

MANAGMENT

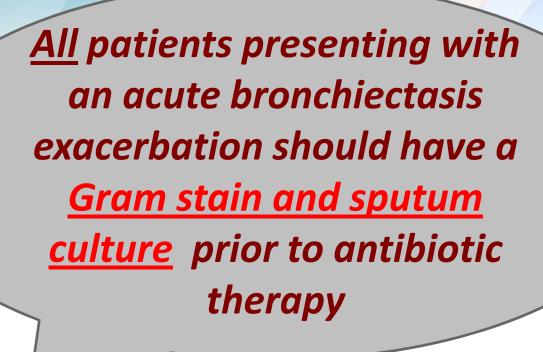
- 1. Antibiotics
- 2. Bronchodilators
- 3. Antiinflammatory Agents
- 4. Airway Clearance Techniques
- 5. Mucolytic Agents and Hydration
- 6. Exercise Training
- 7. Vaccinations
- 8. Immunoglobulin therapy
- 9. gastroesophageal reflux
- 10. Surgery
- 11. Lung Transplantation

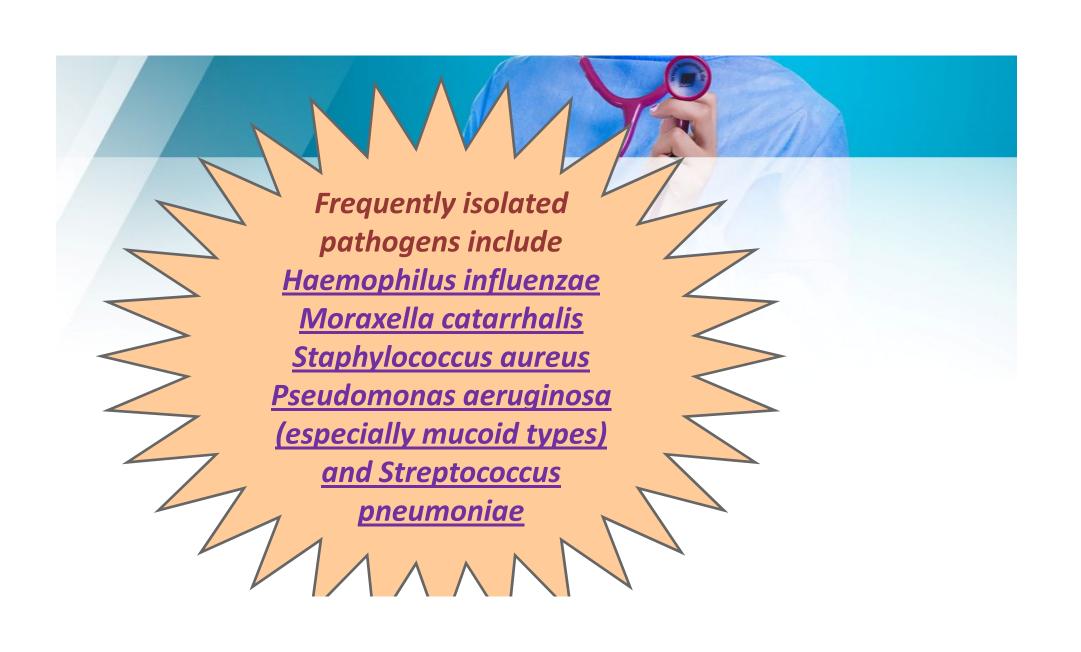












Inpatient



Outpatient



can be treated with an <u>oral</u> antibiotic

Duration of therapy 10 14 days

The initial antibiotic selection is guided by any sputum culture results within the past 12 _24 months

No sputum culture available

sputum cultures
do not show
beta-lactamasepositive H.
influenzae or
Pseudomonas

sputum culture
with
nonpseudomonal
beta-lactamasepositive
organism

Prior sputumgrowing Pseudomonas

fluoroquinolone (levofloxacin, moxifloxacin) amoxicillin 500 mg three times daily or a macrolide

amoxicillinclavulanate, a second or third generation cephalosporin, doxycycline, or a fluoroquinolone

ciprofloxacin, 500 to 750 mg twice daily the efficacy of adding inhaled tobramycin solution (TS) to oral ciprofloxacin has been studied, but it has not been found to be helpful.





Potential indications for inpatient management of bronchiectasis exacerbations

Evidence of severe infection or sepsis
New hypoxemia (oxygen saturation <92%)
Respiratory distress
Tachypnea (respiratory rate ≥25/minute)
High fevers (temperature ≥39°C)
Severe tachycardia (heart rate ≥125/minute)
Hypotension
Altered mental status
Acute fall in urine output or elevation in urea (>20 mg/dL)
Concern for patient frailty
Older age
Multiple difficult-to-manage comorbidities
Malnutrition or frank cachexia
Inability to maintain oral intake

History of exacerbations requiring hospitalization



Need for intravenous therapy without resources for home administration

Concern about adherence to therapy

Mental illness or substance abuse that interferes with management

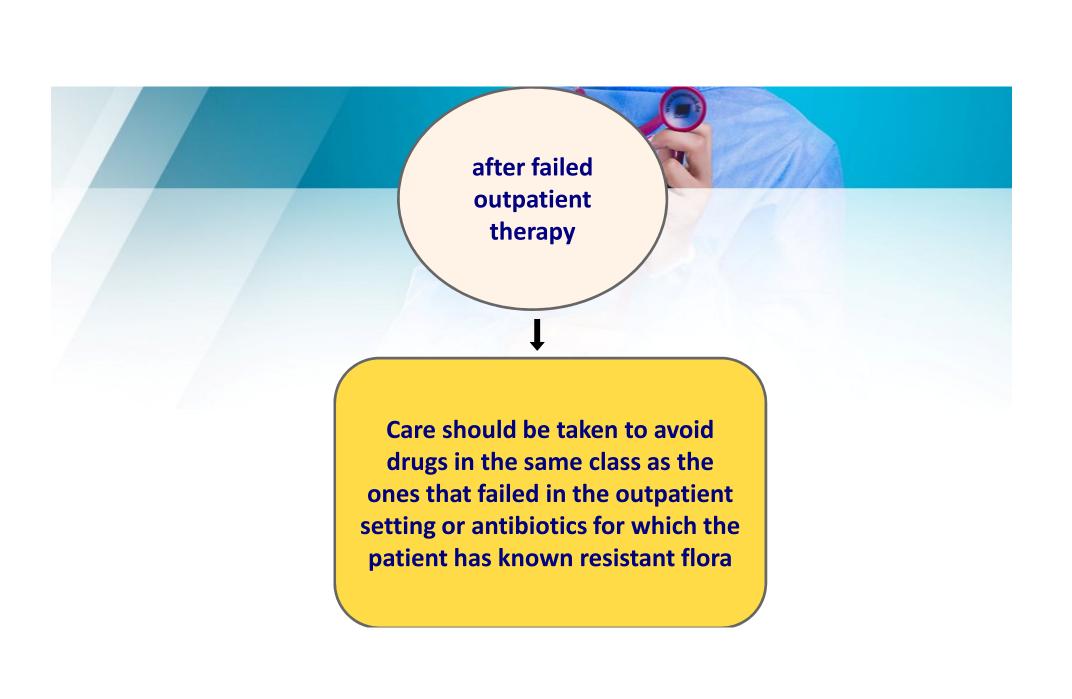
Cognitive or functional impairments

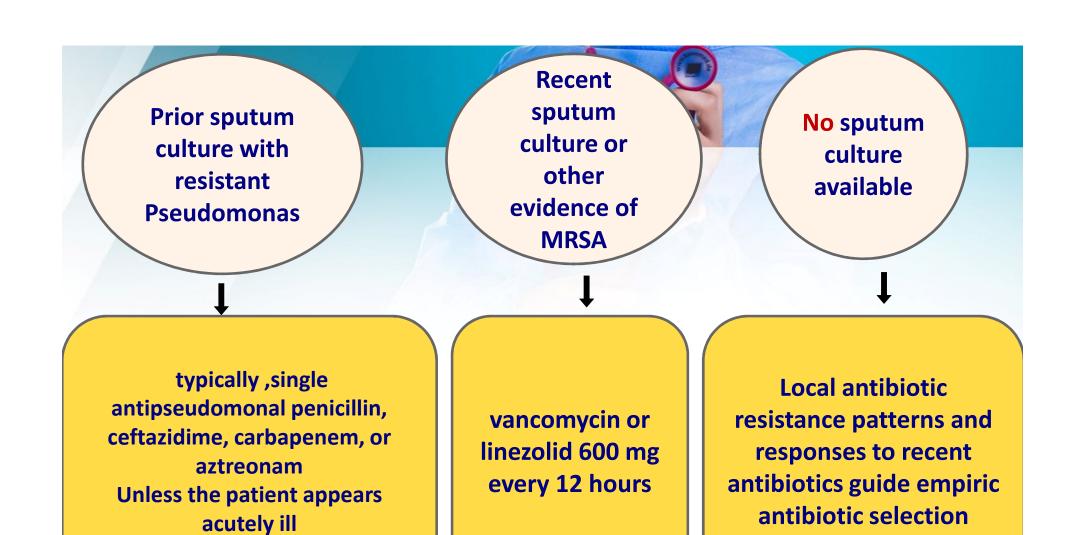
Inability to return for follow-up in the event of clinical worsening

Active hemoptysis

Significant bleeding beyond small-volume or selflimited disease

Clinical worsening despite outpatient treatment







1)Macrolides

500 mg
three times
weekly or
250 mg
once daily
for 6-12
months.

*<u>patients</u> who have recurrent exacerbations and do not have P. aeruginosa infection, we suggest preventive therapy with a macrolide

*For <u>patients</u> with chronic P. aeruginosa in their sputum, inhaled anti- pseudomonal antibiotics are preferred.

*Patients cannot take an inhaled antibiotic and those who continue to have exacerbations despite inhaled antibiotic may benefit from macrolide therapy as an alternative to or in addition to inhaled antibiotic.

Among macrolides, we prefer azithromycin.

2)Nonmacrolide oral antibiotics

amoxicillin 500mg
twice daily,
doxycycline 100
mg twice daily

for patients with three or more exacerbations a year who are not candidates for long-term macrolide administration and do not have airway infection with P. aeruginosa.

3)Inhaled antibiotics

for patients with P. aeruginosa in their sputum and either three or more exacerbations per year or significant morbidity from fewer exacerbations, for patients not infected with P. aeruginosa in whom oral antibiotic prophylaxis is contraindicated, not tolerated, or ineffective.

Inhaled tobramycin:

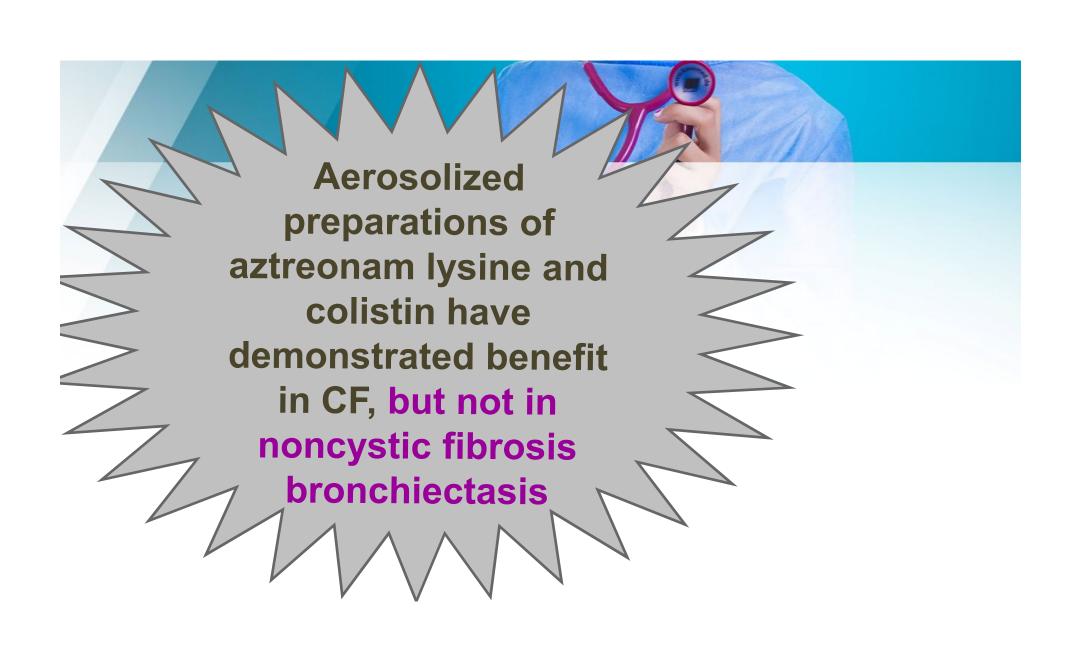
The usual dose for nebulization is ** mg/° mL every ** hours in repeated cycles of ** days on the drug followed by ** days off..

Aerosolized aztreonam lysine:

The dose is 75 mg via eFlow mesh nebulizer three times daily in repeated cycles of 28 days on drug followed by 28 days off drug.

Inhaled colistin:

<u>Aerosolized</u> <u>gentamicin</u>



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2)Airway clearance therapy:

using regular
physiotherapy improve
cough and help patients
to expectorate the
tenacious secretions and
mucous plugs.

Airway Clearance Techniques



Postural drainage and percussion: You sit or lie down in certain positions (postures) to let gravity help mucus drain from different parts of your lungs. Usually someone claps the front or back of your chest (percussion) to loosen and move mucus. This is done in different positions to drain all parts of your lungs.



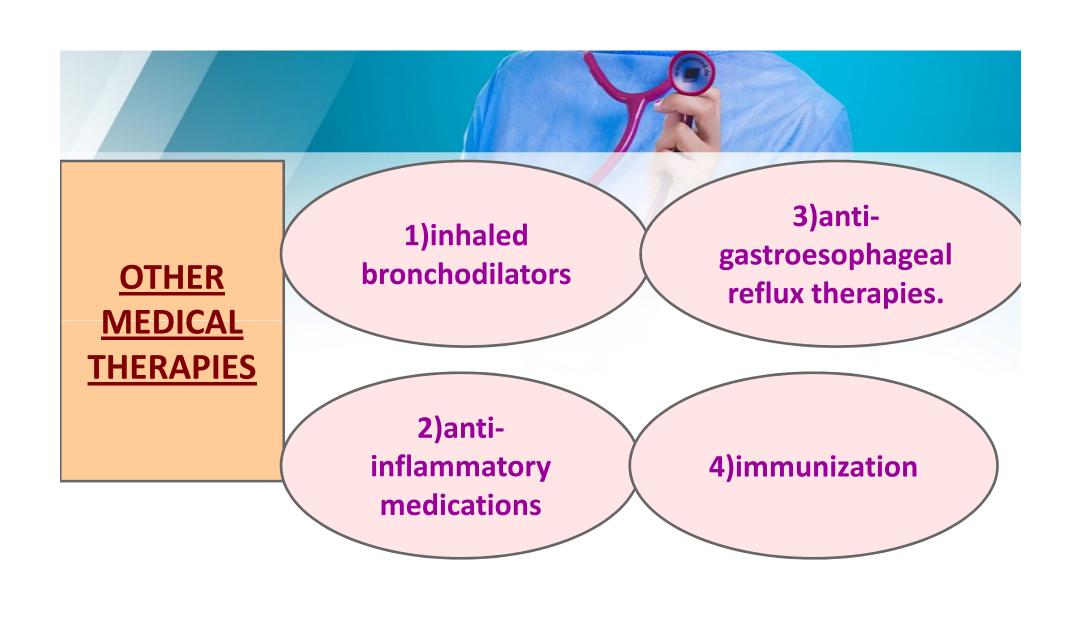
Positive expiratory pressure therapy: A hand-held device creates resistance when you breathe out. This changes pressure in the lungs to loosen mucus



Deep coughing: A deep, controlled cough that is less tiring and more effective in clearing mucus out of the lungs than a "regular" cough.



High-frequency chest wall oscillation: A vest filled with air is attached to a machine that gently squeezes and releases from 5 to 20 times per second. This rapid squeezing and releasing helps vibrate your chest to loosen mucus.



Anti-inflammatory medications

Oral glucocorticoids

for acute exacerbations that are accompanied by wheezing suggestive of concomitant asthma or ABPA In other patients, we avoid it because they depress host immunity, promote bacterial and fungal colonization, and may perpetuate infection.

Inhaled glucocorticoids

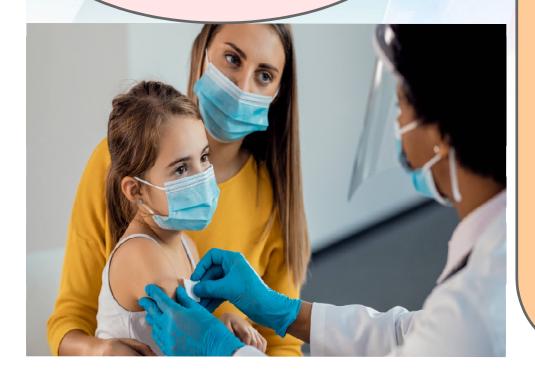
advise against use of inhaled glucocorticoid therapy in the absence of asthma or COPD.

3) Mucolytic agents and airway hydration:

hypertonic ,Inhaled mannitol,Mucolytic agents, (acetylcysteine, Dornase alpha)
Dornase alfa improves pulmonary function (FEV1) and reduces hospitalizations in patients with CF.

4)Systemic hydration: with oral liquids.

Immunizations

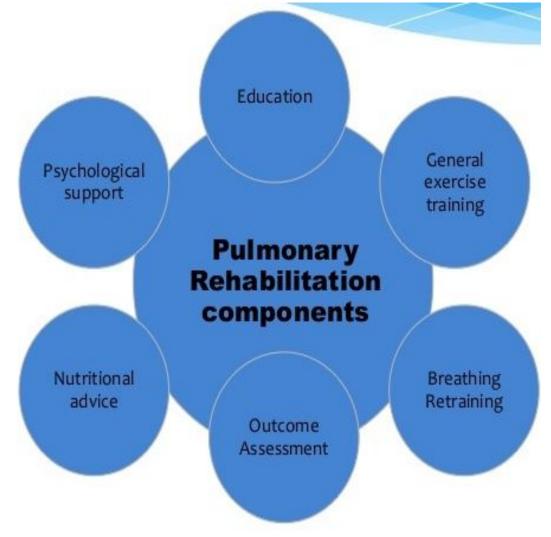


*Seasonal influenza vaccine is administered annually to patients with bronchiectasis, except those unable to receive live attenuated vaccines due to immune deficiency disease.

*Pneumococcal vaccine is typically given to patients with bronchiectasis.

6) pulmonary rehabilitation

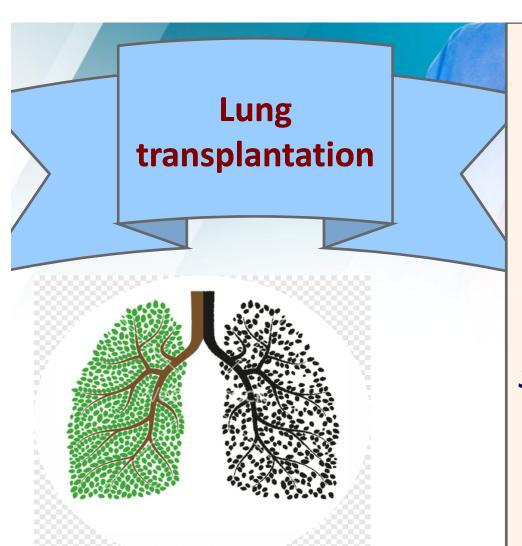






Patient selection

- I. Removal of destroyed lung distal to partial obstruction by a nonmetastatic tumor or the residue of a foreign body.
- II. Recurrent infective episodes due to localized bronchiectasis not responsive to medical therapy.
- III. Overwhelming purulent and viscid sputum production in patients with localized bronchiectasis that is unresponsive to medical therapy.
- IV. Presence of a localized bronchiectatic area suspected of harboring resistant organisms such as (NTM) or multidrug-resistant tuberculosis.
- V. massive hemoptysis (uncontrolled hemorrhage when other measures fail)



The indications for transplant:

1. FEV1<30%.

- 2. severe secondary pulmonary hypertension.
- 3. recurrent massive hemoptysis.
 - 4. ICU admissions requiring noninvasive or invasive ventilation.



Studies

Brensocatib

DPP1 is an enzyme responsible for activating neutrophil serine proteases (NSPs) including neutrophil elastase

Brensocatib is a small molecule, oral, reversible inhibitor of <u>dipeptidyl</u> <u>peptidase 1 (DPP1)</u> being developed by Insmed for the treatment of patients with bronchiectasis and other neutrophil-mediated diseases

WILLOW STUDY



256 patients

87 were assigned to receive placebo,

82 to receive 10 mg of brensocatib,

and 87 to receive 25 mg of brensocatib

subgroups defined according to age, baseline neutrophil elastase concentrations, the number of exacerbations in the 12 months before the trial, the Bronchiectasis Severity Index score, and lung function

Studies

ARIKAYCE

Amikacin
Liposome
Inhalation
Suspension (ALIS)

