CYSTIC FIBROSIS Australia

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A001

CHILDRENS HOSPITAL IN THE HOME FOR CYSTIC FIBROSIS: EVOLVING ROLE OF THE NURSE PRACTITIONER

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Introduction/background: Gold Coast HHS set up a new Children's Hospital in the Home (CHITH) service in late 2018. Cystic fibrosis patients were the majority cohort admitted on this program, however no formal pathways for review during or at completion of treatment were established.

Methods: The nurse practitioner who case manages this cohort collaborated with the CHITH team about the gap in service, identifying who was the most appropriate staff member with availability to manage these admissions. It was identified that the nurse practitioner had the appropriate skills and knowledge to collaboratively manage clinical assessments, reviews, CVAD cares, therapeutic drug management and discharge planning for this cohort. Retrospective audits were carried out to compare this model of care to standard inpatient care.

Results: The CHITH service, with NP collaborative management was non-inferior to inpatient admission, based on lung function, weight and length of stay.

Conclusion: Within this cohort in this HHS, collaborative NP-led care is safe and effective for children admitted to hospital in the home. Nurse practitioner models of care can and should evolve to meet the needs of patients in a complex and ever-changing health care system.

A002

CYSTIC FIBROSIS RELATED DIABETES SCREENING

SCREENING FOR CYSTIC FIBROSIS RELATED DIABETES AT PERTH CHILDREN'S HOSPITAL

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Background and aim: There is considerable variation in recommendations from clinical practice guidelines on when and how to screen for glucose intolerance in Cystic fibrosis (CF). The Australian Standards of Care suggest oral glucose tolerance tests (OGTT) screening for adolescents with risk factors such as unexplained weight loss or growth failure. During 2019, 26% of patients with CF cared for at Perth Children's Hospital, who were 10 years or older had OGTT and 86% had HbA1C measurements.

Research method: A prospective cross-sectional study was performed on 88 patients in the CF clinic, older than 10 years, who are not prescribed insulin or prolonged oral/parenteral steroids. Patients who had not had an OGTT in 2020, were sent a letter in June with instructions on how to organise an OGTT. Results were followed up weekly.

Results: Prior to June, 10 patients had an OGTT. One had impaired glucose tolerance (IGT). Post June, 50 OGTT were performed. Four patients had a positive OGTT suggestive of CF related diabetes (CFRD) and nine had results suggestive of IGT. The patients with results suggestive of CFRD, were reviewed by Endocrinology. One patient was determined not have CFRD, two were started on insulin and the last is being monitored.

In terms of risk factors, the 14 patients with abnormal results, five had suboptimal nutrition, one had undernutrition, and two had overnutrition. None had changes in respiratory status or unexplained weight loss.

Conclusions: Patients with CF can have abnormal OGTT without obvious nutritional risk factors. The Australian Standards of Care may require revision.

A006

AWARENESS, EXPERIENCES AND PERCEPTIONS OF TELEHEALTH IN PEOPLE WITH CYSTIC FIBROSIS DURING COVID-19 PANDEMIC

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Introduction: In Australia telehealth for management of cystic fibrosis (CF) has been predominantly used for outreach care in regional and rural areas. However, in March 2020, the nationwide restrictions imposed by the COVID-19 pandemic necessitated the abrupt and unprecedented transition of CF care almost exclusively to telehealth. As a result, face-to-face clinic visits were replaced by telehealth visits for a majority of patients across CF centers. The aim of our study was to explore perceptions, experiences and awareness of telehealth services of people with CF and their caregivers during COVID-19 pandemic.

Methods: Semi-structured qualitative interviews were conducted with people with CF or their caregivers. An interview guide was developed that focused on experiences of telehealth as well as clinical data collection via telehealth services. Data were audio-recorded and transcribed. We analyzed data into topics and subtopics using conventional content analysis.

Results: Total of 15 participants, six people with CF and nine caregivers, were interviewed. All participants were adults, two of them were male. The mean (SD) age of people with CF was 40.2 (6.9) years. Caregivers' mean (SD) age was 41.8 (6.7) years. In general, most of the study participants thought that telehealth services were as effective as clinic visits. They also believed that telehealth services were more accessible during COVID-19 compared to the routine clinic visits. Conversely, some participants expressed concerns for the disadvantages in telehealth services, including lack of physical examination, access to hospital equipment, and immediate admission to hospital if necessary.

Conclusion: Participants highlighted that telehealth was an excellent option to monitor their conditions; however, meeting clinicians face-to-face in a clinic was still essential. More research is needed to compare standards of care and clinical outcomes for people with CF using telehealth versus clinic-based care.

A012

THE AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY (ACFDR). MAXIMISING REGISTRY DATA VALUE REQUIRES OPTIMISING DATA QUALITY

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Introduction: *Clinical registries* are an *important* tool for understanding disease trajectory, processes of care, and longer-term clinical outcomes. Increasingly, clinical registry data are driving quality improvement and research. A number of processes have been established by the ACFDR to optimise data quality, increasing its value as a useful clinical data resource.

Method: Since the introduction of the ACFDR Data Quality Assurance (QA) program in 2019, each participating centre is required to complete a Minimum Data Set (MDS) for the collection of core data (registration, demographics, genetic mutations, CFTR modulators, transplants), quarterly data (clinical measures, hospitalisations/IV Abx and quarterly sign-off) and annual data (annual general update and sign-off, complications/treatments). Data is captured via direct entry or through template upload obtained directly from the patient's health records, which aids in the timely entry of bulk data. In-built validation and completion checks in the registry also optimise data capture. To minimise data entry error, quarterly site sign off is required, with spot data cleaning maintained through the year by registry staff.

Results: Through the implementation of the Data QA program, the data quality of the ACFDR has been significant improved. Tangible reductions include removing duplicate data entry, biologically unfeasible data, minimising data inconsistency and missing data. These improvements have resulted in the highest completion rate of data collection to date at 96% for the 2020 survey year, up from 94% in 2019 and 88% in 2018.

Conclusion: To ensure data recorded in the registry accurately reflects the health records, continued improvement is required. The implementation of the quality and quarterly checks have been beneficial in improving data quality and completeness and will be an ongoing feature of the ACFDR. Moving forward, onsite data auditing is being piloted in 2021 to further enhance data accuracy and identify potential systematic areas for ongoing improvement.

A016

IMPROVING PROCESSES TO ASSIST CYSTIC FIBROSIS-RELATED DIABETES (CFRD) SCREENING & CFRD COMPLICATION SCREENING, IN LINE WITH BEST PRACTICE, AT AN ADULT CF CENTRE. – A QUALITY IMPROVEMENT ACTIVITY.

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Introduction/Background/Aim: The life expectancy of patients with cystic fibrosis (CF) is improving; yet in the future endocrine complications may become a significant morbidity. Optimal management of CF-related diabetes relies on timely screening, diagnosis, and management. This quality improvement activity firstly audited screening and complication screening rates; secondly explored current barriers and enablers to screening using a theoretical domains framework (TDF) and COM-B behaviour change ('capability', 'opportunity', 'motivation' and 'behaviour') model.

Method(s): A retrospective audit of CFRD screening & complication screening was undertaken for a two-year period 2017-2019. A staff survey was developed and conducted (using TDF validated questionnaire items) to assess barriers to screening and complication screening with key members of the CF MDT, followed by solution mapping workshops to identify interventions to address barriers and using the COM-B model to plan evidence based behaviour change techniques in 3 healthcare areas.

Results: In non CFRD patients, the number of adults attending the Adult CF Centre with an OGTT completed in past 2 years (2017-2019) was 44 / 202 (22%); CFRD complication screening was recorded as complete in 41% of diabetics (24/58) with 6/64 diabetics exempt from screening due to CFRD < 5 years The staff survey assessing barriers identified 3 main healthcare areas: staff knowledge / training , systematic processes & patient education Solution mapping workshops was used to plan interventions based on the COM-B model of evidence based behaviour change techniques.

Conclusion: This multifaceted quality activity assesses healthcare professionals' behaviours in relation to current practice of OGTT screening and diabetic complication screening practices at ACFC, and identifies key areas for change via staff education, systematic process improvements and patient / consumer education. Implementation of the changes to healthcare practices and screening rates will be reviewed annually from January 2022.

A019

ADHERENCE TRENDS IN ADULT CYSTIC FIBROSIS POPULATION BEFORE AND AFTER INITIATION OF CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE REGULATOR MODULATOR THERAPY

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Background: Adherence to maintenance Cystic Fibrosis (CF) medications is low due to a range of social and economic factors despite evidence that medication adherence improves CF patient outcomes, thereby reducing the burden on the healthcare system. CF Transmembrane Conductance Regulator (CFTR} modulator medications are novel in the management of CF in Australia. CFTR modulators currently available on the Pharmaceutical Benefits Scheme (PBS) include Kalydeco, Orkambi and Symdeko. In addition, select patients can access Trikafta on a compassionate basis. The aim of this study was to evaluate medication adherence to CFTR modulators and maintenance CF medications using hospital dispensing records in adult CF patients.

Method: A retrospective quality improvement audit of CF patients prescribed a CFTR modulator for greater than 12 months at a metropolitan tertiary referral hospital (n=141) was conducted. Patients who did not reside in Brisbane or accessed Trikafta through a clinical trial were excluded. Dispensing records of five commonly prescribed non-PBS CF maintenance medications (Vitabdeck, salt tablets, azithromycin, hypertonic saline 6% nebules and dornase alfa nebules) and CFTR modulators were obtained and analysed for the 12 months prior to CFTR modulator initiation to present day.

Results: Dispensing of non-PBS CF maintenance medications was low (average annual dispensing range O - 7.6) across all groups suggesting non-adherence. In contrast, CFTR modulator dispensing was high (average annual dispensing 13); however, this may be due to sponsored delivery of CFTR modulators by the drug company or the patient's perceived benefit from CFTR modulators. For patients prescribed Kalydeco, Symdeko or Trikafta, annual dispensing for three of the five CF maintenance medications reduced by an average of 26% after CFTR modulator commencement. In patients prescribed Orkambi, annual dispensing counts for two of the five CF maintenance medications reduced by an average of 4% after commencement.

Conclusion: Medication dispensing counts suggest adherence to maintenance CF medications in the adult CF population is low, while adherence to CFTR modulators is high. Trends suggest medication adherence may reduce after CFTR modulator initiation.

A020

EVALUATION OF THE LONG-STAY NATIONAL INPATIENT MEDICAL CHART FOR CYSTIC FIBROSIS PATIENTS ADMITTED TO AN ADULT CYSTIC FIRBOSIS WARD

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Background: Due to the progressive nature of cystic fibrosis (CF), hospitalisation is often needed, with the most common cause being CF-related pulmonary exacerbations. Typically, this is managed with a course of intravenous antibiotics for 14 days. Currently, short-stay National Inpatient Medication Charts (NIMC) are used at a metropolitan, tertiary referral hospital adult CF ward, which are designed for 10-day admissions.

Pre-audit data showed medication chart rewrites took an average of nine minutes and thirteen seconds for one chart, and each CF patient required approximately three medication charts. This study aimed to assess medication safety of the long-stay NIMC for CF patients admitted to this ward.

Method: Over three months, CF patients admitted to the ward were alternatively assigned short-stay or long-stay NIMCs for use during admission. The standardised National Safety Medication Chart (NSMC) audit form was used to audit these charts. The data was analysed with the NSMC Audit System.

Results: There were 14 long-stay and 16 short-stay NIMCs. The medicine orders prescribed on the long-stay NIMC were complete and correct 80% of the time, compared to 70% in the short-stay group. The documentation of medication administration by nursing staff was documented correctly 99.7% in the long-stay group, compared to 99.4% in the short-stay group. The venous thromboembolism (VTE) prophylaxis section of the short-stay NIMC was completed 50% of the time. Long-stay charts do not contain a VTE prophylaxis section, but VTE prophylaxis was documented within medical notes in 64% of patients allocated long-stay NIMCs. Long- .

Conclusions: Overall, the results demonstrate that the implementation of the long-stay NIMC within an adult CF ward at a tertiary referral hospital did not negatively affect medication safety, medication administration or VTE prophylaxis documentation. The implementation of the long-stay NIMC will save a significant amount of time and resources for staff.

A021

PRESCRIBING TRENDS IN ADULT CYSTIC FIBROSIS POPULATION

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Background: General practitioners (GP) play an important role as part of the multidisciplinary care required for Cystic Fibrosis (CF) patients. CF patients can access many services from their GP, including the management of non-CF related medical conditions or health concerns as well as prescribing CF medications that are listed on the Pharmaceutical Benefit Scheme (PBS) and available at community pharmacies. The aim of this audit was to review the appropriateness of PBS and non-PBS medication prescribing in the hospital outpatient setting for CF patients.

Method: Electronic prescriptions for outpatient CF patients at a tertiary referral hospital written between August and October 2020 were identified using prescribing software. The prescriber, items prescribed, and origin of the script request were recorded. Handwritten and discharge prescriptions were not included in this audit.

Results: Over three months 285 electronic prescriptions were generated, for a total of 890 medications. PBS items accounted for 54% of medications, of these 69% could have been prescribed by a GP. Of all the PBS medications prescribed, 90% were for CF related conditions whilst 10% were for non-CF related conditions. Non-CF related prescriptions included antidepressants (22%), antiepileptics (6%), antipsychotics (3%), rheumatoid medications (3%), antihypertensives (2%) and diuretics (2%). Of all the non-PBS medications prescribed 99.3% were for CF related conditions.

Conclusions: Increased GP prescribing may reduce the burden on CF specialists in clinic as well as ensuring there are appropriate safety checks and oversight of the prescribing of these medications to optimise patient's health outcomes.

A022

THE ROLE OF THE AUSTRALIAN CYSTIC FIBROSIS (CF) PHARMACIST AND PHARMACY TECHNICIANS

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Introduction : The CF Pharmacist is an integral part of the CF multidisciplinary team (MDT), as endorsed in the United States and European CF guidelines. CF medication regimens can be complex, which contributes to high rates of nonadherence. Australian Hospital Pharmacists, with the support of Pharmacy Technicians, are well placed to support these patients by providing education and tailoring medication regimens to meet their individual needs.

Method : The role of the CF Pharmacist has been well described in European and United States CF Centres, but currently there is no information available detailing the Australian CF Pharmacist's role. The aim of this presentation is to characterise the activities undertaken by CF Pharmacists and Pharmacy Technicians at an Australian metropolitan, tertiary referral centre.

Results : The CF Pharmacist works in both the inpatient and outpatient setting, contributes to MDT meetings and attends Consultant ward rounds. In March 2021, 113 outpatients were seen by a CF Pharmacist, averaging 7 patients per CF outpatient clinic. In March 2021, 442 CF and general thoracic inpatients were seen by a CF Pharmacist, averaging 21 clinical reviews daily. Common roles/responsibilities of the CF Pharmacist include medication history taking/reconciliation, prescription monitoring, facilitation and dispensing, medication review, local policy/procedure production, patient education, therapeutic drug monitoring, contribution to CF research and track PBS applications for high-cost CFTR (CF transmembrane conductance regulator) modulators. At the peak of the COVID-19 pandemic in 2020, a drive-through influenza vaccination clinic and medication collection service were established. Currently, two Pharmacy Technicians support the CF pharmacist by dispensing, supplying imprest to CF ward, updating medications and allergies in the CF database, bill clinic consultations, complete PBS renewal forms for CFTR modulators and assist with data collection for CF research.

Conclusion : The CF Pharmacist and Pharmacy Technicians are well placed in the Australian hospital setting to support CF patients through their medication journey.

A023

THE CLINICAL IMPACT OF TRIPLE CFTR MODULATOR THERAPY IN SEVERE CYSTIC FIBROSIS LUNG DISEASE

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Aim: To assess the response to treatment with elexacaftor/tezacaftor/ivacaftor (elex/tez/iva) on adult patients with severe lung disease (FEV_1<40% predicted) on lung function, exacerbations, weight, exercise tolerance and adverse effects.

Method: This was a prospective observational study of adult patients, previously treated with CFTR modulator therapy, who were able to gain access to elex/tez/iva via managed access program. Six patients who are homozygous F508del with a FEV_1 < 40% participated in this study. The following parameters were measured before and 6 months after initiation of therapy, lung function by spirometry, weight, 6 minute walk test (6MWT) and bloods to monitor liver function. A comparison of the number of pulmonary exacerbations 6 months prior and 6 months post elex/tez/iva treatment initiation was also assessed.

Results: At baseline patients had a mean FEV_1 33.5%, weight 59.8 kg, and 6MWT distance of 546.7 m. There was a significant improvement after 6 months of elex/tez/iva therapy, mean FEV_1 45.33% (p=0.0027), weight 68.78 kg (p=0.0043), and 6MWT 582.8 m (p=0.0014). There was also a decrease in the number of exacerbations requiring antibiotic treatment from a mean 1.42 to 0.50 (p=0.0019). One patient had significant adverse effects including abdominal pain, diarrhoea and headache, but decreased in severity after 4 weeks. Two patients had raised liver enzymes, however were not significant enough to cease therapy.

Conclusion: In patients with severe lung disease, changing to elex/tez/iva therapy has resulted in a significant improvement in FEV_1, weight, and exercise tolerance. There was also a decrease in the number of exacerbations which demonstrates a positive outcome of this therapy. These patients will be continued to be followed up to further assess the long term impact of triple combination therapy.

A031

PREFERRED HEALTH OUTCOME STATES FOLLOWING TREATMENT FOR PULMONARY EXACERBATIONS

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Background: Treatment for pulmonary exacerbations of cystic fibrosis (CF) can produce a range of positive and negative outcomes. Understanding which of these outcomes are achievable and desirable to people affected by disease is critical to agreeing to goals of therapy and determining endpoints for trials. The relative importance of outcomes resulting from treatment of these episodes have not been reported. We aimed to (i) quantify the relative importance of outcomes resulting from treatment for pulmonary exacerbations and (ii) develop patient and proxy carer-reported weighted outcome measures for use in adults and children, respectively.

Methods: An online discrete choice experiment (DCE) survey was conducted. Participants were asked to make a series of hypothetical decisions about treatment for pulmonary exacerbations to assess how they make trade-offs between different attributes of health. Data were analysed using a conditional logit regression model. The correlation coefficients from these data were rescaled to enable generation of a composite health outcome score between 0 and 100 (worst to best health state).

Results: 362 individuals participated (167 people with CF and 195 carers); of these, 206 completed the survey (56.9%). Most respondents were female and resided in Australia. Difficult/painful breathing had the greatest impact on the preferred health state among people with CF and carers alike. Avoidance of gastrointestinal problems also heavily influenced decision-making.

Conclusions: These data should be considered when making treatment decisions for pulmonary exacerbations of CF and determining endpoints for trials. Further research is recommended to quantify the preferences of children and whether these align with those of their carer(s).

A032

COMPUTER MODELLING THE ROOT CAUSE OF RARE CYSTIC FIBROSIS MUTATIONS

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Introduction: Cystic Fibrosis (CF) disease results when an individual has a mutation in both alleles of their Cystic Fibrosis Transmembrane Conductance Regulator (*CFTR*) gene. There are over 300 mutations known to be disease causing [1]. Currently, 40% of the Australian CF population carry one or more of the rare *CFTR* mutations, which are without clinically approved targeted therapies.

The CFTR protein is a chloride channel with a complicated gating mechanism which involves the rearrangement of the channel's disordered R-domain and the binding of ATP. If any one of these functions are affected by a mutation it can cause CF.

In 2018, the atomic structure of the activated, human CFTR chloride channel was resolved [2] This presents an opportunity to simulate the motion of the CFTR protein with an in silico technique known as Molecular Dynamics (MD). With this computational modelling we can investigate why specific mutations to CFTR cause it to misfunction.

Aim: With the use of computer modelling, we aim to characterise all rare CF-causing mutations in the Australian population and help determine which ones would be responsive to existing drugs.

Method(s): We used MD, to simulate the motion of mutant CFTR proteins. By comparing the motion of the mutant CFTR to the healthy one we investigated what structural defects were introduced in the mutant protein.

Results: To date, we have resolved the pathogenesis of 6 rare CF-causing mutations. We discovered that the I37R- and S945L-CFTR mutants produce conformational changes which would effect the rearrangement of the R-domain during the gating cycle. R352Q impedes the motion of chloride through the channel, while G551D and Q1291H/F mutants disrupt the binding of ATP.

Conclusion: With the use of MD we can work out the mechanism of dysfunction of rare CF at the molecular level. This work paves the way to patient specific treatment regimens for rare CFTR mutations.

References

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A033

ELEXACAFATOR-TEZACAFTOR-IVACAFTOR IS BENEFICIAL, WELL TOLERATED AND SAFE FOR CYSTIC FIBROSIS INDIVIDUALS WITH SEVERE END STAGE LUNG DISEASE WITH A PHE508DEL-MINIMAL FUNCTION GENOTYPE

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Background: Elexacaftor-tezacaftor-ivacaftor (ELX-TEZ-IVA) has efficacy in adult CF individuals who have a single Phe508del-*CFTR* mutation and a minimal function (MF) mutation with ppFEV1 40-90%. However, the benefits in those with severe lung disease is not as well characterised. This study evaluated the use of ELX-TEZ-IVA in all eligible adult Phe508del-MF individuals with ppFEV1 <40% at baseline and after 26 weeks of treatment under a single centre managed access program.

Methods: Eligible individuals were evaluated to determine suitability to commence treatment. Patients started on ELX-TEZ-IVA were assessed at baseline (February-March 2020) and after 26 +/- 2 weeks across multiple clinical domains, including changes in pulmonary function, treatment of pulmonary exacerbations, weight, nutritional status, body composition, quality of life and exercise.

Results: Ten patients aged 19-51 years, with Phe508del-MF mutations and ppFEV1 <40% commenced ELX-TEZ-IVA within this cohort. One patient stopped treatment after fourteen days following lung transplantation and the rest remain on treatment to date with no significant side-effects noted. Significant improvements in lung function were demonstrated, with a median change in ppFEV1 of 10% (p=0.009). Intravenous antibiotic days were significantly reduced: median days pre-treatment of 41 to 0 days post-treatment (p=0.03) with 55.6% not requiring any intravenous antibiotic therapy after 26 weeks of treatment. Body composition parameters significantly improved, with median body mass index increasing from 19.3kg/m^2 to 21.6kg/m^2 (p=0.004) and fat-free mass (FFM) from 46.7kg to 50.1kg (p=0.009). The proportion with FFM depletion decreased from 56% to 33%. Quality of life assessed using the Alfred Wellness Score for CF (AWeScore) significantly improved from a median of 62 to 87 (p=0.009).

Discussion: This real-world evaluation study highlights significant benefits across multiple domains post ELX-TEZ-IVA commenced in severe lung disease. Furthermore, the benefits enabled two patients to be removed from the lung transplant waiting list.

A036

PREVALENCE OF TRACHEOBRONCHOMALACIA IS HIGHER THAN PREVIOUSLY REPORTED IN CHILDREN WITH CYSTIC FIBROSIS

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Introduction: Tracheobronchomalacia (TBM) is estimated to be present in 1 in 2100 children. Previous reports suggest the incidence is higher in children with cystic fibrosis (CF) with a prevalence of 6-17%. This has clinical implications with potential to influence airway clearance and lung health.

Aim: To determine the prevalence and clinical associations of TBM in Western Australian children with CF.

Method: Children born between 2001 and 2016 consented to the AREST CF Surveillance Study were included. Operation reports from bronchoscopies performed until the age of four were retrospectively reviewed. Data was collected on designation of bronchoscopy surgeon; free breathing observation during the procedure; the presence of TBM; the persistence of TBM defined as present on more than one bronchoscopy; and its severity. Data on genotype, pancreatic status and symptoms at diagnosis of CF was extracted from the AREST CF database. Associations between categorical variables were compared using Chi Square and continuous variables by Student's t-test.

Results: Of 180 children screened, 167 contributed data. 706 bronchoscopies were undertaken by 43 physicians. Observation of airways in free breathing was reported in 16% of bronchoscopies. Sixty-eight (41%) children had TBM reported on bronchoscopy at least once with 36 (53%) persistent. TBM was reported as moderate/severe in 31 (46%). TBM was associated with homozygous ΔF508 genotype (p=0.017), pancreatic insufficiency (p=0.006) and meconium ileus (p=0.020). Those with TBM were less likely to be asymptomatic at diagnosis (p =0 .006) and more likely to have gastro-intestinal (p=0.001) than respiratory symptoms (p=0.540).

Conclusion: The prevalence of TBM in this cohort is higher than previously described and persistent across multiple bronchoscopies. Gastro-intestinal manifestations of CF and homozygous ΔF508 genotype are associated with the presence of TBM. Documentation of observation of airways during free breathing was low which may have influenced TBM prevalence and severity.

A037

IN ADULTS WITH CF, AIRWAY CLEARANCE TECHNIQUES MAY BE MORE EFFECTIVE THAN COUGH ALONE AT OPTIMISING SPUTUM CLEARANCE: A SYSTEMATIC REVIEW AND META-REGRESSION

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Introduction: Airway clearance techniques (ACT) are standard care for people with CF. However, a meta-analyses of data reported in randomised controlled trials and randomised cross-over trials are not possible due to methodological heterogeneity in the interventions and outcome measures used within these studies. Therefore, the aim of this study was, in adults with CF, to estimate the magnitude of any within-group change in outcome measures used to assess the immediate effects of ACT and determine possible moderators of these effects.

Method(s): Databases were searched to find studies that reported outcomes in adults with CF within 60 minutes of completing ACT. Studies were grouped according to the mechanism of action of ACT (positive expiratory pressure with or without oscillation [PEP] vs. positive pressure [PPD] vs. other techniques [OT] vs. cough alone [C]). The within-group change in commonly reported outcome measures and possible moderators of the change(s) were examined via three-level meta-regression.

Results: Data were extracted from 65 studies (1,120 participants). The most frequently reported outcomes were sputum expectoration (n=46, 71%) and spirometric lung function (n=40, 62%). The pooled estimate of wet weight sputum following any ACT was 12.5g (95% confidence interval [CI] 9.1 to 15.8; n=28 studies) and studies which used PPD, PEP and OT produced greater wet weight sputum compared with C alone ($F_{3,62}$=3.0; p=0.04). Sputum wet weight was not influenced by the clinical stability of participants. The pooled estimate for change in FEV_1 following any ACT was 0.03L (-0.2 to 0.2; n=13 studies). These estimates were affected neither by ACT mechanism of action nor the clinical stability of the study participants.

Conclusion: Sputum wet weight was greater in studies that investigated PPD, PEP and OT than following cough alone. Although commonly used to evaluate immediate effects of ACT, FEV_1 presented a trivial change, if any, following ACT.

A038

MEASURES OF RESPIRATORY MECHANICS AND VENTILATION INHOMOGENEITY DURING AN EXACERBATION IN ADULTS WITH CF

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Introduction/Background/Aim: Lung clearance index (LCI) and airway resistance (Rrs5) and reactance (Xrs5) at 5Hz are emerging clinical outcomes in adults with CF. Measured using tidal breathing, these are thought to be better tolerated during an exacerbation than traditional assessments. The aims of this study were, in adults during an exacerbation, to i) report on ventilation inhomogeneity (LCI) and respiratory mechanics (Rrs5, Xrs5), and ii) investigate associations between these outcome measures and the current gold standard (FEV_1).

Method(s): Participants were recruited within 48 hours of admission with a CF exacerbation to Sir Charles Gairdner Hospital. Upon recruitment participants completed the Alfred Wellness Score for CF (AweScore), multiple breath washout (MBW; LCI), forced oscillation technique (FOT; Rrs5, Xrs5) and spirometry (FEV_1). Associations were explored using Pearson correlation coefficients, ANOVA and multiple linear regression.

Results: Sixteen participants (median [interquartile range] age 26yr [21 to 32], AweScore 47 [35 to 54], ppFEV_1 57% [34 to 79]) were recruited. The mean±SD LCI was 11.7±2.2 turnovers and z-scores for Rrs5 and Xrs5 were 1.58±1.39 and -1.94±0.75, respectively. Linear correlations were noted between FEV_1 (L) and LCI ($r=-0.74$, $p<0.01$), Rrs5 z-score ($r=-0.62$, $p<0.01$), Xrs5 z-score ($r=0.57$, $p=0.02$) and Rrs5-19 ($r=-0.59$, $p=0.02$). Multiple linear regression showed a significant association between FEV_1 (L) and measures obtained via MBW, FOT and the AweScore. Forced expiratory volume in 1 second measured in litres could be estimated using the following equation:

0.39 + (1.32×FRC [L]) - (0.64×MBWtime [min]) + (0.34×Xrs5) + (0.02×AweScore); adjusted $R^2 = 0.85$.

Conclusion: In our sample, during exacerbation, 85% of the variability in FEV_1 could be explained using measures obtained from MBW, FOT and AweScore. These data are preliminary, but suggest that, during exacerbations when spirometry is poorly tolerated, MBW and FOT outcomes may constitute useful alternatives to FEV_1.

A043

RETROSPECTIVE STUDY TO MEASURE THE TIMELINE PROGRESSION OF IMPAIRED GLUCOSE TOLERANCE TO CYSTIC FIBROSIS RELATED DIABETES

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Aim: Children with cystic fibrosis (CF) are at risk of developing impaired glucose tolerance (IGT) and cystic fibrosis related diabetes (CFRD) due to pancreatic scarring over time. There is limited data on the progression of patients who develop CFRD from those with IGT. The primary objective is to measure the time period from when patients progress from IGT to CFRD in a CF cohort.

Method: A retrospective study was conducted in patients with cystic fibrosis at a single tertiary centre (Women's and Children's Hospital) with a diagnosis of IGT and CFRD. Patients with CF undertake an Oral glucose tolerance test (OGTT) as part of routine care annually from 10 years. CFRD and IGT are diagnosed based on the OGTT results. OGTT results were obtained from medical records and electronic medical record.

Results: 11 patients had a diagnosis of CFRD (6 females – 55%). 5 of these patients were diagnosed with CFRD without prior IGT state, with a diagnosis of CFRD ranging from 6 – 14 years of age. 6 of the 11 patients had a mean duration of IGT to CFRD of 3 years (range 2-4 years), 83% (5 out of 6) of whom had homozygous DF508 genotypes.

Conclusion: These results indicate that the period of transition from IGT to CFRD is between 2-4 years. Patients with the homozygous DF508 deletions are at increased risk.

A046

REAL LIFE OUTCOMES OF IVACAFTOR IN AUSTRALASIAN PATIENTS WITH CYSTIC FIBROSIS (CF) WITH AND WITHOUT IVACAFTOR TREATMENT: DATA FROM THE AUSTRALIAN CF DATA REGISTRY AND THE NEW ZEALAND PORT CF REGISTRY 2014-2018.

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Introduction/Background/Aim: Ivacaftor is the first CFTR modulator accessible to patients with CF [1]. Clinical trials showed Ivacaftor-treated patients experienced short to medium term improvement in lung function and nutritional status. Ivacaftor was available for public access to Australian patients in 2014 and New Zealand patients in 2020. Evaluation of the real-life longitudinal impact of Ivacaftor requires a comparable cohort. This study aims to assess the real-life outcomes amongst Australasian patients with comparable CF gating mutations with and without Ivacaftor treatment between 2014-2018.

Method(s): Clinical data were extracted from the Australian CF data registry and New Zealand PortCF registry (2014-2018). All patients with gating mutations eligible for Ivacaftor were included. Clinical parameters include demographics; lung function; body mass index (BMI) and Ivacaftor treatment were included. Between-group comparison between Ivacaftor-treated vs untreated patients was performed.

Results: A total of 357 patients were identified from the Australian and New Zealand CF data registries, comprising 208 patients from Australia and 49 from New Zealand. During the study period, 182/208 (87.5%) Australian patients were on Ivacaftor, whereas all NZ remained untreated. At the end of the study (2018), significant between-group differences in age, lung function and weight were observed. Whilst Ivacaftor-treated patients were significantly older (24.5±12.5 vs 19.7±14.3 years; $p<0.05$), they have significantly higher lung function (Mean FEV1% predicted: 83.1±22.7 % vs 73.5 ±28.6 %; $p<0.001$); and BMI (24.5+/-12.5 kg/m^2 vs 19.3 ± 4.0kg/m^2 ; $p<0.05$) compared to untreated patients.

Conclusion: Our study has provided data on the real-life long-term outcome of Ivacaftor amongst Australasian patients 4 years after market availability in Australia. Ivacaftor-treated patients have significantly better lung function and nutritional status despite being significantly older than untreated contemporaneous patients. Ongoing data analysis is being performed to examine the impact of Ivacaftor on lung function trajectory, complications and survival.

Reference

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A051

CASE REPORT - SILVER BULLET IN THE MANAGEMENT OF CYSTIC FIBROSIS

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Introduction: Treatment of CF has been advanced significantly by the introduction of modulator therapies, which target the underlying cause of the disease. We report such a case to demonstrate the efficacy of Trikafta^{R} (elexacaftor/tezacaftor/ivacaftor and ivacaftor) in dramatically increasing the quality of life (QOL) and life expectancy of an adolescent, through significant and sustained improvements in pFEV1.The case also establishes the superior profile of Trikafta^{R} compared to Orkambi^{R} in severe disease in a patient with homozygous F508del mutation.

Method: Case report of a 17-year-old girl with F508del homozygous CF, pancreatic insufficiency and CFRD showing severe mixed obstructive and restrictive defect who was on Orkambi^{R} for 3.5years, with no significant improvement in her lung function, lowest pFEV1 being 26 % and awaiting lung transplantation. Since commencing compassionate access Trikafta^{R} 12 months ago her pFEV1 has improved from 26% to 44-50% predicted. She has only required one admission for tune up, her BMI has improved from 18 - 23 and her active transplant workup has been suspended.

Result: The primary outcomes were improvement in QOL and predicted increased life expectancy in a patient with severe disease since commencing Trikafta^{R} over a period of 1 year. Clinical improvement was demonstrated by decreased baseline sputum production, improved exercise tolerance, and significant sustained improvement in lung function. Secondary outcomes were suspension of active transplant workup and significant improvement in mental health, with development of an optimistic view of her future. There was also a dramatic reduction in the medication load and frequency of exacerbations and admissions.

Conclusion: Our case report demonstrates that although there is no cure for CF as yet,Trikafta^{R} has the ability to significantly increase the QOL and predicted life expectancy of an adolescent with severe lung disease and has a superior profile over other available modulators.

A054

CLINICAL AND AIRWAY INFLAMMATORY PHENOTYPE IN ADULTS ATTENDING THE PRINCE CHARLES HOSPITAL ADULT CYSTIC FIBROSIS CENTRE (ACFC) COMMENCED ON COMPASSIONATE ACCESS TRIKAFTA®.

EMMA LEDGER, Peter Vardon, Eimear Wright, Vanessa Moore, Michelle Wood, Angela Matson, Robyn Cobb, Daniel J Smith, David W Reid and ACFC team[a,b,c]

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Introduction/Background/Aim: The CFTR modulator Trikafta® ("triple therapy"; elexacaftor-tezacaftor-ivcaftor) has been highly effective in randomised controlled trials. At present, Trikafta is only available in Australia on compassionate grounds for CF patients with very severe lung disease. In this observational study, we characterise the pre-Trikafta clinical and airway inflammatory characteristics in these individuals.

Method(s): An observational study in 23 adult patients commenced on compassionate access Trikafta. Lung function, body mass index (BMI), sputum total cell counts (TCC) and sputum pH were assessed before Trikafta was commenced.

Results: 23 patients provided data for assessment. Seven of the F/F group (see Table) were receiving Orkambi® (Lumacaftor-Ivacaftor) and two were treated with Symdeko® (Tezacaftor-Ivacaftor) at the time of Trikafta commencement. One patient with a residual function *CFTR* mutation was receiving treatment with Symdeko at the time of Trikafta commencement.

	Age	**Sex**	**FEV_1%**	**BMI Kg/m^2**	**Sputum pH**	**Sputum TCC x10^6/mL**
***F/F n=9**	30 (22-45)	5F,4M	31 (21-42)	20.2 (17.1-24.3)	7.3 (6.9-7.6)	18.3 (6.0-37.6)
F/MF n=14[#]**	37 (18-50)	5F,9M	31 (19-42)	22.3 (15.1-34.4)	7.2 (6.6-7.8)	31.9 (3.7-241.1)

*F - F508del *CFTR* mutation, **MF - minimal function *CFTR* mutation ([#]one patient had a residual function *CFTR* mutation). Data are presented as mean and (range).

Discussion: This is an important cohort of patients with very severe CF lung disease. Future studies will focus on clinical and airway inflammatory phenotype predictors for response to Trikafta.

A056

RESPONDING TO COVID-19; EXPERIENCE OF AN ADULT CYSTIC FIBROSIS CENTRE IN AUSTRALIA PROVIDING CARE TO A GEOGRAPHICALLY DISPERSED POPULATION.

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Introduction/Background/Aim: The rapid global emergence and spread of COVID-19 highlights the risks of viral pandemics in the human population with the elderly and those with chronic disease most at risk. Relatively few infections in people with cystic fibrosis (CF) have been reported and clinical outcomes to date have been no worse than the general population, but people with CF especially those with severe disease remain theoretically at higher risk. We report the considerations and strategies introduced to counter the threat of COVID-19 in a geographically dispersed CF population living in Queensland, Australia.

Method(s): An adult CF centre (ACFC) located at The Prince Charles Hospital (TPCH) in sub-tropical Queensland providing care to 300 patients, which is one of the most geographically dispersed in the world with an active telehealth care program already established. TPCH houses a large thoracic department and the anticipation was of large numbers of acute COVID+ patients. The looming influenza season at the time COVID-19 cases began to be reported, the physical distance of patients away from the ACFC, the need to maintain clinical trials activity, the challenges of trans-state jurisdictions and keeping the CF population well-informed on rapidly changing clinical practices were all factors that the ACFC considered in its response.

Results/Conclusion: We report the rapid and comprehensive transition to an almost completely “Virtual CF Centre” within a short period of time and highlight the multi-disciplinary innovations around out-patient care, management of exacerbations, medication provision, influenza vaccination, clinical trials and maintenance of the flow of information to the CF community.

A062

JHH CF ADULT CLINIC IN COVID 19

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Aim: COVID 19 has changed the way we access health care services. John Hunter Hospital Adult Cystic fibrosis clinic director recommended patients self-isolate March to June 2020. Telemedicine was offered to all patients. This study aimed to identify patient reported experiences with service delivery and patient reported outcomes among individuals in the John Hunter Hospital Adult Cystic fibrosis clinic.

Method: 121 patients from the JHH CF clinic were sent an online survey relating to weight status, weight changes, oral intake, exercise habits, physical therapy treatment, and access to medication, exacerbations, mental health and hospital admissions. Experiences and feedback regarding telehealth consults was also collected.

Results: From a 52% response rate, 70% of patients reported maintaining or increasing their weight and oral intake increased in (87%). 32% of patients reported an increase in exercise and 27% increased their airway clearance. Note; there was a 39% decrease in hospital admissions for IV treatments compared to the same time in 2019. Patients reported technological issues such as absent sound and lack of access to accurate home spirometry as the main barrier to telehealth.

Outcome: While telehealth was well received and utilised, patients acknowledged the benefit of face-face consults and would like the option of telehealth or face-face in the future. Specific considerations such as; access to home spirometry, sputum collection practises, access to scales, provision of pathology forms to allow blood collection in advance, proper patient scheduling, robust and reliable telehealth access may assist with improving the patient experience. Implementation of these recommendations and further evaluation is required to monitor the effectiveness of telemedicine at the JHH CF clinic.

A068

COMPARING INTRA-BREATH OSCILLOMETRY AND MULTIPLE BREATH WASHOUT IN CHILDREN WITH CF

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Background and Aim: Multiple breath washout (MBW) is a useful test for detecting early changes in peripheral airways and ventilation inhomogeneity in patients with cystic fibrosis (CF). Unfortunately, the feasibility of MBW is low in very young children (30-40%) and requires lengthy testing sessions. Intra-breath oscillometry (IB-OSC) measures respiratory system resistance (Rrs) and reactance (Xrs) during tidal breathing and has high feasibility (>80%) in very young children. Xrs includes the small peripheral airways and thus, may also reflect ventilation inhomogeneity. Our aim was to examine whether measures of Xrs reflect ventilation inhomogeneity as measured by MBW.

Methods: IB-OSC and MBW were performed on 37 children at the Queensland Children's Hospital, as part of the Early Life Origins of CF (ELO) study. IB-OSC was measured using a 10Hz signal to track within-breath changes, and MBW was performed using the N2 protocol. MBW results were classified as normal or abnormal (lung clearance index (LCI) >7.9) and measures of Xrs (end-expiratory (XeE), end-inspiratory (XeI) and difference (XeE-XeI) were compared between the MBW groups.

Results: Acceptable paired IB-OSC and MBW measurements were achieved by 29 children (62% male), median age 6.5 yrs (IQR 4.6-7.8) and height 119.4 (IQR 105.3-130.3) respectively. Abnormal LCI values were reported in n=12 (41.4%). There were no differences in sex, age, or height between those with normal vs abnormal LCI results. Children with abnormal LCI results had decreased (more negative) Xrs variables when compared to children with normal LCI (XeE -1.86 vs -1.55; XeI -2.59 vs -1.54; XeE-XeI -0.48 vs 0.09).

Conclusion: These preliminary results suggest that IB-OSC Xrs variables reflect ventilation inhomogeneity. As IB-OSC is more feasible than MBW in very young patients with CF, further research is warranted to explore the clinical potential of IB-OSC.

A070

EVALUATION OF THE CLINICAL RESPONSE TO LUMACAFTOR WITH IVACAFTOR IN A REAL WORLD SETTING

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Background: The combination of lumacaftor/ivacaftor (LUM/IVA) was listed on the Pharmaceutical Benefits Scheme in 2018. Data on adherence and the effects of CFTR modulators on extra-pulmonary manifestations of CF remain limited.

Aim: To carryout post-marketing evaluation of LUM/IVA at 6, 12 and 18 months of treatment in patients with mild-moderate CF lung disease.

Methods: A prospective observational study included adults in Western Australia with $ppFEV_1$ ≥40, initiated on LUM/IVA between December 2018 and November 2019. Pulmonary exacerbation frequency, spirometry, weight and incidence of adverse effects were evaluated longitudinally 18-months before and after LUM/IVA initiation. Changes in rhinosinusitis and depressive symptoms were investigated using the Sinonasal Outcome Test (SNOT-22) and Patient Health Questionnaire (PHQ-9). Adherence to LUM/IVA, dornase alfa and azithromycin were assessed using Medication Possession Ratios (MPRs), calculated using hospital pharmacy dispensing data.

Results: Of 31 individuals (mean±SD age 25.7±5.8, median [range] $ppFEV_1$: 84% [49-115%]) prescribed LUM/IVA, 11 (35.5%) discontinued treatment within 18-months.

Mean exacerbation rate declined from 1.06 (95% CI: 0.49, 2.27) to 0.54 (95% CI: 0.22, 1.33) per patient over 18-months of treatment (p=0.339). No change in spirometry was observed, p=0.919. Mean BMI increased (+0.44 kg/m^2, p=0.007).
No significant change from baseline was observed for total SNOT-22 and PHQ-9 scores. Six patients reported low mood, irritability and mood swings following treatment initiation.

At 6 months, 88% of patients had a LUM/IVA MPR ≥80%, compared to 70% of patients at 18-months. Mean dornase alfa MPR decreased from 49% (baseline) to 36% (18-months) and remained unchanged for azithromycin (55-59%).

Conclusion: Although clinically important changes in sinus symptom and depression scores were not observed, a high number of patients separately reported mood-related adverse effects. Adherence was suboptimal, and this together with increased non-adherence with concomitant CF treatments may have implications on efficacy. Interventions to maintain adherence over time may be beneficial.

A076

REAL WORLD OUTCOMES OF CFTR MODULATOR THERAPY IN ADULTS AND CHILDREN

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Introduction/Background/Aim: Recent advances in CFTR modulator therapy have the potential to change the face of cystic fibrosis [CF]. There are currently three CFTR modulators available in Australia: ivacaftor, lumacaftor/ivacaftor and tezacaftor/ivacaftor. In addition, elexacaftor/tezacaftor/ivacaftor is currently available via compassionate access. This retrospective observational study describes real world experience of these therapies in adults and children with CF in a single centre in Melbourne.

Method(s): Data was collected for all adult and paediatric patients commenced on CFTR modulator therapy at Monash CF. Data was collected at baseline and 6 monthly intervals for the duration of treatment, between 18th May 2012 and 1st September 2020. Primary outcomes included lung function, admission days and BMI/ BMI centile over time. Adverse events and reasons for changing or ceasing medications were also analysed.

Results: 74/133 (55%) adult and 55/119 (46%) paediatric patients were commenced on at least one CFTR modulator.

There was a significant improvement in BMI and lung function in all medications in adults, and a non-significant improvement in admission days. There was a significant reduction in admission days for children treated with ivacaftor and lumacaftor/ivacaftor, and a non significant improvement in lung function and BMI centile.

31/86 patients ceased lumacaftor/ivacaftor, 25 due to adverse effects. Of these, 23 changed to tezacaftor/ivacaftor, 18 without significant adverse effects.

Conclusion: Our findings are consistent with those of previous trials, indicating modest lung function benefit, reduction in admission days and improvement in nutrition with use of ivacaftor, lumacaftor/ivacaftor and tezacaftor/ivacaftor in a real world setting. We have documented a higher rate of adverse effects with lumacaftor/ivacaftor compared with phase 3 clinical trials. Our small cohort of patients treated with elexacaftor/tezacaftor/ivacaftor had improvements in all parameters in line with the optimistic results of trials. Tezacaftor/ivacaftor was generally well tolerated by those who experience side effects with lumacaftor/ivacaftor.

A078

GENDER, AGE AND ANATOMICAL EFFECTS APPARENT IN IVACAFTOR IMPROVEMENTS OF BONE MINERAL DENSITY IN CYSTIC FIBROSIS

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Introduction/Background/Aim: Low bone mineral density (BMD) is common in cystic fibrosis (CF). We studied changes in BMD with the CFTR potentiator ivacaftor in patients with the G551D *CFTR* gene mutation.

Method(s): An observational study of BMD in 27 adult patients commenced on ivacaftor. Twenty-three patients had BMD results pre- and post-ivacaftor with first repeat measurements a median of 512 days (IQR 290-773 days) after ivacaftor commencement. Twelve patients had at least two BMD assessments after ivacaftor and five patients had three repeat BMD measurements.

Results: There were significant improvements in lumbar spine BMD, but not in hip BMD on ivacaftor. Females had a greater increase in lumbar BMD with treatment than male patients ($p=0.048$). Lumbar BMD in females increased on average by 0.057 g/cm^3 (95% CI 0.031, 0.084, $p<0.001$), while lumbar BMD increased on average in males by 0.019 g/cm^3 (95% CI -0.009, 0.047, $p=0.19$). Treatment effect was also associated with patient age ($p=0.045$), where the effect was estimated to have reduced by 0.021 g/cm^3 (per 10 years of age).

Conclusion: Ivacaftor improved BMD at the lumbar spine, which was most marked in females and less evident in older patients. Further research needs to determine whether ivacaftor effects are mediated via improved CFTR function on osteoblasts and osteoclasts and how this relates to gender, age and anatomical site.

A079

STRESS PARAMETERS AND CYSTIC FIBROSIS EXACERBATION (SPACE) – A MIXED METHODS STUDY PROTOCOL

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Introduction: Wearable technology has enabled both consumers and clinicians to access a wealth of real-time physiologic data, which can be used to promote healthy lifestyle choices and empower the user in improving their own health. The use of noninvasive biomarkers generated by this technology in identifying and predicting pulmonary exacerbation of CF is currently unknown. We therefore aim to investigate the role of wearables in CF via a two-armed mixed methods approach; a prospective, observational study and qualitative analysis.

Method: A total of 40 adults with CF will be recruited at The Prince Charles Hospital. Twenty will be recruited at day 0 of hospitalisation for pulmonary exacerbation (the “exacerbation” cohort), with another twenty stable patients recruited from clinic (the “stable” cohort). All participants will be asked to wear the *Garmin Vivosmart 4* smartwatch for a period of 120 days. Regular visits will be arranged to download data and correlate with basic observations, symptom severity scores and spirometry. Participants will be encouraged to keep a symptom diary and record any significant life events. Each participant will undergo a semi-structured interview at day 120 exploring their experiences using the device.

Results: Raw data will be obtained and analysed, including sleep data. Mixed statistical methods to analyse continuous variables will be used to identify significant differences between beginning and end of exacerbation throughout the 40 patients. Mixed effects modelling and correlations between clinical status and biometric measurements will also be explored. All qualitative data will undergo thematic analysis to correlate key themes with quantitative assessment of biomarkers.

Conclusion: The integration of mHealth technology into traditional care pathways for individuals with CF is an exciting opportunity. We believe this study will further our understanding of the capability of wearable technology to guide both patients and clinicians in the management of pulmonary exacerbation of CF.

A084

THE JOHN HUNTER CHILDREN'S HOSPITAL CFRD COHORT

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Diabetes is a common complication of cystic fibrosis (CF). Onset of CF related glycaemic abnormality is insidious and associated with declining pulmonary and nutritional status. At JHCH, We perform Oral Glucose Tolerance Test (OGTT) with paired continuous flash glucose monitoring (FGM) via a Libre device annually from 8 years of age to identify such abnormality.

Aim: We aim to describe a retrospective case series of patients diagnosed with CF related glycaemic abnormality on FGM and/or OGTT and to explore any discrepancy between OGTT and FGM results.

Method(s): A total of 51 children were identified with CF related glycaemic abnormality. We accessed their OGTT and FGM results from digital medical records.

Results: 42/51 children were included in the study. 9/52 children excluded as records were incomplete or FGM was not performed within a year of the initial OGTT. 40/42 children with abnormal OGTT also had abnormal FGM and were commenced on insulin. 2/42 children had an abnormal OGTT but a normal FGM and were not commenced on insulin. These 2 children had subsequent normal OGTT and FGM on yearly screening.

Conclusion: Our study showed good correlation between OGTT and FGM. Paired OGTT and FGM were superior in diagnosing CF related glycaemic abnormality. FGM also helps in identification of glycaemic abnormality in "real world" setting and initiate early treatment to optimize lung function.

A086

HAS THE ADDITION OF LUNG CLEARANCE INDEX TESTING IN CHILDREN AFFECTED OUTCOMES AND MANAGEMENT IN CYSTIC FIBROSIS PATIENTS?

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Aim: The primary objective was to investigate if the introduction of annual LCI, measured via MBW since 2017 was associated with improvement in FEV1 within a cohort of paediatric patients at a tertiary CF clinic. The secondary objective was to investigate if inclusion of this testing modality resulted in a change in prescribing patterns.

Method: Data were collected retrospectively from patients that attended John Hunter Children's Hospital CF clinic between 2014-2020 inclusive. Eligible patients' spirometry between 2014-2016 and 2018-2020 were compared via matched pairs testing to evaluate if the inclusion of MBW was associated with an improvement in FEV1. Prescriptions of specific CF-associated medications were reviewed after MBW institution to assess whether LCI was documented as the rationale behind the addition of these medications.

Results: FEV1 were, on average, 2.84% and 3.84% above the maximum acceptable decline per patient in the 2014-2016 and 2018-2020 periods respectively. Spirometry results after the introduction of MBW were demonstrated to be statistically significantly better, on average, than the maximum acceptable level of lung function decline in this cohort ($p = 0.0047$). Of the 8 patients who were prescribed dornase alfa after institution of MBW testing, 6 had documentation referring to LCI as the reason behind introduction of this medication. Of the 2 patients that commenced hypertonic saline, 1 had documentation citing abnormal LCI as reason for its addition. No association between LCI results and azithromycin prescription was found.

Conclusion: This exploratory analysis identified that the addition of annual MBW as a monitoring tool utilised with this patient cohort has been associated with relatively improved mean FEV1 results. LCI results have been cited in patient notes as reason for additional medication prescription. Further research in this area would be valuable to validate the inclusion of LCI in the assessment and ongoing management of paediatric CF.

A087

ASSESSING PATIENT SATISFACTION WITH THE CYSTIC FIBROSIS TELEHEALTH SERVICE

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Introduction: Telehealth (via videocall) was rapidly implemented by the South Australian Adult Cystic Fibrosis (CF) Service in March 2020 due to the Novel Coronavirus 2019 pandemic. Home spirometers were distributed as part of objective patient assessment. Our aim was to assess patient satisfaction with the telehealth service in CF multidisciplinary clinic via online survey.

Methods: All patients of the CF service were invited to complete an online survey designed to assess satisfaction with telehealth, which was undertaken through the consumer organisation Cystic Fibrosis South Australia. A quantitatively driven concurrent mixed-methods design was chosen, with statistical analysis of symmetrical five-point Likert responses using IBM SPSS Statistics for Windows v.26 and coding of unstructured responses for thematic analysis.

Results: To date 57/160 patients (35.6%) completed the survey, with some item non-responses. 37/57 (64.9%) participants were aged ≤35yo, 42/53 (80.7%) had ≥ 2 telehealth appointments, and 46/53 (86.7%) found it easy/very easy to access. 18/48 (37.5%) of participants had technical issues. Despite this 48/52 (92.3%) felt satisfied/very satisfied with engagement of CF team via telehealth. 38/50 (76%) found telehealth more convenient with significant time saved. The thematic analysis added depth to quantitative analysis by identification of six key themes: convenience, ease of access, maintaining therapeutic relationships, comprehensive care, safety, and patient centred care.

Conclusion: Most patients reported high satisfaction with telehealth with a strong preference for availability of hybrid clinics, with the option of telehealth as well as in-person review to augment the CF chronic disease care model.

A090

TRIUMPH WITH TRIKAFTA; AN ADOLESCENT FEMALE'S RESPONSE TO NEW CFTR MODULATOR THERAPY

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A 17 year old female patient with cystic fibrosis (F508del/F508del) at our centre was undergoing workup for lung transplantation prior to a dramatic clinical improvement following compassionate access to Trikafta (Lexacaftor, texacaftor and ivacaftor) in April 2020. Her co-morbidities include pancreatic insufficiency, cystic fibrosis related diabetes, asthma and gastro-oesophageal reflux disease.

Prior to Trikafta, our patient's treatment regimen included airways clearance, supplemental oxygen used intermittently, insulin for CF-related diabetes, rh-DNA-se, inhaled corticosteroids, azithromycin, nebulised anti-pseudomonas antibiotics, pancreatic enzymes, proton pump inhibitors and vitamins. Prior to Trikafta, she had been treated with Orkambi (lumacaftor/ivacaftor) for three years.

Despite this comprehensive treatment regimen and three-monthly hospital admissions, her predicted FEV1 remained at 24-48%. She had significant respiratory compromise with dyspnoea at rest and poor exercise tolerance, along with severe decompensations such as a spontaneous pneumothorax with minimal exertion. The severity of her symptoms and poor life expectancy had a dramatic impact on her quality of life, education and psychological wellbeing.

Within seven weeks of commencing Trikafta treatment her FEV1 rose from 24% to 44% predicted. This was accompanied by reduction in respiratory symptoms and improved energy levels. Following 8 months on treatment she has maintained an FEV1 between 45-50% predicted, she is able to exercise without dyspnoea and is non-productive with airway clearance. Improvement in exocrine and endocrine pancreatic function has facilitated weaning of pancreatic enzymes and insulin and her BMI increased from 19 to 23. She is no longer requiring scheduled hospital admissions.

Treatment with Trikafta has resulted in a remarkable improvement in our patient's quality of life, mental health and prognosis. She has been removed from the transplant waiting list and is planning a bright future.

GI/NUTRITION

A004

PERCUTANEOUS ENDOSCOPIC GASTROSTOMY IN PAEDIATRIC CF PATIENTS: CLINICAL OUTCOMES AND PATIENT EXPERIENCES

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Aim: To investigate outcomes and experiences in paediatric CF patients 12 months after placement of a percutaneous endoscopic gastrostomy (PEG). The primary objective was to determine change in BMI z-score; the secondary objectives were to determine changes in FEV_1, hospital admission days and child/carer-reported experiences.

Method: A retrospective audit was conducted in paediatric patients with CF at the Women´s and Children´s and Monash Children's Hospitals in whom a PEG had been placed between 2014 and 2019. Growth parameters, clinical characteristics and hospital admission days were obtained from medical records. A child and carer questionnaire was developed to explore side effects and experiences post PEG placement.

Results: Thirteen (8 male; 62%) patients were included. Mean age at PEG insertion was 9.48 years (3.61 SD) . Mean BMI z-score increased from -1.10 at PEG insertion to -0.67 12 months post. There was a -0.24 (0.81 SD) mean decrease in BMI z- score in the 12 months prior compared to 0.47 (0.47 SD) mean increase in BMI z-score in the 12 months post (p=0.04). Mean change in FEV_1 (%predicted) was -10.86 (11.81 SD) compared to 4.21 (13.20 SD) (p=0.07) respectively. Mean hospital admission days reduced from 43.77 to 26.31 (p=0.07). Eleven (85%) carers and 8 (62%) children completed the questionnaire. The most common side effects reported by carers and children as 'always' or 'most of the time' were nocturia (30% and 50%, respectively) and bloating (20% and 25%, respectively). Six (55%) carers reported a blocked PEG and 7 (64%) reported an infected PEG site. Both groups reported improvements post PEG insertion in weight and energy levels whilst body image was perceived to be worse. The majority of carers and children would recommend a PEG to other children with CF (90% and 71%, respectively).

Conclusion: These results suggest PEG feeding is effective at improving the nutritional status of paediatric CF patients. Negative outcomes to consider in the decision making process include patient-reported nocturia, bloating and the impact of PEGs on body image.

A005

DOES ROOM SERVICE MEET THE CONSUMER EXPECTATIONS OF A CYSTIC FIBROSIS POPULATION

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Introduction/Background/Aim: Mater's Cystic Fibrosis (CF) population were the first to experience Room Service (RS) in Australia. The on-demand model allows patients to order meals compliant for their individual requirements from a single integrated menu. Evaluation of RS compared with traditional foodservice models for Mater's general population has demonstrated improvements in patient satisfaction, food costs, plate waste and nutritional intake as compared with a traditional food service model 1, 2. However limited studies have explored the success of room service within the CF population and intake data indicates patients are not meeting nutritional requirements. In 2019 patients with CF at Mater Health met on average 47% of energy and 61% of protein estimated requirements from the current room service menu. This study aims to evaluate Mater's RS model and whether it meets the CF population's nutritional needs and consumer expectations.

Method(s): Data is currently being collected to assess patient satisfaction and intake whilst admitted with CF. A modified Meal Ordering Preference Survey (MOPS) has been developed for CF inpatients to evaluate meal ordering behaviours, perceptions and satisfaction with room service. This data will be matched with nutritional intake, evidenced based nutritional recommendations and clinical demographics. This satisfaction data will also be compared against general hospital satisfaction results. Findings from the modified MOPS will guide further qualitative data collection to inform changes to the current model.

Results: Nutritional intake data has been collected and analysis is underway. It is expected that additional qualitative data collection will be completed by June 2021.

Conclusion: Initial data shows that the CF cohort differs from the general hospital population in clinical demographics (increased frequency of admissions, longer lengths of stay and a lower median age). It is anticipated that the differences in clinical demographics will be reflected in satisfaction results. These findings will assist in identifying reasons for inadequate nutritional intake among patients with CF and drive areas for improvement.

A008

REVIEW OF GASTROINTESTINAL SYMPTOMS WITH THE INTRODUCTION OF CFTR MODULATOR THERAPY IN TASMANIAN ADULTS WITH CYSTIC FIBROSIS

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Background: Initial trials for CFTR modulator therapies did not evaluate gastrointestinal (GI) symptoms as primary outcomes. A CF-specific GI symptom questionnaire is yet to be developed. Tools validated in conditions with chronic GI issues are expected to be applicable to the CF population.

Aim: To increase awareness and understanding of GI symptoms to improve management of complex GI issues post CFTR modulator therapy.

Methods: CF patients on lumacaftor/ivacaftor (Orkambi) were recruited to Stage 1, a 12-month retrospective audit. Reported GI symptoms, outcome measures, comorbidities and hospital admissions, changes in treatment, cessation of medication related to GI symptoms were recorded. Patients starting tezacaftor/ivacaftor (Symdeko) were recruited to Stage 2, a prospective 12-month audit. A modified Irritable Bowel Syndrome Severity Scoring System (IBS-SSS) questionnaire at baseline, six-months and twelve-months post was included with Stage 1 measures.

Results: 16 patients were recruited to Stage 1. 62% (n=10) reported GI symptoms with Orkambi. Due to GI symptoms: 56% (n=9) altered treatment, 31% (n=5) had hospital admissions, 25% (n=4) ceased Orkambi. Most reported symptoms were diarrhoea (n=4), bloating (n=4), increased frequency (n=3), increased urgency (n=3) and constipation (n=3). 10 patients were recruited to Stage 2. 70% (n=7) had switched from Orkambi. 50% (n=5) reported GI symptoms, bloating (n=2) being the most common. At 6 months, 70% (n=7) had symptoms improve, 6 of which had switched from Orkambi. At 12 months, 6 patients were still participating. 50% (n=3) had symptoms improve compared to baseline, all of which had switched from Orkambi. Nil hospital admissions, treatment alterations or Symdeko cessation due to GI symptoms.

Conclusion: GI symptoms were more prevalent in patients taking Orkambi, with more than half altering treatment and a quarter ceasing the medication. Symdeko was better tolerated. The modified IBS-SSS tracked changes in symptom severity and highlights the need for a CF-specific GI symptom questionnaire.

A027

PROBIOTIC USE IN ADULTS WITH CYSTIC FIBROSIS IS COMMON AND INFLUENCED BY GASTROINTESTINAL SYMPTOM BURDEN

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Introduction: Gastrointestinal disorders and symptoms, including dysbiosis, impact on morbidity and quality of life in cystic fibrosis (CF)[1]. There is interest in strategies to modulate the microbiota including probiotics. Probiotics may reduce pulmonary exacerbations and decrease gastrointestinal inflammation in CF, though evidence remains inadequate to guide practice, and information on use is limited. This research aimed to characterise probiotic use, beliefs and experiences of adults with CF.

Methods: A cross-sectional mixed methods questionnaire study was conducted in adults with CF (n=205) and a general population healthy Control group (n=158) from Victoria, Australia. Participants were classified as probiotic “Ever Users” or “Never Users”. Outcomes included self-reported probiotic use and beliefs, and factors associated with probiotic use, analysed using logistic regression analysis. Open-ended questions allowed participants to share their experiences and responses were thematically analysed.

Results: 55% of the eligible population completed the questionnaire (78% response rate). 70% of adults with CF had ever used probiotics (supplements and/or foods) compared to 80% of Controls, p=0.03, however there were no differences when stratified by female gender (83% vs 88%, p=0.32) or university level education (83% vs 85%, p=0.69). Key reasons for CF probiotic use were gastrointestinal- and antibiotic-related (75%) rather than immune-related (47%). Probiotic use was primarily self-initiated with most CF participants (73%) not discussing probiotic use with healthcare providers (HCP). Female gender (OR, 95% CI) (2.82 (1.36-5.87, p=0.005)), university-level education (2.73 (1.24-6.01, p=0.01)) and bloating whilst on antibiotics (2.14 (1.04-4.40, p=0.04)) were independently associated with probiotic use in CF; as was female gender in Controls (2.84 (1.20-6.71), p=0.02)).

Conclusion: Probiotics were used by adults with CF for primarily gastrointestinal- and antibiotic-related reasons often without informing HCPs. Further research investigating gastrointestinal outcomes of probiotics will inform practice recommendations guiding their use in CF.

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A028

CLASSIFICATION OF NUTRITIONAL STATUS FOR CHILDREN WITH CYSTIC FIBROSIS: THE USE OF BODY MASS INDEX (BMI) VERSUS FAT FREE MASS (FFM)

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Background: Cystic fibrosis (CF) patients are at risk of fat free mass (FFM) depletion. A correlation exists between FFM and FEV_1, infection and intravenous antibiotics use[1]. BMI is used to classify nutritional status but does not specifically account for fat mass (FM) and FFM. Bioelectrical impedance analysis (BIA) offers the potential to quantify FM and FFM and therefore assess body composition during a CF nutrition assessment.

Aim: In CF children at the Children's Hospital Westmead (CHW);

1. Compare BMI vs. FFM depletion as a method to detect nutritional impairment.
2. Explore predictors of FFM depletion.

Method: Cross-sectional retrospective review of CF children. A validated CF FFM correction equation was applied to BIA data[1]. FFM z-scores were calculated using healthy paediatric reference data[2]. Nutritional impairment was defined as BMI <25thcentile and FFM z-score ≤-2. Predictors of FFM depletion were determined using logistic regression analysis (age and FEV_1) and Chi Square test for categorical data.

Results: 143/158 (91%) of the eligible cohort were studied; 48% female and 79% PI with 50% prescribed modulator therapy. Mean±SD age was 12±4years, FEV_1 92±15%, BMI z-score 0.1±1 and FFM z-score -1.1±-1.0. Nutritional impairment as defined by FFM (34/143, 24%) was higher than BMI (23/143, 17%) with 10/143 (7%) meeting both FFM and BMI criteria. 24/104 (23%) with an acceptable BMI≥25thcentile were FFM depleted. Younger age was a significant predictor of FFM depletion ($p<0.001$) while FEV_1 was not ($p=0.88$). Significantly more males ($p=0.01$, OR=2.83 (95% CI:1.24,6.48)) and fewer patients on a modulator ($p=0.04$, RR=0.83 (95% CI:0.67,1.0) had FFM depletion. Rates of FFM depletion weren't significantly different for those with CF-related diabetes, liver disease, gastrostomy, PI, abscesses and FEV_1≤80%.

Conclusion: Using BMI alone may not capture rates of FFM depletion in children with CF. Younger age, male gender and absence of modulator use were predictors of FFM depletion.

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(1) Papalexopoulou N, Dassios TG, Lunt A, et al. Nutritional status and pulmonary outcome in children and young people with cystic fibrosis. Respir Med. 2018;142:60-65. doi:10.1016/j.rmed.2018.07.016

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A029

COMPARISON OF BODY COMPOSITION PARAMETERS MEASURED WITH DUAL ENERGY X-RAY ABSORPTIOMETRY (DEXA) AND BIOELECTRICAL IMPEDANCE ANALYSIS (BIA) IN CHILDREN WITH CYSTIC FIBROSIS

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Background: Bioelectrical Impedance Analysis (BIA) scales enable body composition to be assessed rapidly in a busy clinic environment. While a correction equation to determine fat free mass (FFM) using a BIA device and dual energy x-ray absorptiometry (DEXA) has been developed in adolescents with CF (FFM_c=-2.844-(0.184xweight)+(1.202xFFM_{BIA})), it is recommended that all BIA scales are validated against DEXA prior to clinical use, especially amongst the younger paediatric CF population[1].

Aim: To assess the validity of BIA scales by comparing BIA FFM (BIA_{FFM}) and BIA FFM corrected (BIA_{FFMC}) values with $DEXA_{FFM}$ for children with CF.

Method: Children with CF with a routine DEXA appointment had BIA completed on the same morning. While not fasted, children were asked to empty their bladder prior to measurement. BIA_{FFM} values were estimated using Tanita-TIMC780MA BIA scales and BIA_{FFMC} values generated by the validated Papalexopoulou CF correction equation[1]. Bland-Altman (BA) plots were produced to assess agreement between the two methods. Difference in means was tested using the paired t-test and Pearson *r* correlation was used to measure the relationship between the 2 methods.

Results: 18 children, mean age 13.9years (range 10-18 years) completed the study. Mean BMI z-score was -0.16 (SD±1.09) and 13/18 (72%) had a BMI 25-75th centile. While BIA_{FFM} had poor agreement with $DEXA_{FFM}$ [mean difference 3.4kg, 95% LOA (-0.03; 6.74)], BIA_{FFMC} and $DEXA_{FFM}$ had good agreement across the FFM range [mean difference of -1.0kg, 95% LOA (-4.63; 2.65)]. BIA_{FFMC} and $DEXA_{FFM}$ correlated well (r=89; p<0.001) and there was no statistically significant difference in mean differences (t=-1.68, p=0.11).

Conclusion: BIA_{FFMC} is a valid measure of FFM for children with CF. Further recruitment to assess the validity across a greater range of FFM is needed before introducing this measure into the CF nutritional assessment process.

References:

(3) Papalexopoulou N, Dassios TG, Lunt A, et al. Nutritional status and pulmonary outcome in children and young people with cystic fibrosis. Respir Med. 2018;142:60-65. doi:10.1016/j.rmed.2018.07.016

A039

ALTERED ESSENTIAL FATTY ACID HANDLING IN THE CYSTIC FIBROSIS RODENT MODEL

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Introduction/Background/Aim: Cystic fibrosis (CF) is caused by mutations in the CF transmembrane conductance regulator (*CFTR)* gene. Patients with CF have low concentrations of essential fatty acids in their circulation and tissues, leading to deficiency. Essential fatty acid deficiency may be critical for CF pathology as cell surface expression of CFTR may be regulated by essential fatty acids. Two rodent models of CF have been developed which lack a functional CFTR (CFTR KO) or express the most common mutation delta F508 (DeltaF508 CFTR). These CF rodent models present with pulmonary and gastrointestinal organ deficits associated with CF. The aim of this study is to investigate circulating concentrations of essential fatty acids as well as genes expressed in the liver which are responsible for essential fatty acid transport and metabolism.

Method(s): Plasma and liver samples were harvested from CFTR KO and DeltaF508 CFTR rats following weaning (between 2 and 4 months of age). Essential fatty acid concentrations were determined using gas chromatography and quantification of mRNA was achieved using real time PCR with the QuantiNova SYBR green master mix (Qiagen). Gene expression was quantified using the 2-ΔΔCq method and normalised to the geometric mean of β-actin as a reference gene.

Results: In the CF rodent models, there was an increase in arachidonic acid: docosahexaenoic acid ratio in deltaF508 CFTR rats in circulation, similar to patients with CF. This was not observed in CFTR KO rats. Further, FADS1, a gene which encodes for the enzymes responsible for essential fatty acid metabolism was altered in both CF rodent models ($p<0.05$), as were genes which encode the essential fatty acid transport proteins FABP3 and FABP5 ($p<0.05$), and FATP1 ($p = 0.06$).

Conclusion: These data suggest that the CF rat models exhibit essential fatty acid deficiency, which may be due to altered hepatic transport and metabolism.

A042

NUTRITIONAL MANAGEMENT OF A NEONATE DIAGNOSED WITH CYSTIC FIBROSIS, TRISOMY21 AND SHORT BOWEL SYNDROME: A CASE STUDY

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Baby IH born 35+6, birthweight 2150g (z-score -1.26, Fenton Charts) diagnosed with Trisomy21 (T21), was transferred to RCH on day 1 (D1) with suspected bowel obstruction, resulting in resection of 40cm ileum, ileostomy formation and diagnosis of microcolon. On D18, he was diagnosed with Cystic Fibrosis (homozygous, N1303K – class II mutation). This case study discusses the nutritional challenges encountered during IH clinical course.

TPN was initiated D1 and enteral feeds (EF) of EBM post-ileostomy formation on D6. Creon-micro mixed with apple puree was introduced D18 at EBM volumes 60ml/kg. High stoma losses (>20ml/kg) requiring fluid replacements resulted in multiple cessations of EBM.

Post stoma reversal, EBM was reintroduced and established by 6 weeks corrected age (CA). Poor growth (weight z-score -2.2, Fenton Charts) and fat malabsorption on fortified EBM at >150ml/kg with >11000iu/kg/day lipase, necessitated a trial of extensively hydrolysed Aptamil Peptijunior and lipase >16000iu/kg/day. However, poor clinical response required the introduction of an amino acid-based formula (Neocate) prior to discharge. Home feeds were Neocate plus Liquigen at 150ml/kg with >16000iu/kg lipase.

At 3-months CA at 3.96kg (z-score -1.83 CA T21 Chart) IH was diagnosed with distal intestinal obstruction syndrome requiring readmission and subsequent laparotomy and stoma formation. Post-operative TPN was commenced and titrated once EF of fortified Neocate were delivered at 150-180ml/kg with >11000iu/kg lipase. Excessive stoma losses, prolapsed stoma, fat malabsorption and faltering growth (weight z-score -2.52 4m CA T21 chart) resulted in an unsuccessful trial of macronutrient adjusted modular feed and resumption of TPN prior to stoma reversal.

IH remained TPN dependent during establishment of EF. At 10 months CA weight restoration (8.49kg, z-score 0.06 10m CA, T21 Chart) allowed discharge home (D236 post-DIOS) on TPN (4/7) and EF of fortified Lipistart (110ml/kg), Creon-micro 16000iu/kg and solids.

A052

MEASURING ENERGY EXPENDITURE DURING PULMONARY EXACERBATIONS OF CYSTIC FIBROSIS USING INDIRECT CALORIMETRY: PRELIMINARY FINDINGS

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Introduction: Energy requirements of people with Cystic Fibrosis (CF) are between 110-200% of the general population. In clinical practice, predictive equations are relied on to estimate energy requirements. Predictive equations cannot account for factors influencing energy expenditure, such as pancreatic function, body composition and clinical status. It is unclear to what extent energy expenditure varies between periods of pulmonary exacerbation (PEX) and clinical stability. This study sought to investigate differences in resting energy expenditure (REE), measured using indirect calorimetry (IC), at commencement and completion of IVABx for PEX and during a clinically stable period. The accuracy of predicted REE (pREE) determined using Schofield equation was also investigated in comparison to measured REE (mREE).

Methods: The mREE (using IC), fat mass (FM) and fat free mass (FFM) (using bio-impedance analysis), and pREE (using Schofield equation) were determined for each participant at three time points: (i) commencement of 14-day IVABx treatment for PEX; (ii) end of IVABx treatment; and (iii) during a clinically stable period (≥six weeks following IVABx treatment). Only the results from time points (i) and (ii) are presented in this preliminary analysis.

Results: Twelve participants (seven male, 10 pancreatic insufficient), aged 32.5±13.4 years and body mass index 21.9±4.9 kg/m^2 were recruited. No statistical difference between mREE at the start and completion of IVABx treatment (95% CI [-265, 208]; p=0.796) was observed. The mREE was significantly higher than pREE at both time points (mean difference 237 kCal; 95% CI [120, 355]; p=0.00024). No significant changes in FM or FFM were detected between the two time points.

Conclusion: Initial findings indicate the Schofield equation underpredicts REE during PEX compared to mREE. Utilisation of IC may assist to individualise nutrition prescription and optimise patient outcomes. Further research with a larger sample size is required to strengthen results and identify further patterns.

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A060

CONSTIPATION AND DISTAL INTESTINAL OBSTRUCTION SYNDROME IN CYSTIC FIBROSIS: A RETROSPECTIVE AUDIT OF THE INCIDENCE, CHARACTERISTICS AND MANAGEMENT IN A COMBINED PAEDIATRIC AND ADULT UNIT

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Aim: To describe the incidence, characteristics and management of constipation and DIOS amongst paediatric and adult CF inpatients over a ten year period.

Methods: Ten year retrospective audit (2011-2020) of children and adults with CF at Monash Health admitted with either constipation or DIOS as their primary or secondary diagnosis.

Results: Between 2011 and 2020, 28 episodes of DIOS were diagnosed in 21 patients (13 male; 62%); 39% (11 of 28) were diagnosed as complete DIOS. Across the same time period, 106 episodes of constipation were diagnosed in 60 patients (25 male; 42%); in 31% (33 of 106) of these episodes, constipation was the primary diagnosis on admission. Of the 21 patients with DIOS and 60 patients with constipation, 5 patients (24%) with DIOS and 18 patients (30%) with constipation experienced a recurrence in the study period. The majority, 100% (21 of 21) and 92% (55 of 60), of patients presenting with DIOS and constipation were pancreatic insufficient. 67% (14 of 21) and 45% (25 of 55) of patients presenting with DIOS and constipation were delta F508 homozygous. Median LOS for the DIOS group was higher than for the primary constipation group, 4.74 days versus 2.94 days, respectively. Few DIOS episodes required surgery; 11% (3 of 28) across the entire cohort. The use of Polyethylene Glycol (PEG)-based laxatives in the treatment of both DIOS and constipation was high; 75% (21 of 28) and 82% (87 of 106), respectively. Adjunct laxative treatments in the management of DIOS and constipation included gastrografin, enemas and osmotic laxatives.

Conclusion: The prevalence of constipation is higher than that of DIOS although when comparing episodes of primary constipation with DIOS the prevalence is similar. Both groups experienced a similar rate of recurrence. DIOS however is associated with a longer hospital stay and higher morbidity. The use of PEG-based laxatives as a curative treatment is high for both constipation and DIOS.

A061

COLORECTAL CANCER SCREENING IN ADULTS WITH CYSTIC FIBROSIS – IS iFOBT WORTH DOING?

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Introduction: Patients with cystic fibrosis (CF) have an increased risk of developing colorectal cancer (CRC). Unlike in the non-CF population, the role of faecal occult blood testing (iFOBT) as a screening test is unclear. As a result, colonoscopy is the current recommended CRC screening test in CF. This is a preliminary report of an ongoing study. This study aims to evaluate the role of iFOBT in CF adults requiring CRC screening.

Methods: An observational, cross-sectional, single-centre study was conducted between February 2019-2021. Adult CF patients from Royal Prince Alfred Hospital who met the criteria for a screening colonoscopy, were recruited and instructed to perform iFOBT before their screening colonoscopy. Incomplete colonoscopies were excluded. iFOBT results (positive or negative) were compared with their colonoscopy findings (normal or abnormal). Abnormal colonoscopy results included both polyps and CRC. Fisher's exact test was performed.

Results: Twenty participants completed the study [mean age 49± 7 (SD) years, 50% female, at least one F508del mutation (90%), pancreatic insufficient (55%), Cystic Fibrosis Related Diabetes (CFRD, 45%), family history of colon cancer (15%), post-bilateral lung transplant (15%), mean FEV1 67±21% predicted among non-transplant participants (n=17)]. Seven (35%) demonstrated positive iFOBT results of whom 4 returned abnormal colonoscopy (57%) results. Of the 13 with negative iFOBT, 6 had abnormal colonoscopy results (46%). Overall, 10 (50%) patients had abnormal colonoscopy findings (polyps 90%, CRC 10%). There was no significant association between iFOBT and colonoscopy results (OR 0.64; 95% CI 0.1-4.1).

Conclusion: The rate of abnormal colonoscopy findings was high, indicating a need for colon cancer screening. These preliminary findings do not support the use of faecal occult blood testing to detect colorectal cancer or pre-cancerous lesions in patients with CF. The results will need to be confirmed in a larger adult CF cohort eligible for screening colonoscopy. Further research on alternative non-invasive CRC biomarkers are likely necessary.

INFECTION / MICROBIOLOGY / IMMUNOLOGY

A009

AZITHROMYCIN AUGMENTS BACTERIAL KILLING IN MACROPHAGES FROM PATIENTS WITH CYSTIC FIBROSIS VIA ERK1/2-MEDIATED PATHWAY

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Introduction. Azithromycin (AZM) treatment benefits patients with cystic fibrosis (CF). We previously reported defective macrophage polarization and function in CF. This study therefore aims to investigate macrophage polarization, functions and macrophage-driven inflammation in CF following treatment with AZM or its non-antibiotic derivative (G5).

Methods. Monocytes from venous blood from patients with CF (n=22) and healthy individuals (n=15) were isolated, differentiated into macrophages and then polarized into pro-inflammatory M1 and anti-inflammatory M2 macrophages with or without AZM or GS-560660 (G5). Percentage of M1/M2 subsets, inflammatory cytokine profile and pathogen clearance mechanisms were studied with and without AZM/G5.

Results. AZM and G5 enhanced bacterial killing and improved defective M2 macrophage polarization in CF. Enhanced bacterial killing following AZM/G5 treatment was observed against E. coli and S. aureus. Mechanistically, augmented bacterial killing was mediated via DUSP1-ERK1/2 pathway. AZM/G5 also inhibited nuclear translocation of NF□B resulting in no change in TNF□ release. Anti-inflammatory cytokine production (IL-10, CCL18) was enhanced following AZM/G5 treatment.

Conclusion. Data from the present study give insight into why AZM benefits patients with CF. Since AZM can induce antibiotic resistance, the therapeutic potential of G5 supress macrophage driven inflammation in CF warrants investigation.

A010

TREATMENT OF HUMAN AIRWAY EPITHELIAL CELLS WITH RHPON2 PREVENTS CELLULAR DAMAGE MEDIATED BY 3OC12-HSL, THE MAJOR *P. AERUGINOSA* QUORUM SENSING SIGNAL MOLECULE

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Introduction/Background/Aim: The major QS molecule produced by *P. aeruginosa*, N-3-oxododecanoyl-L-homoserine lactone (3OC12-HSL), regulates bacterial virulence and biofilm formation. This QS molecule also modulates host inflammatory and stress responses, ultimately leading to tissue damage and improved bacterial survival during lung infections in people with Cystic Fibrosis. The intracellular human enzyme Paraoxonase 2 (hPON2) can hydrolyse 3OC12-HSL and has anti-oxidative properties. In this study we examined whether a recombinant protein form of hPON2 (rhPON2), applied extracellularly, could protect the human airway epithelial cells from 3OC12-HSL-mediated effects.

Methods: We performed high-throughput RNA sequencing and compared expression data from airway epithelial cells treated with 3OC12-HSL to that of cells pre-treated with purified recombinant human PON2 (rhPON2) prior to 3OC12-HSL exposure.

Results: Our results demonstrated that 3OC12-HSL resulted in the upregulated expression of a large number of genes whose proteins are known to be involved in regulating the immune response, unfolded protein response and apoptosis. In support of the gene expression data, we demonstrate that 3OC12-HSL increased the release of cytokines (IL-8) and caspase 3/7 activity, important components of inflammation and apoptosis respectively. Further, 3OC12-HSL treatment significantly upregulated UPR genes such as CHAC1 whose protein degrades the antioxidant glutathione, prompting us to determine the effects of 3OC12-HSL on cellular glutathione levels. We found a reduction of the cellular glutathione levels in airway epithelial cells after exposure to 3OC12-HSL. Importantly, pre-treatment with rhPON2 before exposure to 3OC12-HSL prevented the detrimental effects of 3OC12-HSL in airway epithelial cells.

Conclusion: Collectively, these data show for the first time the effectiveness of extracellular rhPON2 as a 3OC12-HSL-blocking therapy.

A014

REDUCED *PSEUDOMONAS AERUGINOSA* BIOFILM BIOMASS WHEN ANTIBIOTICS AND PHAGES ARE USED IN COMBINATION

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Introduction: Biofilm is a protective mechanism employed by *Pseudomonas aeruginosa* against environmental, immunological and chemical insults which also prevents penetration of agents including antibiotics and some bacteriophages (phages). However, evidence suggests that antibiotics and phage otherwise ineffective when singularly used, may be active when combined. In this study, we investigated the potential of tobramycin and phages in singularity or combined to reduce biofilm biomass.

Methods: Using a checkerboard design, 100 µL of an overnight culture of PA01 ($OD600_{nm}$ 0.01) was pipetted per well in a 96-well microtitre plate and left to incubate statically for 24 hours. Tobramycin and singular phage suspension (φE79 or φ3) were serially diluted (0.016 – 4 µg/mL; 10^2 – 10^6 PFU/mL) and then added onto pre-formed PAO1 biofilms for 20 hours. Following treatment, planktonic PAO1 was removed, and wells washed three times. Biofilms were then stained with 0.1% crystal violet (CV), dissolved in 30% acetic acid and resulting solutions read at 550_{nm} (mean±stdev; n=3).

Results: Compared to tobramycin-only controls, suboptimal concentrations of tobramycin (<0.5 µg/mL; stained biomass at 0.92) when used in combination with phages (φ3 and φE79) across all concentrations demonstrated markedly, but not statistically significant reduction in biomass. Quantification showed that treatment with 0.016 µg/mL of tobramycin alone was ineffective (0.89±0.48) and similar to untreated controls (0.78±0.33). A similar observation was made when treated with only phage (φ3; 0.33±0.97, φE79; 0.31±0.11). Lower biofilm biomass was observed following treatment with 0.016 µg/mL tobramycin and 10^2 PFU/mL of φ3 (0.36±0.06; range; 24.0-51.1% reduction) and φE79 (0.40±0.07; range 14.4-49%reduction).

Conclusion: Due to the inter-assay variability observed, more quantitative tests including the enumeration of viable bacterial load post treatment should be performed. However, this methodology has illustrated the utility of combining antibiotics and phages and may be implemented into antibiotic resistant bacterial infection treatment pipelines that use phage in the future.

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A017

AIRWAY BACTERIA MODULATE NEUTROPHIL GRANULE EXOCYTOSIS

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Introduction/Background/Aim: Neutrophils recruited into cystic fibrosis (CF) lungs develop into a population that abundantly releases tissue damaging enzymes, such as neutrophil elastase (NE)[1]. This population also strongly correlates with severity of lung disease early in life[2], but why this population is activated is unknown, limiting intervention. Evidence points to respiratory viral-bacterial co-infections as a possible cause[3], but this has not been directly studied in CF. To explore this, we developed a laboratory model to characterise responses of airway tissue and neutrophils following respiratory infection with virus and/or bacteria.

Methods: Paediatric primary airway epithelial cells were grown as differentiated cultures representative of native airway tissue. These were challenged individually, or co-infected, with rhinovirus and *Pseudomonas aeruginosa*. After infection, cultures were washed apically with medical saline, akin to bronchoalveolar lavage, to sample the infection microenvironment. Washes were assessed for inflammatory signals from airway tissue, then used to stimulate neutrophils in a published model of neutrophil transmigration to the airways[1].

Results: Epithelial infection microenvironments were characterised by increased antiviral signals with exclusive rhinovirus infection, including IP-10 and RANTES, while *P. aeruginosa* and viral-bacterial co-infection similarly increased pro-inflammatory signals IL-1α/β and TNFα. Neutrophils migrating into *P. aeruginosa* and co-infection washes had elevated CD63, a marker of granules that contain NE, and reduced CD16, a protein that allows neutrophils to engulf bacteria. These changes are reflective of the neutrophil population that develops in CF airways[1,2].

Conclusion: Our findings suggest both primary and secondary bacterial infections trigger the development of NE releasing neutrophils. Recent screening in this model of *Staphylococcus aureus*, *Neisseria lactamica*, and *Prevotella nigrescens*, also indicates that different airway microbes variably prompt this neutrophil subset, suggesting CF airway dysbiosis may be a driver of neutrophil pathological activity. Further studies will characterise mechanisms of this process and identify potential therapeutic targets.

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A018

BACTERIOPHAGE ISOLATION, PURIFICATION AND CHARACTERISATION, FOR THE POTENTIAL TREATMENT OF ANTIMICROBIAL RESISTANT (AMR) PSEUDOMONAS AERUGINOSA RESPIRATORY INFECTIONS IN INDIVIDUALS WITH CYSTIC FIBROSIS

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Introduction/Aim: Cystic fibrosis (CF) mutations facilitate the development of a respiratory micro-environment that favours recurrent pathogenic colonisation, particularly *Pseudomonas aeruginosa*, where in some cases up to 45% of isolates obtained from individuals with CF are multi-drug resistant (Sherrard et al., 2014). Following little investment into new antibiotics, bacteriophages/phages, which are natural viruses that infect/kill bacteria, have re-emerged as a potential therapeutic solution. We hypothesised that phages could be sourced, propagated, purified and subsequently exhibit lytic activity against clinical isolates of *P. aeruginosa*.

Methods: Phages, isolated from filtered environmental samples, were cultured overnight (37°C) with *P. aeruginosa* isolates derived from individuals with CF (n=30). Solutions were dispensed onto Luria-Bertani Broth Agar (LBA) plates harbouring *P. aeruginosa* growth, where plaque formation (indicating phage infection of bacteria), was assessed. Phages were purified via inoculation into a mixture of molten overlay agar and bacteria, which was poured onto LBA (repeated 3 times). Phage host range was determined using a miniaturised spot testing screen, whereby 30 clinical bacterial isolates were exposed to purified phages, allowing bacteria to be classified as phage susceptible, partially susceptible, or resistant, based on their ability to visually kill/lyse bacteria.

Results: Over 177 phages were isolated and successfully purified. All exhibited significant diversity, as illustrated by plaque shape, diameter, edge, halo and turbidity. Phage host range was also diverse, with no single profile identical. All 177 phages induced lysis in greater than 45% of bacterial isolates, 16 induced lysis in 90% of bacterial isolates, and 3 were bactericidal against all isolates.

Conclusion: Collectively, this study illustrates that phages can be isolated, purified and propagated using a simple screening method. It also demonstrates the potential of phage to treat AMR *P. aeruginosa* lung infections in CF, where it is hoped this will facilitate the translation of phage use to a clinical setting.

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A024

UNDERSTANDING *PSEUDOMONAS AERUGINOSA* PHAGE RESISTANCE TO INFORM PHAGE COCKTAIL FORMULATION

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Background/Introduction: The lungs of people with cystic fibrosis (CF) are frequently colonised by *Pseudomonas aeruginosa* and to combat infection, multiple courses of antimicrobials are typically used. However, over time, antimicrobial-resistant *P. aeruginosa* usually develops and is difficult to treat. With limited investment in current pharmaceutical antibiotic pipelines, alternative novel antimicrobials that can treat antimicrobial-resistant infections are urgently required. Lytic bacterial viruses, known as bacteriophages (phages), are one promising alternative, however, bacteria can develop resistance to them. Thus, preventing/overcoming this is an important component in phage therapy clinical translation. To study this further, we initially isolated and characterised phages for an in-house library.

Methods: Phages were isolated directly and through enrichment from environmental sources and host range determined against 70 diverse CF-derived *P. aeruginosa*, using spot assays. Over 250 phages were isolated and appeared diverse according to their zones of clearance (plaques). This diversity was corroborated when phage host range was established and also identified 20 phages active against 70-90% of the *P. aeruginosa* isolates tested. These were sequenced and analysed for novelty, lytic ability and genetic content and results used to identify which phages were suitable for therapeutic application. Certain phages also had their activity tested against PA01 mutants that had putative phage receptors knocked out. Phages acting upon distinct receptors were then exposed to CF-derived *P. aeruginosa* to evolve phage resistance. Phage resistant *P. aeruginosa* were then evaluated to understand phage resistance evolution.

Conclusions: Collectively, data here have assisted in understanding phage-resistance evolution by *P. aeruginosa.* It will also inform the strategic formulation of cocktails that may prevent resistance from occurring.

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A035

DEFEATING BIOFILM: THE ROLE OF ESSENTIAL OILS IN THE BATTLE AGAINST EXTENSIVELY DRUG RESISTANT *PSEUDOMONAS AERUGINOSA*

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Introduction/Background/Aim: Cystic fibrosis-associated *Pseudomonas aeruginosa* (CFPA) is a source of significant morbidity and mortality in CF patients. Evolutionary adaptations such as multidrug resistance (MDR), the capacity to form biofilms, and immune evasion mechanisms are key features of CFPA that enable the successful colonisation of lungs. Current treatments for MDR CFPA are inadequate, and novel approaches are required. In this study, we evaluated the interaction of essential oils and commonly nebulised/inhaled antibiotics when used against MDR CFPA.

Methods: Minimum inhibitory concentrations (MICs) and fractional inhibitory concentration indices (FICIs) of tea tree oil (TTO), 1,8-cineole (major component of *Eucalyptus* oil), tobramycin, colistin, and aztreonam were determined for clinical CFPA isolates (n=20) using a modified broth microdilution assay. The minimum biofilm eradication concentration (MBEC) and fractional biofilm eradication concentration index (FBECI) were also determined using a similar method, with biofilms formed on a purpose-built device (MBEC assay®).

Results: TTO was effective at lower concentrations against MDR CFPA isolates (n=3) that were in a biofilm than they were in a planktonic state (MBEC 8-16 times lower than MIC). CFPA within biofilm was less susceptible to aztreonam and colistin compared to planktonic cells (MBEC 4-16 times higher than MIC) but not tobramycin (MBEC 1-4 times MIC). All combinations of essential oil and antibiotic had indifferent relationships (FICI 0.52-1.83) when tested against planktonic MDR CFPA isolates (n=5). When tested against CFPA isolates (n=3) in biofilm, aztreonam and TTO showed an indifferent relationship (FBECI 0.58-1.13), TTO and colistin had a relationship approaching synergistic (FBECI 0.46-0.69), whereas TTO and tobramycin showed a synergistic relationship (FBECI 0.43-0.47).

Conclusion: The anti-biofilm properties of TTO and the synergistic relationship seen between TTO and tobramycin against CFPA *in vitro* make inhaled TTO a promising candidate as a potential therapeutic agent.

A040

NUTRITIONAL PARAMETERS IN CYSTIC FIBROSIS PATIENTS WITH *MYCOBACTERIUM ABSCESSUS COMPLEX* INFECTION AND DISEASE

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Introduction: Globally, prevalence of non-tuberculous mycobacterium (NTM) infections in cystic fibrosis (CF) is increasing[1]. A proportion of children who isolate NTM from respiratory samples meet criteria for NTM pulmonary disease (NTM-PD) that may require treatment[2]. We hypothesised that poor nutritional parameters are associated with initial acquisition of NTM that predicts progression to NTM-PD.

Method: A single centre retrospective study was performed on CF patients with □1 positive *Mycobacterium abscessus* complex (MABSC) isolate between 2012-2021. Initiation of NTM-specific treatment was used as a surrogate marker for NTM-PD. Weight and BMI parameters at time of first isolate and one-year prior were compared between treated and non-treated patients.

Results: 18 patients were included, of whom 8 (44.4%) required treatment. Mean age at first isolate was 13.1 years, 66.67% were female and 61.1% were homozygous DF508.
Mean BMI z-score and BMI centile 12 months prior to first isolate were lower in MABSC-infected patients (n=8) who received treatment, compared to non-treated patients (n=10), (-0.44(SD□0.4) vs. 0.10(SD□0.6); 34.5th(SD□13) vs. 54.6th(SD□21)). Similarly weight z-score and centile were lower in treated patients (-0.63(SD□0.48) vs. 0.53(SD□0.51); 28.9th(SD□14) vs. 68.9th(SD□17)).
Mean change in individuals' weight and BMI was compared between treated and non-treated groups over 12 months preceding their first isolate: change in weight z-score and centile was -0.06(SD□0.06) vs. +0.14(SD□0.26) and -0.06th(SD□0.3) vs. +0.14th.(SD□0.26) Similarly change in BMI centile and z-score was +0.91th(SD□8.4) vs. +1.54th(SD□11) and -0.02(SD□0.26) and +0.07(SD□0.37).

Conclusion: Declining nutritional parameters prior to acquisition of MABSC infection appear to be associated with progression to NTM-PD. A larger cohort is required to confirm this; however, it would be prudent to ensure vigilant screening for NTM infection and disease for children in this category.

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A047

NOVEL BACTERIOPHAGE ACTIVE AGAINST STAPHYLOCOCCUS AUREUS BACTERIA FROM INDIVIDUALS WITH CYSTIC FIBROSIS EXHIBIT VARIABLE LYTIC CAPABILITIES.

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Background: *Staphylococcus aureus* (*S. aureus*) is one of the first pathogens isolated from children with Cystic Fibrosis (CF) and contributes to airway inflammation. Individuals are often exposed to high levels of antibiotics to treat *S. aureus* infection and, consequently, antibiotic resistant *S. aureus* colonisation establishes. As the need for suitable alternatives to antibiotics is growing; bacteriophages are being screened for therapeutic suitability against *S. aureus*. This study aimed to determine the lytic capabilities of novel bacteriophage active against *S. aureus* from people with CF on solid and in liquid cultures.

Methods: Lytic profiles of 20 novel bacteriophage were generated against *S. aureus* clinical isolates, including methicillin resistant variants, and ATCC strains. Susceptibility was indicated via host-range tests in which 10µl is spot onto agar overlays inoculated with *S. aureus* bacteria. For specificity, each bacteriophage was screened, using this method, against other bacteria such as *Streptococcus* sp. and *Pseudomonas aeruginosa*. Growth-kill curves were performed in liquid growth medium: *S. aureus* grown to OD600=0.1 was infected with bacteriophage at multiplicity of infections (MOI) of 0.1, 0.5, and 1.0. This was done to determine effective dosing and compare bacteriolytic activity over 10 hours of incubation at 37°C.

Results: Environmental bacteriophage exhibited variable lytic profiles against *S. aureus* clinical isolates. Five bacteriophages had narrow host ranges, causing complete lysis in 26% of *S. aureus* isolates including activity against MRSA that others could not infect. In total, environmentally isolated bacteriophage could partially, if not completely, lyse over 90% of the *S. aureus* isolates. Growth-kill curves show that many bacteriophages substantially reduce bacterial density in liquid cultures (<OD600=0.1) at MOIs of 0.1, 0.5, and 1.0.

Conclusion: Novel bacteriophage isolated from environmental sources exhibit strong bacteriolytic profiles against a range of clinical *S. aureus* isolates derived from patients with CF in both solid and liquid growth mediums.

A053

RECOMBINANT HUMAN PARAOXONASE-2 AS AN ANTI-PSEUDOMONAL THERAPY

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Introduction/Background/Aim: In people with CF, dehydrated airway mucus facilitates chronic infection by *Pseudomonas aeruginosa*. During infection, *P. aeruginosa* produces signalling molecules ($3OC_{12}$-HSL) to coordinate formation of antimicrobial, immune cell resistant biofilms and production of host-damaging virulence factors, in a process termed quorum sensing (QS). Additionally, $3OC_{12}$-HSL also has many detrimental effects on host cells. Therefore, inhibition of bacterial QS should reduce *P. aeruginosa* pathogenicity, assisting in clearance of infection and reduce host cell damage. Thus, the *P. aeruginosa* QS system is a potential therapeutic target. Paraoxonase-2 is an intracellular human enzyme able to inactivate $3OC_{12}$-HSL only after it has entered host cells. Our laboratory has produced a recombinant form of human Paraoxonase-2 (rhPON2) that can be administered extracellularly. We aimed to demonstrate that rhPON2 attenuated *P. aeruginosa* QS and virulence, protecting host cells from the adverse effects of *P. aeruginosa* infection.

Method(s): We performed phenotypic assays to assess bacterial biofilm formation, motility, and virulence and utilised a proteomic approach to assess the global effect of rhPON2 on *P. aeruginosa*. RT-qPCR and ELISAs were used to assess expression and secretion of key inflammatory cytokines by airway epithelial cells pre-treated with rhPON2 and infected with *P. aeruginosa*.

Results: Our results demonstrated that rhPON2 treatment of *P. aeruginosa* inactivates $3OC_{12}$-HSL and reduces bacterial motility, biofilm formation and virulence factor production. *P. aeruginosa* proteome analysis revealed that production of proteins necessary for QS, phenazine biosynthesis and cAMP resistance are reduced in response to rhPON2 treatment. Pre-treatment of airway epithelial cells with rhPON2 before *P. aeruginosa* infection significantly reduced host cell inflammatory responses and reduced expression of key *P. aeruginosa* QS regulators.

Conclusion: Together these results demonstrate that rhPON2 protects host cells against *P. aeruginosa* induced inflammation and attenuates bacterial QS and virulence, supporting the use of rhPON2 as an anti-Pseudomonal therapy.

A065

REGIONAL TRANSCRIPTIONAL DIFFERENCES DURING IN VITRO RHINOVIRUS INFECTION IN LUNG ALLOGRAFT RECIPIENTS

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Introduction/Background/Aim: Lung transplant (LTx) recipients are at risk of complications from common respiratory virus infection including rhinovirus (RV). Here, we profiled the gene expression of large and small airway epithelial cells (referred as LAEC and SAEC, respectively) from in vitro RV infection and non-infected. To establish new insights into the molecular mechanisms during this event.

Method(s): Matched primary LAEC and SAEC obtained from LTx recipients (n=4, 45±8.1 years; 1 male) were established in cultures and inoculated with RV at MOI 12 for 24 hours. Next, dual RNA-seq approach was used to determine the viral load and host's gene expression profile.

Results: Meta-transcriptomics analysis confirmed RV infection. Gene expression profile for non-infected LAEC and SAEC identified pro-inflammatory signatures of Interleukin (IL)-17 signalling and surfactant metabolism, respectively. Comparison of the infected LAEC and SAEC to the non-infected counterparts has identified common and unique genes in response
to RV infection. Pathway enrichment analysis of common genes showed conservative antiviral
responses, as IFN and TLR signaling. Additionally, unique genes in LAEC showed strong signature related to interleukin-1 signaling in response to RV infection, whereas unique genes
for SAEC were linked to sulfur compound catabolism.

Conclusion: Comparison of non-infected LAEC and SAEC identified regional hallmarks. RV infection of LAEC activated IL-1 signalling, key mediator of inflammation and fibrosis which might possess detrimental impact on the transplanted lung. Alteration of SAEC transcriptome
signature during RV infection warrants further investigation. Collectively, this study provides starting points for the development of rational targeted therapeutic strategies.

A069

INVESTIGATING THE FUNGAL AND BACTERIAL LOWER AIRWAY MICROBIOME OF PAEDIATRIC PATIENTS WITH CF

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Introduction: Cystic Fibrosis (CF) lung disease is characterised by inflammation and infection which leads to destructive end organ damage[1]. The bacterial component of the CF airway microbiome has reduced diversity of organisms relative to the healthy population. With increasing age, CF patients' lungs become colonised with pathogenic organisms and microbial diversity reduces[2]. Lower community diversity has been shown to correlate with lower lung function[2]. Little is known about the CF fungal component of the lower airway microbiome (mycobiome).

Aims: Examine bacteria and fungi within the lower airway microbiome of paediatric patients referred to a single centre.

Methods: Bronchoalveolar lavage (BAL) samples were collected from patients between 2017 to 2020. BAL microbiota was examined using 16S rRNA or internal transcribed spacer (ITS) region amplicon sequencing on the Illumina MiSeq platform. The hypervariable region V4 of the 16S rRNA gene was amplified using the earth microbiome primers (515F-806R) and the ITS region was amplified using the fITS7-ITS4 primers.

Results: Forty-five children with CF (51% female, Age 0-17 years) and 28 children without CF (50% female, Age 0-17 years) provided BAL samples. Thirty CF patients had BAL samples collected over two or more consecutive years. The lower respiratory system microbiome of children with CF possessed significantly lower bacterial richness and diversity (Simpson index) compared to non- CF children ($p=0.007$). This was not seen within the mycobiome, rather there was wide variability in alpha diversity compared to the non-CF group. Alpha diversity (Simpson Index) was weakly negatively correlated with age in children with CF for both the bacterial microbiome (($R_2=0.157$ $p=0.007$) and mycobiome ($R_2= 0.167$ $p=0.015$). This was not seen in children without CF ($p>0.05$).

Conclusion: These initial results suggest that the microbiome of the lower respiratory system in children with CF in NSW, Australia is less diverse than children without CF and becomes increasingly less diverse with age, aligning with studies across the globe. Further analysis is warranted.

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A075

CLINICAL ASPERGILLUS FUMIGATUS ISOLATES INDUCE SIMILAR INFLAMMATION FROM PRIMARY AIRWAY EPITHELIAL CELLS

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Introduction/Aim: *Aspergillus* is becoming recognised as a pathogen of concern in early cystic fibrosis (CF), where it is associated with lung inflammation and mucus plugging. Unlike bacterial CF pathogens, clinical strains of *Aspergillus* appear to be highly diverse. Whether clinical *Aspergillus* strains greatly vary in their capacity to induce epithelial inflammation is unknown. We utilised two in vitro culture models of primary airway epithelial cells (pAEC) to test our hypothesis that different CF colonising strains of Aspergillus fumigatus would induce different inflammatory responses.

Methods: Ten paediatric CF *Aspergillus fumigatus* clinical isolates were compared, along with two systemically invasive clinical isolates and an ATCC reference strain (46645). Isolates were characterised by ITS (internal transcribed spacer) gene sequencing and screened for antifungal susceptibility. Submerged monolayer pAEC cultures (CF and non-CF; n=3 each) were exposed to conidia from all isolates at multiplicity of infection (MOI) 0.01 and 1. Two CF isolates were then selected for differentiated, air-liquid interface (ALI) cultures of pAEC (CF and non-CF; n=6 each) at MOI 0.01, along with one invasive isolate and ATCC strain. After 24 hours, inflammatory (IL-8 and IL-6) and cytotoxicity markers (LDH) were measured.

Results: Three clusters (A-D) of isolates were identified by ITS and antifungal susceptibility was variable. All isolates exhibited germination over 24 hours but did not induce IL-8 or IL-6 for any strain in monolayer pAEC culture ($p>0.05$). When fully differentiated, only one of the CF isolates caused increased IL-8 levels compared to uninfected, by both CF and non-CF cells ($p<0.05$). Although this was different to uninfected, it was significantly different to the other isolates ($p>0.05$).

Conclusion: Clinical *Aspergillus* isolates appear to be largely homogenous in immunostimulatory capacity and thus the presence of *Aspergillus* is more important to disease than the strain.

A077

DIETARY IRON INTAKE IMPACTS THE MICROBIAL COMPOSITION OF THE MURINE LUNG AND INTESTINAL MICROBIOME.

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Introduction/Background/Aim: Iron is an essential nutrient to many bacterial pathogens as well as the host. In this study we assessed the impact of different dietary iron intakes on the microbiota of the lung and intestinal tract of wildtype mice.

Method(s): Male C57BL/6 mice were fed either a diet supplemented with ferrous ammonium sulphate (FeAS), a high-iron diet or an iron deficient diet for four weeks. Tissues from the lung, duodenum and colon were collected and 16S rRNA gene fragments were pyrosequenced.

Results: Liver iron levels and weight differed significantly between the low and higher iron diets. In the murine lungs, multivariate and univariate methods indicated no association between microbial composition or community diversity and iron diet, but total serum iron levels and weight were associated with richness of the lung microbiome. *Bacteroides* sp. were significantly enriched in the lungs of mice fed the FeAS diet. Analysis of the intestinal microbiome indicated significant changes in overall microbial composition at OTU level between the diets and increased richness in the low iron compared to the iron supplemented mouse groups. In the duodenum, *Leptospira* and *Enterobacteraiceae* were reduced and *Desulfovibrio* species increased in the iron-supplemented diets groups compared to the low iron diet group. In the colon microbiota, *Veillonella*, *Enterobacteriaceae*, *Lactobacillus* and *Prevotella* species were reduced, and *Bacteroidetes family* S24-7 increased, in the higher iron compared to low iron diet mice.

Conclusion: This proof-of-concept study demonstrates associations between differing iron diets and lung and intestinal microbiota in C57BL/6 mice, and also suggests that systemic iron levels may impact on the lung microbiota. Dietary supplementation with iron is common in people with CF, but the impact on the gut and lung microbiota needs to be considered.

A080

REDUCING UNNECESSARY TESTING ON SPUTUM SPECIMENS FROM PATIENTS WITH CYSTIC FIBROSIS: PATHOLOGY STEWARDSHIP IN MICROBIOLOGY

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Introduction/Background/Aim: Once infected, the respiratory tract of patients with Cystic Fibrosis (CF) is commonly colonised for long periods of time with *Pseudomonas aeruginosa* and non-tuberculous *Mycobacteria* species. There is growing evidence on the limited value of *P. aeruginosa* routine susceptibility testing in CF patients with chronic *P. aeruginosa* infection. Furthermore, guideline recommendations on *Mycobacteria* species cultures suggests annual surveillance in stable patients.

Method(s): From August 2019 we ceased routine susceptibility testing on *Pseudomonas aeruginosa* isolates from patients with CF if a previous isolate from the patient had susceptibility testing performed. During this time, we also ceased routine mycobacteria culture if this had been performed within the previous 6 months. A comment was applied to the report inviting the clinician to contact the laboratory if further testing required. Data was extracted on respiratory samples collected between two seventeen-month study periods; one prior to the change in practice (August 2018 to December 2019) and the second following the change (August 2019 to December 2020).

Results: During the study period, our laboratory received 1,912 respiratory specimens for microscopy, culture and sensitivity (M,C,S) testing and 913 respiratory specimens for mycobacterial culture. Between the comparator study period and the period following the change there was a reduction in *P. aeruginosa* sensitivity testing from 95.9% down to 10.7% of isolates and a reduction in mycobacterial cultures by 8.6%. Using this data, we determined the financial and resource savings calculated from the cost of consumables, workload cost and time savings using the Canadian Workload Measurement Standard. A significant financial and resource saving was observed.

Conclusion: Aligning pathology specimen processing with guideline and evidence based testing for *P. aeruginosa* isolate susceptibility and non-tuberculous *Mycobacteria* species infections in CF patients can reduce costs, save resources and be an important laboratory stewardship activity.

MOLECULAR BIOLOGY/PHYSIOLOGY

A044

THE IMPACT OF RHINOVIRUS INFECTION ON MUCIN AND ASSOCIATED PROPERTIES INDUCED BY AIRWAY EPITHELIAL CELLS OF YOUNG CHILDREN WITH CYSTIC FIBROSIS

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Background/Aim: Prior transcriptomic analyses of airway epithelial cells (AEC) from children with cystic fibrosis (CF) revealed unique upregulation of pathways involved in mucin biosynthesis upon infection with rhinovirus (RV). Here, we aimed to assess whether RV infection influenced how CF mucus is different to non-CF mucus. We tested the hypothesis that the alteration of intracellular glycosylation machinery by RV infection contributes to CF mucin differences.

Methods: Fully differentiated air-liquid interface AEC cultures (CF and non-CF) were infected with RV (MOI 0.1) for 24h. The RNA, culture sections and apical washes were collected for downstream analysis using quantitative PCR, immunofluorescence staining and mucin agarose western blotting, respectively.

Results: Gene expression of glycosyltransferase genes *B3GNT2*, *B3GNT8*, *MAN1A1* and *ST8SIA4* were significantly upregulated in CF AEC following RV infection ($p<0.05$); whilst *FUT8*, *GALNT10*, and *NEU3* were significantly downregulated ($p<0.05$). Basal expression of secretory mucin genes *MUC5AC* and *MUC5B* was lower in CF AEC compared to non-CF. Both non-reduced and reduced forms of MUC5AC protein were also significantly lower in CF samples, whilst MUC5B produced by CF samples was mainly in a non-reduced form (10-fold greater than reduced). Upon RV infection, significant induction of *MUC5AC* gene expression (11.2-fold) was observed in CF AEC, though much lower (18.7-fold) compared to non-CF ($p<0.05$). However, no significant change in the level of MUC5B mucin was observed post RV infection. The level of MUC5AC and MUC5B mucin was similar between CF and non-CF AEC following RV infection.

Conclusion: The aberrant expression of glycosyltransferases seen may be likely contributors to altered glycan production on mucins produced by CF cells, influencing the core structure of airway mucous. Our results suggest mucin structure remains dysregulated following RV infection and since RV is a very common infection, provides new insights into the development of mucous pathology in the CF microenvironment.

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A049

IN VITRO CHARACTERISATION OF NASAL EPITHELIAL AND INTESTINAL ORGANOIDS REVEAL RARE *CFTR* MUTATIONS, R352Q AND I37R, TO BE RESIDUAL FUNCTION MUTATIONS RESCUABLE BY CFTR POTENTIATORS

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Background: A significant challenge to making targeted modulator therapies accessible to all patients with cystic fibrosis (CF) is the lack of understanding of the mechanistic dysfunction caused by over 2000 variations in the *CFTR* gene. Patient-derived cell models have been critical in characterisation of modulator responsiveness of rare *CFTR* mutations.

Aim: To characterise the functional defect of two rare *CFTR* mutations, R352Q and I37R, in patient-derived cell models of the airway and gut.

Methods: We created differentiated nasal epithelial cultures and intestinal organoids from CF participants with R352Q/DF508 (n=1), I37R/DF508 (n=1), DF508/DF508 (n=5) and G551D/DF508 (n=2) genotypes, and healthy control participants (n=6). CFTR-mediated chloride transport was assessed in nasal epithelial cultures via an ion transport assay and in intestinal organoids via a forskolin-induced swelling (FIS) assay. Two potentiator agents (VX-770 and GLPG1837), and a CFTR protein-folding corrector (VX-809) were tested. Correlation of data between nasal epithelial cultures and intestinal organoids was determined. Data from R352Q- and I37R- CFTR were compared to mutations with known impacts on CFTR function including folding defect and gating defect, as well as to data from wild-type functioning CFTR.

Results: Data from both cell models showed significant, positive correlation ($r=0.75$, $P<0.0001$) for all CFTR genotypes tested. This cross-validates the CFTR functional data derived from both cell models despite the different cellular source. Both R352Q- and I37R-CFTR demonstrated residual CFTR function which were significantly restored by CFTR potentiators ($P<0.05$) but not CFTR corrector agents.

Conclusion: The combined use of patient airway and gut cell models to characterise rare *CFTR* mutations may improve the specificity and sensitivity of the companion diagnostic platform and aid their translational use in CF.

A074

SERUM-CIRCULATING EXTRACELLULAR VESICLES ARE A POTENTIAL SOURCE OF BIOMARKERS FOR EARLY DETECTION OF CYSTIC FIBROSIS-RELATED DIABETES

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Background/Aim: Cystic fibrosis related diabetes (CFRD) affects between 40-60% of adults with CF. Recent evidence has shown that children with CF can have glucose abnormalities in early childhood before routine screening for CFRD begins (6-10 years of age). The current gold standard test for CFRD is cumbersome and has low sensitivity and specificity for CFRD. Therefore, there is a pressing need for development of new non-invasive early CFRD detection tools. Extracellular vesicles (EV) are carriers of biomolecules including proteins, DNA and RNA and are essential mediators of cell-to-cell communication. In recent years EVs have been shown to harbour effective diagnostic and prognostic biomarkers for pathological conditions including diabetes. However, their potential use as a biomarker for CFRD has yet to be investigated. In this study, we aimed to identify potential protein biomarkers within serumcirculating EVs that may be useful for the diagnosis and/or prognosis of CFRD.

Methods: We isolated EVs from low volumes of serum using commercially available EV isolation columns from adults with CF with CFRD (n=21), impaired glucose tolerance (IGT, n=8) or normal glucose tolerance (NGT; n =13) and healthy non-CF adults (n=9). Presence and size of EVs was confirmed using nanoparticle tracking analysis and electron microscopy. The proteome of EVs was assessed using label-free quantitative mass spectrometry.

Results: We showed that the proteome of serum-circulating EVs of people with CFRD is distinct from those with IGT and NGT which was enriched for proteins important in the adaptive immune system. Furthermore, we show that abundance of the major gel-forming mucin MUC5B is associated with a worsened state of pulmonary function and diabetes in adults with CF.

Conclusion: Collectively, our data shows that EVs are a promising source of biomarkers for CFRD in adults and may be important in predicting the future diabetes status and pulmonary condition of children with CF. The association of MUC5B abundance with CFRD severity and pulmonary dysfunction warrants further investigation.

PHYSIOTHERAPY

A003

TOWARDS RECORDING AIRWAY CLEARANCE, PHYSICAL ACTIVITY AND FITNESS ON THE AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY: A CONSENSUS APPROACH

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Background: Physiotherapy is a cornerstone of cystic fibrosis (CF) management, yet the Australian CF Data Registry (ACFDR) currently does not record physiotherapy-related data. This study aimed to gather opinions from lead Australian CF physiotherapists regarding the importance and feasibility of collecting physiotherapy-related data on the ACFDR.

Method: A three-round online Delphi survey was conducted to gather expert stakeholder opinion and consensus agreement. Lead physiotherapists from all 23 Australian CF centres were invited to participate. Round one explored the potential benefits, barriers and importance of recording three physiotherapy-related domains on the ACFDR: airway clearance, physical activity and fitness. Subsequent rounds were developed based on the findings from the previous round and sought consensus (80% agreement) for the inclusion of physiotherapy-related data on the ACFDR and for the most appropriate methods of collecting such data.

Results: The response rate was >80% for all rounds. Participants agreed that collection airway clearance, physical activity and fitness data on the ACFDR was important and feasible. Findings suggested that airway clearance and physical activity should be collected using self-reported questionnaires, while fitness should be measured using a field-based test.

Conclusion: Australian lead CF physiotherapists believe that collection of airway clearance, physical activity and fitness on the ACFDR is important and feasible. Future work focussed on piloting data collection to ensure its feasibility in real-world clinical settings is needed. This study demonstrates how Delphi methodology can provide a contemporary summary of expert physiotherapist opinion that can help inform nation-wide health service improvement.

A025

BONE HEALTH AND BODY COMPOSITION IN CHILDREN WITH CYSTIC FIBROSIS

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Background: Cystic Fibrosis (CF) can adversely affect bone mineral density (BMD). International guidelines recommend routine screening from eight years. Australian guidelines recommend screening during adolescence or those considered at risk. We aimed to determine the BMD and body composition of children with CF at Perth Children's Hospital (PCH) and current scanning practices.

Methods: We conducted a retrospective, observational study of BMD using Dual Energy Absorptiometry (DXA), collected as routine care for all children with CF (n=130) aged 8-18 years in 2020. BMD z-scores at the lumbar spine and total body less head; body composition; age at first scan and time to follow up scan/s was collected for all scans performed prior to May 2021.

Results: One hundred and four children (80%) had at least one reportable DXA (mean age=12 years 7 months; SD= 2years 11 months; mean body mass index (BMI) =18.8; SD=3.74). Sixty children (58%) had normal BMD, 33 (32%) had a site 'at risk' of low BMD (z-score ≤-1.00 to -1.99) and 11 (11%) had low BMD (z-score ≤ -2). Sixteen children (16%) had two reported DXAs and two children had three, with the average time between scans 3.8 years (SD: 2.5). Of the 18 repeat scans, 16 had no change in BMD, one had improvement (Z-score +1.0) and one had deterioration (Z-score -1.0). Although low to moderate positive correlation exists between BMI and BMD within our cohort (Pearson's correlation (95%CI): Lx spine *r*=0.30 (0.12-0.47); TBLH *r*=0.43 (0.26-0.58)), 41% of children who were not underweight and 40% with normal lean mass had an area at risk of low BMD or with low BMD.

Conclusion: Children with CF with normal BMI or lean mass are still at risk of low BMD. Routine scanning of all children from eight years may be beneficial to monitor bone health.

A034

PERSISTANT AND SEVERE TRACHEOBRONCHOMALACIA IS ASSOCIATED WITH CHANGE TO POSITIVE PRESSURE AIRWAY CLEARANCE TREATMENTS IN CHILDREN WITH CYSTIC FIBROSIS

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Introduction: Tracheobronchomalacia (TBM) reduces airway stability during coughing. This can be problematic for children with cystic fibrosis (CF) who need to perform regular airway clearance (AWC) to remove lung secretions.

Aim: To investigate how TBM findings during bronchoalveolar lavage (BAL) influence airway clearance routines in children with CF.

Methods: Children born between 2001 and 2016 who were under the care of the Perth Children's Hospital CF team were included. Operation reports from BAL performed until the age of four were retrospectively reviewed to determine if TBM was ever present, whether it was persistent (defined as present on more than one BAL) or if it was ever classified as 'severe' (as described on the operation report). The medical records of those with a TBM diagnosis were reviewed and data collected on AWC regimes.

Results: Of 181 children screened, 171 contributed data. 649 BALs were reviewed, mean of 4 per child (1-6). 68 (40%) children had a finding of TBM. Of these, 64 (94%) commenced an AWC routine at CF diagnosis of manual techniques and positioning. Post TBM finding, 42 (62%) children had a change in AWC routine with 31 (70%) reported as directly related to TBM diagnosis. 24 (57%) children changed AWC within 3 months of first TBM finding and 5 (12%) after their second. All were changed to positive pressure (PEP) based therapy.

In those children who had persistent and severe TBM (n = 26) 70% changed to PEP within 3 months and 95% within 12 months. In the remaining 6 children, 1 was already using PEP therapy, and 4 had significant medical or social issues which impacted on their AWC options.

Conclusion: CF children diagnosed with TBM are likely to have their AWC regimes changed to positive pressure therapy. Those with persistent and/or severe malacia were most likely to change routines more quickly.

A048

TELEHEALTH PHYSIOTHERAPY FOR PAEDIATRIC PATIENT'S WITH CYSTIC FIBROSIS TO SUPPORT PHYSIOTHERAPY ROUTINES

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Introduction/Background/Aim: Adherence to treatment routines in patients with chronic respiratory conditions like Cystic Fibrosis (CF) is a challenge and can result in more frequent hospital presentations and reduced quality of life. In 2020, The Royal Children's Hospital (RCH), utilised telehealth physiotherapy in addition to routine outpatient appointments to support adherence with airway clearance (AC) and exercise.

The aim of this audit was to determine who was offered additional physiotherapy telehealth appointments to support AC and exercise during COVID-19, the reason for these appointments and the sessions effectiveness.

Method(s): A quantitative and qualitative single centre retrospective audit of physiotherapy telehealth was completed between March 2020 and February 2021. Routine physiotherapy appointments that were provided via telehealth during this time were not included in the audit. Data collected via chart review included child's diagnosis, date of birth, gender, date and reason for review, referrer, attendance, treatment provided, and therapist reported effectiveness and satisfaction.

Results: 31 patients with CF (age range two – 18 years) were reviewed by a physiotherapist via telehealth to support adherence with airway clearance (AC) and/or exercise. 127 telehealth sessions were offered with a 74% attendance rate. Referral sources included: physiotherapist in most cases (74%), CF Doctor (8%) and CF Nurse (8%). Therapist reported effectiveness themes included improved patient engagement and adherence in AC and exercise.

Conclusion: Physiotherapy telehealth sessions support adherence with physiotherapy routines in paediatric patients with CF. These sessions helped maintain adherence with therapies and was deemed an acceptable way of providing physiotherapy to patients by physiotherapists and patients and their families during the COVID-19 pandemic related restrictions. Further evaluation of patient's perceptions and acceptance of telehealth physiotherapy to support AC and exercise will be important to investigate.

A050

CAN EXERCISE CAPACITY BE MEASURED REMOTELY IN YOUNG PEOPLE WITH CYSTIC FIBROSIS?

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Background: Assessment of exercise capacity annually is recommended as part of routine CF management. In school-aged children, it is measured using CPET and MST-25. In the setting of COVID-19, these modalities are considered high-risk aerosol-generating procedures, interrupting access to testing. Actigraphs (worn as a wrist watch) are validated, medical-grade activity monitors. They can be used as a remote monitoring tool to measure the amount and type of physical activity performed (e.g. sedentary, light, moderate, vigorous).

Aim: To determine whether remote monitoring of physical activity with an Actigraph can be used as a surrogate for formally measured exercise capacity in young people with CF by exploring whether physical activity data from the Actigraph:
1) correlates with exercise capacity measured by formal exercise testing
2) correlates with other clinical measures known to be correlated with exercise capacity

Methods: Actigraphs given to school-aged children during a routine clinic visit (worn on the wrist ~7 days, excluding showering/swimming) over a 6-month period will be analysed. Data downloaded from Actigraphs (METs, step-count, % time in type of activity) will be explored to determine whether correlation exists with formal exercise tests measured by CPET (Bruce Treadmill Protocol) and routine clinical measures known to be correlated with exercise capacity (lung function, lean mass, bone density, glucose tolerance) retrospectively collected from the medical record within the prior 12 months.

Results: To date, 25 children have data from the Actigraph, median age 12 (7-18) years.

Conclusions: Understanding whether remote monitoring of physical activity can be used as a surrogate for exercise capacity will enhance the ability of CF centres to monitor exercise capacity, guide prescription of the amount and type of activity to promote in young people with CF and develop individualised exercise prescription in an evolving COVID-19 landscape where access to formal exercise testing is limited.

A055

A REVIEW OF PHYSIOTHERAPY TELEHEALTH FOR CYSTIC FIBROSIS FOR HOSPITAL IN THE HOME

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Introduction: HiTH Physiotherapy at the RCH has utilised telehealth for airway clearance for patients with cystic fibrosis since 2013. During the Covid-19 pandemic, we were required to increase this mode of delivery. We sought to investigate satisfaction with this service from families.

Method(s): A quantitative prospective and retrospective audit of admissions for CF patients receiving HITH. Data was collected in regards to LOS, age, TH experience (technical issues, effectiveness, impact) and overall satisfaction with HITH TH physiotherapy.

Results: 30 parents responded to the survey in 2020. 83% of patients were aged 5-15yrs. There was 100% satisfaction with TH physiotherapy, with 64% finding it just as effective as face-to-face (FTF) physiotherapy. 90% found it a positive contribution to the admission, especially during the pandemic. 70% had no connectivity issues during TH session. The impact of TH versus FTF was convenience (60%), improved compliance with ACT (43%) and increased clearance of sputum (43%). All respondents recommend TH physiotherapy.

Conclusion: Families are very satisfied with HiTH TH physiotherapy. Our next study will be to investigate what service changes we can implement for adolescents with CF to fit in with increased school demands. The final word goes to a parent: "Really appreciated the extra support we were given here. Staff were fabulous at trying to get best results out of each session. Much appreciated."

A057

TELEHEALTH CYSTIC FIBROSIS PHYSIOTHERAPY ANNUAL REVIEW – PHYSIOTHERPIST PERSPECTIVES

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Background: In response to COVID-19 many Cystic Fibrosis (CF) clinics transitioned majority of outpatient appointments to telehealth. At the Royal Children's Hospital (RCH) all patients with CF over the age of 5 years are reviewed for a physiotherapy annual review where airway clearance, inhalation therapy, exercise, musculoskeletal systems and continence are reviewed. In 2020 the RCH physiotherapy team trialled physiotherapy annual reviews via telehealth.

Method: At RCH in 2020 physiotherapy annual reviews were performed via 'Health Direct' a web-based video call platform from May till the end of 2020.
To evaluate staff perceptions an electronic survey was distributed to physiotherapists between May and October 2020. The survey was developed by the physiotherapy team to evaluate perceptions regarding safety, effectiveness, education, disadvantages related to telehealth physiotherapy annual reviews.

Results: We received 18 responses from May until October 2020. 34% of respondents reported that they strongly agreed (1/18) or agreed (5/18) they were able to complete a safe and effected assessment. 55% of respondents reported that they strongly agreed (3/18) or agreed (7/18) they were able to complete a safe and effective treatment. 82% (15/18) of respondents reported they were able to provide effective education, while 44% (8/18) felt that telehealth was a valuable way of providing physiotherapy care to this patient. Despite these results 62% of respondents reported that they would not recommend continuing telehealth annual reviews post COVID-19.
Three consistent disadvantages of telehealth physiotherapy annual review were highlighted: lack of ability to perform exercise tests and musculoskeletal assessment, inability to trial new airway clearance devices or techniques and technology difficulties.

Conclusion: Telehealth Physiotherapy annual reviews were found to be safe and a good opportunity to perform education. Considerations regarding long-term use should address concerns regarding lack of exercise testing, musculoskeletal examination, trials of new devices and techniques and technical difficulties.

A058

ROYAL CHILDREN'S HOSPITAL (RCH) CYSTIC FIBROSIS MULTIDISCIPLINARY TEAM PERSPECTIVES REGARDING THE USE OF TELEHEALTH FOR CYSTIC FIBROSIS CLINIC

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Background: In response to COVID-19 many CF clinics transitioned to telehealth. We previously described patient and family satisfaction with telehealth clinics, however staff satisfaction remains unknown.

Method: At RCH telehealth clinics occurred via the 'Health Direct' web-based video call platform. Families were sent airway sampling kits including written and video instruction, and were asked to complete anthropometric measurements. Spirometry was completed via telehealth for those who had a home device.
To evaluate staff perceptions an electronic survey was distributed to all members of the CF team who attended 6 randomly selected clinics in September and October 2020. The survey was developed by CF team members to evaluate perceptions regarding safety, effectiveness, education, disadvantages and workload related to telehealth clinics.

Results: The response rate was 78% (42/54) and covered all disciplines of the multidisciplinary team. 72% of responses reported they strongly agreed (5/42) or agreed (25/42) they were able to complete a safe assessment. 71% reported they strongly agreed (6/42) or agreed (24/42) they were able to complete an effective assessment. 86% (36/42) of responses reported they were able to provide effective education, while 63% agreed (22/42) or strongly agreed (3/42) that telehealth clinics provided equivalent health outcomes and 59% (24/42) reported they would recommend continuing telehealth post COVID-19.
Three consistent disadvantages of telehealth were highlighted: lack of objective data and assessments, concerns regarding engagement and communication, and technology difficulties.
55% (5/9) of CF team members reported that time spent preparing and performing follow-up related to the clinic had increased with telehealth.

Conclusion: Telehealth CF clinics were well received by the CF team, and were thought to be safe and effective. Future considerations regarding long-term use should address concerns regarding efficiency, lack of objective measurements, technical difficulties, the impact on engagement and communication and that long term health outcomes are comparable.

A059

COMMUNITY-BASED PHYSICAL ACTIVITY MONITORING USING CONSUMER-GRADE DEVICES FOR CHILDREN WITH CYSTIC FIBROSIS; A LONGITUDINAL OBSERVATIONAL STUDY

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Background: Consumer-based physical activity monitors may be a clinically useful measure of community-based physical activity in children with cystic fibrosis. This study reports on the adherence, attrition, the inter and within-day physical activity variations using activity monitors over 18 months.

Method(s): Retrospective analysis of data collected from 43 children (12.0 +/- 2.3 years; 17 girls) with cystic fibrosis from February 2018 to February 2019. Hourly step count and daily intensity minutes (IM) were recording using the Garmin Vivosmart3®.

Results: Children adhered to wearing activity monitors in the first three months, however only 11 of 43 participants wore their devices at 12 months. More steps and moderate-to-vigorous physical activity were observed on weekdays and school days compared to weekends and school holidays respectively. Children took less steps during periods of hospital admission when compared to outside of hospital. High granularity analysis in hourly increments suggests peak daily step volume occurred on school days before school and during lunch time. Compared to school days, children achieved less steps before 3pm on weekends and school holidays.

Conclusion: This is the first published study on hourly step data for children with cystic fibrosis. Children averaged 44 minutes of moderate-to-vigorous physical activity each day but took less steps per day than previously published data of their healthy peers. Clinicians can utilise activity monitors to promote physical activity in children with cystic fibrosis due to the low-cost and ease of access to high granular data.

A063

TRIAL OF THE AFFLOVEST DEVICE AS AN ADJUNCT TO USUAL AIRWAY CLEARANCE IN A 79 YEAR OLD PATIENT WITH CYSTIC FIBROSIS.

REBECCA NETLUCH

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Aim: To assess the effectiveness of High Frequency Chest Wall Oscillation via the AffloVest as an adjunct to usual airway clearance of a 79 year old female with Cystic Fibrosis who has severe lung disease and high sputum load.

Method: Case report. Pre commencement of the VEST the patient had baseline measurements of Six Minute Walk Test (6MWT), Lung Function via Spirometry, and self-reported effectiveness of sputum clearance. This was repeated at 3 months.
The patient's usual airway clearance included Hypertonic Saline followed by daily manual therapy (Percussion and Vibration + Aerobika Positive Expiratory Pressure Device) with second treatment using Aerobika device. Traditionally the patient finds manual treatment most effective for sputum clearance however 2 x daily treatment in the community was not feasible. Therefore option of AffloVest was considered within the MDT as a viable alternative to trial to facilitate improved clearance with potential implications on QoL and hospital admissions.

Results: At baseline the patient had a 6MWT of 343m (BORG 4-5), Spirometry values of FEV1 = 0.87 (39%) FVC = 1.60 (55%). At three month follow up the 6MWT had improved by 64m to 407m (BORG 1-3). Spirometry values had improved to FEV1= 0.99 (45%) and FVC= 2.10 (73%). Self-reported effectiveness of airway clearance increased from 2/10 (Aerobika only) to 6/10 (VEST and AffloVest). Hospital admissions were unchanged at 2 in the 6 months prior to and post Vest implementation.

Conclusion: Implementation of the AffloVest was well tolerated and had an improvement in the patient's self-reported sputum clearance effectiveness as well as 6MWT, and Spirometry. Review of the patient over a longer timeframe will provide further information on its effectiveness.

A064

NAVIGATING RESTRICTIONS WHILE KEEPING STAFF AND PATIENTS SAFE: HOSPITAL IN THE HOME (HITH) PHYSIOTHERAPY'S RESPONSE TO COVID-19

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Background: The HITH service at SCHN treats acutely unwell patients at home, offering up to bi-daily physiotherapy sessions delivered face-to-face and/or via telehealth. Telehealth was implemented in 2017 and is a well-established mode of treatment delivery for our service. HITH Physiotherapy's response to COVID-19 was to develop a Standard Operating Procedure (SOP) to adapt home visits to the pandemic. Available literature on aerosol generating procedures (AGPs), infection control and airway clearance techniques in light of COVID-19 were reviewed. Other Australian paediatric HITH services were surveyed to gather information on their changes to service delivery. The SOP implemented additional screening to keep staff and patients safe and modified treatment delivery where appropriate.

Aim: To investigate the impact of the COVID-19 SOP on HITH Physiotherapy service delivery.

Method(s): A retrospective review compared the number of patient admissions, average time to transfer to HITH and treatment delivery type in March 2020-2021 to those of March 2019-2020.

Results: The SOP was successfully implemented to ensure staff and patient safety, whilst maintaining high levels of clinical care and adhering to public health guidelines. School visits were ceased to avoid AGPs in public locations. 87 patients were treated by HITH Physiotherapy in March 2020-2021 compared to 104 patients in March 2019-2020. 43.6% of patients were directly admitted to HITH in March 2020-2021 compared to 37.5% in the previous year. Average time to transfer was 7.2 days in March 2020-2021, compared to 8.9 days in March 2019-2020. In March 2020-2021, 55.1% of treatments were delivered via telehealth and 39.1% face-to-face (patient-declined visits account for the remaining %) compared to 57.7% and 42.3% respectively in March 2019-2020.

Conclusion: HITH has demonstrated the ability to adapt to evolving public health guidelines and maintain a high standard of clinical care, while keeping staff and patients safe during the COVID-19 pandemic.

A066

PHYSIOTHERAPY FOR CYSTIC FIBROSIS IN AUSTRALIA AND NEW ZEALAND: A BENCHMARKING SURVEY

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Background: Physiotherapy management of people with cystic fibrosis (CF) is guided by evidence based, best practice recommendations published in the CF standards of care, Australia and Physiotherapy for CF in Australia and New Zealand: a clinical practice guideline. The purpose of this study was to determine whether CF centres in Australia and New Zealand are meeting these standards and identify key areas for improvement.

Methods: An online survey was created based on the key physiotherapy components of the aforementioned publications. The survey was distributed via the pre-established CF Physiotherapy Australia and New Zealand Google group, with one senior representative from each centre asked to complete the survey.

Results: 23 of the 24 surveyed CF centres responded. Only 10% of centres met the recommended Physiotherapy EFT based on the size of their clinics. In regards to inpatient care, 100% of centres met the recommendation for minimum frequency of physiotherapy, assessment within 24 hours of admission, and provision of a weekend physiotherapy service whilst only 60% provided an after-hours physiotherapy service. The standards of care recommend that ‘adequate’ physiotherapy support is available to patients receiving home therapy for exacerbation however only 65% of CF centres provide a Hospital in the Home (HITH) Physiotherapy service.
In reference to outpatient care, 86% of centres provide access to a physiotherapist with expertise in CF management at each clinic visit and only 65% of centres provide formal physiotherapy annual review.

Conclusion: Physiotherapy EFT in CF centres across Australia and New Zealand is well below the recommended standards of care. Pleasingly, despite this, many of the physiotherapy recommendations in regards to care provision are met to an adequate standard. Additional funding and EFT could allow CF centres to provide after-hours physiotherapy, formal physiotherapy annual reviews and hospital in the home services.

A071

A TELEHEALTH EXERCISE SERVICE WAS FEASIBLE AND HAD HIGH SATISFACTION IN ADULTS WITH CYSTIC FIBROSIS

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Background: Exercise training is an integral component of care for people with cystic fibrosis (CF). Due to infection control recommendations, people with CF cannot exercise together, and several available options for exercise training were impacted by the COVID-19 pandemic. This was of particular concern for adults with CF as they found it difficult to meet recommended exercise training guidelines independently, and were therefore at risk of poorer outcomes.

Aim: To determine the feasibility of a physiotherapy led telehealth exercise service for adults with CF, as well as participant satisfaction with the new service.

Methods: Participants were adults with CF, recruited via outpatient clinics after completing a pre-exercise screening questionnaire. Individual and group exercise sessions were offered, and were delivered using a mobile, large-screen telehealth unit based at Sir Charles Gairdner Hospital. A post-exercise participant satisfaction survey was completed after a 3-month period.

Results: Sixteen participants (11 [69%] female, age 31 [11] years, ppFEV1 55 [25]%) utilised the telehealth exercise service over a 3-month period. In total, 64 sessions were offered (41 individual, 23 group), with 102 (6 [1 to 19] per participant) occasions of service generated. Eleven (69%) participants completed the satisfaction survey. All participants (100%) 'strongly agreed' that the organisation of the exercise sessions meeting their expectations. All participants (100%) also responded 'yes' to having telehealth exercise sessions remain as part of their routine CF care *(_"I_ _a_m_ _s_o_ _g_r_a_t_e_f_u_l_ _t_o_ _h_a_v_e_ _v_i_r_t_u_a_l_ _e_x_e_r_c_i_s_e_ _a_s_ _a_n_ _o_p_t_i_o_n_ _t_o_ _m_a_n_a_g_e_ _m_y_ _C_F_ _c_a_r_e_")_*. No participants reported that they preferred 'face-to-face' exercise sessions.

Conclusion: A physiotherapy led telehealth exercise service for adults with CF was feasible, well received and should be considered as an option for CF physiotherapy care for adults with CF in WA.

A072

VALIDATION AND TELEHEALTH APPLICATION OF A PERSONAL, ULTRASONIC SPIROMETER IN CHILDREN WITH CF AND OTHER CHRONIC RESPIRATORY DISORDERS

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Introduction: Accurate lung function monitoring is an essential element in the provision of respiratory assessment in the home during telehealth consultations.

Aim: To investigate the accuracy of two personal spirometers designed for home use. Secondly, to assess the ability of children to perform acceptable and reproducible (A&R) spirometry during telehealth consultations.

Methods: Children with respiratory disorders performed spirometry supervised by a respiratory scientist using a laboratory spirometer and either a personal ultrasonic (SpiroHome) or turbine (Air Next) spirometer. Bland-Altman analysis and intraclass correlation coefficients were calculated for FEV_1 and FVC. Subsequently, the ability of children to perform A&R spirometry using the devices during telehealth consultations supervised by a respiratory scientist was assessed.

Results: The SpiroHome and Air Next were validated against hospital laboratory equipment in 59 and 20 children respectively. There was a strong linear relationship between each device and laboratory equipment, with ICC 0.99 for both devices for both FEV1 and FVC measurements. Bland-Altman analysis revealed a mean bias and limits of agreement (LOA) of the SpiroHome compared to laboratory equipment of -0.01 litres (-0.22 to 0.24) for FEV1 and -0.02 litres (-0.30 to 0.33) for FVC. For the Air Next, the mean bias and LOA were -0.02 (-0.27 to +0.22) for FEV1 and -0.01 litres (-0.3 to +0.27) for FVC. A between device difference in FEV1 %predicted of $\geq$10% was found in 7% of children using the SpiroHome and 10% of children using the Air Next. Supervised spirometry during telehealth produced A&R results on 125/140 (89%) occasions using the SpiroHome and 51/60 (85%) occasions using the Air Next.

Conclusion: There was excellent correlation between the devices and laboratory equipment, however, LOA were wide. Therefore, caution is needed if the devices are used interchangeably with laboratory equipment. A high percentage of A&R spirometry was performed during telehealth, indicating the devices are suitable for this application.

A073

VALIDITY OF PARENT-TAKEN OROPHARYNGEAL SWABS FOR DETECTION OF BACTERIAL PATHOGENS IN CHILDREN WITH CF

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Introduction: In children who cannot expectorate, oropharyngeal swabs (OPS) provide one alternative to sputum samples.

Aims:
1. Compare the pathogen yield of OPS collected by physiotherapists and parents in children with CF.
2. Determine the reliability of physiotherapist taken OPS.

Methods: Children with CF who required an OPS were invited to participate. Children did not participate in the two parts of the study on the same day.

1. A physiotherapist educated a parent on how to perform OPS then collected an OPS on the child whilst the parent observed. This was followed by the parent performing an OPS.
2. A physiotherapist collected two separate OPS on the same child on the same occasion.

All samples were sent for standard CF pathogen culture.

Results: 76 children had an OPS collected by a physiotherapist and a parent. On a separate occasion, 30 children had two OPS collected by a physiotherapist. During comparison of physiotherapist and parent collected OPS, 23/71 paired swabs were positive for the same bacteria, 23/71 paired swabs were both negative for any growth. In 14/71 paired swabs, the physiotherapist taken swab was positive for a bacteria not present on the parent swab. In 11/71 paired swabs, the parent swab was positive for a bacteria not present on the therapist taken swab.
During comparison of bacteria isolated on the two swabs taken by the physiotherapist, 15/30 paired swabs were positive for the same bacteria, 9/30 paired swabs were both negative for any growth. In 6/30 swabs, one swab identified bacteria not isolated on the other swab. On 3/6 of these occasions, *pseudomonas aeruginosa* was only identified on the second swab.

Conclusion: OPS collected either by a physiotherapist and parent or both collected by a physiotherapist produce discrepant results in approximately 35% and 20% of cases respectively. On some occasions, clinically important bacteria are identified only after a second swab is performed.

A082

HOW DO PHYSIOTHERAPISTS USE NON-INVASIVE VENTILATION IN CYSTIC FIBROSIS CENTRES? AN AUSTRALIAN SURVEY.

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Background: Non-invasive ventilation (NIV) is used by physiotherapists as an adjunct to airway clearance and exercise[1,2] and can decrease fatigue during airway clearance[3] and reduce oxygen desaturation during exercise.[4] Details regarding the clinical use of NIV with physiotherapy for people with cystic fibrosis (CF) are not well documented however. This study investigated clinical usage of NIV during physiotherapy for people with CF in Australian centres. Questions explored the clinical indications, contraindications and patient selection criteria for NIV use as an adjunct to physiotherapy in people with CF? Who implements NIV, what settings are used and how are they determined? What outcome measures are used to determine the effectiveness of NIV as an adjunct to physiotherapy and what are the main benefits and complications?

Methods: A purpose-designed survey was sent to 23 Australian CF centres.

Results: Fifteen (65%) centres responded, with 13 reporting using non-invasive ventilation to assist physiotherapy. NIV was most commonly used (85%) in patients with lung function <40% predicted. Indications for use included shortness of breath at rest (100%) and during airway clearance (100%), and fatigue during airway clearance (100%). Physiotherapists were responsible for initiating NIV (62%), setting up (85%) and determining settings (62%). Bi-level ventilation was the only chosen ventilation mode. Benefits included, improved ease of airway clearance (100%), reduced fatigue (92%) and decreased dyspnoea (85%). Only one complication of haemoptysis was reported.

Conclusion: NIV was used during physiotherapy in people with CF with severe disease, mostly during airway clearance to improve tolerability of treatment. Australian physiotherapists initiated NIV when people with CF experienced shortness of breath or fatigue during treatment, aligning with current clinical guidelines. Clinical usage was largely consistent across centres, with numerous benefits and minimal complications reported. Further research is required to explore benefits of NIV use during physiotherapy.

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A083

THE PROGNOSTIC VALUE OF THE MODIFIED SHUTTLE TEST IN CHILDREN WITH CYSTIC FIBROSIS: A RETROSPECTIVE REVIEW

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Background: The Modified Shuttle Test (MST) is a valid field test of exercise capacity in children with Cystic Fibrosis (CF), and is routinely used in Australian paediatric CF annual reviews. However, there is a lack of literature investigating its prognostic utility for decreased exercise capacity. The aim of this study was to determine the prognostic value of the MST in predicting future respiratory exacerbation in children with CF.

Method(s): A retrospective review of MST results collected between 2008-2019 from two Australian CF clinics was carried out. A lack of improvement in MST distance over two consecutive annual tests indicated a decrease in exercise capacity. Prognostic outcomes included: lung function as determined by spirometry, presence of pulmonary infection and hospitalisation for a respiratory exacerbation in the following 12 months. Multivariate regression, including odds ratio analyses, determined the relationship between a reduction in exercise capacity and prognostic outcomes.

Results: 116 children with CF (66 males; mean age 11 years) performed 240 consecutive pairs of MSTs. 83 pairs of MSTs showed no change or a reduction in MST distance. This marker of decreased exercise capacity was a statistically significant, independent predictor of hospital admission in the following 12 months (p-value = 0.025). Odds ratio analysis showed that these children had a 90% increased risk (95% Confidence Interval, 1.084 - 3.306) of having a hospital admission in the following 12 months compared to those whose MST distance increased.

Conclusion: This study demonstrated that an equivalent or reduction in distance achieved in consecutive annual MSTs is associated with impending respiratory exacerbation requiring hospitalisation in the following 12 months in children with CF. As a result, annual MST results may identify children at risk of respiratory exacerbation and thus alert clinicians to the potential need for more intense preventative treatment strategies.

A088

HIGH FREQUENCY CHEST WALL OSCILLATION (HFCWO. HILLROM VEST®) SUPPORTING AIRWAY CLEARANCE UNTIL TRANSPLANT – A CYSTIC FIBROSIS (CF) CASE STUDY

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Male with CF (dF508 homozygous) and history of Meconium Ileus, bronchiectasis, Past M. Abscesses, severe CF Related Liver Disease with portal hypertension, severe gastroesophageal reflux (fundoplication and PEG). Significant early lung disease followed by an extended period of relative stability (despite LFTs ~40%) for six years. He then presented post viral infection, with impaired secretion clearance due to ineffective paroxysmal cough, characterised by airway instability.

Dyspnoeic and hypoxic at rest, requiring supplemental oxygen, his standard airway clearance strategies of combined mouthpiece Positive Expiratory Pressure (PEP) and 6% Hypertonic saline were not possible. QCH CF service infection control policy dictates that non-invasive ventilation devices are single patient use, presenting a challenge to the physiotherapist to support his acute management.

A rigid, barrel shaped chest, enlarged upper abdomen due to hepatosplenomegaly contributed to significant respiratory compromise. Shortness of breath, increased respiratory rate, and reduced inspiratory capacity adversely impacted expiratory flow. Additionally, airway irritability triggered paroxysmal cough compounding inadequate clearance. At this time, he was actively listed for transplant.

Was it about getting air behind secretions in a patient with an increased residual volume (RV) at 246% pred and reduced inspiratory capacity? Or was it about modifying expiratory flow, RV and shear forces to aid clearance?

We opted to use HFCWO (HillROM VEST®) to modify expiratory flow targeting small airways (similar to chest wall strapping, Eberlain 2014, Taher, 2018), while providing modest positive pressure support with the use of EzPAP® via mouthpiece to assist airway stability. Modelled on the Minnesota Protocol, he was able to tolerate optimal settings for the first three compressive cycles upon initial use, and clear with an effective strong cough. By discharge home we were able to combine PEP and HFCWO effectively and he continued to utilise this as his airway clearance strategy until transplant.

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PSYCHOSOCIAL / NURSING / EDUCATION

A007

CYSTIC FIBROSIS PROFESSIONAL DEVELOPMENT FOR EDUCATORS

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An educator is an essential part of children's lives. Children started their education once born, learning from their parents or guardians, their surroundings, then childcare or kindergarten, and then school. 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 3446 people in Australia (Australian Cystic Fibrosis data Registry, 2019), 1854 people under 18 years.

Monash Children's Hospital, The Royal Children's Hospital and Cystic Fibrosis Community Care have a commitment to assist in this professional development for educators. We have adapted the education to different formats according to the feedback from the participants.

This year, we completed five sessions and provided the professional development to over 150 educators from all around Australia (Victoria, ACT, NSW, Qld and WA). These sessions commenced at the beginning of the school year, being held fortnightly. Each session had a defined topic and participants were able to attend all or some of the sessions.

The interactive sessions were all deliver online, with pre and post questionnaires. We plan to continue to improve and amend the topics from the feedback from participants and continue to support and provide education to out patients' educators and teachers.

A013

“I CAN BREATHE”: EXPLORING POST-TRANSPLANT LIFE EXPERIENCES AND SATISFACTION WITH HEALTHCARE AMONG YOUNG PEOPLE WHO HAVE HAD A LUNG TRANSPLANT

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Introduction/Background/Aim: Lung transplantation is an established treatment for endstage
respiratory failure, with cystic fibrosis being the most common indication for lung transplantation in adolescents and young adults. Transplantation is a complex procedure and involves significant lifestyle changes that must be maintained throughout life to support longevity. The perceptions of young people regarding their experience of clinical care and living life with a transplant are underrepresented across the literature. This study aimed to explore the perspectives of young people to: (1) determine the impact lung transplantation has had on their daily lives, and (2) identify the aspects of care that they were satisfied with.

Method(s): A descriptive qualitative study was undertaken via consumer engagement. Sixteen young people who underwent lung transplantation in adolescence were engaged. Indication for lung transplantation varied, with young people having received diagnoses of cystic fibrosis (n = 6), pulmonary hypertension (n = 5), or other conditions (n = 5). Semistructured
interviews and photo elicitation were utilised to gain insights into participants’ experiences, and thematic analysis of interview data was undertaken.

Results: Seven broad themes emerged from the analysis: (1) daily life experiences and satisfaction with post-transplant life; (2) the impact of the COVID-19 pandemic; (3) the journey towards transplantation and preparation for post-transplant life; (4) the processes of and satisfaction with post-transplant care; (5) adjusting to a life of medication; (6) attitudes towards post-transplant restrictions; and (7) words of wisdom for other transplant recipients. Overall, participants expressed a high degree of satisfaction with their care and life after transplant.

Conclusion: The findings of this study have provided compelling insights into the processes of transplant care for young people, and their quality of life after transplant. This study provides directions for healthcare services to follow when delivering care to young people, while also highlighting areas for improvement.

Keywords: Lung transplant; healthcare; patient experiences; young people; consumer engagement

A015

REIMAGINING THE FUTURE OF CYSTIC FIBROSIS ADULT CARE

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Introduction/Background/Aim: Delivery of healthcare will never be the same post COVID-19. This worldwide pandemic provided Cystic Fibrosis (CF) clinicians the opportunity to reimagine how CF healthcare could be delivered. The Mater Health Services (MHS) CF team redesigned their model to deliver outpatient services in patients' home. With ongoing COVID-19 outbreaks and subsequent reductions in outpatient activity evaluation of the new model was paramount.

Method(s): The electronic medical record (VERDI) was utilised for a comprehensive audit. All CF patients cared for by the MHS CF team were included. Data was collected from April to September 2019, 12 months prior to the change in the model of care and for then from April to September 2020. Data collected included: number of clinic visits, influenza vaccination rates, medication pickup and lung function. A survey monkey was completed to determine patient satisfaction.

Results: Results of the audit proved to be interesting. Nonattendance rates for virtual clinic appointments (telehealth) was dramatically reduced from 20% to 5%. Provision of a home spirometer perhaps led to over reporting of symptoms, however FEV1 remained at baseline. 80% of individuals who lived within a 50km radius of the hospital received the influenza vaccine an increase of 20%. A home delivery service for medications proved to be successful with medication pickup(delivery) improving compliance. Finally, patient experience was positive. Patients welcomed the opportunity to continue to feel supported having their healthcare provided at home. They felt more comfortable asking questions in their own environment and that their healthcare was more tailored to need. They felt better informed/educated about their treatment regimes.100% of respondents hoped that the service would be able to continue into the future.

Conclusion: Outpatient review in patients' homes proved to be successful. This reimagined model of care reduced nonattendance rates at clinic, improved influenza vaccination compliance and increased baseline medication compliance. The use of home spirometry needs further validation –with access to consistent spirometry devices, reliable results and reporting of symptoms. These results in addition to positive patient experience has led our team to further investigate integrating this model of care in to our clinics.

A030

AUDIT OF AZITHROMYCIN USE IN A PAEDIATRIC CLINIC: INFORMING FUTURE GUIDELINE REVIEW

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Background and aim: The immunomodulatory and anti-inflammatory properties of azithromycin, a macrolide antibiotic, and its benefits in Cystic Fibrosis (CF) and other chronic airway diseases are well documented[(1,2)]. These include lung function improvement, decline in exacerbations and improvement in weight centiles. Including azithromycin as part of standard care has been strongly recommended by the US Cystic Fibrosis Foundation[(1)]. Research continues to help determine whether short courses or longer term azithromycin use provides more benefit in people with CF. The current CF guideline at the Children's Hospital, Westmead (CHW) recommends patient selection based on lung function and bacterial colonisation with 3 monthly monitoring for side effects and clinical response. To help guide the planned 2021 review of the CHW CF guideline, we aimed to review current azithromycin use and practices (prescription and monitoring).

Method: Retrospective chart audit of azithromycin use in CHW CF patients aged 6-18yrs between 2017-2020. Prescription to suitable patients and monitoring for side effects and effectiveness based on the current clinic guideline document was reviewed.

Results: 39/149 (26%) of patients were identified and prescribed azithromycin during the audit period. Of the remaining 122 age appropriate patients; 20/122 (16%) were eligible to trial azithromycin based on lung function parameters and 82/122 (67%) based on bacterial colonisation. In regards to monitoring of side effects, benefits and adherence, some patients were partially screened and monitored but no patients were fully screened as per the CF CHW practice guideline.

Conclusion: Guidelines and patient monitoring documents exist for azithromycin use at CHW however these tools are poorly utilised. Better strategies for ensuring appropriate patient selection, review and monitoring must be developed between clinicians, patients and their families to ensure more appropriate use of this medication. This is an important role of the CF clinical nurse consultant and pharmacist in conjunction with respiratory consultants.

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A045

ONLINE TUNING INTO KIDS FOR PARENTS OF CHILDREN WITH CYSTIC FIBROSIS: AN ACCEPTABILITY AND FEASIBILITY STUDY

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Background: Parenting a child with a chronic illness, such as cystic fibrosis (CF), can be stressful and challenging[1,2]. One of these challenges is balancing a child's emotional and mental health with their medical needs and physical health. Tuning into Kids (TiK) is an evidence-based parenting program that teaches parents to become "emotion coaches" for their child. TiK is an 8-week face-to-face program that has not previously been available to families through the CF clinic. The online nature has not previously been reported or evaluated.

Aim: This pilot study aims to assess the acceptability and feasibility of delivering TiK via telehealth for CF families at the Children's Hospital Westmead (CHW).

Method(s): CF parents from CHW were recruited via self or team referral to participate in the study. Participation involved attending 8 weekly TiK sessions (2 hours per session) via telehealth. Acceptability and feasibility were assessed via a post participation surveys and qualitative interview.

Results: 6 parents were recruited to this study with preliminary survey results available for 4/6 (67%) of participants. All participants rated the quality of the program as excellent with most or all of their needs having been met. Similarly, all would recommend the program to a friend. 3/4 (75%) were very satisfied with the amount of help received and 1/4 (25%) mostly satisfied. Participants found telehealth an acceptable platform for TiK with all agreeing/strongly agreeing that they liked TiK being online, could maintain attention online and would attend online programs again in the future. Qualitative interview results will be available for reporting shortly.

Conclusion: Preliminary acceptability and feasibility results are promising for the translation of TiK into an online program for CF parents. Results of this pilot study also suggest that other parenting and support programs that were previously unavailable to CF families in the face-to-face setting may be beneficial.

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A085

LIVING WITH CYSTIC FIBROSIS DURING THE COVID-19 PANDEMIC: A SOCIAL CONNECTEDNESS PERSPECTIVE

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Introduction/Background/Aim: Belongingness theory states that it is not just contact with others that satisfies our innate need for belonging, but the sense of quality and meaning we derive from this contact, which is referred to as social connectedness (Kohut, 1984; Lee & Robbins, 1995). Outside of a CF context, social connectedness is widely associated with enhanced mental and physical health (e.g. Deindl et al., 2016; Begun et al., 2018), but there is a lack of research within CF. This study explored social connectedness within the unique context of adults with CF, during the early onset of the COVID-19 pandemic.

Method: Seventeen adults with CF in Western Australia undertook semi-structured interviews via videoconferencing during May 2020. Interview transcripts were thematically analysed.

Results: In a general sense, participants placed significant value on social connectedness in supporting their mental and physical health. While CF creates social connectedness difficulties, participants described an ability to forge meaningful connections. Specific to the COVID-19 pandemic, participants adapted to the reduced in-person contact by increasing online communication. Social support received during the pandemic was both an outcome and enhancer of social connectedness. Overall, participants experienced a sense of being understood in relation to COVID-19, but where there was a lack of understanding it was damaging to a sense of togetherness.

Conclusion: Findings highlight the importance of education and awareness-raising around COVID-19 health implications and contact restrictions for those with CF. This study also offers valuable insights for health professionals in relation to how adults with CF experience social connectedness, both inside and outside of a COVID-19 context, and highlights the need for CF health professionals to offer social connectedness assessments and interventions to help optimise health outcomes.