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Fetomaternal Outcomes in Late Presentation of Large Unrepaired Ventricular Septal Defect with Pulmonary Arterial Hypertension: A Case Report

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Short Report

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Abstract

Introduction

Congenital Heart Disease complicates about 1% of pregnancies worldwide. In sub-Saharan Africa, limited access to paediatric cardiac surgery creates a group of "natural survivors" who often first present during pregnancy. Ventricular septal defects associated with pulmonary arterial hypertension pose significant maternal mortality risks, often exceeding 28%.

Case presentation

A 20-year-old primigravida at 37 weeks of gestation presented with false labour and New York Heart Association Class III heart failure symptoms. Echocardiography revealed a large 1.7-cm perimembranous ventricular septal defect with moderate pulmonary arterial hypertension, with right ventricular systolic pressure of 56 mmHg, classifying her as modified World Health Organization Class IV. Despite multidisciplinary management by a "Pregnancy Heart Team," she rapidly deteriorated to New York Heart Association Class IV. We performed an emergency caesarean section under general anaesthesia due to non-reassuring fetal status. She delivered a live male infant weighing 2600 grams. Postoperatively, we closely monitored her in the intensive care unit. We discharged her home on postoperative day 12, and both the mother and baby are doing well.

Conclusion

Favourable pregnancy outcomes are possible in late-presenting pregnant women with congenital heart disease through early intervention by the "Pregnancy Heart Team." Also, this case highlights the limitations of standard risk-stratification tools for cardiac events in pregnancy in sub-Saharan Africa. Improvements in antenatal screening and increased access to definitive cardiac surgical correction before conception are important for lowering maternal morbidity and mortality in low-resource settings.

Introduction

Congenital Heart Disease (CHD) affects 1% of pregnancies, resulting in 1.5 million cases each year, including 500,000 in Africa and 11,737 in Kenya [1]. While developed countries achieve survival rates exceeding 90% through advanced paediatric cardiac care, 90% of children in sub-Saharan Africa (SSA) lack access to definitive cardiac surgery [1]. This delays diagnosis until childbearing age, creating a population of "natural survivors" [1]. Given the region's high fertility rates, an estimated 10% to 20% of affected individuals survive naturally, adding approximately 30,000 to 60,000 new patients annually to the pool of African Adult Congenital Heart Disease [1]. Although ventricular septal defect (VSD) accounts for 35.6% of global CHD, its pooled prevalence in Kenya is 16.97%, compared with 29.92% across East Africa [2].

Cardiac disease is a leading cause of maternal death, accounting for 10% globally and 9.3% in Kenya [3]. In Kenya, CHD during pregnancy is linked to a 79.8% incidence of adverse cardiac, obstetric, or neonatal events [3]. In low-resource settings such as Kenya, late diagnosis in the third trimester can lead to serious complications such as congestive heart failure in 42.3% of patients and ICU admission in 21.7% [3]. Additionally, neonatal risks include preterm delivery (41.6%) and low birth weight (30.4%), often requiring interventions like labour induction (37.0%) and vacuum-assisted delivery (19.8%) [3].

Management follows the modified World Health Organization (mWHO) classification, though standard risk stratification tools for cardiac events in pregnancy, such as the CARdiac disease in PREGnancy score (CARPREG I), often underestimate complication rates in SSA [4]. In Kenya, patients classified as "low risk" by CARPREG I have demonstrated a 29% cardiac event rate, likely due to late antenatal presentation and poor access to subspecialised care [3]. Uncorrected VSD (mWHO Class II) carries a 15% risk of pre-eclampsia and 9.6% rate of postpartum haemorrhage [4]. These risks are worsened by the physiological demands of pregnancy, including a 50% increase in cardiac output and a 40% increase in plasma volume [5]. These hemodynamic changes can trigger or worsen pulmonary arterial hypertension (PAH), which carries maternal mortality rates up to 28% and a 31 to 35% risk of right ventricular failure [6, 7].

We report a 20-year-old primigravida with a 1.7 cm perimembranous VSD and moderate PAH (mWHO Class IV), managed by a multidisciplinary “Pregnancy Heart Team” at a Kenyan tertiary hospital in early 2026.

Case presentation

Patient information

S.N., a 20-year-old primigravida, presented to the hospital on 14 February 2026. She was at a gestational age of 37 weeks and 6 days, based on her last normal menstrual period of 25 May 2025. She was a self-referral from Kisii with a history of CHD VSD diagnosed in infancy. Corrective surgery was recommended at a tertiary hospital, but it was never performed due to financial constraints. She lacked any recent imaging and had lost her medical records.

Clinical findings

S.N reported three days of non-radiating lower abdominal pain that lacked the frequency or intensity of true labour. She described awareness of her heartbeats and easy fatigability even with mild to moderate activity, consistent with New York Heart Association (NYHA) Class III heart failure. Examination showed fair general condition, no oedema, no jaundice, and no pallor. Her vital signs were stable, with a blood pressure of 126/65 mmHg, a pulse of 95 bpm, and an oxygen saturation of 97%. Cardiovascular assessment revealed a hyperactive precordium with a palpable thrill and a prominent pansystolic murmur. Obstetrical examination showed a full-term symphysis-fundal height; the fetus was in a

longitudinal lie, cephalic presentation, with a fetal heart rate of 142 bpm, while a vaginal exam showed a posterior, closed cervix with no "show" present.

Electrocardiography, echocardiography, obstetric ultrasound, and laboratory assessments

Laboratory results were within normal limits, except for mild anaemia (haemoglobin 11.46 g/dL) and a slightly low total protein of 64.82 g/L (Table 1). An obstetric ultrasound confirmed a single live intrauterine fetus at 36 weeks and 5 days of gestation in cephalic presentation, with a biophysical profile of 8/8 and weighing 3074 grams. Additionally, echocardiography (ECHO) revealed a large 1.7-cm perimembranous VSD with a left-to-right shunt and moderate pulmonary PAH, with a right ventricular systolic pressure of 56 mmHg (Figs. 1–7). Also, an electrocardiogram (ECG) showed left ventricular hypertrophy (Fig. 8).

Table 1
Laboratory investigations results

Laboratory investigation	Result	Reference Range
Full hemogram		
Hemoglobin	11.46 g/dL	[13–18]
Hematocrit	34.50%	[40–54]
White Blood Cell Count	5.56 × 10^9/L	[4–11]
Platelets	258 × 10^9/L	[150–450]
Liver Function		
Total Protein	64.82 g/L	[65–80]
Albumin	35.6 g/L	[35–50]
AST[a]	23.1 U/L	[0–50]
ALT[b]	20.9 U/L	[0–70]
Bilirubin (Total)	16.51 µmol/L	[0–17]
Renal & Electrolytes		
Sodium	135.4 mmol/L	[135–155]
Potassium	3.71 mmol/L	[3.5–5.5]
Urea	2.80 mmol/L	[2.5–7.5]
Creatinine	65.4 µmol/L	[40–107]

Abbreviations note: [a]aspartate aminotransferase, [b]alanine aminotransferase

Timeline and diagnostic assessment: We admitted her on 14 February 2026, following an initial diagnosis of false labour and CHD. The obstetric fetomaternal specialist evaluated her, and by 16 February, her condition had worsened as she experienced dyspnea at rest and palpitations, leading to an updated diagnosis of VSD complicated by PAH. A cardiologist and an anaesthesiologist reviewed her. On 19 February, we induced labour at 1:00 am, but at 3:00 pm, after artificial rupture of membranes at five centimetres of dilation, we noted meconium-stained amniotic fluid, along with fetal bradycardia of 90 bpm, which necessitated caesarean delivery. In the postoperative period, the patient developed a productive cough and mild shortness of breath on day seven, which resolved by day 10. After a stable recovery, we discharged her home on postoperative day 12.

Final diagnosis

A primigravida at term with a large perimembranous ventricular septal defect (VSD), moderate pulmonary arterial hypertension (PAH) mWHO Class IV. She initially had an NYHA Class III heart failure, which progressed to NYHA Class IV symptoms, evidenced by dyspnea at rest.

Therapeutic intervention and outcome

We prescribed Enoxaparin (40 mg), Ceftriaxone (2 g), and Carvedilol (3.125 mg). For delivery, we attempted a double induction using misoprostol and an extra-amniotic catheter, supported by epidural anaesthesia. Due to fetal bradycardia, we performed an emergency caesarean delivery under general anaesthesia. The outcome was a live male infant weighing 2600 grams with APGAR scores of 8 at 1 minute and 9 at 5 minutes. Postoperatively, we stabilised the patient in the ICU with continuous monitoring and individualised medications, including prophylactic anticoagulation.

Follow-up and prognosis: Maternal Outcome: The patient's post-operative recovery was complicated by a respiratory tract infection on day seven, which we successfully treated with broad-spectrum antibiotics. By Day 10, her cough had resolved, and we discharged her on Day 12. The maternal prognosis requires long-term cardiac monitoring. Therefore, we scheduled her for follow-up in the Gynaecological, Cardiothoracic, and Cardiology Outpatient Clinics. She is currently doing well.

Patient perspective

My pregnancy was filled with anxiety due to many years of untreated heart disease. I am thankful for the good care that saved my baby and me, and I hope to finally undergo the heart surgery I need.

Discussion

In SSA, the lack of paediatric cardiac care, including definitive surgery for congenital heart lesions, results in a growing number of "natural survivors," adding approximately 30,000 to 60,000 new patients each year to the pool of "African Adult Congenital Heart Disease" (ACHD) [1, 2]. S.N.'s presentation in her third trimester with NYHA Class III symptoms is typical for Kenya, where 86% of CHD cases are

diagnosed late, often leading to congestive heart failure in 42.3% of cases [3]. While small perimembranous VSDs, the most common subtype at 74%, are often well tolerated, large defects, such as the 1.7 cm lesion in this case (Fig. 1 and Fig. 2) carry a 32% risk of cardiac complications and significantly higher neonatal risks, including 14% preterm birth and 18% low birth weight, as documented in Bangladesh [7].

Cardiovascular physiology and risk stratification

Echocardiography was essential in this case, revealing moderate PAH with a right ventricular systolic pressure (RVSP) of 56 mmHg (Fig. 3). In Group 1 PAH, approximately 50–70% of patients with large VSDs develop PAH [8]. This classified her as mWHO Class IV, a high-risk category where pregnancy is generally discouraged and patients in this group should ideally be counselled on permanent contraception, such as tubal occlusion, before conception [4, 6, 9]. Standard risk-stratification tools such as CARPREG I often underestimate risks in settings such as SSA; for example, Kenyan patients with a CARPREG I score of 0 still experience a 29% cardiac event rate, likely due to limited diagnostic investigations such as Echo and late presentation in pregnancy with 86.0% of patients being identified only in the third trimester or postpartum [3, 10].

The physiological stress of pregnancy is considerable, with a 50% increase in cardiac output and a 40% rise in plasma volume [5]. These changes are accompanied by a 30%–40% decrease in systemic vascular resistance (SVR) [5]. These hemodynamic alterations during pregnancy often exacerbate volume overload in unrepaired defects [5]. This was clearly demonstrated by the high-velocity, mosaic-coloured jet on Colour Doppler (Fig. 4), which confirms the high-volume left-to-right shunt contributing to the patient's pulmonary over-circulation. Such chronic overcirculation eventually causes significant dilatation of the left atrium (Fig. 5) and the main pulmonary artery (Fig. 6), as seen in this patient. The structural impact of the 1.7-cm defect was further evidenced by cardiac chamber remodelling. M-mode echocardiography confirmed significant dilation of the left ventricular internal diameter in diastole, with Left Ventricular Internal Diameter in end-diastole (LVIDd) of 6.50 cm, a hallmark of chronic volume overload [5, 7]. Despite this notable dilation and the high-volume shunt, the patient maintained a preserved ejection fraction of 53.8% (Fig. 7). This illustrates the heart's compensatory mechanisms, specifically the Frank-Starling mechanism, whereby cardiac output increases as end-diastolic pressure rises, prior to the rapid clinical deterioration observed in our case in the 48 hours preceding delivery [4, 5].

Diagnostic challenges and clinical markers

Distinguishing heart failure from the physiological changes of a normal pregnancy, such as mild sinus tachycardia, peripheral oedema, or a gravid uterus-induced left axis deviation, remains a significant diagnostic challenge [5]. In this case, the difficulty was compounded by baseline laboratory investigations that were largely within normal limits, including stable renal function and liver enzymes

(Table 1). While the mild anaemia (haemoglobin 11.46 g/dL) and slightly low total protein (64.82 g/L) identified in Table 1 are common in the third trimester due to haemodilution, these factors can significantly reduce a patient's physiological reserve when superimposed on structural cardiac disease. Specifically, the combination of hypervolemia and decreased oncotic pressure from low albumin levels increases susceptibility to pulmonary oedema [5]. However, persistent electrical changes, such as the left ventricular hypertrophy and strain pattern observed on this patient's ECG (Fig. 8), should serve as red flags. These findings should immediately alert the clinician to an underlying structural pathology, necessitating urgent echocardiographic assessment regardless of how stable the routine laboratory markers may appear [4, 5].

In patients with Group 1 PAH, fixed pulmonary resistance prevents adaptation to these changes. As the left-to-right shunt persists, the right ventricle experiences pressure overload, leading to heart failure [8]. S.N.'s rapid deterioration within 48 hours emphasises the narrow margin of clinical stability in mWHO Class IV patients near term or during intrapartum [4].

Multidisciplinary management

Optimal outcomes in high-risk CHD require a 'Pregnancy Heart Team' to provide specialised fetomaternal management and advanced cardiac monitoring, including continuous pulse oximetry, ECG and cardiotocography (CTG) [9, 10]. While vaginal delivery is preferred to reduce infection and haemorrhage, it requires early neuraxial analgesia to prevent labour-induced catecholamine surges that cause dangerous fluctuations in cardiac output [9, 11]. However, the use of oxytocin boluses must be avoided as they can cause rapid drops in SVR, potentially triggering a shunt reversal [9, 11].

In this case, despite epidural anaesthesia and induction of labour, non-reassuring fetal status, as evidenced by the fetal bradycardia and meconium-stained liquor, necessitated an emergency caesarean section. This aligns with findings in Kenya, where 37.0% of pregnant women with CHD were induced for labour [3] and in China, where 70.1% of pregnant women with PAH underwent Caesarean delivery [12].

Postpartum challenges and prognosis

The first 48 hours postpartum are crucial due to significant fluid shifts and autotransfusion from the contracting uterus, which can cause temporary volume overload [4, 5, 11]. S.N.'s postoperative respiratory infection stresses the vulnerability of the congested pulmonary system in patients with CHD complicated by PAH [4]. Therefore, such patients should be managed in tertiary health centres that offer extended intensive care monitoring to manage these hemodynamic shifts [4, 11]. While her survival represents a clinical success, her long-term prognosis remains cautious. The five-year survival for VSD is approximately 98.4% for those below 40 years of age, as in our case [8]. However, without definitive VSD repair, natural survivors face progressive pulmonary remodelling, which leads to complications such as PAH and even Eisenmenger syndrome [8, 11].

Ultimately, this case shows that positive outcomes can be achieved through multidisciplinary efforts. However, delayed diagnosis and late presentation during pregnancy, like the 53.1% rate in Ethiopia, mainly due to inadequate antenatal care contacts, remain significant barriers to improving maternal care in patients with CHD [13].

Conclusion

The successful pregnancy outcome of this primigravida with a large VSD and moderate PAH highlights the important role of a multidisciplinary "Pregnancy Heart Team." This case illustrates the clinical challenge of "natural survivors" in sub-Saharan Africa, where late diagnosis often delays presentation until the third trimester. While close monitoring resulted in favourable fetomaternal outcomes, the patient's rapid clinical deterioration emphasises the limitations of standard risk-stratification tools for cardiac disease in pregnancy in low-resource settings. Additionally, this case stresses the need for enhanced antenatal screening and specialised cardiology counselling in low-resource settings for patients with congenital heart disease. Also, increasing the number of cardiac surgical centres in sub-Saharan Africa and improving access to definitive surgical correction before conception will help in lowering maternal morbidity and mortality. For survivors like S.N., the focus should now shift to long-term cardiology and cardiothoracic follow-up to improve quality of life.

Declarations

Author Contributions: RNK: Conceptualisation, patient care, and drafting of the initial manuscript. PO, BAO, and DNK: Patient care, literature review, and revision of the manuscript. KOM: Consultant, fetomaternal medicine specialist, and led in patient care, supervision of the case report, and final review. We have all read and approved the final version of the manuscript for submission and publication.

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Data Availability: The data supporting the findings of this case report are included within the article.

Code Availability: Not applicable.

Ethical Considerations: We conducted this case report in accordance with the Declaration of Helsinki. Hospital ethical approval was waived for this case report because it involves standard clinical care and presents no risk to the patient.

Consent to Participate: Not applicable.

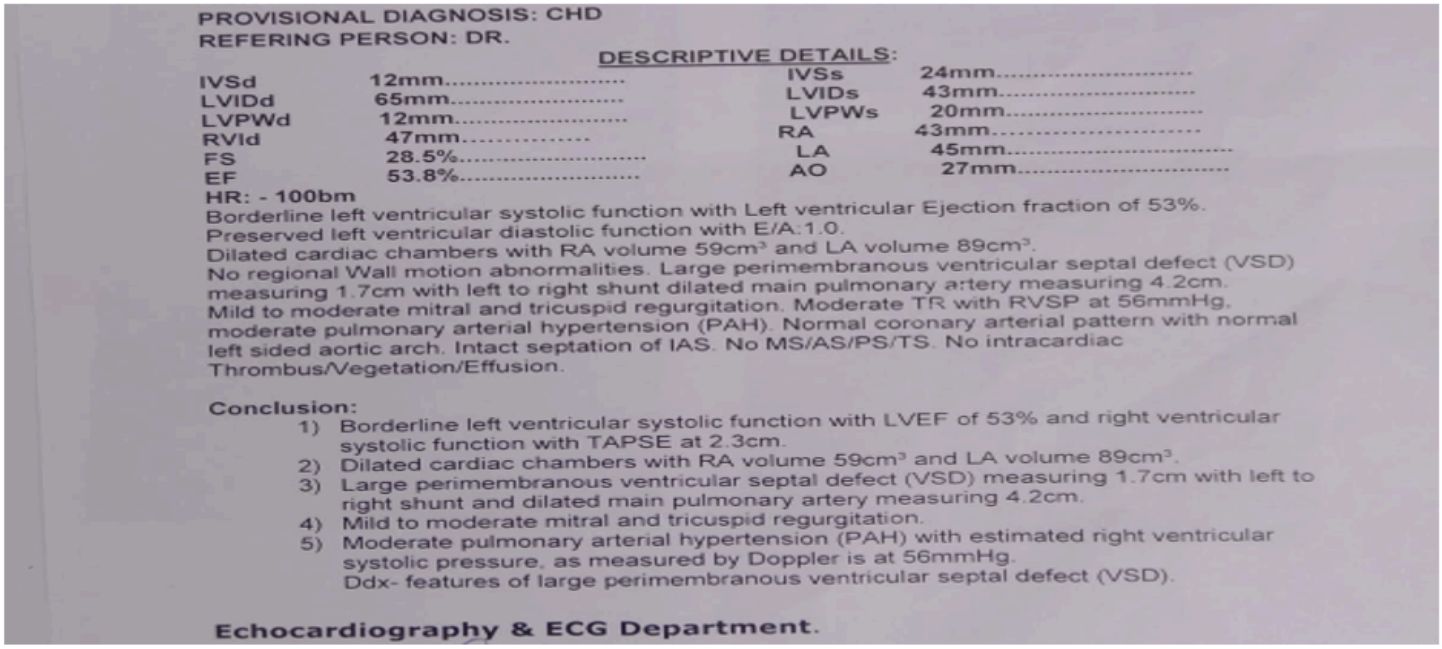
Consent for Publication: We obtained written informed consent from the patient for publishing this case report and the accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of Interests: We declare that we have no known competing financial interests.

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Figures



PROVISIONAL DIAGNOSIS: CHD
REFERING PERSON: DR.

DESCRIPTIVE DETAILS:

IVSd	12mm	IVSs	24mm
LVIDd	65mm	LVIDs	43mm
LVPWd	12mm	LVPWs	20mm
RVId	47mm	RA	43mm
FS	28.5%	LA	45mm
EF	53.8%	AO	27mm

HR: - 100bm
Borderline left ventricular systolic function with Left ventricular Ejection fraction of 53%.
Preserved left ventricular diastolic function with E/A:1.0.
Dilated cardiac chambers with RA volume 59cm³ and LA volume 89cm³.
No regional Wall motion abnormalities. Large perimembranous ventricular septal defect (VSD) measuring 1.7cm with left to right shunt dilated main pulmonary artery measuring 4.2cm.
Mild to moderate mitral and tricuspid regurgitation. Moderate TR with RVSP at 56mmHg, moderate pulmonary arterial hypertension (PAH). Normal coronary arterial pattern with normal left sided aortic arch. Intact septation of IAS. No MS/AS/PS/TS. No intracardiac Thrombus/Vegetation/Effusion.

Conclusion:

1) Borderline left ventricular systolic function with LVEF of 53% and right ventricular systolic function with TAPSE at 2.3cm.
2) Dilated cardiac chambers with RA volume 59cm³ and LA volume 89cm³.
3) Large perimembranous ventricular septal defect (VSD) measuring 1.7cm with left to right shunt and dilated main pulmonary artery measuring 4.2cm.
4) Mild to moderate mitral and tricuspid regurgitation.
5) Moderate pulmonary arterial hypertension (PAH) with estimated right ventricular systolic pressure, as measured by Doppler is at 56mmHg.
   Ddx- features of large perimembranous ventricular septal defect (VSD).

Echocardiography & ECG Department.

Figure 1

Echocardiography reporting results highlighting a perimembranous ventricular septal defect associated with pulmonary arterial hypertension

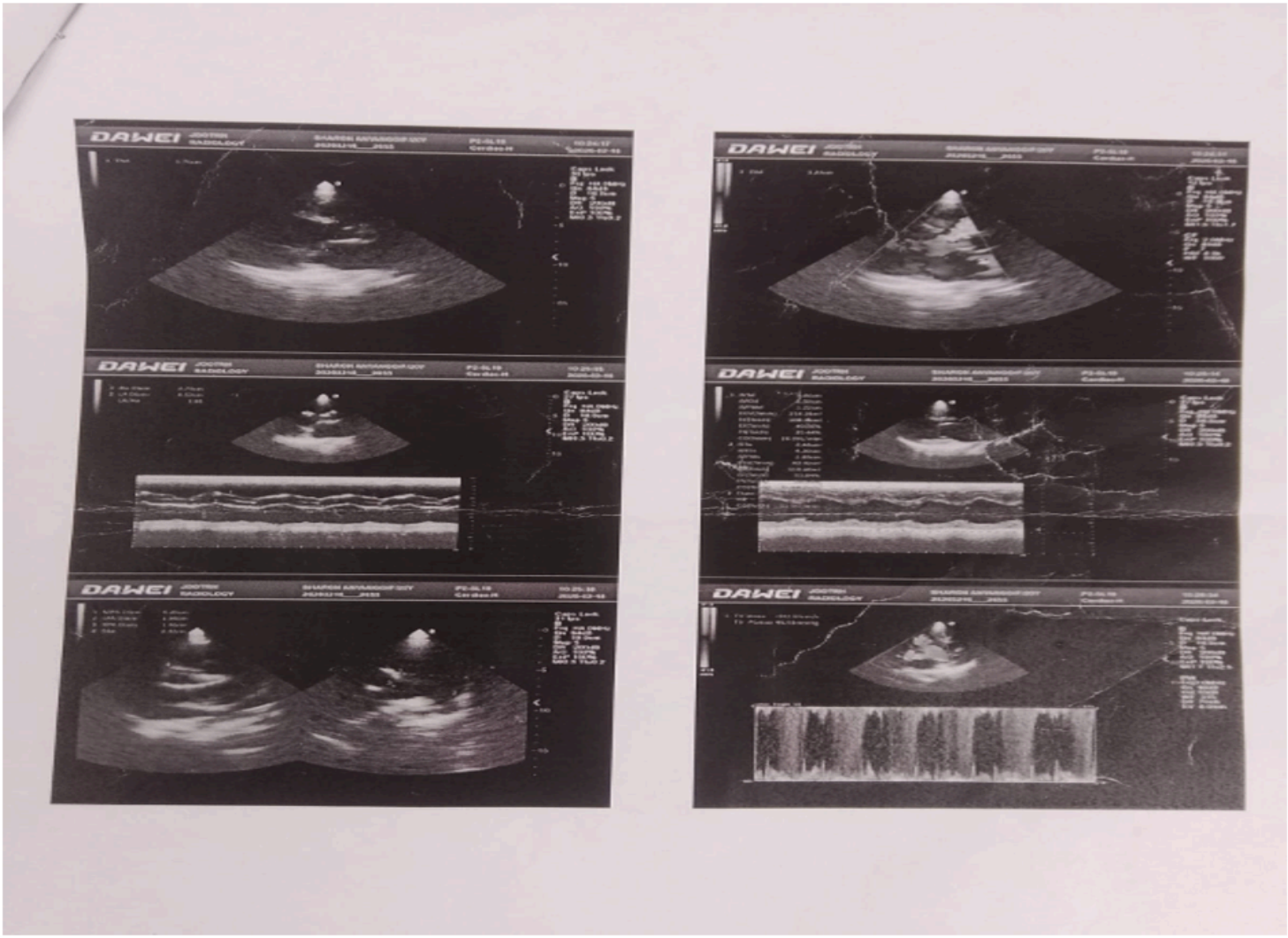


Figure 2

Echocardiogram images illustrating a perimembranous ventricular septal defect associated with pulmonary arterial hypertension

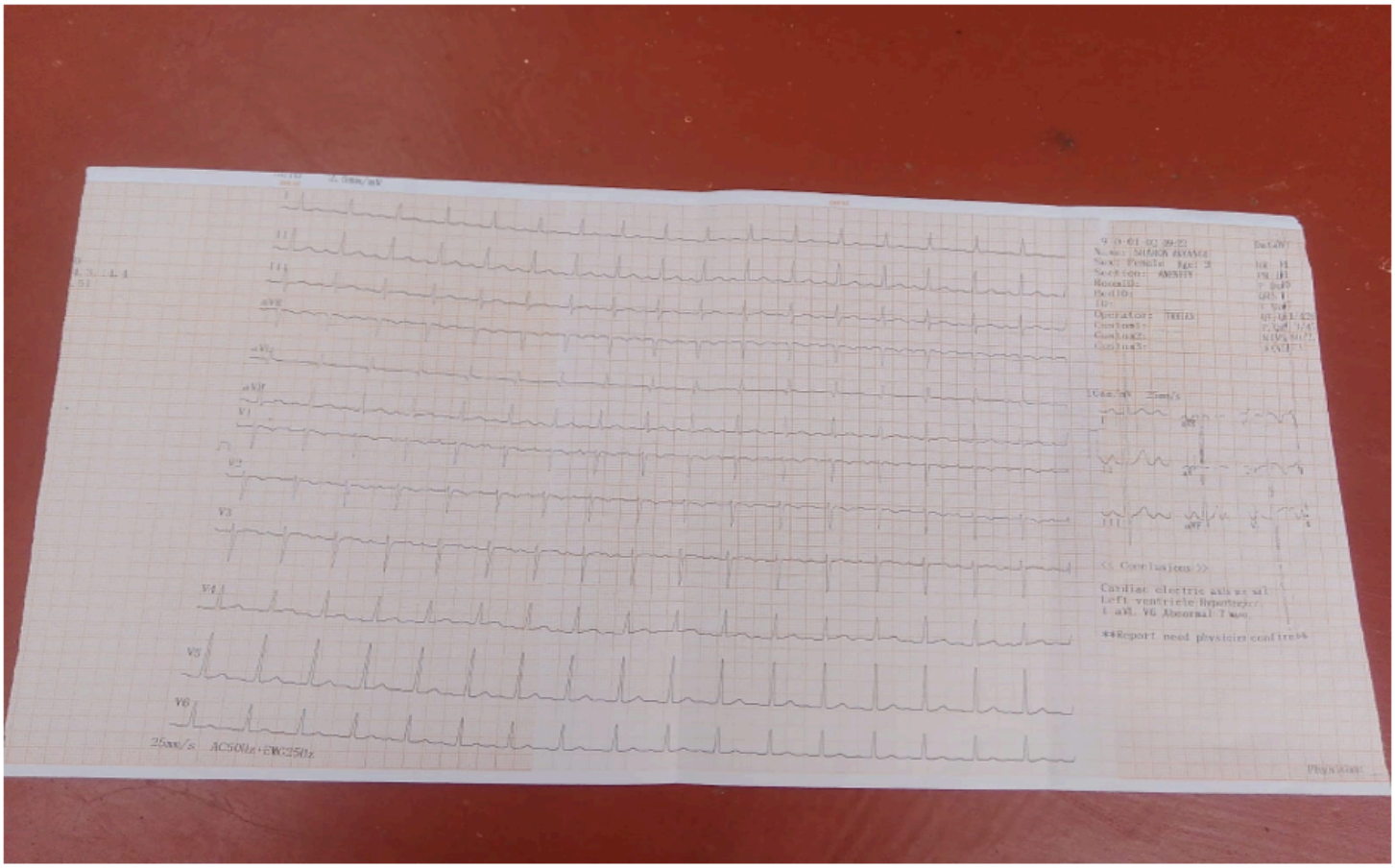


Figure 3

Electrocardiography image showing left ventricular hypertrophy