# *Pseudomonas cepacia* in cystic fibrosis

OVER THE PAST DECADE, *PSEUDOMONAS CEPACIA* HAS BECOME a cause of great concern in patients with cystic fibrosis. Among the many questions about infection with *P cepacia* are:

- Is *P cepacia* the direct cause of the 'fulminant cepacia syndrome' (1)? This uncommon syndrome is quite unlike the frequent exacerbations of chest symptoms seen in carriers of *Pseudomonas aeruginosa*. It consists of high, spiking fever, severe respiratory distress, marked leukocytosis, elevated erythrocyte sedimentation rate and necrotizing pneumonia with a high case fatality rate.
- If *P cepacia* is the cause of the fulminant syndrome, then which antibiotics should be used for treatment, given the resistance of *P cepacia* to many antibiotics (2)?
- Does chronic infection with *P cepacia* increase the rate of lung damage and accelerate the deterioration in pulmonary function (3)?
- Why has the prevalence of chronic carriage of *P cepacia* increased to 20 to 30% in some cystic fibrosis centres in Canada and the United States and decreased in other centres?
- What is the risk of person-to-person spread between carriers and noncarriers of *P cepacia* during hospitalization, clinic visits, and social interactions (4-6)?
- Is the risk of person-to-person spread sufficiently high to require stringent isolation and cohorting of those infected with *P cepacia*?
- If person-to-person spread is not the major source of infection, what are the reservoirs from which patients with cystic fibrosis acquire *P cepacia*?

Definitive answers are not available for these and many other questions concerning *P cepacia* in cystic fibrosis. The statement from the Medical/Scientific Advisory Committee of the Canadian Cystic Fibrosis Foundation entitled the *Epidemiology of Pseudomonas cepacia in cystic fibrosis* in this issue of *The Canadian Journal of Infectious Diseases* (page 163) provides not only an excellent summary of the current state of knowledge, but also points out why *P cepacia* has caused such concern among patients with cystic fibrosis and their families. Based on their review of what is known and not known about the epidemiology of *P cepacia*, the committee has made recommendations on how best to prevent person-to-person spread of *P cepacia*. Their recommendations seem eminently sensible and sufficient to minimize whatever the risk of such transmission may be without creating unnecessary hardship to carriers.

The significance of the statement rests not only on the recommendations on infection control relative to *P cepacia*. Equally important is the clear delineation of the research questions which must be answered in order to determine how *P cepacia* is acquired, what factors determine its incidence and prevalence, and how *P cepacia* causes lung damage in patients with cystic fibrosis. The statement concludes with a call to all those involved in the care of patients with cystic fibrosis to collaborate voluntarily in the first step of a Canada-wide study of *P cepacia* in establishing a central repository of *P cepacia* strains. If cystic fibrosis centres across Canada do heed this request, the creation of such a collection will be invaluable in learning more about the epidemiology of *P cepacia*.

A second article in this issue (page 166) describes a second, essential precondition of any nationwide study of *P cepacia*: the introduction, into all microbiology laboratories associated with cystic fibrosis centres, of optimal methods for microbiological processing of respiratory specimens from patients with cystic fibrosis. One of the problems which has delayed acquisition of knowledge of the epidemiology of *P cepacia* has been the lack of standardization of microbiological methods and the delayed recognition of the specialized methods required to detect *P cepacia* in sputum from patients with cystic fibrosis. Recognition of the need for such standardization of methods by all microbiology laboratories involved with cystic fibrosis clinics is the first and essential step. As pointed out in the article, it will also be necessary to establish accreditation procedures and proficiency testing programs so that the laboratories will be able to provide quality control of their ability to identify *P cepacia* and other pathogens in respiratory secretions of cystic fibrosis patients.

## REFERENCES


1. Isles A, Maclusky I, Corey M, et al. *Pseudomonas cepacia* infection in cystic fibrosis: An emerging problem. J Pediatr 104;206-10.
2. Gold R, Jin E, Levison H, et al. Ceftazidime alone and in combination in patients with cystic fibrosis: Lack of efficacy in treatment of severe infections caused by *Pseudomonas cepacia.* J Antimicrob Chemother 1983;11:331-6.
3. Tablan OC, Maretone WJ, Doershuk EF, et al. Colonization of the respiratory tract with *Pseudomonas cepacia* in cystic fibrosis: Risk factors and outcomes. Chest 1987;91:527-32.
4. LiPuma JJ, Dasen SE, Nielson DW, et al. Person-to-person transmission of *Pseudomonas cepacia* between patients with cystic fibrosis. Lancet 1990;336:1094-6.
5. Tablan OC. Nosocomially acquired *Pseudomonas cepacia* infection in patients with cystic fibrosis. Infect Control Hosp Epidemiol 1993;14:124-6.
6. Burdge DR, Nakielna EM, Noble, MA. Case-control and vector studies of nosocomial acquisition of *Pseudomonas cepacia* in adult patients with cystic fibrosis. Infect Control Hosp Epidemiol 1993;14:127-30.



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