



Complete Summary

GUIDELINE TITLE

Surgical treatments/interventions for pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines.

BIBLIOGRAPHIC SOURCE(S)

Doyle RL, McCrory D, Channick RN, Simonneau G, Conte J. Surgical treatments/interventions for pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Chest 2004 Jul;126(1 Suppl):63S-71S. [67 references] [PubMed](#)

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Pulmonary arterial hypertension

GUIDELINE CATEGORY

Treatment

CLINICAL SPECIALTY

Cardiology
Internal Medicine

Pulmonary Medicine
Thoracic Surgery

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To provide appropriate evidence-based recommendations for the selection and timing of surgical and interventional treatments of pulmonary arterial hypertension

TARGET POPULATION

Patients with pulmonary arterial hypertension

INTERVENTIONS AND PRACTICES CONSIDERED

Surgical Treatment/Interventions

1. Atrial septostomy
2. Pulmonary thromboendarterectomy
3. Alternatives or adjuncts to pulmonary thromboendarterectomy (balloon dilation and medical therapy)
4. Lung transplantation
5. Heart-lung transplantation
6. Bilateral lung transplantation (with repair of the cardiac defect)

MAJOR OUTCOMES CONSIDERED

- Hemodynamic outcomes
- Postoperative functional status
- Survival

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

For the topic on surgical treatments/interventions, the guideline panel considered studies with at least 10 subjects conducted in any patients with known or suspected idiopathic pulmonary arterial hypertension (IPAH), PAH associated with congenital heart disease, and pulmonary hypertension (PH) associated with chronic thromboembolic disease. The guideline panel excluded studies in patients with PH associated with left-sided cardiac disease, such as congestive heart failure or valvular disease, as well as PH associated with high altitude. The guideline panel excluded studies or analysis of studies of corrective surgical procedures for congenital heart disease associated PH.

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

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

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

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 experts 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.

Although the guideline panel has attempted to provide an evidence-based overview of the available data on atrial septostomy, pulmonary thromboendarterectomy, and transplantation, there are important limitations of the evidence due in large part to the quality and number of the studies forming the basis of the recommendations. When specific data were not available, recommendations were based on expert opinion from centers that treat large numbers of patients with pulmonary hypertension and have extensive experience in the procedures examined.

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

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. In select patients with pulmonary arterial hypertension (PAH) unresponsive to medical management, atrial septostomy (AS) should be considered. **Quality of evidence: low; net benefit: intermediate; strength of recommendation: C.**
2. In patients with PAH, AS should be performed only at institutions with significant procedural and clinical experience. **Quality of evidence: expert opinion; net benefit: substantial; strength of recommendation: E/A.**
3. Patients with suspected chronic thromboembolic pulmonary hypertension (CTEPH) should be referred to centers experienced in the procedure for consideration of pulmonary thromboendarterectomy (PTE). **Level of evidence: expert opinion; benefit: substantial; grade of recommendation: E/A.**
4. In patients with operable CTEPH, PTE is the treatment of choice for improved hemodynamics, functional status, and survival. **Level of evidence: low; benefit: substantial; grade of recommendation: B.**
5. In patients with CTEPH deemed inoperable or with significant residual postoperative pulmonary hypertension (PH), balloon dilation, PAH medical therapy, or lung transplantation (LT) may be considered. **Level of evidence: low; benefit: small/weak; grade of recommendation: C.**
6. PAH patients with New York Heart Association (NYHA) functional class III and IV symptoms should be referred to a transplant center for evaluation and listing for LT or heart-lung transplantation (HLT). **Level of evidence: low; benefit: substantial; grade of recommendation: B.**
7. Listed patients with PAH whose prognosis remains poor despite medical therapy should undergo LT or HLT. **Level of evidence: fair; benefit: substantial; grade of recommendation: A.**
8. In patients with PAH who are undergoing transplantation, the procedure of choice is bilateral lung transplantation (BLT). **Level of evidence: low; benefit: intermediate; grade of recommendation: C.**
9. In children with PAH who are undergoing transplantation, the procedure of choice is BLT. **Level of evidence: low; benefit: substantial; grade of recommendation: B.**
10. In adult patients with PAH and simple congenital heart lesions, BLT with repair of the cardiac defect is the procedure of choice. **Level of evidence: low; benefit: intermediate; grade of recommendation: C.**
11. In adult patients with PAH and complex congenital heart disease who are undergoing transplantation, HLT is the procedure of choice. **Level of evidence: low; benefit: substantial; grade of recommendation: B.**

Definitions

Quality of the Evidence

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Strength of Recommendations

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E/D = negative recommendation based on expert opinion only

Net Benefit

Substantial
Intermediate
Small/weak
None
Conflicting
Negative

CLINICAL ALGORITHM(S)

None provided

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

- Development of appropriate recommendations for the selection and timing of surgical and interventional treatments of pulmonary arterial hypertension
- Improved cardiac output and decreased pulmonary arterial pressure and improved functional status
- Atrial septostomy (AS) may serve as palliation or as a bridge to more definitive treatment, such as lung transplantation.
- Pulmonary thromboendarterectomy (PTE) offers a potential surgical cure for a subset of patients with chronic thromboembolic PAH (CTEPH).

POTENTIAL HARMS

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

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

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based clinical practice guidelines. Chest 2004 Jul;126(1 Suppl):63S-71S. [67 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:

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Charles W. Atwood, Jr., MD, FCCP: research support from Respironics, Inc.

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

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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.

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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

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