

Perinatal and Postnatal Outcomes in Fetuses with Pulmonary Atresia and Intact Ventricular Septum (PAIVS)

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Introduction

PAIVS is a very rare condition that requires a high degree of individualized and complex peri- and postnatal medical, interventional, and surgical therapy. This study aims to evaluate the outcomes of fetuses diagnosed with PAIVS.

Methods

This retrospective single-center study analyzes 30 fetuses, including 3 twins and 27 singletons, diagnosed with PAIVS, born between January 2008 and January 2025.

Conclusion

This study highlights the significant perinatal morbidity and mortality associated with PAIVS, despite advancements in prenatal diagnosis and interventions. These findings emphasize the need for improved prenatal risk assessment and enhanced postnatal management to improve survival and long-term outcomes for affected neonates.

Results

The mean maternal age was 30.8 ± 5 years. Perinatal outcomes included 2 intrauterine fetal deaths (IUFD) (6.7%), 2 terminations of pregnancy (TOP) (6.7%), 1 lost to follow-up (3.3%), and 25 live births (83.3%). The mean gestational age at delivery was 37.3 ± 2.7 weeks. The 30-day survival rate among live-born infants was 18 out of 25 (78.3%).

Cardiac surgery was performed in 15 neonates (60.0% of live births) during hospitalization. Two neonates (8.0%) died peri-interventionally – one due to intraoperative electromechanical dissociation and another due to multi-organ failure immediately postoperatively. One neonate (4.0%) died 27 days postoperatively from multiple complications, including myocardial ischemia resulting from a coronary steal phenomenon due to inoperable right-ventricular coronary fistulas, an acute abdomen, and ultimately sepsis.

One pre-interventional death occurred (4.0%). For two neonates, no documentation of the cause of death was available. The six neonates who died postnatally (24.0%) had a mean survival time of 11.5 ± 14.0 days (range: 0-39.0 days).

Prostaglandin E1 treatment was administered in 21 neonates, while 1 did not receive treatment and in 8 cases either death occurred before therapy initiation, or no documentation was available. Catheter-based PDA stenting was performed in 16 neonates. The mean number of cardiac catheter interventions per neonate was 1.6 ± 0.9 . Follow-up data from the center were available in 16 cases.

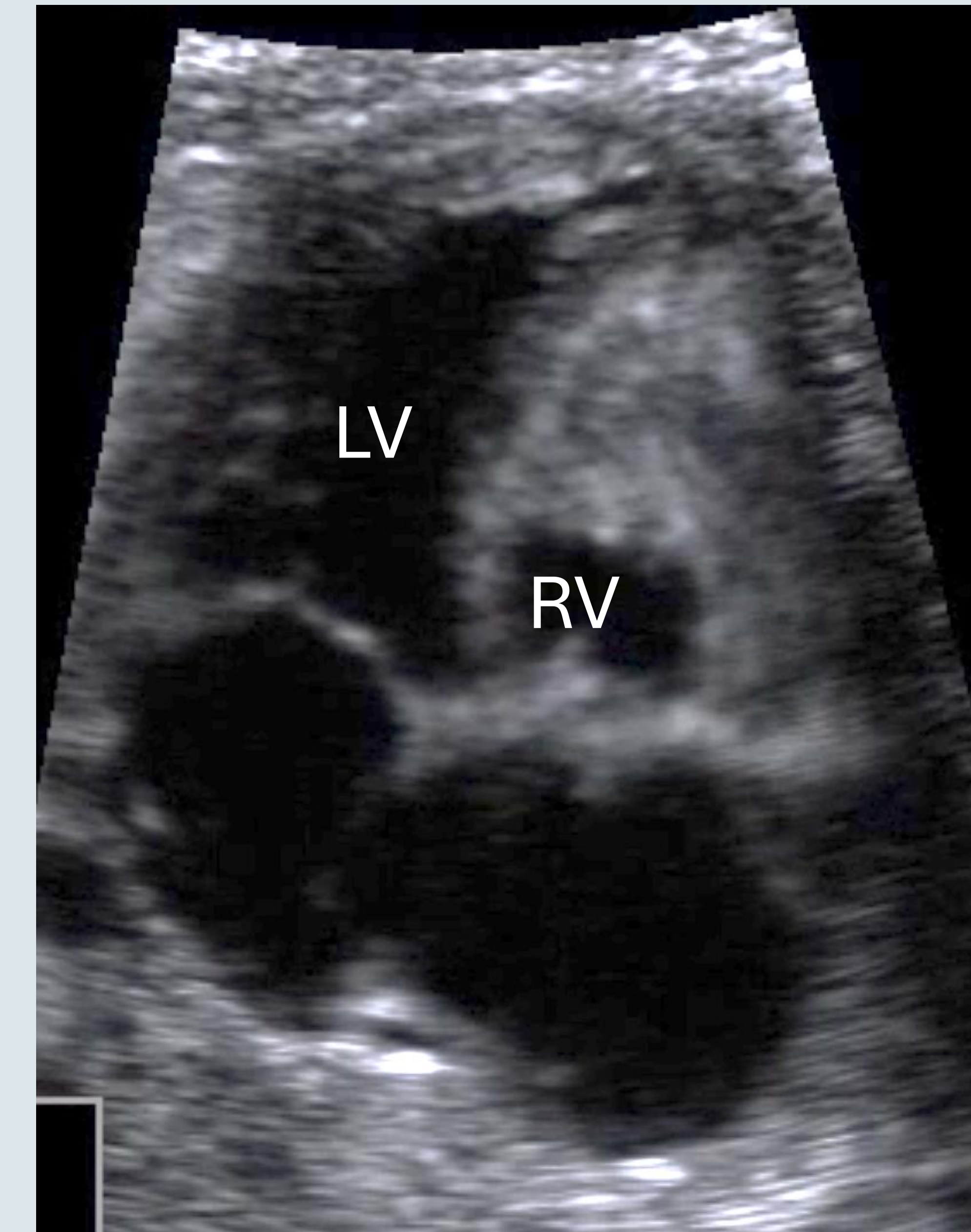


Fig. 1: prenatal echocardiography of PAIVS (GW 38+1): hypoplastic, hypertrophic right ventricle