RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

Unilateral primary lung hypoplasia diagnosed in adulthood

Stamatis Katsenos, MD, PhD, Elvira-Markela Antonogiannaki,MD, Konstantinos Tsintiris, MD

Department of Pneumonology, Arny General Hospital of Athens, Athens, Greece

Corresponding Author:

Stamatis Katsenos, MD, PhD

Department of Pneumonology

Army General Hospital of Athens,

158 Mesogion&Katehaki Avenue,

115 25 Athens,

Greece

Tel: (+30) 210 749 4328

Fax: (+30) 210 749 4095

E-mail: skatsenos@yahoo.gr

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

Abstract

Unilateral primary pulmonary hypoplasia is rare in adulthood. It is usually present in neonates or in early childhood and is characterized by a decreased number of bronchial segmentation and decreased/absent alveolar air space. Most patients have recurrent episodes of wheezing or pneumonia and severe respiratory distress leading to chronic respiratory failure whereas some may occasionally be asymptomatic. Herein, we present a case of left lung hypoplasia in an asymptomatic 28-year-old male, who was admitted for further investigation of an abnormal chest radiograph obtained as part of health evaluation for military service. Thorough work-up, including imaging modalities and bronchoscopy, disclosed a hypoplastic left lung, which was undiagnosed for 25 years. Embryological, clinical and diagnostic aspects are briefly discussed.

***Key words*:** *primary lung hypoplasia;computed tomography;bronchoscopy*

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

Introduction

Unilateral primary pulmonary hypoplasia is rare in adulthood. It is usually present in neonatal period or in early childhood and is characterized by a decreased number of bronchial segmentation and decreased/absent alveolar air space. The majority of patients presents with severe respiratory distress or repeated pulmonary infections and wheezing whereas some may be completely asymptomatic. In this paper, we present a case of left lung hypoplasia in an asymptomatic 28-year-old male, who was admitted for further investigation of an abnormal chest radiograph obtained as part of health evaluation for military service. The disorder had not been detected for 28 whole years.

Case Report

A 28-year old non-smoker male was referred to our department for further evaluation of an abnormal chest radiograph. A plain chest radiograph is compulsory test for Greek recruits as part of health evaluation for their military service. His past medical history was unremarkable. He occasionally reported mild episodes of common cold without seeking any medical attention. Chest clinical examination revealed diminished movement of the thorax on the left side as well as absence of breath sounds over the upper and middle left lung fields. Heart sounds were also heard on the left side. A posteroanterior chest radiograph showed opacification of the left hemithorax with decrease in its size and mediastinal displacement to the left with an increase in volume of right lung. The cardiac contours were indistinct (Figure A). Further imaging evaluation with multislice chest computed tomography demonstrated a hypoplastic left lung with right lung hyperinflation and herniation to the

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

contralateral side as well as displacement of the mediastinum to the left side. Moreover, absence of left main pulmonary artery and its branches into the left upper lobe and lingula as well as a decrease in size of left main stem bronchus were noted. Left lower lobe artery with the corresponding lobe were however present (Figure B). Fiberoptic bronchoscopy was then performed showing stenosis of the left main stem bronchus with absence of left upper lobe and lingula bronchi as well as left superior lower lobe segment (Figure C). The basal segments of left lower lobe were rudimentary (Figure D). Furthermore, the right tracheobronchial tree was completely developed but the orifices of upper lobe and middle lobe bronchus were in different position than usually located because of compensatory right lung expansion and consequent distortion of bronchial structures. A ventilation-perfusion scan of the chest showed complete absence of ventilation and perfusion in left upper lobe and lingula (Figure E). Pulmonary function tests demonstrated moderate to severe obstruction that was mildly responsive to inhaled bronchodilators. In particular, decreased values were noted in expiratory volumes (FVC, 76% of predicted; FEV1, 44% of predicted; FEV1/FVC ratio, 59% of predicted; peak expiratory flow, 25% of predicted) (Figure Fa). The measurement of static lung volumes revealed increased residual volume (RV-200% of predicted value) and total lung capacity (TLC-127% of predicted value). Diffusing capacity of the lung was found moderately reduced (DLCO-51% of predicted value) (Figure Fb). Additional work-up including echocardiography and abdominal CT disclosed no co-existent congenital abnormalities. Therefore, a diagnosis of primary left lung hypoplasia was made and the patient was recommended to ask medical help in the event of respiratory infections. In addition, inhaled b_2-agonists and steroids were administered to the patient despite their minor

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

bronchodilating effect. Preventive vaccination against influenza virus and pneumococcus was also recommended.

Discussion

Pulmonary underdevelopment has been classified into three groups by *Schneider and Schwalbe*.[1-3] In group 1, bronchus and lung are absent (agenesis); in group 2, a rudimentary bronchus is present and limited to a blind-end pouch without lung tissue (aplasia); and in group 3, there is bronchial hypoplasia with variable reduction of lung tissue (hypoplasia).

More specifically, pulmonary hypoplasia is defined as deficient or incomplete development of the lungs. The abnormality is characterized by the presence of both bronchi and alveoli in an underdeveloped lobe and may occur between the 4th and 24th gestational week. It can be categorized into two forms: primary and secondary. Most cases are usually secondary to conditions that limit fetal lung growth whereas primary pulmonary hypoplasia is rare and it is thought to be caused by an embryologic defect of the lung or vascular tissues or an in utero vascular accident.[4] The true prevalence is unknown, but in cases of premature rupture of membranes at 15–28 weeks gestation, the reported prevalence of pulmonary hypoplasia ranges from 9% to 28%.

Primary unilateral pulmonary hypoplasia is usually encountered in a child presenting with life-threatening symptomatology, e.g. early respiratory distress after birth, cyanosis, tachypnea, hypoxia, hypercapnea, and acidosis. However, it may be infrequently present in adults not producing any symptomatology thus rendering its diagnosis problematic.[5] Nevertheless, some adult patients may present with repeated pulmonary infections and wheezing.[6,7]

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

Plain radiographs demonstrate decreased aeration of the affected hemithorax and a small thoracic cage. A common finding is a shift of the mediastinum to the side of the hypoplasia, accentuated during inspiration due to increased compensatory ventilation of the other lung. A retrosternal soft tissue density can also be visible on lateral chest films representing heart and mediastinum that are displaced into the anterior chest. Multidetector CT provides a non-invasive modality of confirming the diagnosis by clearly delineating the deformed and stunted pulmonary vasculature and bronchial tree.[8,9] Bronchoscopy, ventilation-perfusion scanning or CT-angiography can equally identify absence of bronchovascular structures.

Differential diagnosis primarily includes congenital cystic adenomatoid malformation, pulmonary sequestration, congenital lobar emphysema, acquired lung collapse and secondary pneumopathy with associated non-congenital bronchiectasis (pseudo-hypoplasia).[2,10] However, there was no clinical or imaging evidence to support any of these entities in the present case. More frequently, pulmonary hypoplasia is secondary to other fetal developmental abnormalities, such as congenital diaphragmatic hernia, neuromuscular disorders, congenital heart diseases, thoracic cage anomalies (e.g. asphyxiating thoracic dystrophy- Jeune syndrome), genitourinary tract anomalies (e.g. Potter's syndrome-oligohydramnios tetrad), chromosomal aberrations and congenital pulmonary venolobar syndrome (Scimitar syndrome). These disorders occur in neonates and influence adversely morbidity and survival.[11] However, it seems that asymptomatic unilateral primary hypoplasia in adulthood exhibits long survival due to compensatory hypertrophy of the contralateral lung that occupies the ipsilateral hemithorax. The most favorable survival has been observed in cases of left lung hypoplasia because of the satisfactory compensatory hypertrophy of the larger right lung.[2,8] With regard to the present case, we speculate that lung hypoplasia should have

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

been occurred in the first 7-10 weeks of gestational age despite the fact that the individual had no symptoms referable to the chest for 28 consecutive years. It is quite likely that right lung compensatory expansion prevented the presence of chest symptomatology.

Pneumothorax and pulmonary hypertension are common serious complications. Pneumothorax often develops spontaneously or secondary to mechanical ventilation in infants who present with severe respiratory insufficiency in the first few hours of their life. Last but not least, pulmonary infections can cause life-threatening events in patients with this congenital malformation. The underdevelopment of the alveolar tissue results in a small fibrotic and non functioning lung with associated surfactan deficiency and impaired mucociliary clearance thus rendering the host susceptible to greater bacterial or viral proliferation.

In conclusion, unilateral primary lung hypoplasia is very rare in adulthood. Actually, the disease is usually established in perinatal or neonatal period. Symptomatology is generally present despite the occasional accidental discovery of hypoplastic lung in an asymptomatic adult. Multidetector CT is instrumental imaging modality in establishing the diagnosis of pulmonary hypoplasia, since it can discriminate the abnormalities of the bronchovascular structures.

REFERENCES

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited
and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

1. Lee EY, Dorkin H, Vargas SO. Congenital pulmonary malformation in pediatric patients: review and update on etiology, classification and imaging findings. Radiol Clin North Am 2011;49(5):921-948.

2. Berrocal T, Madrid C, Novo S, Gutiérrez J, Arjouilla A, Gómez-León N. Congenital anomalies of the tracheobronchial tree, lung and mediastinum: embryology, radiology, and pathology. Radiographics 2004; 24(1):e17.

3. Schneider P, Schwalbe E. Die morphologie der missbildungen des menschen und der tiere. Jena: Fischer 1912; 3:812-822.

4. Porter H. Pulmonary hypoplasia. Arch Dis Child Fetal Neonatal Ed 1999; 81(1):81F-83F.

5. Pathania M, Lali BS, Rathaur VK. Unilateral pulmonary hypoplasia: a rare clinical presentation. BMJ Case Rep 2013; 2013.

6.Thomas RJ, Lathif HC, Sen S, Zachariah N, Chako J. Varied presentations of unilateral lung hypoplasia and agenesis: a report of four cases. Pediatr Surg Int 1998; 14(1-2):94-95.

7. Comet R, Mirapeix RM, Marin A, Castañer E, Sans J, Domingo C. Pulmonary hypoplasia in adults: embryology, clinical presentation and diagnostic methods. Our experience and review of the literature. Arch Bronconeumol 1998; 34(1):48-51.

8. Georgescu A, Nuta C, Boudari S. 3D imaging in unilateral primary pulmonary hypoplasia in an adult: a case report. Case Rep Radiol 2011; 2011:659586.

9. Beigelman C, Howarth NR, Chartrand-Lefebvre C, Grenier P. Congenital anomalies of the tracheobronchial branching patterns: spiral CT aspects in adults. Eur Radiol 1998; 8(1):79-85.

10. Çay A, Sarihan H. Congenital malformation of the lung. J Cardiovasc Surg 2000; 41(3):507-510.

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

11. Currarino G, Williams B. Causes of congenital unilateral pulmonary hypoplasia: a study of 33 cases. Pediatr Radiol 1985; 15(1):15-24.

FIGURE LEGENDS

Figure A. A posteroanterior chest radiograph showing opacification of the left hemithorax with decrease in its size and mediastinal shift to the left with an increase in volume of right lung. The heart outline is indistinct.

Figure B. Axial chest computed tomography demonstrating a hypoplastic left lung with right lung hyperinflation and herniation to the contralateral side as well as displacement of the mediastinum to the left side. Absence of left main pulmonary artery and its branches into the left upper lobe and lingula as well as a decrease in size of left main stem bronchus is also visible.

Figure C. Fiberoptic bronchoscopy showing stenosis of the left main stem bronchus with absence of left upper lobe and lingula bronchi as well as left superior lower lobe segment. MC=main carina, RMB=right main stem bronchus, LMB=Left main stem bronchus, RUL=right upper lobe, RML=right middle lobe, RB6=superior segment of right lower lobe, RB8-10=basal segments of right lower lobe

Figure D. Fiberoptic bronchoscopy showing basal segments of left lower lobe. LMB=left main stem bronchus, LB8-10=basal segments of left lower lobe.

Figure E. Perfusion and ventilation scintigram (posterior view) showing perfusion and ventilation only in the left lower lobe. R=right lung, L=left lung.

Figure Fa. Spirometry results and flow-volume curve.

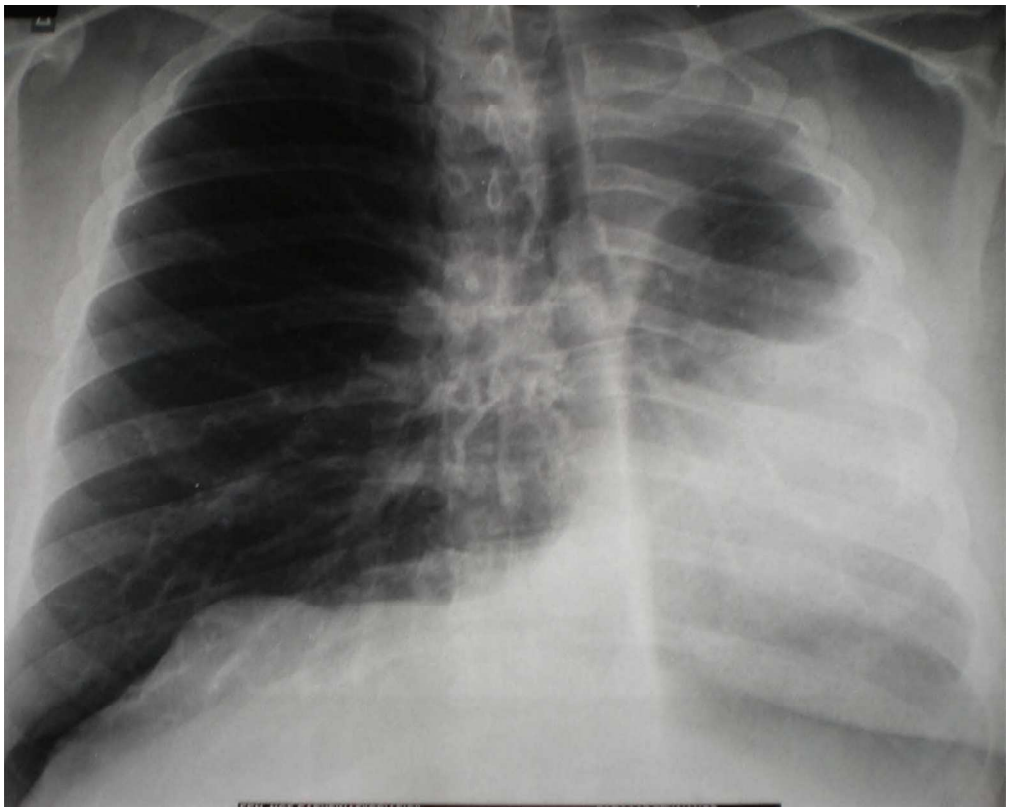
Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740

Figure Fb,c. Static lung volume and diffusing capacity for carbon monoxide measurements.

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

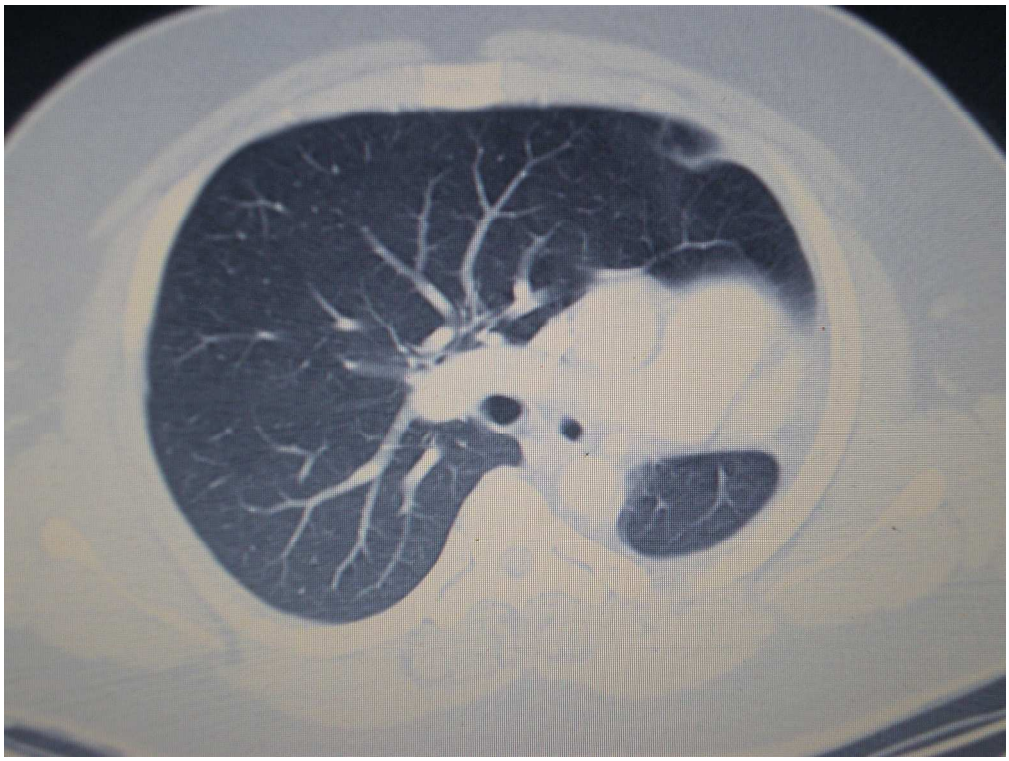
RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740



127x101mm (300 x 300 DPI)

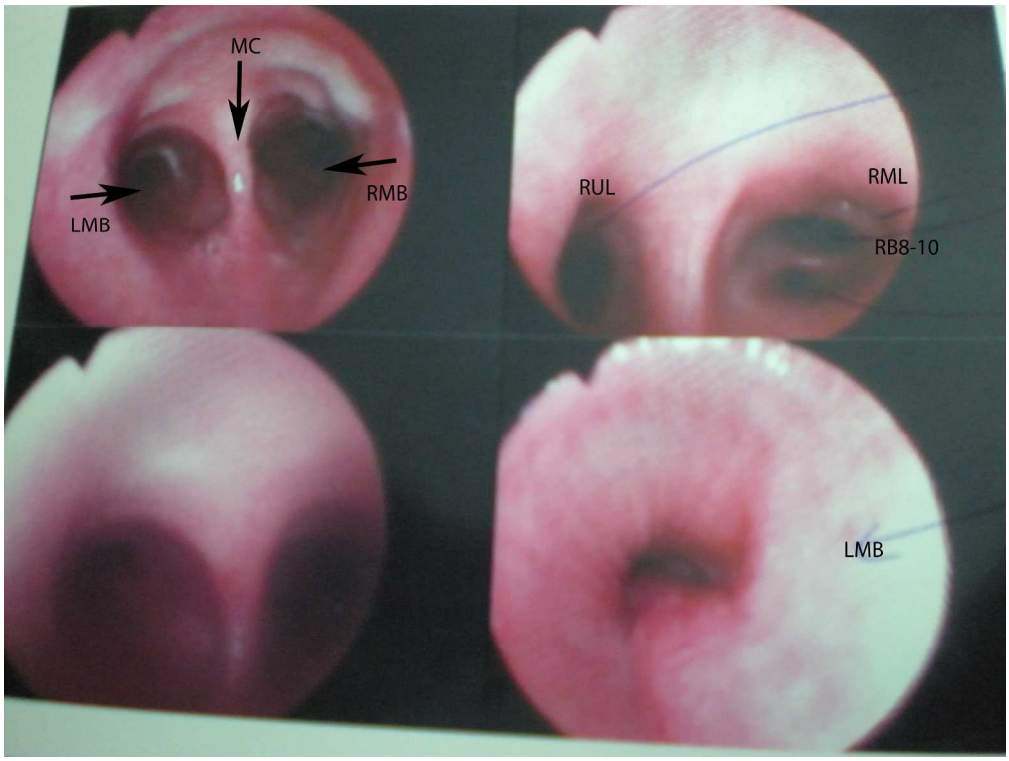
Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited
and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740



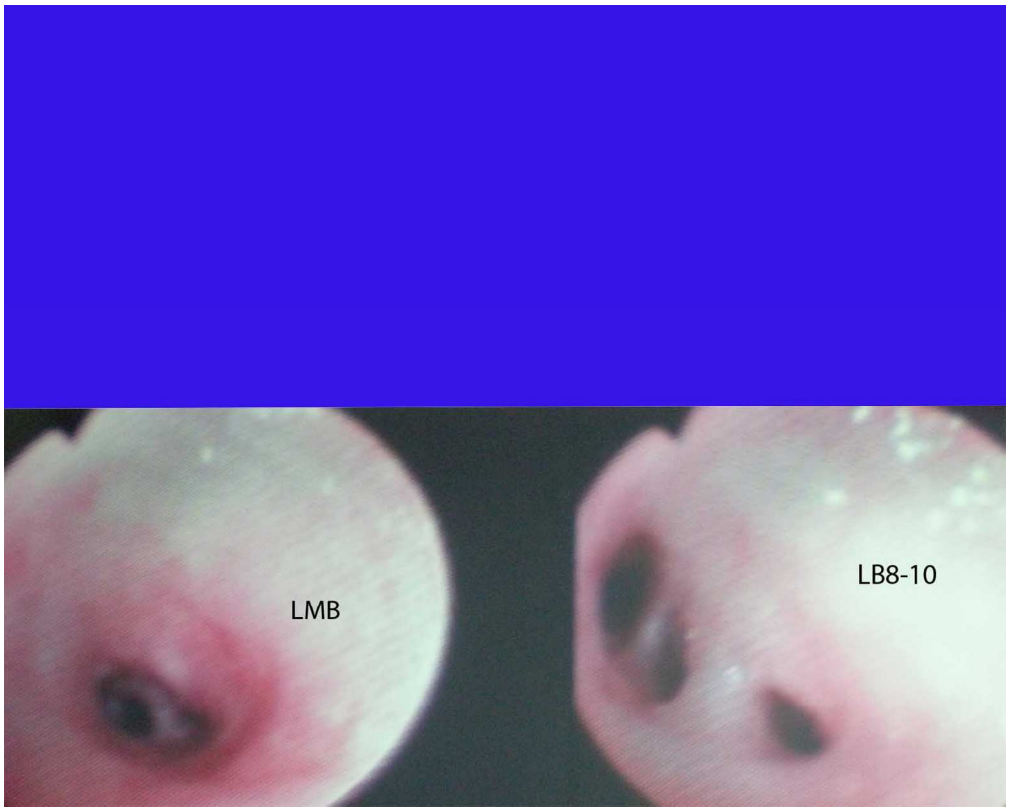
Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited
and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740



Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited
and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

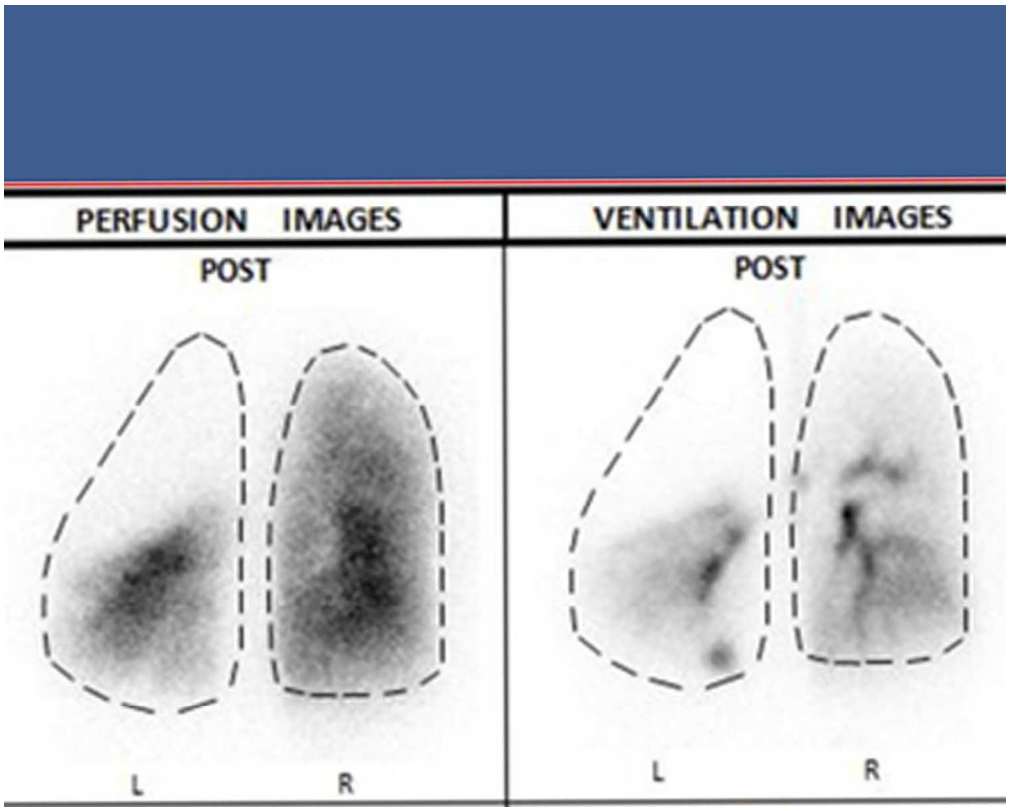
RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740



127x101mm (300 x 300 DPI)

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited
and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.

RESPIRATORY CARE Paper in Press. Published on September 10, 2013 as DOI: 10.4187/respcare.02740



127x101mm (300 x 300 DPI)

Copyright (C) 2013 Daedalus Enterprises
Epub ahead of print papers have been peer-reviewed and accepted for publication but are posted before being copy edited
and proofread, and as a result, may differ substantially when published in final version in the online and print editions of RESPIRATORY CARE.