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Title: Chronic obstructive bronchopulmonary pathology in children with different connective tissue disorders (CTD) phenotypes

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Body: Changes in clinical symptoms of chronic obstructive bronchopulmonary diseases (COBPD) with high prevalence of CTD in children emphasize need for study of this phenomenon. Aim: to study clinical features of COBPD in patients with CTD phenotypes. Materials: 37 children aged 7-18yrs with COBPD and CTD phenotypes: unclassifiable (UP)(n=20), Ehlers-Danlos-like (EDLP)(n=15), Marfan-like (MLP)(n=2)- were observed. Asthma (A) was diagnosed in 32(86.5%), congenital bronchial abnormalities (CBA) - in 5(13.5%) patients. Results: All children with UP had A, associated with allergic rhinitis (AR) in 10(50%) patients, community-acquired pneumonia (CAP) in 5(25%) patients. 2(10%) patients with A and CAP had AR. In patients with EDLP A was diagnosed in 12(80%) patients, in 4(33.3%) of them - with AR, in 5(41.7%) – with CAP, in 3(25%) – with association AR and CAP. All patients with MLP had CBA. Pulmonary hypertension (PH) was in 5(25%) patients with A and UP. 3(15%) patients with A, UP and PH had pulmonary fibrosis (PF). Moderate A was diagnosed in all patients with EDLP, with PH in 7(58.3%) and PF in 4(33.3%) of them. Emphysematous bullae (EB) were detected on CT scans in 3(25%) patients with EDLP, A, PH and PF. CBA in patients with EDLP and MLP were with PH, PF, and with EB in all patients with MLP. Conclusions: 1. Close relationship between severity of A and CTD is revealed. 2. Severity of CBA manifestations in patients with MLP and EDLP depends on CTD severity. 3. Development of complications (PH, PF) in children with A, EDLP and MLP is 2 times more often than in children with UP. 4. It is advisable to consider A associated with AR, CAP and CTD as the clinical phenotypes of A.