Imaging evaluation of hemoptysis in children

Divya Singh, Ashu Seith Bhalla, Prasad Thotton Veedu, Arundeep Arora

Hemoptysis is an uncommon but distressing symptom in children. It poses a diagnostic challenge as it is difficult to elicit a clear history and perform thorough physical examination in a child. The cause of hemoptysis in children can vary with the child's age. It can range from infection, milk protein allergy and congenital heart disease in early childhood, to vasculitis, bronchial tumor and bronchiectasis in older children. Acute lower respiratory tract infections are the most common cause of pediatric hemoptysis. The imaging modalities include chest radiography, multi-detector computed tomography (MDCT), magnetic resonance imaging (MRI) and catheter angiography. MDCT is a rapid multiplanar imaging modality which should be used judiciously to keep radiation dose to a minimum. MRI and catheter angiography have selected application.

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Key words: Hemoptysis; Lower respiratory tract infection; Bronchiectasis; Cystic fibrosis; Foreign body

Core tip: Hemoptysis is a cause of immense concern to the child, the family and the pediatrician. Thorough history and physical examination is necessary to ascertain its presence, which is particularly challenging in the pediatric population. Imaging has an important role in identifying the source of bleeding and its underlying cause. Acute lower respiratory tract infections are the most common cause of pediatric hemoptysis. The imaging modalities include chest radiography, multi-detector computed tomography (MDCT), magnetic resonance imaging (MRI) and catheter angiography. MDCT is a rapid multiplanar imaging modality which should be used judiciously to keep radiation dose to a minimum. MRI and catheter angiography have selected application.

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INTRODUCTION

Hemoptysis is defined as expectoration of blood or blood tinged-sputum due to bleeding from the respiratory tract[1]. Massive hemoptysis is termed as blood loss > 8...
There are several causes of hemoptysis in children (Table 1). The common causes are acute lower respiratory tract infections, bronchiectasis (due to cystic fibrosis, aspiration, ciliary dyskinesia, post infectious), congenital heart disease and foreign body aspiration. Of these, acute lower respiratory tract infections may constitute up to 40% of cases\(^5\). The etiology also varies with the child’s age. Sim \textit{et al}\(^4\) observed that infection, Heiner syndrome (milk protein allergy) and congenital heart disease were the major causes in early childhood; while during late childhood, vasculitis, bronchial tumor and bronchiectasis were far more prevalent.

The lungs receive dual blood supply; one from the high pressure bronchial arteries, the other from the pulmonary arteries with relatively lower pressure. The pulmonary arteries account for 99% of the arterial blood supply to the lungs and take part in gas exchange while the bronchial arteries provide nourishment to the supporting structures of the airways and form the vasa vasora of the pulmonary arteries. The bronchial vasculature is in close proximity to the pulmonary arteries at the level of the vasa vasora where the two systems are connected by thin-walled anastomoses between the systemic and pulmonary capillaries\(^5\). Pulmonary vascular obstructive disorders (congenital heart disease, vasculitis, embolism) open up these anastomoses in regions of the lung that are deprived of their pulmonary arterial blood flow. This subjects these fragile vessels to increased systemic arterial pressure and can cause hemoptysis by rupturing into the alveoli or bronchial airways.

In the setting of tracheobronchial infection, there is inflammation of the airways. As a result, they become congested and friable, which increases their susceptibility to bleed. Chronic inflammation (as in bronchiectasis) can lead to increase in systemic arterial flow due to release of angiogenic growth factors which lead to neovascularization and formation of “leaky” vessels prone to rupture. Approximately 5% of patients with cystic fibrosis can present with massive hemoptysis\(^7\). This is due to hypertrophy of bronchial arteries along with the presence of multiple bronchopulmonary anastomoses. Foreign body aspiration causes hemoptysis due to mechanical trauma or due to associated intense inflammation incited by organic matter. Pulmonary hemosiderosis is an uncommon but significant cause of hemoptysis in children. It is mostly idiopathic; however, it may be associated with an allergy to cow’s milk (Heiner syndrome)\(^8\). Although rare in children, endobronchial or pulmonary parenchymal tumors (carcinoid, bronchial adenoma) may cause significant hemorrhage.

**Imaging evaluation of hemoptysis**

The aim of initial diagnostic evaluation is to identify the immediate source of bleeding along with determination of the primary cause of hemoptysis. Traditionally, the diagnostic algorithms in acute setting have been based on various combinations of conventional radiography, chest computed tomography (CT) and thoracic aortography. MDCT has now made comprehensive visualization of

| Table 1 Causes of hemoptysis in children |
|------------------------------------------|
| Acute lower respiratory tract infections |
| Bacterial                                 |
| Viral                                     |
| Fungal                                    |
| Parasitic                                 |
| Bronchiectasis                            |
| Aspiration                                |
| Cystic fibrosis                           |
| Ciliary dyskinesia                        |
| Post-infectious                           |
| Congenital heart diseases                 |
| Eisenmenger syndrome                      |
| Aplasia/hypoplasia of pulmonary artery or veins |
| Primary pulmonary hypertension            |
| Pulmonary artery narrowing                |
| Infectious                                |
| Inflammatory                              |
| Pulmonary thromboembolism                 |
| Pulmonary arteriovenous malformation      |
| Alveolar hemorrhage syndrome              |
| Idiopathic                                |
| Associated with rheumatologic disease     |
| Pulmonary-renal syndrome                  |
| Neoplasms                                 |
| Bronchial carcinoid                       |
| Bronchial adenoma                         |
| Metastatic                                |
| Foreign body                              |
| Trauma                                    |
| Cryptogenic                               |

mL/kg in 24 h\(^3\). The life-threatening element in massive hemoptysis is asphyxiation due to flooding of the airways by blood. Hence, securing the airway needs immediate attention. Hemoptysis in lesser amounts poses a diagnostic challenge in pediatric patients as it may initially remain unnoticed because children tend to swallow their spu- tum and are unable to provide a clear history or undergo thorough physical examination. It is a cause of immense concern to the child, the parents and the pediatrician. After confirming the presence of hemoptysis, the next step is to establish the cause, so that an appropriate treatment regimen can be adopted. The spectrum of causes of hemoptysis in children is considerably different from that of the adults. Imaging has a pivotal role in evaluation of hemoptysis. There are various modalities which can be resort to; namely, conventional radiography, multi-detector computed tomography (MDCT), magnetic resonance imaging (MRI) and in certain cases, catheter angiography which can also fulfill a therapeutic purpose. The advent of MDCT has paved the way for non-invasive multi-dimensional visualization of the thoracic vasculature, tracheobronchial tree and pulmonary parenchyma. This is of tremendous value as it obviates the need for invasive bronchoscopic procedure with its attendant complications. The following sections illustrate the etiology, pathogenesis and role of imaging in hemoptysis.

**ETIOPATHOGENESIS**

The lungs receive dual blood supply; one from the high pressure bronchial arteries, the other from the pulmonary arteries with relatively lower pressure. The pulmonary arteries account for 99% of the arterial blood supply to the lungs and take part in gas exchange while the bronchial arteries provide nourishment to the supporting structures of the airways and form the vasa vasora of the pulmonary arteries. The bronchial vasculature is in close proximity to the pulmonary arteries at the level of the vasa vasora where the two systems are connected by thin-walled anastomoses between the systemic and pulmonary capillaries\(^5\). Pulmonary vascular obstructive disorders (congenital heart disease, vasculitis, embolism) open up these anastomoses in regions of the lung that are deprived of their pulmonary arterial blood flow. This subjects these fragile vessels to increased systemic arterial pressure and can cause hemoptysis by rupturing into the alveoli or bronchial airways.

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thoracic anatomy possible. It provides high-resolution images of the thoracic and upper abdominal vasculature which aids in diagnosis and also provides a roadmap prior to any intervention. CT findings can forewarn the endoscopist about the presence of peribronchial or endoluminal aneurysms. MRI does not have a role in the acute setting. However, it may serve as a problem-solving tool in certain situations.

**Conventional radiography**

Chest radiography serves as a valuable screening modality. It can help in lateralizing the bleeding with high degree of accuracy and can detect several parenchymal and pleural abnormalities. The commonly used views include frontal and, in some cases, lateral. The utility of lateral radiographs is in case of presence of a radio-opaque foreign body on frontal view, when it can determine if it is in the trachea or esophagus. Lynch et al. observed that addition of a lateral radiograph in children with pneumonia did not improve diagnostic accuracy. Common findings include presence of focal infiltrates which may suggest infection. Unilateral air-trapping with hyperinflation can give a clue towards presence of an unsuspected tracheobronchial foreign body. A radio-opaque foreign body may be seen. Ancillary findings include “tram-track” appearance of bronchiectasis; pulmonary nodules, lymphadenopathy, pleural effusion; cardiomegaly; and vascular redistribution due to pulmonary venous obstruction. Approximately one-third of chest radiographs may be normal in children with hemoptysis. A tracheobronchial source of bleed may eventually be identified in about half of these cases. Therefore, additional follow-up testing is recommended in patients with hemoptysis in whom the underlying cause is not detected by initial radiography.

**MDCT**

The role of MDCT in evaluation of hemoptysis includes: (1) depiction of underlying disease; (2) assessment of consequences of hemorrhage which may be a cause of clinical concern or may conceal the underlying abnormalities; and (3) panoramic visualization of the thoracic vasculature by various reconstruction techniques.

**Technique**

CT technique involves acquisition of multiple sections from the base of the neck to the level of the renal arteries (L2 level). Optimal enhancement of the pulmonary and systemic arteries is achieved by administration of 2 mL/kg body weight of iodinated non-ionic contrast media containing 300 mg I/mL at a rate of 4 mL/s via a wide gauge cannula. The scan should be started during the phase of peak systemic arterial enhancement (scanning delay of 18 s or a threshold of 100 HU in the descending aorta with bolus tracking). Images should be acquired with thin collimation and with the table movement adjusted to allow wide volume coverage during a single breath-hold. Radiation exposure is a significant concern in the pediatric population. Hence, the exposure parameters and kilovoltage need to be adjusted according to the patient’s weight so as to minimize radiation dose with optimal image quality.

**Data processing and interpretation**

Since a large volume of data is acquired, the images are best interpreted at the scanner console or remote workstation. The soft-tissue structures and lung parenchyma can be assessed adequately in axial sections of 5 mm thickness with mediastinal and lung window settings, respectively (Figure 1A and B). High resolution CT images allow detailed evaluation of the pulmonary parenchyma. The tracheobronchial airway can be evaluated on thinner sections and reformatted images.

Two-dimensional maximum intensity projection images (Figure 1C) in the coronal/oblique and sagittal planes can demonstrate the tortuous course of the bronchial arteries from the descending thoracic aorta to the lungs. Intercostal and internal mammary arteries are best visualized in the coronal planes while the inferior phrenic arteries and celiac axis branches are demonstrated well in axial images. Three-dimensional volumetric and shaded-surface display images not only depict the abnormal vessel, but also illustrate its relationship with the surrounding structures, thus providing a preview of the internal anatomy.

Minimum Intensity Projection can be used to generate images of the central airways (Figure 1D) and demonstrate areas of air-trapping within the lungs. These can provide valuable perspective in defining a lesion prior to any intervention. Therefore, a host of reconstructed images need to be analyzed for a thorough CT assessment of hemoptysis.

**STEPWISE STRUCTURAL ASSESSMENT**

**Lung parenchyma**

The pulmonary parenchyma should be evaluated for presence of bronchiectasis, consolidation and ground-glass opacity. The site of hemorrhage can be localized on the basis of presence of fluid density material in the segmental and lobar bronchi and ground-glass opacity with hazy consolidation which represents alveolar hemorrhage. Acute hemorrhage can mask the underlying pathology. Blood clots can also simulate more ominous entities like masses.

**Tracheobronchial tree**

This should be evaluated for presence of any stenosis which may be due to intraluminal (foreign body, neoplasm) or extraluminal (lymphadenopathy, fibrosing mediastinitis) causes. Multiplanar reformatted (MPR) images are accurate in detection of lesions, depiction of degree of narrowing, distal visualization and calculation of distance of the lesion from the carina in selected locations.

**Pulmonary arteries**

The pulmonary arteries should be analyzed for any narrowing due to extrinsic or intrinsic causes. The presence
of accompanying subpleural areas of enhancement can represent lung infarcts. The pulmonary arteries can also show dilatation (Rasmussen aneurysm) and pulmonary arteriovenous malformations.

**Bronchial arteries**

Hemoptysis originates from bronchial arteries in 95% of cases. A bronchial artery diameter of more than 2 mm is considered abnormal. In 70% of individuals, bronchial arteries arise from the descending thoracic aorta between T5 and T6 levels. There are usually one or two bronchial arteries supplying each lung, arising independently or from a common trunk. They are visualized as a cluster of enhancing nodules in the posterior mediastinum just below the level of the aortic arch on axial images. Active bleeding can rarely be detected on CT. Anomalous bronchial arteries are defined as arteries which originate outside the T5-T6 level. Their most common site of origin is the concavity of the aortic arch.

**Non-bronchial systemic arteries**

Non-bronchial systemic arteries can arise from the branches of brachiocephalic arteries, subclavian arteries, axillary, internal mammary and infradiaphragmatic branches from the inferior phrenic artery and celiac axis. On CT, these are seen as dilated tortuous arteries that are not parallel to the bronchi. The presence of pleural thickening greater than 3 mm with enhancing arteries within the extrapleural fat is a pointer of presence of these vessels.

**MRI**

MRI does not have any utility in imaging evaluation of acute hemoptysis. Since it has superior soft-tissue resolution, it is excellent in the evaluation of the mediastinum and hilum in the non-emergent setting. It provides less information about the lung parenchyma. It may be used to demonstrate arteriovenous malformations and congenital anomalies of the pulmonary arteries and delineate the nature of mediastinal soft-tissue in fibrosing mediastinitis. With the introduction of hyperpolarized nuclei like $^3$He and $^{129}$Xe, the horizon of MRI is likely to expand from limited utility in evaluating the pulmonary parenchyma to evaluation of pulmonary structure, function and metabolism with a high sensitivity. Ventilation and dynamic imaging in patients with asthma and cystic fibrosis have shown regional patterns of obstruction and ventilation defects in these individuals. Further knowledge can go a long way in the early diagnosis, monitoring disease progression and evaluation of response to treatment.

**COMMON CAUSES OF PEDIATRIC HEMOPTYSIS**

**Acute lower respiratory tract infections**

Tracheobronchitis, pneumonia and lung abscess can lead to hemoptysis. The infective process may be bacterial, viral, fungal or parasitic in origin. Although tuberculosis is a significant cause of adult hemoptysis, very few cases have been reported in the pediatric literature. Chest radio-
graphs can show pulmonary infiltrates, hyperinflation and cavity with or without air-fluid level (Figure 2A). There may be associated pleural effusion and lymphadenopathy. CT findings can be in the form of consolidation, ground-glass opacity, interstitial thickening, air-trapping, cavity with shaggy walls and air-fluid level, pleural effusion and mediastinal or hilar lymphadenopathy (Figure 2B). CT can also demonstrate complications like empyema (thick enhancing visceral and parietal pleura, “split pleura” sign), bronchopleural fistula, etc.

Parasitic cysts (Echinococcus) can cause hemoptysis by rupturing into the airway. These may be seen as fluid density lesions with a smooth wall and air foci due to communication with the adjacent bronchus (Figure 3A). Detached membranes and daughter cysts can be visualized within the cyst. Concomitant cysts may be seen in other organs, most commonly in the liver (Figure 3B).

The most commonly implicated fungus is Aspergillus. It can have a varied spectrum of presentation, namely aspergilloma, allergic bronchopulmonary aspergillosis (ABPA), semi-invasive aspergillosis, airway or angioinvasive aspergillosis[23,24]. Aspergilloma is the saprophytic colonization of a pre-existing cavity by the fungus and is typically seen as an opacity within a cavity producing the “air-crescent” sign. It is mobile and can show postural change in position. ABPA is a manifestation of type I and III hypersensitivity reaction to the organism and presents as central bronchiectasis with mucous plugged bronchi producing ‘finger-in-glove’ appearance with upper lobe preponderance on radiograph. The mucous plugs have a high-density on CT (Figure 4). There may be centriflobar nodules with “tree-in-bud” appearance. Some patients can also have associated allergic fungal sinonasal disease. Invasive aspergillosis is encountered in immunocompromised patients. Invasive airway disease presents as peribronchial areas of consolidation and multiple branching centriflobular nodules on CT[25]. Nodules with surrounding ground-glass opacity (halo sign) or pleural-based, wedge shaped areas of consolidation (Figure 5) are the hallmark of angioinvasive aspergillosis[26].

**Bronchiectasis**

Bronchiectasis can occur secondary to aspiration, infections, cystic fibrosis and ciliary dyskinesias. On chest radiographs, it manifests as “tram-track”, parallel line opacities, ring opacities and tubular structures (Figure 6A). However, chest radiographs are insensitive for detecting mild to moderate disease. CT (Figure 6B) has a higher sensitivity and on CT imaging, bronchiectasis is characterized by the absence of normal bronchial tapering, the presence of visible bronchi in the peripheral 1 cm of the lung and a bronchoarterial ratio more than 1 (signet ring sign). The etiology can be narrowed by considering the anatomic location and distribution of pathology.
Aspiration tends to involve the lower lobes (right > left). Cystic fibrosis shows lung hyperinflation and interstitial infiltrates with upper lobe preponderance (Figure 7). Bronchiectasis due to ciliary dyskinesias has a lower lobe predisposition[27].

**Congenital heart disease**

Hemoptysis can occur in patients with congenital heart diseases associated with pulmonary artery or venous stenosis or atresia. This is attributed to hemorrhage from enlarged, tortuous aorto-pulmonary collateral arteries and thrombotic lesions in the small pulmonary arteries[28]. Chest radiography may show cardiomegaly with abnormality in cardiac silhouette and a small hilum. An abnormal vascular channel parallel to the right cardiac border (scimitar vein) can be seen in pulmonary venolobar hypoplasia[29]. There may be associated pulmonary volume loss. MDCT is the modality of choice to demonstrate the site and extent of pulmonary artery narrowing and delineate anomalous pulmonary venous drainage (Figure 8). It exquisitely depicts the various aorto-pulmonary collaterals. Other associated cardiac anomalies can also be evaluated[30].
Pulmonary artery narrowing

Chronic pulmonary artery narrowing can occur due to a variety of causes like infections, inflammation and thromboembolism\(^\text{[31]}\). Infections are the most common cause. Narrowing of the pulmonary artery can be caused in the setting of infection by mediastinal lymphadenopathy or fibrosis. Fibrosis may be focal or diffuse. CT finding of focal fibrosis is a calcified soft-tissue mass in the paratracheal and hilar location. It can occur secondary to tuberculosis in the developing countries and histoplasmosis in United States. The diffuse form manifests as an infiltrative, non-calcified soft-tissue mass extending into multiple mediastinal compartments. It can be associated with autoimmune disorders, drugs, or be idiopathic\(^\text{[32]}\). Pulmonary artery narrowing in these cases leads to pulmonary hypoperfusion and consequent bronchial artery hypertrophy while can lead to hemoptysis of varying severity. CT pulmonary angiography is the investigation of choice in this condition as it elucidates pulmonary artery narrowing and bronchial/systemic artery hypertrophy\(^\text{[33]}\).

Pulmonary arteriovenous malformations

Pulmonary arteriovenous malformations (PAVM) are direct communication between the branches of the pulmonary artery and veins without capillary bed. There is a strong association between PAVM and hereditary hemorrhagic telangiectasia\(^\text{[34]}\). Chest radiography is an important tool for diagnosis and follow-up. Classic findings of PAVM are a round or oval well-defined mass, frequently lobulated, ranging in size from 1-5 cm. Two-thirds of these are located in the lower lobe. A connecting vessel may be seen radiating from the hilum. MDCT can identify the PAVM and connecting vessels more accurately (Figure 9). PAVMs have rapid blood flow and hence produce a low intensity signal on MRI. Catheter angiography remains the gold standard in diagnosis of PAVM as it defines the angio-architecture which is necessary before therapeutic embolization or surgical resection.
Idiopathic pulmonary hemosiderosis (IPH) is a rare pulmonary disorder which manifests as a triad of hemoptysis, anemia and diffuse parenchymal infiltrates on chest radiographs[35]. Diagnosis is confirmed by detection of hemosiderin-laden macrophages in broncho-alveolar lavage fluid, sputum or gastric aspirate. Secondary hemosiderosis is associated with systemic vasculitis, bleeding disorders and cardiac disease. Imaging findings are non-specific and need to be correlated with clinical and laboratory data to arrive at a diagnosis of IPH. Chest radiographs may reveal symmetric diffuse or patchy alveolar shadows sparing lung apices, which can clear on follow-up imaging. CT can show diffuse or patchy ground-glass opacity (Figure 10). There can be interstitial thickening in some cases[36].

Foreign body aspiration can be a cause of hemoptysis primarily in patients less than 3 years of age. The aspirated foreign body can be visualized on a radiograph if it is radio-opaque. Associated radiographic findings include non-specific infiltrates, atelectasis, areas of hyperinflation, parenchymal consolidation or bronchiectasis (Figure 11A). Chest radiographs can be normal in 30% cases.

MPR and endoluminal virtual bronchoscopic images derived from MDCT can delineate the shape, location and volume of a foreign body. It can reveal associated pulmonary parenchymal changes (Figure 11B and C). Thus, imaging can help the surgeon plan the bronchoscopy for safe removal of foreign body[37].

Neoplasm
Bronchial neoplasms are a rare cause of hemoptysis (Figure 12). Bronchial carcinoid tumors are the most frequent primary pulmonary neoplasms of childhood. The lesion can be central or peripheral. Radiological findings include hilar or perihilar masses with lobulated margins and associated obstructive changes (atelectasis, consolidation, bronchocele or hyperinflation)[38]. On CT, carcinoid is seen as a well-defined, centrally located mass that narrows or deforms the airway and contains diffuse or punctuate calcification. It shows intense homogenous contrast enhancement. However, all carcinoids do not enhance. There can be associated pulmonary obstructive changes and mediastinal/hilar lymphadenopathy[39].

CONCLUSION
Hemoptysis is a distressing symptom for the child, the...
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Figure 11  Foreign body aspiration. Chest radiograph (A) shows a radio-opaque foreign body in the left main bronchus (arrow) with hyperinflation of the left lung. Axial computed tomography images (B, C) delineate the morphology of the foreign body in the left main bronchus causing luminal compromise. There is associated air trapping in the left lung with patchy consolidation in the apical segment of the lower lobe.

Figure 12  Bronchial carcinoid. Scout computed tomography (CT) image (A) revealing non-visualization of the right main bronchus with volume loss and opacification of the right hemithorax along with bronchiectasis in right lower zone. Axial CT image (B) shows a mass in the right lung with mediastinal infiltration. Coronal Minimum Intensity Projection image (C) shows the outline of the mass projecting in the right main bronchus along with bronchiectasis in the right lower lobe.

Figure 13  Flowchart depicting approach to a child presenting with hemoptysis. MR: Magnetic resonance; CT: Computed tomography.
family and the pediatrician. It poses a diagnostic challenge. Once the presence of hemoptysis has been ascertained, one needs to identify the source of bleeding and primary underlying cause. Acute lower respiratory tract infections are the most common cause of pediatric hemoptysis. The imaging modalities available for the work-up of hemoptysis include chest radiography, MDCT, MRI and catheter angiography. Chest radiographs may be normal in 30% cases. MDCT is a rapid, non-invasive multiplanar imaging modality which should be tailored to keep radiation dose to a minimum for optimal evaluation of hemoptysis in pediatric patients. MRI and catheter angiography have selected application. The use of the various imaging tools available is determined by the clinical presentation and the possible etiology (Figure 1). Maximum diagnostic and therapeutic benefit can be attained by the judicious use of imaging modalities in a child presenting with hemoptysis.

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