Characterising extreme survival in cystic fibrosis: A case series of over-50 year olds

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Objectives: The median age of survival in patients with CF continues to rise. There is a growing cohort of patients older than 50. Their needs may differ from the general adult CF population. This study focuses on this cohort and aims to describe their characteristics and differences from the general adult CF population.

Methods: A case series of patients (n=52) from one UK adult CF centre that had reached age 50 yrs without transplantation. Hospital records were examined and comparisons made with the latest UK CF Trust annual data report.

Results: The median age in the cohort was 52.7 years (range 50.1–73.0); 57.7% were male and 50% were diagnosed as adults (≥16 years old). The presence of homozygous AF508 was lower than the general CF population (30.8% vs 52.6%, p=0.002) and heterozygous AF508 was higher (59.6% vs 38.7, p=0.002). The presence of the class V mutation, c.2657+5G>A, was higher (3.8% vs 0.3%, p=0.01). Pancreatic insufficiency (71.2% vs 87.4%, p<0.001) and ABPA (1.9% vs 11.2%, p=0.03) were less common. The presence of CF-related diabetes (32.7% vs 29.4%, p=0.6), NTM (3.8% vs 4.6%, p=0.99) and liver disease (3.8% vs 4.2%, p=0.99) was similar. Patients required fewer days of IV antibiotic therapy over the 12 month period prior to data capture (median days: 14 vs 24, p<0.01) and had a lower %FEV1 (median: 50 vs 65.1, p=0.01). BMI did not differ (median: 22.7 vs 21.9, p=0.13).

Conclusion: Extreme survival is increasingly recognised in cystic fibrosis. There are important differences between this cohort and the general adult CF population – as such their healthcare needs should be tailored accordingly.

Change pathways: The power of engaging in partnerships to change and enhance care for adults with cystic fibrosis

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Today as health professionals, we are seeing many and varied changes in the care provided for those with cystic fibrosis (CF), together with the options available for the provision of that care. Providing services that will cater to the needs and demands of this cohort of patients is a major challenge. Seven years ago, the Royal Adelaide Hospital CF Service embarked on a pathway of change by engaging in partnerships.

Objectives:
- To understand the main issues from key stakeholders
- To develop and implement processes to address the main issues
- To assess implemented changes in care provision

Methods:
- A Statewide Service was established to facilitate practice change
- The Department of Health commenced a consultative process with consumers and their families
- Partnerships were developed with various stakeholders
- Feedback was sought from consumers and clinicians regarding the current process of service delivery

Outcomes: As a consequence of engaging in positive and appropriate partnerships a unique document was developed to guide clinicians and consumers when negotiating a pathway of change. To facilitate the recommendations outlined in this document, additional funding was sourced; existing resources were increased; the multi disciplinary team expanded, with a resulting increase in activity in relation to all aspects of care provision.

Conclusion: The engagement of partnerships is a powerful tool which can effect change and improved care to a client group. Our story continues to ensure favourable changes are made to improve and maintain positive outcomes and consumer satisfaction for South Australian patients with CF.

Improving communication between adults with cystic fibrosis and their CF physician

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Background: Education of patients with CF is time consuming. A questionnaire, entitled “words to say”, has been elaborated by the French working group in patient education (GETHEM) and designed to allow patients to quickly express their expectations in 8 different fields.

Methods: This prospective study was conducted from October to December 2012 in 100 consecutive patients who completed the self-administered questionnaire during an outpatient visit. They were asked to check words corresponding to the themes they wanted to discuss with their doctor.

Results: Eighteen patients did not answer, claiming they had no question. 48 males and 52 females completed the questionnaire. Their mean age was 34±9 y.o. (18–69), their mean FEV1 was 52.4±19.7% pred. Ten patients were students, 56 were in the labour force and 34 were inactive. Sixty patients were living in couples and 30 had children. The topics of interest were: discomfort associated with CF (n = 86), social and professional life (n = 64), treatment (n = 52), current psychological feeling (n = 51), family life and married life (n = 39), organizing the day (n = 33), procreation (n = 29) and sexuality (n = 19). Among 37 different words, 8 were ticked by more than 25% of the patients: fatigue (n = 59), pain (n = 40), treatment efficacy (n = 38), cough and sputum in terms of discomfort in social life (respectively n = 36 and 31), mood (n = 32), social rights (n = 31) and job (n = 27).

Conclusion: This self-administered questionnaire improves communication between adult CF patients and their caregivers. Their main concerns are the inconvenience associated with CF (primarily fatigue and pain), treatment efficacy and procedures to keep or find a job.

Education and employment: A qualitative exploration of the beliefs, aspirations and experiences of young people with CF

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Objectives: Attainment of age-related developmental tasks in education and employment are now achievable for people with CF, but many still struggle to fulfill their potential. This research explored some of the reasons.

Methods: 6 young people with CF diagnosed before 2 years (3 females, mean age 23) and not currently in full-time employment were interviewed about their educational and employment experiences. Interviews were analysed using Grounded Theory.

Conclusion: All recalled childhood aspirations for education and work, and experiences in trying to achieve them. Four themes emerged:
1. CF caused repeated absences from education and workplace because of routine appointments, illness and time spent on treatment. Impact on education increased with age as they ‘fell behind’ academically and socially from peers;
2. Influenmental others had potential to alleviate or aggravate the impact of CF, with some being understanding and supportive, and others unhelpful, discriminatory and bullying;
3. Participants’ own concerns about health and life expectancy sometimes led to ambivalence about achieving life-goals;
4. Repeated experiences of failure and feeling ‘let down’ by others led to learned helplessness resulting in a resigned withdrawal from efforts to obtain educational qualifications or develop careers.

This research highlights the practical, physical and emotional barriers that young people with CF continue to face in achieving the same success in education and employment as their healthy peers. Teams need to be proactive in supporting the patient in school and workplace, and be alert to the possibility of learned helplessness following disappointment.