

# Conduction Tissue Disposition in Abnormalities of the AV Junction

2<sup>nd</sup> Contemporary Morphology Course  
Abnormalities of the Atrioventricular Junction

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Assistant Staff Cardiologist & Electrophysiologist

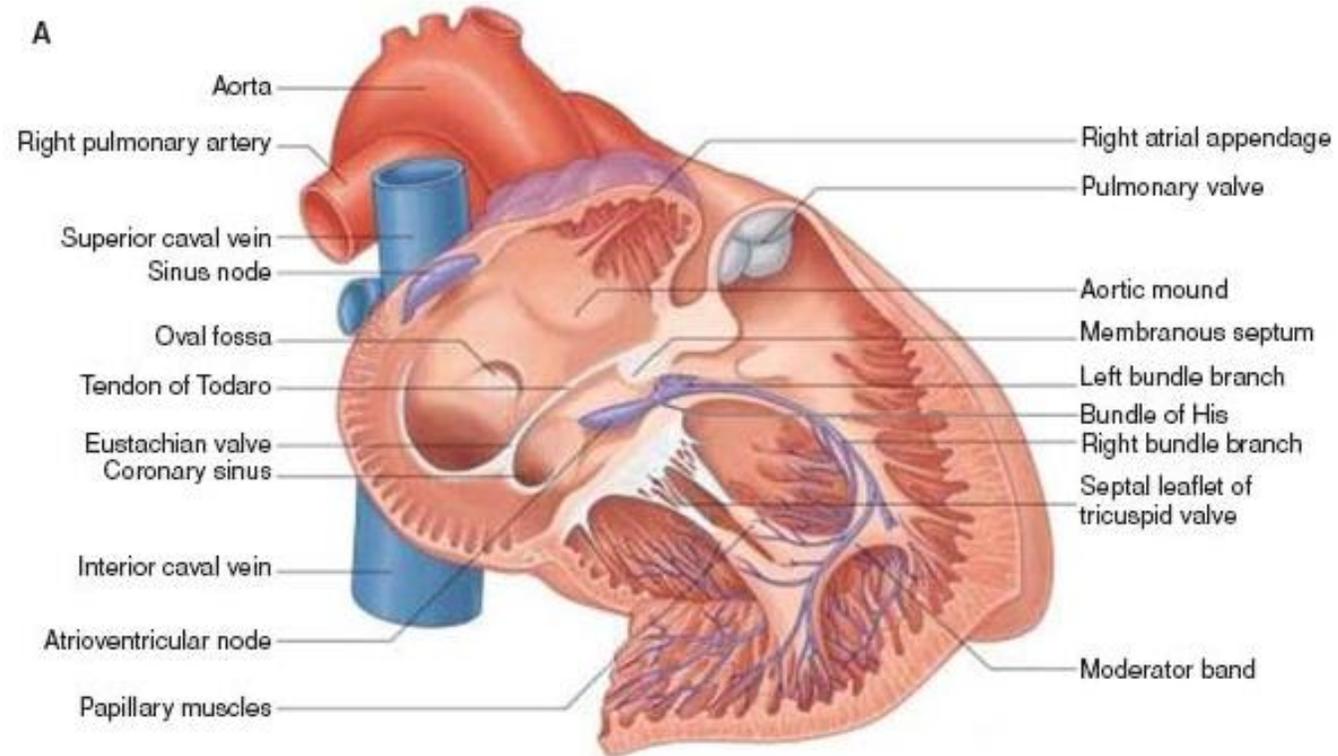
The Hospital for Sick Children

Sunday, December 8<sup>th</sup>, 2019

# Outline

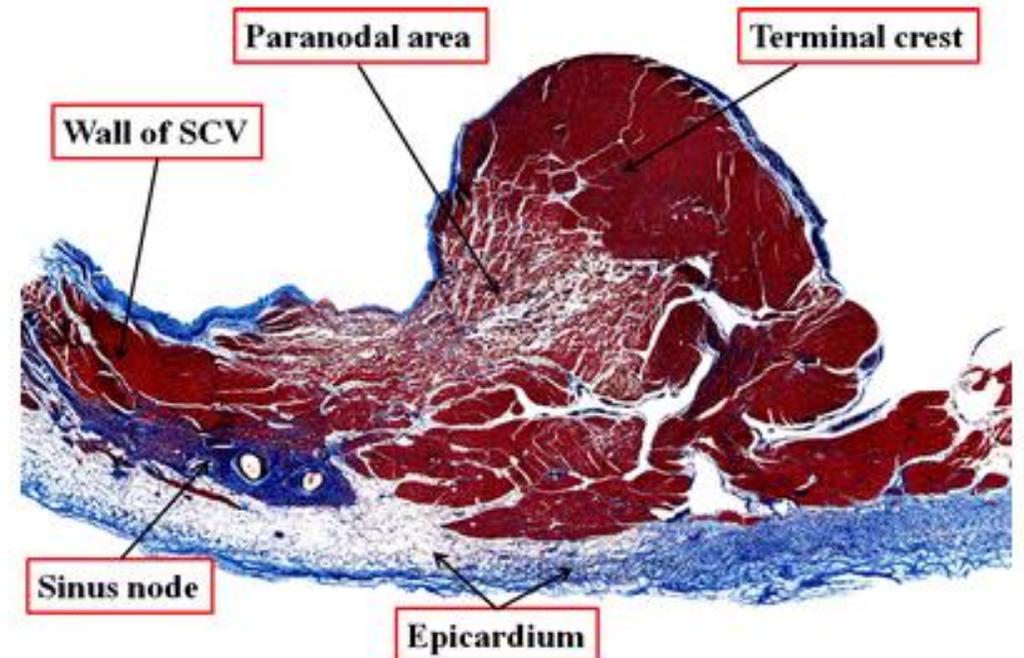
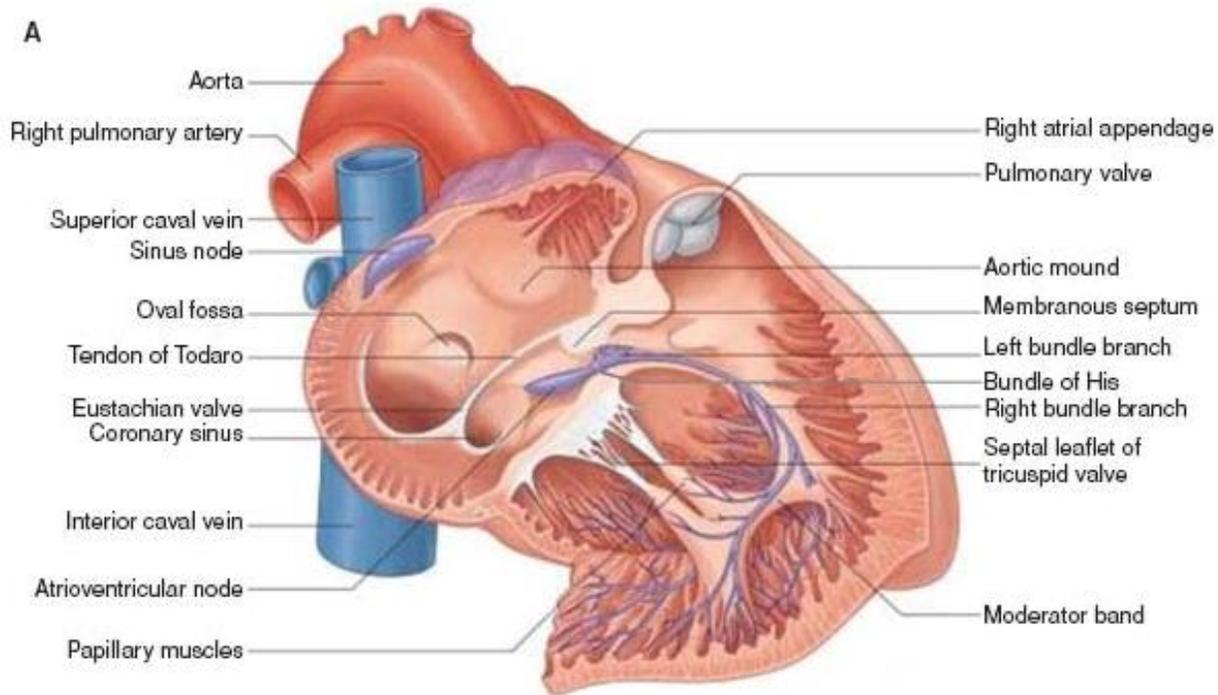
- Structurally normal heart
- Ventricular septal defects (VSD)
- Atrioventricular septal defects (AVSD)
- Ebstein's anomaly of the tricuspid valve
- Atrioventricular and ventriculoarterial discordance
- Univentricular atrioventricular connections

# Structurally Normal Heart



Anderson, J Cardiovasc Trans Res 2013  
Grey's Anatomy 41<sup>st</sup> Ed 2015

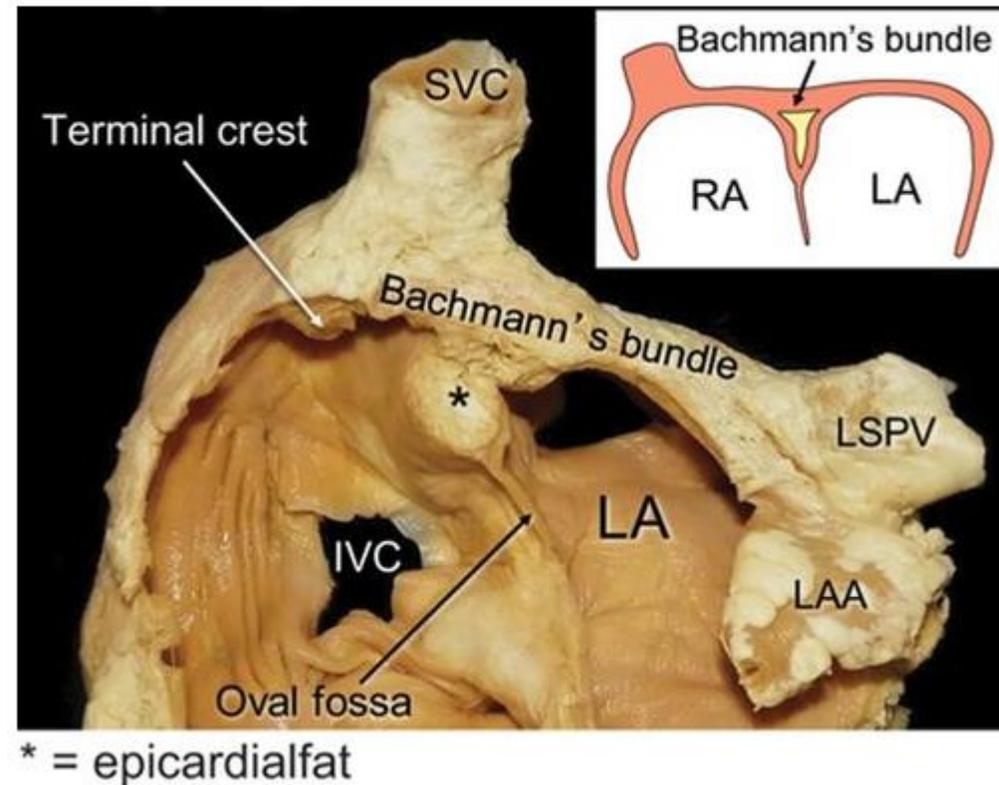
# Structurally Normal Heart: SA Node



Anderson, J Cardiovasc Trans Res 2013

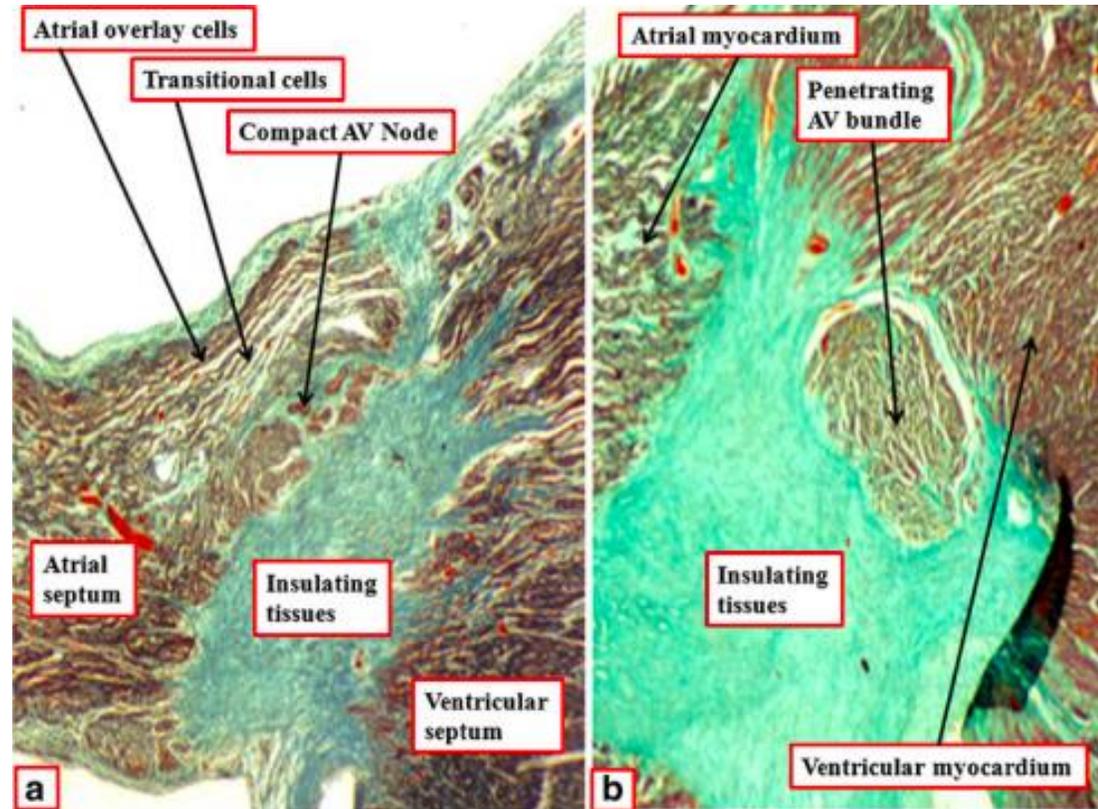
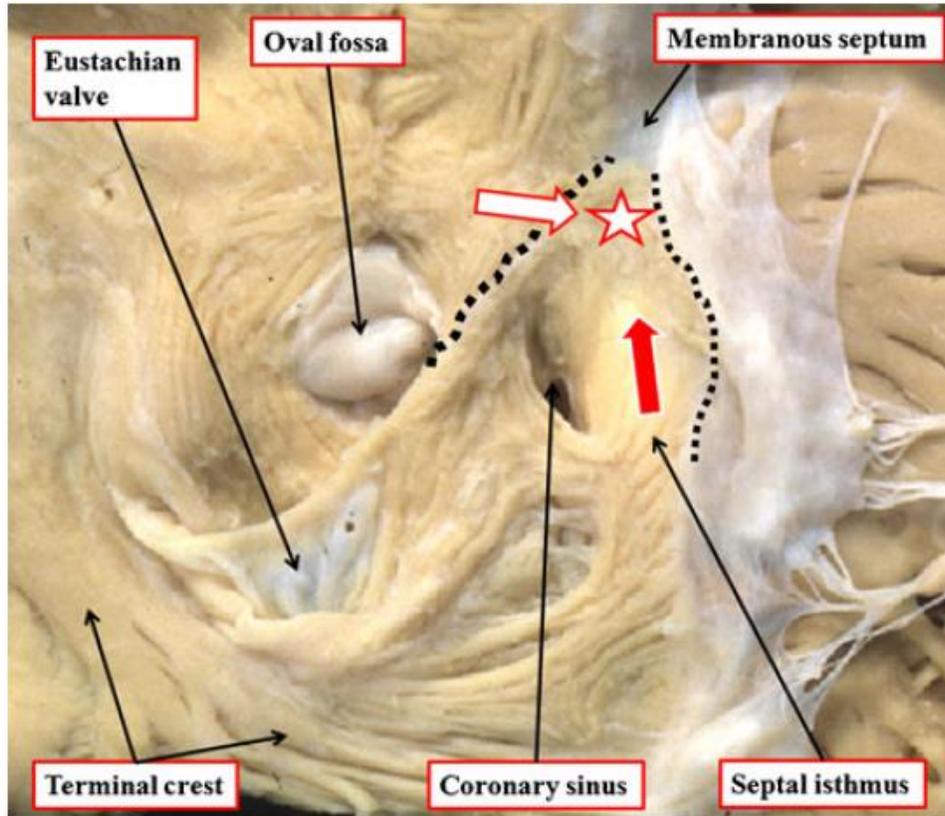
# Structurally Normal Heart: Internodal Atrial Myocardium

- Electrical impulses that emerge from the SA node are propagated more efficiently along the long axis of atrial muscle bundles than it is transversely.
- Conduction to the left atrium is made through Bachmann's bundle with additional pathways through fossa ovalis.



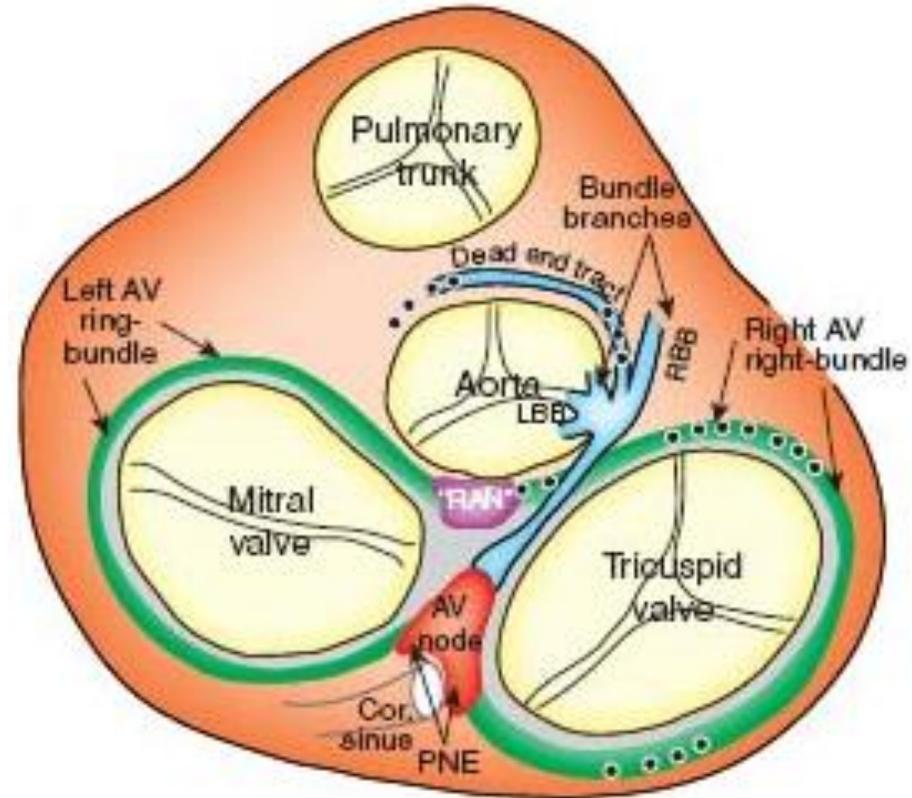
Anderson, J Cardiovasc Trans Res 2013  
Van Campenhout, Circ Arrhythm Electrophysiol  
2013  
Spach, Circ Research, 1971

# Structurally Normal Heart: AV Node



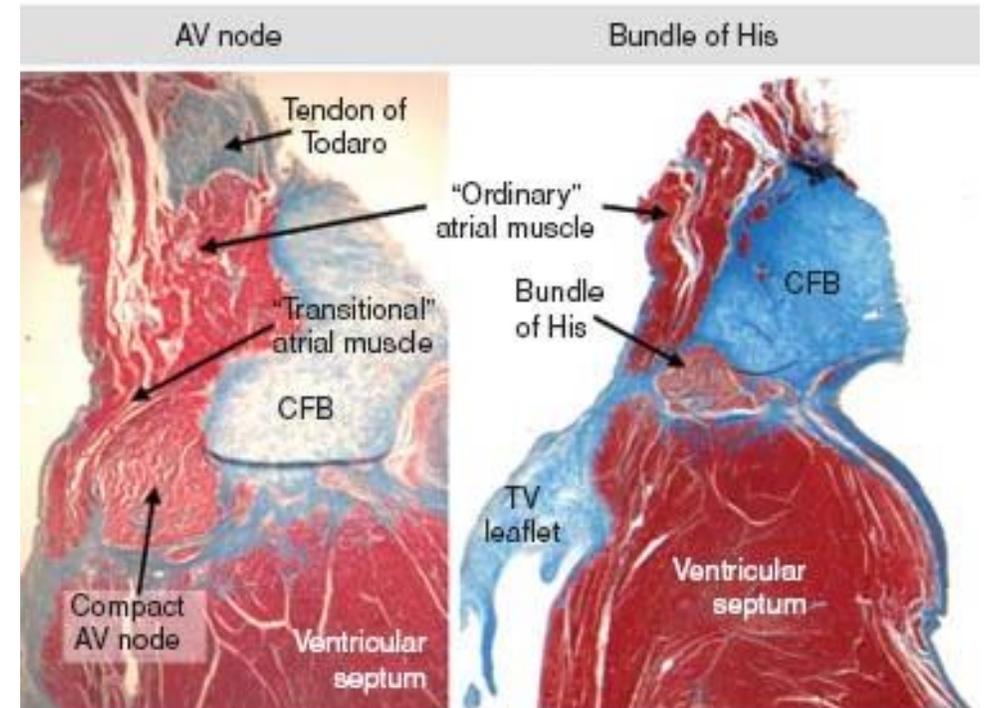
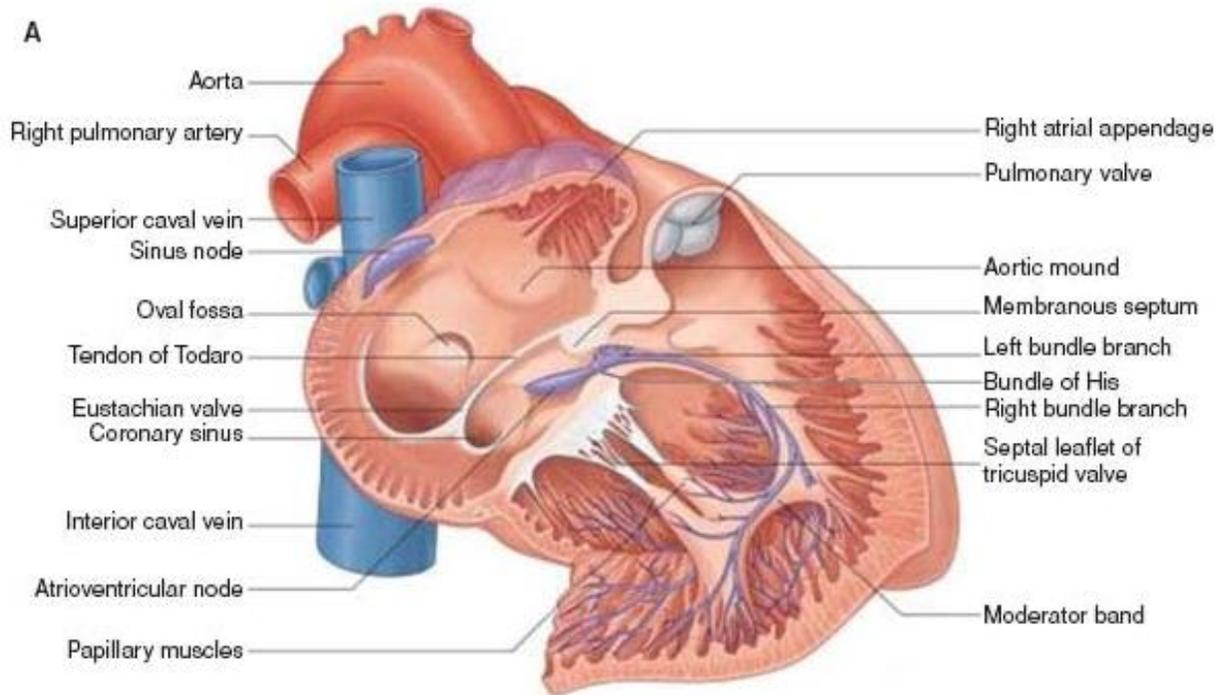
Anderson, J Cardiovasc Trans Res 2013

# Structurally Normal Heart: AV Ring Tissues



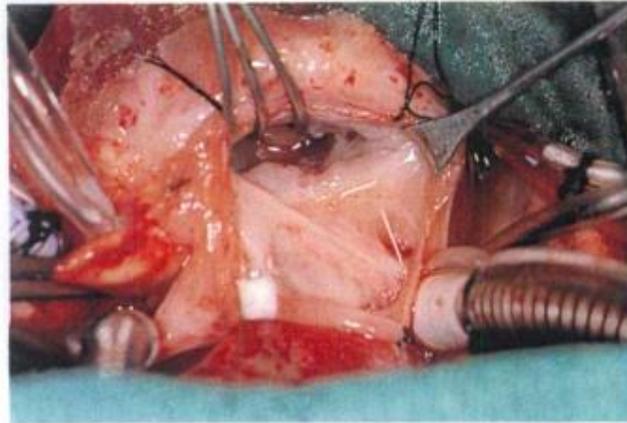
Yanni, Heart Rhythm 2009

# Structurally Normal Heart: Bundle of His

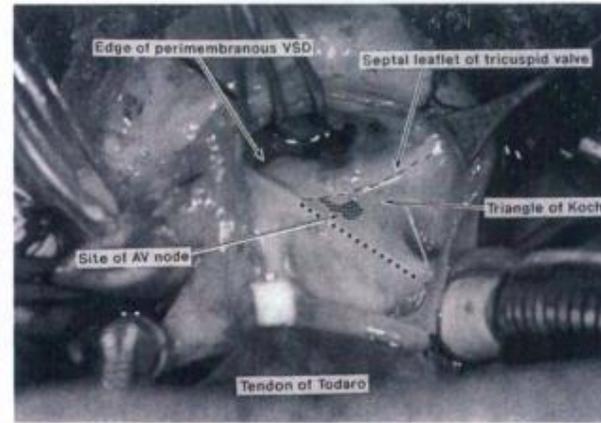


Grey's Anatomy 41<sup>st</sup> Ed 2015  
Moss and Adam's 9<sup>th</sup> Ed 2016

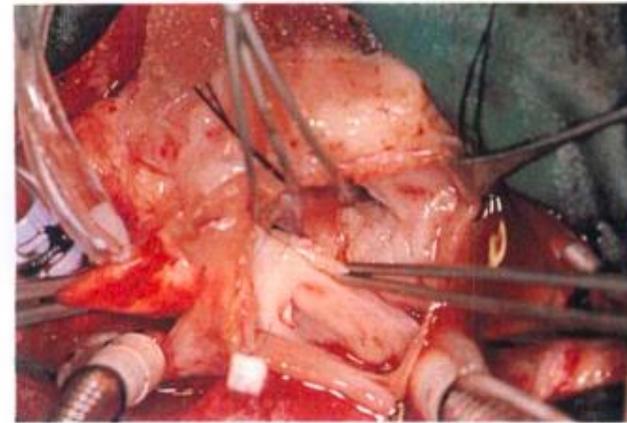
# Ventricular Septal Defects (VSD): Perimembranous



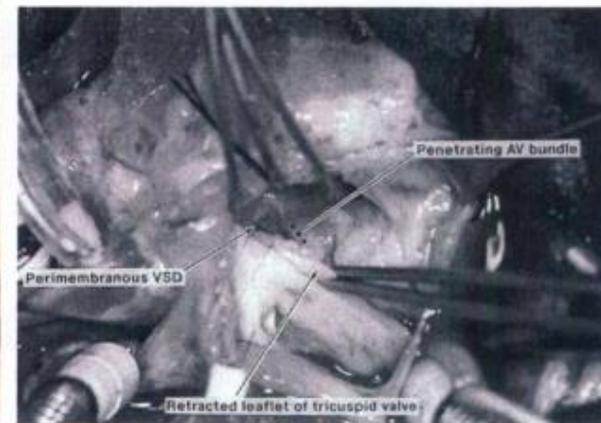
A



B



C

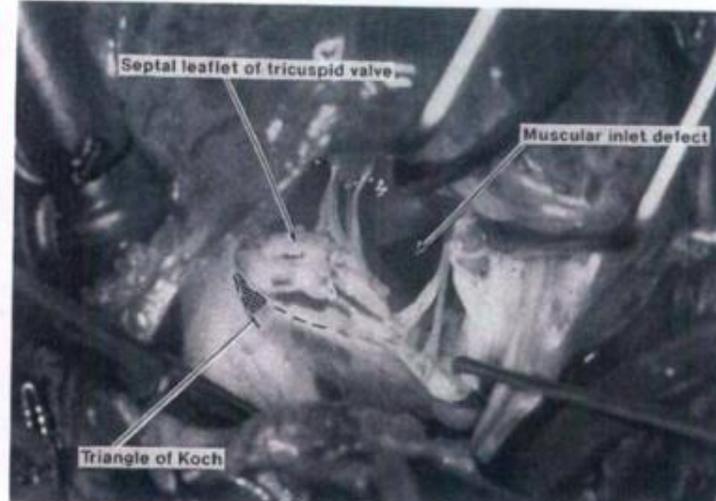
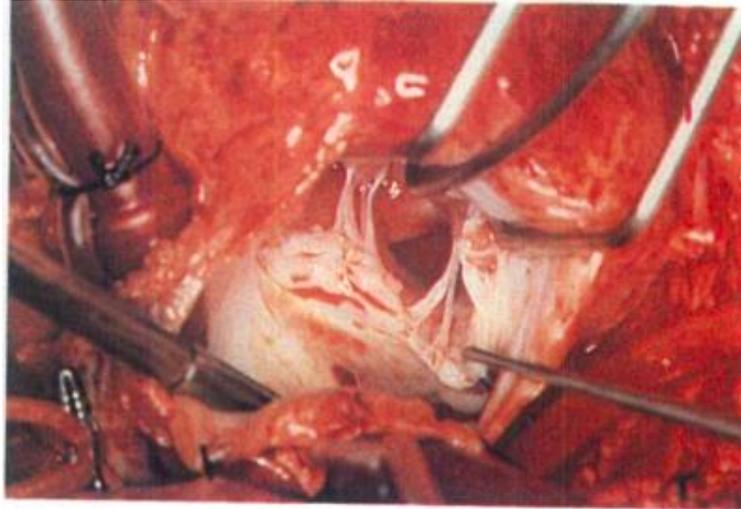


D

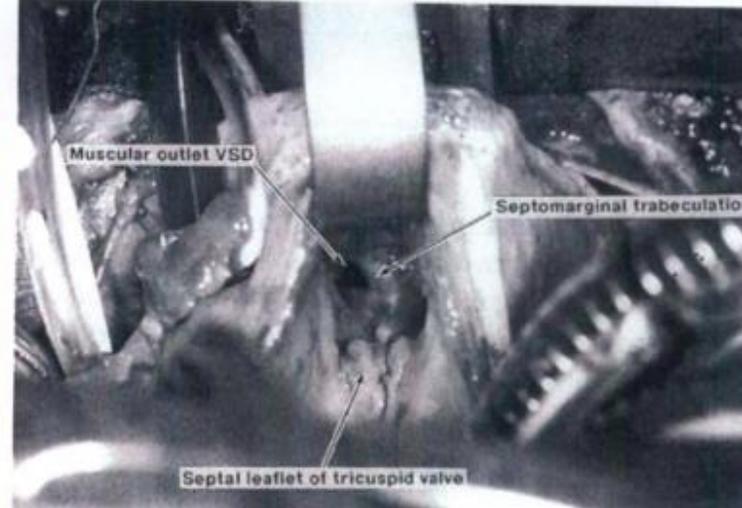
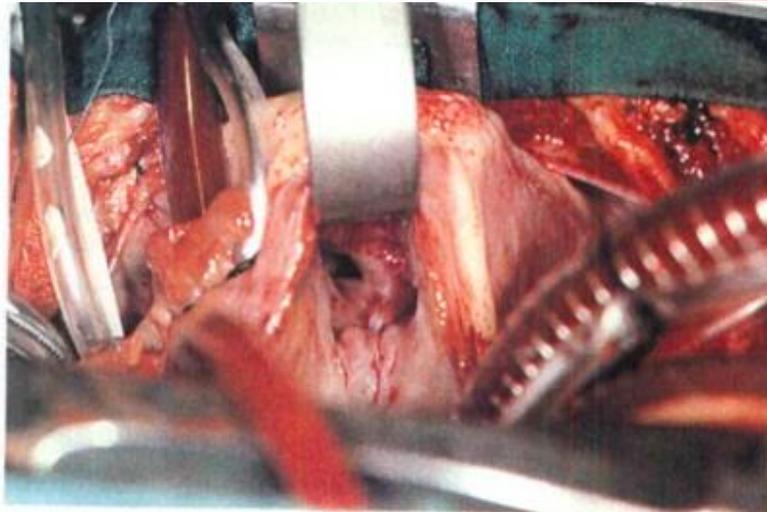
Anderson, J Cardiovasc Surg 1992

# Ventricular Septal Defects (VSD): Muscular

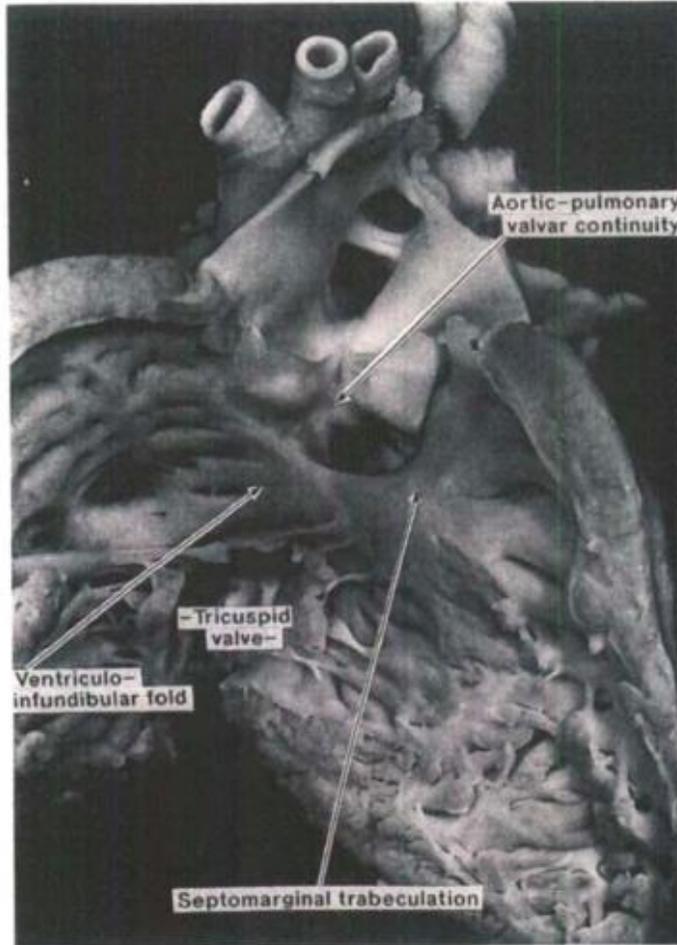
Inlet Extension



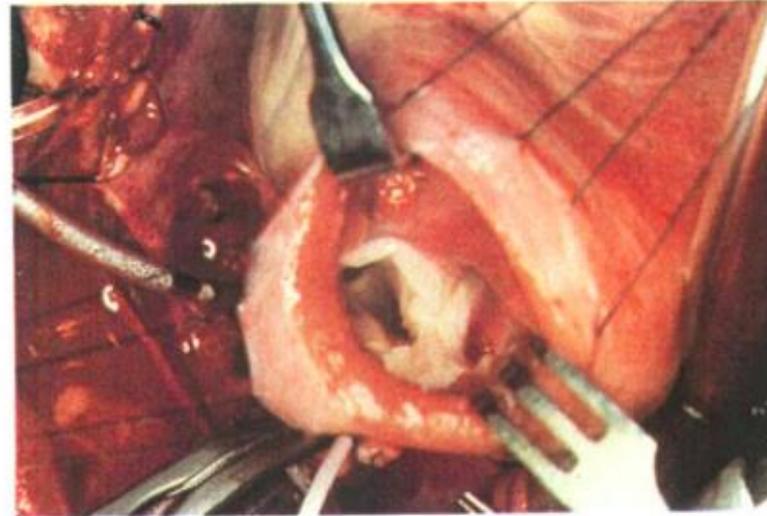
Outlet Extension



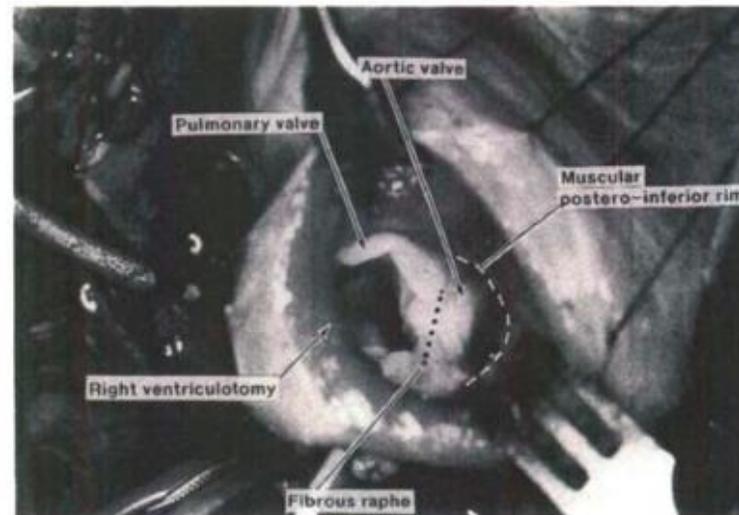
# Ventricular Septal Defects (VSD): Doubly Committed



A



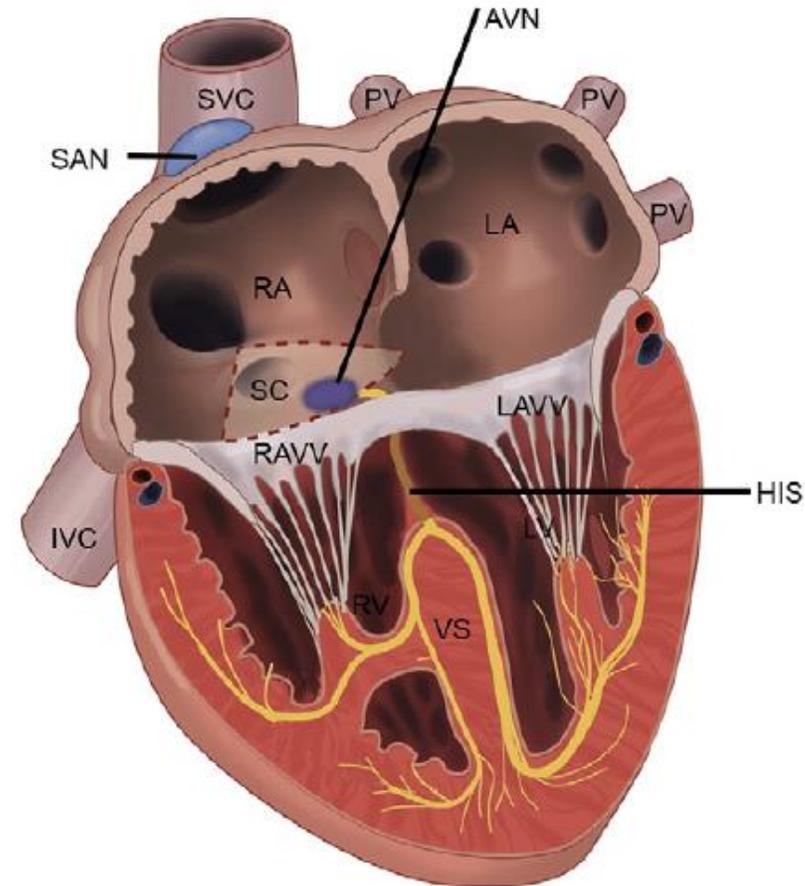
B



Anderson, J Cardiovasc Surg 1992

# Atrioventricular Septal Defects

- AV node is displaced posteroinferiorly, near the CS os
- Bundle of His is displaced inferiorly and penetrates the central fibrous body, traveling towards the LV aspect of the crest of the ventricular septum
- Short distance between AV node and origin of LBB
- Posteroinferior displacement of LBB
- Hypoplasia of anterior LBB



Feldt, Circulation 1970

Calkoen, Int J Cardiology 2016

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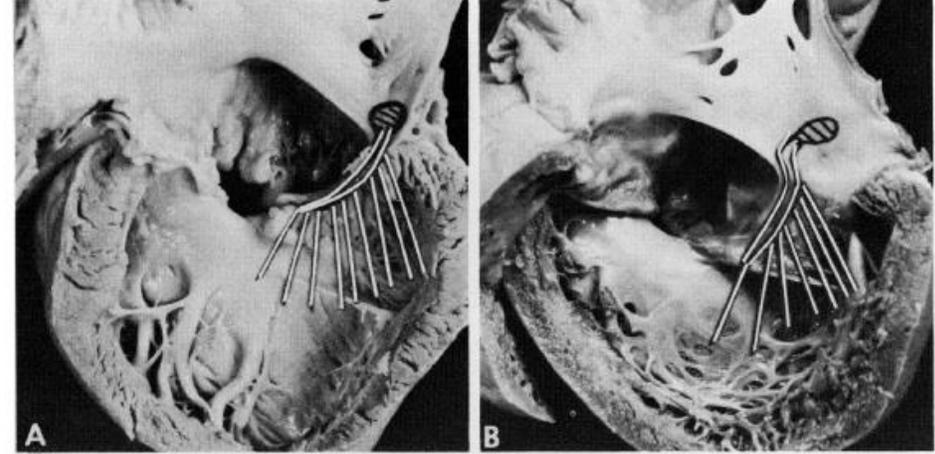
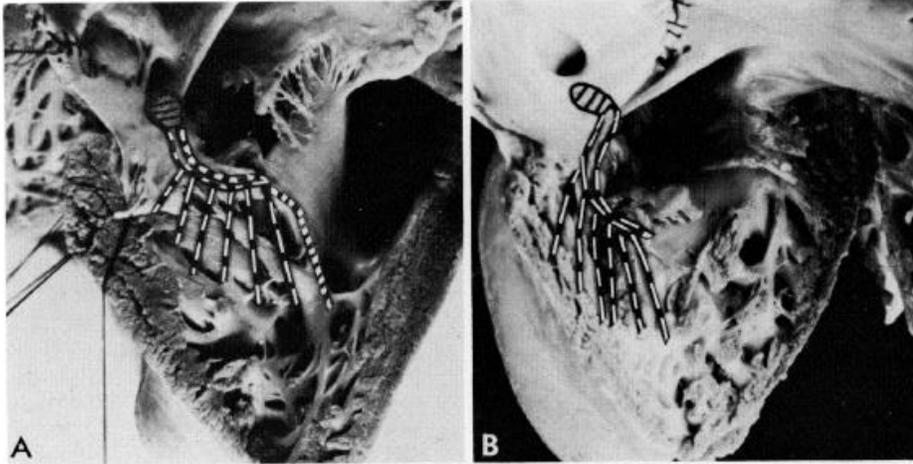


Feldt, Circulation 1970

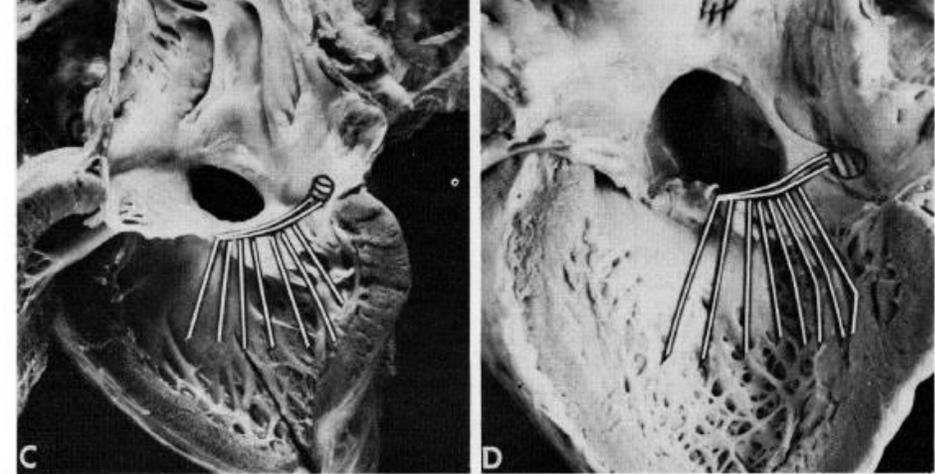
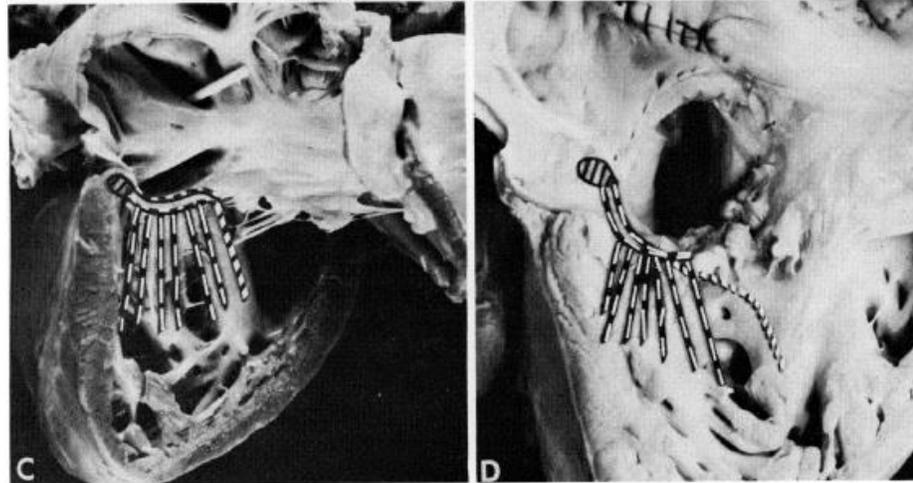
Calkoen, Int J Cardiology 2016

# Atrioventricular Septal Defects

Complete AVSD



Partial AVSD



Right-sided view

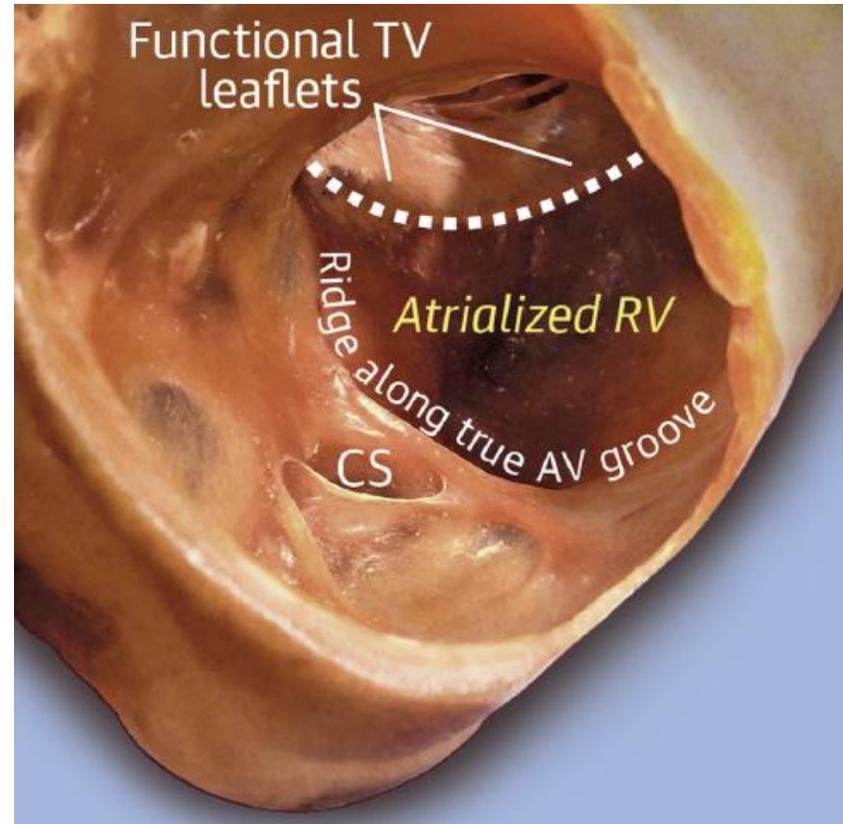
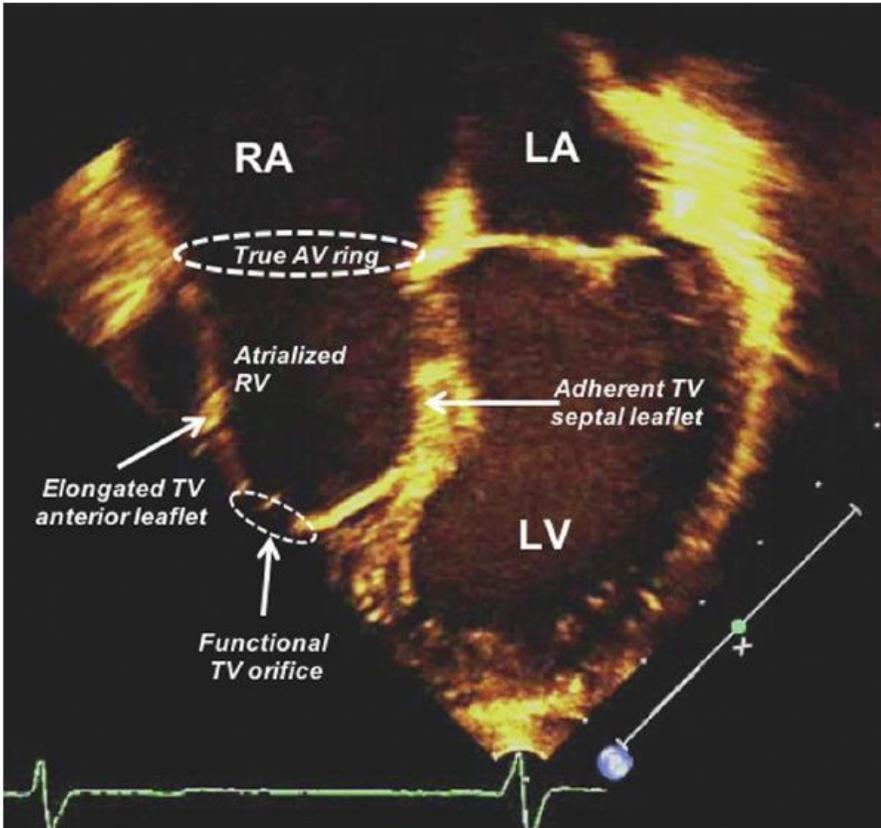
Left-sided view

# Atrioventricular Septal Defects

- AV node is displaced laterally away from CS os in AVSD with malalignment between the atrial and ventricular septums with L-dominance
- AV node is displaced rightwards in L-SVC draining via CS to LA
- AV node is extremely close to CS os in AVSD with poorly developed tendon of Todaro.
- *Site of AV node emergence is located where inferior bridging leaflet crosses ventricular septum*

Seo, J Cardiac Surgery 1992

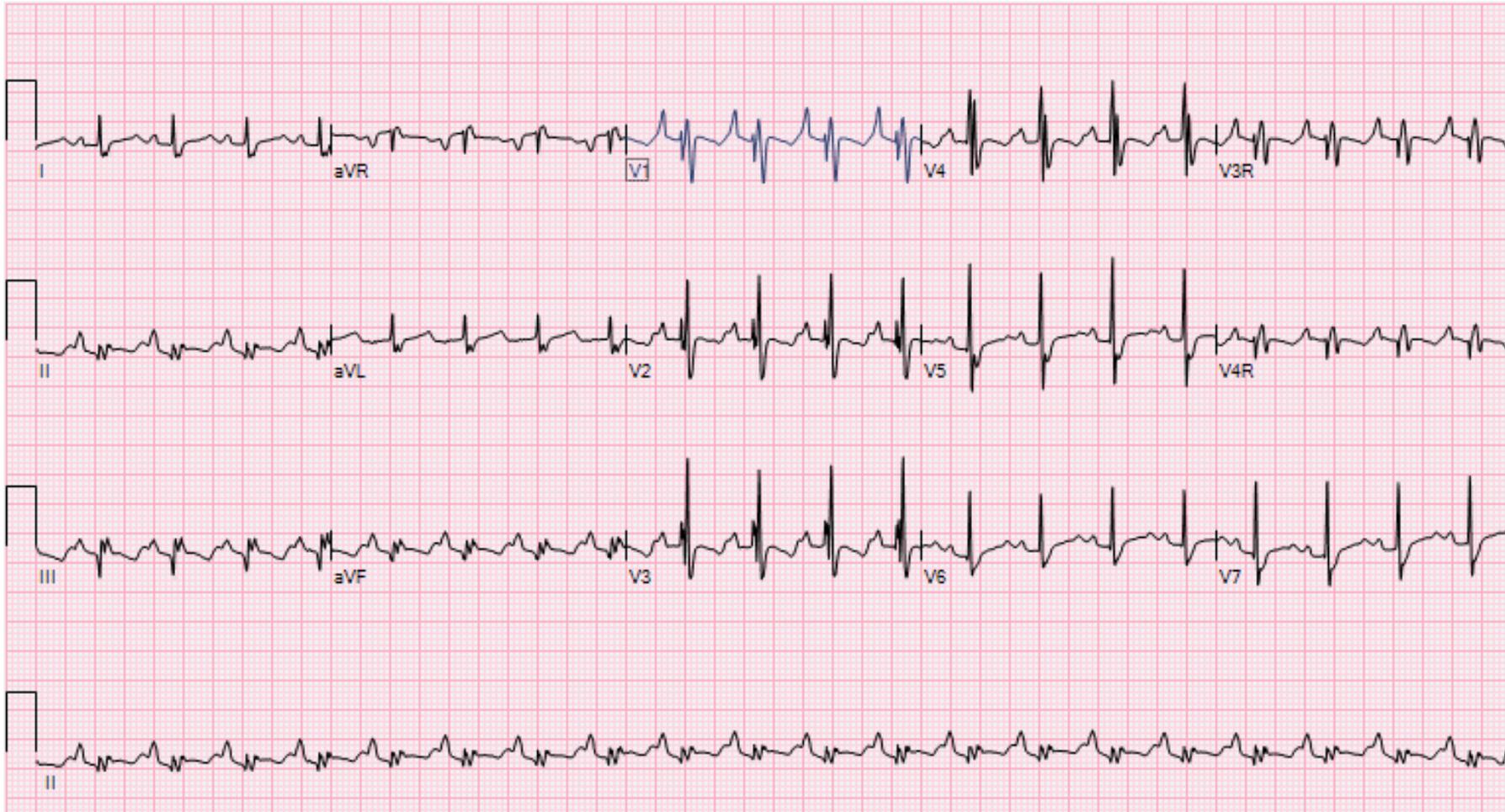
# Ebstein's anomaly of the TV



- SA and AV nodes are in normal locations
- Increased intra-RA conduction delay → PR prolongation
- His in normal location but intra-His and infra-nodal delay → '2<sup>nd</sup> QRS'
- RBB abnormalities

Walsh, JACC: Clin Electro 2018  
Sanchez-Quintana, Rev Esp Cardiol 2010  
Ho, Heart 2000  
Kastor, Circulation 1975

# Ebstein's anomaly of the TV



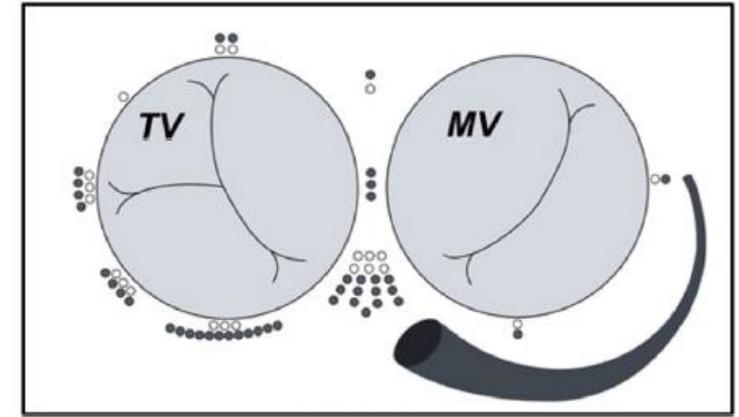
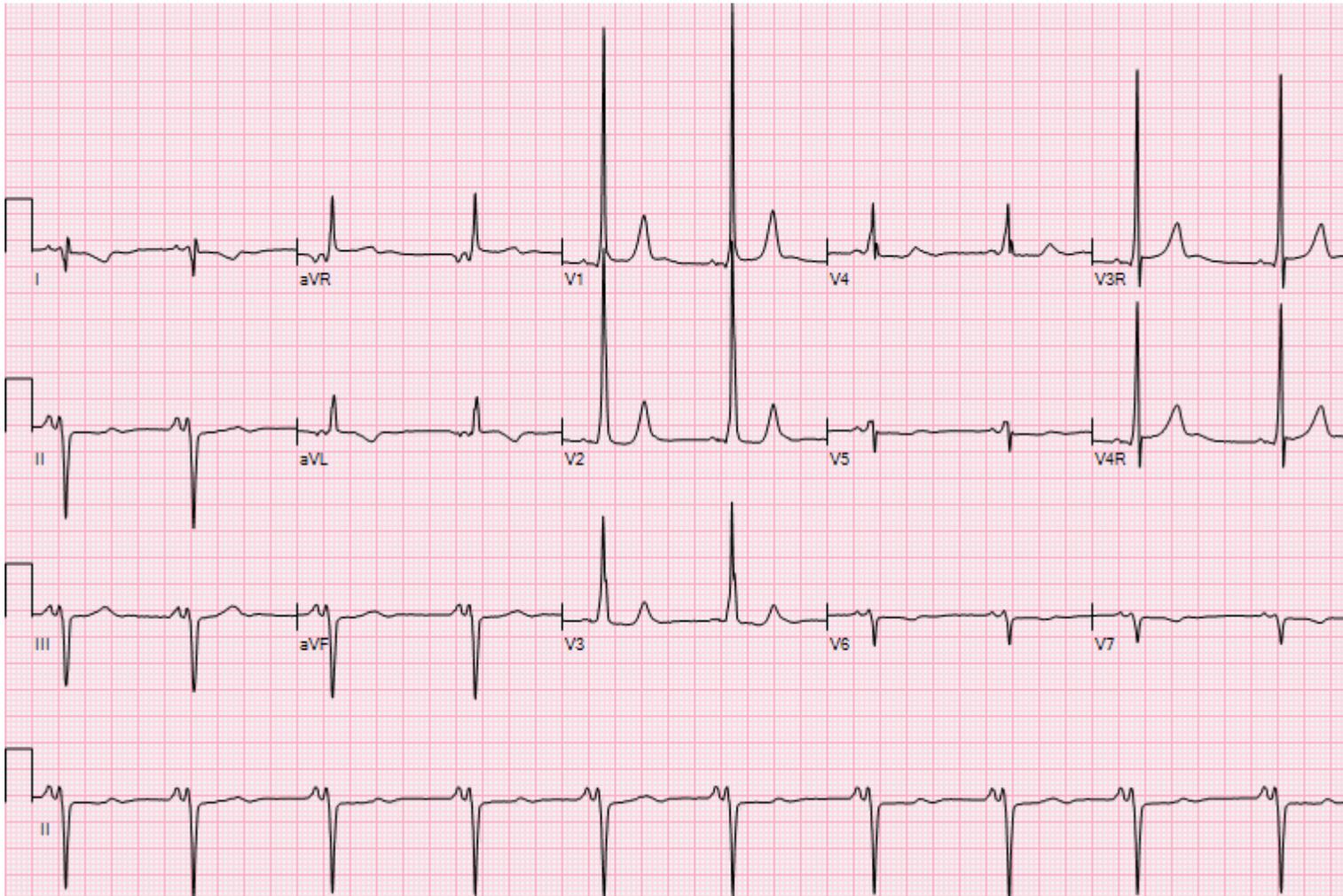
# Ebstein's anomaly of the TV

*Electrophysiologic Measurements in Five Patients with Ebstein's Anomaly of the Tricuspid Valve*

Pt	P-R	P width	P-A	A-H	H-V	BH duration	QRS	Prox ARV	Dist ARV	RVA	RVOT	LVA	VF
D.G. (NSR)	120	110	35	60	25	30	120	50	40	65	60		Yes
D.G. (WPW)							165	95	70	70	100		
D.G. (SVT)				70	60	30	110	35					
W.M.	220	140	65	80	65	30	180				80		Yes
M.R.	180	110	50	95	65	20	165	145	25	70	50		No
W.V.	440	160	50	160*	80	20	120	60	75	45		0	No
A.E.	200	120	40	90	50	15	110	70	80	30			No

Kastor, Circulation 1975

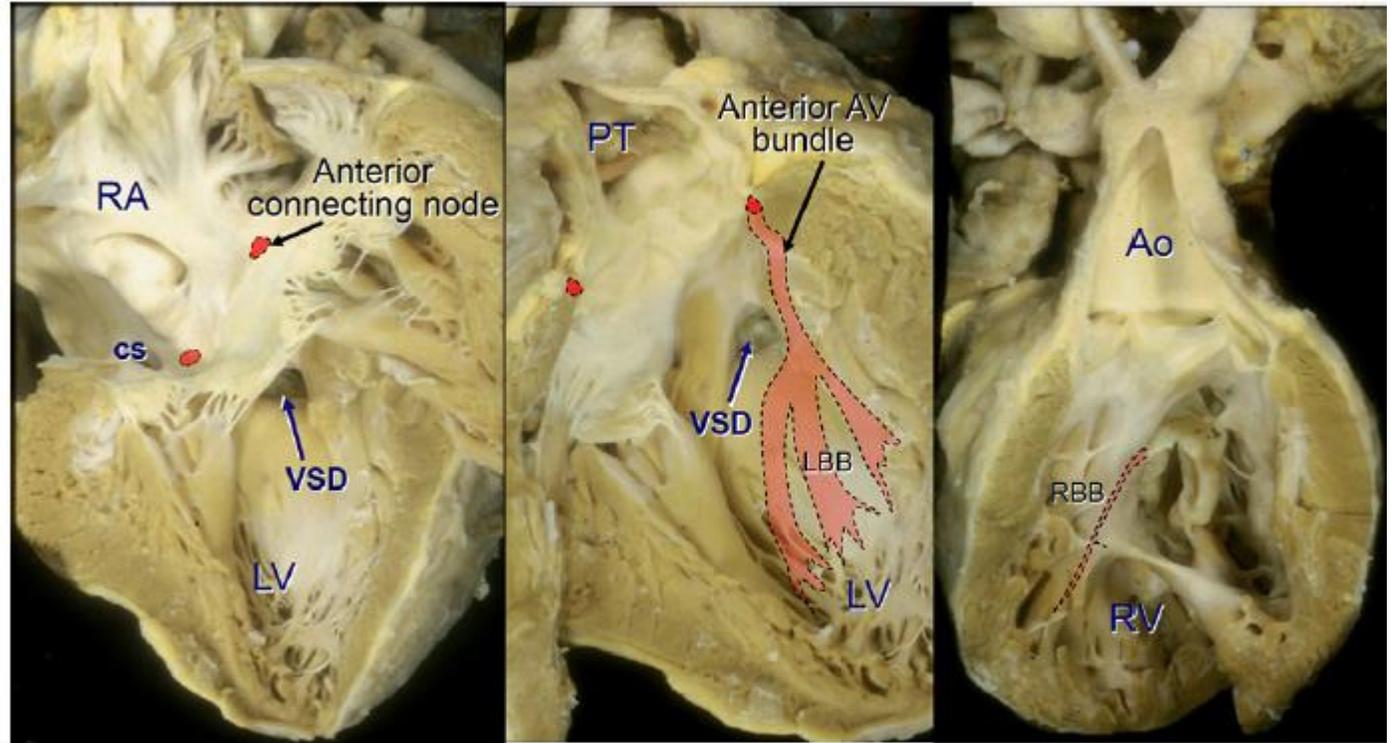
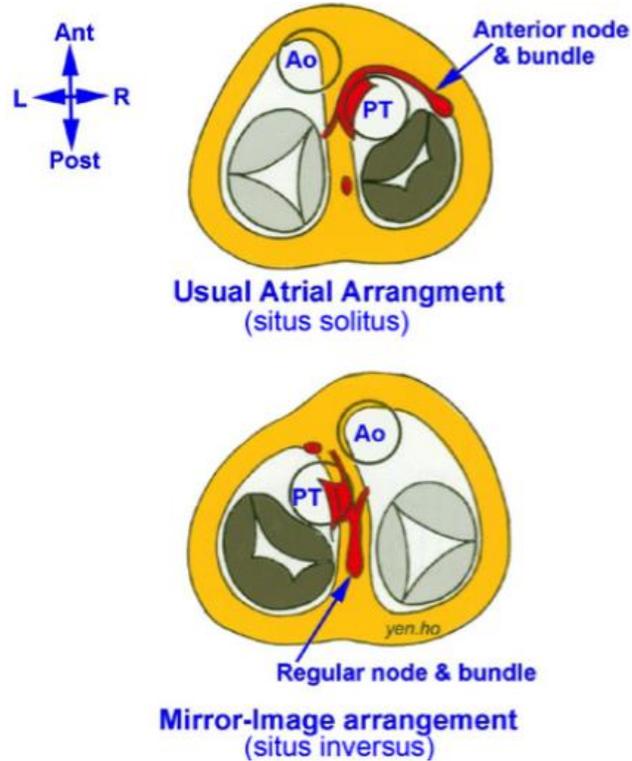
# Ebstein's anomaly of the TV



**TABLE 1 Arrhythmias Encountered in Patients in Ebstein's Anomaly**

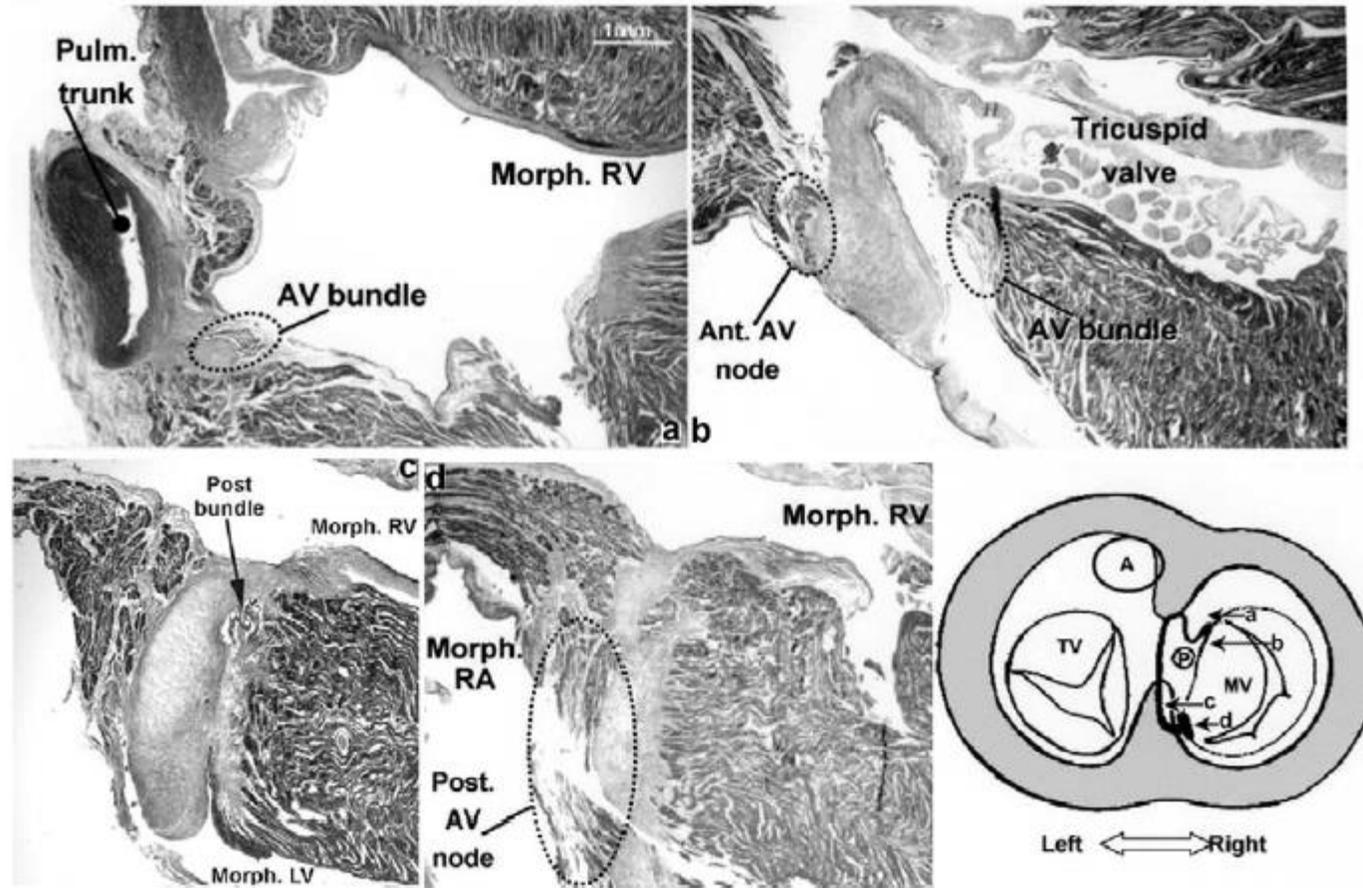
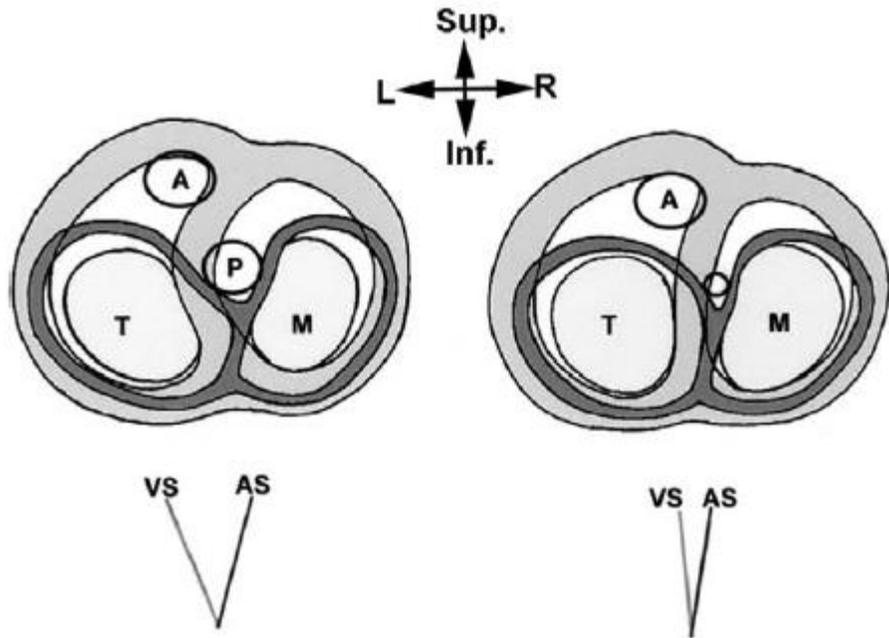
Arrhythmia	Incidence in Ebstein's*
<b>Intrinsic, %</b>	
Accessory AV pathways	10-38
Atriofascicular fibers	5
AV nodal re-entry	8-13
Monomorphic ventricular tachycardia	Rare
<b>Acquired, %</b>	
Atrial macro-re-entry	>20
Focal atrial tachycardia	2-20
Atrial fibrillation	Rare
Polymorphic ventricular tachycardia	Rare
Sudden death, presumed arrhythmic (any mechanisms), %	8-16

# Atrioventricular & Ventriculoarterial Discordance



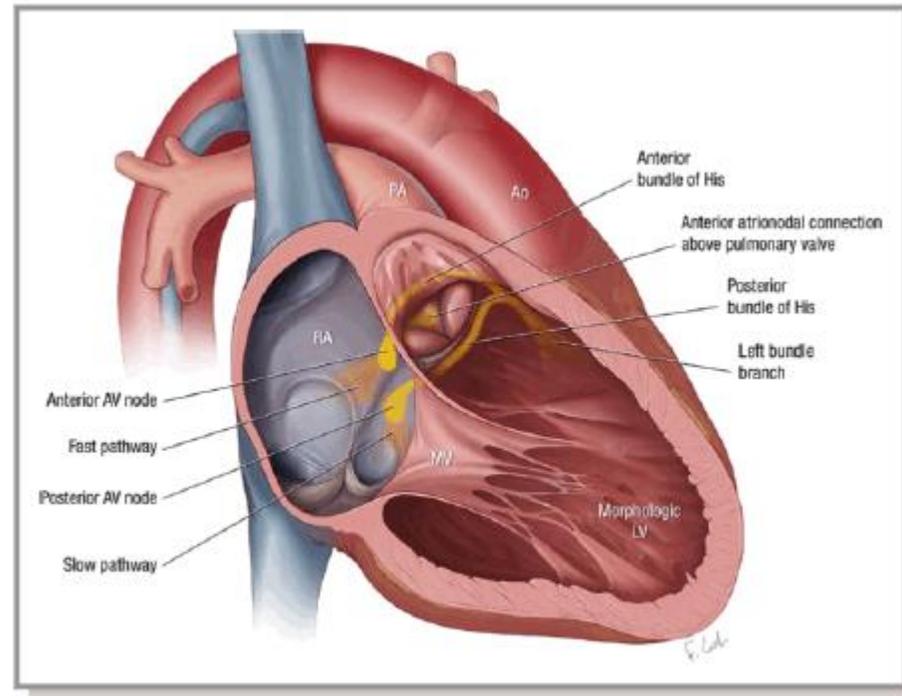
Baruteau, JAHA 2017

# Atrioventricular & Ventriculoarterial Discordance



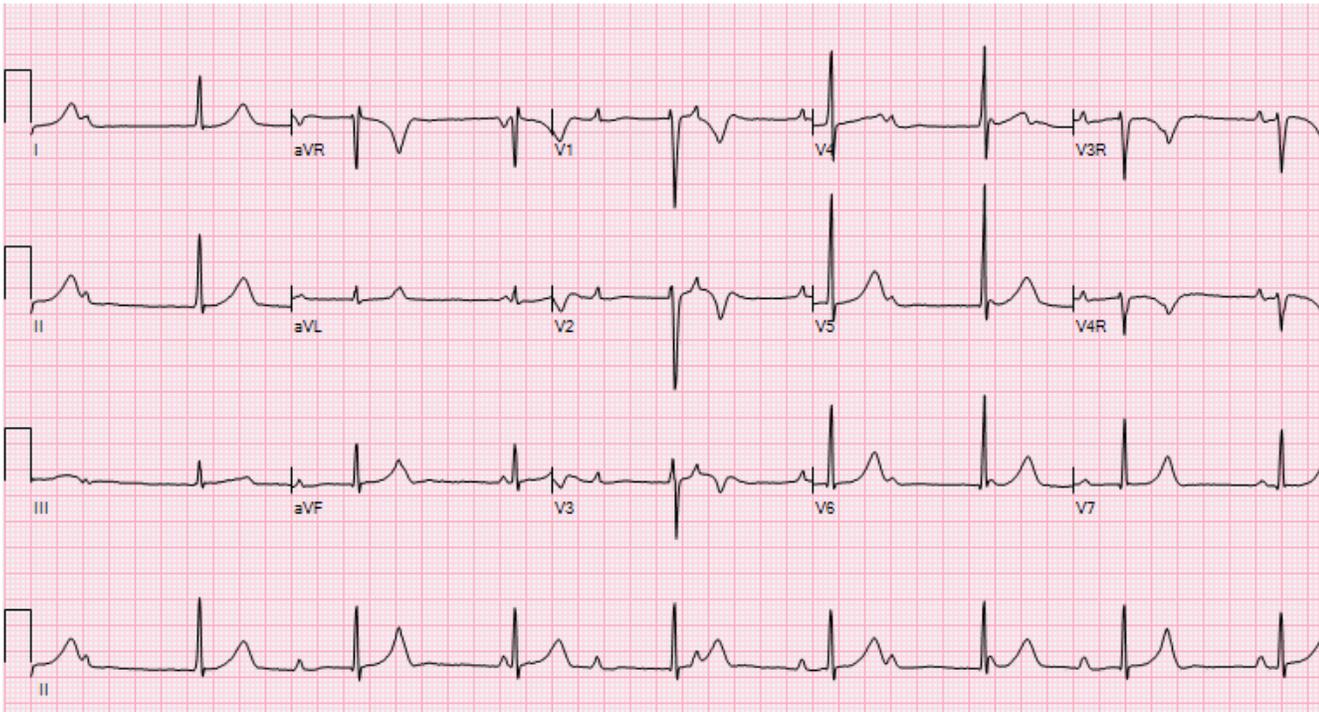
Hosseinpour, Ann Thorac Surg 2004

# Atrioventricular & Ventriculoarterial Discordance



Baruteau, JAHA 2017

# Atrioventricular & Ventriculoarterial Discordance



Variable	Overall (n = 38)	Situs Inversus (n = 8)	Situs Solitus (n = 30)	p Value
Men	21 (55%)	3 (38%)	18 (60%)	0.43
Age at last examination (years)	40 ± 15	40 ± 17	40 ± 14	0.99
Follow-up time (years)	7 ± 7	8 ± 8	7 ± 7	0.93
Surgical palliative repair	13 (34%)	2 (25%)	11 (37%)	0.67
Ventricular septal defect	22 (58%)	4 (50%)	18 (60%)	0.70
Pulmonary stenosis or atresia*	21 (55%)	5 (63%)	16 (53%)	0.71
Ebstein-like anomaly	15 (40%)	0	15 (50%)	0.013
Complete atrioventricular block	18 (47%)	1 (12%)	17 (57%)	0.045

Oliver, Am J Cardiol 2012

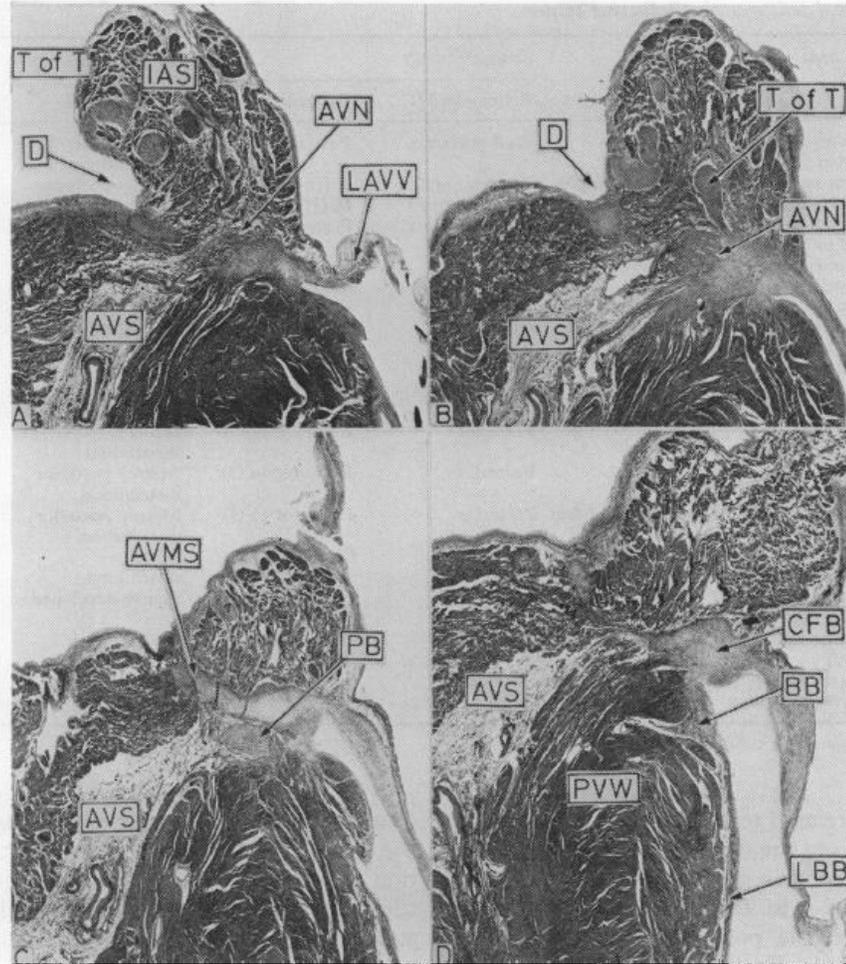
# Double Inlet Left Ventricle (DILV)



- {S,L,L} double inlet left ventricle (leftward rudimentary morphologic RV)
- Bundle of His courses at the superior margin of the bulboventricular foramen, as viewed from the transatrial approach (i.e. LV size)
- In extending the BVF, one can safely resect a wedge of tissue from apical trabecular septum by incising close to the obtuse margin of the ventricular mass.

Anderson, J Thorac Cardiovasc Surg 1996

# Tricuspid Atresia



- AV node located in the floor of the RA, posterior to the tendon of Todaro, and extends anteriorly either medial or lateral to the insertion of the tendon of Todaro.
- Bundle of His arises from anterior or lateral extension of the AV node and after it pierces through the central fibrous body, lies on the right wall of the main ventricular chamber.
- Branching of the bundle of His courses posterior to the outlet foramen.

Bharati, Circulation 1977

Dickinson, British Heart Journal 1979

Thank you!

