

Congenital systemic arteriovenous fistulas: an uncommon fetal presentation

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Introduction

Congenital malformations are the leading cause of infant mortality; congenital heart defects account for 30–50% of these deaths. Systemic arteriovenous fistulas (SAVFs)—direct communications between systemic arteries and veins without capillaries—are rare, usually described in case reports and typically present in infancy or childhood. We aim to present a recent case with a prenatal presentation with severe cardiomegaly.

Case Report

A 27-year-old G3P2 woman was referred at 26 weeks for fetal cardiomegaly. Fetal echocardiography at 35 weeks (late referral) confirmed severe cardiomegaly without hydrops; partial or total anomalous pulmonary venous connection with a supracardiac anomalous connection was initially suspected. Prenatal growth and Dopplers were normal. The patient had an uncomplicated spontaneous vaginal birth at 39 weeks. The male newborn weighed 3080 g, required no initial respiratory support, and was in good general condition.

Postnatal CT angiography confirmed cardiomegaly secondary to a congenital arteriovenous fistula between the left subclavian artery and the ipsilateral jugulo subclavian confluence. Given the favorable clinical course, the infant was transferred from the NICU to his base hospital for follow-up with planned consideration of embolization versus surgery.

Discussion

This rare prenatal presentation of SAVF produced significant cardiomegaly without hydrops, mimicking anomalous pulmonary venous drainage. Normal fetal Dopplers suggest in-utero compensation despite volume overload, but perinatal decompensation remains possible. The anatomic location is exceptional in the literature.

The case underscores the need for timely referral to centers with advanced fetal echocardiography and vascular imaging when cardiomegaly or supracardiac vascular masses are detected to enable monitoring, delivery planning, and coordination for definitive postnatal intervention. Long-term cardiovascular follow-up is advised.



Figure 1. Severe cardiomegaly in the 4c view.

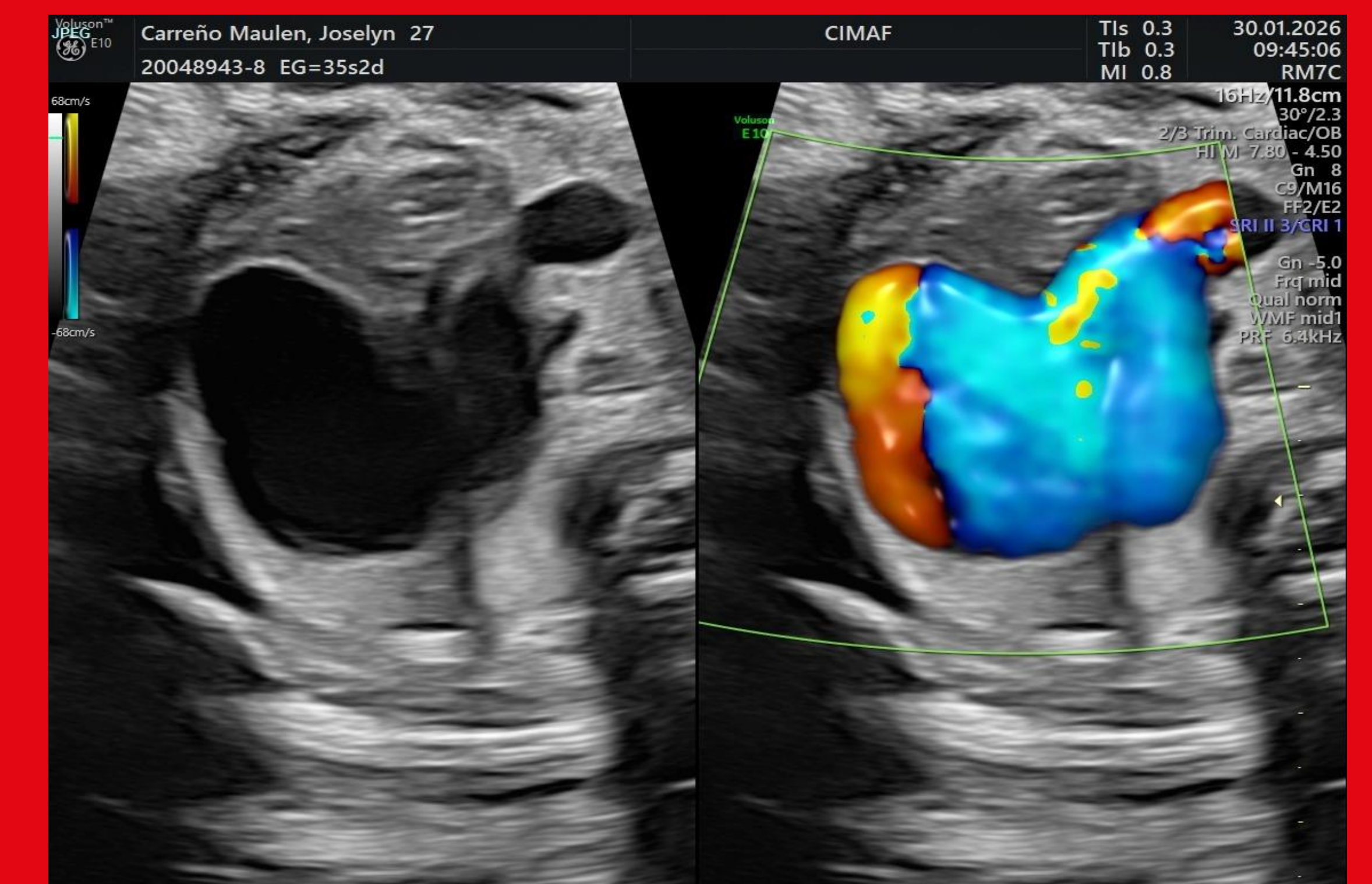


Figure 2. Aneurysmal image with turbulent flow.



Figure 3. Venous aneurysm to which described fistulous tract is connected.