



Complete Summary

GUIDELINE TITLE

Acute idiopathic pulmonary hemorrhage among infants. Recommendations from the Working Group for Investigation and Surveillance.

BIBLIOGRAPHIC SOURCE(S)

Brown CM, Redd SC, Damon SA. Acute idiopathic pulmonary hemorrhage among infants. Recommendations from the Working Group for Investigation and Surveillance. MMWR Recomm Rep 2004 Mar 12;53(RR-2):1-12. [10 references]
[PubMed](#)

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Acute idiopathic pulmonary hemorrhage (AIPH)

GUIDELINE CATEGORY

Diagnosis
Evaluation
Prevention
Risk Assessment

CLINICAL SPECIALTY

Family Practice
Internal Medicine
Pediatrics
Preventive Medicine
Pulmonary Medicine

INTENDED USERS

Health Care Providers
Physicians
Public Health Departments

GUIDELINE OBJECTIVE(S)

- To present the Centers for Disease Control and Prevention's (CDC's) recommended case definitions and surveillance practices for acute idiopathic pulmonary hemorrhage (AIPH) among infants
- To describe how results from surveillance activities can guide efforts to investigate the burden and etiology of AIPH among infants

TARGET POPULATION

Infants

INTERVENTIONS AND PRACTICES CONSIDERED

1. Identification or diagnosis of acute idiopathic pulmonary hemorrhage (AIPH) based on multiple established criteria, such as signs, symptoms, hospitalization status, need for intubation and mechanical ventilation, and results of chest x-ray (CXR) or computed tomography (CT)
2. Classification of AIPH (clinically confirmed, probable or suspected)
3. Investigation of suspected clusters of AIPH among infants by the state health departments in coordination with the Centers for Disease Control and Prevention (CDC)
 - Collection and reporting of clinical information by pediatric intensive care unit (PICU) and neonatal intensive care unit (NICU) staff
 - Home assessment to gather pertinent risk-assessment data
4. Retrospective analysis by CDC of state-level mortality and hospitalization data based on International Classification of Diseases (ICD) codes
5. Case surveillance as indicated

MAJOR OUTCOMES CONSIDERED

- Numbers of infants with pulmonary hemorrhage and/or acute idiopathic pulmonary hemorrhage
- Positive predictive value (PPV) of International Classification of Diseases, Ninth Revision (ICD-9) codes

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases
Searches of Unpublished Data

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Not stated

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Three meetings of panelists were convened to advise The Centers for Disease Control and prevention (CDC) staff regarding investigation of acute idiopathic pulmonary hemorrhage (AIPH) among infants. The Case Definition Panel included three pediatric pulmonologists, one pediatric intensive care specialist, one pediatric pathologist, two epidemiologists, and one environmental epidemiologist. The purpose of this panel was to recommend a case definition for use in public health surveillance for AIPH to facilitate case finding. Case finding will facilitate documentation of the burden of the condition and identification of possible etiologic agents or risk factors.

The Surveillance Implementation Panel included one pediatrician, one pediatric pulmonologist, one forensic pathologist, three epidemiologists (including one state epidemiologist), and four environmental epidemiologists (including one state

epidemiologist and one toxicologist). Its purpose was to recommend a standard approach for public health surveillance for AIPH.

The Home/Indoor Environment and Laboratory Investigation Panel included two mycologists, one biochemist, two microbiologists, two industrial hygienists, two toxicologists, and one environmental epidemiologist. Its purpose was to recommend standard approaches and protocols for environmental data collection, laboratory analysis, and data interpretation during public health surveillance for AIPH.

For each of the three areas, group discussion led by a moderator was based on prepared questions. Participants produced written summaries, which form the basis of the recommendations provided in this report.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not stated

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Case Definition

Case Classification and Severity Criteria

Acute idiopathic pulmonary hemorrhage (AIPH) is the sudden onset of pulmonary hemorrhage in a previously healthy infant in whom differential diagnoses and neonatal medical problems that might cause pulmonary hemorrhage have been ruled out. 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, severe respiratory distress or failure, requiring mechanical ventilation and chest radiograph (CXR), and usually demonstrate bilateral infiltrates.

AIPH among infants and sudden infant death syndrome (SIDS) potentially share similar risk factors (e.g., age group and maternal cigarette smoking). Also, in

certain cases, SIDS is associated with pulmonary hemorrhage found at autopsy. Thus, factors that are known risk factors for SIDS should be identified when evaluating an infant possibly having AIPH. Potential information sources for case-identification and case-status classification during an investigation of pulmonary hemorrhage are provided in Table 1 of the original guideline document.

Clinically Confirmed Cases of AIPH Among Infants

Criteria for a confirmed case include pulmonary hemorrhage in a previously healthy infant aged ≤ 1 year with a gestational age of ≥ 32 weeks, with no history of neonatal medical problems that might cause pulmonary hemorrhage, and whose condition meets all of the following three criteria:

- Abrupt or sudden onset of overt bleeding or obvious evidence of blood in the airway, including
 - epistaxis, hemoptysis, or frank blood in the airway below the larynx at visualization, not caused by any medical procedure (e.g., laryngoscopy or intubation);

or

- identification of hemosiderin-laden macrophages ($>20\%$ of pulmonary macrophages containing hemosiderin on bronchoalveolar lavage or biopsy specimen). A source of bleeding from the nose and oropharynx should be ruled out at the time of admission.
- Severe-appearing illness leading to acute respiratory distress or respiratory failure, resulting in hospitalization in a pediatric intensive care unit (PICU) or neonatal intensive care unit (NICU) with intubation and mechanical ventilation.
- Diffuse unilateral or bilateral pulmonary infiltrates visible on CXR or computerized tomography (CT) of the chest. CXR or chest CT findings should be documented within 48 hours of examination of the infant.

A previously healthy infant should

- have been discharged from the hospital after birth with an uneventful course before the occurrence of bronchoalveolar hemorrhage;
- never have been previously intubated, nor required respiratory support with oxygen;
- not have evidence of physical abuse;
- not have any abnormality identified on admission or follow-up bronchoscopy that would explain the bleeding; and
- not have neonatal medical problems that can cause pulmonary hemorrhage.

The Centers for Disease Control and Prevention (CDC) will adhere closely to this case definition, requiring that all the criteria be met for a confirmed case. The definition for a clinically confirmed case excludes pulmonary hemorrhage among older children and infants with restricted access to a PICU. Because no criteria exist for postmortem examinations, this definition excludes infants who die before hospital and PICU admission, whose illness might have met the case definition. However, the definitions for probable and suspect cases (see the following) will

capture the majority of these cases and allow identification of illness among infants who die before examination by a physician.

Probable Cases of AIPH Among Infants

Criteria for a probable case include a previously healthy infant aged ≤ 1 year with a gestational age of ≥ 32 weeks,

- who has a sudden onset of bleeding from the airway, with or without respiratory distress, with or without intubation, and with or without pulmonary infiltrates on CXR or chest CT;
- or
- who died and had evidence of bleeding from the airway found on autopsy or postmortem; had been in respiratory distress; would or should have been intubated in the opinion of a clinician; and would have had infiltrates on CXR or chest CT.

Suspected Cases of AIPH Among Infants

Criteria for a suspected case include a previously healthy infant,

- who died and had evidence of bleeding from the airway found on autopsy or postmortem;
- or
- who either did not have chest imaging studies or had imaging studies that indicated no pulmonary infiltrates.

Respiratory distress or intubation is not required for a suspected case.

Severity Classification Scheme for AIPH Among Infants

Because of the potential for variation in symptoms among infants for each of the criteria, different case combinations might be related to the timing or duration of symptoms, disease severity, pathologic processes, or etiologic agents associated with AIPH among infants. A discussion of the proposed case-classification categories for AIPH among infants is provided in Tables 2 and 3 in the original guideline document.

A summary of clinical features of AIPH among infants and neonatal medical problems and differential diagnoses that should be ruled out before classifying a case as AIPH among infants are included in the original guideline document. Other differential diagnoses associated with pulmonary hemorrhage are also listed in the original guideline document.

Feasibility Study To Determine the Concordance of International Classification of Disease (ICD) Codes for Pulmonary Hemorrhage with the CDC Case Definition

CDC will retrospectively review cases of pulmonary hemorrhage to determine the public health impact of AIPH among infants and to generate hypotheses regarding the importance of risk factors possibly associated with AIPH among infants. If that review indicates that AIPH among infants is a separate clinical entity and that these cases have occurred in clusters, or that an increase in incidence or mortality is associated with these cases, CDC will initiate prospective surveillance and case ascertainment to identify cases for epidemiologic studies designed to confirm or disprove associations between pulmonary hemorrhage host factors, environmental factors, and biologic agents, including such molds as *Stachybotrys chartarum*.

Retrospective Review by Using Existing Data Sources

Refer to the original guideline document for a discussion of retrospective reviews that have been performed by CDC, possible benefits of retrospective review of AIPH, and the CDC's plan for retrospective review.

Investigation of Suspected Clusters of AIPH Among Infants

If an apparent cluster of cases of pulmonary hemorrhage occurs, CDC recommends that state health departments initiate an investigation. CDC staff will work with each state, upon request, to evaluate case reports to assist the epidemiologic and environmental investigation, if any. State health departments can use existing protocols for outbreak or cluster investigations and collect information to determine if cases meet the CDC case definition for AIPH among infants. Because these cases probably will be identified in pediatric intensive care units (PICUs), CDC recommends that if the PICU staff identifies any suspected cases, they report them to their state epidemiologist.

For each case of AIPH in an infant, CDC recommends that PICU and NICU staff collect clinical information to certify case status, demographic information, and reports regarding the status of the patient's home. PICU and NICU staff also should carefully document illnesses that are similar clinically to AIPH, even if another specific etiology is confirmed, because they might offer additional information or indicate the need to re-assess the case definition.

If performed, environmental assessment of the home to gather pertinent risk-assessment data should use standard protocols designed by trained environmental health professionals. At a minimum, the assessment should involve visual inspection, including checks for dampness, water damage, obvious mold, evidence of pests, and environmental tobacco smoke. Depending on the assessed need for further evaluation and the resources available, additional investigation might include determining moisture content, settled dust sampling, air sampling for different allergens and biologically active compounds, and other investigations as needed.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Case findings of acute idiopathic pulmonary hemorrhage (AIPH) will facilitate documentation of the burden of the condition and identification of possible etiologic agents or risk factors.

POTENTIAL HARMS

Not stated

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

The Centers for Disease Control and Prevention (CDC) will retrospectively analyze state-level mortality and hospitalization data based on International Classification of Disease (ICD) codes and will retrospectively review discharges for pulmonary hemorrhage in selected pediatric intensive care units (PICUs). These studies will

- distinguish between the clinical findings associated with different symptoms of acute idiopathic pulmonary hemorrhage (AIPH) among infants;
- determine whether ICD codes capture cases that meet the CDC-recommended case definition for AIPH among infants;
- determine whether AIPH among infants is a distinct recognizable clinical entity;
- determine the proportion of cases ascertained retrospectively through ICD-9 codes that meet the clinical case definition by estimating the positive predictive value (PPV) of ICD-coded data; and
- define the magnitude of AIPH among infants and the need for conducting etiologic studies.

CDC will review the Cleveland and Chicago case series to determine the degree to which the present case definition applies to them. In addition, CDC will evaluate the present case definition on the basis of data from initial surveillance findings and modify it as appropriate.

If these reviews establish that AIPH among infants is a public health problem on the basis of increasing numbers or clusters of cases geographically or temporally, targeted prospective case surveillance will be initiated. If prospective surveillance is initiated, CDC will maintain a database of current cases of AIPH among infants, reported by PICUs that meet the case definition. The database will serve as a

source of cases for case-control studies to determine etiology. CDC will work with state and local health departments to investigate clusters of AIPH among infants' cases.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Staying Healthy

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Brown CM, Redd SC, Damon SA. Acute idiopathic pulmonary hemorrhage among infants. Recommendations from the Working Group for Investigation and Surveillance. MMWR Recomm Rep 2004 Mar 12;53(RR-2):1-12. [10 references]
[PubMed](#)

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2004 Mar 12

GUIDELINE DEVELOPER(S)

Centers for Disease Control and Prevention - Federal Government Agency [U.S.]

SOURCE(S) OF FUNDING

United States Government

GUIDELINE COMMITTEE

Working Group for Investigation and Surveillance

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available from the Centers for Disease Control and Prevention (CDC) Web site:

- [HTML Format](#)
- [Portable Document Format \(PDF\)](#)

Print copies: Available from the Centers for Disease Control and Prevention, MMWR, Atlanta, GA 30333. Additional copies can be purchased from the Superintendent of Documents, U.S. Government Printing Office, Washington, DC 20402-9325; (202) 783-3238.

AVAILABILITY OF COMPANION DOCUMENTS

None available

PATIENT RESOURCES

None available

NGC STATUS

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