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Title: Ventilation inhomogeneity in children with cystic fibrosis and primary ciliary dyskinesia

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Body: Rationale: Ventilation inhomogeneity (VI) measured by multiple breath inert gas washout (MBW) are common findings in cystic fibrosis (CF) children with normal spirometry (1). Recently this was shown also in primary ciliary dyskinesia (PCD) (2). The lower morbidity in PCD vs. CF would suggest that small airway involvement is less severe in PCD. We therefore compared MBW indices reflecting peripheral airway involvement in children with CF and PCD. Methods: A cross sectional study was performed in 24 children with PCD and 25 with CF, matched by age and FEV1. N2 MBW in triplets (Exhalyzer D, EcoMedics AG) and spirometry were performed within one occasion in clinically stable patients. Lung Clearance Index (LCI), an index of global VI, and specific indices of VI arising in the conductive (Scond) and the acinar (Sacin) airway zones (2) were calculated and presented as z-scores based on new normative data (Houltz B et al, ERS abstract 2012). They were compared between patient groups using Mann-Whitney Test. Results: There was no significant difference in LCI, Scond and Sacin (z-scores) between the two patient groups, although LCI, z-scores tended to be higher in PCD (Table 1).

Table 1. Demography and Results.

	CF	PCD	P value
N (males)	25 (12)	24 (7)	-
Age, yrs	12.3 (2.1)	13.0 (3.1)	0.48
BMI	17.1 (2.0)	18.1 (2.8)	0.25
FEV1, % pred.	99.3 (10.5)	95.4 (12.5)	0.57
LCI, z-scores	8.1 (6.5)	11.6 (6.6)	0.07
Scond*VT, z-scores	10.6 (6.1)	11.0 (6.0)	0.83
Sacin*VT, z-scores	6.0 (5.6)	8.5 (6.4)	0.18

Results expressed as mean (SD), unless otherwise stated.

Conclusion: In contrast to our expectations severity of peripheral airway involvement is similar in children with CF and PCD. (1) Respiration 2009;78:339; (2) Thorax 2012;67:49.