

Role of magnetic resonance lymphangiography in the early complication management of post-Fontan palliation: A case report

Fontan palyasyonu sonrası erken komplikasyon yönetiminde manyetik rezonans lenfanjiyografinin yeri: Olgu sunumu

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ABSTRACT

Lymphatic complications are associated with increased morbidity and mortality in single-ventricle patients. T2-weighted magnetic resonance lymphangiography is an imaging modality of increasing importance both to identify the risk status before Fontan palliation and to reveal lymphatic complications that may occur in the postoperative period. In this article, we present the management of a pediatric case who was followed and treated for prolonged chylothorax after Fontan palliation and discuss the use of magnetic resonance lymphangiography in this process.

Keywords: Fontan palliation, lymphangiography, lymphatic complications, magnetic resonance imaging.

In proportion to the increasing life expectancy of patients with a single ventricle, the complications observed after single-ventricle palliation also increase in inverse proportion to the mortality rate.^[1] The rise of central venous pressure following the Fontan procedure increases lymphatic fluid production but complicates lymphatic circulation. In this context, the dilatation of lymphatic structures disturbs the lymphatic pump function and results in the occurrence of early and late complications such as pleural effusion, plastic bronchitis, and protein-losing enteropathy.^[2] In general, T2-weighted magnetic resonance lymphangiography (MRL) plays a significant role in the prediction and follow-up of these complications.^[3]

ÖZ

Tek ventrikül hastalarında lenfatik komplikasyonlar, artmış morbidite ve mortalite ile ilişkilidir. T2 ağırlıklı manyetik rezonans lenfanjiyografi hem Fontan palyasyonu öncesi risk durumunu belirlemek, hem de ameliyat sonrası dönemde oluşabilecek lenfatik komplikasyonları ortaya koymak konusunda önemi her geçen gün artan bir görüntüleme yöntemidir. Bu makalede, Fontan palyasyonu sonrası uzamış şilotoraks nedeniyle takip ve tedavi edilen pediatrik bir olgunun yönetimi sunuldu ve bu süreçte manyetik rezonans lenfanjiyografinin kullanımı tartışıldı.

Anahtar sözcükler: Fontan palyasyonu, lenfanjiyografi, lenfatik komplikasyonlar, manyetik rezonans görüntüleme.

In this article, we present the management and follow-up processes of a pediatric case followed for prolonged chylothorax after the Fontan procedure.

CASE REPORT

A three-year and seven-month-old male patient infant weighing 21 kg with transposition of the great arteries, large ventricular septal defect, pulmonary stenosis, left ventricular hypoplasia underwent a patent ductus arteriosus (PDA) stent and balloon atrial septostomy at the age of six days and a Glenn shunt procedure at the age of 3.5 months. He was admitted to the ward for preparation before the Fontan palliation. In routine T2-weighted MRL performed for evaluation before Fontan palliation,

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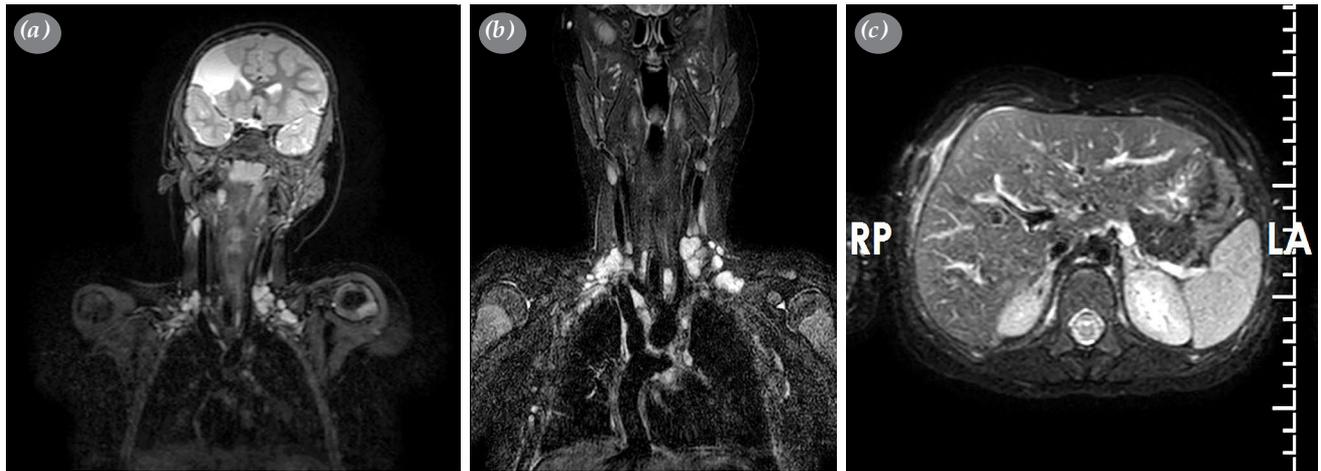


Figure 1. (a) T2-weighted imaging shows abnormally increased lymphatic channels in the supraclavicular region that do not extend into the mediastinum. (b) Lymphatic expansions reaching both supraclavicular regions and the mediastinum as seen by T2-weighted magnetic resonance imaging lymphangiography. (c) T2 hyperintensity signal changes seen in axial cross-section in pleura and retroaortic region.

the case was evaluated as type 2 according to lymphatic malformation grading (Figure 1a).^[4] In the angiographic evaluation, the mean pulmonary artery pressure was 15 mmHg, pulmonary vascular resistance index (PVRI) was 1.11 WU/m², systemic vascular resistance index (SVRI) was 23.39 WU/m², and PVR/SVR ratio was 0.05. Echocardiographic evaluation revealed no valvular regurgitation. Systemic ventricular ejection fraction was 66%. An extracardiac Fontan operation with 18-mm polytetrafluoroethylene (PTFE) tubular graft was

decided. The pulmonary artery, Fontan, and Glenn pressures measured during surgery were 14 mmHg, 19 mmHg, and 20 mmHg, respectively, and it was decided to perform a fenestration. Fenestration was performed with a 4.8-mm punch. In the postoperative follow-up, the pleural drainage was more than 5 mL/kg/h and the drainage turned into chylous fluid after Day 4. On Day 7, long-chain fatty acids were removed from the patient's diet, and total parenteral nutrition and a diet containing medium-chain triglycerides were initiated. On Day 10, somatostatin

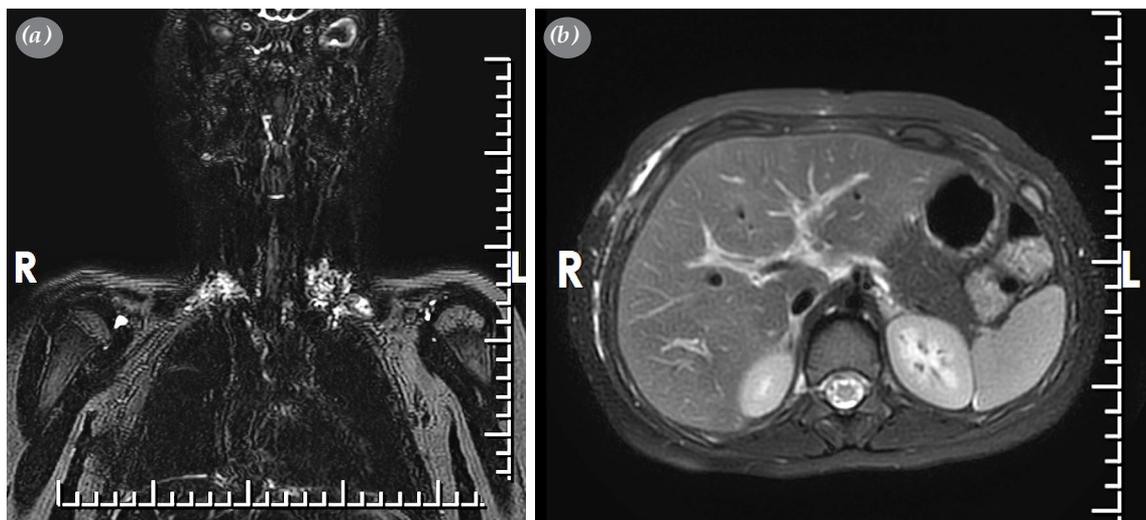


Figure 2. (a) T2-weighted magnetic resonance imaging results obtained after the fenestration dilatation process revealed that the lymphatic changes had disappeared. (b) The signal changes in the pleura and retroaortic region had significantly regressed.

infusion was started while drainage continued. On Day 15, it was decided to perform a control MRL due to persistent chylothorax. The MRL revealed that the lymphatic malformation grading, evaluated as type 2 in the preoperative period, progressed to type 3. Furthermore, T2 hyperintensity signal changes were observed in the portal hilus, around the duodenum toward the mesenteric root, and in the retroaortic region in sections passing through the upper abdomen (Figures 1b, c). These findings were attributed to an increase in Fontan pressure and the patient underwent diagnostic angiography, which revealed pulmonary artery pressure of 19 mmHg, pulmonary vascular resistance index value of 1.62, high levels of WU/m² and Fontan pressure (19 mmHg) and narrowing of the released fenestration. A 5 to 6-mm fenestration was thought to be sufficient, and the fenestration was expanded with a 5×20 mm coronary balloon (Shunmei®, China). After that procedure, the Fontan pressure decreased to 17 mmHg, saturation dropped from 95 to 92%, and the chylous drainage was drastically reduced. The Fontan diet was resumed. On Day 3, the thorax tube was removed. The patient was discharged six days after the procedure and returned one month later for follow-up MRL. As a result of that imaging, the lymphatic malformation grade was type 2, and the pleural and pericardial effusions and T2 hyperintensity signal changes previously seen in the abdomen disappeared (Figures 2a, b). A written informed consent was obtained from the parent of the patient.

DISCUSSION

In recent years, T2-weighted MRL is an increasingly important imaging method for evaluating the preoperative risk status of patients with a single ventricle and detecting possible lymphatic anomalies and postoperative complications.^[3-5] Biko et al.^[4] categorized lymphatic distributions ranging from 1 to 4 in a study which aimed to reveal the relationship between the T2-weighted MRL results and surgical results of patients who underwent the Fontan procedure. Types 1 and 2 were classified as being of low risk and types 3 and 4 as high risk in terms of lymphatic complications.^[4]

In our clinic, we perform T2-weighted MRL for every patient for whom the Fontan procedure is planned. The lymphatic anatomy can be clearly visualized with STIR and iso-two-dimensional (2D) sequences, particularly for the neck and upper thoracic region. However, the evaluation of lymphatic channel system in these regions is only possible with

two-dimensional FSE T2-weighted imaging, which can be performed with thin-section coronal and axial orientations in the thoracic and abdominal regions.

Subsequent management of patients identified as high-risk by MRL may include further evaluation of lymphatic abnormalities, close monitoring of clinical and hemodynamic status, reassessment of the timing of Fontan surgery, consideration of alternative treatment options (such as transplantation) and potential lymphatic interventions. Management decisions should be made in a multidisciplinary approach based on the individual situation of each patient.^[4,5] As a matter of fact, although the patient was evaluated as low risk in the MRL evaluation performed before the Fontan operation in our patient, it was thought that the reason for the chylothorax that started on postoperative Day 4 and did not respond to medical treatments was the narrowing in the fenestration. The fact that the findings regressed in the MRL evaluation performed after balloon dilatation procedure to the fenestration also supported this.

Previous studies have usually focused on the long-term follow-up and complications of single-ventricle palliation; however, no data could be found in the literature regarding the follow-up and management of early-period complications via T2-weighted MRL.^[3,4] The clear differences between our patient's pre-, postoperative, and post-discharge lymphangiography images confirm that T2 MRL is an effective and valuable imaging method for both early-period and late-period lymphatic complications after single-ventricle palliation.

In conclusion, T2-weighted magnetic resonance lymphangiography proved to be a valuable tool not only in preoperative risk stratification but also in the early identification and management of postoperative lymphatic complications following Fontan palliation. Its use may enhance clinical decision-making and improve patient outcomes when integrated into a multidisciplinary care approach.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Data collection and/or processing, literature review, writing the article: D.K.; Idea/concept, analysis and/or interpretation, materials: S.B.; Design: İ.C.T.; Critical review: A.C.H.; Control/supervision, references and fundings: E.Ö.

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REFERENCES

1. Rychik J, Atz AM, Celermajer DS, Deal BJ, Gatzoulis MA, Gewillig MH, et al. Evaluation and management of the child and adult with fontan circulation: A scientific statement from the American Heart Association. *Circulation* 2019;140:e234-84. doi: 10.1161/CIR.0000000000000696.
2. Savla JJ, Kelly B, Krogh E, Smith CL, Krishnamurthy G, Glatz AC, et al. Occlusion pressure of the thoracic duct in fontan patients with lymphatic failure: Does dilatation challenge contractility? *World J Pediatr Congenit Heart Surg* 2022;13:737-44. doi: 10.1177/21501351221119394.
3. Kelly B, Mohanakumar S, Ford B, Smith CL, Pinto E, Biko DM, et al. Sequential MRI evaluation of lymphatic abnormalities over the course of fontan completion. *Radiol Cardiothorac Imaging* 2024;6:e230315. doi: 10.1148/ryct.230315.
4. Biko DM, DeWitt AG, Pinto EM, Morrison RE, Johnstone JA, Griffis H, et al. MRI evaluation of lymphatic abnormalities in the neck and thorax after fontan surgery: Relationship with outcome. *Radiology* 2019;291:774-80. doi: 10.1148/radiol.2019180877.
5. Collins JD, Thompson SM. Noncontrast MR Lymphangiography to identify progression of lymphatic abnormalities over the course of fontan completion. *Radiol Cardiothorac Imaging* 2024;6:e240201. doi: 10.1148/ryct.240201.