

Morphology of Ebstein Anomaly and Tricuspid VALve

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 Our plans for the future

www.rbht.nhs.uk/morphology

Morphology of Ebstein and TV

- Normal TV
- Normal TV development
- Index case of Ebstein anomaly
- Morphology of the TV in Ebstein
- Associated malformations in Ebstein anomaly
- Other TV malformations

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Normal TV

Hingeline (annulus)

Anatomical components: Hingeline, leaflets, tendinous cords, papillary muscles, atrial and ventricular walls

Tendinous cords
 Rough zone & deep cords
 Free-edge cords
 Basal cords

Leaflet cords

Interleaflet cords

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Ebstein

Wilhelm Ebstein (1836-1912)
 Professor of Medicine, Göttingen, from 1874

University of Göttingen

From: van Son et al, Eur J Cardiothorac Surg 2001

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Heart of Joseph Preshler

Dilated RA
 FO open, multiple openings
 Absent Thebesian valve
 No chordae and pap muscles
 Fibrous membrane sac
 Malformed leaflet

Described by Wilhelm Ebstein in 1866
 Illustrations by Dr Wyss

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Ebstein W, Ueber einen sehr seltenen Fall von Insuffizienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. Arch Anat Physiol Wissensch Med. 1866:238-254

- A severe malformation of the tricuspid valve
- Absence of the Thebesian valve
- A patent foramen ovale

'Ebstein's disease'
 Arnstein, 1927
 Yater and Shapiro 1937



Delamination



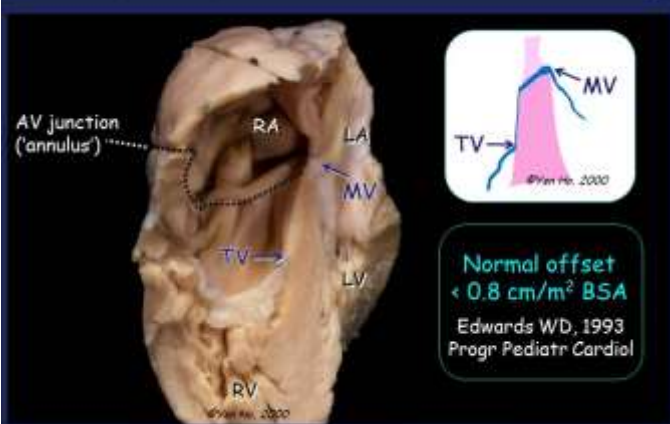
The pathognomonic feature:

"apical/downward displacement" of the TV

Reduced separation of the leaflet from the myocardium (lack of delamination) such that the leaflet is not upwardly mobile to the level of the 'annulus' (hingeline)



Extreme: leaflet agenesis



Normal offset
 $< 0.8 \text{ cm}^2 \text{ BSA}$
 Edwards WD, 1993
 Progr Pediatr Cardiol



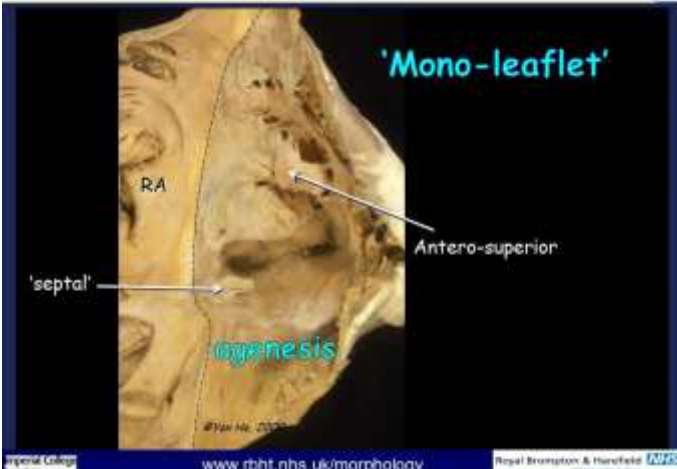
The pathognomonic feature:

"apical/downward displacement" of the TV

- Which leaflets are affected?
- How are they affected?
- What are the anatomic variations?



How malformed?
 Severity of regurgitation?

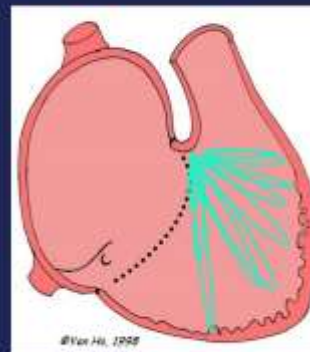


Ebstein: spectrum of abnormality

- Annular dilatation
- Location of effective orifice
- Nature of distal attachments (ant-sup leaflet)
- Thinning of ventricular wall at inlet (atrialization)
- Size of functional right ventricle
- Degree of valvar dysplasia

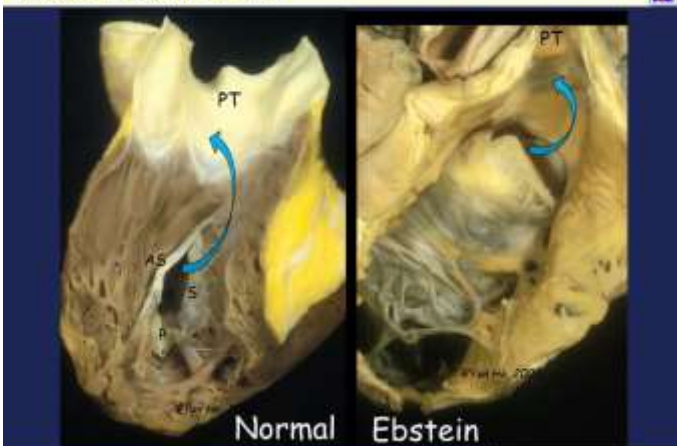
Ebstein: effective orifice

Location of effective orifice - rotation



23 heart specimens

Ebstein: effective orifice

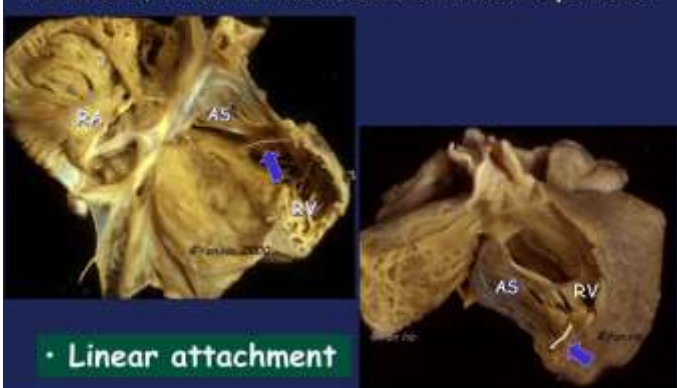


Ebstein: spectrum of abnormality

- Location of effective orifice
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- Thinning of ventricular wall at inlet
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- Degree of valvar dysplasia

Ebstein: distal attachments of leaflets

Nature of distal attachments of antero-sup. leaflet



• Linear attachment

Ebstein: distal attachments of leaflets

Nature of distal attachments of antero-sup. leaflet



'hammock'

- Location of effective orifice
- Nature of distal attachments
- Thinning of ventricular wall at inlet
- Size of functional right ventricle
- Degree of valvar dysplasia

Primarily a TV malformation

A Morphologic spectrum of severity

- Apical displacement of septal + inferior leaflets
- Distal attachments of the antero-sup leaflet
- Location of effective valvar orifice
- RV size, functional RV (inlet vs trab+outlet)
- and associated defects

Other TV malformations

Leaflet Dysplasia



Redundant leaflets, prolapse



Imperforate valve membrane ('tricuspid atresia')



Tricuspid Valve comprises more than leaflets

- Leaflets, commissures, cords, pap muscles, hingeline, membranous septum, atrial & ventricular myocardium
- Abnormality can affect one or more components

Thank you

Yen Ho