



Case Report Education

## Mind the right ventricle: Tackling right heart dysfunction in left ventricular diverticulum with omphalocele

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### ABSTRACT

Left ventricular diverticulum (LVD) is a rare congenital anomaly characterized by an abnormal outpouching of the left ventricular wall. It is often identified during childhood, as it is commonly associated with midline thoracoabdominal defects and other congenital heart abnormalities. Here, we present a 10-month-old boy with a LVD and omphalocele posted for surgical management who had difficulty weaning in the post-operative period. We discuss the unique challenges that necessitate meticulous planning and execution to optimize right ventricular function and prevent pulmonary arterial hypertension in such patients.

**Keywords:** Left ventricular diverticulum, Omphalocele, Right ventricular dysfunction

### INTRODUCTION

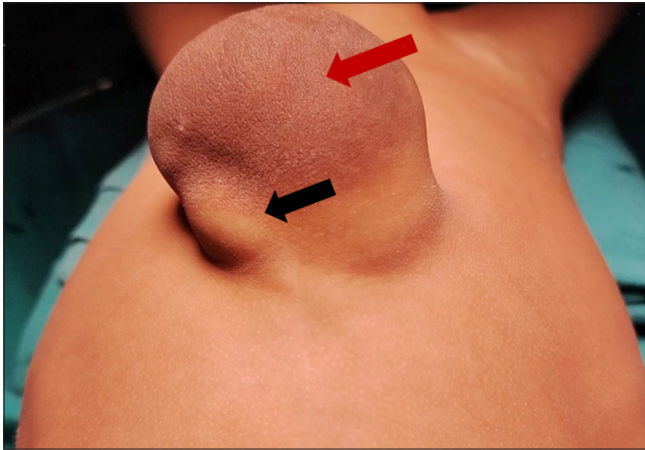
Congenital left ventricular diverticulum (LVD) is a rare anomaly, with an incidence of approximately 0.4%. It is characterized by an outpouching of the left ventricle, which includes the endocardium, myocardium, and pericardium.<sup>[1,2]</sup> Omphaloceles are defects in the ventral abdominal wall that is particularly associated with cardiac defects, in up to 80% of cases. In cases where a severe congenital heart defect (CHD) is present along with omphalocele, the management becomes extremely challenging.<sup>[3]</sup> Over the past few decades, the significance of right ventricular (RV) dysfunction in determining the clinical outcomes of various diseases has become increasingly evident with more than 50% of patients in specific conditions being affected by it, including left heart failure (HF), pulmonary arterial hypertension (PAH), CHDs, and cardiomyopathies.<sup>[4]</sup> We present a unique case of managing post-operative RV dysfunction in a child with a congenital left ventricular (LV) diverticulum associated with omphalocele. This case represents a rare and clinically significant presentation of a congenital LV diverticulum associated with an omphalocele, complicated by post-operative RV dysfunction – a triad that has been sparsely documented in the literature. Furthermore, this case underscores the underrecognized risk of acute RV failure in the post-operative period, despite the primary pathology being left-sided. The detailed perioperative anesthetic and hemodynamic strategies employed here contribute valuable insights for managing such complex congenital anomalies.

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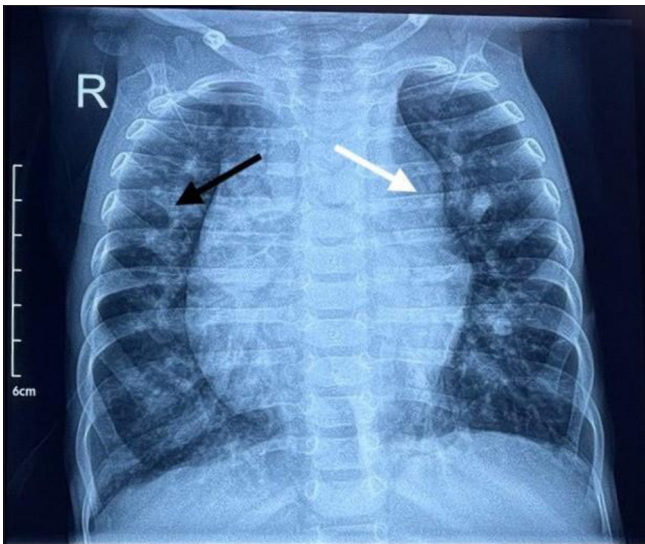
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## CASE REPORT

A 10-month-old male infant presented to our hospital with a growing pulsatile mass at the umbilical region along with a reducible umbilical hernia [Figure 1]. The child had a history of feeding difficulty, recurrent upper respiratory tract infections, and failure to thrive. There was no history of embolic events, syncope, arrhythmia, or HF. Physical examination revealed high-pitched pansystolic murmur at the lower left sternal border. Chest radiograph [Figure 2] showed cardiomegaly with increased pulmonary vascular markings. Transthoracic echocardiography showed a large 10-mm mid-

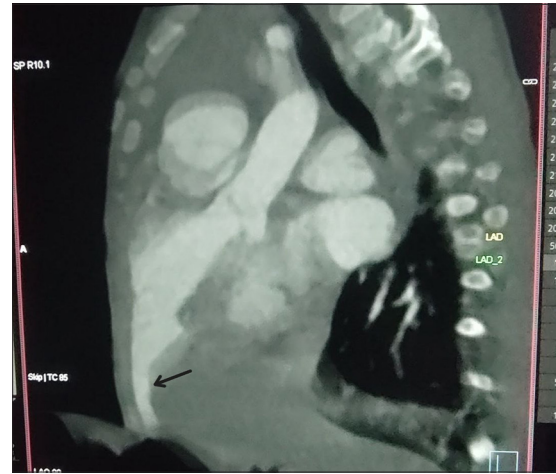


**Figure 1:** A 10-month-old male infant presenting with a pulsatile mass in the umbilical region. This preoperative image shows the mass in the umbilical region measuring  $6 \times 4$  cm (red arrow), with a prominent pulsatile left ventricular diverticulum (black arrow) and the remaining part which was reducible.

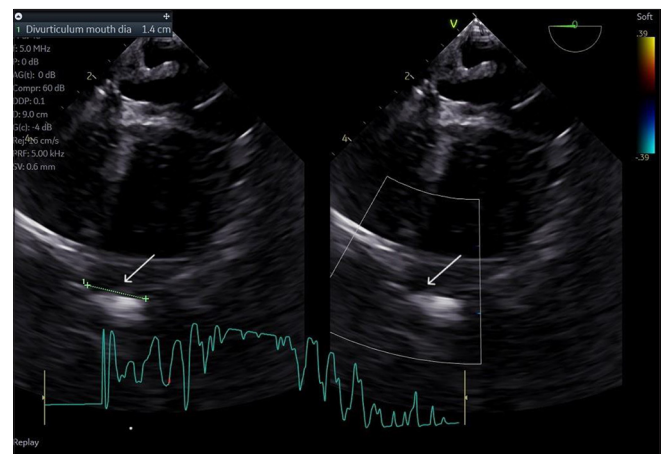


**Figure 2:** A 10-month-old male infant diagnosed with a left ventricular diverticulum. This pre-operative chest radiograph shows cardiomegaly with increased pulmonary vascular markings (black arrow) and a prominent pulmonary artery (white arrow). Note that no sternal deficiency or diaphragmatic hernia is visible here.

muscular ventricular septal defect (VSD) with predominantly left to right shunt. A 6-mm diverticulum from the left ventricular apex extending downward subcutaneously up to the umbilicus was noted. The apex of the diverticulum was seen over the umbilical hernia site. The left atrium and left ventricle were dilated. RV dysfunction was noted and there was no clot in the diverticulum. Computed tomography [Figure 3] confirmed the finding of tubular structure arising from the apex of the left ventricle, measuring 3 mm in width and 4.1 cm in length extending up to the umbilical hernia site which measured 4 cm in width and 3.1 cm long containing colon loops with no features of obstruction.



**Figure 3:** A 10-month-old male infant presenting with a pulsatile umbilical mass. Pre-operative sagittal slice computed tomography angiograph of the thoracoabdominal region is showing a tubular, contrast-filled outpouching extending from the apex of the left ventricle (black arrow) suggestive of left ventricular diverticulum.



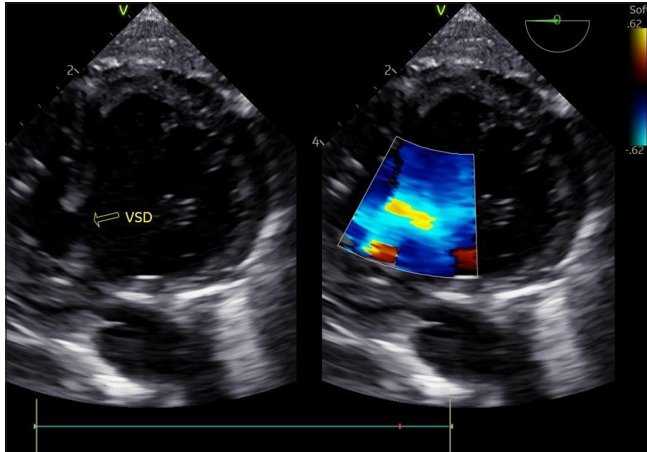
**Figure 4:** A 10-month-old male infant diagnosed with a left ventricular diverticulum. Intraoperative transesophageal echocardiography with a mid-esophageal four-chamber view showing the diverticulum (arrow) as an outpouching of the left ventricle with its diameter being measured. The diverticulum is visualized arising from the apical region of left ventricle with an orifice of size 1.4cm in diameter.

Intra-operative transesophageal echocardiography [Figures 4 and 5] showed a patent foramen ovale with left to right shunt, mild tricuspid regurgitation, a 12-mm perimembranous VSD with left to right shunt and a LVD with a wide ventricular connection measuring 1.2 cm.

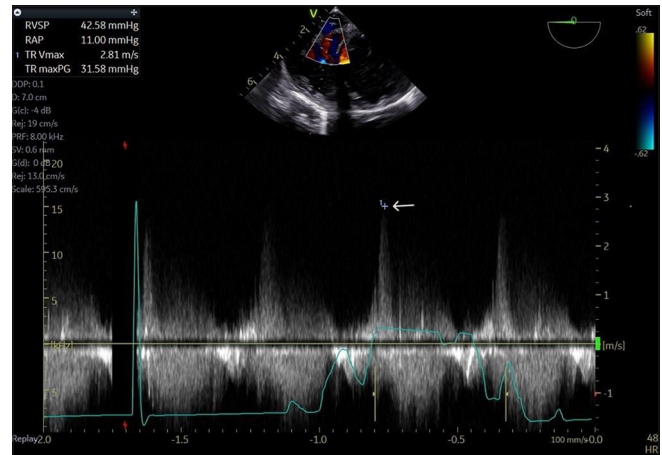
The child underwent VSD closure with excision of the diverticulum [Figure 6] under cardiopulmonary bypass support and umbilical hernia repair surgery. Given the significant risk of spontaneous rupture, thromboembolic

events, and progression of cardiac dysfunction associated with an LVD, the surgical team opted for a single-stage procedure comprising diverticulum excision and umbilical hernia repair. This decision was guided by the anatomical continuity of the diverticulum with the hernia sac and the potential for hemodynamic compromise if managed in separate surgical settings. The operation was performed under cardiopulmonary bypass to allow precise myocardial repair and to mitigate the risk of intraoperative circulatory instability.

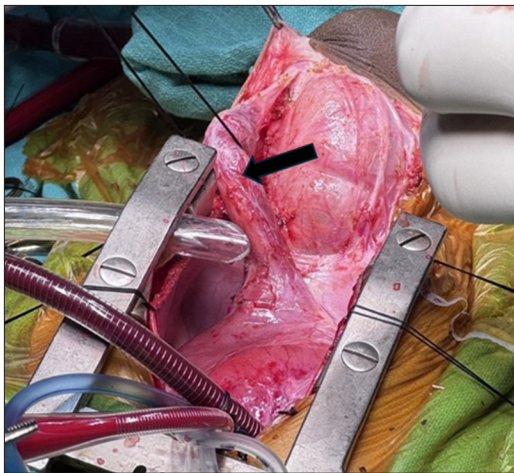
General anesthesia (GA) was induced with intravenous fentanyl (2 mcg/kg), midazolam (0.05 mg/kg), sevoflurane,



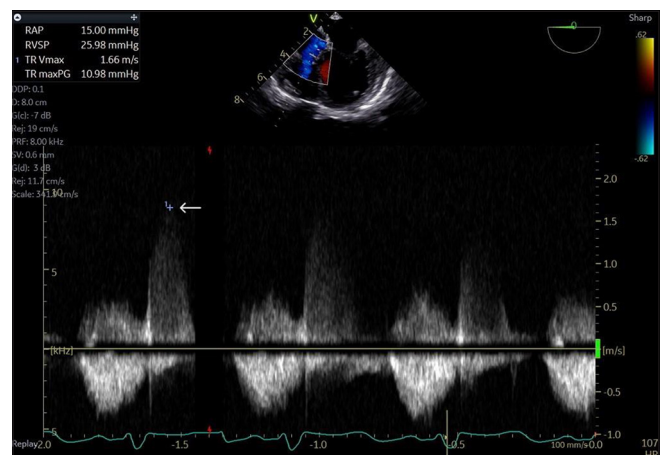
**Figure 5:** A 10-month-old male infant diagnosed with a left ventricular diverticulum. This intraoperative transesophageal echocardiography transgastric mid-papillary short axis view showing a perimembranous ventricular septal defect (arrow) with a color doppler interrogation demonstrating a left to right shunt.



**Figure 7:** A 10-month-old male infant diagnosed with a left ventricular diverticulum. This immediate post-operative echocardiography showing elevated right ventricular systolic pressure (arrow showing tricuspid regurgitation peak velocity of 2.81m/s) suggesting right ventricular dysfunction and pulmonary artery hypertension.



**Figure 6:** A 10-month-old male infant diagnosed with a left ventricular diverticulum. This intra-operative surgical image showing the left ventricular diverticulum as a tubular, vascularized structure (black arrow) originating from the apex of the left ventricle and extending anteriorly toward the abdominal wall. The image also shows the placement of perfusion cannulas and retraction for optimal surgical field visualization.



**Figure 8:** A 10-month-old male infant diagnosed with a left ventricular diverticulum. Postoperative echocardiography image during intensive care unit stay showing normal right ventricular systolic pressure (arrow showing tricuspid regurgitation peak velocity of 1.66m/s) after management of right ventricular dysfunction.

and rocuronium (1 mg/kg). After endotracheal intubation, GA was maintained using sevoflurane with oxygen. Invasive monitoring included the placement of both central venous and arterial catheters to facilitate continuous assessment of pre-load, systemic arterial pressure, and response to pharmacologic interventions.

Postoperatively echocardiography [Figures 7 and 8] showed severe RV dysfunction due to which there was difficulty weaning off ventilator amounting to prolonged mechanical ventilation and intensive care unit (ICU) stay. The patient remained on mechanical ventilation for 5 days. To support RV contractility and reduce afterload, a continuous infusion of milrinone (0.5 mcg/kg/min) and dobutamine (5 mcg/kg/min) was administered for 72 h. RV function began improving by day 5 and normalized echocardiographically by post-operative day 7. In the ICU, the patient required prolonged ventilation and inotropic support due to persistent RV dysfunction. Inhaled nitric oxide and sildenafil were initiated for PAH management. Gradual improvement was noted with normalization of RV pressures by post-operative day 7.

## DISCUSSION

The management of RV dysfunction (RVD) in a child presenting with LVD, peri-membranous VSD, and omphalocele requires a collaborative, multidisciplinary approach. Special attention must be given to perioperative care, particularly to mitigate factors that can exacerbate RVD and PAH. In this complex congenital case, the causes of RVD are multifactorial and can be attributed to (i) increased RV afterload due to increased pulmonary blood flow from the VSD leading to PAH, (ii) impaired myocardial contractility resulting from structural abnormalities because of the LVD, which can further affect RV contractility due to ventricular interdependence, and (iii) diastolic dysfunction occurring as a result of combination of volume overload and myocardial anomalies impairing the right ventricle's ability to fill adequately during diastole.

Omphalocele, a congenital defect where abdominal organs protrude through the umbilicus, often necessitates staged surgical repair to minimize complications. Large omphaloceles are associated with pulmonary hypoplasia and subsequent PAH.<sup>[5]</sup> Surgical intervention for omphalocele can further exacerbate right heart dysfunction through increased intra-abdominal pressure, which may impair venous return, reduce preload, and compress the diaphragm. These changes can elevate pulmonary arterial pressures, straining the right ventricle and predisposing it to failure.<sup>[6]</sup> This report builds upon recent findings that emphasize the high incidence of cardiac anomalies in neonates with omphalocele and supports the need for early, coordinated multidisciplinary intervention to optimize outcomes. A recent study by Țarcă *et al.*, reinforces

the importance of this association and the need for vigilant cardiovascular assessment and follow-up in such patients.<sup>[5]</sup>

To address these challenges, surgical teams often adopt staged repair approaches for large omphaloceles to prevent acute increases in intra-abdominal pressure. Central venous pressure (CVP) monitoring and real-time hemodynamic assessments guide intraoperative and post-operative management. Maintaining adequate lung expansion and preventing excessive PAH are paramount.

Effective anesthesia strategies to preserve right ventricular function begin with a thorough preoperative assessment of hemodynamics and the severity of pulmonary arterial hypertension (PAH), typically using echocardiography or right heart catheterization.<sup>[7]</sup> Intraoperatively, agents such as sevoflurane and opioids are preferred for their minimal effects on hemodynamics, while avoiding ketamine and nitrous oxide. Ventilation strategies include low tidal volumes (6–8 mL/kg) and high respiratory rates to prevent volutrauma. Hypercapnia and hypoxia should be avoided to prevent exacerbating PAH. Hemodynamic support includes CVP monitoring and inotropes such as milrinone or dobutamine to support RV function.<sup>[8]</sup> Postoperatively, ICU monitoring for PAH crises is essential, with pain managed using multimodal analgesia to avoid respiratory depression. Pulmonary vasodilators such as inhaled nitric oxide and sildenafil help manage PAH, while fluid management must avoid overload to prevent right HF.

In summary, the interplay between congenital cardiac defects, pulmonary hypertension, and omphalocele-associated changes in physiology underscores the importance of meticulous perioperative planning. Timely interventions and vigilant monitoring can significantly improve outcomes in these complex cases. A limitation of our report is the lack of long-term follow-up due to loss of follow-up post-discharge, which restricts assessment of cardiac remodeling and late RV function.

## CONCLUSION

This case report emphasizes the need for a heightened awareness of the potential for RV dysfunction in patients with LVD in the perioperative period. Comprehensive pre-operative assessment, judicious selection of anesthetic agents, meticulous intraoperative monitoring, and proactive hemodynamic management are essential components to optimize patient care and improve surgical outcomes.

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