count for the clinical manifestation of the disease(1,9). The photodermatitis of this disease is devastating and often causes permanent disfiguration as seen in our case. The excretion of burgendy red urine in CEP begins at birth or shortly thereafter and continues throughout life(1,9). Spelenectomy is often beneficial in CEP with definite decrease in photosensitivity and reduction in urinary porphyrin excretion(10). It was, however, not undertaken in our case.

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Congenital Lobar Emphysema

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Congenital lobar emphysema (Synonym –Panlobular emphysema of infancy) is the postnatal over distention of one or more lobes of a histologically normal lung, usually presenting with respiratory distress in infancy(1,2). This condition is rare but should

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be considered in the differential diagnosis of respiratory distress in the neonatal period. Early surgical intervention carries good prognosis. We are reporting 6 cases of congenital lobar emphysema who had presented with respiratory distress in early life.

Case Reports

The details of all the cases are summarized in *Table I*

Case 1: A 2-day-old term male baby weighing 2900 g, delivered normally presented with history of breathing difficulty since 12 hours of age. Chest radiography showed emphysema of left upper lobe with shifting of the mediastinum to the right. Bronchoscopy revealed that the left main bronchus was collapsing spontaneously (bronchomalacia). Since the mother refused surgical intervention, the child was managed conservatively with intravenous fluids and antibiotics and was discharged after one week.

Case 2: A one-month-old female child weighing 3400 g presented with cough and breathing difficulty of 7 days duration and fever with poor feeding of 3 days duration. Chest examination revealed the presence of bilateral crepitations. Chest radiography showed emphysematous changes in left upper lobe with herniation to the right side. The child was improving on conservative management but left the hospital after 10 days against advice and was lost to follow-up.

Case 3: A two-month-old female child presented with history of breathing difficulty since 4th day of life. The child was treated outside for three such attacks with antibiotics, IV fluids and blood transfusion. The fourth attack of respiratory distress was complicated with oliguria, puffiness of face and pedal edema for which she was referred to this hospital. On examination, the baby was pale, icteric with edema feet and ecchymotic patches on the left forearm. The child had intermittent apneic spells on the second day, developed extensive sclerema on the third day and expired on the fourth day. Other investigations revealed increased serum bilirubin and hepatic enzymes with normal renal and hematological parameters.

Case 4: A full-term female baby delivered by forceps without any complications developed tachypnea and chest retractions two hours after birth. Respiratory system examination revealed hyper resonance and diminished breath sounds on the right side. The child improved with conservative management. She was discharged after seven days and is normal on regular follow-up.

Case 5: A post-term male baby weighing 3000 g was delivered normally with moderate birth asphyxia. *X*-ray chest showed complete situs inversus with emphysema (left side) with herniation of lung to right (*Fig. 1*). A mucus plug was removed from the right main bronchus by bronchoscopy. The child was treated with IV fluids, antibiotics and decongestive measures. He expired on the third day after showing initial improvement. Autopsy revealed complete situs inversus with a bilobed right lung and a trilobed left lung with emphysematous changes. There was dextrocardia with transposition of great arteries, single atrium and patent ductus arteriosus (*Fig. 2*).

Case 6: A two-month-old male child was admitted with history of respiratory distress since birth. He was treated outside with various antibiotics. Respiratory system showed hyper resonant lung fields with occasional rhonchi on both sides and diminished breath sounds on the right side. *X*-ray chest showed left lobar emphysema with herination of lung to right side (*Fig. 3*). The child responded to conservative manage-

TABLE I—*Summary of All Cases of Congential Lobar Emphysema*

Case No.	Age	Sex	Presenting symptoms and signs	Additional features	*X*-ray	Bronchoscopy findings	Management	Outcome
1.	2 days	M	Dyspnea, tachypnea, trachea to right intercostal retractions + subcostal retractions, hyper-resonant left hemithorax, breath sounds diminished in right hemithorax	Liver 5 cm, Spleen 2 cm	Emphysema left upper lobe with mediastinal shift to right	Normal trachea and right bronchial tree, Left main bronchus had evidence of bronchomalacia	Conservative	Discharged
2.	1 mo	F	Cough, dyspnea, fever with poor feeding, tachypnea, Intercostal retractions + subcostal retractions	Patent ductus arteriosus	Emphysema left upper lobe with herniation to right	Not done	Conservative	Satisfactory improvement but left against advice
3.	2 mo	F	Dyspnea with grunting crepitations and rhonchi in both lung fields	Septicemia	Lobar emphysema right side, shift of mediastinum to left	Not done	Conservative	Expired
4.	1 day	F	Tachypnea, intercostal retractions + subcostal retractions + central cyanosis, hyper-resonant right hemithorax	–	Emphysema right side with herniation of lung to left	Not done	Conservative	Discharged
5.	1 day	M	Tachypnea, intercostal retractions + subcostal retractions + cyanosis	Congestive cardiac failure, dextrocardia transposition of great vessels, patent ductus arteriosus and single atrium	Complete situs inversus with emphysema left side, and herniation of lung to right	Mucus plug in right main bronchus	Conservative	Expired
6.	2 mo	M	Dyspnea, tachypnea, central cyanosis chest retractions, hyper-resonator left hemithorax	–	Left lobar emphysema with herniation of lung to right	Normal trachea and right bronchus, left main bronchus had evidence of bronchomalacia	Conservative	Discharged

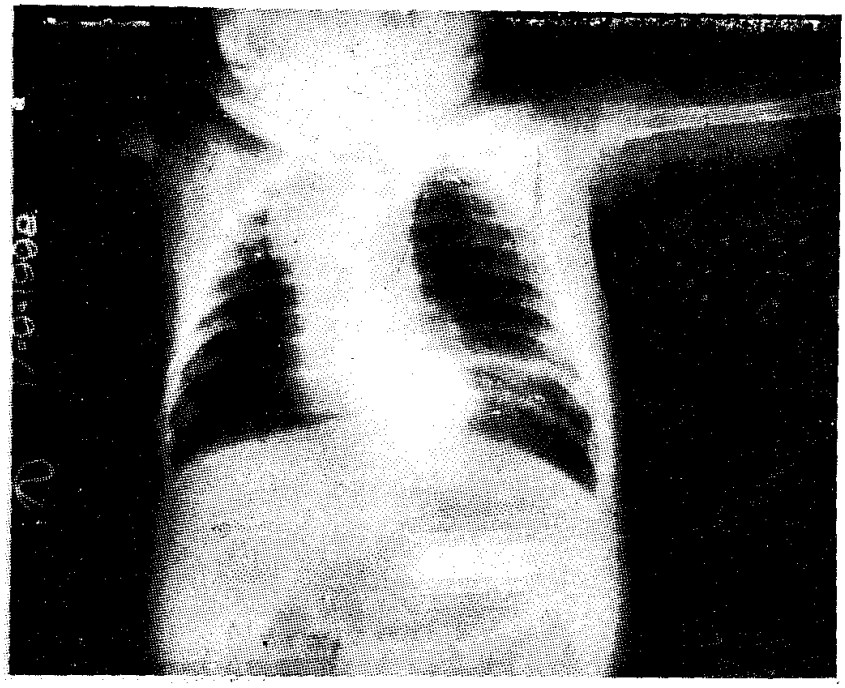

Fig. 1. X-ray chest of Case No. 5 showing lobar emphysema left lung with herniation of the lung to right side with situs inversus.

Fig. 2. Autopsy photograph of Case No. 5 showing trilobed left lung and bilobed right lung with absent atrial septum and ductus arteriosus.

ment within seven days and is on regular follow-up.

Discussion

Congential lobar emphysema (CLE) usually manifests with respiratory distress or with recurrent chest infections in early infancy(2,3). Two of our cases had recurrent chest infections. The over inflated lobe compresses the adjacent structures producing atelectasis of the nearby pulmonary segments and shifting of mediastinum with herniation of lung to the opposite side. CLE results secondary to partial bronchial obstruction which acts like a valve. The obstruction may be due to a mucus plug, pliable bronchial cartilage, aberrant vessel causing compression, bronchial cyst, bronchial stenosis and atresia, polyalveolar lobe or foreign body aspiration(4,5). There may be more fundamental tissue abnormalities at the alveloar level(1). However, in most instances the etiology is not ascertained.

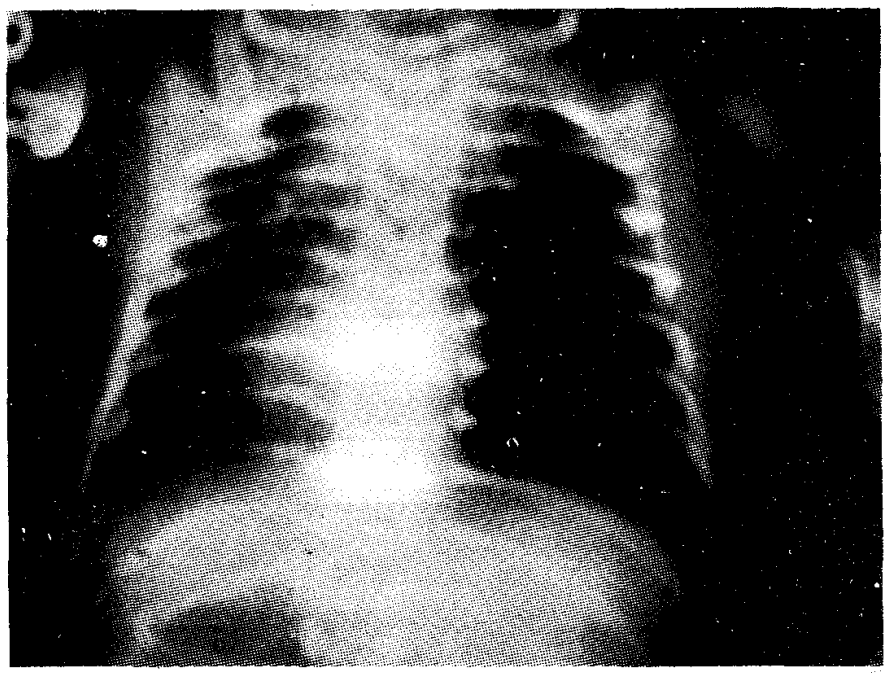

Fig. 3. X-ray chest of Case No. 6 showing lobar emphysema left side with herniation of lung to right.

Out of the above six cases, one had a mucus plug and two had bronchomalacia.

Four of our cases presented with breathing difficulty and tachypnea within few hours after birth and one each on the 4th and 22nd day of life. The age of presentation can vary from early neonatal period to 10 years(6). Diagnosis of this condition depends mainly on clinical suspicion and the radiological evidence of lobar emphysema in the postnatal period. Increased clinical awareness of this condition and frequent use of roentgenography helps in early diagnosis. Rarely, lung scintigraphy or cardiac catheterization may be required to establish an etiology and to diagnose any associated anomalies(2). Cardiac anomalies like pulmonary valve agenesis has been reported with CLE. But complete situs inversus with single atrium, transposition of great vessels and patent ductus arteriosus has not been reported earlier. Thoracotomy and resection of the emphysematous lung may be life saving in selected cases. Surgical resection of the affected lobe is advised when the PO_2 falls below 50 mm of Hg and PCO_2 exceeds 50 mm of Hg(7). Two of our cases having manifestations of respiratory distress since birth were poor risk cases for surgical intervention. They expired in spite of active management. One baby developed apneic spells with septicemia and sclerema. The other four patients responded to conservative management satisfactorily. Intermittent positive airway pressure may be sufficient for treating mild cases of CLE(4,6). Long term prognosis has been similar with surgical and medical treatment(7). Earlier presentation in the neonatal period has been associated with poor prognosis(6). Multiorgan failure, septicemia and sclerema are associated with higher mortality(8).

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NOTES AND NEWS

PEDIATRIC NEPHROLOGY UPDATE

A Pediatric Nephrology Update is being organized by the Bangalore Children's Hospital and Nephrology Chapter of IAP on *15th December 1993* from 9.00 a.m. to 4.30 p.m. at the Holiday Inn, Bangalore.

Faculty includes eminent pediatric nephrologists like Dr. J. Brodhel (Germany), Dr. N. Yashikawa (Japan), Dr. K.J. Sheth (USA),

Topics include Approach to UTI in Children, Therapy of Glomerulopnephritis, Childhood Hypertension, Pathology of Glomerulonephritis and panel discussion on Controversies in Pediatric Nephrology.

Registration: Rs. 100/- only; Postgraduates Rs. 75/-.

Cheques payable to 'Sackhumvit Trust', Bangalore.

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