As cystic fibrosis (CF) is an incurable disease, it has an impact on patients, their families. These children are at greater risk of having significant emotional or behavioural problems. Family demographics also have impact on their quality of life.

Aims: To assess demographic measures and factors which affect psychosocial status of our patients.

Methods: Parents of CF children completed a questionnaire on age at diagnosis, adherence to routine visits and treatments, occupational and school problems, social activities, family structure and income, living conditions and problems with health insurance.

Results: Data of 85 CF patients aged between 4 and 368 months (median 84 months) were obtained. Median age at diagnosis is 4 months. 44.7% were not going to school. 41.2% were at elementary education, 3.5% attending to university. 77.9% had 4 or more routine clinic visits annually. 62.4% had problems at school. 14.1% had psychological assistance. 35.3% had social activities like sports, music, painting and dancing. One patient smoked cigarettes and 57.6% of parents smoked cigarettes at home. 30.6% didn't apply to their diet and 23.5% to physiotherapy.

Their families consisted of median 4 persons; at least one person had a job and median income was $494. 18.8% of mothers and 31.8% of fathers were graduated from university. 1.2% of mothers had no formal education. 35.3% lived in rented houses and 42.4% used stoves for heating up their houses. 96.4% of patients' health expenditures were covered by government.

Conclusion: Cigarette smoking, noncompliance with physiotherapy and diet, and family education are problems in Turkish CF patients. Information and support should be available to patients and their parents, to assist them in coping with this chronic illness.

Cultural restraints and barriers to physical activity for male CF adults

D. Greenop, M.J. Walshaw, M.J. Ledson. Regional Adult CF Unit, The Cardiothoracic Centre, Liverpool, UK

Introduction: Physical exercise is seldom a solitary activity but requires the participation or presence of other people, and as a social activity it is hypothesised that it is linked to the dominant cultural discourses of health and fitness. While the social structure of femininity militates against female participation in rigorous physical exercise, such activity is more acceptable to males who have less resistance to this aspect of self-care. We aimed to explore the material, social and cultural barriers to physical activity for adult CF males.

Methods: Using qualitative techniques, an independent research student (DG) compared the attitudes to exercise of 20 male CF adults (mean age 26 [range 18–43]) with 20 females (mean age 25 [range 18–39]) who had similar lung function (group data: 5 normal FEV1, 13 mild impairment, 14 moderate and 9 severe) selected by the CF team.

Results: While males with mild disease were more physically active than similar females, this trend reversed with increasing disease and the ability to exercise rigorously ceased. Of the 17 participants with an FEV1 < 50% predicted (8 male), no males used public health promoting facilities, carried out housework, shopping, or went dancing, only 2 exercised through walking, and 5 did no exercise at all (for 9 females: 5, 2, 3, 2, 6, and 3 respectively).

Conclusions: Although this study supports the above hypothesis, it is apparent that adult CF males are influenced by cultural discourses of masculinity that may militate against participation in less rigorous physical activities such as walking. Thus, while the social construction of masculinity favours males when well, it disfavours them when unwell. The dominant masculine discourse creates a stigmatising effect (inducing insecurity/loss) for many adult males as their CF advances.

A review of pulmozyme use in children on Teesside

P. Lindsey, E. Hampton, S. Kumar. James Cook University Hospital, Middlesbrough, UK

Background: Following the Pulmozyme Early Intervention Trial (Quan et al 2001) our use of Pulmozyme in children increased.

Aim: To establish parents’ perceptions of their child's health whilst using Pulmozyme and to identify antibiotic use for one year before and after starting treatment.

Method: Children under 18 who commenced treatment within the last 5 years and who had been diagnosed at least 1 year before starting Pulmozyme were included. Routine lung function measurements were documented. During clinic visits or routine telephone contact parents were questioned regarding their child’s use of Pulmozyme. Questions related to timing of administration, their perception of concordance with treatment and their child’s health status. A review of the case notes identified the antibiotic treatment of exacerbations.

Results: Thirty six (57%) of patients attending clinic had been prescribed Pulmozyme. Of these 24 fitted the above criteria. Fifteen (63%) maintained an FEV1 > 75% throughout the review. 83% of parents were questioned. Timing of Pulmozyme administration ranged from 30 minutes to 8 hours pre physiotherapy with 67% taking it immediately on return from school. Parents and children found concordance easy and reported significant subjective benefits. Complete information on antibiotics was available for 18 patients, too few for statistical analysis. There were a median of 4 oral and 2.5 intravenous antibiotic courses before treatment and 2.5 and 2 after. Fifteen of 18 patients had a reduction in at least 1 type of antibiotic treatment.

Conclusions: Parents perception of their child’s health status during treatment with Pulmozyme was positive. This is reflected in maintenance of FEV1 and reduction in antibiotic use. Pulmozyme is easily adapted into family routine and concordance is reportedly high in comparison to other treatments.

Adherence to airway clearance therapies in patients with Cystic Fibrosis

A.C. Modi1, M.K. Sontag2, J.M. Koenig2, F.J. Accurso2, A.L. Quittner2 & Investigators and Coordinators of the Airway Secretion Clearance Study. 1 Cincinnati Children’s Hospital Medical Center; 2 University of Colorado at Denver and Health Sciences Center; 3 University of Miami, USA

Airway clearance therapies (ACT) are important in clearing mucus from the lungs of patients with CF. We examined adherence rates among patients randomized to three ACTs: postural drainage and percussion (PD&P), the FlutterTM and high-frequency chest wall oscillation (HFCWO). This is one arm of a multi-center study of ACT.

Adherence data was collected on 139 patients, (M age = 14.2 yrs; 54% male; M baseline FEV1 % predicted = 82.2%). Patients were stratified by age (7–12, 13–17, and over 18 yrs) and randomized to PD&P (31%), Flutter (39%), and HFCWO (39%). Every 4 months patients completed a Daily Phone Diary, a computerized 24-hr recall of daily activities, including ACT adherence.

We found differences in adherence after randomization for all patients using PD&P (64%) vs. both Flutter (47%, p < 0.01) and HFCWO (54%, p < 0.10). Differences were also found between Flutter (49%) and HFCWO (60%) at 1-yr follow-up (p < 0.10). We found differences in adherence for adolescents after randomization, between Flutter (35%) and PD&P (69%, p = 0.03), but not HFCWO (50%). Adherence for Flutter (32%) was significantly lower than PD&P (81%, p = 0.03) and HFCWO (66%, p < 0.01) 1-yr later. No statistical differences were found between ACTs for children or adults.

Data suggest that ACT type does not appear to affect adherence in children and adults. However, adolescents appeared to be more adherent to HFCWO and PD&P compared to Flutter, and adherence increased over time for these ACTs. Better adherence to these treatments may be related to the amount of supervision by parents of adolescents.

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