Pulmonary Atelectasis in Children

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Pulmonary atelectasis is a common problem in clinical practice, especially in cases involving children, and is mainly caused by resorptive atelectasis, compressive atelectasis, and adhesive atelectasis. Diagnosis of this problem requires the recording of complete histories, physical examinations, and investigations that demand interpretative experience. Chest radiographs using both anterior-posterior and lateral views are also important in diagnosis. New investigative techniques, such as F-18 FDG-PET/CT imaging to differentiate atelectatic lungs from tumors, have been developed. Many treatment modalities lack evidence-bases but possess clinical benefits. These modalities include chest physiotherapy, bronchodilators, recruitment maneuvers and nebulized or direct tracheal application of recombinant human DNase I (rhDNase I) that can be used with regular patients and patients with cystic fibrosis.

Keywords: Pulmonary atelectasis, chest radiograph, chest physiotherapy

Introduction

Pulmonary atelectasis is a common health problem, especially in association with developing lungs. Children are predisposed to pulmonary atelectasis due to their smaller and more collapsible airways, more compliant chest wall, and incomplete development of collateral ventilation through pores of Kohn and channels of Lambert. However, early diagnosis and management of this pathology can lead to full recovery.
Definition

Pulmonary atelectasis refers to collapsed and non-aerated regions of the lung parenchyma. It is mostly related to abnormalities of chest walls and lung parenchyma. 1,2

Causes

Pulmonary atelectasis in childhood is mainly caused by resorptive atelectasis, compressive atelectasis, and adhesive atelectasis.

Resorptive atelectasis is the most common cause in children due to airway obstructions that lead to non-ventilation of distal airways. 1,2 Gas in the regions of the sites of the obstructions are completely absorbed by the pulmonary blood flow through those regions. 1,3 Airway obstructions may be due to exogenous causes, such as foreign body aspiration, or due to endogenous causes, such as polyps, papillomas, mucus plugs in patients with cystic fibrosis, and bronchial wall edema in patients with asthma. 1,2 The absorption of alveolar gas is also implicated in the development of intraoperative atelectasis, even in the absence of airway obstructions due to the use of high inspired fractions of oxygen. 4

Compressive atelectasis is caused by (i) extrinsic bronchial compression, (ii) intra-thoracic compression, and (iii) chest wall defects and neuromuscular disease. Extrinsic bronchial compression is the most common cause due to the enlargement of hilar or mediastinal lymph nodes by diseases such as tuberculosis, lymphomas, and metastatic tumors. 1,2 Intra-thoracic compression is a result of cardiomegaly, pneumothorax, chylothorax, or hemothorax which occupy space in the mediastinum and compress the lung parenchyma. 1,2 Abnormalities of the diaphragm caused by diaphragmatic hernia and neuromuscular diseases such as Guillain Barre syndrome can produce atelectasis by compression of the bronchi by displaced viscera. 1,2

Adhesive atelectasis is caused by a surfactant deficiency or dysfunction that leads to increased surface tension in acute respiratory distress syndrome (ARDS), near-drowning, hyaline membrane disease, pneumonia, pulmonary edema, and patients with cardiopulmonary bypasses. 1,2,5

Pathophysiology

Atelectasis is caused by a complete airway obstruction. The gases trapped by a complete obstruction are absorbed by the pulmonary blood flow. The rate of gas absorption depends on the gas solubility – respirable air nitrogen is absorbed in 2 to 3 hours but 100% oxygen is absorbed in 2 to 3 minutes. 1 For this reason atelectasis is more common in patients who receive high concentrations of oxygen, such as those receiving treatment for hyaline membrane disease, or intra- and post-operative patients, especially those having general anesthesia. 1,4,6,7 Intra-operative patients or patients in intensive care units have low functional residual capacity (FRC) and are prone to atelectasis due to two reasons, firstly, the positive pressure ventilation by which the thoracic central blood shifts to the abdomen and increases abdominal pressure and upward movement of the diaphragm, and secondly, neuromuscular blockages and diaphragm paralysis. 3,4,6,7

Compressive atelectasis is caused by direct compression of the lung parenchyma by extrinsic compressive factors such as congenital heart disease, vascular malformation, and cardiomegaly. Also, heart disease with pulmonary edema may cause atelectasis from restrictive defects. 1,8

Adhesive atelectasis is caused by a surfactant deficiency or dysfunction that leads to increased surface tension and unstable alveoli. 1,2,5 Cardiac surgery with cardiopulmonary bypasses induces profound changes in surfactant systems resulting in adhesive atelectasis. 5 Atelectasis produces alveolar hypoxia and pulmonary vasoconstriction to prevent ventilation-perfusion mismatching and reduces arterial hypoxia. 1,9 This vascular response is less effective when a large part of the lung is collapsed.
If the blood cannot be diverted, it flows through the atelectatic non-ventilated lung and produces intrapulmonary shunting.\textsuperscript{1,8,9} The loss of an aerated lung may increase the risk of pneumonia and ventilator-induced lung injury by overstretching of the aerated lung.\textsuperscript{1,9}

### Clinical manifestations

#### Medical history

The clinical manifestations of atelectasis are non-specific and a full medical history may help to identify its etiology. Medical histories to identify the etiology of atelectasis are summarized in table 1.

<table>
<thead>
<tr>
<th>Etiology of atelectasis</th>
<th>Medical history</th>
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<tbody>
<tr>
<td>Broncho-pulmonary dysplasia</td>
<td>Neo-natal history of hyaline membrane disease (HMD)</td>
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<td></td>
<td>History of mechanical ventilation</td>
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<tr>
<td>Cystic fibrosis</td>
<td>Failure to thrive</td>
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<td></td>
<td>Steatorrhea</td>
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<td>Recurrent aspiration</td>
<td>Frequent vomiting or coughing during and immediately after feeding</td>
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<tr>
<td>/Gastro-esophageal reflux disease</td>
<td>Recurrent migrating or rotating atelectasis</td>
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<tr>
<td>Neuromuscular disease</td>
<td>Persistent atelectasis</td>
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<tr>
<td>Bronchial tumor</td>
<td>Hemoptysis</td>
</tr>
<tr>
<td>Asthma</td>
<td>Recurrent wheezing</td>
</tr>
<tr>
<td></td>
<td>Persistent atelectasis (especially at right middle lobe)</td>
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*Data adapted from Peroni DG et al.\textsuperscript{1}

If a foreign body is lodged in a large airway, patient may have a history of choking and cyanosis. If lodged in a smaller airway, there may be an asymptomatic period followed by infection.\textsuperscript{1} Post-operative atelectasis mostly presents within 24 hours after an operation and can persist for several days.\textsuperscript{10} In intensive care units, displacement of an endo-tracheal tube into the right main bronchus is common and may cause atelectasis. Regular checking of the position of the endo-tracheal tube is needed.

In asthma, the pathology of bronchial inflammation, bronchial debris, and mucus plugs cause airway obstructions. These are frequently in the right middle lobe, leading to the term right middle lobe syndrome. The right main bronchus is longer than other bronchi and starts at the bronchus intermedius after a sharp angle that leads to the retention of intrabronchial secretion\textsuperscript{1} and causes atelectasis. A patient with right middle lobe syndrome due to asthma with acute symptomatology may recover spontaneously but may lack re-expansion of the atelectatic lung.\textsuperscript{11} Therefore atelectasis may persist unnoticed for a prolonged period of time and lead to irreversible bronchiectasis.\textsuperscript{11}

#### Signs and symptoms

The signs and symptoms of atelectasis are often non-specific depending on the severity and etiology of the disease. Usually, the signs and symptoms are pleuritic chest pains, coughs and dyspnea. The presence of atelectasis does not change the clinical picture unless the lesion is large.\textsuperscript{1,3,12} Suggestive signs of atelectasis are...
localized, reduced breath sounds, a constant wheeze, reduced chest wall expansion or asymmetrical chest movements, dullness on percussion, mediastinal or cardiac displacement, and elevation of the diaphragm. In resorptive atelectasis, removal of an obstruction results in the rapid disappearance of symptoms and provides a clue for diagnosis.

Investigations

Once atelectasis is suspected, investigations to document its presence, extent, distribution, and severity should be performed.

Chest radiographs involving anterior-posterior and lateral films are important in diagnosis. They may show lines, opacification, crowded pulmonary vessels, crowded air bronchograms, deviation of inter-lobar fissures, deviations of mediastinum or hemidiaphragm, reduction of intercostals spaces, shifts of trachea to atelectatic lungs, and hyper-lucency of adjacent lung tissue. Atelectasis and consolidations may be found in patients with pneumonia. With consolidations the alveoli are full of exudates and there is no significant loss of lung volume, but atelectasis develops because of mucus plugs or surfactant dysfunction and result in loss of lung volume.

Atelectasis may occur in any lobe or segment of the lung, but the right and left lower lobes are the most frequently involved. The presence of atelectasis also can be noted in a specific lobe because of the relationship to the heart or to the diaphragm. Lower lobe atelectasis, resulting in dense opacification of that lobe, can obliterate the normal contour of the adjacent diaphragm. Similarly, in the right middle lobe or in the lingula, atelectasis can obliterate the normally visible right or left heart borders (silhouette sign), respectively. In right middle lobe atelectasis, results may be normal or show only a dense band without lung volume loss, and it may be difficult to differentiate diagnosis with that of pleural thickening. Abnormalities on chest radiographs are most apparent in the lateral view. The volume loss resulting from the collapse of the right middle lobe is seen as a triangle of increased density between the minor fissure and the lower half of the major fissure. In recurrent aspiration syndrome in infants, the most frequently affected lobes are the posterior areas of the upper and lower lobes. In older children who spend more time standing vertically, the lower lobes of the lingula and the right middle lobe are more frequently affected. In patients with neuromuscular disease, atelectasis is located mostly in the dorsal broncho-pulmonary segments due to gravity-related lung compression by the heart and intra-abdominal organs caused by the persistent supine position.

Computed tomography (CT) can be used in the diagnosis of atelectasis and has greater sensitivity than plain radiographs. CT is used with peripheral or round atelectasis that may be mistaken for tumors, and in acute respiratory distress syndrome (ARDS) to determine the distribution of atelectasis. It is also informative in the evaluation of bronchial patency and lymph node enlargement. F-18 FDG-PET/CT imaging is a new technique that uses different metabolic characterization and differentiates atelectatic lungs not involved with tumors from tumor-infiltrated atelectatic lungs, from inflammation, and infection. There is a positive relationship between the density of collapsed lungs and the intensity of FDG uptake. FDG uptake in atelectasis is higher than in normal lungs and generally lower than in tumor tissue.

In patients with total or near total opacification of the hemithorax, chest ultra-sound shows high sensitivity to distinguish between pleural effusion, consolidation, and atelectasis without ionizing radiation, and can be done at the bedside quickly and economically. Several studies suggest this approach can ease the use of CT scans, especially in critically ill patients.

Fiberoptic bronchoscopy (FOB) with broncho-alveolar lavage (BAL) is a useful and safe tool in the investigation and treatment of infants and children with intra-bronchial obstructions, especially in persistent...
atelectasis and middle lobe syndrome.\textsuperscript{1,10,11,14,17} Measurement of arterial oxygen tension by non-invasive methods, such as a pulse oximeter, or by invasive method, such as arterial blood gas, is helpful to evaluate the severity of atelectasis.

Treatments

Treatment of atelectasis depends on mechanism, causes, duration, and severity of the disease. There is a lack of evidence-based studies to guide the management but many modalities have clinical benefits.

Chest physiotherapy is the traditional first-line therapy for atelectasis.\textsuperscript{2,3} For children, deep breathing, coughing exercises, incentive spirometry, percussion, and vibration are effective ways to treat atelectasis.\textsuperscript{3} A study of 57 ventilated children revealed that chest physiotherapy with saline lavage and simulated coughing were successful in the improvement of 84\% of patients’ lung expansion.\textsuperscript{18}

Nebulized bronchodilators are traditionally recommended for the management of atelectasis. This modality can expand the diameter of the airways, increase sputum volume, and improve secretion clearance.\textsuperscript{1,2}

When atelectasis persists or there is no response to conservative treatment due to intra-luminal obstruction, fiberoptic bronchoscopy should be used to investigate and remove the cause of the obstruction.\textsuperscript{1,3} A study involving 26 children in a pediatric intensive care unit revealed that fiberoptic bronchoscopy successfully resolved 74\% of cases of atelectasis.\textsuperscript{19} Another study showed that the rate of atelectasis re-expansion after one instance of alveolus lavage was higher in the cases whose courses of disease were under 3 weeks than in those cases whose courses of disease were over 3 weeks.\textsuperscript{17}

Nebulized or direct tracheal application of recombinant human DNase I (rhDNase) reduces the visco-elastic properties of purulent secretions by breaking down the highly polymerised deoxyribonucleic acid. This treatment is approved only for the treatment of patients with cystic fibrosis and was described in the management of a mechanically ventilated child with asthma.\textsuperscript{1,2,10} A randomized trial of 75 infants with respiratory syncytial virus bronchiolitis showed an improvement in chest radiograph scores in the patients given nebulized DNase.\textsuperscript{20} A retrospective descriptive study of 25 non-cystic fibrosis pediatric patients who received rhDNase for atelectasis found that individual improvement was observed in 17 patients (68\%).\textsuperscript{21}

Surfactants may be used in adhesive atelectasis but this use has not been developed extensively as the required volumes are large and expensive.\textsuperscript{2}

Recruitment maneuvers are designed to expand atelectatic lungs and keep them open by positive end-expiratory pressure (PEEP) or continuous positive airway pressure (CPAP).\textsuperscript{3,4,10} The probability of success of these maneuvers is difficult to predict, but in post-operative atelectasis prediction is easy and this post-operative state has good results.\textsuperscript{3} In lung injury, prediction of response may be possible on the basis of the topographical distribution of the injury, as evidenced by CT.\textsuperscript{3} Patients with neuromuscular diseases tend to have ineffective coughs and difficulty expelling secretions. Several devices are available to assist these patients, including intermittent positive pressure breathing, a mechanical insufflator-exsufflator, and non-invasive bi-level positive pressure ventilation.\textsuperscript{10}

Oxygen therapy is indicated for patients with dyspnea or desaturation to maintain normal oxygenation but there must be an awareness of resorptive atelectasis from high concentration oxygen.

Treatment of atelectasis may need more than one modality or medication, as in the case of patients with cystic fibrosis requiring antibiotics with regular chest physiotherapy. Treatment of the causes of atelectasis is important to prevent irreversible conditions, such as bronchiectasis that may need surgical pulmonary resection.
Conclusion

Atelectasis is common in children and is benign and reversible if patients receive early detection and management. However, non-specific signs and symptoms may lead to a lack of diagnosis resulting in persistent atelectasis and irreversible bronchiectasis. Patients with underlying pulmonary disease or cardiopulmonary dysfunction may experience a decline in pulmonary function and have severe sequelae. Early diagnosis and management can prevent this condition. New diagnostic techniques, such as chest ultrasound and F-18 FDG-PET/CT imaging, are available but require experienced radiologists.

Many treatment modalities have clinical benefits but there is a need to support these approaches with evidence-based studies.

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References
