

The dental patient with a congenital bleeding disorder

Précis: With support and understanding of the diseases, dentists can be involved in the dental management and treatment of patients with congenital bleeding disorders. This paper outlines a simple algorithm for practitioners to use (**Table 1**).

Abstract: Congenital bleeding disorders account for approximately one in 10,000 births.¹ Dentists are often anxious about delivering treatment to this special group of patients. In the Irish Republic, patients with inherited bleeding disorders have their dental care co-ordinated centrally at the National Centre for Hereditary Coagulation Disorders (NCHCD), St James's Hospital, Dublin. Dental care is normally integrated with routine outpatient haematological appointments. This ensures regular monitoring of oral health and the early treatment of any hard/soft tissue pathology. This article describes, in simple diagrammatic form, the normal coagulation mechanism² (**Figures 1 and 2**), explains common coagulation terms (**Appendix 1**), and examines the three most common congenital bleeding disorders: haemophilia A, haemophilia B, and von Willebrand disease. General recommendations based on the current literature are provided with respect to procedures that are appropriate to perform in a general dental practice setting. Although not discussed in this article, it is important to note that non-coagulation bleeding disorders also exist. These include: hereditary haemorrhagic telangiectasia; blood vessel wall defects resulting from connective tissue disorders such as Marfan syndrome and Ehlers-Danlos syndrome; and, platelet disorders such as Bernard-Soulier syndrome, resulting in defective platelet adhesion.

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Christopher Vinall,
BDS, MFDRCSI
Medical student
Trinity College Dublin

Professor Leo FA Stassen,
FRCS(Ed), FDSRCS, MA, FTCD
Consultant Oral and Maxillofacial Surgeon.

Address for correspondence:
Professor Leo FA Stassen
National Maxillofacial Unit,
St James's Hospital,
James's Street,
Dublin 8.
Email: vinallc@tcd.ie

Oral health and orthodontic considerations in children with juvenile idiopathic arthritis: review of the literature and report of a case

Abstract: Juvenile idiopathic arthritis (JIA) is a severe disease of childhood, which comprises a diverse group of distinct clinical entities of unclear aetiology. Some abnormality of the immune system is present in all JIA cases. In its most severe clinical form, JIA may show localised and/or systemic complications, including functional impairment of the affected sites. This may result in variable growth and developmental anomalies. In many JIA cases, where the temporomandibular joint (TMJ) is affected, mandibular growth may be restricted, thus leading to the development of mandibular hypoplasia and/or retrognathism. As a result, it is not uncommon for JIA patients to present with skeletal Class II and open bite malocclusions. Furthermore, in JIA cases with unilateral TMJ involvement, craniofacial asymmetry may occur. In such cases, early orthodontic intervention facilitates both the skeletal and the occlusal rehabilitation.

Increased prevalence of dental caries and periodontal disease in JIA cases may be attributed to a combination of aetiological factors, including difficulties in executing good oral hygiene, unfavourable dietary practices and side effects from the long-term administration of medication. In addition, an association between periodontal disease and JIA has been reported based on their similar pattern of clinical dysregulation of the inflammatory process. This paper presents a brief description of JIA, with special reference to dental health and orthodontic treatment considerations. In addition, a case is presented where the appropriate orthodontic intervention led to the establishment of a normally functioning, as well as an aesthetically pleasing, occlusion.

KEY WORDS: juvenile idiopathic arthritis, craniofacial growth, TMJ, oral health, malocclusion, orthodontic treatment.

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Philippos N Synodinos

Orthodontist

Athens, Greece.

Ioannis Polyzois

Periodontist

Athens, Greece.

Correspondance:

Ioannis Polyzois

Dublin Dental School & Hospital,

Lincoln Place,

Dublin 2.

Email: ioannis.polyzois@dental.tcd.ie