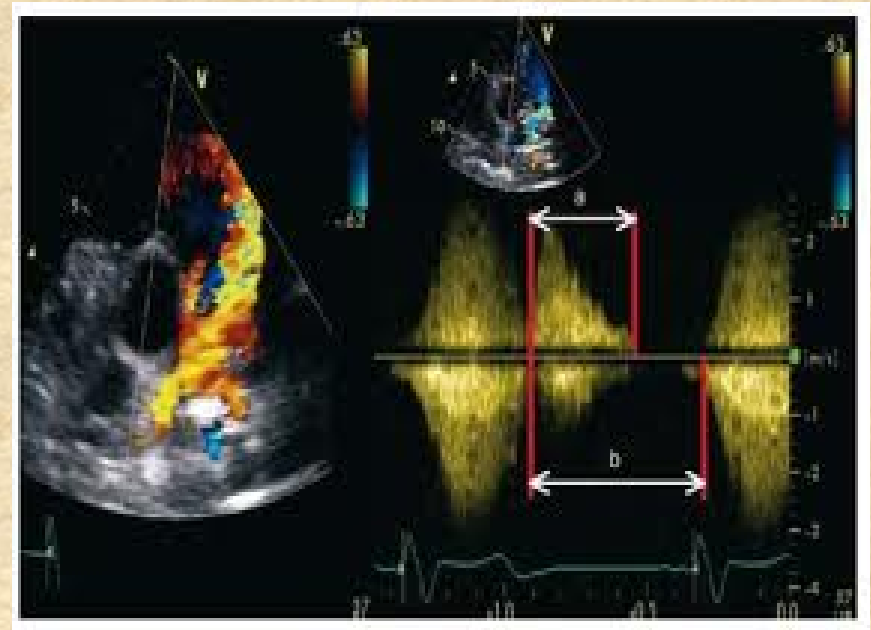


# Substrate Plus Trigger

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.

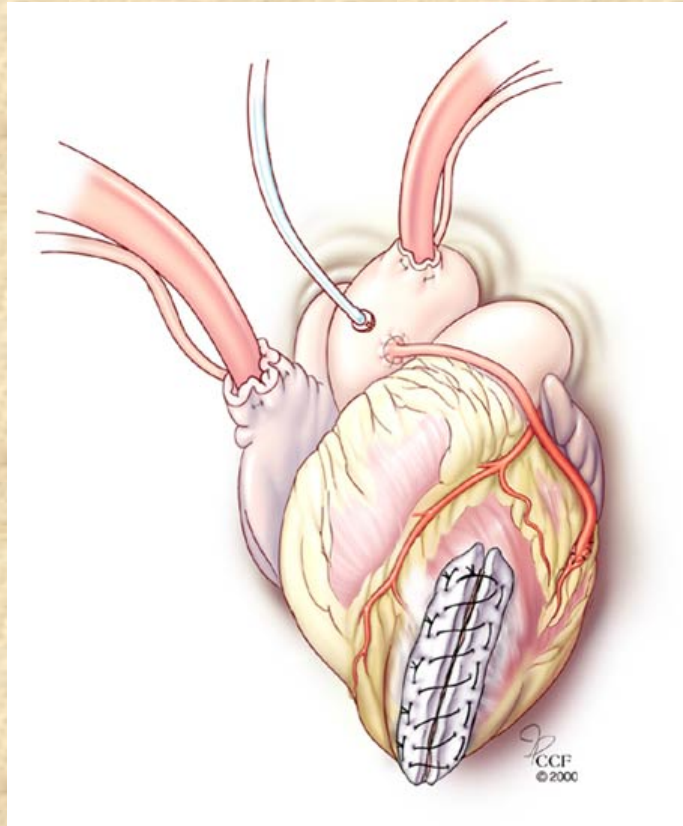


Substrate



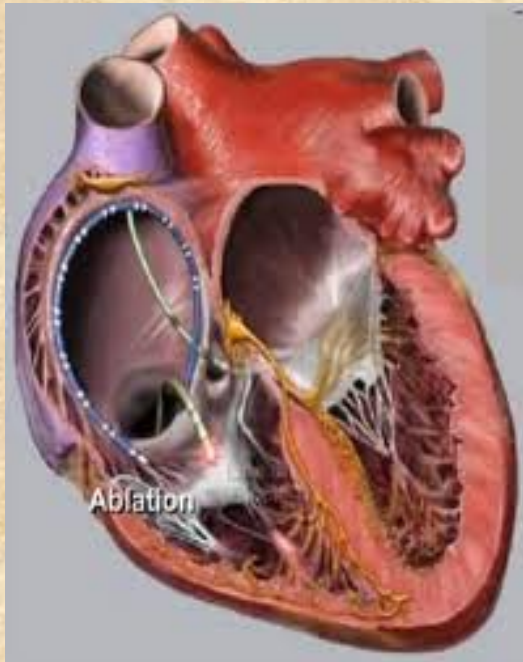
Trigger

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.



Ablation



Revision



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*Thank You*