CASE PRESENTATION

Familial Congenital Lobar Emphysema

By Khalil Hassan Ras Romani*
F. Al-Hilli**

TWO BAHRAINI brothers with congenital lobar emphysema of the left upper lobe who presented with respiratory distress few hours after birth are reported. The condition is rare and has only been described in one other report in the literature. To the best of our knowledge this is the first report of two brothers with congenital lobar emphysema of the left upper lobe successfully treated with lobectomy almost during the first week of life. One of the infants developed mild degree of emphysema in the right middle lobe one week after surgery. The diagnosis of the condition when suspected clinically can be confirmed radiologically and by studying the pathological material. The clinical presentations, aetiology, pathological findings and management are also reviewed.

INTRODUCTION

Overinflation of a pulmonary lobe is one of the main causes of respiratory distress in the newborn and young infants (4,12). This rare condition has been recognized as congenital lobar emphysema (also called infantile lobar emphysema, regional infantile emphysema and congenital obstructive emphysema). It usually affect an upper lobe or right middle lobe (6,12) leading to compression of the rest of the ipsilateral lung with herniation across the middle line to compress the contralateral lung. There is also mediastinal shift. A variety of anatomic lesions may produce this picture and the mechanism is that air can enter the affected lobe during inspiration but cannot escape freely during expiration. The disease is usually fatal but prompt lobectomy is curative if it is recognized during early infancy.

Although expiratory wheezing has been described in siblings of infants with congenital lobar emphysema (6), the familial tendency of the disease is not clear. To the best of our knowledge there is only one report in the literature of two brothers with congenital lobar emphysema of the left upper lobe and only one of these survived after lobectomy (6). The present communication provide the second report of two brothers who presented few hours after birth with respiratory distress due to congenital lobar emphysema of the same left upper lobe. It also provide the first report of these brothers who underwent successful emergnecy lobectomy.

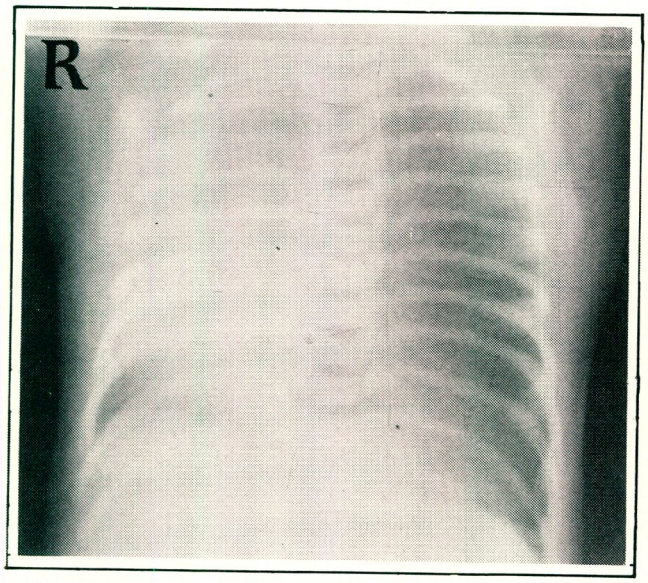
* Department of Surgery, Salmaniya Medical Centre, Bahrain.

** Department of Pathology, Salmaniya Medical Centre, Bahrain.

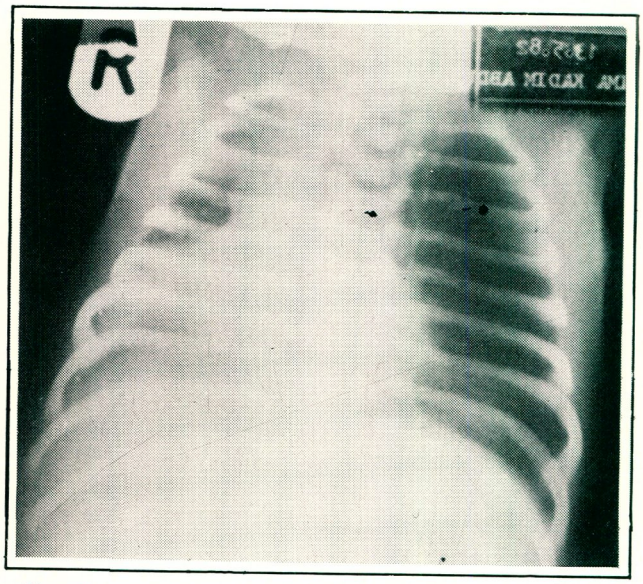
CASE REPORTS

Case No. 1

A ten days old Bahraini male infant was referred to the Paediatric Surgical Unit with severe respiratory distress. He was the product of full term normal pregnancy weighing 3.6 Kilograms at birth. He has 4 other normal siblings. Mild respiratory distress was noticed 5 hours after birth which improved with oxygen therapy. Three days later, the distress became progressive and on the tenth day his condition deteriorated suddenly and developed cyanosis, tachypnea and dyspnea. The left side of the chest was hyperresonant to percussion and breath sounds were diminished with bilateral wheezing. Chest X-ray showed hyperaereation of the left upper lobe with herniation across the anterior mediastinum to the right side and collapse of the left lower lobe. The right lung was also collapsed and compressed with right mediastinal shift (Figure 1). Emergency posteriolateral thoracotomy was performed through the fifth intercostal space. The left upper lobe was severely emphysematous and herniated across the anterior mediastinum to



Chest X-ray on the tenth day after birth (Case No.1) showing hyperaereation of the left upper lobe with herniation across the mediastinum. There is also mediastinal shift to the right with collapse of most of the right lung and left upper lobe.*

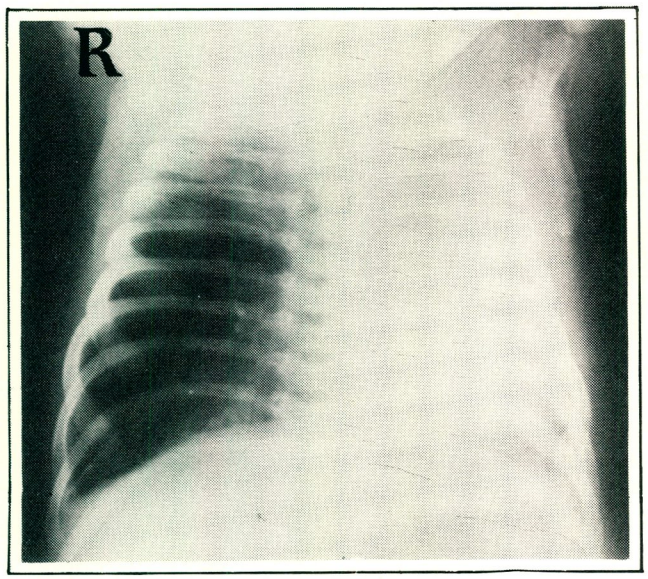


Chest X-ray on the seventh day after birth (Case No.2) showing hyperaereation of the left upper lobe and collapse of the left lower lobe. There is also herniation of the emphysematous lobe across the anterior mediastinum with mediastinal shift to the right and collapse of the right lung.

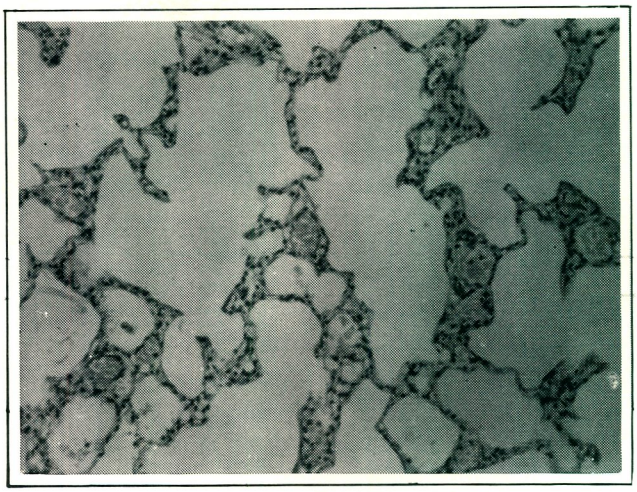
the right side. The left lower lobe was collapsed. The left upper robe was retracted downward and backward. The pleura was incised parallel and below the concavity of the aortic arch in the region of the left vagus. The incision was curved forward and downward between the lung edge and the phrenic nerve infront. The pulmonary artery was mobilized by opening its sheath and with careful dissection it was dissected from the anteriosuperior aspect of the left main bronchus. Three branches of the left pulmonary artery to the upper lobe were identified and divided between ligature. The left main fissure was opened with ease and the left upper lobe was dissected from the left lower lobe. From the left pulmonary artery which traverse the base of the interlobular fissure, the two branches to the linguela were identified and divided between ligature. The upper lobe was then retracted backward and the tributaries of the left pulmonary vein from the upper lobe and the linguela were dissected and divided. The main bronchus to the left upper lobe and linguela was then divided between clamps and the proximal part was closed. Left intercostal drain was inserted and the wound was closed. The baby had an uneventiful post-operative recovery. He is now two years old and had grown normally. During the first year of his life he presented once with tachypnea and wheezes and in 3 occasions with no symptoms but chest auscultation revealed scattered wheezes. During the second year, he was completely asymptomatic with clear chest X-ray.

Case No. 2

Seven days old Bahraini infant, the brother of Case No. 1 was referred to the Paediatric Surgical Unit with the suspicion of congential lobar emphysema. He was the product of full term normal delivery weighing 3.6 Kilograms at birth. He has four other normal siblings. The infant was noticed to be tachypnic four hours after birth which improved with oxygen therapy. On th fourth day of life his tachypnea increased and he gradually developed dyspnea. On examination he was tachypnic with laboured breathing. The left side of the chest was hyperresonant, breath sounds were reduced and wheezes audible. Chest X-ray showed hyperaereation of the left upper lobe with herniation across the anterior mediastinum. The right lung was compressed by the emphysematous lobe and by the shifted mediastinum (Figure 2). Emergency left upper lobectomy through posteriolateral thoracotomy was performed. The immediate postoperative chest X-ray showed right pneumothorax, but with the insertion of right intercostal tube, a good recovery resulted. One week after surgery the baby developed post-feeding tachypnea with audible wheezes on the right side of the Chest. Chest X-ray showed hyperaereation of the right middle lobe with moderate degree of herniation across the mediastinum and collapse of the right lower lobe (Figure 3). He was treated conservatively with oxygen therapy with each attack of tachypnea. The infant is now two months old and thriving well but occasionally he develops tachypnea with feeding. There was no progressive change on chest X-ray.



Chest X-ray one week after lobectomy of left upper lobe (Case No.2) showing hyperaereation of the right middle lobe with herniation across the mediastinum with severe mediastinal shift to the left. The lower right lobe shows partial collapse.



Microscopic appearence of emphysema with overdistension of the alveoli. The alveolar septae are widely separated and had clubbed endings.

PATHOLOGICAL EVALUATION

At operation, the emphysematous lobe was apparent by its over distension and the difficulty in collapsing it. The unaffected part of the lung was atelectatic. Both specimens reported in this paper were received in the laboratory fixed in 10 per cent formol saline. Reinflation and quantitative studies of the alveoli, arteries and bronchi were therefore not possible. Reconstruction from serial sections of bronchial pathways to demonstrate developmental bronchial anomalies was also not carried. No evidence of obstruction to the airways could be demonstrated. Microscopically the alveoli of the emphysematous parts appeared over-distended with air and the alveolar septae were widely separated and had clubbed endings (Figure 4). In the collapsed parts there were many spaces filled with polymorphs suggesting pulmonary infection and few alveoli were lined by hyaline membrane. In all the sections examined, the bronchial cartilage plates were all normal with no evidence of hypoplasia. In both cases, no convincing pathological aetiological factor was demonstrated as to the cause of congenital lobar emphysema.

DISCUSSION

Congenital lobar emphysema is predominently a condition of early infancy and for this reason it has been generally assumed to be a congenital disease. Occasionally it is not recognized until the forth month of life or later and in these cases it is probable that acquired respiratory infection plays a significant role in the pathogenesis. The familial tendency has been pointed out in siblings of infants with congenital lobar emphysema (6). But the significance of this tendency is not clear.

The severity of symptoms in infants with congenital lobar emphysema varies greatly . Some infants develop severe respiratory distress in the first few days of life while others progress with slower onset during later infancy, perhaps as coincident with respiratory infection. The symptoms usually appear shortly after birth with expiratory wheezing and tachypnea followed by dyspnea and, in severe cases cyanosis. Physical examination may reveal asymetery of the thorax due to unilateral over-inflation. The percussion note is increased, breath sounds reduced and rales and wheezes may be audible. The course is rapid, progressive and nearly always ends fatally unless the lobe is resected (11). Massive air trapping and over-distension within the involved lobe makes the latter physiologically useless and result in compression atelectasis of the ipsilateral lung and interference with the venous return to the heart. The X-ray manifestations are distinctive and present little difficulty in diagnosis. There is a markedly over-distended lobe with collapse of the normal lobes on the affected side and mediastinal shift into the uninvolved side with compression of the contralateral lung. Bronchoscopy and bronchography

are unnecessary and may be dangerous in a cyanotic baby. But on occasions when radiology is not diagnostic and in conditions in which the presence of foreign body is strongly suspected, bronchoscopy might be helpful.

Congenital lobar emphysema may affect any pulmonary lobe. Hendren and McKee (6) reviewed 106 cases collected from the English and French literature and reported seven cases of their own. In 46 cases the disease was reported to involve the left upper lobe, in 38 the right middle lobe, in 24 the right upper lobe, in two the lower lobes and in three more than one lobe.

Congenital lobar emphysema can arise from a variety of anatomic lesions : (a) Compression of the bronchi by extrinsic factors such as abnormally large or abnormally situated blood vessels (10, 15, 19, 21), enteric duplication or bronchogenic cyst (23, 24). (b) Intrinsic intrabronchial obstruction from foreign bodies, infection (11), inspissated secretions (23) and congenital redundant fold of the bronchial mucous membrane (16). (c) Increased collapsibility of the lobar bronchus during expiration due to congenital absence or hypoplasia of the bronchial cartilage (1, 20). This concept has been challenged (13) because the variability of the state of development of the bronchial cartilage in early life is not widely known. It would seem however, that the segmental cartilage anomalies are always overlooked because the involved segment is either left behind or clamped across during lobectomy and this renders microscopic examination impossible. (d) Developmental abnormalities of the alveoli resulting in loss of the elastic recoil (1) and congenital bronchial atresia and stenosis (9, 20). (e) Localized neuromuscular dysfunction (20). In their review of 113 cases with congenital lobar emphysema, Hendren and McKee (6) noted that 25 per cent of the cases were due to abnormalities of the bronchial cartilage, 13 per cent to intrinsic obstruction, 8 per cent to extrinsic obstruction, 4 per cent of diffuse bronchial abnormalities and 50 per cent to unknown causes. In the reported cases of this paper no convincing pathological cause could be demonstrated.

Applying quantitative techniques of the pulmonary alveoli, airways and arteries to a small number of formaline inflated specimens with lobar emphysema, three pathological type were described (5, 7, 8, 22); (a) Polyalveolar type in which the number of alveoli is increased, but the airways, acini and arteries are normal for age in number, size and structure. This suggest that the emphysematous condition represent gigantism of the alveolar region. (b) Overinflation of a normal lobe. In this type the alveolar number is normal for age but the volume of an alveolus is increased and hence there is a total increase in lung volume. The arteries and bronchi are normal in number and structure. This type suggest that the emphysema could follow bronchial obstruction. (c) Congenital emphysema with hypoplasia of the lung. In this type the involved lung is small in volume with reduced number of bronchi and arteries. The total alveolar number is less than normally found at birth but the alveoli are eight times the normal volume.

The treatment of choice is surgical removal of the affected lobe. This might be indicated urgently, as in our cases, or electively in milder forms. Anaesthesia must not be started until all are in readiness for expeditious thoracotomy. Excessive positive pressure must be avoided during induction of anaesthesia lest the over distended lobe get even larger and cause further embarassment to the cardiopulmonary function in an already precariously ill infant (3). At operation the affected lobe is apparent by its over distension. The unaffected part of the lung which must be saved is atelectatic.. No emphysematous portion should be left as this will result in continued respiratory embarassment. This is because of the progressive nature of the disease. The results of lobectomy are excellent and in the review of Hendren and McKee (6) in 101 cases with congenital lobar emphysema treated surgically by lobectomy there were only four deaths. The mortality rate following lobectomy was calculated as 7 per cent resulting from brain damage from postoperative hypoxia, congenital heart disease, postoperative pneumothorax and aspiration of mucous (14). Conservative surgery in the form of thoracocentesis, bronchial dilatation, removal of aberrant blood vessel and tube thoracotomy have also been tried in eight cases with three deaths (6).

There are few advocates of conservative management but the results are uncertain. Needle aspiration of the emphysematous lobe had been suggested as a definitive treatment but pneumothorax is a hazard and the ultimate outlook is not clear. Death after aspiration has been reported along with many fatal mishaps (6). Nonethless it is of interest to note the unique report of Roghair (17) of two children with dyspnea from what appeared to be congenital lobar emphysema. The children received no treatment and were followed into adult life and were found to be asymptomatic with normal chest X-ray appearences. This report

encouraged us to avoid further surgical intervention in case No. 2 after he developed emphysema in the right middle lobe, particularly that his symptoms did improve with oxygen therapy.

There are few reports of long term follow up of infants with congenital lobar emphysema treated by lobectomy. Sloan (18) followed six patients till adulthood. All the children had grown up and developed normally with leading active role in life and some with competitive sport activities. The pulmonary physiological tests showed decreased vital capacity proportional to the amount of lung tissue removed and normal or slightly increased total lung volume suggesting compensatory growth and over distension of the remaining lung (18). Hyperinflation of another lobe has also been described (2). Tapper et al (22) also reported a follow up of over 10-20 years and described decreased specific airway conductance, low forced expiratory volume and low maximum mid-expiratory flow rate. Other late signs and symptoms include recurrent episodes of wheezing as reported by Hendren and McKee (6) in 4 of their 7 patients. In our present cases, the first baby was followed for 2 years. In his first year he presented once with cough, tachypnea and wheezes and on three occasions with no symptoms but chest auscultation revealed scattered wheezes. During the second year he was completely asymptomatic with clear chest X-ray. The second baby developed congenital lobar emphysema of the right middle lobe causing tachypnea after feeding; this improved with oxygen therapy. The X-ray findings were persistent but not progressive. In their review of 113 cases, Hendren and McKee (6) found only three cases with congenital lobar emphysema in another lobe.

ACKNOWLEDGEMENT

We wish to thank Dr. W.F. Whimster for the review of microscopic section of case No. 1 and Dr. P. Rao for his help in the anaesthesia of both cases.

REFERENCES

1. Bolande, R.B. ; Schneider, A.F. ; Boggs, J.D. : Infantile lobar emphysema ; an aetiological concept. Arch. Path., 1956 ; 61 : 289.
2. Brooks, J.G. Pediatrics, Librairie du Liban, 1980, p 225.

3. Cote : The anaesthetic management of congenital lobar emphysema. J. Anaesthesiol., 1978 ; 94 : 296.
4. Fagan, C.J. ; Swischuk, L.E. : The opaque lung in lobar emphysema. Am. J. Roentgenol. Radium Ther. Nucl. Med., 1972 ; 114 : 300
5. Henderson, R. ; Hislop, A. ; Reid, L. : New pathological findings in emphysema of childhood : 3. Unilateral congenital emphysema with hypoplasia and compensatory emphysema of contralateral lung. Thorax, 1971 ; 26:195.

6. Hendren, W.H. ; McKee, D. M. : Lobar emphysema of infancy. J. Paeditr. Surg., 1966 ; 11 : 24.
7. Hislop, A.; Reid, L.: New Pathological findings in emphysema of childhood. : 1. Polyalveolar lobe with emphysema. Thorax, 1970 ; 25 : 682.

8. Hislop, A.; Reid, L. : New Pathological findings in emphysema of childhood : 2. Overinflation of a normal lobe. Thorax, 1971 ; 26 : 190.

9. Lacquet, L.K. ; Fornhoff, M. ; Dierickx, R. ; Buyssons, N. : Bronchial atresia with corresponding segmental pulmonary emphysema. Thorax, 1971 ; 26 : 68.

10. Leape, L.L. ; Ching, N. ; Holder, T.M. ; Lobar emphysema and patent ductus arteriosus ; development of emphysema while under observation. Pediatrics, 1970 ; 46 : 97.

11. Leape, L.L. ; Longino , L.A. : Infantile lobar emphysema. Pediatrics, 1964 ; 34 : 246.

12. Lincoln, J.C.R. ; Stark, J. ; Subramanian, S. ; Aberdeen, E. ; Bonhan — Carter, R.E; Berry, C.L. ; Patu, M.C. ; Waterson, D.J. Congenital lobar emphysema. Ann. Surg., 1971 ; 1973 : 55.

13. Millard, M. : Lung, pleura and mediastinum. In : Anderson, W.A.D. and Kissane, J.M. eds. Pathology. vol. II. St. Louis : C.V. Mosby Co., 1977 : 1038.

14. Murray, G.F. ; Collective review. Congenital lobar emphysema. Surg. Gynecol. Obstet., 1967 ; 124 : 611.

15. Pierce, W.S. ; de Paredes, C.G. ; Friedman, S. ; Waldhausen, J.A. : Concomitant congenital heart disease and lobar emphysema in infants ; incidence, diagnosis and operative management. Ann. Surg., 1970 ; 172 : 951.

16. Robertson, R. ; James, E.S. : Congenital lobar emphysema. Pediatrics, 1951 ; 8 : 795.

17. Roghair, G.D. : Nonoperative management of lobar emphysema : Long term follow-up. Radiology, 1972 ; 102 : 125.

18. Sloan, H. : Lobar obstructive emphysema in infancy treated by lobectomy. J. Thorac. Surg., 1953 ; 26 : 1.

19. Stanger, P. ; Lucas, R.V. ; Edwards, J.E. : Anatomic factors causing respiratory distress in acyanotic congenital cardiac disease, special reference to bronchial observation. Pediatrics, 1969 ; 43 : 760.

20. Stovin, P.G.I. ; Congenital lobar emphysema. Thorax, 1959 ; 14 : 254.

21. Strunge, P. : Infantile lobar emphysema with lobar agenesia and congenital heart disease. Acta Paediatr. Scand., 1972 ; 61 : 209.

22. Tapper, D ; Schuster, S. ; McBride, J. ; Eraklis, A. ; Wohl. M.E. ; Williams, A.; Reid, L. : Polyalveolar lobe. Anatomic and physiologic parameters and their relationship to congenital lobar emphysema. J. Pediatr. Surg., 1980 ; 15 : 931.

23. Thomson, J.; Forfar, J.O. ; Regional obstructive emphysema in infancy. Arch. Dis. Childh. , 1958; 33:97.

24. Weichert, R.F. Lindsey, E.S.; Pearce, C.W.; Waring, W.W.: Bronchogenic cyst with unilateral obstructive emphysema. J. Thorac. Cardiovasc. Surg. , 1970 ; 59 : 287.