Nasal polyposis in Cystic Fibrosis: experience from CF Center, UHC Zagreb

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Cystic fibrosis (CF) is a lethal autosomal recessive disease, caused by mutations in the CF transmembrane conductance regulator gene (CFTR). CFTR mutations affect epithelial cells in the lungs, sinuses, pancreas, liver, kidneys, intestine and sweat glands, causing abnormally viscous mucus production, thickening of digestive fluids and salty sweat. The consequences for the respiratory system are mucus build-up, decreased mucociliary clearance and tissue inflammation. A change in microbioma follows, with S. aureus and P. aeruginosa being prevalent in most patients, as they have the capacity of biofilm formation causing chronic colonization. This represents the basis for recurrent infection. At the same time, pancreatic insufficiency leads to malabsorption of fat-soluble vitamins, i.g. vitamin D3, a powerful immunomodulator implicated in both pulmonary and sinus pathophysiology. CF is marked by a high incidence of nasal polyposis, even in the pediatric population and ENT follow-up is mandatory. CF nasal polyposis is a distinctive form of nasal polyposis and its treatment and follow up present many challenges.

CF affects one out of every 3000 newborns. The total number of patients with CF in Croatia is 175, and the majority of them now refer to our CF Center of Zagreb University Center where they are approached by a multidisciplinary team: both pediatric and adult pulmologists, gastroenterologists, endocrinologists, rhinologists, microbiologists and nutritionists. We aimed to review our experience with CF patients from a rhinologist point of view and present the prevalence and specifics of nasal polyposis in cystic fibrosis patients from our CF Center.

Key words: nasal polyposis, cystic fibrosis, sinus surgery, vitamin D3 deficiency