Ventilation Scintigraphy with Submicronic Radioaerosol as an Adjunct in the Diagnosis of Congenital Lobar Emphysema

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When clinical examination and routine chest x-ray do not adequately explain neonatal respiratory distress, lung scintigraphy using a submicronic aerosol particle may be most helpful. Three cases illustrating this point are presented. Discussion centers around the diagnosis of an atypical case of congenital lobar emphysema (CLE), and differentiating between CLE, foreign body aspiration and compensatory hyperinflation in neonates with respiratory distress. Conservative and surgical treatment options for CLE are also illustrated.

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The assessment of regional lung function in infants is difficult when either clinical or simple radiographic examinations are performed. The utility of a submicronic aerosol containing technetium-99m diethylenetriaminepentaacetate ([^{99m}Tc]DTPA) in assessing ventilation in infants has recently been reported (*1*). The major advantage of this method over radioactive xenon is that it does not requires patient cooperation and, in contrast to krypton-81m, the nuclide and aerosol generator are widely available.

The purpose of the present report is to demonstrate the usefulness of ventilation scintigraphy with a submicronic radiolabeled aerosol in the diagnosis of congenital lobar emphysema (CLE). The first example is a typical case of CLE in infancy. The second report describes an unusual presentation of CLE, while the third case illustrates how the ventilation scintigram altered the clinical diagnosis and led to the correct management of the patient.

METHODS

The ventilation scintigram is performed by having the child breath for 2 min from a 2-l anesthetic bag containing a submicronic aerosol of [^{99m}Tc]sulfur colloid ([^{99m}Tc]SC) as previously described (*1*). The colloid used* is on average only 0.2–0.8 microns in diameter, thus enabling the labeling of the submicronic aerosol. The advantages of [^{99m}Tc]SC colloid over [^{99m}Tc]DTPA have been described by Coates et al. (*2*).

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CASE REPORTS

Case 1

This infant was born by cesarian section after a full term and uncomplicated pregnancy. The child was well until 5 wk of age when he developed a febrile illness with no other symptoms except for difficulty in feeding. Clinical examination was normal except for the rectal temperature of 39.5°C and decreased air entry in the right upper thorax. The chest radiograph (Fig. 1A) revealed mild generalized hyperinflation of both lungs but the left upper lobe appeared overinflated relative to the other lobes. This latter impression was further confirmed on fluoroscopy. A ventilation scintigram was then obtained. This demonstrated absent ventilation to the left upper lobe (Fig. 1B). A diagnosis of congenital lobar emphysema was made.

The fever resolved spontaneously, and the child was subsequently discharged. He has been followed for 2 yr and no complications have occurred. There has not been any respiratory distress. Lobectomy has not been performed.

Case 2

This infant was delivered by Cesarian section in a peripheral hospital after an uneventful pregnancy. The Apgar scores were 5 and 7 at 1 and 5 min. Subsequently the infant became bradycardic and bradypneic. The infant became cyanotic in room air and intercostal retractions and rales were noted upon examination of the chest. A chest radiograph (Fig. 2A) showed an area of increased opacity in the right lung. Congenital pneumonia was suspected and after a septic workup, the infant

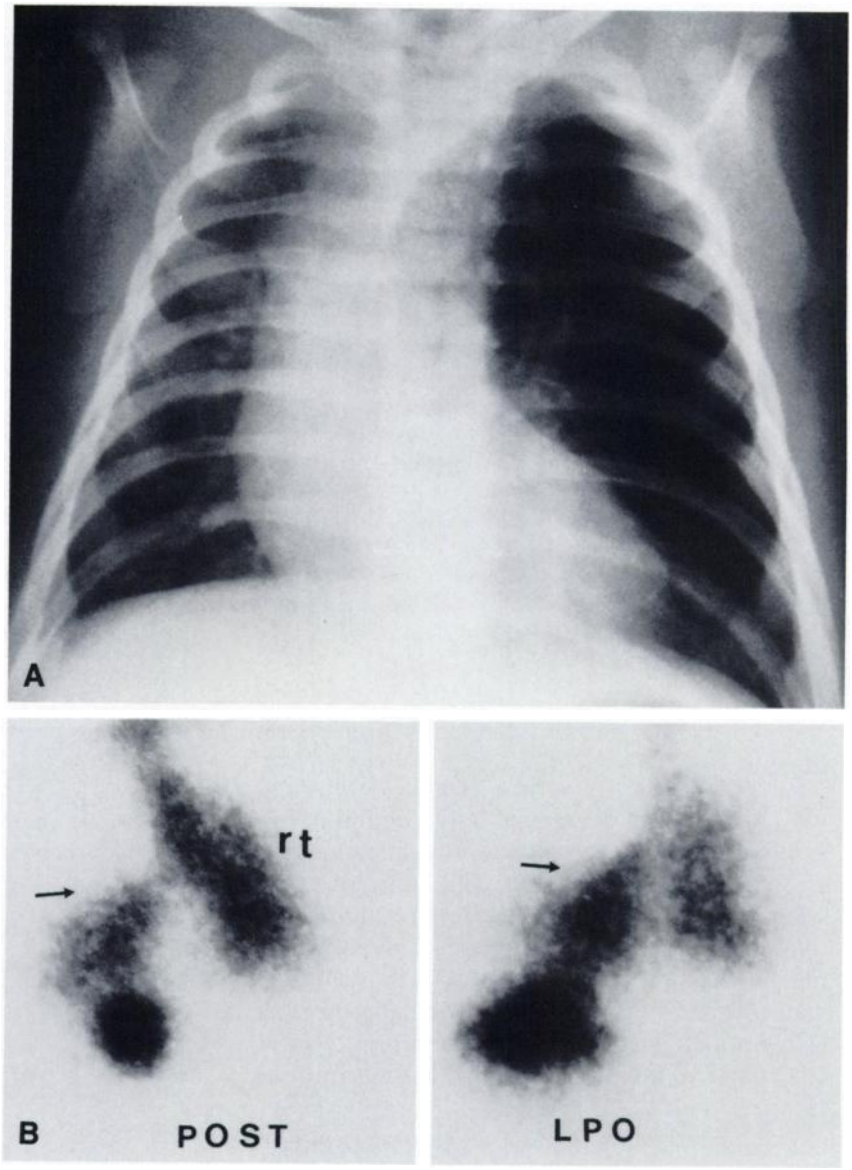

FIGURE 1
A: Case 1. Chest x-ray. There is hyperlucency of the left upper lobe with herniation across the midline and compression of the heart and mediastinum towards the right. B: Posterior, left posterior oblique ventilation scintigraphs show a ventilation defect in the region of the left upper lobe. Note some of the Tc-S colloid has been swallowed and is seen in the esophagus and stomach.

was started on antibiotics. The infant continued to require supplemental oxygen and developed increasing respiratory distress. She was then transferred to our Medical Centre.

Examination on admission revealed a respiratory rate of 34 and mild intercostal indrawing. Decreased air entry was appreciated on the right but there were no adventitial sounds heard. The remainder of the examination was normal. A chest radiograph on admission demonstrated partial clearing of the opacity in the right lung and ventilation/perfusion scintigrams showed absence of ventilation and perfusion in the right middle lobe (Fig. 2B). A diagnosis of congenital lobar emphysema was suspected. Repeat chest radiograph demonstrated progressive clearing of the opacity in the right lung with the development of the classic hyperinflation of the right middle lobe which is expected with CLE.

Over the next month the child developed increasing hyperinflation of the right middle lobe with virtual collapse of the right upper lobe and right lower lobe on the chest radiograph. Computerized axial tomography (Fig. 2C) showed hyperinflation of the right middle lobe causing shift of the mediastinum to the left. Clinically the child developed increasing respiratory distress, and underwent lobectomy (*3*). Pathologic examination confirmed the diagnosis of congenital lobar emphysema. Her recovery was uneventful.

Case 3

This infant was born prematurely at 30 wk gestation. He required intubation and assisted ventilation for severe hyaline membrane disease. Following extubation at 14 days of life he developed right middle and lower lobe collapse associated with overinflation of the right upper lobe. This was unresponsive to medical therapy. Bronchoscopy was performed 1 wk later and no obstructing lesion could be identified. The infant was subsequently discharged at 2 mo of age. Over the next 2 mo the chest radiograph remained unchanged despite aggressive bronchodilator therapy and physiotherapy. Surgical consultation was obtained and the possibility of congenital lobar emphysema of the right upper lobe with subsequent compres-

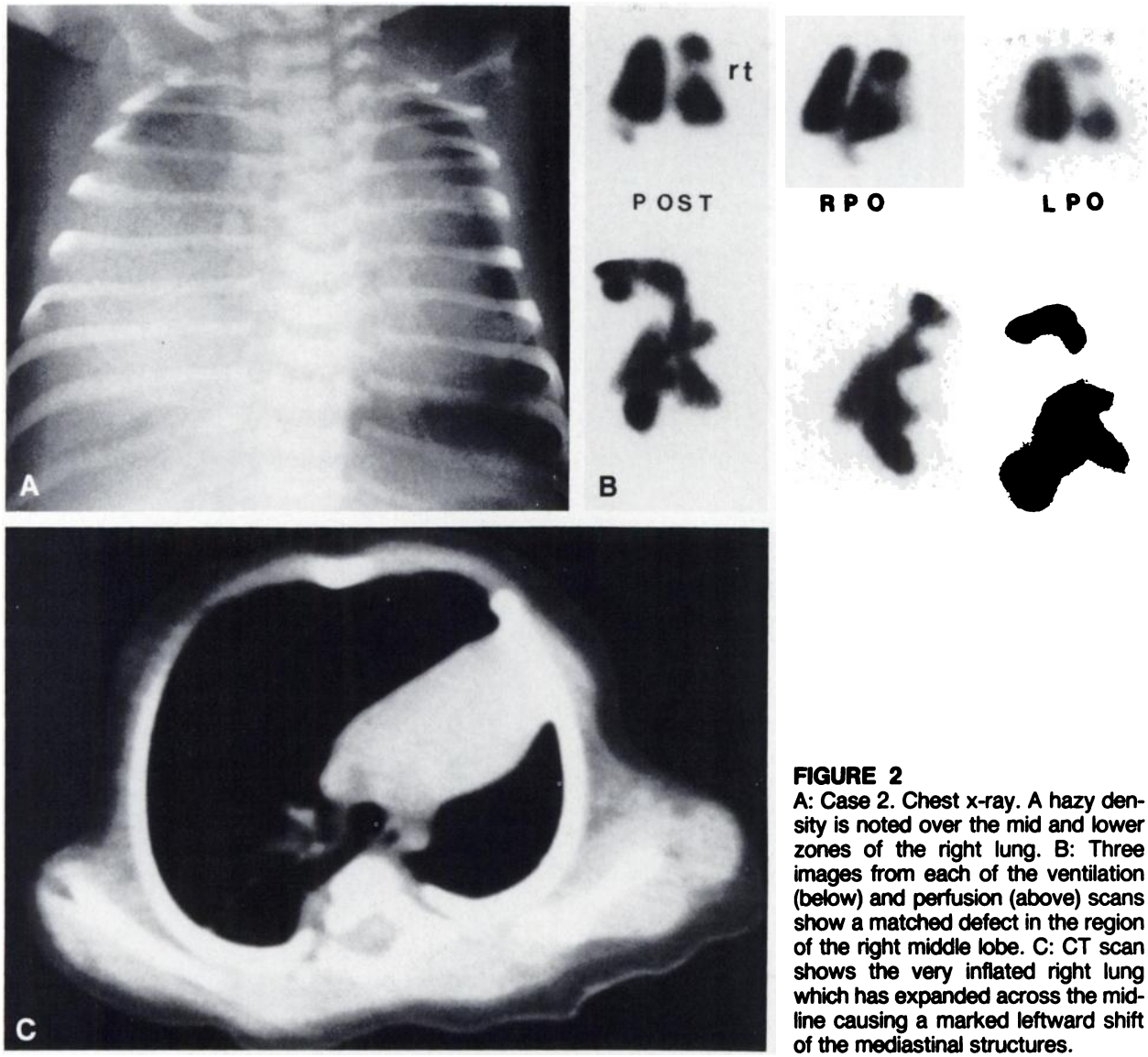

FIGURE 2
A: Case 2. Chest x-ray. A hazy density is noted over the mid and lower zones of the right lung. B: Three images from each of the ventilation (below) and perfusion (above) scans show a matched defect in the region of the right middle lobe. C: CT scan shows the very inflated right lung which has expanded across the midline causing a marked leftward shift of the mediastinal structures.

sion of the right middle and lower lobes was suggested (Fig. 3A). Ventilation and perfusion scintigrams were obtained which showed normal ventilation and perfusion to the right upper lobe thus ruling out CLE (Fig. 3B). Perfusion and ventilation were reduced in the right base. Continuing medical therapy did not result in re-expansion of the right lower lobe. A repeat bronchoscopy demonstrated stenosis of the bronchus intermedius presumably due to repeated suctioning as an infant. Balloon dilatation of the stenotic area resulted in re-expansion of the right middle and lower lobes (Fig. 3C).

DISCUSSION

The infant or child presenting with respiratory distress presents a pressing challenge to the physician and an emotional burden to the parents. When clinical examination and the chest x-ray do not readily point to a diagnosis, lung scintigraphy may be helpful.

Once there is clinical suspicion of CLE, the diagnosis will often be supported by the chest radiograph with radiolucency of the involved area. Secondary compressive effects of the overexpanded lobe (such as tracheal or mediastinal shift away from the affected lobe, atelectasis of adjacent lobes or herniation of the expanded lobes across the midline) are additional features pointing to the diagnosis (*4*). However, as Case 2 illustrates, the initial chest x-ray may not show this "typical appearance." The child may present with a region of increased density on the chest x-ray than hyperlucency. This can be mistaken for pneumonia. This "atypical" presentation of CLE has been reported before (*5,6*) and if the secondary compressive signs are present should suggest the diagnosis. The density is thought to be due to fetal lung fluid which is slow to clear from the affected lung. Within several days the fluid clears and the more typical pattern of a radiolucent lobe with sparse bronchovascular markings is appreciated. In our case the density was not in a classic radiographic lobar pattern

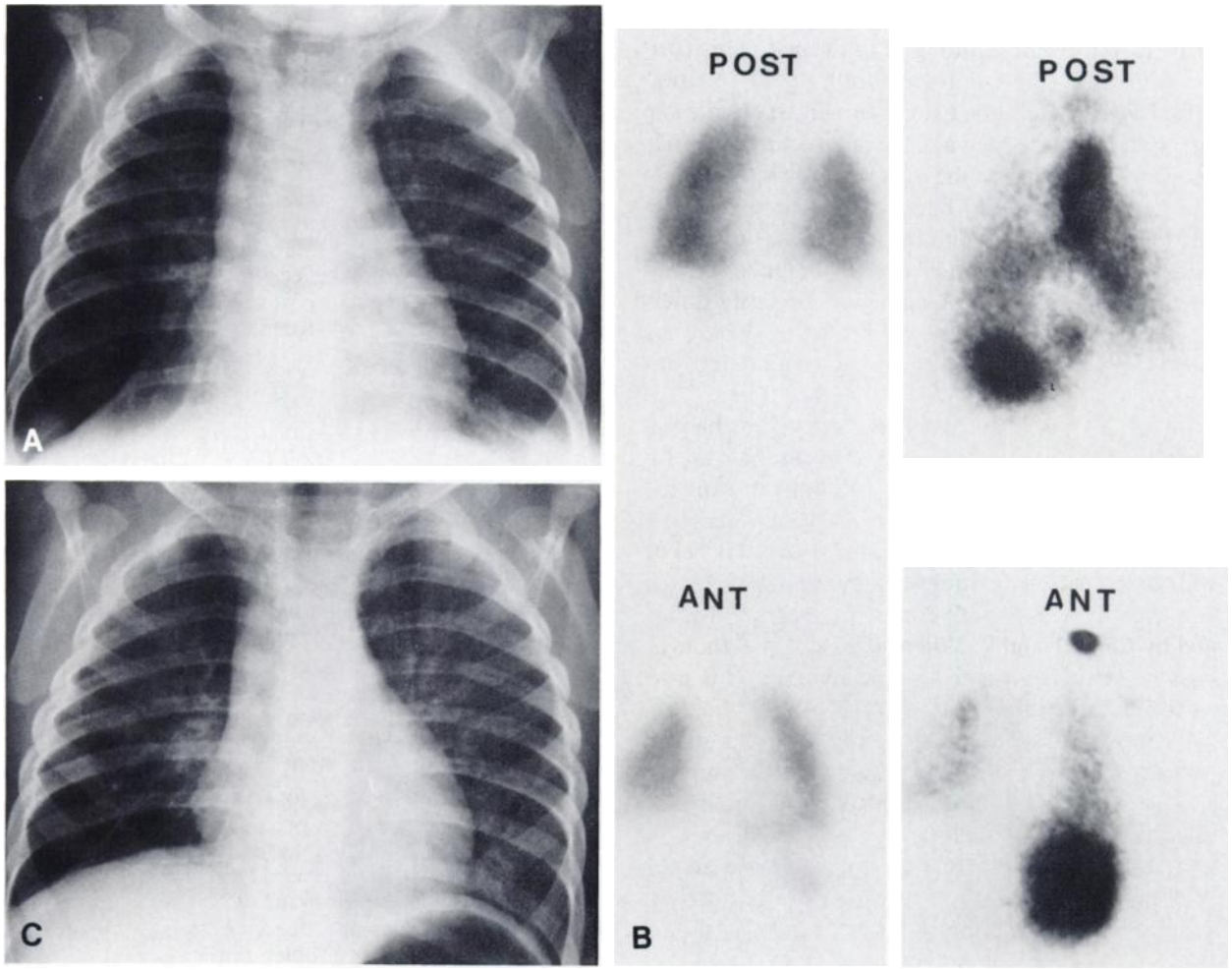

FIGURE 3
A: Case 3. Chest x-ray shows increased opacity in the right lower lung zone consistent with collapse of the right middle and lower lobes. The remaining right lung is hyperlucent compared to the left lung. B: Anterior and posterior views from the perfusion (left) and ventilation (right) scans fail to show decreased ventilation or perfusion in the right upper lobe. Perfusion and ventilation are reduced at the right base. C: Chest x-ray. Postoperatively the collapsed right lobes have now undergone re-expansion.

as has been described by others. Lung scintigraphy demonstrated the typical lobar pattern and pointed to the diagnosis before the fluid cleared and spared the infant additional and possibly costly and invasive diagnostic testing.

Other diagnostic maneuvers for CLE which have been utilized include fluoroscopy, inspiration and expiration chest films, bronchography, bronchoscopy, angiography, CT and even exploratory thoracotomy (*7–10*). The advantages of the lung ventilation and perfusion scintigram over some of these techniques are apparent. First, it is a safe procedure. The radiation dose from the aerosol ventilation study is ~20 mrad to the lungs of an infant, i.e., approximately that of a chest x-ray (*1*). Gonadal dose is 0.75 mrad. For the perfusion study radiation dose to the lung is 1 mrad/μCi[^{99m}Tc] macroaggregated albumin injected, gonadal dose is 0.035 mrad/μCi. A ventilation scan may also obviate invasive procedures such as bronchoscopy. If an assessment of regional pulmonary blood flow is required a perfusion scintigram can be obtained by using an intravenous injection of ^{99}Tc-labeled macroaggregates of albumin (*1*). The perfusion scintigram will usually obviate the need for angiography, especially when combined with echocardiography to rule out associated congenital heart disease. Finally, the ventilation scintigram using a submicronic aerosol requires little patient cooperation and no sedation. This is not the case when radiogas is used.

The three principal disorders in the differential diagnosis of a radiolucent lobe would include a CLE, a foreign body, and compensatory hyperinflation secondary to atelectasis of other regions of the lung. Lung scintigraphy is helpful in differentiating these disorders. When a foreign body obstructs a major bronchus the ventilation scan will show a lobar or segmental defect

beyond the site of obstruction. Perfusion is still often present to the region although diminished. This contrasts with CLE where both perfusion and ventilation are decreased to the same extent. In our experience the lung scintigram has been valuable in directing the bronchoscopist's attention to the area with the most probable high yield for foreign body, especially to the right upper lobe area which is difficult to visualize in infants. Compensatory hyperinflation of one lung lobe secondary to atelectasis of other lung regions is easily differentiated from CLE as illustrated by Case 3 where the hyperinflated lobe had appropriate ventilation and perfusion.

Lung scintigraphy has also been utilized in the preoperative evaluation of CLE to ensure that there are no other abnormal areas which are not apparent on the chest x-ray, for example, bronchomalacia of other lobes (*10*). A recent communication reports segmental "lobar emphysema" detected with lung scintigraphy (*12*).

The treatment alternatives for CLE are well illustrated by Cases 1 and 2. Whereas once it was thought all cases should be treated surgically there are now reports of conservative treatment with no ill effect (*3, 13*). The earlier the presentation, and the more severe the respiratory distress, the more likely it is that surgery will be needed. It is really the "behavior" of the hyperinflated lobe as reflected clinically by repeated bouts of infection and respiratory embarassment due to atelectasis of adjacent lobes or compression of the opposite lung which dictates the necessity of surgery as shown by Case 2. When conservative management of CLE is undertaken the ability to quantitate the ventilation to various regions of the lungs (*1*) will aid in making appropriate clinical decisions.

In conclusion, the child with respiratory distress that is not adequately explained by clinical or routine chest x-ray may benefit from lung scintigraphy. This technique can be useful when the plain chest radiograph is insufficient to differentiate between foreign body aspiration, compensatory hyperinflation and atypical CLE.

NOTE

*Supplied by Chedoke-McMaster Regional Radiopharmacy.

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