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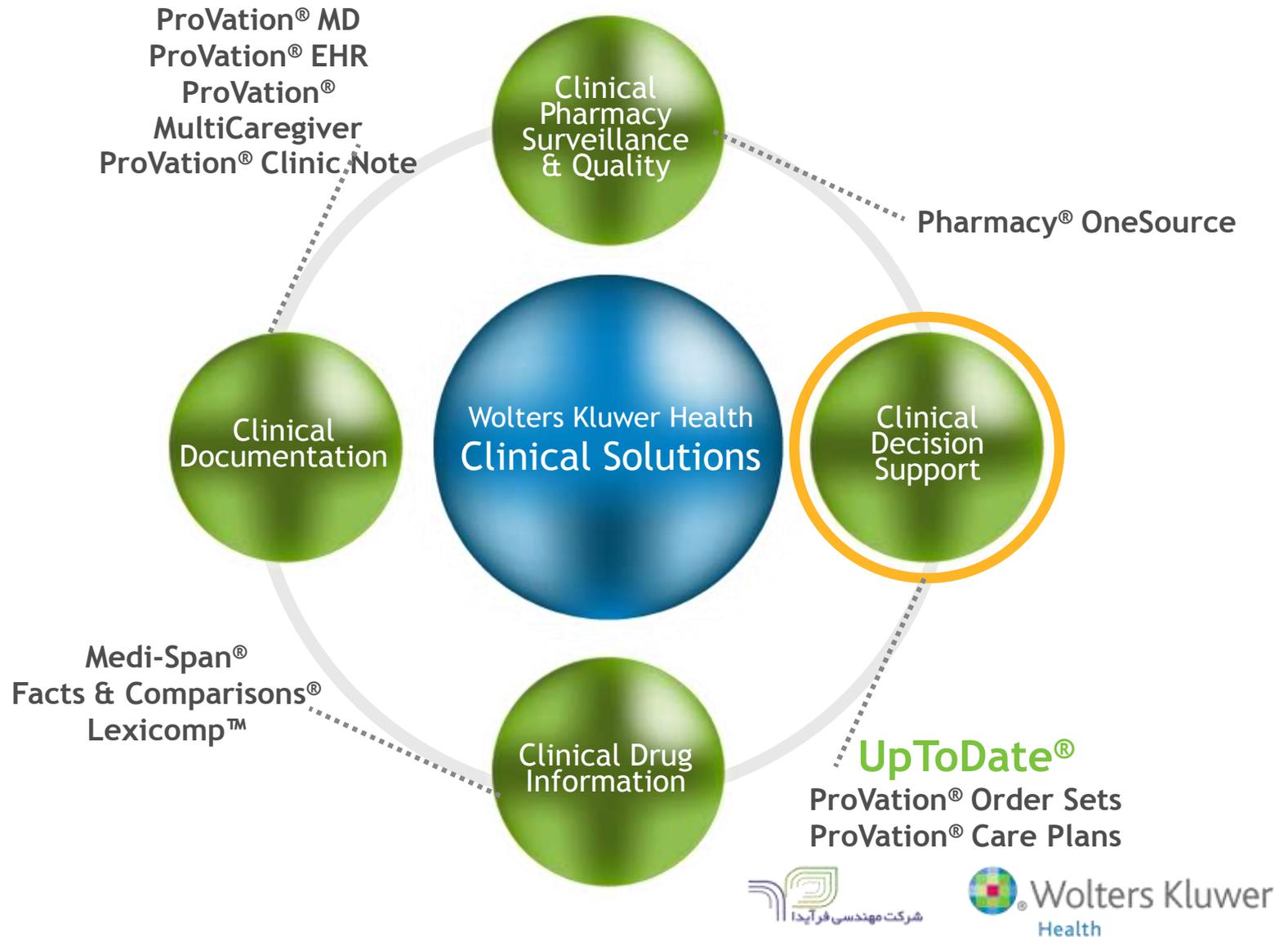
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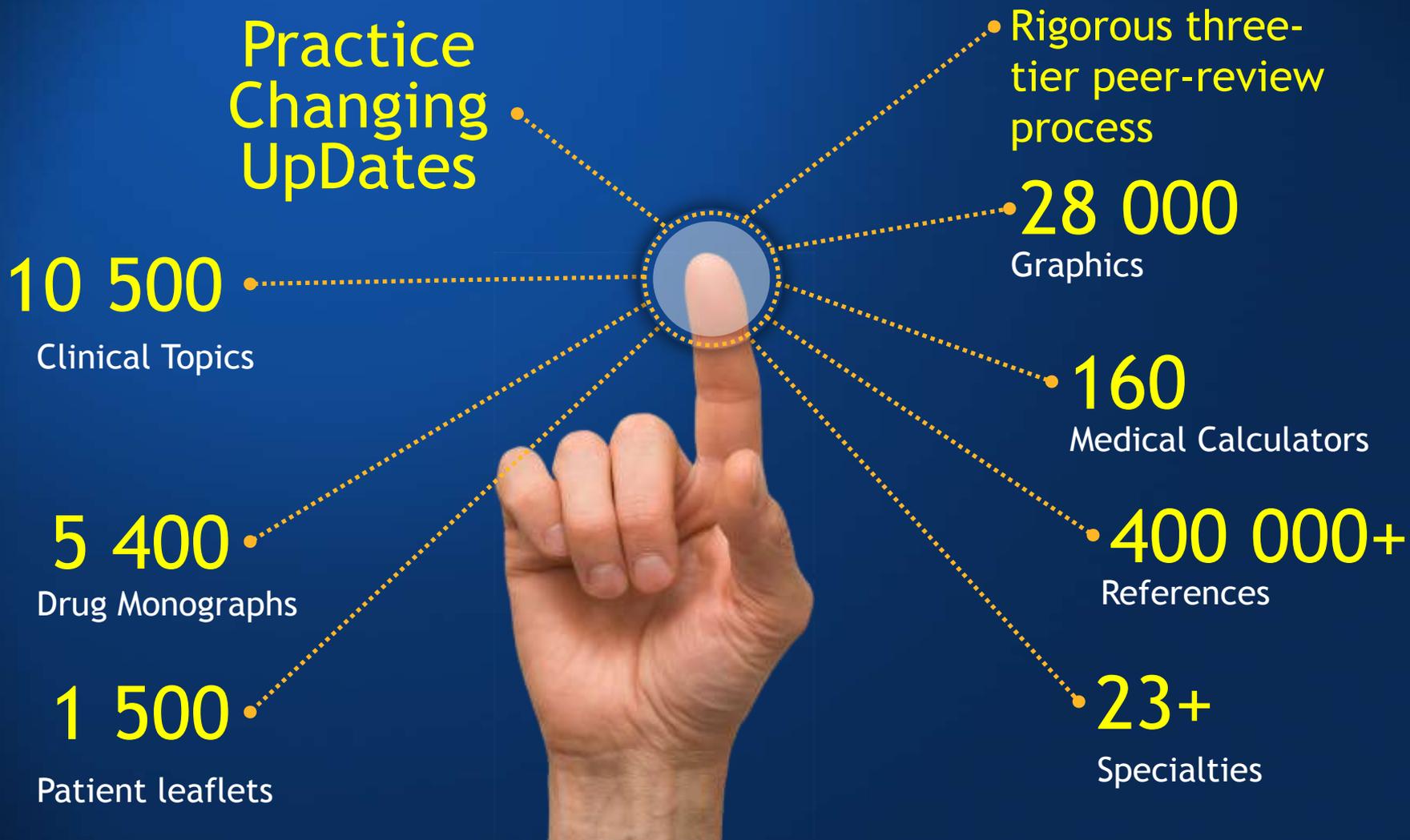
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Cystic fibrosis: Antibiotic therapy for lung disease

Author
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Disclosures

All topics are updated as new evidence becomes available and our [peer review process](#) is complete. Literature review current through: Jan 2014. | This topic last updated: Sep 18, 2013.

INTRODUCTION — Cystic fibrosis (CF) is a multisystem disorder caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, located on chromosome 7 [1]. (See "[Cystic fibrosis: Genetics and pathogenesis](#)".)

Pulmonary disease remains the leading cause of morbidity and mortality in patients with CF [2-5]. One of the major drivers of CF lung disease is infection [6,7]. The approach to treating infection in CF is multifaceted, involving antibiotics, chest physiotherapy, inhaled medications to promote secretion clearance, and anti-inflammatory agents. Undoubtedly, improved use of antibiotics is responsible for a substantial portion of the increased survival that has occurred in patients with CF ([figure 1](#)) [4,6].

The use of antibiotics to treat CF lung disease will be reviewed here. Treatments other than antibiotics for CF lung disease and the diagnosis, clinical manifestations, and investigational therapies for CF are discussed separately. (See "[Cystic fibrosis: Overview of the treatment of lung disease](#)" and "[Cystic fibrosis: Clinical manifestations and diagnosis](#)" and "[Cystic fibrosis: Clinical manifestations of pulmonary disease](#)" and "[Cystic fibrosis: Investigational therapies](#)".)

PATHOGENS — Chronic bacterial infection within the airways occurs in most patients with cystic fibrosis (CF) ([table 1](#)); the prevalence of each bacterial type varies with the age of the patient ([figure 2](#)).

Pseudomonas aeruginosa — For reasons that are poorly understood, the CF airway is particularly susceptible to Pseudomonas aeruginosa (P. aeruginosa), with infection occurring as early as the first year of life. The prevalence of Pseudomonas aeruginosa (P. aeruginosa) increases as patients age, such that more than 73 percent of adults are chronically infected [8]. With prolonged infection, P. aeruginosa converts to a mucoid phenotype by the production of alginate. This mucoid phenotype is seen infrequently in populations without CF but is manifested by over 66 percent of patients with CF. (See "[Clinical microbiology, microbiology, and pathogenesis of Pseudomonas aeruginosa](#)".)

Chronic infection with P. aeruginosa is associated with accelerated loss of pulmonary function and decreased survival [9,10].

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SUMMARY AND RECOMMENDATIONS

- Cystic fibrosis (CF) lung disease is characterized by persistent infection with *Pseudomonas aeruginosa* (P. aeruginosa) are the most prevalent pathogens (Grade 1B).
- The clinical course is frequently complicated by acute pulmonary exacerbations that impair lung function. Exacerbations are treated with antibiotics, given either orally or intravenously, based on the sensitivities of the infecting bacteria (table 2). Current practice is to treat with two antibiotics if P. aeruginosa is cultured from respiratory secretions, and two antibiotics for P. aeruginosa are [piperacillin-tazobactam](#), [ticarcillin-clavulanate](#), [ceftazidime](#), [imipenem](#), [meropenem](#), or [levofloxacin](#). For other pathogens, [azithromycin](#), [amikacin](#), or a fluoroquinolone (eg, [ciprofloxacin](#)), depending on antibiotic susceptibility test results. (See 'Antibiotic selection' above.)
- The pharmacokinetics of many antibiotics differs in patients with CF as compared with normal individuals. Patients with CF generally require larger and/or more frequent dosing for penicillins, cephalosporins, sulfonamides, and fluoroquinolones. Starting doses of aminoglycosides should also be larger than those recommended for individuals without CF, but dosing must be adjusted based on pharmacokinetic analysis of serum levels because of considerable interindividual variation in clearance rates. (See 'Dosing' above.)
- In the absence of an acute pulmonary exacerbation, we generally suggest not administering chronic or intermittent systemic antibiotics to patients with CF (Grade 2C), EXCEPT for the following:
 - We recommend the chronic use of [azithromycin](#) for patients 6 years and older who have clinical evidence of airway inflammation such as chronic cough or any reduction in forced expiratory volume at one minute (FEV1), regardless of the patient's P. aeruginosa infection status (Grade 1B). To avoid induction of antibiotic resistance, azithromycin should not be given to patients infected with nontuberculous mycobacteria. (See 'Chronic oral antibiotics' above and "Cystic fibrosis: Overview of the treatment of lung disease", section on 'Macrolide antibiotics'.)
 - For patients older than six years with persistent P. aeruginosa infection and moderate or severe lung disease, we recommend chronic treatment with inhaled [tobramycin](#) (Grade 1A). We also suggest this treatment for patients with mild lung disease and persistent P. aeruginosa infection (Grade 2B). Inhaled [aztreonam](#) lysine is a reasonable alternative. Either inhaled tobramycin or aztreonam lysine are given for one month, on alternate months. (See 'Inhaled antibiotics' above.)
- We suggest not scheduling elective hospitalizations for antibiotics and intensified chest physiotherapy ("clean out") (Grade 2C). (See 'Hospitalizations' above.)

Based on the body of evidence, and the expertise of the leading specialty experts we make graded recommendations on the next course of action

- We recommend the chronic use of [azithromycin](#) for patients 6 years and older who have clinical evidence of airway inflammation such as chronic cough or any reduction in forced expiratory volume at one minute (FEV1), regardless of the patient's P. aeruginosa infection status ([Grade 1B](#)). To avoid induction of antibiotic resistance, azithromycin should not be given to patients infected with nontuberculous mycobacteria. (See '[Chronic oral antibiotics](#)' above and "[Cystic fibrosis: Overview of the treatment of lung disease](#)", section on 'Macrolide antibiotics'.)
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Grade 1A recommendation

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Explanation:

A Grade 1 recommendation is a strong recommendation. It means that we believe that if you follow the recommendation, you will be doing more good than harm for most, if not all of your patients.

Grade A means that the best estimates of the critical benefits and risks come from consistent data from well-performed, randomized, controlled trials or overwhelming data of some other form (eg, well-executed observational studies with very large treatment effects). Further research is unlikely to have an impact on our confidence in the estimates of benefit and risk.

Recommendation grades

1. Strong recommendation: Benefits clearly outweigh the risks and burdens (or vice versa) for most, if not all, patients
2. Weak recommendation: Benefits and risks closely balanced and/or uncertain

Evidence grades

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- C. Low-quality evidence: Evidence from observational studies, unsystematic clinical observations, or from randomized trials with serious flaws

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- We recommend the chronic use of **azithromycin** for patients 6 years and older who have clinical evidence of airway inflammation such as chronic cough or any reduction in forced expiratory volume at one minute (FEV1), regardless of the patient's P. aeruginosa infection status (**Grade 1B**). To avoid induction of antibiotic resistance, azithromycin should not be given to patients infected with nontuberculous mycobacteria. (See '[Chronic oral antibiotics](#)' above and "[Cystic fibrosis: Overview of the treatment of lung disease](#)", section on '[Macrolide antibiotics](#)'.)
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- AbobotulinumtoxinA: Aminoglycosides may enhance the neuromuscular-blocking effect of AbobotulinumtoxinA. *Risk C: Monitor therapy*
- Amphotericin B: May enhance the nephrotoxic effect of Aminoglycosides. *Risk C: Monitor therapy*
- BCG: Antibiotics may diminish the therapeutic effect of BCG. *Risk X: Avoid combination*
- Bisphosphonate Derivatives: Aminoglycosides may enhance the hypocalcemic effect of Bisphosphonate Derivatives. *Risk C: Monitor therapy*
- Capreomycin: May enhance the neuromuscular-blocking effect of Aminoglycosides. *Risk C: Monitor therapy*
- CARBOplatin: Aminoglycosides may enhance the ototoxic effect of CARBOplatin. Especially with higher doses of carboplatin. *Risk C: Monitor therapy*
- Cephalosporins (2nd Generation): May enhance the nephrotoxic effect of Aminoglycosides. *Risk C: Monitor therapy*
- Cephalosporins (3rd Generation): May enhance the nephrotoxic effect of Aminoglycosides. *Risk C: Monitor therapy*
- Cephalosporins (4th Generation): May enhance the nephrotoxic effect of Aminoglycosides. *Risk C: Monitor therapy*
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- Gallium Nitrate: Aminoglycosides may enhance the nephrotoxic effect of Gallium Nitrate. *Risk C: Monitor therapy*
- Loop Diuretics: May enhance the adverse/toxic effect of Aminoglycosides. *Risk C: Monitor therapy*
- Neuromuscular-Blocking Agents: Aminoglycosides may enhance the effect of Neuromuscular-Blocking Agents. *Risk C: Monitor therapy*
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Risk Rating	Action	Description
A	<i>No Known Interaction</i>	Data have not demonstrated either pharmacodynamic or pharmacokinetic interactions between the specified agents
B	<i>No Action Needed</i>	Data demonstrate that the specified agents may interact with each other, but there is little to no evidence of clinical concern resulting from their concomitant use.
C	<i>Monitor Therapy</i>	Data demonstrate that the specified agents may interact with each other in a clinically significant manner. The benefits of concomitant use of these two medications usually outweigh the risks. An appropriate monitoring plan should be implemented to identify potential negative effects. Dosage adjustments of one or both agents may be needed in a minority of patients.
D	<i>Consider Therapy Modification</i>	Data demonstrate that the two medications may interact with each other in a clinically significant manner. A patient-specific assessment must be conducted to determine whether the benefits of concomitant therapy outweigh the risks. Specific actions must be taken in order to realize the benefits and/or minimize the toxicity resulting from concomitant use of the agents. These actions may include aggressive monitoring, empiric dosage changes, choosing alternative agents.
X	<i>Avoid Combination</i>	Data demonstrate that the specified agents may interact with each other in a clinically significant manner. The risks associated with concomitant use of these agents usually outweigh the benefits. These agents are generally considered contraindicated.

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Cystic fibrosis: Antibiotic therapy for lung disease

Author

Richard H Simon, MD

Disclosures

All topics are updated as new evidence becomes available. Literature review current through: January 2014.

INTRODUCTION — Cystic fibrosis (CF) is caused by a mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, located on chromosome 7q31.2.

Pulmonary disease remains the leading cause of morbidity and mortality in CF. The approach to treating CF lung disease focuses on increasing mucociliary secretion clearance, and anti-inflammatory therapy. The use of antibiotics to treat CF lung disease is based on clinical manifestations, and investigations of the role of bacterial infection in the disease.

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Cystic fibrosis: Antibiotic therapy for lung disease

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Curr Opin Pulm Med, 2004 Nov;10(6):510-4.

Update on cystic fibrosis epidemiology.

Goss CH, Rosenfeld M.

Author information

Abstract

PURPOSE OF REVIEW: With the improving survival of cystic fibrosis (CF) patients, the clinical spectrum of this complex multisystem disease continues to evolve. Epidemiologic studies have provided important insight into the disease course, prognosis, and complications. This review summarizes recent advances in our understanding of predictors of survival and outcome and modifiers of disease in CF. This review is not meant to be comprehensive, but highlights selected studies, many of which have particular relevance to the growing number of older CF patients.

RECENT FINDINGS: Survival rates of US CF patients improved remarkably over the past 15 years, but most of the improvement was limited to patients 2 to 15 years of age. Both median household income and ambient air pollutants were found to be important modifiers of disease, echoing research reported in other chronic lung diseases. Genotype classified according to functional mutation class was highly associated with outcome (class I, II, and III mutations were associated with the highest mortality). Of the emerging pathogens, *B. cepacia* complex and *B. gladioli* are the most prominent. A small but significant percentage of patients have been shown to acquire new *B. cepacia* complex or *B. gladioli* strains with time.

SUMMARY: Epidemiologic research in cystic fibrosis continues to inform patient care and clinical research, and to generate new hypotheses regarding pathophysiology. Survival and outcomes continue to improve in this multisystem disease. With continued improving survival, epidemiologic studies will be critical to tracking changes in prognosis and outcome.

PMID: 15510059 [PubMed - indexed for MEDLINE]

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Cystic fibrosis: Antibiotic th

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Defects in the CFTR gene in cystic fibrosis

Class IV: Defective conduction

Class III: Defective regulation

Class II: Defective processing

Class I: Defective protein production

Schematic representation of the biosynthesis and function of CFTR in an epithelial cell and of mechanisms of dysfunction associated with different cystic fibrosis mutations. CFTR: cystic fibrosis transmembrane conductance regulator; ER: endoplasmic reticulum; PKA: phosphokinase A; NBO1 and NBO2: nuclear binding folds.

Adapted from Welsh, MJ, Tsui, L-C, Boat, T, Beaudet, AL. Cystic fibrosis. In: *The Metabolic and Molecular Basis of Inherited Disease*, Scriver, CR, Beaudet, AL, Sly, WS, et al (Eds), McGraw-Hill, New York, 1995, p. 3801.

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Beaudet, AL, Sly, WS, et al (Eds), McGraw-Hill, New York, 1995, p. 3801.

1500 Patient Support Leaflets

The Basics

1 to 3 page long
Written in plain language.
Best for a general overview
Answer the 4 or 5 most important questions

Beyond the Basics

5 - 10 pages long
More detailed than "The Basics"
Better for readers who are comfortable with some technical medical terms.



IMPORTANT - All leaflets are written by the same editorial experts

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cystic fibrosis children

All Topics



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Cystic fibrosis: Antibiotic therapy for lung disease

cystic fibrosis children

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Patient information: Cystic fibrosis (The Basics)

Patient information: Cystic fibrosis (The Basics)

Written by the doctors and editors at UpToDate

What is cystic fibrosis? — Cystic fibrosis is a disease that some children are born with. It causes thick mucus and other fluids to build up and clog different parts of the body, including the lungs, pancreas, liver, and intestine (figure 1).

The thick mucus in the lungs causes people with cystic fibrosis to get frequent lung infections. Over time, these infections damage the lungs. The thick fluids in the pancreas and liver keep the intestine from absorbing certain nutrients from food. This affects a child's growth and causes abnormal bowel movements.

Cystic fibrosis is caused by an abnormal gene. To get the disease, people need to get the abnormal gene from both their mother and father. If people get the abnormal gene from only 1 parent, they will not have cystic fibrosis. But they will have a chance of passing on the abnormal gene to their children.

Cystic fibrosis is a life-long condition. As of now, doctors can't cure the disease, but they can use different treatments to help with symptoms.

People with cystic fibrosis don't live as long as people without the disease. But better treatments are helping people with cystic fibrosis live longer. To help manage your child's disease for as long as possible, it's important to work closely with his or her doctor.

What are the symptoms of cystic fibrosis? — People can have different symptoms at different times. Most people start having symptoms as a baby or young child. A few people start having symptoms as teens or adults. A person's symptoms usually get worse over time.

Common symptoms of cystic fibrosis include:

- Not growing or gaining weight normally
- A long-lasting cough — The cough usually brings up mucus (it sounds "wet"). Some people cough up blood.
- Trouble breathing or breathing that sounds like whistling (wheezing)
- Frequent infections of the lungs or sinuses — The sinuses are hollow areas in the bones of the face.
- Skin that tastes salty (for example, you might taste salt when you kiss your child)
- Belly pain, diarrhea, or constipation (trouble having bowel movements)

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Contents: Patient Information

UpToDate offers different levels of patient education materials to meet the varying information needs of your patients.

The Basics

"The Basics" are short (1 to 3 page) articles written in plain language. They answer the 4 or 5 most important questions a person might have about a medical problem. These articles are best for people who want a general overview.

NEW - The Basics articles are also available in Spanish.

[View all The Basics](#)

Beyond the Basics

"Beyond the Basics" articles are 5 to 10 pages long and more detailed than "The Basics". These articles are best for readers who want a lot of detailed information and who are comfortable with some technical medical terms.

[View all Beyond the Basics](#)



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TOPIC OUTLINE

ASTHMA

- Highly selective COX-2 inhibitors in aspirin-exacerbated respiratory disease (January 2014)
- Anti-IgE (omalizumab) therapy improves asthma control in occupational asthma (August 2013)
- Predicting asthma response to anti-IgE therapy (omalizumab) (August 2013)

COPD

- Combination therapy with umecclidinium and vilanterol for COPD (January 2014)
- Reassuring data about the tiotropium soft mist inhaler (October 2013)

CRITICAL CARE

- Score to predict neurologic status following in-hospital cardiopulmonary resuscitation (January 2014)
- Futile therapy in the ICU (November 2013)
- Statins in patients with ventilator-associated pneumonia (October 2013)
- Crystalloids versus colloids for hypovolemic shock (October 2013)
- Beta blockade as a therapy in septic shock (October 2013)
- Universal contact precautions in the intensive care unit (October 2013)
- FDA approval of telavancin for HAP and VAP (October 2013)

What's new in pulmonary and critical care medicine

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Disclosures

All topics are updated as new evidence becomes available and our [peer review process](#) is complete.

Literature review current through: Jan 2014. | This topic last updated: Jan 14, 2014.

The following represent additions to UpToDate from the past six months that were considered by the editors and authors to be of particular interest. The most recent What's New entries are at the top of each subsection.

ASTHMA

Highly selective COX-2 inhibitors in aspirin-exacerbated respiratory disease (January 2014)

Patients with aspirin-exacerbated respiratory disease (AERD) often have severe hypersensitivity reactions to nonsteroidal antiinflammatory drugs (NSAIDs), which are directly related to inhibition of the enzyme COX-1. Although highly selective COX-2 inhibitors are theoretically safe, observational studies described patients who appeared to react to these agents. In a new meta-analysis of placebo-controlled blinded trials, over 400 patients with AERD were challenged with highly selective (celecoxib, rofecoxib) or relatively selective (ie, meloxicam, nabumetone, nimesulide) COX-2 inhibitors [1]. Whereas 1 in 13 patients had reactions to the relatively selective agents, there were no reactions to the highly selective agents. This analysis supports the safety of highly selective COX-2 inhibitors in patients with AERD. (See "[NSAIDs \(including aspirin\): Allergic and pseudoallergic reactions](#)", section on 'Highly selective COX-2 inhibitors'.)

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Topic Feedback

Practice Changing UpDates

TOPIC OUTLINE 

INTRODUCTION

ONCOLOGY, GYNECOLOGY
(FEBRUARY 2013, MODIFIED
FEBRUARY 2014)

- Chemotherapy plus bevacizumab for recurrent, advanced, or metastatic cervical cancer

GASTROENTEROLOGY AND
HEPATOLOGY (DECEMBER 2013)

- Sofosbuvir and simeprevir for chronic genotype 1 hepatitis C infection

HEMATOLOGY (DECEMBER 2013)

- Obinutuzumab plus chlorambucil for previously untreated chronic lymphocytic leukemia

ONCOLOGY, GENERAL SURGERY
(OCTOBER 2013)

- New ASCO/CAP criteria for HER2 positivity

RHEUMATOLOGY, ADULT PRIMARY
CARE, FAMILY MEDICINE,
CARDIOLOGY (AUGUST 2013)

- Cardiovascular risk of NSAIDs

INFECTIOUS DISEASES (AUGUST
2013)

- Treatment of AIDS-related CMV retinitis

INFECTIOUS DISEASES, ADULT
PRIMARY CARE, FAMILY MEDICINE
(JULY 2013)

- Pre-exposure prophylaxis against HIV infection for injecting drug users

GYNECOLOGY, ADULT PRIMARY
CARE, FAMILY MEDICINE (MAY 2013,
MODIFIED JUNE 2013)

Practice Changing UpDates

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Disclosures

All topics are updated as new evidence becomes available and our [peer review process](#) is complete.
Literature review current through: Jan 2014. | **This topic last updated:** Jan 14, 2014.

INTRODUCTION — This section highlights selected specific new recommendations and/or updates that we anticipate may change usual clinical practice. Practice Changing UpDates focus on changes that may have significant and broad impact on practice, and therefore do not represent all updates that affect practice. These Practice Changing UpDates, reflecting important changes to UpToDate over the past year, are presented chronologically, and are discussed in greater detail in the identified topic reviews.

ONCOLOGY, GYNECOLOGY (FEBRUARY 2013, MODIFIED FEBRUARY 2014)

Chemotherapy plus bevacizumab for recurrent, advanced, or metastatic cervical cancer

- For women with recurrent cervical cancer, those with advanced disease who are not surgical candidates, and those who present with metastatic disease, we recommend first-line treatment with chemotherapy plus bevacizumab rather than chemotherapy alone ([Grade 1B](#)).

Women with recurrent, metastatic, or advanced cervical cancer should receive treatment consisting of a platinum-based combination chemotherapy plus the angiogenesis inhibitor bevacizumab. This recommendation is based on the results of the GOG 240 study, in which over 450 women were randomly assigned to chemotherapy with or without bevacizumab [1]. Compared to chemotherapy alone, treatment incorporating bevacizumab resulted in significant improvements in overall survival, progression-free survival, and the overall response rate. However, chemotherapy plus bevacizumab increased the rates of hypertension and a higher incidence of fistula formation, bleeding, and thromboembolic events. Based on the survival benefits, we recommend the administration of bevacizumab in addition to chemotherapy as a first-line treatment for metastatic cervical cancer. However, the costs of bevacizumab may require scrutiny in comparison to its benefits and risks, especially in underdeveloped areas. (See "[Management of recurrent or metastatic cervical cancer](#)", section on 'Chemotherapy plus bevacizumab as first-line treatment'.)

GASTROENTEROLOGY AND HEPATOLOGY (DECEMBER 2013)

Sofosbuvir and simeprevir for chronic genotype 1 hepatitis C infection

- Most patients with chronic genotype 1 HCV infection who are candidates for and desire therapy should be treated with peginterferon, weight-based ribavirin, and a direct-acting antiviral (DAA). For these patients, we recommend the DAAs sofosbuvir or simeprevir rather than telaprevir or boceprevir ([Grade 1B](#)).

Thank you!

