



## BACKGROUND

Mitral valve (MV) development is a complex process which is not completely understood. Isolated double orifice mitral valve (DOMV) is very rare. DOMV is often associated with other congenital cardiac anomalies, most commonly atrioventricular septal defect.

## CASE

- 67-year-old man with history of usual interstitial pneumonia was referred to our institution for progressive shortness of breath out of proportion to the patient's lung pathology.
- Transthoracic echocardiogram (TTE) found bicuspid aortic valve (AV) with a mean gradient of 51 mmHg.
- Computed tomography (CT) and transesophageal echocardiogram (TEE) were ordered as part of the workup for AV replacement.
- Incidentally the anterior and posterior leaflets of the MV were fused together. This resulted in mild mitral regurgitation. The point of fusion lay more towards the anterolateral commissure, resulting in a DOMV with two unequal-sized orifices. At the point of leaflet fusion, an accessory scallop was attached to the middle scallop of the anterior leaflet (A2).
- Also noted on the CT was stenosis of the posterior superior pulmonary vein

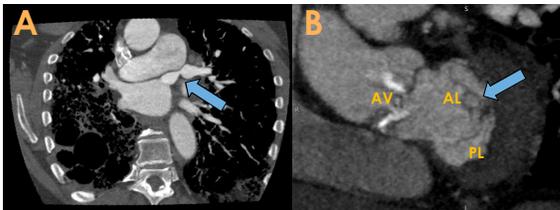


Fig. 1 CTA chest  
A – Pulmonary Stenosis (arrow)  
B – Calcified AV as well as anterior (AL) and posterior (PL) MV leaflets fused with an accessory scallop (arrow)

## REFERENCES

- Zalstein, E; Hamilton, R; Zucker, N; Levitas, A; Gross, G. Presentation, natural history, and outcome in children and adolescents with double orifice mitral valve. The American Journal of Cardiology. 2004. 93(8). 1067-1069.
- HOVE, J. R., KOSTER, R. W., FOROUHAR, A. S., ACEVEDO-BOLTON, G., FRASER, S. E. & GHARIB, M. 2003. Intracardiac fluid forces are an essential epigenetic factor for embryonic cardiogenesis. Nature, 421, 172-7.

# FUSED ACCESSORY SCALLOP CAUSING ASYMMETRIC DOUBLE ORIFICE MITRAL VALVE

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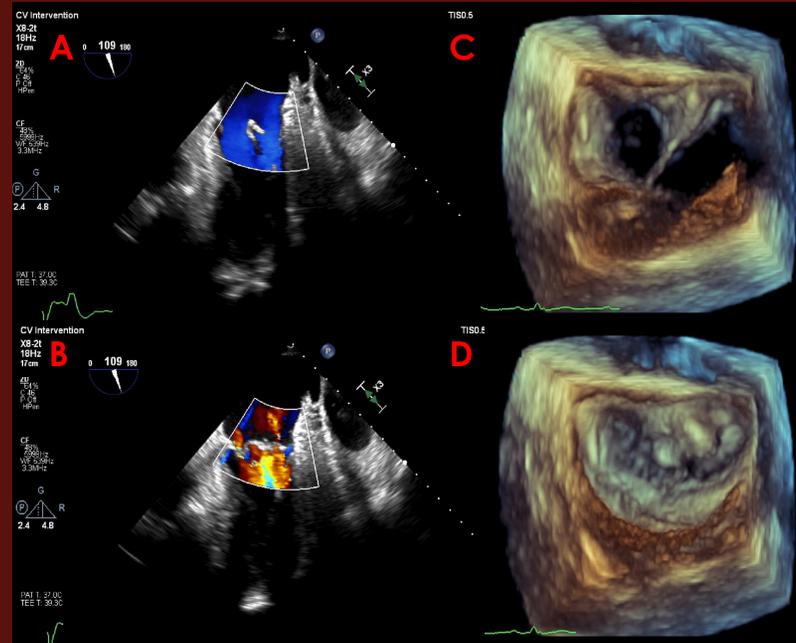


Fig. 2  
A – TTE in diastole. Color doppler showing mitral inflow around the leaflet fusion  
B – TTE in systole. Color doppler showing dual regurgitant jets  
C – TEE in diastole showing asymmetric dual orifices  
D – TEE in systole showing dual regurgitant jets

## DISCLOSURE INFORMATION

The authors have nothing to disclose and no conflicts of interest

## DISCUSSION

- One case series of 47 patients with DOMV noted only 6.5% incidence of isolated DOMV<sup>1</sup>. In the remainder of patients such as ours, the anomaly was associated with other congenital abnormalities.
- This case demonstrates association of DOMV with primary pulmonary vein (PV) stenosis and bicuspid AV. It also shows a novel etiology of DOMV; accessory mitral valve scallop.
- Though secondary PV stenosis is seen frequently after atrial fibrillation ablation, primary stenosis is a pathological result of embryogenesis. Studies suggest that restricting blood flow such as with PV stenosis into fetal cardiac chambers plays a role in abnormal valvulogenesis<sup>2</sup>. At this point, it is unclear if this is a mere coincidence of association or a syndrome that will become more recognizable with the widespread adoption of new cardiovascular imaging modalities.

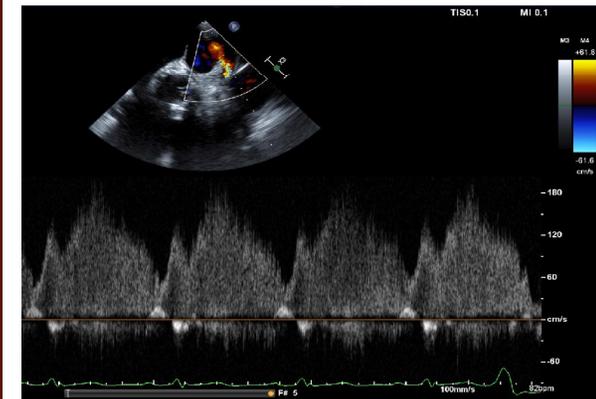


Fig. 3 – Color doppler of flow through the stenotic pulmonary vein showing increased velocity

## CONCLUSION

Several cases of DOMV in association with various congenital abnormalities have been previously reported. To our knowledge, this is the first case of DOMV, associated with pulmonary stenosis and bicuspid aortic valve. In addition the double orifices were created by a unique culprit i.e. accessory scallop.