Case Report

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Anaesthesia Section

Anaesthetic Management of a Neonate with Ebstein's Anomaly Undergoing a Meningomyelocele Surgery: A Case Report

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ABSTRACT

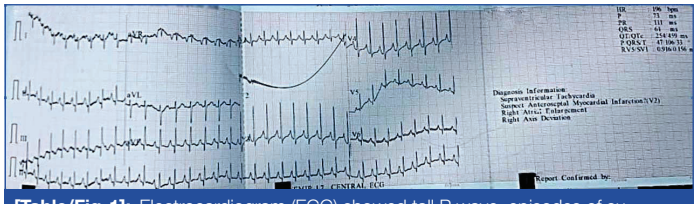
Ebstein's Anomaly (EA) is a congenital heart defect characterised by the downward displacement of the posterior and septal leaflets of the tricuspid valve toward the right ventricular apex. Hereby the authors discuss a 16-day-old female neonate with EA who underwent surgery for a ruptured meningocele. The baby, born at full term, had an Appearance, Pulse, Grimace, Activity and Respiration (APGAR) score of 7 at one minute and 9 at five minutes. Upon further observation, she showed multiple episodes of breath-holding and cyanosis. On examination, an enlarged head and swelling on the lumbar spine led to an ultrasound that confirmed hydrocephalus and meningocele. An Electrocardiogram (ECG) showed supraventricular tachycardia, and a chest X-ray revealed cardiomegaly. The diagnosis of EA was confirmed by two-dimensional (2D) echocardiography. General anaesthesia with inhalational induction was planned. Perioperative considerations for this patient included right-to-left shunting with the possibility of paradoxical emboli, prolonged onset time for intravenous drugs due to pooling and recirculation of blood in the right atrium, increased Pulmonary Vascular Resistance (PVR), perioperative pulmonary hypertension, right and left ventricular dysfunction, and arrhythmias. Key cardiovascular goals for this patient included maintaining normal sinus rhythm, ensuring adequate preload while avoiding significant changes in Systemic Vascular Resistance (SVR) or PVR, avoiding medications and factors that depress myocardial function, and promptly identifying haemodynamic instability or inadequate tissue perfusion with appropriate corrections. The surgery for excision and repair of the meningocele was uneventful, and the neonate was sent to the Neonatal Intensive Care Unit (NICU) for further monitoring.

Keywords: Cardiomegaly, Meningocele, Paediatric anaesthesia, Tricuspid regurgitation

CASE REPORT

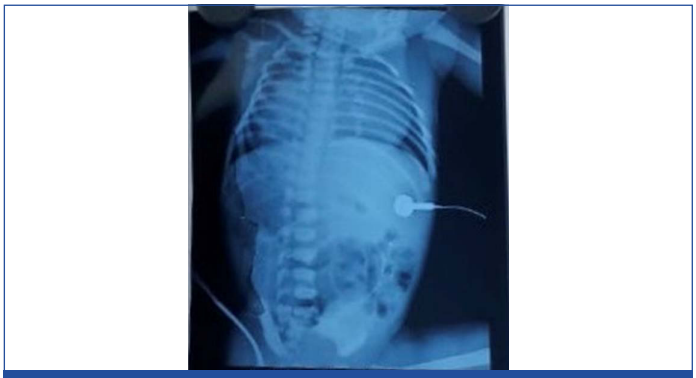
This is a case of 16-day-old female neonate with Ebstein's Anomaly (EA) and hydrocephalus, planned for the repair of a ruptured meningocele. The patient was a Dichorionic Diamniotic (DCDA) twin, delivered by emergency Lower Segment Caesarean Section (LSCS) due to Premature Rupture of Membranes (PROM). The patient was full term, appropriate for gestational age, and cried immediately after birth, with an APGAR score of 7 at one minute and 9 at five minutes after birth.

The patient experienced multiple episodes of breath-holding and cyanosis and was admitted to the NICU for further evaluation. On examination, the baby was active, moving all limbs and accepting feeds, with multiple episodes of breath-holding and cyanosis. The weight was 2.6 kg. The patient had a heart rate of 152 beats per minute, with a saturation of 86% in all four limbs on room air, cyanosis during crying and a pansystolic murmur. Her ECG showed tall P waves, episodes of supraventricular tachycardia, and right axis deviation [Table/Fig-1]. Her chest X-ray revealed cardiomegaly with Right Ventricular (RV) and right atrial enlargement [Table/Fig-2].



[Table/Fig-1]: Electrocardiogram (ECG) showed tall P wave, episodes of supraventricular tachycardia and right axis deviation.

Her 2D echocardiogram revealed Ebstein's anomaly of the tricuspid valve, with a 1.4 cm apical displacement of the valve, severe Tricuspid Regurgitation (TR), and a small patent ductus arteriosus.



[Table/Fig-2]: Chest X-ray revealed cardiomegaly with Right Ventricle (RV) and right atrial enlargement.

The Right Ventricle (RV) systolic pressure was 16 mmHg plus the right atrial pressure, with no pulmonary valve stenosis. The patient was given an injection of alprostadil for three days after birth to maintain the patency of the ductus arteriosus.

An ultrasonography scan was performed for the skull, whole abdomen, Kidneys, Ureter, Bladder (KUB), and local site. It revealed dilatation of the lateral and third ventricles, suggestive of hydrocephalus, and a cystic lesion measuring 3×2.7 cm in the lumbosacral region, with a few echogenic areas, without herniation of spinal components, suggestive of meningocele. Investigations showed a Haemoglobin (Hb) level of 24.3 g/dL, with normal blood counts, normal renal function, blood sugar, coagulation profile and serum bilirubin. A blood gas sample showed pH=7.414, Partial Pressure of Oxygen (pO_2)=40.7 mmHg, Partial Pressure of Carbondioxide (pCO_2)=30.1 mmHg, Base Excess (BE)=-5.3 mmol/L, bicarbonate=19.5 mmol/L, sodium=137.7 mmol/L and potassium=3.74 mmol/L. The saturation on the Arterial Blood Gas (ABG) sample was 77.4%, while at that

time, the neonate's saturation was 86% on the monitor, suggesting it was probably a venous/mixed sample. It was unable to confirm whether the sample taken was arterial or venous.

General anaesthesia was planned for the excision and repair of the meningocele. The preoperative orders included the continuation of the antibiotics given in the NICU (injection gentamicin 13 mg once daily and injection piperacillin-tazobactam 260 mg three times daily). The patient remained in the NICU and could not be given breast milk, as the mother was unable to express adequate feeds; therefore, the NICU team kept the neonate on formula milk.

The patient was kept Nil Per Oral (NPO) for six hours. A high-risk consent was obtained from the patient's attendant, including consent for postoperative NICU stay and mechanical ventilation. In the operating room, a pulse oximeter and 5-lead electrocardiography were connected, and a 24 G cannula was already in-situ. Injection atropine 10 mcg/kg was used for premedication. An Intravenous (i.v.) fentanyl dose of 2 mcg/kg was given as an analgesic. Incremental inhalation induction was performed using sevoflurane. Injection atracurium 0.5 mg/kg was used for intubation and as a relaxant throughout the surgery. The Endotracheal Tube (ETT) was secured with gentle laryngoscopy using a size 3.0 mm inner diameter tube. The patient was placed in a prone position for surgery. Anaesthesia was maintained on sevoflurane at 1.5-2.5% in 100% oxygen through a Jackson Rees circuit.

Throughout the intraoperative period, heart rate fluctuations from 135 bpm to 160 bpm were observed, along with Saturation of Peripheral Oxygen (SpO_2) changes from 90% to 94%, while maintaining End-tidal CO_2 ($ETCO_2$) levels of 22-25 mmHg. Injection paracetamol 7.5 mg/kg was used as an analgesic, along with local anaesthetic infiltration. The intraoperative period lasted 45 minutes, with a total i.v. fluid input of 40 mL N/5 + 10% D and minimal blood loss. Following surgery, the patient was reversed in a supine position using injection glycopyrrolate 0.01 mg/kg and Injection Neostigmine 0.05 mg/kg and was extubated. Recovery from anaesthesia was uneventful. The baby was active, crying, with a heart rate of 164 bpm, and maintaining SpO_2 at 96% on nasal prongs at three liters per minute. The patient was shifted to the NICU for monitoring.

DISCUSSION

Ebstein's Anomaly (EA) is considered a rare congenital heart disease, with an estimated incidence of about 1 in 200,000 live births [1]. It is characterised by the downward displacement of the posterior and septal leaflets of the tricuspid valve toward the right ventricular apex. The improper alignment causes TR. The extent of TR depends on the severity of the downward displacement, the abnormal attachment of the anterior leaflet, and the overall integrity of the leaflets [2].

The RV undergoes partitioning into two sections: the 'atrialised' RV and the 'functional' RV. The 'atrialised' RV receives the backward flow caused by TR, leading to dilation along with the right atrium [3]. In severe cases of the disease, an increase in right atrial pressure surpassing that of the left atrium causes a right-to-left shunt, thereby exhibiting cyanosis.

Roughly half of individuals with Ebstein's anomaly exhibit an accessory pathway associated with Wolff-Parkinson-White syndrome. This can predispose them to abnormal heart rhythms, such as atrioventricular re-entrant tachycardia, delayed intra-atrial conduction, right bundle branch block, and ventricular pre-excitation [4].

The fundamental principles in managing cases of Ebstein's anomaly include maintaining appropriate preload and afterload, preserving sinus rhythm, and preventing an increase in right-to-left shunting. This shunting can result from a decrease in SVR, an increase in PVR, or elevated intrathoracic pressure. Right-to-left shunting may also predispose patients to paradoxical emboli during the perioperative period and increase the risk of systemic complications, including stroke or brain abscess [4]. Additionally, it is essential to avoid tachycardia, as it can impair right ventricular filling. In infants, the primary approach to treatment involves supportive measures aimed at decreasing PVR and alleviating hypoxemia. For symptomatic infants experiencing either heart failure or cyanosis, inhaled nitric oxide can be beneficial, and the infusion of prostaglandin E1 (PGE1) can help maintain the patency of the ductus arteriosus, thereby reducing PVR [5].

In this case, a thorough preoperative evaluation was conducted, including a detailed cardiac assessment. This included assessing the severity of TR, the presence of cyanosis, the degree of right ventricular dysfunction, and any associated cardiac anomalies. Continuous monitoring of vital signs, including ECG, heart rate, oxygen saturation, and end-tidal CO_2, was performed. Sinus rhythm was maintained, and careful fluid management was implemented to ensure adequate preload without exacerbating pulmonary congestion. Factors that increase PVR, such as hypoxia, hypercarbia, acidosis and hypothermia, were avoided. Measures were taken to prevent an increase in right-to-left shunting, such as avoiding factors that increase intrathoracic pressure, including high peak pressures, Positive End Expiratory Pressure (PEEP), and excessive coughing or straining.

Avoiding or limiting agents such as propofol or barbiturates, which have direct negative inotropic or vasodilatory effects, is recommended, along with careful titration of volatile anaesthetic agents. Sevoflurane was used for the induction and maintenance of anaesthesia, while intravenous agents like opioids were titrated carefully to avoid haemodynamic instability [6]. Close monitoring in the postoperative period is essential to detect and manage any haemodynamic variations or respiratory compromise promptly. This includes admission to a paediatric intensive care unit.

Gurrieri C et al., reported a case series of 12 patients in which six patients underwent surgical correction for EA before conception, while three patients had tachyarrhythmias [7]. Among all the patients, five were on cardiac medications before pregnancy, two of whom required medication adjustments during pregnancy due to worsening tachyarrhythmias. Of the total, seven patients underwent normal vaginal delivery, three had planned caesarean sections, one was delivered with assisted forceps, and the last experienced precipitous labour. Neuraxial anaesthesia was planned for all caesarean deliveries. They concluded that risk stratification is crucial for the management of pregnant patients with EA. Patients with EA pose challenges due to the disease's pathology itself, along with risks related to pregnancy. Vaginal delivery is suggested as the preferable mode of delivery unless there is an obstetric indication. Uterotonic agents should be used cautiously. Neuraxial analgesia and anaesthesia can be employed as a preferable and safe approach for managing pregnant patients with Ebstein's anomaly (EA) [7].

Overall, the anaesthetic management of neonates with Ebstein's anomaly for non cardiac surgery requires meticulous planning, vigilant monitoring, and collaboration among healthcare providers to achieve successful outcomes, while minimising perioperative risks.

CONCLUSION(S)

Perioperative considerations for EA include right-to-left shunting with the potential for paradoxical emboli, a high onset time for intravenous drugs due to pooling and recirculation of blood in the right atrium, increased PVR and perioperative pulmonary hypertension, right and left ventricular dysfunction, and arrhythmias. Key cardiovascular goals for such patients are maintaining normal sinus rhythm, ensuring adequate preload, avoiding significant changes in PVR or SVR, avoiding medications and factors that depress myocardial function.

REFERENCES

[1] Jost CHA, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. Circulation. 2007;115(2):277-85.

[2] Boston US, Ken B, Kumar TKS, Knott-Craig CJ. 107. Neonatal Ebstein's Anomaly. In Raja, Shahzad G. (ed.). Cardiac Surgery: A Complete Guide. Switzerland: Springer 2020.
[3] Holst KA, Connolly HM, Dearani JA. Ebstein's Anomaly. Methodist Debakey Cardiovasc J. 2019;15(2):138-44.
[4] Phi K, Martin DP, Beebe A, Klamar J, Tobias JD. Anaesthetic care during posterior spinal fusion in an adolescent with Ebstein's anomaly. J Med Cases. 2020;11(3):68-72.
[5] Atz AM, Munoz RA, Adatia I, Wessel DL. Diagnostic and therapeutic uses of inhaled nitric oxide in neonatal Ebstein's anomaly. Am J Cardiol. 2003;91(7):906-08.
[6] Khatib SK, Tadwalkar GV. Anaesthetic implications of Ebstein's anomaly: A report of two cases. Anaesth Pain & Intensive Care. 2012;16(1):61-64
[7] Gurrieri C, Sharpe EE, Connolly HM, Rose CH, Arendt KW. Peripartum anaesthetic management in patients with Ebstein anomaly: A case series. Proc (Bayl Univ Med Cent). 2023;36(3):346-50.

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