## Case Discussion

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Warren (Sonny) Jackman
Heart Rhythm Institute
University of Oklahoma Health Sciences Center Oklahoma City, OK

## Disclosure

Consulting:

- Biosense Webster
- Boston Scientific
- Spectrum Dynamics
- VytronUS
- ACT

Lecture Honoraria:

- Biosense Webster
- Boston Scientific
- AtriCure
- Biotronik
- St Jude Medical

1. Relationship Between Accessory Pathways and Hypertrophic CM - PRK-AG2 Mutation
2. 2 Accessory Pathways - Differential Pacing
3. Left Atrial Appendage - LV Connections
4. Questions: Mapping Systems, Surgery

## PRK-AG2 Mutation

- Glycogen storage disease (increased AMP kinase activity)
- Autosomal dominant mutations of the ү2 regulatory subunit of AMP-activated protein kinase (modulates glucose uptake and glycolysis)
- Histology: myocytes are enlarged with vacuoles containing glycogen derivatives, but myocyte disarray (characteristic of HCM) is absent and interstitial fibrosis is minimal
- HCM, preexcitation, atrial fibrillation, progressive AV block

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6. R Posteroseptal AP

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## AHA 2002

## Atrial Appendage - Ventricle Connections

- Study Population (9 pts)
- 8 RAA-RV connections
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## Atrial Appendage - Ventricle Connections

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- No structural heart disease
- 1-3 prior failed ablation procedures


## Atrial Appendage - Ventricle Connections

## Endocardial Recordings

1. Absence of a distinct AP activation potential
2. The site of earliest activation was displaced away from the annulus
3. Earliest activation was recorded from the atrial appendage

## Atrial Appendage - Ventricle Connections

Catheter Ablation Results

- Elimination of AP conduction in 6 of 9 pts (Surgery in 3 pts - Tamponade in 1)
- Required separation of a segment of the atrial appendage attached to the ventricular connection
- Large number of RF applications

4-17 (median 8)



Pre-RF1
Pre-RF8
Post Ablation

$R A$
$\mathrm{~s}^{\prime}$

$\mathrm{RAA}_{p} \sqrt{A_{A}}$
RAA $_{d}$


RV



