RFCA in Ebstein’s Abnormality

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Ebstein’s anomaly

- Described by Wilhelm Ebstein in 1866.
- 1/200,000 live births, <1% of all cases of congenital heart ds.

1. Downward displacement and adherence of the septal and posterior leaflets
2. Redendant anterior leaflet
3. Atrialized RV
4. Dilated tricuspid annulus
Associated malformations

- ASD, PFO: in 80-94%
- Congenitally corrected transposition of the great arteries, bicuspid aortic valve, subaortic stenosis, coarctation, pulmonary artery hypoplasia/atresia...
Arrhythmias in Ebstein’s anomaly

• AV or atriofascicular accessory pathway
  • Intraatrial reentrant tachycardia
  • Ectopic atrial tachycardia
  • Atrial fibrillation

PACES/HRS Expert Consensus. *Heart Rhythm* 2014
Typical ECG features in Ebstein

- Rhythm: ectopic atrial rhythm, AF/IART 40%
- PR: 1\textsuperscript{st} degree AV block
- QRS: low amplitude, multiphasic, RBBB
- Atrium: RAE (Himalayan P wave)
- Negative delta: II, III, aVF, V1-4
Himalayan P wave suggesting severe RAE

Sharma et al. Internal and Emergency Medicine 2011
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PACES/HRS Expert Consensus

• Class I
  – RFCA is recommended for preexcitation, and high-risk or multiple accessory pathways, as commonly encountered in Ebstein anomaly

• Class IIa
  – Concomitant atrial arrhythmia surgery should be considered during cardiac surgery

• Class IIb
  – It may be reasonable to perform EP studies in patients with Ebstein anomaly before surgery

PACES/HRS Expert Consensus. *Heart Rhythm* 2014
Accessory pathways in Ebstein anomaly


W Wei et al. Europace 2014
Tricuspid annuli in Ebstein anomaly

Usual ablation target area
Case: WPW in Ebstein anomaly

- A 34-year-old male
- C/C : Intermittent palpitation for 1 years
- Echo : Ebstein anomaly, moderate to severe TR
- Scheduled to perform tricuspid valve repair

Courtesy of Professor Uhm (Severance)
Resting ECG
Echocardiography
Right Ventriculography
Antegrade mapping of AP
Retrograde mapping of AP
Delta wave disappeared during RFCA
RFCA for right posterior AP

Atrialized RV
Intracoronary wire can be guidance

Shah et al. *JCE* 2004
Challenging issues

• Atrialized RV
  – fragmented and low-amplitude EGM
  – Difficult to distinct between atrial and ventricular activation potentials as well as the identification of AP potentials.
  – Success rate: 60-70%

Cappato et al. *Circulation* 1996