Complete Summary

GUIDELINE TITLE

Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines.

BIBLIOGRAPHIC SOURCE(S)

McGoon M, Gutterman D, Steen V, Barst R, McCrory DC, Fortin TA, Loyd JE. Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Chest 2004 Jul;126(1 Suppl):14S-34S. [170 references] PubMed

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EVIDENCE SUPPORTING THE RECOMMENDATIONS
BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS
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SCOPE

DISEASE/CONDITION(S)

Pulmonary arterial hypertension

GUIDELINE CATEGORY

Diagnosis
Evaluation
Screening

CLINICAL SPECIALTY

Cardiology
Internal Medicine
Medical Genetics
Pulmonary Medicine
Rheumatology

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

- To review evidence for screening in susceptible patient groups and the approach to diagnosing pulmonary arterial hypertension when it is suspected
- To provide specific recommendations for applying this evidence to clinical practice

TARGET POPULATION

Patients with suspected pulmonary arterial hypertension and asymptomatic individuals at risk for pulmonary arterial hypertension

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation/Screening

1. Genetic testing and professional genetic counseling
2. Electrocardiogram
3. Chest radiography
4. Doppler echocardiography
5. Testing for connective tissue disease and human immunodeficiency virus (HIV) infection
6. Ventilation/perfusion (V/Q) scanning
7. Contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI)
8. Pulmonary angiography
9. Testing of pulmonary function and arterial blood oxygenation
10. Testing of pulmonary function testing with diffusing capacity of the lung for carbon monoxide
11. Lung biopsy (not recommended routinely because of risks)
12. Right-heart catheterization
13. 6-min walk test

MAJOR OUTCOMES CONSIDERED

Sensitivity, specificity, and predictive value of screening and diagnostic tests for pulmonary arterial hypertension

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

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

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Note from National Guideline Clearinghouse (NGC): The Center for Clinical Health Policy Research at Duke University identified and evaluated evidence on this topic, working with the guideline development panel to formulate key questions suitable for systematic literature synthesis.

Search Strategy

Computerized searches of the MEDLINE bibliographic database from 1992 to October 2002 were conducted. The developer searched using the term hypertension, pulmonary. The search was limited to articles concerning human subjects that were published in the English language and accompanied by an abstract. In addition, the developer searched the reference lists of included studies, practice guidelines, systematic reviews, and meta-analyses, and consulted with clinical experts to identify relevant studies missed by the search strategy or published before 1992.

Study Selection

Two physicians (one with methodologic expertise and one with content area expertise) reviewed the abstracts of candidate articles and selected a subset for review in full text. Full-text articles were again reviewed by two physicians to determine whether they were study reports or review articles and were pertinent to at least one of the key questions. The selection criteria differed for each topic.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Expert Consensus
Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Quality of the evidence is rated as follows

Good = evidence based on good randomized controlled trials or meta-analyses

Fair = evidence based on other controlled trials or randomized controlled trials with minor flaws

Low = evidence based on nonrandomized, case-control, or other observational studies

Expert opinion = evidence based on the consensus of the carefully selected panel of experts in the topic field. There are no studies that meet the criteria for inclusion in the literature review.

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

A comprehensive review of published studies was performed to provide an evidence-based analysis, including an assessment of the sensitivity and specificity of the methods used clinically to detect and diagnose pulmonary arterial hypertension. Each of the diagnostic methods and strategies were examined, including those that are utilized for the confirmation of conditions associated with pulmonary arterial hypertension. Novel diagnostic techniques and future directions for the field were then considered. The summary evidence tables can be viewed on-line at http://www.chestjournal.org/content/vol126/1_suppl/.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Informal Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

An international panel of 19 experts representing five medical specialties was assembled. Representatives from other medical and patient advocacy associations were also invited to join the panel (including the American College of Cardiology, American College of Rheumatology, and the Pulmonary Hypertension Association). These experts convened on several occasions, including the culminating panel conference in September 2003, in which they deliberated over the composition of the final recommendations and grading of the current state of the evidence, benefits to the patient, and the strength of the recommendations.

Guideline development was led by an executive committee including the chair, the leader of the methodology support group, and the American College of Chest Physicians project manager, which supervised the guideline development process, methodologic issues, panel composition, structure of the final document, and activities of the writing committees. Each writing committee, led by a group leader who served as primary author and editor of that chapter, conferred with the methodology team on inclusion/exclusion criteria, relevant research questions, and important literature that was not readily identified. These individuals continue with their responsibilities to assist in the development of the implementation tools.

When the evidence was insufficient for evidence-based recommendations, the panel used informal group consensus techniques to develop recommendations based on the expert opinion of the panel. With every member of the panel attending the final conference, the expert-based opinions are truly representative of geographically diverse and multispecialty inclusive practice patterns of the complete panel.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Strength of Recommendations

A = strong recommendation
B = moderate recommendation
C = weak recommendation
D = negative recommendation
I = no recommendation possible (inconclusive)
E/A = strong recommendation based on expert opinion only
E/B = moderate recommendation based on expert opinion only
E/C = weak recommendation based on expert opinion only
E/D = negative recommendation based on expert opinion only

Net Benefit is Defined as Follows

Substantial
Intermediate
Small/weak
None
Conflicting
Negative

COST ANALYSIS

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

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The writing groups and the executive committee of the panel extensively reviewed each chapter during the writing process. The final conference provided an opportunity for the entire panel to review the latest drafts. Following final revisions and one final review by the executive committee, each chapter of the guidelines was reviewed and approved by the American College of Chest Physicians (ACCP) Health and Science Policy Committee, the ACCP Pulmonary Vascular NetWork, and then by the ACCP Board of Regents. The guidelines have not been field tested.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Rating schemes for level of evidence, strength of recommendation, and net benefit follow the major recommendations.

1. Genetic testing and professional genetic counseling should be offered to relatives of patients with familial pulmonary arterial hypertension (FPAH). Level of evidence: expert opinion; benefit: intermediate; grade of recommendation: E/A.
2. Patients with idiopathic pulmonary arterial hypertension (IPAH) should be advised about the availability of genetic testing and counseling for their relatives. Level of evidence: expert opinion; benefit: intermediate; grade of recommendation: E/A.
3. In patients with a suspicion of pulmonary arterial hypertension (PAH), electrocardiogram (ECG) should be performed to screen for a spectrum of cardiac anatomic and arrhythmic problems; it lacks sufficient sensitivity to serve as an effective screening tool for PAH, but contributes prognostic information in patients with known PAH. Quality of evidence: low; benefit: small/weak; strength of recommendation: C.
4. In patients with a suspicion of PAH, a chest radiograph (CXR) should be obtained to reveal features supportive of a diagnosis of PAH and to lead to diagnoses of underlying diseases. Quality of evidence: low; benefit: intermediate; strength of recommendation: C.
5. In patients with a clinical suspicion of PAH, Doppler echocardiography should be performed as a noninvasive screening test that can detect pulmonary hypertension (PH), though it may be imprecise in determining actual pressures compared to invasive evaluation in a portion of patients. Quality of evidence: fair; benefit: substantial; strength of recommendation: A.
6. In patients with a clinical suspicion of PAH, Doppler echocardiography should be performed to evaluate the level of right ventricular systolic pressure and to assess the presence of associated anatomic abnormalities such as right atrial enlargement, right ventricular enlargement, and pericardial effusion. Quality of evidence: expert opinion; benefit: intermediate; strength of recommendation: E/B.
7. In asymptomatic patients at high risk, Doppler echocardiography should be performed to detect elevated pulmonary arterial pressure. Quality of evidence: expert opinion; benefit: intermediate; strength of recommendation: E/B.
8. In patients with suspected or documented pulmonary hypertension (PH), Doppler echocardiography should be performed to look for left ventricular systolic and diastolic dysfunction, left-sided chamber enlargement, or valvular heart disease. Quality of evidence: good; benefit: substantial; strength of recommendation: A.
9. In patients with suspected or documented PH, Doppler echocardiography with contrast should be obtained to look for evidence of intracardiac shunting. Quality of evidence: fair; benefit: intermediate; strength of recommendation: B.
10. In patients with unexplained PAH, testing for connective tissue disease and human immunodeficiency virus (HIV) infection should be performed. Quality of evidence: expert opinion; benefit: intermediate; strength of recommendation: E/A.
11. In patients with PAH, ventilation/perfusion (V/Q) scanning should be performed to rule out chronic thromboembolic pulmonary hypertension (CTEPH); a normal scan result effectively excludes a diagnosis of CTEPH. Quality of evidence: low; benefit: substantial; strength of recommendation: B.
12. In patients with PAH, contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) should not be used to exclude the

diagnosis of CTEPH. Quality of evidence: low; benefit: negative; strength of recommendation: D.

13. In patients with PAH and a ventilation/perfusion (V/Q) scan suggestive of CTEPH, pulmonary angiography is required for accurate diagnosis and best anatomic definition to assess operability. Quality of evidence: expert opinion; benefit: substantial; strength of recommendation: E/A.
14. In patients with PAH, testing of pulmonary function and arterial blood oxygenation should be performed to evaluate for the presence of lung disease. Quality of evidence: low; benefit: substantial; strength of recommendation: B.
15. In patients with systemic sclerosis, pulmonary function testing with diffusing capacity of the lung for carbon monoxide (DLCO) should be performed periodically (every 6 to 12 months) to improve detection of pulmonary vascular or interstitial disease. Quality of evidence: fair; benefit: intermediate; strength of recommendation: B.
16. In patients with PAH, lung biopsy is not routinely recommended because of the risk, except under circumstances in which a specific question can only be answered by tissue examination. Quality of evidence: expert opinion; benefit: substantial; strength of recommendation: E/A.
17. In patients with suspected PH, right-heart catheterization is required to confirm the presence of PH, establish the specific diagnosis, and determine the severity of PH. Quality of evidence: good; benefit: substantial; strength of recommendation: A.
18. In patients with suspected PH, right-heart catheterization is required to guide therapy. Quality of evidence: low; benefit: substantial; strength of recommendation: B.
19. In patients with PAH, serial determinations of functional class and exercise capacity assessed by the 6-minute walk test provide benchmarks for disease severity, response to therapy, and progression. Quality of evidence: good; benefit: intermediate; strength of recommendation: A.

Definitions

Quality of the Evidence

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Net Benefit

Substantial
Intermediate
Small/weak
None
Conflicting
Negative

CLINICAL ALGORITHM(S)

A clinical algorithm is provided in the original guideline document for patient evaluation.

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate recommendations for screening and diagnosis of susceptible patient groups for pulmonary arterial hypertension.

POTENTIAL HARMS

None stated

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- The information provided in the guideline should be used in conjunction with clinical judgment. Although the guideline provides recommendations that are based on evidence from studies involving various populations, the recommendations may not apply to every individual patient. It is important for the physician to take into consideration the role of patient preferences and the availability of local resources.

- The American College of Chest Physicians (ACCP) is sensitive to concerns that nationally and/or internationally developed guidelines are not always applicable in local settings. Further, guideline recommendations are just that, recommendations not dictates. In treating patients, individual circumstances, preferences, and resources do play a role in the course of treatment at every decision level. Although the science behind evidence-based medicine is rigorous, there are always exceptions. The recommendations are intended to guide healthcare decisions. These recommendations can be adapted to be applicable at various levels.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

Implementation tools are being developed, including a quick reference guide in print and personal digital assistant format, and educational slide presentations for physicians and other health-care practitioners.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness
Staying Healthy

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

McGoon M, Gutterman D, Steen V, Barst R, McCrory DC, Fortin TA, Loyd JE. Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Chest 2004 Jul;126(1 Suppl):14S-34S. [170 references] PubMed

ADAPTATION

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

DATE RELEASED

2004 Jul

GUIDELINE DEVELOPER(S)

American College of Chest Physicians - Medical Specialty Society

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GUIDELINE COMMITTEE

American College of Chest Physicians (ACCP) Expert Panel on Pulmonary Artery Hypertension

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

The following participants have disclosed information regarding potential or real conflicts of interest and commitment:

Steven H. Abman, MD: scientific advisory board for INO Therapeutics; consultant for Pfizer.

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David B. Badesch, MD, FCCP: consultant or Speaker's Bureau for Glaxo Wellcome/GlaxoSmithKline, Actelion, InterMune, Encysive, Myogen, Astra-Merck, Astra-Zeneca, Exhale Therapeutics/CoTherix, Forrest Labs, INO Therapeutics, Berlex; research support from Glaxo Wellcome/GlaxoSmithKline, United Therapeutics, Boehringer Ingelheim, Actelion, Encysive, ICOS/Texas Biotechnologies/Encysive, Myogen, INO Therapeutics, Scleroderma Foundation, National Institutes of Health, National Heart, Lung, and Blood Institute, United Therapeutics, Pfizer, American Lung Association.

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Ramona L. Doyle, MD, FCCP: Speaker's Bureau for Actelion; clinical research for Actelion, Myogen, United Therapeutics.

David D. Gutterman, MD, FCCP: stock options with Johnson & Johnson; relative who is a Vice-President at GlaxoSmithKline.

James E. Loyd, MD, FCCP: relationships with GlaxoSmithKline, United Therapeutics, Actelion, ICOS/Texas Biotechnology, Westat, PRA International, Pfizer, Exhale Therapeutics.

Michael D. McGoon, MD: past research support from Glaxo Wellcome, United Therapeutics, Actelion; research support from Texas Biotech/Encysive, Myogen, Pfizer, Medtronic.

Vallerie V. McLaughlin, MD, FCCP: consultant for Actelion, United Therapeutics, Exhale Therapeutics; Speaker's Bureau for Actelion; research funding from Actelion, United Therapeutics, Pfizer, Encysive/Texas Biotechnologies, Glaxo Wellcome, Exhale Therapeutics, Myogen.

Stuart Rich, MD: research funding from Actelion, Pfizer, United Therapeutics, Encysive, Myogen; consultant for Actelion, Pfizer, United Therapeutics, GlaxoSmithKline.

Lewis J. Rubin, MD, FCCP: consultant for Actelion, Myogen, Schering, Exhale Therapeutics, United Therapeutics, Pfizer, Celgene; investigator for Actelion, Myogen, Exhale, Pfizer, Celgene; no stock holdings or other ownerships or positions.

Gerald Simonneau, MD: consultant and investigator for Glaxo Wellcome, Pfizer, Actelion, Schering, Myogen, United Therapeutics.

Virginia D. Steen, MD: relationships with Arthritis Foundation, Scleroderma Foundation, Actelion.

Fredrick M. Wigley, MD: research funding from Biogen, Pfizer, Actelion; consultant to Genzyme.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available to subscribers of Chest - The Cardiopulmonary and Critical Care Journal.

Print copies: Available from the American College of Chest Physicians, Products and Registration Division, 3300 Dundee Road, Northbrook IL 60062-2348.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

Background Articles

- Rubin, LJ. Diagnosis and management of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Introduction. Chest 2004 Jul;126(1 Suppl):7S-10S.
- Rubin LJ. Diagnosis and management of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Executive summary. Chest 2004 Jul;126(1 Suppl):4S-6S.
- McCrory DC, Lewis SZ. Methodology and grading for pulmonary hypertension evidence review and guideline development. Chest 2004 Jul;126(1 Suppl):11S-13S.

Electronic copies: Available to subscribers of Chest - The Cardiopulmonary and Critical Care Journal.

Print copies: Available from the American College of Chest Physicians, Products and Registration Division, 3300 Dundee Road, Northbrook IL 60062-2348.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on August 27, 2004.

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