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treatment of antibiotic association

PATHOGENS — Chronic bacterial infection within the airways occurs in most patients with CF (table 1), the prevalence of which varies with patient age (figure 2).

Pseudomonas aeruginosa — For reasons that are poorly understood, the CF airway is particularly susceptible to *Pseudomonas aeruginosa*, with infection occurring as early as the first year of life. The prevalence of *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 the *P. aeruginosa* isolated from patients with CF. (See "Epidemiology, microbiology, and pathogenesis of *Pseudomonas aeruginosa* infection".)

Chronic infection with *P. aeruginosa* is an independent risk factor for accelerated loss of pulmonary function and decreased survival [9,10]. Conversion of *P. aeruginosa* to the mucoid phenotype worsens prognosis. Transmissible strains of *P. aeruginosa* have been detected in CF populations in Europe, Australia, and Canada, and some of these strains are associated with a worse prognosis compared with non-transmissible strains [11]. (See "Infection prevention and control" below.)

Staphylococcus aureus — *Staphylococcus aureus* is the bacterial pathogen most frequently identified in respiratory secretions of CF infants and children. It remains a significant pathogen throughout adulthood. In children under six years of age infected with *P. aeruginosa*, co-infection with *S. aureus* has an independent and additive effect on airway inflammation [7].

Methicillin-resistant *Staphylococcus aureus* (MRSA) — Methicillin-resistant *S. aureus* (MRSA) has become more prevalent in the CF population, increasing from 2.1 percent in 1996 to 26 percent in 2009 [8]. Regarding the role of MRSA in patients with CF:

- One study reported that acquisition of MRSA was associated with a slightly greater rate of decline in pulmonary function (as measured by forced expiratory volume at one second [FEV1]) in children, but not in adults [12], while another study reported that MRSA had no effect on the rate of FEV1 decline [13].
- A study of nearly 20,000 CF patients in the United States found that MRSA was associated with 1.3 times the risk of death compared with individuals never infected with MRSA [14]. Multivariate analysis showed that MRSA was an independent risk factor whose effect could not be explained by its association with other known risk factors including age, sex, diabetes, pancreatic status, FEV1 at baseline, and socioeconomic status, or co-infection with *Burkholderia cepacia* complex or *P. aeruginosa*.

***Burkholderia cepacia* complex** — Advances in bacterial genetics have revealed that *B. cepacia*, which was originally thought to be a single species, is now known to constitute multiple separate species, each of which is a member of the *B. cepacia* complex [15]. The species most commonly isolated from the sputum of CF patients are *Burkholderia multivorans* and *Burkholderia cenocepacia* [16-18].

Chronic infection with *B. cepacia* complex bacteria is associated with an accelerated decline in pulmonary function and shortened survival in CF [19-21]. Lung transplantation in patients infected with *B. cepacia* complex is associated with recurrent and often severe infection with poor outcomes, particularly for those carrying *B. cenocepacia* [22-24]. Infection with *B. cenocepacia* is considered to be a contraindication to transplantation in many centers. (See "Bacterial infections following lung transplantation", section on "Burkholderia cepacia".)

Other pathogens — Other pathogens have been identified in respiratory secretions of CF patients, with varying prevalence (table 1 and figure 2) [25]. These include:

- Non-infectious *Aspergillus* spp.

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