

A 6-year-old boy with cystic fibrosis is brought to the office for shortness of breath. He has had a worsening, productive cough over the past week and now has fever, chills, and difficulty breathing. His vaccinations are up to date. Weight is at <5th percentile and height is at the 10th percentile. His temperature is 38.9 C (102 F), blood pressure is 122/80 mm Hg, pulse is 118/min, and respirations are 28/min. Pulse oximetry shows an oxygen saturation of 91%. Physical examination shows a tired-appearing boy with nasal flaring, intercostal retractions, and a barrel-shaped chest. Auscultation reveals diffuse bilateral wheezing and crackles. Clubbing of the fingers and toes **exhibit** is present. Sputum culture results are pending. What is the most likely causative organism of this patient's pneumonia?

- ☐ A. *Burkholderia cepacia* complex
- ☐ B. *Cryptococcus neoformans*
- ☐ C. *Histoplasma capsulatum*
- ☐ D. *Klebsiella pneumoniae*
- ☐ E. *Legionella pneumophila*
- ☐ F. *Listeria monocytogenes*
- ☐ G. *Mycobacterium avium* complex
- ☐ H. *Pneumocystis jirovecii*
- ☐ I. *Pseudomonas aeruginosa*
- ☐ J. *Staphylococcus aureus*
- ☐ K. *Streptococcus pneumoniae*



Media Exhibit

1 of 1





A 6-year-old boy with cystic fibrosis is brought to the office for shortness of breath. He has had a worsening, productive cough over the past week and now has fever, chills, and difficulty breathing. His vaccinations are up to date. Weight is at <5th percentile and height is at the 10th percentile. His temperature is 38.9 C (102 F), blood pressure is 122/80 mm Hg, pulse is 118/min, and respirations are 28/min. Pulse oximetry shows an oxygen saturation of 91%. Physical examination shows a tired-appearing boy with nasal flaring, intercostal retractions, and a barrel-shaped chest. Auscultation reveals diffuse bilateral wheezing and crackles. Clubbing of the fingers and toes **exhibit** is present. Sputum culture results are pending. What is the most likely causative organism of this patient's pneumonia?

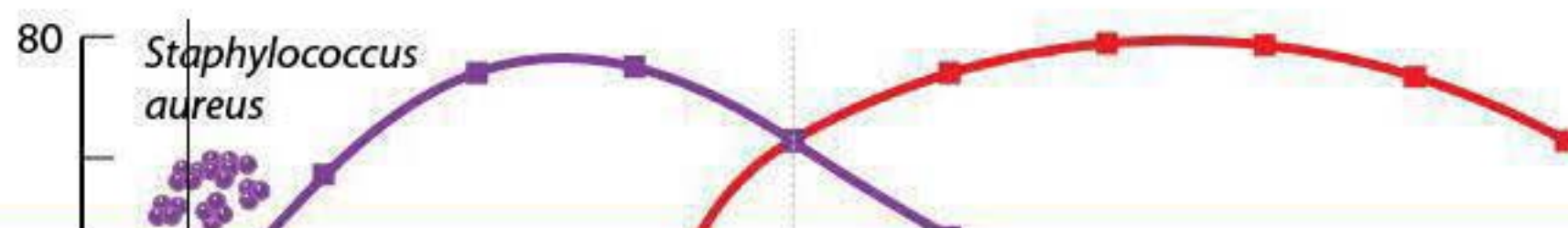
- ☐ A. *Burkholderia cepacia* complex [2%]
- ☐ B. *Cryptococcus neoformans* [0%]
- ☐ C. *Histoplasma capsulatum* [0%]
- ☐ D. *Klebsiella pneumoniae* [1%]
- ☐ E. *Legionella pneumophila* [0%]
- ☐ F. *Listeria monocytogenes* [0%]
- ☐ G. *Mycobacterium avium* complex [0%]
- ☐ H. *Pneumocystis jirovecii* [1%]
- ☐ I. *Pseudomonas aeruginosa* [54%]
- ☒ J. *Staphylococcus aureus* [23%]
- ☐ K. *Streptococcus pneumoniae* [19%]

[Proceed to Next Item](#)

Explanation:

User Id:

Rates of bacterial colonization in cystic fibrosis based on age





A 6-year-old boy with cystic fibrosis is brought to the office for shortness of breath. He has had a worsening, productive cough over the past week and now has fever, chills, and difficulty breathing. His vaccinations are up to date. Weight is at <5th percentile and height is at the 10th percentile. His temperature is 38.9 C (102 F), blood pressure is 122/80 mm Hg, pulse is 118/min, and respirations are 28/min. Pulse oximetry shows an oxygen saturation of 91%. Physical examination shows a tired-appearing boy with nasal flaring, intercostal retractions, and a barrel-shaped chest. Auscultation reveals diffuse bilateral wheezing and crackles. Clubbing of the fingers and toes **exhibit** is present. Sputum culture results are pending. What is the most likely causative organism of this patient's pneumonia?

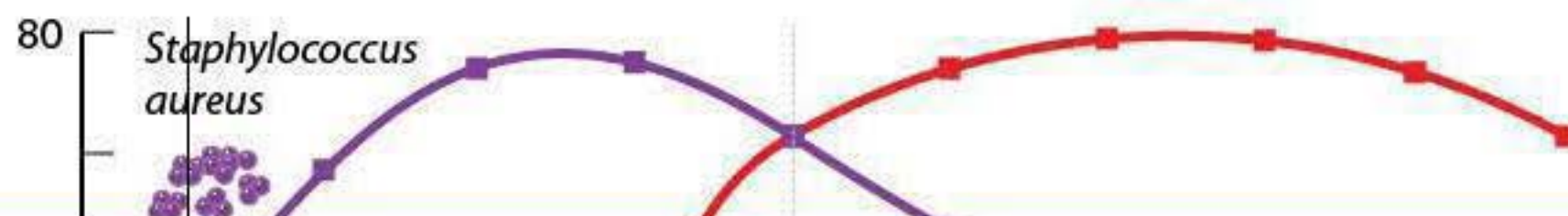
- ☐ A. *Burkholderia cepacia* complex [2%]
- ☐ B. *Cryptococcus neoformans* [0%]
- ☐ C. *Histoplasma capsulatum* [0%]
- ☐ D. *Klebsiella pneumoniae* [1%]
- ☐ E. *Legionella pneumophila* [0%]
- ☐ F. *Listeria monocytogenes* [0%]
- ☐ G. *Mycobacterium avium* complex [0%]
- ☐ H. *Pneumocystis jirovecii* [1%]
- ☐ I. *Pseudomonas aeruginosa* [54%]
- ☒ J. *Staphylococcus aureus* [23%]
- ☐ K. *Streptococcus pneumoniae* [19%]

[Proceed to Next Item](#)

Explanation:

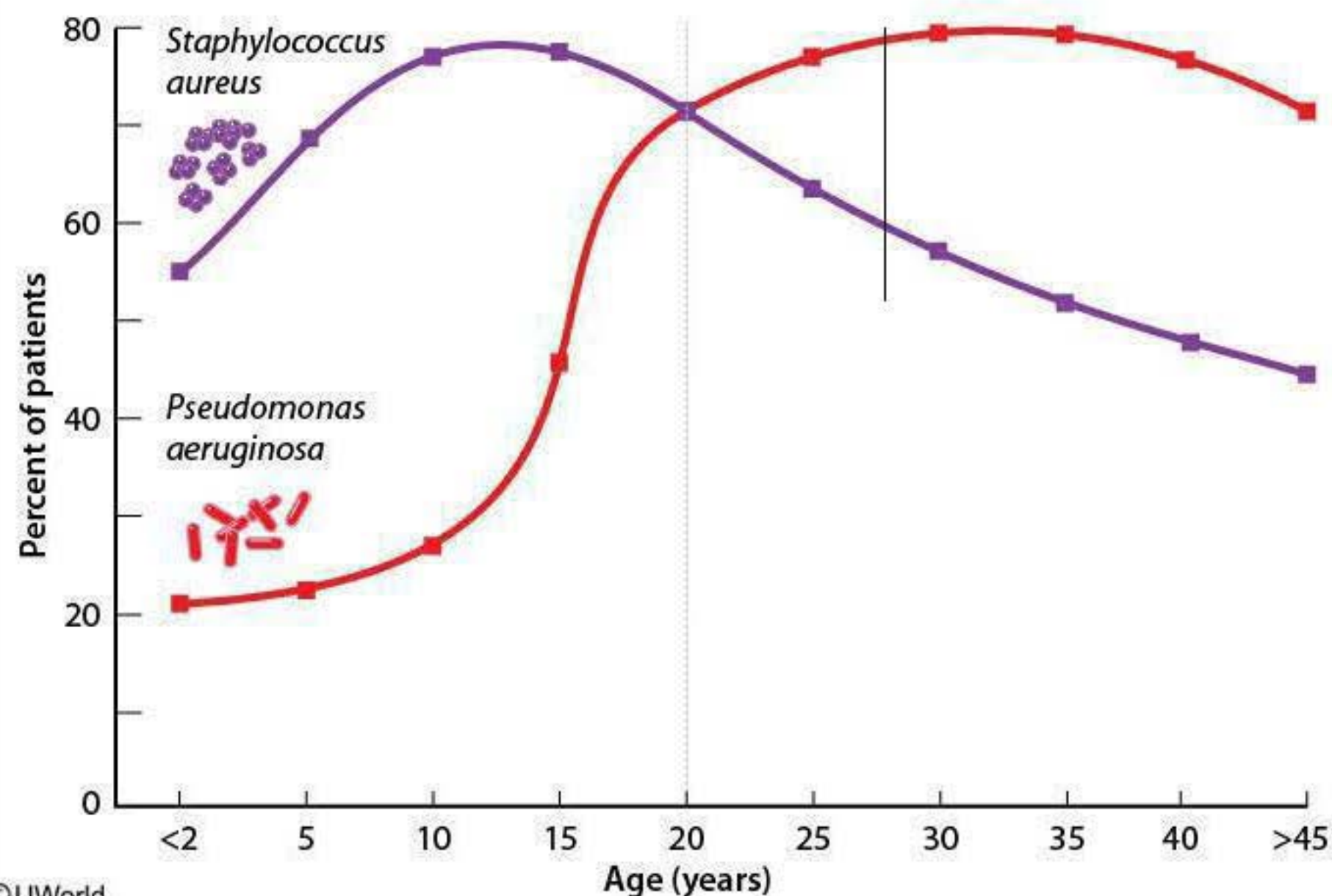
User Id:

Rates of bacterial colonization in cystic fibrosis based on age





Rates of bacterial colonization in cystic fibrosis based on age



©UWorld

**Cystic fibrosis** (CF) is an autosomal recessive disorder characterized by accumulation of inspissated secretions in multiple organs. Most patients have chronic malabsorption and diarrhea as well as **recurrent pneumonias** that lead to irreversible **bronchiectasis**, chronic hypoxia, and progressive respiratory failure. Hyperinflation of the lungs causes the rib cage to remain partially expanded, giving the appearance of a "**barrel chest**". Bulbous enlargement of the tips of the digits (**clubbing**) occurs as well.

Patients with CF are at high risk of infection from unusual and resistant bacteria, viruses, and fungi due to exposure to multiple antibiotic courses. The most common pathogen isolated from sputum cultures in infants and young children is *Staphylococcus aureus*, a gram-positive coccus that grows in clusters. *S aureus* colonization decreases with age but remains a significant source of disease in adults with CF.

Colonization and infection with *Pseudomonas aeruginosa* (**Choice I**) can occur as early as infancy and is the most common cause of CF-related pneumonia in adults. Given its association with a precipitous decline of pulmonary function and increased risk of death,



©UWorld

Age (years)

**Cystic fibrosis** (CF) is an autosomal recessive disorder characterized by accumulation of inspissated secretions in multiple organs. Most patients have chronic malabsorption and diarrhea as well as **recurrent pneumonias** that lead to irreversible **bronchiectasis**, chronic hypoxia, and progressive respiratory failure. Hyperinflation of the lungs causes the rib cage to remain partially expanded, giving the appearance of a "**barrel chest**". Bulbous enlargement of the tips of the digits (**clubbing**) occurs as well.

Patients with CF are at high risk of infection from unusual and resistant bacteria, viruses, and fungi due to exposure to multiple antibiotic courses. The most common pathogen isolated from sputum cultures in infants and young children is *Staphylococcus aureus*, a gram-positive coccus that grows in clusters. *S aureus* colonization decreases with age but remains a significant source of disease in adults with CF.

Colonization and infection with *Pseudomonas aeruginosa* (**Choice I**) can occur as early as infancy and is the most common cause of CF-related pneumonia in adults. Given its association with a precipitous decline of pulmonary function and increased risk of death, *Pseudomonas* should be treated for during initial management but is not the most common pathogen in children.

(**Choice A**) *Burkholderia cepacia* complex refers to several similar species that colonize a small percentage of CF patients. Colonization is associated with accelerated pulmonary decline and decreased survival.

(**Choices B, C, and H**) Patients with deficient cell-mediated immunity (eg, CD4 count <200/ $\mu$ L) are at high risk for fungal pneumonia from *Cryptococcus neoformans*, *Histoplasma capsulatum*, and *Pneumocystis jirovecii*. These fungi are rare in CF; aspergillosis is more common.

(**Choices D and E**) *Klebsiella pneumoniae* and *Legionella pneumophila* are gram-negative rods that cause pneumonia but are less commonly implicated in CF.

(**Choice F**) *Listeria monocytogenes* is a gram-positive rod that causes septicemia and meningitis in neonates and elderly adults. It is an extremely uncommon cause of pneumonia.

(**Choice G**) Nontuberculous mycobacteria (eg, *Mycobacterium avium* complex, *M abscessus*) are becoming more prevalent in CF but are still less common than other bacterial pathogens.

(**Choice K**) *Streptococcus pneumoniae* is the most common cause of pneumonia in otherwise healthy patients. It rarely causes disease in patients with cystic fibrosis, though the pneumococcal vaccine should be administered routinely as in this patient.



Colonization and infection with *Pseudomonas aeruginosa* (**Choice I**) can occur as early as infancy and is the most common cause of CF-related pneumonia in adults. Given its association with a precipitous decline of pulmonary function and increased risk of death, *Pseudomonas* should be treated for during initial management but is not the most common pathogen in children.

(**Choice A**) *Burkholderia cepacia* complex refers to several similar species that colonize a small percentage of CF patients. Colonization is associated with accelerated pulmonary decline and decreased survival.

(**Choices B, C, and H**) Patients with deficient cell-mediated immunity (eg, CD4 count <200/ $\mu$ L) are at high risk for fungal pneumonia from *Cryptococcus neoformans*, *Histoplasma capsulatum*, and *Pneumocystis jirovecii*. These fungi are rare in CF; aspergillosis is more common.

(**Choices D and E**) *Klebsiella pneumoniae* and *Legionella pneumophila* are gram-negative rods that cause pneumonia but are less commonly implicated in CF.

(**Choice F**) *Listeria monocytogenes* is a gram-positive rod that causes septicemia and meningitis in neonates and elderly adults. It is an extremely uncommon cause of pneumonia.

(**Choice G**) Nontuberculous mycobacteria (eg, *Mycobacterium avium* complex, *M abscessus*) are becoming more prevalent in CF but are still less common than other bacterial pathogens.

(**Choice K**) *Streptococcus pneumoniae* is the most common cause of pneumonia in otherwise healthy patients. It rarely causes disease in patients with cystic fibrosis, though the pneumococcal vaccine should be administered routinely as in this patient.

#### Educational objective:

*Staphylococcus aureus* is the most common pathogen isolated in infants and young children with cystic fibrosis (CF). *Pseudomonas aeruginosa* is the most common cause of CF-related pneumonia in adults and contributes to life-threatening decline of pulmonary function.

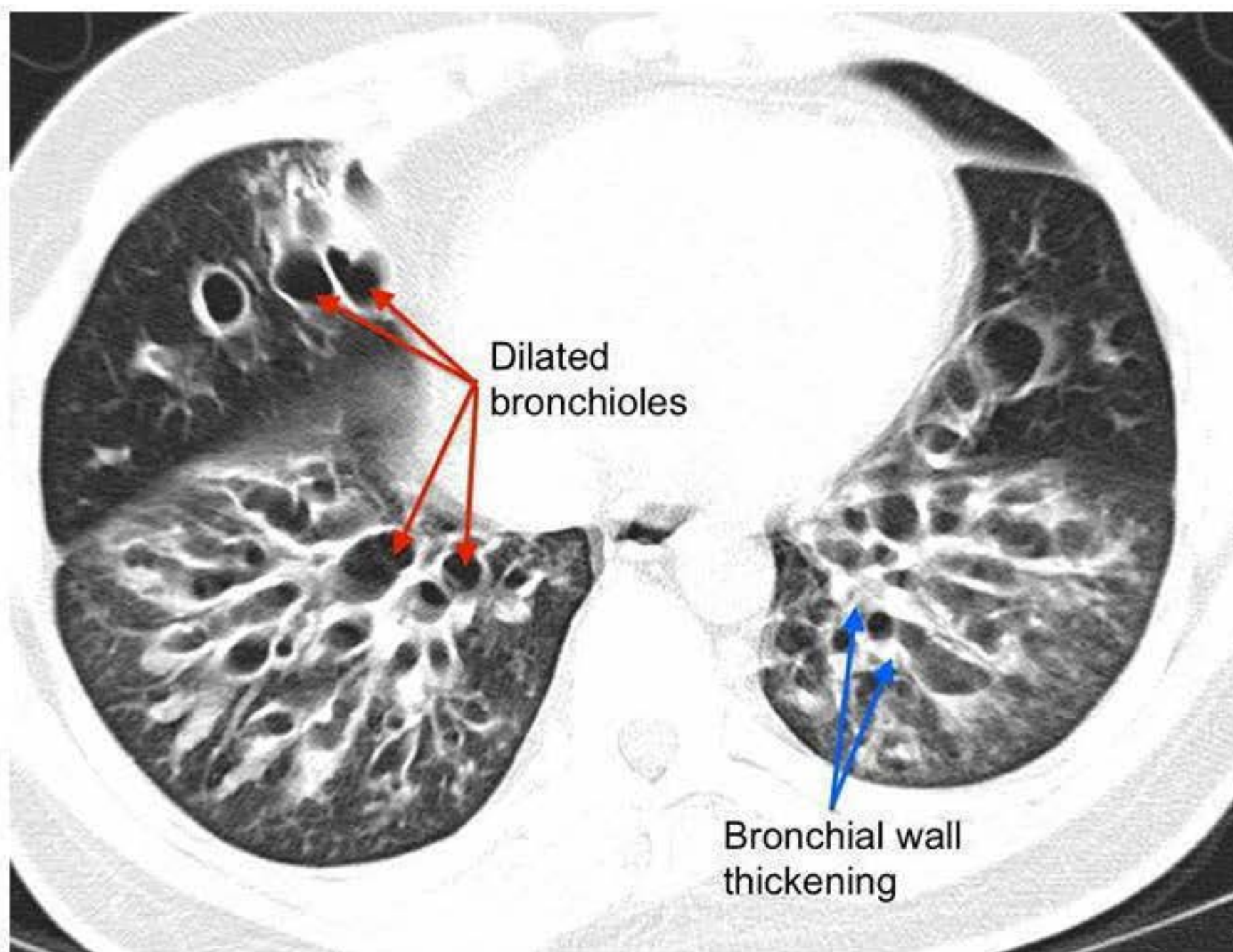
#### References:

1. [Staphylococcus aureus in early cystic fibrosis lung disease.](#)
2. [Changing Epidemiology of the Respiratory Bacteriology of Patients With Cystic Fibrosis.](#)



Media Exhibit

ectasis





Media Exhibit

Fibrosis Features

### Cystic fibrosis features

