07.06 - Paediatric respiratory epidemiology

32158
Otologic features in patients with Primary Ciliary Dyskinesia- an EPIC-PCD study

Orphan diseases, Chronic diseases

M. Goutaki[1], Y. T. Lam [1], M. Alexandru[2], A. Anagiotos[3], M. Armengot[4], M. Boon[5], A. Burgess[6], N. Caversaccio[7], S. Crowley[8], S. A. D. Dheyauldeen[9], N. Emiralioglu[10], E. Erdem[11], O. Gunaydin[12], E. G. Haarman[13], A. Harris[14], H. Ismail-Koch[6], C. Kempeneers[15], B. Karadag[11], S. Kim[16], P. Latzin[17], N. Lorent[18], U. Ozcelik[10], A. L. M. Poirrier[19], I. Rangnau[20], A. Reula[21], J. Roehmel[22], C. Van Gogh[23], P. Yiallouros[24], J. F. Papon[2]

[1]Institute of Social and Preventive Medicine, University of Bern - Bern (Switzerland), [2]1. AP-HP, Hôpital Kremlin-Bicetre, Service d'ORL et de Chirurgie Cervico-Faciale, Le Kremlin-Bicêtre 2. Faculté de Médecine, Université Paris-Saclay, Le Kremlin-Bicêtre - Paris (France), [3]Department of Otorhinolaryngology, Nicosia General Hospital - Nicosia (Cyprus), [4]1. Department of Otorhinolaryngology, and Primary Ciliary Dyskinesia Unit, La Fe University and Polytechnic Hospital 2. Medical School, Valencia University - Valencia (Spain), [5]Department of Paediatrics, University Hospital - Leuven (Belgium), [6]Primary Ciliary Dyskinesia Centre, Southampton Children's Hospital, Southampton NHS Foundation Trust - Southampton (United Kingdom), [7]Department of Otorhinolaryngology, Head and Neck Surgery, University Hospital of Bern - Bern (Switzerland), [8]Paediatric Department of Allergy and Lung Diseases, Oslo University Hospital - Oslo (Norway), [9]1. Department of Otorhinolaryngology, Head and Neck Surgery, Oslo University Hospital 2. Faculty of Medicine, University of Oslo - Oslo (Norway), [10]Department of Pediatric Pulmonology, Hacettepe University, School of Medicine - Ankara (Turkey), [11]Department of Pediatric Pulmonology, Marmara University, School of Medicine - Istanbul (Turkey), [12]Department of Otorhinolaryngology, Hacettepe University, School of Medicine, - Ankara (Turkey), [13]Department of pediatric pulmonology, Emma Children's Hospital, Amsterdam UMC, Vrije Universiteit - Amsterdam (Netherlands), [14]Primary Ciliary Dyskinesia Centre, NIHR Respiratory Biomedical Research Centre, University of Southampton - Southampton (United Kingdom), [15]Division of Respirology, Department of Pediatrics, University Hospital Liège - Liège (Belgium), [16]AP-HP, Hôpital Kremlin-Bicetre, Service d'ORL et de Chirurgie Cervico-Faciale, Le Kremlin-Bicêtre - Paris (France), [17]Paediatric Respiratory Medicine, Children's University Hospital of Bern, University of Bern, Switzerland - Bern (Switzerland), [18]Department of Respiratory Diseases, University Hospital - Leuven (Belgium), [19]Department of Otorhinolaryngology, University Hospital Liège - Liège (Belgium), [20]Department of Otorhinolaryngology, Head and Neck Surgery, Charité-Universitätsmedizin - Berlin (Germany), [21]1. Biomedical Sciences Department, CEU-Cardenal Herrera University, Castellón 2. Molecular, Cellular

and Genomic Biomedicine Group, IIS La Fe - Valencia (Spain), [22]Department of Pediatric Pulmonology, Immunology and Critical Care Medicine, Charité-Universitätsmedizin - Berlin (Germany), [23]Department of Otolaryngology/ Head and Neck Surgery, Amsterdam University Medical Center - Amsterdam (Netherlands), [24]1. Medical School, University of Cyprus 2. Pediatric Pulmonology Unit, Hospital 'Archbishop Makarios III' - Nicosia (Cyprus)

Background: Ear problems are common in primary ciliary dyskinesia (PCD), but little is known on their spectrum and severity. We aimed to characterize otologic disease in PCD patients using data from the ENT Prospective International Cohort of PCD patients (EPIC-PCD), a multicentre prospective observational clinical cohort.

Methods: We prospectively included patients with a routine ENT examination and a completed FOLLOW-PCD symptoms questionnaire at the same visit. We compared reported symptoms and clinical findings between children and adults.

Results: We included 208 patients (108 males) with median age 16 years (range 0-63). 114 (55%) reported ear pain, 61 (29%) ear discharge and 105 (50%) hearing problems. Reported symptoms did not differ by age. On examination 20/179 (11%) patients had tympanic perforation, 28/174 (16%) retracted membrane and 52/175 (30%) otitis media with effusion (OME). Tympanic sclerosis was found in 41/165 (25%) patients and 18 patients had grommets. OME was more common in children and sclerosis in adults.

68 out of 101 patients had an abnormal tympanogram. Audiometry was performed in 131 patients, showing hearing loss in 63 (48%), usually mild and bilateral. 2 adults had severe hearing loss. Reported hearing problems and audiometry results were discordant in 22 patients; 15 reported normal hearing with abnormal audiogram while 7 patients reported hearing problems with a normal audiogram.

Conclusion: This is the largest study describing otologic disease in PCD. Ear problems were common in patients of all ages although hearing loss was not perceived by some patients. This emphasizes the need for standardised ENT follow-up for all PCD patients.

Funding: SNF PZ00P3_185923