Pulmonary Hemorrhage/Hemoptysis in Children

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Summary. Pulmonary hemorrhage and hemoptysis are uncommon in childhood, and the frequency with which they are encountered by the pediatric pulmonologist depends largely on the special interests of the center to which the child is referred. In those centers caring for children with cystic fibrosis or congenital heart disease, these will be by far the most common causes of hemoptysis. Other causes of hemoptysis are far less common, such as bleeding from localized lesions in the upper airway or tracheobronchial tree. Even less common is bleeding into the lungs as part of a systemic disease, usually with renal involvement (pulmonary-renal syndromes), such as systemic lupus erythematosus or Goodpasture’s syndrome. Bleeding into the lungs in children with a bleeding diathesis probably only occurs in immunosuppressed children after transplantation. When no other cause is found for pulmonary hemorrhage, the presumed diagnosis is idiopathic pulmonary hemosiderosis. This review discusses the various causes of hemoptysis and pulmonary hemorrhage, and the appropriate investigations to aid in determining the correct diagnosis. The management and prognosis of idiopathic pulmonary hemosiderosis, based on cumulative experience from published reports, are considered in more detail.


Key words: hemosiderosis; hemoptysis; diagnosis; management; children.

INTRODUCTION

Pulmonary hemorrhage and hemoptysis in children are uncommon, even though bronchiectasis, which is not so uncommon, especially in cystic fibrosis, may present with bleeding. Pulmonary hemorrhage and hemoptysis may also be due to very rare conditions such as pulmonary-renal syndromes and idiopathic pulmonary hemosiderosis (IPH). Since bleeding into the lung can be dangerous and even life-threatening, it is important to have a good understanding of its likely causes, investigation, and management. The incidence of hemoptysis in children is impossible to determine with any accuracy since, even if almost all children with this worrying symptom present for medical attention, they may present to physicians in different specialties and different types of institution. The frequency with which hemoptysis is a presenting symptom will then depend on the mix of patients being treated at the institution in question. Thus Coss-Bu et al.1 reviewed 228 children and young adults who presented to a major referral center (Texas Children’s Hospital) over a 10-year period. Hemoptysis was due to cystic fibrosis (CF) in 65% of these patients, to various types of congenital heart disease in 16%, and to a whole variety of other problems in the rest. Excluding the patients with CF, in 46% of the remaining 79 children, hemoptysis was due to congenital heart disease, 16% to infectious causes other than CF, 8% to neoplasms, and 14% to various other causes. The very large number of children with CF and congenital heart disease in this series obviously reflects the medical and surgical interests and expertise of this particular institution. In our own department, which is a tertiary pediatric pulmonary service with major interests in congenital lung anomalies, pulmonary infection, and asthma, we have undertaken 2,148 bronchoscopies in children aged less than 18 years over the past 6.5 years. Of these, hemoptysis was the primary indication for bronchoscopy in only 17 children (0.8% of bronchoscopies). Of these 17 children, the final diagnosis was bleeding due to upper airway disease in 5 (29.4%), bleeding from a tracheostomy site in 3 (17.6%), no cause found in 3 (17.6%), and a variety of other causes

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Received 23 October 2003; Revised 9 November 2003; Accepted 9 November 2003.

DOI 10.1002/ppul.20020
Published online in Wiley InterScience (www.interscience.wiley.com).
in the remaining 6 children. Of course, the incidence of hemoptysis in the younger child may be underestimated, as young children usually swallow their sputum.

A summary of the conditions which are known to be associated with hemoptysis in children, graded approximately by the frequency with which they occur in the general population, is given in Table 1.

### RELATIVELY COMMON CONDITIONS

**Foreign-Body Aspiration, Infection, Cystic Fibrosis, Etc.**

While the aspiration of a foreign body in childhood is relatively common, especially in boys in the 1–3-year age group, hemoptysis due to this cause is uncommon even with the aspiration of sharp objects such as pins. Among our 17 children with hemoptysis, only one was due to the aspiration of a foreign body, and this was of vegetable origin. Bleeding is much more likely to occur during the removal of a vegetable foreign body which has been in place long enough to become infected. Chronic lung infection, especially with bronchiectasis due to cystic fibrosis (or tuberculosis where this is common), is quite likely to present as hemoptysis in older children, as noted in the series of Coss-Bu et al. An unrecognized aspirated foreign body may, of course, be the cause of the chronic lung infection.

### Upper Airways, Gastrointestinal Bleeding

Rather surprisingly, in our own experience, bleeding thought to be hemoptysis was quite likely to come from the nose or from infected tonsils or adenoids. Even so, a normal chest radiograph in a child with suspected hemoptysis is no guarantee that the bleeding is from the upper airway, and bronchoscopy will almost always be indicated. Hematemesis, also uncommon in children, may be very difficult to distinguish from hemoptysis and should always be considered in the differential diagnosis.

### Trauma: Accidental, Surgical, and Otherwise

Direct trauma to the chest due to motor accidents or other causes can cause pulmonary contusion and occasionally pulmonary hemorrhage. In a child with tracheostomy for whatever cause, bleeding from around the stoma is not particularly uncommon in our experience, and may present as hemoptysis or a blood-stained aspirate during suctioning. Perhaps the most important cause of traumatic bleeding in children is due to accidental or deliberate suffocation. A significant number of infants who die from sudden infant death syndrome (SIDS) are found with blood in the mouth or lungs at autopsy. Southall et al. noted bleeding from the nose or mouth in 11 of 39 children undergoing covert video surveillance for alarming life-threatening events (ALTEs) and suspected child abuse, compared with none of 46 infants with ALTEs due to natural medical causes.

### Cardiac Causes

Pulmonary arterial or venous hypertension due to congenital or acquired heart disease was a prominent cause of hemoptysis in those children who did not have cystic fibrosis in the series of Coss-Bu et al. Pulmonary hemorrhage may be the first symptom in a child with idiopathic primary pulmonary hypertension, although this is much less common than congenital heart disease. Unless there is another obvious cause of pulmonary hemorrhage, the child should undergo echocardiography and be examined by a pediatric cardiologist.

### Tumors, Anomalies, Etc.

Among the rare causes which the pediatric pulmonologist can expect to encounter, tumors such as pulmonary adenoma or carcinoid tumors of varying degrees of malignancy may present as hemoptysis, usually in an older
child. However, hemoptysis due to a tumor was found in only 2.6% of the patients of Coss-Bu et al.¹ and in 6% of our much smaller series. Caution should be used in approaching tumors in the lung in children, as they may bleed massively and it may be difficult to control the bleeding through a fiberoptic bronchoscope in a small airway. Congenital vascular anomalies in the lung, such as multiple arteriovenous fistulae, are known to be a cause of hemoptysis in adults and are often associated with hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). In the series reported by Swanson et al.,⁴ there was at least one child, as the age range was given as 5–83 years, but the mean age of presentation was 40 years.

**Bleeding Diathesis**

Nonpulmonary causes of bleeding from the lungs include bleeding due to primary hematologic disorders, but this is quite uncommon. Although hemoptysis has been reported occasionally in adults with hemophilia, this does not seem to have been reported in childhood. Immunocompromised children, especially after solid organ and bone marrow transplantation, sometimes develop acute respiratory distress due to pulmonary hemorrhage.

**RELATIVELY RARE CONDITIONS**

A small number of conditions present with pulmonary hemorrhage of varying degrees in children, all of which are so rare that even the pediatric pulmonologist is likely to encounter them only occasionally. One group of conditions comprises pulmonary hemorrhage associated with systemic diseases of the “autoimmune” type also involving the kidneys, and is termed the “pulmonary renal syndromes.” The other condition (or conditions?) in which there appears to be no systemic involvement and in which the pathology is localized to the lungs is termed “idiopathic pulmonary hemosiderosis” (IPH).

**Pulmonary-Renal Syndromes**

The specific pulmonary-renal syndromes are those in which severe pulmonary and renal involvement appear to be the major features of the disease and include Goodpasture’s syndrome (GPS), systemic lupus erythematosus (SLE), Wegener’s granulomatosis (WG), and microscopic polyangiitis (MP). In GPS and SLE, diffuse pulmonary hemorrhage occurs as part of the pulmonary vasculitis, while in WG and MP, bleeding is more likely to occur from cavitary lesions. In addition, pulmonary hemorrhage was also reported in children with Henoch-Schonlein purpura (HSP), a relatively common condition in childhood who have renal involvement,⁵ and this should probably be included in the specific pulmonary-renal syndromes. There are also nonspecific pulmonary-renal syndromes in which the pulmonary problem is not an integral part of the disease, such as pulmonary edema, embolism, or infection in a child with renal disease. In a survey of pulmonary-renal syndromes in children collected in a major pediatric center in Bern, von Vigier et al.⁶ described 21 children, all but 5 of whom had nonspecific pulmonary-renal problems. Of the 5 with specific syndromes (less than one child per year), 2 had SLE, 2 had WG, and 1 had MP. In their review of the literature on specific pulmonary-renal syndromes in children, von Vigier et al.⁶ were able to find 13 with GPS, an unstated larger number with SLE, 28 with WG or MP, and 11 with HSP.

An excellent discussion of specific pulmonary-renal syndromes was recently included in a Case Record of the Massachusetts General Hospital⁷ concerning a child with hemoptysis. Table 2 was adapted from the table in this paper and expanded to include HSP and the usual treatment modalities.

**Goodpasture’s Syndrome**

This condition is an immune complex disease in which the damage is almost entirely confined to the kidneys and

| TABLE 2—Some Features of Pulmonary-Renal Syndromes¹ |
|-----------------|----------------|-----------------|----------------|
| GPS | SLE | WG | MP | HSP |
| Pulmonary hemorrhage | ++++ | + to ++ | +++ | +++ | ++++ |
| Glomerulonephritis | ++++ | +++ to ++++ | ++++ | ++++ | ++++ |
| Upper airway involvement | | | | | |
| Skin rash | 0 to + | + to ++ | ++++ | ++ | ++ |
| Arthralgia | 0 | ++++ | +++ | ++++ | +++ |
| Elevated ESR | 0 to + | ++++ | ++++ | ++++ | + |
| Abdominal involvement | 0 | 0 | 0 | 0 | ++++ |
| Positive serology | anti-GBM, p-ANCA (rarely) | ANA, anti-DS-DNA, p-ANCA (rarely) | c-ANCA, p-ANCA (rarely) | p-ANCA, c-ANCA | IgA+, IgM+ |
| Treatment | CS +, CT +, plasmaphoresis | CS +, CT + | CS +, CT + | CS +, CT ? | CS |

¹GPS, Goodpasture’s syndrome; SLE, systemic lupus erythematosus; WG, Wegener’s granulomatosis; MP, microscopic polyangiitis; HSP, Henoch-Schonlein purpura; GBM, glomerular basement membrane; ANA, antinuclear antigen; DS-DNA, double-stranded DNA; ANCA, antineutrophilic cytoplasmic antigen; CS, corticosteroid; CT, cytotoxic.
lungs. The children have glomerulonephritis and pulmonary hemorrhage, with little or no involvement of other systems. The diagnosis is confirmed by the finding of antiglomerular basement membrane antibodies in peripheral blood, and there is rarely any need for renal or lung biopsy. While much more common in adults, \(^8\) GPS has been reported for over 20 years in children with typical clinical and immunologic findings. \(^9\) Management includes the use of plasmapheresis in addition to corticosteroids. The response appears to be good if started early in the disease, although there are very few reports of the outcome in children.

**Systemic Lupus Erythematosus**

In a study of SLE between 1980–1995, Font et al.\(^{10}\) noted that some 8% of their patients were children or adolescents with a mean age of onset of 11 years, and of these 91% were girls. The disease in children resembled that in adults, with rash, arthritis, fever, and nephropathy being common. Diffuse alveolar hemorrhage is an uncommon but well-documented manifestation of SLE in children.\(^{11}\) Of 34 children in the study of Font et al.,\(^{10}\) only 2 (6%) had lung involvement. The diagnosis is confirmed by the finding of antinuclear antibodies (ANA) and antihistone antibodies in peripheral blood, and there is rarely any need for renal or lung biopsy. While much more common in adults, \(^8\) GPS has been reported for over 20 years in children with typical clinical and immunologic findings. \(^9\) Management includes the use of plasmapheresis in addition to corticosteroids. The response appears to be good if started early in the disease, although there are very few reports of the outcome in children.

**Wegener’s Granulomatosis and Microscopic Polyangiitis**

These are closely related, if not identical, conditions which are distinguished from one another on the basis of serological reactions. In WG, the cytoplasmic staining anti-nuclear cytoplasmic antibody (cANCA) is normally present in serum, while the perinuclear staining antibody (pANCA) is absent. In MP, both types of ANCA can be present. In adults, these conditions are relatively common ways in which the lungs are involved in systemic disease, but WG and MP were found in only 3 children over a 7-year period in a Swiss study.\(^6\) Pulmonary granulomatous disease, sometimes with cavitation, upper airway lesions, skin vasculitic lesions, and renal involvement, is a common clinical manifestation of these conditions. Hemoptysis may occur due to bleeding from cavitary lesions. Rottem\(^{14}\) compared the clinical manifestations of WG in 23 children and 135 adults who were mostly being treated with corticosteroids and cyclophosphamide. WG was similar in childhood-onset and adult-onset patients, but childhood-onset patients were complicated five times more often by subglottic stenosis and twice as often by nasal deformity. Similar observations on children with WG were reported by Belostotsky et al.,\(^{15}\) who analyzed data on 17 children collected over 17 years at a major referral center.

**Henoch-Schonlein Purpura**

Pediatricians are familiar with this common condition of childhood, which usually presents with a typical rash over the lower extremities, abdominal pain, and joint symptoms, all of which resolve spontaneously or in response to corticosteroids if necessary. Just occasionally the disease is more serious with renal involvement, and on very rare occasions diffuse pulmonary hemorrhage may occur. Sporadic case reports of HSP with pulmonary hemorrhage in children have appeared in the literature, and although these children were very seriously ill, successful outcomes were reported with treatment by corticosteroids or cytotoxics.\(^5,16\)

**Idiopathic Pulmonary Hemosiderosis**

Idiopathic pulmonary hemosiderosis (IPH) is a rare condition which is distinct from the pulmonary-renal syndromes in that the pathology is confined to the lungs and there is no renal or systemic involvement. Over a 17-year period at the Children’s Hospital of Los Angeles, 17 children with IPH were diagnosed,\(^17\) making approximately one case of IPH per year in a major referral center. IPH in childhood used to be considered to have a very poor prognosis, but there is evidence that the prognosis has been improved by more effective treatment which emphasizes the importance of the correct diagnosis and management. Whereas some 60% or more children with IPH died in earlier published series, the 5-year projected survival in the study of Saeed et al.\(^{17}\) in 1999 was 86%. We reviewed reports of IPH in some 75 children in a survey of the literature from 1957–2000.\(^18\) Subsequently, Le Calinche et al.\(^{19}\) reported a further 15 children followed up for at least 10 years, although 3 of these also appeared to develop systemic disease. But nothing is ever simple in medicine, especially in these rare pulmonary hemorrhagic diseases of childhood, because there have also been reports of what appears to be an epidemic of an unusual form of IPH which affected 30 very young infants over a 7-year period treated at the Rainbow Babies and Children’s Hospital in Cleveland. These infants will be considered separately from the 90 children with “typical” IPH.

**Clinical Presentation**

Taking all 90 children with typical IPH together, almost 70% presented with respiratory symptoms under 6 years of age (Table 3). The clinical picture is of recurrent hemoptysis which can vary from life-threatening episodes...
requiring transfusion to intermittent blood-streaked sputum, and varying degrees of anemia depending on the severity of the hemoptysis. In severe attacks, there is hypoxia and dyspnea, and the child may suffer from chronic fatigue. Between attacks the child is well, apart from the effects of any residual anemia; recovery from an episode of bleeding may be surprisingly rapid and complete. The radiological findings in diffuse alveolar hemorrhage are usually symmetric and predominate in the perihilar regions and lower lung zones, with sparing of the apices and costophrenic angles. Radionuclide imaging can be helpful in detecting active pulmonary bleeding, using technetium 99m-labeled red blood cells or technetium 99m-labeled sulfur colloid. Increased activity can be found within the lung, corresponding to active bleeding, although this is rarely needed to make the diagnosis. Hematologic investigation will usually show microcytic, hypochromic anemia, unless the bleeding is recent. Reticulocytosis may be present, depending on available iron stores. In children old enough to perform tests of lung function, the typical picture is of restrictive lung disease with reduced forced vital capacity (FVC), reduced forced expiratory volume in 1 sec (FEV₁), and a normal FEV₁/FVC ratio. If there is enough blood in the lungs, there will be an elevation of single-breath carbon monoxide (CO) uptake as a result of the binding of CO to hemoglobin in intra-alveolar blood. Investigations to exclude other causes of pulmonary hemorrhage, such as the pulmonary-renal syndromes, or cardiac disease will be negative in IPH.

Heiner’s Syndrome

In 1962, Heiner et al.²⁰ published a study of 7 infants in whom they believed that pulmonary hemosiderosis was due to cow’s milk allergy, since all had precipitins to cow’s milk in their serum. Careful reading of their classic paper shows that these 7 infants with high levels of precipitins were selected by screening 2,200 patients with respiratory symptoms. The symptoms in these children were not those normally expected in IPH, and all had nonspecific complaints of chronic lung and upper respiratory tract disease. However, 4 infants had hemoptysis, 5 had anemia, and iron-laden macrophages in “gastric or bronchial aspirates” were found in 4. The children apparently improved on a milk-free diet, although it seems that 6 subsequently became tolerant of milk. In their discussion, Heiner et al.²⁰ noted that they also studied 7 other patients with hemosiderosis who did not have precipitins, and in an addendum, they added that they found another 26 patients with precipitins comparable to those in the paper by screening serum from 1,284 patients with similar symptoms. Of these 26, 8 had respiratory or gastrointestinal symptoms relieved by a milk-free diet, but no details were given. Some 13 years later, Boat et al.²¹ reported another 6 infants with a similar clinical picture and high titers of milk precipitins identified by screening the sera of 160 children with idiopathic chronic lung disease. All 6 children had typical manifestations of “milk-induced pulmonary hemosiderosis” (their term) with recurrent pulmonary infiltrates, hemosiderin-laden pulmonary macrophages in 5, intermittent wheezing in 5, eosinophilia in 4, anemia in 4, and failure to thrive in 4. Three children went on to develop obvious upper airway obstruction and cor pulmonale. Elimination of cow’s milk from the diet, and adenoidectomy when indicated, resulted in improvement in all 6 patients. However, in a subsequent study these investigators could find no difference in the quantity or type of milk precipitins in 4 children with presumed pulmonary hemosiderosis compared with other children with chronic lung diseases, gastrointestinal disease, and upper airway disease.²² Since the 1970s, enthusiasm for cow’s milk allergy as the cause of IPH has waned somewhat, and in the very large majority of the 90 children with IPH collected from the literature and summarized in Table 3, no such association was present. To be fair to the original investigators, they also voiced doubts about the association and pointed out that some of their patients with hemosiderosis did not have milk precipitins, while many children with milk precipitins did not have hemosiderosis.

**Bronchoalveolar Lavage**

The most useful investigation in IPH consists of bronchoscopy and bronchoalveolar lavage (BAL). Bronchoscopy is the most direct way to evaluate hemoptysis and determine the site of bleeding, if it is from a localized source visible by bronchoscopy and the bleeding is currently active. However, even in the absence of gross bleeding, the demonstration of hemosiderin-laden macrophages in BAL is good evidence that bleeding has occurred recently and that the disease process is still active (see below). The finding of hemosiderin in macrophages from gastric aspirates is also suggestive of the
diagnosis but is less direct and arguably more traumatic for the child than BAL.

**Pathology**

Lung biopsy is rarely, if ever, justified to make the diagnosis of IPH, as it can be made on the basis of the clinical picture and the exclusion of other causes of pulmonary hemorrhage. Occasionally, lung biopsy will be necessary when it is impossible to exclude systemic diseases. The lung histology in IPH shows many red blood cells, seen in alveoli and the interstitium during active bleeding, and there is an absence of any evidence of vasculitis. After a few days, macrophages in both areas contain hemosiderin.

**Treatment and Prognosis**

In our review of the literature from 1957–2000, we found 7 papers relating to IPH containing enough information to be suitable for analysis, but in only 3 of these were there more than 1–2 children, and of the total of 75 children with IPH, data on follow-up were available in only 68. To this can now be added the 15 children from the series of Le Clainche et al., for whom data are available. However, this series only concerns children known to have survived at least 10 years, and there are no data on those who may have died during this time. The results for these 90 children, for whom follow-up data were available in 83, are summarized in Tables 3 and 4. Of the 83 children with IPH, some 72% were alive at time of follow-up, and for those for whom data were available, 59% were alive 2 years after diagnosis. For those alive at time of follow-up and for whom information on treatment was available (58 children), 65.5% were still on treatment. The treatments used and their efficacy are summarized in Table 4. In 92% of children, the initial treatment was with prednisone, pulse methyl prednisolone, or their equivalents. A smaller number were treated initially with hydroxychloroquine, cytotoxics, or some combination of prednisone and these other agents. The initial treatment was regarded as successful in 61% of cases, and when this was ineffective, a change in treatment or the addition of a second agent was successful in some 85% of the previous treatment failures. In some cases, treatment was regarded as successful even if the child had a residual pulmonary handicap or if the bleeding returned when treatment was stopped, and as already noted, some 65.5% still needed treatment at time of follow-up. In the study of Le Clainche et al., 5 of 13 (38%) children about whom there was information were still on treatment at least 10 years after diagnosis. Because of the reported association between cow’s milk allergy and hemosiderosis, many children with IPH are placed on milk-free diets, but unless there is unequivocal evidence of true milk allergy, this is quite unjustified.

In those who recover from IPH, lung function can apparently be completely normal if no further bleeding occurs. Some care is required in accepting the apparently good prognosis in some reports, since this often refers to survival and not to functional status. Thus in their study of IPH, Hacking et al. concluded that “the good prognosis seen in these patients is different to previous case reports indicating a greater than 50% mortality.” In their study of 11 children none died, but only 2 were reported well at follow-up, 7 were exercise-limited, and 2 were oxygen-dependent: hardly a great functional success. However, their patients had only ever been treated with prednisolone, and 5 of those who were exercise-limited or oxygen-dependent only received it for 3 months. In the series of Le Clainche et al., 4 of 15 patients (27%) were apparently completely well, although one was still on treatment and 8 others had a few symptoms even though their chest radiograph or lung function was abnormal. One of our own children with IPH presented at age 13 years, had active bleeding treated with hydroxychloroquine and courses of prednisone for the next 3 years, and was recruited into the army at age 18 years with a normal chest radiograph, totally normal lung function, and no treatment.

**The Cleveland Epidemic of IPH**

The infants with pulmonary hemorrhage followed by Dearborn et al. comprised 30 seen between 1993–2000 who were less than 1 year of age. They mostly presented with severe diffuse alveolar hemorrhage, and all known causes of pulmonary hemorrhage were excluded by investigation. Some 75% required transfusions and ventilatory support and 5 died, but the bleeding lessened or stopped in the others, and most have been able to stop treatment with corticosteroids. The investigators felt strongly that the condition could be due to exposure to a toxogenic fungus such as *Stachybotrys chartarum*, which

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**TABLE 4— Treatments and Efficacy in 83 Children With IPH From a Review of Literature**

<table>
<thead>
<tr>
<th>Initial treatment</th>
<th>% total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prednisone/PMP</td>
<td>72</td>
</tr>
<tr>
<td>Hydroxychloroquine</td>
<td>1</td>
</tr>
<tr>
<td>All combinations</td>
<td>4</td>
</tr>
<tr>
<td>None</td>
<td>6</td>
</tr>
<tr>
<td>All initial treatments</td>
<td>% treated</td>
</tr>
<tr>
<td>Success</td>
<td>47</td>
</tr>
<tr>
<td>Failure</td>
<td>31</td>
</tr>
<tr>
<td>All subsequent treatments</td>
<td>% treated</td>
</tr>
<tr>
<td>Success</td>
<td>11</td>
</tr>
<tr>
<td>Failure</td>
<td>2</td>
</tr>
</tbody>
</table>

1PMP, pulse methyl prednisolone.
was present in some 90% of the homes of the infants.31

There were, however, other features, apart from the very young age and severe course in some of these infants, which are not seen in typical IPH, such as developmental delay, seizures, and hemoglobinuria. This suggests that they should be considered as a separate group from typical IPH until the picture becomes clearer.

Factitious Hemoptysis

Last but not least, things are not always what they seem, especially with emotionally labile adolescent children. There have been a number of reports over the years of adults with a form of Munchausen’s syndrome which presented as factitious hemoptysis, but it has now appeared in children. Bjornson and Kirk32 described a 12-year-old girl who presented initially with a history of recurrent hematemesis and later with hemoptysis for which she underwent numerous investigations. Ultimately, the cause of bleeding was found to be biting of her lip and poking her finger into her nostril. A remarkably similar case was recently seen by a colleague (Dr. H. Blau, personal communication). This concerned a 14-year-old emotionally unstable girl with a 1-week history of coughing up blood, about half a cup, twice a day. Her chest radiograph and lung function were normal, and a sample of “hemoptysis” consisted of blood-stained saliva. Appropriate further enquiries revealed that the girl made repeated small cuts in her tongue with a razor blade, squeezed out the blood between her teeth, collected it with saliva, and then “coughed” it out. Why? She was very impressed with a film in which the heroine died of tuberculosis.

An Approach to the Child with Hemoptysis

Remembering that hemoptysis and pulmonary hemorrhage, which are uncommon in children, may be due to causes which are basically either very common or very rare, a balanced approach to the diagnostic evaluation of the problem is required. An approach to the child who presents with suspected hemoptysis or pulmonary hemorrhage is summarized in Figure 1. As in all medicine, this should be based on taking a careful history, examination, and appropriate special investigations. Unfortunately, neither the history nor physical examination is likely to help with a definitive diagnosis of the nature of the problem except when the association is very obvious, such as with advanced cystic fibrosis or in a child known to suffer from a systemic disease associated with pulmonary hemorrhage. In almost all other situations, investigations will be needed. A plain chest radiograph is mandatory as this may well indicate the site and extent of the bleeding, but a normal radiograph is no guarantee that there has been no pulmonary hemorrhage or that any bleeding was of extrapulmonary origin. Computerized tomography with contrast may be needed to define cavitary lesions or the very rare pulmonary arterio-venous malformations which can cause hemoptysis.

The definitive investigation is bronchoscopy, which should include careful inspection of the upper airway and bronchoalveolar lavage (BAL) where appropriate. During active bleeding, the site or at least the approximate location of the bleeding can be determined, and pediatric pulmonologists have even been known to pass the bronchoscope into the esophagus to look for bleeding from the upper gastrointestinal tract. When there is no active bleeding and no obvious cause such as a foreign body, BAL should be performed to look for hemosiderin-laden macrophages (HLMs) as an indication of previous pulmonary hemorrhage. While this test is widely recognized and recommended for use in the evaluation of suspected pulmonary hemorrhage, there is surprisingly little information on when the test is likely to be positive and how long it is likely to remain positive. Sherman et al.33 described 4 infants aged 4 weeks to 4 months studied at various times after acute pulmonary hemorrhages (interestingly, very similar to those later reported from Cleveland by Dearborn et al.30). These data suggest that HLMs were not found until some 50 hr after an acute bleed, and were still present 5 days after the bleed, but not at autopsy in one infant 12 days after the bleed. The number of infants was too small to make any really valid conclusions, and there do not appear to be any better data in the adult literature. The most useful study to date was undertaken experimentally in mice by Epstein et al.,34 who clearly showed that HLMs began to appear on day 3 (2.8% total cell count) after the acute introduction of blood into the lungs, peaked (60%) at day 6, remained high until day

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**Fig. 1. Schematic representation of investigation of a child who presents with probable hemoptysis or pulmonary hemosiderosis.**

- CF, cystic fibrosis; BAL, bronchoalveolar lavage; HLM, hemosiderin-laden macrophages; IPH, idiopathic pulmonary hemosiderosis.
If the plain chest radiograph, bronchoscopy, and BAL (performed between 3 to about 14 days after the suspected bleed) are all normal, it is reasonable to wait to see if there are any further episodes of suspected bleeding, which can be investigated similarly.

If there is evidence of bleeding into the lungs, not due to a localized lesion in the airway, and the child does not have CF or bronchiectasis from another cause, a cardioligic evaluation including echocardiography should be performed. In the absence of significant cardiac pathology, investigations for possible pulmonary-renal syndromes should be performed. These may of course be performed much earlier if the child is known to have a systemic disease or if there are suggestive findings such as a rash, blood, or cellular casts in the urine. In most cases, the investigations listed in Table 2 will be needed. If there is no evidence of systemic disease, it is likely that the cause of the pulmonary hemorrhage is IPH, which is essentially a diagnosis by exclusion. Open lung biopsy is rarely needed to arrive at the most likely diagnosis, which can almost always be reached by less invasive procedures.

CONCLUSIONS

Hemoptysis and pulmonary hemosiderosis in children are very rare but may indicate the presence of severe disease which can be fatal if not managed appropriately. If the more common causes such as cystic fibrosis, localized airway lesions, and cardiac diseases are excluded, then the cause of bleeding may be one of the very rare pulmonary-renal syndromes or idiopathic pulmonary hemosiderosis.

REFERENCES


