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YOU WOULDN’T BEAD ABOUT IT!

BENJAMIN GERHARDY, Lucy Burr, Rebecca Keating

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We present the case of a 27 year old male, F508 homozygote genotype, who presented with major haemoptysis. A computed-tomography angiographic study of the bronchial vessels did not reveal a specific source of bleeding. He underwent two bronchial artery embolisations over three days but fresh haemoptysis persisted after these interventions. Bronchoscopy demonstrated fresh blood at the right upper lobe orifice and he subsequently had a third bronchial artery embolisation directed at this area, which led to cessation of the haemoptysis. Reinstigation of chest clearance therapies in the following days led to expectoration of particles within his mucus. Microscopic examination of these particles confirmed that they were the Embozene® embolization particles, both 500micrometer and 700micrometer diameter (identifiable due to colour differences). These beads were continuously expectorated over approximately 1 week without any relapse in bleeding.

To the best of our knowledge and a literature search this is the first documented case of microbead expectoration following bronchial artery embolization.
NEBULISED TRANEXAMIC ACID: A REVIEW OF THE FIRST 3 YEARS OF USE

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Introduction: Haemoptysis is common in adults with Cystic Fibrosis (CF). Tranexamic acid (TA) is an antifibrinolytic agent, which blocks binding of plasminogen and plasmin to fibrin, to inhibit breakdown of clots. Recently topical (nebulised) therapy with TA has been shown to reduce haemoptysis in a case series of haemoptysis due to multiple different causes (Chest, 2016). Following these cases, we commenced use of nebulised TA in haemoptysis in adults with CF. This abstract summarises the last 3 years’ experience in adults with CF at Westmead Hospital.

Methods: Data were retrospectively collected from patient records, including time, volume, precipitating factors of the haemoptysis, together with the details of the TA utilisation. Adverse effects and any changes to patient’s usual therapy were noted.

Results: 26 CF adults, 14 males, age 27.0±8.1 (mean±SD) years with an episode of haemoptysis were trialled on nebulised TA, in addition to usual management. TA was found to be generally well tolerated, with chest tightness being reported in 6 patients, resolving with bronchodilators. Resolution of haemoptysis occurred in the majority of patients within 48 hours. A home haemoptysis management plan, including guidance for further TA therapy, was provided to 19 adults with CF for outpatient use.

Conclusion: This anecdotal case series supports the use of TA in the management of haemoptysis in adults with CF. Further randomised controlled studies in adults with CF with mild-moderate haemoptysis are required.
SAYING "NO" TO PBS LISTED ORKAMBI? UPTAKE IN A TERTIARY CF CENTRE

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Introduction: In October 2018, Lumicaftor/Ivacaftor (OrkambiTM) was listed on the Pharmaceutical Benefits Scheme (PBS). A 6-month systematic roll-out of this drug was undertaken at Royal Prince Alfred Hospital, Sydney, Australia. This retrospective review describes the uptake of OrkambiTM in our CF clinic cohort and also provides an explanation on those who declined it.

Methods: Two hundred and forty pre-transplant patients were screened from our clinic cohort. Those who were homozygous for the Phe508del mutation were provided an overview on OrkambiTM by CF physicians and nursing staff during their routine clinic appointments. This included a review of the modest benefits on lung function, potential side effects and complications, contraindications, and the importance of contraception in females. Patients were also informed that they needed to be enrolled in the Australian CF Data Registry.

Results: Ninety-eight patients were homozygous for the Phe508del mutation (40%). By April 2019, 54 of these patients (55%) had been initiated on OrkambiTM. Due to liver disease (n=6) and very poor lung function (FEV1<20%, n=1), 7 patients (7%) were not commenced on it. Sixteen patients (15%) are awaiting stabilisation of Liver function results, gastroenterology or psychiatry review, or further discussion on contraception/breastfeeding prior to commencement of OrkambiTM. A total of 5 patients (5%) declined the drug as they were contemplating pregnancy in the next year (n=1), refused contraception (n=1) or preferred not to initiate it due to its modest benefit on lung function (n=3). An additional 16 patients (16%) were informed about OrkambiTM but have not returned to clinic for review. Thirteen patients (13%) experienced side effects and discontinued the drug.

Conclusion: Only a minority of patients in our centre said “No” to OrkambiTM due to refusal to initiate contraception, consideration of pregnancy and modest effects on lung function.
CNS IMAGING STUDIES IN CYSTIC FIBROSIS PATIENTS PRESENTING WITH SUDDEN NEUROLOGICAL EVENTS

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Background: Cystic fibrosis (CF) is a multi-system disorder, with lung disease the major cause of morbidity and mortality. Depending upon genotype, individuals with CF experience a range of extrapulmonary disease that significantly adds to their morbidity. Acute neurological events may present as an extrapulmonary complication. These events can be secondary to a range of different aetiologies. As life expectancy increases, this range of extrapulmonary disease is affecting consideration for lung transplantation.

Methods: A retrospective analysis of the medical records of CF patients attending a large teaching hospital between 2000 and 2018 was performed. Patients presenting with acute neurological events, excluding headaches, who had MRI Brain imaging were evaluated.

Results: Acute neurological presentations, excluding headaches, were reported in twenty-seven index patients. Of these, sixteen patients had MRI Brain imaging for review. Three patients suffered pathology secondary to vascular events, both ischaemic and haemorrhagic; four patients had evidence of ischaemia or infarction not consistent with a vascular territory stroke and the remaining patients experienced a range of different neurological events. The most common presentation amongst these patients was seizure activity, followed by a transient motor or sensory deficit.

Conclusions: Neurologic complications are recognised amongst individuals with CF. Although rare, they can be secondary to a range of different aetiologies; including hypoxia, vitamin E & K deficiency, antibiotic adverse effects, fluid depletion, use of psychotropic agents or stroke from paradoxical emboli. An underlying cause for the neurological presentations in this report was identified in 12 out of the 16 patients with available imaging. The remaining 4 patients had MRI evidence of recent or remote ischaemia or infarction that was not consistent with a single vascular territory stroke. These findings could be secondary to dysfunctional cell energetics that are present in individuals with CF. Additional studies are required to further evaluate this association.
ASSESSMENT OF HIGH FREQUENCY HEARING LOSS IN ADULTS WITH CYSTIC FIBROSIS

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**Introduction:** Aminoglycoside therapy is an important component of CF management, despite the known risk of sensorineural hearing loss. Currently the incidence of hearing loss in adults with CF at Westmead is unclear. Recently high frequency audiometry has been suggested to be a more sensitive measure of hearing loss. This project describes both normal and high frequency pure tone audiometry, in our adults with CF. In addition, determine if hearing loss correlated with total course of aminoglycosides use.

**Methods:** CF adults were recruited from the CF clinic. In a quiet room, pure tone audiology examined frequencies of 500Hz to 6000Hz, whilst high frequency audiology tested frequencies of 8000Hz to 16000Hz. Hearing loss was determined with a threshold of >25dB hearing level at any frequency in either ear. Those subjects who were given amikacin were analysed separately. The number of tobramycin courses were obtained retrospectively using the hospital pharmacy dispensary. The number of IV tobramycin courses were divided into 3 groups, 0 courses, 1-9 courses and 10+ courses.

**Results:** 140 adults with CF adults, mean age 27 years were recruited from the CF service at Westmead. Of the 5 adults given amikacin 3 noted clinically significant hearing loss. Of the 135 who did not receive amikacin, normal tone audiometry failed to demonstrate any relationship between tobramycin use and hearing loss. In contrast, high frequency audiometry demonstrated that 64 had hearing loss in either or both ears, with a dose response curve when comparing the groups.

**Conclusion:** This study confirms the toxicity of amikacin. Exposure to iv tobramycin is associated with high frequency hearing loss which can be missed when only normal frequencies (500Hz-6000Hz) are assessed. High frequency audiometry should be considered for the assessment of tobramycin toxicity.
STANDARDISATION OF TOBRAMYCIN ADMINISTRATION TO IMPROVE ACHIEVING THERAPEUTIC LEVELS

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Introduction: Therapeutic Tobramycin levels are essential for effective treatment. Often patients require changes to dosing and repeat levels due to sub or supra therapeutic levels. This may be due to inconsistencies in delivery methods and timing of blood collection for levels.

Aim: To evaluate whether standardisation of tobramycin administration & monitoring improve the likelihood of achieving therapeutic levels with initial assay.

Methods: A standardised approach to administering intravenous tobramycin was implemented:
• Stat dose day 1 if administered before 1800hrs.
• Daily dose 0630hours diluted in 30mls fluid infused over 30 minutes plus 10ml flush.
• Maintenance fluids first night if patient fasted for line insertion to ensure adequate hydration (excluding hospital in the home (HITH) patients).
• Levels day 3 of admission (or later if falls on weekend).

Tobramycin levels were compared over an 8 month period in 2016 & 2018, pre and post introduction of the standardised approach to determine whether therapeutic levels were achieved more consistently with initial assay. Adherence to the protocol was assessed. Data analysed using chi square test.

Results: 24 & 46 patients were identified for the 2016 & 2018 periods respectively. Assessment of protocol adherence in the 2018 group revealed that 45/46 (98%) had the correct delivery method of tobramycin, 40/46 (87%) had assays performed on or after day 3, 14/46 (30%) had a stat dose on day 1 and 3/31 (9%) had maintenance fluids first night. In the 2016 group 6/24 (25%) achieved a therapeutic level with the initial assay, compared to 23/46 (50%) in the 2018 group which reached statistical significance (p = 0.04).

Conclusion: Implementation of the standardised approach doubled those achieving therapeutic levels on the first check to 50%. This may be further improved with full adherence to the new protocol.
RECOVERY TO BASELINE PULMONARY FUNCTION AFTER EXACERBATION IN CYSTIC FIBROSIS PATIENTS AT THE PRINCE CHARLES HOSPITAL

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Introduction/Aim: Pulmonary exacerbation (PEXs) occur intermittently in patients with CF, defined by increasing chest symptoms and usually a fall in lung function. Unfortunately up to 25% fail to recover their baseline lung function following PEX treatment. The aim of this study was to determine what proportion of patients achieve a return to baseline lung function within 3 months following admission PEX.

Methods: Retrospective chart audit of every CF hospital admission to The Prince Charles Hospital (TPCH) for 2017. Recovery to baseline lung function was deemed to have occurred if the patients best FEV1 in the 3 months post discharge equalled or was greater than 90% or more of the patients best FEV1 in the 6 months prior to admission.

Results: A total of 133 patients experienced 304 admissions for PEX during the study period. Of these, 14 admissions from 9 patients were excluded because they received lung transplants (n=8 admissions from 6 patients) or died (6 admissions from 3 patients) within 3 months of the PEX. A further 22 admissions from 19 patients had insufficient data to determine recovery status. A total of 268 admissions from 118 patients were included in analyses.

The study population comprised 69 (58.5%) males with mean age 31 (SD 10) years. On their first admission, 105 patients recovered lung function giving a proportion of 0.890 (95% CI: 0.818-0.935). The median number of admissions during the study period was 2. Median length of stay (LOS) was 13 days amongst patients who recovered lung function at the first admission compared to 15 days (p=0.01) and 3 admissions (p<0.001) in those who did not.

Overall, lung function was recovered for 222 of 268 admissions (0.828; 95% CI: 0.767-0.876).

LOS, CRP change, FEV1 3-month post admission, and presence of Aspergillus differed significantly at the 5% level by recovery status. Recovery was less likely if Aspergillus or Achromobacter were present. Of the six organisms of interest (Pseudomonas-mucoid, Pseudomonas, Staphylococcus aureus, Aspergillus, Stenotrophomonas and Achromobacter), the median number present in admissions not recovering was 3 compared to 2 in those recovering.

The most common combination of antibiotics were Tobramycin and Ceftazidime (48.5%). Fosfomycin or Aztreonam were more likely to have been used if recovery did not occur.

Pancreatic status, CF related diabetes and CF related liver disease did not differ between recovery status.

Conclusion: Overall, lung function is recovered in 82.8% of admissions to TPCH adult cystic fibrosis unit in patients having PEXs. This recovery rate is greater than that reported in the literature.
OUTCOMES OF DOMICILIARY INTRAVENOUS AND PHYSIOTHERAPY FOR CHILDREN WITH CYSTIC FIBROSIS - THE WESTMEAD CHILDREN’S COHORT

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Introduction: Domiciliary treatment is attractive in terms of reducing length of stay in the hospital and reducing the risk of nosocomial infections for people with cystic fibrosis. We examined the efficacy of 3 treatment arms in the treatment of a pulmonary exacerbation.

Methods: A retrospective study reviewed 4 years of admissions of children with cystic fibrosis managed at the Children’s Hospital at Westmead between 2015 and 2018. There were 3 distinct categories of treatment-complete treatment in the hospital (hospital), at least 5 days of treatment in the hospital and the remainder at home (mixed) and those receiving treatment completely at home (home).

Results: 246 respiratory exacerbation admissions were divided as follows: Hospital (N: 143), mixed, (N: 60) and home (N: 40). The hospital cohort had lower lung function [FEV1%] and weight on admission and a larger drop in FEV1% from baseline and weight loss prior to admission (P<0.0001). The mixed cohort showed the highest improvement in return to baseline FEV1% at 92% (p = 0.06) while the hospital cohort showed most improvement in FEV1% (P<0.0001) and weight. In terms of predicting the return to baseline, every drop of 1% in FEV1% decreased the odds of improving to baseline by 10% (OR: 0.907 CI 0.875-0.939),

Conclusion: A mixed admission which allows proper assessment to formulate a plan that can be used at home under the continued vigilance of the hospital in the home programme provides better outcomes in terms of returning baseline lung function. The sickest patients (hospital) showed more improvement in terms of lung function and weight gain whilst the mildest gains were made in the less unwell patients (home). Careful patient selection is important for models of domiciliary care.
VV-ECMO AS A BRIDGE TO TRANSPLANT IN CF PATIENTS: HOW THE IMPOSSIBLE CAN BE MADE POSSIBLE - A CASE SERIES OF WA EXPERIENCE

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Cystic Fibrosis (CF) has accounted for almost 25% of WA lung transplants since 2004. Patients are often challenging due to complex microbiology and CF-related co-morbidities. Psychosocial dynamics may differ from other transplant cohorts and early referral facilitates care transition, clinical assessment, patient education and psychology input. The two cases described are patients who for different psychosocial reasons presented in extremis for lung transplantation, requiring bridging with ECMO.

Case 1
19 year old female with co-morbid diabetes (DM), pancreatic insufficiency (PI) and low BMI presented to the Adult CF Unit with a pulmonary exacerbation (FEV1 20.4% predicted) and type 2 respiratory failure (T2RF) in Oct 2011. She had not attended for some years based on her family’s strong religious beliefs. Referral to the transplant unit was made during that admission, followed by rapid assessment and listing. Shortly afterwards she represented and required NIPPV followed by intubation and VV-ECMO for 47 days before bilateral lung transplantation. Pre-transplant ICU stay was complicated by sepsis, acute kidney injury and two cardiorespiratory arrests during ECMO circuit changes.

Case 2
25 year old female with PI and DM was initially referred to the transplant team in October 2015 following a severe pulmonary exacerbation (discharged on nocturnal O2). The patient and her parent were not ready to contemplate transplant and disengaged from transplant appointments. Almost 3 years later another exacerbation with T2RF prompted re-referral to the transplant team. She was assessed and listed whilst an inpatient in the CF unit. Deterioration prompted emergency transfer to the transplant unit ICU, intubation and VV-ECMO for 16 days before bilateral lung transplantation.

Psychosocial barriers may delay referral and engagement in the transplant process, leading to late listing and high risk transplant surgery. Through strong collaboration between the CF, transplant and ICU teams both patients were successfully bridged with ECMO. Both required prolonged rehabilitation before discharge but are currently alive with good allograft function.
MEASURING HEALTH-RELATED QUALITY OF LIFE IN PATIENTS WITH CYSTIC FIBROSIS

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Background: Demographics, treatments and clinical outcomes of cystic fibrosis (CF) patients have changed dramatically in the last few decades. However, there is a growing need to address psychosocial factors in the management of CF. Measuring health-related quality of life (HRQOL) using patient-reported outcome measures (PROMs) has enabled researchers and clinicians to evaluate the overall health and well-being of CF patients. Incorporating a PROM into the Australian Cystic Fibrosis Data Registry (ACFDR), which holds data from over 90% of Australia’s CF population, would allow the measurement of long term HRQOL outcomes of treatments and complications and facilitate HRQOL life research in CF. The aim of this study was to gain an understanding of the use of PROMs in CF and to evaluate whether existing PROMs used in CF are suitable for incorporation in the ACFDR.

Methods: A systematic literature search was conducted to identify suitable PROMs in CF adult and paediatric patients. The search focused on English language, full peer-reviewed publications from 2008 to 2018, used the MESH and non-MESH keywords and/or their variations to identify relevant articles.

Results: Twenty-seven different PROMs were identified. Most common PROMs were designed specifically for CF. Equal numbers of studies were conducted on adult (32%, n=31), paediatric (35%, n=34) and both (27%, n=26) populations. The majority of studies (75%, n=73) were of observational design. The two most widely used PROMs, the Cystic Fibrosis Questionnaire-Revised (CFQ-R) and the Cystic Fibrosis Quality of Life Questionnaire (CFQoL) demonstrated superior psychometric properties and acceptability in English-speaking populations. No PROMs were used within a clinical registry setting previously.

Conclusion: Findings of this review provide a rationale for the next stage of the study, during which we will undertake qualitative work with patients and clinicians to determine clinical utility and acceptability of the most frequently reported CF-specific instruments CFQ-R and CFQOL.
REVIEW OF A PAEDIATRIC CF CENTRE TO ASSESS CHANGES IN OUTCOME OVER 18 YEARS

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Background: Management of children with Cystic Fibrosis (CF) evolves with new medications and strategies. Aim: To determine whether these changes have impacted patient outcomes over an 18 year period.

Methods: Comparison of demographics and outcomes of children with CF under full care in one centre (Starship Children’s Hospital New Zealand) in 2000 and 2018.

Results: In 2000 there were 50 (31 males) mean age 8.5 years, 53% homozygous dF508, with mean FEV1 81% predicted and in 2018 54 (30 males), mean age 9.5 years, 38% homozygous dF508 with mean FEV1 85% predicted. Late diagnoses had increased from 6% to 15% of the clinic. The use of hypertonic saline had gone from 0% to 67%, RhDNase from 6% to 33%, and TOBI from 0% to 9%. Even in 2018 there are no children on ivacaftor. Chronic Staphylococcus aureus infection was reduced from 73% to 54%, Pseudomonas aeruginosa was reduced from 50% to 11% while Burkholderia cepacia remained at 2%. MRSA had increased from 3% to 6%, as had Stenotrophomonas maltophilia from 4% to 19%. BMI <25% had decreased from 29% of children to 18% associated with an increase from 12% to 22% on nutritional supplementation with only 4-6% of patients with gastrostomy.

Conclusions: Across an 18 year period there are now lower numbers of children homozygous dF508, more late diagnoses, less with chronic Staphylococcus and Pseudomonas infection but more with Stenotrophomonas infection associated with an increase nebulised airway clearance and antibiotic therapies but no significant increase in FEV1.

Conflicts of interest: No
LONGITUDINAL STUDY ON THE REAL WORLD CLINICAL OUTCOMES IN AUSTRALIAN PATIENTS ON IVACAFTOR

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Background: Ivacaftor was listed under the Pharmaceutical Benefits Scheme on 1st December 2014 for Australian patients with cystic fibrosis (CF) harbouring G551D/ gating mutation. The real-life clinical outcomes in Australian patients prescribed long-term Ivacaftor is unclear.

Aim: This post-approval, observational study aims to evaluate the ‘real-life ‘impact of Ivacaftor in Australian patients in the 2 year post-approval period using the Australian Cystic fibrosis Data registry (ACFDR).

Method: Comparison of lung function and nutritional status in patients before and after ivacaftor treatment using the ACFDR data was performed.

Results: In total, 251 patients with G551D/gating mutation was identified in the ACFDR. Of 251 patients, 164 had treatment information available -130/164 patients were treated with Ivacaftor, 34/164 were identified as not receiving Ivacaftor. Of the 130 Ivacaftor-treated patients, 103 had clinical data available for comparison. Compared to the pre-treatment baseline, significant within-person improvement in FEV1 (L) (Mean difference: 199ml; [95CI: 140-259ml]; p<0.001)); FEV1 % (Mean difference: 2.4%; [95%CI: 1.0-3.8%;p:0.001]) and weight (Mean difference: 3.1 kg; [95%CI:2.2-3.9kg; p<0.001]) were noted after Ivacaftor commencement. At 1-year follow-up, sustained pulmonary and nutritional improvement was noted (compared to baseline): Mean FEV1 (L) difference: 282ml (95%CI: 205-359ml; p<0.001); mean FEV1 % difference: 2.5% (95%CI: 0.7-4.3%; p<0.001); mean weight difference: 3.5kg (95%CI: 2.2-4.7; p<0.001). At 2-year follow-up, sustained pulmonary and nutritional improvement was noted compared to baseline: Mean FEV1 difference: 262ml (95%CI: 124-400ml; p<0.001); Mean weight difference: 6.6kg (95%CI: 4.5-8.7kg; p<0.001). Longitudinal trends was analysed with mixed-effect modelling. No significant difference in the rate of lung function change was noted before and after Ivacaftor.

Conclusion: Whilst general limitations of registry data analysis applies, this study demonstrated real-life clinical benefits in Australian patients treated with Ivacaftor up to 2 year follow-up. The impact of Ivacaftor on disease trajectory and survival requires a larger comparator group and further study is in progress.
SURVIVAL OF PATIENTS WITH CYSTIC FIBROSIS: A LONGITUDINAL STUDY USING AUSTRALIAN PATIENT REGISTRY DATA

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Background. Recently published international studies indicate increasing median survival in cystic fibrosis (CF) patients, which possibly reflects a number of factors, including improvement and refinements in CF management and centre-based multidisciplinary care (Stephenson A, et al, Ann Intern Med. 2017;166:537-546). The aim of this study was to test and provide a standardised approach for estimating survival among patients participating in the Australian Cystic Fibrosis Data Registry (ACFDR).

Methods. This population-based cohort study used prospectively collected data from 23 Australian CF centres participating in the ACFDR from 1998 to 2016. To ensure accuracy of the survival data, the ACFDR was linked with the records from the National Death Index (NDI). Period survival analysis was used to calculate median age of survival estimates over time. Median age of survival was calculated for each 5-year window beginning with the period 1999-2003 up until 2009-2013. The overall median survival of the cohort was also calculated using the Kaplan-Meier method. Survivor curves were estimated from a mathematical formula that uses the proportion of individuals alive at each age who die before their next birthday.

Results. Between 1998 and 2016 the ACFDR followed 4,507 patients. More than half (52.6%) of the patients were males, 3.1% had undergone lung transplantation, majority of the patients (86.9%) were diagnosed under the age of 2 years with the mean (SD) age at diagnosis of 1.9 (6.7) years. Survival results will be presented at the conference following validation of death data with NDI linkage.

Conclusions. Survival statistics, estimated using data from national CF patient registries are widely used to inform the CF community and monitor disease progression and survival of CF populations. Using a standardized approach to survival calculations will provide greater confidence in international comparisons and in the identification of risk factors associated with survival.
DEVELOPMENT OF A CAUSAL MODEL OF PULMONARY EXACERBATIONS OF CYSTIC FIBROSIS

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Background: Optimising the management of pulmonary exacerbations of cystic fibrosis (CF) may help preserve lung function and prolong life. Treatment of exacerbations is complex however, with wide variation in practice and little consensus on what is optimal. A clearer understanding of the underlying causal mechanisms may help to inform research prioritisation and design, and clinical decision-making.

Methods: Australian and New Zealand CF experts, young and adult CF patients, their care-givers, and modelling experts used a modified Delphi approach to co-develop a model which articulates how various factors are thought to interplay in influencing the short-term outcomes of exacerbations. The model provides an explicit causal framework for integrating expert domain knowledge, knowledge from lived experience with CF, and evidence from the literature, with new data.

Results: The provisional BEAT CF causal model for pulmonary exacerbations comprises four sub-models covering background factors, treatments, acute disease processes, and outcomes. Background factors like age, mutation type, and nutritional status were considered to be driving the pulmonary disease stage. The disease stage, together with the presence of specific airway pathogens and use of various treatments, were considered to impact upon the outcome of exacerbations which, in turn, can be measured as measures of lung function and the presence of various symptoms.

Discussion and future work: We used a novel method to elicit causal understandings of the complex interplay of factors in pulmonary exacerbations. Future work will attempt to refine and prioritise the relative importance of disparate outcome measures from a patient perspective. The causal model will be used for research prioritisation, design and analysis of BEAT CF which aims to optimise management of pulmonary exacerbations. Backed by data from BEAT CF, we anticipate the final model will eventually be deployed as a consumer and clinician-friendly decision-support tool to inform complex and personalised decision-making in CF.
THE ALFRED WELLNESS SCORE IN ADULTS WITH CYSTIC FIBROSIS: STABILITY, VALIDITY AND RESPONSE TO PULMONARY EXACERBATIONS

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We recently developed the Alfred Wellness Score (Awescore), a brief patient-reported outcome measure of 5 domains with two items each.

Objectives: (1) To assess Awescore stability and validity compared to the CFQ (Cystic Fibrosis Questionnaire); (2) to assess the effects of pulmonary exacerbation on Awescore compared to baseline values.

Methods: In two separate studies participants completed (1) Awescore and CFQ (time 1, time 2) one month apart while clinically stable; and (2) the Awescore during clinical stability and at diagnosis of a pulmonary exacerbation using recognised criteria.

Results:
(1) No significant difference was seen between the Awescore measures for 20 participants (time 1: mean 65 [SD 14], time 2: 65 [13]; mean difference -0.2 [95%CI -3.5 to 3.1]); these scores were significantly and strongly correlated (Pearson's r=0.854, p=<0.005). The CFQ scores (time 1: mean 813 [SD 125], time 2: 789 [131]) were moderately correlated with the Awescore (Pearson's r=0.632; p=0.003).

(2) Sixty patients completed the Awescore during clinical stability and exacerbation (age mean 33 [SD 10] years, BMI 22 [2] kg/m2, FEV1 median 50 [IQR 40,65] %predicted). A significant reduction in Awescore from clinical stability (mean 76 [SD 10], range 48-95) to exacerbation (47 [13], 17-69) was demonstrated (mean difference -29 [95%CI -32 to -25]) and observed in all domains (p<0.0005). The median (IQR) for baseline vs exacerbation for the domains were: Cough /Sputum 7(6,8) / 7(6,8) vs 4(3,5) / 4(3,5); Energy /Exercise 8(6,9) / 7(6,9) vs. 4(3,5) / 4(2,5); Appetite /Weight 8(8,9) / 8(7,9) vs. 5(3,7) / 6(4,8); Anxiety /Mood 9 (7,10) / 8(8,9) vs. 7(5,8) / 5(4,7); Sleep /General Health 8(6,8) / 7(7,8) vs. 4(3,6) / 5(3,6).

Conclusion: The Awescore is a stable and valid measure when compared with the CFQ. Pulmonary exacerbations impact negatively and significantly on all health domains of wellness in adults with CF.
EMERGING REGISTRY USES REQUIRES ADAPTABLE SYSTEMS:
REINVENTING THE AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY

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Objectives: The Australian Cystic Fibrosis Data Registry (ACFDR) has collected annual and encounter data on Australian patients with cystic fibrosis (CF) since 1998. Over this time it has tracked the demographic characteristics and outcomes of approximately 3,500 patients. However, emerging registry requirements include capturing emerging complications and treatments for CF patients; collecting and comparing outcomes internationally; and integration of registry data with health service and clinical trial platforms. Over 12 months, the ACFDR has been reinvented to accommodate all these needs.

Methods: The ACFDR’s dataset was completely reviewed to identify, assess and prioritise the data elements needed to support best practice care for CF patients. Methods used to undertake the review included comparison with registry datasets from the UK, US, ECFS and Canada, structured Steering Committee consultation, and testing and piloting of the new dataset. Criteria for data item inclusion included data entry burden and feasibility for collection. REDCap was used to develop the new database.

Results: Key changes to the ACFDR dataset included a patient summary dashboard; updated diagnostic information (aligned with international harmonisation work); addition of a patient consent module and a CFTR modulator module; additional microbiology information; and quarterly data completion checkboxes to support regular data entry. The database also has improved integration of data from electronic medical records and other databases including clinical trials.

Conclusion: A holistic approach to data element review of a data registry, incorporating international harmonization, is required to ensure that clinical registries capture the most relevant data for contemporary CF diagnostic and treatment practices. Data element reviews require significant consultation with clinical experts and database users to ensure a balance between the collection of clinically meaningful and feasible information.
HEALTH RELATED QUALITY OF LIFE IN CHILDREN WITH CYSTIC FIBROSIS AT 5 YEARS AND ASSOCIATIONS WITH HEALTH OUTCOMES

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Background: The impact of early CF disease and its management on health-related quality of life (HRQOL) in preschool children is poorly characterised, and data on relationships between HRQOL and clinical health outcomes in young children with CF are limited. We aimed to characterise and compare parent and child-reported HRQOL and evaluate relationships with clinical outcomes at age 5 years.

Methods: Subjects were participating in the multicenter Australasian Cystic Fibrosis Bronchoalveolar Lavage (ACFBAL) trial investigating BAL-directed vs. standard CF therapy. Children aged 5 years and their parents rated HRQOL using the Pediatric Quality of Life Inventory (PedsQLTM) and Cystic Fibrosis Questionnaire-Revised (CFQ-R) questionnaires.

Results: PedsQL and CFQ-R questionnaires were completed by 141 primary caregivers and 135 and 130 children, respectively. There was no evidence of a difference in HRQOL between children randomized to BAL-directed vs. standard CF therapy. Children rated poorer generic HRQOL than healthy children. Correlation between child and parent-reported HRQOL was poor. For domains where evidence of a difference between child and parent-reported HRQOL was found, parents tended to rate HRQOL higher on average. Nutritional status, FEV1, pulmonary exacerbations (PEs) and hospitalization rates predicted HRQOL at age 5 years. Higher FEV1 z scores predicted higher parent-reported HRQOL in CFQ-R respiratory (p=0.004), physical (<0.001), emotional (p=0.007) subscales and PedsQL total (p=0.011) and physical domains (p=0.009). Higher PE rates were associated with lower (worse) parent-reported CFQ-R respiratory (p=0.003), PedsQL total (p=0.021) and physical (p=0.019) scores, and with lower child-reported CFQ-R respiratory score (p=0.055).

Conclusion: HRQOL in young children with CF is lower than healthy children. The impact of CF on HRQOL is evident by age 5.
PROTOCOL FOR A DISCRETE CHOICE EXPERIMENT TO EVALUATE PREFERENCES OF PATIENTS WITH CYSTIC FIBROSIS AMONG ALTERNATIVE TREATMENT-RELATED HEALTH OUTCOMES

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Introduction: Clinical decision making is a complex process. Patient preference information regarding desirable health states should inform treatment and is critical to agreeing on goals of therapy. Cystic fibrosis (CF) is a common, inheritable multi-system disorder for which the major manifestation is progressive, chronic lung disease. Intermittent pulmonary exacerbations are a hallmark of disease and these drive lung damage that results in premature death. We suspect that clinicians make assumptions, most likely implicit assumptions, about outcomes that are desired by patients who are treated for pulmonary exacerbations. The aim of this study is to identify and quantify the preferences of patients with cystic fibrosis regarding treatment outcomes.

Methods and analysis: We will develop a discrete choice experiment (DCE) in collaboration with people with CF and their carers and evaluate how patients make trade-offs between different aspects of health-related status when considering treatment options.

Ethics and dissemination: Ethics approval for all aspects of this study was granted by the Western Australia Child and Adolescent Health Service Human Research Ethics Committee [RGS903]. Weighted preference information from the DCE will be used to develop a multi-attribute utility instrument as a measure of treatment success in the upcoming Bayesian Evidence-Adaptive Trial to optimise management of CF (BEAT-CF). Dissemination of results will also occur through peer-reviewed publications and presentations to relevant stakeholders and research networks.
COMMENCEMENT OF PBS ORKAMBI (LUMICAFTOR/IVACAFTOR) AT A LARGE ADULT CYSTIC FIBROSIS (CF) CENTRE: A REAL WORLD EXPERIENCE

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Background: Orkambi was listed on the PBS (Pharmaceutical Benefits Scheme) on 1st October 2018 for eligible patients homozygous for F508del. We describe commencement of PBS Orkambi at a large adult CF centre.

Method: The Prince Charles Hospital CF database/pharmacy records of 110 patients homozygous for F508del (42F and 68M) were analysed.

Results: 25 patients (23%) were transitioned from Special Access Scheme (SAS) Orkambi to PBS Orkambi. 9 patients were ineligible due to current pharmaceutical trial participation. 27 patients (24%) declined or were unable to consider Orkambi therapy for a range of reasons (see table). 11 patients have not attended clinic since 1st October 2018. 2 patients remain undecided and 4 patients have delayed commencement of Orkambi (1 is undergoing fertility treatment and 3 with severe lung disease require admission for commencement). 28 patients (25%) commenced Orkambi for the first time after PBS listing and have continued therapy. The mean time to commencement after PBS listing was 6 weeks (0-24 weeks). 4 patients new to Orkambi were intolerant (2 rash, 1 behaviour change, 1 chest tightness) and ceased. Counselling and prescribing utilised significant amounts of staff time. (Table 1 to be inserted here detailing reasons for declining Orkambi).

Conclusions: A significant number of F508del/F508del patients were unable to utilise PBS Orkambi therapy (52%). This contrasts with our experience with Ivacaftor therapy (100% uptake by eligible group). Those who have commenced Orkambi under the PBS scheme have mostly tolerated the therapy well. The transition of a large group of patients to PBS Orkambi used significant staff resources.
Background: Lumacaftor/ivacaftor (LUM/IVA) combination therapy received PBS licence in October 2018. Outcomes on the patient experience of treatment initiation outside of clinical trial are limited to date.
Aim: To report on our centre’s experience of LUM/IVA initiation in patients with mild-moderate CF lung disease.

Results: We report immediate post-dose respiratory-related outcome in 29 patients with mild-moderate cystic fibrosis (CF) lung disease (median [range] ppFEV1: 86.2% [51%-120.6%]) prescribed LUM/IVA. Whilst 96% patients experienced a decline in ppFEV1 from baseline at 2-hours (mean relative change [95%CI]: -7%; -9.4% to -4.8%, p< 0.001), only 18% reported non-severe respiratory-related symptoms within 4-hours of first dose. 50% and 80% recovered to pre-LUM/IVA ppFEV1 at 1-month and 3-month follow up respectively. Compared to pre-treatment, the mean relative change in ppFEV1 at 3 months was +3.73% (95%CI: -11% to +18%, p 0.061). Within the first 3 months, 11% patients reported chest tightness, 18% were treated for a pulmonary exacerbation.29% patients had non-respiratory adverse reactions with 14% reporting mood changes, 0.03% oesophageal reflux and 0.03% dysmenorrhea. The latter 2 patients were prescribed a proton pump inhibitor (PPI) and hormonal oral contraception (OCP) respectively, both known to interact with LUM/IVA. 2/28 (7%) ceased LUM/IVA, 1 secondary to low mood and 1 dysmenorrhea. One patient moved state after 1 month. 18% had known CF liver disease. There was no observed deterioration in liver function.

Conclusion: Our results highlight that a drop in ppFEV1 from baseline is common on initiation of LUM/IVA, but the majority are asymptomatic. Most patients had improvement in ppFVE1 at 3 months. LUM/IVA respiratory-related adverse events are common but rate of cessation of treatment is low. We observed a link between adverse effects and known LUM/IVA drug interactions (PPI and OCP). Longer term follow up will allow further evaluation of adverse effects and safety.
The life expectancy of individuals with cystic fibrosis (CF) has increased significantly. Therefore more adults are likely to consider having children. Over the past decades the number of pregnancies has increased. Managing pregnancy in a woman with CF is complex and requires a multi-disciplinary approach to ensure the best outcomes for the baby and mother.

The use of CFTR modulators has resulted in improving and maintaining lung function and overall quality of life. However very little is known about the effects of CFTR modulators in pregnancy. There are a few case reports in the literature on the use of these medications in pregnancy, but more information is needed about the effects of the CFTR modulators and the number of women who continue to take them while pregnant.

In this case series we discuss three women who had successful pregnancies while taking CFTR modulators. All three women were counselled on the lack of information on the use of CFTR modulators in pregnancy and breastfeeding. Each woman made an informed decision to continue to take these medications. The pregnancies were managed as high risk requiring shared care input from obstetrics and respiratory specialists. The clinical outcomes for both babies and mothers will be discussed including lung function results and number of exacerbations.

Patient N pre pregnancy FEV1 2.04L and 3 months post pregnancy FEV1 1.86L (nil exacerbations). Patient D (baby 1 pre FEV1 1.93L and 3 months post FEV1 1.87L (1 exacerbation)) (baby 2 pre FEV1 1.96L and 2 months post FEV1 1.73L (nil exacerbations)). Patient A pre FEV1 1.18L and 4 months post FEV1 1.24L (3 exacerbations). In this era of CFTR modulators, more women with CF are likely to have children. Therefore there is a need for communication of outcomes on the use of CFTR modulators in pregnancy and breastfeeding.
TO BE OR NOT TO BE ON CFTR THERAPIES DURING PREGNANCY: RISKS TO BE CONSIDERED

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Safety data for CF transmembrane conductance regulator (CFTR) modulator therapy in pregnancy remains limited. We report a 29-year-old female with severe CF-related lung disease and diabetes who continued ivacaftor therapy through two naturally-conceived pregnancies. Both babies were born without evidence of congenital malformations, and maternal health remained unremarkable post-partum.

The patient was stabilised on ivacaftor one year prior to her first pregnancy, with significant improvements in ppFEV1 (30 to 46) and reduction in pulmonary exacerbations. Ivacaftor was ceased at week five of the patient’s first pregnancy due to limited safety data, coinciding with a rapid decline in ppFEV1 to 37 requiring hospitalisation for intravenous antibiotics. The patient made an informed decision to restart ivacaftor at week 10 of her pregnancy. Her lung function returned to baseline and diabetes control improved; both were maintained for the remainder of her pregnancy (ppFEV1 46, HbA1c 5.7% prior to delivery).

During her second pregnancy, the patient continued ivacaftor, opting for a reduced dose during the first trimester. Lung function and HbA1c remained stable throughout the pregnancy, without pulmonary exacerbations (ppFEV1 42, HbA1c 6.3% prior to delivery).

This case highlights the beneficial effects of CFTR therapy on maternal health and pregnancy outcome, and highlights the decision-making process required to continue these treatments during pregnancy. Discontinuation of ivacaftor therapy in this case was associated with a rapid deterioration in FEV1 and diabetes control. Maintaining the health of the mother during pregnancy, alongside improving fetal outcomes should be the primary focus of care. The lack of safety data surrounding CFTR therapy in pregnancy requires patient involvement in making informed decisions, particularly where long-term risks and benefits to mother and baby may not be established. Continued reporting of successes and failures of CFTR modulators in pregnancy can provide ongoing surveillance and help shape decisions for other similar cases.
INITIATION OF IVACAFTOR/LUMACAFTOR THERAPY IN PAEDIATRICS – EXPERIENCE BEYOND THE CLINICAL TRIAL

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Participants in clinical trials of new therapies, are required to fulfil exhaustive inclusion and exclusion criteria. For CFTR modulator therapies; this usually means lung function that is neither too ‘high’ nor ‘low’, no significant co-morbidity, and, no concomitant use of certain medications. Once registered and widely available, all patients who meet the prescribing criteria, are able to commence therapy, irrespective of meeting criteria that would have precluded them from a clinical trial of the same therapy. Thus, ‘clinical’ initiation of therapy has the potential to evoke a number of adverse effects and have an impact (beyond administrative), on already stretched clinical services.

Prior to commencing ivacaftor/lumacaftor therapy in our clinic, patients were required to agree to set pre- and post-initiation measures (including a 2 hour post-dose observation) to ensure their safety, and allow for documentation of adverse effects. Between October 2018 and April 2019, 28 patients who were naïve to ivacaftor/lumacaftor, commenced therapy using the age-appropriate dose. Patients ranged in age from 6 to 17 years and had a baseline FEV1 between 50 and 110 %predicted.

At least one documented adverse effect on Day 1, Week 2 or Month 3 review was experienced or reported by 79% of patients. Decrease in FEV1 of >10% of baseline occurred in 43%, increase in liver function enzymes to >2x baseline value in 7%, and 57% reported ‘medication-related’ symptoms (rash, shortness of breath, chest tightness). None discontinued therapy, but some required clinical review, phone consultation, additional pharmacotherapy (36%) or dose-reduction with eventual titration up to the listed dose (32%).

In conclusion, adverse effects associated with initiation of ivacaftor/lumacaftor therapy in children and young people in the clinical setting are common. There is likely an impost on clinical workload which needs to be anticipated in the period immediately following initiation of CFTR modulator therapy.
IVACAFTOR USE IN CYSTIC FIBROSIS PATIENTS WITH RESIDUAL FUNCTION MUTATIONS AND POOR LUNG FUNCTION

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Introduction: In Australia, Ivacaftor was made available via a compassionate access program for Cystic Fibrosis (CF) patients with specific residual function mutations. Patients were eligible to receive Ivacaftor if they had a stable forced expiratory volume (FEV1) of <40%, were being assessed for lung transplantation, or demonstrated a rapid decline in lung function over 6 months. This case series describes the cohort of patients who qualified for this program.

Methods: All 250 pre-transplant CF patients from our centre were assessed for eligibility throughout the duration of the program (Jan-Dec 2018). Pre and post treatment spirometry and body mass index (BMI) were obtained. Subjective symptoms, transplant status and CFTR modulator compliance were recorded.

Results: Four patients with the following genotypes were enrolled: 2789+5G>A/N1313K, F508del/Pro67Leu and F508del/R117h (n=2). In April 2019, the duration of Ivacaftor use for these participants ranged from 7-11 months. All patients reported increased energy levels, reduced sputum production and no intolerance to Ivacaftor. The pre-treatment median FEV1 was 1.03L, 36% (range 0.99–1.17L, 21-39%), forced vital capacity (FVC) was 2.27L, 57% (range 1.65-2.55L, 48-76%) and BMI was 22 kg/m2 (range 20-24 kg/m2). Post-treatment median FEV1 was 1.11L, 38% (range 1.12–1.47L, 22-56%) after 1 month of Ivacaftor use and 1.2L, 42% (range 0.96 – 1.36L, 21-50%) at their most recent visit, while the most recent BMI increased to 23.6 kg/m2 (range 22-25 kg/m2). Three patients were being worked up (n=2) or awaiting listing (n=1) for lung transplantation. Two patients did not continue with their lung transplant work up and one patient had his listing for transplant delayed by the transplant service. Compliance was monitored through patient reporting and time calculation between script requests.

Discussion: In CF patients with residual function mutations and poor lung function, use of Ivacaftor improved symptoms, stabilized or improved lung function and weight, and was tolerated well.
SELF-MONITORING BLOOD GLUCOSE (SMBG) LEVELS IN ADULTS WITH CYSTIC FIBROSIS RELATED DIABETES (CFRD) - A COMPARISON BETWEEN FINGER LANCET CAPILLARY (FLC) TESTING AND FLASH GLUCOSE MONITORING (FLASH GM): A PILOT STUDY

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Background: CFRD is the most common co-morbidity associated with CF. Local CFRD prevalence is 121 adults (35%). Finger lancet capillary (FLC) is standard monitoring for daily blood glucose levels and guides insulin treatment. Clinical guidelines suggest self-monitoring blood glucose (SMBG) four to eight times a day. Approximately 70% of diabetics do not achieve monitoring recommendations and difficulties are frequently reported at clinical reviews. Flash GM offers an alternative arm sensor that continuously measures interstitial fluid glucose for 14 days, negating the need for FLC testing.

Method: A cross-sectional observational study piloted five adults with CFRD. Consenting participants completed a questionnaire on SMBG, trialled flash GM system for 14 days and provided feedback via questionnaire.

Subjects: Five subjects were selected at CFRD clinic. Inclusion criteria included: aged 18 years old and above with CFRD requiring insulin, and able to provide informed consent. Exclusion criteria included pregnancy, indeterminate glucose tolerance/ impaired glucose tolerance, or informed consent not provided.

Results: Five adults with CFRD consented to participate. All patients reported preferred use of flash GM to SMBG. Subjective improvements of participants included improved diabetic control, hypoglycaemic avoidance, improved management of diabetes with physical activity and positive improvements to lifestyle.

Conclusion: The pilot study provided patients & CF staff experience in using flash GM. All patients subjectively preferred flash GM. No concerns were reported with self-insertion or adherence of device to skin. No concerns regarding treatment of hypoglycaemic events where reported. Currently the device and sensors are only available for private purchase (NDSS subsidy is pending).

Results from this pilot study indicate that Flash GM is safe for use in adults with CFRD, this device may assist in CFRD self-management by improving barriers to glucose monitoring, further studies with a larger cohort are recommended.
DISCREPANCIES IN NATIONAL AND INTERNATIONAL CYSTIC FIBROSIS STANDARDS OF CARE: TIME FOR CONSENSUS?
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Background: Increased survival over the past 50 years of people born with the genetic disorder cystic fibrosis (CF) has seen a rapid increase in the CF population in countries with well-developed healthcare systems. The increase has been most marked in adults with more adults than children in countries with well-developed healthcare systems. However, this increase in numbers has not been matched by additional resources in terms of health professional personnel or physical facilities to accommodate these patients. In this study, we examined the recommendations on staffing levels for CF centres around the world to investigate how consistent they were.

Method: We searched publication databases such as PUBMED, EMBASE, CF association websites and data registries to identify consensus statements on staffing levels in adult CF centres. We reviewed the most recent standard of care guidelines for the recommended number of staff per each allied health professional of the CF Multidisciplinary Team. Comparison of staffing levels for each CF team discipline will be presented in table format for each country that had published recommendations.

Results: There were substantial differences with respect to medical, nursing, physiotherapy, dietetic, social work, psychology and pharmacy staffing recommendations depending on country and number of staff per patient. Variations in recommendations between paediatric and adult care exist, and some countries have separate multidisciplinary teams for outpatient and inpatient care. Microbiologists and medical trainees were not always included within the multidisciplinary team.

Conclusion: Consideration of inclusion of other allied health professionals within the CF multidisciplinary team and consensus of level of staffing recommendations would be beneficial. Without this, people with CF could potentially be adversely affected if the allocated resources fail to keep up with the number of patients and complexity of disease.
BENEFITS OF A CYSTIC FIBROSIS CONSUMER REFERENCE GROUP IN THE PAEDIATRIC CLINICAL AND RESEARCH SETTINGS

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Background: Consumer involvement in paediatric clinical and research services is important. Western Australia has a combined clinical and research cystic fibrosis (CF) Consumer Reference Group (CRG) that provides valuable input into services provided by Perth Children’s Hospital and Telethon Kids Institute.

Methods: The Child and Adolescent Health Service (CAHS) cystic fibrosis CRG provides a consumer perspective to both clinical and research aspects of cystic fibrosis and advises about priorities, practice and policies. It aims to enhance the quality and relevance of activities through consumer participation. At each meeting, issues relevant to children’s experience in the CF clinic are discussed, and researchers present their proposals for input and advice. In the ARESTCF program, consumer input is a requirement for all research projects conducted on data and / or samples collected from study participants. The committee is also asked for input into documentation such as research consent forms or questionnaires and they may provide letters of support to Ethics Committees.

Results: Input from CRG is being captured and will be analysed in terms of the impact on the program. The CRG has advocated for resources for role plays for children in anxious situations; had discussions around mental health screening for unresolved grief; and are currently developing guidelines for both consumers and scientists to consider during research project presentations. The CRG has allowed input to be provided for each research proposal prior to the proposal being presented to the Scientific Management Committee for approval. This has not only saved time in the approval process, but has allowed for valuable input into research proposals and has allowed the research to be tailored to meet consumer needs as well as scientific integrity.

Conclusion: The CRG has provided valuable input into the clinical and research programs across Perth Children’s Hospital and Telethon Kids Institute.
IMPROVING INPATIENT NUTRITION OUTCOMES USING THE PRINCIPLES OF TOYOTA PRODUCTION SYSTEM (TPS)

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**Background:** Hospital admissions for children with Cystic Fibrosis (CF) is an opportune time to recover loss in lung function and weight, the later relies on an acceptable food service model. CF patient feedback has been negative with a plate waste of 65% and an average weight gain of 0.36% per day of admission. In 2018 the Respiratory team partnered with Toyota to apply the Toyota Production System (TPS) in order to improve nutritional outcomes for CF inpatients.

**Objectives:** Primary aim was to increase patient weight gain during a hospital admission, secondary aim was to increase patient and carer meal satisfaction scores.

**Method:** A project leadership team was established, members attended TPS Dojo training, followed by three site visits by Toyota to SCH each month for three days with weekly teleconferences in between. Patient and carer satisfaction was self-reported using a happy face (overall satisfaction with the meal and consumption of 50% or more) or a sad face. Patient/carers were asked to nominate the reason for a sad face from set domains.

**Results:** There was 60% sad faces (from 99 traditional meals served) with the two most common reasons being ‘there were no suitable options’ and ‘the meal did not taste good’. Further root cause analysis determined that a more in depth review of the statewide menu was necessary, but was beyond the scope of this project. A more immediate solution was developed to provide CF patients with a selection of alternate “short order“ meals using a room service model. Sad faces reduced to 0% (out of 198 new short order meals served) and average weight gain per day increased to 0.42%.

**Conclusion:** The concepts and principles of TPS as applied to improving the nutritional status of children with CF admitted to SCH proved to be successful.
IMPROVING DISPENSING SERVICES TO PATIENTS WITH CYSTIC FIBROSIS AT THE WOMEN'S AND CHILDREN'S HEALTH NETWORK

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**Background:** In late 2015, the Women’s and Children’s Health Network (WCHN) commenced an action research project aiming to improve dispensing services for patients with cystic fibrosis (CF). Prior to the project, families frequently reported waiting longer than 90 minutes for their prescriptions to be dispensed. This was highlighted as the most significant issue for WCHN CF consumers via a CF Australia Peer Review survey in 2014. The project adopted a multidisciplinary and consumer engagement strategy to:
- identify barriers and opportunities for improvement within the dispensing service and
- develop recommendations to improve efficiencies for Pharmacy and the pharmacy experience for families of patients with CF.

**Methodology:**
- Analytical review of dispensing data
- Observational review of workflows in Pharmacy
- Targeted interview-based, hard-copy and electronic consumer surveys

**Results:**
- 59% of prescriptions took longer than 60 minutes to be dispensed (12% took longer than 120 minutes)
- Supply quantities were usually for one month's treatment despite the majority being long-term medicines.
- There was sub-optimal work prioritisation within Pharmacy.
- An influx of prescriptions was received mid-morning, coinciding with staff rostered breaks.

**Changes implemented:** By January 2017, the following recommendations were implemented:
- Increasing quantities supplied per dispensing occasion.
- Streamlining workflows to improve prioritisation of 'waiting' prescriptions.
- Providing patients with options to hold their prescriptions in Pharmacy and email orders as required.
- Improving visibility of prescription status by upgrading the electronic prescription tracking system.
- Rescheduling staff breaks.

**Evaluation:** A formal post-project evaluation conducted in July 2018 found that:
- 90% of prescriptions were dispensed within 60 minutes (1% took longer than 120 minutes), and this was sustained over a 12-month period.
- 85% of consumer families surveyed (n=46) noticed an improvement in dispensing turnaround times.
- 70% stated they used the email order system
- 89% considered the Pharmacy service to be either good or very good.
EFFECT OF TRANSFER FROM A PAEDIATRIC TO ADULT CYSTIC FIBROSIS CENTRE ON CLINICAL STATUS AND HOSPITAL ATTENDANCE

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Objective: Transfer from paediatric to adult services could lead to clinical deterioration; few studies have examined this. We sought to examine the clinical impact of a structured individualised transition process in patients with cystic fibrosis (CF).

Design: Medical records of all patients with CF in Western Australia who transferred from a paediatric to adult CF centre between 2008 -2012 were reviewed. Data were extracted for two years before and after transfer. The number of CF outpatient visits, inpatient days and home intravenous antibiotic therapy (HIVT) days were recorded at yearly intervals before and after transfer. Sputum culture results at transfer were compared to culture results at one and two years after transfer. All respiratory function and anthropometric data over the 4 years were extracted.

Setting: Perth Children’s Hospital, Tertiary Paediatric Centre, Perth, Western Australia and Sir Charles Gairdner Hospital, Adult Cystic Fibrosis Service, Perth, Western Australia.

Results: Forty-two patients with CF were transferred between 2008-2012. Mean age at transfer was 18.9 years (range 17-22). Compared to one-year pre-transfer, the frequency of outpatient visits at one and two years post-transfer increased. After transfer, there was no change in BMI, HIVT days or inpatient days, and no acceleration in the expected decline in FEV1.

Conclusion: This study found that transition from a paediatric to an adult CF centre using a structured, individualised transition process was not associated with accelerated clinical deterioration.

Key Words: transition; cystic fibrosis; transfer; adolescents; young people; paediatric CF centre; adult CF centre; lung function
OPTIMISING THE ENDOCRINE REFERRAL PATHWAY IN CYSTIC FIBROSIS CARE

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Cystic Fibrosis (CF) bone disease is a common extra-pulmonary complication of CF and its prevalence is increasing with improved life-expectancy. Optimising bone health requires screening and early management of suboptimal bone density and endocrine risk factors (pubertal delay, growth, malabsorption). Prior to 2016, our endocrinology reviews occurred within CF clinics. Formal referral to external endocrinology clinics was then required due to clinic restructure. In the absence of formal referral criteria, screening and referral practices were audited for 2016-2018 using a referral algorithm recently developed. A CF-related diabetes screening and referral pathway is already well established as part of routine CF care.

Aim: Using the referral criteria of the algorithm to identify gaps in screening and referral between 2016-2018.

Methods: A retrospective review of the 2016-2018 CF cohort aged >10 years was performed to identify current screening and referral for bone density (DXA), pubertal delay (breast/testicular development, menarche, serum sex steroids), height (SDS) and malabsorption (25OHVitD, calcium, parathyroid hormone).

Results: 130 children aged >10 years attended the clinic between 2016-2018. DXA scans were completed in 46% of children. Pubertal screening was poorly performed with no physical exam of testicular development in males, and breast development and menarche documented in only 42% and 21% of age-appropriate females respectively. Fifteen percent of patients did not have hormonal markers of puberty collected. Forty-nine patients fulfilled criteria for formal referral: 10 had low bone density (7%); 26 pubertal delay (18%); 2 had a height <2SDS (2%); 3 low calcium (3%); and 8 high parathyroid hormone (5%). Only 7 (14%) were referred appropriately using 2019 algorithm criteria.

Conclusion: Significant deficiencies in screening and referral have been identified. The impact of the algorithm with formal referral criteria will be assessed in the future.
USE OF MOBILE TECHNOLOGY TO IMPROVE COMMUNICATION

THEMATIC ANALYSIS OF TEXT-BASED MESSAGING BETWEEN CYSTIC FIBROSIS PARENTS AND STAFF

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Background: For patients with Cystic Fibrosis (CF) and their families, coordinating regular medical appointments, and facilitating continuity of care between multidisciplinary team (MDT) members can be challenging. Online patient portals have been used to support communication between patients/families and health teams, and have been well-received in the paediatric setting (Britto et al 2013). Research to date has explored their ease of use and design features with little research into the impact on communication with hospital staff. The My Health Memory (MHM) app was piloted with the CF cohort at the Children’s Hospital Westmead (CHW) from May 2017. Initial functions of the app included a text-based messaging system where communication between a patient/parent and staff is integrated directly into the patient’s electronic medical record (EMR).

Aim: Review the communication (text-messaging) exchanged between patients and healthcare providers at CHW via the MHM app.

Methods: Thematic content analysis of 872 messages exchanged between CF patients/parents and the MDT in the first 12 months of use.

Results: 872 messages were exchanged during the study period with 484/872 (55%) from hospital staff and the remaining 391/872 (47%) from patients/parents. 52/872 (6%) of messages were administrative to outline use of the text-messaging service. The main theme identified was treatment management and adherence (approximately 50% of messages). Message content varied but included; script requests, medications enquiries, clinic/admission planning, test results and symptom management. More detailed communication was noted around respiratory or gastrointestinal symptoms. Parents frequently sought reassurance from their healthcare professionals regarding appropriate symptom management and further follow-up. Additional themes pulled from analysis included patient education, quality of life issues, and MHM app experiences/feedback.

Conclusions: The MHM app has provided an additional avenue for communication amongst CF patients/parents and the MDT. Further research is underway to evaluate other app components in addition to patient/parent and staff satisfaction.
ONE VS TWO BLOOD SAMPLES FOR THERAPEUTIC DRUG MONITORING OF TOBRAMYCIN

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Background: Therapeutic drug monitoring (TDM) for once daily dosing of intravenous tobramycin requires application of pharmacokinetic (PK) methods to predict overall exposure (Area Under Curve (AUC)) and maximum plasma concentration (Cmax) to adjust dosing. The RAH uses SeBA-GEN (Sequential BAyesian Algorithm for GENtamicin) for gentamicin and tobramycin. (1). Although two plasma tobramycin concentrations are routinely used, SeBA-GEN has the ability to calculate PKs and recommend dosing using one sample only, thus minimising many problems associated with obtaining two samples including logistics, patient discomfort and cost.

Aim: The aim of this project was to compare PKs of tobramycin and dosing using one and two blood concentrations.

Method: Blood for tobramycin levels were collected at 1 and 4-6 hours after beginning of the 30 minute infusion. Patient and tobramycin data were entered into SeBA-GEN using (i) only the first sample, (ii) only the second and then (iii) both of the measured concentrations. Clearance (CL), AUC, Cmax, and the recommended dose to obtain TDM targets were estimated. Assuming ≤ 10% difference in PKs and thus dose to be clinically insignificant, power analysis suggested 50 patients were needed.

Results: Data from the first 30 patients (median age 31 years, 57% male, median dose 7.3mg/kg) were collected. Median values of Cmax using sample one, two and both samples were 27.85, 28.45 and 27.55mg/L; median AUC values were 89.2, 89.5 and 89.9 mg/L.hr and median CL were 5.48, 5.35 and 5.33 L/hr respectively. Complete data including statistical analysis and dose recommendations for 50 patients will be presented.

Conclusion: Differences in Cmax, AUC, CL and thus predicted doses using one vs two blood samples were clinically insignificant. We recommend using two samples for first TDM and then only one sample taken between 4-6 hours for follow up TDM.

DIFFERENCES IN CLINICAL OUTCOMES OF PAEDIATRIC CYSTIC FIBROSIS PATIENTS WITH AND WITHOUT MECONIUM ILEUS

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**Background:** Meconium ileus (MI) affects up to 20% of newborns with cystic fibrosis (CF). We compared clinical outcomes between Australian paediatric CF patients with and without meconium ileus (non-MI).

**Methods:** This was a retrospective case-control study of MI and non-MI patients in New South Wales, Australia, from 1988 to 2010. MI patients were matched 1:1 with pancreatic insufficient non-MI patients for age, sex and CF clinic. Clinical measurements, nutrition and gastrointestinal outcomes over this period were compared between groups using linear mixed models for continuous variables to account for age.

**Results:** There were 162 matched pairs (N=324, 52% female) with mean (SD) age of 15.3 (8.2) and 14.9 (7.9) years for MI and non-MI patients respectively (P=0.6). MI patients had poorer FEV1% compared to non-MI patients (estimate (SE) -7.5% (2.2), P=0.0009). There were no significant differences in P. aeruginosa isolation rates; however S. aureus isolation rates were lower in MI patients (72%) compared to non-MI (82%) (OR 0.6 [0.3-1.0], P=0.03). Chronic colonisation rates for P. aeruginosa and S. aureus were not significantly different between groups. MI patients aged 2-20 had significantly lower BMI Z-scores over time (estimate (SE) -0.2 [0.1], P=0.02). MI patients were more likely to receive oral feed supplements (OR 2.8 [1.4-6.1], P=0.003) and gastrostomy formation (OR 4.4 [1.1-24.6], P=0.02).

**Conclusions:** CF patients with MI may have worse lung function, growth and nutrition than non-MI patients over time. Meconium ileus may be an early prognostic factor for CF.
CLINICAL OUTCOMES OF AN AUSTRALIAN COHORT WITH CYSTIC FIBROSIS-RELATED LIVER DISEASE

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Background: Cystic fibrosis-related liver disease (CFLD) is the leading non-pulmonary cause of mortality in cystic fibrosis (CF). We aim to compare respiratory, endocrine and nutritional outcomes of adults and children with severe CFLD and those without (noCFLD) matched for age, sex, genotype, pancreatic insufficiency and treating centre.

Methods: A retrospective review (1998-2016) of all Australian children and adults with CF, pancreatic insufficiency and severe CFLD compared with matched noCFLD controls was undertaken. Generalised linear mixed model analyses were used to assess clinical measurements performed throughout the study period.

Results: 166 patients with severe CFLD and 166 with noCFLD were identified. Percent predicted forced expiratory volume in 1 second (FEV1%) was significantly lower in those with CFLD than noCFLD controls across all ages (estimate (SE) -6.05% (2.12), p=0.004). Frequency of lung transplantation, pulmonary microbiology and mortality were not significantly different between groups. Median (IQR) number of hospital admissions per year were higher in those with CFLD than noCFLD for both respiratory (0.57 (0.2-1.33) vs. 0.4 (0.1-0.89), p=0.002) and gastrointestinal admissions (0.09 (0-0.2) vs. 0 (0-0.05), p<0.001).

Amongst those with CFLD there was a higher frequency of osteoporosis (16.2% vs. 8.4%, OR=2.1 (95%CI 1.01-4.52), p=0.04), and intermittent insulin dependent diabetes (38.5% vs. 19.2%, OR=2.61 (95%CI 1.55-4.47), p=0.0001). In those with CFLD, there was increased use of nasogastric (16.6% vs. 12.6%, OR=2.51 (95% CI 1.06-6.46), p=0.03) and gastrostomy nutritional supplementation (22.9% vs. 13.2% OR=1.93 (95% CI 1.05-3.63), p=0.03). There was no difference in anthropometric z-scores between CFLD and noCFLD.

Conclusion: Patients with CFLD have a phenotype associated with greater co-morbidities than those with noCFLD. CFLD is associated with worse lung function, increased hospitalisations, need for greater nutritional support, increased risk of CF-related bone disease and diabetes when compared to a matched cohort of noCFLD.
CARBOHYDRATE COUNTING ACCURACY IN ADULTS WITH CYSTIC FIBROSIS RELATED DIABETES

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Aim: Poorly controlled Cystic Fibrosis Related Diabetes (CFRD) is associated with adverse impacts on lung function and nutritional status. Insulin therapy is the only recommended medical treatment. Carbohydrate Counting (CC) is used to guide insulin doses and can assist in achieving optimal post prandial blood glucose levels. This study aimed to determine the prevalence of individuals with CFRD who carbohydrate count and to assess the accuracy of CC in hospitalised patients.

Methods: A cross-sectional, mixed-methods, descriptive study recruited individuals with CFRD hospitalised at an Australian tertiary hospital. Consenting patients completed a short questionnaire including their barriers to CC. Patients were asked to estimate the carbohydrate content of their ordered meals provided by hospital foodservices. The study dietitian assessed each meal’s estimation against the actual content.

Results: 17 individuals were recruited to this study; seven had a fixed insulin regimen, and ten had a flexible insulin regimen. Patients in the fixed insulin group reported lower levels of confidence in their ability to carbohydrate count (p<0.001) and placed less importance on CC (p<0.001). 53% of the fixed insulin group and 41.7% of the flexible insulin group’s estimations of the carbohydrate content of the hospital food items were accurate.

Conclusion: Less than two thirds of those patients recruited to this study used CC as a tool to guide insulin dosing, and patients estimated accurate carbohydrate values in only 46% of meals provided. Further research is warranted to investigate the most suitable method to assist accurate carbohydrate content estimations in a hospital setting.
GUT MICROBIOTA IN CHILDREN WITH CYSTIC FIBROSIS: A TAXONOMIC AND FUNCTIONAL DYSBIOSIS

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Objectives: Intestinal dysbiosis has been observed in children with cystic fibrosis (CF), yet the functional consequences are poorly understood. We investigated the functional capacity of intestinal microbiota and inflammation in children with CF and matched healthy controls (HC).

Methods: Stool samples were collected from children (aged 0.8–18 years) with CF and age and gender matched HC. Microbial communities were investigated by iTag sequencing of 16S rRNA genes (V1-V3 region) and functional profiles predicted using Tax4Fun. Inflammation was measured by faecal calprotectin and M2-pyruvate kinase. P<0.05 or q<0.05 (multiple-testing corrected) considered significant.

Results: 27 CF and 27 HC subjects were included (52% male, mean age 8.2 (5.0) and 7.7 (5.3) years, respectively, p=0.2). 24 (89%) CF children were pancreatic insufficient. Paediatric CF gastrointestinal microbiota demonstrated lower richness and diversity (Shannon index; mean difference (95% CI) of -0.74 (-1.00 to -0.49), p=2.2×10^-6) compared to HC. CF samples exhibited a marked taxonomic and functional dysbiosis when compared to HC, evidenced by: (i) a marked increase in the relative abundance of Proteobacteria (genera Escherichia, Shigella and Enterobacter) in CF samples (q<0.05); (ii) the abundance of 56 KO pathways were significantly different between CF and HC pairs (p=0.00019); and (iii) we observed an enrichment of genes involved in short-chain fatty acid (SCFA) (i.e. propanoate and butyrate), antioxidant (i.e. glutathione), amino acid (i.e. phenylalanine) and nutrient (i.e. vitamin B1, B5 and coenzyme A) metabolism in CF samples. Ten genera (including Acidaminococcus) and six KEGG pathways (including benzoate degradation) positively correlated with calprotectin (q<0.05). We observed correlations between several intestinal genera (including Ruminococcaceae) and both growth z-scores and FEV1% (q<0.05).

Conclusion: These taxonomic and functional changes provide insights into paediatric CF gastrointestinal disease. Exploration of potential CF gastrointestinal therapeutics including antioxidants, SCFAs, amino acids and gut microbiota modulators is warranted.
ARE WE ACHIEVING ENERGY DENSITY AT THE EXPENSE OF MICRONUTRIENT DENSITY?

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Background: Children with CF have been reported to consume significantly more energy-dense, nutrient-poor foods than controls where there are now concerns of inadequate micronutrient intake. There are no current or comprehensive dietary studies assessing micronutrient intake in CF children.

Objectives: To evaluate micronutrient intake in children with CF compared to recommended dietary intakes (RDIs), and age- and sex-matched controls.

Methods: Dietary intake of 13 micronutrients was measured in CF children aged 2-18 years and age- and sex-matched controls using a validated food frequency questionnaire (The Australian Child and Adolescent Eating Survey).

Results: CF children (n=82) consumed significantly more energy than controls (n=82) [3142(2531-3822) kcal vs 2216(1660-2941) kcal; p<0.001]. Absolute intake in CF children was significantly higher in all micronutrients except vitamin C and folate, however energy-adjusted intake was significantly lower for all micronutrients except vitamin A, sodium, calcium and phosphorous. Energy-adjusted intake in primary school CF children was significantly less than controls in 8/13 micronutrients. Overall, median intakes exceeded the RDIs for all micronutrients however CF children fell short of the RDIs for folate (26.8%), iron (15.9%) and calcium (9.8%). In pre-school, 50% of CF children and 91.7% of controls did not meet the iron RDI. High school CF and control children failed to meet RDIs for 7/13 and 9/13 micronutrients respectively.

Conclusion: Increased intake of most micronutrients in CF children was largely attributed to higher energy consumption. However, micronutrient density of the diet declined with increasing age, where high school children failed to meet RDIs for most key micronutrients.
WEIGHT AND BODY COMPOSITION CHANGES FOLLOWING ONE YEAR OF TREATMENT WITH LUMACAFTOR-IVACAFTOR IN SEVERE CYSTIC FIBROSIS LUNG DISEASE

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The impact of lumacaftor-ivacaftor (LUM-IVA) on body composition is not widely studied, particularly in severe cystic fibrosis (CF) lung disease.

Aim: To evaluate changes in body composition in adults with CF with FEV1< 40% predicted over 12 months of LUM-IVA treatment.

Methods: Data were analysed for 24 CF adults (13 male, baseline age: 32.6±8.6 years, FEV1%predicted: 34.7±7.4%, BMI: 20.3±2.7kg/m2), who received LUM-IVA for 12 months under a managed access program. Body composition (tetrapolar multifrequency BIA, SECA, Germany) was measured at baseline, 1, 6 and 12 months. Absolute changes (Δ) in BMI, weight, fat-free mass (FFM) and fat mass (FM); and %Δ in weight from baseline were determined. Analyses were performed using linear mixed effects regression modelling and Pearson’s correlation.

Results: No weight change was seen at one month. Weight increased significantly in the first 6 months (mean±SD %Δweight0-6mth: 3.9±1.7%, p=0.03) and was maintained by 12 months (%Δweight 0-12mth +4.9±7.3, p=0.003). BMI increased significantly at 6 months (+0.8±1.3kg/m2, p=0.005), plateauing by 12 months (ΔBMI 6-12mth: 0.1±1.1kg/m2, p=0.65). Fat mass increased significantly at 6 months (ΔFM-0-6mth: 1.8±2.5kg, p=0.003), but plateaued by 12 months (ΔFM6-12mth: 0.7±1.9kg, p=0.12). No changes were seen in mean FFM (ΔFFM-0-12mth: 0.02±2.2kg, p=0.96), indicating overall preservation of FFM. Lower baseline BMI was associated with higher weight gain (r= -0.54, p=0.006) and FFM (r= -0.57, p=0.005) at 12 months. The % of patients with BMI<18.5kg/m2 decreased from 33% at baseline to 13% at 12 months (p=0.003, McNemar’s test).

Conclusions: Gains in weight, BMI and fat mass seen over the first six months on LUM-IVA are attenuated by one year. In this cohort, underweight patients were more likely to improve BMI and body composition. Mechanisms underlying body composition changes require further investigation, including improved appetite, salt/hydration or exercise, or via amelioration of catabolism.
IMPLEMENTATION OF ROOM SERVICE FOR ADULTS WITH CYSTIC FIBROSIS

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Introduction: Room service is a patient centred model of food service which allows patients to order meals to their rooms at preferred times. This model has been shown to improve nutritional intake, patient satisfaction and reduce plate waste and patient meal costs when compared to a traditional hospital foodservice model [1]. The quality of hospital food is an important aspect of patient care, especially for patients with Cystic Fibrosis (CF) who often have higher nutritional requirements and longer average lengths of stay. The Prince Charles Hospital Brisbane, introduced room service in May 2019. This review investigates CF patient satisfaction with the provision of hospital food service pre- and post implementation of a room service model.

Methods: 10 CF inpatients completed the validated Acute Care Hospital Foodservice Patients Satisfaction Questionnaire (ACHFPSQ) and a modified version of the Food and Menu Survey pre- and post the implementation of room service. Scores were measured on a scale of 1 to 5 (5 = very satisfied). Measures recorded components of food quality, meal service quality – food temperature, hunger and food quantity. Clinical demographic data was also collected and analysed.

Results: Prior to the implementation of room service in 2017, CF patients rated overall food service satisfaction as 3.2/5. The average food quality was scored 3.25, and the meal service quality 3.93. Additionally, 4/10 patients reported that they only sometimes receive enough food, and 70% of patients felt hungry between meal times. Satisfaction results post room service implementation will be collected in June 2019.

Conclusion: It is expected that a room service model of care will improve satisfaction with hospital food service for patients with CF, which is predicted to in turn positively influence energy and protein intake and ultimately nutritional status. Further discussion to be included on completion of room service implementation surveys (June 2019).
A NON-RANDOMISED SINGLE CENTRE COHORT STUDY, COMPARING STANDARD AND MODIFIED BOWEL PREPARATIONS, IN ADULTS WITH CYSTIC FIBROSIS REQUIRING COLONOSCOPY

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Background: Adults with cystic fibrosis (CF) have five to ten-fold higher risk (25 to 30-fold risk post-transplant) of colorectal cancer (CRC) than the general population. Whilst international guidelines highlight necessity of intensive colonic lavage to assist optimal examination; limited publications have reported practical aspects achieving this. In this study, we compare two bowel preparation regimens: standard and modified CF bowel preparations.

Methods: Non-randomised adults with CF attending a single centre, requiring colonoscopy investigation were selected. Between 2001 and 2015, 70 adults with CF had an initial colonoscopy procedure. After five exclusions, standard bowel preparation was prescribed for 27 patients, modified CF bowel preparation for 38 patients. Demographic and clinical data were collected.

Results: Significant differences occurred between groups; modified CF bowel preparation group having a higher proportion of "excellent/good" gastrointestinal (GI) cleanse than standard bowel preparation (50.0% versus 25.9%) and lower rates of "poor" cleanse (10.5% versus 44.5%) between groups (p<0.006). Rates of "fair" GI cleanse visualisation were similar between the two groups (39.4% versus 29.6%). Detection rates of adenomatous polyps at initial colonoscopy was higher in modified CF bowel preparation than standard preparation groups (50.0% versus 18.5%, p<0.01). Positive adenomatous polyp detection rate in patient’s aged >40 years was higher (62.5%) than those <40 years (24.3%) (p<0.003). Colonic adenocarcinoma diagnosis was similar in both groups.

Conclusion: This study primarily highlights that standard colonoscopy bowel preparation is often inadequate in patients with CF, and that colonic lavage using modified CF bowel preparation is required to obtain optimal colonic visualisation. A higher rate of polyps in CF patients over 40 years of age was evident; supporting CFF guidelines in recommending colonoscopy screening at 40 years of age, which is earlier than Australian National CRC screening in the non-CF population.

Further studies of optimal and practical CF bowel preparations are required.
INTRADIALYTIC PARENTERAL NUTRITION IMPROVES NUTRITIONAL STATUS IN A COMPLEX CYSTIC FIBROSIS AND END STAGE RENAL DISEASE PATIENT: A CASE REPORT

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This case study reports on a 38 year old female double lung transplant recipient with a complex medical history including cystic fibrosis (CF) and End Stage Renal Disease (ESRD) on haemodialysis who commenced on intradialytic parenteral nutrition (IDPN) due to severe malnutrition and inadequate oral diet and enteral nutrition (PEG). The patient had gastroparesis and severe abdominal pain post prandially which limited oral/enteral nutrition input. IDPN was added to increase nutrition input without contributing further to gastrointestinal symptoms. SMOF Kabiven (986mL bag) was commenced at a rate of 100ml/hr for 4 hours during haemodialysis sessions (3 days per week).

The patient was reviewed regularly by dietitians at haemodialysis unit and/or during hospital admission(s). The patient’s dry weight, biochemical results, nutrition impact symptoms and input were collected prior to commencing IDPN, and at 1 week, 1, 3, 6 and 12 months post IDPN commencement. Six monthly PG-SGA scores showed an improvement from severe (C24) to moderate malnutrition (B7). The patient’s dry weight increased by 4.5kg over a 12-month period. The patient was interviewed about the use of IDPN in the management of her nutritional needs. “I think it has impacted it (my quality of life) in a positive way in that I have gained some weight since starting IDPN and through gaining weight I’ve got more energy to do things.... I’m actually getting out and enjoying some life.” The patient said she would recommend IDPN to others in a similar situation.

Contradicting nutritional recommendations and limited evidence-based guidelines available for people with cystic fibrosis complicated by chronic kidney disease make this population group a challenge to manage. The use of IDPN for this patient with a background of cystic fibrosis, end stage renal disease and severe malnutrition was successful and may provide a useful adjunct therapy for other cases.
IDENTIFYING AND PRIORITISING ADULT PATIENTS WITH CYSTIC FIBROSIS FOR COLORECTAL CANCER SCREENING WITH COLONOSCOPY

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**Introduction:** As the life expectancy of persons with cystic fibrosis (CF) has increased, concerns regarding the risk colorectal cancer (CRC) have arisen. A recent meta-analysis identified a 5 to 10 times greater risk of CRC in adults with CF compared to the general population. Our aim was to address this risk by developing a management plan for colonoscopy prioritisation and screening of CF patients at our centre based on the presence of additional risk factors including age, anaemia, iron deficiency and family history of CRC.

**Methods:** We accessed the adult CF clinical database to identify patients who will be 38 years or older by the end of 2019. We used the clinical database iSOFT to obtain the most recent haemoglobin (Hb) levels, iron studies profiles and colonoscopy reports. Anaemia was defined as Hb <135g/L for men and <115g/L for women. Iron deficiency was based on ferritin <30ug/L and iron deficiency not excluded by transferrin saturation <13% for men and <14% for women.

**Results:** 52 patients were aged 38 years or older. Five (9.6%) patients were found to have anaemia. Eight (15.4%) patients had iron deficiency without evidence of anaemia. Iron deficiency could not be excluded in a further four (7.6%) patients. We also identified 9 (17.3%) patients who had not had annual blood tests. Seven patients have had colonoscopies within the past five years. Family history collection is ongoing.

**Conclusions:** We have identified a subgroup of patients who require further assessment of anaemia or iron deficiency by endoscopic investigation. Those with iron deficiency +/- anaemia will be prioritised first, followed by those with anaemia then positive family history. Patients for whom iron deficiency cannot be excluded or have missing annual bloods will have repeat blood tests. Screening of asymptomatic patients will then be offered to those aged 40 and above.
HIGH DOSE VITAMIN A USE IN CYSTIC FIBROSIS PATIENTS

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Introduction: Cystic fibrosis (CF) patients with pancreatic insufficiency usually receive fat-soluble vitamin supplementation. The 2017 Nutrition Guidelines for CF recommends a standard adult dose of 2500-5000 IU, with an upper limit of 10,000 IU. There is however insufficient evidence on the safe upper limit for vitamin A (VA) supplementation in CF. VA toxicity is associated with hepatotoxicity, alopecia and dry skin. This retrospective analysis evaluates high dose (50,000 IU) daily VA use in a tertiary adult CF centre.

Methods: A subgroup of CF patients on high dose VA from the Royal Prince Alfred Hospital CF Centre, were audited. Demographic, clinical and laboratory data over a 5 year period for each patient was reviewed. The latter included VA levels, C-reactive protein (CRP) and liver function tests (LFT’s). VA levels between 1.4-4 umol/L were considered normal. The medication possession ratio (MPR) for VA was obtained from outpatient pharmacy dispensing records.

Results: 40 patients were included in this analysis (60% male, median age 65 years [IQR 35-74]). The median VA level in this cohort was 1.5 umol/L [IQR 0.2-3.6] with 21/40 (52%) having at least a single reading below 1.4. LFT’s and CRP are listed in Table 1. Pharmacy dispensing records demonstrated compliance ranged from 33-66%. All patients were on pancreatic enzyme supplementation. No evidence of toxicity was identified in their medical record. One patient had critically low VA levels and peripheral vision loss due to documented non-compliance with VA supplementation.

Table 1. Biochemical monitoring of VA

<table>
<thead>
<tr>
<th>Marker</th>
<th>Total cohort n=40</th>
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<tbody>
<tr>
<td>CRP</td>
<td>8.9 (0.1-78) mg/L</td>
</tr>
<tr>
<td>AST</td>
<td>28 (11-99) U/L</td>
</tr>
<tr>
<td>ALT</td>
<td>32 (8-122) U/L</td>
</tr>
<tr>
<td>ALP</td>
<td>109 (35-609) U/L</td>
</tr>
<tr>
<td>GGT</td>
<td>38 (6-591) U/L</td>
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Conclusion: Low normal or sub-therapeutic VA levels were observed despite daily high dose VA supplementation. This could be due to poor compliance. In patients with good compliance, no toxicity was detected.
NASOGASTRIC TUBE FEEDING FOR PATIENTS WITH CYSTIC FIBROSIS AT THE CHILDREN’S HOSPITAL WESTMEAD

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Background: Nasogastric tube feeding (NGTF) is considered for children with Cystic Fibrosis (CF) who cannot meet their growth potential with oral intake alone. We reviewed our unit’s NGTF outcomes by determining: (i) frequency of NGTF versus gastrostomy feeding, (ii) impact of short-term NGTF on nutritional outcomes and (iii) rates of progression to gastrostomy insertion post NGTF.

Method: Data was retrospectively extracted from the Australian CF Data Registry (ACFDR) and electronic records for CHW CF patients from 2007-2017. Exclusion criteria: age (<2years), disability diagnoses, PICU admission, solid-organ transplant, NGTF <7days. Primary outcome measures: anthropometry at baseline (best 12months prior), admission, discharge and post-discharge (1 and 3-6 months); and gastrostomy rates. Fishers test used for statistical analyses.

Results: 83 admissions amongst 46 patients met the study criteria. There were 1-5 NGTF admissions per patient (20/46, 43% >1NGTF admission). Mean length of stay was 15days. Between 2007-2017 patients (percentage total clinic size) receiving NGTF significantly increased from 1/273 (<1%) to 12/207 (6%), (p=0.0003). Significant increases also seen in gastrostomy feeding (12/273 (4%) to 15/207 (7%)), (p=0.0197). Mean (SD) BMI z-score on admission was -1.4 (0.6), returned to baseline at discharge -0.7 (0.6), dipped at 1month post-discharge -1 (0.6), but was maintained at 3-6months post-discharge -1.0 (0.6). Most patients (13/16, 81%) progressed to gastrostomy insertion after NGTF early in the audit period (2007-2012). Subsequently in the later period (2013-2017) this reduced to 8/30 (27%) of patients.). Of the 22 patients without gastrostomy, 13/22 (59%) had acceptable nutrition (BMI>25th centile), 4/22 (18%) sub-optimal (BMI 10-25th centile) and 5/22 (22%) had undernutrition (BMI<10th centile) at transition to adult care or end of the audit period. Median time (range) from last NGTF to transition or end of audit was 10 (1-49) months.

Conclusion: Aggressive short-term NGTF may reduce early gastrostomy insertion rates for paediatric patients with CF.
TELEHEALTH: IMPROVING NUTRITION SUPPORT FOR PAEDIATRIC CF PATIENTS AND PARENTS IN THE HOME

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Background: A strategic priority at the Sydney Children Hospital Network is delivering care close to home. As part of a broader integrated care program, telehealth services have been encouraged across the network since 2017. The Cystic Fibrosis (CF) team at the Children’s Hospital Westmead (CHW) have been recognised as leaders in utilizing this service to improve patient care. The CF nutrition team have offered telehealth to CF patients/families since June 2018.

Aim: Evaluate use and CF patient/parent/staff satisfaction with the nutrition telehealth service.

METHOD: An electronic survey was developed to assess patient/parent/staff satisfaction and identify areas for improvement. Parent/patient participants from June 2018 were identified via the hospital’s patient management system and invited to complete the survey.

Results: 50 sessions were completed amongst 15 participants June 2018-April 2019. Most participants were adolescents (12/15, 80%). Service utilisation increased from a mean of 3.2 (2018) to 7.8 (2019) sessions/month. Sessions per patient/parent ranged from 1-13. Additional staff (nursing, physio, social work or psych) was present at 8/50 (16%) of sessions. 12 patients/parents and 8 staff completed the satisfaction survey. The main identified reasons for instituting telehealth follow-up was weight loss/poor weight gain and adherence challenges. Participant satisfaction was rated high: 4.8/5 by patients/parents and 4.1/5 by staff. Patients/parents also rated information/support provided as 4.8/5 and identified no challenges with the service. Staff identified adolescents forgetting appointments as the main challenge. Most staff (7/8, 88%) agreed/strongly agreed that staffing was adequate for current service provision. There was a mixed response and no consensus regarding service expansion.

Conclusion: Telehealth has proven to be a useful platform at CHW to support CF patients/families address CF nutrition challenges in the home. A staffing enhancement may be required to expand the service longer term.
NUTRITION FOR THE COMPLEX CF NEONATE WITH SIGNIFICANT GASTROINTESTINAL COMPLICATIONS – CASE STUDY

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Baby ZD was born at 33 weeks gestation with Cystic Fibrosis (DF508/V520F and heterozygous for alpha-1 antitrypsin) weighing 1600g (z-score 0.92, Fenton charts). Antenatal ultrasound indicated a probable bowel obstruction. On day 1, she underwent laparotomy and resection of 22cm of ischemic jejunum, 20cm from the duodenal-jejunal flexure, with end-to-end anastomosis. Acholic stool pellets noted intra-operatively were washed out with rectal N-Acetyl Cysteine.

ZD was initially TPN dependant. Early trophic feeding was not initially tolerated; however, breastfeeds were introduced by 8 weeks with full feeding established by 10 weeks. Nutritional challenges included vomiting and faltering growth despite concentrated and fortified 150-170ml/kg 100kcal/100ml extensively hydrolysed Pepti-Junior feeds.

Despite Ursodeoxycholic acid, ZD developed worsening conjugated hyperbilirubinaemia prompting further investigations for biliary atresia. She had no gall bladder on ultrasound, and no biliary excretion on DISIDA scan, and progressed to a cholangiogram and liver biopsy. The bile duct was absent on cholangiogram and the biopsy showed features of large duct obstruction and cholestasis. A Kasai procedure was performed at 11 weeks (weight 2.4kg, z-score -3.94 corrected). Intestinal absorption was further compromised by a relative short bowel, secondary to the Roux-en-Y.

Post-operative TPN resulted in excellent catch up growth. Bowels were opened daily however abdominal distension and vomiting continued; daily ClearLax resolved the abdominal distension, reduced vomiting and ZD transitioned to oral/enteral feeds. By 3.5 months ZD was tolerating 3mmol/kg sodium, fat soluble vitamins, ClearLax, Creon Micro and minimum 150ml/kg Pepti-Junior feeds that were concentrated and fortified to 115kcal/100ml. She weighed 3.19kg (z-score -3.62 corrected).

At 10 months, ZD’s weight and length is tracking along the 3rd centile, with all feeding milestones achieved (solids introduced at 6 months with baby lead weaning from 7 months), vomiting has ceased, and LFTs normalised.
NON-PROPHYLACTIC SUPPLEMENTATION OF VITAMINS AND MINERALS IN AUSTRALIAN CHILDREN WITH CYSTIC FIBROSIS

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Background: Optimising nutrition in patients with cystic fibrosis (CF) is positively associated with improved lung function and survival. Current Australasian CF guidelines (2017) encourage routine supplementation of fat soluble vitamins (A, D, E, K), particularly in pancreatic insufficient (PI) patients. The Royal Children's Hospital (RCH) Melbourne, is the only Australian paediatric centre not prophylactically supplementing vitamins and minerals (vits/mins) for CF patients with VitABDECK, but supplement based on annual nutritional biochemistry. We aimed to determine if this practice is supported, by investigating trends in supplementation recommendation in our cohort.

Method: Retrospective data of all vits/mins recommendation/usage and annual nutrition biochemistry results was collected from clinical notes in patients reviewed by dietitians May 2018-April 2019 (n= 255) and compared to pathology reference ranges and RCH guidelines.

Results: Of 255 patients (148 male, 107 female), 217/255 were PI and 38/255 were pancreatic sufficient (PS). Of the cohort, 87/255 (34.1%) were recommended/using VitABDECK- 83/87 were PI. The primary age groups using VitABDECK were patients aged 13-18 years of age (38/87), and 6-12 years of age (34/87). Of the 217/255 PI patients, 133/217 (61.2%) were not recommended/using VitABDECK and 75/133 (56.4%) of these patients were supplementing ≥2 vits/mins with products other than VitABDECK. Of those not recommended/using VitABDECK, 9/133 patients did not complete annual nutrition biochemistry. Of the remaining 124 patients, 35/124 (28.2%) required no nutrient supplementation based on annual nutrition biochemistry, 50/124 (40.3%) required single nutrient supplementation and 39/124 (31.4%) had ≥2 low nutrients contained in VitABDECK.

Conclusion: Most patients were PI and were not recommended VitABDECK based on review of annual nutrition biochemistry findings, this suggests our current practice of prescribing vits/mins by need only is valid, however close monitoring is required to ensure safety.
AUDIT OF THE PREVALENCE, PROGRESSION AND RISK FACTORS FOR CYSTIC FIBROSIS BONE DISEASE IN THE WA ADULT POPULATION WITH CF

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Introduction: Bone disease is common in people with cystic fibrosis. The CF conductance regulator (CFTR) gene mutation interferes with osteoblastic bone formation by downregulating proteins such as osteoprotegrin and prostaglandin E2. Whilst previous studies have examined CF bone disease (CFBD) in children and young adolescents, CFBD progression and risk factors in older adults is less well understood. CFTR modulators improve CFTR protein function and lung function in patients with CF, however its effects on BMD are unclear.

Method: This is a retrospective observational study on a cohort of adult patients attending the Western Australia adult CF centre at Sir Charles Gairdner Hospital. All patients with ≥ 1 DEXA scans performed between 2000 and 2018 were included in this study. Patient clinical characteristics were extracted from the Australian CF data registry. Serum vitamin D, testosterone and calcium levels were retrieved from the hospital electronic database.

Results: A total of 132 patients were included in this study. 11/132 had osteoporosis, 47/132 had osteopenia and 58/132 had CFBD. 44/132 patients were on CFTR modulator therapy.

Femur and hip T scores were positively correlated with FEV1, FVC and BMI. No correlation between BMD and Vit D, Ca, BMI was noted.

For patients on CFTR modulators, there was decline in Hip BMD despite introduction of CFTR modulator therapy, with no significant changes in rate of Spine or Femur BMD.

Conclusion: Prevalence of CFBD in our cohort was high (44%). Low BMD in adults is significantly associated with low lung function, therefore optimisation of lung function is a priority in preventing CFBD.

Amongst 22 adults on CFTR modulators, no significant changes in spine or hip BMD was noted. With PBS listing of Orkambi in 2018, the potential impact of Orkambi in CFBD can be further characterised in adults with a wide spectrum of CF disease severity.
PILOT IMPLEMENTATION OF A MORE FLEXIBLE FOOD SERVICE MODEL IN CYSTIC FIBROSIS (CF) TO IMPROVE HOSPITAL FOOD INTAKE

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Background: CF inpatients have longer and more frequent hospital admissions and higher nutrition requirements than other adult inpatient populations. Multiple system and patient factors contribute to not meeting nutrition requirements, despite having access to adequate nutrition from hospital food including a range of short-order meal items. Patients self-source a high proportion of food to make up gaps not met by their hospital food intake.

Aim: To implement additional flexible food service options for CF adults to reduce barriers to adequate nutrition from hospital food.

Methods and Service Development Approach: A food service model offering additional flexible food options supplementing the usual hospital CF diet was introduced as a pilot on the two primary CF wards. This included a ‘Snack Pantry’ (SP) (10 non-perishable food items, 24-hour access), kept in a secure location to which nursing staff and dietitians have access. SP items provided an average 990kJ and up to 1500mg sodium per item. Patients ordered via their nurse. CF dietitians were responsible for monitoring and replenishing stock levels. ‘Meals on Demand’ (MOD) (frozen meals, stored and re-heated in hospital kitchen, delivered within 30 minutes of ordering) was also piloted. MOD could be ordered in addition to regular hospital meals, any time from 9am-5pm. Orders for MOD were placed by ward clerk or nursing staff and picked up from the kitchen by ward support staff.

Results/Outcomes: The SP was well-utilized and has since been permanently implemented on the two CF wards. The MOD was poorly utilized and discontinued after the pilot phase.

Conclusion: Food service innovations, such as a SP, can be successfully implemented on CF wards to reduce barriers to adequate nutrition. Future directions include evaluation of the SP including patient satisfaction, contribution to nutritional intake, reviewing of stock control methods, and trialling a SP in other clinical areas.
MEASURING ENERGY EXPENDITURE DURING PULMONARY EXACERBATIONS OF CYSTIC FIBROSIS USING INDIRECT CALORIMETRY

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Introduction: The energy demands of patients with Cystic Fibrosis (CF) can vary greatly depending on pancreatic function, clinical and nutritional status. There is little understanding of how often to calculate energy requirements to capture changes during CF pulmonary exacerbation (PEX) and meet nutritional needs for optimal outcomes. Furthermore, there is little published on how predictive equations can be practically applied to reflect potential changes in energy expenditure. This study aimed to measure within-individual variation in resting energy expenditure (REE) via indirect calorimetry (IC) during both pulmonary exacerbations (PEX) requiring IVABx and during a period of clinical stability; and compare predicted REE (pREE) determined using equations with measured REE (mREE) during both PEX and clinical stability.

Methods: 5 participants (3 female, 2 male) with mean age and BMI of 34.4 ± 13.14yo and 20.46 ± 1.89 kg/m² respectively, had mREE (using IC), body composition (using bio-impedance analysis) and pREE (using the Schofield equation) determined at three time points: (i) the start of a 14 day IVABx treatment course for PEX; (ii) end of the IVABx treatment; and (iii) during a period of clinical stability.

Results: mREE during PEX treatment was significantly greater than pREE (mean difference: 272kCal; 95%CI:17-528; p=0.04). mREE during clinical stability also tended to be greater than pREE but was not statistically significant (mean difference: 332kCal; p:0.06). No significant changes in weight, fat mass, fat-free mass or mREE were observed from the start and end of IVABx treatment.

Conclusion: Preliminary results suggest that predictive equations underestimate REE in CF adults during PEX, when compared with mREE. Results varied for within-individual changes in mREE over the PEX treatment period, but were not statistically significant. This study highlights the potential role for IC to support optimising nutritional outcomes. Further research with a larger sample size in this group is required.

References:
YOU ARE WHAT YOU EAT: IDENTIFYING CORRELATIONS BETWEEN DIETARY INTAKES AND THE GASTROINTESTINAL MICROBIOTA AND INFLAMMATION IN CYSTIC FIBROSIS (THE EARTH STUDY)

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Introduction/Aim: Intestinal dysbiosis in cystic fibrosis (CF) is likely multifactorial and diet is an underexplored microbial modulator. This study aims to 1) explore the relationship between the CF (high-calorie, high-fat) diet and gastrointestinal microbiota, and 2) examine the interplay between these dietary intakes and intestinal inflammation (measured using faecal calprotectin).

Methods: This is a preliminary cross-sectional analysis of a prospective, longitudinal, observational cohort study at Sydney Children’s Hospital Randwick. Children with CF and healthy controls (HC) completed a clinical survey, food frequency questionnaire (ACAES), and provided stool samples. Samples underwent 16s rRNA sequencing (V4 region) and faecal calprotectin was measured. Analyses were performed in RStudio (v3.4.4).

Results: 33 CF (16 female (48.5%); median age [IQR]=8.8 years [4.6-12.0]) and 27 HC (17 female (63.0%); median age [IQR]=12.6 years [10.1-15.1]) subjects were recruited. Stool alpha diversity (Shannon Index) was significantly lower in CF compared with HC (mean (SD)=2.39 (0.72) vs 3.67 (0.30) respectively; p<0.001). ANCOM stool analysis revealed that relative abundances of Enterococcus and Enterobacter were higher, while Akkermansia were lower (FDR<0.05) in CF compared to HC. CF faecal calprotectin was significantly higher than HC (mean (SD)=88.4 (4.8) vs 11.3 (4.8) respectively; p=0.002). CF dietary intake revealed: (i) meat positively correlated with intestinal Cyanobacteria (R=1, q<0.001), (ii) saturated fat positively correlated with calprotectin (R=0.83, p=0.04) and (iii) resistant starch negatively correlated with calprotectin (R=-0.83, p=0.04).

Conclusion: These preliminary results, based on a robust study methodology, provide insight into the relationship between dietary intakes and gastrointestinal health, and associated microbial influences. Increased intestinal inflammation and dysbiosis in children with CF is associated with dietary intake (such as meat, saturated fat, and resistant starch). The health impact of dietary interventions in CF may extend beyond the traditional high-calorie diet.
DOES ROOM SERVICE MEET THE NUTRITIONAL NEEDS AND CONSUMER EXPECTATIONS OF A CYSTIC FIBROSIS POPULATION?

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Mater Health, was the first hospital group in Australia to implement Room Service (RS), making Mater’s Cystic Fibrosis (CF) population the first to experience this foodservice model. The on-demand RS model allows patients to order freshly prepared meals appropriate for their nutritional requirements from a single integrated menu. Since RS implementation in 2013, Mater has undertaken extensive evaluation of the model, adding to a growing body of evidence confirming the benefits of RS. The literature demonstrates improvements in patient satisfaction, food costs, plate waste and nutritional intake, including as a percent of patient requirements. However, to date there are no studies that have evaluated the RS model specifically within a CF Population.

The increased frequency of admissions, longer lengths of stay and the lower median age differentiate the CF cohort from many general hospital populations. The impact of these characteristics on the success of RS in the CF population needs to be explored. In addition, the changing nutritional needs of the CF population, and recent release of the Nutritional Guidelines for Cystic Fibrosis in Australia and New Zealand (2017) further warrants evaluation of RS within the CF population.

Our study aims to evaluate whether Mater’s RS model meets the CF population’s nutritional needs, adheres to evidence based nutritional recommendations and meets CF consumer expectations.

Data collection has commenced, including average number of meals ordered per patient per day, plate waste, menu items chosen and nutritional intake compared with individual requirements. This data will be compared against general hospital population data, as well as CF nutritional recommendations. Patients’ ordering patterns, perceptions and satisfaction with RS will also be explored, through surveys and semi structured interviews. Data collection is expected to be completed in September 2019 and will inform future quality improvements and design of patient centred foodservice models.
THE ALTERED INTESTINAL VIROME IN CHILDREN WITH CYSTIC FIBROSIS

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Objectives: The intestinal virome is yet to be explored in children with cystic fibrosis (CF). Bacteriophages influence microbial species diversity and biochemical cycles. We investigated the composition and function of intestinal viral communities in children with CF and healthy controls (HC).

Methods: We performed a case-control study on children with CF and age and gender matched HC. Stool samples were enriched for viral DNA/RNA by viral extraction, random amplification and purification before sequencing (Illumina MiSeq). Taxonomic assignment of viruses was performed using Vipie. Functional annotation was performed using Virsorter. Inflammation was measured by faecal calprotectin and M2-pyruvate kinase (M2-PK). P<0.05 or q<0.05 (multiple-testing corrected) considered significant.

Results: Eight CF and eight HC subjects were included (50% male, mean age 6.9±3.0 and 6.4±5.3 years, respectively, p=0.8). All CF subjects were pancreatic insufficient. Samples contained an average 568,964±201,042 reads mapped to known viruses (0.038±0.11%), viral dark matter (75.6±27.5%), bacterial ribosomes (23.7±27.2%) and humans (0.63±0.88%). No overall significant difference in Shannon index CF and HC was identified (mean difference (95%CI) -0.29 (-1.30 – 0.72), p=0.6). Taxonomy-based beta diversity (presence-absence Bray-Curtis dissimilarity) was significantly different between CF and HC (R2=0.12, p=0.001). Myoviridae, Faecalibacterium phage FP Taranis and unclassified Gokushovirinae were significantly decreased in CF compared with HC (q<0.05). In children with CF (compared to HC), the relative abundance of: (i) a peptidoglycan-binding domain of the peptidoglycan hydrolases (COG3409) was significantly increased (q<0.05) and (ii) capsid protein (F protein) (PF02305.16) was significantly decreased (q<0.05). Picornavirales, Picornaviridae, and Enterovirus positively correlated with weight and BMI (r=0.84, q=0.01). Single-stranded DNA viruses negatively correlated with M2-PK (r=-0.86, q=0.048).

Conclusion: Children with CF have an altered intestinal virome compared to matched HC. Intestinal viruses and their functions may have important clinical implications. Further exploration of Faecalibacterium phage, Gokushovirinae and phage lysins is warranted.
EXAMINING THE ASSOCIATION BETWEEN NUTRITIONAL OUTCOMES DURING A PULMONARY EXACERBATION OF CYSTIC FIBROSIS WITH THE INTERVAL TO THE NEXT EXACERBATION

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Introduction: Treatment of pulmonary exacerbations (CFPEx) aims to restore lung function and nutritional status. One recent study suggests weight gain during treatment of an exacerbation increases the time to the next CFPEx.

Aim: To examine the weight changes during CFPEx treatment in an adult CF cohort and its potential association with time to next CFPEx.

Methods: Retrospective review of CFPEx treatment in all patients who received intravenous antibiotics at the WA adult CF centre between 2016-2017. One episode of CFPEx per patient was randomly selected for inclusion.

Results: 80 patients received CFPEx treatment between 2016-2017. 46/80 (57.5%) patients had comprehensive clinical data documented and were included. At the start of treatment, patients’ lung function and weight were significantly lower than baseline (Median FEV1% difference: -10% [95%CI: -9 to -16%; p<0.05], Median weight difference: -0.6 kg [95CI: -0.4 to -2.0; p:0.005]. At the end of treatment, lung function significantly improved (Median FEV1% change: +9% [95%CI: 6-11 %]; p<0.001), but no significant weight change was observed (Median difference: -0.1 kg [95CI: -0.6 to 0.3kg], p:0.5). Patients’ end-of-treatment weight remained significantly lower than baseline (Mean difference: -0.7kg; [95%CI: -0.6 to -3.5 kg]; p:0.005). Suboptimal weight recovery was particularly observed in hospitalised patients (Mean difference: -1.6kg [95%: -0.7 to -2.7kg]; p:0.002) and not in patients receiving home IV antibiotic therapy (Mean difference: -0.6kg [95%CI: -1.8 to 0.6 kg]; NS). Weight gain during CFPEx was not significantly associated with time to next CFPEx.

Conclusion: Our study highlights challenges in achieving optimal nutritional target during CFPEx in adults with CF. Moving forward with this data we need to identify potential barriers to optimal nutritional intake during PEx. We did not observe a significant association between weight change and time to next CFPEx. Further studies with a larger patient sample is required to understand this potential association.
THE ALTERED INTESTINAL PROTEOME IN CHILDREN WITH CYSTIC FIBROSIS

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Objectives: Intestinal dysbiosis exists in cystic fibrosis (CF) and host-expressed proteins serve as reporters on host-microbiota interaction. We aimed to characterise the human intestinal proteome in children with CF and healthy controls (HC). Furthermore, we aimed to explore associations between intestinal microbiota and the human intestinal proteome in CF.

Methods: A pilot study analysing the faecal proteome of CF and HC children. Stool samples were chemically and mechanically processed to extract proteins before liquid chromatography-mass spectrometry (LC-MS/MS). Scaffold v4.7.5 was used. Differentially abundant proteins tested by ANOVA (Benjamini-Hochberg). Microbial communities were investigated by iTag sequencing of 16S rRNA genes. P<0.05 or q<0.05 (multiple-testing corrected) considered significant.

Results: 12 CF (83% pancreatic insufficient (CF-PI); 45% female; mean age 8.2±4.5 years) and 5 HC (100% female; mean age 8.3±4.9 years) subjects included. 324 unique human and 312 unique bacterial proteins identified between all samples. 29 human proteins were differentially abundant between CF-PI, CF pancreatic sufficient (CF-PS) and HC, 11 being proteases decreased in CF-PI (q<0.05). Proteins associated with immune function (cluster of Ig mu chain C, leukocyte elastase inhibitor, galectin-3-binding protein and Ig kappa chain V-ll region RPMI 6410), inflammation (meprin A subunit beta), enterocyte damage (angiotensin-converting enzyme 2) and antioxidants (glutathione reductase, mitochondrial) were significantly elevated in CF-PI subjects. Lysozyme C (bacteriolytic enzyme) was significantly decreased in CF-PI subjects. Shannon index was significantly lower in CF compared to HC, 3.05±0.66 vs 4.08±0.27 respectively, p=0.0004. CF subjects had significantly higher relative abundances of Proteobacteria, Escherichia Coli and Enterobacter (q<0.05). Glutathione synthetase negatively correlated with the log-relative abundance of several bacterial genera in CF-PI samples (spearman correlation, q<0.05).

Conclusion: The human intestinal proteome appears altered in CF children. In CF-PI subjects, there appears to be upregulated immune function, inflammation and increased cell damage. Further investigation into host-expressed proteins and specifically glutathione is warranted.
RETROSPECTIVE AUDIT OF THE EFFECTIVENESS OF AN APPETITE STIMULANT (PERIACTIN) IN PATIENTS WITH CF ATTENDING THE WOMEN’S AND CHILDREN’S HOSPITAL

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Aim: To conduct a retrospective audit to assess the effectiveness of cyproheptadine (periactin®) as an appetite stimulant in improving the nutritional status of patients with CF at risk of malnutrition at the Women’s and Children’s Hospital (WCH).

Methods: A retrospective audit was conducted to assess change in BMI z-score in the 12 months prior to, compared to the 12 months post, commencement of periactin.

Inclusion criteria: All WCH patients with CF prescribed periactin from 2013-2018 for a period ≥12 months.

Exclusion criteria: patients who were concurrently prescribed oral or enteral nutrition support.

Pharmacy records were used to identify those prescribed periactin. Growth parameters (weight, height), pancreatic status, CF genes, lung function, CF Related Diabetes (CFRD) status +/- treatment, use of oral or enteral supplementation, CFTR modulator use and periactin dose were obtained via medical records and lung lab reports. BMI and BMI z-scores were calculated.

Results: Fifteen patients were included in the analysis. Fourteen (93%) were pancreatic insufficient. The daily dose of periactin prescribed ranged from 0.1-0.32mg/kg. No patients had CFRD. No patients were on CFTR modulators.

Mean BMI z-score at commencement of periactin was -1.06. Mean BMI z-score after 12 months on periactin was -0.15. There was a -0.52 mean change in BMI z-score in the 12 months prior to commencement of periactin, compared to a 0.91 mean change in BMI z-score in the following 12 months on periactin (p=0.0002).

Mean change in FEV1(%predicted) in the 12 months prior to commencement of periactin was -6.2 compared to +2.79 in the following 12 months on periactin (p=0.07).

There was no correlation found between dose of periactin and change in BMI z-score (r=0.031).

Conclusion: The results of this retrospective audit indicate periactin is effective at improving the nutritional status of paediatric CF patients who are at risk of malnutrition.
IL-1 IS ASSOCIATED WITH EPITHELIAL NECROSIS IN CYSTIC FIBROSIS AIRWAYS FOLLOWING RHINOVIRUS INFECTION

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Introduction: Necrosis of airway epithelial cells (AEC) resulting in airway inflammation driven by interleukin (IL) 1 is a characteristic finding in cystic fibrosis (CF), driven by mucus obstruction of the airway and a response to human rhinovirus (RV) infection. As little is known about IL-1 and pathogenesis of CF lung disease and RV is a common early life infection, this study aimed to assess cellular and inflammatory responses of CF and non-CF AEC infected with RV.

Method: CF and non-CF AEC (n=9) were infected with RV (MOI 3) for 24 hours and viable, necrotic and apoptotic events assessed via flow cytometry (% total events). IL-1α, IL-1β, IL-1Ra and IL-8 were measured in cell culture supernatants (pg/mL). Data was natural log transformed, Friedman and Mann-Whitney tests used for significant differences (mean±SD), and Spearman for correlations (p<0.05).

Results: RV infection resulted in lower viable events in non-CF AEC (4±0.2 vs 3.2±0.6, p<0.05), increased necrotic events in non-CF and CF AEC (2±0.3 vs 2.4±0.4 & 2.1±0.2 vs 2.4±0.4; p<0.05) and increased apoptotic events in non-CF AEC (3.2±0.3 vs 3.5±0.1, p<0.05). RV infection also increased IL-1α and IL-1β protein in non-CF (3.9±0.5 vs 6±0.5 & 0.8±1.4 vs 2.9±0.7; p<0.05) and CF AEC (3.7±0.7 vs 5.5±0.5 & 0.8±1.5 vs 2.9±1.3; p<0.05) supernatant. IL-1α and IL-1β in supernatant positively correlated with necrosis (r=0.80 & r=0.77 respectively; p<0.0001) in CF but not non-CF AEC after RV infection. RV infection increased IL-1Ra protein in non-CF (7.1±0.1 vs 8.9±0.3, p<0.05) and CF AEC (7.4±0.4 vs 8.6±0.3, p<0.05) supernatant, although IL-1Ra was higher in non-CF AEC (p<0.05). RV infection increased IL-8 protein in non-CF and CF AEC (8.2±0.6 vs 9.6±0.3 & 8.1±0.4 vs 8.9±0.6; p<0.05).

Conclusion: RV infection of CF and non-CF AEC increased necrotic events and in CF AEC was associated with IL-1, suggesting this pathway as a novel anti-inflammatory target.

Support: CFA, CFWA, USCF, BMBF
A MULTICENTRE, OBSERVATIONAL CASE-CONTROL STUDY TO DETERMINE THE EFFICACY AND SAFETY OF LUMACAFTOR/IVACAFTOR IN PATIENTS WITH SEVERE LUNG DISEASE AND CYSTIC FIBROSIS

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Background: Lumacaftor/ivacaftor (LUM/IVA) has been shown to improve percent predicted FEV1 (ppFEV1) and reduce exacerbation frequency in patients with ppFEV1 40 – 90. However, there is limited safety or efficacy data on its use in patients with ppFEV1 < 40.

Aim: To determine the safety and efficacy of LUM/IVA in patients > 12 years old with cystic fibrosis (CF), homozygous for F508del CFTR mutation and with ppFEV1 < 40.

Methods: A retrospective cohort design was used. Data was collected from patients > 12 years of age with CF, homozygous for F508del CFTR mutation and with ppFEV1 < 40, and compared with data from age- and sex-matched controls with CFTR mutations leading to severe CFTR dysfunction and ppFEV1 < 40. Seven Australian CF centres contributed patient data. The primary outcome was the rate of pulmonary exacerbations requiring the use of intravenous antibiotics over a 12-months. Secondary outcomes included: mean rate of change in ppFEV1; time to first exacerbation; death; lung transplantation; treatment-emergent adverse events, and discontinuation of LUM/IVA.

Results: Data was collected from 102 patients; 72 patients on LUM/IVA and 30 matched controls. Treatment substantially reduced mean (sd) exacerbations; 1.49 (1.74), compared to controls 3.06 (2.45), p<0.001. A reduction in incidence rate ratio of 0.485 95%CI 0.318 to 0.740), when adjusted for prior exacerbations the effect was greater. However, side effects were common and treatment was discontinued in 43%.

Conclusion: In this cohort of patients with CF and ppFEV1 < 40, when tolerated, LUM/IVA markedly reduced the frequency of pulmonary exacerbations when compared to matched controls. However, there was a high discontinuation rate due to adverse events.
THE OTOTOXICITY OF INTRAVENOUS AMIKACIN WHEN USED IN THE TREATMENT OF NON TUBERCULOUS MYCOBACTERIA INFECTION IN CYSTIC FIBROSIS

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Background and objective: Pulmonary non-tuberculous mycobacteria (NTM) is an important pathogen in cystic fibrosis (CF) patients with mortality rates up to 36% at 5 years. Current antibiotic eradication regimens are complex with multiple agents required. The aminoglycoside Amikacin is widely accepted as a component of treatment however has known ototoxicity. Despite the significant amount of literature regarding aminoglycoside-induced sensorineural hearing loss, there is limited known on the effect of amikacin on hearing loss in the context of NTM management in CF.

Methods: We performed a retrospective study of CF patients treated with amikacin for NTM between January 2008 to December 2018 at a single site tertiary paediatric hospital in Adelaide, Australia. The aim was to describe the prevalence of hearing loss in these patients and to determine whether there was a dose response relationship between total cumulative antibiotic exposure, pharmokinetic parameters (area under the curve and maximum serum concentration) and hearing loss.

Results: Eleven patients with CF received NTM eradication inclusive of IV amikacin within the defined time period for a mean duration of 68.41 days (range 27-120). Sensorineural hearing loss was identified in six patients (55%) with all hearing loss within the high or extended high frequency range of mild-moderate severity. Only one patient with hearing loss had resolution post cessation of IV amikacin. Patients with hearing loss had higher mean total cumulative doses (1555.71/mg/kg vs. 1282.54/mg/kg), longer duration of treatment (72.17 vs. 64.67 days), higher mean AUC levels (210.07 mg.hr/L vs. 185.57mg.hr/L) and higher mean Cmax levels (95.92 mg/L vs. 82.99mg/L) comparative to those without hearing loss.

Conclusion: High tone sensorineural hearing loss is common in CF patients undergoing treatment for NTM eradication. This retrospective study showed an association between total cumulative dose, treatment duration and higher levels of IV amikacin (measured by AUC and Cmax) with ototoxicity.
AN IN VITRO STUDY TO RATIONALISE COMBINATION THERAPIES FOR CYSTIC FIBROSIS TREATMENT

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The vicious cycle of persistent inflammation in association with recurrent infections that leads to pulmonary failure is the primary cause of death in cystic fibrosis (CF) patients. Consequently, current CF therapy is mainly focused on inhaled antibiotics (i.e. tobramycin) in combination with oral anti-inflammatory drugs (i.e. high dose ibuprofen) and occasional oral/intravenous antibiotics (i.e. azithromycin). However, according to the current CF consensus treatment guidelines, there is a knowledge gap in the understanding of potential interactions between these drugs in terms of their efficacy and mechanism of action, resulting in variable treatment outcomes. In fact, a recent clinical trial of 263 CF subjects has shown that oral anti-inflammatory azithromycin antagonises the antimicrobial efficacy of inhaled tobramycin. Based on this recent evidence, this study aims to investigate other potential interactions between tobramycin and other anti-inflammatory drugs used in CF treatment. To achieve this aim, an in vitro inflammation study of two combinations of tobramycin with either ibuprofen or azithromycin on IL-8 expression was performed using CuFi-1 and BEAS-2B pulmonary epithelial cell lines. Both combinations demonstrated a significant increase in IL-8 concentration in the two cell lines compared to the untreated control (cells stimulated with lipopolysaccharide (LPS) only), suggesting a drug-drug interaction resulting in enhanced inflammatory effect. This preliminary study aims to rationalize combination therapies for CF treatment, which in turn, could potentially change current clinical practice, improve standard of care and overall, reduce treatment burden on CF patients.
UTILISATION OF THE 2003 CYSTIC FIBROSIS FOUNDATION CONSENSUS CONFERENCE CRITERIA IN THE DIAGNOSIS OF ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS IN CYSTIC FIBROSIS AT A TERTIARY HOSPITAL: A RETROSPECTIVE AUDIT

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Background: Allergic bronchopulmonary aspergillosis (ABPA) is an important complication in Cystic Fibrosis (CF) that can result in worsening lung function. Despite the release of the 2003 Cystic Fibrosis Foundation (CFF) Consensus Conference minimal diagnostic criteria, the diagnosis of ABPA in CF remains challenging.

Objective: To determine how many paediatric CF patients at a tertiary hospital who were commenced on ABPA therapy satisfied the 4 minimal diagnostic criteria outlined by the 2003 CFF Consensus Conference.

Methods: We conducted a retrospective audit of paediatric CF patients at the Royal Children’s Hospital, Melbourne who were treated for a new ABPA diagnosis between November 2012 and November 2018. Using medical records, the number of criteria each patient met at diagnosis was determined. The diagnosis of ABPA was defined as the commencement of prednisolone therapy for a minimum of 2 weeks specifically for ABPA. Patients with confounding conditions, incomplete or complex medical records were excluded.

Results: We identified 39 patients with current or prior ABPA from the 2017/2018 CF clinical annual review. 19 patients were excluded: diagnosis prior to November 2012 (8), mistakenly included (4), confounding infections (4), incomplete medical records (3). Of the 20 patients included, 15 (75%) fulfilled 4/4 criteria at diagnosis. Of the 5 patients who met <4 criteria, four patients met 3/4 criteria and one patient met 2/4 criteria. Failure to meet criterion 4 (positive Aspergillus precipitins or radiological changes) was the most commonly failed criterion (3/5), with no clinical deterioration (2/5) and IgE below threshold (1/5) also identified.

Conclusion: Our results show that despite international criteria, ABPA treatment is initiated in a significant proportion of paediatric patients based on other factors. Our results highlight the differences in diagnostic practices in one centre alone, and further studies are needed to investigate diagnostic variations across multiple centres and reasons behind these differences.
COMPARISON OF AIRWAY MICROBIOLOGY IN PAEDIATRIC CYSTIC FIBROSIS PATIENTS WITH AND WITHOUT SUPERIMPOSED ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS

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Background: Allergic bronchopulmonary aspergillosis (ABPA) is an important complication in Cystic Fibrosis (CF). The relationship between airway microbiology and the development of ABPA is poorly understood, as is the effect of steroid treatment on airway microbiology.

Objective: To compare the airway microbiology in the 12 months before and after diagnosis of ABPA in CF, as well as to age matched CF controls without ABPA for the same time periods.

Methods: Twenty paediatric CF patients at the Royal Children's Hospital, Melbourne were treated for a new ABPA diagnosis between November 2012 and November 2018. The sputum cultures in the 12 months pre- and post-ABPA diagnosis were collected for each ABPA patient. Two age matched CF patients without ABPA, and who had at least one valid sputum culture for the pre- and post-ABPA dates of interest, were selected for each ABPA patient (control group). The airways microbiology in the ABPA group was compared to that of the control group for the two 12 months periods.

Preliminary Results: Analysis is still underway. In the year prior to diagnosis, 45% (5/11) of patients in the ABPA group reported positive cultures for Aspergillus fumigatus, compared to the 23% (5/22) reported for the control group. In the ABPA group, 27% (3/11) had positive cultures for Pseudomonas aeruginosa in the pre-ABPA diagnosis period and this rose to 45% (5/11) for the post-ABPA diagnosis period. This is in comparison to the control group, where 9% (2/22) were positive for P. aeruginosa in first 12 months, and 23% (5/22) were positive in the second 12 months of interest.

Conclusion: Our preliminary results suggest that patients with ABPA have higher rates of P. aeruginosa culture both in the lead up to diagnosis and in the year following diagnosis.
EFFECT OF FIRST STEROID TREATMENT COURSE ON CLINICAL MARKERS OF ALLERGIC BRONCHOPULMONARY ASPERGILLOSI IN PAEDIATRIC CYSTIC FIBROSIS

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Background: Allergic bronchopulmonary aspergillosis (ABPA) is an important complication in Cystic Fibrosis (CF); although, its diagnosis is challenging due to its similar presentation to infective exacerbations. The 2003 Cystic Fibrosis Foundation (CFF) Consensus Conference released four minimal diagnostic criteria in hopes of overcoming these difficulties. However, they have been heavily criticised in literature.

Objective: To compare the FEV1 response to steroid therapy in patients with a new ABPA diagnosis who satisfied the CFF’s 4 minimal criteria versus those who did not satisfy the criteria.

Methods: Twenty paediatric CF patients at the Royal Children’s Hospital, Melbourne were treated for a new ABPA diagnosis between November 2012 and November 2018. The patients were classified into the groups “Complete Criteria”, for those who met 4/4 criteria at diagnosis, and “Incomplete Criteria”, for those who met <4 criteria at diagnosis. For each patient, the change in FEV1 percent predictive (∆FEV1), defined as the difference between the last FEV1 prior to ABPA diagnosis and the first FEV1 post steroid cessation, was calculated. ∆FEV1 was compared between the two groups.

Preliminary Results: Analysis is still underway. At diagnosis, 15 patients met 4/4 criteria, and 5 patients met <4 criteria. Of the 7 patients analysed so far in the Complete Criteria group, ∆FEV1 ranged from -7.0% to +39.7%, with an average ∆FEV1 of +17.1%. ∆FEV1 for the 3 patients analysed so far in the Incomplete Criteria group, and the criteria they did not satisfy, were: -3.8% (criterion 4 not satisfied), -5.3% (criterion 1 not satisfied), and +25.7% (criterion 4 not satisfied). The average ∆FEV1 for this group was +5.5%.

Conclusions: Patients who satisfy the CFF’s full criteria for the diagnosis of ABPA in CF are more likely to benefit from steroid treatment compared to those who do not fulfil the criteria.
HIGH PREVALENCE OF ANTIBODIES THAT INHIBIT KILLING OF
P. AERUGINOSA IN A CYSTIC FIBROSIS COHORT

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Rationale: Pseudomonas aeruginosa is the principle pathogen implicated in progressive and recurrent respiratory infections in cystic fibrosis. Recently, impaired serum-mediated killing of P. aeruginosa was associated with increased severity of respiratory infections in patients with bronchiectasis. This inhibition was mediated by high titres of O-antigen-specific IgG2 antibodies. Patients with these ‘inhibitory antibodies’ have been successfully treated with plasmapheresis.

Objectives: To determine the prevalence and nature of ‘inhibitory antibodies’ in patients with CF and P. aeruginosa infection without cognate isolates.

Methods: IgG2 titres were measured in 75 serum samples obtained from patients with CF against eight P. aeruginosa serotypes. To confirm the inhibitory capacity of serum, serum bactericidal assays were performed.

Measurements and Main Results: 24 of 75 patients had serum that inhibited healthy control killing of P. aeruginosa. Interestingly, in a small number of patients with low IgG2 titres, increased antibody affinity appeared to mediate inhibition of killing. Moreover, in two patients with low IgG2 titres but inhibitory serum, we found that high titres of O-antigen specific IgA also inhibited killing of P. aeruginosa.

Conclusions: Inhibitory antibodies are highly prevalent (33%) in patients with CF and P. aeruginosa. Both titre and affinity of these antibodies is important for the inhibition. IgA specific for O-antigen can also inhibit serum-mediated killing even in the absence of IgG2. Thus, diagnostic screens for ‘inhibitory antibodies’ will need to account for titre, affinity and multiple isotypes binding to P. aeruginosa O-antigen.
MONOCLONAL ANTIBODY THERAPY IN CYSTIC FIBROSIS PATIENTS WITH ASTHMA: A MONASH HEALTH EXPERIENCE

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Background: Monoclonal antibodies have emerged as a novel therapy in asthma however the evidence on their use and benefits in cystic fibrosis (CF) patients with asthma is lacking. Monash Health CF unit has had experience in using monoclonal antibodies in treating CF patients with asthma.

Aim: To assess the changes in exacerbation rate and lung function in cystic fibrosis patients on monoclonal antibodies.

Methods: Patients were identified from monoclonal antibody therapy database in Monash CF Unit. Data were collected from electronic medical records: patient characteristics, exacerbation rate per month (12 months before treatment and up to 12 months after commencement), and lung function (baseline and up to 12 months after commencement). Changes in exacerbation rates and lung function at 3, 6 and 12 months were analysed.

Results: 26 patients received monoclonal antibodies (omalizumab=14, benralizumab=8, mepolizumab=4) with median age 26 years (range 10 to 51) and 15 males (58%). Exacerbation rates were lower at 3, 6 and 12 months compared to baseline (median 0.25/month (0.17-0.33) vs 0 (0-0.33), 0.17 (0-0.33) and 0.08 (0.08-0.25), p<0.05). There were significant improvements at 3 months in FEV1 (median 1.57L (IQR 1.31-1.88) vs 1.65L (1.46-2.15) p=0.01) and FVC (median 2.69L (IQR 2.35-3.56) vs 2.89L (2.52-3.69) p=0.05) but not at 6 or 12 months after commencement.

Conclusion: In CF patients with asthma, monoclonal antibody therapy is associated with a lower exacerbation rate and an improvement in lung function. Further study is needed to confirm these findings as this treatment may improve outcomes in CF patients.
TWO CASES OF ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS IN CYSTIC FIBROSIS SUCCESSFULLY TREATED WITH MEPOLIZUMAB

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Allergic bronchopulmonary aspergillosis (ABPA) is common in cystic fibrosis (CF). (1,2) Treatment is challenging and relapse rate is high. Oral steroids and antifungals are standard therapy for ABPA. However, long term systemic steroid often results in significant side effects. Furthermore, drug interaction between azoles and CFTR modulators is a potential concern. Mepolizumab, an anti-IL-5 monoclonal antibody, can benefit patients with severe eosinophilic asthma (3) and there have been case reports of Mepolizumab use in ABPA in patients with asthma. There is no literature on Mepolizumab treatment for ABPA in CF.

Case Presentation: We present two cases of recurrent ABPA successfully treated with Mepolizumab in adults with CF. Both patients had significant respiratory symptoms and ABPA recurrence, based on CF foundation consensus recommendations (4). Patient one had failed to tolerate Omalizumab and antifungal treatment, patient two was on Itraconazole. Following addition of Mepolizumab, patient one successfully ceased Prednisolone within 2 months (now month 7) and patient two has ceased after 5 months. Subsequent diabetic control improved in both patients. Eosinophils reduced from 1.2 to 0.04 x10^9/L, and 0.27 to 0.04 x10^9/L in patient 1 and 2 respectively. FEV1 increased +5% at 6 months in patient 1. Patient 2 had no FEV1 improvement, however both report significant improvement in chest tightness and reduced their inhaled corticosteroid dosage. Both patients have tolerated Mepolizumab without side effects.

Conclusion: To our knowledge, this is the first case report of successful Mepolizumab treatment for ABPA in CF. Mepolizumab has led to symptomatic and serological improvement in both patients, facilitating more rapid steroid (oral and inhaled) wean. The cost of treatment is $1638/month per patient, and the cost / benefit outcome for longer term use remains unknown. However, Mepolizumab is well tolerated and may serve as an important adjunctive treatment alternative for difficult to control ABPA in CF.
KINETIC EXPRESSION OF GENES AND RECEPTORS DURING MONOCYTE TO MACROPHAGE DIFFERENTIATION IN CYSTIC FIBROSIS

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Introduction/Aim: Previous studies reported cystic fibrosis transmembrane conductance regulator (CFTR)-dependent defective alternative polarization (M2) of macrophages in patients with cystic fibrosis (CF). Later it was shown that defects also reside in the CF monocytes, the precursor cells of macrophages. This study aimed to identify differences in expression of genes, receptors and other regulators associated with monocyte-to-macrophage differentiation between controls, CF and cells treated with the CFTR inhibitor CInh-172.

Methods: Kinetic expression of M1 and M2 genes, miR-155 and miR-146b were analysed by real-time PCR during monocyte to macrophage differentiation. Surface expression of IL-4Rα, IL-13Rα1, CD14, CD68 and TLR4 were analysed by flow cytometry.

Results: Expression of Cox-2 and IRF4 were minimal during monocyte to macrophage differentiation in CF and control cells. Surface expression of IL-4Rα and IL-13Rα1 increased progressively in control cells during macrophage differentiation but not in CF cells. Expression of CD14 decreased gradually whereas CD68 expression increased progressively during macrophage differentiation in all groups. TLR4 had higher expression in CF macrophages compared to control and CInh-172 treated cells. Expression of miR-155 was consistently higher during macrophage differentiation in CF compared to control cells and miR-146b expression was similar in M0 macrophages both in control and CF.

Conclusion: Expression of CD14 and CD68 follows similar pattern in Control, CF and CInh-172 during monocyte to macrophage differentiation. The failure to upregulate surface expression of IL-4Rα and IL-13Rα1 in CF monocytes as they differentiate into macrophages may be explained, in part, by the higher expression of miR-155 during this process. The link between miR-155 expression and lack of CFTR function remains to be explored.

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Declaration of interest: None
DEGREE OF STRUCTURAL LUNG DISEASE IN PAEDIATRIC CYSTIC FIBROSIS PATIENTS WITH AND WITHOUT SUPERIMPOSED ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS

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Background: Allergic Bronchopulmonary Aspergillosis (ABPA) is an important complication in Cystic Fibrosis (CF) that can result in worsening pulmonary disease. The Brody Score is a semi-quantitative analysis that assesses the degree of structural lung disease (SLD) from High Resolution Computer Tomography (HRCT) scans.

Objectives: To quantify the extent of SLD in CF patients diagnosed with ABPA and compare this to PFT changes. To assess longitudinal changes in these parameters in patients who have multiple time points available. To compare these data with CF age-matched controls without ABPA.

Methods: 25 patients from the Royal Children’s Hospital CF clinic, Melbourne who had undergone ABPA treatment and been investigated by at least one HRCT were identified. HRCT scans were assessed for the extent of SLD using a modified Brody Score that assessed bronchiectasis, peri-bronchial thickening and air trapping. FEV1 %predictive results closest to each HRCT were collected. For comparison, the same data were collected for age-matched CF controls with HRCTs performed at similar time intervals.

Results: The mean modified Brody Score of the first HRCT was higher in patients with ABPA compared with CF-only patients, representing a higher degree of SLD (29.3% vs 18.7%). FEV1 was also lower in the ABPA group (78.4% v 90.2%). 13 of the 25 ABPA patients had a second HRCT scan after diagnosis, with the mean time between scans 26 months (ABPA group) and 29 months (CF-only group). Increase in degree of SLD was 13.3% and 3.9%, respectively. FEV1 increased in ABPA group and decreased in CF-only group (85.7% v 82.5%).

Conclusion: Patients in our CF cohort with ABPA had a greater degree of SLD at a cross-sectional time point compared to their peers without ABPA and had an increased rate of progression of SLD over time. The progression of SLD did not correlate with changes in FEV1.
DISTRIBUTION OF STRUCTURAL LUNG DISEASE IN PAEDIATRIC CYSTIC FIBROSIS PATIENTS WITH SUPERIMPOSED ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS

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Background: Allergic Bronchopulmonary Aspergillosis (ABPA) is an important complication in Cystic Fibrosis (CF) that can result in worsening pulmonary disease. The Brody Score is a semi-quantitative analysis that assesses the degree of structural lung disease (SLD) from High Resolution Computer Tomography (HRCT) scans. The distribution of bronchiectasis in ABPA has previously been described as centrally predominant, without propensity to any particular lobe.

Objectives: To identify any lobe predominance of SLD in paediatric CF patients with ABPA, using a modified Brody Score. To quantify the contribution of each component of the modified Brody Score to the total score. To compare the distribution of bronchiectasis and peri-bronchial thickening centrally versus peripherally.

Methods: 25 CF patients from the Royal Children’s Hospital, Melbourne who had undergone ABPA treatment and been investigated by at least one HRCT were identified. HRCT scans were analysed for the extent of SLD using a modified Brody Score that assessed bronchiectasis, peri-bronchial thickening and air trapping. Each lobe was scored separately for the presence and extent of these components, as well as for the distribution of the components centrally or peripherally.

Results: The right upper lobe and right lower lobe had the greatest degree of SLD based on the modified Brody Score (38.6% each). The other lobe scores were left lower lobe (30.6%), left upper lobe (28.8%), right middle lobe (24.8%), and left lingula (15.0%). The greatest contributor to total score was bronchiectasis (41.6%), followed by peri-bronchial thickening (34.8%) and air trapping (25.1%). 78.2% of the bronchiectasis and peri-bronchial thickening was distributed in the central lung, with the remaining 21.8% located peripherally.

Conclusion: There does not appear to be any lobe predominance of SLD in paediatric CF patients with ABPA. Bronchiectasis was the greatest contributor to the quantified SLD. Bronchiectasis and peri-bronchial thickening were more prominent centrally than peripherally.
HIGH INCIDENCE OF NON-TUBERCULOUS MYCOBACTERIA-POSITIVE CULTURES AMONG CHILDREN WITH CYSTIC FIBROSIS IN AUSTRALIA

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Chronic nontuberculous mycobacterial (NTM) infections have rapidly emerged in people with CF, posing a significant threat to their survival. This prospective study aims to determine the prevalence and incidence of NTM in Australia as part of the ongoing National NTM in CF study. A risk based cohort study is underway in 19 CF centres in Australia. Recruits are consenting adult or paediatric individuals with CF who produce a respiratory sample as part of a routine clinical visit (sputum, bronchoalveolar lavage or induced sputum). Samples are collected at baseline, six and 12 months and sent for mycobacterial culture. Nationally 576 adult and 341 paediatric recruits have provided up to 3 sputum samples for mycobacterial culture (September 2016 – Mar 2019). Preliminary findings indicate that in the national cohort of the NTM in CF study, NTM infection is higher in paediatric (12.9%) compared to adult recruits (8.1%). Mycobacterium abscessus group (MABS) infections (57.9%) are the predominant NTM infection in paediatric recruits followed by M. avium complex (MAC) infections (35.1%). In contrast, MAC species are the most commonly isolated NTM species (46.6%) in adults, followed by MABS (39.7%). This pattern of infection varies from state to state within Australia. In the National cohort, NTM positive recruits are significantly younger than the NTM-negative recruits (p< 0.05). Recruits infected with MABS have a significant reduction in mean FEV1% pred (60.7% ± 20.0 SD) when compared to age matched NTM negative recruits (66.6% ± 19.3 SD) (p<0.05). Younger people with CF are more likely to acquire MABS. Participants with MABS infections have reduced lung function compared to age matched peers without NTM infections or those infected by slow growing mycobacterial species. It is not known if poor lung function is a marker for acquisition of MABS or a result of MABS infection.

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ADULT CYSTIC FIBROSIS (CF) PATIENTS PREFER TO PERFORM IMPULSE OSCILLOMETRY (IOS) IN COMPARISON TO SPIROMETRY

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Introduction/Aim: Spirometry is the gold standard method for monitoring lung function in CF. IOS is an alternative diagnostic tool to monitor lung function. The aim of this study is to compare patient preference and assess patient discomfort among adult CF patients in using IOS and spirometry.

Methods: Two prospective studies are being conducted simultaneously to compare the sensitivity of IOS and spirometry in detecting lung function improvements in CF patients commencing on ORKAMBI and receiving treatment for pulmonary exacerbations respectively. Patients were given questionnaires after lung function measurements to assess for patient preferences between the two diagnostic methods. The patients were also asked to rate their level of discomfort for each test. The same questionnaires were used in both of the studies. All patients who have participated in both of these studies to date were included.

Results: Total of 43 patients (n=43) were recruited; 22 patients from CF exacerbation study and 21 from ORKAMBI study. The mean±SD age was 31.6±8.4 years; 32 patients were males (74%). The majority of patients (79%, n=34) preferred performing IOS. Eight (19%) patients preferred spirometry in comparison to IOS while one patient (2%) had no preference.

Ten patients (23%) reported experiencing discomfort while performing spirometry compared to two patients (5%) using IOS. Spirometry had an overall mean rating score of 2.6±2.3 compared to mean rating score of 0.7±1.0 in IOS, which was statistically different (p<0.0001).

Conclusion: Adult CF patients prefer to perform IOS. More patients experienced discomfort when performing spirometry.
CLINICAL FACTORS WHICH INFLUENCE THE RELEASE OF POTENTIALLY INFECTIOUS COUGH AEROSOLS IN PEOPLE WITH CYSTIC FIBROSIS

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Objectives: Our group has investigated the extent of Pseudomonas aeruginosa (Pa) aerosol release during cough in people with CF (Knibbs, Thorax, 2014; Wood, AJRCCM, 2018; Stockwell, AJRCCM, 2018) and we have found that the concentration of Pa cough aerosols is highly variable between patients. The only clinical factor consistently associated with the release of Pa cough aerosols is the concentration of Pa in the sputum. Therefore, we wanted to determine if additional clinical factors are associated with cough aerosol production by using merging the study datasets.

Methods: The merged Pa cough aerosol study datasets were analysed to identify factors associated with cough aerosol production. Participants were categorised based on the amount of Pa cough aerosol release: high producer if count was ≥10 colony forming units (CFU) or low producers if the count was <10 CFU (Jones-Lopez, AJRCCM, 2013).

Results: Fifty-two people with CF and chronic Pa infection performed 68 cough tests. Pa sputum concentration was the only factor associated with Pa aerosol concentration (r=0.61; p<0.001). No associations were seen for gender (p=0.78), FEV1% predicted (r=-0.025; p=0.86), BMI (r=-0.23; p=0.11), age (r=0.22; p=0.13), modulator use (p=0.47), C-reactive protein level (r=-0.24; p=0.87) or other CF respiratory pathogens (p=0.16). The sputum concentration association was also seen in 12 subjects that were recruited to multiple (2-3) cough tests. Over time, the cough aerosol category did not change in 9 subjects (8/9 remained high producer; 1/9 remained low producer) but did change in 3 subjects (2/3 changed from high to low producers; 1/3 changed from low to high producer).

Conclusions: Pa sputum concentration is the most consistent clinical factor which predicts for Pa cough aerosol production. Production of cough aerosol (high/low producers) was unchanged over time for most longitudinal participants.

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POTABLE WATER MAY BE A POSSIBLE ACQUISITION SOURCE OF MYCOBACTERIUM ABSCESSUS GROUP INFECTION IN PEOPLE WITH CYSTIC FIBROSIS

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Introduction: Mycobacterium abscessus group (MABS) are emerging respiratory pathogens in people with cystic fibrosis (CF) and their acquisition and transmission pathways are poorly understood. Up to 25% of MABS strains in people with CF are unique genotypes and may be acquired environmentally (Bryant, Science 2016). Potable water is a known environmental source of MABS (Thomson, J Clin Microbiol. 2013). This study investigates the presence of MABS in potable water of hospitals and homes in Queensland.

Methods: Potable water was sampled from hospitals and homes in Queensland. Hospital: Twelve water points were sampled at The Prince Charles Hospital (TPCH) over five time points (2014-17) and a single time point at the former Royal Children’s Hospital (RCH) (2015). Home: A minimum of three water points were sampled; shower, bathroom sink and kitchen sink taps. The participants were categorised into four groups: 1) CF and active MABS respiratory infection (<2 years since positive culture; n=10), 2) CF and past MABS respiratory infection (>2 years since positive culture; n=7) or 3) CF and never infected with NTM (n=8), and 4) homes of people with no lung disease (n=8). All water samples were incubated for 8 weeks. The genomes of presumptive NTM isolates were sequenced and compared to CF respiratory isolates.

Results: Hospital: The TPCH longitudinal water sampling had a variable MABS detection rate (range 0-25% per time-point). At the RCH, MABS was detected in 2/12 (17%) water points. Home: MABS was detected in 6/136 (4%) water points. MABS genomes from the water and CF respiratory isolates were very closely related MABS at times.

Conclusions: The genetic relatedness of the water and respiratory MABS isolates demonstrates that potable water may be a potential source of MABS respiratory infection in some people with CF.

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A ‘REAL-LIFE’ STUDY ON BACTERIAL AIR CONTAMINATION IN AN ADULT CYSTIC FIBROSIS CLINIC

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Background: Respiratory pathogens from patients with cystic fibrosis (CF) can contaminate clinical environments and increase the risk of health-care related cross-infection. A recent study suggested that viable cough-generated Pseudomonas aeruginosa aerosols from patients with CF travelled further and persisted longer than previously thought. To date, there is a paucity of real-life data to inform clinicians on the optimal between-patient time interval in the CF outpatient clinic to minimise the risk of airborne transmission.

Aims: To assess the prevalence of air bacterial contamination in a tertiary adult CF clinic. The primary endpoint was the rate and duration of air contamination during and after spirometry in a routine CF clinic.

Methods: Continuous air sampling was performed in a CF outpatient clinic room where patients perform spirometry during clinic visits. Active air sampling was performed with the Surface Air System and passive sampling with settle plates. Sputum samples were collected whenever available. Standardised quantitative culture was performed on air and sputum samples. MALDI-TOF was used to identify all bacterial isolates. Fungal colonies were identified based on morphological characteristics.

Results: Air sampling was performed during 10 CF clinics over a 3-month period. A total of 64 clinic visits by 35 patients were captured. Air bacterial contamination (Pseudomonas aeruginosa, Staphylococcus aureus, Achromobacter xylooxidans) was detected during spirometry in 12/64 (19%) clinic visits by 10/35 (29%) patients. Aerosolised viable P. aeruginosa was detectable up to 20 min after spirometry. Background air sampling was done in the same room outside clinic hours. None of these samples were positive for CF-related bacterial pathogens.

Conclusions: Aerosolised viable CF pathogens were detectable in 19% of the clinic visits up to 20 min after spirometry. This data supports the current infection control guideline which recommends at least a 30-min interval between consecutive patients in the CF outpatient clinic.
A LONGITUDINAL STUDY INTO NEUTROPHIL PHENOTYPES AND LUNG DISEASE OUTCOMES IN EARLY CYSTIC FIBROSIS.

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Introduction: We have recently established that a pathological phenotypic shift occurs within polymorphonuclear neutrophils (PMNs) upon recruitment to CF airways, and this can occur in early life. Reprogrammed PMNs display reduced phagocytic receptor (CD16) and hyperexocytosis of protease containing granules (CD63), which is associated with CT scan-based outcomes. Over 3 years, we are characterising the longitudinal phenotypes of airway and blood PMN in young children with CF. We report here our interim progress.

Methods: Bronchoalveolar lavage (BAL) and blood (where possible to allow baseline PMN phenotyping) was obtained from children with CF less than 3 years old, as a sub-study of the AREST CF program. The cellular portion of BAL and blood were stained for flow cytometry with the following markers; viability, CD45 (leukocyte), CD3 (T cell), CD66b (PMN), CD115 (monocyte/macrophage), CD163 (monocyte/macrophage), Siglec-8 (eosinophil), CD16 (PMN phagocytosis), CD63 (PMN exocytosis), CD33 (PMN maturity), and neutrophil elastase (key factor in CF structural lung disease). Samples were acquired using a BD Fortessa cytometer and analysed for phenotypic shift (CD16, CD63 expression) and surface neutrophil elastase expression. AREST CF clinical parameters including CFTR mutation class, age, gender, airway microbiology, and PRAGMA-CF CT score were also collected.

Results: To date, 29 children <3 years old have been recruited to the study, with 16 at 3-months of age. We have assessed BAL from 56 visits, resulting in multiple assessments for 17 participants to date. Matched blood has been assessed for 41 of 56 (73.2%) visits. The 18-month interim analysis of the dataset is schedule for June.

Conclusion: We demonstrate it is possible to longitudinally characterise PMN phenotypes in children with CF under 3 years of age, during a critical period of disease. We are on track for recruitment and aim to complete multi-variate analyses of extent of PMN reprogramming and disease outcomes in 2021.
POLYMICROBIAL INFECTION AND NEUTROPHILIC DISEASE IN CYSTIC FIBROSIS AIRWAYS

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Introduction/Aim: Cystic fibrosis (CF) lung damage is driven by a cycle of infection, pro-inflammatory signalling, and neutrophilic reprogramming. However, the mechanisms behind this process are poorly characterized. We created a multi-component in vitro system to model CF inflammatory responses and airway neutrophil recruitment, specifically in the context of polymicrobial infections. This was used to assess epithelial and neutrophil responses to rhinovirus and Pseudomonas aeruginosa infection.

Methods: Submerged monolayers of primary CF airway epithelial cells (3M:1F; ages≤5yr) were infected individually and in combination with rhinovirus strain RV1b (MOI 0.5) and a mucoid P. aeruginosa clinical isolate (MOI 0.001). After 48 hours, cell culture supernatants were harvested and epithelial secreted cytokines quantified by ELISA. Filtered supernatants were also applied to an in vitro model of neutrophil transmigration to the airways. Migrated neutrophils were harvested 10 hours post stimulation and assessed by flow cytometry.

Results: Both infection with RV1b or RV1b+P. aeruginosa significantly increased production of proinflammatory cytokines IL-8 and IL-1β compared to uninfected controls or bacterial infection alone (p<0.01). Production of CCL5 was significantly increased in viral infections (p<0.03). Biofilms formed upon P. aeruginosa infection, however, more non-aggregated planktonic bacteria were observed with RV1b+P. aeruginosa co-infection. In the transmigration model, neutrophils migrated in similar numbers towards all supernatants. However, neutrophils migrating towards bacterial or co-infection supernatants were more likely to reprogram to a CF specific proinflammatory phenotype (p<0.01) and had reduced CD16 staining (p<0.01), indicating reduced phagocytic ability.

Conclusion: Results highlight the role of respiratory viruses in CF as triggers of airway inflammation and promoters of secondary bacterial infection. Coinfection induced the greatest shifts towards aberrant neutrophil phenotypes, suggesting that polymicrobial infections may be implicated in CF neutrophilic disease. This model permits investigation of CF airway responses to diverse pathogenic insults and the mechanisms that drive neutrophil reprogramming and inflammation.
CaEDTA COMBINED WITH NEBULISED TOBRAMYCIN IMPROVES BACTERIAL CLEARANCE AND LUNG FUNCTION IN CYSTIC FIBROSIS PATIENTS WITH CHRONIC PSEUDOMONAS AERUGINOSA INFECTION

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Background: Iron is crucial for Pseudomonas aeruginosa survival, and elevated iron levels in CF airways contribute to the establishment and persistence of P. aeruginosa lung infection. In vitro and animal studies show that iron removal by chelators improves antimicrobial efficacy, although the utility of this approach in humans for the treatment of P. aeruginosa lung infection is yet to be established.

Objectives: To study the safety and efficacy of combining calcium ethylene diamine tetra-acetate (CaEDTA) with inhaled tobramycin in CF patients with chronic P. aeruginosa infection undergoing treatment for pulmonary exacerbation.

Methods: All patients (n=24) received standard of care IV antibiotic treatment together with nebulised tobramycin 250mg BID for 2 weeks, followed by a further 4 weeks of nebulised tobramycin BID. In addition, patients were randomised to receive 75mg CaEDTA (n=12) or saline (n=12) QID for 2 weeks (while on IV antibiotics) and BID for the following 4 weeks, with a 4-week safety follow-up period subsequently.

Results: Mean P. aeruginosa sputum count (log10 CFU/g) in the CaEDTA vs saline group was reduced by 2.0 vs 0.4 at 2 weeks and by 1.5 vs 0.9 at 6 weeks, respectively. Lung function (ppFEV1) in the CaEDTA vs saline group showed a mean improvement of 16 vs 5 %points at 2 weeks, 11 vs 2 %points at 6 weeks, and 6 vs 2 %points at 10 weeks, respectively. Adverse events were similar in both groups, and none were specifically attributed to the study drug.

Conclusions: In this pilot study in CF patients, adding CaEDTA to nebulised tobramycin was safe, improved sputum clearance of P. aeruginosa and led to greater improvement in lung function compared to patients on inhaled tobramycin alone. The study provides proof of concept for combined use of inhaled CaEDTA and tobramycin in treatment of pulmonary exacerbations and the findings warrant further exploration of the potential of this combination in larger clinical studies.
ALTERED NEUTROPHIL REPROGRAMMING IN CF AIRWAYS MAY BE TRIGGERED BY CF EPITHELIUM DIRECTLY

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Introduction: We have demonstrated polymorphonuclear neutrophils (PMN) are reprogrammed upon migration into CF airways with increased granule release (CD63) and decreased phagocytic receptor expression (CD16), which is recapitulated by CF clinical samples in an in vitro neutrophil migration model. Whether PMN reprogramming is driven by interaction with CF airway tissue directly is unknown. We further adapted the model and tested the hypothesis that migration through CF epithelia or CF fibroblasts can directly induce PMN reprogramming.

Methods: 12-well Alvetex inserts were coated (hydrogel or collagen type I). For primary airway epithelial cells (pAEC), 0.25 x 10^6 cells were seeded (n=3 CF, n=2 non-CF), and cultured at air-liquid interface (ALI) in PneumaCult ALI media (StemCell Technologies) for 28 days. For fibroblasts, 0.5 x 10^6 primary normal human lung (NHLF) or CF human lung (CF-HLF) fibroblasts were seeded and cultured for three days in fibroblast growth media (Lonza). For H441 epithelial cell line, 0.25 x 10^6 cells were seeded and cultured at ALI in growth media (Corning) for 14 days. Once cultures were established, inserts were inverted and 0.25 x 10^6 healthy donor PMN migrated into the apical compartment containing 100 nM leukotriene B4 for 10 hours before assessment by flow cytometry.

Results: Comparing PMN following migration through the existing H441 model, H441 co-cultured with fibroblasts, and fibroblasts alone, we observed no significant difference in CD16 or CD63 expression between culture types or fibroblast phenotypes (Kruskal-Wallis, p>0.05). However, PMN migrated through non-CF pAEC featured lower CD63 expression compared to PMN migrated through CF pAEC (t-test, p<0.05). No difference was observed for CD16.

Conclusions: Migrating PMN may respond differently to CF epithelium than non-CF epithelium. Future work aims to verify the signalling pathways occurring between pAEC and PMN during migration and to incorporate lung fibroblasts for a model more reflective of airway tissue.
ALTERED INNATE IMMUNE RESPONSES OF CYSTIC FIBROSIS AIRWAY
CELLS TO RHINOVIRUS

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Background: Viruses are the predominant respiratory infection in early life and can cause exacerbations in CF. Prior studies of rhinovirus (RV) infection in primary CF airway epithelial cells (pAEC) have largely explored inflammatory and antiviral pathways. Whether virus infection contributes to other airway pathologies in CF such as abnormal mucus accumulation is unknown. To address this question, we compared transcriptional responses to RV infection between non-CF and CF pAEC.

Methods: pAEC cultures from children with CF (n=7) and non-CF controls (n=5) (age; all <5 years) were infected with RV, RNA extracted, and RNA-sequencing performed (Illumina HiSeq2500). Sequence reads were mapped to the human reference genome using HISAT2 and gene counts were performed using SummarizeOverlaps. Differential gene expression (DEG) analysis was performed using DESeq2 to identify the transcriptomic responses of non-CF and CF pAECs to rhinovirus infection.

Results: We identified 877 DEGs in non-CF (613 upregulated and 238 downregulated) and 1379 DEGs in CF (828 upregulated; 525 downregulated) pAECs in response to RV infection, predominantly, relating to interferon and inflammation pathways. A total of 21 unique biological pathways were identified in CF. One non-canonical enriched pathway of interest was O-linked glycosylation of mucin. Specifically, genes encoding n-acetylgalactosaminytransferases and sialyltransferases (GALNT10, GALNT11, ST6GALNAC1, ST6GALNAC2, ST6GALNAC5) were downregulated (1.6-2 fold) while n-acetylgalactosaminytransferase and galactosyltransferases (B3GNT8, B4GALT1, B4GALT5) were upregulated (1.8-1.9 fold). This was accompanied by increased expression of membrane-tethered mucin genes (MUC1, MUC15; 3.2-4.4-fold). We are currently assessing the impact of RV on secretory mucin expression (MUC5AC and MUC5B) in differentiated pAEC cultures.

Conclusion: CF pAEC elicit more complex responses upon virus insult than previously thought. Changes in glycosyltransferases gene expression could affect mucin production and secretion, suggesting a mechanism by which viral infection can affect the airway microenvironment early in life

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CLOSTRIDIUM DIFFICILE INFECTION IN ADULTS WITH CYSTIC FIBROSIS

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Introduction: Clostridium difficile is the most common cause of hospital-associated infectious diarrhoea. Cystic fibrosis (CF) patients have significant antimicrobial and hospital exposures, which predispose them to C. difficile infection (CDI). Asymptomatic carriage of C. difficile is common in some CF centres, however, the epidemiology of CDI in this population is poorly understood. Apart from asymptomatic carriage, C. difficile can cause severe colitis in CF patients.

Aim: To assess C. difficile carriage and infection in adults with CF.

Methods: We performed prospective screening for C. difficile in stool samples collected from asymptomatic (no abdominal symptoms) adults with CF. Stools were cultured on ChromID C. difficile agar, and in-house PCR assays were used to determine the ribotype (RT) and toxin profile of isolates. Stools of symptomatic CDI patients were routinely tested using the BD MAX C. difficile tcdB assay.

Results: Following culture, 32/65 stools (49%) from 22/45 asymptomatic patients (49%) were positive for C. difficile, yielding 33 isolates. Of these, 18 (55%) from 14/45 patients (31%) were toxigenic. Significant toxigenic C. difficile RTs included RTs 046 (4), 012 (3), 014 (2), 020 (2) and 017 (1), and non-toxigenic isolates included RTs 039 (10) and 010 (2). The BD MAX assay was positive for 13/59 stools (22%) from 11/33 symptomatic patients (33%). RT data were available for eight strains, which included RTs 017 and 078.

Discussion: The prevalence of both toxigenic and non-toxigenic C. difficile among asymptomatic patients was high (31/24%, respectively vs. 0-15% in healthy adults) and similar to symptomatic patients (33%). Half the RTs found in symptomatic patients were also present in asymptomatic patients, suggesting a lack of association between RT and clinical presentation. Given the high strain diversity, both groups of patients likely acquired C. difficile from sources outside the hospital and developed CDI following admission. The appearance of RT 017 was unexpected. The significance of these observations warrants further investigation.
CORRELATING THE LUNG MICROBIOME WITH DIETARY INTAKE IN CYSTIC FIBROSIS – ANOTHER WAY TO LOOK AT PHENOTYPIC OUTCOMES?

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Introduction/Aim: Previous literature has recently demonstrated respiratory dysbiosis in CF and has highlighted a correlation between infant breastfeeding and respiratory microbial diversity. This suggests a potential link between dietary intake and respiratory microbiome and a potential gut-lung axis. This study aims to explore the interplay between the respiratory microbiota and both the CF (high-calorie, high-fat) diet and intestinal microbiota.

Methods: This is a preliminary cross-sectional analysis of a prospective, longitudinal, observational cohort study at Sydney Children’s Hospital Randwick. Children with CF and healthy controls (HC) completed a food frequency questionnaire (ACAES), and provided an airway (sputum/oropharyngeal swab) and stool sample. We performed 16s rRNA sequencing (V4 region). Analyses were performed in R (v3.4.4).

Results: 33 CF (16 female (48.5%); median age [IQR] 8.8 [4.6-12.0]) and 27 HC (17 female (63.0%); median age [IQR] 12.6 [10.1-15.1]) subjects were recruited. Alpha diversity (Shannon index) was not significantly different between CF and HC. Beta diversity (relative abundance Bray-Curtis dissimilarities) revealed significant clustering between CF and HC (p=0.009). The relative abundance of Sphingobacteria was significantly lower in CF compared to HC (FDR<0.05). Pearson correlations of CF respiratory (r) and stool (s) phyla revealed strong correlations between Candidate division SR1 (r) and Fusobacteria (s) (R=0.92, p<0.001); Deinococcus-Thermus (r) and Deinococcus-Thermus (s) (R=0.85, p<0.01); Euryarchaeota (r) and Proteobacteria (s) (R= 0.95, p<0.001); and Tenericutes (r) and Fusobacteria (s) (R=0.99, p<0.001). In CF, respiratory Fusobacteria negatively correlated with a number of micronutrient intakes (p<0.05), including: (i) fibre (R=-0.99), (ii) provitamin A (R= 0.95), and (iii) retinol (R=-0.97).

Conclusion: These preliminary baseline data demonstrate clear deviations in the respiratory milieu of a paediatric CF population, and provide support for a potential gut-lung axis. Diet as a potential respiratory microbial modulator warrants further exploration.
PREVALENCE OF MULTIPLE ANTIBIOTIC RESISTANT PSEUDOMONAS IN AN ADULT COHORT WITH CYSTIC FIBROSIS

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**Background:** Increasing prevalence of multi-antibiotic resistant Pseudomonas aeruginosa (MARPA) has been recognised in patients with Cystic fibrosis. Based on the Australian Clonal Pseudomonas study samples collected between 2007-2010, the prevalence of MARPA prevalence in Australian CF centres was 31%. The latest antimicrobial resistance trends in CF P. aeruginosa isolates is unknown.

**Aim:** To assess the latest antibiotic resistance pattern of P. aeruginosa in an adult cohort with cystic fibrosis.

**Method:** Sputum samples were prospectively collected from all patients attending the WA adult cystic fibrosis centre. Five P. aeruginosa isolates were randomly selected for antimicrobial susceptibility testing (AST) by disc diffusion method to 12 commonly used antipseudomonal antibiotics according to CLSI guideline. Clinical parameters of the patient were retrieved from the centre’s patient registry.

**Results:** Four hundred and sixteen P. aeruginosa isolates were collected from 84 adults (3-5 isolates/patient) between 2017-2018. Of 416 isolates tested, 73/416 (18%) were resistant to all beta-lactam antibiotic tested; 227/416 (55%) were resistant to fluoroquinolone antibiotics; 275/416 (66%) were resistant to aminoglycoside. In total, 175/416 (42%) isolates from 58/84 adults (69%) were identified as MARPA. Patients infected with MARPA were older (48 years [SD: 31] vs 42 yrs [SD:29] and have more severe lung function impairment (FEV1: 54% [SD: 16%] vs 62% [20%]; p<0.05) compared with patients without MARPA.

**Conclusion:** A significant proportion of adults attending our centre harboured MARPA infection. Patients infected with MARPA were older and have more severe lung disease than those without MARPA infection. Our study demonstrated a higher prevalence of MARPA (42%) than previously detected in 2007-2010 (31%). This may suggest an ongoing rise in MARPA prevalence in an aging adult cohort with CF due to cumulative antibiotic exposure. Further genotype testing of these isolates are underway to identify potential clonal relationships of MARPA isolates.
STATISTICAL MODEL FOR COMPUTATIONAL DETECTION OF ASPERGILLUS INFECTION

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Aspergillus fungal infection is an important but under-recognized contributor to early cystic fibrosis (CF) lung disease and changes in detection methods are needed to improve patient outcomes. Lung infection by opportunistic pathogens drives irreversible CF lung disease. Although the prevalence of common CF pathogens Staphylococcus aureus and Pseudomonas aeruginosa decreased due to aggressive treatment prevalence of Aspergillus has increased. Unfortunately, culture-based methods in clinical pathology for Aspergillus diagnosis are highly variable. We have developed a statistical model to computationally identify bronchoalveolar lavage samples where Aspergillus infection was missed by the culture-based methods. From published literature we expect 40-60% of samples to have been missed. Consistent with the previous findings, in a pilot study of the 5th year patients, we identified 45 out of 103 patients (43%) as having an Aspergillus infection score greater than 0.5 which we used as a cutoff for decision making. We are currently confirming our model-based findings with qPCR.
IS IMPULSE OSCILLOMETRY (IOS) BETTER THAN SPIROMETRY FOR MONITORING TREATMENT OUTCOMES IN ADULT CYSTIC FIBROSIS (CF) PULMONARY EXACERBATIONS? A PILOT STUDY

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Introduction/Aim: Spirometry is the gold standard for detecting and monitoring lung function decline in CF exacerbation. IOS is a diagnostic tool that detects airway resistance and requires significantly less patient effort to perform than spirometry. The aim of this study is to compare IOS and spirometry in evaluating the effectiveness of intravenous antibiotic treatment for CF exacerbation.

Methods: CF patients commencing intravenous antibiotic treatment for pulmonary exacerbations were prospectively recruited into the study. IOS and spirometry were performed at the commencement of IV antibiotics, weekly throughout treatment and at the cessation of treatment.

Results: Nineteen patients aged 32.9±9.3 years (mean±SD) were recruited into the study to date. Mean pre-exacerbation FEV1 and FVC were 2.62±1.1L (65.6±24.6% predicted) and 4.2±1.3L (82.9±19.4% predicted). Subjects received antibiotics for 16.2±4.7 days. Using linear mixed effects model with repeated measurement over time, there were significant improvement in all spirometry parameters. There was no significant difference in the IOS resistance parameters, but the results showed there was a trend in the decline in resistance values with antibiotic treatment. There was a significant correlation between spirometry and IOS resistance values which demonstrated that as airflow obstruction decreased with the administration of antibiotics, airway resistance also improved. No significant difference was seen in inflammatory markers before and after the administration of treatment.

Conclusion: Spirometry remains the gold standard for monitoring lung function in the CF exacerbation group but the linear mixed effects model demonstrates that IOS may be a useful additional monitoring tool. The trend in the resistance values declining with antibiotic treatment indicates that this may be significant with more patient numbers. This study is still ongoing.
COMPARISON OF SINUS AND LOWER AIRWAY BACTERIAL COLONISATION

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Introduction: In people with Cystic Fibrosis (CF), the paranasal sinuses can be colonised with bacteria including Pseudomonas aeruginosa (PsA) and Staphylococcus aureus (SA). Whilst upper airway suction is used commonly in children, the comparison of upper versus lower airway colonisation has not been performed in adults with CF. This audit compared the cultures of paranasal sinus wash and lower airway sputum samples. In cases where specific sinus treatment was introduced based on the results of the sampling, treatment regimen was also audited.

Method: Paired samples of sinus wash and lower airway were taken from adults with CF. Subjects performed sinus wash using a FloTM Sinus Care Kit reconstituted with sterile water. The results of the sinus wash and paired sputum cultures were retrospectively compared.

Results: Samples were collected from 20 CF adults, 12F, age 26.7±8.6 (mean±SD) years, during a period of clinical stability. In the paired samples, 3 subjects grew normal flora in both samples, and 7 subjects grew the same bacterial species in both samples. Sinus wash cultures grew PsA (10/20), SA (5/20) and Burkholderia cepacia complex (2/20). Sputum samples grew PsA in 10 subjects and SA in 4. Of particular interest were subjects who cultured different bacteria from the sinus wash compared to the lower airway, including 1 subject who repeatedly cultured Burkholderia multivorans from the sinus wash but not the lower airway. Specific sinus treatment was introduced for 4 subjects where it was considered clinically relevant to attempt eradication of sinus pathogen.

Conclusion: This simple, non-invasive sinus wash method can identify pathogens growing in paranasal sinuses and therefore may be a useful clinical tool to direct eradication regimens. Different organisms can be present in the sinuses as compared to the lower airway highlighting the need for regular sinus surveillance.
SEARCH FOR TRANSCRIPTIONAL AND METABOLIC MARKERS OF THE HOST-RHINOVIRUS INTERACTION USING PRIMARY AIRWAY EPITHELIAL CELLS FROM HEALTHY CHILDREN AND THOSE WITH CYSTIC FIBROSIS (CF)

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Introduction/Aim: Acute exacerbation episodes in Cystic Fibrosis (CF) are typically associated with rhinovirus (RV) infection. Responses by CF lung tissue to RV are aberrant. To gain insight into the host-rhinovirus interaction signature after infection, we used both a Whole Transcriptomics Sequencing (WTS) strategy to determine the viral load and host’s mRNA expression along with a metabolic approach to profile the metabolites associated with the viral infection of primary bronchial epithelial cells obtained from healthy children (H) and those with CF.

Methods: WTS and hydrophilic interaction liquid chromatography (HILIC) coupled to mass spectrometry were used to identify the differences between RV coverage and transcripts along with metabolites produced by H (3.9 ± 1.5 years; n=8) and CF (2.6 ± 1.8 years; n=8; all p.Phe508del/p.Phe508del) primary epithelial cells at 24 hours post infection with human rhinovirus 1B. Multivariate analyses were performed to identify potential transcripts and metabolites biomarkers that could discriminate between infected and non-infected cells.

Results: Viral genome coverage was performed to confirm the viral infection and assess viral load. The RV coverage in uninfected cells was less than 0.5X. Infected cells of H and CF children presented 44.4X and 101.6X of RV, respectively. At the transcriptional level, RV infection was associated with genes related to RIG-I-like (IFIH1 and DDX58) and NOD-like (OAS3 and OAS2) receptor signaling pathways. At the metabolomic level, increases in the level of Nicotinamide (Vitamin B3 pathway), Pyridoxal (Vitamin B6 pathway) and Uracil were observed.

Conclusion: We have identified several host-derived transcripts and metabolites associated with RV infection independent of the host’s genetic background. Although functional analysis are required to understand the biological meaning underlying these changes, these candidates may be used as biomarkers of RV infection. Future analysis will help to understand whether their production can be targeted for antiviral purposes.
VARIATION IN TREATMENT PREFERENCES OF PULMONARY EXACERBATIONS AMONG AUSTRALIAN AND NEW ZEALAND CYSTIC FIBROSIS PHYSICIANS

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Background: Pulmonary exacerbations contribute to progressive decline in lung function in cystic fibrosis (CF). Despite advances in CF management and survival, the optimal treatment of exacerbations remains unclear. Understanding the variability in treatment approaches among physicians might help prioritise clinical uncertainties to address through clinical trials.

Methods: Physicians from Australia and New Zealand who care for people with CF were invited to complete a web survey of treatment preferences for pulmonary exacerbations. Six typical clinical scenarios were presented; three to paediatric physicians and another three to adult physicians. For each scenario, physicians were asked to choose from a list of treatment options and to provide reasons for their choices. They were also asked whether and how they would change treatment if there was a poor initial response after seven days.

Results: There were 49 respondents; 31 paediatric and 18 adult CF physicians; more than half reported 10+ years of experience. There was considerable variation in primary antibiotic selection; none was preferred by more than half of respondents in any of the scenarios. For adjunctive antibiotic therapy, respondents consistently preferred IV tobramycin and a third antibiotic was rarely prescribed, except in one adult scenario. Most prescribed hypertonic saline nebulisation and twice-daily chest physiotherapy in most scenarios while dornase alfa prescription was more variable. Most CF physicians (>80%) preferred to change therapy if there was no early response. Adult physicians were more likely (>57%) to prescribe antibiotics based on their professional opinion rather than hospital or department policy, compared to paediatric physicians.

Conclusions: Variation exists among CF physicians in their preferred choice of primary antibiotic and use of dornase alfa. Preferences are driven by professional opinion, possibly reflecting a lack of evidence upon which to base policy recommendations. Evidence from high quality clinical trials is needed to inform physician decision making.
ANTISENSE OLIGONUCLEOTIDE MEDIATED SPLICE MODULATION TO IMPROVE CFTR FUNCTION OF INTRON 9 5T POLYMORPHISM

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Introduction: Over 2000 mutations in the Cystic fibrosis transmembrane conductance regulator (CFTR) gene causes cystic fibrosis (CF) with variable clinical phenotypes. The length of the poly T tract in intron 9 influences exon 10 selection and can manifest as mild or severe disease depending on other CFTR mutations. Manipulation of CFTR pre-RNA splicing using antisense oligonucleotides (AOs) is a potential therapy for selected CF causing mutations. We aim to develop splice modulating AOs to rescue CFTR function in CF patients that carry the shorter 5T polymorphism in intron 9.

Methods: Multiple AOs targeting CFTR intron 9 and the flanking exons; 9 and 11 were designed and initially optimised using 2’-O-Methyl modified bases on a phosphorothioate backbone (2OMe) and transfected into primary airway epithelial cells from a child with p.508del/Arg117His;5T CF. After 48 hours RNA was collected, and PCR was used to determine the ratio of altered transcript compared to full-length product. CFTR protein size was determined by western blot analysis. CFTR functional outcomes were measured using Ussing chamber utilising Air-Liquid Interface primary cell cultures.

Results: Of the 32 2OMe AOs tested for exon 10 inclusion, none reduced the intron 9 5T induced exon 10 skipping. Of the 8 AOs designed to skip exon 9, the highest efficiency was 24% from both the p.Phe508del allele and intron 9 5T allele. Of the 6 AOs designed to skip Exon 11, the highest efficiency was 22% from the intron 9 5T allele. CFTR protein size was determine on western blot and CFTR function was determined by response to Forskolin (Isc).

Conclusion: We propose that skipping the exons flanking exon 10 (9 and/or 11) on the CFTR 5T allele could improve CFTR function in CF patients carrying selected mutations, either alone or in combination with current therapeutics.

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THE UNIQUE MICROBIOMES OF THE DUODENUM, COLON AND LUNG IN THE G551D MOUSE MODEL OF CYSTIC FIBROSIS

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**Background:** An abnormal lung microbiome is a characteristic feature of the genetic disorder cystic fibrosis (CF). The interplay between the gut and lung microbiomes is assuming importance in CF as the relevance of a healthy gut microbiome to maintenance of systemic metabolic functions and host immunity becomes increasingly apparent. Murine models offer the opportunity to study the gut and lung microbiomes in CF in the absence of antibiotic pressures that confound studies in human patients.

**Methods:** We performed a phylogenetic assessment based on 16S rDNA sequencing of the microbiota of the lung, duodenum and colon in nine-week old CF mice (CftrG551D/G551D) and wild type littermates maintained in identical environments and fed the same diets.

**Results:** The CF mice demonstrated significantly reduced diversity of the colon microbiota and trends towards reduced diversity in the duodenum in the CF compared to WT mice. In the lungs, there were significant differences between CF and WT mice in the relative abundances of Alkalibacterium, Lactobacilli, Ralstonia and Microbacterium, (p<0.05). There were sex-dependent differences in the relative abundance of individual genera in the lungs in the CF mice and overall community composition assessed by principal component, redundancy and canonical correspondence analyses.

**Discussion:** Alterations in the gut and lung microbiota may be inherent features of CF that are present from a very early stage in disease pathogenesis.
PHENOTYPE CHARACTERISATION OF PHE508DEL AND CFTR KNOCKOUT RATS: AN UPDATE

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Introduction: Over the last two years we have generated and bred cystic fibrosis (CF) rats with Phe508del and CFTR knockout (KO) genotypes. Our primary purpose for developing the CF rat model is for pre-clinical development of airway-directed gene therapy. The aim of this study was to provide a phenotype characterisation update for both the Phe508del and KO CF rat models.

Methods: Functional measures including nasal potential difference and pulmonary function testing were used to assess defects in both the upper and lower airways. RNAscope®️ in situ hybridisation and quantitative PCR were used to determine the CFTR mRNA expression levels in the lungs. A range of histological analyses were also performed to detect pathologies in organs including the gastrointestinal tract, pancreas and testes.

Results: Both models recapitulate CF-related pathologies in a range of organs, however Phe508del rats appear to show milder CF phenotypes. KO rats demonstrate a higher mortality rate compared to Phe508del rats due to intestinal obstructions. In the airways of both models, electrophysiological defects are present and mRNA CFTR expression in the lungs are substantially reduced when compared to wild-type animals. Neither model demonstrates the overt lung disease that is typically seen in humans. Multi-focal exocrine pancreatic degeneration is observed in a proportion of KO rats. Moreover, the colon exhibits excess mucus production in both genotypes. The male reproductive tract is severely malformed in Phe508del and KO rats with degeneration of the vas deferens. Spermatogenesis is also significantly impaired in KO rats.

Conclusions: Our Australian-developed Phe508del and KO rat models demonstrate CF-like pathologies in a range of organs including the airways, pancreas, colon and male reproductive tract. While an overt lung phenotype is not observed in “pathogen-free” rats, bacterial challenge studies with Pseudomonas aeruginosa are currently underway to induce chronic infection and subsequent lung disease.
ANTIOXIDANT THERAPY WITH Γ-GLUTAMYLCYSTEINE (GGC) A POTENTIAL NOVEL THERAPY IN CYSTIC FIBROSIS

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Background: CF is characterised by deficiencies in both intra- and extracellular glutathione (GSH) levels. Lipopolysaccharide (LPS) from Pseudomonas aeruginosa is a major virulence factor involved in lung disease progression in CF. The GSH depletion is exacerbated by LPS which further contributes to cellular oxidative stress and the inadequate control of inflammatory pathways in CF patients. Therapeutic strategies to increase GSH using thiols GSH and the cysteine pro-drug NAC have not demonstrated relevant improvements in CF clinical outcomes.

Objective: We have previously shown that the immediate pre-cursor to glutathione, γ-glutamylcysteine (GGC) is effective in increasing intracellular levels of GSH in vivo in healthy humans. In this study, we will ascertain GGC potential as a novel therapy in CF.

Methods: Airway cells isolated from CF and healthy children were used to create a primary epithelial cell model for LPS induced oxidative stress. GGC, GSH and NAC were tested as therapeutic or prophylactic treatments in LPS free cell models and in cells challenged with LPS. Parameters including cell viability, cell proteome, stress granule, tight junctions and production of GSH and reactive oxygen species (ROS) were assayed.

Results: Therapeutic or prophylactic GGC treatment ameliorated the loss in cell viability of the LPS challenged cells and ameliorated the effects of LPS challenge on the proteome and canonical pathways. These pathways included those associated with the inflammatory response, oxidative stress, tight junctions, mitochondrial dysfunctional and metabolism. Therapeutic treatment of LPS challenged cells with GGC increased intracellular GSH levels, while suppressing oxidative stress related cellular responses through lowering of both ROS generation and the resolution of stress granules. Prophylactic treatment of LPS challenged cells with GGC was shown to provide effective oxidative stress protection.

Conclusion: We suggest GGC supplementation may have potential as both a prophylactic and therapeutic treatment to mitigate the incidence and severity of the symptoms suffered by CF patients during P. aeruginosa infection.
POTENTIATOR (IVACAFTOR AND GLPG1837) THERAPY IMPROVES FUNCTION FROM A RARE CFTR MUTATION R352Q IN PATIENT DERIVED AIRWAY CELL MODEL AND INTESTINAL ORGANOIDS

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Background: CFTR is a chloride ion channel present on the apical membrane of epithelial cells. Over 2000 mutations in the CFTR gene have been identified, of which ~300 disrupt normal function. R352Q is a rare missense mutation located in transmembrane helix 6 within transmembrane domain 1, and is proposed to disrupt channel gating and conductance. In vitro studies have shown that R352Q is responsive to potentiator therapy (VX-770-Kalydeco®) in an immortalised non-polarised heterologous expression system1. Validation of this observation in patient-derived primary cell models is lacking.

Objective: To validate the efficacy of the clinically available potentiator, Ivacaftor (VX-770), and two other potentiators (GLPG1837 and Genistein), in restoring R352Q-CFTR function in patient-derived rectal and nasal cell models.

Methods: We created matched rectal organoids2 and conditionally reprogrammed human nasal epithelial (CRC-HNE) cell models3 from a patient with R352Q/F508del CFTR genotype. CFTR-mediated chloride secretion was assessed in rectal organoids by a Forskolin-induced swelling (FIS) assay. CRC-HNE cells were differentiated at air-liquid interface and used to validate the organoid FIS data via an ion transport assay (Physiologic Instruments, CA).

Results: In rectal organoids, stimulation by CFTR potentiators VX-770 and GLPG1837 exhibited significant swelling compared to the vehicle condition (vehicle vs VX-770, P<0.0001 and vehicle vs GLPG1837, P<0.0001). Treatment with the corrector VX-809 displayed an increase in organoid swelling with and without potentiator treatment. Stimulation with potentiator in differentiated HNE cells validated the organoid FIS data.

Conclusion: Data from rectal organoids and CRC-HNE cells evidenced higher residual CFTR function compared to F508del homozygous organoids and wildtype-HNE controls, respectively. Concordant results were observed in CFTR correction with modulators obtained from matched differentiated-HNE cells. Our findings indicate that CFTR potentiators are efficacious in achieving CFTR functional restoration in patient-derived primary cell models with the R352Q mutation.

References:
1) Van Goor et al, 2014
2) Dekkers et al, 2016
3) Martinovich et al, 2017
NOVEL CFTR MODULATOR COMBINATIONS: EVALUATION OF THE CORRECTOR PTI-801, POTENTIATOR PTI-808, AND AMPLIFIER PTI-428 IN CF SUBJECTS

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Currently-approved CFTR modulator therapies have variable clinical efficacy leaving room for additional clinical benefit. PTI-801 and PTI-808 represent a novel CFTR corrector and potentiator, respectively. PTI-428, a CFTR amplifier, selectively increases the amount of immature CFTR protein and provides additional substrate for correctors and potentiators to act upon. In vitro, in human bronchial epithelial cells from F508del homozygous donors, the combinations of PTI-801+PTI-808 and PTI-801+PTI-808+PTI-428 increased CFTR chloride transport activity by 193% and 369% compared to that of tezacaftor+ivacaftor, respectively, suggesting a superior in vitro response to a currently approved modulator combination.

To determine safety, tolerability, PK and effect on FEV1 and sweat chloride of co-administration of the novel CFTR modulators PTI-801, PTI-808 and PTI-428, dose combinations of PTI-801+PTI-808 and PTI-801+PTI-808+PTI-428 are being evaluated in randomized, double blinded, placebo-controlled clinical studies in subjects with CF, age ≥18 years, with FEV1 40-90% of predicted.

Initial data suggest a generally well-tolerated safety profile and clinical benefit with both the doublet combination PTI-801+PTI-808 and the triplet combination PTI-801+PTI-808+PTI-428. In the preliminary analysis, absolute changes in percent predicted FEV1 of 6.6 and 8 percentage points versus placebo were observed following a treatment period of 14 days for doublet and triplet, respectively. Reductions in sweat chloride of -13 mM and -24 mM versus placebo were observed at day 14, for doublet and triplet, respectively.
CHANGE IN LOW-DOSE CHEST COMPUTED TOMOGRAPHY (CT) SCORES AFTER 72 WEEKS OF TEZACAFTOR/IVACAFTOR (TEZ/IVA) IN PATIENTS WITH CYSTIC FIBROSIS (CF) AND ppFEV₁ ≥70%: AN EXPLORATORY PHASE 2 STUDY


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Objective: To explore TEZ/IVA effects on CF-CT scores at week 72 in homozygous F508del patients.

Methods: Patients aged ≥12 years with percent predicted FEV₁ (ppFEV₁) ≥70% at screening were randomised to TEZ 100 mg qd/IVA 150 mg q12h or placebo for 72 weeks. Primary endpoint: absolute change from baseline in total CF-CT score at week 72; secondary endpoint: TEZ/IVA safety over 72 weeks; other endpoints: CF-CT subscore changes, absolute change in ppFEV₁ at week 72. Sample size was not based on a power analysis.

Results: Twenty patients received ≥1 TEZ/IVA dose, and 21 placebo. At baseline, mean (SD) ppFEV₁ was 91.4 (16.0) for TEZ/IVA and 86.6 (12.7) for placebo; total CF-CT scores were similar between groups (38.3 [22.9] and 43.7 [34.0], respectively). TEZ/IVA numerically improved mean change from baseline in total CF-CT score at week 72 [mean (95% CI) difference vs placebo: −1.5 (−7.5, 4.5)]. Serious adverse events (AEs): TEZ/IVA, 8 patients (40.0%); placebo, 13 (61.9%). No patient on TEZ/IVA withdrew because of an AE. There were no new safety concerns. Mean (SD) change from baseline in ppFEV₁ at week 72 was 1.2 (8.4) percentage points with TEZ/IVA vs −3.6 (9.0) with placebo. TEZ/IVA numerically lowered the mean change in bronchiectasis, hyperinflation and mucus plugging CF-CT subscores at week 72: mean (95% CI) differences vs placebo were −0.6 (−3.1, 2.0), −0.5 (−3.1, 2.0) and −0.8 (−2.1, 0.6), respectively. Mean (95% CI) differences vs placebo in peribronchial thickening and parenchymal opacity subscores at week 72 were 0.2 (−1.9, 2.3) and 0.2 (−1.0, 1.3), respectively.

Conclusion: In this exploratory study, TEZ/IVA numerically improved CT scores and maintained ppFEV₁ over 72 weeks in homozygous F508del patients with ppFEV₁ ≥70%, suggesting that TEZ/IVA may reduce structural lung disease progression. TEZ/IVA was generally well tolerated, consistent with its established safety profile.

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GENERATING A PRIMARY CELL CULTURE MODEL WITH IMPROVED SENSITIVITY FOR HIGH THROUGHPUT SCREENING ASSAY


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Rationale: A recent advance in CF therapy is the discovery and approval of small molecules to correct or modulate CFTR production and function. The clinical responses can vary between individuals, suggesting the need for personalised therapy. Intestinal organoids are one promising personalised in vitro model but may be less palatable in children than nasal brushing. However, low endogenous expression of CFTR by basal AECs limits potential for high throughput CFTR assays in this culture setting. To address this, we are optimising a CRISPR/dCas9 method to stably increase CFTR expression in AEC to improve the sensitivity of personalised, monolayer-based assays.

Method: Airway cells were obtained from healthy children and those with CF (≤6 years) by cytology brush and conditionally reprogramed primary cell cultured established. Cells were then stably transduced using retroviral vectors to express yellow fluorescence protein (YFP), and lentiviral vectors to express dCas9 protein and sgRNAs targeting the CFTR gene. Cells stably expressing all three proteins (YFP, blue fluorescence protein BFP and mCherry) were selected via sorting. CFTR expression was then confirmed at gene and protein level.

Results: Conditionally reprogrammed AECs retained their proliferative capacity after stable transduction for YFP expression. However, minimal signal was observed for both healthy and CF AEC in the monolayer CFTR assay. We have created four sgRNAs targeting the CFTR gene and produced lentiviral vectors for these sgRNAs and dCas9 protein. We are currently characterising cells transduced with the CRISPR/dCas9 system, including CFTR expression.

Summary: This study is demonstrating a feasible primary cell model from children with CF that can be grown, expanded in culture and subsequently used as a personalised high throughput screening assay of CFTR. We will be using cultures in the future to assess CFTR correction using known molecules ivacaftor and lumacaftor as controls and potentially other new small molecules.
CLINICAL PREDICTORS AT INFANCY MAY BE BETTER AT PREDICTING SCHOOL-AGED LUNG DISEASE COMPARED WITH INFANT MULTIPLE BREATH WASHOUT

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Introduction: Studies show that a significant proportion of infants with cystic fibrosis (CF) have diminished lung function which tracks into school-age. In this study we aimed to investigate whether MBW outcomes measured in infants diagnosed following newborn screening are associated with worse lung function and structural lung disease at school-age.

Methods: Infant MBW was collected in 65 individuals with CF (0-2 years). Clinical outcomes such as infections, respiratory hospitalizations and symptoms were also available in these participants. At school age (7–10 years), children returned for MBW and spirometry testing, and chest computed tomography (CT) measurements. We determined if lung clearance index (LCI) from the MBW test at infancy predicted FEV1, FVC and FEV1/FVC measured by spirometry, LCI at school-age, and chest CT outcomes.

Results: Using a simple mixed effects model, a 1 unit increase of mean infancy LCI measurements was associated with an increased school-aged LCI by 1.21 units (p=0.030; 95% CI: 0.15-2.12). However, when full models that took into account other known clinical predictors such as CF genotype, pancreatic insufficiency status, respiratory hospitalizations and infection status were used, associations were no longer seen for infant LCI with school-aged LCI (coefficient 0.092; p=0.59). Having a delta F508 heterozygous genotype, pancreatic insufficiency or infection with Haemophilus influenzae or Staphylococcus aureus was significantly associated with both an increase in LCI and percentage of bronchiectasis and total structural lung disease at school-age.

Conclusion: LCI measured in infancy was associated with LCI at school-age. However other clinical factors such as CF genotype, infections with proinflammatory pathogens and pancreatic insufficiency status confounded its ability to predict worse lung disease at school-age.
CHARACTERISING THE DYNAMICS OF TRAPPED AIR ON CT IN YOUNG CHILDREN WITH CYSTIC FIBROSIS

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Background: Trapped air on chest computed tomography (CT) reflects small airways disease and has the potential to be used as an early measure of lung disease in cystic fibrosis (CF). However, little is known about the dynamics of trapped air in young children with mild disease. We hypothesised that persistent trapped air is common and increases with age, and that trapped air is likely to become permanent once it remains stable over two years.

Methods: Children in the Australian Respiratory Early Surveillance Team for CF (AREST CF) birth cohort contributed CT scans from consecutive annual visits. We analysed expiratory CT scans acquired one year apart to determine the proportion of trapped air that remains topologically stable in a two-year time period. CT scans were matched using deformable image registration to evaluate how individual voxels of trapped air evolve over time. We measured the lung volume proportion of stable (TAstable), new (TAnew) and reversed (TAreversed) trapped air. Mixed effects model analysis was used to determine the association between trapped air and age.

Results: We analysed 56 CT triplicates (155 scans) from 45 CF patients with mean (SD) age of 4.48 (1.26) years. TAstable was present in all children with CF but proportions varied between patients [Mean (SD): 9.88 (12.47)%]. Log-transformed TAstable was not associated with age [0.07(-0.06,0.19), p=0.318]. Trapped air that remains topographically stable over two years [Mean (SD) TAstable: 57.11 (20.87)%] was more likely to remain present in the following visit than dynamic trapped air [Mean (SD) TAnew: 35.84 (17.26)%] (p<0.0001).

Conclusion: We detected stable and dynamic “phenotypes” of trapped air on CT which may represent permanent or potentially reversible airway abnormalities, respectively. Further investigation is needed to understand the underlying mechanisms of these trapped air “phenotypes” and improve our understanding of early CF pathobiology.
Background: Multiple breath inert gas washout (MBW) aims to assess lung function during normal tidal breathing, but 100% oxygen (O2) exposure changes breathing pattern during Nitrogen (N2) MBW in infants. SF6 MBW was recommended for infancy. The persistence of this effect and the effect of aging is unclear but important to determine to evaluate N2MBW suitability for older age groups. We studied the magnitude of effect during the preschool age range (defined internationally as 2-6 years of age) in Cystic Fibrosis subjects.

Methods: Clinically indicated tests in subjects ≥2 years old, experienced enough to provide a stable prephase breathing period of ≥30s (ideally 60s) were analysed. MBW was performed with validated commercial N2MBW equipment (Exhalyzer D, ECO Medics AG, Switzerland) according to ATS preschool technical standards. Facemask interface was used to optimise breathing stability. Outcomes were assessed on transition from prephase (room air breathing, 21% O2) to washout phase (100% O2 exposure) included change in tidal volume (VT), minute ventilation (VE), end tidal carbon dioxide (etCO2), respiratory rate (RR) and inspiratory drive (VTin/Tin). Results were compared to previously published infant data.

Results: 41 MBW test visits across CF children aged 2-5 years were analysed. Magnitude of change in breathing pattern was less pronounced in preschool subjects, than previously reported in infants (VT change -32.9%, VE change -32.6%, VTin/Tin change -27.8%). Across the preschool age range, the magnitude of effect with 100% oxygen exposure decreased with age between age 2 and 5 years for VT (-19.6% to -13.8%), VE (-28.0% to -20.9%), etCO2 (+7.2% to +4.5%) and VTin/Tin (-22.4% to -19.5%). In contrast to infants, no visible change in end-expiratory lung volume occurred in preschool subjects.

Conclusions: Detectable differences in breathing pattern with 100% O2 exposure persist into the preschool age range but appear to decrease with age.
PHYSIOTHERAPY
ABSTRACTS
VARIABLE AGREEMENT BETWEEN WEARABLE HEART RATE MONITORS AND 3-LEAD ELECTROCARDIOGRAM DURING EXERCISE IN ADULTS WITH CYSTIC FIBROSIS

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In people with cystic fibrosis (CF), greater cardiopulmonary fitness is associated with increased survival and improved quality of life. Wearable activity monitors are a popular method of monitoring exercise. Heart rate (HR) measurements may be used to indicate exercise intensity and monitor exercise. We assessed the agreement of HR recordings of the Fitbit Charge HR™, Polar®️ H7 HR sensor and Masimo SET®️ Rad-5v pulse oximeter with the 3-lead electrocardiogram (ECG) during continuous and interval exercise.

Adults with CF admitted with an exacerbation completed 15 minutes of exercise on a cycle ergometer on two occasions whilst wearing the previously-mentioned devices. Firstly, participants cycled at 30% of estimated peak workload (Wpeak). Secondly, participants cycled at 1 minute intervals of 60% of Wpeak interspersed with 2 minutes of ‘rest’. HR readings on all devices were recorded at minute intervals. The agreement of HR recordings were analysed using the Bland Altman method.

The Polar® H7 HR sensor had the best agreement with 3-lead ECG, with a bias of 0.10 ± 1.17 bpm and 0.13 ± 0.93 bpm during continuous and interval exercise, respectively. The Masimo SET® Rad-5v pulse oximeter also had good agreement, with a bias of 1.00 ± 7.21 bpm and 0.92 ± 4.68 bpm, respectively. The Fitbit Charge HR™ demonstrated poor agreement, with a bias of 9.31 ± 17.30 bpm and 4.73 ± 12.81 bpm during continuous and interval exercise, respectively.

The Fitbit Charge HR™ is not recommended for assessing HR during exercise in adults with CF. Findings support the use of the Polar® H7 for accurate HR monitoring.
PHYSIOTHERAPY: AT WHAT COST? PARENTAL EXPERIENCE OF PHYSIOTHERAPY FOR INFANTS WITH CYSTIC FIBROSIS

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Background: Physiotherapy is reported to be one of the most burdensome aspects of cystic fibrosis (CF) care. Commonly parents are responsible for performing physiotherapy for their infant child from diagnosis. In older age groups, parental experience of CF care and quality of life impact the health, quality of life and adherence to physiotherapy of their child. It is unknown how parents of infants experience physiotherapy as part of CF care.

Objectives: The objective of this study is to explore the physiotherapy experience of parents of infants with CF.

Methods: Semi-structured, open-ended interviews with parents of infants (aged 0-2yrs) with CF were conducted by a physiotherapist researcher. Interviews were audio recorded, transcribed and coded into domains through a cyclical process of interpretation and analysis, guided by the principles of philosophical hermeneutics.

Results: Ten mothers and three fathers were interviewed, with four of the mothers being full time caregivers. The infants with CF had a mean (standard deviation (SD)) age of 20 (3) months, and parent participant ages ranged from 27 to 44 years. Physiotherapy is an ever-present experience of pressure, doubt, and guilt. Physiotherapy challenges participants’ perception of identity and is a source of conflict. Managing the expectations of physiotherapy comes at a cost to time and relationships, with these sacrifices perceived as an expected and necessary part of meeting the physiotherapy needs of their child. Despite the perceived burden and sacrifices, physiotherapy is experienced by participants as an opportunity to positively impact the health of their child and is a significant priority.

Conclusions: Sacrifice, guilt and opportunity are all experiences that parents of infants with CF perceive in relation to physiotherapy. Clinician awareness of the perceptions and experiences of physiotherapy for parents of infants with CF may help enhance the personalisation of physiotherapy prescription and minimise burden.
THE POSITIVE EXPIRATORY PRESSURE THERAPY RECORDER FOR ASSESSING COMPLIANCE (PEPtrac): VALIDITY AND CLINICAL FEASIBILITY

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Background: Previous studies have investigated the pressure and oscillation properties generated by commonly utilised positive pressure airway clearance technique devices (ppACT) in the laboratory setting, however the properties of these devices when used unsupervised by patients has not been reported. We investigated the properties of the PariPEP S®, Acapella DH® and Aerobika® when used unsupervised by people with CF.

Method: Adults with CF were recruited at the Royal Adelaide Hospital to use the PariPEP S®, Acapella DH® or the Aerobika®. After a single supervised session, participants used the ppACT device unsupervised ≥1/day for one week. Properties were measured and stored using an electronic device (PEPtrac).

Results: Eighteen participants (M:F = 10:8, age 30 [6] yrs) were recruited. There were 32, 38 and 40 sessions recorded for the PariPEP S®, Acapella DH® and Aerobika® respectively. Mean (SD) expiratory duration was 4.8 (1.2), 3.7 (0.8) and 2.9 (1.1) sec for the PariPEP S®, Acapella DH® and Aerobika® respectively, with a mean expiratory pressure of 13.0 (4.0), 13.6 (3.6) and 11.0 (3.2) cmH2O, with significant differences between devices for duration and pressure (p < 0.001). The mean (SD) oscillation frequency for the Acapella DH® and Aerobika® was 18.7 (3.4) and 16.5 (3.0) Hz, with an oscillation amplitude of 5.3 (1.5) and 6.4 (1.7) cmH2O (p < 0.001 between devices). There was significant variability between participants within each ppACTs device for mean expiratory duration and pressure, and oscillation frequency and amplitude for the two oscillatory ppACTs (p < 0.001).

Conclusions: There were significant differences in pressure and oscillation properties between the PariPEP S®, Acapella DH® and Aerobika® devices, and significant between-user variability. Larger studies are required to confirm these findings and to assess whether this variation affects clinical outcomes.

Acknowledgments: This work was supported by an Australian Government Research Training Program Scholarship
PRESSURE AND OSCILLATION PROPERTIES GENERATED BY COMMONLY USED AIRWAY CLEARANCE DEVICES IN ADULTS WITH CYSTIC FIBROSIS DURING UNSUPERVISED USE.

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1 Royal Adelaide Hospital, 2 La Trobe University, 3 Adelaide University, 4 Central Adelaide Local Health Network, 5 Alfred Health

Background: Previous studies have investigated the pressure and oscillation properties generated by commonly utilised positive pressure airway clearance technique devices (ppACT) in the laboratory setting, however the properties of these devices when used unsupervised by patients has not been reported. We investigated the properties of the PariPEP S®, Acapella DH® and Aerobika® when used unsupervised by people with CF.

Method: Adults with CF were recruited at the Royal Adelaide Hospital to use the PariPEP S®, Acapella DH® or the Aerobika®️. After a single supervised session, participants used the ppACT device unsupervised ≥1/day for one week. Properties were measured and stored using an electronic device (PEPtrac).

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Conclusions: There were significant differences in pressure and oscillation properties between the PariPEP S®, Acapella DH® and Aerobika® devices, and significant between-user variability. Larger studies are required to confirm these findings and to assess whether this variation affects clinical outcomes.

Acknowledgments: This work was supported by an Australian Government Research Training Program Scholarship
A STATIC STRETCHING PROGRAM CAN INCREASE MUSCLE LENGTH AND RANGE OF MOTION IN ADULTS WITH CYSTIC FIBROSIS ADMITTED WITH AN EXACERBATION.

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Background: Adults with cystic fibrosis (CF) admitted to hospital with acute exacerbations spend increased time in sedentary positions. This can lead to increased muscle tightness and decreased range of motion in muscle groups. A daily static stretching program was developed for adults admitted to Sir Charles Gairdner Hospital with exacerbations of CF, and we investigated whether completion of this program daily improved muscle length and range of motion (ROM).

Research Question: Does implementing a daily static stretching program increase the ROM and/or muscle length for adults admitted with an exacerbation of CF?

Methods: Participants were randomised to the intervention or control group. The intervention group completed a prescribed set of three static stretches (per a handout) targeting hamstrings, calves and thoracic extensors, under physiotherapist supervision daily during their inpatient stay. The control group were given the same handout and asked to complete the stretches in their own time, without supervision. Outcomes were assessed on day one and on day of discharge, including hip ROM, 90/90 hamstring length, ankle dorsiflexion ROM, knee to walk calf length and thoracic extension length.

Results: Eleven participants (5 female, aged [mean ± SD] 27 ± 8.4, FEV1 % pred. 60 ± 20) completed the study. Five (45%) were allocated to the intervention group. There were significant improvements in right hamstring length (7cm vs. 1.5cm, p < 0.01, 95% CI 2 – 9), ROM (9o vs. 1o, p = 0.01, 95% CI 2 – 13) and thoracic spine extension (1.4cm vs. 0.4cm, p = 0.045 95% CI 0 – 2) in the intervention group compared with control. There was no significant improvement in calf length or ROM.

Conclusion: The implementation of daily static stretching for adults admitted with an exacerbation of CF can improve muscle length and ROM in the hamstrings and thoracic extension during an inpatient stay.
FEASIBILITY AND EFFICACY OF THE INNOSPIRE GO NEBULISER IN CHILDREN WITH CF - PATIENT, PARENT AND CARER REPORTED OUTCOMES

JEN CORDA
Royal Children’s Hospital

In September 2018 the Philips InnoSpire Go mesh nebuliser became available in Australia. Those patients and their families who purchased the new nebuliser were asked to complete a survey relating to the nebulisers feasibility and efficacy as well as the families overall satisfaction with the machine. At the time of this abstract submission a total of 11 surveys have been returned detailing families opinions. Preliminary results show that families feel that the device is:
- As effective or more effective
- As easy or easier to use
Compared to their previous nebuliser.
They all have reported that they are likely to use the machine on an ongoing basis.
Further collection of surveys will occur between now and the conference date, with full evaluation of results presented.
AN AUDIT OF THE INTRODUCTION OF THE AEROBIKA FOR AIRWAY CLEARANCE THERAPY (ACT): CLINICAL EXPERIENCE AND PATIENT REPORTED OUTCOMES IN ADULTS WITH CYSTIC FIBROSIS

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The Aerobika was introduced in Australia in 2016 as an alternative oscillating positive expiratory pressure (OscPEP).

**Aims:** To evaluate the physiotherapy practice and patient reported outcomes (PRO) of the Aerobika for regular ACT over two years in adults with CF.

**Methods:** Patients were provided with an Aerobika and educated about its use and cleaning. Resistance settings and dosage were individualised. Patients recorded their experiences on visual analogue scales (VAS) with anchors from -5 (less effective) to 0 (no difference) to +5 (more effective).

**Results:** Eighty patients (47 female) trialled the Aerobika for regular ACT over the past two years. Data presented as mean (SD), range. Age: 34.3(10.5) 19-67 years; FEV1 51.8(18.4) 23-96% predicted; FVC 70.0(15.7) 46-113%; BMI 21.4(2.5) 17.7-28.5). PRO were completed after use for at least 3 months. Usual ACT devices prior to Aerobika: PEP 44/80=55%; mask 39%; mouthpiece 61%; OscPEP 41/80=51%; Flutter 78%; Acapella 22%. Mucolytics 46/80=58%; hypertonic saline 6%=65%; 3%=20%; 0.9%=15%. All used AD or huffing for sputum expectoration. VAS scales: Resistance setting: +1(most)=46%; mid setting=29%; -5 (least)=25%. Effectiveness compared with usual ACT +2.3(2.1)-5 to +5; Sputum volume cleared: +1.8(2.1)-5 to +5; How clear/free breathing: 2.1(1.8)-5 to +5; Effort +2.1(2.0)-2 to +5; Tiring: 1.8(1.9)-3 to +5. Time: +1.6(2.4)-5 to +5; Easy to use: 4.1(1.6) -2 to +5; Use for regular ACT: 3.8(2.2) -5 to +5; Adherence increase: 1.3(3.4) -5 to +5; Did not like Aerobika 8/80 (found PEP more effective). Combination with mucolytics: 71/80, HS 6% = 46%; 3% = 17%; 0.9% = 37%. Nebuliser: AeroEclipse 51%; AeronebGo 46%; EFlow 3%. BPA free plastic is durable.

**Conclusions:** Patients were enthusiastic about trialling the Aerobika. They found it easy to use and at least as useful for sputum clearance as usual ACT. They particularly valued combining it with mucolytics to save time while achieving effective airway clearance therapy.
AN AUDIT OF THE CLINICAL EXPERIENCE OF THE USE OF THE METANEB IN CF: FEASIBILITY, SAFETY AND PATIENT REPORTED OUTCOMES

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We introduced Intrapulmonary Percussive Ventilation (IPV) using the Metaneb in Melbourne in 2016.

**Aim:** To evaluate the clinical experience, feasibility, safety and patient reported outcomes after using the Metaneb for airway clearance therapy (ACT) in adults with CF.

**Methods:** Patients with persistent mucus plugging not improving with their usual ACT techniques were selectively treated with the Metaneb providing constant positive expiratory pressure (CPEP) and constant high frequency oscillations (CHFO). Two to three minute cycles of each modality with pauses for expectoration and rests (as required) were provided. Dosage was individualised. Patients recorded their experiences on visual analogue scales (VAS) with anchors from -5 (most negative outcome) to 0 (no difference) to +5 (most positive outcome) after treatment with the Metaneb compared with their usual ACT.

**Results:** Thirty four patients (14 male) were treated with the Metaneb. Data is presented as mean (SD), range. Age: 35.9(11.2)20-76 years; FEV1: 46.2(16.8)25-87% predicted; FVC: 65.9(15.0)47-105%; BMI 22.0(4.0)17.7-40.2; VAS scales: Effectiveness of airway clearance: 3.4 (1.2) 1 to 5; Sputum volume cleared: 2.8 (1.5) 0 to 5; How clear/free breathing after treatment: 3 (1.6) 0 to 5; ACT time taken: 0.3 (2.1) -3 to 3; How tiring Metaneb treatment: 1.1 (2.9) -3 to 5; How easy breathing with Metaneb: 1.6 (1.9) -1 to 5; Preference for Metaneb with exacerbations: 4.6 (1.1) 1 to 5; Mucolytics used: Isotonic saline 66%, HS 33% of patients; CPEP and CHFO pressures: 10-20cmH2O; preferred CHFO frequency: 230 RPM (n=32). Treatment time was similar to usual ACT (PEP/Oscillating PEP/Autogenic drainage/Forced Expirations). Clinical reasons for selecting the Metaneb: refractory mucus plugging (n=32); Pregnancy (n=3); bowel obstruction (n=1); abdominal surgery (n=3). There were no adverse events.

**Conclusions:** Treating selected adults with CF with the Metaneb was feasible, safe and more effective in clearing refractory mucus plugging than usual ACT in selected patients.
TRANSITIONING TO ADULT PHYSIOTHERAPY SERVICES: EXPERIENCES OF ADOLESCENTS WITH CYSTIC FIBROSIS IN WESTERN AUSTRALIA

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Introduction: The transition of adolescents with cystic fibrosis (CF) from paediatric to adult services is a stressful experience for patients and families. Two of the most common concerns of patients prior to transition are leaving well-known caregivers and meeting a new care team. Previous studies have identified meeting the adult CF team prior to transition as a way to significantly reduce the level of concern and improve patient satisfaction.

In June 2018 Princess Margaret Hospital for Children (PMH) moved to the Queen Elizabeth II Medical Centre site. Re-named the Perth Children’s Hospital (PCH), this brought the paediatric CF service to the same location as the adult centre at Sir Charles Gairdner Hospital (SCGH).

Previously, formal transition clinics at PMH had been attended by an adolescent, their family, and a SCGH consultant and physiotherapist. With both centres now being at the same location, there was an opportunity to change the experience of transitioning from paediatric to adult physiotherapy services. Replacing the formal introduction and handover meeting, physiotherapists from each service now meet at PCH to handover during a joint airway clearance treatment.

Methods: A sample of patients who have transitioned from PMH/PCH to SCGH and the physiotherapists involved in this process will be interviewed to discuss their experiences on the transition process before and after the relocation of PMH to PCH.

Results: Patients’ and physiotherapists’ experiences of these different methods of transitioning physiotherapy services will be available for presentation in poster format at the time of the Australian Cystic Fibrosis Conference in 2019.

Discussion: This new process of transitioning care between paediatric and adult physiotherapists has enabled a relaxed and informal environment. The transitioning adolescent is now able to meet the adult physiotherapist for the first time with the paediatric physiotherapist in familiar surroundings while completing a familiar task, reducing anxiety and improving patient satisfaction.
THE ALFRED STEP TEST EXERCISE PROTOCOL (A-STEP): OPTIMISING AS A MAXIMAL EXERCISE TEST FOR ADULTS WITH CYSTIC FIBROSIS

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We developed and feasibility tested the A-STEP, an incremental, maximal exercise test for adults with CF across the range of lung function and age that replaces the submaximal 3 Minute Step Test. We identified a number of potential A-STEP limiting factors.

Aim: To identify limitations that impact optimal usefulness of the A-STEP. METHODS: We reviewed each subject’s (n=40) individual test to identify any parameters affecting A-STEP performance.

Results: Maximal criteria: In the safety/feasibility study maximal was determined if subjects reached one of: 1 ≥ 90% age predicted HR max; 2 ≥ 9 or 10/10 for maximal shortness of breath 3 ≥ 9 or 10/10 for maximal Leg Fatigue. Eighty percent achieved at least one criteria: 142.5%; 242.5%; 355%. To be a maximal test, modified instructions ensure all future subjects achieve at least one maximal criteria before stopping. Safety: All subjects safely completed between 6 - 15 Levels. An additional level was added as 1 subject completed the 15 Level test. SpO2 fell below 80% (criteria for stopping) in 1 subject who quickly recovered without supplemental oxygen. There were no adverse events when performed by a trained physiotherapist. Preparation: Moderate step movement occurred at high speeds, a non-slip mat resolved this. Instructions have been included to limit movement artefact of oximetry and ensure subjects are well hydrated pre-test. Bronchodilators should be individualised as may result in pre-test tachycardia. Subjects should be reminded to wear appropriate clothing, as this resulted in submaximal testing in a small number.

Conclusion: A-STEP limitations were identified and the protocol altered around objective physiological criteria, number of test levels, safety concerns, subject preparation and physical aspects of carrying out the test. The optimised A-STEP is low cost, easy to use, time efficient, without floor and ceiling and can be used safely for adults with stable CF.
PHYSIOTHERAPEUTIC MANAGEMENT OF LOBECTOMY IN PAEDIATRIC CYSTIC FIBROSIS: A CASE REPORT

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**Background:** Cystic fibrosis (CF) patients can develop pulmonary lesions resulting in localised complications that are non-responsive to conservative management. Resection of damaged tissue may prevent spread of microorganisms and slow disease progression (Lucas et al., 1996).

Over a 60 year period, less than 170 cases of pulmonary resection surgery have been published in the paediatric CF population (Villac Adde et al., 2018). Within our centre, partial lung resection has been performed twice over a ten year period.

Post-operative physiotherapy management is challenging due to sparsity of supporting evidence and risk of anastomotic breakdown and/or pneumothorax with the use of positive expiratory pressure (PEP) therapy. Intervention becomes a balance between optimisation of secretion clearance and physical function whilst avoiding damage to healing tissue. This case examines some of the considerations for physiotherapy intervention in a paediatric patient who underwent lobectomy.

**Discussion:** A 5yo female (dF508 homozygous) underwent a left upper lobe lobectomy. Her regular physiotherapy routine included the use of PEP. Pre-operative physiotherapy management focused on exploring alternative airway clearance techniques (without PEP) and play to enhance safety and reduce anxiety.

Factors impacting post-operative physiotherapy intervention included development of subcutaneous emphysema, sub-optimal pain relief and intravenous therapy location. Post-operative physiotherapy management included deep breathing exercises, mobilisation and thoracic mobility exercises. PEP therapy was successfully reintroduced two weeks post operatively following radiological resolution of subcutaneous emphysema and absence of pneumothorax.

**Conclusion:** This case will present management of a paediatric CF patient post lobectomy ensuring effective and safe airway clearance. It will integrate consumer feedback and promote discussion of optimal physiotherapy management in this rare population.

**Discussion points**
- Airways clearance and exercise in a patient post lobectomy including precautions and contraindications to physiotherapy treatment.
- Patient desensitisation and preparedness for surgery through role play.
- Post-operative clinical course and improvements in objective measures.

**References:**

CONTROLLED INHALATION IMPROVES EFFICIENCY OF AEROSOL DELIVERY IN CYSTIC FIBROSIS

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Background: Inhaled therapies are used extensively in the treatment of cystic fibrosis (CF). With progressive impairment of lung function, aerosol deposition of inhaled drug occurs more centrally in the lung. The aim of this study was to determine whether long slow inhalations with a dosimetric nebuliser might improve penetration of aerosol into the peripheral lung areas.

Methods: A randomised cross over study was used to compare two aerosol delivery systems in 5 subjects with moderately severe CF lung disease (aged 12-18 years; mean FEV1 72%; range 63-80%). Each delivery system utilised different inhalation modes, hence delivery time of the isotope labelled normal saline was adjusted, so the total inspiratory time with each device was comparable. The pattern of aerosol distribution was compared between a) tidal breathing using a standard delivery system (Pari BOY® SX compressor and PARI LC SPRINT®) and b) long slow inhalations using a dosimetric delivery system (AKITA® JET system and PARI LC SPRINT®). Distribution and total aerosol dose deposited corrected for attenuation was obtained using gamma scintigraphy.

Results: Both systems delivered comparable volumes to the lungs (225.66µl vs 202.61µl, p=0.45) however the standard system delivered a greater volume to the orogastric regions (185.74µl vs 51.98 µl, p=0.013) and a greater volume overall (407.97µl vs 254.59µl, p=0.024). The dosimetric system was associated with a higher proportion of the delivered dose depositing in the lungs (74%), compared to standard system (32%)(p<0.0001). There was no difference in the mean peripheral to central ratios (2.13 vs 2.16, p=0.90).

Conclusion: Long, slow inhalations with the dosimetric system was more aerosol efficient as it significantly reduced orogastric deposition and exhaled wastage. The lack of difference in the peripheral to central deposition ratio might be explained by a disproportionate amount of larger droplets deposited centrally, achieved by the long slow inhalations.
“SHORT AND SWEET... I REALLY ENJOYED IT!”: TOLERANCE AND EXPERIENCES OF A HIGH INTENSITY INTERVAL TRAINING PROGRAM IN PEOPLE LIVING WITH CYSTIC FIBROSIS

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Introduction: In people with cystic fibrosis (CF), higher exercise capacity is associated with better quality of life, reduced risk of hospitalisation and improved survival. Guidelines recommend that this population complete 30-60 minutes of moderate intensity continuous exercise daily, which can be difficult due to time-constraints and intolerable symptoms. This study aimed to report the tolerability and experiences of a low-volume high intensity interval training (HIIT) program in adults with CF.

Methods: This study formed part of a single-blinded randomised controlled trial whereby adults with CF were randomly allocated to the experimental (10-minutes of cycling-based HIIT thrice weekly for 8 weeks; intensity progressed based on participant tolerance) or control group (weekly contact via a preferred method). For the experimental group, tolerability (attendance, completion and post-exercise muscle soreness [Visual Analogue Scale]) was recorded following each session. Upon program completion, participants were asked to undertake a semi-structured ‘debrief’ (audio-recorded, transcribed, coded and thematically analysed).

Results: Seven adults with CF (aged 29±5 years, FEV1 69±18% predicted) were allocated to the experimental group. Attendance and completion of the HIIT program was high (93% and 100%, respectively). One episode of mild muscle soreness (VAS:10mm) post-training was reported. Prominent debrief themes were; excellent tolerability (“I was never sore...I always felt good after, more energy”), high exercise enjoyment (“I really enjoyed it [HIIT]...I am sad that it is finishing, I probably will incorporate it into my routine going forward” and “short and sweet...I looked forward to it!”) and low time commitment/easy routine (“the fact that it was quick, it’s going to be over soon, it’s like, I can do this” and “the sessions weren’t ‘easy’ but the fact that it was a small amount of time really worked for my schedule”).

Conclusion: There is potential for HIIT to be a tolerable, low-burden and well-accepted method of exercise training for people with CF.
EXERCISE TESTING AND TRAINING PRACTICES IN AUSTRALIAN AND NEW ZEALAND CYSTIC FIBROSIS CENTRES

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Background: Exercise testing and training are considered fundamental components of care in people living with cystic fibrosis (CF). While the body of evidence that supports annual exercise testing and regular exercise training in this population is substantial, there are currently no published data pertaining to the uptake and importance placed on these practices within Australian and New Zealand CF centres.

Aim: To determine the extent, scope and importance placed on exercise testing and exercise training within CF centres across Australia and New Zealand.

Methods: Information pertaining to exercise testing and training practices was sought by administering a survey to eligible CF centres across Australia (n=22) and New Zealand (n=18). The survey comprised five sections (46 questions) and was sent via an online link (Qualtrics) to a health professional (preferably with direct knowledge of involvement in exercise testing and training practices) at each centre. Response rate (RR) was optimised using the Dillman approach.

Results: A RR of 80% (32/40) was achieved. Each state/territory in Australia, except the Northern Territory, was represented by at least one centre. Eight of the 12 major regions in New Zealand were represented by at least one site. Regarding tests of exercise capacity, field tests were performed more commonly than laboratory tests (28/32 [88%] versus 11/32 centres [34%]; difference: 54%; p<0.0001). Most (88%) respondents perceived field tests to be at least ‘somewhat’ important whereas 90% of respondents perceived laboratory tests to be ‘a little’ to ‘somewhat’ important. Regarding exercise training, the importance of regular participation in physical activity and/or exercise was discussed by at least one health professional in the CF team at every clinic appointment and/or annual review. Some form of outpatient exercise training offered to most patients (24/32; 75%).

Conclusion: This survey captures the current practices of exercise testing and training in CF centres across Australia and New Zealand.
WHOLE BODY VIBRATION TRAINING IN CYSTIC FIBROSIS: FACE TO FACE VS. TELEHEALTH

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**Background:** Bone deficits, fractures, and sarcopenia are significant sequelae in cystic fibrosis (CF). Whole Body Vibration Training (WBVT) is an emerging adjunct with positive impact on bone and muscle response in CF. Our project aimed to determine the feasibility and effectiveness of WBVT in children with CF, especially in relation to bone and muscle parameters. Additionally, we hypothesised Telehealth treatment reviews would reduce travel and financial demands on families.

**Methods:** Participants: 15 pre-pubertal outpatients with CF, mean age 7.9, QCH CF Unit. Intervention (n=9) : standardised 12 week WBVT program incorporating airway clearance techniques (ACT); 5 sessions/week; Controls (n=6): usual care over 12 weeks. ACT was included in the required WBVT rest periods, minimising time burden on participants. Primary outcome: lean body mass via dual-energy X-ray absorptiometry (DXA), converted to lean body mass index (LBM/ht2). Secondary outcomes: Quality of life (QOL), spirometry, densitometry data; functional muscle strength measures and posture. Physiotherapy reviews were performed at 4 and 8 weeks, either face to face at QCH (n=3) or via telehealth (n=6) to assess technique.

**Results:** Our primary outcome of LBMI didn’t demonstrate significant changes over 12 weeks, however, three DXA data sets demonstrated significant changes in the WBVT group, which were not seen in controls: bone mineral content Z-scores of total body and total body less head and importantly, the bone content for amount of muscle z-scores. No significant differences were shown between groups in QoL physical functioning and treatment burden domains, which supports the feasibility of WBVT home programs.

**Conclusion:** Optimising musculoskeletal health prior to puberty is critical. Integrated WBVT and airway clearance are feasible with telehealth or clinic reviews. Three months WBVT may reduce musculoskeletal deficits in young individuals with CF without increased treatment burden, that warrants further investigation.
AN AUDIT OF PAEDIATRIC CYSTIC FIBROSIS (CF) ADMISSIONS WITH HOSPITAL IN THE HOME (HiTH).

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Introduction: Patients with CF at the Royal Children’s Hospital complete part or all of their tune-up admissions with HiTH. In 2015, a day 7 medical and physiotherapy review at home was established, including spirometry. In discussion with the CF team, length of admission was based on this review.

We aimed to examine the day 7 review to determine:
• how often the review occurred
• whether the CF team documented discussing the review outcome with the family
• reasons for extending admission beyond 10 days

Methods: CF admissions under HiTH from 2015-2018 were reviewed retrospectively via the patients’ electronic medical record (EMR). Data was collected on tune-up location, length-of-stay, day of transfer to HiTH, whether day 7 review occurred and if it was discussed with the family. Sputum culture results were collected for patients with admissions >10 days.

Results: 366 tune-ups occurred under HiTH (124 patients). Average length-of-stay was 13.47 days. 81% of patients transferred to HiTH prior to day 7 had a review completed; 84% of these occurred on day 7 and 28% of reviews were discussed with families. 68% of admissions extended beyond 10 days: most commonly due to ongoing cough/unwell (51%) and sub-baseline pulmonary function (20%). The most common bacteria identified were pseudomonas aureginosa (31%), haemophilus species (30%), MSSA/staph aureas (20%), aspergillus (20%), candida species (20%), mycobacterium avium complex (13%) and stenotrophomonas (9%).

Post audit, day 7 review key performance indicators (KPIs) were established:
• review completed for 95% of admissions
• CF team to contact families (with EMR documentation) for 80% of reviews

Conclusion: Day 7 review of CF patients admitted under HiTH obviates the need for in-hospital review and enhances length-of-stay decision making. Improved communication with families at home is required to ensure they feel connected to the hospital during a HiTH admission. Future audit is required to examine KPI adherence.
Daily airway clearance (ACT) is prescribed from diagnosis in the management of CF to minimise infection, inflammation and airway damage. Many factors impact on the effectiveness of ACT including adherence, technique, lack of knowledge and social influences. Telehealth provides a platform on which healthcare services are provided without attending the hospital, interfering less with school and parental work commitments, improving cost and time-effectiveness for families and giving insight into home environments. This platform was incorporated into physiotherapy management of CF children in March 2018.

Aim: Review the physiotherapy management of CF children via telehealth and patient satisfaction with the support provided from March 2018 – April 2019.

Methods: A retrospective review was performed to identify key areas of physiotherapy management provided via telehealth. Families who received at least 5 telehealth sessions were asked to complete a Telehealth Satisfaction Questionnaire based on Parmanto’s Telehealth Usability Questionnaire [1] which explored usefulness, ease of use, interface/interaction quality, reliability and future use using a sliding scale of agreement between 0-100. Data presented as median (range).

Results: Thirty-two patients were reviewed via telehealth, aged 13 (2-17) years, and attended 7 (1-93) sessions: 12/32 (38%) for adherence; 11/32 (34%) for technique optimisation; 5/32 (16%) for education; and 2/32 (6%) for social reasons or to support home intravenous antibiotic treatments. Twenty-one of the 32 (66%) received more than 5 sessions and 16/21 (76%) returned satisfaction surveys. Overall satisfaction was 100 (40-100), improved access to healthcare scored 100 (71-100), provision of needs 100 (80-100), equivalence to seeing clinician in person 96 (40-100) and future use 100 (95-100).

Conclusion: Patient satisfaction with physiotherapy telehealth support was high for a range of factors impacting on the effectiveness of ACT.

ARE PHYSIOTHERAPY STUDENT-LED TELEHEALTH SESSIONS EFFECTIVE IN IMPROVING ADHERENCE TO AIRWAY CLEARANCE TECHNIQUES IN ADOLESCENTS WITH CYSTIC FIBROSIS?

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Airway clearance techniques (ACT) become more complex in adolescence and adherence to prescribed regimens declines. To elicit change in adherence, participation in supervised physiotherapy sessions at least twice a week is required. Telehealth eliminates the need to attend the hospital and provides insight into the home environment where ACT are performed.

Aim: Assess whether physiotherapy student-led telehealth sessions improves adherence to ACT regimens over 6 months in CF adolescents.

Methods: Adolescents with poor adherence to prescribed ACT regimes were offered telehealth sessions. To facilitate multiple weekly supervised ACT sessions, physiotherapy students provided concurrent individual ACT sessions to adolescents with supervision from the CF physiotherapist. At baseline and after 1 and 6 months, Quittner’s Treatment Adherence Questionnaire (TAQ) [1] was completed. Quittner’s Airway Management Skills Checklist (AMSC) [1] for ACT effectiveness was completed at baseline and 6 months. Minimum/maximum FEV1% in the 6 months prior to telehealth and during 6 months of telehealth were compared. Data presented as median (range).

Results: 5 adolescents were recruited, aged 15 (12-16) years. All adolescents reported having done no ACT in the two weeks prior to commencement of telehealth. ACT regimens had 5 (3-6) treatments taking 30 (20-40) minutes. Adolescents attended 42 (22-59) sessions. After one month adherence was 43% (32-93) and after 6 months 44% (36-100), an increase of 4% (-12 – 20) despite changes in treatment number to 5 (4-6) treatments and time to 40 (20-45) minutes. Minimum/maximum FEV1% did not change with minimum values of 81% (65-102) and 81% (63-94), and maximum values of 87% (74-108) and 87% (69-94) respectively. At baseline AMSC scores were 85% (42-94) and at 6 months 100% (83-100), an improvement of 15% (6-56).

Conclusion: Physiotherapy student-led telehealth sessions improved adherence and effectiveness of ACT in adolescents with poor adherence to prescribed physiotherapy regimes.

ANALYSIS OF THE IMPACT OF PHYSICAL ACTIVITY ON 10-YEAR HEALTH OUTCOMES IN ADULTS WITH CYSTIC FIBROSIS

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Background: For adults with Cystic Fibrosis (CF), higher levels of physical activity are associated with improved exercise capacity and quality of life, as well as reduced decline in lung function and bone mineral density. To date, the impact of physical activity on long-term health outcomes, including transplantation, survival, hospitalisation and CF-related diabetes, is unknown.

Aim: Analyse differences in 10-year health outcomes (lung transplantation, survival, hospitalisation, lung function, bone mineral density and glucose tolerance) according to moderate-vigorous physical activity (MVPA) in adults with CF.

Methods: Between 2007-2008, 104 adults from the RPA CF clinic reported their MVPA (mean age 29±10 years, 45 female, FEV1 63±24 %predicted) in a research study on exercise behaviour. Participants were prospectively classified as achieving or not achieving physical activity recommendations (defined as ≥150 minutes of MVPA/week). Health data were collected from RPA hospital records over the following 10-year period. Participants were censored at the time of transplantation or death. The mean follow-up period was 7.5±3.2 years (range 0-10).

Results: 48 adults were prospectively classified as achieving physical activity recommendations (“high activity”) and 56 were not (“low activity”). The demographic and clinical characteristics of the groups were similar at baseline for age, sex, lung function, bone mineral density, glucose tolerance and hospitalisations in the previous year. There was a trend for reduced incidence of transplantation or death in the “high activity” compared to the “low activity” group (27% v 43%, p=0.09). There was significantly longer time to transplantation or death in the “high activity” compared to the “low activity” group (p<0.05), taking 3.1 years longer until 25% of participants had lung transplantation or died. Data for other health outcomes will be analysed and presented at the 2019 Australasian CF conference.

Conclusion: Higher levels of MVPA increased time to transplantation or death in adults with CF.
RELATIONSHIP BETWEEN MODIFIED SHUTTLE TEST, QUALITY OF LIFE AND PARTICIPATION IN CHILDREN WITH CYSTIC FIBROSIS; A CROSS-SECTIONAL, RETROSPECTIVE STUDY

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Background: Modified shuttle test (MST) is an effective tool in assessing functional exercise capacity in adults and children with cystic fibrosis (CF). Understanding the relationship between MST, lung function and quality of life (QoL) can help clinicians better interpret each assessment findings.

Aim: To evaluate the inter-relationship between MST parameters, lung function, and QoL measures in children with CF.

Method: We performed a retrospective cross-sectional analysis of children with CF ages 8 to 18 year. Data was collected at Queensland Children’s Hospital as part of an ongoing randomised controlled trial. Outcome measures included MST distance (MST(distance)), lung function via spirometry, and cystic fibrosis questionnaire – revised (CFQ-R).

Results: Fifty-one children with CF (24 female, 27 males; ages 8.10 to 17.34 years) with mean forced expiratory volume in 1 second percentage predicted (FEV1%predicted) of 86.58% predicted (40-112%). Moderate to good relationship between MST(distance), age (r=0.422, p<0.01), height (r=0.412, p<0.01), weight (r=0.329, p<0.05) and physical functioning domain (r=0.518, p<0.01) of the CFQ-R and no other domain. Correlation between MST(distance) and FEV1%predicted was insignificant. Correlations between physical functioning and respiratory domains of the CFQ-R, FEV1%predicted and FVC%predicted were observed. A multiple regression model was applied to predict FEV1%predicted from age, MST(distance) and BMI. These variables predicted FEV1%predicted (F[3,45]=5.381, p<0.01, R2=0.264). All three variables were significant in the prediction model (p<0.05). MST(distance) was a stronger predictor and accounted for more variability in predicting the physical functioning domain of the CFQ-R than FEV1%predicted (MST(distance): F[1,48]=17.501, p<0.01, R2=0.285; FEV1%predicted: F[1,47]=4.639, p<0.05, R2=0.09).

Conclusion: We observed moderate-to-good correlations between MST(distance) and lung function, anthropometric measures and the physical domains of QoL in children in CF with moderate to normal lung function. Improvement in MST may therefore benefit overall lung function. The model developed cannot accurately predict improvements in lung function due to high variability.
HOSPITAL IN THE HOME (HITH) TELEHEALTH AND HOME VISIT HYBRID PHYSIOTHERAPY MODEL

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HITH within the Sydney Children’s Hospital Network (SCHN) has become an important part of the care continuum for CF patients, with increasing numbers of patients accessing the service and creating the need for alternative methods of increasing efficiency such as delivering care via Telehealth.

**Aim:** To expand the HITH service to provide patient centred physiotherapy care within the home via telehealth. Our goal was to provide physiotherapy consultations via videoconferencing to patients deemed appropriate, as an adjunct to face to face sessions. This project aims to assess the feasibility, efficiency and acceptability of telehealth as a service delivery medium.

**Method:** Bidaily physiotherapy treatments were provided, within Metropolitan Sydney one face to face and one telehealth session was provided each day. Out of area patients received two telehealth sessions, where previously no physiotherapy was accessible. Outcome measures included family satisfaction survey, travel time saved and frequency of BD visits enabled.

**Results:** The Telehealth 12 month pilot project commenced December 2017 and had a direct impact on health economic indicators and the productivity of physiotherapy within HITH. In the six months preceding implementation of telehealth a mean of 137 occasions of service (OOS) per month were performed, in the 2018 corresponding period this increased to 170 OOS. Telehealth led to increased efficiency with therapists now delivering approx. 2 extra sessions per day, facilitating BD physiotherapy for 36 admissions during the pilot year, compared to 4 in the year prior. Telehealth therapy sessions for in area patients saved over 13 days or 319 hours and 14 693km of driving from Dec 2017 to Dec 2018. Patient/Carer satisfaction with Telehealth was reported as high (9.1/10).

**Conclusion:** Telehealth is a feasible, acceptable and efficient method of delivering physiotherapy to CF patients, and we have begun a study assessing effectiveness of this type of treatment delivery.
RAAP: RESPIRATORY ADMISSION AVOIDANCE PHYSIOTHERAPY

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Early intensive physiotherapy for CF patients through HITH may avoid admission for respiratory exacerbations. There is potential to reduce the severity of patient health decline, reducing bed occupancy, IV medication, burden of care and long term reductions in respiratory function (Sanders, 2010). Early intervention community base physiotherapy has been shown to decrease hospitalisation and ED presentations in a CF population (Ledger et al, 2013, Norman et al, 2011).

**Aim:** To avoid hospitalisation and reduce admission related costs (patient and hospital), by preventing further decline in clinical status through early home based physiotherapy intervention.

**Method:** Referrals accepted for patients at risk of hospitalisation from CF teams across SCHN for a 2 week physiotherapy only RAAP admission to optimise home treatment. Receiving bidaily sessions week one and daily week two. Medical reviews were completed pre, mid and post RAAP admission. This program aimed to be family centred, allowing patients to receive treatment at home or school.

**Outcomes:** Hospitalisation avoided (no hospitalisation within 6 weeks post discharge from RAAP), patient/carer satisfaction survey, estimated admissions/bed days saved.

**Results:** The pilot feasibility study ran from March 2017 to June 2018, nine of 12 patients avoided hospitalisation, equating to 126 bed days potentially saved. Average time to next admission was 166 days and four have not been readmitted. The three unsuccessful RAAP admissions included a post-transplant patient, a viral infection mid admission and exacerbation too severe for HITH, and were all hospitalised by day 14. Satisfaction was high (9.4/10), perceived effectiveness equally as high (9.3/10) and timing/logistics rated slightly lower (7.8/10).

**Conclusion:** This program was shown to be a feasible and acceptable pathway for preventing further decline in health status leading to hospitalisation in the majority of accepted patients. Further expansions of this program are reliant on additional resources allowing for preventative models of care.
WHAT RELATIONSHIP DO PHYSICAL ACTIVITY AND NUTRITIONAL PARAMETERS HAVE IN RESPIRATORY HEALTH STATUS OF ADULTS WITH CYSTIC FIBROSIS?

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Background: Previous studies have linked optimal nutrition with a reduction in the decline in respiratory health status in cystic fibrosis (CF). Similarly, regular moderate-vigorous physical activity (PA) may preserve respiratory function and quality of life (QOL) in adults with CF. However, few studies have investigated the combination of energy expenditure, physical activity, nutritional requirements in adults with Cystic Fibrosis (CF), and their combined association with QOL.

Aim: To correlate PA, muscle strength, energy intake and expenditure with indicators of QOL in adults with CF.

Method: Prospective recruitment of 47 clinically stable adults with CF attending TPCH Adult Cystic Fibrosis Centre between 2013-2018. PA was assessed using a “SenseWear Pro 3 Armband” (SWA), worn for 5-7 days. Six minute walk test (6MWT) completed, with recording of distance (6MWD), nadir saturations and subjective breathlessness and quadriceps strength tested. Current lung function, weight and body mass index (BMI) were recorded. Three day food diary and Cystic Fibrosis Questionnaire-Revised (CFQ-R) QOL questionnaire completed. Nutritional, PA and physiological parameters were correlated with QOL measures.

Results: The cohort mean CFQ-R score was 67.5±15.3%, with a mean 6MWD of 632±130m, peak breathlessness of 3.9±2.0 and nadir SpO2 of 91±6%. Mean BMI was 22.26. Univariate analysis demonstrated CFQ-R correlated with 6MWD (r= 0.38, p<0.05), peak dyspnoea (r= -0.57, p<0.01), nadir SpO2 (r= 0.43, p<0.05), FEV1(%) (r= 0.56, p<0.01) and hospital admissions (r= -0.36, p<0.05). Conversely, PA and nutritional intake did not correlate with CFQ-R domains for HRQL or hospitalisations. Quadriceps strength and total energy expenditure (TEE) correlated with daily average kilojoule intake (r=0.41, p<0.01 and r=0.48, p<0.01 respectively).

Conclusion: 6MWT and FEV1 percentage predicted, but not PA or nutritional intake correlate with QOL and risk of hospitalisation.
PATIENT PERCEIVED EFFECTIVENESS AND LONG TERM PERFORMANCE OF AUTOGeneric DRAINAGE COMPARED WITH STANDARD AIRWAY CLEARANCE TECHNIQUES

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Introduction: Cystic Fibrosis (CF) is characterised by abnormally thick airway secretions causing chronic lung infections. Airway clearance techniques (ACT) are a standard component of modern CF management. However, no single ACT has been shown to be consistently superior, so individuals perform a variety of techniques over time. Autogenic drainage (AD) is an effective technique for mobilising sputum and can be performed without the use of any devices. However, AD can be difficult for both physiotherapists and patients to learn and perform effectively, which may affect the decision to continue with the technique long term.

Methods: 20 Adults with CF were taught AD in multiple sessions. Once individuals were confident performing AD, the subjective effectiveness of the technique was compared with their previous ACT (eg. positive expiratory pressure, percussion and modified postural drainage). A questionnaire using a visual analogue scale was completed for the different physiotherapy techniques used.

Results: 17 patients continued to use AD for at least 6 months, median age of 25 (±6.1). The majority of adults required higher mental concentration (14 /17) when performing AD, but found it equally or more effective (14/17) compared with their previous techniques. A retrospective audit found that lung function was not significantly different after 6 months of AD, compared to previous ACT. Despite the perceived effectiveness, some individuals reverted back to their previous technique in the long term, or combined aspects of AD with their previous technique.

Conclusion: AD appears to be an effective airway clearance technique, but requires more concentration to perform. Despite perceived benefits, some individuals use alternative techniques for long term airway clearance. Further studies are required to assess the efficacy of this airway clearance method in the long term.
PSYCHOSOCIAL/NURSING/EDUCATION ABSTRACTS
SET UP FOR FAILURE - "NON-ADHERANCE VERSUS NON-AFFORDABILITY"

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Mater Health

Due to advances in medical treatments individuals with Cystic Fibrosis (CF) are now surviving into their third and fourth decades of life however, there is an associated increase in complex co-morbid conditions. Management of these conditions often requires an arsenal of symptomatic treatments and a multitude of prophylactic daily regimes (1). "Non-adherence" to these regimes and associated deterioration in baseline condition has been extensively reported in the literature (2-14).

But are we as health care professionals setting our patients up for failure, prescribing treatment regimens that CF individuals are unable to afford and therefore are unable to “adhere” to? This case study outlines 4 different CF patient scenarios in our current “young adult/ adult” clinic demonstrating the diversity in costs depending on your social situation over a twelve month period. These case studies include:

- Female patient in her 40’s, single, working fulltime from home, paying off a mortgage
- Female patient in her 20’s, single mother of 3 children (no child support), renting
- Male patient in his 30’s, married, tradesman with his own business, paying off a mortgage
- Male patient in his 20’s, working for months at a time overseas, living with family when in Australia

These 4 examples clearly outline the enormous financial burden that is currently a reality for many of our patients even when having access to government assistance. It further demonstrates the importance of patient consultation when prescribing treatments, tailoring treatments to what is affordable and thereby not inappropriately labelling our patients as “non-adherent”.

AUSTRALIAN AND NEW ZEALAND CYSTIC FIBROSIS NURSING CLINICAL PRACTICE RESOURCES

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Background: Information resources such as clinical practice guidelines, protocols and information documents are widely used by CF services. Resources take time to develop and update. Differences in resources between centres contribute to variation of care which is associated with poorer health outcomes.

Aims:
1. To determine the use of information resources across key clinical areas between CF centres in Australia and NZ.
2. To determine who develops and maintains the resources.
3. To gauge willingness for developing standardised, shared resources.

Methods: A REDCap survey was distributed to CF nurses across Australia and NZ through the CF Nurses Google Group. The survey addressed the scope and number of CF related clinical practice resources used by each CF centre in Australia and NZ across five areas of care: transition, infection control, new diagnosis, annual review and hospital admissions.

Results: 22 responses were received. Respondents represented all states and territories in Australia and NZ across adult, paediatric and dual site CF centres, except South Australia and the South Island of NZ. Sixteen (73%) respondents saw the responsibility of developing and updating information resources falling on the nursing and allied health team. Twenty one (95%) respondents believe the task of keeping resources updated is challenging. All respondents reported using resources from other CF centres and organisations. Twenty (90%) of respondents indicated willingness to share information resources with other centres. Twenty (90%) respondents were willing to contribute to developing standardised shared resources.

Discussion: Clinical information resources are widely used across all CF centres. Resources are mostly developed and maintained by nursing and allied health teams. Informal sharing of resources occurs frequently. Nursing staff appear to be interested and willing to collaborate to develop standardised resources to reduce unnecessary repetition in updating and creating resources at individual centres.
On Wednesday 13th March 2019, Monash Children’s Hospital, The Royal Children’s hospital and Cystic Fibrosis Community Care joined together to present the first Victorian Professional Development day for Educators.

Children spend 6hrs a day, 195 days per year, at school. During this time their teachers are responsible for them to provide education but also to manage any particular care each child might require.

Imagine being a teacher of a child with a chronic illness. Not only are you as the teacher responsible for this child’s education but also for their management of their illness during their school year. Cystic Fibrosis is a chronic illness which affects more than 3379 people in Australia (Australian Cystic Fibrosis data Registry, 2015). Of these, 1787 people are under the age of 19, therefore being educated at a school or by educators. It is therefore very important to educate these educators for these children with cystic fibrosis.

Previously, both Monash Health and The Royal Children’s Hospital have provided separate days. However, there was a limited number of people able to attend from rural areas. We are now combining our resources and our knowledge to present this educational day. Not only was the day presented live at Monash Health and The Royal Children’s Hospital, but we also streamed live to 5 ‘hubs’ throughout rural Victoria and southern New South Wales. This live streaming allowed for rural teachers to attend who may not have attended in the past.
DEMORALISATION IN CYSTIC FIBROSIS

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Introduction: People with Cystic Fibrosis (CF) face relentless symptoms with substantial and difficult treatment regimes. The psychological burdens CF people endure, such as elevated levels of depression, can lead to wilful cessation of treatment with resultant increased morbidity and early death. This giving-up is the clinical hallmark of demoralisation, where a person is overwhelmed by helplessness and hopelessness.

Aim: This study measured demoralisation and depression in an adult CF cohort, hypothesising that the personality trait of neuroticism would be a moderator in the relationship between demoralisation and depression.

Methods: 113 adults with CF (F=53, M=60) completed a health-related quality-of-life measure (CFQ-R), along with scales for depression (PHQ-9), demoralisation (DS-1) and personality (BFI-2). A 3x5 MANOVA and discriminant function analysis compared depression and demoralisation. Any moderating effects of neuroticism were identified with a regression moderation test, while a mediation test considered the effect of depression.

Results: Moderately elevated scores of depression (M=4.96, SD=4.55) and demoralisation (M=20.81, SD=14.16) were identified. Upon comparison of individual participants, 82% with severe depression had severe demoralisation, while only 45% of those with severe demoralisation had severe depression. This identified a group of patients who are highly demoralised but not depressed. Despite neuroticism not being a moderator in the relationship between demoralisation and depression, depression was found to have a significant indirect mediation effect (b=.539, BCa CI [.339, .759]) on the relationship between neuroticism and demoralisation.

Discussion: This study identified a new conceptual model of depression as a mediating influence on the relationship between neuroticism and demoralisation. Additionally, identifying a group of demoralised, but not depressed people questions the utility of current mental health assessments that screen for depression but not demoralisation. Establishing effective screening and interventional strategies for these at-risk individuals would provide a foundation to address demoralisation in people with CF.
TERTIARY TO COMMUNITY CARE FOR PEOPLE WITH CYSTIC FIBROSIS

CHARLOTTE BURR & KATHRYN PEKIN
PERTH CHILDREN’S HOSPITAL AND CYSTIC FIBROSIS WA

The WA CF Model of Care (2013) outlines a partnership-based model of service delivery that has a clinical governance framework utilising evidence-based guidelines and protocols to facilitate safe and effective tertiary-led care into the community sector.

Specialist clinical care is provided in the tertiary hospitals. Cystic Fibrosis WA (CFWA) as the community support provider assist with treatment plans set out by the interdisciplinary teams in the tertiary hospitals for people with cystic fibrosis (CF).

Community outcomes include assistance with airway clearance, assistance developing routines and self-management, exercise routines, and general support as directed.

Monthly meetings and regular contact as required ensures that this partnership model offers continuity of care.
EMOTIONAL WELLBEING SCREENING WITHIN QUEENSLAND CHILDREN'S HOSPITAL

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Since the 2017 Cystic Fibrosis Conference there has been an increasing focus on the emotional wellbeing of children and young people living with CF, as well as their parents. The TIDES study evidenced that significant elevations in symptoms of depression and anxiety were found among patients and parent caregivers across countries, suggesting that screening for symptoms of Depression and Anxiety should be performed annually and addressed systematically (Quittner, A. et al.)

Within Queensland Children’s Hospital (QCH) staff have been completing an Emotional Wellbeing Screen (GAD-7 and PHQ-9) with families since early 2017, focusing on adolescents (over 12 years) and their parents. Completion of the Screen was initially ad-hoc and sporadic as it was being distributed by different team members and was reliant on Consultants to score. Over time, it was determined that the Social Workers would assume responsibility of distributing, collecting and scoring the tool.

In November 2018 Cystic Fibrosis Australia (CFA) funded the ‘Mental Health Roadshow’ with both Alexandra Quittner and Anna Georgiopolous running workshops to upskill staff in screening for emotional wellbeing. Since this time the team at QCH have extended the scope of screening to include all parents from the point of diagnosis and adolescents 12 years plus.

This poster will examine the data collected by QCH and will identify trends as well as to consider the impact of the Mental Health Roadshow in terms of positively impacting on the provision of emotional wellbeing screening within Queensland.
ANXIETY AMONG CHILDREN AGED 6-12 WITH CYSTIC FIBROSIS AND THEIR PARENTS

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Introduction: Individuals with cystic fibrosis (CF) are at high risk of experiencing anxiety, which is in turn related to lower treatment adherence and poorer health outcomes. Additionally, up to half of the parents/caregivers of children with CF experience anxiety, potentially affecting their parenting behaviour and their children. However, research to date has focused on CF participants aged 13 years and older, leaving anxiety among those with CF aged 6-12 years largely unstudied. The relationship between parenting factors and child anxiety in the CF population is not well understood, nor are the anxiety-related factors procedural anxiety and parental fear of disease progression.

Methods: First, a systematic review of the CF literature collated existing knowledge on a) anxiety in children aged 6-12 years, b) wellbeing and efficacy of parents, and c) the relationship between parenting factors and child anxiety. Second, a cross-sectional survey administered through the Queensland Children’s Hospital CF Service to children with CF aged 6-12 and their parents assessed incidence rates of, and relationships between: child anxiety, child procedural anxiety, child depression, parental mental health, parental fear of disease progression, and treatment adherence.

Results: The systematic review indicated that anxiety is highly prevalent in children with CF, and that parent wellbeing and efficacy is lower than in the general population. However, the overall finding was that there is a vast dearth of research in this area and that no studies to date have analysed the relationship between parenting factors and child anxiety in children aged 6-12 with CF. Preliminary data on the incidence of, and relationships between, the factors assessed in the survey will be discussed.

Conclusion: Anxiety is an important issue in children aged 6-12 with CF, and further research on suitable interventions is required. Such interventions should also address procedural anxiety and fear of disease progression.
EXPLORING THE NEED FOR ENHANCED MENTAL HEALTH SERVICES IN A PAEDIATRIC CYSTIC FIBROSIS CLINIC

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Quittner et al (2014) reported rates of anxiety and depression in the CF population and caregivers 5 times higher than the general population. In response, the international CF community made recommendations for mental health care of people with CF. The CF clinic at the Children’s Hospital at Westmead provides care for 210 patients with a large multidisciplinary team (MDT) including 0.5FTE social worker. This falls well below recommendations of 2.0 FTE social worker and 1.5FTE psychologist for clinics greater than 150 patients (Australian CF ACI model of care, 2016). The aim of this study was to assess CF family and CF MDT satisfaction with access to psychosocial services within the CF clinic.

A satisfaction survey conducted in 2018 was completed by 49% of families. They were asked about current mental health services. 53% of respondents (n=56) indicated they were rarely or never asked about their child’s mental health. While 68% of families rated their child’s mental health as excellent, 63% indicated psychological support through the CF Clinic would be very (27%) or extremely (36.5%) useful. Furthermore, 66% of respondents indicated that such support would be very (23%) or extremely useful (42%) for parent/caregivers and/or siblings.

The CF MDT were surveyed to explore their perception of current mental health services. 92% felt ill-equipped to provide mental health care and 85% said they had insufficient time to do so. 80% believed mental health affected a caregiver’s ability to provide care and 90% believed a child’s mental health affected their ability to comply with treatments. All rated the clinic “below average” or “far below average” in meeting the mental health needs of patients and families within clinic.

These results have reinforced earlier work and guided a submission to the hospital to improve mental health services through the provision of increased staff and time allocation.
DEVELOPMENT OF AN ENZYME REPLACEMENT APPLICATION: A PROOF OF CONCEPT PROTOTYPE

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There are currently no efficient, accurate digital resources for the adjustment of pancreatic enzyme replacement therapy for achieving optimal nutrition status for children and adults who are pancreatic insufficient with cystic fibrosis in Australia. Through a partnership between a large tertiary hospital facility and university, a successful submission was made to transform the idea of developing a mobile-phone based tool for patients that can quickly calculate the amount of fat in a meal or snack and calculate the number of enzymes required into an application.

Currently, following education from a specialist dietitian, individuals typically estimate the number of enzymes required using multiple strategies to calculate the amount of fat in the meal or snack, then apply a predetermined ratio of number of enzymes per grams of fat. This technique is often inaccurate due to a poor understanding of the fat content of certain foods and requires a certain level of mathematical literacy. Poorly matched pancreatic enzyme replacement therapy may result in fat, energy and nutrient malabsorption, weight loss, low fat-soluble vitamin levels and can contribute to steatorrhea, abdominal pain or distension, gas and bowel obstructions.

The idea for an enzyme calculator is unique. Current mobile applications are limited to calculating the fat content, with no existing apps including the next step of calculating the number of enzymes required based on fat intake. A specialist dietitian is still required to advise the individual of their personalised enzyme to fat ratio that would need to be entered into the application to assist the calculation.

Information technology students and specialist dietitians have worked together integrating knowledge and innovation tools to develop a responsive web application, with the goal of providing proof of concept to assist with securing funding in the future for development of a native application to enable nationwide usability of this enzyme dose calculator.
MOBILE DEVICE UTILISATION LIFTING ADHERENCE AND TREATMENT ENGAGEMENT IN CYSTIC FIBROSIS (MODULATE-CF)

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Introduction: As survival in CF has improved, treatment complexity has increased and patients’ ability to complete prescribed treatments has decreased, with estimates that CF patients complete less than half of their prescribed physical and medication therapies. Regular engagement with a specialist CF centre result in better outcomes, however patients living in remote locations do not have easy access to specialist care. This challenge is most keenly felt in Australia where patients may reside 1000’s of kilometres from a specialist CF Centre.

Aim: To develop and determine the uptake and impact of a CF specific educational website and parallel health tracking web application on CF patient knowledge and adherence to maintenance therapies.

Methods: The Prince Charles Hospital (TPCH) Adult CF Centre (ACFC), in collaboration with UQ centre for online health and Supporters of 65 Roses Inc developed a web-based health tracking application which allows patients to record health management plans, medications and monitor key health metrics, including lung function and nutritional status. In parallel, a website was developed to provide CF specific health education and information on CF services provided through the TPCH ACFC. On access to the website, adult CF patients are asked to complete questionnaires on knowledge, confidence to self-manage and adherence to CF care at baseline, followed by access to online resources. Patients are supported in recording treatment plans by the multidisciplinary team. Follow-up questionnaires are completed at 1 and 6 months following utilisation of resources to determine the impact on patient confidence and adherence with CF care.

Results: Patient recruitment commenced in early May 2019, with a target of recruiting 50 patients. It is anticipated that recruitment will be completed and 1 month follow-up data will be available for all patients by August 2019, which will be discussed.
AN INNOVATIVE SMARTPHONE APPLICATION TO SUPPORT THE SOCIAL CONNECTEDNESS AND WELLBEING OF YOUNG PEOPLE LIVING WITH CYSTIC FIBROSIS

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Objectives: To develop and test a smartphone application, CyFi Space, to support the social connectedness and wellbeing of young people living with cystic fibrosis (CF).

Methods: A sequential mixed-methods approach was employed. Following the development of CyFi Space, 22 young people with CF aged 12-17 years attending CF clinics at 2 paediatric hospitals in Australia piloted CyFi Space for 6 weeks before completing an online survey to test the app’s usability and acceptability. Eleven online group interviews with a sub-sample of participants (n=20) explored the app’s perceived strengths and weaknesses and areas for improvement.

Results: The CyFi Space app contained 6 features: i) Alvi, a virtual buddy, that asked users how they were feeling, and suggested actions based on the user’s response; ii) wellness tips; iii) entertaining videos; iv) an online chatroom; v) CF information and resources; and vi) a support services page. CyFi Space was rated highly useable by study participants: 96% felt comfortable using the app, 91% learnt to use the app quickly, and 86% found it easy to use. Acceptability of CyFi Space was rated moderate: 77% agreed CyFi Space was fun to use and would recommend the app to other young people with CF. While there was strong support for CyFi Space, participants suggested improvements, such as accessing the chatroom from within the app rather than being directed to an external search engine, personalising the app's images and videos, and allowing users to turn off notifications.

Conclusion: Interventions that improve young people’s psychosocial health are vital in addressing the social isolation and mental illness often experienced by young people with CF. While CyFi Space has the potential to benefit the lives of young people with CF, changes recommended by study participants need to be incorporated into the app before it is distributed more widely.
A PILOT MENTORING PROGRAM FOR CYSTIC FIBROSIS DIETITIANS – A PRELIMINARY EVALUATION

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Introduction: Enhancing the knowledge, skills and abilities of dietitians new to the area of Cystic Fibrosis (CF) across Australia, has been identified as a key strategy for the successful implementation of the 2017 Nutrition Guidelines for CF in Australia and New Zealand. This project aimed to assess the preliminary impact of a pilot mentoring program for CF dietitians with less than three years of experience in CF nutrition.

Methods: Two senior CF dietitians developed the pilot mentoring program structure. The program included monthly mentoring sessions, a reading list and the completion of a knowledge based quiz and case studies, designed specifically for an adult or paediatric CF population. Four CF dietitians with less than 3 years experience in CF Nutrition (mentees) were paired with two senior CF dietitians with an average of 9.5 years working in CF (mentors). The matching of mentees and mentors was based on experience in adult vs paediatric CF nutrition care. Prior to commencing the program, mentees completed a short questionnaire on their expectations of the program. This preliminary evaluation will be completed six months into the program to assess overall mentee satisfaction, quality and usefulness of the program, mentee confidence with CF nutrition care and more detailed feedback on key aspects of the program.

Results: Expectations of the program from mentees included guidance with complex clinical cases, quality initiatives, research and time management in a busy caseload. Preliminary results are still being collected and will be available in June 2019.

Conclusion: It is expected that the introduction of a CF dietitian mentoring program for staff new to the area of CF will enhance the knowledge, skills and abilities of these dietitians – fostering growth and building capacity in this challenging nutrition field.
SYSTEMIC SCREENING FOR PREVALENCE OF DEPRESSION AND ANXIETY IN A WESTERN AUSTRALIAN ADULT COHORT WITH CYSTIC FIBROSIS

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Background: Individuals with Cystic Fibrosis (CF) are at increased risk of symptoms of depression and anxiety. This study sought to measure the prevalence of depression and anxiety symptoms in adults attending Sir Charles Gairdner Hospital (SCGH) CF clinic.

Methods: Over six months in 2017-2018, all adults attending the clinic were provided with two self-administering, standardised mental health screening tools. The Patient Health Questionnaire 9 (PHQ 9) identifies depressive symptoms, and the Generalised Anxiety Disorder (GAD 7) identifies anxiety symptoms.

Results: 132/185 (71%) adults completed the PHQ 9 and 126/185 (68%) completed the GAD 7. Psychological symptoms of anxiety, depression or both were identified in 48/132 (36%) of patients. Symptoms of depression (score ≥ 5) were reported in 41/132 (31%) of patients, of which 23 (17%) reported moderate or severe depressive symptoms (score ≥ 10). Symptoms of anxiety (score ≥ 6) were reported in 34/126 (27%) of patients, of which, 17 (13%) reported moderate or high anxiety symptoms (score ≥10). Anxiety and depressive symptoms were significantly correlated (Pearson correlation: 0.78; p<0.001). No significant correlation was found between lung function, body mass index (BMI) or age and symptoms of anxiety and depression.

Conclusion: We found that 36% of the patient cohort require mental health monitoring or support, and that anxiety and depressive symptoms are significantly correlated. It is suspected that prevalence may be greater than 36%, due to under-reporting of symptoms and not capturing patients who do not regularly attend clinic. No significant correlation was found between physical parameters and symptoms of anxiety and depression. This finding highlights the pitfall of underestimating mental health difficulties of patients who are physically ‘well’, and the need for non-targeted screening. Future work should develop mental health support pathways within the hospital and community.
SOCIAL MEDIA: EVALUATION OF A CYSTIC FIBROSIS CLINICAL SERVICE FACEBOOK GROUP FOR COMMUNICATION WITH FAMILIES

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Background: The Cystic Fibrosis (CF) team at The Children’s Hospital at Westmead (CHW) have been running a closed Facebook group for CF clinic families since May 2017. The group is a platform for sharing service updates, education resources, research and fundraising. Two years on from launching the group, we explored the uptake, member engagement, popular content, benefits, challenges and areas for improvement.

Method: A 3-pronged evaluation was conducted using Facebook analytics and a patient and staff satisfaction survey. Satisfaction survey results were compared with a pre-evaluation survey, completed in 2017.

Results: The CF CHW Facebook group has 262 members, reaching 75% of families. 52/262 (20%) members completed the satisfaction survey. Positive feedback was received with 37/52 (72%) rating the group great or excellent and 46/52 (88%) reporting to feel somewhat or more engaged with the CF service. One family commented that the page is an “easy way to feel part of the CF community and get information”. The percentage of families feeling informed with clinic activities doubled since the 2017 pre-evaluation survey to 26/52 (50%). Popular content according to Facebook analytics & survey responses included clinic service information, nutrition, research and physiotherapy. Average posts per week were 3-4 from administrators and 5-6 from members. Posts with photos of patients or staff received 2.9 times more ‘likes’. Administrators reported to spend on average, less than one hour per week on content development and monitoring. Perceived benefits included fundraising and easy dissemination of information/resources to families. ‘Lack of time’ was identified as a barrier to further content development.

Conclusion: The CF CHW Facebook group has been successful in connecting CF families and improving their sense of engagement with the CF service. It has also proven to be a suitable platform for communication and information sharing amongst staff and families.
NEW ZEALAND CF COMMUNITY INSIGHT SURVEY 2018

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Background: In 2018 Cystic Fibrosis New Zealand (CFNZ) wanted to refresh the organisation’s strategic direction to better support the CF community.

Aim: To gather information using a Community Insight Survey on the wellbeing of the CF community; the extent to which CF impacts on their quality of life, their aspirations for the future and support needed.

Methods: A customised online survey was developed utilising the NZ General Social Survey and the CF-R Supports Needs Survey. This enabled comparison with the wider NZ population and with other CF populations. The survey went to 405 people with CF (PWCF)/families (of an estimated 505 PWCF in NZ). The response rate of 52% provided statistically reliable data.

Results: Both the ‘self-reported health status’ and ‘satisfaction with life’ of both PWCF and parents was lower than the NZ population. The better the health of PWCF and parents, the more likely they were to be satisfied with life. Parents with very young PWCF were most likely to report being less satisfied with life. There were no significant differences between the health status or satisfaction with life by gender or age for PWCF. Physical issues had more impact than emotional issues on what PWCF wanted to accomplish and their quality of life. Living outside a main centre meant CF impacted more on PWCF lives.

Respondents told CFNZ that their aspirations for PWCF were for:
- Easier and equitable access to the best medicines and treatment for children and adults across the country
- the day-to-day management of CF to be less demanding, expensive and time-consuming
- the day-to-day experience and functioning to be less impacted by CF.

Conclusions: These valuable insights from the CF community have provided a strong direction for CFNZ and its services.
A METACOGNITIVE INTERVENTION OF NARRATIVE IMAGERY (MINI) FOR YOUNG PEOPLE WITH CYSTIC FIBROSIS: A PILOT STUDY

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Background: Recent medical advances have enabled young people with Cystic Fibrosis (CF) to live healthier and longer lives but have continued to require demanding self-care regimens and frequent hospital stays. These treatments intrude into patients’ daily lives, and can trigger negative emotions, family tension and non-adherence to treatment. Development of psychological treatments has not kept pace with these demands. Given the multi-faceted nature of CF challenges, psychological interventions for young people with CF should be holistic and involve the family. This study aimed to gather preliminary evidence for a new integrative psychotherapy for CF to meet these needs (Metacognitive Intervention of Narrative Imagery, MINI), which included narrative and meta-cognitive therapies and mental imagery.

Methods: 13 patients with CF aged 10-17 years were recruited for an uncontrolled pilot study over 8 weeks. MINI comprised three 1-hour sessions over 4 weeks. In Session 3, patients shared the storybook they had created during the treatment with their family. Patients were assessed for anxiety, depression, general emotional state, and self-efficacy at baseline, 4, and 8 weeks.

Results: Patients gave highly favourable rating of sessions and patients and their parents also rated the storybook highly. The intervention resulted in notable reductions in anxiety and improvements in emotional state and self-efficacy.

Conclusions: This pilot study provides preliminary evidence that MINI may offer clinical benefits to young people with CF, although results need confirmation in randomised controlled trials. Since MINI followed a set, structured script, it has potential for wide dissemination.
CONNECTION AND COMMUNITY: STARLIGHT'S LIVEWIRE ONLINE PROGRAM

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Adolescence is a complex time of rapid physical, psychological, and social development. Having a chronic health condition can severely impact this trajectory. Young people in this situation have increased risks of experiencing negative psychosocial outcomes such as lower overall life satisfaction, poorer mental health, and lower self-efficacy. For adolescents, the social isolation created by Cystic Fibrosis can be further compounded by a lack of peers with similar experiences, and the limited availability or access to social support services.

The Starlight Children’s Foundation (Starlight) delivers programs in partnership with health professionals, to support children, adolescents and their families who are living with a serious illness, disability or health condition. After identifying a gap in the availability of online peer support programs for chronically ill adolescents and their siblings, Starlight’s Livewire program was established in 2008.

Livewire Online supports adolescents via an online 24/7 peer community. It offers the opportunity to connect with others who understand the complexity of living with significant health conditions, promoting the voice of lived experience through live chat, private messaging, information provision, community and entertainment content, projects, and blogs. As an online service, Livewire.org has the benefit of being accessible from anywhere, at any time.

Working in partnership with a range of organisations, Livewire provides a safe space for connection and a platform for social interaction, self-expression, understanding, and personal development. Feedback from parents has indicated that they credit Livewire with benefits in the adolescent’s wellbeing, resulting from the opportunity for connection and inclusion, and the cultural encouragement of positive coping mechanisms.

This poster presentation will outline the different components and benefits of the online community for both adolescent with cystic fibrosis and their siblings. It will discuss how Livewire partners with others to help provide additional expertise and support.
IMPLEMENTING AN ADHERENCE PROTOCOL IN THE CYSTIC FIBROSIS CLINIC – A MIXED METHODS IMPLEMENTATION STUDY

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Background: Supporting patients to adhere to their prescribed treatments remains a challenge for health professionals despite advances in the management of cystic fibrosis. The outpatient cystic fibrosis clinic presents an opportunity for clinical teams to address adherence issues. However, opportunities to intervene are often missed due to systemic barriers within the clinic setting.

Comprehensive adherence protocols have been proposed in the cystic fibrosis literature to increase the visibility, consistency and efficiency of adherence work. These protocols embed adherence tools into existing clinic encounters. The aim of this study was to explore the feasibility, appropriateness and acceptability of implementing an adherence protocol into a tertiary, pediatric health care setting.

Methods: This mixed method study was designed using an implementation science approach. Focus groups and qualitative interviews were conducted with staff, patients and parents to explore the barriers and facilitators to adherence work in a CF clinic. This informed modifications to an existing comprehensive adherence protocol (CF MyWay). The local protocol was implemented as routine care for three months within the clinic. Routine auditing and feedback to the clinical team was provided.

Results: Interim analysis of focus group and interview data highlighted key factors that may influence staff, parent and patient behaviour around adherence. From the thematic analysis, themes have begun to emerge including the impact of the clinic environment, the impact of communication and collaborative care planning. Pre-implementation modifications to the CFMyWay protocol included changes to communication systems, localisation of adherence resources and digitisation of assessments. Descriptive analysis and audit data from the implementation are currently being collected.

Discussion: This presentation will describe the process of implementing an adherence protocol into routine care in the cystic fibrosis clinic. The adherence protocol will be presented. Key factors that influence staff fidelity will be shared as well as learnings regarding the use of an implementation science approach.
ACCEPTABILITY OF USING PERSONALISED MINI-ORGAN-AVATARS TO ACCELERATE DRUG DISCOVERY AND PREDICT DRUG RESPONSE IN CYSTIC FIBROSIS PATIENTS

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Background: Individual patient derived pre-clinical cell models (organoids/Avatars) have helped guide drug discovery for a number of diseases including Cystic Fibrosis (CF). This technology may also help guide treatment choices at an individual level using personalised medicine. However, little is known about the acceptability of patient derived cell models to the CF community or the wider population.

Methods: We used a cross sectional online explainer video and questionnaire. Our questionnaire was adapted from a recently published study on willingness to use patient-derived xenografts in cancer care [1]. Acceptability was examined in five domains; willingness to use organoids, perceived advantages and disadvantages of organoids, maximum acceptable out-of-pocket costs, maximum acceptable turnaround time, acceptable source of tissue. Between March and April 2019, participants were recruited via email invitation, in person at CF clinic and using social media.

Findings: 92 individuals completed the online questionnaire. The cohort included CF adults (13%), parents of children under 18 with CF (35%), non-CF parents (29%) and non-CF adults (23%). 98% of respondents found that the advantages of patient derived stem cell models outweighed the disadvantages. Responses were not significantly different between the groups of participants. The most endorsed advantage was that organoids might help doctors choose the right drug more quickly, without having to test drugs on the patient. The least endorsed disadvantage was that the patient may be recommended a treatment that is different to the most common treatment or one that is incompatible with their current effective treatment regimen. 91% of participants chose the nose as the site they would most likely consider being sampled.

Interpretation: Using patient derived organoids is extremely acceptable to the majority of adults who completed the questionnaire. Participants were mostly likely to choose the least invasive procedure to obtain samples.

SOCIAL WORK AT FIRST SIGHT - ESTABLISHMENT OF A SOCIAL WORK OUTPATIENT CF CLINIC

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Background: In 2014 the Cystic Fibrosis Australia Peer Review Report detailed the lack of social work services available to Adult CF patients in Western Australia. Coupled with concern regarding unmet psychosocial needs, an outpatient CF social work clinic was established in a fiscally challenging environment. In January 2017, a two session per week social work CF outpatient clinic commenced, aimed at operating under a self-funding model.

Aim: To outline the benefits and challenges of clinic establishment. We will examine the viability of the clinic, and whether the needs of stakeholders are being met.

Methods: Social work services are offered to patients who are referred by themselves, the CF treating team or identified by the social worker as likely to benefit from input. The clinic targets patients transitioning from paediatric care, patients struggling with treatment adherence, and patients with complex psycho-social needs.

Results: The clinic has been utilised by 22% of the hospital’s CF patient cohort. 2% of patients self-referred, 4% were referred by the paediatric social worker during transition, and 93% were referred by the multi-disciplinary team, with doctors being the greatest referrers. 5% of patients identified as Aboriginal and 60% of all patients were female. Patients aged 19 to 28 years comprised 44% of the clinic cohort and had the highest number of contacts with the social worker. Over the period of the study, the average time of social work contact varied from 2.5 to 7.3 hours per patient over all age groups.

Conclusion: This CF social work clinic is currently utilised by 1 in 5 patients. Greatest use of the social work clinic has been by patients aged 19 to 28 years. The service primarily provides psychosocial support and counselling interventions in relation to treatment adherence. Yet to be undertaken is a satisfaction survey of this patient group.
CYSTIC FIBROSIS AND FAMILY ACTIVITY: PILOT STUDY EVALUATING THE EFFECTS OF A SURFING INTERVENTION ON HEALTH AND WELLBEING

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**Introduction:** Diagnosis and treatment of Cystic Fibrosis (CF) can cause emotional stress and physical burden on families. Physical activity in natural environments can have a beneficial effect on physical and psychological health. Surfing is a playful, ocean-based activity that may benefit patients with CF. Family-based interventions built around surfing may potentially facilitate the positive physical and psychological wellbeing of families with CF.

**Aim:** To assess the feasibility of innovative a family activity interventions built around surfing for families with children with CF. Compare surfing with another outdoor activity, golfing, and no intervention.

**Method:**
- A randomised, controlled, parallel group pilot study.
- Participants: Children with CF and their families.
- Three groups were compared: surfing (intervention), golfing (comparison), waitlist control.
- Interventions: 15 x 1 hour lessons of surfing or golfing over 7 weeks.
- Outcome measures: CFQ-R, Pediatric Inventory Parents (PIP), Habitual Activity Evaluation Scale (HAES), lung function (FEV1), evaluation questions.
- Questionnaires were administered pre- and post-intervention, 3-months after.

**Results:** Fifteen families participated (5 families per group), with a total of 16 children with CF (f=6, m=10), median age = 9 (range =7-18 years). Nine out of 10 families completed the 15 lessons. One family withdrew from golfing due to the child with CF becoming unwell during the course of the study. HAES baseline median activity was “active” (range: “somewhat inactive” to “very active”). Baseline FEV1 median = 99.4% (range = 83.1-117.5) With preliminary results no significant change in FEV1 was seen.

Preliminary data suggest trends toward improvement in CFQ-R, Emotional State domain, and reduction in perception of difficulty of care from the PIP, for surfing intervention.

Greater positive feedback by families for surfing compared with golfing and intent to continue surfing, post study.

**Discussion:** Preliminary results suggest that family activity interventions built around surfing are feasible and beneficial for families with children with CF.