

Goofs, Coups and Things I Have Learned in the Past Year

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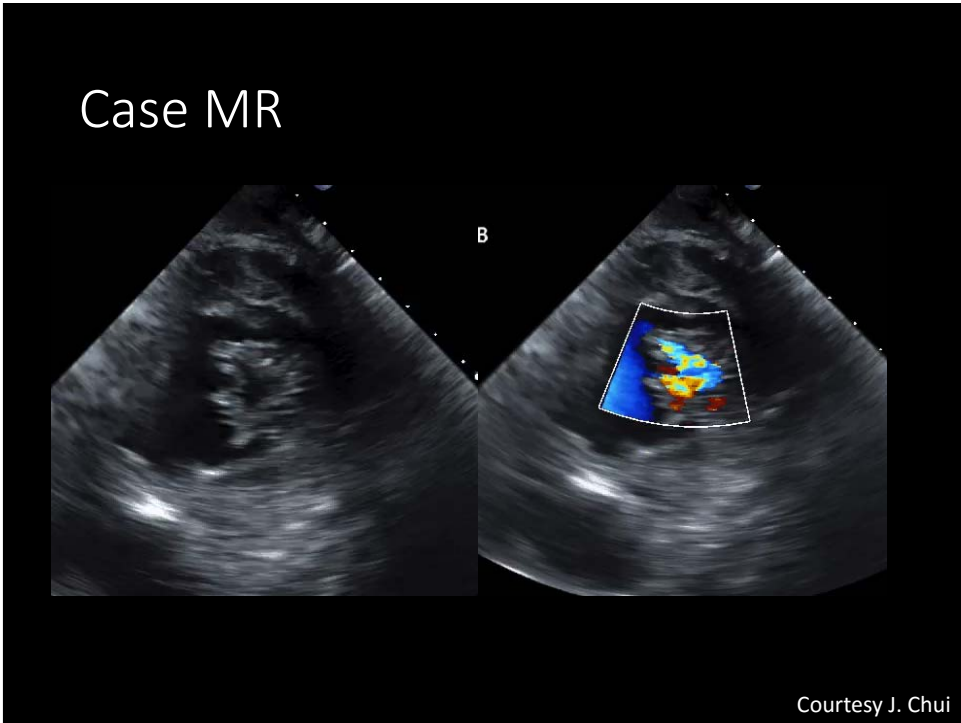
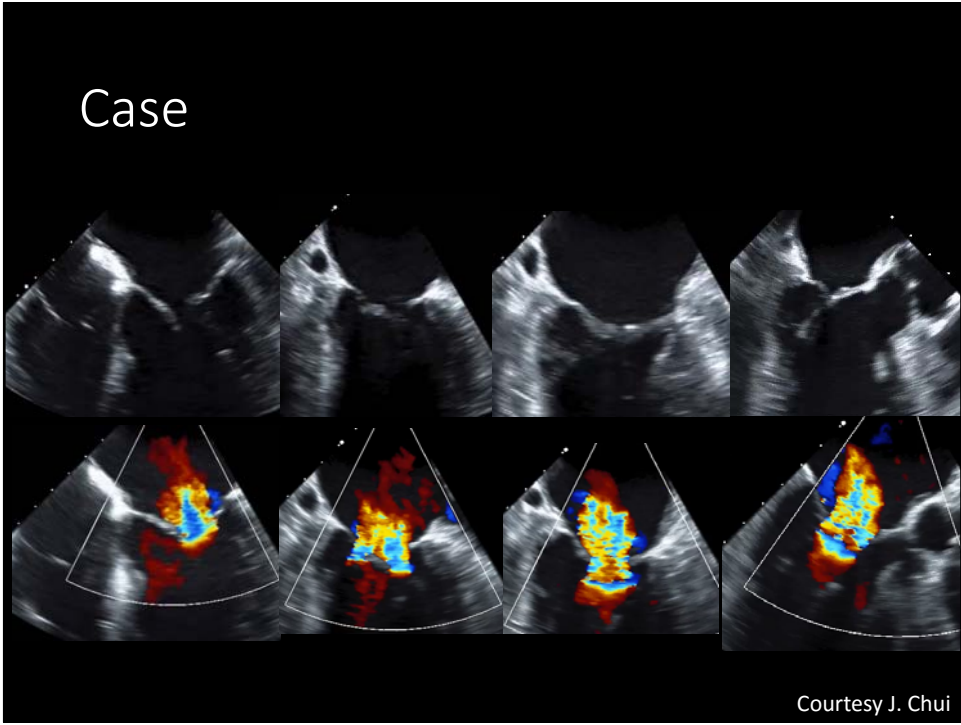
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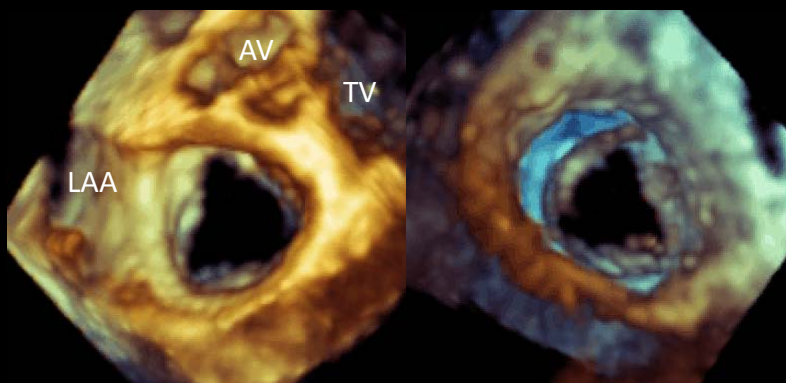
Acknowledgements: Jason Chui, Robert H. Anderson and Roberto Lang

Case

- 59 year old morbidly obese patient who presented for CABG
- Pre-op TTE was technically difficult and mild-to-moderate MR was reported
- Intraoperative TEE revealed a central jet of MR

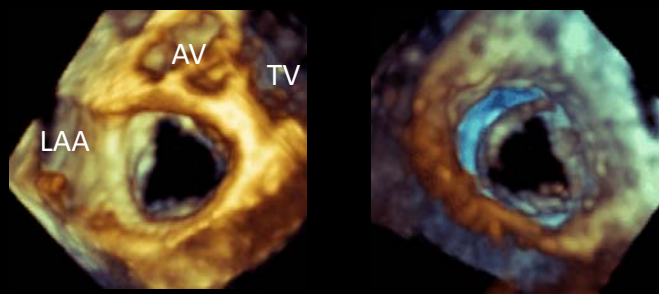


Case



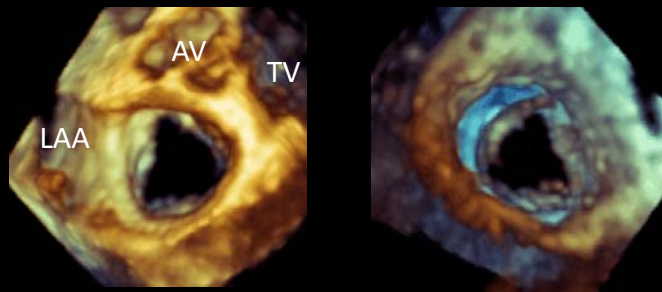
Courtesy J. Chui

How would you report this valve?



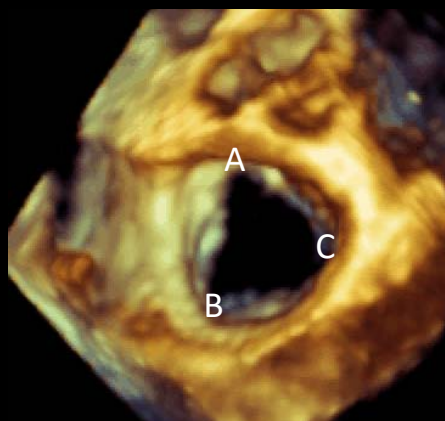
- a) Mitral valve with posterior leaflet cleft
- b) Cleft of the aortic/anterior mitral valve leaflet
- c) Left sided trifoliate AV valve with AVSD and intact septal structures
- d) Trileaflet mitral valve
- e) Need more information

How would you report this valve?

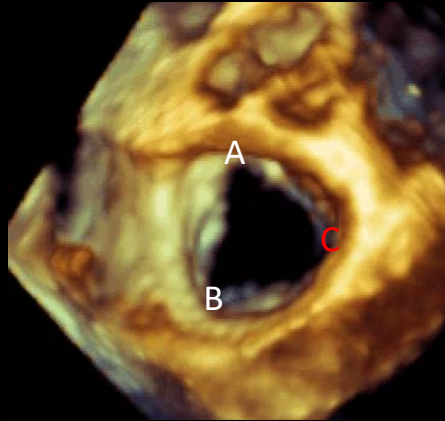


- a) Mitral valve with posterior leaflet cleft
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- d) Trileaflet mitral valve
- e) **Need more information**

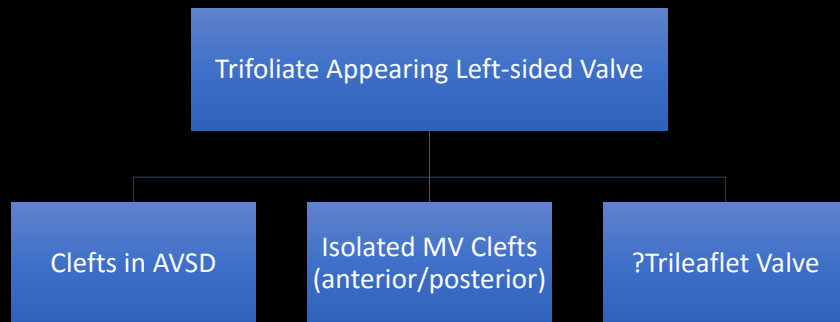
Where is the abnormality?



Where is the abnormality?



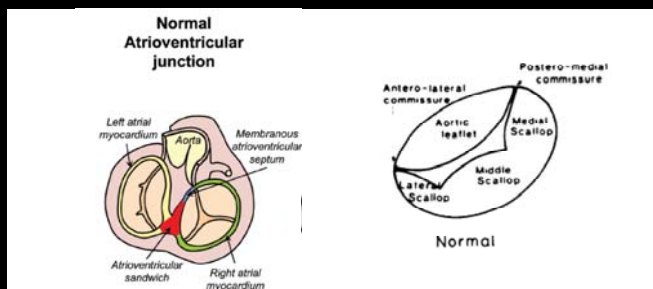
Differential



Normal Mitral Valve and AV Canal Development

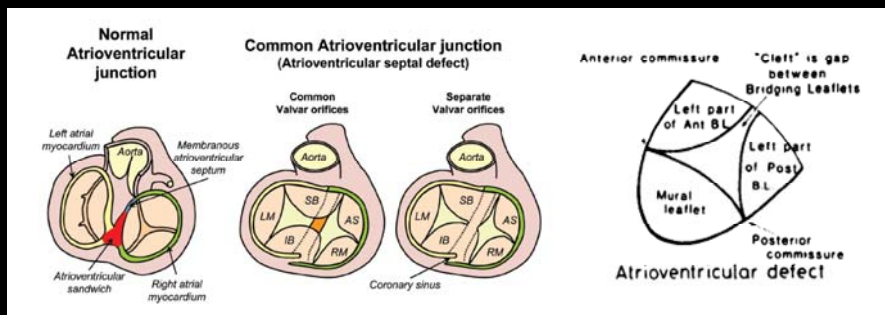
Normal development requires:

1. Division of the atrioventricular canal, followed by
2. Commitment of the developing aortic root to the left ventricle



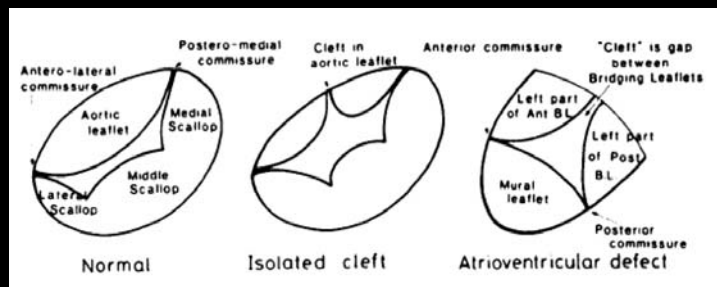
Adachi et al. Asian Cardiovasc Thorac Ann 2008;16:497-502
 Smallhorn et al. Br Heart J 1982;48:331-342

Abnormal Mitral Valve and AV Canal Development



Adachi et al. Asian Cardiovasc Thorac Ann 2008;16:497-502
 Smallhorn et al. Br Heart J 1982;48:331-342

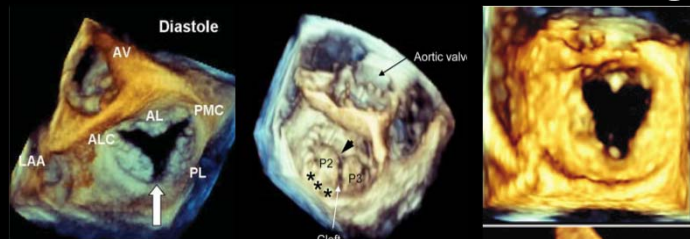
Isolated anterior mitral valve cleft is different from those associated with AV canal defects



- Cleft points to the LVOT
- Mitral valve is otherwise normal

Adachi et al. Asian Cardiovasc Thorac Ann 2008;16:497–502

Mural/Posterior Leaflet Clefting



- Extremely rare but long-reported
- Cleft is oriented away from the LVOT
- Appearance is similar to AVSD with intact septum
 - ? distinct morphological entity, or a 'forme fruste' of an AVSD with intact septum
- Features
 - well-formed septal structures supporting the trifoliate atrioventricular valve
 - lack of any inlet-outlet disproportion of the ventricular septum
 - the mural leaflet in AVSDs is significantly smaller than that found in patients with isolated clefting of the mitral valve

Vignallou JB, et al. Eur Heart J Cardiovasc Imaging. 2012 Jun;13(6):535.

Biaggi P, et al. J Am Soc Echocardiogr. 2010 Oct;23(10):1114.e1-4.

Jouni H, et al. Eur Heart J. 2014 Jun 21;35(24):1623.

Trileaflet Mitral Valve



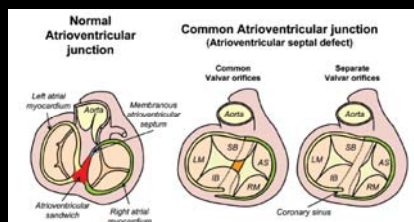
- Only recently reported with 7 cases diagnosed with 3D echo
 - Three equidistant commissures or zones of apposition
 - a central coaptation point
 - the presence of a displaced accessory papillary muscle
 - concordant atrioventricular and ventriculo–arterial connections
- All cases reported an association of significant LVOT obstruction or subaortic stenosis
- ?Novel phenotype in hypertrophic cardiomyopathy that is distinct from isolated clefting of the mural leaflet of the valve

D'Ancona G, et al. *Eur Heart J*. 2015 Jul 7;36(26):1697

Butler T, et al. *Int J Cardiol*. 2015 Dec 15;201:549-51.

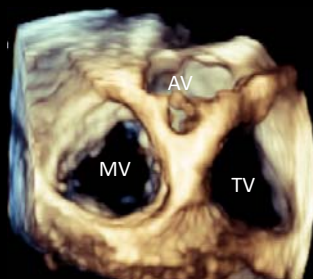
Moya-Mur JL, et al. *Eur Heart J Cardiovasc Imaging*. 2015 Jun;16(6):692.

AVSD Assessment: Sprung Aorta



Case

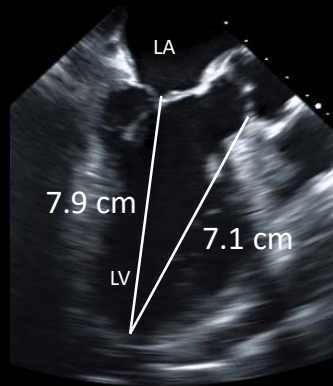
Example of an Sprung Aorta



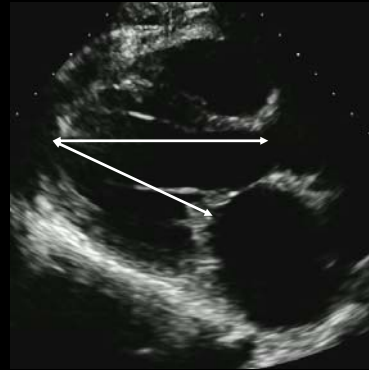
Adachi et al. *Asian Cardiovasc Thorac Ann* 2008;16:497–502

AVSD Assessment: Inlet-Outlet Disproportion

Case

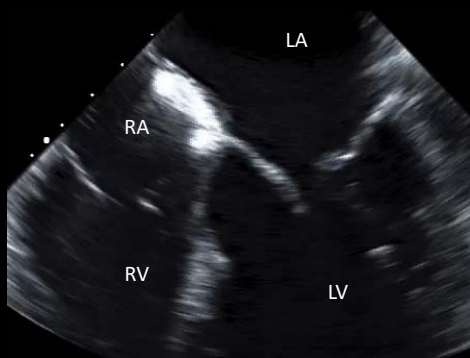


Example of Inlet-Outlet Disproportion

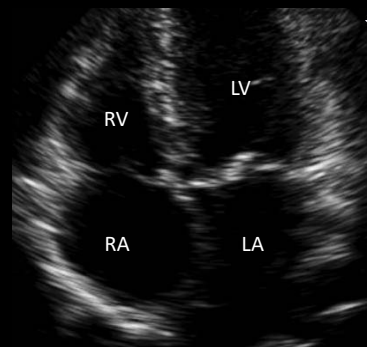


AVSD Assessment: AV Valve Offset

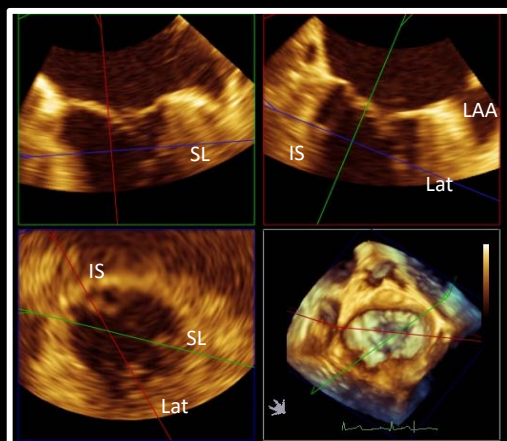
Case



Example of No Offset



Assess Papillary Muscles



- 3 papillary muscles and attachments,
 - infero-septal,
 - supero-lateral, and
 - lateral,
- Correspond to the zones of apposition between the three distinct leaflets
- Supero-lateral papillary muscle is small and can be labeled as an accessory papillary muscle

What did I learn?

- 3D echocardiography improves visualization but there has to be an understanding of the embryologic development and nomenclature for diagnosis
- Differential would include isolated cleft of an otherwise normal mitral valve versus trifoliate left AV valve in the setting of an AVSD with intact septum
- Unclear if trileaflet mitral valves are a new entity versus cases of mis-identification

What did we call it?

- Isolated cleft of the aortic/anterior leaflet



Thank you for listening!