Managing patients in their own home: experiences from an adult CF service

(Amy Smith and Charlie Hardaker, Bristol Adult CF Centre)

A new strain of Coronavirus, COVID-19, was first identified in December 2019 and has since resulted in a worldwide pandemic. The most common symptoms affect the respiratory system causing high temperatures and a persistent cough. In a small proportion of the population with COVID-19, treatment in a hospital setting for oxygen therapy, CPAP or admission to intensive care and mechanical ventilation is required. Although we still do not know the extent of how COVID-19 affects persons with Cystic Fibrosis (PwCF), they have been identified as an extremely high risk group of individuals and have been advised by the UK Government to self-isolate at home for 12 weeks from the 22nd of March 2020.

PwCF experience numerous symptoms affecting multiple organs but predominantly the respiratory system. Due to CFTR gene defect, there is an imbalance of absorption and secretion of sodium and chloride ions across epithelial cell channels which results in a built up of thick, sticky secretions in the airways. In order to maintain and slow down the deterioration in lung function, PwCF are encouraged by Physiotherapists to maintain a daily airway clearance regime alongside inhaled medications and exercise.

At the Bristol Adult Cystic Fibrosis Centre (BACFC), one of the airway clearance techniques used is intermittent positive pressure breathing (IPPB) or mechanical insufflation – exsufflation (MI-E) using the NIPPY Clearway device which has been found to aid airway clearance and lower work of breathing. IPPB mode aids clearance by increasing tidal volumes and ventilating peripherally behind secretions with altered flow rates to allow for movement of secretions to proximal airways. In MI-E mode, as well as large inspiratory breaths, this allows the therapist to set an expiratory breath while the patient is coughing or performing a forced expiratory technique to generate an expiratory bias leading to the expulsion of secretions and reduce effort for the patient. Due to the limited number of NIPPY Clearway devices, PwCF mostly have access to this method of airway clearance when they come into hospital for an admission.

A single case study report (Forster et al. 2014) compared airway clearance techniques including non-invasive ventilation device (NIV), IPPB using a Bird Mark 7 ventilator and MI-E using a NIPPY Clearway device. These clearance techniques were compared using flow and pressure breath profiles, regional lung ventilation via Electrical Tomography and ease of sputum clearance using a 10-point visual analogue scale (VAS) pre and post treatment. Results demonstrated that lung recruitment occurred with all techniques however suggested IPPB and MI-E have preferred expiratory flow bias and therefore further facilitate airway clearance. The patient-reported VAS also favoured using the NIPPY Clearway device for ease of clearance. Due to PwCF currently self-isolating at home and only attending hospital for essential appointments or urgent admissions, this limits the amount of equipment they have access to at home. As a result the multi-disciplinary team are transferring care into the community where possible to be able to maintain social-shielding measures and safety of PwCF. The Physiotherapy team at BACFC have purchased additional NIPPY Clearways to loan to patients in order to avoid admission and allow optimisation of airway clearance during the COVID-19 pandemic. In order to distribute the devices, patients with a clinical need to use IPPB or MI-E were highlighted. This included patients with either a forced expiratory volume in one second (FEV₁) of \leq 40% or have 4 or more admissions annually and use either IPPB or MI-E during an exacerbation of their CF where their home clearance routine is no longer effective. We aim to distribute and set up the devices in the patient's home with additional follow up appointments virtually via phone or video calls.