398 Cystic fibrosis and tobacco smoke exposure

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Introduction: The prognosis of CF patients is mainly influenced by lung affection. Literature described that passive tobacco exposure increases the risk of pulmonary infections and exacerbations, and the deterioration of lung function and nutritional status.
Methods: We realized a survey in our CF unit to evaluate tobacco exposure in our young patients and sensitize their parents about the harm of smoking.
CF unit nurse assessed tobacco exposure using a questionnaire addressed to parents about smoking (age of first cigarette use, number of pack per day, smoking during pregnancy or child presence).
Results: Fifty nine patients were included [mean age:10.5 (0.4–18) years, 31 patients ΔF508/ΔF508, mean FEV1: 78.9% (±24), 5 patients chronically colonized by *Pseudomonas aeruginosa*]. One hundred ten parents answered the questionnaire. Thirty three percent of our patients (36/110) had at least one smoking parent (11% smoking mother, 22% smoking father). The majority was smoker since adolescence. Diagnosing CF pushed one parent to begin smoking while 6 other parents stopped smoking after the diagnosis was made ($p < 0.05$).
Conclusions: CF patients' parents seemed not to be enough informed about the risks of tobacco exposure. They should receive the proper medical and psychological support to encourage them quit smoking. In our survey, which is a small sample, the majority affirmed not smoking in the presence of their child but this could not be confirmed by a simple questionnaire. A larger cohort should be studied including urinary nicotinine levels. Beside improvement in life expectancy brought by new treatments available, smoking cessation should be considered in our management.

399 Progress in cystic fibrosis transition care in Canada

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Transition from pediatric to adult health care is a major life event for patients with a chronic disease such as Cystic Fibrosis (CF). Transition care is the work undertaken to prepare youth for the adult health care system and the management of their health condition in adulthood. In Canada, CF transition care is often "championed" by nurses, but it is unknown if, or to what extent, it is practiced in individual CF clinics. We surveyed by questionnaire all 27 Canadian pediatric CF clinic nurse-coordinators; 23 surveys (85%) were completed and analyzed.
All clinics reported transfer of patients to a distinct adult CF clinic at or about age 18 years, with rare exceptions causing delay. Although almost half of the pediatric clinics share one or more team members with their local adult clinic, most responding clinics view adult CF care as distinct from pediatric care, and approximately 75% of pediatric clinics engage in practices to help prepare their patients for adult CF healthcare. However, of these, only 22% follow a formal protocol with set goals, and documenting progress. The remainder either follow an "informal" transition program with a wide variety of practices in whom it is not possible to determine whether patients are receiving adequate preparation for adult care; or (in 25% of clinics) no formal or informal transition program.
Survey results were presented at a Canadian CF nurse's meeting. The commitment to providing CF transition care was reaffirmed and CF clinics with formal protocols were identified to serve as sources of information and inspiration for clinics interested in formalizing their own approach to transition care. At least two CF clinics are now in the process of developing their own programs. The survey has identified gaps in CF care in Canada and served as a stimulus for improvement.

400* Quality of life in women with cystic fibrosis and urinary incontinence

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Introduction: Urinary incontinence (UI) is well recognised in females with cystic fibrosis, however previous studies have not compared UI with quality of life (QoL).
Aim: To assess the impact of UI on QoL in women with CF.
Methods: Women with CF aged >17 yrs, attending the Royal Brompton Hospital Adult CF Centre were invited to complete two anonymous postal questionnaires: the CFQoL, and an assessment of occurrence, severity and impact of UI [using a 10 cm visual analogue scale (VAS)].
Results: Eighty-eight women aged 17 to 40+ yrs returned completed questionnaires. Age of menarche (mean: 13.8 yrs, range 9–22 yrs), menopause ($n = 5$), and pregnancies over 24 weeks gestation ($n = 30$) were recorded. 68% of women reported UI, with 45% of these reporting that it occurred at any time and 5% experienced whole bladder emptying during a chest infection. The VAS showed a mean score of 3.8, with only 15% of respondents requesting help for their UI. Precipitating factors included coughing (95%), sneezing (42%), laughing (38%) and huffing (23%). CFQoL results showed significantly lower scores for women with UI compared to those without, in a number of domains including chest symptoms ($p = 0.006$), social function ($p = 0.02$), interpersonal relationships ($p = 0.02$), emotional responses ($p = 0.02$) and body image ($p = 0.02$).
Conclusion: As with previous studies this population report a similar incidence of UI (68%), with coughing being the commonest cause. Although perceived as a relatively minor problem (low VAS score), women with UI report this as a significant impact on many aspects of their QoL. However, in spite of this impact on QoL, many women are reluctant to be investigated or treated for UI. This may in part be due to the perception of the problem compared to having CF and the already large burden of treatment in CF.

401* Optimal delivery of end of life care for CF patients, families and staff

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Adults with CF face challenges including the ongoing hope for lung transplantation (Tx) until the final days of life. Little information is available to inform a CF-specific, palliative care (PC) model. We performed a multi-phase study of the differential patient, family and staff experiences to develop a PC CF model of care. Surveys were derived from focus groups. Patients ($n = 82$), bereaved family members ($n = 18$) and CF staff ($n = 39$) completed the surveys using analog scales. Patients and families had less understanding of PC yet supported it as part of CF care. All preferred a sequential rather than a parallel model, where PC and Tx planning co-occur. Patients actively tried not to think about end of life as health deteriorated. Information that was direct and honest, covering what to expect and options for care and written information was supported. Timing of discussions were important and at Tx referral was preferred. For staff, tensions between providing comfort to patients vs active treatment in the hope of transplantation was apparent. Training to manage the emotional aspects is needed. The development of a PC and CF model of end of life care requires further information and counselling support, earlier in the course of their illness than previously recognised. Differences in unmet needs, expectations for care and clinical outcomes exist across the groups.
Supported by: The Australia CF Trust.

Table 1. Median score out of 10 or % predicted FEV1

	Patients (82)	Families (18)	Staff (39)
PC part of care	8	10	10
PC knowledge limited	6	6	3
PC only if no Tx	2	4	1
CF team to initiate discussions	5	5	5
CF to discussions at FEV1	20%	30%	30%