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Cystic Fibrosis Frequently Asked Questions

Question 10: Could the Burden of Care with Cystic Fibrosis Impact on Educational Outcomes?

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With advances in the management of cystic fibrosis (CF), affected children are living longer and reaching adulthood. This places greater emphasis on a child's ability to fulfill one of the main goals of childhood, that is, to prepare for adulthood. Despite medical advances, there's a high treatment burden required to maintain health, potentially compromising the young person's ability to achieve their full academic potential, impacting on their future career options.

The life expectancy of children diagnosed with CF today is approximately 40 years, a considerable increase compared to the 1960s, when most children didn't reach their teenage years [1–4]. Better survival has been achieved as a result of intensive therapy with an increased treatment burden, based on management guidelines, requiring hospital admissions and greater nutritional requirements. Since the introduction of newborn screening [NBS] programmes, more than 63% of CF cases are diagnosed in the first few weeks of life [2]. This has led to an improved life expectancy and a new focus on educational outcomes and preparedness for participation in the workforce as adults [5].

However, there are multiple reasons to suspect that children with CF are at a significant disadvantage when it comes to schooling. This includes extensive treatment regimens, hospital admissions and debilitating symptoms which result in prolonged absences and potentially compromise the child's ability to actively participate in class.

TREATMENT BURDEN OF PHYSIOTHERAPY

Management of CF is extensive and can be very time consuming, with parents spending up to 3 hours each day on treatment with their children [Fig. 1][6]. Treatment typically includes daily use of dornase alpha and/or mannitol and hypertonic saline, to thin and assist in the clearance of mucus [7–9]. The indicators of disease severity will usually predict the extent and intensity of interventions that will be required [Table 1]. For some children, β2 adrenergic agonists are used to reduce the occurrence of bronchospasm resulting from inhaled medication [7]. During acute pulmonary exacerbations of illness, children are required to take regular antibiotic therapy [6]. At home, this is usually through a nebulizer, generally twice a day, which can take approximately 40 minutes with standard nebulisers per day [9]. Furthermore, as children become older they are expected to take on increasing responsibility for their care as part of their evolving autonomy, but this can be problematic with regard to adherence [10].

With progression of lung disease or more serious infections, such as with Pseudomonas aeruginosa (PA), hospital admissions

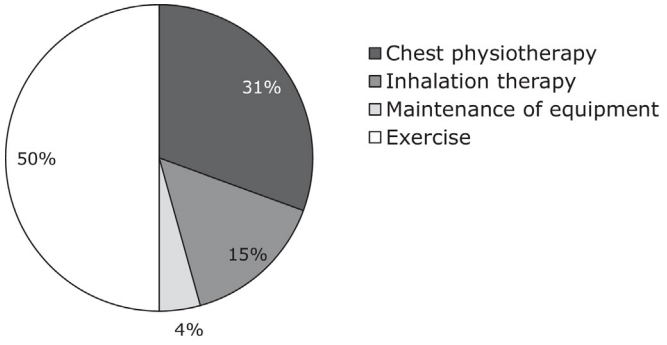

Fig. 1. Proportion of time spent on particular CF therapy. Derived from Hafen et al. [55].

Table 1
Indicators for Severity of Disease. Derived from Konstan et al. [8].

Indicators of Disease Severity
Low FEV_1
Inadequate nutrition
Pancreatic insufficiency
Infection with Pseudomonas Aeruginosa
Recurrent exacerbations

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Table 2
Estimated times spent performing airway techniques. Derived from Samuels [54].

Technique	Age	Frequency (per day)	Duration (min)
Series of breathing techniques	≥5	1-5	20-40
Positive expiratory pressure	≥9	1-5	20-40
Flutter	≥9	1-5	20-40
Autogenic drainage	>13	1-5	20-60

are required to obtain intravenous access, initiate antibiotic therapy, monitor antibiotic levels, review physiotherapy routines and assess nutritional status. The admissions may last for up to three weeks [11]. For children chronically infected with PA, cycled courses of twice daily inhaled antibiotics are added to their already comprehensive treatment regime [12].

With the increasing use of aminoglycoside antibiotics both intravenously and via nebulization, often from an early age, there remain concerns surrounding the potential for ototoxicity, vestibular dysfunction and tinnitus resulting from the cumulative doses received [11]. This can adversely affect children at school, by compromising their ability to hear and comprehend content taught in class. In addition, for the treatment of chronic allergic broncho-pulmonary aspergillosis [ABPA] which may affect 6%-15% of older children and adults with CF, the use of oral steroids is associated with impaired memory, potentially impacting the person's ability to consolidate material studied [13–16].

Despite physiotherapy being one of the mainstays of CF management, adherence is often disappointing with only 40-55% adherence reported in one recent adult study [9]. According to Goodfellow, as children get older, they are more likely to have poorer adherence to physiotherapy regimens [10]. Methods for mucus clearance consist of percussion, positive expiratory pressure and postural drainage depending upon age and personal preference [7]. Ideally, these airway clearance practices, together with regular exercise are encouraged up to twice daily and intensified during times of pulmonary exacerbation [Table 2]. According to Samuels and colleagues, expected times for airway clearance techniques are generally between 20-60 minutes every day [Table 2]. However, these times are more likely to occur under ideal conditions and are unlikely to represent the realistic time burden on patients. Procrastination, distractions and time to set up and clean equipment are additional factors that need to be considered in the home setting with children and adolescents.

TREATMENT BURDEN OF PANCREATIC INSUFFICIENCY

The vast majority of children with CF have pancreatic insufficiency, with approximately 67% of children affected from birth [17]. By 12 months of age, almost 90% of children will require pancreatic enzymes to treat fat malabsorption [18]. The presence of nutritional deficits from a young age has the potential to negatively impact upon the development of neural pathways during infancy and childhood, possibly influencing the school outcomes of affected children [19]. More specifically, extremely low levels of vitamin E during early childhood have been linked with impaired brain development [20]. With approximately 20% of teenagers developing CF related diabetes, there is the risk of hypoglycaemia, resulting in an imbalance between caloric intake [by mouth or with enteral feeds] and insulin dose [4]. Monitoring blood sugars becomes an important additional burden that they must accommodate in their already time-consuming routine.

Due to malabsorption and increased energy needs, children with CF are required to have 120-150% of the recommended daily caloric intake of children without CF, with almost half of their calories taken from fat [21]. This can place considerable pressure

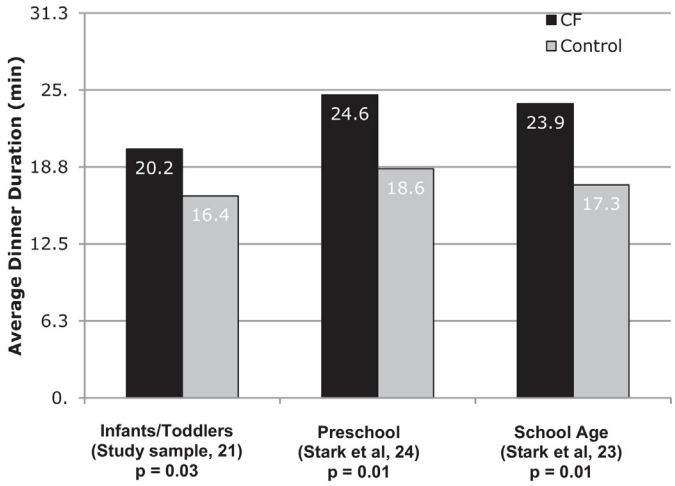

Fig. 2. Average Mealtime duration for CF patients compared to controls at different ages. Derived from Powers et al. [21].

on parents and children to meet yet another treatment requirement. According to Stark and colleagues, children with CF took approximately 24 minutes to complete dinner, compared to 17 minutes for controls [Fig. 2] [22]. While this study details the time children with CF spend during dinner, it fails to take into account other meals of the day, including snacking and the time some children and parents require to set up nocturnal feeds. Interestingly however, another study completed by Stark showed that school-aged children with CF didn't demonstrate a higher occurrence of disruptive eating behaviour, compared to gender and age-matched controls [23]. Rather than mealtimes for children with CF becoming prolonged due to disruptive behaviour, it may rather be that parents and children are attempting to increase their caloric intake by spending longer at the dinner table [24].

PSYCHOSOCIAL STRESSORS

According to Grieve and colleagues, teenagers with CF are persistently challenged with serious stressors associated with their wellbeing, which has the potential to be detrimental to their educational attainment [25]. Worsening lung function during times of acute pulmonary exacerbations and worsening daily respiratory symptoms as the disease progresses compromises the child's energy levels, limits them from participating actively in school, curricular and co-curricular activities [25,26]. While developing and maintaining friendships is very important for children and adolescents, as is the desire to fit in, with the excessive time burden of managing the disease, patients with CF are possibly inhibited from meeting the social needs of childhood and adolescence [26,27]. This is particularly challenging for adolescents, with their illness isolating them from their classmates and peers, stemming from debilitating symptoms or lengthy hospital admissions [26].

With the physical, psychological, social and financial challenges associated with a life-long illness such as CF, caregivers of children with CF are at an increased risk of depression and anxiety [28]. In an Italian study of 225 parents with children with CF, aged from 13-17 years, more than half of the parents who described extreme stress were clinically depressed [29]. These findings are supported by a study in the USA, of the parents of 88 children with CF, aged 1-11 years [30]. This has serious implications because the children whose parents had elevated levels of anxiety and/or depression were more likely to be poorly adherent with treatment regimens [28,30].