Life-Threatening Peripartum Cardiomyopathy (PPCM) in a Previously Healthy Parturient – Interplay Between Genetic Susceptibility and Pregnancy-Related Stress

Lauren McHenry M.D., M.S., Amritpal S. Randhawa B.S., Prabdit Sidhu M.D., M.P.H., Daniel Conti M.D., Rostam Bakhtari M.D., Maria Gutierrez M.D.

UC Davis Anesthesiology and Pain Medicine

BACKGROUND

1:2,000
births worldwide
Incidence

LVEF < 45%
Without identifying cause

50%
Rate of complete recovery of LVEF within 6 months

Peripartum cardiomyopathy (PPCM) is a rare, life-threatening cause of heart failure in late pregnancy or early postpartum

Multifactorial etiology - hemodynamic and oxidative stress, inflammation, vascular damage, and hormonal shifts

Underlying genetic susceptibility for dilated cardiomyopathy (DCM) may overlap with pregnancy-related physiologic stress to precipitate disease

Multiple anesthetic challenges - hemodynamic instability, uteroplacental perfusion, malignant arrythmias, need for mechanical circulatory support

High morbidity and mortality, with many patients requiring ICD, LVAD, or cardiac transplantation

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CASE PRESENTATION

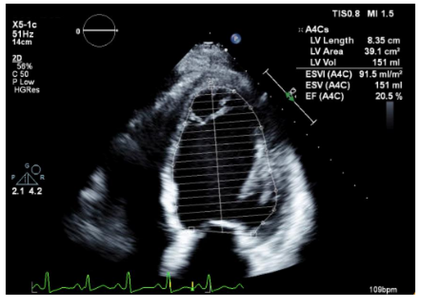
19-year-old previously healthy G1P0 female at 38w4d

2-week viral prodrome → acute progressive dyspnea & chest pain

INITIAL WORKUP

35	7,368	8.9 / 27	209	NEG
ng/L	pg/mL	g/dL / %	x10^3/uL	COVID · Flu · RSV
Troponin	**BNP**	**Hgb / Hct**	**Plt**	**Resp. panel**

TTE: LVEF 20% + dilated ventricles, 1-2+ MR → concern for PPCM



Right Heart Catheterization

RA 12 | RV 39/12(21) | PA 37/26(32) | PCWP 22 | CO/CI 3.5/2.1
SVR 1668 dyn·s·cm^{-5} | PVR 2.8 WU

PREOPERATIVE MANAGEMENT

- L&D floor → Cardiac ICU for optimization & delivery planning
- Multidisciplinary team: OB/Gyn, OB and cardiac anesthesia, cardiology, CT surgery, critical care
- Telemetry + invasive monitoring with Swan-Ganz catheter + arterial line

Delivery Plan

Vaginal delivery with assisted second-stage vs primary cesarean → Urgent CS when patient entered spontaneous labor overnight

INTRAOPERATIVE MANAGEMENT

Titrated Neuraxial Block with Epidural

3cc test dose + repeat 0.5% bupivacaine boluses → surgical block

Pre-Cannulation for VA ECMO

6Fr sheath R femoral vein + 6Fr sheath L femoral artery

✓ **Uncomplicated Low Transverse CS**

Transferred to ICU on with epinephrine support at 0.1 mcg/kg/min

POSTPARTUM COURSE

- Preserved CO/CI; elevated filling pressures → diuresis overnight
- Epinephrine weaned off; femoral ECMO sheaths removed
- Discharged POD #5 with Life Vest wearable defibrillator
- Guideline directed medical therapy (GDMT) initiated
- Persistent severely reduced LVEF < 20% with recurrent HF exacerbations
- LV thrombus → anticoagulation initiated
- Genetic testing revealed pathologic variant in DSP (desmoplakin) gene
- Referred for heart transplantation

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DISCUSSION

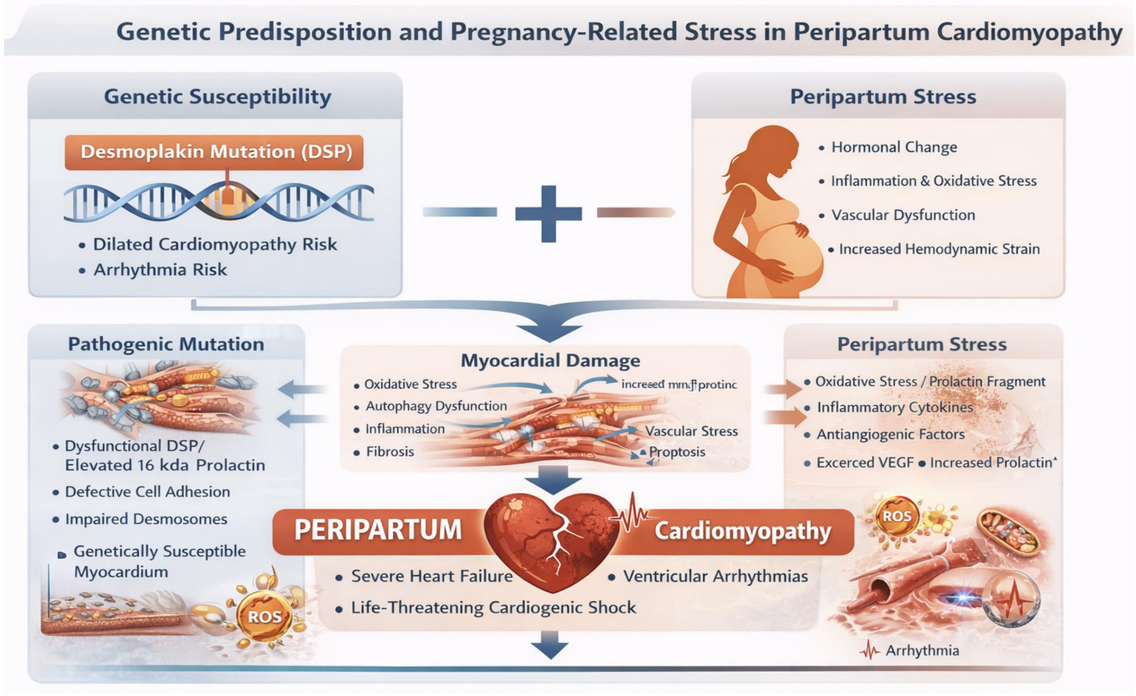
DSP (desmoplakin) is a major structural protein component of desmosomes which are essential for cellular adhesion.

DSP mutations are associated with arrhythmogenic right ventricular cardiomyopathy (ARVC), dilated cardiomyopathy (DCM), biventricular cardiomyopathy and left ventricular arrhythmogenic cardiomyopathy.

Up to 15% of patients with PPCM carry heterozygous loss-of-function mutations in genes implicated in nonischemic DCM, such as TTN (Titin), FLNC (filamen C), and DSP, suggesting a shared disease continuum in which similar genetic susceptibility is unmasked by different environmental or physiologic stressors.

⚠ "Multiple hit" model ⚠

Patients with DSP mutations have structurally vulnerable cardiomyocytes and while these genetic defects may remain clinically silent under normal conditions, the profound physiological stress of pregnancy can precipitate overt cardiomyopathy.



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