Common and unusual dental development abnormalities in a patient with bicuspid aortic valve

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Abstract

Bicuspid aortic valve (BAV) is the most common congenital abnormality of the heart. In this condition, instead of three cusps, the aortic valve has two cusps. Children with congenital heart diseases are at increased risk of developing oral diseases, such as: higher number of decayed teeth, developmental anomalies, periodontal disease, malocclusion, dental crowding, as well as susceptibility to develop infective endocarditis from bacteremia caused by chronic poor oral health. However, little information is available regarding oral manifestations and their management in patients with congenital heart defects, despite the importance of these diseases. This paper presents oral manifestations associated with BAV in a young patient, alongside the general features of the condition. The presented case with BAV brings together features of a complex pathology and multidisciplinary treatment, which was conducted over a long period of time and still continues nowadays.

Keywords: bicuspid aortic valve, oral manifestations, developmental anomalies.

Introduction

Bicuspid aortic valve (BAV) is the most common congenital abnormality of the heart, in which, instead of three cusps, the aortic valve has two cusps [1–5]. BAV disease is not only a disorder of valvulogenesis, but also represents coexistent aspects of a genetic disorder of aorta and/or cardiac development [1]. Cardiac and valve morphogenesis occurs very early in fetal development [3], being of mesodermal origin [6].

BAV may be associated with many other cardiac and coronary arteries malformations and is also included in several genetic diseases, such as Shone’s, Williams, and Turner’s syndromes [1, 7, 8].

The symptoms of BAV develop insidiously and can vary usually from asymptomatic valve in infancy or childhood, to severe valve disease in adulthood [1, 4, 9]. Typical symptoms during adulthood are exertional syncope, angina and dyspnea [1, 9–11].

Congenital heart diseases are associated with developmental dental abnormalities (anodontia, microdontia, enamel hypoplasia, shape alterations) [7, 12–14]. Dental abnormalities result as an embryologic disorder in the anatomical development of the tooth buds [3]. They occur by disrupting any of the physiologic processes of dental development (initiation, proliferation, histodifferentiation, morphodifferentiation and mineral matrix formation) and can be detected as alterations in teeth number, shape, structure and size [15, 16].

Patients with congenital heart diseases present increased risk of oral diseases (caries, periodontal disease), having poor oral health [17], and being more prone to developing systemic effects of oral diseases [12, 18].

Evidence suggests a genetic inheritance pattern in both BAV and some dental development abnormalities (anodontia, microdontia, etc.) [9, 19]. Nevertheless, there is little information available regarding oral manifestations and their management in patients with congenital heart defects, despite the importance of these diseases [12].

Aim

The present paper aims to bring new information regarding oral conditions, such as dental perturbances, that patients with BAV are exposed to develop, issue that is not thoroughly covered by the current literature. With this respect, the case with BAV reported below brings together a complex general and oral pathology requiring a long-term treatment.

Case presentation

A case of a patient with BAV followed from infancy to adulthood is hereby presented. The general clinical features of the cardiac disease slowly increased in severity over time, such that in adulthood the patient presented the typical symptomatic triad (exertional syncope, angina and dyspnea).

During infancy, the only oral manifestation observed was anodontia of the upper primary lateral incisors.
The worsening aortic stenosis led to failure to thrive (low weight and height) and delayed wrist bones ossification observed after wrist radiography (skeletal age – seven years was less than the biological age – nine years) (Figure 1).

Periapical radiography of 6.5 (second left upper primary molar) at age of nine years revealed the presence high up in the maxillary bone of an inverted premolar forming its root toward the alveolar ridge, and tending to emerge into the maxillary sinus (Figure 2). The tooth was extracted (Figure 3) and the edentulous space was subsequently reduced by orthodontic treatment (Figure 4).

Clinical examination revealed the absence of all mandibular permanent incisors (subsequently restored by a prosthetic appliance – Maryland bridge), and of the upper lateral permanent incisors (edentulous space subsequently reduced by orthodontic treatment) (Figure 4).

The clinical oral examination also revealed that the permanent upper central incisors had developmental shape anomalies (chisel shape) (Figure 5).

Alongside the aforementioned oral manifestations, the clinical and radiological examination revealed an initial periodontal disease (Figure 4), condition frequently associated with congenital heart disease.

**Discussion**

The presented case falls within the general clinical picture of BAV, but exhibits a series of particularities.

The general signs and symptoms, as well as the disease’s evolution, were concordant with data in the specialty literature. Still, our patient presented a series of general conditions, which were not previously reported to be commonly associated with BAV, i.e., mild hypothyroidism and allergic eczema of the palms. Thus, hypothyroidism might have increased the delay in the somatic development caused by BAV, manifested by an appreciably lower height and weight as reported to the patient’s chronologic age [20].

With regards to the oral aspects, alongside the manifestations that frequently occur in patients with BAV, the presented case exhibited clinical features not common in these patients, i.e., dental shape anomalies (chisel shaped teeth), number anomalies (multiple teeth agenesis) and position anomalies (inverted tooth). All these dental anomalies are produced by the disturbance of the morphological stages and physiological processes of odontogenesis.

Dental agenesis is a developmental anomaly that represents the congenital absence of one or more teeth [21] and is produced by the disturbance of the initiation and dental bud stages and of the cellular proliferation process. What is noteworthy is that this patient had both anodontia of the upper lateral primary incisors (which is very rare, 0.5–0.9% of the population) and anodontia of six permanent incisors (which is more frequent, 1.6–9.6% of the population) [22]. The presence of this type of condition in both dentitions, primary and permanent, might suggest that the disruptive factor has acted for a long
period of time. Also, taking into account the fact that the patient’s brother also presented anodontia of one permanent tooth and the patient’s father presented milder features of anodontia, such as microdontia and conical shape of the permanent upper incisors (father was probably a carrier of the affected gene and transmitted it to his son) [23, 24], one may conclude that the etiological factor possibly had a genetic component, as in the case of BAV [25].

Dental shape developmental anomalies (chisel shape) are produced by disruptions of the bell stage and morpho-differentiation process, and their treatment is usually prosthetic if they seriously affect patient’s esthetics.

Inverted teeth are very rare. This developmental anomaly is represented by a tooth found in malposition included in the maxillary bone, a tooth whose roots are directed toward the alveolar ridge, which may/may not erupt in the mouth [26]. Some of the etiological factors of tooth inversion are systemic conditions like nutritional or endocrine disorders, genetic factors, and also local factors, such as previous trauma to the affected site during tooth growth, abnormal location of tooth bud during initiation, and follicular tooth sac inflammation [27]. The treatment in the case of inverted teeth is usually represented by its extraction, followed by orthodox treatment for the gap’s closure.

Conclusions

Children with BAV are at increased risk of developing various oral diseases, such as developmental dental abnormalities and periodontal disease. Taking into account the possibility of occurrence of oral diseases requiring multidisciplinary treatment, patients with BAV should be advised to establish interceptive and prophylactic measures necessary for prevention and early treatment.

Conflict of interests

The authors declare that they have no conflict of interests.

Author contribution

All authors contributed equally in the elaboration of the study.

References