INVESTIGATING THE COMPLEXITY OF THE RELATIONSHIP BETWEEN GASTRO-OESOPHAGEAL REFUX AND CF LUNG DISEASE

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Objective To evaluate ultrashort echo time (UTE) magnetic resonance imaging (MRI) as a feasible endpoint in a multicenter trial using a subset of patients aged 6 to 11 years with cystic fibrosis (CF) homozygous for the F508del mutation treated with lumacaftor/ivacaftor (LUM/IVA) combination therapy (NCT02514473).

Methods UTE MRI scans were obtained at study baseline (n=10); a second scan was completed at week 24 in 9 subjects (4 active treatment, 5 placebo) at 3 institutions using MRI hardware from 2 vendors. One of the platforms was 3D radial UTE (Johnson et al. Magn Reson Med. 2013;70:1241–1250); the other was 3D “stack of stars” UTE (Lederlin et al. J Magn Reson Imaging. 2014;40:839–847). Scans were obtained during 5 min of tidal breathing without the use of intravenous contrast. MRI scans were scored by 2 independent readers using the Brody score after supervised training on UTE MRI scans. Readers were blinded to time point and treatment group. Scores are presented as mean (SD); no statistical testing was performed.

Results Despite the lower image quality of MRI compared with computed tomography, potential treatment effects were seen on the UTE MRI images (figure 1A). Mean total Brody score decreased from 41.1 to 32.5 with treatment, a change from baseline (SD) of 8.4 (11.2) with active treatment, but increased from 31.3 to 34.6, a change from baseline of 3.3 (8.2) with placebo (figure 1B). Mucus plugging subscore decreased by 5.0 (5.1) from 8.5 to 3.5 with active treatment but increased by 1.4 (4.4) from 4.2 to 5.6 with placebo. There were no noticeable changes in other subscores (bronchiectasis, peripheral thickening, parenchyma, or hyperinflation).

Conclusion In this analysis from an exploratory substudy in patients with CF aged 6 to 11 years homozygous for F508del, UTE MRI was a feasible approach for detecting the effect of LUM/IVA, despite the small sample size, short duration of treatment, and limitations in image quality. As optimisation in UTE MRI technology improves image quality, monitoring disease course in patients with CF may improve.

Please refer to page A259 for declarations of interest in relation to abstract P254.