Unicuspid Aortic Valve Replacement In A Patient With Profound Developmental Delay

Matthew DeMatteo, DO, Joseph Hinchey, MD, PHD
Department of Anesthesiology, Hartford Hospital, Hartford, CT
Department of Anesthesiology, University of Connecticut, Farmington, CT

INTRODUCTION

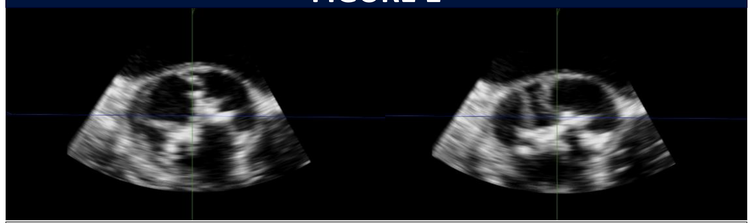
Unicuspid aortic valves (UAV) are rare congenital malformation which typically present in the third to fifth decade of life with severe aortic stenosis or regurgitation. We present a case of acute congestive heart failure secondary to severe aortic stenosis in the setting of UAV in a patient with profound intellectual delay.

CASE REPORT

A 33-year-old male with past medical history of autism spectrum disorder (nonverbal at baseline), and seizure disorder presented to an outside hospital with shortness of breath. A chest x-ray was consistent with congestive heart failure. He was treated with IV Lasix and initiated on a nitroglycerin infusion for hypertension. An EKG showed new onset a-fib and an echocardiogram was performed revealing aortic stenosis and reduced systolic LV function with an EF of 29%. The patient was transferred to Hartford hospital for further management. He was stabilized on milrinone and scheduled for aortic valve replacement and maze procedure.

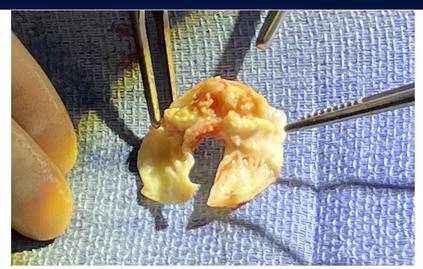
Given the patients profound intellectual delay, a perioperative analgesic and anxiolytic plan was developed. Preoperatively the patient was given midazolam prior to placement of an arterial line and induction. After induction, central line and single shot parasternal blocks were placed. Intraoperatively the patient received fentanyl, ketamine, and hydromorphone. Postoperatively, an emergence plan utilizing dexmedetomidine was discussed with the ICU team.
Intraoperative TEE showed a unicommissural unicuspid aortic valve which was confirmed on direct visualization. After discussion with the patient's parents, his primary care givers, the patient received a mechanical aortic valve replacement, and maze procedure.

FIGURE 1



Left: intraoperative TEE mid-esophageal short access view showing unicuspid aortic valve during systole
Right: Intraoperative TEE mid-esophageal short access view showing unicuspid aortic valve during diastole

FIGURE 2



Macroscopic view of unicuspid, unicommissural aortic valve

DISCUSSION

UAV is an extremely rare congenital malformation with an estimated prevalence of 0.02% in adults. Typically these lesions will present in the third to fifth decade of life with a predominance of cases being in male patients (4:1). Most cases present with severe aortic stenosis or regurgitation.

DISCUSSION

UAV can be unicommissural, or acommissural. They can be identified with TTE, cardiac CT, MRI or with TEE being the gold standard with a sensitivity and specificity of 75 and 86% respectively. Although in our patient this was not identified on TTE, unicommissural unicsupid aortic valve was identified on TEE and confirmed with direct visualization of the valve surgically. These lesions are best identified during systole when there is no cusp separation leading to an eccentric "teardrop" opening.

Management of these congenital malformations can vary with severity of disease, and age of presentation. Congenital aortic stenosis due to UAV is most often treated with balloon valvuloplasty, surgical valvotomy, or commissurotomy. Valve replacement is generally not recommended until the patient has reached adulthood. Given that our patient presented as a young adult, after discussion with his parents, it was decided to do a replacement with a mechanical valve to attempt to avoid reoperation in the future.

The patient being nonverbal with severe intellectual delay presented an interesting challenge in developing a perioperative anxiolytic and analgesic plan. The patient would be unable to communicate pain post operatively, and extubation in ICU was planned. A multimodal strategy was developed including, anxiolytics, sedatives, regional anesthesia, opioid and non opioid medications.

REFERENCES

1. Singh S, Ghayal P, Mathur A, et al. Unicuspid unicommissural aortic valve: an extremely rare congenital anomaly. *Tex Heart Inst J.* 2015;42(3):273-276. Published 2015 Jun 1. doi:10.14503/THIJ-13-3634
2. Sjostad BD, Witt CM, O'Leary PW, Maleszewski JJ, Scott CG, Dearani JA, Pellikka PA. Unicuspid Aortic Valve: Demographics, Comorbidities, Echocardiographic Features, and Long-Term Outcomes. Circulation. 2019 Nov 26;140(22):1853-1855. doi: 10.1161/CIRCULATIONAHA.119.041835. Epub 2019 Nov 25. PMID: 31765262.
3. Ingason, A.B., Sigfusson, G. & Torfason, B. Congenital aortic stenosis due to unicuspid unicommissural aortic valve: a case report. *J Cardiothorac Surg* **13,** 61 (2018). https://doi.org/10.1186/s13019-018-0755-0
4. Sniecinski, Roman M. MD*; Shanewise, Jack S. MD†; Glas, Kathryn E. MD*Transesophageal Echocardiography of a Unicuspid Aortic Valve, Anesthesia & Analgesia: March 2009 - Volume 108 - Issue 3 - p 788-789 doi: 10.1213/ane.0b013e318193cbd8