

**Treatment of persistent chylothorax after surgery for heterotaxy syndrome**

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**OBJECTIVE:** Chylothorax is a serious complication after congenital heart surgery and it is mostly encountered after univentricular repair. Although conservative management usually enough for treatment, some patients may need surgical intervention. Ductus thoracicus ligation with right thoracotomy is the preferred approach. Heterotaxy syndrome is a lateralization disorder and there is no data about the localization of the ductus, which makes a confusing situation in those patients who need surgical intervention due to persistent chylothorax.

**METHODS:** Three patients who had univentricular heart and heterotaxy syndrome underwent cavopulmonary anastomosis (n=2) or Kawashima operation (n=1). Their ages were 7 months, 3 years and 1,5 years respectively. One patient had previous modified BT shunt and one patient had bilateral banding because of associated aortic arch hypoplasia as a first stage palliation. Dextrocardia was present in two of the patients. Chylothorax developed at 15 days after the operation in 1 patients who underwent Kawashima operation. Others readmitted to the hospital due to chylothorax two weeks after uneventful postoperative period and hospital discharge. Despite maximum medical treatment including high dose of octreotid infusion, high amount of chylouse drainage (>30 ml/day) persisted more than two weeks in all patients. Two patients with right or left sided unilateral persistent chylothorax underwent surgery. Ductus could not be localized and decortication and talk application of the ipsilateral side was performed. MR lymphangiography was done for the last patient who had bilateral persistent drainage to detect the ductus location for proper intervention.

**RESULTS:** Two patient responded well for surgical intervention. MR lymphangiography revealed both sided, but left dominant thoracic duct in the other patient. Surprisingly, chylothorax decreased dramatically after MR study in this patient. We speculated that lymphatic vessels might be occluded by contrast agent Lipiodol.

**CONCLUSIONS:** Persistent chylothorax in patients with heterotaxy syndrome is difficult to manage, because of the unknown location of the thoracic duct. Decortication and talk application for the ipsilateral thoracic cavity may be an effective intervention. MR lymphangiography might be helpful for ductus localization in patients with bilateral persistent drainage.