https://doi.org/10.47108/jidhealth.Vol8.Iss1.399
Jain AP, et al., Journal of Ideas in Health (2025); 8(1):1236-1239
Journal of Ideas in Health

Navigating the complexities of anesthetic management in a preterm neonate with double outlet right ventricle posted for noncardiac surgery

Ashish P Jain[1*], Akhilesh Chhaya[1], Swapnali Suryavanshi[1], Mohammad Nadeem Shaikh[1], Deep Vadnagara[1], Yatin Bharad[1]

Abstract

Background: Double outlet right ventricle (DORV) is an uncommon congenital heart defect characterized by the simultaneous emergence of both the aorta and pulmonary artery from the right ventricle. This condition is frequently linked with other congenital anomalies, such as imperforate anus, which presents considerable challenges for anesthesiologists. This case report outlines the effective anesthetic management of a preterm neonate diagnosed with DORV and imperforate anus, who was scheduled for an urgent sigmoid colostomy.

Case presentation: A 6-day-old preterm female weighing 2.2 kg, exhibited DORV along with a subaortic ventricular septal defect, mild pulmonary stenosis, adequate biventricular function, and moderate pulmonary hypertension. Additionally, she presented with an imperforate anus accompanied by a recto-vaginal fistula. The preoperative assessment involved echocardiography, electrocardiography, and various laboratory tests. The pediatric cardiologist advised that the patient's oxygen saturation should be maintained within the range of 80-85% throughout the emergency colostomy procedure. Anesthetic management involved premedication with glycopyrrolate, ondansetron, fentanyl, and dexamethasone. The patient was induced with sevoflurane and intubated with a 2 mm endotracheal tube. Anesthesia was maintained with a mixture of air and oxygen, sevoflurane, and atracurium. Hypothermia was prevented using a warmer, and fluid management was guided by the patient's hemodynamic parameters. After the 1-hour surgery, the patient was transferred to the neonatal intensive care unit, where she was initially supported with synchronized intermittent mandatory ventilation and later extubated

Conclusion: The effective anesthetic management of this preterm neonate with DORV and imperforate anus necessitated a comprehensive understanding of the associated cardiac and anorectal anomalies, meticulous preoperative planning, and a collaborative multidisciplinary approach to perioperative care. The anesthesiologist was instrumental in ensuring the patient's safety and enhancing the surgical conditions throughout the emergency colostomy procedure.

Keywords: Double Outlet Right Ventricle (DORV), Imperforated Anus, Preterm Neonate, Neonatal Emergency, Congenital Anomalies, Multidisciplinary Approach, India

Correspondence: Ashish P Jain (drashishpjain@gmail.com)
[1]Parul Institute of Medical Sciences and Research, Parul University, Vadodara, Gujarat 391760, India.

How to cite: Jain A, Chhaya A, Suryavanshi S, Shaikh M, Vadnagara D, Bharad Y. Navigating the complexities of anesthetic management in a preterm neonate with double outlet right ventricle posted for noncardiac surgery. J Ideas Health. 2025 Feb. 28 ;8(1):1236-1239. DOI: 10.47108/jidhealth.Vol8.Iss1.399

Article Info: (**Case Report**)
Received: 16 December 2024
Revised: 04 February 2025
Accepted: 18 February 2025
Published: 28 February 2025

Journal Home page: https://www.jidhealth.com
e ISSN: 2645-9248

Background

Double outlet right ventricle (DORV) is an uncommon congenital heart defect characterized by the emergence of both the aorta and the pulmonary artery from the right ventricle [1]. This intricate cardiac anomaly represents about 1-2% of all congenital heart diseases. DORV frequently coexists with other congenital anomalies, such as imperforate anus, which is a rare form of anorectal malformation [2,3]. The anesthetic management of a preterm neonate with DORV and imperforate anus presents considerable challenges for the anesthesiologist.

Case presentation

The current patient is a 6-day-old preterm female weighing 2.2 kg, who was delivered vaginally. Upon birth, she received vaccinations, and an imperforate anus was observed, prompting notification of specialists. After a series of investigations, she was diagnosed with DORV, imperforate anus, and a rectovaginal fistula, leading to the decision to proceed with an emergency sigmoid colostomy.

Details of Double outlet right ventricle (DORV)

The patient in this case report was diagnosed with DORV with a subaortic ventricular septal defect (VSD), mild pulmonary stenosis, good right and left ventricular function, and moderate

pulmonary arterial hypertension [1,2]. DORV is characterized by the presence of both the aorta and the pulmonary artery predominantly arising from the right ventricle, with variable relationships between the great arteries and the ventricular septal defect [1]. The associated VSD and pulmonary stenosis can lead to cyanosis and right ventricular hypertrophy [2].

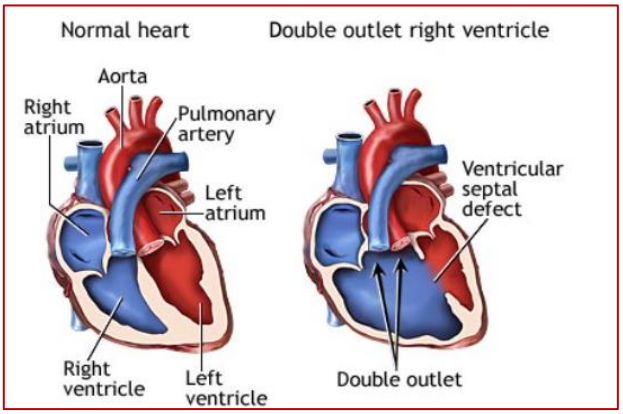

Figure 1: Normal heart vs heart with DORV (Source: https://medlineplus.gov/ency/article/007328.htm) [3]

Details of Imperforate Anus

Imperforated anus is a rare anorectal malformation that occurs in approximately 1 in 5,000 live births [4]. It is characterized by the absence of a normal anal opening and is often associated with other congenital anomalies, including cardiac defects [5]. In the present case, the patient had a recto-vaginal fistula, which is a common type of imperforate anus [4,5].

Anesthesia Management

Preoperative Preparations

The preoperative evaluation and optimization of this preterm neonate with DORV and imperforate anus were crucial for the successful anesthetic management. The patient's preoperative workup, including echocardiography, electrocardiography, and laboratory tests, was performed to assess the severity of the cardiac and other congenital anomalies. The pediatric cardiologist recommended maintaining the patient's oxygen saturation (SpO2) between 80-85% during the surgical procedure to ensure adequate oxygenation and prevent complications. SpO2 >90% should be avoided as it can increase left to right shunting of oxygenated blood due to pulmonary vasodilatation [6].

Intraoperative Management

After informed consent, patient was taken into operation theatre. IV line was secured. The patient received premedication to optimize hemodynamic stability and prevent complications: Inj Glycopyrrolate: 40 mcg, Inj Ondansetron: 0.1 mg, Inj Fentanyl: 3 mcg and Inj Dexamethasone: 0.4 mg intravenously. The patient was induced with sevoflurane and intubated with a 2 mm endotracheal tube. Anesthesia was maintained with a mixture of air and oxygen to maintain the target SpO2 in 80-85% range, along with sevoflurane and inj atracurium (loading dose 0.5mg/kg followed by 0.1mg/kg IV). Hypothermia was managed using full body draping and external warmer to maintain the patient's body temperature within the normal range. Fluid management was guided by the patient's hemodynamic parameters and third space loss. A dial flow was used to ensure adequate fluid administration. Ventilation was performed throughout the procedure using a neonatal mode workstation (Draeger Fabius Plus), with an approximate tidal volume of 20 mL delivered. Closed circuit system made sure that loss of humidity and body heat are minimum. Depth of anesthesia was maintained throughout the surgery. After the surgery, the patient was transferred to the neonatal intensive care unit (NICU), where she was initially supported with synchronized intermittent mandatory ventilation (SIMV) and later extubated. The patient was kept nil by mouth for 48 hours and then gradually transitioned to breastfeeding to prevent hypoglycemia and ensure adequate nutrition. Appropriate antibiotic prophylaxis was administered, and the patient's laboratory values, including elevated C-reactive protein and liver enzymes, were closely monitored and managed. Fluid management was guided by the patient's hemodynamic parameters to maintain adequate perfusion and prevent complications.

NICU Stay and Care

The patient was closely monitored in the NICU, where she received respiratory support and was gradually transitioned to exclusive breastfeeding. The parents were counselled about the need for future surgeries to correct the VSD and imperforate anus.

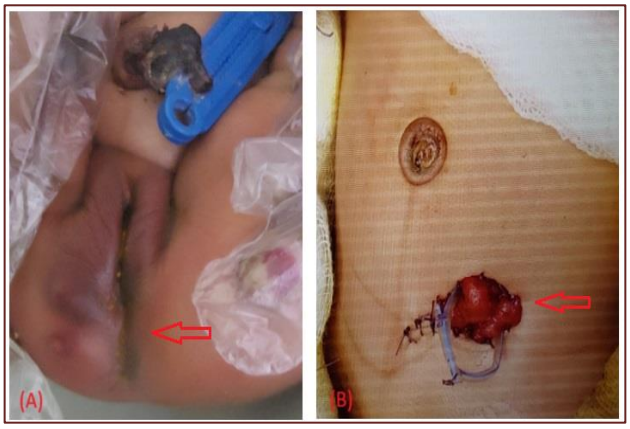

Figure 2: (A): Imperforated anus; (B) Post Colostomy

Discussion

The successful anesthetic management of a preterm neonate with double outlet right ventricle (DORV) and imperforate anus requires a thorough understanding of the underlying cardiac and anorectal anomalies, as well as careful preoperative planning and a multidisciplinary approach to perioperative care. Compared to the anesthetic management of other cyanotic congenital heart diseases, such as tetralogy of Fallot (TOF), the anesthetic considerations for DORV are complex due to the variable relationships between the great arteries and the ventricular septal defect (VSD) [7,8]. In DORV, the aorta and pulmonary artery can have different spatial relationships, leading to a wide range of physiological presentations, from cyanosis to congestive heart failure [7]. This necessitates a tailored approach to anesthetic management, as highlighted in the case report by McClung et al. [9]. Similar to the present case, the anesthesiologists in the McClung et al. study had to carefully manage the patient's oxygenation and ventilation to maintain the target oxygen saturation (SpO2) range recommended by the pediatric

cardiologist [9]. The use of a mixture of air and oxygen, along with close monitoring of the patient's hemodynamic parameters, was crucial in ensuring adequate tissue perfusion and preventing complications [4]. The presence of associated congenital anomalies, such as the imperforate anus in this case, adds further complexity to the anesthetic management. As highlighted in the case report by Singh et al. [10], neonates with congenital cardiac anomalies and difficult airways pose significant challenges to the anesthesiologist, requiring a tailored approach to airway management and ventilation. Compared to the anesthetic management of neonates undergoing other types of surgery, such as vitreoretinal surgery for retinopathy of prematurity Saxena et al. [11] or thoracoscopic surgery [12], the management of a neonate with DORV and imperforate anus requires additional considerations. These include the need for careful hemodynamic monitoring, maintenance of target SpO2 levels, and the management of associated congenital anomalies [1,2,4]. Furthermore, the anesthetic management of neonates with ventricular septal defects (VSDs) has been discussed in several studies [7,8]. While the present case involved a DORV with a subaortic VSD, the anesthetic principles of maintaining hemodynamic stability, optimizing oxygenation, and managing the associated anomalies are similar [1,2]. The importance of a multidisciplinary approach to the perioperative care of these complex patients, as seen in the present case, has been emphasized in various studies [8,9,10]. The collaboration between the anesthesiologist, pediatric cardiologist, and pediatric surgeon is crucial in ensuring the best possible outcome for the patient.

Conclusion

The successful anesthetic management of a preterm neonate with DORV and imperforate anus requires a thorough understanding of the underlying cardiac and anorectal anomalies, careful preoperative planning, and a multidisciplinary approach to perioperative care. The anesthesiologist plays a crucial role in ensuring the patient's safety and optimizing the surgical conditions during the emergency colostomy procedure, drawing on principles from the management of other complex congenital heart diseases and neonatal surgeries.

Abbreviation

DORV: Double Outlet Right Ventricle; IV: Intravenous; NICU: Neonatal Intensive Care Unit; SPO2: Peripheral Oxygen Saturation; SIMV: Synchronized Intermittent Mandatory Ventilation; TOF: Tetralogy of Fallot; VSD: Ventricular Septal Defect.

Declaration

Acknowledgment

None

Funding

The authors received no financial support for their research, authorship, and/or publication of this article.

Availability of data and materials

Data will be available by emailing drashishpjain@gmail.com

Authors' contributions

All authors were equally participated in designing, supervising, the study and conceiving the idea. They worked together in data analysis, interpreted the results and curated and drafted the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

We conducted the research following the declaration of Helsinki. The ethical approval was obtained from Parul Institute of Medical Sciences and Research, Parul University, India.

Consent for publication

Not applicable

Competing interest

The authors declare that they have no competing interests.

Author Details

[1]Parul Institute of Medical Sciences and Research, Parul University, Vadodara, Gujarat 391760, India.

References

1. Bell-Cheddar Y, Devine WA, Diaz-Castrillon CE, Seese L, Castro-Medina M, Morales R, Follansbee CW, Alsaied T, Lin JHI. Double outlet right ventricle. Frontiers in Pediatrics [Internet]. 2023 Sep 25;11. Available from: https://doi.org/10.3389/fped.2023.1244558
2. Karmegaraj B, Kumar S, Srimurugan B, Sudhakar A, Simpson JM, Vaidyanathan B. 3D / 4D spatiotemporal image correlation (STIC) fetal echocardiography provides incremental benefit over 2D fetal echocardiography in predicting postnatal surgical approach in double-outlet right ventricle. Ultrasound in Obstetrics and Gynecology [Internet]. 2020 Feb 5;57(3):423–430. Available from: https://doi.org/10.1002/uog.21988
3. Double outlet right ventricle: MedlinePlus Medical Encyclopedia [Internet]. Available from: https://medlineplus.gov/ency/article/007328.htm
4. Khatib N, Belossesky R, Marwan O, Weiner Z. Fetal bowel calcifications: A sign of anal atresia with rectourethral fistula. Journal of Clinical Ultrasound [Internet]. 2010 Apr 29;38(6):332–334. Available from: https://doi.org/10.1002/jcu.20706
5. Eftaiha SM, Melich G, Pai A, Marecik SJ, Prasad LM, Park JJ. Sacral nerve stimulation in the treatment of bowel dysfunction from imperforate anus: A case report. International Journal of Surgery Case Reports [Internet]. 2016 Jan 1;24:115–118. Available from: https://doi.org/10.1016/j.ijscr.2016.05.018
6. Nursing guidelines: Oxygen saturation SpO2 level targeting in neonates [Internet]. Available from: https://www.rch.org.au/rchcpg/hospital_clinical_guideline_index /oxygen_saturation_spo2_level_targeting_premature_neonates/
7. Kim CY, Kim WH, Kwak JG, Jang WS, Lee CH, Kim DJ, Lim C, Chang WI. Surgical Management of Left Ventricular Outflow Tract Obstruction after Biventricular Repair of Double Outlet Right Ventricle. Journal of Korean Medical Science [Internet]. 2010 Jan 1;25(3):374. Available from: https://doi.org/10.3346/jkms.2010.25.3.374

8. Spaeth JP. Perioperative Management of DORV. Seminars in Cardiothoracic and Vascular Anesthesia [Internet]. 2014 Mar 21;18(3):281–289. Available from: https://doi.org/10.1177/1089253214528048
9. McClung H, Prasad R, Schwartz R. Anesthetic management of tracheoesophageal fistula repair in a newborn with a double-outlet right ventricle. Journal of Clinical Anesthesia [Internet]. 2011 Dec 1;23(8):643–645. Available from: https://doi.org/10.1016/j.jclinane.2011.01.010
10. Singh A, Garg K, Ganesh V, B NN, Singh D. Anaesthesia challenges in a neonate having supracardiac total anomalous pulmonary venous return with incidental stridor for non-operating room anaesthesia (NORA): feed and wrap technique as a rescue. Cureus [Internet]. 2022 Oct 16; Available from: https://doi.org/10.7759/cureus.30357
11. Sinha R, Talawar P, Ramachandran R, Azad R, Mohan V. Perioperative management and post-operative course in preterm infants undergoing vitreo-retinal surgery for retinopathy of prematurity: A retrospective study. Journal of Anaesthesiology Clinical Pharmacology [Internet]. 2014 Jan 1;30(2):258. Available from: https://doi.org/10.4103/0970-9185.130050
12. Liu H, Wang F, Zhang J, Gao Z. Risk factors for anesthesia-associated postoperative capillary leakage after thoracoscopic surgery in neonates: A single-center observational study. Frontiers in Pediatrics [Internet]. 2023 Jan 4;10. Available from: https://doi.org/10.3389/fped.2022.1051069.