The use of NIV within a regional adult Cystic Fibrosis Centre


Introduction: Studies show Non-Invasive Ventilation (NIV) in Cystic Fibrosis (CF) has reduced respiratory failure and aided secretion clearance. Our aim was to examine the use of NIV in CF patients over 12 months (Jan–Dec 2008).

Method: A retrospective observational study of adult CF inpatients treated with NIV was performed. Indication for NIV treatment, tolerance, complications and perceived benefit were recorded.

Results: 20 patients were treated with NIV over 35 episodes, 9 male with a mean age of 31 years (SD 9). Median predicted lung function on admission was 23% (10–53%). 13 patients had 1 NIV episode; 7 patients had 2–6 episodes. NIV was used to aid sputum clearance in 5 patients (12 episodes) (34%), control hypercapnic symptoms in compensated type II respiratory failure in 18 patients (17 episodes) (49%) and to reverse acidosis in acute type II respiratory failure in 5 patients (6 episodes) (17%).

NIV was tolerated well in 18 patients (33 episodes) (94%), with subjective benefit in all episodes. In patients with raised PaCO2, reduction of headache was reported (49%) and to reverse acidosis in acute type II respiratory failure in 5 patients (6 episodes) (17%).

NIV was used successfully to control symptoms of hypercapnic respiratory failure and sputum retention and was generally well tolerated.

Discussion: This 4 month randomised cross-over trial compared taking Pulmozyme at 4pm, with taking it immediately following the evening session of chest physiotherapy. The primary outcome was FEF25 with secondary outcomes being FEV1 and subjective feedback to assess whether there were any side effects from taking Pulmozyme late at night.

A large number of subjects could not be recruited or had to be withdrawn from the study due to increased respiratory symptoms. The total number of patients who completed the study was 5, with a mean age of 11 years old. The results of this study did not show any significant differences between the two timing of Pulmozyme with p=0.557 for FEF25 and p=0.868 for FEV1. Therefore no conclusions as to which of the two different timings of Pulmozyme in the most effective at reducing airflow obstruction can be drawn from this study.

There were no reported increase in overnight symptoms from any subjects from taking Pulmozyme in the evening, so it could be concluded that Pulmozyme is safe to administer at that time.

There have been several short term studies investigating this research question. This study attempted to carry out a longer term study with a similar study design as previous shorter term research. However the difficulty in subject recruitment and the high drop out rates reported here have previously also been reported by investigators attempting a longer term study with this design. The authors conclude that this type of study design is not appropriate for a longer term investigation to answer this research question.

A randomised, cross-over trial of upright sitting versus alternate side lying during nebulised delivery of medication in cystic fibrosis

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The pattern of deposition of inhaled medications is non-uniform in people with CF. Deposition follows tidal ventilation, so inhalation in sitting tends to under-dose the apices. One theory is that inhalation while in alternate side lying (ASL) would improve homogeneity of deposition by preferentially dosing each lung (including the apex) when it is dependent. Before testing this theory, we sought to determine whether there any slowing of delivery time with the ASL strategy.

Method: 24 adults (mean±SD age 30±9y, 13F) with stable CF lung disease (FEV1 ≤ 55% in 3 episodes) (13%). All 5 patients (12 episodes) started on NIV to aid sputum clearance and 1 patient with acute respiratory failure reported increased sputum clearance with NIV. 2 patients were unable to tolerate the mask and felt no subjective benefit. The most common complications reported by those patients who tolerated NIV were: a) hypoxia (422%), b) pressure sores 3 (17%), c) difficulty sleeping 2 (11%) and d) dehydratation 1 (1%).

Conclusion: NIV was used successfully to control symptoms of hypercapnic respiratory failure and sputum retention and was generally well tolerated.

The delivery time did not significantly differ between ASL (18.54±3.80min) and ASL (17.96±3.53min), a difference of 0.58 min (95%CI 1.40 to –0.24). There was no significant correlation between delivery time and FEV1, FVC or height. In ASL, increasing the time between turns to 2min did not significantly affect the dose delivered on each side, however turning each 3min resulted in less uniformity (mean ratio West Midlands 1.24 to 1.40).

Discussion: ASL during inhalation therapy does not prolong delivery time. We are now investigating the effect of ASL on uniformity of deposition with radioaerosol scans.

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A rheological investigation of cystic fibrosis sputum

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All sputum samples were collected from patients and rheological measurements undertaken within 24 hours. All measurements were taken at a temperature of 25°C, using an Anton Paar Physica MCR 501 rheometer. All samples were equilibrated in the rheometer for a period of 1 hour before measurements were taken.

The intention was to freeze samples when supplied, but data illustrating the complex viscosity of three examples of the mucus, on which measurements were taken immediately, followed by freezing for a period of 4 days, showed that freezing of the mucus can result in significant changes of the gel structure. One example, ETO801763 showed little change in the values of the complex viscosity but another example, ETO801764, showed a considerable decrease, whilst a third example, ETO801765, gave increased values.

Data illustrating the complex viscosity of a large number of samples will be presented. Large variations in the values of the complex viscosity were observed, ranging from almost 10^13 mPas to 3000 mPas. Although the overall general pattern of behaviour of the samples is a reduction in the value with increasing frequency of oscillation, there were exceptions to this pattern, where an increase is initially seen in some cases at low levels of oscillation but, in others, a sharp increase is observed at very high levels.

In conclusion, consideration should be paid to the danger of complicating effects of cryo-preservation prior to physical analysis of cystic fibrosis sputum. This data also suggests that components within the sputum generate unpredictable and highly variable rheological data in different patients. Unravelling the biochemical basis of this variability may assist the development of improved therapeutic approaches to the treatment of cystic fibrosis.