

On line Table 1: Lobar Distribution of HRCT Scan Abnormalities in the PCD Study Population (n = 64).

Variables	Right superior lobe	Middle lobe	Right lower lobe	Culmen	Lingula	Left lower lobe
Extent of bronchiectasis	71,9 §	78,1	89,1	65,6 *,§	65,6 *,§	93,8
Severity of bronchiectasis	73,4	79,7	90,6	65,6 *,§	67,2 *,§	93,8
Peribronchial thickening	50 *,§	67,2	79,7	48,4 *,§	48,4 *,§	78,1

Data are presented in % patients. * : p<0.0001 vs. right lower lobe. §: p<0.0001 vs. left lower lobe.

On line Table 2: Main phenotypes and genotypes characteristics of PCD patients.

Patient	Sex	Situs inversus	Ethnicity	Consanguinity	Familial history of PCD	Nasal NO (nL/min)	Ultramicroscopic ciliary defect	Genetic anomaly	Extrapulmonary Impairment	Sterility/hypofertility
1	M	Yes	M/E	No	Yes	ND	ODA	DNAI1		U
2	M	No	Eu	No	No	0	ODA + IDA	None		No
3	F	No	WI/SSA	Yes	No	6	ODA	DNAH5		U
4	M	Yes	WI/SSA	No	Yes	17	ODA + IDA	LRRC50=DNAAF1		Yes
5	M	Yes	M/E	Yes	Yes	15*	ODA	None		No
6	M	No	M/E	No	No	38	IDA	CCDC39		Yes
7	M	Yes	Eu	No	No	0	Normal	DNAH11		U
8	F	No	Eu	No	Yes	103	ODA	ND		U
9	M	Yes	M/E	Yes	No	ND	ODA + IDA	DYX1C1		U
10	F	Yes	M/E	No	No	ND	ODA + IDA	LRRC50=DNAAF1		Yes
11	F	Yes	M/E	No	No	ND	ODA + IDA	LRRC50=DNAAF1	Psychomotor impairment	U
12	M	No	M/E	No	Yes	ND	CC	RSPH4A		No
13	M	No	M/E	Yes	Yes	ND	CC	RSPH9		U
14	F	Yes	M/E	Yes	Yes	19	ODA	DNAI1		U
15	M	Yes	M/E	No	Yes	11	ODA	DNAI1		U
16	F	Yes	Eu	No	No	ND	Normal	DNAH11		No
17	F	No	Eu	No	No	105	CC	Hydin		U
18	F	Yes	Eu	No	No	ND	ND	ND		U
19	F	Yes	Eu	No	No	0	ODA	ND	Polycystic kidney disease	U
20	F	No	SA	No	No	17*	Normal	Hydin		U
21	M	Yes	Eu	Yes	Yes	ND	ODA + IDA	None	Polycystic kidney disease	Yes
22	M	Yes	M/E	Yes	No	75	ODA	DNAI1		U
23	F	No	Eu	Yes	No	ND	ODA	DNAI2		U
24	M	No	Eu	No	No	ND	CC	None		U
25	M	No	Eu	No	No	119	CC	None		U
26	M	No	Eu	No	Yes	98	ODA + IDA	RPGR	Psychomotor impairment, Retinis pigmentosa	U

27	M	No	Eu	No	Yes	ND	ODA + IDA	RPGR	Retinis pigmentosa	No
28	F	No	Eu	Yes	No	ND	IDA	None		Yes
29	F	Yes	Eu	No	No	ND	ND	ND		No
30	M	Yes	Eu	No	No	11	ODA + IDA	None		No
31	F	Yes	Eu	No	No	ND	ODA + IDA	None		No
32	F	No	Tu	Yes	No	14	CC	None		U
33	F	Yes	M/E	Yes	No	ND	IDA	CCDC39		U
34	M	No	Eu	No	No	93	CC	ND		U
35	M	No	Eu	Yes	Yes	86	ODA + IDA	RPGR	Retinis pigmentosa	U
36	M	No	Eu	No	Yes	25	ODA + IDA	RPGR		U
37	M	No	Eu	No	No	211	ODA + IDA	None	Psychomotor impairment, polydactylia	U
38	F	No	Eu	No	Yes	ND	ND	ND		No
39	M	Yes	Eu	No	No	ND	ND	ND		U
40	F	Yes	Eu	No	No	402*	ND	ND		No
41	M	No	Eu	Yes	No	150	Normal	ND		U
42	F	No	Eu	Yes	No	11	ODA	DNAI1		No
43	M	No	M/E	No	No	54	CC	None		Yes
44	M	Yes	M/E	Yes	Yes	ND	ODA	DNAH5		No
45	M	Yes	Asia	No	No	ND	ND	ND		U
46	M	No	Eu	No	No	2	ODA + IDA	None		U
47	M	No	M/E	No	Yes	6	CC	RSPH9		U
48	F	No	Eu	No	No	19*	ODA	DNAI1		U
49	F	No	Europe	No	No	14*	ODA + IDA	None		No
50	M	Yes	M/E	No	No	ND	ODA + IDA	ND		Yes
51	F	No	Eu	No	No	ND	CC	None		U
52	F	Yes	Eu	No	No	ND	ODA	DNAH5		Yes
53	F	Yes	Eu	No	No	26	ODA	ND		Yes
54	M	Yes	Eu	No	No	ND	ND	ND		Yes
55	M	Yes	Eu	No	Yes	ND	ODA	DNAH5		Yes
56	M	No	Eu	No	No	ND	CC	ND		Yes

57	F	No	Eu	No	No	190*	CC	RSPH1		U
58	M	Yes	Eu	No	No	ND	ND	ND		No
59	F	No	M/E	No	No	ND	ND	ND		Yes
60	M	Yes	Eu	No	Yes	22	IDA	CCDC39		Yes
61	F	No	Eu	Yes	No	99*	ODA + IDA	None		No
62	F	Yes	Asia	No	No	5	ODA	ND		No
63	F	Yes	Eu	No	No	ND	ND	ND		No
64	M	Yes	WI/SSA	No	No	ND	IDA	CCDC39		Yes
65	M	No	Eu	No	No	4	CC	RSPH4A		No
66	M	No	Eu	No	No	9	CC	RSPH4A		No
67	M	Yes	Eu	No	No	ND	ODA + IDA	ND		Yes
68	M	No	Eu	No	No	11	IDA	ND		Yes
69	M	No	Eu	No	No	55	CC	RSPH1		Yes
70	F	Yes	M/E	No	No	74	ND	ND		U
71	M	No	M/E	No	No	13*	IDA	CCDC40		U
72	F	Yes	Asia	Yes	No	ND	ODA + IDA	None		U
73	M	Yes	M/E	No	No	2	IDA	CCDC39		Yes
74	M	No	Eu	No	No	85	ODA + IDA	None		U
75	M	No	Tu	No	No	210*	ODA	DNAH5		No
76	M	Yes	Eu	No	No	ND	Normal	DNAH11		U
77	M	No	Eu	No	No	6	IDA	CCDC39		U
78	F	No	S/A	No	No	18	ODA	DNAAF2		U

Abbreviations: M/E= Maghreb/Egypt, WI/SSA = West Indies/Sub-Saharan Africa, Eu = Europe, SA = South America, T = Turkey

ODA = ODA defect, IDA = absence of IDA with axonemal disorganization, ODA + IDA = absence of both dynein arms, CC = abnormal central complex

PI : Psychomotor impairment, RP : Retinis pigmentosa, PKD : Polycystic kidney disease

ND = no data, U = unknown, * : several measurements available

On line figureS1 : Respiratory function of PCD patients.

Figure s1A shows the linear regression on courses of FEV₁ (mL) from their first lung function measurement in adulthood (≥ 18 years). Each regression line represents one patient.

Figure s1B shows the relationship between last FEV₁ available (% predicted) and age (years); Spearman rank correlation coefficient $\rho = -0.246$ and $p = 0.029$.

Figure s1C shows the relationship between last FEV₁ available (% predicted) and BMI (kg/m^2); Spearman rank correlation coefficient $\rho = -0.283$ and $p = 0.012$

Figure s1A

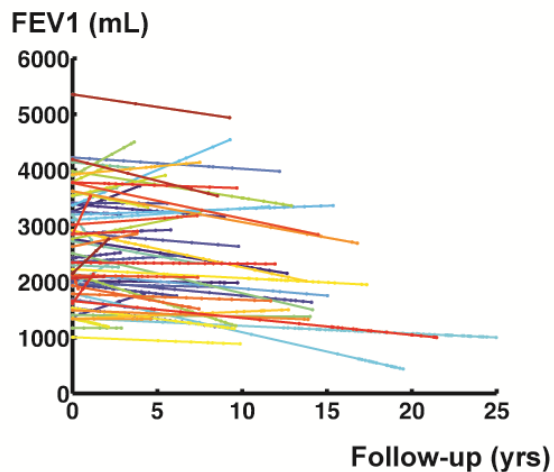


Figure s1B

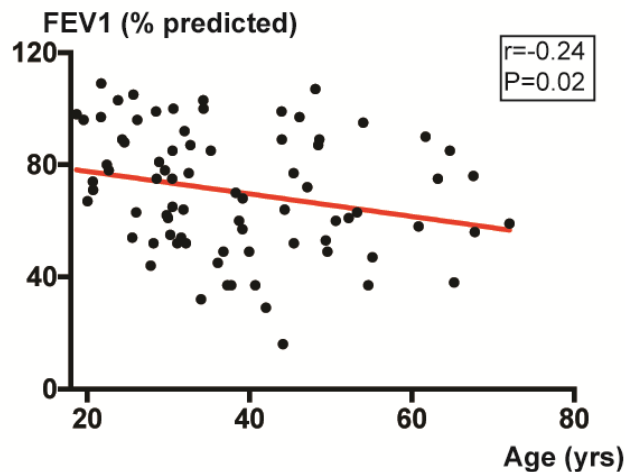


Figure s1C

