JACC: CASE REPORTS VOL. 29, 2024

CASE REPORT

CLINICAL CASE: CARDIO-OBSTETRICS 2023

A Complex Adult Congenital Heart Disease Case in Pregnancy

A Multidisciplinary Approach

Elise Rosenthal, MD,[a] Kerly Guerrero, MD,[b] Manoj Gupta, MD,[c] Kfier Kuba, MD,[d] Diana S. Wolfe, MD, MPH[b,e,f]

ABSTRACT

Multidisciplinary teams decrease the likelihood of adverse pregnancy outcomes in high-risk pregnant cardiac patients. We present the case of a patient with complex congenital heart disease and a mechanical mitral valve, whose treatment included warfarin until delivery despite the discovery of placental hematomas. A multidisciplinary approach mitigated both maternal and fetal adverse pregnancy outcomes. (J Am Coll Cardiol Case Rep 2024;29:102170) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 27-year-old woman, gravida 1 para 0, with repaired complex congenital heart disease, including a prosthetic mitral valve and receiving warfarin, presented at 18 weeks of gestation because of painless vaginal bleeding.

LEARNING OBJECTIVES

- To understand the utility of a cardio-obstetrical team in optimizing care for pregnant patients with mechanical heart valves.
- To review the treatment of pregnant patients receiving therapeutic anticoagulation with warfarin, including the need for careful monitoring and delivery planning.
- To understand the role of placental complications in the management of anticoagulation in pregnant patients.

MEDICAL HISTORY

She had a transitional atrioventricular canal defect with severe left-sided atrioventricular valve regurgitation and a giant left atrium (Video 1, **Figure 1A**), which was repaired surgically but she experienced an immediate postoperative complication requiring revision of an atrial septal patch and left-sided valvuloplasty, and dual-chamber epicardial pacemaker placement at age 23 years in 2013. In 2014, for severe left- and right-sided atrioventricular valve regurgitation, her left-sided atrioventricular valve was surgically replaced with a 29-mm St. Jude mechanical prosthesis, and her tricuspid valve was repaired surgically. A prepregnancy echocardiogram showed a small residual atrial septal defect, borderline low left ventricular systolic function with an ejection fraction

From the [a]Department of Obstetrics and Gynecology, Division of Maternal Fetal Medicine, UT Southwestern Medical Center, Dallas, Texas, USA; [b]Department of Obstetrics and Gynecology, Division of Maternal Fetal Medicine, Montefiore Medical Center and Albert Einstein College of Medicine, Bronx, New York, USA; [c]Department of Pediatric Cardiology, Children's Hospital at Montefiore and Albert Einstein College of Medicine, Bronx, New York, USA; [d]Department of Obstetrics and Gynecology, Division of Maternal Fetal Medicine, Long Island Jewish Medical Center-Northwell Health, New Hyde Park, New York, USA; [e]Maternal Fetal Medicine Cardiology Joint Program, Montefiore Medical Center and Albert Einstein College of Medicine, Bronx, New York, USA; and the [f]Department of Medicine, Cardiology, Montefiore Medical Center and Albert Einstein College of Medicine, Bronx, New York, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received September 8, 2023; accepted October 17, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.102170

ABBREVIATIONS AND ACRONYMS

LMWH = low-molecular-weight heparin

MFM = maternal fetal medicine

UFH = unfractionated heparin

of 50%, and normal mechanical mitral valve inflow gradient (Videos 2 and 3, **Figures 1B and 1C**). Early in pregnancy, she contacted her cardiologist, and warfarin 2 mg nightly was continued, with an INR goal of 2.5 to 3.5. Her care was comanaged by our Maternal Fetal Medicine (MFM)-Cardiology Joint Program.

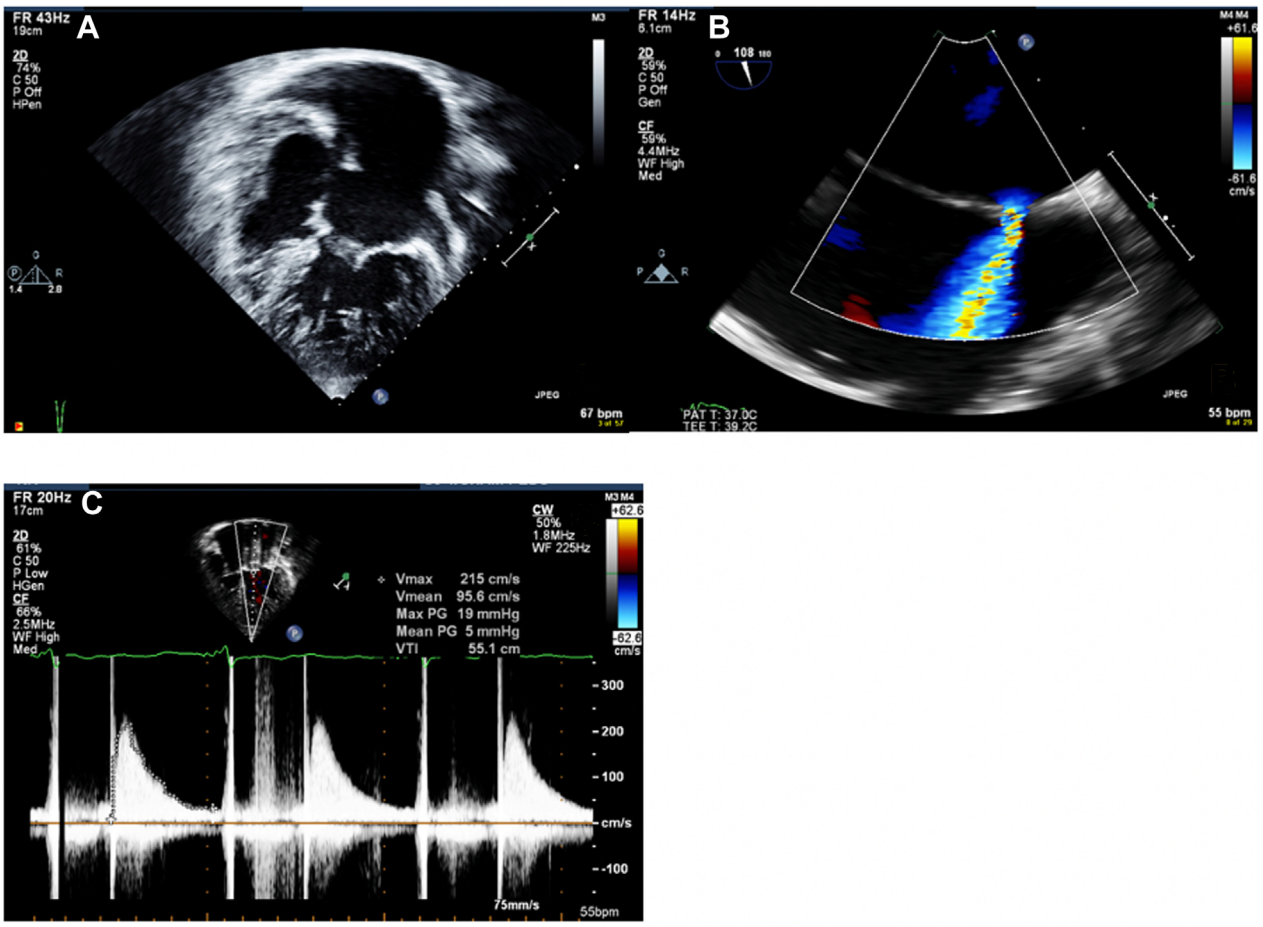
DIFFERENTIAL DIAGNOSIS

Vaginal bleeding can be due to cervicitis, postcoital bleeding, cervical dilatation, subchorionic hemorrhage, and abruption. Chronic abruptions are often painless and were suspected in this patient, who was receiving an anticoagulant agent.

INVESTIGATIONS

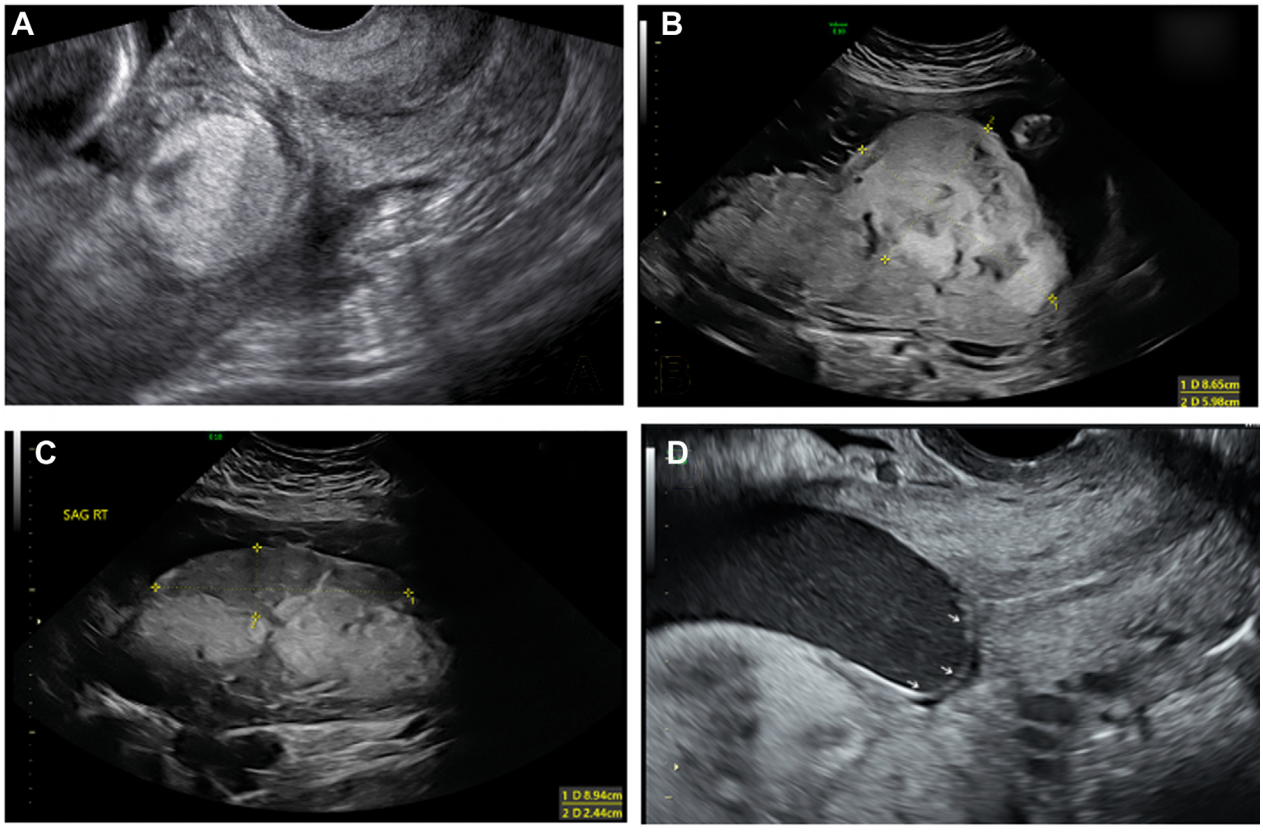
Initial transthoracic echocardiography revealed a left ventricular ejection fraction of 53% and a gradient of 3 mm Hg across the prosthetic mitral valve. At 18 weeks, she presented with painless vaginal bleeding. Her warfarin dose was 4 mg nightly, and her INR was 3.9. Ultrasonography revealed a placental hematoma (**Figure 2**). After an admission and multidisciplinary meeting, her warfarin dose was adjusted, and she was discharged. Fetal anatomy and

FIGURE 1 **Preoperative and Prepregnancy Anatomy**



(A) Preoperative transthoracic echocardiogram (TTE) showing partial atrioventricular canal with large primum atrial septal defect (ASD), severely dilatated left atrium, and hypertrabecular left ventricle. (B) Prepregnancy TEE, bicaval view showing small residual ASD with left-to-right flow. (C) Mitral valve gradient showing mean gradient of 5 mm Hg across the prosthetic mitral valve.

FIGURE 2 **Obstetrical Ultrasound Images**



Ultrasound images at 18 weeks (A) and 25 weeks (B, C) showing placental hematoma, and at 25 weeks (D) showing membrane tenting adjacent to a placental hematoma.

echocardiographic findings were normal. By 25 weeks, 2 placental hematomas developed (Figure 2).

MANAGEMENT

The multidisciplinary plan included serial antepartum fetal testing; on labor and delivery, transitioning anticoagulation to unfractionated heparin (UFH) before neuraxial anesthesia or cesarean delivery; antibiotics for endocarditis prophylaxis;[1] telemetry; pacemaker settings to be changed from DDI to VVI; prophylactic defibrillator pad placement; and an emergency cart located near the patient's room (Figure 3). In case of cesarean delivery, her pacemaker would be asynchronously paced, and post partum she would be admitted to the cardiac care unit and undergo transition to warfarin with UFH bridge.

She had light vaginal bleeding throughout pregnancy, but at 30 weeks she presented with significant bleeding. Her admission vital signs included blood pressure 106/57 mm Hg, heart rate 50 to 60 beats/min, and oxygen saturation 96%. Examination revealed no elevation of jugular venous pressure, mechanical S1, and 3/6 systolic murmur at the left sternal border in the second rib space. The result of vaginal examination was significant for a closed cervix with light bleeding. She was treated by our MFM-cardiology team in anticipation of delivery. Warfarin was discontinued, weight-based intravenous UFH was administered, and serial coagulation study determinations remained stable.

On hospital day 2, she had an acute hemorrhage requiring emergent cesarean delivery under general anesthesia. Her estimated blood loss was 1200 mL, and she received 1 unit of packed red blood cells. Postoperatively, UFH was initiated, and bridged to warfarin on an inpatient basis. Her postpartum course was complicated by hemorrhage related to supratherapeutic anticoagulation, requiring admission to

FIGURE 3 Maternal Fetal Medicine-Cardiology Joint Program Delivery Plan (Abbreviated) Used in the Care of the Patient Described in This Report

IDT MFM-Cardiology Joint Program Delivery Plan

Attendees: Services represented
[x] MFM [x] Cardiology [x] L&D Att [x] L&D Director [x] Patient safety
[x] Anesthesia [x] NICU [x] L&D nursing [x] Blood Bank [x] CICU

Summary of Delivery Plan

- **Anticoagulation transition to UFH drip before and after neuraxial anesthesia or cesarean delivery**
- **Pacemaker settings:** In case of cesarean, her pacemaker would have a magnet placed over it and be asynchronously paced at 85bpm.
- **Emergency crash cart near L&D room**
- **Crossmatch for 4 units of PRBC's on admission**
- **Intrapartum monitoring:**
  - Telemetry: [x] Yes [] No
  - Lines: [] Central line [] Arterial line [x] Peripheral lines (2)
  - Fluid monitoring/goal: [x] Strict I/Os
  - Endocarditis prophylaxis: [x] Yes [] no
- **Anesthesia plan:**
  [] Epidural [x]Spinal [] Combined S/E [] General
- **Risk of decompensation in labor:**
  [x] High [] Moderate [] Low
- **Medications to avoid:** No contraindications
- **In case of hemorrhage:** No contraindications
- **In case of preeclampsia:** Can use magnesium sulfate for seizure prophylaxis

Postpartum Plan:
- **Need for routine recovery in CCU:** [x] Yes [] No
- **Anticoagulation plan:** UFH bridge and start warfarin on POD#1 if no signs/symptoms of bleeding

Disclaimer: The above is intended to serve as guidelines and not intended to be a standard of care. Care should be based on the judgment of the physician based on the individual patient's condition.

the intensive care unit and 2 additional units of packed red blood cells. She was discharged home on postoperative day 15.

DISCUSSION

In the risk stratification of a pregnant patient with congenital heart disease, the cardiac, obstetrical, and perinatal perspectives must be considered.[2] The World Health Organization classification is the most commonly used scoring tool for stratifying patients with congenital heart disease; according to that system, classified our patient was in class III, associated with significantly increased risk for maternal mortality or severe morbidity and a 19% to 27% chance of a cardiac event in pregnancy.[3] Her cardiologist considered that she was in NYHA functional class I to II. According to the ZAHARA (Zwangerschap bij Aangeboren HARtAfwijking-Pregnancy in Women With Congenital Heart Disease) risk classification, she had a 25% to 70% increased risk for an adverse pregnancy outcome.[4] She was counseled on her high risk of maternal morbidity and mortality before and during pregnancy with our MFM-Cardiology Joint Program, including the option of pregnancy termination.

Pregnancy increases the risk of thrombosis with mechanical heart valves, and antepartum and intrapartum management requires careful planning. Warfarin is the gold standard anticoagulant for patients with mechanical valves throughout pregnancy, including in the first trimester. However, even with adequate anticoagulation, the risk of thromboembolic complications remains increased in pregnancy, with rates of ≤2.7%, and there is a risk of significant fetal and maternal adverse effects.[5]

There is emerging literature regarding the safety and efficacy of low-molecular-weight heparin (LMWH) for anticoagulation for pregnant patients with mechanical heart valves. LMWH may be a suitable alternative to warfarin because it does not cross the placenta and has improved live birth rates and perinatal outcomes compared with warfarin.[5] Recent guidelines by cardiac societies, including the American College of Cardiology/American Heart Association and the European College of Cardiology, have recommended the use of LWMH with peak anti-Xa level monitoring. However, these recommendations are based on prior experience with the use of this therapy rather than on randomized clinical trials. In 1 study, monitoring of peak anti-Xa levels with goals between 0.77 and 1.2 U/mL in patients with mechanical heart valves was associated with subtherapeutic trough anti-Xa levels in 80% of patients.[6] Although LMWH is associated with increased risk of thrombosis, at 8.7%,[5] case reviews of outcomes associated with valve thrombosis suggest an association with inadequate dosing, improper monitoring, or subtherapeutic peak anti-Xa levels.[6] The Society for Maternal Fetal Medicine recommends the monitoring of both peak and trough anti-Xa levels every 1 to 2 weeks, with a goal trough level of >0.6 U/mL and goal peak level of 1 to 1.2 U/mL for mechanical mitral valves.[7]

The obstetrical risks of anticoagulation in pregnancy include increased rates of pregnancy loss, placental abruption, and postpartum hemorrhage.[3,7] Additionally, warfarin use is associated with fetal anomalies that are collectively referred to as warfarin embryopathy. Furthermore, a maternal history of congenital heart disease increases risk of that in the fetus.[1] Our patient had normal fetal anatomy and fetal echocardiogram. She patient experienced placental hematomas consistent with chronic placental abruption in the setting of supratherapeutic INR, which ultimately led to premature delivery.

Finally, contraceptive counseling is particularly important in the care of medically complex patients. Contraceptive methods should be discussed throughout a patient's reproductive lifespan, including at prepregnancy counseling and during pregnancy. Our patient was again counseled on her significant cardiac and obstetrical risks in a

subsequent pregnancy, including a high risk of cardiac event. Long-acting reversible contraceptive methods were recommended.[8]

FOLLOW-UP

The patient underwent the implantation of an etonogestrel contraceptive at her postpartum visit. The neonate was discharged on day of life 38 after an uncomplicated admission requiring respiratory and feeding support.

CONCLUSIONS

With multidisciplinary team planning, complications were managed efficiently with mitigation of morbidity, leading to favorable outcomes despite premature delivery. Although anticoagulation for mechanical heart valves can be challenging in pregnancy, counseling and a multidisciplinary approach can improve management.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Elise Rosenthal, UT Southwestern Medical Center, 5323 Harry Hines Boulevard, G6.206, Dallas, Texas 75390, USA. E-mail: Elise.Rosenthal@utsouthwestern.edu. @elisearosenthal.

REFERENCES

1. Hollier LM, Martin JN, Connolly H, et al. ACOG Practice Bulletin No. 212: pregnancy and heart disease. *Obstet Gynecol*. 2019;133(5):e320–e356.

2. Wolfe DS, Hameed AB, Taub CC, Zaidi AN, Bortnick AE. Addressing maternal mortality: the pregnant cardiac patient. *Am J Obstet Gynecol*. 2019;220(2):167.e1-e167.

3. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, et al. ESC Scientific Document Group. 2018 ESC guidelines for the management of cardiovascular diseases during pregnancy. *Eur Heart J*. 2018;39(34):3165–3241.

4. Drenthen W, Boersma E, Balci A, et al. Predictors of pregnancy complications in women with congenital heart disease. *Eur Heart J*. 2010;31(17):2124–2132.

5. D'Souza R, Ostro J, Shah PS, et al. Anticoagulation for pregnant women with mechanical heart valves: a systematic review and meta-analysis. *Eur Heart J*. 2017;38(19):1509–1516.

6. Goland S, Schwartzenberg S, Fan J, Kozak N, Khatri N, Elkayam U. Monitoring of anti-Xa in pregnant patients with mechanical prosthetic valves receiving low-molecular-weight heparin: peak or trough levels? *J Cardiovasc Pharmacol Ther*. 2014;19(5):451–456.

7. Pacheco LD, Saade G, Shrivastava V, Shree R, Elkayam U, Publications Committee. Society for Maternal-Fetal Medicine Consult Series #61: Anticoagulation in pregnant patients with cardiac disease. *Am J Obstet Gynecol*. 2022;227(2):B28–B43.

8. Curtis KM, Tepper NK, Jatlaoui TC, et al. U.S. Medical Eligibility Criteria for Contraceptive Use, 2016. *MMWR Recomm Rep*. 2016;65(No. RR-3):1–104.

KEY WORDS cardio-obstetrics, high-risk pregnancy, mechanical mitral valve

APPENDIX For supplemental videos, please see the online version of this paper.