FIGURE 1. A, Initial preoperative angiogram showing no significant disease in the left coronary tree. B, Angiogram on presentation showing severe slit-like narrowing of the proximal left anterior descending artery (*arrow*) and total occlusion of the left circumflex artery (*broken arrow*). C, Post-intervention angiogram. D and E, Computed tomographic angiography of consecutive sagittal sections of the mediastinum after stenting, demonstrating the lower pseudoaneurysm (*broken arrow*) impinging on the left main artery (D, *arrow*) and proximal left anterior descending artery (E, *arrow*). The upper pseudoaneurysm (*asterisk*) is also seen in both sections.

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Persistent interstitial pulmonary emphysema requiring pneumonectomy

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Persistent interstitial pulmonary emphysema (PIPE) is a rare but serious disorder of neonates characterized by abnormal accumulation of air in the pulmonary interstitium. Although localized PIPE has the potential for curative resection, generalized PIPE affecting both lungs carries a 100% mortality.¹ Classically associated with antecedent mechanical ventilation, it can occur in the absence of prior ventilatory support.²

CLINICAL SUMMARY

A growth-restricted (0.4 growth percentile) male twin was delivered by means of caesarian section at 36 weeks and 2 days' gestation. He required feeding and temperature support but no ventilatory assistance. On day 14 after delivery, he showed signs of respiratory distress. Chest radiographic analysis showed hyperinflation of the left lung with cystic air spaces and contralateral shift of the mediastinum. Computed tomographic (CT) analysis of the chest confirmed hyperinflation and multiple cystic air spaces throughout the left lung. The right lung (Figure 1, A) showed marked compression by the displaced mediastinum. The differential diagnosis was believed to be either congenital cystic adenomatoid malformation (CCAM) or congenital lobar emphysema. Oxygen (fraction of inspired oxygen, 0.3), physiotherapy, and intravenous antibiotics were instituted.

On day 22, the child's condition deteriorated, with increasing fraction of inspired oxygen requirements and signs of respiratory distress requiring intubation and ventilation. Selective intubation of the right main bronchus was undertaken to prevent further distension of the left lung, and he was referred for surgical management.

Rigid bronchoscopy was undertaken to exclude a bronchial cause; however, this showed normal anatomy. A size 5 Fogarty catheter was used as a left bronchial blocker. At left thoracotomy, the lung was bilobed; however, the parenchyma was diffusely cystic, interspersed only with small areas of normal parenchyma. The islands of normal lung parenchyma were nonsegmental in nature, and pneumonectomy was performed.

Histologic examination showed preservation of bronchial anatomy and normal alveolar density, with severe interstitial emphysema in both lobes comprising numerous dilated air-filled spaces expanding the interstitium, with some air-filled

spaces lined by a histiocytic reaction indicating chronicity of changes and therefore a diagnosis of PIPE (Figure 2).

The patient was extubated on postoperative day 3. Two days after extubation, he showed signs of respiratory distress. Chest radiographic analysis showed right upper lobe collapse, and noninvasive continuous positive pressure ventilation was commenced. By day 7 postoperatively, a CT scan of the chest showed the right lung to be fully inflated with cystic air spaces, which is consistent with PIPE (Figure 1, B). He responded to conservative management, and by day 23 postoperatively, his right lung was radiographically normal, and he continues to thrive 9 months postoperatively.

DISCUSSION

PIPE is a rare but potentially fatal disorder of neonates characterized by multiple air-filled cysts within the interstitium of the lung thought to result from rupture of the alveolus at its junction with perivascular connective tissue.³

The differential diagnosis is that of neonatal cystic lung disease, including CCAM and congenital lobar emphysema. PIPE is characterized on CT scanning by characteristic lines and dots within a radiolucent mass representing extensive cystic air spaces, whereas CCAM is characterized by large, air-filled, multiloculated cysts, usually with a soft tissue mass-like or nodular component. The radiologic features of CCAM are highly variable and can include multiple small cysts with focal areas of emphysematous change, making preoperative diagnostic distinction difficult.

Despite its association with prior mechanical ventilatory support, there are numerous reports of its occurrence without prior ventilatory support.² The Valsalva maneuver associated with crying is associated with high airway pressures. In a study of maximal airway pressures in crying neonates,

FIGURE 1. A, Enhanced computed tomographic scan showing a markedly inflated left lung. The entire upper lobe and most of the lower lobe appear as cystic air spaces, with marked displacement of the mediastinum to the right and compression of the right lung. The right lung, apart from compression-related effects, appeared structurally normal, with no identifiable cystic air spaces. B, Computed tomographic scan 13 days after left pneumonectomy. The inflated right lung now shows cystic air spaces similar in nature to the preoperative left lung, which is consistent with persistent interstitial pulmonary emphysema.

FIGURE 2. A, Macroscopic examination of the lung showing normal bronchial anatomy. There was diffuse multicystic change throughout the parenchyma, with the largest cyst measuring approximately 20 mm. B, Microscopic sections showing dilated air-filled spaces lying within the interstitium. Histiocytic giant cells lined many of the cystic spaces, indicating a degree of chronicity of approximately 7 days or more.

the mean expiratory pressure was 53 cm H₂O,⁴ and we suggest that this mechanism might account for its development in infants in the absence of ventilatory support. The development of PIPE at low mean airway pressures is postulated to reflect increased sensitivity of the underdeveloped lung to stretch.⁵ The misconception that mechanical ventilation is a prerequisite for the development of PIPE might lead to delayed diagnosis or misdiagnosis.

Diffuse PIPE affecting multiple lobes carries a poor prognosis.¹ Nonoperative management includes selective bronchial intubation and lateral decubitus positioning. Surgical resection of affected areas should be undertaken to prevent the cycle of positive pressure damage in those with disease progression. In a series of infants with PIPE, 12 of 22 patients had diffuse disease affecting both lungs. None underwent resection, and the mortality of this group was 100%.¹ However, survival is reported after treatment with partial liquid ventilation after failure of conventional management.⁶

PIPE should be considered as a differential diagnosis in neonates with respiratory distress, irrespective of preceding

mechanical ventilatory support. Early surgical intervention should be considered in individuals who have progressive disease. This is, to our knowledge, the first report of survival after the treatment of generalized PIPE by means of pneumonectomy.

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