

**Objective:** To evaluate exercise capacity using gold standard objective measure, Cardio Pulmonary Exercise Test (CPET) and HRQoL using Cystic Fibrosis Questionnaire Revised (CFQ-R) in a cohort of Danish people with CF after more than a year of receiving treatment with ETI.

**Materials and methods:** Retrospective cohort including people with CF from the age of 12, receiving ETI for more than a year. CPET and CFQ-R is performed at baseline and follow-up. Descriptive statistics are presented including paired sample t-test. Respiratory outcomes VO<sub>2</sub> peak (ml/kg/min) and VO<sub>2</sub> peak (ml/min) and scores from CFQ-R will be evaluated.

**Results:** 156 persons performed CPET at baseline and follow-up. Mean follow-up time 16 month (10–22 month), mean age 29.5, 51% female and 59% with chronic infections. Mean BMI and FEV<sub>1</sub>% pred at baseline 23.0 and 74% respectively. Mean VO<sub>2</sub> peak (ml/kg/min) at baseline and follow-up 31.9 and 32.5 respectively. Mean diff 0.6, 95% CI [–0.03; 1.25], p = 0.06. Mean VO<sub>2</sub> peak (ml/min) at baseline and follow-up 1972.5 and 2137.6 respectively, mean diff 165.1, 95% CI [123.7; 206.5], p < 0.001.

**Conclusion:** The results show a borderline significant change in VO<sub>2</sub> peak (ml/kg/min) and a significant change in VO<sub>2</sub> peak (ml/min) after mean 16 month ETI treatment. The results are preliminary when submitting this abstract and will be presented at ECFS in Vienna with more results included.

#### WS01.05

##### Development of a musculoskeletal screening tool for children and young people with cystic fibrosis (Addenbrooke's MST): initial findings

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**Objectives:** Development of the Addenbrooke's Musculoskeletal Screening Tool (MST) for children and young people (CYP) with CF.

It is recommended by the Association of Chartered Physiotherapists in CF that Musculoskeletal (MSK) screening is carried out in all children from 7 years of age. Currently there is no specific tool for CYP with CF, only the Adult Manchester MST is available in CF (Ashbrook, Taylor and Jones, 2011).

**Methods:** A paediatric MST was constructed by reviewing the Manchester MSK Screening Tool, the pGALS and recent literature surrounding both paediatric and CF related MSK conditions. The tool was developed with support from paediatric CF specialist physiotherapists, paediatric MSK specialist physiotherapists and respiratory consultants. This tool was then used over a one year period on a total of 58 CYP.

**Results:** The MST was well accepted by clinicians and CYP, taking up to 5 minutes to complete. There were 81.8% more positive MSK screens in the year using this Addenbrooke's MST (20) compared to the previous year using the Manchester MST (11). There were also 6 more referrals, all deemed appropriate by MSK specialist physiotherapists. MSK advice was given to 83% more children in the year using AMST (11) compared to the year using MMST (6). Urinary incontinence appears to be under reported in this population when comparing to other studies with only two positive screens.

Kyphosis (as diagnosed by plumb line) was particularly prevalent in this population (20%) and those with kyphosis had a significantly reduced FEV<sub>1</sub>% (p = 0.008) and FVC% (p = 0.034), as well as tighter pectoralis major (p = 0.002).

**Conclusion:** A screening tool designed for CYP with CF is important in identifying specific conditions to this population. There is an increased number of appropriate referrals when using this tool and interesting initial evidence on the use of pectoralis major length as an outcome measure, however further research and validation is required.

#### WS01.06

##### Inducing sputum in the adult cystic fibrosis post-modulator era

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**Objectives:** CFTR modulators have dramatically changed the landscape of CF, leading to reduced frequency of available sputum sampling for

pathogen surveillance. Induced sputum (IS) samples via a variety of physiotherapy methods present an alternative to cough swabs to monitor microbial growth. The aim was to evaluate the efficacy of an IS service at the All Wales Adult CF Centre (AWACFC) for microbial surveillance.

**Methods:** Patients underwent an IS if they were unable to provide a sputum sample within the previous 12 months. Each session took approximately 60 minutes of physiotherapy time. Methods were chosen by the patient and included exercise, Metaneb<sup>®</sup>, oscillatory PEP and manual techniques. Retrospective data was collected from 15 months of IS sessions and cough swabs in Cardiff including method used, samples produced and microbial yield.

**Results:** A total of 45 IS were completed. Success rate for the patient producing at least 1 sample was 87%. Exercise combined with hypertonic saline was the most common method used (N = 30). Exercise had the biggest success rate versus other IS methods combined (80% vs 71%). 74% of IS samples isolated at least 1 specific microbe compared to 16% of cough swabs from the same cohort. Isolates grown included *Mycobacterium chelonae*, *Pseudomonas aeruginosa*, *Haemophilus influenzae*, *Staphylococcus aureus* and *Stenotrophomonas maltophilia*. 1 patient reported vomiting post IS, but otherwise all other sputum inductions were tolerated well with no other reported adverse events.

**Conclusion:** IS resulted in increased microbial surveillance in the absence of spontaneously produced sputum. IS was well tolerated with a high success rate. Exercise was the most common method used and had a higher success rate. Sputum induction sessions also presented an opportunity for the physiotherapy team to evaluate airways clearance techniques.

## WS02 – Fertility, pregnancy and gender-related topics

#### WS02.01

##### Decreased fertility in female cystic fibrosis patients: peering into the endometrial factor using cutting-edge organoid models

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Female CF patients suffer from sub-to infertility, often facing problems to become pregnant. Underlying reasons remain understudied. In particular, it is largely unknown whether dysfunction of the endometrium is involved, the womb's inner lining and key tissue for embryo implantation and development. This gap is mainly due to lack of appropriate study models. Therefore, we develop and apply tissue-mimicking organoid models to in-detail decipher molecular and functional aberrations in fertility-deficient CF endometrium as compared to healthy (fertile) endometrium. First, we established endometrial organoids (EMO) from the *Cftr<sup>tm1Eur</sup>* mouse model. These organoids show a smaller lumen than wildtype (WT) EMO, and do not swell in the FIS assay, both validating the CFTR defect. Currently, we are comparing expression of endometrial functionality/fertility markers in CF versus WT EMO, as well as their responsiveness to estrogen (E2) and progesterone (P4), which *in vivo* regulate the estrous cycle. Second, we develop(ed) organoids from CF patient endometrium. Before, we have shown that EMO from healthy endometrium can reliably reproduce all menstrual cycle phases under defined E2/P4 exposure. Currently, we are deciphering whether and how menstrual cycle phases, including the receptivity stage, are different between CF and healthy EMO. We already observed dissimilarities in the proliferative (E2) and secretory (P4) phase, such as increased apoptosis and decreased fertility marker expression, respectively. Taken together, we establish(ed) organoid models from CF endometrium as novel and powerful tools to gain insight into the endometrial factor in CF fertility deficiency. Importantly, the organoids will