Journal of Cystic Fibrosis 11 (2012) 180–186

Journal of Cystic Fibrosis

www.elsevier.com/locate/jcf

Original Article

Patient indicators of a pulmonary exacerbation: Preliminary reports from school aged children map onto those of adults☆

J. Abbott [a,*], A. Holt [a], A.M. Morton [b], A. Hart [c], G. Milne [d], S.P. Wolfe [e], S.P. Conway [b]

[a] School of Psychology, University of Central Lancashire, Preston, PR1 2HE, UK
[b] Adult Cystic Fibrosis Unit, St. James' Hospital, Leeds, LS9 7TF, UK
[c] School of Medicine and Health, University of Lancaster, LA1 4YW, UK
[d] Children's Cystic Fibrosis Unit, Ninewells Hospital, Dundee, DD1 9SY, UK
[e] Paediatric Cystic Fibrosis Unit, Leeds General Infirmary, Leeds, LS1 3EX, UK

Received 5 September 2011; received in revised form 9 November 2011; accepted 19 November 2011
Available online 14 December 2011

Abstract

Background: Despite the importance of identifying and managing a pulmonary exacerbation, and its use as an outcome measure in interventions, there is no standardised definition in cystic fibrosis. In achieving standardised criteria it is important to identify patient-reported indicators.
Methods: Interviews were undertaken with 35 school aged children. They reported symptoms experienced during a pulmonary exacerbation in two ways: the first symptoms they become aware of, and how they recognised when they were improving. Interviews were taped, transcribed verbatim and the data analysed thematically.
Results: For many children, the onset of an exacerbation was characterised by 'cold' symptoms, tiredness, and changes in cough. For those with moderate or severe disease, sleep disruption, activity induced breathlessness, changes in mood, sputum volume and lack of appetite were common. When describing improvement children focused initially on activities they were now able to perform accompanied by improvements in tiredness and cough. Those with moderate or severe disease also reported improvements in sleep and mood, breathlessness, sputum volume and colour.
Conclusions: Child-reported indicators of a pulmonary exacerbation tend to map onto those reported by adults. These results provide the rationale for the development of a single scale for school age children and adults that could be sensitive to progressive stages of CF disease.

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

1. Introduction

Pulmonary exacerbations (PE) are a prominent characteristic of cystic fibrosis (CF) disease. They are associated with a poor health-related quality of life [1,2] disease progression [3] and survival [4]. Therefore, it is essential that exacerbations of pulmonary symptoms are recognised by the patient, diagnosed and the severity determined by the clinician [5], early appropriate treatment initiated, and the course of the exacerbation and response to treatment assessed.

Despite the importance of identifying and managing a pulmonary exacerbation, and its use as an outcome measure in interventions, there is no standardised definition in CF. In an Australian survey there was a lack of consensus among clinicians who managed CF patients as to the variables considered when diagnosing a pulmonary exacerbation. Clinicians agreed with only half of the listed signs, symptoms and investigations derived from definitions used in published research studies [6]. Furthermore, when diagnosing a PE clinicians who managed both children and adults rated over 20% of signs and/or symptoms as having different levels of importance [6].

Some attempts have been made to establish clear criteria in CF with the use of scoring systems [7–12] but there is a lack of consistency across scales [6,13,14]. From a combined pool

☆ Data presented at the 33rd ECFS Conference, Valencia, Spain (2010).

* Corresponding author at: School of Psychology, University of Central Lancashire, Preston, PR1 2HE, UK. Tel.: +44 1772 893790.
E-mail address: jabbott@uclan.ac.uk (J. Abbott).

1569-1993/$ -see front matter © 2011 European Cystic Fibrosis Society. Published by Elsevier B.V. All rights reserved.
doi:10.1016/j.jcf.2011.11.005

of more than 30 symptoms from eight scales, only aspects of cough (cough/frequency and/or day/night cough) were assessed on all eight scales. Seven of the eight scales assessed sputum production and/or volume and five included decreased appetite. Furthermore, the rationale for including symptoms in scales was not always clear and no scale has undergone appropriate clinimetric evaluation.

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. Gaining information directly from children and adults is a sensible way forward given that a clinician's diagnosis of a PE is heavily influenced by patient reports. Asking the patient is nothing new, but such information can be used to provide a formal, standardised, valid and reliable way of gaining the patients' perspective as to the factors that constitute a PE, and supply information not captured by other means as there are some things that only the patient can know (for example, fatigue and nausea).

Little data have been collected directly from children or adults with CF concerning pulmonary exacerbations. Data used to develop the symptom scales on the CF-specific quality of life instruments [15–19] go someway towards this but their original purpose was not to identify an exacerbation. A promising scale is being developed and evaluated in the US [20]. This is a patient-derived respiratory symptom assessment and the rationale for the inclusion of symptoms has been based on qualitative work. Twenty-five interviews (13 children and 12 adults) using the Day Reconstruction Method (based on symptoms from the previous day) and 9 cognitive interviews were undertaken. Thematic analysis of the data enabled the development of a 16 item daily symptom diary (cough, chest tightness, difficulty breathing, wheeze, coughing up mucus, fever, chills/sweats, fatigue, sleep, school/work attendance, reduction in activities, more time sitting/lying down, worry, frustration, sad/depressed and cranky). The importance of qualitative work is often undervalued but it is essential to generate accurate patient-reported information.

Likewise, interviews were undertaken with 49 adults with CF in the UK [21]. This was a purposefully selected sample with representation from key demographic and clinical variables. 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. A range of systemic and respiratory symptoms were reported and their relative importance varied by severity of disease. 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. 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. This current work reports on interviews with children employing similar methodology and analyses. It aimed to (a) identify the salient characteristics of a pulmonary exacerbation as reported by school aged children with cystic fibrosis and (b) examine whether their descriptions of a pulmonary exacerbation map onto the results from adults.

2. Methods

2.1. Sampling

School aged children with cystic fibrosis were recruited from two specialist CF Centres in the UK. Purposive sampling ensured representation of key variables including age, gender, CF FEV_1 percentiles, nutritional status, microbiology, number of clinically recorded exacerbations requiring hospital admission or home IV treatment in the past year and whether the child was experiencing an exacerbation at the time of interview. All exacerbations had been diagnosed by a CF paediatrician and defined clinically as a change in symptoms not responding to outpatient therapy. Symptoms included increased cough, fatigue, volume and colour of sputum, and/or decreased activity and weight loss. The treatment protocol for exacerbations was generally two IV antibiotics, increased physiotherapy and attention to nutrition where needed.

Thirty-eight children were invited to take part in the study, all of whom had experience of pulmonary exacerbations. Those children experiencing an exacerbation at the time of interview were being managed in hospital and were interviewed towards the end of their treatment. For outpatients, FEV_1 was measured at the clinic visit at which the interview took place. For those hospitalised with an exacerbation, FEV_1 was recorded on the day of discharge. The study was approved by the North Staffordshire Local Research Ethics Committee. Both parental and child consent/assent were obtained.

2.2. Conceptual approach, interviews and data analysis

Interviews were undertaken by a single researcher (A Holt). A semi-structured interview format was used which allowed the child flexibility to follow their own experiences and also for the researcher to probe areas of interest [22]. Children were asked to report symptoms experienced during a pulmonary exacerbation. They were asked to report the first symptoms they become aware of, and how they subsequently recognised when they were improving. Parents of children under 12 years were allowed to provide information in the interviews but the experiences and perceptions of the children were always elicited first.

A grounded theory approach utilizing constant comparative analysis [23] was employed. The interview schedule was amended as new concepts emerged from ongoing analysis. This provided systematic prompts to ensure focused and appropriate questioning around children's experiences of pulmonary exacerbations. Interviews were taped, transcribed verbatim and the data analysed thematically. A rigorous method of working through the interview transcripts was devised as were methods of coding and categorizing the data. Data collection and coding took place concurrently. Line by line coding of the transcripts was undertaken.