ARRHYTHMIAS IN EBSTEIN ANOMALY

February 20, 2025

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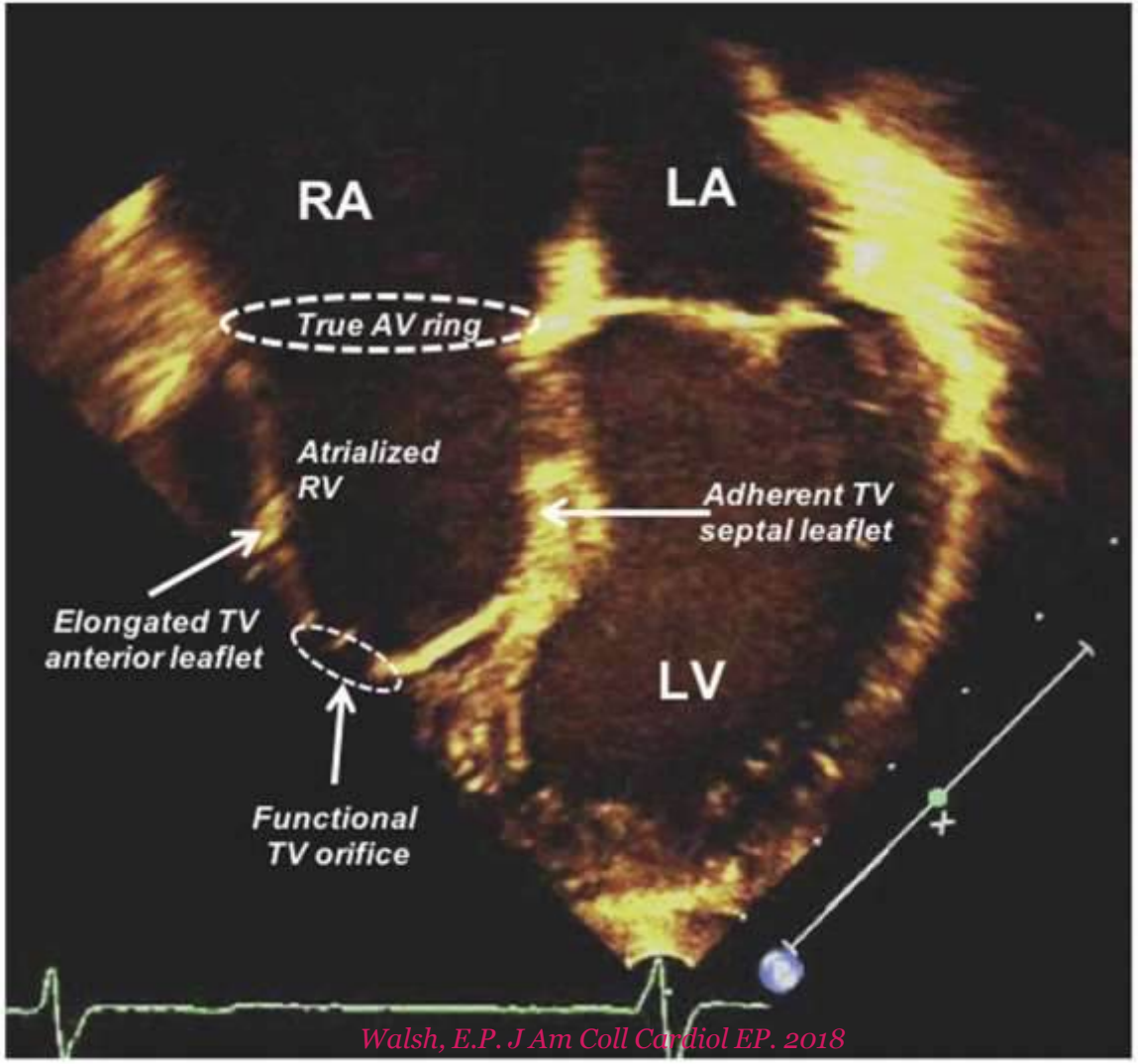
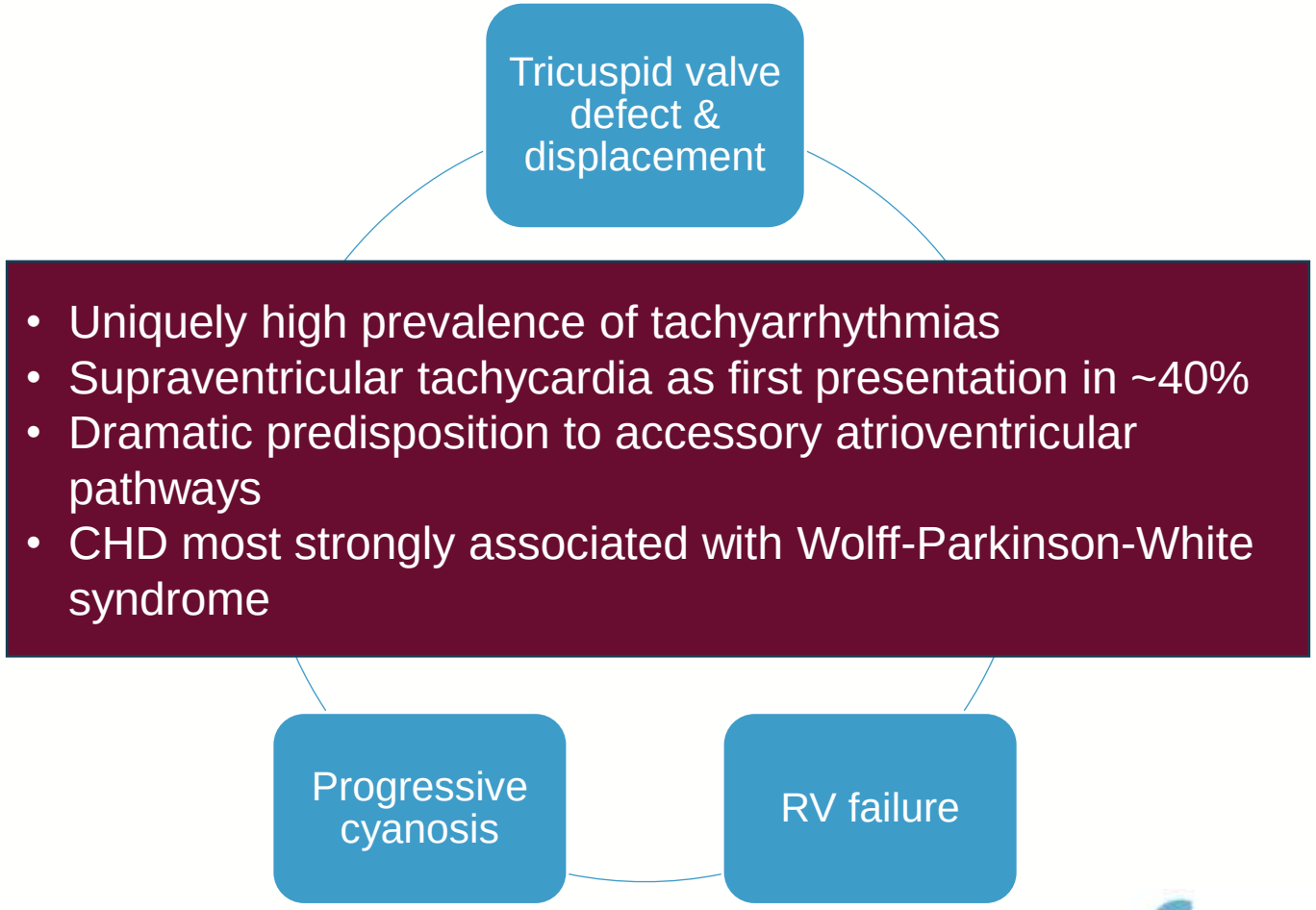
DISCLOSURES

- Research grant from Medtronic External Research Program



Walsh, E.P. J Am Coll Cardiol EP. 2018



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ARRHYTHMIAS IN EBSTEIN ANOMALY

Intrinsic substrate

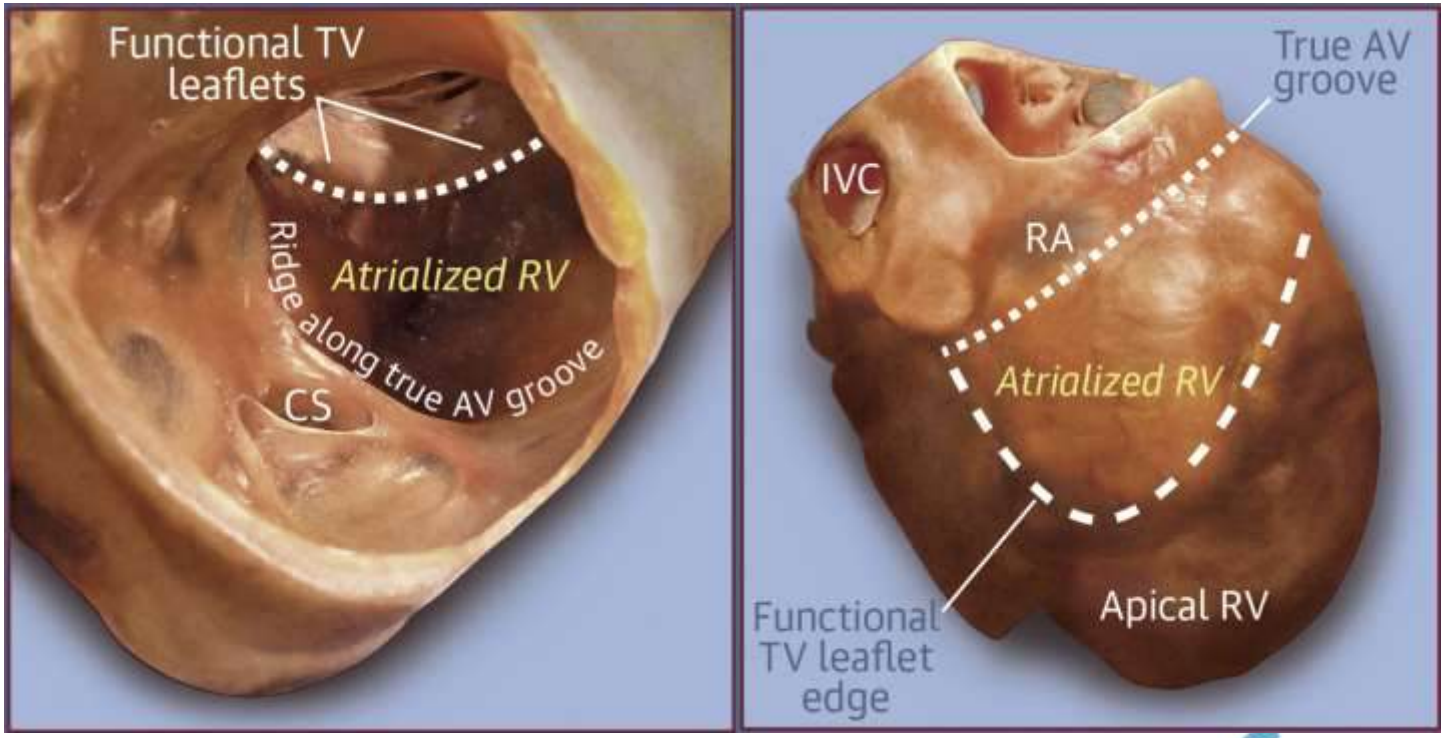
- AV accessory pathway mediated reentrant SVT: 10-38%
- Atriofascicular pathways: reentrant SVT, preexcited SVT: 5%
- AV nodal reentry: 8-13%
- Monomorphic VT from atrialized RV, possibly macro-reentry: Rare

Acquired substrate

- Atrial macroreentry (“atrial flutter”) >20%
- Focal atrial tachycardia: 2-20%
- Atrial fibrillation: Rare
- Polymorphic VT: Rare

Sudden arrhythmic death (any mechanism): 8-16%

ANATOMY OF EBSTEIN MALFORMATION OF TRICUSPID VALVE



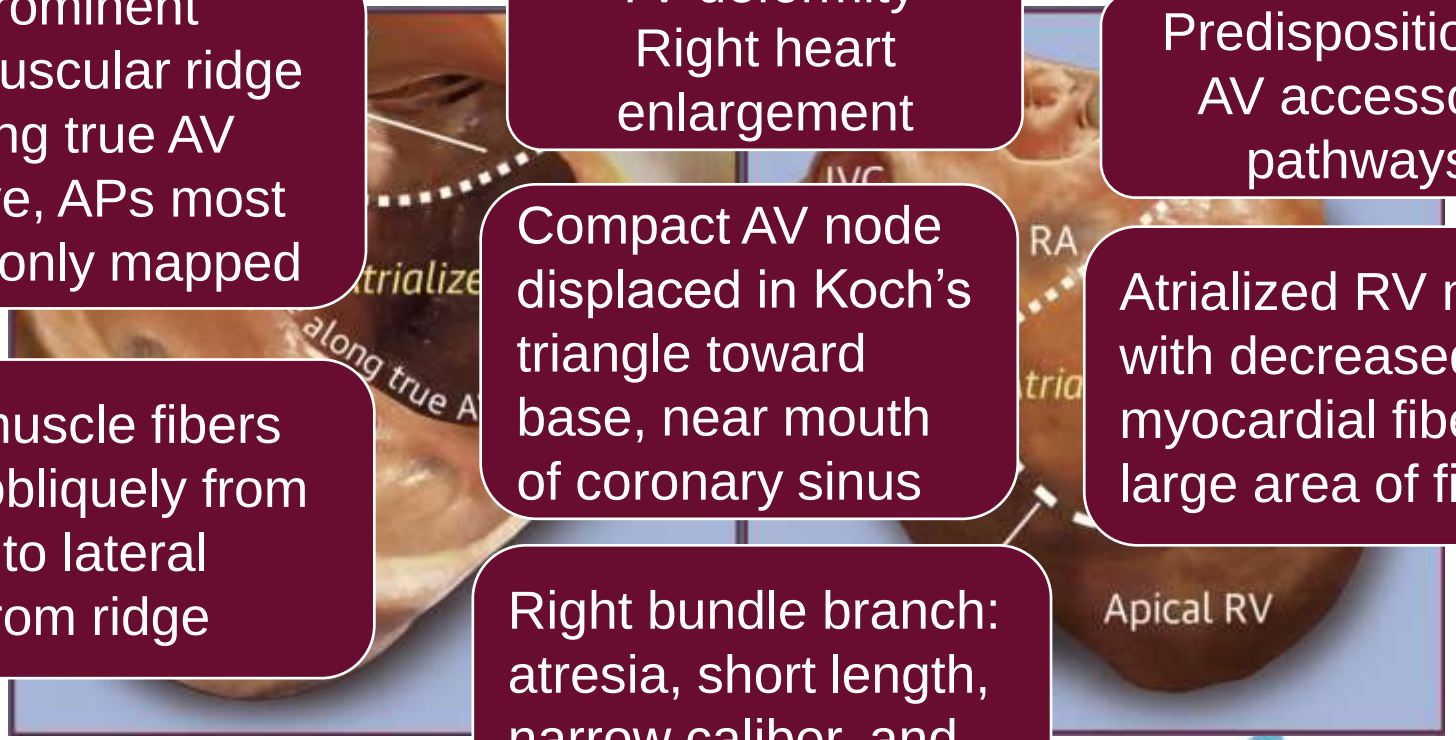
Walsh, E.P. J Am Coll Cardiol EP. 2018

ELECTROPHYSIOLOGIST'S POINT OF VIEW

Prominent fibromuscular ridge along true AV groove, APs most commonly mapped

TV deformity
Right heart enlargement

Predisposition to AV accessory pathways



Compact AV node displaced in Koch's triangle toward base, near mouth of coronary sinus

Atrialized RV muscle with decreased myocardial fiber and large area of fibrosis

Atrial muscle fibers travel obliquely from medial to lateral away from ridge

Right bundle branch: atresia, short length, narrow caliber, and fibrosis

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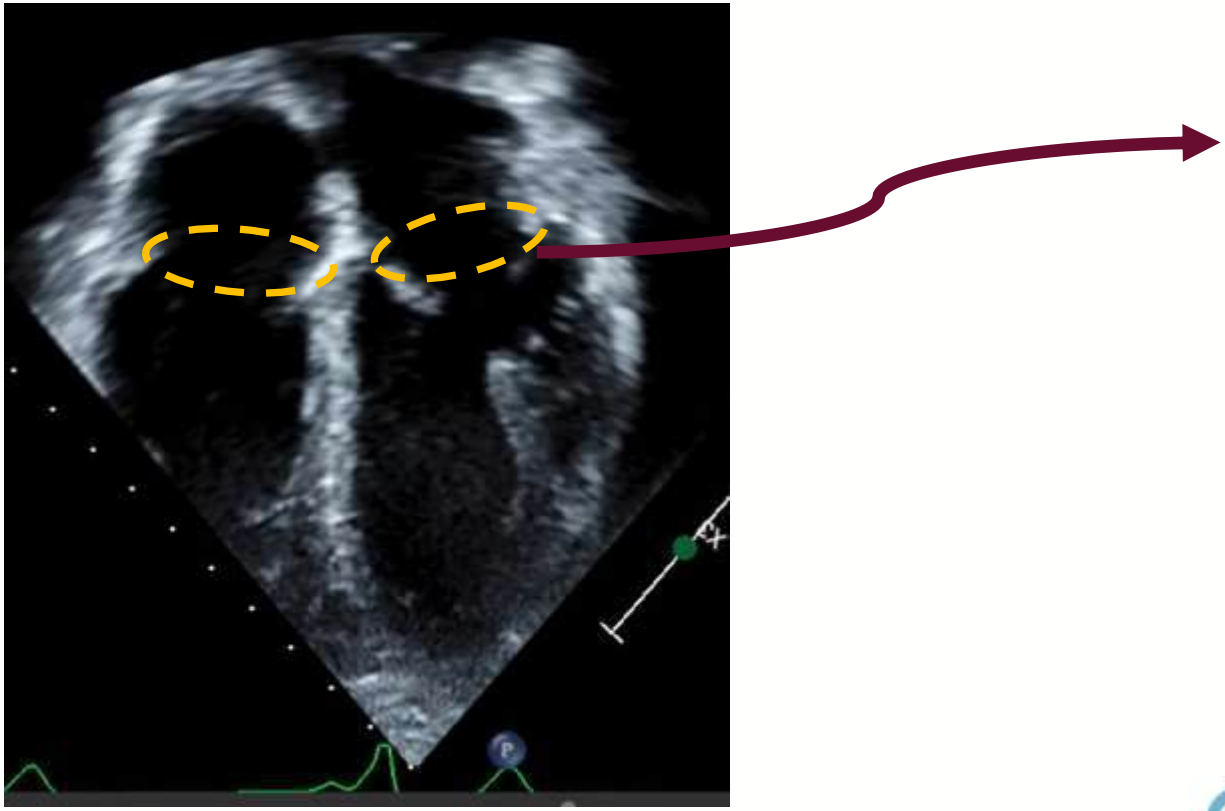
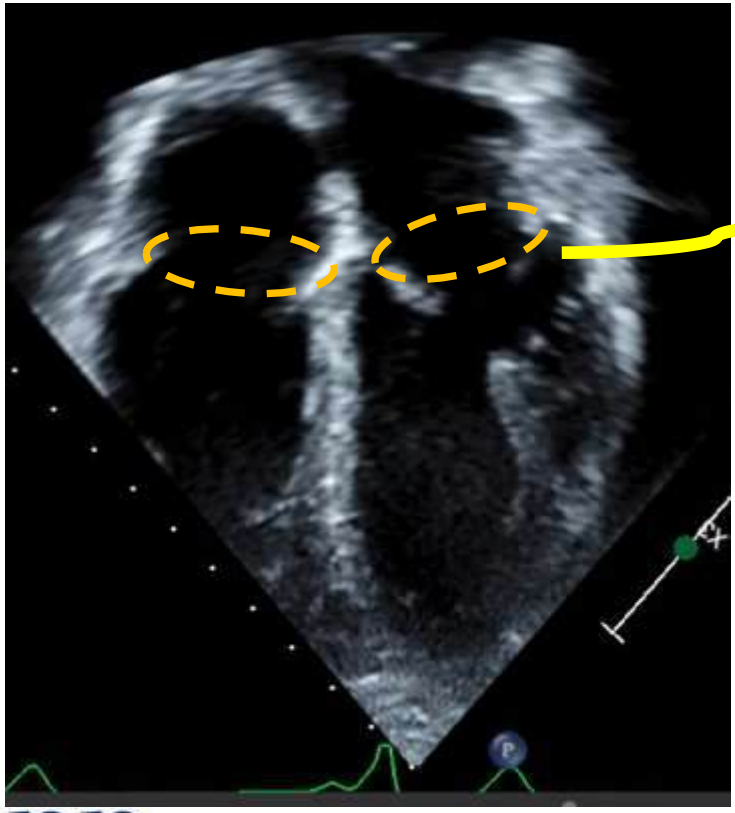
AV ACCESSORY PATHWAY LOCATION: “TRUE AV RING”

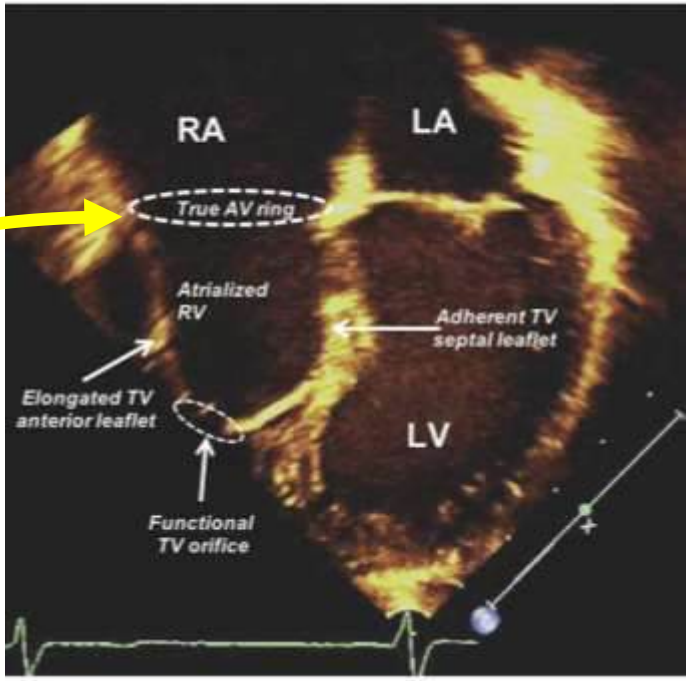


AV ACCESSORY PATHWAY LOCATION: “TRUE AV RING”

NORMAL



EBSTEIN ANOMALY



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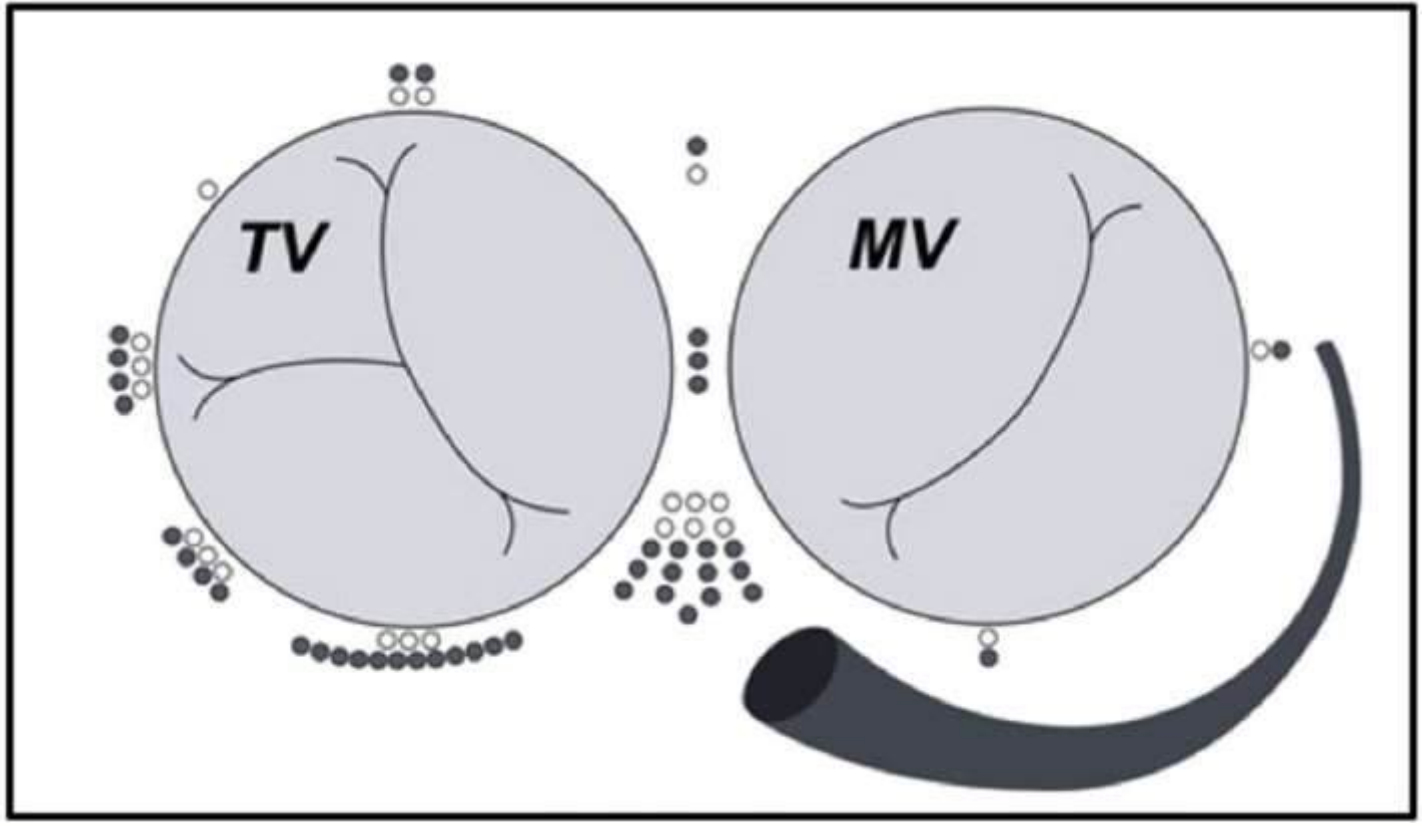
UNIQUE PREDISPOSITION TO

Atrioventricular

ACCESSORY PATHWAYS

ACCESSORY PATHWAYS (AP) IN EBSTEIN ANOMALY

- Incidence estimated to be 10-38%
- ~50% multiple separate pathways or complex insertion patterns
- Vast majority mapped to posterolateral, posterior, and posteroseptal sites along the abnormal TV ring



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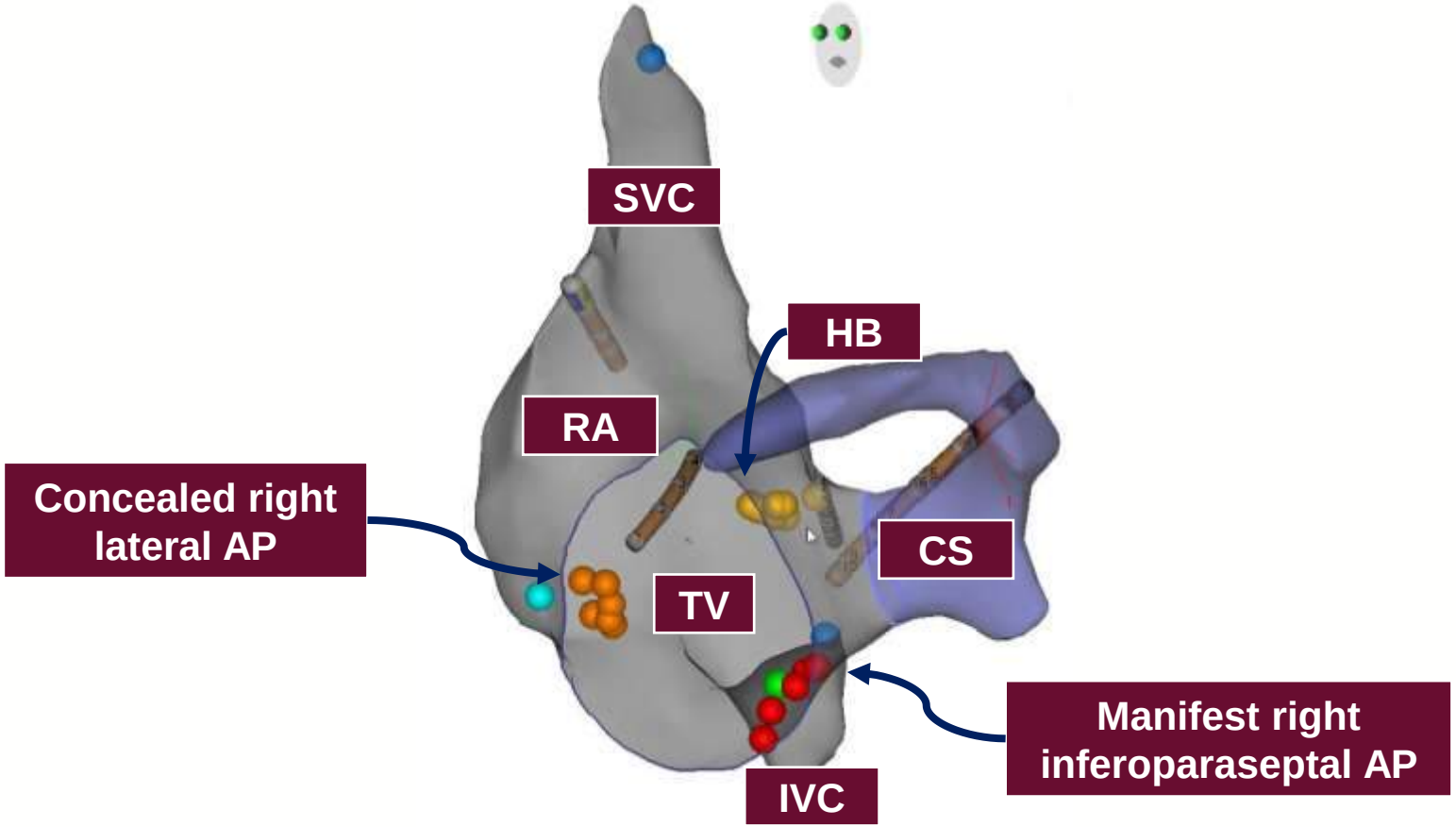
ACCESSORY PATHWAYS (AP) IN EBSTEIN ANOMALY

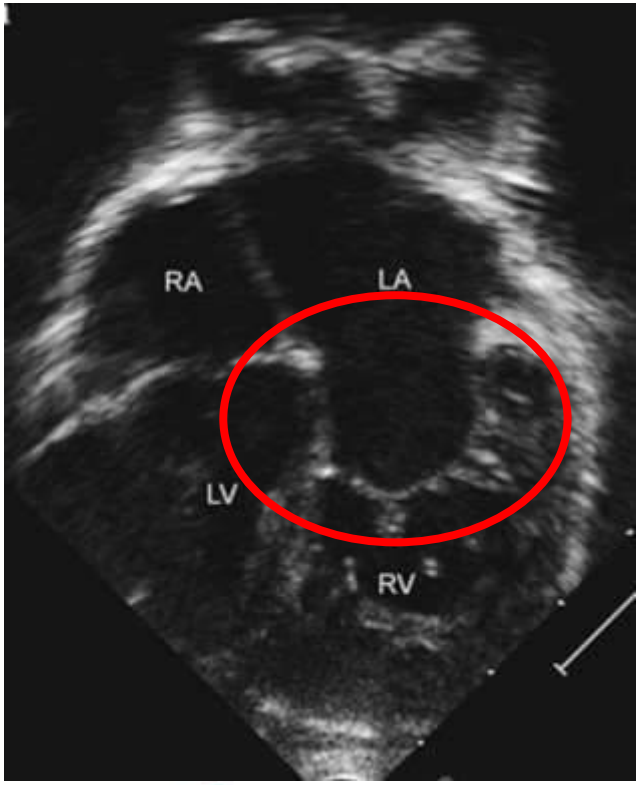
- Incidence estimated to be 10-38%
- ~50% multiple separate pathways or complex insertion patterns
- Vast majority mapped to posterolateral, posterior, and posteroseptal sites along the abnormal TV ring
- >75% of the APs are capable of bidirectional conduction and generate manifest pre-excitation (WPW pattern) on ECG
- Atriofascicular fibers also reported commonly (5-8%); location similar to that found in structurally normal hearts

EBSTEIN ANOMALY OF TRICUSPID VALVE: 3D MAP

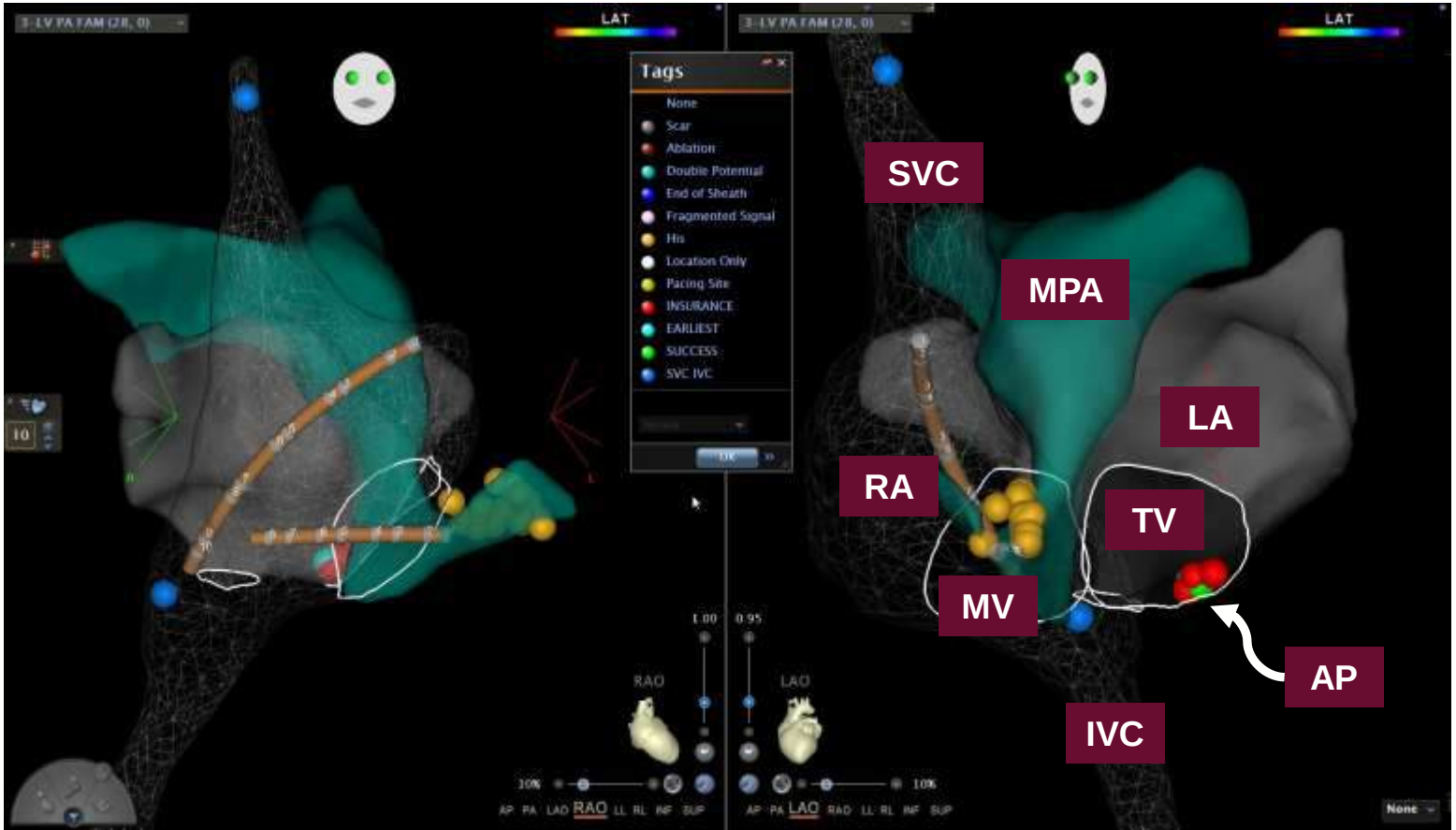


CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES WITH EBSTEIN LIKE MALFORMATION OF LEFT SIDED TRICUSPID VALVE

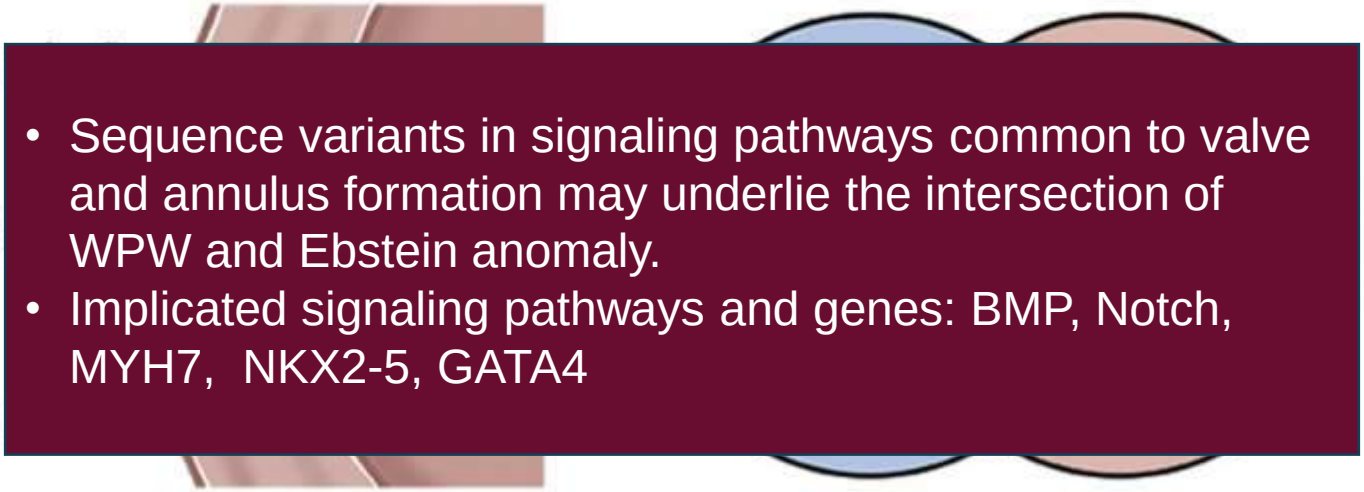
- Septal and posterior TV leaflets can be displaced inferiorly toward the cardiac apex
- Anterior leaflet will usually not display the elongated sail-like deformity seen in classic Ebstein
- Degree of atrialization for the RV not as dramatic
- Elevated incidence of ‘accessory pathways’, tend to cluster in the ***left*** posterolateral AV groove



CC-TGA, EBSTEIN LIKE ANOMALY OF LEFT SIDED TRICUSPID VALVE: 3D MAP



OVERLAP OF AP / WPW AND EBSTEIN ANOMALY: KEY MOLECULAR SIGNALING PATHWAYS



- Sequence variants in signaling pathways common to valve and annulus formation may underlie the intersection of WPW and Ebstein anomaly.
- Implicated signaling pathways and genes: BMP, Notch, MYH7, NKX2-5, GATA4

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He BJ, et al. JACC: Clinical Electrophysiology 2021.

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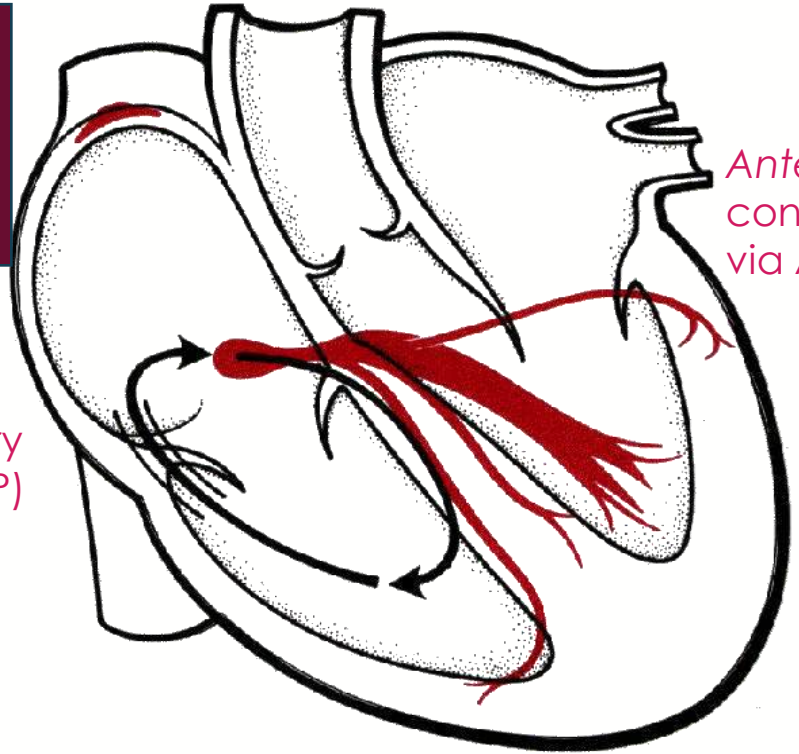
ATRIOVENTRICULAR ACCESSORY PATHWAY MEDIATED REENTRANT SVT

Orthodromic reentry induced in >90% with APs and antidromic reentry in ~ 23%



Antegrade conduction via AV node-HPS

Retrograde conduction via accessory pathway (AP)

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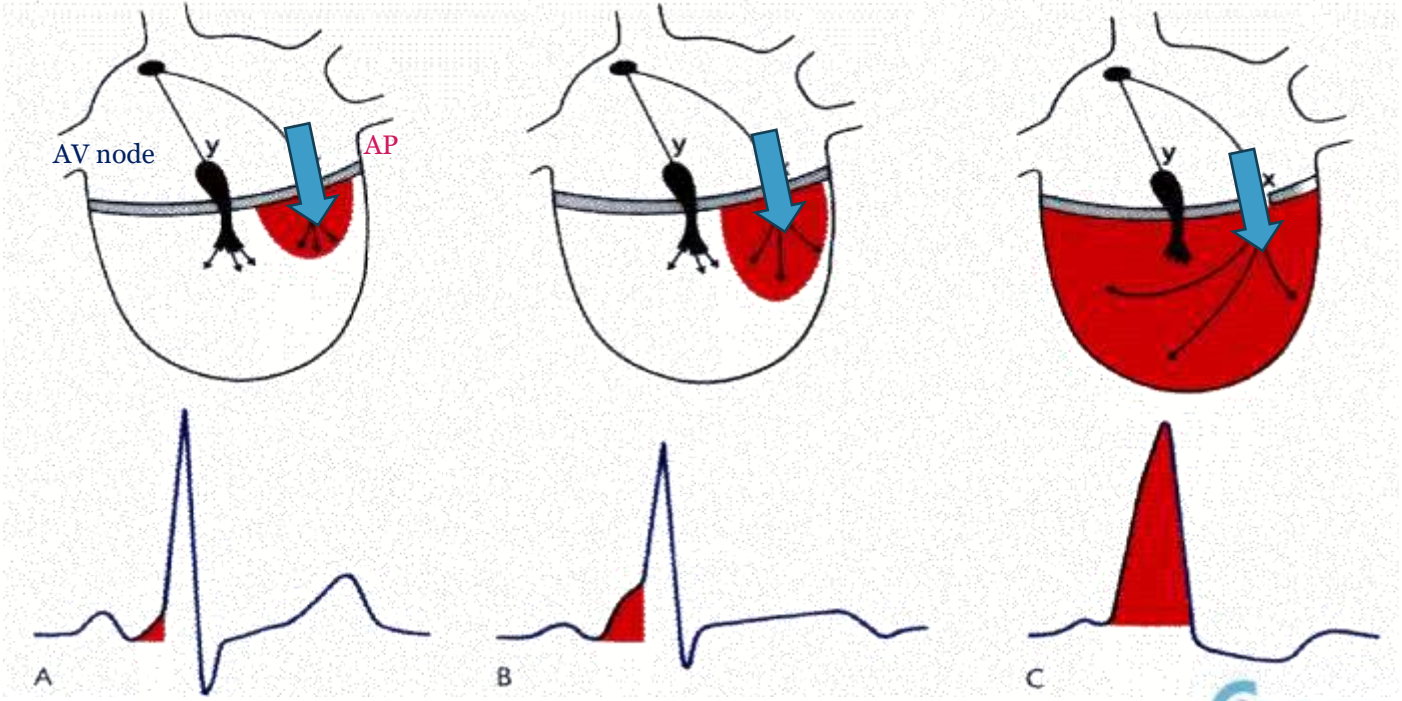
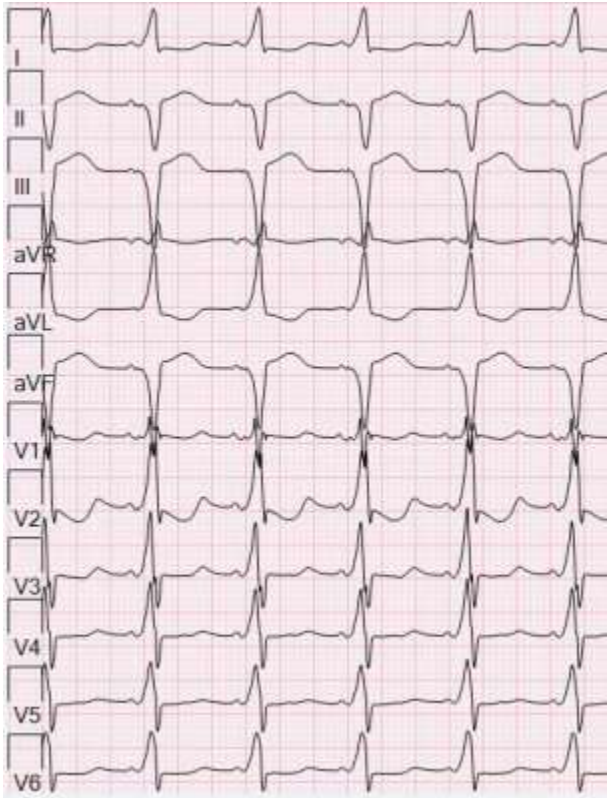
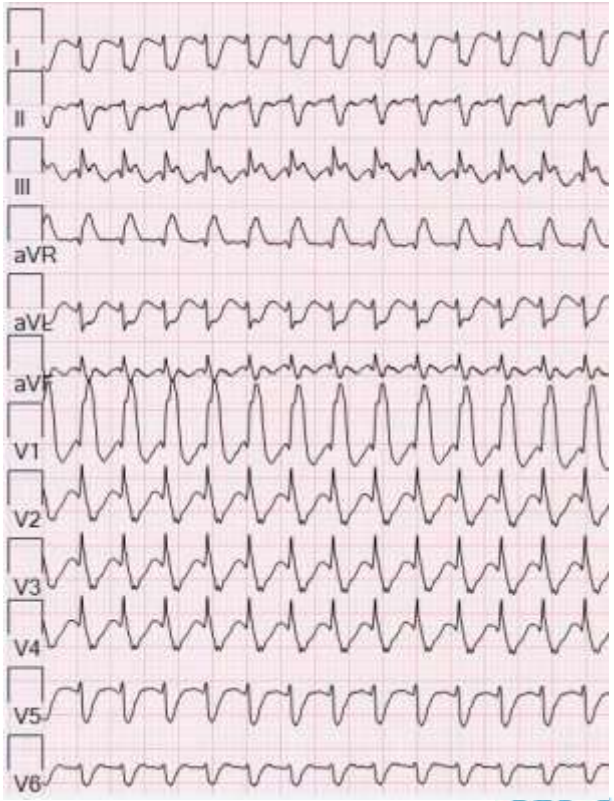
ATRIOVENTRICULAR ACCESSORY PATHWAY WITH ANTEGRADE CONDUCTION: WOLFF-PARKINSON-WHITE PATTERN IN SINUS RHYTHM



Sinus rhythm



Reentrant SVT



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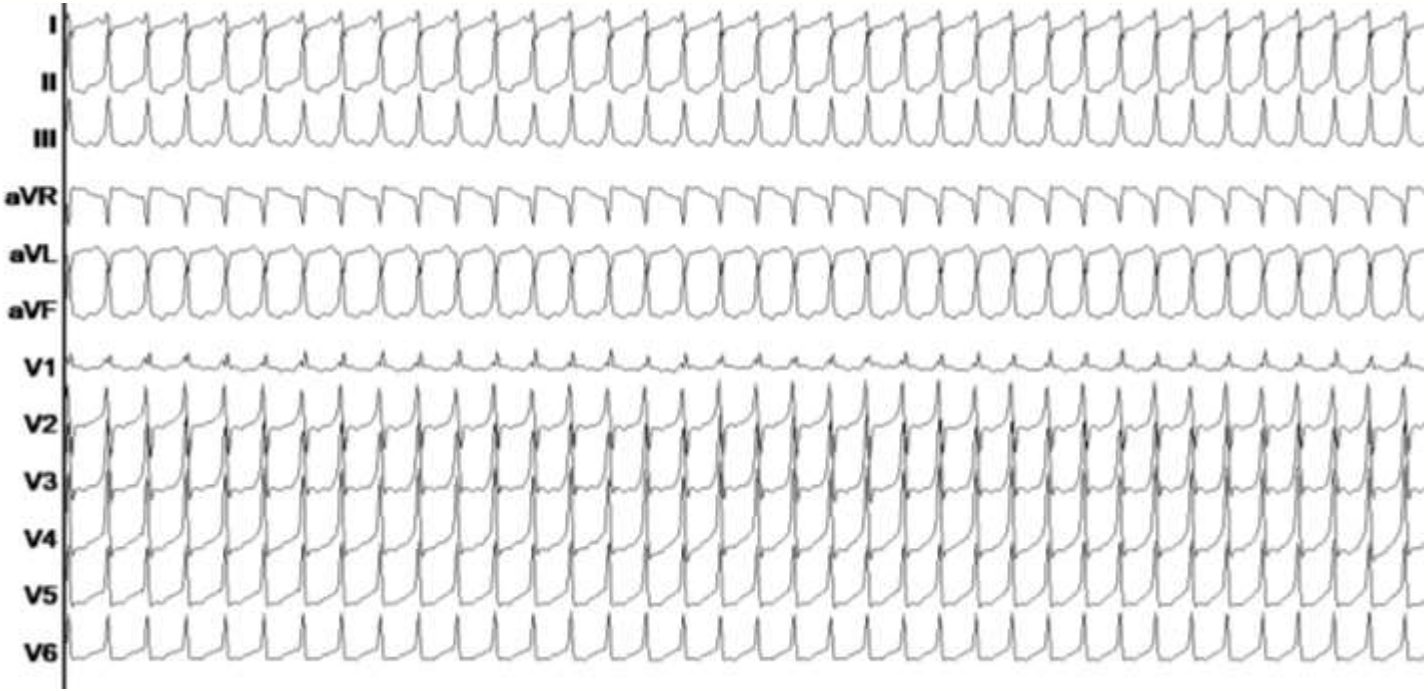
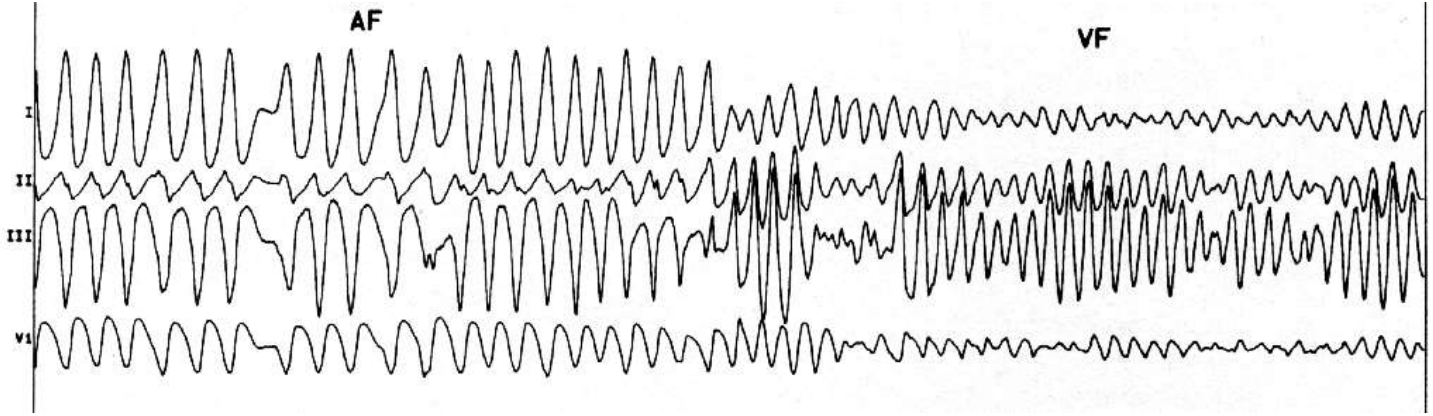
ORTHODROMIC ATRIOVENTRICULAR REENTRANT TACHYCARDIA



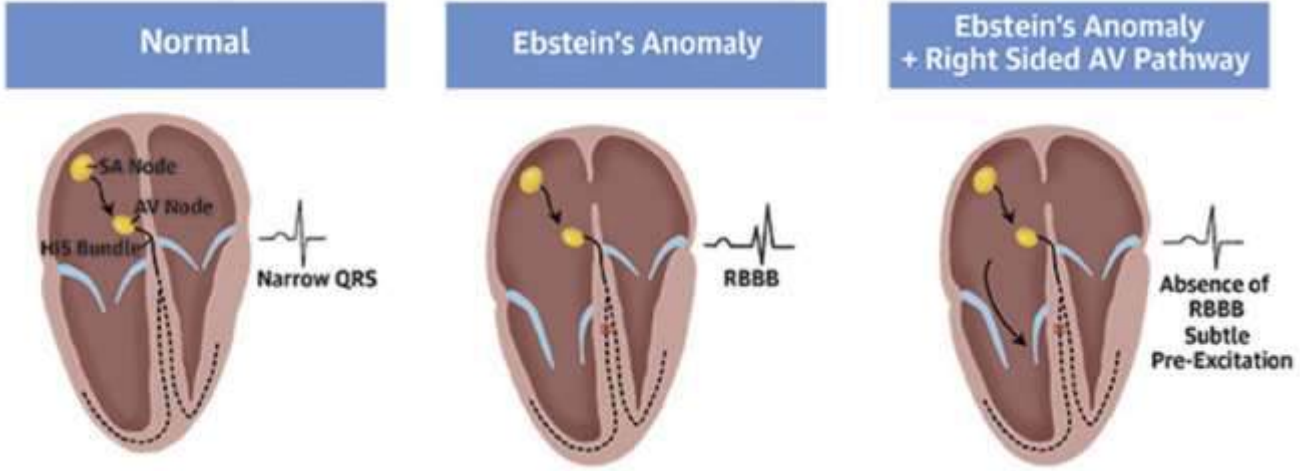
ATRIAL FIBRILLATION WITH RAPID CONDUCTION VIA AV ACCESSORY PATHWAY: PRE-EXCITED ATRIAL FIBRILLATION

PRE-EXCITED ATRIAL FIBRILLATION DEGENERATION TO VENTRICULAR FIBRILLATION

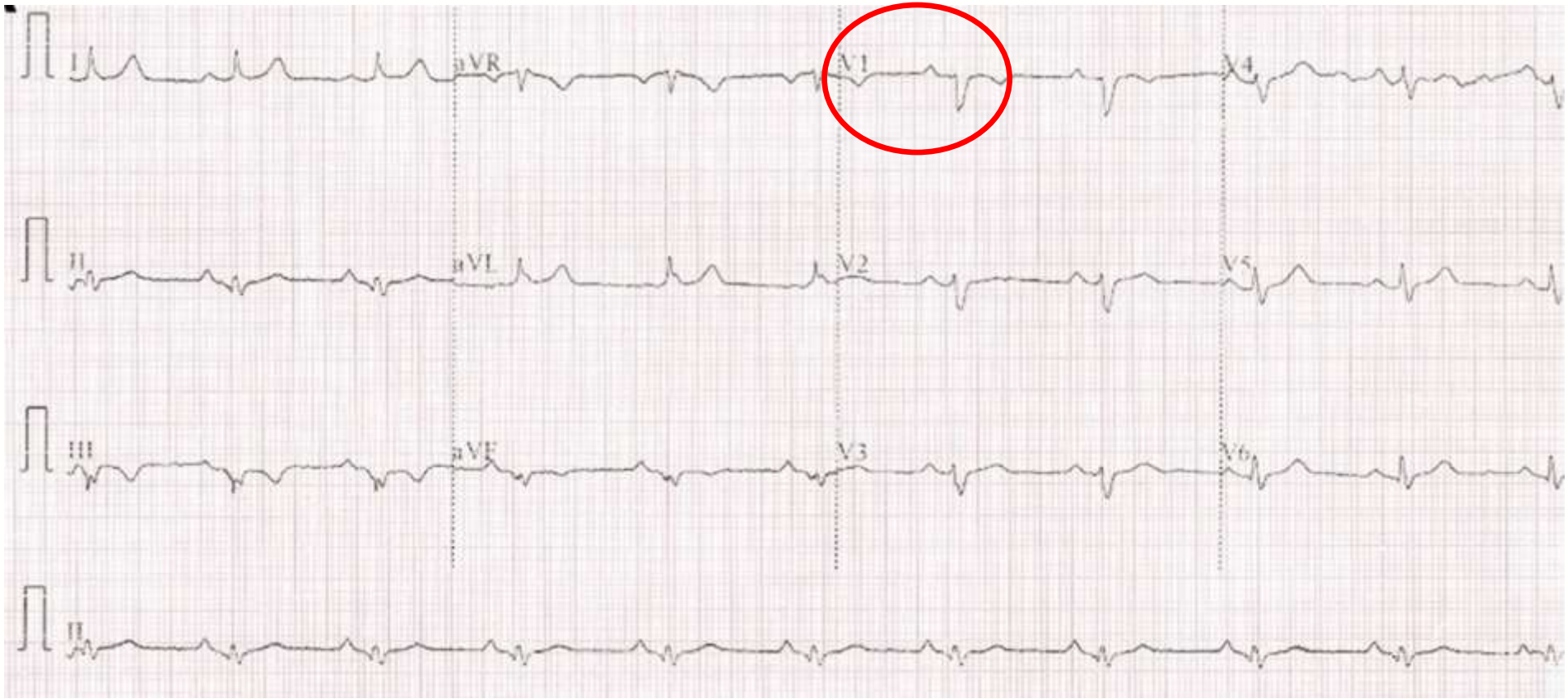


ECG PATTERN IN EBSTEIN ANOMALY

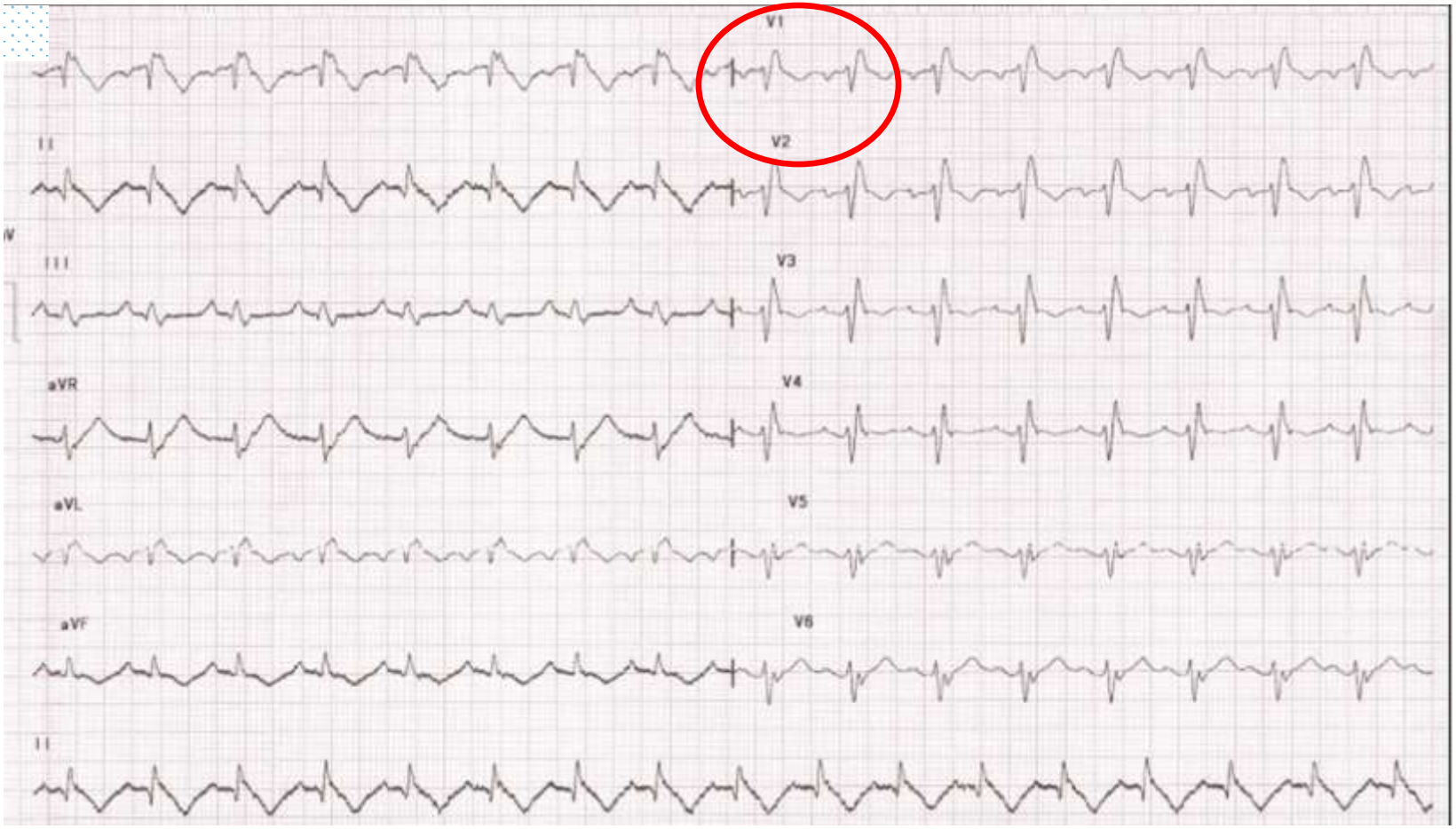
- Vast **majority (71 to 94%) will exhibit RBBB** in the absence of preexcitation (WPW)
- **Absence of the typical RBBB** (“pseudonormalized”) may be clue to presence of preexcitation from right sided AP
- Many will also have first-degree AV block in the absence of preexcitation



PRE-ABLATION OF RIGHT POSTERIOR AV ACCESSORY PATHWAY



POST-ABLATION OF RIGHT POSTERIOR AV ACCESSORY PATHWAY



ARRHYTHMIA MANAGEMENT: GUIDELINES & PRINCIPLES

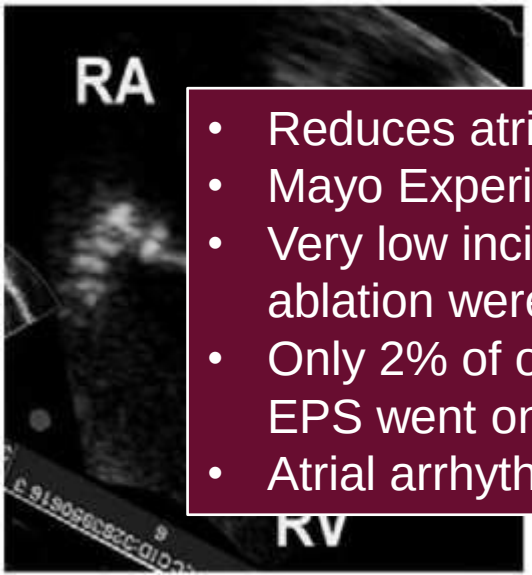
- General approach:
  - Individualized management combining medical therapy, catheter ablation, and/or surgical intervention
  - Hemodynamic optimization
- Pre-op EPS & Catheter ablation
- Arrhythmia surgery: Left atrial or biatrial Maze
- Women with EA contemplating pregnancy need pre-pregnancy arrhythmia assessment

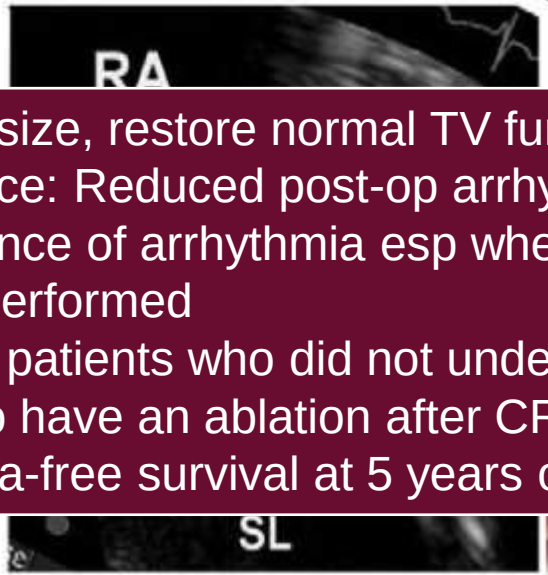
ROLE OF PRE-OP ELECTROPHYSIOLOGIC EVALUATION

Increasing consensus to consider a comprehensive EP study as a prelude to intracardiac operations

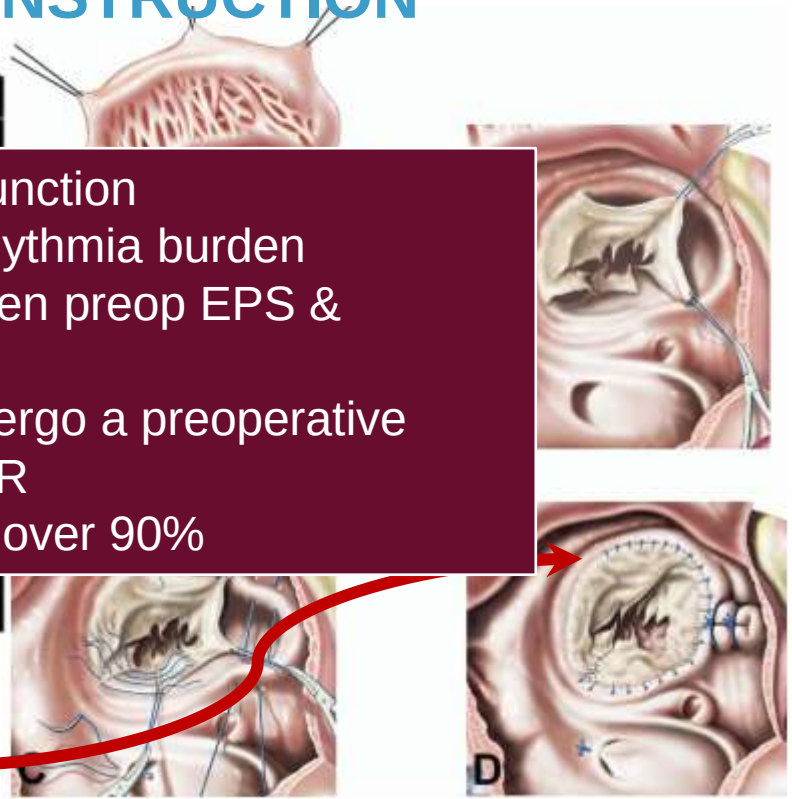
- Both concealed and manifest accessory pathways (preexcitation) could become problematic after surgery
- Post-operative catheter manipulation and ablation near the freshly reconstructed TV could affect operative results and long-term TV function
- Cone reconstruction can act as a frustrating mechanical barrier to catheter access in the region where ablation is most likely to be needed
- When an AP is identified preoperatively but cannot be successfully ablated, surgical ablation offers an opportunity

IMPACT OF CONE RECONSTRUCTION





- Reduces atrial size, restore normal TV function
- Mayo Experience: Reduced post-op arrhythmia burden
- Very low incidence of arrhythmia esp when preop EPS & ablation were performed
- Only 2% of our patients who did not undergo a preoperative EPS went on to have an ablation after CR
- Atrial arrhythmia-free survival at 5 years over 90%



Restricts access to true AV ring, location of AV accessory pathway

da Silva JP et al. J Thorac Cardiovasc Surg 2007
Wackel et al. Congenital Heart Disease 2018

CHALLENGES: CATHETER ABLATION OF AP IN EBSTEIN ANOMALY

- **Electrophysiological**
  - Fractionated low-amplitude ventricular electrograms recorded from atrialized RV
  - Oblique atrial muscle fiber orientation along AV groove distorting activation patterns Multiple accessory pathways
- **Anatomic**
  - Dilated RA/RV/AV groove
  - Difficulty identifying the true AV groove
  - Ridge tissue along the true AV groove that interferes with catheter tip navigation
  - Inability to retroflex catheter tip under TV leaflets to improve stability
  - Very thin AV groove tissue in spots that may increase risk of coronary artery injury
- **Hemodynamic**
  - Intolerance of SVT, even when rates only modestly elevated
  - Right-to-left shunting with potentially increased thromboembolic risk

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Reich JD, et al. J Cardiovasc Electrophysiol 1998
Roten L, et al. J Cardiovasc Electrophysiol 2011

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OUTCOMES OF CATHETER ABLATION OF AP

- Overall acute success rates are high, ~90–95% in experienced centers.
- Recurrence rate after a first procedure for AP ablation continues to be significantly high (20% to 40%)
- Repeat ablations may be necessary, and success rates generally remain good with iterative procedures

ACQUIRED ATRIAL ARRHYTHMIAS

- Atrial macro–re-entry is the most common; classic cavotricuspid isthmus-dependent or circuits around atriotomy incisions or septal patches in postoperative patients
- Atrial Flutter and Atrial Fibrillation *(Mayo, Martin de Miguel et al. JACC Advances 2022)*:
  - Very prevalent due to chronic RA enlargement.
  - Atrial flutter/tachycardia seen in ~21% and AF in ~18% of adults at baseline.
  - Cumulative incidence of new atrial arrhythmias continues to rise with age (16% AF; 22% AFL in 10 years)

SUDDEN ARRHYTHMIC DEATH

- 8-16%
- Predictors:
  - A prior history of VT
  - Heart failure
  - history of prior TV surgery (included older era surgery)
  - Severe RV enlargement and dysfunction, which can distort and compromise LV performance through ventricular-ventricular interaction
  - LV noncompaction
- Sudden cardiac death (SCD) is also a recognized risk, often due to rapid conduction of atrial fibrillation over an atrioventricular accessory pathway leading to VF

Vacca JB, Bussmann DW, Mudd JG. Am J Cardiol 1958
Koyak Z, Harris L, de Groot JR, et al. Circulation 2012
Attenhofer Jost CH, Tan NY, Hassan A et al. Eur Heart J 2018

ONGOING RESEARCH, FUTURE DIRECTIONS

- **Atrial Myopathy & Arrhythmia Risk:**
  - Quantifying atrial dysfunction as a predictor of arrhythmias
  - Novel echocardiographic measures like right atrial strain and advanced MRI volumetrics may help identify patients at highest risk for AF/AFL
- **Genetic Insights:**
  - Future studies may clarify if certain genotypes carry higher arrhythmic risk (e.g., closer monitoring for AF in those with an “atrial myopathy” gene profile).
- **Hybrid interventions:**
  - Development of hybrid EP-surgical suites allows combined procedures

CONCLUSIONS

- Arrhythmias are extremely common, stemming from the unique anatomy (atrialized RV, enlarged RA) and high prevalence of accessory pathways
- It is hoped that heightened appreciation for its unique anatomic features will improve the long-term outcomes of ablation procedures for this population
- More patients with Ebstein anomaly live into older adulthood, thanks to surgical advances in TV reconstruction
- Comprehensive EP study and catheter ablation prior to surgical intervention is highly beneficial


Thank you!