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Sinonasal features in patients with Primary Ciliary Dyskinesia - an EPIC-PCD study

Orphan diseases, Chronic diseases, Rhinitis

Y. T. Lam¹, J. F. Papon², M. Alexandru², A. Anagiotos³, M. Armengot⁴, M. Boon⁵, A. Burgess⁶, N. Caversaccio⁷, S. Crowley⁸, S. A. D. Dheyauldeen⁹, N. Emiralioglu¹⁰, E. Erdem¹¹, O. Gunaydin¹², E. G. Haarman¹³, A. Harris¹⁴, H. Ismail Koch⁶, B. Karadag¹⁵, C. Kempeneers¹⁶, S. Kim¹⁷, P. Latzin¹⁸, N. Lorent¹⁹, U. Ozcelik¹⁰, C. Pioch²⁰, A. L. M. Poirrier²¹, A. Reula²², J. Roehmel²⁰, C. Van Gogh²³, P. Yiallourous²⁴, M. Goutaki²⁵

¹Institute of Social and Preventive Medicine, University of Bern - Bern (Switzerland), ²1. AP-HP, Hôpital Kremlin-Bicetre, Service d'ORL et de Chirurgie Cervico-Faciale 2. Faculté de Médecine, Université Paris-Saclay - Le Kremlin-Bicêtre (France), ³Department of Otorhinolaryngology, Nicosia General Hospital - Nicosia (Cyprus), ⁴1. Department of Otorhinolaryngology, and Primary Ciliary Dyskinesia Unit, La Fe University and Polytechnic Hospital 2. Medical School, Valencia University Medical School, Valencia University - Valencia (Spain), ⁵Department of Paediatrics, University Hospital - Leuven (Belgium), ⁶Primary Ciliary Dyskinesia Centre, Southampton Children's Hospital, Southampton NHS Foundation Trust - Southampton (United Kingdom), ⁷Department of Otorhinolaryngology, Head and Neck Surgery, University Hospital of Bern - Bern (Switzerland), ⁸Paediatric Department of Allergy and Lung Diseases, Oslo University Hospital - Oslo (Norway), ⁹1. Department of Otorhinolaryngology, Head and Neck Surgery, Oslo University Hospital 2. Faculty of Medicine, University of Oslo - Oslo (Norway), ¹⁰Department of Pediatric Pulmonology, Hacettepe University, School of Medicine - Ankara (Turkey), ¹¹Department of Pediatric Pulmonology, Marmara University, School of Medicine, Istanbul - Istanbul (Turkey), ¹²Department of Otorhinolaryngology, Hacettepe University, School of Medicine - Ankara (Turkey), ¹³Department of pediatric pulmonology, Emma Children's Hospital, Amsterdam UMC, Vrije Universiteit - Amsterdam (Netherlands), ¹⁴Primary Ciliary Dyskinesia Centre, NIHR Respiratory Biomedical Research Centre, University of Southampton - Southampton (United Kingdom), ¹⁵Department of Pediatric Pulmonology, Marmara University, School of Medicine - Istanbul (Turkey), ¹⁶Division of Respiriology, Department of Pediatrics, University Hospital Liège - Liège (Belgium), ¹⁷AP-HP, Hôpital Kremlin-Bicetre, Service d'ORL et de Chirurgie Cervico-Faciale - Le Kremlin-Bicêtre (France), ¹⁸Paediatric Respiratory Medicine, Children's University Hospital of Bern, University of Bern - Bern (Switzerland), ¹⁹Department of Respiratory Diseases, University Hospital - Leuven (Belgium), ²⁰Department of Pediatric Pulmonology, Immunology and Critical Care Medicine, Charité-Universitätsmedizin - Berlin (Germany), ²¹Department of Otorhinolaryngology, University Hospital Liège - Liège (Belgium), ²²1. Biomedical Sciences Department, CEU-Cardenal Herrera University 2. Molecular, Cellular and

Genomic Biomedicine Group, IIS La Fe - Valencia (Spain), ²³Department of Otolaryngology/ Head and Neck Surgery, Amsterdam University Medical Center - Amsterdam (Netherlands), ²⁴1. Medical School, University of Cyprus 2. Pediatric Pulmonology Unit, Hospital 'Archbishop Makarios III' - Nicosia (Cyprus), ²⁵1. Institute of Social and Preventive Medicine, University of Bern 2. Paediatric Respiratory Medicine, Children's University Hospital of Bern, University of Bern - Bern (Switzerland)

Background: Most patients with primary ciliary dyskinesia (PCD) have sinonasal symptoms, but yet little is known on their frequency and severity. We aimed to describe sinonasal features in PCD patients using data from the Ear-nose-throat (ENT) Prospective International Cohort of PCD patients (EPIC-PCD), a multicentre observational clinical cohort.

Methods: We included patients who had a routine ENT clinical examination and completed the FOLLOW-PCD standardised symptoms questionnaire at the same visit. We compared reported symptoms and clinical findings in children and adults.

Results: We included 208 patients (108 males) with median age 16 years (range 0-63). 178 (86%) patients reported chronic nasal symptoms that in 73 (35%) persisted. Specifically, 116 (56%) reported blocked nose, and 129 (62%) rhinorrhoea. 18 of 72 adults reported anosmia compared to only 3 of 136 children. 133 (64%) patients reported headaches, which in 26 appeared when bending down. Clinical examination showed blocked nose in 88 (42%) patients. Nasal mucosa was evaluated in 190 patients and showed erythema in 64 and oedema in 61. 32 of 182 patients had nasal polyps (19 adults and 13 children) and 66 of 174 had hypertrophic turbinates. The Sinonasal Outcome Test-22 was available for 57 patients with a moderate median score of 39 (range 4-86), that did not differ by age.

Conclusion: The EPIC-PCD is the largest prospective cohort describing sinonasal features in PCD patients. Sinonasal symptoms are common in PCD patients, which underlines the need of ENT input in the multidisciplinary care. Sinonasal disease worsens with age as more adults present with nasal polyps and have impaired sense of smell.

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