

Evolution of Critical Aortic Stenosis Secondary to Bicuspid Aortic Valve: From In-Utero Left Heart Dysfunction to Successful Postnatal

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Background

Bicuspid aortic valve (BAV) arises from abnormal valvulogenesis of the left ventricular outflow tract. In utero, BAV presenting as critical aortic stenosis (AS) can lead to progressive left ventricular (LV) dysfunction and potential evolution toward a hypoplastic left heart-type physiology.

Case Presentation

A 23-year-old primigravida was referred at 32 weeks’ gestation with suspected evolving hypoplastic left heart syndrome (HLHS). Fetal echocardiography revealed a bicuspid aortic valve, critical aortic stenosis (AS), and mild mitral stenosis. This was accompanied by significant left ventricular (LV) systolic dysfunction and a dilated, globular LV with mild septal endocardial fibroelastosis. No extracardiac anomalies were detected. Given the advanced gestational age, conservative antenatal management with serial surveillance was elected. At 36 weeks' gestation, a 2.2 kg male infant was delivered via Cesarean section at a tertiary cardiac center

Postnatal Course

Postnatal echocardiography confirmed critical AS with a severely dilated, hypokinetic LV and minimal forward flow across the left ventricular outflow tract (LVOT)—a failing, dilated LV rather than a “stagnant” hypoplastic left heart.

Intervention

An early and successful transapical percutaneous balloon aortic valvuloplasty was performed to decompress the left ventricle(Fig. 3). This relieved the critical LVOT obstruction and pressure afterload. Repeated post-procedure echo revealed mild-moderate residual AS and mild AR with improved ventricular function

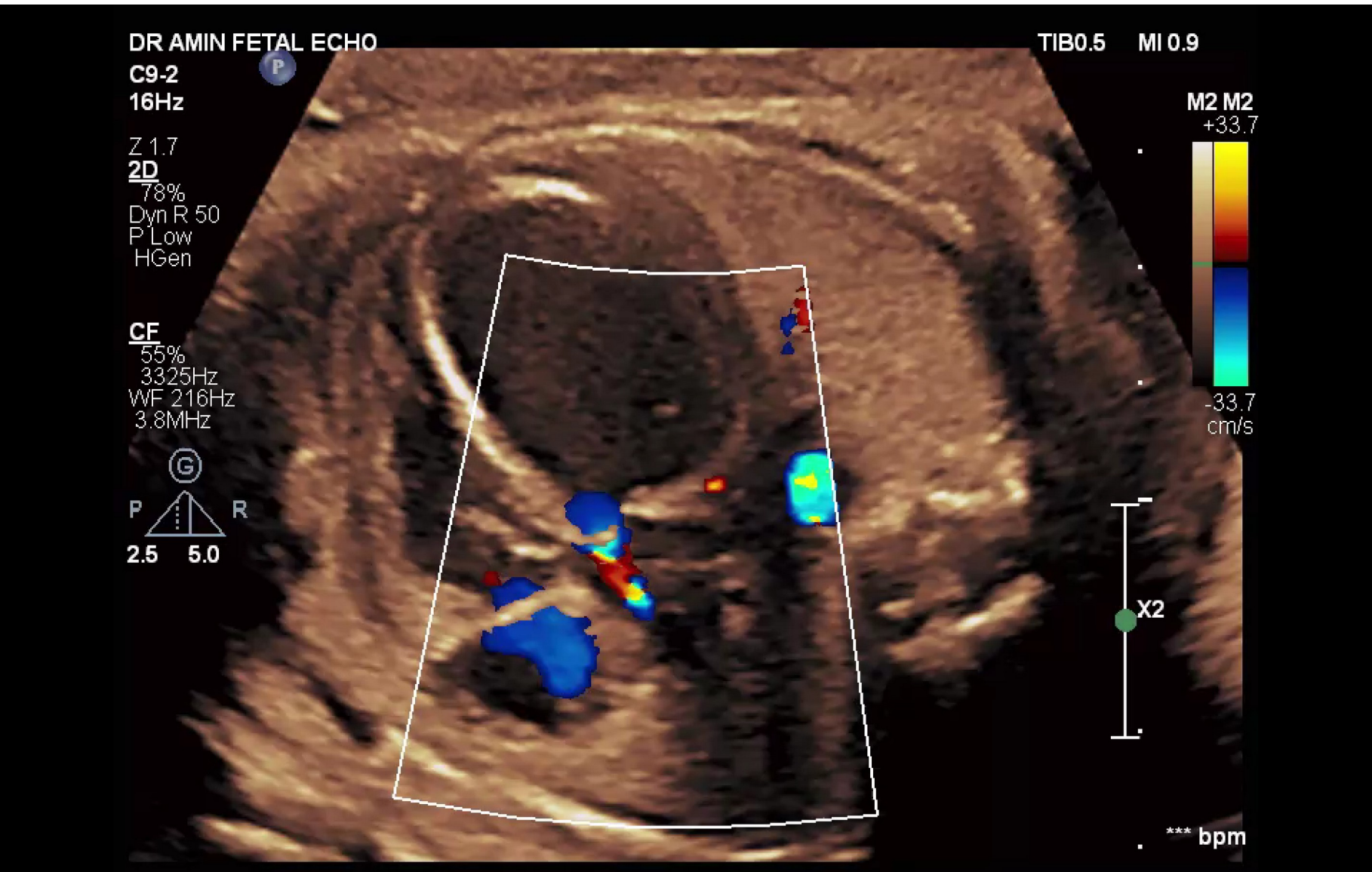


FIGURE 1 : Fetal echocardiogram demonstrating a dilated, globular left ventricle (LV) with mild septal endocardial fibroelastosis due to critical aortic stenosis

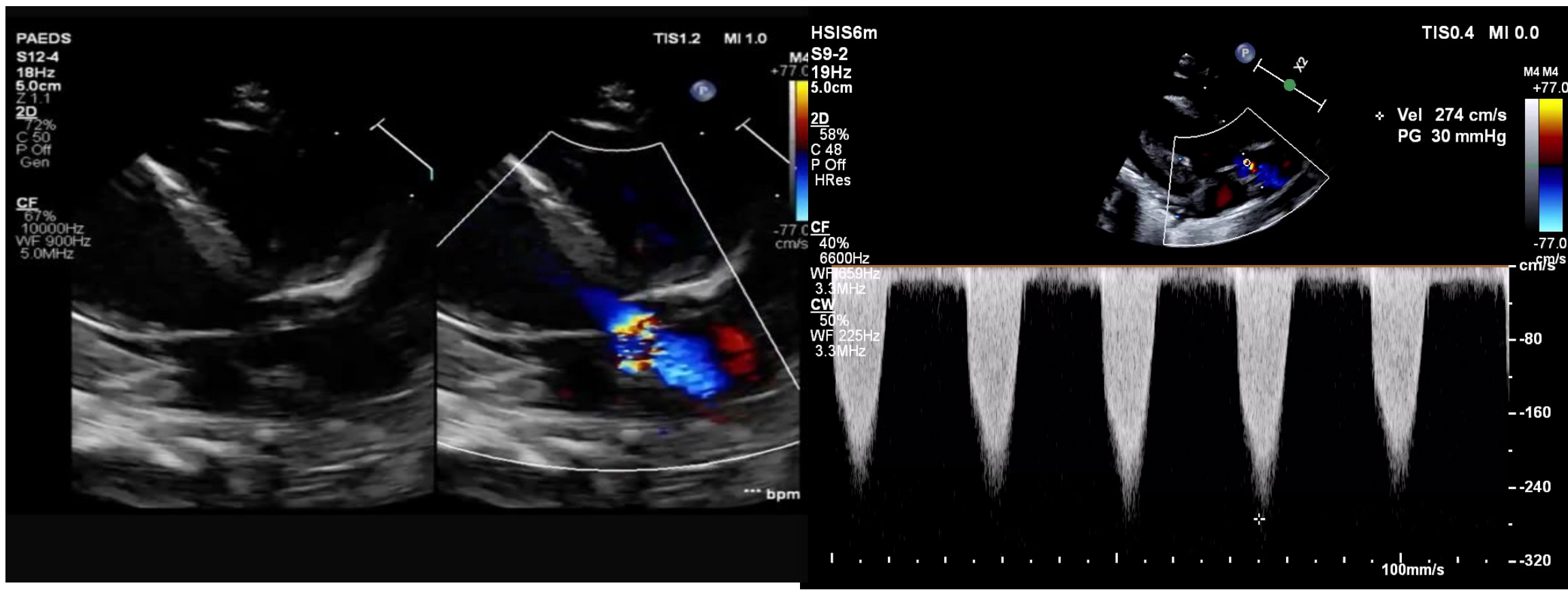


FIGURE 2: Postnatal Echocardiography showing presence of Critical Aortic Stenosis with minimal forward flow across LV

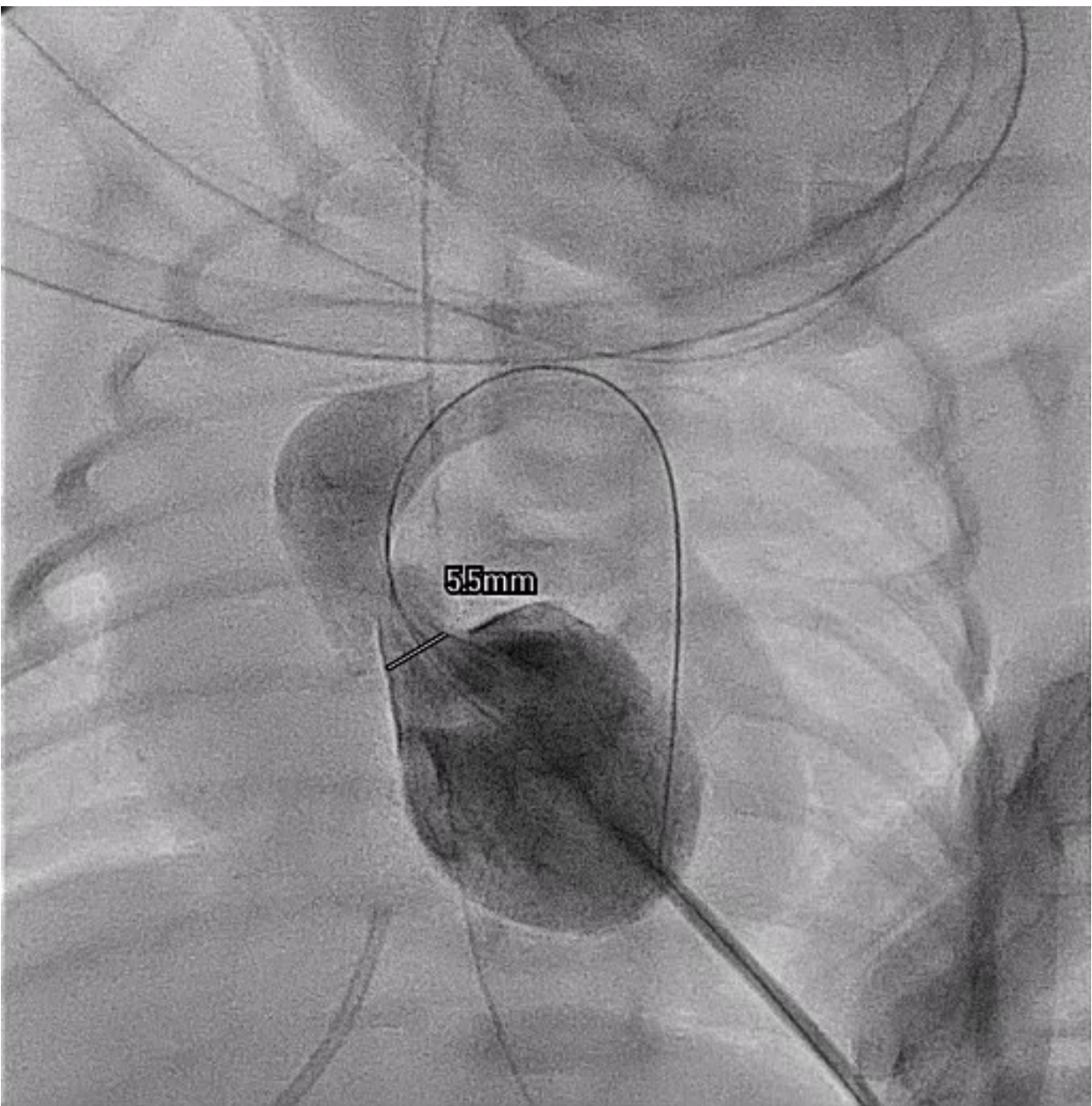


FIGURE 3: Transapical percutaneous balloon aortic valvuloplasty (PBAV)

Recovery & Follow-up

Discharged well with normalized ventricular function after 49 days in the NICU. At the 3-month follow-up, he was thriving well. However, transthoracic echocardiography demonstrated significant residual AS, mild aortic regurgitation (AR) and mild LV hypertrophy, though LV systolic function remains preserved. A repeat elective cardiac intervention was planned

Discussion

Serial fetal echocardiography is key to distinguishing a “stagnant” HLHS trajectory from a failing, dilated LV secondary to critical AS — a window in which prompt postnatal intervention can salvage LV function and preserve a biventricular circulation.

Conclusion

Prenatal identification of critical AS is essential, to monitor the in utero progression of left-heart obstructive lesions. A strategic delivery at a tertiary cardiac center, combined with prompt support from paediatric interventional cardiology, is paramount for optimizing outcomes, salvaging LV function, and preserving a biventricular circulation.