Intralobar pulmonary sequestration associated with left main coronary artery obstruction and mitral regurgitation: A case report

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Abstract

A 4-year-old boy with left intralobar pulmonary sequestration associated with left main coronary artery obstruction (LMCAO) and severe mitral regurgitation (MR) was admitted to our hospital. Since the patient presented with dyskinesia of the cardiac apex and increased left ventricular end-diastolic volume (LVEDV), left main coronary artery reconstruction and mitral annuloplasty were performed. The enlargement of the left ventricle was improved after sequential surgeries._There was a risk of deterioration of MR and regrowth of LVEDV due to shunt blood flow; therefore, left lower lobectomy and aberrant artery division were performed. This is a very rare case of a patient with pulmonary sequestration associated with LMCAO and severe MR.

Introduction

Pulmonary sequestration accounts for approximately 5% of congenital lung diseases. The shunt volume due to aberrant arteries is usually mild; however, sometimes, there are cases of increased shunt volume causing left ventricular volume overload and symptoms of heart failure.[1-6] Extralobar pulmonary sequestration is often associated with malformations of other organs; however, intralobar pulmonary sequestration is rare. Herein, we report the case of a patient with left main coronary artery obstruction (LMCAO), severe mitral regurgitation (MR), and left intralobar pulmonary sequestration, who was successfully treated with sequential surgical treatment.

Case

A 4-year-old boy was diagnosed with acute pneumonia, and a dilated heart shadow was found on X-ray. Physical examination revealed Levine II/VI systolic murmurs. Laboratory investigations showed a high brain natriuretic peptide (BNP) level (96.5 pg/mL). Computed tomography (CT) showed an overinflated area and multicystic changes in the left lower lobe of the lungs (Figure 1a). Three-dimensional CT and cardiac catheterization showed an aberrant artery arising from the descending aorta (Figures 1b and c). Blood from the sequestration returned to the left atrium via the left inferior pulmonary vein (Figures 1b and d). No connection between the normal bronchus and the sequestrated lungs was observed.

Therefore, the patient was diagnosed with intralobar pulmonary sequestration. Transthoracic echocardiography (TTE) showed dyskinesia of the cardiac apex and severe MR. Cardiac catheterization revealed systemic blood flow, pulmonary artery wedge pressure, left ventricle pressure, end-diastolic pressure, left ventricular end-diastolic volume (LVEDV), and left ventricular ejection fraction of 4.2 mL/min, 14 mmHg, 101 mmHg, 9 mmHg, 306% of normal, and 44%, respectively. Moreover, a grade III MR was revealed. Coronary angiography (CAG) showed 99% occlusion of the left coronary artery and severe stenosis of the main trunk, and it was diagnosed as LMCAO (Figure 2a). The anterior descending and circumflex branches were imaged in a retrograde manner through the collateral route from the right coronary artery. The undeveloped coronary arteries from the apex to the lateral wall were also imaged (Figure 2b).

Left main coronary artery reconstruction and semicircular mitral annuloplasty were performed for LMCAO, left ventricular dysfunction, and MR (Figure 3). The intraoperative findings confirmed that blood flow from the left inferior pulmonary vein was abundant. Left main trunk (LMT) incision was performed, an inverted U-shaped incision was made in the left sinus of Valsalva, a flap was anastomosed to the anterior wall of the LMT, and the remaining anterior LMT plasty was performed with a 12 × 8 mm trapezoidal main pulmonary artery wall patch. The intraoperative findings showed a prolapse in the A2 and A3 of the mitral valve. The posterior leaflet was extended throughout. Semicircular mitral annuloplasty was performed according to the size of the anterior leaflet. The defect of the main pulmonary artery was repaired with autologous pericardium. The patient left the intensive care unit the day after surgery and was discharged from the hospital on the eleventh postoperative day.

There was a risk of deterioration of MR and regrowth of LVEDV due to excess shunt blood flow; therefore, early surgical intervention for pulmonary sequestration was scheduled. Thoracoscopic left lower lobectomy and aberrant artery division were performed 65 days after the heart surgery. The intraoperative findings confirmed an abnormal blood vessel branching from the descending aorta and draining via the inferior pulmonary vein. The patient successfully recovered and was discharged from the hospital on the sixth postoperative day. Written informed consent was obtained from this patient's parents during both surgeries.

TTE 2 days and 10 months after left lower lobectomy and aberrant artery division suggested that the remaining moderate MR was probably due to papillary muscle dysfunction. A remarkable regression of the left ventricular dilation was confirmed, and the reduced wall motion of the apex also improved (Figure 4). Cardiac catheterization performed 1 year after surgery revealed that no stenosis was observed in the left main coronary artery, left anterior descending artery, and left circumflex artery and anterograde development of the coronary artery from the apex to the lateral wall that was scarce before surgery (Figure 2c). Further, no collateral vessels originating from the right coronary artery were noted (Figure 2d). The pulmonary artery pressure and pulmonary artery wedge pressure improved, suggesting an improvement in cardiac function. In terms of papillary muscle dysfunction, the MR is currently being treated with angiotensin-converting enzyme inhibitors to prevent left ventricular enlargement. The pediatric post-repair moderate MR will be improved as the left ventricle will be developed by remodeling in the future because there is no residual coaptation failure of the mitral valve.

Discussion

Pulmonary sequestration is a rare congenital malformation characterized by nonfunctioning lung tissue separated from the normal lung tissue and fed by an aberrant artery.[7)] There is no clear traffic between the normal bronchus and the sequestrated lung. However, it has been reported that emphysematous changes occurred in both the sequestrated lung and the adjacent normal lung, and that traffic was caused by abnormal bronchi and fistulas.[8)] It is probable that the hyperinflated region was generated by the same mechanism in this case as well. Intralobar pulmonary sequestration is associated with malformations of other organs in 14% of the cases and cardiac malformations, including macrovasculature in 2.0%.[9)] Three cases of intralobar pulmonary sequestration associated with heart malformation that were performed lobectomy were reported, although none with LMCAO was found. A 25-year-old man diagnosed with aortic valve stenosis (bicuspid valve) 5 months after birth underwent aortic commissurotomy.

However, intralobar pulmonary sequestration was not diagnosed, and congestive heart failure developed. Left lower lobectomy was performed, following which the clinical symptoms improved.[10] A 2-month-old infant presented with cyanosis and severe respiratory failure. Scimitar syndrome, patent ductus arteriosus, and intralobar pulmonary sequestration of the right lower lobe were detected, and the patient underwent right lower lobectomy at 8 years of age.[11] In another report, a newborn baby had tetralogy of Fallot and intralobar pulmonary sequestration. Right lower lobectomy was performed at 15 months.[11] In the present case, it was considered that excess shunt blood flow from the sequestrated lungs exacerbated MR and left ventricular dysfunction in addition to LMCAO. Since coil embolization to treat pulmonary sequestration has been reported, there may have been the option of preoperative coil embolization.[12]

Echocardiography revealed persistent MR; however, left ventricular end-diastolic diameter, left ventricular end-systolic diameter, and the serum level of BNP improved after left main coronary artery reconstruction and mitral valve annuloplasty, followed by left lower lobectomy and ligation of the aberrant artery (Figure 3). Although cardiac surgery significantly contributed to the left cardiac blood supply and preload, it is possible that the ligation of the aberrant artery and the resection of a sequestrated lung contributed to the improvement of cardiac functions and prevented future exacerbations of heart failure.

Since multiple surgeries are burdensome in children, pulmonary surgery can be performed at the same time as cardiac surgery. However, it is necessary to change the operative position during surgery. The risk of lengthy surgery, postoperative bronchial fistula after lobectomy, and the possibility of infection, particularly in the sternum, may be increased. Furthermore, LMCAOs have an early critical stage in infancy. In the left main coronary artery atresia, the collateral blood flow from the apex to the lateral wall might not keep up with the rapidly growing myocardial muscles, resulting in myocardial ischemia.[13)] Similar outcomes are possible in this case. In the present case, the patient had a lower limit of normal cardiac functions. Therefore, we believe that the timing of sequential surgeries was appropriate.

Conclusion

After sequential surgeries, including left main coronary artery reconstruction and mitral valve angioplasty followed by left lower lobectomy and ligation of the aberrant artery, the enlargement of the left ventricle was improved. It is expected to contribute to the prevention of future exacerbations of left heart failure.

Compliance with ethical standards

Conflict of interest The authors have declared that no conflict of interest exists.

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Figure Legends

Figure 1. Chest CT and cardiac catheterization of the sequestrated lungs. **a)** An overinflated area and multicystic changes in the left lower lobe. **b)** An aberrant artery arising from the descending aorta (red arrow) and blood flow from this area returning to the left inferior pulmonary vein (blue arrow). **c, d)** An aberrant artery arising from the descending aorta (red arrowheads) and blood flow from the sequestrated lungs to the inferior pulmonary vein (blue arrowheads). CT: computed tomography

Figure 2. Pre- and postoperative cardiac catheterization of the bilateral coronary arteries. **a)** Preoperative left CAG. The left coronary artery was 99% occluded and severely stenotic in the main trunk (red arrow). **b)** Preoperative right CAG. The anterior descending and circumflex branches were retrogradely imaged via the collateral route (yellow arrows). **c)** Postoperative left CAG showing the development of the left anterior descending and circumflex arteries. **d)** Postoperative right CAG. No collateral arteries perfused the lesion from the apex to the lateral wall. CAG: coronary angiography

Figure 3. Left main coronary artery reconstruction and semicircular mitral annuloplasty were performed for LMCAO, left ventricular dysfunction, and MR. **a**) LMT incision was performed, an inverted U-shaped incision was made in the left coronary sinus, and a flap was

anastomosed to the anterior wall of the LMT. **b**) The remaining anterior LMT plasty was performed with a main pulmonary artery wall patch. LMCAO: left main coronary artery obstruction; LMT: Left main trunk

Figure 4. Postoperative evaluation of cardiac functions. The serum BNP levels along with LVDd and LVDs (estimated by echocardiography) were improved by sequential surgical interventions. BNP: brain natriuretic peptide; LMT: left main trunk; LVDd: left ventricular end-diastolic diameter; LVDs: left ventricular end-systolic diameter

Fig. 1

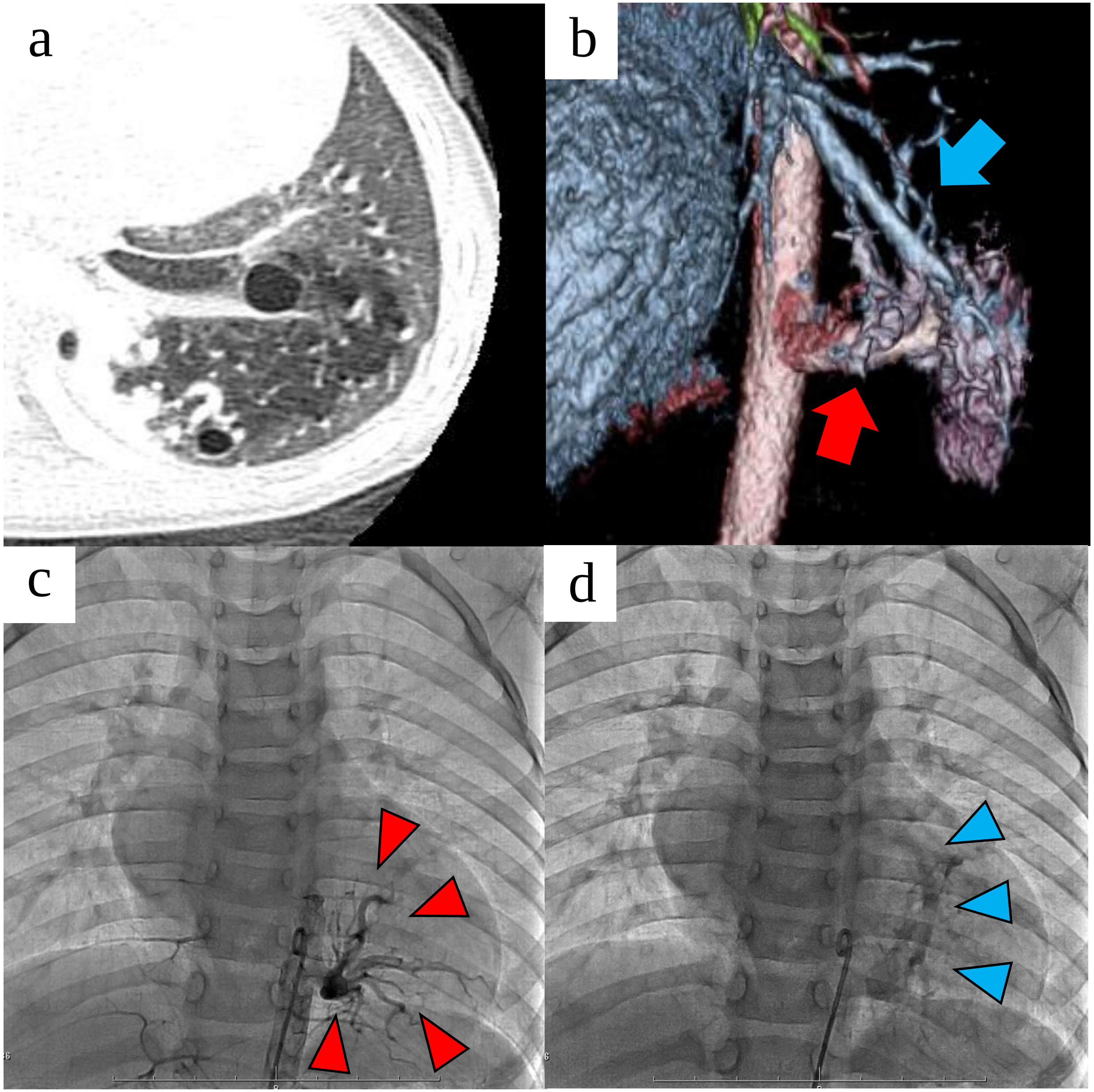


Fig. 2

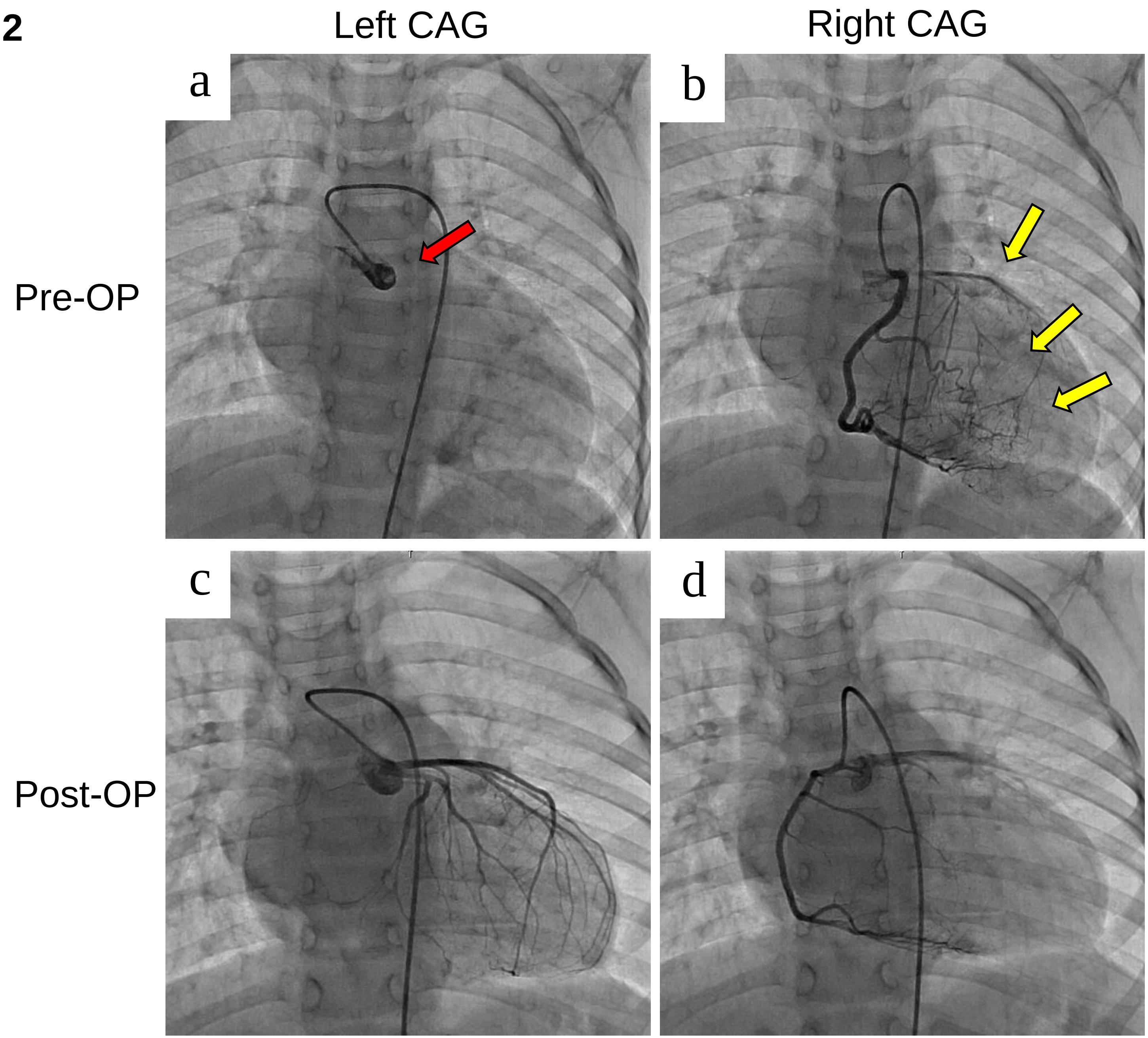
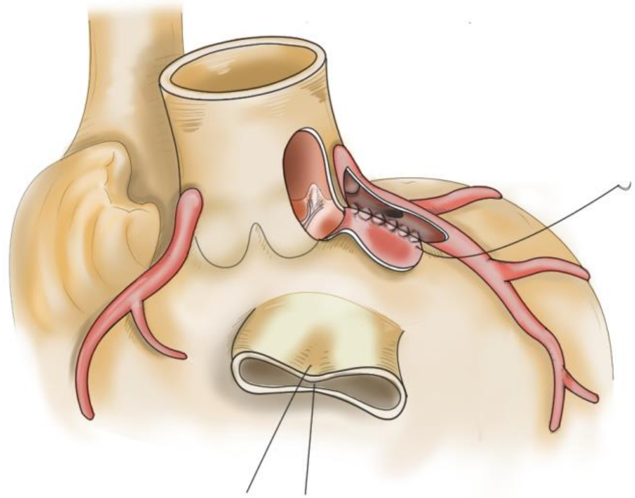


Fig. 3

a



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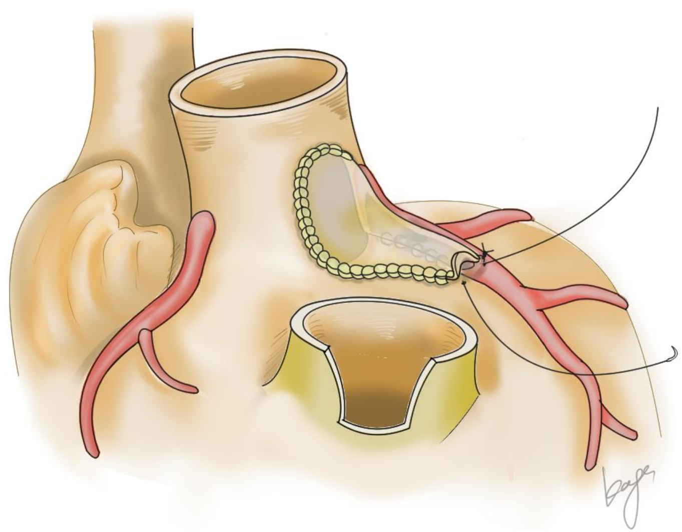
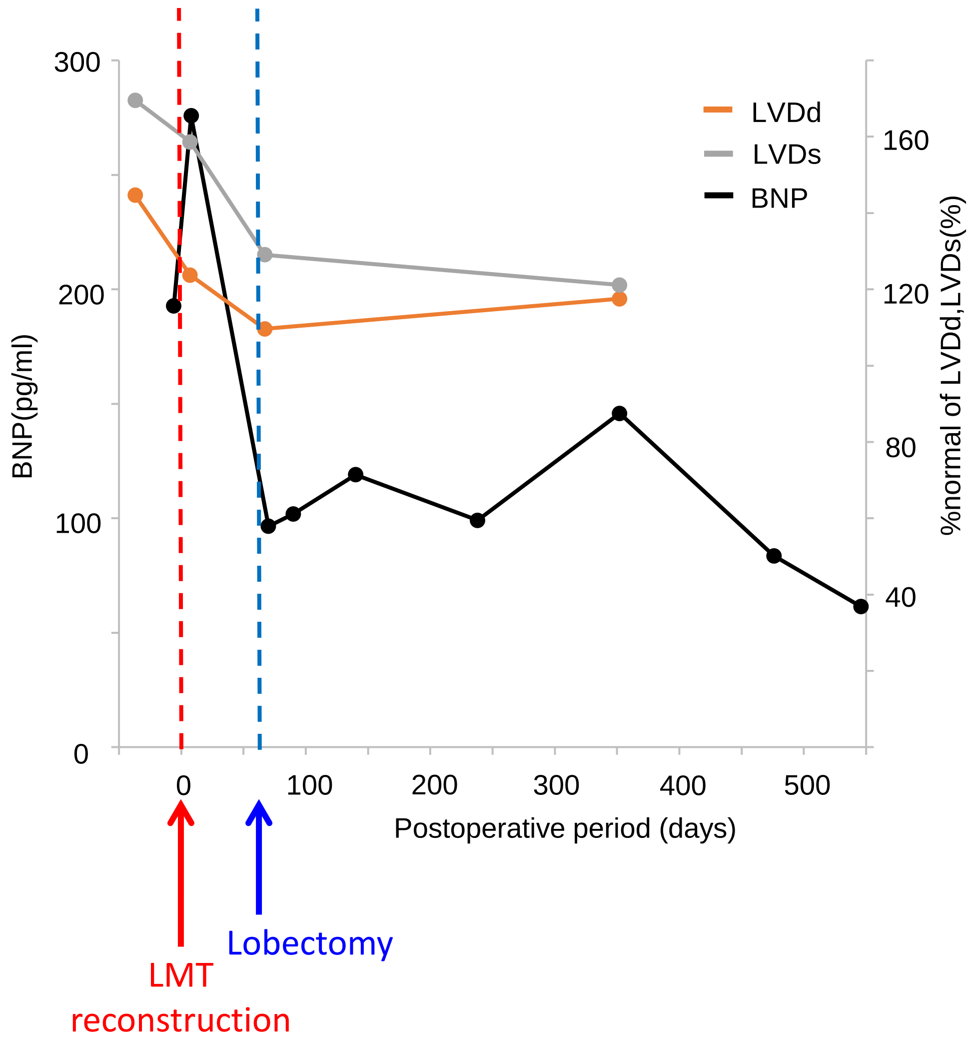


Fig. 4


300
200
100
0
BNP(pg/ml)
LVDd
LVDs
BNP
160
120
80
40
%normal of LVDd,LVDs(%)
0
100
200
300
400
500
Postoperative period (days)
Lobectomy
LMT
reconstruction