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Case Report

Ebstein Anomaly in a Term Neonate Complicated by *Burkholderia cepacia* Culture-Proven and Multiorgan Failure: A Rare Case Report of Late-Onset Sepsis

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ABSTRACT

Ebstein anomaly is a rare congenital malformation of the tricuspid valve that can present in the neonatal period with severe cyanosis, heart failure, and cardiopulmonary instability. Neonates with complex congenital heart disease requiring prolonged intensive care are particularly vulnerable to hospital-acquired infections, which may significantly worsen clinical outcomes. We report the case of a term neonate with antenatally suspected Ebstein anomaly who developed early cardiac decompensation soon after birth, requiring intensive care management and respiratory support. During the neonatal intensive care unit stay, the infant developed culture-proven multidrug-resistant *Burkholderia cepacia* sepsis associated with multiorgan dysfunction, including hepatic impairment, coagulopathy, and neurological involvement. The patient required ventilatory support, inotropic therapy, anticonvulsants, and targeted antimicrobial treatment. Following prompt multidisciplinary management and intensive supportive care, the infant gradually improved and was discharged in stable condition. This case highlights the increased susceptibility of neonates with congenital heart disease to severe nosocomial infections and emphasises the importance of early recognition, aggressive supportive therapy, and strict infection control measures to improve outcomes.

Keywords: *Burkholderia cepacia*, Ebstein abnormality, Late-onset sepsis, Multiorgan failure, Neonatal, Congenital heart disease, Ventricular septal defect.

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INTRODUCTION

An atrialization of a portion of the right ventricle and variable degrees of tricuspid regurgitation result from the Ebstein anomaly, a rare congenital disorder of the tricuspid valve, which is characterised by the downward displacement of the septal and posterior leaflets into the right ventricle (Attenhofer *et al.*, 2007). With an estimated incidence of roughly 1 in 200,000 live births, it accounts for less than 1% of all congenital cardiac disorders (Celermajer *et al.*, 1994). Depending on the degree of tricuspid regurgitation, the degree of valve displacement, right ventricular function, and related cardiac abnormalities like atrial or Ventricular Septal Defects (VSD), the clinical appearance might vary significantly (Stulak *et al.*, 2007). The illness is frequently severe in infants and can show up as pulmonary hypertension, heart failure, cyanosis, and an enlarged heart.

Neonates may require mechanical ventilation and inotropic assistance in the most severe cases due to considerable cardiopulmonary impairment (Kumar, 2021). Further deteriorating the prognosis and raising morbidity during the newborn period is the occurrence of extra lesions like pulmonary hypertension or VSD (Stoll *et al.*, 2011). Intensive care unit admissions of newborns with congenital heart disease are especially vulnerable to late-onset sepsis because of the length of stay, invasive procedures, central lines, and mechanical ventilation (Shane *et al.*, 2017). Globally, late-onset sepsis continues to be a major cause of morbidity and death in NICU settings (Mahenthiralingam *et al.*, 2005). Increasingly, gram-negative, non-fermenting *Burkholderia cepacia* is being identified as an opportunistic hospital-acquired infection. Multiple antibiotic resistance is inherent, and it has been connected to epidemics in neonatal intensive care units, especially in critically unwell and ventilated newborns (Sousa *et al.*, 2011; Dizbay *et al.*, 2009). Although it more commonly affects immunocompromised

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hosts, severe infections have also been reported in neonates with underlying health issues (Dizbay *et al.*, 2009).

In addition to being difficult to cure, severe congenital heart defects and multidrug-resistant hospital-acquired infections raise the chance of multiorgan failure. Ebstein anomaly, which is made worse by culture-confirmed *Burkholderia cepacia* late-onset sepsis, is extremely rare in newborns. We report the successful multimodal intensive care therapy of a full-term newborn with Ebstein anomaly who experienced acute cardiac distress, multiorgan failure, and culture-confirmed late-onset sepsis caused by *Burkholderia cepacia*.

CASE DESCRIPTION

A vaginal birth of a full-term male baby followed a simple labour at an outside facility. The baby started crying as soon as he was born. Prenatal examinations had raised the possibility of congenital heart disease, and postnatal echocardiography verified the Ebstein abnormality. To assess the baby, a 2D echocardiography was first carried out at a pediatric cardiac centre. After being discharged, he was given instructions for follow-up. The infant experienced respiratory distress, poor circulation, and congestive heart failure symptoms on the seventh day of life. He was admitted to the neonatal critical care unit and transferred to a more advanced facility for further care. There was no record of birth trauma, perinatal hypoxia, or maternal fever.

The labour proceeded without any complications, and there was no significant family history of congenital heart disease or genetic issues. A blood culture that identified *Burkholderia cepacia* confirmed the infant's late-onset sepsis during the NICU stay. Severe liver dysfunction suggestive of neonatal cholestasis, anaemia, coagulopathy, thrombocytopenia, respiratory failure, convulsions, and septic shock all made the clinical course worse. The infant had a scheduled cardiology follow-up, was on heart medication, was able to tolerate feedings, and was hemodynamically stable at discharge.

Clinical Findings

On the seventh day of life, the full-term male infant exhibited severe respiratory distress, poor eating, and poor circulation all signs of congestive heart failure. Lethargy, chest retractions, and rapid breathing were all signs of the baby's illness. Support in the form of inotropics was necessary due to the evident hemodynamic instability. With a markedly enlarged right atrium, mild to moderate mitral regurgitation, moderate pulmonary hypertension, a moderate-to-large muscular ventricular septal defect with preserved biventricular function, and a downward displacement of the tricuspid valve (Table 1).

The infant developed acute respiratory failure during hospitalisation, and high-frequency oscillatory ventilation was initiated due to increasing oxygen requirement and carbon dioxide retention (Figure 1). After that, there was a gradual switch to room air and then nasal oxygen. Levetiracetam and phenobarbitone were required to treat the infant's neurological seizures. Although the initial electroencephalogram suggested encephalopathy, a follow-up EEG before discharge showed no encephalopathy.

The clinical result was further worsened by thrombocytopenia, coagulopathy, transfusion-worthy anemia, and abnormal liver function tests with conjugated hyperbilirubinemia suggestive of neonatal cholestasis. Due to evidence of stomach distension throughout the hospital stay, feedings had to be temporarily stopped before being gradually restarted. The baby was able to handle full oral feedings, was hemodynamically stable upon discharge, and maintained oxygen saturation in room air.

Follow-up and Outcomes

Following extensive multidisciplinary management, the infant showed a consistent clinical recovery. As the baby's respiratory state improved, high-frequency oscillatory ventilation was replaced with normal ventilation, extubation to nasal oxygen was performed, and eventually, adequate oxygen levels in room air were maintained. Inotropic assistance helped achieve hemodynamic stability and was progressively decreased. The use of pulmonary vasodilators and diuretics reduced the symptoms of congestive heart failure. Levetiracetam and phenobarbitone were used to treat the seizures, and no more incidents happened.

In advance of discharge, a follow-up electroencephalogram revealed normal results. Blood component therapy helped to resolve blood-related problems such as anaemia, coagulopathy, and thrombocytopenia. Tests of liver function revealed improvement when neonatal cholestasis patients received supportive treatment. The infant responded well to the gradual reintroduction of enteral feeding, which was followed by full oral feeding. The infant was hemodynamically stable, seizure-free, able to tolerate complete meals, and sustaining oxygen saturation in room air at the time of discharge. For continued therapy of the ventricular septal defect and Ebstein abnormality, close monitoring with a pediatric cardiologist was advised (Figure 2).

DISCUSSION

Ebstein anomaly is a congenital heart defect characterized by downward displacement of the tricuspid valve leaflets. This results in atrialization of the right ventricle and varying degrees of tricuspid regurgitation (Attenhofer Jost *et al.*, 2007). With a clinical presentation that varies greatly, from asymptomatic persons to critically ill babies, it represents less than 1% of congenital cardiac abnormalities (Celermajer *et al.*, 1994; Stulak *et al.*, 2007). Because of the condition's substantial tricuspid regurgitation, decreased effective pulmonary blood flow, enlarged heart, and right atrial enlargement, babies frequently exhibit

Table 1: Clinical Findings.

System	Clinical Findings
General	Ill-looking neonate, lethargy during septic phase
Respiratory	Severe respiratory distress, tachypnea, chest retractions, required intubation and HFOV
Cardiovascular	Ebstein anomaly, severe TR, mild–moderate MR, moderate PPHN, grossly dilated RA, moderate–large VSD, cardiomegaly
Neurological	Seizures, EEG suggestive of encephalopathy (initially), improved on treatment
Hematological	Thrombocytopenia, coagulopathy, anemia requiring PRBC, FFP, platelet transfusions
Hepatic	Elevated transaminases, conjugated hyperbilirubinemia, neonatal cholestasis
Gastrointestinal	Abdominal distension, temporary NPO, gradual feed advancement
Outcome at Discharge	Hemodynamically stable, room air saturation maintained, tolerating feeds



Figure 1: Term neonate with Ebstein anomaly requiring mechanical ventilation and intensive monitoring during NICU stay.



Figure 2: Clinical appearance of the neonate at follow-up after stabilization and recovery.

severe hemodynamic instability (Stulak *et al.*, 2007; Warnes *et al.*, 2008).

Clinical results are further deteriorated in severe cases by pulmonary hypertension and related intracardiac abnormalities, such as a ventricular septal defect (Celermajer *et al.*, 1994; Stoll *et al.*, 2011). The infant had a moderate-to-large ventricular septal defect, right atrial enlargement, and pulmonary hypertension. Severe tricuspid regurgitation further worsened respiratory distress and early heart failure, and early congestive heart failure. It might be difficult to treat neonates with Ebstein's anomaly who exhibit symptoms. Surgical intervention is reserved for patients who do not respond to medical therapy. Severe cases require aggressive stabilization, including ventilatory support, inotropes, and pulmonary vasodilators. (Kumar, 2021; Stoll *et al.*, 2011). In this instance, stabilising the cardiopulmonary state during the acute phase required early mechanical ventilation and inotropic support.

In intensive care units, invasive procedures, central venous access, and mechanical ventilation enhance the risk of late-onset sepsis in infants with complex congenital heart disease (Shane *et al.*, 2017; Mahenthiralingam *et al.*, 2005). Globally, nosocomial infections remain a major cause of morbidity and mortality in neonatal intensive care units (Mahenthiralingam *et al.*, 2005). The infant's development of culture-proven late-onset sepsis during the NICU stay in this instance further complicated the clinical outcome. *Burkholderia cepacia* is a gram-negative, non-fermenting bacillus that is becoming increasingly recognized critically ill as an opportunistic nosocomial pathogen (Sousa *et al.*, 2011). Due to its inherent resistance to several antimicrobial drugs, it has been linked to epidemics in neonatal intensive care units, especially in critically ill and ventilated newborns (Sousa *et al.*, 2011; Dizbay *et al.*, 2009). Severe infections in infants with underlying comorbid problems have been reported, even though they are more frequently linked to immunocompromised hosts (Dizbay *et al.*, 2009). Our patient's congenital cardiac condition, prolonged ventilation, and invasive monitoring likely increased their risk of contracting *B. cepacia*. This case presented a significant clinical challenge due to the dual diagnoses of Ebstein's abnormality and multidrug-resistant late-onset sepsis. Septic shock combined with a systemic inflammatory response might worsen heart failure, raise mortality risk, and involve several organs in neonates who already have congenital heart defects (Shane *et al.*, 2017; Mahenthiralingam *et al.*, 2005). Blood abnormalities, coagulopathy, neurological symptoms, and liver dysfunction were all linked to sepsis in our case, and these factors were all consistent with multiorgan dysfunction syndrome.

A multidisciplinary approach, intensive supportive care, focused antibiotic treatment, and early detection were necessary for a favorable outcome. This example highlights the need for stringent infection control procedures in neonatal intensive care units, particularly for infants with severe congenital cardiac defects. Ebstein anomaly combined with culture-confirmed *Burkholderia cepacia* late-onset sepsis in full-term newborns is extremely rare. This case demonstrates the vulnerability of infants with congenital heart disease to severe hospital-acquired infections. It also highlights the importance of early diagnosis and coordinated management to improve survival.

Recent reports have further emphasised the clinical variability and management challenges associated with Ebstein anomaly. Dessalegne *et al.* described a case in a young female, highlighting

that disease severity depends on anatomical variation and functional impairment rather than age alone (Dessalegne *et al.*, 2024). Additionally, recent studies on neonatal intensive care unit infections have identified *Burkholderia cepacia* as an emerging multidrug-resistant pathogen responsible for outbreaks, often associated with contaminated medical equipment and prolonged hospitalisation (Bharara *et al.*, 2020). These findings reinforce the importance of strict infection control practices and early targeted therapy in vulnerable populations such as infants with congenital heart disease.

CONCLUSION

The complex connection between structural congenital heart disease and severe hospital-acquired infections during the neonatal era is demonstrated by the current instance. When culture-confirmed *Burkholderia cepacia* hospital-acquired infection is combined with Ebstein abnormalities, which are characterised by severe tricuspid regurgitation and pulmonary hypertension, it can lead to multiorgan dysfunction and increased mortality risk. Early diagnosis of cardiac decompensation, cautious hemodynamic support, multimodal intensive care, and timely introduction of targeted antibacterial medication all contributed to the successful survival of this seriously unwell full-term newborn. To enhance survival in these high-risk groups, our study emphasises the need for coordinated cardiac and critical care therapy as well as the need for greater monitoring of opportunistic infections in neonates with congenital heart disease.

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ABBREVIATIONS

2D: Two-Dimensional; ***B. cepacia*:** *Burkholderia cepacia*; **CHF:** Congestive Heart Failure; **CHD:** Congenital Heart Disease; **CLD:** Chronic Liver Disease; **EEG:** Electroencephalogram; **HFOV:** High-Frequency Oscillatory Ventilation; **ICU:** Intensive Care Unit; **LOS:** Late-Onset Sepsis; **MR:** Mitral Regurgitation; **NICU:** Neonatal Intensive Care Unit; **PH:** Pulmonary Hypertension; **TR:** Tricuspid Regurgitation; **VSD:** Ventricular Septal Defect; **QoL:** Quality of Life; **NICU:** Neonatal Intensive Care Unit; **MODS:** Multiorgan Dysfunction Syndrome; **LFT:** Liver Function Test; **SpO_2:** Peripheral Oxygen Saturation.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

PATIENT PERSPECTIVE

The parents expressed relief and gratitude for the multidisciplinary care provided. They acknowledged the prolonged NICU stay as emotionally challenging but were reassured by the infant's gradual recovery and stable discharge condition.

INFORMED CONSENT

Informed consent was obtained from the patient's parents for publication of this case report and accompanying clinical information. Identifying information has been anonymised.

ETHICAL APPROVAL

Approval for publication of this case report was obtained from the treating consultant, Dr Rama Rao, a General Surgeon at Aster Ramesh Hospital, Andhra Pradesh. The manuscript contains no identifiable patient information.

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