Case Report:
A Family with Brugada Syndrome

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Nothing to Disclose
Proband

• 39 year old man referred for an abnormal EKG
• No cardiac risk factors, no significant medical history, no palpitations or syncope.
• Father died of CHF at young age with multiple myeloma. Mother AAW age 75. Two brothers and one sister AAW. No family h/o SCD.
• BP 140/98 HR 58 Lungs clear, RRR S1 S2, no edema
Proband

- EKG

- Echo and MRI: Normal

- EP Study: Inducible VF, + procainamide challenge
**Proband**

- ICD placed for primary prevention of SCD.  
  - CORRECT?

- Genetic testing showed T353I mutation in SCN5A that leads to decreased inward Na⁺ current
Pedigree
Son of Proband

  - CORRECT?
Son of Proband

  – CORRECT?
Sister of Proband

- 45 year old woman with chest pain. Echo and MRI normal. ICD offered but patient refused.
  – CORRECT?
Niece of Proband

- 22 year old woman, Von Willebrand syndrome and h/o palpitations, with aborted SCD watching horror movie. Recovered neuro function, ICD placed.
  – CORRECT?
Daughter of Proband

- 22 year old woman, asymptomatic, offered and accepted ICD. Received 2 shocks for SVT during argument.
  - CORRECT?
Summary

• Indications for ICD in Brugada syndrome
  – Aborted SCD
  – EKG pattern?
  – Inducible VF?
  – Family h/o SCD?
  – Gender?
  – Age?
  – Mutation?