

The 2nd Contemporary Morphology Course with Specimens and 3D Print Models
CONGENITAL HEART DISEASES IN YOUR HANDS

♥ **Abnormalities of the Atrioventricular Junction** ♥

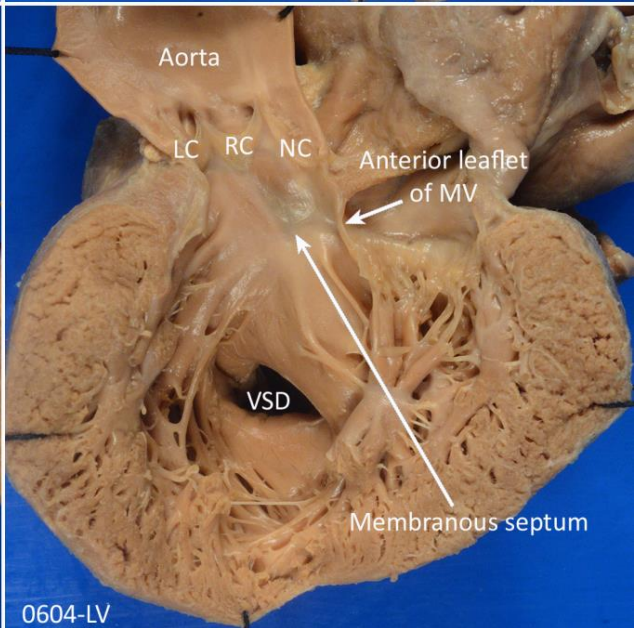
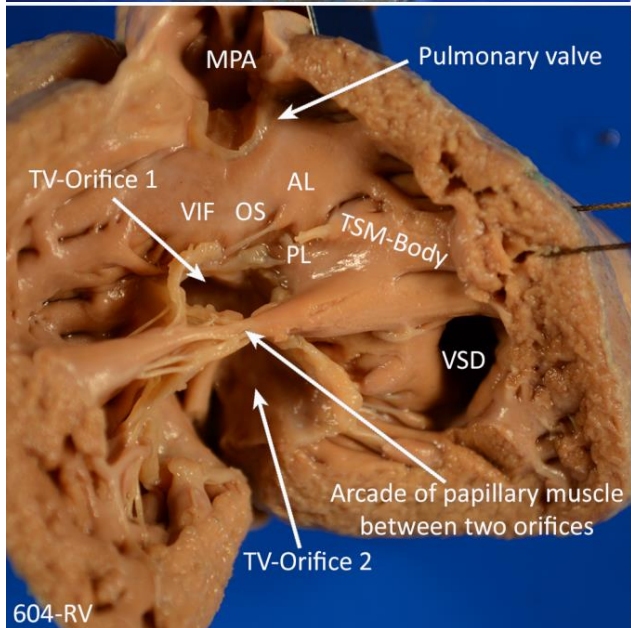
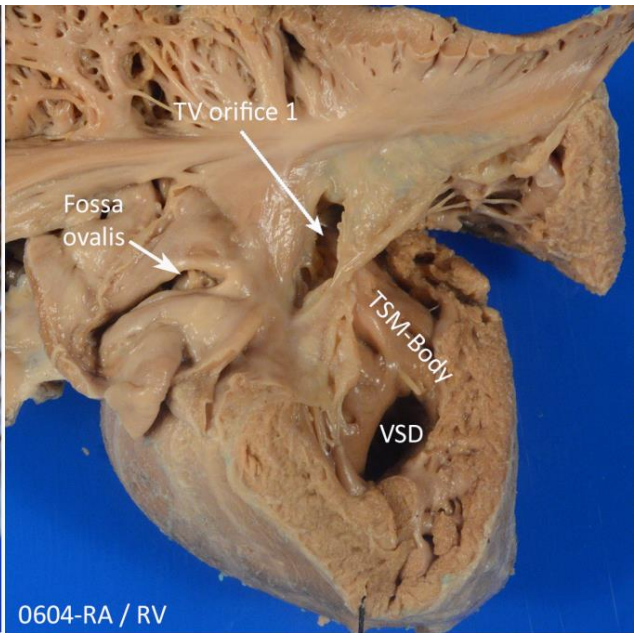
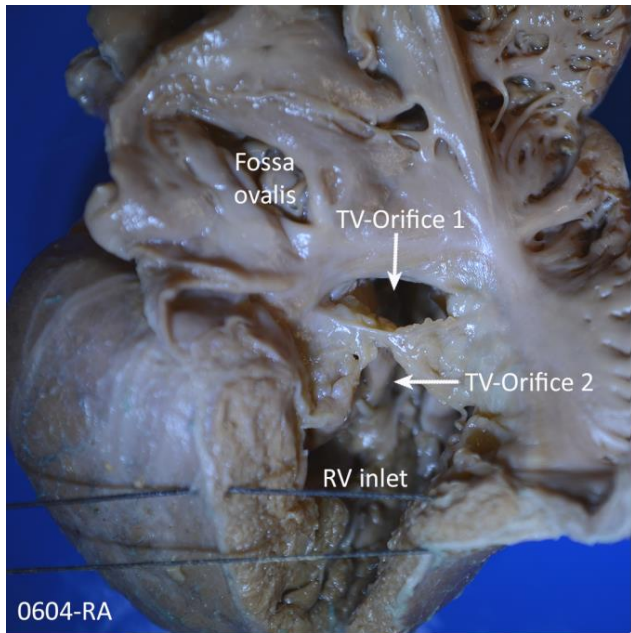
December 6-7, 2019

Peter Gilgan Centre for Research and Learning (PGCRL)

Hospital for Sick Children, Toronto, Canada

CASES DESCRIPTIONS
TRICUSPID VALVE ABNORMALITIES

□	Specimen: 0604	Categories: VSD / Dual Orifice TV
Pathology	<ul style="list-style-type: none"> • Muscular VSD involving the apical trabecular part of the septum • Coarctation of aorta • Double orifice tricuspid valve 	
Findings	<ul style="list-style-type: none"> • VSD in apical trabecular septum posterior to trabecula septomarginalis • The tricuspid valve orifice is divided into anterior (TV orifice 1) and posterior orifice (TV orifice 2). A kind of papillary muscle arcade is formed below the connecting tongue of the valve leaflet tissue between the two orifices. 	





Specimen: 1050

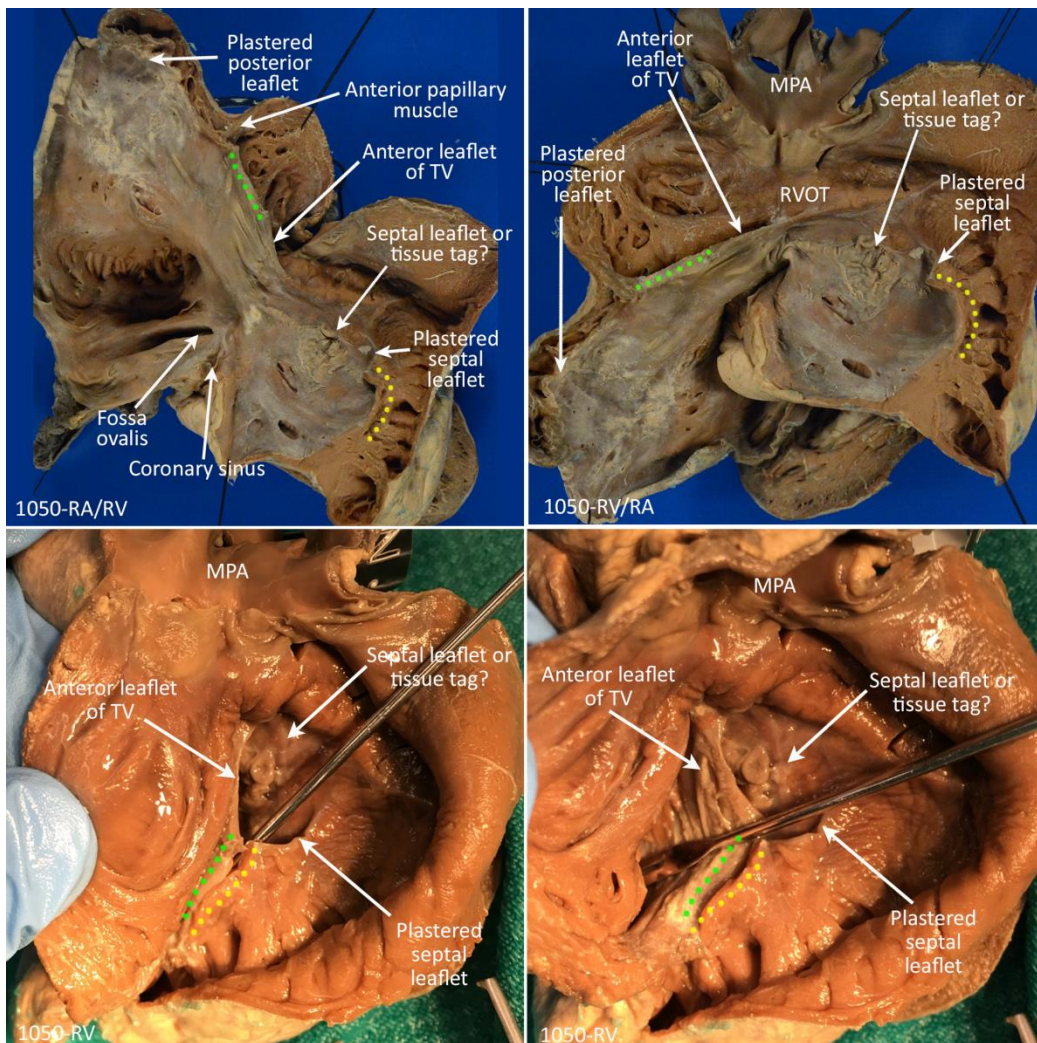
Categories: Ebstein's anomaly

Pathology

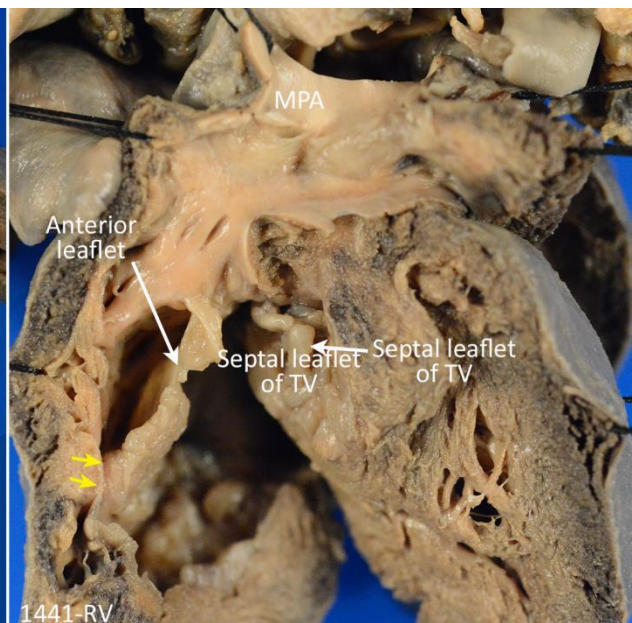
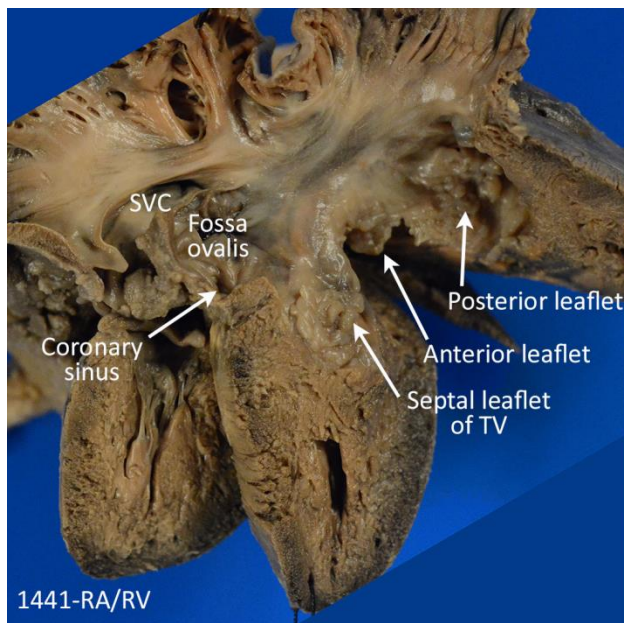
- Stenotic Ebstein's anomaly of tricuspid valve

Findings

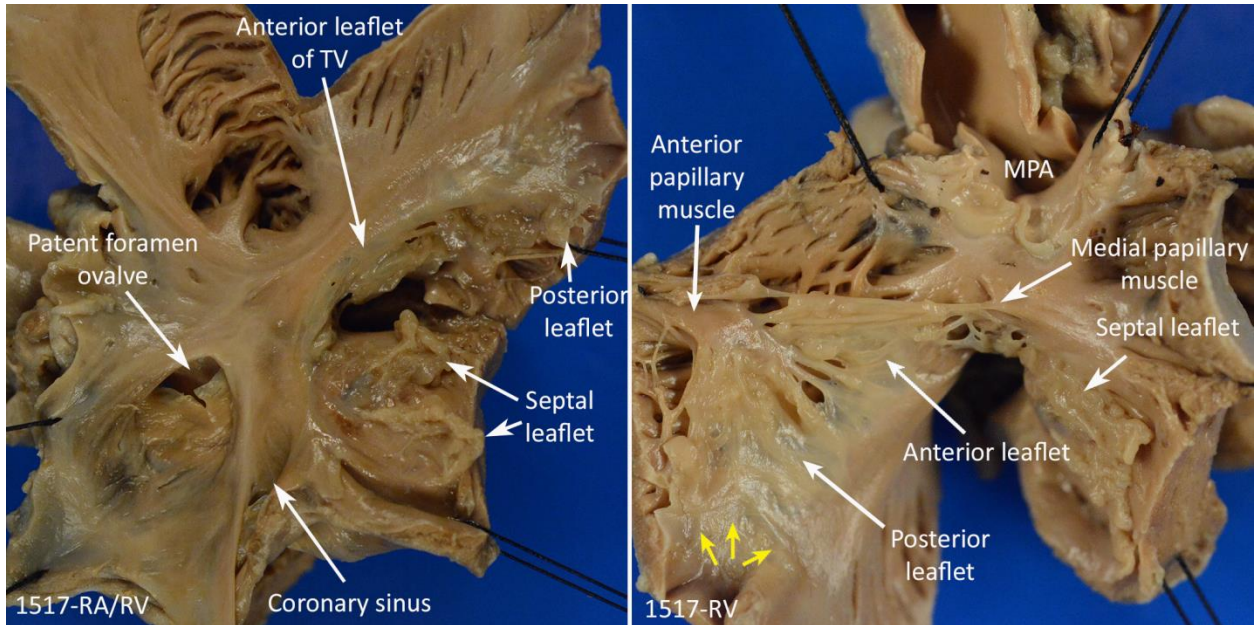
- Displaced attachment of markedly deficient septal and posterior leaflets. Muscularized septal leaflet. Fused septal and posterior leaflet was dissected. Green and yellow dotted lines indicate cut margins of the fused septal and anterior leaflets. In lower panel, the dissected fused leaflets are approximated together.
- Large anterior leaflet showing normal attachment
- Fusion of anterior leaflet with displaced posterior and septal leaflets to form a tricuspid sac. No boundaries between the fused valve leaflets. A probe is placed through the stenotic tricuspid valve orifice in left lower panel and on the ventricular aspect of the anterior leaflet in right lower panel. Note the small size of the valve orifice.
- Dilated right ventricular outflow tract. Endocardial fibroelastosis of the atrialized part of the right ventricle with a irregular patch of fibrous tissue tag on the ventricular septum near tricuspid valve orifice. .
- Large atrialized right ventricle. Patent foramen ovale



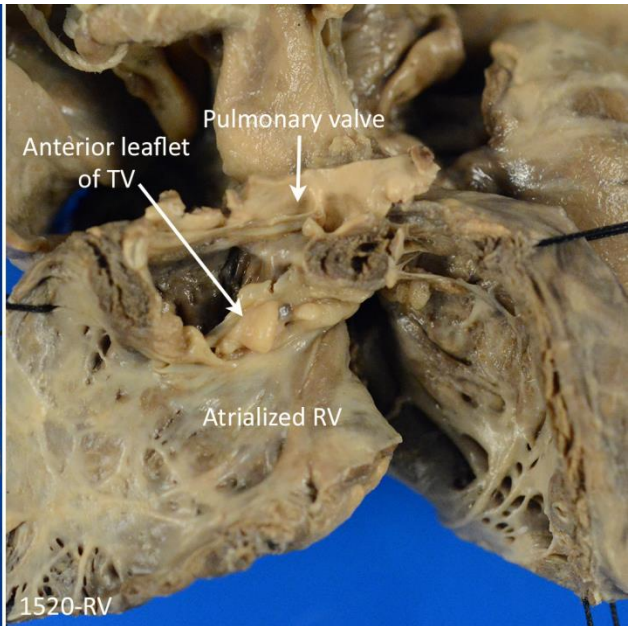
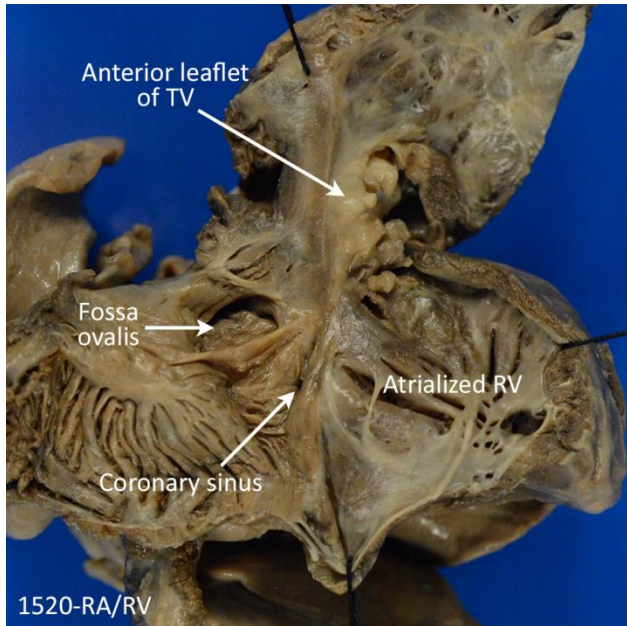
□	Specimen: 1441	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly of dysplastic tricuspid valve. 	
Findings	<ul style="list-style-type: none"> • Dysplastic tricuspid valve • Displaced attachment of septal leaflet • Severely dysplastic large anterior and posterior leaflets of tricuspid valve, directly attached to the free wall of the right ventricle (yellow arrows in right panel). • Small right ventricular cavity • Muscular VSD 	



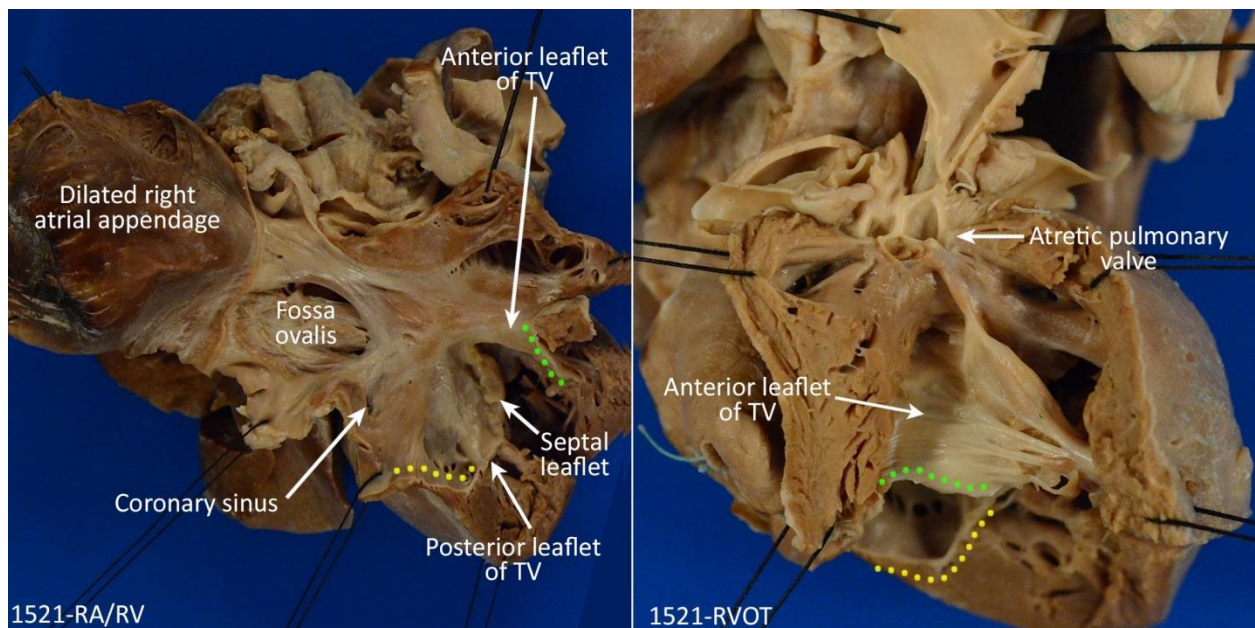
□	Specimen: 1517	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly of tricuspid valve 	
Findings	<ul style="list-style-type: none"> • Displaced attachment of dysplastic septal leaflet. • Minor displacement of relatively large posterior leaflet. Only the far posterior part of the posterior leaflet shows displaced attachment (yellow arrows in right panel). • Large anterior leaflet • Partial fusion between anterior and posterior leaflets 	



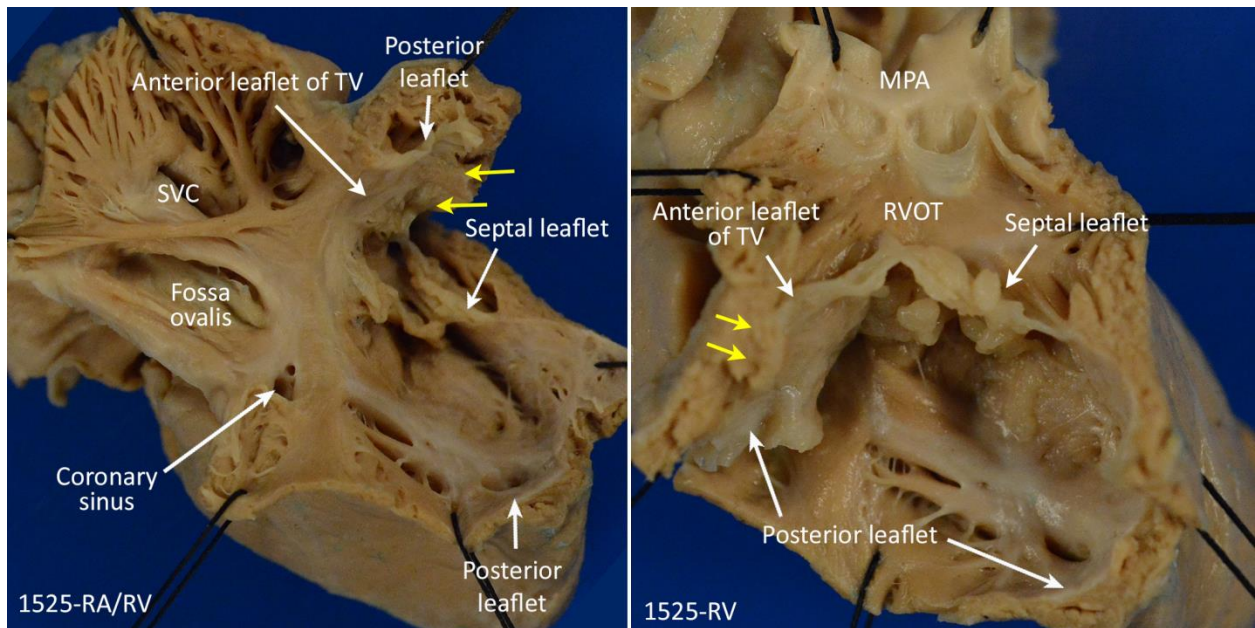
□	Specimen: 1520	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Severely stenotic Ebstein's anomaly of tricuspid valve 	
Findings	<ul style="list-style-type: none"> • Marked displaced attachment of septal and posterior leaflets with fusion • Severely dysplastic anterior leaflet is the only functioning part of the tricuspid valve. • Large atrialized part of the right ventricle showing endocardial fibrosis • Functioning part of RV consisting of small outflow component of the right ventricle 	



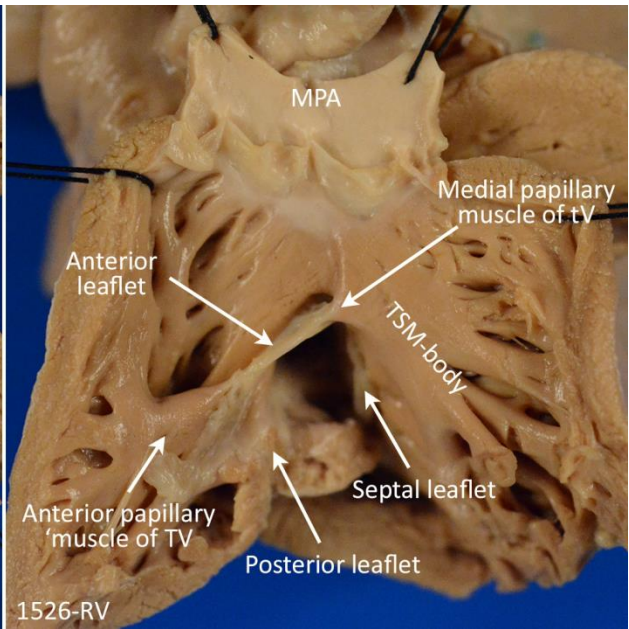
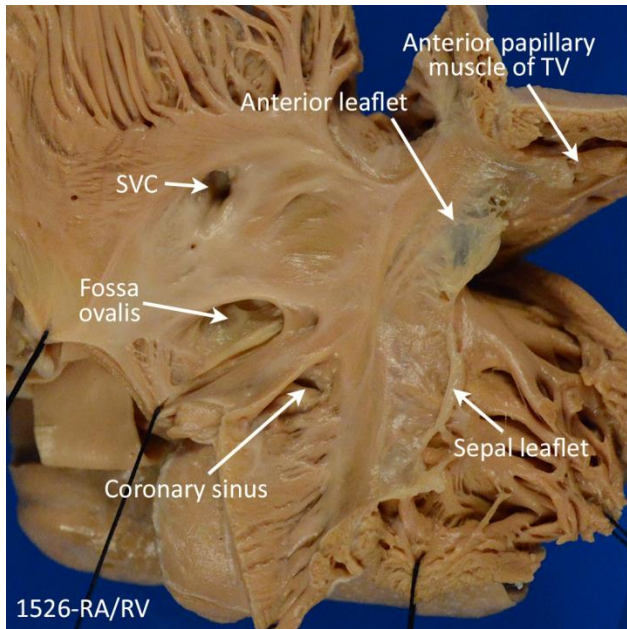
□	Specimen: 1521	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Stenotic Ebstein's anomaly of tricuspid valve with pulmonary atresia. S/P Repair attempt 	
Findings	<ul style="list-style-type: none"> • Displaced attachment of deficient septal and posterior leaflets with rolled margins • Large sail-like terior leaflet, not displaced. • Fusion between displaced septal and posterior leaflets and non-displaced anterior leaflet forming a stenotic sac in the right ventricle. No identifiable boundaries between the leaflets. Green and yellow dotted lines indicate cut edges of the fused anterior and posterior leaflets. • Atritic pulmonary valve • Marked dilatation of right atrium. Patent foramen ovale. Tiny fenestrations in the lower part of the fossa ovalis. • Right ventricular hypertrophy 	



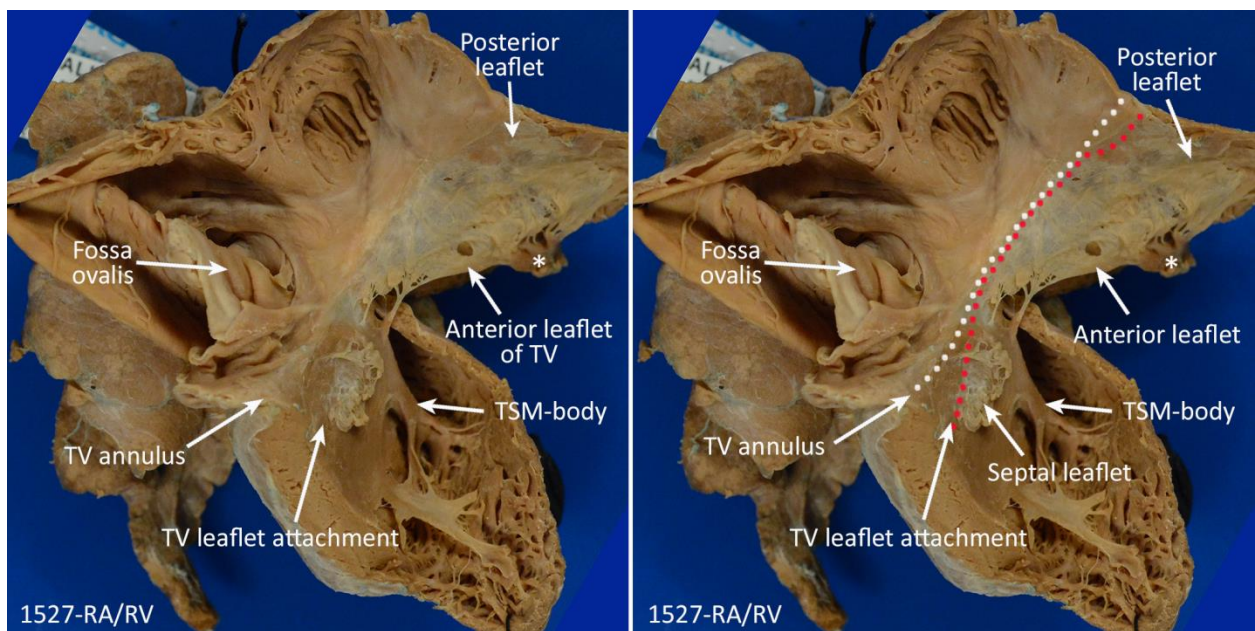
□ Specimen: 1525	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Stenotic Ebstein's anomaly of severely deficient tricuspid valve leaflets
Findings	<ul style="list-style-type: none"> • Severely vestigial septal leaflet of tricuspid valve firmly attached to the septum and inferior free wall. • Dysplastic anterior leaflet and anterior part of posterior leaflet with normal annular attachment. • Free edge of fused anterior and posterior leaflets attach directly to the right ventricular free wall (yellow arrows in both panels). • Leaflets fuse directly to apical myocardium leaving no ventricular space in apical region. • Right atrial dilatation



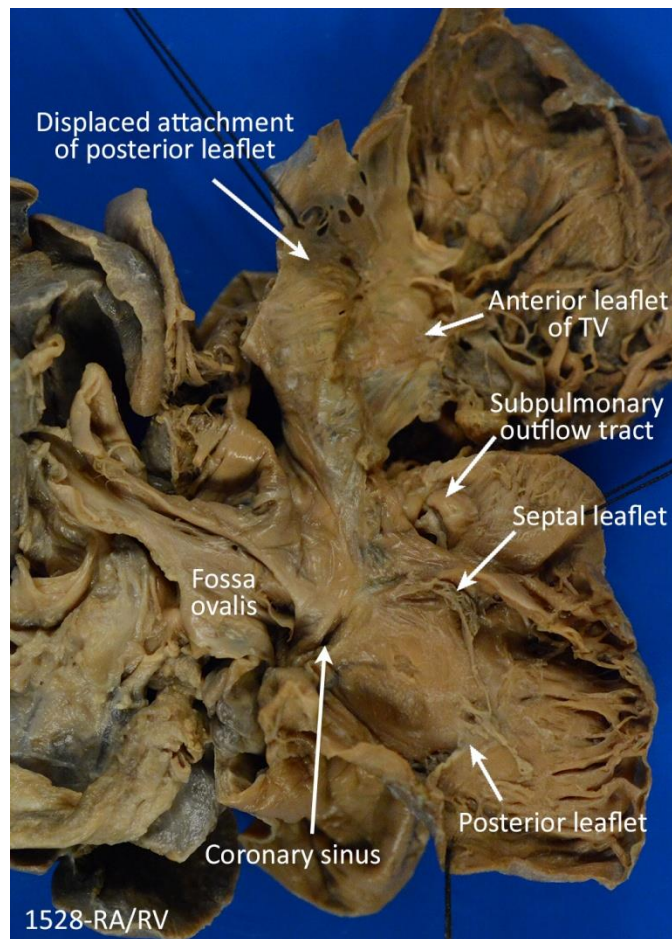
<input type="checkbox"/> Specimen: 1526 Categories: Dysplastic valves	
Pathology	<ul style="list-style-type: none"> • Dysplastic tricuspid valve without displaced attachment
Findings	<ul style="list-style-type: none"> • Dysplastic tricuspid valve leaflets with rolled-up edges. • Direct insertion of anterior and posterior leaflets to anterior papillary muscle.



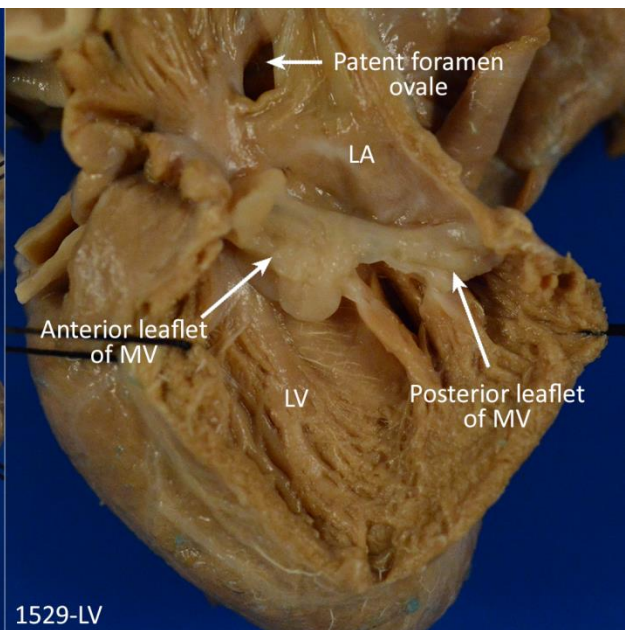
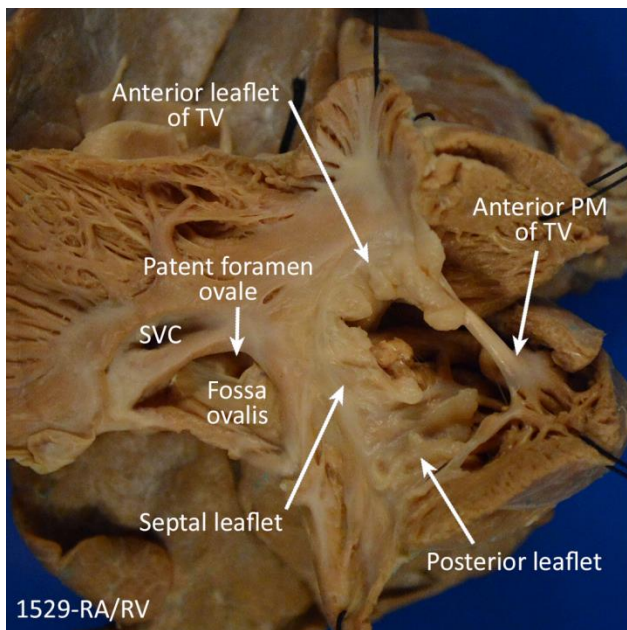
<input type="checkbox"/> Specimen: 1527	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly of tricuspid valve
Findings	<ul style="list-style-type: none"> • Displaced attachment of septal leaflet (marked by red dotted line on the right panel photo) in relation to tricuspid valve annulus (marked by white dotted line) • Large anterior and posterior leaflets are fused and inserts to the anterior papillary muscle (asterisk) through short chords. • Right atrial dilatation • Right ventricular dilatation • Mildly dilated left ventricle



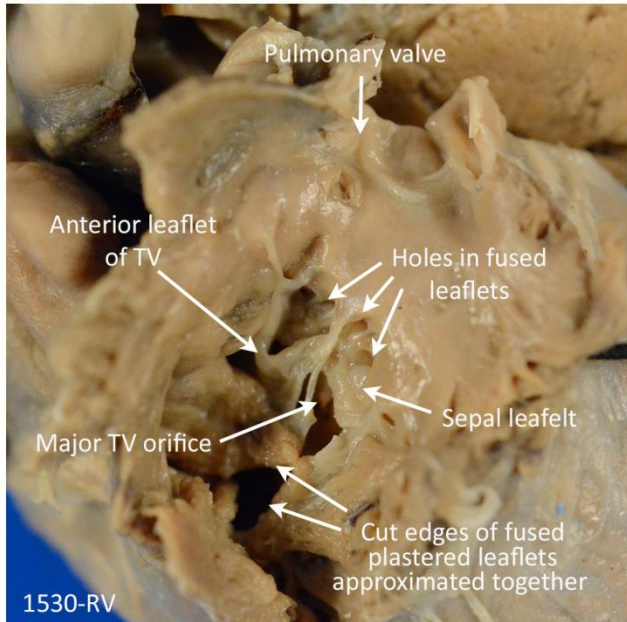
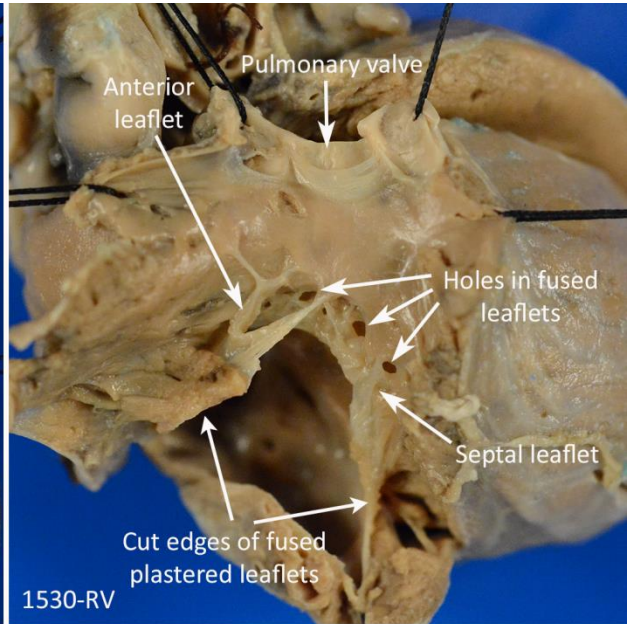
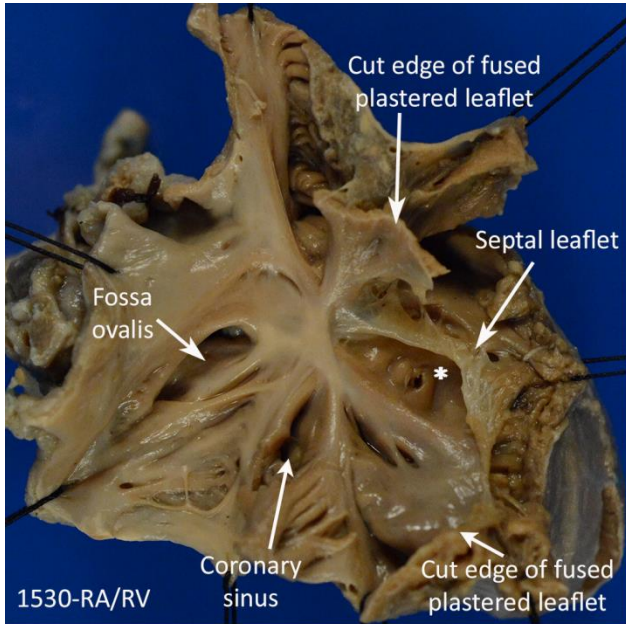
□	Specimen: 1528	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly of tricuspid valve with pulmonary valve stenosis 	
Findings	<ul style="list-style-type: none"> • Severely attenuated septal leaflet of tricuspid valve • Pulmonary valve stenosis • Right atrial dilatation • Dilated functional right ventricle with thin wall. 	



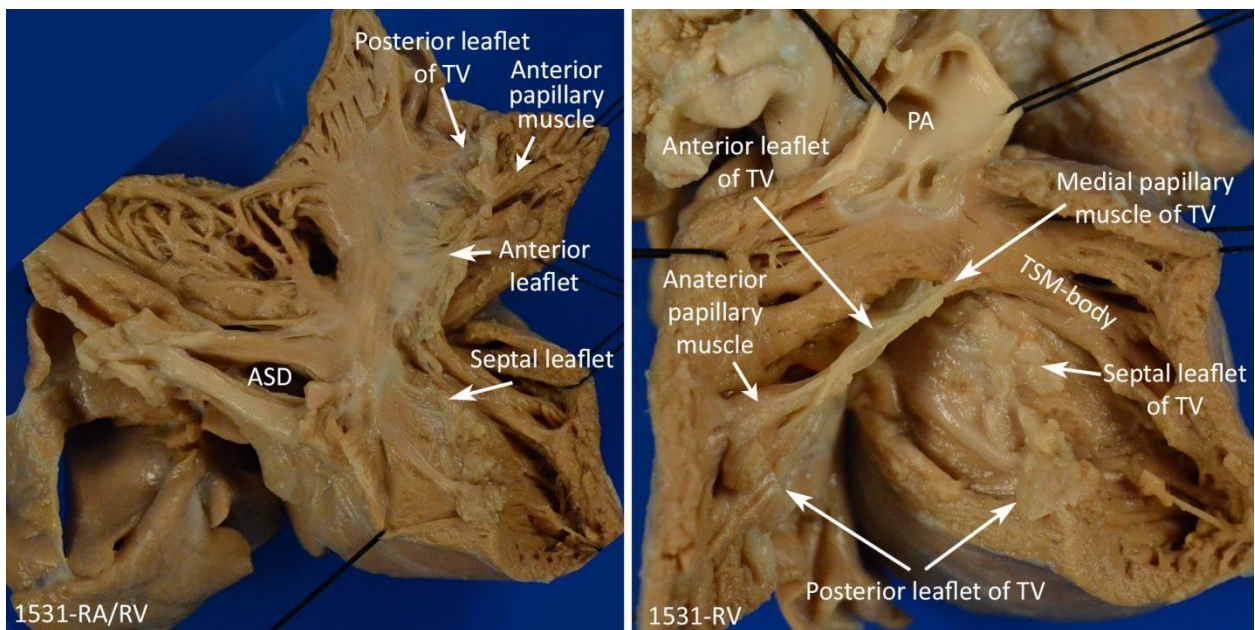
<input type="checkbox"/> Specimen: 1529 Categories: Dysplastic valves	
Pathology	<ul style="list-style-type: none"> • Dysplastic tricuspid and mitral valves
Findings	<ul style="list-style-type: none"> • Thick and dysplastic tricuspid and mitral valve leaflets • No displaced leaflet attachment of tricuspid valve • Dilated right and left atria • Dilated and hypertrophied right ventricle • Mildly thick semilunar valve leaflets • Patent foramen ovale • Small aorta



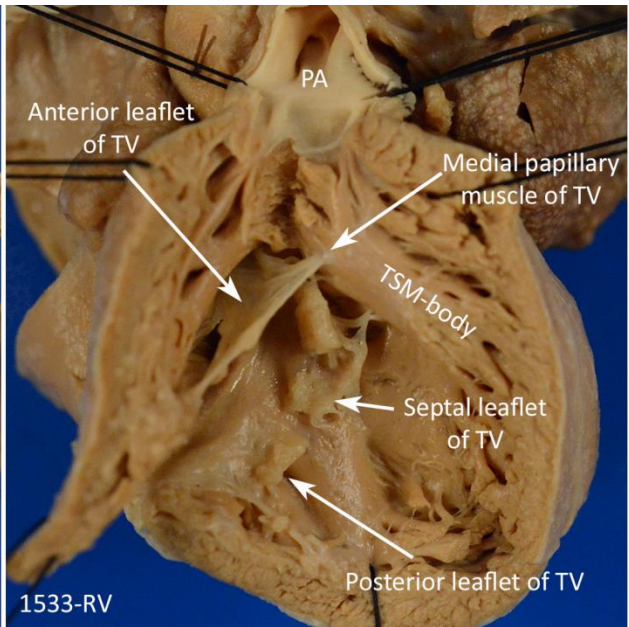
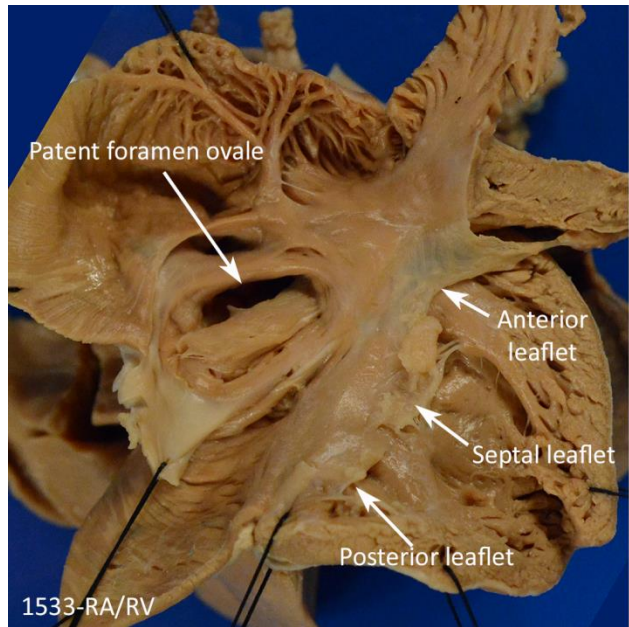
□ Specimen: 1530	Categories: Ebstein's anomaly
Pathology	• Stenotic Ebstein's anomaly
Findings	<ul style="list-style-type: none"> • Apically displaced attachment of dysplastic septal leaflet. Muscularized posterior leaflet. • Fused anterior, septal and posterior leaflets forming pouch-like sac along the septum. A part of the pouch behind the fused leaflets is marked by an asterisk. • Cut edges of fused leaflets are approximated in lower panel. Note small size of major orifice of the tricuspid valve and a tiny additional holes in the fused leaflets.



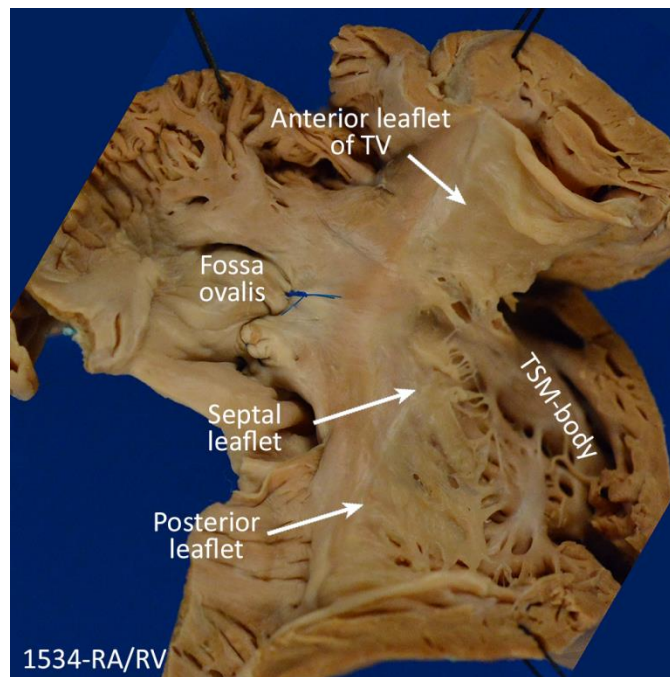
□ Specimen: 1531	Categories: Ebstein's anomaly
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly with small muscular VSD
Findings	<ul style="list-style-type: none"> • Apically displaced attachment of dysplastic septal leaflet and medial aspect of posterior leaflet • Normal attachment of mildly dysplastic sail-like anterior leaflet and adjacent part of posterior leaflet. • Short chords inserting to anterior papillary muscle of tricuspid valve • Atrial septal defect



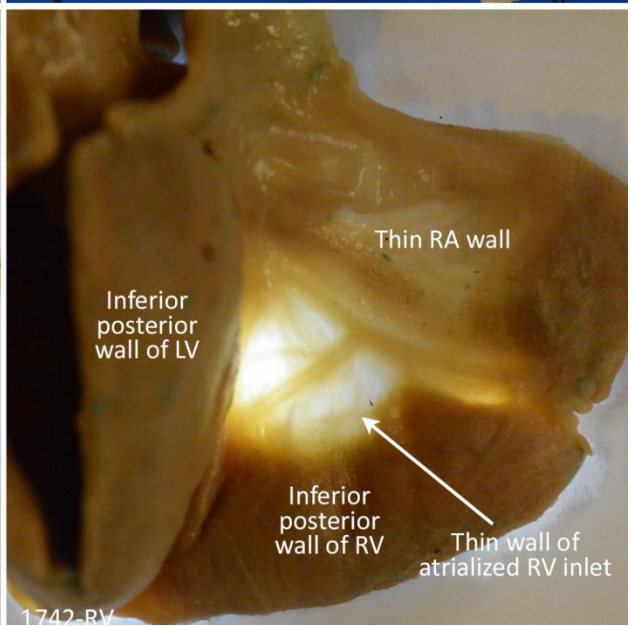
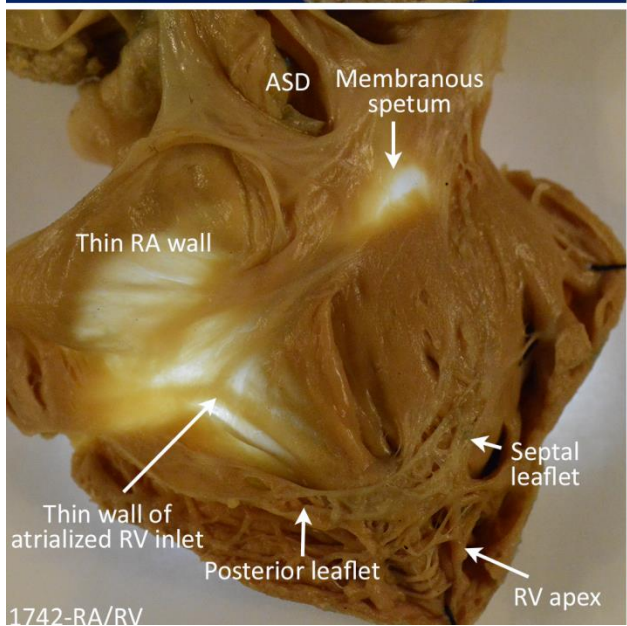
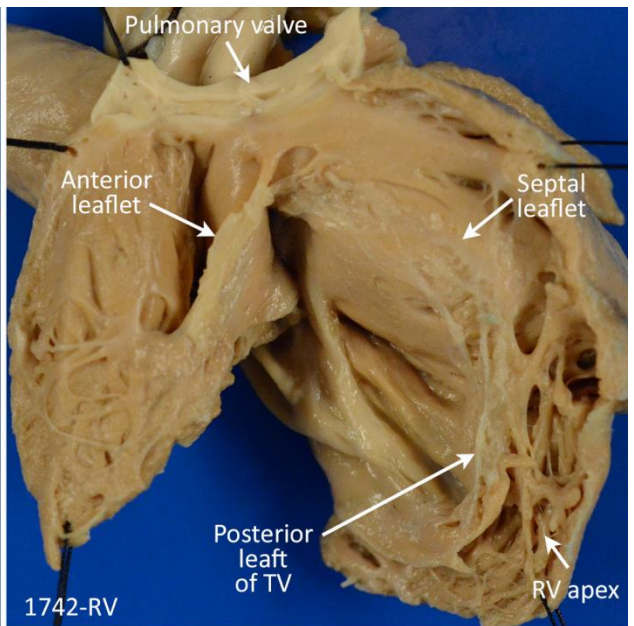
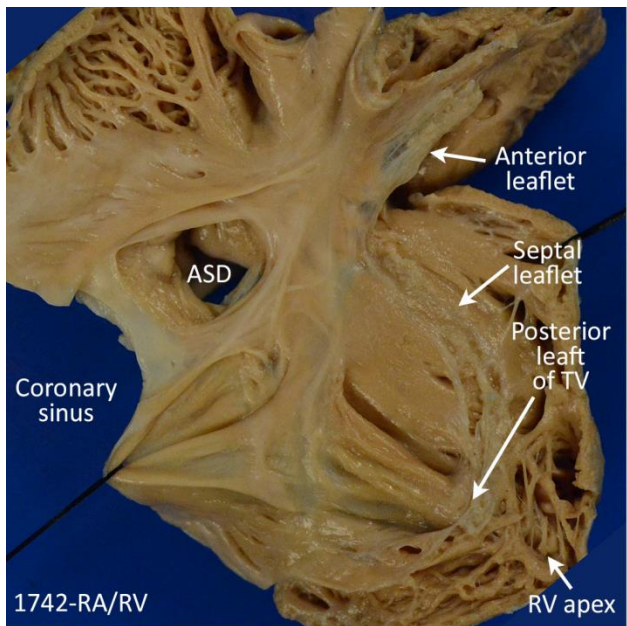
<input type="checkbox"/> Specimen: 1533 Categories: Ebstein's anomaly	
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly. Post-repair
Findings	<ul style="list-style-type: none"> • Mildly displaced attachment of septal and posterior leaflets of tricuspid valve. • Patent foramen ovale



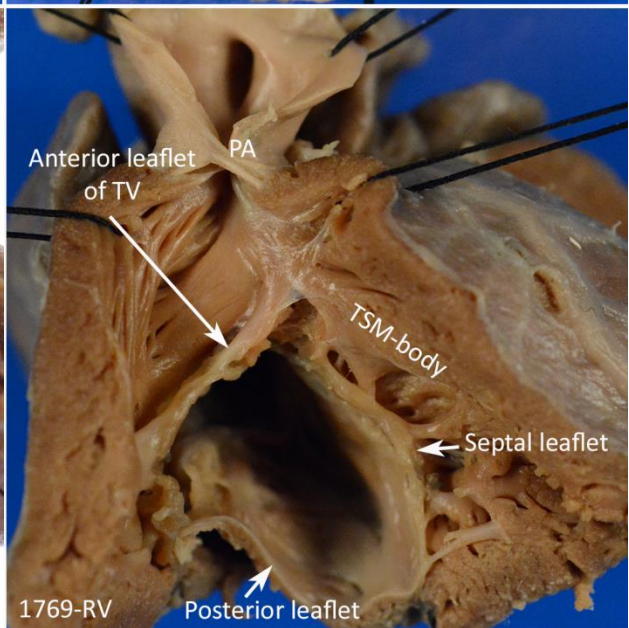
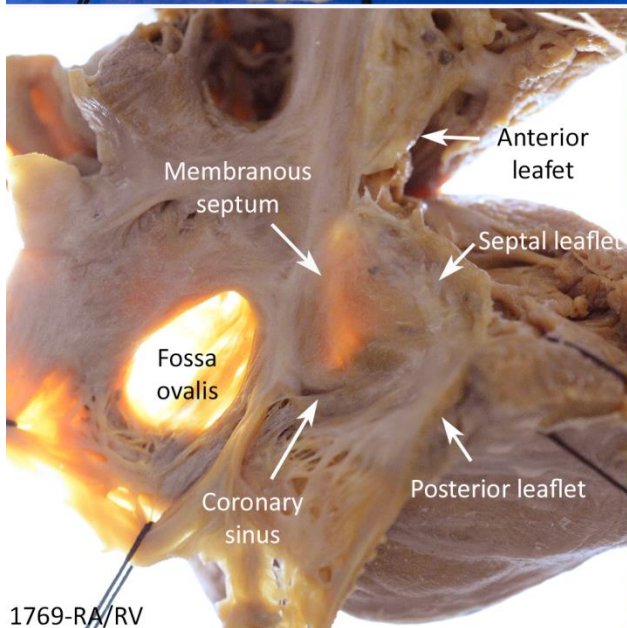
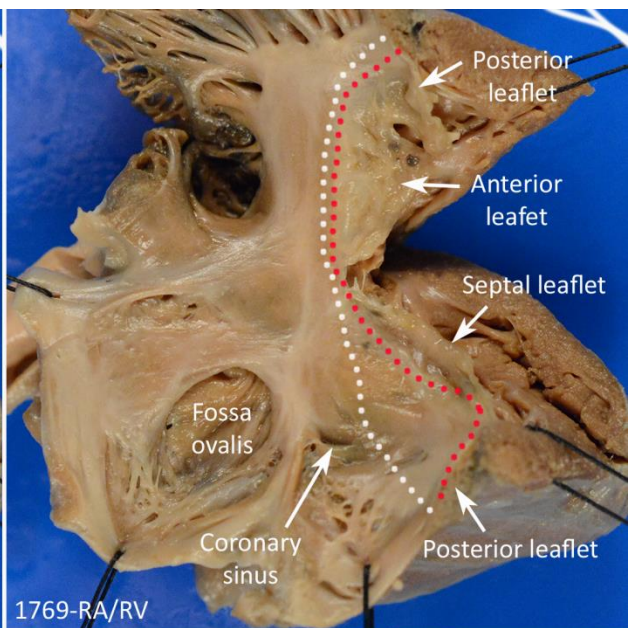
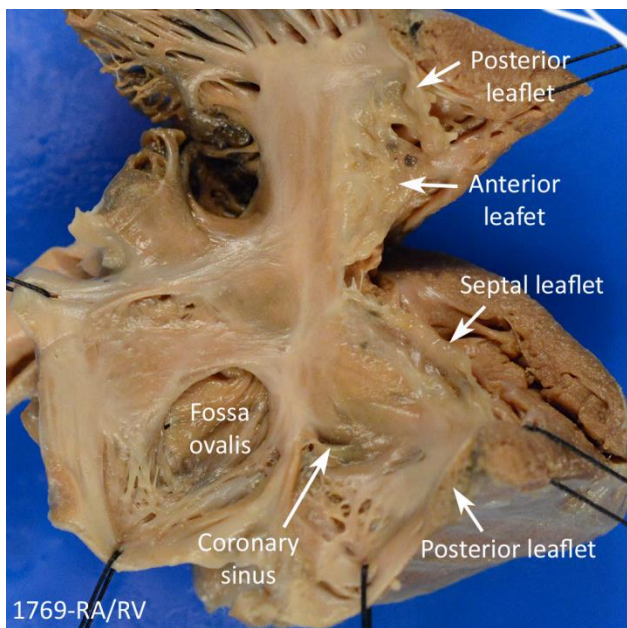
□ Specimen: 1534	Categories: Dysplastic valves
Pathology	<ul style="list-style-type: none"> • Dysplastic tricuspid valve
Findings	<ul style="list-style-type: none"> • Moderately severe dysplasia of anterior leaflet and mildly dysplastic septal and posterior leaflets of tricuspid valve • Mildly displaced attachment of septal leaflet toward the apex • Pulmonary vein stenosis, stented



<input type="checkbox"/> Specimen: 1742 Categories: Ebstein's anomaly	
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly with very thin atrialized part of right ventricle
Findings	<ul style="list-style-type: none"> • Markedly displaced attachment of small dysplastic septal and posterior leaflets • Very thin wall of the atrialized part of the right ventricular inlet transilluminated from outside (left lower panel) and from inside (right lower panel). Anatomical atrioventricular junction is demarcated by the coronary artery branch. • Dilated right atrium. Atrial septal defect. • Dilated right ventricle.



<input type="checkbox"/> Specimen: 1769 Categories: Ebstein's anomaly	
Pathology	<ul style="list-style-type: none"> • Ebstein's anomaly with hypoplastic septal leaflet and severely attenuated posterior leaflet.
Findings	<ul style="list-style-type: none"> • Displaced attachment of septal and posterior leaflets of tricuspid valve (marked by red dotted line. Tricuspid valve annulus is marked by white dotted line).. • Normal attachment of anterior leaflet of tricuspid valve • Hypoplastic septal leaflet. Severely attenuated posterior leaflet. • Large membranous septum as shown in transilluminated photo in left lower panel.



<input type="checkbox"/> Specimen: 2408 Categories: Ebstein's anomaly	
Pathology	<ul style="list-style-type: none"> • Explanted stenotic Ebstein's anomaly. S/P Tricuspid valve plasty
Findings	<ul style="list-style-type: none"> • Displaced attachment of septal and posterior leaflets of tricuspid valve. • Normal attachment of anterior leaflet of tricuspid valve • Fusion of most parts of the free edges of the leaflets leaving a few small openings, forming a so-called tricuspid valve sac • Small openings in the fused valve leaflets are the only egressing routes for right atrial flow to the right ventricle.

