

Breathe deep: Which patients could benefit from inhaled steroids in cystic fibrosis?

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A specific variation in the glucocorticoid receptor gene is associated with lung disease progression in cystic fibrosis, research published this week in the online open access journal Respiratory Research reveals. This finding adds weight to previous research suggesting that specific subgroups of patients with cystic fibrosis may benefit from glucocorticoid treatment.

Patients with cystic fibrosis show wide variability both in terms of the inflammatory burden of the lung and in their response to inhaled glucorticoids. As such, the effectiveness of this therapy in patients with cystic fibrosis remains uncertain. However, previous research has suggested that specific subgroups of patients may benefit from treatment with inhaled glucocorticoids.

In several inflammatory diseases, variations in sensitivity to glucocorticoids have been found to be associated with single nucleotide polymorphisms in the glucocorticoid receptor gene. So, a team from Hôpital Trousseau, Assistance Publique Hôpitaux de Paris, Inserm and Université Pierre et Marie Curie (all based in Paris, France) set out to investigate the effect of four polymorphisms (TthIII, ER22/23EK, N363S and BclI) in the glucocorticoid receptor gene on disease progression in 255 young people with cystic fibrosis.

The BclI glucocorticoid receptor gene polymorphism was found to be significantly associated with a decline in lung function, as measured by the forced expiratory volume in 1 second and the forced vital capacity.



The deterioration in lung function was more pronounced in patients with the BclI GG genotype than in those with the CG and CC genotypes.

The authors write: "The association of BclI polymorphism and lung disease progression in cystic fibrosis gives support to the concept that specific subgroups of patients with cystic fibrosis may benefit from the use of glucocorticoids preferably by the inhaled route. If true, this should allow discriminatory prescribing which is of tremendous importance."

Source: BioMed Central

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