A CASE REPORT OF COMPLEX CONGENITAL HEART DISEASE: GERBODE DEFECT WITH A BICUSPID VALVE AND COARCTATION OF THE AORTA

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Case Vignettes

Background:
Congenital Gerbode defect is a rare phenomenon that entails a deformity in the ventricular septum that allows for the communication of blood between the left ventricle (LV) and right atrium (RA). We report a rare case of a patient with a congenital Gerbode defect with an associated bicuspid aortic valve and a coarctation of the aorta (CoA), which is the first such case report according to our knowledge. We discuss how the co-existence of these congenital abnormalities affected the management of his Gerbode defect.

Case Presentation:
The patient was a 38-year-old male with a past medical history of hypertension and pediatric heart murmur who presented to our hospital with shortness of breath, abdominal distention, and leg swelling. Echocardiography demonstrated a ventricular septal defect with blood flow from the left ventricle to the right atrium, consistent with the Gerbode defect. He required multiple hospitalizations for decompensated heart failure, and during a latter hospitalization, he was found to have CoA with a gradient of 35-40mmHg. Percutaneous repair of his CoA led to symptomatic improvement.

Discussion
The pathophysiology of Gerbode defect involves shunting of the blood from the LV to the RA due to the significant pressure gradient between the two chambers. Our patient had concomitant CoA, as well as a bicuspid aortic valve, which further hindered his cardiac output and augmented the pressure gradient between LV and RA, leading to greater shunting. Without correction, this process can lead to right heart failure, resulting in systemic venous congestion, and ultimately left sided heart failure. The timing and type of repair depends on a myriad of factors, including associated comorbidities, hemodynamics, clinical picture, and surgical risk. In this case, repair of the Gerbode defect was initially deferred as stenting of his coarctation of the aorta led to significant symptomatic improvement.

Conclusion
The management of Gerbode defect is complex, and a full hemodynamic and congenital abnormality workup should be performed to provide optimal management.