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Conclusion: Our center was having difficulty obtaining home spirometry results in a timely fashion for TH visits. This project shows that a simple intervention (a pre-clinic phone call and subsequent call if needed) improved our results, with 76% of CF patients submitting results to the team before or on the day of the TH visit. Some of the reasons patients did not submit spirometry results included patients did not have their device readily available, the device had not been set up in time for the TH visit, and patients wanted to perform maneuvers with the therapist. Proactive communication with patients before clinic can help to overcome these barriers. The team will continue to explore strategies to communicate with families and address barriers to getting diagnostic data to the providers. Moving forward, we have set a new goal to receive spirometry results from at least 80% of patients before the TH visit.

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266

Physiotherapy during the COVID-19 pandemic: Support for the shielding patient by the shielding physiotherapist

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Background: The COVID-19 pandemic led to a rapid change in the delivery of care for patients with cystic fibrosis (CF). Clinic face-to-face appointments quickly changed to virtual reviews and assessments. The Royal Belfast Hospital for Sick Children is the regional pediatric CF unit for Northern Ireland, with 200 patients identified by screening (age 0–18). All patients were advised to shield from March 2020. Shielding was a term introduced by the Department of Health to protect clinically extremely vulnerable individuals who were identified as being at highest risk of severe illness from COVID-19. They were advised to protect themselves by no leaving their homes and minimizing all face-to-face contact. An advanced practitioner physiotherapist in CF (part time 14 hours/week) at our clinic was advised to shield due to an autoimmune condition. We present the support she was able to offer the pediatric service over 12 months (April 2020–March 2021).

Methods: The physiotherapist was provided with technology and access from home to be able to carry out virtual and telephone assessments with patients, liaise with CF multidisciplinary team members and physiotherapy colleagues, and complete the necessary patient records on digital systems. Results: Over the time period, the physiotherapist carried out 149 video consultations and 77 telephone assessments with 74 patients. Six patients with moderate to severe lung disease had more than 6 video consultations each over the period, with a maximum of 29 for an individual patient. The content of the virtual assessments included airway clearance assessment and treatment, postural assessment, Pilates sessions, and equipment troubleshooting. Annual assessments were also carried out virtually, with follow up attendance for exercise testing, and with the introduction of home spirometry, the physiotherapist was able to assist patients using this independently for the first time at home. Following the introduction of modulator therapy in Northern Ireland, she was able to review patients at home within the first few days of commencement. She attended 15 video meetings with the CF physiotherapy team, 3 video meetings with the physiotherapy manager, and 40 CF multidisciplinary team meetings held weekly. The physiotherapist was able to perform joint virtual sessions with junior staff and students, thereby contributing to their education and supervision (n = 6). In addition, the physiotherapist was able to maintain her mandatory training over the year by completing e-learning (n = 3) and attending video training (n = 4).

Conclusion: Virtual assessments offered by a physiotherapist who is shielding are a feasible and efficient means of providing ongoing support and advice to patients during the COVID-19 pandemic as well as sharing the challenges of shielding with them. This support augments the physiotherapy CF service and should be supported and encouraged by managers.

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Tissue oxygen utilization and ventilatory parameters during exercise in patients with cystic fibrosis: The role of HbA_{1c}

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Background: Minute ventilation/carbon dioxide (VE/VCO₂) slope during exercise is a measure of how efficiently the body can clear CO₂, with high values predicting mortality in various patient populations. Patients with cystic fibrosis (CF) exhibit both an impairment in oxygen uptake kinetics during exercise and an increase in VE/VCO₂ slope over time without a decline in exercise capacity. CF also affects the pancreas, and about 50% of adults with CF develop cystic fibrosis-related diabetes (CFRD). It is plausible that glycosylated hemoglobin, measured by HbA_{1c}, can interfere with the utilization of oxygen by the working tissue and the removal of CO₂ by the lungs during exercise in patients with CF. This study sought to test the hypothesis that higher HbA_{1c} negatively impacts oxygen utilization and removal of CO₂ during exercise in patients with CF.

Methods: This study included 17 patients (8 male, 9 female) with CF. Anthropometric and HbA_{1c} measurements were obtained. Pulmonary function testing was performed. A maximal exercise test on a cycle ergometer using the Godfrey protocol was conducted to obtain indices of exercise capacity (VO₂ max, VO₂ at ventilatory threshold (VT)) and ventilatory parameters (VE/VCO₂ slope and end-tidal CO₂ (petCO₂)). Near-infrared spectroscopy was placed on the vastus lateralis during exercise to measure oxygenated and deoxygenated hemoglobin.

Results: Overall, there is a negative relationship between HbA_{1c} and VO₂ max ($r^2 = -0.486$, P = 0.048), and patients with an HbA_{1c} less than 5.7% (n = 7) had greater VO₂ max (P = 0.015) than those with HbA_{1c} greater than 5.7% (n = 8) after controlling for lung function, sex, age, and BMI. There was also a negative relationship between HbA_{1c} and petCO₂ at maximal exercise ($r^2 = -0.598$; P = 0.01) and a positive relationship between HbA_{1c} less than 5.7% had lower VE/VCO₂ slope ($r^2 = 0.638$; P = 0.006). Patients with an HbA_{1c} less than 5.7% had lower VE/VCO₂ slope (P = 0.01) and higher petCO₂ (P = 0.001) than those with an HbA_{1c} greater than 5.7% after controlling for lung function, age, sex, and BMI. In a subset of patients (n = 13) with near-infrared spectroscopy data, HbA_{1c} was negatively correlated with muscle deoxygenated hemoglobin at VT ($r^2 = -0.664$, P = 0.04) after controlling for lung function. Patients with HbA_{1c} less than 5.7% (n = 8) had significantly less deoxygenated hemoglobin at the muscle than those with HbA_{1c} less than 5.7% (n = 5, P = 0.03).

Conclusion: For the first time, we have demonstrated that glycosylated hemoglobin can impair ventilation efficiency and CO_2 elimination during exercise in patients with CF. HbA_{1c} is also negatively associated with the amount of deoxygenated hemoglobin at the muscle during exercise. It is likely that glycosylation inhibits the ability of the hemoglobin molecule to release oxygen at the working tissue and CO_2 at the lungs. This leads to decreased use of oxygen and increased retention of CO_2 , both of which contribute to exercise intolerance in patients with CF.

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268

Unite to THRIVE: A virtual wellness intervention with PT and psychology

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Background: PT and psychology are integral parts of the Children's Hospital of Colorado CF multidisciplinary team. Both disciplines strive to provide patient education, home intervention, screening, and optimization of quality of life through participation in health and wellness strategies. The need for peer connection, education, stress reduction, and home management was identified by the CF PT and psychologist at the start of the pandemic [1]. Historically, individuals with CF have not been able to connect with peers secondary to infection control [2], which has limited group therapy options. The pandemic accelerated the use and implementation of telemedicine to provide care to patients with CF throughout the