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Cesarean Delivery in a Parturient With "Repaired" Congenital Mitral Dysplasia and Severe Functional Mitral Stenosis: A Case Report

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We present the anesthetic management of a 23-year-old gravida 2 para 0-0-1-0 with a history of mitral valve replacement secondary to congenital mitral dysplasia. The hemodynamic changes of pregnancy had resulted in severe functional mitral stenosis of her mechanical valve, which was significantly undersized for her current body surface area. Her complex cardiac history required our multidisciplinary team to focus on peripartum anticoagulation management, extracorporeal membrane oxygenation (ECMO) preparation under a dural puncture epidural (DPE) technique, and managing the hemodynamic changes that are critical for a congenital heart disease parturient. (A&A Practice. 2020;14:e01203.)

GLOSSARY

ADH = antidiuretic hormone; **BMI** = body mass index; **BSA** = body surface area; **CHD** = congenital heart disease; **CD** = cesarean delivery; **CVP** = central venous pressure; **DPE** = dural puncture epidural; **ECMO** = extracorporeal membrane oxygenation; **HIPAA** = Health Insurance Portability and Accountability Act; **ICU** = intensive care unit; **INR** = international normalized ratio; **LMWH** = low-molecular-weight heparins; **LVEF** = left ventricular ejection fraction; **MMV** = mechanical mitral valve; **MV** = mitral valve; **MV meanPG** = mitral valve mean pressure gradient; **NYHA** = New York Heart Association; **PACU** = post-anesthesia care unit; **PIV** = peripheral intravenous; **PPM** = patient-prosthetic mismatch; **PVP** = peripheral venous pressure; **RVSP** = right ventricular systolic pressure; **TEC** = thromboembolic complications; **TTE** = transthoracic echocardiogram

Congenital mitral valve (MV) dysplasia is a rare subset of left-sided congenital heart disease (CHD) characterized by a wide spectrum of morphologic abnormalities but presents primarily as either a regurgitant or stenotic lesion.[1,2] Clinically significant congenital MV lesions are estimated to affect 0.4% of CHD patients of the general population.[2] Current opinion recommends MV repair versus replacement in affected children to reduce the risk of long-term sequelae such as heart block, endocarditis, valve thrombosis, and, importantly, as in our patient, the development of patient-prosthetic mismatch (PPM). PPM is considered after the insertion of a prosthetic valve when the effective orifice area of the prosthesis is less than that of a normal human valve, which results in an increased postoperative transvalvular gradient.[3] Our patient had a 21-mm St. Jude prosthetic valve implanted as a child that had never been replaced and that was significantly undersized for her body surface area (BSA). While her valve appeared to provide adequate hemodynamic function during her ordinary adult life, it proved woefully insufficient to deal with the increased cardiac output and other hemodynamic changes of pregnancy. The patient provided Health Insurance Portability and Accountability Act (HIPAA) authorization for publication.

CASE DESCRIPTION

A 23-year-old gravida 2 para 0 weighing 112 kg (body mass index [BMI] 42.5) with a history of congenital mitral (MV) dysplasia was scheduled for a primary cesarean delivery (CD) at 36 weeks. She had a mechanical mitral valve (MMV) placed at age 4 years that had never been replaced, and she remained on life-long warfarin. The patient had no known cardiac complications until she reached 11 weeks gestational age, at which time she was hospitalized for significant volume overload and New York Heart Association (NYHA) class IV symptoms. She was found to have severe functional mitral stenosis (mitral valve mean pressure gradient [MV meanPG] of 16 mm Hg) due to her MMV, which was undersized for her current BSA 2.14 m2. Inpatient treatment included aggressive diuresis, which reduced her symptoms to NYHA II, and she was discharged home on diuretics in preparation for a planned CD at a later date. She was maintained on warfarin due to the increased incidence of thromboembolic complications (TEC) in patients on low-molecular-weight heparins (LMWH) with mechanical valves, and at the time of presentation, the patient was past the fetal organogenesis period.[4] Serial transthoracic echocardiograms (TTE) throughout pregnancy revealed worsening mitral stenosis with an MV

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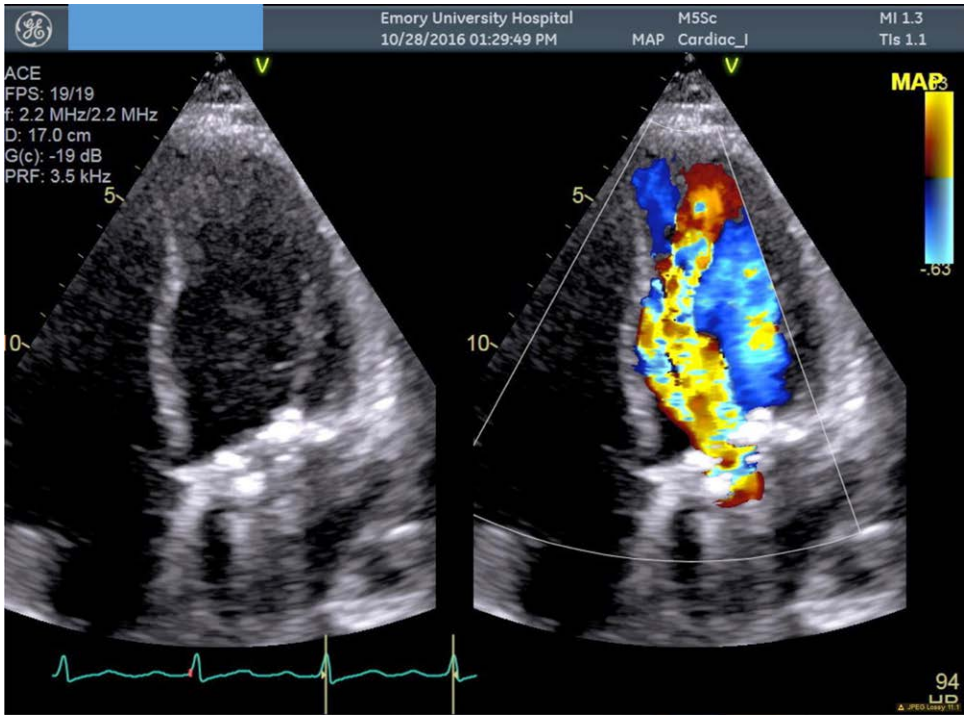

Figure 1. TTE at 24 wk reveals undersized MMV causing "functional" mitral stenosis with significant turbulent flow seen on Doppler. Turbulent flow is represented by the large jet that extends from the MMV into the left ventricle and aliasing seen on color flow Doppler. MMV indicates mechanical mitral valve; TTE, transthoracic echocardiography.

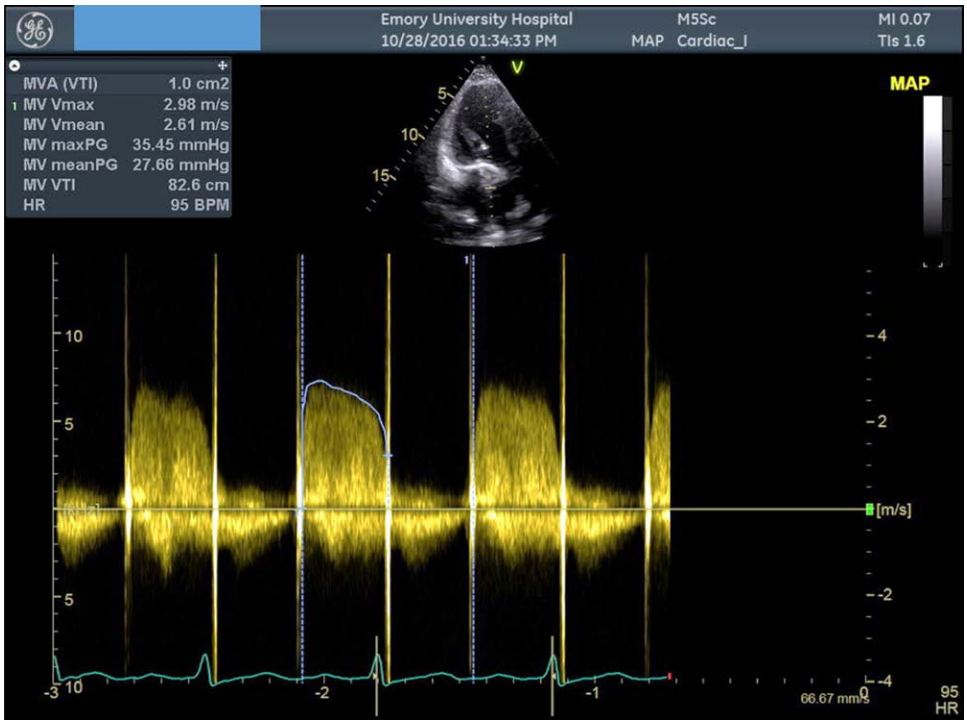

Figure 2. Continuous-wave Doppler reveals the severely elevated MV meanPG 27 mm Hg across the MMV suggesting severe mitral valve stenosis (TTE at 24 wk). Normal MV meanPG is <5 mm Hg, and MV meanPG >10 mm Hg suggests severe mitral stenosis. MMV indicates mechanical mitral valve; MV meanPG, mitral valve mean pressure gradient; TTE, transthoracic echocardiography.

meanPG of 25–27 mm Hg (Figures 1 and 2), moderate pulmonary hypertension (right ventricular systolic pressure [RVSP] 40–45 mm Hg), and gradually decreasing left ventricular ejection fraction (LVEF)—dropping from 60% to 40% over a 1.5-month period. Because of the decrease in LVEF, coupled with her worsening NYHA III symptoms, a decision was made to proceed with a CD at $35^{2/7}$ weeks. She was admitted 6 days before her scheduled CD to allow transition from warfarin to a heparin infusion and to allow final planning between multidisciplinary teams. The heparin was discontinued 6 hours before her CD. Anesthesia for the CD was provided using a dural puncture epidural (DPE), arterial and peripheral venous pressure (PVP) monitoring, and targeted vasopressor and fluid therapy. A 17-G Tuohy needle was guided in the epidural space, and a 26-G Gerti Marx spinal needle was used to provide a dural puncture; no medications were

injected intrathecally, and the spinal needle was removed once cerebrospinal fluid was observed. After placement of an epidural catheter, a T10 level was obtained with 10 mL of 0.5% bupivacaine in divided doses, after which cardiothoracic surgery placed femoral access wires in case there was an emergent need to transition to extracorporeal membrane oxygenation (ECMO). On obtaining a T4 level of anesthesia with further epidural administration of 9 mL 0.5% bupivacaine containing 100 µg of fentanyl, an uneventful CD, requiring minimal vasopressor support, resulted in delivery of an infant weighing 2660 g with Apgars of 8/9. Adequate uterine tone was achieved with carefully titrated intravenous doses of *n*-methylergonovine maleate rather than using an oxytocin bolus or infusion. Methylergonovine was given in 20-µg/mL aliquots with a total dose of 40 µg over 4 minutes to achieve optimal uterine tone. Epidural morphine 3 mg was administered for postoperative pain relief. Estimated blood loss was 800 mL, and replacement fluids included 500 mL of crystalloid and 500 mL of 5% albumin. The epidural catheter and femoral wires were removed 1 hour after finishing the CD, and she recovered in the post-anesthesia care unit (PACU) for 2 hours, followed by transfer to the cardiac intensive care unit (ICU) for continued observation and postoperative diuresis. The heparin infusion was restarted 6 hours after her CD. Her postpartum course was complicated by a rectus sheath hematoma during reinitiation of warfarin therapy. A supratherapeutic international normalized ratio (INR) 6 was observed on day 4 of warfarin reinitiation and was treated with 2 units of fresh frozen plasma to achieve INR 2.5–3.0. The patient received serial CT scans of the abdomen and pelvis to ensure the hematoma was not enlarging, and it resolved without surgical intervention. A gradual hemoglobin decline from 12.5 to 8.0 g/dL prompted the administration of 4 U packed red blood cells over her 2-week hospital course to maintain a hemoglobin of 10 g/dL. The patient was discharged from the hospital 18 days after her CD on warfarin with plans to replace her MMV with a bioprosthetic valve at 6 months after delivery. A follow-up TTE at 1 month postpartum demonstrated that the turbulent flow across her MMV significantly improved, and the MV meanPG decreased to 8 mm Hg with the return of prepregnancy physiology (Figures 3 and 4).

DISCUSSION

Our patient with congenital MV dysplasia presented with NYHA IV symptoms at 11 weeks gestational age. At that time, her symptoms were attributed to her PPM with serial TTEs revealing an MV meanPG of 9 mm Hg (prepregnancy) to 16 mm Hg (11 weeks) to 27 mm Hg (24 weeks). As evident by her prepregnancy MV meanPG, our patient was considered to have moderate "functional" mitral stenosis from her MMV being placed as a child and now undersized for her adult BSA. It is the cardiac physiologic changes of pregnancy that include hypertrophy and increased cardiac output that worsened her PPM and symptomology. These symptoms prompted early cardiac surgery consultation with the decision for ECMO preparation during neuraxial placement or anytime during or after her CD. A scheduled, late-preterm CD was chosen for mode of delivery to minimize time without anticoagulation and to avoid the episodic increase in preload with unpredictable uterine contractions. Consideration was made to continue her ECMO access wires while in the ICU, but our patient tolerated postdelivery autotransfusion without significant hemodynamic instability, so femoral wires were removed shortly after the procedure.

Intraoperatively, we placed a preinduction arterial line and large bore peripheral intravenous (PIV) access (18, 16 g) to guide resuscitation with goals to maintain euvolemia. The 16-g PIV was used to monitor the trend in PVP. PVP is

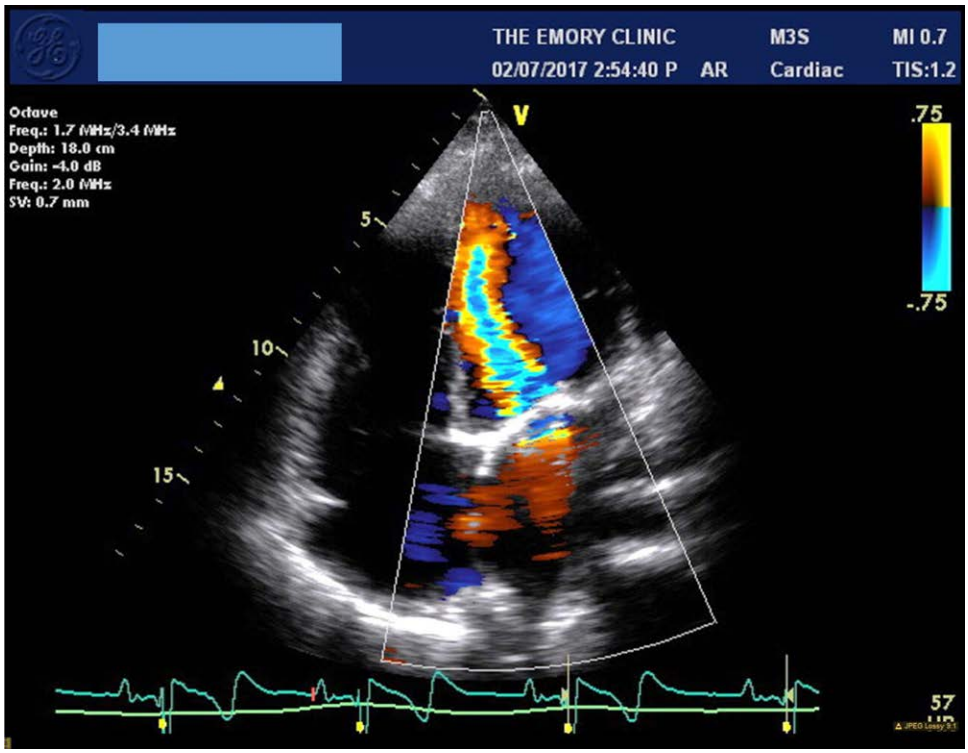

Figure 3. TTE 1 mo postpartum reveals improvement in turbulent flow across the MMV. Turbulent flow improvement is suggested by the decrease in aliasing with interval decrease in the size of turbulent jet. MMV indicates mechanical mitral valve; TTE, transthoracic echocardiography.

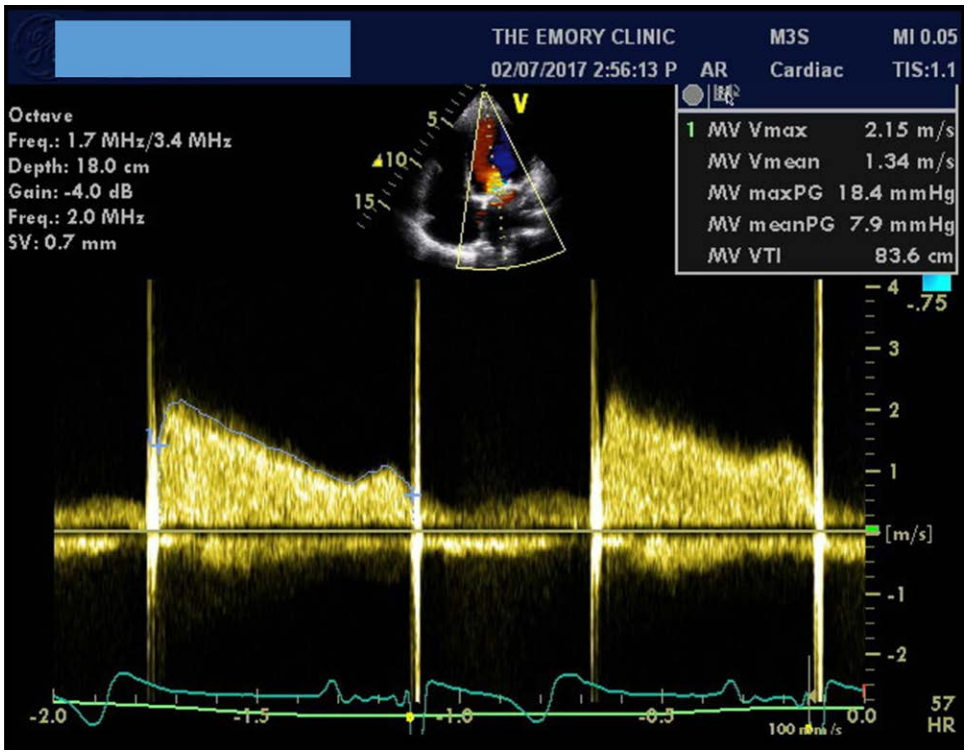

Figure 4. TTE 1 mo postpartum reveals reduced MV meanPG 8 mm Hg across the MMV indicating less severe functional mitral stenosis and return of prepregnancy physiology. MMV indicates mechanical mitral valve; TTE, transthoracic echocardiography.

sensitive for early detection of hypovolemia and displays a strong correlation with central venous pressure (CVP) monitoring.[5,6] In our case, the patient refused central venous line placement, and intraoperative TTE was not readily available, so the PVP was a useful additional monitor in conjunction with the arterial line and physical symptoms. The trend in PVP was maintained between 8 and 10 mm Hg with small boluses of crystalloid when the PVP was <5–6 mm Hg. The patient remained on oral labetalol the morning of her CD, and external defibrillator pads were placed on the patient before DPE dosing in case there was a need for prompt cardioversion secondary to arrhythmias. A DPE was utilized in an effort to provide denser analgesia with less local anesthetic and perhaps provide higher patient satisfaction as compared to a traditional epidural.[7,8] Excellent intraoperative pain control was considered especially important in this patient, as both mitral stenosis and pulmonary hypertension can be negatively impacted by physiological responses to pain—particularly with tachycardia and increased pulmonary vascular resistance. Additionally, it was felt that the DPE technique provides for a surer placement of the catheter into the epidural space and thereby lessens the chances for failed block and the resulting delays in a surgical procedure requiring complex coordination between multiple services.

Choice of uterotonic also played a significant role in this CHD patient. Oxytocin can cause profound tachycardia, hypotension, ischemia, and cardiac arrest, particularly when administered as a bolus.[9] Additionally, the structural similarity between oxytocin and antidiuretic hormone (ADH) allows oxytocin to bind and activate ADH receptors[10] causing water retention and pulmonary edema,[11] making the 500–1000 mL volume usually delivered with oxytocin infusions undesirable. Both the volume load and water retention effects of oxytocin make this uterotonic unfavorable for cardiac patients; hence, we chose methylergonovine. Methylergonovine is a drug that may have fewer cardiac side effects than oxytocin[12] and no effect on water retention. Intravenous boluses of dilute methylergonovine provide a fast onset of myometrial contraction with negligible systemic and pulmonary hypertensive responses at such dilute concentrations.

This case highlights clinical decisions made for a complex CHD parturient, including ECMO preparation, anticoagulation for a MMV, and DPE anesthesia. The postoperative hematoma is a teaching point that stresses warfarin reinitiation should be done cautiously. Perhaps our initial restart dose of 5 mg warfarin (baseline dose) was too aggressive to begin on postoperative day 1, and it subjected our patient to several units of blood products. Finally, a carefully designed neuraxial-based anesthetic plan can assure intraoperative and postoperative comfort, provide optimal hemodynamic goals, and reduce the risks of postdelivery cardiac decompensation, even in classically high-risk CHD patients.

DISCLOSURES

Name: Erica M. Johnson, MD.
Contribution: This author helped prepare the abstract, introduction, case report, and discussion. This author was the primary anesthesiology provider for the case as an obstetric anesthesiology fellow.
Name: James A. Dolak, MD, PhD.
Contribution: This author helped with the design of the case, provided editorial help and reference guidance, and also was involved with final approval of this current report.
This manuscript was handled by: Markus M. Luedi, MD, MBA.

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