231 A study of treatment activities and perceived treatment burden in adults with cystic fibrosis

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Objective: Advances in cystic fibrosis (CF) management over the past 20 years have lead to an increase in life expectancy such that CF is emerging as a chronic disease requiring complex treatment strategies. In this study, we investigated the relationship between daily treatment activities and perceived treatment burden in CF adults from a single UK centre.
Methods: A questionnaire was designed using the CF Questionnaire-Revised (a CF-specific measure of healthcare-related quality of life) to measure self-reported daily treatment activities and percentage perceived treatment burden score (PPTBS), with higher scores representing greater perceived treatment burden.
Results: Among the twenty respondents, the mean reported time to complete daily therapies was 130 minutes (range 0–480 minutes) and the median total number of daily medications was 5 (range 1–8). The mean PPTBS was 60% (SD 16%), with no significant difference in PPTBS based on gender or age. In a multivariable model, PPTBS trended towards a negative association with FEV_1 ($p = 0.08$) however was not significantly influenced by the total time spent on treatments or number of daily medications (total, nebulised or oral).
Conclusion: Daily treatment activities exhibit a broad range which may reflect variability in disease severity and/or treatment adherence in the respondent group. Interestingly, perceived treatment burden did not appear to associate with the total time spent on daily treatments. The observed trend towards a negative association with FEV_1 suggests that factors other than the time burden of treatment regimen act to increase PPTBS in CF adults with more severe lung disease.

232 The paediatric cystic fibrosis service at the Leeds Children's Hospital – a patient and parent perspective

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Objectives: To undertake a Quality Improvement Questionnaire of parents attending our paediatric CF Centre, and compare results with our staff's perceptions of our service.
Methods: A modified version of a questionnaire used by the Stanford CF center (CA) was used. A similar questionnaire was used to survey the staff. In addition there was opportunity for people to give their opinions on good aspects of the service and suggest ways in which it could be improved.
Results: A total of 122 (76%) completed questionnaires were obtained from the patient group and a total of 10 (56%) from the staff. 99% in the patient group agreed or strongly agreed in recommending the service compared to 90% in the staff. 87% in the patient group had aspects of the service that they felt stood out compared to 90% for staff. Patients liked the friendly, approachable and knowledgeable staff and the timely advice they received where staff felt the expertise and MDT working of the team stood out. 30% of the patients had suggestions for improvements compared to 60% of staff. Patients saw seeing the same doctor, shorter waiting (average clinic time 65mins) and more flexible appointment times as issues. Staff wanted more physical space in clinic and better staffing levels. Only 45% of patients were offered genetic counseling with the staff feeling 90% would have been.
Conclusion: Both patients and staff show a high level of satisfaction with the service with less than a third of patients feeling improvements could be made. Patient priorities for potential improvements do not necessarily concur with those of staff. Moving forward we must balance these to further enhance the service.

233 Three-year experience of a new clinic dedicated to the care of screened infants with equivocal CF related diagnosis

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Objectives: CF newborn screening, whilst detecting infants with classical CF, has also led to the detection of infants with borderline sweat chlorides and rare CFTR mutations. Uncertainty remains over the best diagnostic classification and management of these infants with local, regional and national variations in practice. 3 years ago we established a specific clinic for such children in our area that we named "The CFTR Variant Clinic".
Methods: We present our 3 year experience of this clinic.
Results: We currently follow 10 children aged 1–11 yrs within this clinic [genotypes F508del/R117H(7T) (×4), F508del/intron 8 polyT 5T (×2), F508del/C866Y, F508del/R170H, G542X/P988R and p.Val456ala/p.Tyr569Asp]. All initial sweat chlorides were normal or equivocal (25–58 mmol/l): one became abnormal after interval repeat. The children old enough to do spirometry all have FEV1s of 80–120% predicted. Not all chest x rays have remained normal. One child needed surgery as an infant for a meconium pseudocyst. All children are pancreatic sufficient and are following normal growth centiles. Some children have had repeated bacterial isolates (predominantly *S. aureus*, *H. influenzae* and *S. pneumoniae*) from respiratory secretions. No child has ever isolated *Ps. aeruginosa* and none has needed intravenous antibiotics.
Conclusion: We believe that diligent long term follow up of such screened infants is warranted. This should be provided by teams with experience in CF. We believe that by sharing longitudinal clinical outcome data between centres and between nations we will ultimately be able improve the current difficulties faced by both clinicians and families when considering these children.

234 Cystic fibrosis: new trends in ophthalmological evaluation

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Background: Cystic fibrosis (CF) is characterized by hypoxia that affects several organic tissues. Retinal ganglion cells may suffer for the hypoxic status, and this may lead to alterations of retinal nerve fiber. To date less is known about the relationship between hypoxia and the retinal activity.
Methods: Twenty-two eyes in CF patients (mean age 30±13 SD years old) were analyzed. A complete ocular evaluation and visual field exams of the 30 central degrees were done using the Frequency Doubling Technology Perimetry (FDT). FEV1%, SpO_2%, and Ht% have been calculated. FDT has allowed to evaluate the Mean Defect (MD) and Pattern Standard Deviation (PSD) of each exam.
Results: We found a relationship between: MD and HT% (r value –0.18), MD and FEV1 (r value –0.68), and MD and SpO_2% (r value –0.08). Moreover, there were correlations between: PSD and HT% (r value 0.29), PSD and FEV1 (r value 0.71), and PSD and SpO_2% (r value –0.31).
Conclusions: There is a statistically significant correlation between FDT alterations and retinal trophism. The oxygen supply alterations might determine hypoxia of the ganglion cells causing a decrease of receptive activity. This method could be useful to evaluate the progression of the CF disease.