A COMPARISON BETWEEN THE CLINICAL FEATURES OF ALPHA-1 ANTI-TRYPSIN DEFICIENCY (A1ATD) PATIENTS

Introduction

Alpha-1 antitrypsin deficiency (A1ATD) is a hereditary disorder affecting about 1 in 3000 people in the UK commonly associated with early-onset emphysema. There are two common deficiency alleles - PiS and PiZ. PiZZ patients have severe AATD, with levels of 10–15% normal. PiSZ patients have less severe deficiency (≈ 40% normal) and are generally thought to have a minimal risk. We hypothesised that if PiSZ patients were at lower risk of COPD than PiZZ, and their lung disease would be more characteristic of usual COPD than that of PiZZ patients.

Method

104 PiSZ patients and 638 PiZZ patients from the UK AATD registry (ADAPT) were compared for their demographics, lung function, risk factors for COPD (e.g. smoking, occupation), co-morbidities associated with COPD, index status (i.e. if diagnosed due to lung disease or family screening) and CT densitometry (where available). Outcome in terms of lung function decline and mortality was also assessed. Univariate statistics were used to guide subsequent regression analyses.

Results

Emphysema was more likely in PiZZ than PiSZ patients (OR 11.0 (5.7–21.3); p < 0.001) in the regression analysis after accounting for age, pack years and lung index status. PiZZ patients also had significantly worse FEV1 and DLCO than PiSZ patients in similar regression models (both p < 0.01). Emphysema was more severe in both upper and lower zone (both p < 0.01), and proportionately greater in the lower zone (UZ/LZ VI = 1.5 v 1.2) in PiZZ patients. Mortality and DLCO decline were also greater in PiZZ patients.

Conclusion

PiSZ patients have a milder form of AATD associated with better lung function. The data suggests the pattern of emphysema is closer to usual COPD than classical AATD. Further analyses comparing PiSZ to PiMM are now ongoing.
CANNABIS LUNG CAUSING DEBILITATING EMPHYSEMA: ARE WE ON THE VERGE OF AN EPIDEMIC?

Narendra Babu Chinnappa, Kasia Zalewska, Damian Mckeon. 

Introduction and objectives Cannabis (or marijuana) is the world’s most widely-used illicit drug, according to UN drug report 2012 prevalence of cannabis use between 15–64 years of age is around 1.7% in Europe and 2.6% in USA. 1 It is particularly prevalent amongst adolescents and young adults. As societies reconsider the legal status of cannabis, policy makers and clinicians require sound knowledge of the acute and chronic effects of cannabis. There has been surprisingly little research into its effects on respiratory health. In a rural region of North Wales we have noticed an increasing number of young patients presenting with precocious bullous emphysema associated with very high tobacco and cannabis usage.

Methods A series of 8 patients presenting through the Emergency Department with an exacerbation of COPD were noted to have precocious COPD associated with high cannabis use. The age was between 35–48, all had both physiological and radiological signs of advanced emphysema. All had at least 10–20 years of cannabis usage smoking more than 5 ‘joints’ per day. Of these, 4 patients were significantly impaired to require long term oxygen therapy, and one is actively listed for a single lung transplant. All had normal levels of alpha 1 antitrypsin and chymotrypsin.

Results We found young patients with debilitating COPD secondary to cannabis use i.e. as less as 10 years of use. 2 We postulate that cannabis smoking leads to severe COPD in young patients independent of genetic susceptibility, which is on the verge of increase.

Conclusions The addition of cannabis to the tobacco, and high usage at a young age is leading to increase in the incidence of COPD in general and bullous emphysema as a phenotype in particular. We are concerned that the dangers of cannabis inhalation and these risks are not being appreciated by the wider health community. More research is needed to know the mechanisms of the inflammatory response secondary to cannabis smoking.

REFERENCES
1. UN drug report 2012, United Nations Office on Drugs and Crime