SHORT REPORT

Pneumothorax in cystic fibrosis: a retrospective case series

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Pneumothorax is a known complication in cystic fibrosis (CF), associated with poor outcome. Records of CF patients with pneumothorax at the Royal Children's Hospital, Melbourne between 1990 and 2004 were reviewed, and the characteristics, sputum culture results, lung function, treatment, and outcome for the 11 patients who had pneumothoraces were described.

Pneumothorax as a complication in cystic fibrosis (CF) was first reported in 1962, when Bernard *et al* found six patients with pathological sweat tests among 14 individuals with spontaneous pneumothorax.[1] In this study we describe the characteristics of CF patients at the Royal Children's Hospital (RCH), Melbourne who were diagnosed with pneumothoraces over a 15 year period.

METHODS AND RESULTS

We reviewed all medical records at the RCH with a diagnosis of CF and pneumothorax between January 1990 and December 2004. We excluded patients with pneumothorax post-thoracic surgery, including lung transplantation. Recurrence of pneumothorax was defined as a pneumothorax that occurred on the ipsilateral side >7 days after complete radiological resolution of the first pneumothorax.

Presence of *Pseudomonas aeruginosa* was recorded for all patients with these data extracted from the medical records. The sputum results were also examined for other bacteria, including *Burkholderia cepacia*. In addition, presence of Melbourne epidemic *P aeruginosa* (*MPA*) was recorded for all records after 1999, as surveillance was only introduced after this time. Sputum results for *MPA* were extracted from the specific *MPA* database. Lung function tests were obtained using a Jaeger body plethysmograph; results were obtained for each patient as close as possible to one year prior to the first pneumothorax (median days (range): 370 (90–423)) and as close as possible to one year after the first pneumothorax (median days (range): 328 (5–400)).

There were approximately 500 children living in the area of Victoria, Australia who had CF between 1990 and 2004 who were cared for at the RCH. In this time period there were 4446 admissions to the RCH due to CF related problems. For 15 of these admissions there was a diagnosis of one, and for one of these admissions, a diagnosis of two pneumothoraces. In total there were 17 pneumothoraces experienced by 11 patients. The characteristics, sputum culture results, lung function, treatment, and outcome of the patients are shown in table 1, with details for each pneumothorax. Spirometry results are summarised in table 2.

The most recent first presentation of a pneumothorax occurred in September 2000. The median length of follow up after last pneumothorax was 569 days (range 9–2555 days). The mean age (SD; range) of first pneumothorax was 16.6 years (2.3; 12.7 to 19.6 years). Ten of the 11 subjects were positive for PA.

DISCUSSION

The prognosis after a spontaneous pneumothorax was poor in our cohort. Flume's analysis revealed that a pneumothorax increases the probability of dying within two years.

The findings of no first presentation of a pneumothorax in our clinic since September 2000 might be due to the availability of new treatments, such as inhaled tobramycin, or the introduction of the newborn screening programme, resulting in a shift of the first presentation of a pneumothorax towards the adult group of CF patients.

In contrast, Flume *et al* reported an association of pneumothorax and inhaled tobramycin with an odds ratio of 1.6, probably due to an acute drop in FEV_1 after inhaling tobramycin, although there have been no reported cases of pneumothorax in CF patients following nebulised therapy.[2] However, it is known that *P aeruginosa* causes excessive inflammation in CF, which may settle into a vicious cycle of airway obstruction, infection, and excess inflammation, resulting in alveolar hyperinflation and lung destruction. Structural changes may then lead to increased transpulmonary pressure differences, resulting in lung rupture into the pleural space. It might be therefore that the findings by Flume *et al* have been confounded by its use in a more severe patient group.

Newborn screening was introduced in the RCH in 1989 and the first of these children are now in their teenage years. It might therefore be the case that early diagnosis of CF may preserve the lung structure. A comparative study however between a screened population and a CF cohort based on clinical diagnosis would be necessary to address this hypothesis more in detail.

The mean rate of decline of $\%FEV_1$ over the two year period around the pneumothorax was 6% predicted/year. This rate of decline is slightly higher in comparison to an average rate of decline of $\%FEV_1$ in our CF Unit of approximately −4% predicted/year. But as decline in lung function appears to be closely related to *P aeruginosa* and *B cepacia* colonisation, it might be therefore more likely to be due to infecting organisms then occurrence of a pneumothorax. However, pneumothorax occurred more often in moderate to severe lung disease (table 1) in this study, though it may occur in mild lung disease. But numbers were too small to reliably assess a comparison with appropriately matched (age, sex, microbiology, year of pneumothorax, medical treatment) controls.

Most of the CF patients with pneumothorax in this study were positive for *P aeruginosa*. Flume *et al*, in the analysis of the US Cystic Fibrosis Foundation (CFF) Patient Registry database, recently reported that presence of *P aeruginosa* is associated with a twofold increase in the odds of experiencing a pneumothorax.[2] In addition, different CF clinics, including our unit at the RCH, have recently reported the detection of epidemic *P aeruginosa* strains, but with different results regarding the clinical outcome and without specific analysis regarding pneumothoraces.

On average 300 different CF patients consulted the RCH each year between 1990 and 1998. Data extracted from the CF database revealed that the proportion positive for PA in each year ranged from 16% to 26%. Sputum culture results

Table 1 Characteristics of CF patients with pneumothoraces

Case	Sex	Age at diagnosis (years)	Sputum cohort	Localisation (time between Px)	FEV_1 before Px (% pred)	FEV_1 after Px (% pred)	Therapy for each pneumothorax	Outcome (time after first pneumothorax)
1	M	16	*MPA+*	Left	39%*	30%	Thoracoscopy with ligation of cyst and pleurodesis	Lung transplantation (2555 days)
2	F	18	*MPA+*	Right	53%*	Missing	Initial chest tube, followed by thoracoscopy with apex oversew	Death (9 days)
3	F	19	*MPA+*	Right	61%*	52%	Initial chest tube, then thoracoscopy with pleurodesis	Lung transplantation (1640 days)
4	M	15	*PA+ (MPA−)*	1. Left 2. Left (12 days)	45%*	40%	1. Chest tube 2. Chest tube, followed by thoracoscopy and stapling of pleural bleb	Death (569 days)
5	F	16	*PA + (MPA−)*	Left	50%*	Missing	Chest tube	Death (271 days)
6	F	15	*B cepacia*	1. Right 2. Right (34 days) 3. Left (1825 days)	48%*	38%	1. Chest tube, followed by thoracoscopy and pleurodesis 2. Observation 3. Pleurodesis	Alive (2095 days)
7	M	12	*PA+*	Bilaterally	45%*	33%	Bilaterally chest tubes	Death (791 days)
8	F	13	*PA+*	1. Left 2. Right (32 days)	37%*	Missing	1. Chest tube 2. Chest tube	Death (43 days)
9	M	18	*PA+*	1. Right 2. Right (66 days)	61%*	Missing	1. Observation 2. Chest tube	Death (82 days)
10	M	18	*PA+*	Right	73%*	54%	Chest tube	Alive (2400 days)
11	F	19	*PA+*	Left	64%*	33%	Needle aspiration	Lost to follow-up after 270 days

PA−, negative for *P aeruginosa*; *PA+*, positive for *P aeruginosa*; *MPA−*, negative for *MPA*; *MPA+*, positive for *MPA*; Px, pneumothorax.
MPA status recorded for cases 1 to 5 only.
*CF lung disease was classified into mild (FEV_1 ⩾66% predicted), moderate (FEV_1 46–65% predicted), and severe (FEV_1 ⩽45% predicted).

Table 2 Spirometry results

Measurement	Before Px (n = 11)	Time of Px (n = 8)	After Px (n = 7)
FEV_1, l mean (SD; range)	1.8 (0.46; 0.77 to 2.38)	1.4 (0.93; 0.28 to 3)	1.2 (0.3; 0.82 to 1.56)
FEV1 % pred mean (SD; range)	52.4 (11.2; 37 to 73)	38.6 (11.8; 15 to 52)	40.0 (9.5; 30 to 54)
FVC, L mean (SD; range)	2.3 (0.82; 1.16 to 4.2)	1.9 (0.51; 1.28 to 2.9)*	2.0 (0.51; 1.5 to 2.88)
FVC % pred mean (SD; range)	71.7 (13.5; 53 to 100)	57.8 (10.9; 44 to 74)*	58 (17.1; 36 to 90)

* n = 7 (FVC missing in one individual).
Px, pneumothorax.
The median (range) rate of decline in FEV_1 per year was 6% predicted (2.7 to 16.4) for the seven subjects with complete spirometry results.

for patients consulting after introduction of surveillance in 1999 for *MPA* (as well as *P aeruginosa*) revealed that for the years 1999, 2002, and 2004, 36%, 31%, and 33%, respectively, were positive for *P aeruginosa* and 21%, 14%, and 11%, respectively, were positive for *MPA*. It is possible that CF patients with pneumothoraces are more likely to have had *P aeruginosa* than their counterparts who have not. Because we do not know the number of person-years at risk that each CF patient contributed to these data it is not possible to formally compare the rate of *PA* between patients who went on to have a pneumothorax and those that did not.

The collapsed lung can be stiff and take a long time to re-expand.[3] The initial lack of response to treatment such as observation, needle aspiration or chest tube, resulted in an overall failure rate (initial failure plus recurrence) of 50% in our study. These findings support the idea of an early, more definite procedure such as surgical or chemical pleurodesis, and are reflected in the current recommendation of surgical intervention after the first episode of a pneumothorax in those patients fit to undergo surgery.[3 4]

To prevent a pneumothorax, patients with CF should avoid manoeuvres which will create marked fluctuations in intrapleural pressure, such as weight lifting. In the recovering phase, air travel should be avoided for at least two weeks following complete resolution of a pneumothorax.[5]

In conclusion, our results implicate that an aggressive therapeutic approach for *P aeruginosa* infection may prevent the occurrence of a pneumothorax, a clinical feature that per se reduces the life expectancy in those with CF.[2] For the paediatric group of patients, this finding supports the importance of early detection and treatment of *P aeruginosa* infection.

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