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Title: Air liquid interface culture can alter ciliary beat pattern in epithelium from primary ciliary dyskinesia patients

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Body: Introduction: Primary Ciliary Dyskinesia (PCD) is a rare inherited heterogeneous disorder of cilia, impairing mucociliary clearance. PCD is diagnosed by low nasal nitric oxide concentration, abnormal airway ciliary function with corresponding ultrastructural defects, excepting atypical cases. PCD may be differentiated from secondary dyskinesia by assessing differentiated ciliated epithelium at air liquid interface (ALI). Aim: To evaluate ciliary beat pattern (CBP) on PCD airway epithelium before and after ALI culture. Method: Ciliary function of nasal brushing epithelium from 9 PCD patients with ultrastructure defects confirmed by TEM, was analysed (n=10 cell clusters) at 37ºC by high-speed video microscopy (100x oil objective). Confluent basal epithelial cells on 0.3 mg/ml Purecol (Nutacon) coated 12 mm transwell inserts were differentiated over 21 days in 1:1 BEGM:DMEM with SingleQuots (Clonetics, Lonza) and 100 nM retinoic acid. Ciliated ALI membranes were excised and cilia and ciliary function re-examined. Results: Three ALI cultures (33.3%) retained their CBP compared to the original epithelium: 2/9 completely static cilia; 1/9 static with twitching cilia. Six ALI cultures (66.7%) continued to exhibit abnormal CBP but the phenotype changed following ALI culture: 5/9 static with twitching cilia became completely static on ALI; 1/9 dyskinetic with static cilia became completely static on ALI and additionally dynein arm defects altered from 60 to 98% after culture. Conclusions: The diagnosis of PCD remained clear before and after ALI culture, however we observe that ciliary function and percentage defect rate may alter in epithelial cell culture at ALI.