

Hemoptysis in Children

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Context: Pulmonary hemorrhage and hemoptysis are uncommon in childhood, and the frequency with which they are encountered by the pediatrician depends largely on the special interests of the center to which the child is referred. Diagnosis and management of hemoptysis in this age group requires knowledge and skill in the causes and management of this infrequently occurring potentially life-threatening condition.

Evidence acquisition: We reviewed the causes and treatment options for hemoptysis in the pediatric patient using Medline and Pubmed.

Results: A focused physical examination can lead to the diagnosis of hemoptysis in most of the cases. In children, lower respiratory tract infection and foreign body aspiration are common causes. Chest radiographs often aid in diagnosis and assist in using two complementary diagnostic procedures, fiberoptic bronchoscopy and high-resolution computed tomography. The goals of management are threefold: bleeding cessation, aspiration prevention, and treatment of the underlying cause. Mild hemoptysis often is caused by an infection that can be managed on an outpatient basis with close monitoring. Massive hemoptysis may require additional therapeutic options such as therapeutic bronchoscopy, angiography with embolization, and surgical intervention such as resection or revascularization.

Conclusions: Hemoptysis in the pediatric patient requires prompt and thorough evaluation and treatment. An efficient systematic evaluation is imperative in identifying the underlying etiology and aggressive management is important because of the potential severity of the problem. This clinical review highlights the various etiological factors, the diagnostic and treatment strategies of hemoptysis in children.

Key words: Children, Hemoptysis, Management, Pulmonary hemorrhage, Review.

Pulmonary hemorrhage was not commonly described in children in the early texts, although it was noted to occur. Hemoptysis is defined as the expectoration of blood or blood-tinged sputum from the lower respiratory tract. Although common in adults, blood tinged sputum is a rare presenting symptom in children. The diagnosis of pediatric hemoptysis can be challenging. Children tend to swallow their sputum; therefore, hemoptysis may go unnoticed unless the bleeding is substantial(1). Coupled with this, the inability to provide a complete history and to cooperate with a thorough physical examination may further compound the diagnostic dilemma. Thus, hemoptysis can serve as a source of significant anxiety for the patient, the family, and the pediatrician.

Most important in children is to first establish that the child is experiencing hemoptysis. Extra-pulmonary bleeding, such as those arising from the nose, or the gastrointestinal tract, may be incorrectly attributed to hemoptysis. This is known as pseudo-hemoptysis. As the diagnostic and treatment strategies differ markedly, the two sources must be differentiated. The blood in hemoptysis is bright red in color and may be admixed with sputum and frothy. The blood in hematemesis is dark red or brown and may be mixed with food particles(2). One also has to differentiate factitious hemoptysis from real hemoptysis. Once the distinction is made, the physician can proceed to uncover the underlying cause. Hemoptysis is classified as non-massive or massive based on the

volume of blood loss; however, there are no uniform definitions for these categories. Hemoptysis is considered massive if blood loss is more than 200 mL per day(3).

Pulmonary hemorrhage may even be present in a previously healthy infant in whom neonatal medical problems have been ruled out. This pulmonary hemorrhage can appear as hemoptysis or blood in the nose or airway with no evidence of upper respiratory or gastrointestinal bleeding. Patients have acute respiratory distress or failure, requiring mechanical ventilation and chest radiography, and usually demonstrate bilateral infiltrates. Centers for Disease Control and Prevention (CDC) has given criteria for a confirmed case of pulmonary hemorrhage in a previously healthy infant aged ≤ 1 years of age with gestational age of ≥ 32 weeks, with no history of neonatal medical problems, and whose conditions meets all the following three criteria(4):

- (i) Abrupt or sudden onset of overt bleeding or obvious evidence of blood in the airway, including epistaxis, hemoptysis or frank blood below the larynx at visualization.
- (ii) Severe-appearing illness leading to acute respiratory distress or respiratory failure resulting in admission in PICU or NICU with intubation and mechanical ventilation.
- (iii) Diffuse unilateral or bilateral pulmonary infiltrates visible on chest X-ray or CT scan of thorax, and these findings should be documented within 48 hours of examination of infant.

Criteria for probable case of pulmonary hemorrhage in the infant include a previously healthy infant aged ≤ 1 year with a gestational age of ≥ 32 weeks, with(4):

- (i) Who has sudden onset of bleeding from the airway, with or without respiratory distress, with or without intubation and with or without pulmonary infiltrates on chest X-ray or CT scan;
 - (a) Who died and had evidence of bleeding from the airway found on autopsy or postmortem.

ETIOLOGY

The etiology of hemoptysis in children is as varied as in adults. The various causes of hemoptysis in children are summarized in **Table I**. Important causes of massive hemoptysis in children are: bronchiectasis, pulmonary tuberculosis, CHD, bronchial AV malformation, foreign body aspiration, cystic fibrosis, bronchial adenomas, DIC and tracheostomy-related.

Acute lower respiratory tract infection, either in the form of pneumonia or tracheobronchitis, accounts for almost up to 40% of the cases(5). In a 10 year retrospective study of hemoptysis in children, Coss-Bu, *et al.*(6) observed pneumonia to be the most frequent cause accounting for 31% of the episodes of hemoptysis in children. The presence of an infectious process (e.g. necrotizing pneumonia, tuberculosis, lung abscess, infected bronchiectasis) leads to destruction of lung parenchyma and erosion of blood vessels, resulting in hemoptysis. Infections have been reported as the most common etiology of hemoptysis in several studies in children(7,8). Although tuberculosis used to be commonly implicated, few cases have been reported in pediatric literature(5,9). Only one study by Crocco, *et al.*(10) reported high prevalence of hemoptysis in

TABLE I CAUSES OF HEMOPTYSIS IN CHILDREN

• Infections - pneumonia, tracheobronchitis
• Cystic fibrosis
• Congenital heart diseases - ASD, VSD, complex cyanotic heart diseases, tetralogy of fallot, truncus arteriosus, transposition of the great arteries
• Tuberculosis
• Foreign body aspiration
• Nasopharyngeal bleeding
• Tracheostomy-related
• Bronchiectasis
• Pulmonary neoplasms
• Pulmonary hemosiderosis
• Factitious hemoptysis
• Miscellaneous – invasive aspergillosis, pulmonary arteritis, catamenial hemoptysis, hydatid cysts in lungs
• Unknown causes

tuberculosis (80%). In countries like India where the prevalence of tuberculosis is high, this infection should never be neglected and should rank much higher. Another important cause of hemoptysis in children is bronchiectasis, which can be unilateral or bilateral. The bronchiectasis occurs due to the repeated respiratory tract infections since childhood. Chronic bacterial endobronchial infection and inflammation of the mucosa damage and destroy mucociliary defences, and this leads to secretion stasis, which in turn propagates further bacterial infection, and increases airway inflammation and bronchial dilatation(11). The infection is usually bacterial in nature and consists of *Streptococcus pneumoniae*, *Staphylococcus aureus*, *M. catarrhalis*, *klebsiella* species, or *Pseudomonas aeruginosa*. Aspergillus infection of the lungs either in the form of allergic bronchopulmonary aspergillosis (ABPA) or invasive aspergillosis can occur in children and some cases of hemoptysis have been reported(12).

Congenital heart disease can be a source of profuse bleeding in a child. With the advent of corrective cardiac surgery, the incidence of hemoptysis in this setting has declined significantly(5). Hemoptysis in CHD occurs most frequently with pulmonary vascular obstructive disease, but it can also occur in conjunction with enlarged collateral bronchial circulation. Hemoptysis in this condition is caused by erosion of a tortuous dilated bronchial artery into a bronchus, from rupture of an atherosclerotic bronchial artery plaque, or from localized pulmonary infarction at the bronchopulmonary anastomosis(13). Recently, life threatening hemoptysis has been reported in a child due to aortic pseudoaneurysm, which was proved by the aortic biopsy showing neutrophilic infiltration of the mucosa(14).

Foreign body aspiration is always considered in the differential diagnosis of pediatric hemoptysis. The bleeding in this case results from the mechanical trauma to the respiratory epithelium or the ensuing inflammatory reaction, especially to vegetable matter(15). Tom, *et al.*(16) identified foreign bodies as the second most common cause of hemoptysis. We have observed four children with hemoptysis, who had vegetable matter impacted in the bronchi, which were successfully removed with bronchoscope.

In the Western countries, hemoptysis is relatively common in patients with cystic fibrosis (CF), especially with increased survival into adulthood, and advances in medical treatment. Approximately 5% of patients with CF may present with massive hemoptysis due to bronchiectasis. In one study(2), CF contributed to 65% of cases of pediatric hemoptysis in a 10-year retrospective study. Also the Patients with CF also had longer hospitalization compared to other causes of hemoptysis, reflecting the chronic nature and the multitude of problems associated with CF. There is hyperplasia, tortuosity and dilatation of bronchial arteries due to chronic inflammation, and hemorrhage results from erosion of these dilated, thin walled bronchial vessels after successive pulmonary infections(5).

Neoplasms of the respiratory tract are rare in children, but endobronchial or pulmonary parenchymal tumors may cause significant bleeding. Tumors that may cause hemoptysis include bronchial carcinoid, bronchial adenoma, endobronchial metastasis, mediastinal teratomas, tracheal tumours, or bronchial arteriovenous malformations in children(17). Hemoptysis is a well-recognized complication of long-term tracheostomies. Wetmore, *et al.*(18) reported that significant bleeding may occur in approximate 10% of the patients with long-term tracheostomy. Fabian and Smitheringale(19) found tracheostomy related hemoptysis to be second most common cause of hemoptysis (15.5%). Typically, the bleeding is described as pink or red-tinged secretions on suctioning the tracheobronchial tree.

Idiopathic pulmonary haemosiderosis is a rare cause of diffuse alveolar hemorrhage of unknown etiology. It occurs most frequently in children, has a variable natural history with repetitive episodes of diffuse alveolar hemorrhage, and has been reported to have a high mortality. The recurrent episodes of diffuse pulmonary hemorrhage may present as hemoptysis(20). Many patients develop iron deficiency anemia secondary to deposition of haemosiderin iron in the alveoli. Recently, Kabra, *et al.*(21) described hemoptysis in about 58% of children in patients with pulmonary hemosiderosis. Most of these patients had small and recurrent hemoptysis. Examination of sputum and broncho-

alveolar lavage fluid can disclose hemosiderin-laden alveolar macrophages (siderophages), and the lung biopsy shows numerous siderophages in the alveoli, without any evidence of pulmonary vasculitis, nonspecific/granulomatous inflammation, or deposition of immunoglobulins. In some of these patients normo-complement urticarial vasculitis has been observed in children, and this may predispose for the hemoptysis(22).

Other causes of hemoptysis are far less common, such as bleeding from localized lesions in upper airways or bleeding into the lungs as like part of a systemic disease systemic lupus erythematosus, Goodpasture's syndrome, pulmonary thromboembolism, hydatid cyst, and even duplication cyst of the stomach can cause hemoptysis in children(23-26). Isolated pulmonary arteritis can lead to massive hemoptysis in children(27). Recently, catamenial hemoptysis has been described in a 12 year old child and fiberoptic bronchoscopy revealed presence of endobronchial endometriosis(28).

Factitious hemoptysis is considered in the differential diagnosis if no etiology is discernible after a thorough evaluation, especially when the medical history or patient's behavior is unusual(29). Covert biting of the buccal mucosa has been attributed to cause hemoptysis in these children(30). Psychological counseling is usually required in such children.

DIAGNOSIS

Hemoptysis in children should be evaluated systematically. The investigations begin with a detailed medical history and physical examination. First, the hemoptysis has to be differentiated from hematemesis. Historic clues are useful for differentiating hemoptysis from hematemesis (**Table II**). Patient history also can help identify the anatomic site of bleeding, differentiate between hemoptysis and pseudo-hemoptysis, and narrow the differential diagnosis. Once true hemoptysis is suspected, the investigations should focus on the respiratory system.

The physician should always inquire about the possibility of foreign body aspiration, including

choking or coughing episodes, and new onset wheezing. A history of chronic lung disease or CHD is also important. This is followed by thorough examination of the neck and head. Special attention should be given to the oral cavity and nasopharynx as the potential sources of bleeding. Lung examination may reveal localized wheezing, suggesting foreign body, or rales or decreased breath sounds, which may be associated with an infectious process(5).

Routine blood test with complete haemogram has to be done in all the children. This is imperative because children generally tend to swallow blood and the amount of bleeding is likely to be underestimated. As an infectious etiology is common, sputum is evaluated for bacterial, fungal and mycobacterial organisms. Sputum culture will be helpful for identifying the pathogens and sensitivities to various antibiotics.

Chest radiography serves as a valuable screening technique. Unilateral air trapping with hyperinflation may suggest the diagnosis of foreign body aspiration(1). Focal or interstitial infiltrates may help the diagnosis of infection. Other helpful findings include pulmonary nodules, hilar adenopathy, pleural effusion and cardiomegaly. In approximately one third of children with hemoptysis, chest

TABLE II DIFFERENTIATING FEATURES OF HEMOPTYSIS AND HEMATEMESIS

Hemoptysis	Hematemesis
<i>History</i>	
Absence of nausea and vomiting	Presence of nausea and vomiting
Lung disease	Gastric or hepatic disease
Asphyxia possible	Asphyxia unusual
After the episode, sputum is always blood tinged	After the episode, sputum is always clear
<i>Sputum examination</i>	
Frothy	Rarely frothy
Liquid or clotted appearance	Coffee ground appearance
Bright red to pink	Brown to black
<i>Laboratory Parameters</i>	
Alkaline pH	Acidic pH
Mixed with macrophages and neutrophils	Mixed with food particles

radiographs may be normal. High Resolution Computed Tomography (HRCT) can be useful in further delineation of chest radiography findings. Contrast studies are helpful to differentiate between vascular structures and solid masses(2). In recent years, HRCT scan of thorax has become the most accurate and sensitive noninvasive diagnostic tool for the evaluation of bronchiectasis(31).

If the etiology of hemoptysis is not discovered after aforementioned workup, and if the bleeding is recurrent, bronchoscopy which may be rigid or fiberoptic, is indicated to identify source of bleeding(9,10). Fiberoptic bronchoscopy can be performed with sedation and allows more detailed evaluation of distal bronchial tree. However, it does not permit effective ventilation and removal of blood clots. In contrast, rigid bronchoscope offers ventilation and helps localize site of bleeding. It is also ideal for suctioning of clotted blood and is also more effective for removal of airway foreign bodies(32). In one study, rigid bronchoscopy was performed 24 times in 18 patients for diagnostic and therapeutic reasons. The various findings included: blood, mucosal inflammation, purulence, tracheal abrasions, gradation tissue and bronchial mass. The diagnostic yield was 61%(5). The diagnostic yield of bronchoscopy in hemoptysis ranges from 40% to 100% in various studies(1,9).

Cardiac evaluation should be considered in patients with hemoptysis unexplained by pulmonary causes, even in the absence of overt cardiac symptoms(33). Echocardiography should be performed for the evaluation of any suspected congenital cardiac disease. Pulmonary thromboembolism is a rare cause of hemoptysis in children, and a combination of diagnostic procedures must be used to identify a suspected or confirmed case of pulmonary thromboembolism in children, including ventilation perfusion studies(24). When no other cause is found for pulmonary hemorrhage, the presumed diagnosis is idiopathic pulmonary hemosiderosis(21). In these patients, sputum and bronchoalveolar lavage fluid can disclose haemosiderin-laden alveolar macrophages (siderophages).

Figure 1 provides an algorithm for the evaluation of hemoptysis in children.

TREATMENT

Management of the child with hemoptysis depends on two important issues – the underlying causes and the severity of the bleeding. The three goals of therapy are: to prevent asphyxiation, stop the bleeding, and treat the primary cause. Most of the cases are self-limited and will resolve spontaneously.

Minor hemoptysis

Minor hemoptysis is managed symptomatically, by giving cough suppressants like dextromethorphan and oral/parental haemostatic agents like ethamsylate or Botropase, reassurance of the patient and parents, and the treatment of the underlying cause. If a specific etiology is identified, appropriate therapy of the underlying disease should be initiated. Pulmonary infections are treated with appropriate antibiotics. Cystic fibrosis exacerbations are managed with antibiotics and corticosteroids. Tracheostomy-related trauma is managed by modifying the suctioning technique, using soft red rubber catheters, and humidification. Pulmonary tuberculosis should be treated with anti-tubercular therapy. Idiopathic pulmonary hemosiderosis is treated with prednisolone. Kabra, *et al.*(21) recently treated children with pulmonary hemosiderosis with prednisolone and hydroxychloroquin followed by inhaled corticosteroids, and found favorable response in majority of the patients.

Massive hemoptysis

In small number of cases, the child may present with life-threatening hemorrhage (>8 mL/kg every 24 hours or 200 mL every 24 hrs). Massive hemoptysis can quickly progress to acute respiratory distress in a child. These children require multiple procedures to stabilize the airways and to control blood loss(34). Intravenous fluids and blood products are given to prevent cardiovascular collapse from the exsanguination of blood. Reversal of any coagulation disorder and protecting the non-bleeding lung from aspiration by placing the bleeding side down and selectively intubating the good lung, in cases of massive hemoptysis, should be undertaken(35). The use of cough suppressants containing codeine has been controversial as they have the potential to alter the level of consciousness

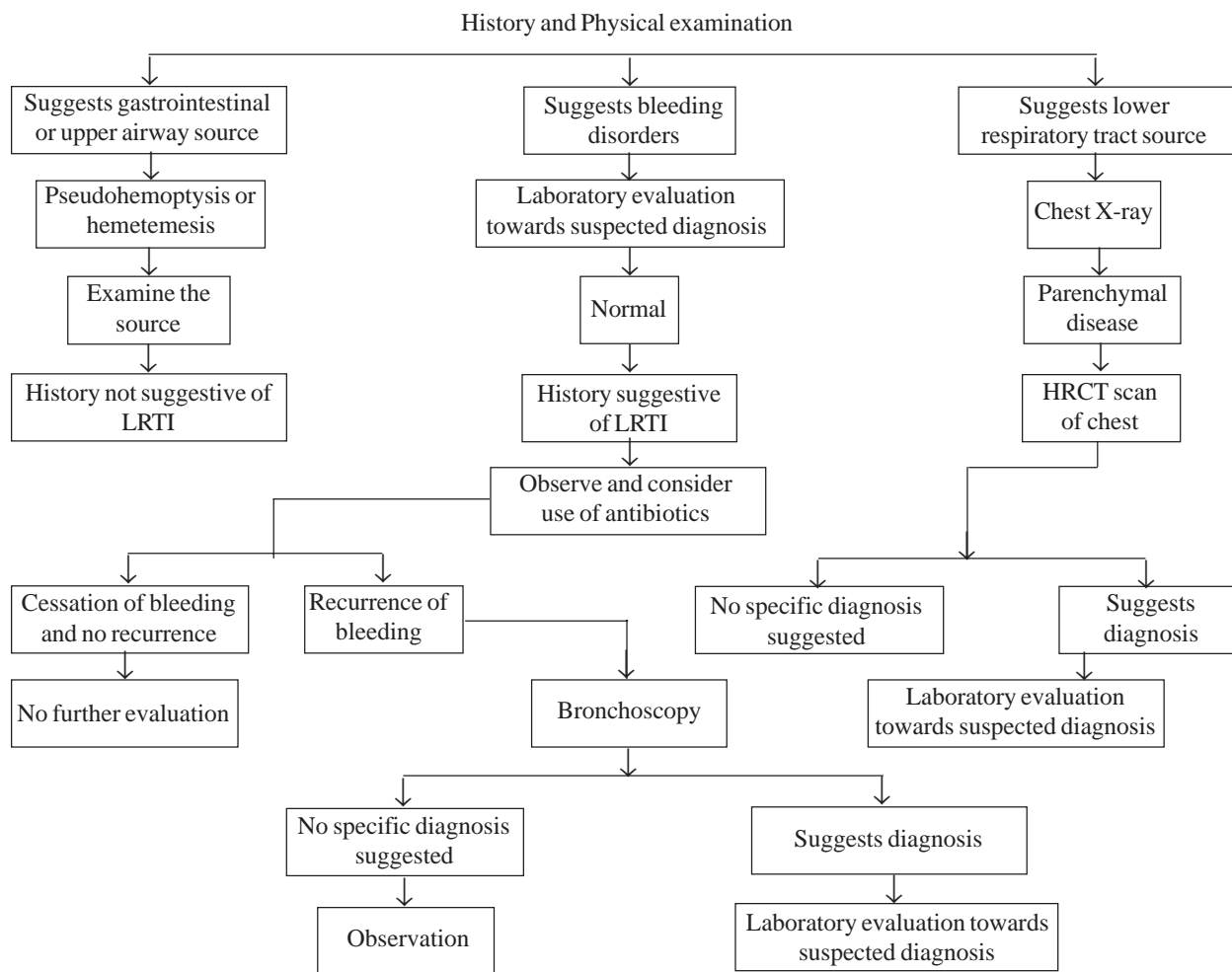


FIG. 1 Algorithm for diagnosing hemoptysis in children.

and hence lead to the risk of aspiration. However, judicious use and careful titration should avoid this problem(36). The various options available for massive hemoptysis are: endoscopic balloon occlusion of a lobe or main bronchus, topical airway vasoconstrictors, use of Nd-YAG laser, CO₂ laser bronchoscopy, endoscopic tumor excision, transcatheter embolisation of bronchial vessels and lobectomy. The foremost objectives in management of massive hemoptysis are to protect the airways, maintain oxygenation, stop the bleeding and maintain sufficient blood volume. This is critical because most of the deaths occur due to asphyxiation, not exsanguination. The airway should be kept patent with an endotracheal tube or rigid bronchoscope in cases of severe respiratory distress.

The bleeding should be localized without delay. Although there is debate regarding timing of bronchoscopy, most of the authors favor early bronchoscopy(35). This approach is associated with the best success of identifying the bleeding site. In most cases, fiberoptic bronchoscopy via an existing endotracheal tube is the easiest and safest approach. If fiberoptic bronchoscopy fails to identify the bleeding site, an experienced endoscopist can perform rigid bronchoscopy if time permits. Rigid bronchoscopy protects the airways and helps in localization of bleeding site. Topical vasoconstrictors, such as oxymetazoline or epinephrine (1:20000), iced 0.9% saline, fibrinogen-thrombin glues or Botropase are applied through the bronchoscope to curtail the bleeding. In addition,

endoscopic tamponade can be performed by balloon catheter or with bronchoscopic pressure(2). Patients with alveolar hemorrhage syndromes, blood dyscrasias, or those on anticoagulant or antiplatelet therapy should receive appropriate medical therapy rather than invasive interventions.

If the hemorrhage continues despite endoscopic measures, or if the bleeding site cannot be localized, emergency arteriography is carried out(5). Selective bronchial artery embolisation (BAE) is effective in the emergency management of life-threatening hemoptysis(37). Bronchial arteriography identifies the bleeding vessel and selective embolisation with absorbable gelatin sponge, steel or platinum coils, or polyvinyl alcohol particles controls the hemorrhage. In one study(34), there was immediate cessation of hemorrhage in 77% of the cases; while long term control beyond 3 months was achieved in 45% of patients. Another study(37) found bronchial embolisation to be superior to medical management for resolution of life threatening bleeding. BAE is effective in controlling hemoptysis in most of the cases although recurrent bleeding is not uncommon. BAE is technically challenging and requires angiographic skills that are not always available in pediatric hospitals(38). Complications are not uncommon and may be severe or even fatal. The most serious complication of embolisation therapy is neurological damage due to embolisation of the spinal arteries(5). Recently, del Gregorio, *et al.*(39) have reported their experience with bronchial artery embolisation in patients with hemoptysis; about 31% of these patients were having bronchiectasis. In this study, clinically successful outcome with BAE was observed in 91.1% of the cases, and repeat embolisation was required in 6.7% of the cases. Recurrence of hemoptysis was observed in 22.3% of cases, but only 10.4% of patients required repeat embolisation. Minor complications that did not require treatment were observed in 28% of cases. Thus, BAE is a nonsurgical treatment that is safe and effective in patients with massive hemoptysis(39).

If bronchial artery embolisation fails to control hemoptysis, surgical management should be considered. Massive hemoptysis carries greater than 50% mortality without surgical intervention. Surgery is offered to patients who can withstand general

anesthesia and pulmonary resection. Immediate identification of the bleeding site by bronchoscope is followed by resection of the bleeding source(40). Segmentectomy or lobectomy is preferred to pneumonectomy, as the latter carries a much higher mortality rate. One study(41) reported 83% survival in 65 resections performed for massive hemoptysis in this setting. Simrali, *et al.*(42) recently published their experience with surgery for bronchiectasis in 176 children of less than 16 years with hemoptysis. They performed various procedures including segmental resection, lobectomy, pneumonectomy and bilateral resection in patients with bronchiectasis. Complete resection was performed in almost 94% of the cases. The patients were followed for a mean period of 4.3 years. Successful outcome was observed in 73% of cases, while improved outcome was seen in another 23% of the cases. It was observed that there was good improvement in the quality of life even after pneumonectomy and bilateral resection of the lungs. Hemoptysis could be controlled in all of these cases. It was concluded that with acceptable mortality and morbidity rates and high chances of cure after complete resection, surgical treatment is a successful and reliable method of treatment in childhood bronchiectasis that yield marked improvement in the quality of life(42). Chan, *et al.*(27) has also treated isolated pulmonary arteritis with lobectomy, as the hemoptysis could not be controlled with the medical measures alone. Hence, surgical options have to be considered if massive hemoptysis remains uncontrolled in spite of medical therapy.

Figure 2 provides an algorithm for the management of hemoptysis in children.

Hemoptysis in children is rare, but if occurs, it is a frightening symptom for the parents. The patient's history should help determine the amount of blood and differentiate between hemoptysis, pseudo-hemoptysis, and hematemesis. A focused physical examination can lead to the diagnosis in most cases. Mild hemoptysis often is caused by an infection and can be managed on an outpatient basis with close monitoring. Massive hemoptysis requires immediate hospitalization and proper therapeutic interventions have to be initiated at the earliest to stop the bleeding and to prevent its recurrence.

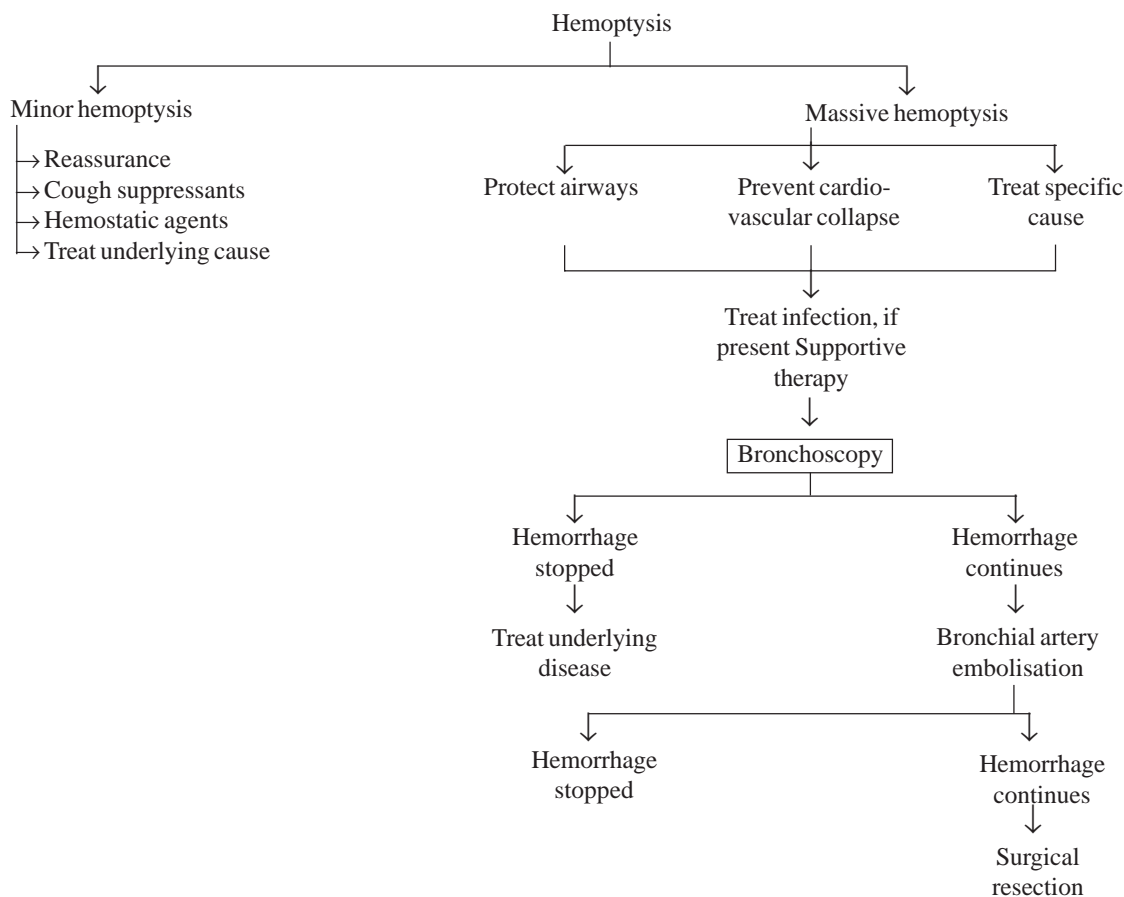


FIG. 2 Algorithm for management of hemoptysis in children.

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