THE EPIDEMIOLOGY OF PSEUDOMONAS CEPACIA IN PATIENTS WITH CYSTIC FIBROSIS

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Pseudomonas cepacia has emerged as an important nosocomial pathogen colonizing and infecting the respiratory tract of patients with cystic fibrosis (CF). Although assessment of outcomes associated with P. cepacia colonization has been difficult, controlled studies have shown that colonized patients experience more adverse outcomes compared with those not colonized. In the United States, an increasing trend in national incidence and prevalence of P. cepacia colonization has been shown, but cases have been unevenly distributed in a few centers. These estimates, however, may be biased by intercenter differences in laboratory methods for detecting P. cepacia in patient sputum. The source and mode of transmission of P. cepacia have not been adequately demonstrated, and may vary from center to center. Until further studies elucidate the epidemiology of P. cepacia in patients with CF, it may be prudent for CF centers to consider the use of selective media to isolate P. cepacia from sputa of patients with CF, to conduct investigations of clusters of P. cepacia-colonized patients, and to consider adopting infection control precautions recommended for control of multiply resistant gram negative organisms.

INTRODUCTION

Cystic fibrosis (CF) is a genetic disorder characterized by chronic pulmonary disease, pancreatic insufficiency, and increased concentration of electrolytes in sweat (26, 56, 79). Patients with CF develop progressive pulmonary obstruction and recurrent infections which culminate in their premature death. Various microbial agents have been implicated in causing pulmonary exacerbations in patients with CF (17, 31, 48, 65, 70, 78). However, S. aureus and P. aeruginosa have been the most prevalent bacterial isolates in studies of the pulmonary microbial flora in CF, especially those involving cultures of lung tissue just before patient death or at autopsy (14, 26, 29, 30, 32, 35, 53). The recognition of P. cepacia as a significant respiratory pathogen in patients with CF has been recent. Before 1980, reports of isolation of P. cepacia from respiratory secretions of patients with CF were rare (7, 62). Although Laraya-Cuasay et al. reported in 1977 that P. cepacia was isolated from 6.3% of 6364 cultures of respiratory secretions of patients with CF at St. Christopher's Hospital for Children (SCHC), Philadelphia, during 1973-1976 (46), its significance was unknown. In 1984, however, Isles et al. at the Hospital for Sick Children (HSC) in Toronto, Canada, published their 10-year experience with P. cepacia and reported that the prevalence of P. cepacia

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in their CF patients almost doubled, from 10% in 1971 to 18% in 1981 (36). In the same report, the authors described a syndrome of rapid clinical deterioration and death, perceived to be unique to some patients colonized or infected with P. cepacia.

Epidemiologic investigations conducted by the U.S. Centers for Disease Control (CDC) in 1983 and 1984 at 2 hospitals (SCHC [51] and Rainbow Babies and Children’s Hospital (RBCH) in Cleveland) reporting >5% annual incidence of P. cepacia colonization, confirmed the strong association between P. cepacia and CF (72, 73). At SCHC, of 339 patients with CF who had sputum cultures from 1979-1983, 61 were positive for P. cepacia; in contrast, only 5 of 1425 non-CF patients with sputum cultures done during the same time period were culture-positive for P. cepacia (p<0.0001, Fisher’s exact test [FET] one-tail (72). Similarly, at RBCH, between 1981 and 1983, 104 of 519 patients with CF were colonized with P. cepacia but none of about 1000 non-CF patients had P. cepacia in cultures of their respiratory secretions, p<0.0001, FET one-tail (73).

In addition, a survey of 126 U.S. CF centers in 1985 revealed that the percentage of centers detecting P. cepacia in respiratory secretions of CF patients had increased from 21% in 1981 to 42% in 1984 (CDC unpublished data). Overall, the increase was seen in all groups of centers categorized by size, but in each year, a higher proportion of the larger centers (>200 patients) recovered P. cepacia from the sputa of their patients with CF (Figure 1).

Incidence and Prevalence

The overall incidence of P. cepacia colonization in the United States, estimated from the above survey, increased from 3 per thousand in 1981 to 6 per thousand in 1984; prevalence increased from 11 per thousand in 1981 to 16 per thousand in 1984 (Figure 2). However, wide variations in size-specific and center-specific incidences were seen (Figure 3). Less than 2% of centers were reporting incidence of >5%. In any single year, new cases were reported from only 13%-30% of 126 centers, mostly from a few centers with >200 patients. Of 110 centers with ≤200 patients, no new cases were reported in 90% in 1981, 92% in 1982, 84% in 1983, and 74% in 1984; of 16 centers with >200 patients, no new case was reported in 75% in 1981, 70% in 1982 and 1983, and 65% in 1984. This uneven distribution

![Figure 1](image1.png)

Figure 1. - Percentage of CF Centers isolating P. cepacia from sputum cultures of patients with CF, 1981-1984, by CF Center Size.

![Figure 2](image2.png)

Figure 2. - Overall incidence and prevalence of P. cepacia colonization of CF patients 1981-1984.

![Figure 3](image3.png)

Figure 3. - Incidence of P. cepacia colonization of CF patients, by CF Center Size, 1981-1984.