

# Congenital lobar emphysema surgical versus conservative management

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## Summary:

**Background:** Congenital lobar Emphysema (CLE) is a rare yet serious condition which requires early detection and management, and usually present during infancy.

## Objectives:

**Patients and methods:** This retrospective study aimed to review and evaluate methods adopted for the management of congenital lobar emphysema cases and it aimed to report and to document any observation which might need further research. The study retrospectively reviewed medical records of 53 cases whom were admitted to the department of thoracic surgery-medical city teaching complex over a period of 4 years (from June 2005 to June 2010). All of them had been diagnosed and managed as cases of congenital lobar emphysema based on their clinical picture and radiologic examination.

**Results:** The commonest age group affected were infants in the second month of life (40 patients) (75.47%), 10 patients (18.86%) within the next 3 months, 2 cases (03.77%) were over 6 months old and only one case (01.88%) was over 1y. The number of male patients in this study was higher than the female patients (38 versus 15) with a male to female ratio was 2.53:1. The affected lobes were left upper lobe in 39 patients (73.60%), right middle lobe in 9 patients (16.98%), right upper lobe in 3 cases (5.66%), and right upper and middle lobes in 2 cases (3.77). Postoperative complication encountered in 6 patients; chest infection (2 patients), bronchopleural fistula and empyema (2 patients), wound infection (1 patient), and postoperative atelectasis (1 patient). Each treated accordingly. No intraoperative mortality was recorded, yet postoperative mortality happened in 3 cases, two cases due to respiratory failure and other one due to septicaemia. The significant finding in this review was that, 8 cases out of the 53, during the period of observation and conservative management, they eventually developed complete clinical improvement of their signs and symptoms and complete resolution of their radiologic findings. All other patients (45) required surgical resection, were lobectomy was the commonest type of surgical resection (39 patients), combined lobectomy and wedge resection performed in (6) patients.

**Conclusions:** A group of cases diagnosed with CLE can benefit from a period of observation and monitoring aiming for conservative management

**Key words:** Congenital lobar emphysema, lobectomy, conservative management.

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## Introduction:

Congenital malformations of the lung are rare and vary widely in their presentation and severity (1). Multiple causes for bronchial obstruction have been suggested and include intrinsic and extrinsic causes. The aetiology may always be linked to some form of bronchial obstruction and air trapping (2, 3). Emergency surgical lobectomy is the only treatment for CLE with severe respiratory distress, but non-surgical management may be appropriate in infants with only mild to moderate respiratory distress, infants with CLE who are not clinically in respiratory distress (4, 5).

## Patients and methods:

This is a retrospective study among 53 infants apart from 1 case who was over 1 year old. All were admitted to Thoracic

Surgical Department of Medical City Teaching Hospital (from June 2005 to June 2010). Preoperative assessment included medical history taking, clinical examination, routine chest x-ray, chest CT scan, complete blood picture, and blood biochemistry. Preoperative management included; O<sub>2</sub> supply and monitoring of vital signs. Then based on their clinical status and respiratory symptoms, eight patients were kept under close monitoring and observation while the other 45 patients underwent surgical resection. Single lumen endotracheal tube for ventilation during anaesthesia was used. Standard posterolateral thoracotomy incision and fifth intercostal space was used for all patients. Classical lobectomy was performed in (39) cases while combined wedge resection or bi-lobectomy in 6 cases. Chest closed after inserting two drainage tubes, an anterior apical and posterior basal. All patients were kept on minimum suction on the underwater seal

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bottles. Postoperatively patients received oxygen, prophylactic antibiotics and physiotherapy. Postoperative hospital stay was variable, depending on postoperative events, complications and chest tube removal times.

**Results:**

This study included 38 boys and 15 girls with male to female ratio of 2.53:1

Five patients had been referred and admitted as cases of lobar emphysema based on their clinical and chest x-ray findings. Later after doing chest-CT they have been found to be cases of agenesis of lobe and secondary physiological hyperinflation of other lobes. They have been excluded from the study.

The patients fall on 4 age groups, (1w – 3m), infants (3m – 6m.), and (6 m. -1y), as shown in Table (1). The youngest patient was 3 weeks old and the oldest was over 1y.

Forty three children (81.1%) were symptomatic at birth and the remaining 10 developed symptoms later on.

The commonest clinical findings were dyspnoea (77.77%), tachypnea (66.66%) and cough (42.22%) (Table 2).

Chest x-rays showed hyperinflation of affected lobes in all patients.

CT-Chest done for all the patients The affected lobes were left upper lobe in 39 patients , right middle lobe in 9 patients, right upper lobe in 3 cases, and two lobes were affected (right upper and middle lobe) in 2 cases.

Forty five patients underwent surgery which varied between lobectomy in 39 patients, and combined bilobectomy and wedge resection in 6 patients (were as bilobectomy had been done in 2 cases whereas lobectomy and wedge resection performed in 4 cases).

Conservative management was successful in eight (8) patients (table 3).

Intraoperative complications were not reported in any case.

There were no intraoperative mortality while postoperative mortality reported in 3 patients due to respiratory failure (n=2) and septicaemia (n=1).

Postoperative complications were chest infection (in tow patients), bronchopleural fistulae and empyema (in tow patients) and wound infection in 1 case and postoperative lobar atelectasis are found in 1 case. The complications were treated accordingly in the ward and patients had been discharge well.

**Table (1): Age distribution of patients with congenital lobar emphysema**

Age	No.	%
1w.-3m.	40	75.47
3m. -6m.	10	18.86
6m. – 1y	2	03.77
>1 y	1	01.88

**Table (2): Clinical findings of patients.**

Signs and Symptoms	No.	%
Dyspnoea	43	77.77
Cough	27	42.22
Tachypnea	37	66.66
Cyanosis	17	37.77
Fever	15	15.55
Intercostal and subcostal recession	10	22.22
Irritability	4	08.88

**Table (3): The data of the conservatively treated cases**

	Sex	Referral diagnosis	Age at time of diagnosis	Symptoms at the time of diagnosis	Lobe affected	Management
1	M	Pneumothorax	4 months	Fever,sob,cough	RML	<ul style="list-style-type: none"> <li>• Oxygen mask</li> <li>• Antibiotics</li> <li>• Steroids</li> <li>• Bronchodilators</li> <li>• Physiotherapy</li> </ul>
2	M	CLE	3.5 months	Fever,sob,cough,tachpnoea	LML	
3	F	CLE	3 Months	Fever,sob,tachypnoea,cough	RML	
4	M	Upper airway obstruction	2 Months	Sob, tachypnoea,fever,cough	RML	
5	M	Pneumothorax	2.5 months	Sob, tachypnoea,fever,cough	LUL	
6	F	CLE	5 Months	Sob, tachypnoea,fever,cough	RML	
7	M	CLE	2 Months	Fever,sob,tachypnoea,cough	LUL	
8	F	CLE	5 Months	Fever,sob,cough,tachpnoea	RML	

**Discussion:**

Congenital lobar emphysema (CLE) is a life-threatening cause of acute respiratory distress in neonates (1, 9). The onset of symptoms is usually seen in early infancy and is rare after 6 months of age (6). Similar findings appeared in the present study, where forty(40) out of fifty three(53) patients (75.47%) were in the 1st three months of neonatal period. while only one patient (01.88) presented after 1years (7, 8). Gender distribution was similar to other studies where a male to female ratio reflected a male predominance (9, 10). Being a cause of acute respiratory distress in neonates, it was not surprising that the dyspnoea and tachypnea were the commonest presenting symptoms (9). Plain chest radiographs usually establishes the diagnosis (8), where hyperinflations of the affected lobes were shown in all patients. Yet chest CT scan was very useful in the exclusion of five cases of lobar agenesis that had plain CXR very similar to lobar emphysema. CT was also useful in confirming the radiographic findings and correctly delineates the affected lobe by showing stretched, attenuated, spread-out vessels in the hyper lucent lobe, which help to differentiate it from compensatory emphysema. It can also exclude an intra-thoracic mass or vascular ring (11). Anatomical distribution of the affected lobe was similar to many other studies, where the left upper lobe (73.6%) was the commonest among other lobes. And the right upper lobe was the least to be affected. (12, 13, 14, 15). Patients underwent surgical intervention. In general the lesion was confined to one lobe as it was seen in 39 patients (73.60%) and treated by lobectomy. Less frequently more than one lobe may be involved (16); such finding had been found only in 6 patients (11.30%) in this study. Therefore they required resection of more than one lobe Compared to Buckner DM (9) where as 86% need lobectomy and there is no case of bilobectomy, whereas bilobectomy was reported by Waleed MH (12). What is significant in this study was that conservative treatment succeeded in 8 patients (15%) with congenital lobar emphysema, emphasizing the opinion brought about by other studies (17,18,19,20, 21), that there is a place for observation and conservative treatment in congenital lobar emphysema. Whether this might reflect a different underlying pathology or a minor form of CLE was not verifiable. Most patients had smooth postoperative period with rapid relief of symptoms, and chest roentgenograms showed full expansion of the lung and the mediastinum became central. The postoperative course was uneventful for the majority of the patients who had been discharged well within 7 days, this indicate the good tolerance of infants to pulmonary resection. Six patients (11.32%) developed postoperative complications, where chest infection happened in two patients, with very good response to antibiotics and physiotherapy. Same measures used to treat one patient with post-operative atelectasis. Bronchopleural fistulae and empyema developed post operatively in two patients treated by tube thoracostomy with application of minimal suction. Wound infection occurred in only one patient and treated by change of the antibiotics and frequent change of dressing. Three patients died post operatively consisting a mortality rate of (5.66%), two of them due to respiratory

failure and one due to septicaemia, which is higher than that in other studies(9,22,23,24,25) and this may be due to delayed referral and delayed surgical treatment, but still less compared to other recent study which was 16.6% (26).

**Conclusion:**

A group of cases diagnosed with CLE can benefit from a period of observation and monitoring aiming for conservative management. Yet the criteria for their selection need to be standardised

Plain CXR can over diagnose cases of CLE, which requires a confirmation by chest CT for better diagnosis

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