

**Title: Enhancing wellbeing and quality of life in cystic fibrosis through holistic nursing practices: a case study**

Kirsty Tointon BA (Hons), BSc (Hons)  
Student Health Visitor, Bournemouth University  
(Developed from work submitted whilst a second year student of BSc Child Health Nursing programme, Bournemouth University)

Jane A Hunt PhD RSCN SRN PGCEP PGCRDS  
Senior Lecturer Children's & Young People's Nursing  
Faculty of Health and Social Sciences  
Bournemouth University

**Abstract:**

Cystic fibrosis is one of the most common life limiting autosomal recessive genetically inherited conditions affecting Caucasians. It is caused by mutations of chromosome 7 responsible for encoding amino acid protein 1480, the cystic fibrosis transmembrane conductance regulator (CFTR) protein. The most common mutation, affecting 75% of patients with CF in the UK, is that of delta F508 which results in the omission of phenylalanine at position 508 in the 1480 chain. However, other rarer mutations exist resulting in differing presentations of the condition. Ellen, a teenager with a rarer presentation of CF, and her family, are drawn upon in this case study to discuss living with CF and its impact upon quality of life and well-being in life limiting conditions of childhood.

## **Introduction**

Well-being and quality of life (QoL), whilst high on national and international political agendas in recent times (e.g. The Children's Society 2015, UNICEF 2011), can be abstract and subjective, are complex terms frequently intertwined with each other, with life satisfaction, social capital and with happiness (e.g. Hean et al. 2013, Taylor et al 2008, Ferguson 2006). Savage et al. (2009) identify QoL as a multidimensional construct, relating to individuals' perceptions of the impact of illness and treatment on their health, well-being or physical, psychological and social functioning. Similarly, well-being has been defined as: '*a dynamic state that is enhanced when people can fulfil their personal and social goals... both in relation to objective measures, such as household income, educational resources and health status; and subjective indicators such as happiness, perceptions of quality of life and life satisfaction*' (Statham & Chase 2010), with broad consensus that it refers to the quality of people's lives (The Children's Society 2015). Whatever the complexity and lack of clarity of definition of these two terms, the extent to which having a 'life-limiting' illness, defined as having: '*no reasonable hope of cure*' (Together for Short Lives 2013:4), impacts on well-being and QoL is explored here. To this end, this case study explores the impact that living with cystic fibrosis (CF) has on one family and identifies how this could affect their well-being and QoL.

## **Cystic fibrosis**

One of the most common life limiting, genetically inherited conditions affecting Caucasians, CF is a multisystem, autosomal, recessive inherited disorder (Ren et al. 2011, Winter 2010). It affects exocrine glands, impacting upon sweat production and regulation, the gastro-intestinal tract, most notably the pancreas, and reproductive systems; lung disease continues to be the major cause of morbidity and mortality (Ren et al 2011). CF is caused by mutations of a gene in chromosome 7 that encodes amino acid protein 1480, the cystic fibrosis transmembrane conductance regulator (CFTR) protein (Peckham et al 2006). Amino acids block the formation of CFTR protein, thus if any are missing or irregular the CFTR protein may cease to function correctly (Mayall et al. 2009).

The most common gene mutation, affecting 75% of patients with CF in the UK, is that of delta F508, resulting in the omission of phenylalanine (an essential amino acid) at position 508 in the 1480 chain (Thomas & Pedersen 1993). A rarer mutation of R117H, accounting for 0.8% of mutant genes, changes arginine (another essential amino acid) to a histidine (an alternative essential amino acid from which histamine is derived) in a transmembrane domain of the protein, alters conduct of the ion channel, frequently maintaining pancreatic function (Pont-Kingdon et al.2004).

Varying gene mutations affect CFTR function in different ways. The normal CFTR gene functions as a controlled chloride channel, which may then adjust the activity of other chloride and sodium channels at the cell surface (Ren et al 2011, Mayall et al 2009) to produce a watery mucous which protects the linings of involved organs, enabling bacteria excretion (Cystic Fibrosis Trust 2011). People with CF, however, have irregular transport of chloride and sodium across the respiratory epithelium, resulting in thickened, viscous airway secretions (Ratjen 2003), trapping harmful bacteria, causing lower respiratory infections (Cystic Fibrosis Trust 2011). Mutations in both copies of the CFTR gene and the genetic context in which a mutation occurs plays a significant role in defining CF type (Katkin 2015).

Treatment for CF is dependent upon the nature of the gene mutation type and extent of symptoms and organs affected by the disease (Ren et al 2011); it primarily aims to maintain optimal health and reduce lung infection risks and malnutrition (Mayall et al. 2009). Pancreatic-enzyme supplements are required for those with pancreatic insufficiency as well as a high calorie diet, due to their body's inability to absorb fat (Ren et al. 2011). Medication and other treatments, such as physiotherapy, are part of arduous daily regimens, aiming to dislodge mucous and prevent lung infections (Bryon & Wallis 2011). Eventually, however, capacity and elasticity of the lungs are lost, resulting in eternal damage and lung function decline, which can be fatal.

### **Ellen: The case study**

Ellen<sup>1</sup> is 15 years old and in Year 11 at school, preparing for her GCSE examinations. She was diagnosed with CF at the age of 12 years after her mother had been repeatedly told for years that her respiratory symptoms amounted to nothing more than a cough and sinusitis. Ellen inherited the most common gene mutation, delta F508, from her father and a R117H mutation from her mother; the latter resulting in Ellen having pancreas-sufficient CF. Thus, there are no health implications to her digestive system, although her respiratory symptoms are severe. She lives with her younger sister Janie and their mother Nella. Janie also has CF and was only diagnosed after screening following Ellen's diagnosis. After separating from Nella, their father, Jon, moved away and subsequently rarely sees his daughters. Nella invariably cares for her daughters independently. Her parents' separation, along with enforced frequent school absenteeism, due to her illness, and peer bullying have contributed to Ellen having anxiety, currently controlled with Sertraline. Ellen is routinely admitted to her local hospital every three months for a two weeks course of intra-venous flucloxicilin and amoxicillin, or sooner if and when her lung function deteriorates.

### **Planning Ellen's nursing care**

Nursing care is holistic rather than diseased-focused (Manley et al. 2011). Thus, carrying out a comprehensive, systematic nursing assessment of psychological, social, spiritual and physical needs, implicit within nursing's professional practice standards (NMC 2015), facilitates QoL and well-being needs being met. Being mindful of this, nurses caring for Ellen ensure that regular assessment is conducted to re-evaluate her holistic nursing requirements.

Adolescence, according to Erikson, is a time of conflict between identity and role confusion when a sense of self needs be developed (Pressley & McCormack 2007). Psychological challenges associated with adolescence are typically heightened for Ellen and others like her due to demands associated with CF (Ernst et al. 2010). Vulnerable teenagers are potentially at risk of developing mental illness (Ernst et al. 2010) and studies have highlighted that anxiety and depression are common in adolescence with CF (Quittner et al. 2008). Consequently, prompt identification and treatment of patients with significant mental illness should constitute routine CF care (Besier & Goldbeck 2012). Thus, regular re-evaluation of Ellen's psychological wellbeing comprises an important element of her holistic nursing care.

Symptoms of anxiety are indicators of poor psychological well-being, having strong negative associations with individuals' perceptions of their QoL (Besier and Goldbeck 2012). Ellen's nurses recognised lack of contact with her father and her parents' separation, along with

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<sup>1</sup> All names throughout this work are fictitious to maintain anonymity

bullying at school and absenteeism, contributed to her anxiety, resulting in her nursing and wider multidisciplinary team having referred her to a psychologist. Consequently, alongside her medication, Ellen requires much nursing of her anxiety, whereby her nurses took time to talk with and listen to her. Despite her anxieties, Ellen possesses much positivity, humour and acceptance of her disease and its management and, as studies have indicated, such coping strategies potentially enhance QoL (Hayes et al. 2006, Wong & Heriot 2008). Comparing early studies with more recent ones, adjustment, coping and acceptance of adolescents with CF have enhanced, thought to be associated with improved treatment and longer life expectancy (Britto et al 2004, Sawyer et al. 2005). Thus, it is essential to appreciate how adolescents with CF, such as Ellen, maintain their well-being and QoL amidst 'normal' adolescent psychological challenges.

School absences for children and adolescents with CF are well documented (Twynam et al 2010). Due to frequent treatment Ellen misses much school, invariably resulting in both educational and peer relationship difficulties and repeated bullying, a phenomena reported in young people with CF (Twyman et al. 2010). Low self-esteem is also a predictor of increased bullying risks (Egan & Perry 1998). Since Ellen's self-esteem is challenged, her social vulnerability and educational struggles contribute to both bullying and social isolation. Since Ellen sees little of her father and endures difficult peer relationships, her only social support lies with her mother and sister, resulting in feelings of social isolation. To maintain Ellen's social well-being and QoL, Ellen's holistic nursing care requires close working relationships between children's nurses working in hospitals and in the community and school nurses, to minimise her social isolation.

Spirituality has a direct effect on well-being and is fundamental to personal perspectives of 'place in the world' (Wright & Neuberger 2012); it also enhances healing. Lying at the heart of nursing, spiritual care is paramount (RCN 2011). Ellen's parents' separation, her social isolation from peers, educational difficulties arising from school absences, bullying, complexities of 'normal' adolescence and those specifically associated with CF, and her anxieties, all contribute to significant spiritual needs which need to be met. Through acknowledging these spiritual needs, Ellen's nurses aim to maintain her spirituality through being empathetic and offering opportunities to talk about her concerns.

Given no pancreatic involvement, the aim of Ellen's physical nursing care, both during hospital admissions and whilst at home, is to manage her respiratory disease. Common respiratory symptoms in adolescents with CF include: a persistent cough, sinusitis, mucous amassing, resulting in inflammation, bacteria accumulation and subsequent pneumonia (Twyman et al. 2010; Peckham et al. 2006) and these are frequent concerns for Ellen. Hence, she is prescribed Colistimethate Sodium via a nebuliser twice daily, Tobramycin podhaler twice daily, Salbutamol evohaler as required and hypertonic sodium chloride solution for physiotherapy, required twice daily. These daily treatment regimens are physically arduous for Ellen and she finds it particularly difficult to 'keep up' with her friends. These physical pressures undoubtedly reduce Ellen's low sense of self-worth, QoL and well-being and increase her anxieties. Acknowledging the demands this has on Ellen, nurses ensure that they continued to encourage, support and empathise with these pressures, reassuring her that she is not alone.

### **Living with CF: Family influences**

CF impinges upon the lives of families of affected adolescents too (Branstetter et al 2008). Fujita and Diener (2005) have, for example, identified the importance of support from family

members in coping. Therefore, nurses in their holistic practices of Ellen endeavour to acquaint themselves with all her family members. This is particularly important as an association between parental coping and child adjustment is likely to influence an adolescent's overall well-being and QoL.

How well parents, siblings and affected children adjust to CF is not only reflective of the severity of the illness, but also by family members' acceptance and parental coping strategies (Casier et al. 2010, Wong & Heriot 2008); it is behavioural and cognitive efforts that people use which influence their coping strategies in stressful situations (Lazarus & Folkman 1984). Much of Ellen's extended family feel some sense of responsibility and guilt for her inherited life-limiting condition. Nonetheless, Nella views her rare gene mutation as responsible for her daughters retaining their pancreatic function. This is a positive coping strategy, which is important in parental adjustment to CF (Wong & Heriot 2008). In contrast, Jon displays behavioural disengagement through not seeing Ellen and Janie often. Not only will this invariably impact negatively on Ellen, it is likely to also have a damaging effect on Jon's own well-being and QoL.

Psychological distress endured by siblings of a child with a life limiting condition is well documented. Well siblings often view increased attention on their ill sibling as a personal rejection (Tew 2010). Despite also having CF, Janie remembers feeling jealous when Ellen was first diagnosed. Janie perceived those close to her to be preoccupied with Ellen, leaving her in her sister's shadows, mirroring the thoughts and feelings of siblings in similar situations (Williams et al. 2009). However once diagnosed herself, Ellen and Janie became extremely close. Janie recognises in light of her initial feelings, she may have found her relationship with her sister and mother much harder without her own diagnosis of CF. The sisters are able to understand the difficulties each face, and the treatments they endure daily together. Having each other gives them strength and seemingly enhances Janie's well-being and QoL too.

### **Quests in achieving well-being and QoL**

In a quest to achieve a good sense of well-being and a sound QoL, in the presence of life limiting conditions such as CF, we argue that holistic nursing practices are paramount. Within this holistic nursing framework, acceptance-based interventions whereby nurses within a wide interdisciplinary team of health professionals facilitate individuals to accept their health condition (Hayes et al 2006), are beneficial to improving well-being and QoL. Acceptance of a life limiting condition is measured as a means of increasing action which is directed towards important life goals (Hayes et al 2006). Similarly, a sense of optimism, associated with acceptance, may help to reduce or prevent altogether the onset of anxiety and depression (Ey et al. 2005). Ellen does experience, and is currently receiving treatment for, anxiety. However, the holistic nursing practices of her nurses facilitate continuing acceptance of her condition and feelings of optimism to enable her to achieve a good sense of well-being and a positive QoL, reflecting findings from studies about adolescents with CF published during the last decade (Britto et al. 2004, Sawyer et al 2005). These are similarly reflected in both her mother, Nella, and her sister Janie resulting in a family approach to achieving well-being and QoL.

### **Conclusion**

Well-being and QoL are complex to define yet are currently high on international and national agendas; both are paramount to consider in holistic nursing practices of adolescents with life-

limiting conditions such as CF. This case study has explored the impact of living with CF upon the well-being and QoL for one adolescent and her family. Adopting a holistic nursing framework, it has been established that, even in the presence of anxiety, rather than the severity of an adolescent's condition negatively impacting on their QoL and well-being, individuals' perceptions greatly influence acceptance, optimism and the development of individuals' coping strategies. Ellen's well-being and QoL has been enhanced with the support and positive strategies of her mother and sister, her nurses and the wider health care team, holistic interventions and her own positive coping strategies. Both Nella and Janie demonstrate similar positive strategies and traits. More negative strategies can potentially impact on Ellen's well-being and QoL too, as the case study highlights, with Jon's disengagement from his daughters. Both positive and negative coping strategies can exist within one family unit. Thus, it is essential for nurses to be mindful of every individual within a family network to ensure appropriate holistic nursing care is provided, where possible, to each family member, to enhance their well-being and QoL.

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