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Title: Multicentre longitudinal analysis of body mass index, lung function and sputum microbiology in primary ciliary dyskinesia

Dr. Marco 11326 Maglione maglione84@libero.it MD [1], Prof. Dr Andrew 11327 Bush a.bush@rbht.nhs.uk MD [2], Dr. Kim 19892 Nielsen kgn@dadlnet.dk MD [3], Dr. Claire 19893 Hogg c.hogg@rbht.nhs.uk MD [2], Dr. Silvia 19894 Montella amina2004@virgilio.it MD [1], Dr. June 19903 Marthin junekm@mail.dk MD [3], Dr. Angela 19914 Di Giorgio digiorgioangela@libero.it MD [1] and Dr. Francesca 19915 Santamaria santamar@unina.it MD [1]. [1] Department of Pediatrics, Federico II University, Naples, Italy ; [2] Department of Pediatric Respiratory Medicine, Royal Brompton Hospital, London, United Kingdom and [3] Department of Pediatrics & Adolescent Medicine, Copenhagen University Hospital, Copenhagen, Denmark .

Body: BACKGROUND: No studies have longitudinally assessed paired body mass index (BMI) and lung function in primary ciliary dyskinesia (PCD). Data about the relationship between sputum pathogens and spirometric evolution are also lacking. METHODS: We aimed to determine longitudinal BMI and spirometry changes in 162 PCD patients from London, UK (n=75), Naples, Italy (n=24) and Copenhagen, Denmark (n=63). Annual BMI and FEV_1 were analyzed over blocks of 2, 4 and 6 consecutive years. Isolation of P. aeruginosa (PA) from sputum was recorded in order to assess impact on lung function progression. RESULTS: Median age at first spirometry was 9 years (range, 4.2-27.2). Mean Z scores of first measured BMI and FEV_1, were -0.02, and -1.37, respectively. There were no significant changes in BMI and FEV_1 slopes, over any time block. During the follow-up PA was not related to lung function evolution, even though patients with at least one isolation of PA showed a trend towards worse lung function at 2 years.

		FEV1 Z score slope*				
	N	2 yrs	N	4 yrs	N	6 yrs
Ever P. aeruginosa	69	-0.06 (-0.19 to 0.08)	51	-0.09 (-0.17 to -0.01)	38	-0.07 (-0.14 to 0.004)
Never P. aeruginosa	93	0.07 (-0.04 to 0.17)	59	-0.02 (-0.08 to 0.05)	43	-0.05 (-0.10 to 0.002)
p		0.053		0.10		0.76

*Mean and 95% CI.

CONCLUSIONS: PCD subjects receiving centralized care show steady BMI and stable spirometry over medium term follow-up. Evolution of lung function could not be significantly correlated to PA isolation, although a trend was found towards decline of FEV_1 after 2 years of follow-up.