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SURGICAL MANAGEMENT OF CONGENITAL LOBAR EMPHYSEMA*

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SUMMARY: Doğan R, Demircin M, Sarıgül A, Paşaoğlu İ, Göçmen A, Bozer AY. (Department of Thoracic and Cardiovascular Surgery, and the Chest Diseases Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara Turkey). Surgical management of congenital lobar emphysema. Turk J Pediatr 1997; 39: 35-44.

Here in nine patients with congenital lobar emphysema who had been treated surgically in the previous 10 years are reported. The ages of the patients at diagnosis ranged from 26 days to 11 months. The six patients whose symptoms started in the neonatal period had more severe dyspnea, cyanosis and respiratory distress. Tube thoracostomy was performed in two of three patients who had been misdiagnosed initially. The affected side was the left upper lobe in five patients, the right upper lobe in three, and the right middle and upper lobes in one patient. Lobectomy was performed in all cases. Dysplasia of the bronchial cartilage was found in six patients and bronchial atresia of the left upper lobe was found in another infant as the etiologic cause of the condition. Although the possibility of conservative management in congenital lobar emphysema has been reported recently, we believe that surgery is the treatment of choice in patients who have persistent or progressive, severe respiratory distress in spite of medical treatment. *Key words: congenital lobar emphysema, pulmonary emphysema, respiratory tract.*

Congenital lobar emphysema is an uncommon condition in which overinflation of the involved lobe can cause severe respiratory distress. Since the first report on the successful treatment of lobar emphysema by lobectomy by Gross[1] in 1945, the condition of lobar emphysema in infancy and childhood has been recognized with increasing frequency.

Clinically the condition may present with varying severity. In approximately one-third of cases, severe respiratory distress occurs soon after birth, and rapid deterioration may follow[2-4]. The remaining two-thirds of patients have a slower onset, usually at one or two months of age, often coinciding with a respiratory infection. Occasionally asymptomatic or stable cases have been reported, but characteristically in patients who were not symptomatic until they were older than six months of age[3-6]. This may be confusing to the physician faced with the problem of diagnosing this rare condition.

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The natural history of the disease is unknown for the most part, since surgery to remove the overinflated lobe has been performed in the majority of patients. Only a few isolated case reports of nonsurgical treatment were recorded in the literature before 1990[7-10]. More recently, Kennedy[11] and Stigers[12] showed that most patients with congenital lobar emphysema could be managed medically with good results. However, the treatment of choice is surgical removal of the affected lobe in patients who have persistent or progressive, severe respiratory distress that does not respond to medical treatment.

In the past 10 years, nine patients with congenital lobar emphysema were surgically treated at the Thoracic and Cardiovascular Surgery Department. Here in we present our experience with the diagnosis, etiology, prognosis and treatment of this condition.

Material and Methods

In this study, nine children with congenital lobar emphysema who had undergone surgical treatment in our department between 1985 and 1995 were analyzed. Of the nine patients, two were diagnosed at Dr. Sami Ulus Children's Hospital. The remaining seven children were diagnosed at Hacettepe Children's Hospital.

Six of the nine patients were boys and three were girls. Their ages ranged from 26 days to 11 months (Table I). The history was typical in most, with dyspnea dating from early infancy and intermittent attacks of cyanosis. On admission all were severely ill. Chest x-rays showed a markedly emphysematous lobe of the lung causing compression atelectasis of the adjacent lobes and shifting of the mediastinum to the opposite side of the thorax (Figs. 1-4). The affected side was the left upper lobe in five patients, the right upper lobe in three, and the right middle and upper lobes in one patient. In all patients, physical examination revealed tachypnea, cyanosis, intercostal retraction, and decreased breath sounds over the involved hemithorax. Chest radiographs of Cases 5, 6 and 8 were initially interpreted as pneumothorax, and diagnostic aspiration was performed at another institution. Tension pneumothorax developed with a marked increase in respiratory distress with cyanosis. A chest drain was inserted with no further air drainage and no clinical improvement (Cases 5 and 8).

To exclude any associated cardiac defect or vascular abnormalities that could accompany congenital lobar emphysema, two-dimensional echocardiography was performed in all patients. Except for patent foramen ovale in Cases 3, 7 and 9, no additional anomaly was found. Time-consuming examinations requiring sedation and intubation, such as angiography and computed tomography, could not be done. However, in order to determine the presence or absence of abnormalities of the bronchial tree mucous plug or a foreign body, bronchoscopy was performed in all cases with Starz (Karl-Starz Endocopy 6MB and Co, Tuttlingen, West Germany) pediatric rigid bronchoscopy equipment under general anesthesia with controlled ventilation. No foreign body aspiration or mucous plaque was found. However, in Case 3, the left upper lobe bronchus was found to be a blind-ended pouch filled with a small amount of mucoid material.

Table I: Patients with Congenital Lobar Emphysema Symptomatology, Management and Results

No.	Patients	Sex	Age of onset of symptoms	Age at operation	Presenting features	Additional anomalies	Affected lobe	Treatment	Results
1 SA	1638116	F	2.5 months	3 months	Tachypnea Cyanosis	–	RUL	Lobectomy	Well at 3 years
2 RB	1671903	M	2 months	2.5 months	Tachypnea Cyanosis	–	RUL	Lobectomy	Lost to follow-up after first check-up
3 ÇÖ	1671830	M	10 months	11 months	Cough, fever, wheeze	PFO, CPD	LUL	Lobectomy	Well at 3 years
4 AA	1690080	M	10 days	2 months	Cough, cyanosis, dyspnea, congestive heart failure, fever	–	RUL RML	Bilobectoy	Died on 8th day with sepsis
5 CA	1865479	M	15 days	2 months	Fever, cough, tachypnea	–	LUL	Tube thorocostomy Lobectomy	Well at 6 years
6 ÖT	1877143	F	4 days	10 days	Respiratory distress, cyanosis, iatrogenic pneumothorax	–	LUL	Lobectomy	Well at 3 months
7 MÖD	1896478	F	2 days	26 days	Respiratory distress, tachypnea, cyanosis	PFO	LUL	Lobectomy	Well at 1 year
8 EÜ	2065800	M	15 days	1 month	Cyanosis Tachypnea, tube thoracostomy	–	RUL	Lobectomy	Well at 2 years
9 MÇ	2037484	M	2 days	2 months	Tachypnea	PFO	LUL	Lobectomy	Well at 3 years

RUL : Right Upper Lobe.
LUL : Left Upper Lobe.
RML : Right Middle Lobe.
PFO : Patent Foramen Ovale.
CPD : Congenital Pericardial Defect.

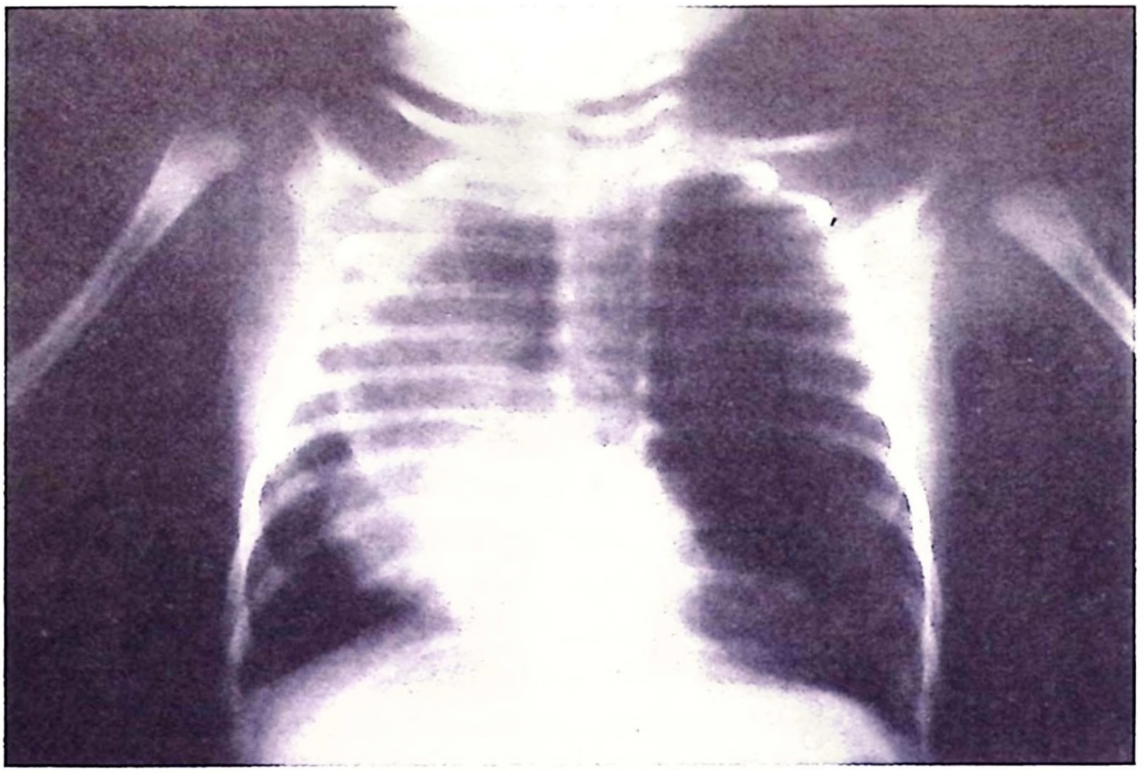

Fig. 1: The emphysematous left upper lobe is herniated across the mediastinum into the right side. Note the mediastinal shift to the right (Case 7).

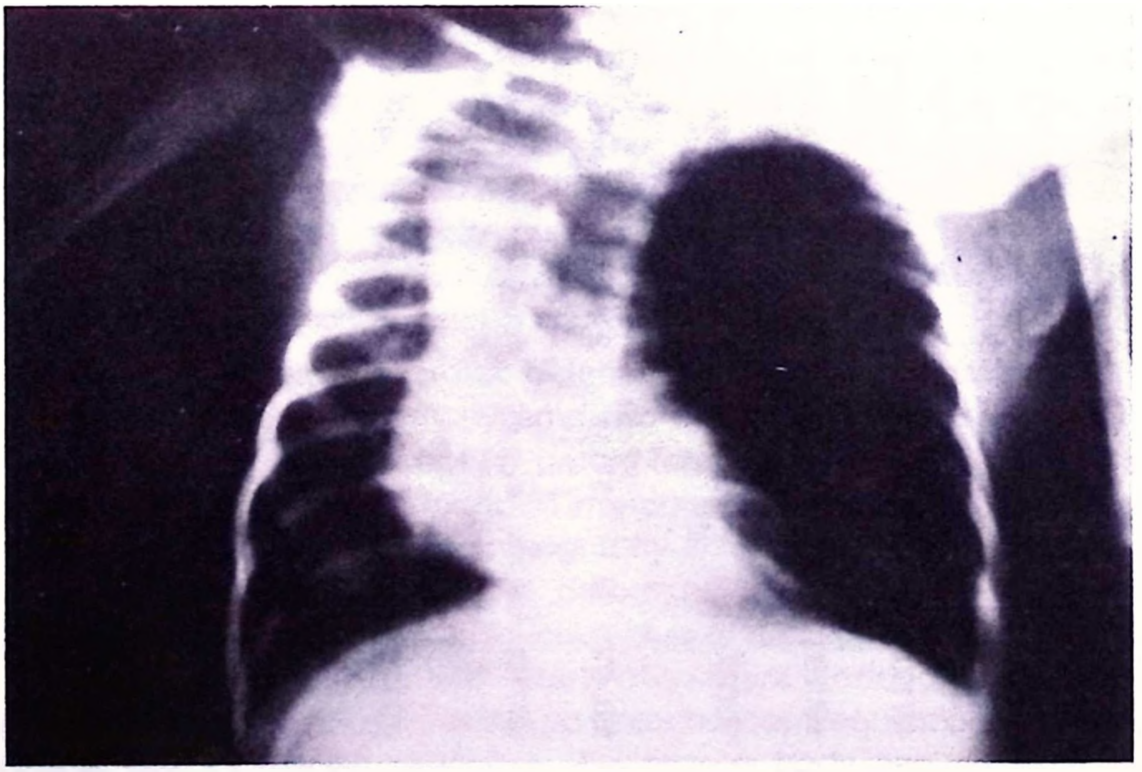

Fig. 2: Hypertanslucent left chest. Initial diagnosis was tension pneumothorax (Case 6). Congenital left upper lobe emphysema.

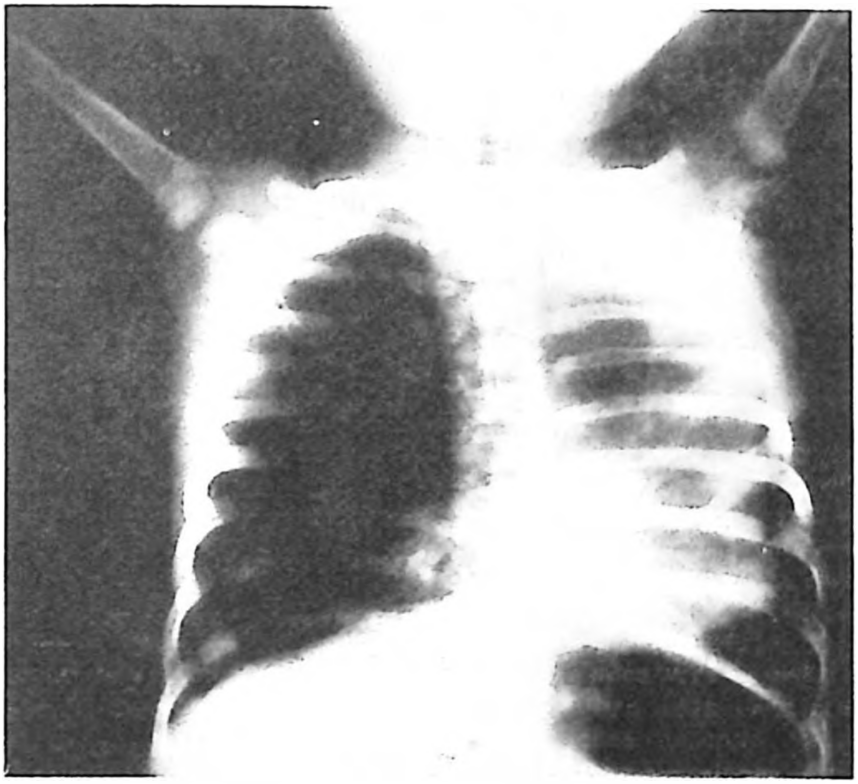

Fig. 3: Infant with congenital lobar emphysema of the right upper lobe. The involved lobe is hyper-aerated and has crossed the mediastinum into the left hemithorax. The right lower lobe is compressed (Case 8).

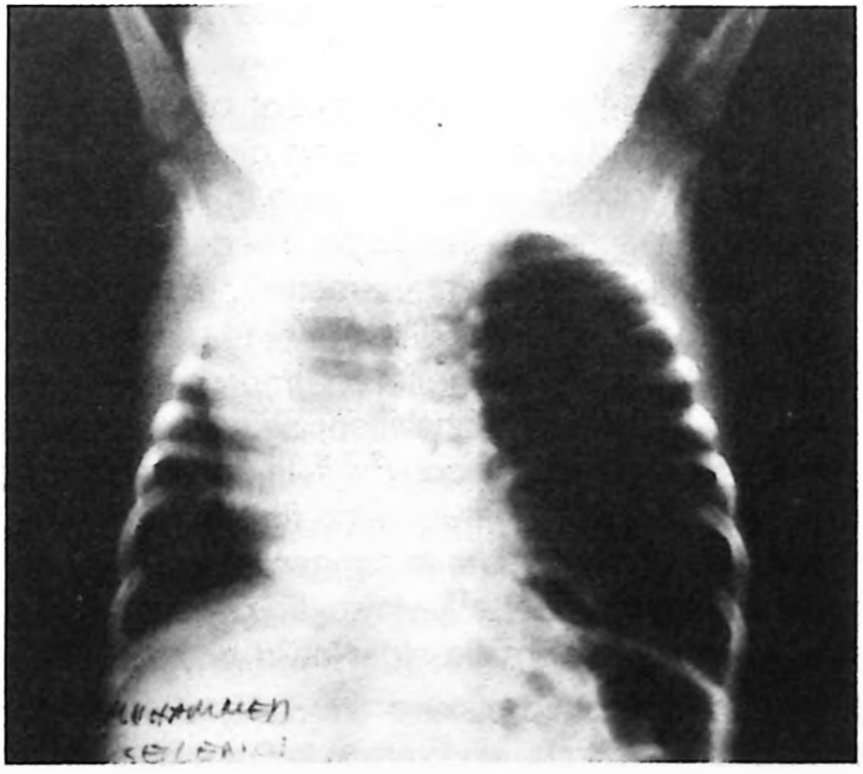

Fig. 4: Infant with overdistended left upper lobe due to congenital lobar emphysema, with displacement of the heart and mediastinum to the right (Case 9).

Following bronchoscopy, which was performed in the operating room, the children were intubated. At the time of anesthetic induction, positive pressure ventilation, which may precipitate air trapping and cardiorespiratory collapse, was avoided. Thereafter the chest was opened as quickly as possible. Grossly, the affected lobes seemed like sponge rubber, not deflating bouncing back into shape after being compressed. The remaining lung tissue was atelectatic. Before resection, the mediastinum was carefully examined for lesions that could have obstructed the bronchus. Lobectomy was performed in all patients. On histopathological examination of the resected lobes, sponge-like lung structure and emphysematous alveoli were detected. Although the gross appearance of the bronchial cartliages was found to be dysplastic in six patients at operation, the operative findings were not confirmed pathologically. In Case 3, the proximal bronchial cartilage of the left upper lobe was found to be absent after ligation of the pulmonary artery and veins. The left upper lobe bronchus ended with a blind pouch containing fetal mucoid secretions. A congenital pericardial defect measuring 2 x 1 cm was also seen in this case.

One patient died on the eighth postoperative day. This patient who had been severely ill preoperatively, developed sepsis and, despite antibiotic and resuscitative measures, suffered a cardiac arrest and died. The survivors were followed up by the Pediatric Chest Diseases Division. One patient was lost to follow-up after the first postoperative check-up. The remaining seven patients were found to be well at regular follow-up visits.

Discussion

Infantile congenital emphysema is an infrequent but progressive variety of obstructive emphysema leading to uniform massive distension of the involved lobe. Severe respiratory distress occurs soon after birth in approximately one-third of patients. The severity of dyspnea depends on the degree of over-inflation the affected lobe. Cyanosis, wheezing and rib retraction are common. Approximately 12 percent of patients are in critical condition prior to treatment[5]. The symptoms can progress rapidly with crying and agitation. Improvement is rarely achieved with medical treatment, i.e., oxygen inhalation, airway humidification, antibiotic administration, suctioning and bronchoscopy[2, 3, 6]. In approximately half of patients, symptoms develop later, sometime between the first week of life and six months of age[2, 13]. Only five percent of cases with congenital lobar emphysema present with symptoms after six months of age[14]. However, congenital lobar emphysema may be diagnosed in an asymptomatic older child or adult[13, 15].

Approximately two-thirds of reported patients are male, and almost all patients are white. The condition is reported to be uncommon in black and premature infants[6, 14]. Murray[5] reported a 14 percent incidence of associated congenital heart lesions and lack of familial incidence. Although it is rare, some cases have bilateral involvement[16, 17].

In the present series the onset of the symptoms was typically in early infancy. The symptoms are those of respiratory distress. The seven neonates who presented with severe respiratory distress supported the hypothesis that the earlier the presentation the more severe the symptoms. Our male/female ratio (2/1) was similar to that reported in the literature. Congenital lobar emphysema usually affects the left upper lobe, followed by the right middle lobe. However, in our series the left upper lobe was most often affected. (5/10), followed by the right upper lobe (4/10). Right-middle-lobe involvement was seen in only one infant.

The cardinal radiographic sign of congenital lobar emphysema is expansion of the involved lobe. During the first few days of life, bronchial obstruction often results in delayed clearance of fetal lung fluid from the affected lobe. During this period the affected lobe may be seen to be opaque. This radiologic finding occurs in two-thirds of patients who present with symptoms at birth[12, 13, 18]. In infants with progressive respiratory distress and the characteristic x-ray findings of lobar hyperinflation, mediastinal displacement away from the emphysematous side, and controlateral atelectasis, congenital lobar emphysema should be suspected[3, 5, 19]. The diaphragm is depressed and flattened on the affected side. There may be anterior herniation of the lobe through the mediastinum, with consequent posterior displacement of the heart. In severe cases the over-expanded lung may herniate into the opposite hemithorax. These radiological findings and the clinical picture of an infant in respiratory distress are generally diagnostic, and no further examination is necessary. Congenital lobar emphysema should be differentiated from giant air cysts of the upper lobes, diaphragmatic hernia and pneumothorax. A mistake in diagnosis can be avoided by careful interpretation of the chest x-ray, fluoroscopy, lung isotope scanning and barium swallowing[2, 16, 19].

Fluoroscopy helps to establish the diagnosis in an older child with less severe clinical signs or when there are atypical radiologic features; compensatory emphysema and other causes of non-obstructive emphysema should also be considered. When other causes of obstructive emphysema are suspected, a barium swallow may exclude extrinsic compression to a bronchus such as aberrant vessels, bronchogenic cyst, etc[19]. Bronchography may help to identify the obstructed bronchus and show either an intrinsic or extrinsic obstruction. It must be noted that these investigative procedures are not indicated and may be dangerous for an infant with respiratory distress[6, 12, 19]. More invasive investigative procedures such as arteriography may occasionally be required to show anomalous vessels causing extrinsic compression of a bronchus[20, 21]. Bronchoscopy may be valuable to rule out the presence of a mucous plug, mucosal fold or foreign bodies causing either obstructive emphysema or atelectasis with compensatory emphysema, but it would be hazardous for an infant in severe respiratory distress[17, 19]. In an older child with an acute onset of symptoms, bronchoscopy is indicated to rule out an endobronchial mass or an aspirated foreign body, and should be planned to precede immediate thoracotomy.

Radionuclide ventilation/perfusion scintigraphy is useful in evaluating the function of the affected and compressed lobes, and is safe with a minimal absorbed radiation dose[11, 12, 19, 22, 23]. Perfusion studies are accomplished by intravenous injection of ^{99m}Tc-MAA[11, 12, 19, 23]. Ventilation studies may be performed in infants utilizing Krypton 81-m or submicronic aerosols containing either ^{99m}Tc-DTPA or ^{99m}Tc-SC[11, 23]. In case in which plain radiographs or CT indicate congenital lobar emphysema, the scintigraphic demonstration of a reduction in both ventilation and perfusion to the affected lobe can confirm the diagnosis of congenital lobar emphysema; however, CT and ventilation/perfusion scintigraphy are difficult to perform in a small infant. Each of these studies takes approximately six minutes. Although Kr-81m ventilation/perfusion scans are obtained during continuous breathing in an acceptable period of time (1-3 minutes), they have not been used in our center. Moreover, Kr-81m has some disadvantages, such as it is not conveniently available on a regular basis, and it may not be as effective as Xe-133 (the agent routinely used in our center) in the assessment of air trapping.

The etiology of congenital lobar emphysema has not been clearly defined. It has been suggested that several mechanisms may be involved: infection damaging the alveoli, developmental abnormality of alveoli[24], an obstruction of the lobar bronchus perhaps from inspissated secretion[25], bronchomalasia resulting from undue flexibility or absence of cartilage[26], or the most recently described pathologic entity-polyalveolar lobe[27]. In polyalveolar lobe the alveolar number is increased fivefold. The airways and arteries are normal for age in number, size and structure, suggesting that the condition represents gigantism of the alveolar region[28]. Although it is rare, the clinical features of congenital lobar emphysema may arise from bronchial atresia of the left upper lobe[29]. However, most authors now agree that in approximately one-half of cases no definite causes could be found[3, 14, 20].

In the past, congenital lobar emphysema was considered to be strictly a surgically treatable disease. This point of view was based on the belief that removal of the affected lobe would improve pulmonary function by releasing compressed normal lung parenchyma, and prevent recurrent infection and sudden distention of the affected lobe. Most trials of medical treatment have not been encouraging and have been associated with high mortality rates[3, 5, 6, 17, 26, 30]. When treated by medical therapy alone, 50 percent of patients died from progressive respiratory distress, and 75 percent of the survivors had persistent emphysema[6].

Conservative treatment for lobar emphysema has been recommended by some authors. Korngold[7] reported two surviving cases treated by needle aspiration. Later, Roghair[8] reported two cases of long-term follow-up of medically treated infants with infantile lobar emphysema. Eigen et al.[9] suggested that lung growth and function in patients with congenital lobar emphysema who were treated conservatively were not different from those of children treated surgically.

Observation of these authors shows that the medically treated patients did not have an increased incidence of pulmonary infection and did not experience sudden deterioration. In 1977 Shannon et al.[10] reported 16 patients treated medically. Two of these patients died from bronchopulmonary dysplasia; surgical resection was required in three of the 14 surviving patients, and the remaining 11 were successfully managed medically. In 1991 Kennedy et al.[11] reported 12 children with congenital lobar emphysema who were managed medically with long-term follow-up. More recently Stigers et al.[12] also reported eight infants with congenital lobar emphysema. Three patients required surgical resection for progressive worsening of respiratory distress; the remaining five patients were managed medically with eventual remission of symptoms. However, it would not be appropriate for us to enter a discussion about the conservative treatment and follow-up of such patients because we do not have patients treated this way in our clinic.

Based on the reports in the literature and our own experience, it can be concluded that patients with congenital lobar emphysema can be managed medically. The urgency of treatment and the treatment modality depends on the severity of the patient's respiratory disease. Attempts at needle aspiration must be avoided, for this almost always results in a tension pneumothorax that can be fatal. Conservative treatment may be considered in selected infants and older children with mild symptoms. In patients who have persistent or progressive, severe respiratory distress that does not respond to medical therapy, the treatment of choice is surgical removal of the affected lobe.

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