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Original Article

When is too little care, too much harm in cystic fibrosis? Psychological and ethical approaches to the problem

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Abstract

Some parents of children with cystic fibrosis (CF) do not adhere to treatments recommended by the CF team. This can be a challenging issue for CF clinicians and can create conflict between the parents and treating team. Both parents and treating team believe they are acting in the best interests of the child, but do not share a common opinion as to what that entails. In this paper we present an understanding of the psychological framework of parents' illness representation that may foster a better understanding by CF clinicians of how to approach parents who hold a conflicting opinion regarding optimal care. Continuing to work with families towards optimal care is a moral obligation, but the key ethical decision is when to intervene to protect the child. In this paper we introduce the concept of the zone of parental discretion as an ethical tool to help decide the best way forward when parents do not accept medical advice on the optimal care of their child with CF.

Keywords: Cystic fibrosis; Psychological; Ethics

1. Introduction

The care of children with cystic fibrosis (CF) is complex and time consuming for parents. It involves giving daily medications, airway clearance and a strict diet [1,2]. There are regular clinic attendances, investigations and for some, hospital admissions. Not every family adheres to recommended treatment [3]. The consequences of consistently missing treatments are more rapidly apparent than others. Failure to give pancreatic enzyme, fat-soluble vitamin or salt replacement will result in malnutrition, diseases of vitamin deficiency and hyponatraemic dehydration. Death could result in a matter of weeks, certainly months. Failure to treat acute pulmonary exacerbations will result in accelerated damage to the airways [4]. Other therapies, such as daily airway clearance, mucolytics and regular antibiotic treatment for chronic lower airway infection improve long-term outcomes, but the effect of missing these is not as immediately apparent [5,6].

The reasons parents do not adhere to CF treatment recommendations vary. Some lack personal or financial resource or suffer social disorganisation [7]. Children's behaviour around therapies can be difficult contributing to poor adherence [8]. Some parents deliberately refuse to give certain elements of treatment. In some cases the consequences are rapidly apparent and child protection services can be activated. However, the effect of poor adherence is not always directly apparent and navigating a path through this is difficult. Despite the best efforts of the team, parents' failure to provide CF treatments can leave CF clinic staff feeling responsible for poor outcomes, creating moral distress [9].

The aim of this paper is to present three cases that highlight parental deliberate non-adherence with recommended treatments. We ask the question "how little care constitutes too much harm?"

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in patients with CF. We introduce the concepts of illness representation and the zone of parental discretion as ways of considering the problem [10]. The cases presented are "fictionalised", to maintain patient confidentiality, but elements from the cases are based on our collective experience.

Case 1. A 3 month infant presented with failure to thrive. He was missed by newborn screening (elevated immunoreactive trypsinogen, but negative on 5 CFTR gene mutation screen). He had pancreatic exocrine insufficiency and sweat chloride 102 mmol/L (CF range > 60 mmol/L). His father refused all therapies, including nutritional support with pancreatic enzyme, fat-soluble vitamin and salt replacement and prophylactic antibiotics. Despite prolonged encounters with both parents, the father could not be persuaded to adhere with our recommendations. The case was referred to child protection and a court ordered immediate admission to hospital with administration of all recommended investigations and therapies. The child was discharged into the care of the family two weeks later, with close supervision by child protection. The mother agreed to administer all medications and undertake airway clearance.

Case 2. A twelve year old male with pancreatic insufficient CF (p. F508del homozygous, sweat chloride 101 mmol/L) transferred to our clinic. He was taking pancreatic enzyme replacement, fat-soluble vitamin and salt replacement. His body mass index (BMI) was 15 (10th percentile for age). He had advanced bronchiectasis on CT scan and was chronically infected with *Staphylococcus aureus* (susceptible to methicillin). His FEV_1 was 65% predicted for height. He was poorly adherent with dornase alpha and airway clearance. His father refused to give him oral antibiotics and refused admission to hospital for intravenous antibiotics. The patient's scheduled vaccinations had been given, but annual influenza vaccine was refused. He was also given Echinacea, garlic and "Chinese herbs". He was brought to CF clinic regularly, but despite personal engagement, empathic listening and provision of the clear rationale for antibiotic use, his father's approach could not be altered. His father had an entrenched health belief that followed natural therapies and avoidance of antibiotics that he considered harmful. The family changed clinic physicians on a number of occasions. After parental separation when the patient was 16 years old, with maternal custody, the patient commenced on regular admissions to hospital and was given oral and inhaled antibiotics at home. His FEV1 at 16 years was 40% predicted.

Case 3. A three year old child with CF (NBS positive, p. F508del homozygous, sweat chloride 96 mmol/L). She was given pancreatic enzyme, fat-soluble vitamin and salt replacement and was well nourished (BMI 16, 50th percentile for age). Cough swabs regularly cultured haemophilus influenzae which was also on her last two annual bronchoalveolar lavages. Her chest CT demonstrated mild bronchiectasis, airway thickening and gas trapping. She received dornase alpha. Her parents refused to give her antibiotics in the belief that antibiotics would affect her developing immune system and alter normal gut flora.

In the three cases there is a gradation of harm created by the parents' refusal to adhere with recommended CF treatment. The first case demonstrates clear harm to the child by withholding nutritional support which would result in malnutrition and, if unchecked, death in a matter of weeks to months. In the second case the patient has CF lung disease that is more advanced than many patients of his age and demonstrates an accelerated decline in lung function. However, the accelerated lung function decline cannot be definitively attributed to the lack of antibiotics, crucial to considering the third case.

2. Psychological framework to understand why parents refuse to provide optimal CF care

These three cases suggest a difference in opinion between the parent and the treating team. Our patient, however, is the child but the parents are the ones asked to deliver care. This creates a complex therapeutic relationship that has been explored elsewhere [11]. In this section of the paper we provide a framework for understanding why parents might have these beliefs about CF management. It is an ethical requisite to keep trying to help parents to accept optimal treatment – but it needs to be done in a way that has some chance of success, rather than entrenching disagreement.

The diagnosis of a chronic illness that results in a shortened life-span, requires parents to form a representation of what this illness will mean to their child and family. An "illness representation" (IR) or "health belief" is an individual's own implicit, common sense belief about his or her illness [12]. The Illness Representation creates the knowledge base on which to form their behavioural response. Concordance of the illness representation with the medical advice, is linked to the likelihood of adherence to treatment, and ultimately predicts how a parent/patient will adjust to the illness [13]. By understanding a parent's illness representation, clinicians can understand parent's motivations for their behaviour, reduce their own moral distress, and engage families in a way that may change their approach to recommended treatments.

An individual's illness representation is mediated by factors independent of the knowledge imparted by a medical professional. The Common Sense Model of Self-regulation has five features of illness representation: Identity, Timeline, Cause, Cure/Control and Consequences [14]. *Identity* refers to the label, severity and symptom profile the individual believes is associated with the illness (e.g. this is not life-threatening). *Timeline* refers to the expected timeframe for illness (e.g. a parent may assess CF as being either a series of brief, acute illnesses; a chronic condition, or a cyclical condition). *Cause* relates to the individual's attribution about what caused the illness (e.g. bacteria, virus, genetics, being run down) and what maintains the symptomology. *Cure/control* refers to whether the individual believes there is a cure for this illness (the presence of medicines such as ivacaftor may influence an individual's belief about cure), how well it can be controlled by the individual (e.g. 'if I take my treatments as directed CF will not progress), or whether the impact of CF on their quality of life can be controlled; *Consequences* refer to the individual's beliefs about what the consequences of the illness are (e.g. death, shortened life,