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Ciliary dyskinesia might be secondary to chronic inflammation in cystic fibrosis

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Background Mucociliary clearance (MCC) results from an effective interaction between the mucus layer and the normal coordinated ciliary beating. Ciliary dyskinesia, or an abnormal ciliary beating, is defined as an abnormal ciliary beat frequency (CBF) and/or ciliary beat pattern (CBP), and can be primary, or secondary to chronic infection or inflammation. In cystic fibrosis (CF), MCC is impaired due to dehydrated mucus, but recent data suggested a ciliary dyskinesia in CF patients, but it is unknown if this is primary, or secondary to chronic inflammation.

The aim of this study is to evaluate if ciliary dyskinesia is primary or secondary in CF patients.

Methods Ciliated epithelial samples were obtained by nasal brushing from 10 CF patients and 5 healthy volunteers. Beating cilia were recorded using digital high-speed videomicroscopy at 37 °C. Ciliary functional analysis (CFA) was assessed by CBF and by the percentage of dyskinetic CBP (%DK). CFA was repeated after Air-Liquid Interface (ALI) cell culture.

Results Our results confirm that ciliary dyskinesia is increased in CF patients compared with healthy volunteers, with no significant difference in CBF (13.3 ± 2.9 Hz vs. 12.5 ± 1.1 Hz, $P = 0.542$), but a higher %DK (31.4 ± 8.2% vs. 17.4 ± 7.1%, $P = 0.007$). However, after ALI cell culture, there is no difference between CF patients and healthy volunteers in CBF (15.6 ± 2.1 Hz vs. 15.0 ± 2.3 Hz, $P = 0.860$), or in %DK (11.7 ± 2.3% vs. 16.2 ± 7.5%, $P = 0.371$).

Conclusions Our pilot study confirms that ciliary dyskinesia is present in CF patients, and may contribute to impaired MCC. However, after ALI cell culture, CFA is similar between CF patients and healthy volunteers, suggesting that ciliary dyskinesia may be secondary to chronic respiratory inflammation in CF patients.

Disclosure of interest The authors declare that they have no competing interest.

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Lack of sensitivity and specificity for PCD diagnosis when ciliary videomicroscopy is performed at room temperature

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Introduction Primary ciliary dyskinesia (PCD) is a heterogenic inherited ciliopathy in which respiratory cilia are stationary or dyskinetic. Digital high-speed videomicroscopy (DHSV) is highly sensitive and specific for PCD diagnosis, but lacks standardization. Various laboratories perform DHSV using different temperature,which may influence ciliary functional analysis (CFA), including ciliary beat frequency (CBF) and beat pattern (CBP). Recent data suggest that CBF increases with temperature, but the relationship between CBP and temperature has not been extensively studied. However, CBF should not be used without CBP assessment in diagnosing PCD, given its lack of sensitivity and specificity. The aim of this study is to evaluate the effect of temperature during DHSV on CFA.

Methods Ciliated epithelial samples were obtained by nasal brushing from 14 patients referred to our PCD diagnosis center (7 with a confirmed PCD diagnosis (PCD), and 7 with an excluded PCD diagnosis (non-PCD) and from 5 healthy volunteers, to establish our laboratory normal values. Beating cilia were recorded using DHSV at 37 °C and at room temperature(25 °C) within 9 hours after sampling. CFA was assessed by CBF and the percentage of abnormal CBP.

Results When DHSV is performed at 37 °C, our result show a 100% sensitivity and specificity of the percentage of abnormal CBP to diagnose PCD, with a result above normal values for all PCD patients, and within or under normal values for all non-PCD patients.

However, when the percentage of abnormal CBP is measured at 25 °C, a PCD diagnosis might be missed in 1 PCD patient with a borderline value and might lead to a wrong PCD diagnosis in 3 non-PCD patients.

Table 1

	CBF (Hz)		Abnormal CBP (%)	
	37°C	25°C	37°C	25°C
Normal values (±SD)	12.8 ± 1.7	7.1 ± 1.9	19.3 ± 6.2	8.6 ± 7.1
Patients				
PCD 1	0	0	100	100
PCD 2	5.8	3.8	76.5	73.9
PCD 3	4.0	3.2	82.5	84.6
PCD 4	10.3	5.1	69.4	77.8
PCD 5	9.4	17.4	92.5	20
PCD 6	1.5	6.9	93.1	65
PCD 7	7.7	11.7	96	60
Non PCD 1	17.6	10.6	22.2	8
Non PCD 2	14.9	10.1	17.1	26.1
Non PCD 3	10.9	5.8	24.6	22.9
Non PCD 4	17.4	11.6	8.3	14.3
Non PCD 5	13.7	5.3	16	43.5
Non PCD 6	14.7	7.1	6.7	9.1
Non PCD 7	16.8	10.3	11.4	8.7

Table 1: Results of CBF and the percentage of abnormal CBP in patients with a confirmed PCD diagnosis (PCD) and with an excluded PCD diagnosis (non-PCD). Green: Values within our normal range for temperature; Red: values outside our normal range for temperature; Orange: borderline values (outside but near our normal range for temperature).