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## IMAGES AND CASE REPORTS IN HEART FAILURE

# High-Output Heart Failure From Growth of Vascular Malformations in Multiple Gestation Pregnancy

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### CASE PRESENTATION

A 29-year-old woman (gravida 2, para 1) at 26 weeks of gestation with monochorionic diamniotic twins presented with 6 weeks of dyspnea, orthopnea, palpitations, lower extremity edema, and enlargement of a facial birthmark (Figure 1). She had a history of heart failure as a newborn secondary to cerebral arteriovenous malformation (AVM), which was resected at 10 weeks of age with no further heart failure symptoms. She had previously carried a singleton pregnancy to term without complications. Family history was significant for cutaneous capillary malformations (CMs) in her mother, sister, and brother.

She presented with a temperature of 37°C, a heart rate of 131 beats per minute, a blood pressure of 129/84 mmHg, a respiratory rate of 22 breaths per minute, and an oxygen saturation of 98% on room air. Exam demonstrated a pulsatile 12×7.5 cm left-sided facial mass (Figure 1), a 4×2 cm pulsatile left-sided scalp mass, a jugular venous pressure of 10 cm of water, a II/VI systolic murmur at the left sternal border, and 2+ bilateral lower extremity pitting edema. Laboratories revealed a hemoglobin of 8.6 g/dL, cardiac troponin I of <0.04 ng/mL, B-type natriuretic peptide of 93 pg/mL, and a thyroid stimulating hormone of 0.55 mIU/mL. ECG demonstrated sinus tachycardia at 132 beats per minute. Transthoracic echocardiogram revealed an ejection fraction of 65% to 70%, biatrial enlargement, and a calculated noninvasive cardiac output (CO) of 16.5 L/min (normal CO for twin gestation, 7–8 L/min).<sup>1</sup> Magnetic resonance angiography with ferumoxytol revealed a CO of 14 L/min, and high-flow

vascular lesions of the left cheek, left skull vertex, and right lower lateral neck (Figure 2).

The patient was diagnosed with high-output heart failure and was initially managed for 3 weeks with intravenous and oral diuretics in the inpatient and outpatient settings. Despite diuresis, she developed progressive heart failure and subsequently underwent embolization of her cheek and skull vertex vascular malformations with N-butyl cyanoacrylate glue and polyvinyl alcohol particles, resulting in ≈50% flow reduction by angiography (Figure 3). The patient symptomatically improved postoperatively. Transthoracic echocardiogram after the procedure demonstrated a calculated noninvasive CO of 11.9 L/min. On postembolization day 5, she developed thrombocytopenia and acute kidney injury. Given the concern for pre-eclampsia, she underwent cesarean section at 30 weeks and 1 day with resolution of heart failure symptoms. Twins A and B were born with birthweights of 1300 and 1700 g, respectively. Twin A was born with hypospadias and a small ventricular septal defect. Transesophageal echocardiography immediately after delivery demonstrated a calculated noninvasive CO of 9.3 L/min. The patient was discharged on oral diuretics. Her facial engorgement fully resolved 2 weeks postpartum (Figure 1). At time of writing, the twins are 23 weeks old and doing well.

### DISCUSSION

Here, we describe a case of high-output heart failure secondary to enlarging vascular malformations in the setting of multiple gestation pregnancy. CMs manifest as

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**Figure 1. Superficial appearance of left cheek vascular malformation.**

Vascular malformation shown (A) before pregnancy, (B) on initial presentation (26 wks, 1d), (C) before embolization (29wks, 3d), (D) 4 d after delivery, and (E) 2wks after delivery.

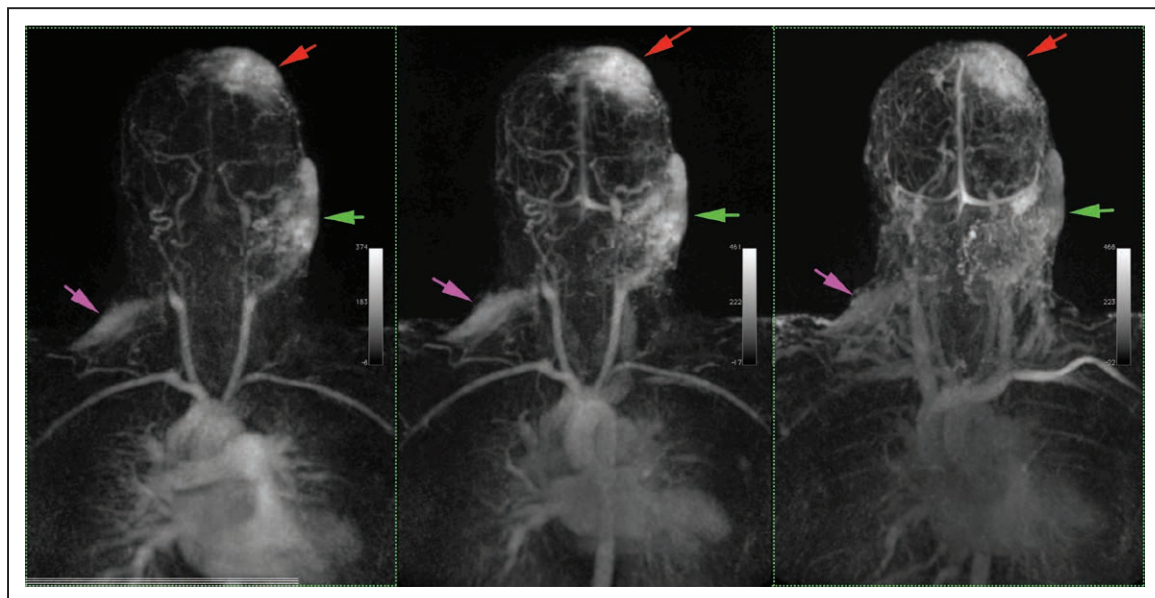
red patches on the head and neck and are found in 0.3% of newborns.<sup>2</sup> More rarely, CMs associate with high-flow AVMs in an autosomal dominant disorder called CM-AVM syndrome.<sup>3</sup> The patient's presentation, lesion appearance, and family history of cutaneous CMs (mother, sister, and brother) point to a clinical diagnosis of CM-AVM syndrome. This disorder frequently associates with loss-of-function mutations in *RASA1* and *EPHB4* and has high penetrance and variable expressivity.<sup>4-6</sup> The patient is still considering genetic testing.

AVMs are hormone sensitive and increase their size and flow in response to hormonal changes. As a result, they may enlarge during adolescence or pregnancy.<sup>3,7,8</sup> The cardiovascular changes associated with pregnancy increase the risk for heart failure in those with vascular malformations. In the first trimester, women experience a drop in peripheral vascular resistance due to systemic vasodilation.<sup>1</sup> Heart rate and stroke volume rise, resulting in an increase in CO. This rise can approach 145% of normal in singleton pregnancies and can reach 7 to 8

L/min in twin gestations.<sup>1,9</sup> The increased estrogen and progesterone from the patient's multiple gestation pregnancy likely significantly contributed to the increased size and flow of her existing CM-AVMs.

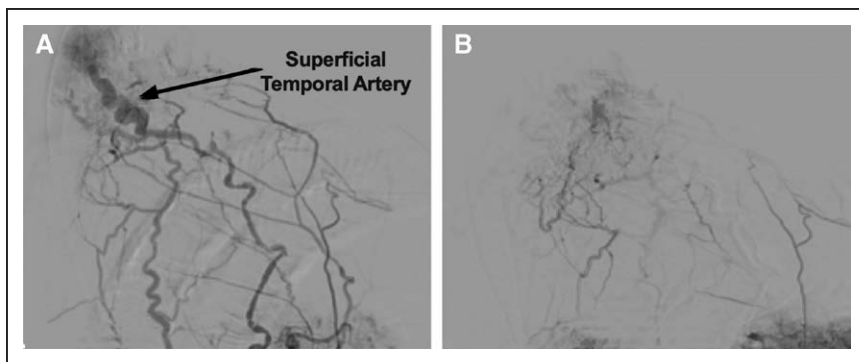
Vascular malformations in pregnancy are typically managed with a conservative watch and wait strategy. Given the patient's clinical deterioration, embolization of the AVMs was performed. To our knowledge, this represents the first report of CM-AVM embolization during pregnancy. Despite significant postoperative reduction in arteriovenous shunting and mild symptomatic improvement, the patient developed signs of superimposed preeclampsia, ultimately requiring preterm delivery. The regression of her AVMs postpartum confirmed their hormonal sensitivity.

Acute decompensated high-output heart failure in pregnancy due to congenital vascular malformations is rare and can pose significant management challenges. Endovascular embolization may have a role in the management of such conditions following the failure of conservative medical therapies. The postoperative development of preeclampsia



**Figure 2. Magnetic resonance angiography time resolved series at 27 wk gestation demonstrating left cheek (green arrow), left scalp (red arrow), and right shoulder (pink arrow) high-flow vascular malformations.**

The 3 frames show progressive evolution of enhancement from arterial to late venous phases.



**Figure 3. Angiography of the vascular malformation and surrounding blood vessels.**

**A**, Before embolization: left external carotid artery angiogram demonstrates a hypervascular blush overlying the left parietal scalp primarily supplied by a hypertrophied branch of the superficial temporal artery. **B**, After embolization: left external carotid artery angiogram shows decreased opacification of the vascular malformation nidus. The hypertrophied superficial temporal artery branch has been occluded.

necessitating urgent delivery in this report, however, highlights the complexities inherent in such cases.

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**Disclosures**

Dr Hogeling is a Principal Investigator on a study sponsored by Celgene. The other authors report no conflicts.

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