RESPIRATORY DYSFUNCTION IN PATIENTS WITH THE MARFAN SYNDROME

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The Marfan syndrome (MS) is inherited in an autosomal dominant way. MS is characterised by the high degree of penetration and expression of the pathological gene. Fibryline, the gene product, is a protein that acts in the composing of elastic fibers. Therefore, MS is a multisystem disorder that affects connective tissue. The aim of this paper was to evaluate the frequency of occurrence of factors influencing abnormalities in respiratory tract functioning. The study encompassed 45 patients with MS ranging in age from 2 to 54 years. Both clinical examination and morphological measurements were performed in each case. Cephalometric and chest anthropometrical measurements were performed. All measurements were standardized to the mean values for healthy population. Based on the measurements recorded, the following indexes were evaluated: width/length index of the head, morphological index of the face, and index of chest flattening. The occurrence of chest deformities was also evaluated. The study indicate that several factors can result in pulmonary disease in patients with the Marfan syndrome.