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Bilateral congenital lobar emphysema: a rare entity and a therapeutic challenge

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Abstract

Background: Bilateral congenital lobar emphysema is an extremely rare condition, and detailed workup of these patients is very important. Operative intervention is usually based on radiological, biochemical, and clinical details. Bronchoscopy gives an added advantage in cases with diagnostic dilemmas and makes the treatment more evidence based. We here present three cases and review the literature for this rare entity.

Material and methods: We retrospectively collected data about all the cases of bilateral CLE operated by the authors at various centres over the period of 3 years from January 2016 to December 2018.

Results: We managed three cases of bilateral CLE, and all three had unique details based on which treatment was planned. We operated three cases of this very rare entity and compared our findings with 23 cases from the literature reviewed.

Conclusion: We propose that bronchoscopy should be an essential component in the management of all cases of CLE. It helps in defining pathology, the severity of the disease, and the decision on which side to be operated first and avoids unnecessary simultaneous lobectomies. We recommend case selection on basis of radiological, biochemical, clinical, and bronchoscopy criteria. Based on this, an approach of sequential lobectomies is less risky and should be advocated in most of the patients.

Keywords: Bilateral emphysema, Congenital lobar emphysema, Posterolateral thoracotomy

Background

Congenital lobar emphysema (CLE) is a rare developmental anomaly of the lung characterized by over-distension and air trapping in the affected lobe secondary to bronchomalacia or absent cartilage [1–16]. It presents as respiratory distress due to ventilation-perfusion mismatch because of compression atelectasis on the ipsilateral or the contralateral side [1–4]. Bilateral involvement is exceptionally rare and has been reported only in 13 scattered case reports and series with 22 cases reported [1–16]. Of these, 5 were bilobar and 15 were bilateral involvement (Table 1). Bilateral CLE often presents as a diagnostic and therapeutic dilemma and approach guidelines are not protocol based. We present our experience of three cases and review the available English literature of this rare entity.

Material and methods

We retrospectively collected data about all the cases of bilateral CLE operated by the authors at various centres over the period of 3 years from January 2016 to December 2018. We operated three cases of this very rare entity and compared our findings with available cases from the literature reviewed. We searched Google Scholar and PubMed using the terms 'Bilateral' and 'Congenital Lobar Emphysema' and also searched from larger series of bilateral or bilobar cases of congenital lobar emphysema. We reviewed 25 cases of bilateral CLE found (Table 1).

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Table 1 Review of literature

S.No.	Author	No.Of patients	Age/Sex	Lobes involved	CT Scan	Bronchoscopy	Treatment Done	Survival	Radionuclide scanning	SURGICAL DECISION BASED UPON	TREATED BEFORE AS	Intervention done before referral
1	Floyd et al	one	NA	RML/LUL	NA	NA	Bilateral lobectomy done months apart	Survived	NA	NA	NA	NA
2	May et al	one	NA	RML/LUL	NA	NA	Bilateral lobectomydone months apart	Survived	NA	NA	NA	NA
3	Schramel et al.	one	NA	RML/LUL	NA	NA	One staged bilateral lobectomy	Survived	NA	NA	NA	NA
4	Tournier et al.	one	NA	RML/LUL	NA	NA	One staged bilateral lobectomy	Survived	NA	NA	NA	NA
5	Ekkelkamp and Vos	one	NA	RML/LUL	NA	NA	One staged bilateral lobectomy	Survived	NA	NA	NA	NA
6	Schiller et al	one	NA	RML/RLL/LLL	NA	NA	Died before surgery	Died	None	NA	NA	NA
7	Stigers et al	two	NA	RML/LLL	NA	NA	Unilateral lobectomy	Survived	None	NA	NA	NA
			NA	RLL/LLL	NA	NA	Conservative management	Survived	None	NA	NA	NA
8	Maiya et al	one		RML/LUL	Hyperinflation of left upper lobe	Bronchomalacia right main and left middle bronchi	Bilateral lobectomy done in same admission	Survived	None	Radiological features and symptoms	None	None
9	Kumar et al	one	6mth/M	RML/lingula	hyperinflation left lingula and right middle lobe	None	One staged bilateral lobectomy	Survived	None	Radiological features and symptoms	Recurrent Pneumonia	None
10	Ghribi et al	three	.5mth/M	RML/LUL	hyperinflation left upper lobe and right middle lobe	None	Bilateral lobectomy done months apart	Survived	Perfusion defect in the LUL and a hypoperfused RML	Radiological features and symptoms	None	None
			4mth/M	RUL/RML	hyperinflation of the right upper and middle lobes	None	One staged bilateral lobectomy	Survived	None	Radiological features and symptoms	None	None
			.5mth/M	RUL/RML	none	None	Bilateral lobectomy done in sameadmission	Died	None	Radiological features and symptoms	None	None
11	Abushahin	one	two mth/M	RML/LUL	hyperinflation of the right middle and left upper lobe	Bronchomalacia of left bronchus and bronchus intermedius	Bilateral lobectomy done in same admission	Survived	None	Radiological features and symptoms	bronchiolitis	none

Table 1 (continued)

S.No.	Author	No.Of patients	Age/Sex	Lobes involved	CT Scan	Bronchoscopy	Treatment Done	Survival	Radionuclide scanning	SURGICAL DECISION BASED UPON	TREATED BEFORE AS	Intervention done before referral
12	Cataneo et al	two	7mth/M	RUL/RML	hyperinflation of the right upper and middle lobes	None	Bilateral lobectomy done in same admission	Survived	None	Radiological features and symptoms	LRTI	NA
			onemth/M	RUL/RML	hyperinflation of the right upper and middle lobes	None	Bilateral lobectomy done in same admission	Survived	None	Radiological features and symptoms	LRTI	NA
13	Perea	Four	onemth/M	RML/LUL	hyperinflation left upper lobe and right middle lobe,Aberrant bronchi	compression of left bronchus and Right middle bronchus, improvement on ballon occlusion of right bronchus	Unilateral lobectomy	Survived	None	Bronchoscopy findings	Respiratory distress	Tracheostomy/ bronchoscopy
			neonate/M	RML/LUL	Overinflation of RML and LUL with herniation across midline	bronchomalacia and compression of both the right middle lobe and left upper lobe bronchi	Unilateral lobectomy	Survived	None	Bronchoscopy and radiological findings	Respiratory distress	Conservative
			neonate/M	RML/LUL	Overinflation of RML, LUL	compression with near complete effacement ofthe right middle and left upper lobe bronchi	Bilateral lobectomy done in same admission	Survived	None	Bronchoscopy and radiological findings	Respiratory distress	Conservative
			7mth/M	RML/LUL	Diffuse emphysema in RML, LUL also with herniation	left upper lobe bronchus narrowing improved with high levels of PEEP	Unilateral lobectomy	Survived	None	Bronchoscopy and radiological findings	Asthma,viral bronchiolitis	Bronchoscopy
	OUR SERIES	Three	6mth/M	RML/LUL	hyperinflation of RML,LUL	Normal	Bilateral lobectomy in same admission	Survived	None	Radiological features and symptoms	Reactive airway disease	Conservative
			7mth/M	RML/LUL	hyperinflation of RML,LUL	Normal but improvement on balloon occlusion of left bronchus	Bilateral lobectomy done months apart	Survived	None	Bronchoscopy and radiological findings	Bronchiolitis	Conservative
			4mth/M	RML/LUL	hyperinflation of RML,LUL	left upper bronchus narrowing right middle lobe mucus plug	Unilateral lobectomy	Survived	None	Bronchoscopy and radiological findings	Viral Brochiolitis	Conservative
14	Lei et al	One	neonate/M	RML/LLL	hyperinflation of RML followed by LLL	endobronchial inflammation	Unilateral lobectomy	Died	None	Bronchoscopy and radiological findings	Neonatal Pneumonia	Ventilation
15	Sawant et al	One	4mth/M	RML/LUL	hyperinflation of RML,LUL	None	Bilateral done in same sitting	Survived	None	Radiological features and symptoms	None	None

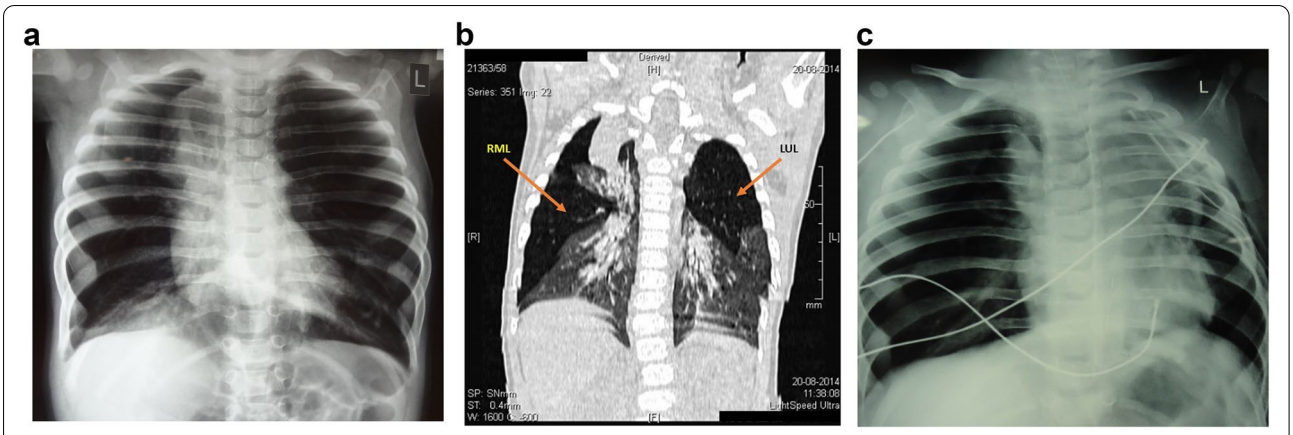

Fig. 1 **a** Pre-operative chest X-ray for case 1. **b** CT image for case 1. **c** Chest X-ray after a left upper lobectomy

Results

We managed three cases of bilateral congenital lobar emphysema. As all the cases had unique details important to the pathophysiology and treatment protocol of bilateral CLE, we are describing them separately.

Case presentation

Case 1

A 6-month-old male child presented to us with a history of increased work of breathing with subcostal retractions since birth. He had a history of hospitalization for the same complaints at least 3 times in the last 6 months. Each time managed as reactive airway disease with nebulization and steroids. There was no antenatal abnormality. He was born at term by normal vaginal delivery and the immediate postnatal period was uneventful. Parents noticed an increased respiratory rate in the first few days of life.

On this admission, at presentation, he was afebrile, maintaining saturation at room air, and was hemodynamically stable. His heart rate was 130/min, blood pressure was 94/52 mmHg, and respiratory rate was 38/min with subcostal retractions. On auscultation, bilateral air entry was equal with bilateral wheeze present, and heart sounds were normal without any murmur. On investigation, his complete blood picture including haemoglobin, total leukocyte count, platelet count, and serum electrolytes including urea and creatinine was within normal range. Chest X-ray showed opacity in the right upper lung field with hyperinflated lung on the left side (Fig. 1a). A CT scan of the chest revealed hyperlucent and expanded right upper lobe, right middle lobe, and left upper lobe with a paucity of vascular markings suggestive of congenital lobar emphysema. Additionally, there was consolidation in the posterior segment of the right upper lobe and the basal segment of the left lower lobe. There was evidence of compressive atelectasis of the bilateral lower lobe and enlarged thymus (5.4×1.6 cm) extending to the anterior superior mediastinum (Fig. 1b).

In view of the bilateral involvement of CLE, a diagnostic bronchoscopy followed by a sequential lobectomy was planned, and as on chest X-ray, where the left upper lobe was predominantly involved, a left-side thoracotomy was planned. The diagnostic bronchoscopy reveals no intraluminal pathology. A left-side thoracotomy through the 4th intercostal space was done; on entering the pleural cavity, the left upper lobe was emphysematous and the left lower lobe was normal. A left upper lobectomy was done and the child was shifted to a paediatric intensive care unit (PICU) on a ventilator. The child recovered well and was extubated 2 days later and started on NG tube feeds. Post-operative chest X-ray showed expansion of the left lower lobe and right-side emphysema became more pronounced (Fig. 1c). On the 4th post-operative day, the child was found to have increased work of breathing with breathlessness and saturations were between 70 and 80% on room air. High-flow oxygen started and ABG done which showed severe respiratory acidosis and X-ray of the chest revealed hyperinflation on the right side. The child was immediately intubated and a central line was placed. The child was taken up for a right-side thoracotomy after initial resuscitation. During surgery, the right-side middle lobe was found to be hyperinflated and emphysematous and enormously enlarged herniating to the left side of the thorax. The right upper lobe and lower lobe were collapsed. A right middle lobectomy was done and the child was shifted back to the paediatric intensive care unit on a ventilator. The child recovered well and extubated on the second post-operative day and was discharged on the 5th day without any complications. Chest X-ray at the time of discharge revealed good bilateral lung expansion with no residual emphysema, atelectasis,

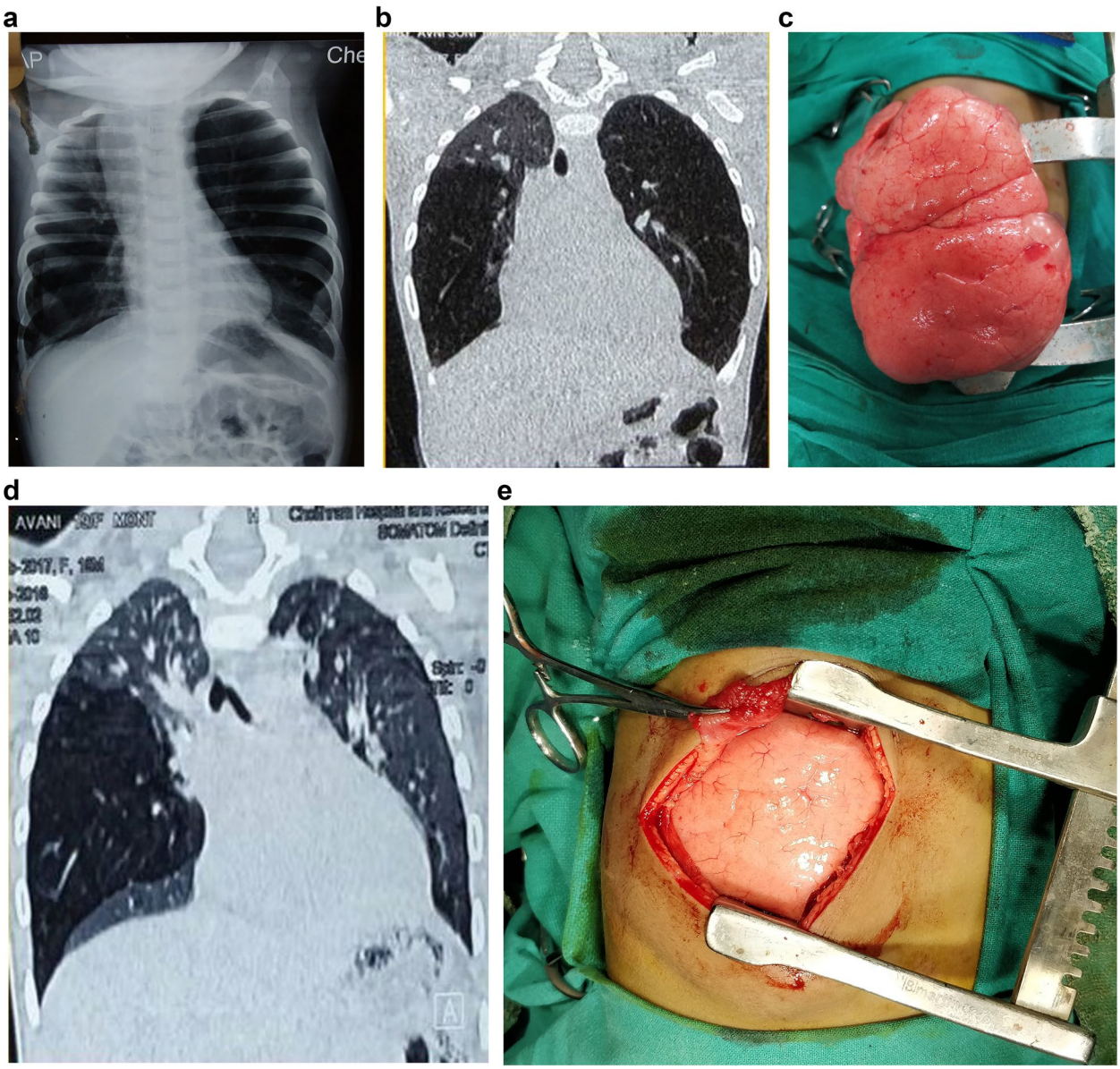

Fig. 2 **a** Pre-operative X-ray for case 2. **b** CT image for case 2. **c** Emphysematous lobe found intra-operatively. **d** CT showing right middle lobe emphysema. **e** Emphysematous right middle lobe

or mediastinal shift. On follow-up, the child is doing well without any respiratory distress. He is feeding well and gaining weight.

Case 2

A 7-month-old female was admitted with complaints of cough and increased work of breathing. She had a history of two previous admissions for similar complaints, when she was diagnosed with viral bronchiolitis and managed conservatively on nebulization and antibiotics. A chest radiograph was suggestive of hyperinflation of the right middle lobe and left upper lobe, which was more pronounced on the left side (Fig. 2a). A CECT showed hyperinflation of the left upper lobe with herniation to right along with hyperinflation of the right middle lobe (Fig. 2b). The underlying ipsilateral normal lobes were collapsed on both sides. Bronchoscopy was suggestive of occlusion of both the left upper and right middle bronchi. The right middle bronchus opened to an increased PEEP. An occlusion of the left upper bronchus with a Fogarty catheter lead to improved ventilation and a slight opening of the right middle bronchus as well. Based on these

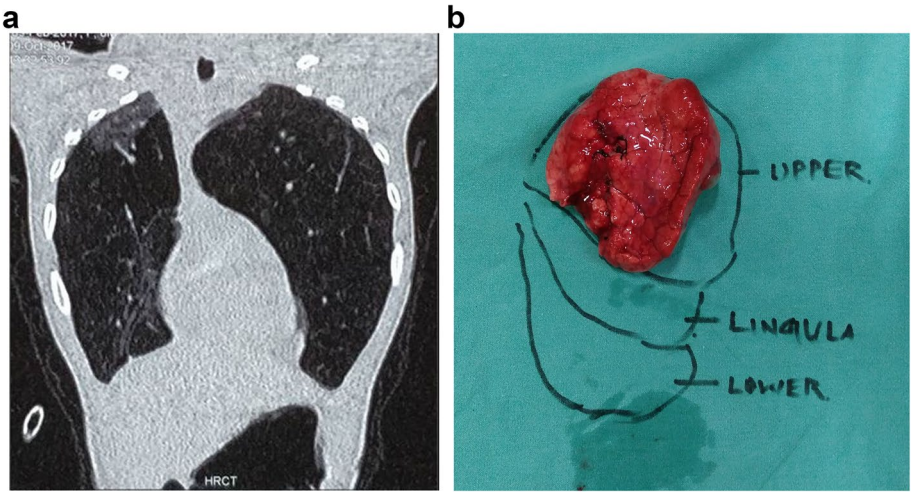

Fig. 3 **a** CT image showing bilateral emphysema. **b** Operative specimen for case 3

findings and radiological findings of more pronounced left upper lobe hyperinflation and herniation, a left upper lobectomy was done. A left upper lobectomy was performed via a left posterolateral thoracotomy (Fig. 2c). The patient did well post-operatively and was extubated on the second post-operative day. The chest X-ray still showed right hyperinflation with an expansion of the remaining left lobes. The infant, however, was asymptomatic and oxygen requirements reduced gradually. She was optimized nutritionally by instituting nasogastric tube feeds and was discharged on the 10th post-operative day on oral feeds. The parents were explained regarding the indications for operating upon on the right side and were kept under close follow-up.

Despite that, she had four episodes of respiratory tract infections with mild symptoms managed on an outpatient basis elsewhere with antibiotics and nebulization. The parents were again counselled for the need for a right middle lobectomy. Repeated chest X-rays showed persistent right middle lobe hyperinflation. A repeat CECT showed good left-sided lung expansion, but there was pronounced right middle lobe hyperinflation and herniation towards the left (Fig. 2d). The parents after 6 months of left lobectomy were advised for a right middle lobectomy based on the radiology and symptomatology. A right middle lobectomy was done via a right posterolateral thoracotomy (Fig. 2e).

The child was discharged uneventfully on the 4th post-operative day and has been symptom free on follow-up.

Case 3

A 4-month-old male child was referred from another hospital where he was treated for respiratory distress for 10 days. He was admitted with cough and subcostal retractions and the mother gave a history of the child remaining unwell since birth. He was having recurrent episodes of respiratory tract infections and was labelled asthmatic based on the family history of asthma. Chest X-ray was suggestive of bilateral pulmonary pruning. He was oxygen dependent and was maintaining saturation on low-flow oxygen by nasal prongs. A CECT showed lobar emphysema on the left upper lobe and right middle lobe which was more on the left causing herniation of the left upper lobe to right (Fig. 3a). Suspecting a diagnosis of bilateral lobar emphysema, a bronchoscopy followed by lobectomy was planned. Bronchoscopy revealed the presence of occlusion on the left side upper lobe bronchus with mucus impaction on the right-side main bronchus. A mucus plug was removed from the right side. On occlusion of the left upper lobe bronchus with a Fogarty catheter, there was a reduction in the ETCO2. Thus, based on the radiological and bronchoscopy findings, the child underwent left upper lobectomy via a left posterolateral thoracotomy (Fig. 3b).

Post-operatively, the right middle lobe showed decreased hyperinflation and the left side showed complete expansion of the other lobes. The child was ventilated for 3 days and extubated uneventfully. He was maintained on nebulization and free-flow oxygen which was weaned off subsequently. The child was discharged on the 12th post-operative day after achieving full feeds. The parents were explained for a need of close follow-up for the right-side residual hyperinflation.

During follow-up, the child is asymptomatic and feeding well with satisfactory weight gain. A repeat chest X-ray showed normal bilateral lung fields with no evidence of hyperinflation even on the right.

Discussion

CLE is diagnosed at birth in about 25% of cases and by 1 month of age in about 50% of cases. The diagnosis is sporadic after 6 months of age [8–12]. Most of the cases reported including the ones in our series had repeated previous admissions, with a low degree of suspicion amongst paediatricians, as the symptoms mimic many medical conditions like upper respiratory tract infections, asthma, viral bronchiolitis, etc. [10–16]. During these admissions, most of these cases were managed conservatively; however, a few underwent invasive procedures like tracheostomy and bronchoscopies, before being referred for surgical intervention (Table 1) [8–13].

There are various reasons described for CLE, and the principal mechanism is that the affected bronchus allows passage of air on inspiration but only limited expulsion of air on expiration leading to over-expansion of the affected lobe [1–8]. This air trapping may be due to (a) dysplastic bronchial cartilages creating a ball valve effect, (b) endobronchial obstruction from mucus plug or extensive mucosal proliferation and infolding, (c) extrinsic compression of bronchi from aberrant cardiopulmonary vasculature or enlarged cardiac chambers, and (d) diffuse bronchial abnormalities which may or may not related to infections [1–8]. However, in approximately 50% of cases, the aetiology is unknown [7–12]. The most common lobe involved in CLE is the left upper lobe (40–50%), followed by the right middle lobe (30–40%), right upper lobe (20%), lower lobes (1%), and multiple sites for the remainder [8–16]. In the literature reviewed, 5 were bilobar involving the right middle lobe/right lower lobe and 20 were of bilateral, with right middle lobe/left upper lobe involvement being the most common (Table 1) [1–13].

The severity of the disease is dependent on the magnitude of abnormality, which cannot be quantified in many cases [13]. Conventionally, three vague variables are used to decide whether surgery is required in patients with CLE. They are age at presentation, severity and frequency of symptoms, and radiology [10–13].

Previous reports have emphasized that children presenting before 2 months of age should be operated and the older age groups should be managed conservatively. The age of presentation can be taken as the reflection of the severity of the disease [10–16].

Reports also describe that those with severe symptoms should be operated but those who are asymptomatic and mildly symptomatic should be managed conservatively [14]. These reports however do not mention the criteria to differentiate between mild and severe symptoms [10–16]. They also do not pay any attention to the frequency of these mild symptoms as recurrent mild symptoms can also hamper the quality of life of many patients and can hamper the ultimate growth potential. They further add that patients on conservative treatment can deteriorate and this poses a life-threatening risk even in asymptomatic and mildly symptomatic cases [14]. In an Indian scenario, where a patient may not have access to immediate expert management of these life-threatening scenarios and may not reach the right specialist for mild symptoms, conservative treatment is a dangerous option.

Radiological findings are frequently used to diagnose CLE which actually is a pathological diagnosis [1–13] (Table 1). Radiology is used to decide which side needs to be operated first in cases of bilateral CLE [8–13]. The lobe which is more hyperinflated and thus herniating to the opposite side is the one operated first. Also, the amount of compression of other ipsilateral lobes is one of the radiological criteria defining the side to be operated upon [8–16].

A more objective way, however, to ascertain whether a patient with CLE requires operative management or can be observed conservatively can be by bronchoscopic findings [10–14]. Findings during bronchoscopy can also help us appreciate the fate of surgery. If the offending lobe bronchus is collapsed and opens up on giving PEEP during bronchoscopy, it is more likely to be managed conservatively [13]. If on occlusion of the offending lobe bronchus the ABG improves and ventilatory requirements decrease, that means doing lobectomy would be beneficial. Doing bronchoscopy can also make us differentiate between acquired emphysema due to mucus impaction from congenital cases [13]. It can make us differentiate between extraluminal causes and inherent congenital cartilaginous defects [13]. Based on the above criteria, one can also very well decide which lobe needs to be operated first in cases of bilateral CLE. Thus, in many instances, bronchoscopy can give objectivity to our cases of bilateral and unilateral CLE and should routinely find a place in the management protocols.

There are a multitude of surgical scenarios described for cases of bilateral CLE, and they are as follows:

1. Bilateral lobectomies in the same sitting (6/25 cases; Table 1)
2. Unilateral lobectomies, conservative or follow-up for contralateral (6/25 cases; Table 1)
3. Bilateral lobectomies in same admission (7/25 cases; Table 1)
4. Bilateral lobectomies done months apart (4/25 cases; Table 1)

The surgical scenarios have been described for various courses during the admission. Maiya et al. [8] recommended two-stage sequential lobectomies as it is practical and less risky and with less post-operative pain, and they believe that an infant tolerates sequential thoracotomies better than simultaneous bilateral thoracotomies.

We also recommend the same approach; however, it may not be possible in all cases. In seven cases, there was a need for contralateral lobectomy during the same admission as there was sudden contralateral hyperinflation causing worsening of symptoms and delayed weaning from the ventilator [8, 11–13, 15].

Thus, we propose that bronchoscopy should be an essential component in the management of all cases of CLE. It helps in defining pathology, the severity of the disease, and the decision on which side to be operated first and avoids unnecessary simultaneous lobectomies. We recommend case selection on basis of radiological, biochemical, clinical, and bronchoscopy criteria. Based on this, an approach of sequential lobectomies is less risky and should be advocated in most of the patients.

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Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
For all the three patients presented in the study, consent for publication was duly obtained (submitted as supplementary files in the submission).

Competing interests
The authors declare that they have no competing interests.

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