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Pediatric Hemoptysis

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Abstract
Hemoptysis or expectoration of blood is one of the respiratory symptoms in children, hemoptysis is hard to detect because it is often swallowed. The common causes of hemoptysis in adults include bronchiectasis, tuberculosis, pulmonary mycosis, and lung cancer. On the other hand, hemoptysis in children is often caused by tracheobronchitis, pneumonia, bronchiectasis in cystic fibrosis, and foreign body aspiration. Bleeding of the pulmonary artery or bronchial artery is the cause of hemoptysis. Upper airway examination is important to rule out epistaxis as the cause of hemoptysis. The primary objectives of hemoptysis management are asphyxia management, bleeding cessation, and treatment of the underlying disease. When all other treatments fail to stop the bleeding, surgery is recommended. ***(J Respirol Indones 2021; 41(3): 214–20)***
Keywords: *hemoptysis in children, pediatric hemoptysis*

Hemoptisis pada Anak

Abstrak
Batuk darah atau hemoptisis merupakan salah satu gejala penyakit paru. Pada anak, batuk darah sulit terdeteksi karena pada umumnya anak sering menelan dahak. Penyebab tersering batuk darah pada pasien dewasa adalah bronkiektasis, tuberkulosis, infeksi jamur di paru, dan kanker paru. Sedangkan pada anak, penyebab tersering batuk darah antara lain, trakeobronkitis, pneumonia, bronkiektasis pada fibrosis kistik dan aspirasi benda asing. Hemoptisis dapat terjadi akibat pecahnya arteri pulmoner atau arteri bronkial. Pemeriksaan saluran napas atas penting untuk menyingkirkan sumber perdarahan akibat epistaksis. Tujuan utama tatalaksana hemoptisis adalah memastikan jalan napas tetap terbuka untuk mencegah asfiksia, menghentikan perdarahan, dan menatalaksana penyakit dasarnya. Pembedahan merupakan pertimbangan terakhir jika tatalaksana lainnya tidak dapat menghentikan perdarahan. ***(J Respirol Indones 2021; 41(3): 214–20)***
Kata kunci: *batuk darah pada anak, hemoptisis pada anak*

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INTRODUCTION

Coughing up blood or hemoptysis is one of the symptoms of pulmonary disease. In children, the incidence of hemoptysis is difficult to estimate because, in general, children often swallow phlegm. Acute idiopathic pulmonary hemorrhage (AIPH) is the discovery of blood in the airway of a child aged ≤1 year without predisposition to other diseases and respiratory distress that can cause respiratory failure.[1] The etiology of hemoptysis in pediatric patients is different from that in adult patients. In adult patients, tuberculosis is one of the most common diseases, causing symptoms of coughing up blood. However, tuberculosis in pediatric patients is a systemic disease and rarely causes blood coughing. Therefore, information about coughing blood in pediatric patients is also important to be known by a pulmonary doctor. This library review covers the definition, etiology, pathophysiology, diagnosis, and management of hemoptysis in children.

DEFINITION

Hemoptysis or coughing blood is the expectoration of blood coming from the lower airway.[1] Another source specifically mentions that hemoptysis is the expectorant blood or bloody phlegm derived from the airways under the vocal cords.[2] Hemoptysis should be distinguished by hematemesis (vomiting blood) and epistaxis. Table 1 illustrates the differences between hemoptysis and hematemesis.[3] Department of Pulmonology and Respiratory Medicine Faculty of Medicine, Universitas Indonesia – Persahabatan Hospital using massive hemoptysis criteria as follows:[2]

a. Coughing up blood ≥600 ml/24 hours and does not stop during observation.
b. Coughing up blood ≥250 ml but <600 ml/24 hours and laboratory tests show hemoglobin <10 g% while coughing up blood continues.
c. Coughing up blood ≥250 ml but <600 ml/24 hours hemoglobin levels 10 gr%, and observation for 48 hours, with conservative treatment, this process has not stopped.

No consensus classifies the degree of hemoptysis in children. The American Thoracic Society distinguishes hemoptysis in patients with cystic fibrosis into scant (<5 ml), mild-to-moderate hemoptysis (5–240 ml and massive hemoptysis (>240 ml).[3] Department of pulmonology and respiratory medicine, Faculty of Medicine, Universitas Indonesia – Persahabatan Hospital applied the following criteria for massive hemoptysis in adults, which is the expectoration of blood: at least 600 mL in 24 hours, or 250–600 mL in 24 hours with hemoglobin (Hb) <10 gr/dL and still ongoing during observation, or 250–600 mL in 24 hours with Hb >10 gr/dL and still ongoing in 48 hours.[4]

Table 1. The Differences in Hemoptysis and Hematemesis

No	Hemoptysis	Hematemesis
Historical		
1	No complaints of nausea and vomiting	Nausea and vomiting
2	There is a history of pulmonary disease	There is a history of gastrointestinal disease
3	Asphyxia	Rarely asphyxia
Phlegm examination		
1	Foaming	Rarely foaming
2	Liquid or blood clot	Like coffee powder
3	Bright red or pink	Brownish to blackish
Laboratory		
1	Alkaline pH	Acidic pH
2	A mixture of macrophages and neutrophils	Mixed food

ETIOLOGY

The most common causes of hemoptysis in adult patients are bronchiectasis, tuberculosis, fungal infections in the lungs, and lung cancer.[5,6] Research at Persahabatan Hospital showed data from 323 hemoptysis patients admitted to the emergency room of Persahabatan Hospital caused by tuberculosis (64.4%), bronchiectasis (16.7%), and lung cancer (3.4%). Other research conducted at The Persahabatan Hospital also showed that tuberculosis (75.6%) is the main cause of inpatient hemoptysis and IGD, followed by the previous history of pulmonary tuberculosis (16.7%).[7] In children, hemoptysis can be caused by various diseases, such as the following.

a. Pulmonary Disease
   1. Tuberculosis
   2. Pneumonia

3. Aspergillosis
4. A parasitic infection (*P. Westermani*)
5. Bronchiectasis
6. Cystic fibrosis
7. Pulmonary abscess
8. Tumors (Adenoma, carcinoids, hemangiomas, metastases in the lungs, immunoblastic sarcoma, Kaposi sarcoma)
9. Aspirations of foreign bodies
10. Diffuse Alveolar Damage (DAD)
11. Acute Respiratory Distress Syndrome (ARDS)
12. Tracheobronchitis
13. Contusion or trauma
14. Bronchogenic cysts

b. Cardiovascular Disease
1. Heart failure
2. Eisenmenger syndrome
3. Mitral stenosis
4. Veno-occlusion disease
5. Venous artery malformations
6. Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome)
7. Pulmonary embolism
8. Pulmonary hypertension

c. Immune System Disorders
1. Purpura Henoch-Schonlen
2. Idiopathic Pulmonary Hemosiderosis
3. Goodpasture syndrome
4. Granulomatosis Wegener
5. Systemic Lupus Erythematosus
6. Polyarteritis nodosa
7. Heiner syndrome
8. Pulmonary alveolar proteinosis
9. Sclerosis tuberous
10. lymphangiomyomatosis or lymphangioleiomyomatosis

d. Other Conditions
1. Gastroesophageal reflux
2. Hyperammonemia
3. Kernicterus
4. Intracranial bleeding (in premature infants)
5. Poisoning
6. Diffuse alveolar injury (Smoke inhalation)
7. Post spinal cord transplant
8. Coagulation disorders

Among these diseases, the most common causes of hemoptysis in children are tracheobronchitis and pneumonia.[5] Foreign bodies are also one of the most common causes.[8] Aspiration of foreign bodies often occurs in children under 3 years of age. It is generally not immediately detected until symptoms such as chronic cough, pneumonia or hemoptysis occur.[10] In addition, tracheostomy-related bleeding is also a common cause.[9]

PATHOPHYSIOLOGY

The lungs are fertilized by two arteries, the pulmonary artery and the bronchial artery. Pulmonary arteries make up 99% of the lungs and play a role in gas exchange. The bronchial artery provides nutrients for the airways, mediastinum glands, nerves, visceral pleura, esophagus, vasa vasorum of the aorta, pulmonary arteries, and pulmonary veins.[6,7]

Hemoptysis can occur due to bleeding of the pulmonary artery or bronchial artery. Pulmonary arteries have a large volume with low pressure. The branched pulmonary artery follows the bronchi to the terminal bronchi. This branching of the pulmonary artery then branches out to form a capillary pad (anastomosis) that envelops the alveolus before returning to the left atrium through the pulmonary vein. The bronchial artery has a smaller volume but has high systemic pressure. This artery is an aortic branch or intercostal artery. Bleeding coming from the pulmonary artery is usually slow due to its lower pressure. Heavy bleeding usually originates from the bronchial arteries due to high hydrostatic pressure, increasing the rate of bleeding.[9]

The basic disease that causes hemoptysis can be estimated by knowing the source of the bleeding. Alveolar bleeding is usually caused by an autoimmune disease or post-spinal cord transplant. Bleeding in the airways can occur due to airway hemangioma, pulmonary artery-venous malformations (hereditary hemorrhagic telangiectasia), and collateral bronchial arteries that appear in patients with chronic infections, especially

cystic fibrosis. Pulmonary bleeding can occur locally as well as diffuse. Chronic infections or inflammation are common causes of hemorrhagic voices in isolated bronchial lesions.[1]

Massive hemoptysis usually occurs due to bleeding in the bronchial arteries.[7] Massive hemoptysis is also biased due to erosion caused by chronic inflammation of the airways located adjacent to the bronchial arteries. Blood derived from this erosion is generally bright red and spurts out. This is due to the dilation of the bronchial artery and systemic artery pressure. Within 48–72 hours of bleeding, alveolar macrophages convert iron inside erythrocytes into hemosiderin. Macrophages containing hemosiderin are in the alveolus for several weeks until finally eliminated from the alveolus.[1]

It can be used to distinguish between acute or chronic bleeding. Pulmonary bleeding is also often followed by an increase in neutrophils and other pro-inflammatory mediators. In cases of recurrent pulmonary bleeding or chronic bleeding, pulmonary fibrosis is one of the most commonly found abnormalities.[1]

Chronic infections and inflammation that occur in children with bronchiectasis or chronic fibrosis can cause airway damage, encourage the formation of new blood vessels (neovascularization), and cause dilation and fragility of the walls of the bronchial arteries. These fragile blood vessels form close to the mucosa where the inflammation occurs, making it easy to bleed from coughing or infections.[8]

When the child coughs strongly, coughing up blood can occur due to mechanical trauma of the airways. However, coughing up blood like this is generally mild and self-healing. Airway injury can also occur during the suction of tracheostomy pipes or endotracheal pipes.[8] In cystic fibrosis patients, endobronchial bleeding generally indicates the erosion of the secondary airway wall due to infection.[1] In children with pulmonary hypertension, hemoptysis occurs due to the rupture of pulmonary blood vessels. However, this is rare.[9]

DIAGNOSIS

Diagnosis of pulmonary bleeding is established if blood or hemosiderin is found in the lungs. Anamnesis and physical examinations are important for directing the causes of bleeding in the lungs, extra lungs (gastrointestinal tract and upper airway), or coagulation disorders. One of the most common complaints in children of greater age is a feeling of “bubbling” on the chest wall.[1]

Other symptoms that can be found in children who have experienced a lot of blood volume loss are cyanosis, respiratory failure, and shock. Laboratory examinations are conducted to evaluate whether or not coagulation disorders exist. In finding the source of bleeding, an upper airway examination is important to rule out the possibility of bleeding due to epistaxis.[5] Nasopharyngoscopy can help to perform such examinations. The examination modalities that can be used to evaluate the source of bleeding and basic diseases include thoracic photos, chest angiogram CT, bronchoscopy, and echocardiography. Biomarkers can be examined to rule out possible causes of autoimmune diseases.[5]

MANAGEMENT

The important thing to note in the treatment of hemoptysis is the amount of blood excreted and the primary disease that causes hemoptysis. The main purpose of hemoptysis management is to ensure that the airway remains open to prevent asphyxia, stop bleeding, and manage primary diseases.[10] Most cases do not require intervention because hemoptysis in children usually heals itself spontaneously. Calming children and families are one of the earliest things that can be done when hemoptysis occurs. Mild hemoptysis can be treated according to its symptoms, such as the administration of hemostatic agents, as well as overcoming its basic diseases and administration of antibiotics suitable for basic infectious diseases, antibiotics and corticosteroids in cystic fibrosis patients, anti-tuberculosis drugs (Anti-TB) for children with tuberculosis, and modifying suction

techniques in children with hemoptysis related to tracheostomy.[11]

Massive hemoptysis can progress to acute respiratory distress syndrome (ARDS) quickly. In children with massive hemoptysis can be carried out intubation and mechanical ventilation. Mechanical ventilation with PEEP (Positive End-Expiratory Pressure) also serves as a tamponade for active bleeding in addition to improving oxygenation. Other procedures for acute periods include oxygen administration, blood transfusions, and resuscitation fluid administration.[1,10]

Transfusions are administered in the case of hypotension or when the hematocrit decreases significantly. Antitussive drugs should not be administered. The child is laid on the side of the sick lung to prevent blood from filling the healthy side of the lung. In cystic fibrosis patients, vitamin K should be administered in patients with prothrombin disorders (PT).[12]

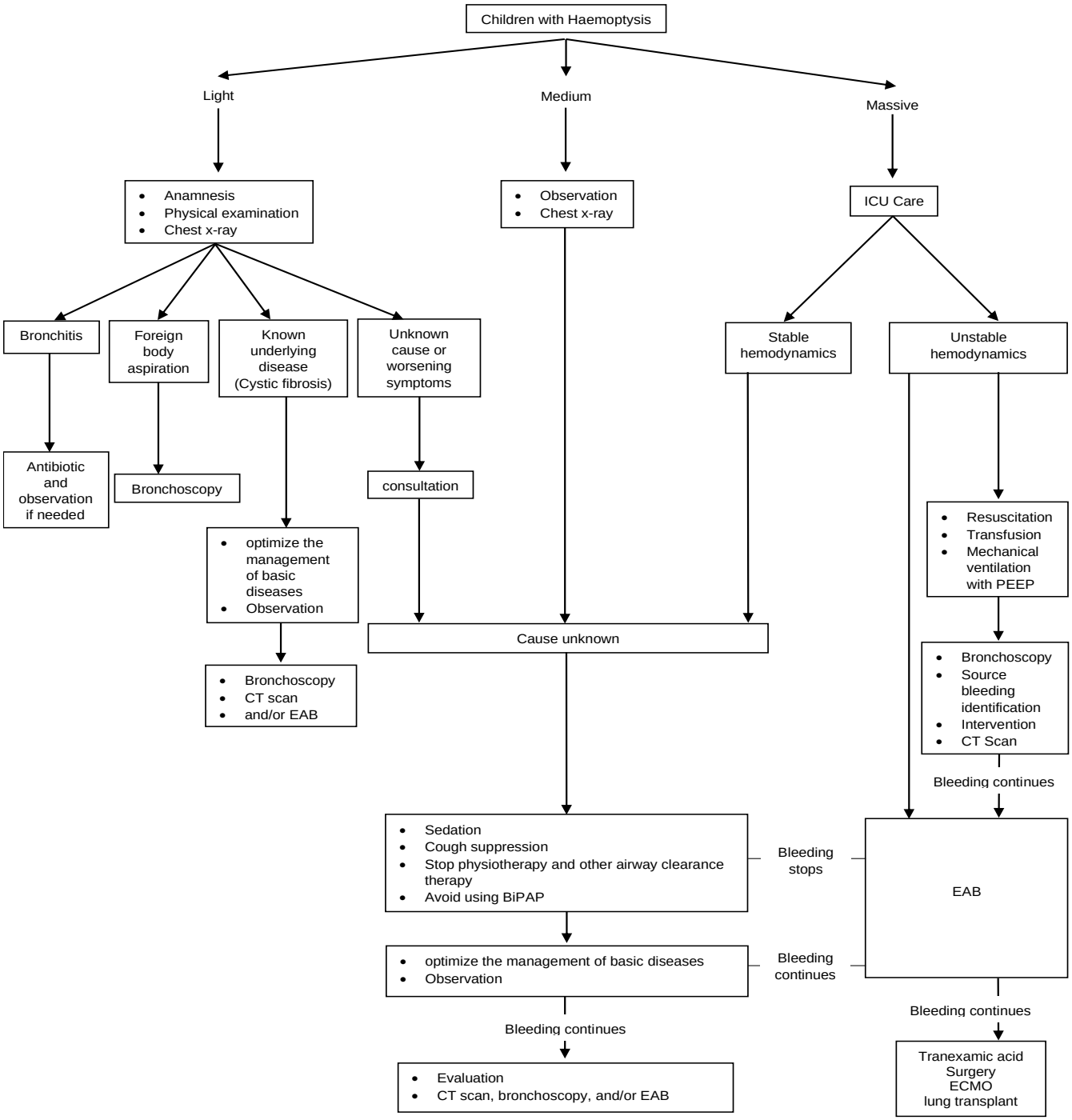

Figure 1. Hemoptysis Management Flow in Children.

Anti-coagulation drugs should be discontinued. Ticarcillin, salicylates, and nonsteroidal anti-inflammatory drugs (NSAIDs) can interfere with platelet function and aggravate hemoptysis.[12] The efficacy of anticoagulation agents such as tranexamic acid in treating hemoptysis is still unknown. However, the results of a new study found that the use of inhaled tranexamic acid may help stop non-massive hemoptysis.[14]

Bronchoscopy is a gold standard for seeing the location of the bleeding directly. However, in practice, bronchoscopy rarely shows the source of bleeding.[4] Other roles of bronchoscopy include removing foreign differences, taking biopsy specimens, and taking specimens for examination of microorganism cultures. Rigid bronchoscopy is better than flexible bronchoscopy because rigid bronchoscopy can be performed, such as administering vasoconstriction agents and endobronchial tamponades without interfering with ventilation.[10]

Meanwhile, the advantage of flexible bronchoscopy is that it can reach deeper branches of the bronchi compared to rigid bronchoscopy. Endobronchial tamponade techniques can be performed using Fogarty balloon catheters, double-lumen bronchus-blocking catheters, and modification of J angiography wire measuring 0.035 inches, or with pulmonary artery catheter balloons. Topical vasoconstrictors can be administered, such as oxymetazoline or epinephrine (1:20,000), saline 0.9% cold, or fibrinogen/thrombin (Botropase).[10]

Laser neodymium-yttrium-aluminum-garnet (Nd-YAG), laser CO2, and argon plasma coagulation (APC) can also be done to stop bleeding. However, the data on laser use in hemoptysis treatment in children is still limited. Angiography with or without embolization and surgery is performed in patients with vascular disorders if intervention with bronchoscopy is unsuccessful.[1]

Embolization of the bronchial artery is useful to control persistent hemoptysis in cystic fibrosis patients. The American Thoracic Society recommends that cystic fibrosis patients experiencing massive and clinically unstable hemoptysis should be Embolization Artery Bronchial (EAB).[12]

In cystic fibrosis patients, bronchoscopy should not be performed first in patients with massive hemoptysis if the EAB has not been performed. Bronchial arteriography helps determine the location of the bleeding and becomes a map of the direction leading to the location of bleeding. When performing angiography and embolization, the arteries that supply blood to the spinal cord need to be considered. The anterior spinal artery is the main artery that supplies the spinal cord. The arteries receive their blood supply from the same truncus that supplies the superior intercostal arterial branching and the right bronchial artery. The left bronchial artery rarely contributes to the anterior spinal artery. Therefore, when embolizing, especially the area of the right bronchial artery, it is necessary to pay attention not to disturb the anterior spinal artery.[15]

Embolization can be performed with a gelatin sponge that absorbs, rolled steel or platinum, or polyvinyl alcohol particles. The most effective treatment for overcoming massive hemoptysis, whether originating from the bronchial or non-bronchial circulation, is pulmonary artery embolization. The level of EAB in stopping bleeding is 73–99%, with a repeated incidence of hemoptysis 10–55% in monitoring for 46 months. Complications of EAB include neurological damage due to embolization of the spinal artery and recurrent hemoptysis.[6]

Lobectomy in children with hemoptysis is avoided to maintain the pulmonary functions of the child.[1] Surgery in patients with massive hemoptysis is performed as a last resort if other procedures do not manage to overcome the cough of blood.[12]

CONCLUSIONS

Hemoptysis is rare in children. However, massive hemoptysis can lead to death in children. Infections, particularly lower airway infections, aspiration of foreign bodies, and bronchiectasis,

particularly bronchiectasis in cystic fibrosis, are the most common causes of hemoptysis in children. Hemoptysis is usually mild and can heal itself, but in massive hemoptysis, the main principle is to prevent asphyxia and manage the primary disease. Bronchoscopy and EAB can be performed to stop bleeding. Surgery is performed if other procedures are unsuccessful.

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