

TABLE S1: Characteristics of included studies describing spirometric indices of PCD patients by country, including studies with overlapping population

Study	Publication Year	Participants n	Age	Type of study	Study design	Diagnostics	Symptoms stratified by age	Longitudinal data	FEV1	FVC
Single-centre/regional studies										
Belgium										
BOON et al.	2015	39	16.1 (11.1-19.6) [¶]	P	Case-control	EM, HFVM	-	-	+	-
BOON et al.	2014	122	17.7 (9.5-28.1) [¶]	R	Case series	EM, HFVM, gen	-	-	+	+
BOON et al.	2014	11	16 (7-62) ⁺	R	Case series	EM, HFVM, gen	+	-	+	-
HELLINCKX et al.	1998	11	15.2±7 [#]	P	Case series	EM, HFVM	+	+	+	-
Brazil										
OLM et al.	2011	10	13.4±4.7 [#]	P	Case-control	EM+HFVM	+	-	+	+
Canada										
RATJEN et al.	2016	35	13.7 (5.8-17.9) ⁺	P	Case-control	EM, gen	-	-	+	+
RASHAKRISHNAN ET AL	2012	10	12.1 (7.8-15.4) ⁺	P	Case-control	EM, nNO, gen	-	-	+	+
Cyprus										
YIALLOUROS et al.	2015	20	NR (0.1-58.4) ⁺	R	Case series	EM or HFVM+nNO	+	+	+	+
Denmark										
ALANIN et al.	2016	24	24 (10-65) ⁺	P	Case series	EM, HFVM, gen	-	+	+	+
GREEN et al.	2016	28	12.4 (10.7-14.6) [¶]	P	Case-control	EM, HFVM	-	-	+	+
LOOMBA et al.	2016	17	13.8 ± 3.5 [#]	R	Case-control	EM, HFVM, nNO	-	-	+	+
MARTHIN et al.	2010	74	9 (4.4-43.7) ⁺	R	Case series	EM, HFVM, nNO	-	+	+	+
MADSEN et al.	2013	44	14.8 (6.5-29.7) ⁺	P	Case-control	EM, HFVM, nNO	-	-	+	+
GREEN et al.	2012	27	11.3 (6.3-18.5) ⁺	P	Case series	EM, HFVM	-	-	+	+
ELLERMAN et al.	1997	24	21± 13.6 [#]	P	Case series	HFVM	+	+	+	+
GROTH et al.	1989	14	23.5 (12-44) ⁺	P	Case-control	Clinical, EM	-	-	+	+
France										
FRIJA-MASSON et al.	2017	78	34.8 (28.6-47.1) [¶]	R	Case series	EM, gen or Kartagener	NA	+	+	+
VALLET et al.	2013	41	6 (3-11) [¶]	R	Case series	EM, HFVM	-	+	+	+
MAGNIN et al.	2012	20	7.5 (7.1-8.7) [¶]	R	Case series	EM	-	+	+	+
MAHUT et al.	2006	16	13.5 (12-17) [¶]	R	Case series	Clinical, EM	+	-	+	-
Germany										
TOLUSAKOW et al.	1981	21	NR (3-43) ⁺	R	Case series	clinical suspicion	-	-	+	+
Israel										
AMIRAV et al.	2016	13	10 (5-43) [¶]	R	Case series	all tests, including gen	-	-	+	-
COHEN-CYMBERKNOH et al.	2014	14	15.9±8.6 [#]	R	Case series	EM, HFVM, nNO, gen	-	-	+	-

Age is reported in years as mean ±SD[#], mean (range)⁺, median (IQR)[¶], or median (range)⁺ depending on the available information; NR, not reported
R, retrospective; P, prospective; EM, electron microscopy; HFVM, high frequency video-microscopy; nNO, nasal nitric oxide; gen, genetic analysis; NA, not applicable
Studies appearing in bold are those included in the meta-analysis

Italy											
	PIFFERI et al.	2015	45	14 (22.25) [¶]	P	Case-control	EM, HFVM, nNO	-	-	+	+
	MIRRA et al.	2014	22	10.5 (2-34) ⁺	P	Case series	EM, HFVM	- (not by age)	-	+	+
	VALERIO et al.	2012	24	7.6 (0.1-19) ⁺	P	Case series	EM, HFVM	-	-	+	+
	PIFFERI et al.	2012	50	In adults: 30.5 (18-47) ⁺ children: 11 (6-17) ⁺	P	Case series	EM, HFVM	-	-	+	+
	MAGLIONE et al.	2012	20	11.6 (6.5-27.5) ⁺	R	Case series	EM, HFVM	-	-	+	+
	MONTELLA et al.	2009	13	15.2 (10.4-29.3) ⁺	P	Case series	EM, HFVM	+	-	+	+
	SANTAMARIA et al.	2008	14	15 (7-27) ⁺	P	Case-control	Clinic, EM	-	-	+	+
Korea											
	KOH et al.	2000	19	12 (7-16) ⁺	P	Clinical trial	EM	NA	-	+	-
Saudi Arabia											
	AL SAADI et al.	2013	22	10.86±3.09 [#]	P	Case-control	EM+HFVM	NA	-	+	-
Sweden											
	MOSSBERG et al.	1983	24	30 (19-47) [*]	P	Case series	clinical suspicion	NA	-	+	-
Turkey											
	GOKDEMIR et al.	2014	24	12.9±2.7 [#]	P	Clinical trial	EM or Kartagener	NA	-	+	+
The Netherlands											
	PAFF et al.	2013	25	10.7 (7.1–14.5) [¶]	P	Case-control	according to consensus	NA	-	+	-
United Kingdom											
	SHAH et al.	2016	118	23.5 (10-36) [¶]	R	Case series	EM, HFVM, nNO,	-	+	+	-
	SUNTHER et al.	2016	150	11.4 (6-16.2) ⁺	R	Case series	EM, HFVM, nNO	-	+	+	+
	IRVING et al.	2013	33	24.66 [*]	P	Case-control	EM, HFVM, nNO	-	-	+	+
	SHOEMARK et al.	2008	20	40 (32-45) ⁺	P	Case-control	EM, HFVM	-	-	+	-
	PARASKAKIS et al.	2007	24	12 (8-16) ⁺	P	Case-control	EM, HFVM, nNO	-	-	+	-
	ZIHLIF et al.	2006	23	10.3 (9-14) [¶]	P	Case-control	EM, HFVM, nNO	-	-	+	+
	BUSH et al.	2006	19	9.5 ± 3.0 [#]	P	Case-control	EM, HFVM, nNO	-	-	+	+
	ZIHLIF et al.	2005	20	10.8 (9-14) [¶]	P	Case-control	EM, HFVM, nNO	-	-	+	-
	CSOMA et al.	2003	15	10.3 (7-14) [*]	P	Case-control	EM, HFVM	-	-	+	-
	HORVATH et al.	2003	14	35±4.6 [#]	P	Case-control	EM, HFVM	NA	-	+	+
	NARANG et al.	2002	31	11.0 (5.5-17.3) ⁺	P	Case-control	HFVM, EM	-	-	+	-
	KARADAG et al.	1999	21	10.8±3.2 [#]	R	Case-control	EM, HFVM	NA	-	+	+
	PHILLIPS et al.	1998	12	11 (7-15) ⁺	P	Case-control	HFVM, EM	-	-	+	+
USA											
	NOONE et al.	1999	12	34 (14-71) ⁺	P	Case series	Clinical, EM	- (not by age)	-	+	+
	REGNIS et al.	2000	12	36 (13-70) ⁺	P	Case-control	Clinical, EM	- (not by age)	-	+	+

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Multi-centre/international studies

Denmark, Italy, UK

MAGLIONE et al.	2014	158	8.7 (4.2–17.4)	R	Case series	according to consensus	NA	+	+	+
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Netherlands, Italy

SANTAMARIA et al.	2008	20	14.3 (4.6-27.5) ⁺	P,R	Case series	EM, LM	-	-	+	+
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Germany, Switzerland

NYILAS et al.	2017	49	14.7±6.6 [#]	P	Case-control	all tests	-	-	+	+
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NYILAS et al.	2016	20	13 ± 2.7 [#]	P	Case-control	according to consensus	-	-	+	-
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NYILAS et al.	2015	47	14.7 ± 6.9 [#]	P	Case-control	according to consensus	-	-	+	-
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Canada, USA

DAVIS et al.	2015	118	8 (5-11) [¶]	P	Case series	EM or gen	NA	-	+	-
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NOONE et al.	2004	78	adults: 36 (19-73) ⁺ children: 8 (1-17) ⁺	P	Case series	EM, HFVM, gen	+	-	+	-
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International PCD registry

WERNER et al.	2016	201	20 (1-75) ⁺	P	Case series	all tests, including gen	-	+	+	+
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