Ehlers-Danlos Syndromes

Subjects: Pathology

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Ehlers–Danlos syndromes (EDS) are a clinically and genetically heterogeneous group of hereditary connective tissue disorders involving joint hyperlaxity, cutaneous hyperelasticity, and tissue fragility.

Keywords: Ehlers Danlos ; Rare Diseases ; oral health

1. Introduction

A revised classification containing 13 subtypes was published by the International EDS Consortium, with a number of clinical conditions to guide and improve the diagnosis of each subtype[1]. In the European Union, a disease is considered ‘rare’ if it affects fewer than one in two thousand people[2]. In the case of EDS, the prevalence varies, depending on the type: between 1:30,000 (classical type) and < 1:1,000,000 (arthrochalasia type), whereas for the rarest form (the periodontitis type), the prevalence is unknown[3].

EDS’ oral conditions have been described as periodontitis[1][4][5], temporomandibular dysfunctions[1][6], bleeding tendencies[7][8], enamel hypoplasia[1][9], shape abnormalities of the teeth and changes in the number of teeth[1][8], a high palate[1][10], dysgnathia, and malocclusion[1][11], as well as decreased effects of local anesthesia[12][13].

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