Infantile lobar emphysema is a rare congenital anomaly that often requires lobectomy in early infancy.\(^1\) Although the etiology of the condition is most commonly idiopathic, an association has been found between infantile lobar emphysema and congenital heart disease.\(^2\)–\(^4\) We encountered an infant who had ventricular septal defect (VSD) and atrial septal defect (ASD) and infantile emphysema of the right upper lobe. The infant was intubated due to respiratory distress and thereafter was dependent upon mechanical ventilation. We successfully performed a total correction of the cardiac anomalies combined with a right upper lobectomy. The resection of the right upper lobe was performed under cardiopulmonary bypass through a median sternotomy. There was no major airway bleeding or mediastinitis. The patient was extubated on postoperative day 12 and discharged on postoperative day 29.

**Keywords:** congenital heart disease, infantile lobar emphysema, concomitant operation

### Introduction

Infantile lobar emphysema is a rare congenital anomaly that often requires lobectomy in early infancy.\(^1\) Although the etiology of the condition is most commonly idiopathic, an association has been found between infantile lobar emphysema and congenital heart disease.\(^2\)–\(^4\)

We encountered an infant who had ventricular septal defect (VSD) and atrial septal defect (ASD) and infantile emphysema of the right upper lobe. The infant was intubated due to respiratory distress and thereafter was dependent upon mechanical ventilation. We successfully performed a total correction of the cardiac anomalies concomitant with lobectomy of the right upper emphysematous lobe.
86.0%; left, 91.0%. There were no pulmonary arteriovenous fistulae. Because the saturation of the left pulmonary vein was relatively low, we concluded that closure of VSD and ASD under cardiopulmonary bypass (CPB) entailed a high risk, and postponed surgical intervention.

However, he was intubated due to respiratory distress 11 days after cardiac catheterization, and the hyperinflation of the right emphysematous lobe worsened (Fig. 1C) and he became dependent on the mechanical ventilation. Cardiac catheterization (under controlled ventilation) was performed for reevaluation (4 weeks after the first catheterization). It revealed decreased pressure in the pulmonary artery (58/17 mmHg) and an increased left-to-right shunt (Qp/Qs: 6.1). Saturations of the pulmonary veins were as follows: right upper, 89.5%; right lower, 91.9%, left, 98.4%. At this point, we decided that he could tolerate a cardiac operation.

When he was five months old (weight, 4.8 kg), cardiac surgery was performed through a median sternotomy. He was placed on CPB in the standard fashion using ascending aortic and bicaval cannulation. After ligation of the ductus arteriosus, we performed surgical closure of the VSD and ASD under cardiac arrest. After the cardiac repairs, a thoracic surgery team removed the right upper emphysematous lobe through the same median sternotomy under CPB with a beating heart. The resection of the lobe was performed without difficulty through a pleurotomy (without performing a thoracotomy or single-lung
VSD with Infantile Lobar Emphysema

ventilation). The duration of CPB and aortic cross-clamping was 192 and 47 min, respectively. After CPB was discontinued, bronchoscopy was performed, and we confirmed that there was no major airway bleeding.

Postoperatively, it took several days to re-expand the compressed right middle and lower lobes, and he was successfully weaned from nitrogen monoxide on postoperative day 6, and extubated on postoperative day 12. There were no pulmonary hypertensive episodes or mediastinitis. Postoperative echocardiography revealed good cardiac function and persistent mild pulmonary hypertension. He was discharged 29 days after the surgery with oxygen at 1 L/min for the treatment of pulmonary hypertension. A review at 6 months post-surgery showed that he had gained another 2 kg, and a chest X-ray showed normal bilateral lung fields.

Discussion

Infantile lobar emphysema is a rare anomaly of lung development that usually presents with respiratory distress and pulmonary lobar hyperinflation. Almost all cases are diagnosed within the first 6 months of life irrespective of any associated lesions. Conservative or non-operative treatment is possible in asymptomatic or mildly symptomatic cases, however, lobectomy is the universally accepted treatment in moderately or severely symptomatic cases.

Congenital heart disease can be associated with infantile lobar emphysema, however, there has been some debate as to the appropriate management of these infants regarding whether the correction of the cardiac defect should be performed first, or whether the lobectomy should be performed first, or whether they should be performed simultaneously.

In the present patient, preoperative mechanical ventilation aggravated the lobar emphysema. Based on this, we judged that he would be dependent upon mechanical ventilation after cardiac surgery unless a lobectomy was performed. Moreover, the second cardiac catheterization revealed pulmonary hypertension with pulmonary overcirculation. Therefore, it was concluded that circulatory management might be difficult following a lobectomy without a cardiac correction. It may be argued that pulmonary artery banding concomitant with a lobectomy might be less invasive than a total correction with a lobectomy. However, a total correction of cardiac anomalies concomitant with a lobectomy simplifies the postoperative management.

Although a thoracotomy is generally the preferred treatment for infantile lobar emphysema, a median sternotomy with CPB makes it possible to decompress the lung, and to resect the emphysematous lobe without the need for single lung ventilation or an additional thoracotomy. To the best of our knowledge, such a concomitant surgery has not been reported previously.

The advantages of this concomitant surgery are as follows: (1) only one midline skin incision is needed and single-lung ventilation is unnecessary; (2) postoperative circulatory and respiratory management is simplified. However, this procedure has some disadvantages: lobectomy performed under CPB entails a considerable risk of mediastinitis due to concomitant lung resection, and airway bleeding due to heparinization. In our patient, sputum culture was negative, therefore we judged the risk of postoperative mediastinitis to be relatively low. In terms of airway bleeding, if it was difficult to control intraoperatively, we planned to use extracorporeal membrane oxygenation until the bleeding was controlled.

Conclusion

In conclusion, we successfully performed a total correction of VSD and ASD concomitant with lobectomy of a right upper emphysematous lobe through a median sternotomy.

Disclosure Statement

None declared.

References