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Title: Metabolic and respiratory functioning of patients with cystic fibrosis during the years preceding death

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Body: In patients with cystic fibrosis (CF) FEV_1 is used as a predictor of survival. The median mortality within two years is greater than 50% if FEV_1 is 30% of predicted. In comparison to other diseases, such as COPD, the association between hypercapnia and mortality in CF is not fully understood. Patients with CF also develop metabolic alkalosis but data is limited with regards the metabolic state within the last years of life. Aims: To determine whether there is a relationship between hypercapnia, reduced lung function and mortality in patients with CF during the years preceding death. Methods: All adult patients with CF who died of respiratory failure between 2007 and 2012 were studied. Capillary blood samples, lung function, and date of NIV or oxygen commencement were collated from records. The best FEV_1 percentage predicted and FVC percentage predicted value were recorded at six monthly intervals from the date of death. Results: 15 patients with CF died from respiratory failure during this period, 10 female and 5 male, with a median age of 30 years old (range 17-46 years). The average time from the FEV_1 falling below 30% and the date of death was 2.5 years (range 0-7.5 years). Only 77% patients developed hypercapnia prior to death. In 7 of these patients significant metabolic alkalosis contributed to the hypercapnia. Hypercapnia developed on average 14 months before death (range 0-3.5 years). Home NIV was used in 8 patients; 5 of these required intermittent acetazolamide treatment for symptomatic hypercapnia. Conclusion: Once hypercapnia has developed the outcomes can be poor. As previously reported metabolic alkalosis is a frequent finding in this cohort.