Title: Current knowledge on eating experiences and behaviours in Cystic fibrosis: Exploring the challenges and potential opportunities for interventions.

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Cystic Fibrosis (CF) is an autosomal recessive inherited disorder affecting up to 100000 people worldwide, and is the most common genetic disease in Caucasian North European populations. In the European Union 1 in 2000-3000 newborns have CF and in the United States of America the incidence is 1 in every 3500 births [1] and is caused by defects in a recessive gene, the cystic fibrosis transmembrane conductance regulator (CFTR). The resulting imbalance in fluids and salts increases the production of thick, sticky mucus and impairs the functioning of the respiratory tract, inhibiting the clearance of microorganisms leading to recurrent infections. Lung disease is progressive and is the most significant life limiting complication of CF [2]. CF also impacts on digestive system organs (liver, pancreas and intestine) causing pancreatic ducts to become blocked, preventing the production of digestive enzymes, leading to the maldigestion and malabsorption of nutrients [3, 4]. Secondary conditions such as diabetes mellitus and chronic liver damage are common and other complications also include pneumothorax, haemoptysis and osteoporosis [3, 5]. Over the past two decades, life expectancy for people with CF has increased in the UK from 18 years to 47 years [6]. This increased life expectancy is a result of developments in treatments, and as medications to treat the basic defect in CFTR develop further, life expectancy is likely to continue to rise [2]. This timely advancement also brings new challenges for patients and clinicians alike as complications of CF are seen to increase with age.

CF remains a progressive disease and daily treatments are focused on managing symptoms, for the respiratory system this includes physiotherapy, nebulised bronchodilators, intravenous and/or oral antibiotics, steroids and mucolytics. For the digestive system, enzymes and nutritional supplements are required, and for more than a third of adults with CF related diabetes (CFRD) tablets or insulin are required [7]. The symptoms resulting from this genetic disorder require complex medical management which is burdensome and time consuming, treatments take up to four hours each day, with frequent hospital stays even when a person is relatively well. It is extremely challenging for people to find the time to complete all treatments every day, particularly when having to prioritise prescribed treatments within a daily routine which may include school, work, family and social activities. In addition, the motivation to undertake all treatments may be reduced as the positive association between current treatment regimens and future health is not always salient [8]and health is likely to progressively deteriorate regardless of adherence efforts [9] Treatment adherence is a major concern [10] poor adherence causes an increase in outpatient visits and hospital admissions, creating incorrect conclusions about treatment efficacy.

The damaging effects of CF frequently result in a compromised nutritional status due to a number of factors. Most people with CF have an increased energy need, between 150-200% of usual

requirements due to increased resting energy expenditure [11] and damage caused to lungs makes breathing more difficult which increases energy requirements. Pancreatic insufficiency, affecting up to 90% of the CF population, leads to malabsorption of fat and a need to ingest more fatty food. Enzymes are needed at every meal and snack, but even with this medication, many experience a number of difficult digestive symptoms including greasy and bulky stools, frequent and difficult bowel movements, constipation, nausea, swollen painful abdomen, heart burn and loss of appetite, all of which make it difficult to meet and maintain energy requirements [12].

These symptoms impact strongly on eating behaviours and nutritional and weight status, and this is of vital importance because optimal nutritional status is closely linked to respiratory status. Weight and body mass index (BMI) are well established in the literature as being independent predictors of mortality in CF [13,14]. The importance of optimal nutritional status for health and for quality of life [3] means that dietetic treatments, counselling and support, constitute a major part of treatment, and in the UK dieticians are important members of paediatric and adult multidisciplinary teams. From birth there is a necessary focus on following a prescribed dietary regimen alongside all other treatments including regular weighing and monitoring of weight, having to eat an elevated amount of calories each day, eating particular (high fat) foods and drinking plenty of water. Trying to comply with the recommendations can prove to be a very difficult challenge for both children and adults, especially when one considers that lack of appetite is a main symptom for many people. Nutritional supplements and enteral tube feeding are often needed to achieve adequate energy intake and malnutrition remains a significant problem within CF [15].

Such an intense focus on diet, weight and exercise for disease management inevitably impacts on the development of eating behaviours, and eating difficulties in childhood including poor appetite, longer mealtimes, slower pace of eating and food refusal are well documented [16]. There is limited evidence investigating eating behaviours within adolescent and adult CF populations, research focuses on the psychopathology of eating, with initial early findings suggesting that people with CF may be at risk of developing eating disorders [17]. Later studies suggested that formal eating disorders such as Anorexia Nervosa, Bulimia Nervosa and Binge Eating were not more prevalent, but that disturbed eating attitudes and behaviours were evident, specifically using exercise to influence shape, and a higher influence of weight on self-evaluation [18]. Given the serious consequences of low body weight on morbidity and mortality some authors called for regular screening for disturbed eating in people with CF.

There are a number of challenges with investigating disordered eating behaviours within CF, and indeed in other chronic lifelong conditions. The focus on diet and eating as part of a

treatment regimen necessitates a preoccupation with eating, and being highly attentive to eating and diet is considered good adherence. Therefore interpreting research relying on self-report measures is problematic, as it is subject to respondent bias as participants may answer in a way that denotes their compliance and adherence, and even if this is not the case, an elevated focus on eating and body weight may be considered adaptive for health rather than disordered.

In a recent study, experiences and practices on disturbed eating behaviours in CF were sought from health care providers [19], this methodology circumvents the issue of potential patient responder bias. Heath care providers reported that almost half of patients misused pancreatic enzymes (though this may be poor adherence rather than disordered eating) and engaged in food restriction behaviours, while almost a third engaged in binge eating and/or skipping meals. Authors highlighted the need for a CF specific disordered eating screening tool, a standard protocol for disordered CF eating and more specific training for health providers around disordered eating.

There is a paucity of research in regards to eating experiences and behaviours for people with CF, the available literature focuses on eating disorders and disordered eating, and does not reach any clear conclusions for all the reasons discussed above. The narrative that is clear, is that rules around eating and diet are very much in the clinical domain, what and how they eat become less of a personal life choice for people with CF, rather it is dictated by CF, clinical teams, parents and relatives. Enteral tube feeding is one of the treatment options available when insufficient calorie intake is prolonged, and while this can be highly effective in improving wellbeing [12], it is perceived by some people with CF as an ever present threat. Having to resort to enteral feeding was seen in recent study to epitomise a failure to self-regulate eating and illustrated the lack of personal control and choice over eating and an inability to keep oneself alive [20].

A fuller understanding of the eating behaviours in people with CF is needed to identify the prevalence of problematic eating, but also how to better support eating behaviours to achieve the maintenance of a healthy weight and to align any interventions with good psychological wellbeing. Egan et al's [20] research on everyday eating experiences of people with CF highlighted clearly the amount of time and cognitive effort expended in self-regulating eating, from thinking about eating, preparing to eat and actually consuming food. In this qualitative interview study participants described using a number of strategies to try and reduce the effort needed to eat. These strategies included avoiding food preparation by using take-away, eating out, or having partners prepare food, eating easy to swallow foods such as yoghurt and soup, and eating easily available foods that need no preparation (crisps and chocolate). Distraction and multitasking, such as eating whilst watching a film were also used in order to eat sufficient calories. Collectively these strategies present a picture

of avoidance and inattentive or distracted eating, which are the very opposite to what is usually considered adaptive and healthy eating, namely mindful eating.

This research also identified a preoccupation with weight gain, body image and dietary health implications, and eating was frequently seen as another treatment to endure. Overall in this study there was an overriding lack of pleasure in eating, even when weight was stable and eating was not particularly difficult, it was not pleasurable and still created anxiety. The anxiety felt was related to previous experiences of how quickly health can fluctuate, from being able to self-regulate eating when relatively well, a person with CF can quickly become very ill, experiencing reduced appetite and increased nausea at a time when they need to consume more calories to maintain weight. This results in weight loss which then exacerbates symptoms and can bring the (usually unwanted) prospect of enteral feeding into the treatment plan.

This triad of experiences, lack of pleasure in food and eating, anxiety around eating and the fluctuating nature of CF strike at the core of attempts to understand the nuances between disordered eating attitudes and behaviours and usual eating behaviours for someone with CF. Interventions to support eating experiences and behaviours need to be adaptive to fluctuations in health status which alter nutritional needs, requiring a change in how a person regulates their eating according to the current clinical assessment of the individual. They also need to address the fundamental issue of how to eat well when there is no pleasure in eating. Given the lack of current evidence on eating in CF, it is necessary to look at the wider literature on eating, particularly around modifying eating behaviours, to help in the development of effective interventions that enable self-regulation in a chronic condition that is constantly at risk of fluctuating and requiring different interventions.

Self-regulation, which is a primary aim for people with CF, has been explored extensively in association with eating and is increasingly problematic across the western world. Researchers have attempted to address the problems of obesity and related diseases, to enable lower calorific consumption, and therefore, weight loss and maintenance, by regulating food consumption [21]. While the problems around self-regulation are evidential for much of the general population, findings also propose that the main issue for obesity is living in an ‘obesogenic environment’, whether it related to a macro- or micro- level approach of environmental influence to overconsume. At a macro-level, researchers have proposed public health initiatives of increasing the availability of smaller portions and alternatives to sugary soft drinks [22]. Research evidence at a micro-level shows that overconsumption is evident when serving oneself on a larger plate or sitting too close to a bowl of chocolates. It is suggested that the obesogenic environment bears the most accountability

for the increasing numbers in obesity [23, 24]. Other research has focused on the occurrence of multi-tasking (such as watching TV while eating), and indicates that not being attentive to the food leads to inaccurate recalling of how much food was consumed, and to higher consumption compared to people who are not paying attention to another activity while eating [25, 26].

All such research has been classified as standing under the umbrella of mindless eating, and has been given much attention in tackling the obesity epidemic. While this research shows how mindless eating poses a problem to the general population, Mantzios, Egan and Patchell [27] proposed that there is potential to enhance calorific intake by utilising the research conducted with the general population, and to enable people with CF to mindlessly enhance their calorific intake. In essence, they suggested that the cognitive and environmental factors that lead the general public to overconsume, may be a method of clinical and nutritional counselling and advice to help people with CF consume more calories when needed.

Over the past ten years there has been emerging evidence of over-nutrition in people with CF leading to being overweight and obese [28]. This is due to a number of clinical and treatment factors including earlier diagnosis of CF and of pancreatic insufficiency, and improved medical therapies, especially, but not confined to, CFTR targeted therapies. In addition, dietary and eating behaviour advice to consume a higher fat diet than non CF people that has been endorsed over a lifetime by health providers may not be so consistently relevant, as the Western diet and the obesogenic environment have increased intake of high fat foods in the general population. The applicability of mindless eating in response to under nutrition has been suggested and a fuller exploration of how mindfulness and mindful eating may support weight regulation in other health scenarios for people with CF is indicated.

Kabat-Zinn [29] described mindfulness as experiencing the present moment, whether it is pleasant or unpleasant, with qualities of awareness and non-judgment. Similarly, mindful eating adopts the fundamentals of mindfulness practices, and applies them to eating- and food- related experiences. Mindful eating has been effective in increasing the pleasure around eating, while decreasing fat and sugar consumption, grazing, and motivations to eat that are not aligned to internal feelings of hunger and satiety [30, 31, 32, 33, 34]. Furthermore, while some researchers have made clear propositions around eating less and making more informed choices [35, 36, 37], others have instigated the usefulness of mindful eating when eating is problematic or disordered[38,39]. Egan and Mantzios [40] proposed how mindful eating may form an additional tool for achieving optimal weight for people with CF, and how it could propose an element of self-regulation around eating and food. Their assumption was that mindful eating assists self-regulation

by reducing the automatic, affect, and cognitive alignments when eating, which are primarily responsible for eating and weight difficulties. Currently, self-regulation may be achieved through combining current findings and the need for both mindless and mindful eating in the repertoire of eating practices for clinicians and people with CF.

Levels of anxiety and depression are similar between individuals with CF and the general population [41] however the impact of even slightly increased levels are detrimental for a person with CF as they are linked to reduced lung function, quality of life and adherence. Anxiety, stress and depression are also associated with emotional, external and restrained eating [42], however this is so far largely unexplored in the CF literature. In a recent study, Egan & Mantzios' findings [43] suggested that higher levels of emotional eating (i.e., eating in response to emotions) significantly predicted higher BMI in a sample of people with CF. While a higher BMI is often the aim and is desirable, an increase in levels of emotional eating is not desirable for holistic health and wellbeing.

Emotional eating is more problematic in changing as a health behaviour (when compared to environmental influences), and may form a platform that would lead to other problematic eating such as binge eating and grazing. When mindfulness and mindful eating were tested conditionally in assessing their impact on the relationship between emotional eating and BMI, it was found that both mindfulness and mindful eating moderated this relationship to the extent of making it non-significant (44). This research proposes the potential of mindfulness and mindful eating in creating a better eating environment for people with CF.

Within the literature on eating in people with CF, there is clear evidence for problematic eating experiences and attitudes. A strong focus on eating, high levels of anxiety, reduced or non-existent pleasure in eating, and a difficulty self-regulating healthy weight are all very apparent. Whether these factors constitute disordered eating or not is debatable. Certainly being highly aware and attentive to weight and diet may be considered adaptive for the physiological health of people with CF, but the anxiety and lack of pleasure in eating are detrimental to both physiological and psychological health and wellbeing. The pressing need is find holistic ways of supporting healthy eating behaviours which assist in managing co-morbid conditions such as CFRD, and osteoarthritis, maintain optimal weight rather than simply focusing on losing or gaining weight and tackle the psychological and social barriers to pleasurable eating. Given the complex fluctuating symptoms and extensive burden of treatments for people with CF this is a considerable challenge.

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