CASE REPORT

Journal of Indonesian Thoracic Cardiac and Vascular Surgery *(JINATCVS)* 2025,
Volume 2, Number 1: 16-20

Published By :
The Indonesian Association of Thoracic and Vascular Surgeons

Postoperative chylothorax intervention in a pediatric patient with complex congenital cardiovascular diseases: a case report

Yunanto Kurnia[1*], Haryo Aribowo[1], Yuletta Adny Ambarsari[1], Ihsanul Amal[1], Trianingsih[2], Yosua Martin[1], Yoga Arditya[1], Andrea Radyaputri[3]

[1]Department of Thoracic and Cardiovascular Surgery, Sardjito General Hospital, Yogyakarta, Indonesia;
[2]Department of Radiology, Sardjito General Hospital, Yogyakarta, Indonesia;
[3]Universitas Gadjah Mada, Yogyakarta, Indonesia.

*Corresponding to:
Yunanto Kurnia;
Department of Thoracic and Cardiovascular Surgery, Sardjito General Hospital, Yogyakarta, Indonesia;
yunantokurnia@ugm.ac.id

Received: 2025-01-02
Accepted: 2025-02-17
Published:2025-03-15

ABSTRACT

Background: Postoperative chylothorax is a chyle extravasation into the pleural space due to disruption of the thoracic duct within 30 days of surgery. When chest tube output is ≥1,100 ml over any 24 hours, ≥1 L/day for more than five days, or ≥2 L after two days of optimal conservative therapy such as dietary modification and pharmacological therapies, operative therapy or surgical intervention should be considered. Here we present a case of refractory chylothorax after pediatric surgery that did not respond to conservative treatment, and aim to investigate the recommended treatment guideline.

Case Presentation: A 3-year and 9-month-old pediatric patient with dextrocardia mirror-image, double outlet right ventricle (DORV), hypoplastic left ventricle, malposition of the great arteries, and severe pulmonary stenosis underwent a right-sided pulsatile Bi-directional Cavo-pulmonary Shunt (BCPS) and concurrent patent ductus arteriosus ligation. The patient was re-admitted to the hospital following the procedures due to productive cough, dyspnea, and decreased appetite. The chest x-ray revealed a suspicion of BCPS obstruction and left-sided milky pleural effusion. Thoracic duct ligation was performed. The patient's condition improved.

Conclusion: The management of post-operative chylothorax is based on the chyle production. Patients not responding to conservative treatment may need secondary invasive interventions with undetermined optimal timing. However, the current paradigm suggests that 5-14 days of conservative treatment is adequate to evaluate the need for invasive treatment. High-risk patients may require early invasive treatment, hence the need for risk factor assessment, including superior vena cava high pressure, premature birth, low postoperative albumin, and low body weight and height.

Keywords: Chylothorax, Conservative, Thoracic, Treatment.
Cite This Article: Kurnia, Y., Aribowo, H., Ambarsari, Y.A., Amal, I., Trianingsih., Martin, Y., Arditya, Y., Radyaputri, A. 2025. Postoperative chylothorax intervention in a pediatric patient with complex congenital cardiovascular diseases: a case report. *Journal of Indonesian Thoracic Cardiac and Vascular Surgery* 2(1): 16-20

INTRODUCTION

A chylothorax is a medical condition of chyle extravasation into the pleural space as a result of possible obstruction or injury of the thoracic duct. The pleural fluid often has a milky appearance and is characterized by elevated triglycerides of >110 mg/dl or the presence of chylomicrons.[1] Postoperative chylothorax is described as the chylothorax that develops within 30 days of surgery. The amount of the product may be classified based on the maximum cumulative output of all inserted chest tubes for 24 hours. Chylothorax was classified as high (≥20 ml/kg/day) or low (<20 ml/kg/day). When chest tube output is ≥1,100 ml over any 24 hours, ≥1 L/day for more than five days, or ≥2 L after two days of optimal conservative therapy, operative therapy or surgical intervention should be considered.

Lifestyle and dietary modification, along with medical therapies such as pleural fluid drainage and the use of somatostatin analogue, may be considered for chylothorax cases with lesser amounts of chest tube product. Somatostatin and octreotide, as somatostatin analogues, function to inhibit somatostatin receptors in lymphatic vessels, thus constricting the lymphatic vessels and reducing the lymphatic fluid secretion. The previous research showed that surgical duct ligation is more effective than thoracic duct embolization, with the ability to cannulate the cisterna chyli being the limiting factor.[2]

The increased incidence of postoperative chylothorax is associated with the increased number of pediatric cardiac surgeries.[3,4] The proposed mechanisms of postoperative chylothorax are damage to the thoracic duct, disruption of the accessory lymphatic vessel, and increased pressure in the superior vena cava.[4]

Despite the magnitude of postoperative chylothorax after pediatric cardiac surgery, the lack of a definitive guideline leads to practice variation. The "Chylothorax Work Group" has developed a consensus to address this problem. The group's intention in developing the consensus is not essentially to be a definitive guideline, but to act as a platform to drive further studies into a definitive guideline.[5]

The standard approach to managing chylothorax typically begins with

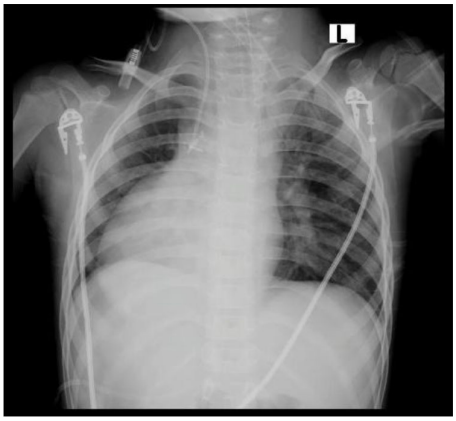

Figure 1. The patient's chest x-ray photo post-Bidirectional cavo-pulmonary shunt (BCPS) on November 2nd 2023, with dextrocardia mirror-image, widened vascular pedicle, rounded cardiac apex, and clear, normal lungs.

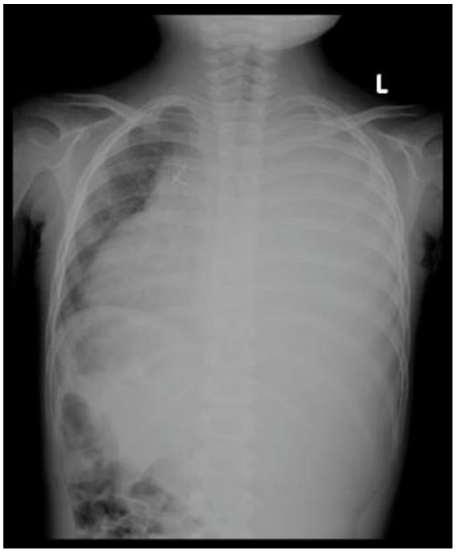

Figure 2. The patient's thorax x-ray on the first day of admission after surgery on November 16th 2023, showed a large opacity of the left lung, indicating massive left-sided pleural effusion. This finding highlighted the urgency for further action to manage the effusion and the patient's complaints.

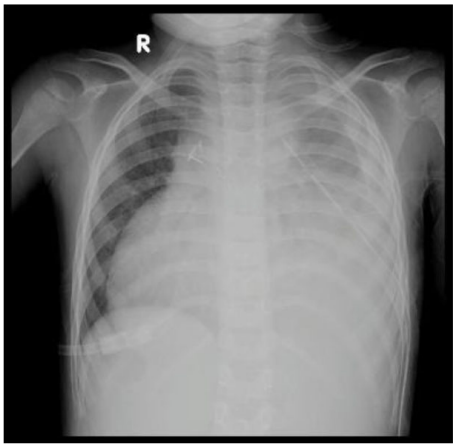

Figure 3. Post-water seal drainage (WSD) thorax x-ray photo on November 18th 2023, showed improvement in the previous pleural effusion marked by the clearer lucency on the upper left lung.

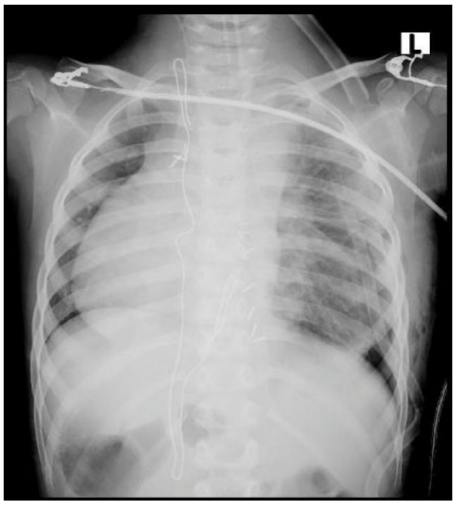

Figure 4. Post-duct ligation thorax x-ray photo on December 4, 2023, showed a prominent reduction in the left pleural effusion marked by increasing lucency of the left lung.

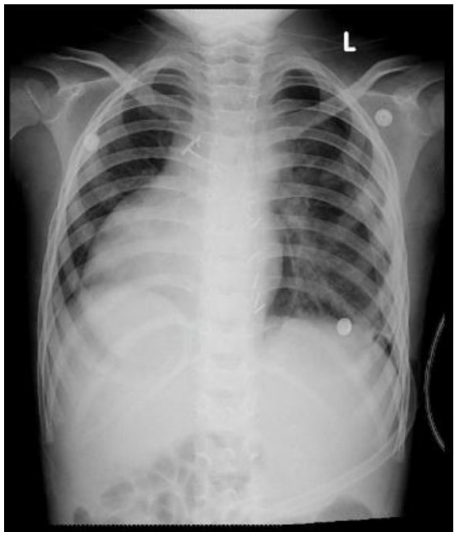

Figure 5. The patient's thorax x-ray image before being discharged from the hospital on December 9, 2023, showed dextrocardia mirror-image and clear, normal lungs.

conservative therapy. When patients do not respond adequately, the condition is classified as refractory chylothorax and may require more invasive interventions. However, the appropriate timing for transitioning to invasive treatment remains a matter of debate. In this report, we present the first documented case of refractory chylothorax following a Bi-directional Cavo-pulmonary Shunt (BCPS) in a pediatric patient at Dr. Sardjito General Hospital, the highest referral center in the Special Region of Yogyakarta, which failed to improve with conservative measures. Based on those mentioned above, this case study aims to explore current evidence-based recommendations for managing such complex situations.

CASE PRESENTATION

The patient was a 3-year-old and 9-month-old female. Six months prior to the admission, she was diagnosed with dextrocardia mirror-image, double outlet right ventricle (DORV), hypoplastic left ventricle, malposition of the great arteries, and severe pulmonary stenosis. Two weeks before the admission, the patient underwent a right-sided BCPS procedure with maintenance of an open main pulmonary artery (pulsatile BCPS) and concurrent patent ductus arteriosus (PDA) ligation. Intraoperative measurements revealed a pulmonary artery pressure of 18 mmHg. There was no significant post-operative complication observed in the patient as shown in the chest x-ray photo (Figure 1). After being discharged, she was then given a prescription of sildenafil, furosemide, aspirin, and captopril.

The patient presented in the outpatient clinic with a productive cough, dyspnea, and decreased appetite without significant impact on routine activities. Clinical examination disclosed cyanosis, particularly evident in the oral region. Facial edema was apparent without nasal flaring or chest wall retractions. Left lung vesicular sounds were inaudible, with ronchi detected in the right lung. A grade 3 systolic murmur was auscultated along the right parasternal line. The extremities exhibit warmth, cyanosis, and clubbing of the fingers. The heart rate was 140 beats per minute, the respiratory rate was 40 breaths per minute, the body temperature was 36.8 °C, and the oxygen saturation was 72%.

Those findings led to the suspicion of an obstruction of the previously performed BCPS. A chest x-ray was obtained as shown in Figure 2, revealing a substantial left-sided pleural effusion. Subsequently, a chest tube was promptly inserted on the day of admission, followed by a decrease in pleural effusion volume within 2 days, as shown in Figure 3, and the product confirmed the presence of milky pleural effusion. Laboratory analysis of the effusion exhibited a positive Rivalta test, LDH of 358, and triglyceride levels of 1,200. Starting from the 5th day of care, the patient was given medium-chain triglyceride (MCT) based enteral nutrition. On the next day, the chest tube output was 330ml/day (22.5ml/kg/day), and we started giving total parenteral nutrition (TPN) to the patient. Intravenous octreotide was initiated on the 9th day with an initial dose of 1 mcg/kg/day. The octreotide dose was then increased to 3 mcg/kg/day on the following day. On the 12th day of care or the 6th day since the start of TPN, considering the persistent drainage was more than 20 ml/kg/day, it was deemed necessary to proceed with a thoracic duct ligation. Concurrently, the octreotide dose was increased to 6 mcg/kg/day.

On the 18th day of care (12th day of TPN), the thoracic duct ligation was performed. Intraoperative observation showed that the left lung was not able to fully inflate because of the fibrosis and adhesion in the surrounding tissue. The procedure led to a reduction in chest tube drainage to a range of 30-60 ml/day (2.04 - 4.09 ml/kg/day) and the more improved thorax x-ray image as shown in Figure 4. The chest tube was removed five days after the procedure was done. The patient reported minimal dyspnea, absence of cough, and exhibited stable vital signs as well as hemodynamics encouraged by normal thorax x-ray (Figure 5).

DISCUSSION

Chylothorax is one of the significant complications following congenital cardiac surgery in pediatric patients, although rare. Several mechanisms underlying postoperative chylothorax include direct injury of the lymphatic vessels or the thoracic duct, hypertension of the systemic vein, as well as vein thrombosis. Surgical incisions on the neck and thorax or needing mobilization over the thoracic duct may injure the duct, which commonly drains to the systemic venous circulation at the subclavian vein and the internal jugular vein junction. This often occurs in patients undergoing procedures for patent ductus arteriosus (PDA) or coarctation of the aorta. As a consequence, the chyle from the duct drains to the mediastinum or passes through the pleural cupola, accumulates in the pleural spaces, creating effusion, and induces an inflammatory reaction.[3,4,6]

Increased central venous pressure (CVP), higher than lymphatic pressure, resulting in chylothorax, may also appear in patients of bidirectional or total cavopulmonary shunt and Fontan procedures, similar to the patient in our case.[3,4,7,8] The increase in systemic venous hydrostatic pressure induces congestion, which disrupts the normal flow of the lymphatic circulation and stimulates more lymph production, specifically from the liver. The definition of high venous pressure remains debatable, but some evidence reported 15 mmHg as the cut-off value due to worse hemodynamic outcomes when the value exceeds CVP >15 mmHg, as minimum as 50%-75% of the lymphatic flow is also preserved with a CVP <15 mmHg.[7,8] Moreover, because the vena cava is anastomosed directly to the pulmonary artery, the arterial pressure (PAP) will also affect the CVP. The recommended PAP for a successful BCPS or Fontan procedure is <15 mmHg.[9] However, a more recent study reported that intraoperative CVP must be preserved at 15-20 mmHg to allow passive pulmonary perfusion and optimal cardiac output.[10] Therefore, the decision to manage the patient's CVP should be based on their clinical conditions and comorbidities.

Despite the common incidence and comorbidities, currently, there is no standard guideline for postoperative chylothorax management. An initiative called "Chylothorax Work Group" has tried to construct a consensus about some recommendations on the management of post-operative chylothorax in pediatric congenital heart disease, and it is meant to be a starting point for further study about chylothorax and eventually lead to definitive guidelines.[5]

The management concept of post-operative chylothorax divides chylothorax into low-volume and high-volume chylothorax based on the quantity of the chest tube output. Conservative treatment, such as pleural drainage, fat-modified diet with medium-chain fatty acids (MCFAs) with subsequent *nil-per-os* (NPO), and pleurodesis to obliterate the pleural space, is considered suitable for the initial approach. The first conservative treatment phase includes a diet with MCFAs for 5-7 days, which will reduce the chyle production due to their direct absorption into the portal venous system instead of the intestinal lymphatic system. This reduction in the flow of lymph also promotes the spontaneous closure of the lymph leak. The second phase is oral feeds termination, followed by TPN for 5-7 days. On the other hand, chemical pleurodesis can be used to reduce the need for surgical therapy.[4,11,12]

Cases that do not respond well to conservative treatment are categorized as refractory chylothorax and may need further medications, such as octreotide for 5-7 days if the chest tube drainage remains >20 ml/kg/day, reduction of central venous pressure up to <15 mmHg, or secondary invasive interventions.[4,5,7,11] Octreotide, a somatostatin analogue used for the patient in our case, was expected to stimulate the chylous fistula closure. Octreotide is generally safe despite being used with high doses and for up to 3 weeks. The recommended dose of octreotide is 0,5-20 mcg/kg/hour or 6 mg/day intravenously or 100 mcg every 8 hours if given as a subcutaneous injection.[12,13] If required, an initial dose of 1 mcg/kg/hour can be increased daily by 1-10 mcg/kg/hour up to a maximum dose of 20 mcg/kg/hour, such as in persistent chylothorax.[14] In our case, the patient did not respond to a similar initial dose, hence the dose was up-titrated before it was eventually decided that surgery would be performed.

Another alternative management besides conservative treatment includes embolization of the thoracic duct.[3,12] Percutaneous duct embolization or disruption has been considered an important non-surgical high-output chylothorax management, with a success

rate up to 80%. The interruption of the duct will trigger thrombus formation and inflammation, promoting closure of the leak. Albeit being a therapy option for the patient in our case, duct embolization using a catheter would not be feasible to do safely due to the small diameter of the patient's thoracic duct.

Therefore, the patient in our case finally received a secondary intervention of thoracic duct ligation after the 12th day of TPN. Ligation of the duct is the first-line surgical therapy.[3] Thoracic duct mass ligation targets the duct to be sealed as it drains into the right hemithorax through the aortic hiatus. This procedure may be performed using open thoracotomy or a minimally invasive surgery technique via robotic or video-assisted thoracoscopy, which has been proven to have better results.[12]

Currently, there is no ideal time to switch to secondary invasive treatment on refractory chylothorax, conservative therapies, which are standardized in all centers. Waiting longer to see whether the patient responds to conservative treatment can minimize the rate of invasive treatment, but not without its risks. A more extended period of high-volume chylothorax can lead to severe complications such as infection (plastic bronchitis), thrombosis, deficiency of protein, immunologic deficiencies, and even death.[3,4,7,15] In addition, because in our case the chest tube product of 20 ml/kg/day did not show improvement, and the patient continued to be fed with low low-fat diet, long observation, especially for pediatric patients, will potentially worsen their clinical manifestations, nutritional status, as well as mental status. The risk associated with invasive treatment is also considered small compared to the benefits, one of which is right ventricular failure prevention by BCPS, hence mortality reduction, especially in the cases of persistent high-output chylothorax.[4,16] The decision to give early surgical intervention for our patient was expected to improve the chylothorax sooner as well. Therefore, there came a paradigm shift towards earlier utilization of invasive treatment, with success rate evidence up to 90%.[12]

Despite the traditional practice of 3-4 weeks of conservative treatment before considering invasive treatment, evidences suggest that as early as 5-7 days up to 14 days of conservative treatment is enough to evaluate the need for invasive treatment.[3,15] A previous study reported that maintenance of conservative treatment with NPO and octreotide up to more than 10 days before switching to surgery resulted in poor postoperative outcomes.[3] Several studies recommended secondary invasive treatment if the chest tube output exceeds >100 ml/kg/day or >1 L/day for 5-7 days, >100 ml/day continuously for >2 weeks, remains unchanged for 1-2 weeks, or the patient deteriorates (in hemodynamic, nutritional, or metabolic aspect) despite on consecutive NPO or TPN.[3–5,11,17] There is a portion of patients (13-27%) with high volume chylothorax unresponsive to 7 7-day NPO strategy, hence requiring further invasive treatment.[15,18] Our patient reported above is one of the patients who did not respond well to >7 consecutive days of TPN and developed complications such as hypoalbuminemia, lymphopenia, and fibrosis of lung tissue observed intraoperatively.

Other predictive factors besides chest tube output and chylothorax duration are needed to determine a portion of patients who have a higher risk of developing high-volume and refractory chylothorax. Patients with higher risk may need to be considered to receive early invasive treatment to minimize complications of prolonged high-volume chylothorax. Previous studies suggested that some risk factors are associated with failure of conservative management and needing early surgical treatment, such as high venous pressure in the SVC, low postoperative albumin, premature birth, high volume loss, as well as low body weight and height.[4,7,19]

CONCLUSION

Chylothorax poses a significant postoperative complication in pediatric cardiac surgery, yet a standardized guideline for its management is currently lacking. The timing for transitioning to invasive treatment is contentious, with a paradigm shift favoring earlier utilization of invasive treatment to minimize complications associated with prolonged high-volume chylothorax. Further studies are needed to determine other predictive factors of developing refractory chylothorax besides chest tube output and chylothorax duration.

ETHICAL APPROVAL

This study has been approved by the Medical and Health Research Ethics Committee of the Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada, with the approval number of KE/FK/0221/EC/2024.

FUNDING

This study did not accept funding from external sources, grants, or third-party support.

CONFLICT OF INTEREST

The authors have fulfilled the ICMJE uniform disclosure form. The authors do not have a conflict of interest to declare.

AUTHOR CONTRIBUTION

YK, HA, YAA, IA, and T were involved in establishing the research concept, designing, and supervising the process of manuscript writing. YM, YA, and AR conducted the study and analyses. All authors prepared the manuscript and agreed to this final version of the manuscript to be submitted to this journal.

REFERENCES

1. Riley LE, Ataya A. Clinical approach and review of causes of a chylothorax. Respir Med. 2019;157:7-13. doi:10.1016/j.rmed.2019.08.014.
2. Reisenauer JS, Puig CA, Reisenauer CJ, Allen MS, Bendel E, Cassivi SD, et al. Treatment of Postsurgical Chylothorax. Ann Thorac Surg. 2018;105(1):254-262. doi:10.1016/j.athoracsur.2017.07.021.
3. Lee SK, Choi KH. Incidence of Postoperative Chylothorax Following Congenital Heart Surgery in Pediatric Patients: A Single-Center Experience. Heart Surg Forum. 2020;23(4):E430-E434. doi:10.1532/hsf.3043.
4. Ahmed MA Sr. Post-Operative Chylothorax in Children Undergoing Congenital Heart Surgery. Cureus. 2021;13(3):e13811. Published 2021 Mar 10. doi:10.7759/cureus.13811
5. Lion RP, Winder MM, Amirnovin R, Fogg K, Bertrandt R, Bhaskar P, et al. Development of consensus recommendations for the management of post-operative chylothorax in paediatric CHD. Cardiol Young. 2022;32(8):1202-1209. doi:10.1017/S1047951122001871.

6. Albåge A. Editorial: postoperative chylothorax-a cause for concern. Acta Neurochir (Wien). 2017;159(10):2023-2024. doi:10.1007/s00701-017-3297-7.
7. Lo Rito M, Al-Radi OO, Saedi A, Kotani Y, Ben Sivarajan V, Russell JL, et al. Chylothorax and pleural effusion in contemporary extracardiac fenestrated fontan completion. J Thorac Cardiovasc Surg. 2018;155(5):2069-2077. doi:10.1016/j.jtcvs.2017.11.046.
8. Kim G, Ko H, Byun JH, Lee HD, Kim H, Sung SC, et al. Risk Factors for Prolonged Pleural Effusion After Extracardiac Fontan Operation. Pediatr Cardiol. 2019;40(8):1545-1552. doi:10.1007/s00246-019-02183-3
9. Carrel T. Commentary: Vis a tergo-a push from behind-is of paramount importance for the optimal function of a bidirectional cavopulmonary shunt. J Thorac Cardiovasc Surg. 2020;160(6):1541-1542. doi:10.1016/j.jtcvs.2020.04.128.
10. Salik I, Mehta B, Ambati S. Bidirectional Glenn Procedure or Hemi-Fontan. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2022.
11. Wagh S, Deshmukh S, Ankolikar S. Conservative treatment of postoperative chylothorax: a case report. J Basic Clin Physiol Pharmacol. 2018;29(4):417-419. doi:10.1515/jbcpp-2017-0082.
12. Martucci N, Tracey M, Rocco G. Postoperative Chylothorax. Thorac Surg Clin. 2015;25(4):523-528. doi:10.1016/j.thorsurg.2015.07.014.
13. Bui A, Long CJ, Breitzka RL, Wolovits JS. Evaluating the Use of Octreotide for Acquired Chylothorax in Pediatric Critically Ill Patients Following Cardiac Surgery. J Pediatr Pharmacol Ther. 2019;24(5):406-415. doi:10.5863/1551-6776-24.5.406.
14. Kalomenidis I. Octreotide and chylothorax. Curr Opin Pulm Med. 2006;12(4):264-267. doi:10.1097/01.mcp.0000230629.73139.26.
15. Winder MM, Eckhauser AW, Delgado-Corcoran C, Smout RJ, Marietta J, Bailly DK. A protocol to decrease postoperative chylous effusion duration in children. Cardiol Young. 2018;28(6):816-825. doi:10.1017/S1047951118000392.
16. Koirala R, Panthee N, Pradhan S, Rajbhandari N, Shrestha DK, Chettri S, et al. In Hospital Outcomes after Bidirectional Cavopulmonary Anastomosis: 18 Year Experience from a Single Center. Nepalese Heart Journal. 2020;17(2):13–19. doi:10.3126/njh.v17i2.32673.
17. Samanidis G, Kourelis G, Bounta S, Kanakis M. Postoperative Chylothorax in Neonates and Infants after Congenital Heart Disease Surgery-Current Aspects in Diagnosis and Treatment. Nutrients. 2022;14(9):1803. doi:10.3390/nu14091803.
18. Shin YR, Lee H, Park YH, Park HK. Chylothorax after Surgery for Congenital Cardiac Disease: A Prevention and Management Protocol. Korean J Thorac Cardiovasc Surg. 2020;53(2):41-48. doi:10.5090/kjtcs.2020.53.2.41.
19. Zhang RG, Liu YM, Yao ZY, Fang JX, Li Y, Zheng ML, et al. Risk Factors of Chylothorax After Congenital Heart Surgery in Infants: A Single-Centre Retrospective Study. Ther Clin Risk Manag. 2024;20:161-168. doi:10.2147/TCRM.S436991.