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Short communication

What defines a pulmonary exacerbation? The perceptions of adults with cystic fibrosis

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Abstract

Background: There is no standardised definition of a pulmonary exacerbation in cystic fibrosis (CF). In attempting to achieve standardised criteria it is important to identify patient-reported indicators.
Methods: Interviews were undertaken with 47 adults with CF. Participants were asked to report symptoms experienced during a pulmonary exacerbation in two ways: the first symptoms they become aware of, and how they subsequently recognised when they were improving.
Results: A range of systemic and respiratory symptoms were reported. Their relative importance varied by severity of disease. The severity and subsequent improvement of an exacerbation was often described as limitations on their activities.
Conclusion: These preliminary data suggest that patient-reported indicators of a pulmonary exacerbation may not be the same for all adults with CF. Whether different indicators are associated with specific demographic or clinical variables remains to be evaluated.

Keywords: Pulmonary exacerbation; Respiratory symptoms; Patient-reported outcome; Qualitative data

1. Introduction

In cystic fibrosis (CF) pulmonary exacerbations are a key outcome measure of disease, clinical research and care. They are associated with a poor health-related quality of life [1,2] disease progression [3] and survival [4]. Despite the importance of identifying a pulmonary exacerbation, there is no standardised definition in CF and there is a lack of consensus among clinicians. Standardised and valid criteria are important for an accurate diagnosis of a pulmonary exacerbation, and the determination of severity [5], so that (1) appropriate treatments can be initiated and (2) a pulmonary exacerbation can be considered a meaningful outcome measure in interventions.

Recent reviews [6,7] have noted that three research groups [8–10] have indicated that patient-reported symptoms were more predictive of a pulmonary exacerbation than physical examination and laboratory data and argue that diagnostic criteria should focus on signs and symptoms. Scales that have combined patient-reported signs and symptoms, clinical evaluations and laboratory results have been developed for clinical trials but their validity, reliability and sensitivity have not been evaluated [11,12].

As part of the process of achieving standardised criteria it is important to clearly identify patient-reported indicators of a pulmonary exacerbation for different levels of disease severity. This work aimed to identify the salient characteristics of a pulmonary exacerbation as reported by adults with cystic fibrosis, in order to contribute to the ongoing discussion on defining a pulmonary exacerbation in CF disease.

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2. Methods

2.1. Sampling

Adults (age 16 years +) with cystic fibrosis were recruited from two CF Centres in the UK. Purposive sampling ensured representation of key variables including age, gender, in/out-patient, small CF Unit/specialist CF Centre (treating > 50 patients), FEV1% predicted, BMI, microbiology, number of clinically recorded exacerbations in past year, CF related diabetes and nutritional and transplant status. Sixty people were invited to take part in the study. All patients had previous experience of pulmonary exacerbations but not all patients were experiencing a PE at the time of interview. Those experiencing a PE were interviewed towards the end of treatment. All exacerbations had been diagnosed by a CF physician and defined clinically as an increase in respiratory symptoms (cough, volume and colour of sputum, or breathlessness) associated with a fall in spirometry compared to usual values for the patient. The study was approved by the North Staffordshire Local Research Ethics Committee.

2.2. Interviews

Interviews were undertaken by a single researcher (A Holt). Participants were asked to report symptoms experienced during a pulmonary exacerbation in two ways: the first symptoms they become aware of, and how they subsequently recognised when they were improving. Voluntary reported symptoms were elicited from the patients to encourage them to describe their symptoms in their own words.

2.3. Conceptual approach and data analysis

A grounded theory approach utilizing constant comparative analysis [13] was employed. The interview schedule was amended as new concepts emerged from ongoing analysis to ensure focused and appropriate questioning. Interviews were taped, transcribed verbatim and the data analysed thematically. Data collection and coding took place concurrently. Line by line coding of the transcripts was undertaken. Codes were attached to specific words, phrases, or sentences. These codes were then grouped into conceptual categories [14,15]. This was done independently by two researchers (JA, A Holt) and the coding of concepts and categories agreed following discussions.

Descriptions of symptoms were investigated for different disease severity groups: mild disease (FEV_1 > 70%), moderate disease (FEV_1 = 40–70%) and severe disease (FEV_1 < 40%).

3. Results

Forty-seven adults with cystic fibrosis agreed to be interviewed. Thirteen people declined (lack of time/had to get back to work = 5; didn't want to talk about CF = 5; no reason given = 3). The demographic and clinical characteristics of those who declined were similar to those interviewed. The characteristics of the sample are given in Table 1.

A range of systemic and respiratory symptoms were experienced during a pulmonary exacerbation. Descriptions of symptoms were similar regardless of whether or not the person was being treated for an exacerbation at interview. There is some evidence that the first symptoms patients became aware of varied by severity of disease (Table 2). For many patients the onset of an exacerbation was characterised by fatigue and changes in sleep, cough, sputum, appetite, mood and daily activities. Those with mild disease typically reported 'cold' symptoms whereas those with severe disease found it more difficult to recognise the onset of an exacerbation. They typically reported greater levels of fatigue (e.g., sleeping during the day), greater effort required to cough and breathe, nausea/vomiting (related to sputum/cough) and chest pain. For some (especially those with severe disease) it was easier to describe improvements in symptoms following treatment rather than symptoms at the start of a pulmonary exacerbation. They often did not realize how far they had deteriorated until they started to notice an improvement.

There was more consistent reporting of symptoms across severity groups when describing improvements from an exacerbation. An improvement was primarily described in terms of the activities they were able to perform (e.g., return to work, shopping) followed by improvements in fatigue, sleep, sputum, cough and mood (Table 3). Those with severe disease

Table 1
Characteristics of the sample (n = 47).

	Mild (FEV_1 % > 70) N = 12	Moderate (FEV_1 % 40–69) N = 18	Severe (FEV_1 % < 40) N = 17	All patients N = 47
FEV_1 % (range)	89.8 (72–105)	56.1 (41–68)	29.7 (15–39)	54.5 (15–105)
Age (years, range)	24.0 (17–35)	27.2(17–47)	26.9 (19–34)	26.8 (17–47)
BMI (range)	21.7(19.2–23.6)	21.9 (18.1–29.3)	19.0 (14.4–22.7)	21.0 (14.4–29.3)
Recorded exacerbations in past year (range)	1.3 (0–3)	2.6 (0–6)	3.5 (0–9)	2.6 (0–9)
Diabetes (n)	2	6	11	19
Enteral tube feeds (n)	1	1	8	10
Oral supplements (n)	2	8	10	20
Venous access device (n)	4	8	14	26
P. aeruginosa (n)	4	12	17	33
B. cepacia complex (n)	0	2	2	4
Inpatients (n)	3	6	16	25
Specialist CF Centre [treating > 50 patients] (n)	6	11	14	31

Mean FEV1, age, BMI and exacerbations and numbers of people with key variables for each disease severity group.