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Young people with cystic fibrosis' concerns for their future: When and how should concerns be addressed, and by whom?

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Introduction

Cystic fibrosis (CF) is the UK's most common autosomal recessive genetic disorder, with historically few children surviving childhood. Better understanding of the disease process has led to rapid improvements in treatment and care, with the majority of people affected now living into adulthood. Indeed, the current median survival age of 31 in the UK (UK Cystic Fibrosis database and CF Trust, 2006) is expected to rise to around 50 years for those born in the last decade (Dodge et al, 2007). Most adolescents with CF undergo developmental tasks common to all, such as further/higher education, commencing employment and developing intimate relationships, yet the salience of these decisions is likely to be more acute for those with a limited lifespan. Adolescents with CF additionally face challenges such as disclosing their condition, the possibility of requiring organ transplantation, and accepting that they may be infertile.

In response to CF patients' care needs, transition services have been developed to bridge paediatric and adult services. All services are staffed by multidisciplinary team (MDT) members including medical consultants, specialist CF and general nurses, physiotherapists, dieticians and others, aiming to address young peoples' physical health, their mental wellbeing, and psychosocial concerns. However, the confidence or effectiveness of MDT members feel able to address current and anticipated psychosocial issues relevant to these young people in transition or early adult care is unknown. We consequently evaluated two CF transition services (Lowton et al, 2005) to examine this issue further.

Methods

Fifty young people aged between 13 and 24 years, registered with either centre, were interviewed about their expectations or experiences of transition care. Forty-four of these also completed the CF Quality of Life questionnaire (CFQuol) (Gee et al, 2000). In addition, 23 paediatric and adult MDT members from physiotherapy, dietetics, general and specialist nursing and medicine (from Senior House Officer to Consultant), from both centres were interviewed individually about their expectations and experiences of their service.

Although some young people did appear to be in ‘denial’ about the severity of their condition, most had a generally positive attitude towards living with the disease and outlined future plans. However, three main concerns were noted. First, young people questioned how leaving the family home for university or work might affect easy access to a specialist adult MDT. Second, concerns were raised about the impact of CF on current and future intimate relationships, with uncertainty surrounding their fertility. Third, anticipated future health concerns centred on the possibilities of organ transplant and reduced life expectancy. In particular, many young people reported having been affected by misinterpreting media information, early deaths of others with CF, or internalising fears of dying young. Whilst one centre offered young people out-of-hours access to specialist nurses, and young people generally perceived CF nurses as approachable, some reported hesitancy in contacting staff they knew well about these issues.

Quantitative data confirmed interview findings. The majority (57.9%) of young people interviewed scored less than 50 on the *future concerns* domain of the CFQuol, indicating some concerns, particularly in relation to life expectancy, organ transplantation and fertility.

Similar proportions of those yet to transition, compared to those who had, achieved less than 50 in this domain (58.8% v 57.1%).

As more young people with CF survive into middle age, when and how to address their concerns surrounding a longer-term future requires the MDT to consider what support is in the young person's best interests both at and after transition. Although MDT members reported being aware of patients' worries, staff from both centres reported that psychological input was more easily available to paediatric patients than adults. Furthermore, how psychological needs of young people were addressed during transition varied between MDT members. Together with personal attributes and opportunities of space and time, there was interplay between the young person's perceptions of their relationship with MDT members, and professional's perception of the situation. For example, several young people described the specialist CF nurses to be so 'like family' that they did not want to 'bother them' with their worries. Similarly, the CF nurses appeared to perceive their role as protective towards the young people, which may have inhibited them from encouraging expression of worries. In contrast, although regular one-to-one physiotherapy sessions were described by one therapist as 'intimate', the relationship that young people reported having with physiotherapists was not perceived as familial. Some young people therefore appeared to use physiotherapy sessions as an opportunity to talk about their concerns, including end-of-life issues and funeral planning. The way in which physicians addressed young people's concerns also appeared to depend on how they were perceived by the young person and their reaction to this.

Discussion

Currently the MDT appears to address young people's concerns for their future both reactively and informally, either through staff members having rapport with individual young people who feel able to express their concerns, or coincidentally through the nature of the healthcare professional's role, rather than it being part of a standard package of care. The decision not to address certain issues during transition or into adult care may be based on the MDT's clinical judgement of appropriateness and the intention of acting in the patient's best interests. Whilst there is some evidence that young people's avoidant or repressive coping strategies may be associated with a better psychological outcome (Oxley & Webb, 2005), these may become detrimental to health, and can create conflict for practitioners who have a duty of care.

Staff do report emotional difficulties in discussing dying and death with this patient group (Lowton, 2005), and the uncertainty of the disease course coupled with vast differences in end-of-life approaches, ranging from organ transplantation to hospice care, serve to complicate attempts to construct any clear MDT policy. That some staff report difficulty in identifying when and how to initiate discussion, and young people report internalising their worries, suggests a need to review how MDT response and resourcing can be optimised in adult CF services. As adults begin to outnumber children with the condition, members of the MDT, whatever their professional background, need to be supported to find more systematic ways of identifying when the help of a team member might be of use to the young adult with concerns for their future wellbeing.

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