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Transplacental Everolimus rescue strategy in a Giant Fetal Cardiac Rhabdomyoma : Preventing progression to duct dependent systemic circulation after birth.

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Background:

- Large fetal cardiac rhabdomyomas can cause severe in-utero hemodynamic compromise by obstructing blood flow.
- Transplacental mTOR inhibitor therapy is an emerging rescue treatment that may reverse circulatory deterioration.
- We report a case demonstrating its effectiveness in a fetus with a massive cardiac rhabdomyoma causing systemic outflow obstruction.



CASE PRESENTATION



A **43-year-old primigravida** with an IVF conception was referred at **32 weeks** for a **massive fetal intracardiac mass**.

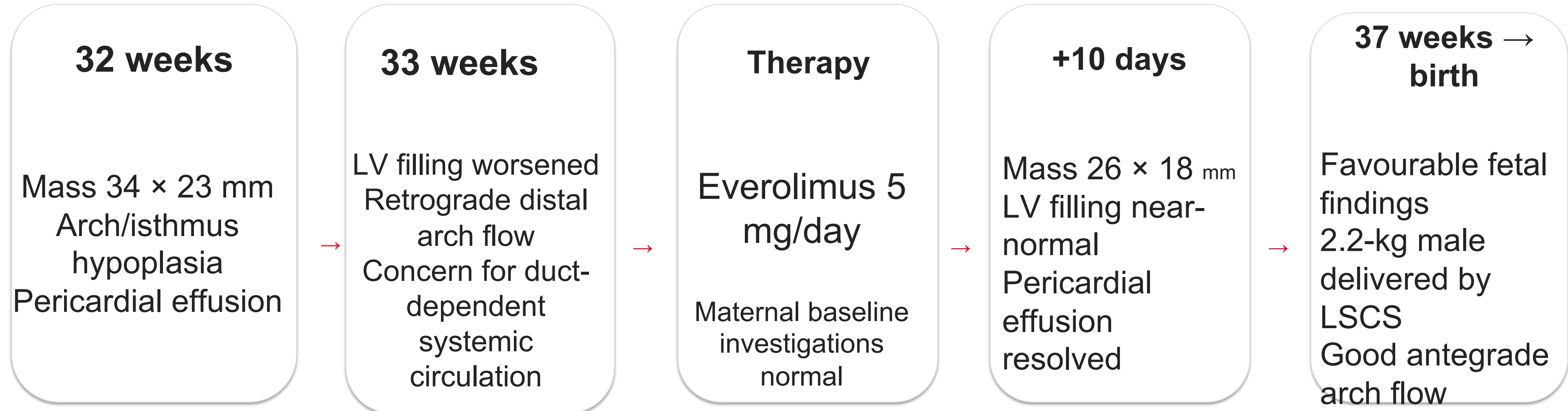
FETAL ULTRASOUND FINDINGS

 34 × 23 mm Hyperechogenic mass involving mid & apical left ventricle	 5 × 4 mm Second lesion in right-ventricular apical region	 Multiple Echogenic cortical tubers	 3 mm Transverse aortic arch hypoplasia (Z score −3.8)	 2.5 mm Aortic isthmus hypoplasia (Z score −4.5)	 6.4 mm Pericardial effusion
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At presentation: | No hydrops • No LV inflow/outflow obstruction

Clinical course and intervention:



Postnatal outcome:

Birth	37 weeks • 2.2 kg male • APGAR 7/9
Echo	20 × 16 mm mass • no inflow/outflow obstruction • good antegrade aortic arch flow
Treatment	Postnatal everolimus 0.4 mg twice daily after normal baseline labs
2 weeks	Further reduction to 14 × 8 mm; dose subsequently adjusted to serum drug levels
2 months	Tumor size further reduced: Everolimus stopped

Therapeutic response at a glance

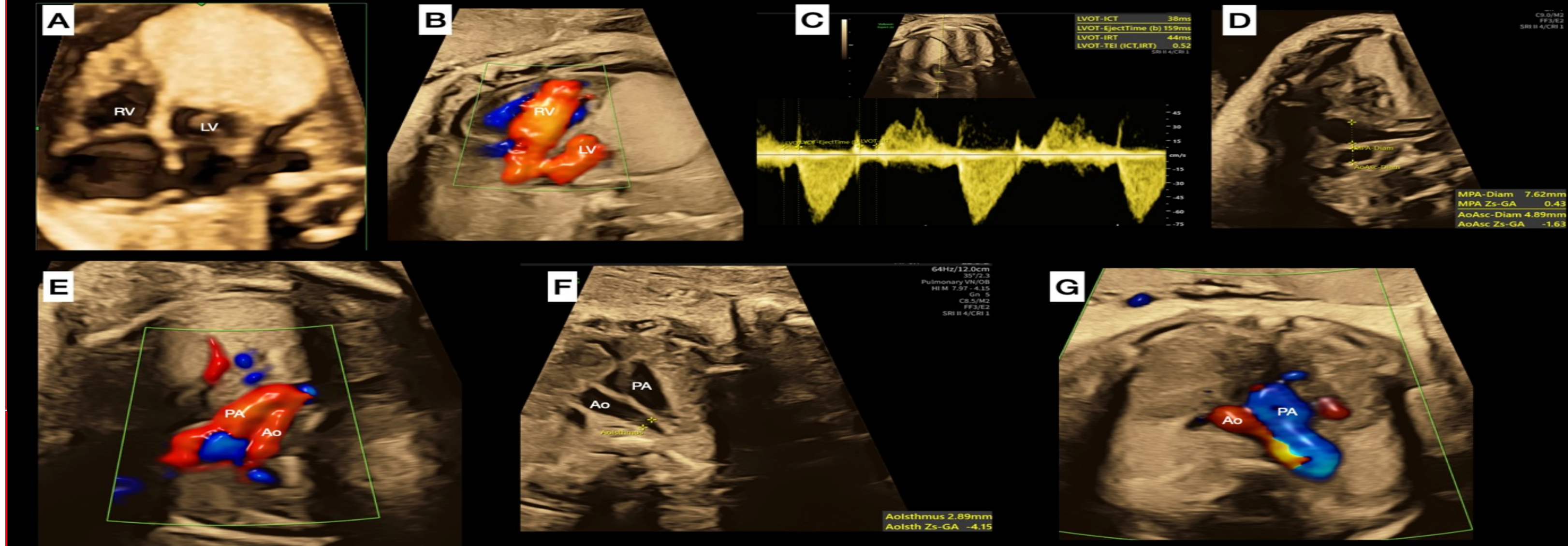
Tumour size	34 × 23 mm	26 × 18 mm	14 × 8 mm
LV filling	Significantly impaired	Near-normal	Good postnatally
Pericardial effusion	6.4 mm	Resolved	—

Decision for transplacental therapy

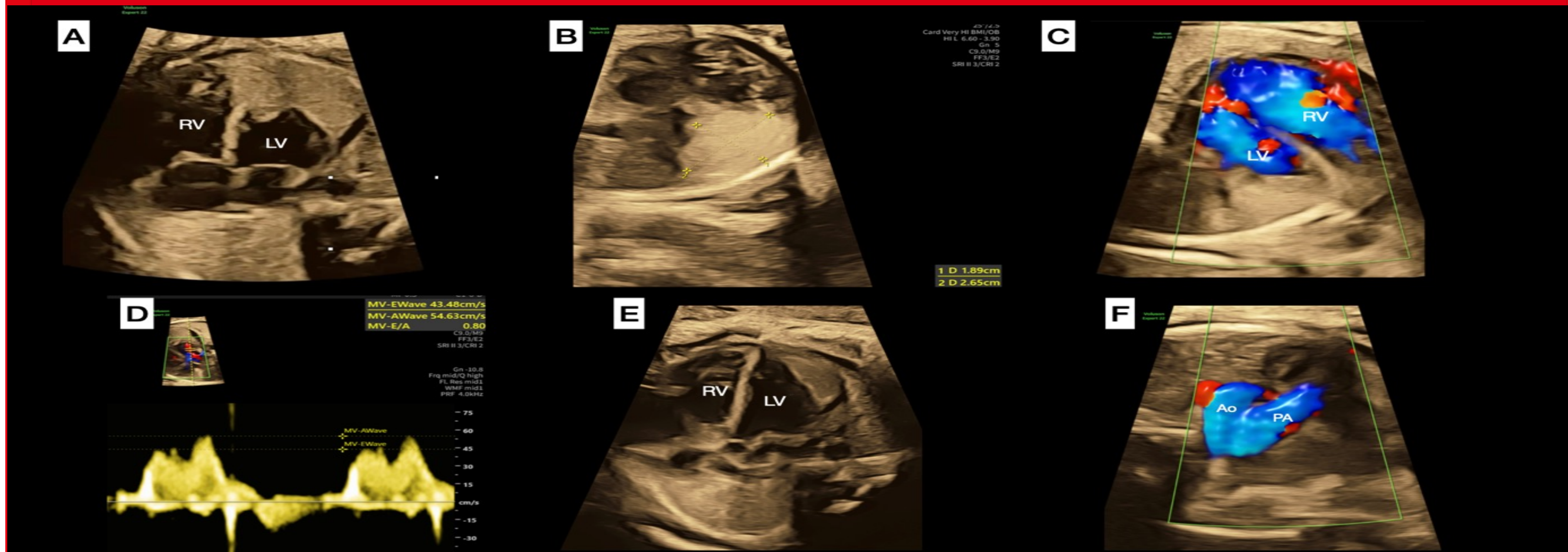
- After one week of surveillance, there was significant compromise of LV filling with retrograde flow in the distal aortic arch, indicating severely reduced antegrade LV output and raising concern for evolving duct-dependent systemic circulation.
- Following multidisciplinary discussion, transplacental everolimus was initiated at 5 mg once daily after normal baseline CBC, lipid profile, renal and liver function tests.

Conclusion:

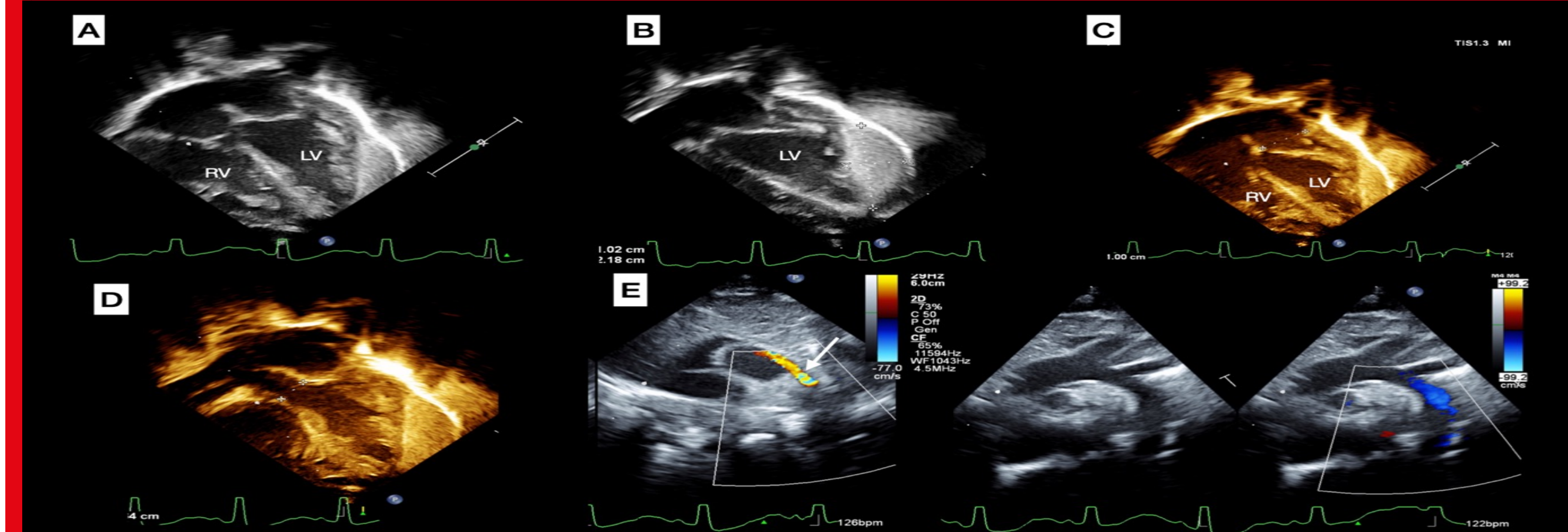
- Transplacental everolimus was associated with rapid regression of a massive fetal cardiac rhabdomyoma, improvement in LV filling, resolution of pericardial effusion and preservation of antegrade systemic flow.
- In this high-risk fetus, antenatal targeted therapy may have helped prevent progression to duct-dependent systemic circulation.
- Careful multidisciplinary selection and close maternal–fetal surveillance remain essential, and further evidence is needed to define the role of fetal mTOR inhibition.



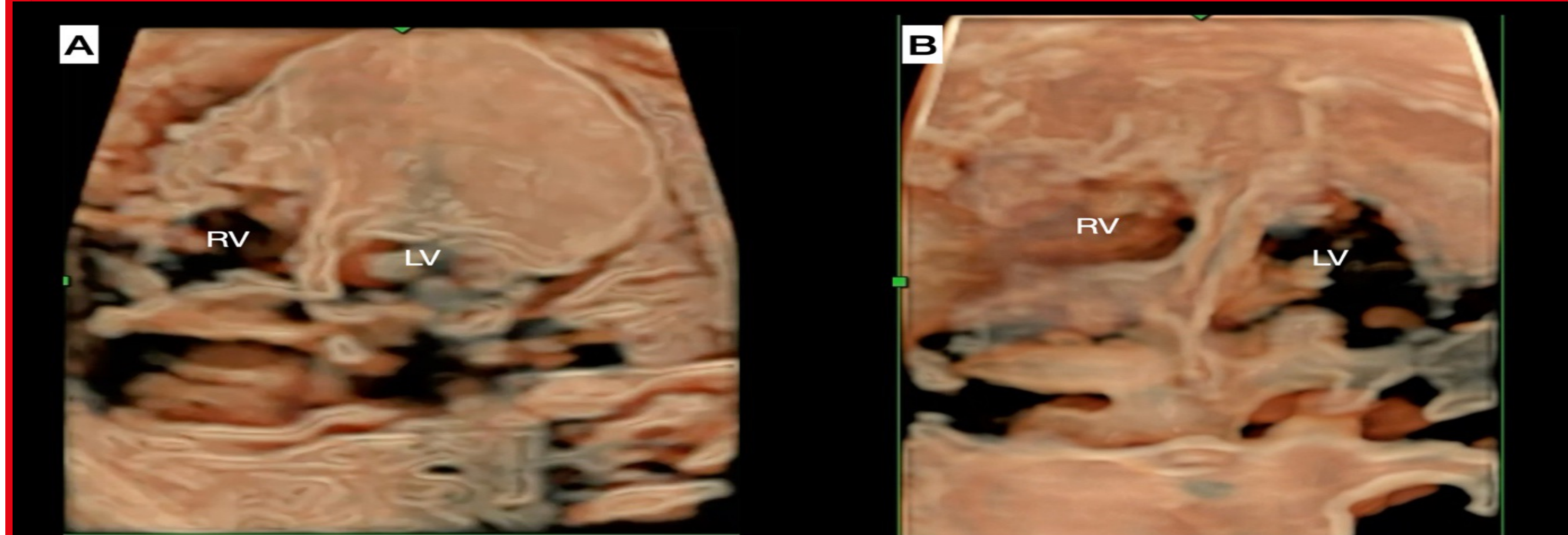
Fetal echo t23 mm Intra cardiac mass involving the mid and apical segments of the left ventricle. B and C. showing impaired al ECHO assessment at 33 weeks showing A. 34 × LV filling. E and F showing disproportionate Great arteries with a small aortic isthmus.



Fetal ECHO assessment 10 days post Initiation of Trans placental Everolimus showing substancial reduction in size of Intra-cardiac mass with improved LV inflow and good antegrade flows across arch.



Transthoracic ECHO assessment of the neonate immediately after birth showing the significant regression of the Intra-cardiac mass with good LV inflow and unobstructed arch flows.



4D STIC rendering Fetal ECHO image showing LV Intra-Cardiac mass prior to Transplacental Everolimus therapy at 32 weeks
LV Intra -cardiac mass post Transplacental therapy