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VALVULAR HEART DISEASE

CASE REPORT: CLINICAL CASE

# Pregnancy With Severe Valve Disease in Low-Resource Populations

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## ABSTRACT

A 32-year-old pregnant woman from a remote area presented with chest pain and was diagnosed with critical aortic stenosis. Her care was managed by a multidisciplinary team with virtual consultations arranged until her admission. She successfully delivered with all necessary backup facilities in place to manage potential complications. (JACC Case Rep 2024;29:102438)



## HISTORY OF PRESENTATION

A 32-year-old woman presented to the valve clinic at 22 weeks of gestation with complaints of chest pain and shortness of breath. Upon physical examination, her heart rate was 100 beats/min, with a regular rhythm, and her blood pressure measured 100/70 mm Hg, accompanied by a respiratory rate of 20 breaths/min. The patient appeared pale, and multiple xanthomas were noted over her knee, back, and feet (**Figures 1A to 1C**). An ejection systolic murmur of grade III at the aortic area was detected upon auscultation. Pulmonary, abdominal, and neurological examinations yielded unremarkable findings.

### LEARNING OBJECTIVES

- To understand that pregnancy can be continued with better outcomes in critical aortic stenosis patients through collaborative team management involving a cardiologist, obstetrician, and anesthetist experienced in cardiac anesthesia, along with the backup support of cardiac surgery and critical care teams.
- To appreciate the importance of establishing standardized care for high-risk pregnancies with valvular heart diseases in developing countries.
- To acknowledge the crucial role of telemedicine in the follow-up of high-risk pregnant patients with valvular heart conditions during antenatal care in resource-limited settings.

## PAST MEDICAL HISTORY

This marked her fifth pregnancy, with 2 healthy children and a history of 1 abortion and 1 neonatal death. However, she had not undergone any detailed workup. Additionally, she had a family history of sudden cardiac death, including her sister at the age of 4 years and her father at the age of 35 years.

## DIFFERENTIAL DIAGNOSIS

Based on the presenting signs and symptoms, a suspected diagnose of peripartum cardiomyopathy and valvular heart disease were made.

## INVESTIGATIONS

A 12-lead electrocardiogram was conducted, revealing ST-segment changes in leads $V_2$ to $V_6$ (**Figure 2**).

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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.

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**ABBREVIATIONS AND ACRONYMS**

**CT** = computed tomography

**TTE** = transthoracic echocardiography

Transthoracic echocardiography (TTE) was then performed, indicating normal biventricular systolic function which also revealed a calcified aortic valve and aortic root, along with critical valvular and supravalvular aortic stenosis. Measurements showed a mean pressure gradient of 106 mm Hg, peak pressure gradient of 163 mm Hg across the aortic valve, and an aortic valve area of 0.3 $cm^2$, determined by the continuity equation method. Further measurements included an annulus diameter of 14 mm, sinus diameter of 20 mm, sinotubular junction diameter of 15 mm, and ascending aorta diameter of 22 mm (**Figures 3A, 3B, and 4**).

She had previously undergone cardiac computed tomography (CT) before being referred to the valve clinic, during which she was unaware of her pregnancy. The CT scan was reviewed, revealing a tricommissural aortic valve, along with a small and calcified aortic root and ascending aorta, demonstrating significant supra-aortic stenosis (**Figures 5A and 5B**). The calcium scores were also assessed, with aortic valve calcium score measuring 1,600 AU and left main coronary artery calcium score measuring 2 AU (**Figures 6A and 6B**).

Her routine laboratory results indicated anemia (hemoglobin 9 g/dL) and hypercholesterolemia, with total cholesterol levels measuring 505 mg/dL, high-density lipoprotein at 26 mg/dL, low-density lipoprotein at 518 mg/dL, and triglycerides at 148 mg/dL. Previous medical records also included biopsy reports of skin lesions diagnosed as xanthomas. Additionally, her obstetric and fetal ultrasound scans showed normal findings. Immediate pre-delivery TTE showed consistent findings with ejection fraction (biplane) of 55% (Videos 1 to 4).

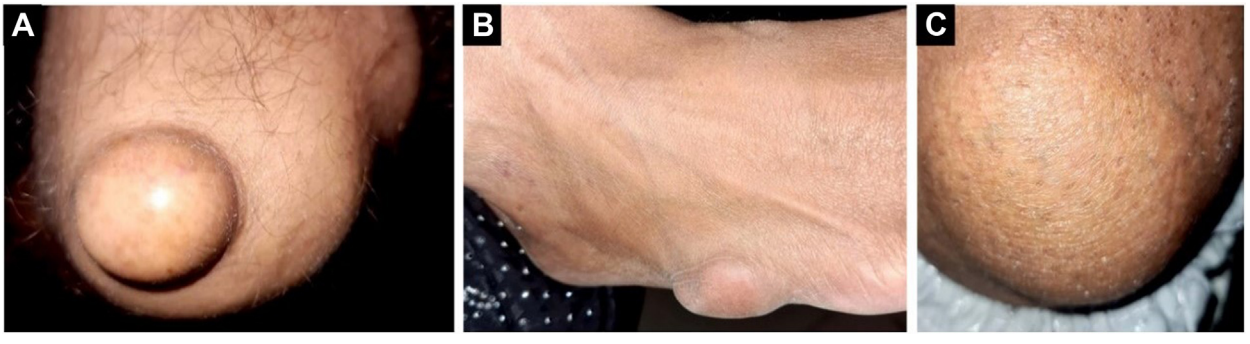
## MANAGEMENT

Medical management was determined because the patient was already in her second trimester, the valve was heavily calcified, and the annulus size was small. She received counseling regarding her diagnosis, and a same-day obstetrician visit was arranged. She was prescribed bisoprolol 5 mg/d and folic acid 5 mg/d. She was transfused with RBC concentrate followed by intravenous iron therapy for anemia. During delivery, the hemoglobin level was 12 g/dL. For familial hypercholesterolemia, in the absence of options such as plasmapheresis, we initiated high-intensity statin therapy and screened all family members, starting them on statins as well.

Given her need to travel to her hometown (575 km from our hospital), it was decided to conduct weekly virtual follow-ups via telephone. The patient was provided with the contact numbers of 2 fellows to reach out to in case of any emergency outside of scheduled telemedicine follow-ups, ensuring direct access to medical assistance when needed. Vitals were monitored remotely by the patient's husband as well as by local paramedical staff, ensuring accuracy. This framework ensured continuous remote monitoring and support, enabling prompt intervention and care continuity during the pregnancy.

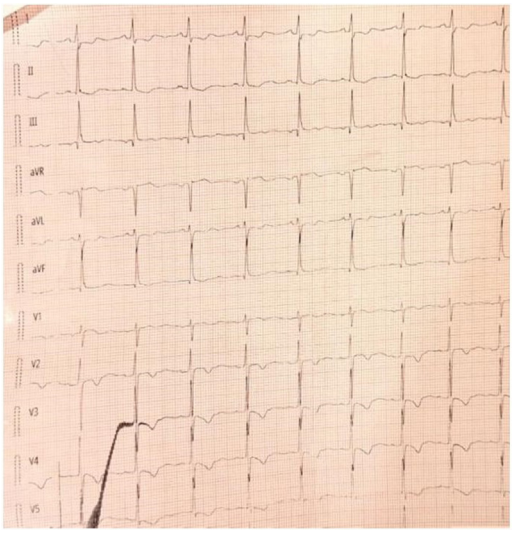
During one of these sessions, she reported a high-grade fever, which was diagnosed as falciparum malaria and successfully managed. Throughout these follow-ups, direct communication was maintained between the multidisciplinary team to monitor the patient's condition and plan her delivery. Subsequently, she was called for admission at the 30th week of gestation and was hospitalized for 4 weeks.

**FIGURE 1** Xanthoma




Xanthomas on knee (A), foot (B), and thigh (C).

**FIGURE 2** Electrocardiogram




12-lead electrocardiogram at presentation showing normal sinus rhythm.

The cesarean section was planned with a multidisciplinary team approach, comprising cardiologists, an obstetrician, and a cardiac anesthesiologist. Critical care backup arrangements were also put in place, along with a ventilator facility to address any potential complications. Additionally, the neonatal intensive care unit was informed about the case to prepare for the neonate.

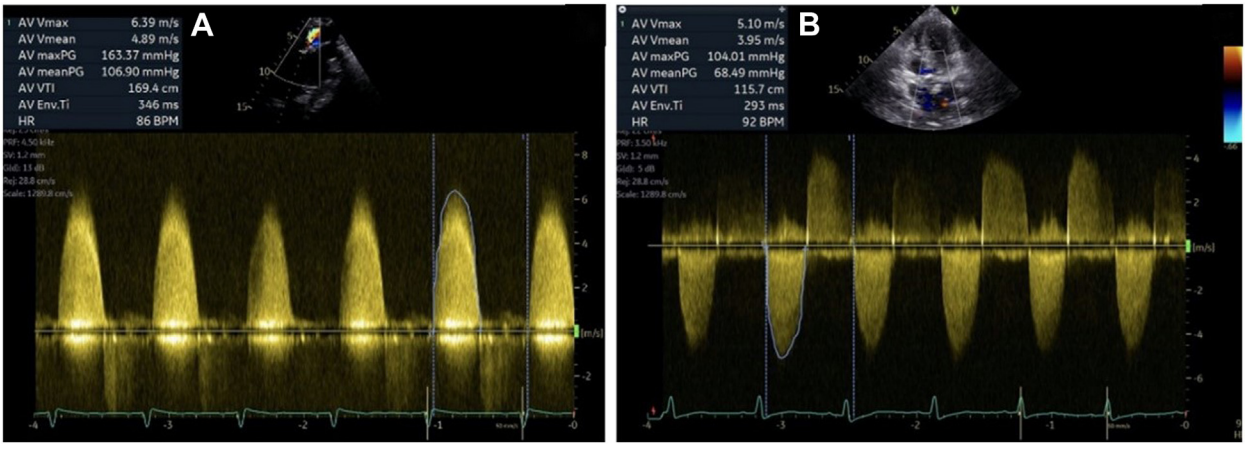
Epidural anesthesia was administered below the L4 level to prevent hemodynamic instability. The cesarean section proceeded swiftly, with a delivery time of <5 minutes. Immediate assessment of the newborn was conducted by a pediatrician, who then transferred the baby to the neonatal intensive care unit. Tubal ligation was performed during the procedure after obtaining informed consent. Although phenylephrine and epinephrine were available as backup measures for hypotension, they were ultimately not required.

## FOLLOW-UP

The patient did not experience tachycardia or hypotension during the perioperative period. Following delivery, she was placed under observation in the high dependency unit, and her hospital stay was uneventful. She was discharged on the third postoperative day with plans for aortic valve replacement in the coming months. A baby boy weighing 1.5 kg was delivered with a good APGAR score.

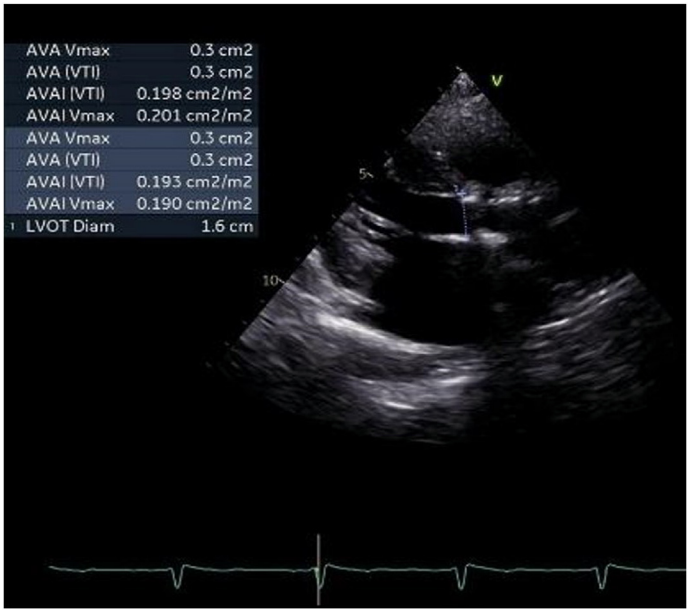
All of her siblings underwent screening, revealing normal echocardiograms, but they were diagnosed with familial hypercholesterolemia and initiated on statin therapy (rosuvastatin 20 mg at bedtime). Subsequent echocardiography after delivery indicated decreased gradients compared to the previous study, with a mean pressure gradient of 68 mm Hg and a peak pressure gradient of 104 mm Hg (Figure 4). Additionally, during the postnatal period, she developed an infected lipoma wound, which required surgical debridement. It was decided to let this heal and

**FIGURE 3** Aortic Valve Gradient: Transthoracic Echo Findings




Transthoracic echocardiograms showing gradients across aortic valve during pregnancy (A) and post delivery (B).

**FIGURE 4 Continuity Equation Measurement of Aortic Valve Area**




Transthoracic echocardiogram showing aortic valve area by continuity equation mesuring 0.3 $cm^2$.

bring her back for valve replacement in a few weeks. Additionally, she was uptitrated on rosuvastatin and ezetimibe, with a dosage of 20/10 mg at bedtime.

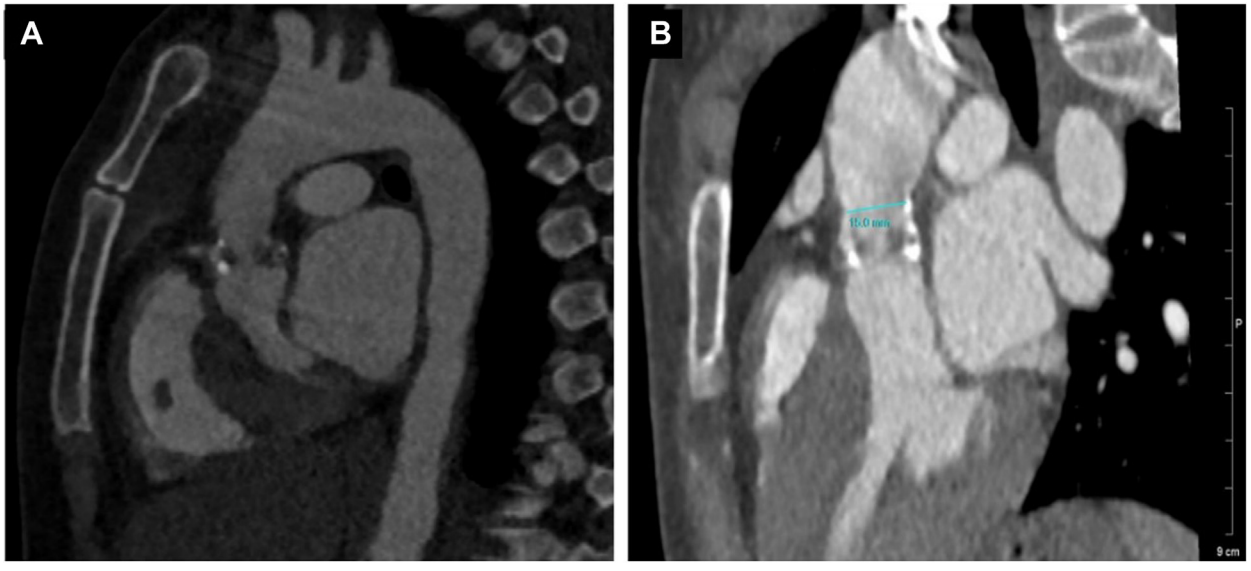
Two months after delivery, the patient returned with intermittent angina and was admitted for surgery. Coronary angiography revealed left main coronary artery stenosis of 40% to 50% and right coronary artery stenosis of 50% to 60%.

The patient underwent aortic valve replacement with an 18 mm bi-leaflet valve and coronary artery bypass grafting for intermediate left main coronary disease and aortic root expansion. Unfortunately, the patient had a complicated course, her ejection fraction dropped intraoperatively and ultimately the patient passed away on the second postoperative day.

## DISCUSSION

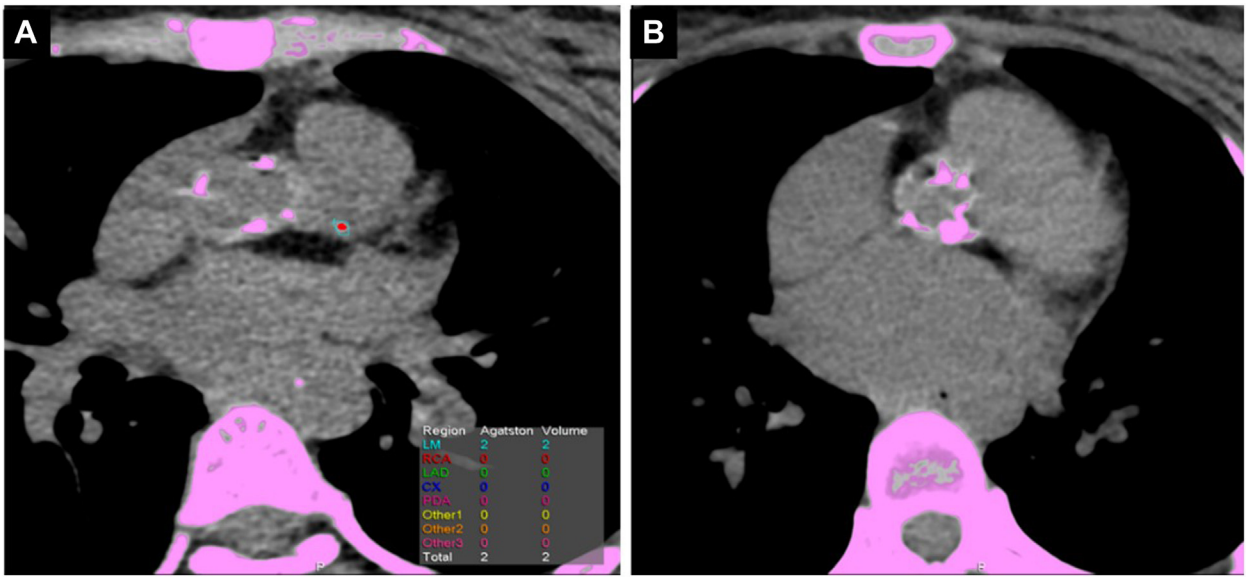
In patients with severe aortic stenosis, the maternal mortality rate is reported at 17.4%, whereas the perinatal mortality rate stands at 31.6%.[1] In resource-limited settings lacking screening measures, severe valvular lesions may remain undetected before pregnancy, potentially leading to maternal or perinatal mortality. It is crucial to raise awareness within the community about valvular heart diseases and

**FIGURE 5 CT Imaging: Aorta and Aortic Root**




Cardiac computed tomography showing aorta and aortic root (A) and calcificaiton of aorta (B).

**FIGURE 6** Cardiac CT: Aortic and Left Main Coronary Calcium Scoring




Cardiac computed tomography showing calcium scoring of aorta (A) and left main coronary artery (B).

their history, as thorough examinations can aid in screening valvular patients. Additionally, there is an established association between hypercholesterolemia and valvular or supravalvular aortic stenosis.[2]

The main modes of therapy for severe aortic stenosis include medical treatment, valve replacement, and termination of pregnancy. Medical management involves strict bed rest, beta-blockers, cautious use of diuretics, and avoidance of drugs that reduce afterload.[3] Cesarean section is indicated for delivery in severe aortic stenosis patients.[4] Optimal outcomes necessitate multidisciplinary care before, during, and after delivery.[5] A cardiac surgery team equipped with a standby cardiopulmonary bypass machine is essential for ensuring a safe delivery.[6]

Telemedicine plays a crucial role in advancing patient care.[7] In resource-limited countries, telemedicine holds the potential to closely monitor high-risk valvular pregnancies when a nearby specialized health care facility is not accessible. Establishing a network between multidisciplinary teams is essential for comprehensive management and improved outcomes for pregnant patients with valvular conditions.

During pregnancy, an increase in gradients can be expected due to the increased plasma volume and hyperdynamic state.[8] Post-delivery assessment of gradients across the aortic valve is recommended.

## CONCLUSIONS

A successful outcome in this pregnancy with critical aortic stenosis was achieved through a multidisciplinary team approach, close monitoring with telemedicine follow-up, and hospitalization from the 30th week of gestation until delivery. Unfortunately, the patient did not survive the surgery. Screening of her siblings facilitated the diagnosis of familial hypercholesterolemia within the family, enabling early initiation of treatment.

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

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**KEY WORDS** aortic stenosis, multidisciplinary, pregnancy, team approach, telemedicine

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


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