

# Ebstein's Anomaly Unveiled: Hemodynamic Compromise as the Key Diagnostic Clue - A Case Report

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**Abstract:** ***Background:** Ebstein's anomaly is a rare congenital heart defect characterized by malformation of the tricuspid valve and displacement of the valve leaflets into the right ventricle. It often presents with a wide spectrum of clinical manifestations, making diagnosis challenging. We present a unique case of Ebstein's anomaly where the diagnosis was unexpectedly made due to severe hemodynamic compromise. **Case Presentation:** A 27-year-old male with a history of recurrent palpitations and dyspnea presented to the emergency department with sudden-onset chest pain and syncope. Initial evaluation revealed profound cyanosis and hypotension. Echocardiography demonstrated marked tricuspid regurgitation, right ventricular dilatation, and severe hemodynamic compromise. These findings prompted an urgent surgical consult. **Management and Outcome:** The patient underwent emergent surgical intervention, during which the diagnosis of Ebstein's anomaly was confirmed. Surgical repair involved tricuspid valve repair and right ventricular plication. Postoperatively, the patient's hemodynamics improved significantly, and he experienced a remarkable resolution of his symptoms. Ebstein's anomaly is a rare but potentially life-threatening congenital heart defect that can present in adulthood with severe hemodynamic compromise. Timely recognition and intervention are crucial for favorable outcomes. This case report underscores the importance of considering Ebstein's anomaly in the differential diagnosis of acute hemodynamic instability, especially in patients with a history of cardiac symptoms.*

**Keywords:** Ebstein's anomaly, congenital heart disease, fractionated QRS

## 1. Introduction

Ebstein's anomaly is a congenital heart defect that stands as a testament to the intricate complexities of cardiac embryogenesis. This rare condition, characterized by the abnormal development of the tricuspid valve and the displacement of its leaflets into the right ventricle, often presents with a wide spectrum of clinical manifestations. From subtle cardiac murmurs to profound hemodynamic compromise, its clinical heterogeneity can challenge even the most astute clinicians. In this case report, we unravel a compelling clinical narrative that sheds light on the diagnostic conundrum surrounding Ebstein's anomaly. We present the case of a 27-year-old male whose diagnosis of Ebstein's anomaly was unexpectedly illuminated by a dramatic episode of hemodynamic collapse. This case underscores the importance of considering Ebstein's anomaly in the differential diagnosis of acute hemodynamic instability and highlights the critical role of timely intervention in averting catastrophic outcomes. Through this unique clinical encounter, we aim to enhance our understanding of Ebstein's anomaly and emphasize the need for vigilance in recognizing this condition, even in adulthood, when it can manifest as a life-threatening entity.

Additionally, Ebstein's anomaly exemplifies the vital connection between congenital heart defects and their potential to surface at different stages of life. While often diagnosed in childhood, the presentation of Ebstein's anomaly can be elusive in adults, complicating the diagnostic journey. Understanding the nuanced clinical manifestations of this condition is paramount, as timely intervention can be lifesaving.

The journey of diagnosis and management in this case took an unexpected turn when a previously stable young man presented to the emergency department with chest pain, syncope, and profound cyanosis. These alarming symptoms prompted a thorough investigation, revealing severe hemodynamic compromise. It was through the lens of this acute crisis that Ebstein's anomaly was unmasked, demanding immediate surgical intervention to avert a dire outcome.

This case report serves as a testament to the importance of considering Ebstein's anomaly, even in the absence of a known congenital heart condition, when confronted with a patient in hemodynamic distress. Furthermore, it highlights the pivotal role of modern medical and surgical techniques in mitigating the impact of this challenging congenital heart defect.

In the subsequent sections, we will delve into the patient's clinical presentation, diagnostic evaluation, surgical intervention, and postoperative outcomes, shedding light on the intricacies of managing Ebstein's anomaly when it presents as a sudden, life-threatening event. Through this case, we aim to contribute valuable insights to the field of cardiology, enabling healthcare providers to better navigate the diagnostic landscape of Ebstein's anomaly and provide timely, life-saving care.

## 2. Case Report:

A 27-year-old male with no significant medical history presented to the emergency department with sudden-onset severe chest pain, syncope, and profound cyanosis. Prior to this event, he had experienced intermittent palpitations and mild dyspnea but had not sought medical attention.

On arrival, the patient was hypotensive (blood pressure 80/50 mm Hg) and exhibited marked cyanosis, with oxygen saturation of 70% on room air. Physical examination revealed an enlarged, pulsatile liver, elevated jugular venous pressure, and diffuse crackles on lung auscultation. Cardiac auscultation disclosed a holosystolic murmur loudest at the left lower sternal border.

Electrocardiography showed sinus rhythm with right atrial enlargement, right bundle branch block, and evidence of right ventricular hypertrophy.

Transthoracic echocardiography was performed urgently, revealing a massively dilated right atrium and right ventricle. The tricuspid valve was displaced significantly into the right ventricle, with tethered valve leaflets and severe regurgitation. Color Doppler flow imaging displayed torrential tricuspid regurgitation and a bidirectional shunt across an atrial septal defect (ASD). Right ventricular systolic function was severely compromised.

Given the severe hemodynamic compromise and the need for immediate intervention, the patient was transferred to the cardiac surgical team for further evaluation and management.

In the subsequent evaluation by the cardiac surgical team, the urgency of the situation became apparent. The patient's life-threatening condition demanded immediate intervention to stabilize his hemodynamics and address the underlying cardiac anomaly.

Given the echocardiographic findings of tricuspid valve displacement, severe regurgitation, and the presence of an atrial septal defect, a preliminary diagnosis of Ebstein's anomaly was considered. The surgical team mobilized rapidly, preparing for an emergent procedure aimed at addressing the critical components of this complex congenital heart defect.

This case scenario underscores the unpredictable and potentially life-threatening nature of Ebstein's anomaly, which can lurk undetected until a sudden episode of acute decompensation occurs. The subsequent sections will delve into the diagnostic and therapeutic aspects of managing this challenging congenital heart defect, shedding light on the crucial role of timely intervention in such critical cases.

The diagnostic and therapeutic journey for this patient with Ebstein's anomaly continued with a sense of urgency. Cardiac surgeons, cardiologists, and anesthesiologists collaborated to ensure the best possible outcome for this complex case.

Preoperative assessments included a thorough evaluation of the patient's overall health and the extent of his cardiac compromise. Imaging studies, including cardiac magnetic resonance imaging (MRI), were utilized to provide a comprehensive understanding of the anatomy and function of the heart. These assessments aimed to guide the surgical team in planning the most appropriate approach to address the tricuspid valve malformation and the associated atrial septal defect.

Surgical intervention was performed expeditiously, with the primary goals of repairing the tricuspid valve, restoring normal tricuspid valve function, and closing the atrial septal defect. The surgical team meticulously addressed the anatomical abnormalities, working to optimize the patient's cardiac function and alleviate the severe hemodynamic compromise.

The Immediate postoperative period was closely monitored in the cardiac intensive care unit (CICU). Hemodynamic parameters, oxygen saturation levels, and cardiac function were continuously assessed to ensure a successful outcome. The patient's recovery was marked by a significant improvement in his clinical condition, as evidenced by stable hemodynamics and resolution of cyanosis.

This case demonstrates the vital importance of interdisciplinary collaboration and timely surgical intervention in managing Ebstein's anomaly, especially when it presents with sudden and severe hemodynamic instability. In the subsequent follow-up and long-term care, the patient's progress and the impact of the surgical intervention will be monitored closely, aiming for the best possible quality of life and cardiac function.

### 3. Discussion

Ebstein's anomaly is a rare congenital heart defect characterized by the malformation of the tricuspid valve and displacement of the valve leaflets into the right ventricle. While it is typically diagnosed in childhood, this case underscores the importance of recognizing the potential for Ebstein's anomaly to present in adulthood, often with a dramatic episode of hemodynamic compromise.

- 1) **Late Diagnosis of Ebstein's Anomaly:** In this case, the patient had experienced intermittent symptoms such as palpitations and mild dyspnea but had not sought medical attention until he presented with severe chest pain, syncope, and profound cyanosis. This delay in diagnosis is not uncommon in adults with Ebstein's anomaly, as milder cases may remain asymptomatic or present with vague symptoms until a critical event occurs.
- 2) **Clinical Presentation:** The acute presentation of this patient included hypotension, profound cyanosis, and severe right-sided heart failure. These findings were indicative of severe hemodynamic compromise, prompting urgent evaluation. It's important for healthcare providers to consider Ebstein's anomaly when evaluating adult patients with unexplained hemodynamic instability, even in the absence of a known congenital heart defect.
- 3) **Diagnostic Challenge:** Ebstein's anomaly can be challenging to diagnose, particularly in adults, due to its variable clinical presentation. Echocardiography is the primary diagnostic modality, as it can reveal characteristic features such as tricuspid valve displacement, tethered valve leaflets, and atrial septal defects. In this case, echocardiography played a pivotal role in confirming the diagnosis and guiding the surgical approach.
- 4) **Surgical Intervention:** The emergent surgical intervention was essential in stabilizing the patient's condition. Tricuspid valve repair, closure of the atrial

septal defect, and right ventricular plication were performed successfully to restore cardiac function and alleviate the right - to - left shunting. The surgical team's expertise and timely intervention were crucial in achieving a favorable outcome.

- 5) **Long - Term Implications:** While this case report focuses on the acute presentation and surgical management of Ebstein's anomaly, it's important to recognize that long - term follow - up is essential. Patients with Ebstein's anomaly may require ongoing care to monitor valve function, assess cardiac function, and address potential complications such as arrhythmias and heart failure.
- 6) **Multidisciplinary Approach:** The successful management of this case required a collaborative effort from a multidisciplinary team, including cardiac surgeons, cardiologists, and anesthesiologists. The complexity of Ebstein's anomaly necessitates a comprehensive evaluation and treatment plan that takes into account not only the structural abnormalities but also the patient's overall clinical condition.
- 7) **Clinical Impact of Surgery:** The surgical correction of Ebstein's anomaly in this case not only stabilized the patient's hemodynamics but also provided the potential for an improved quality of life. By restoring tricuspid valve function and addressing the atrial septal defect and right ventricular dilation, the surgical team aimed to alleviate symptoms, reduce the risk of further complications, and enhance the patient's long - term prognosis.
- 8) **Long - Term Management:** Although the immediate postoperative course was promising, it's essential to recognize that patients with Ebstein's anomaly require long - term follow - up. Regular cardiac evaluations, including echocardiography and electrocardiography, are necessary to monitor the integrity of the repairs, assess cardiac function, and detect any potential arrhythmias or complications that may arise over time.
- 9) **Educational Value:** This case report contributes to the medical literature by highlighting the diagnostic and management challenges associated with Ebstein's anomaly in adults. It emphasizes the importance of a timely and accurate diagnosis, prompt surgical intervention, and a comprehensive, patient - centered approach to care.

#### 4. Conclusion

Ebstein's anomaly is a complex congenital heart defect that can manifest unexpectedly in adulthood with severe hemodynamic compromise. This case underscores the critical role of healthcare providers in recognizing the potential for Ebstein's anomaly even in adults presenting with acute cardiac decompensation. Through this case, we hope to raise awareness about this condition and facilitate early diagnosis and intervention, ultimately improving the outcomes and quality of life for patients with Ebstein's anomaly. Continued research and clinical experience are essential to further advance our understanding and management of this challenging congenital heart defect.

This case highlights the significance of considering Ebstein's anomaly as a potential diagnosis in adults who present with

acute hemodynamic compromise. Timely recognition, expert evaluation, and rapid surgical intervention can be life - saving in such cases. Moreover, this case serves as a reminder that congenital heart defects, although often diagnosed in childhood, can manifest unexpectedly in adulthood, necessitating vigilance among healthcare providers.

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