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SHORT COMMUNICATION

Taurodontism: A minor diagnostic criterion in Laurence-Moon/Bardet-Biedl syndromes

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Abstract

Objective. The objective of the study was to present the prevalence of taurodontism in the permanent dentition in individuals with Laurence-Moon/Bardet-Biedl syndromes (LM/BBS). ***Methods and participants.*** Thirty-nine individuals were studied, which comprises the whole population of known LM/BBS patients in Norway. Data were collected retrospectively. Panoramic radiographs (OPG) were evaluated to document taurodontism. ***Results.*** Taurodontism was found in 82.9% of the individuals with LM/BBS. The second mandibular molars had the highest (72.3%) prevalence of taurodontism and the first mandibular molars the lowest (58.2%). ***Conclusion.*** This study suggests that taurodontism should be included as a minor diagnostic criterion for the Laurence-Moon/Bardet-Biedl syndromes (LM/BBS).

Key Words: *taurodontism, Laurence-Moon/Bardet-Biedl syndromes (LM/BBS)*

Introduction

LM/BBS

Laurence-Moon-syndrome and Bardet-Biedl-syndrome have traditionally been referred to as one single condition with the name Laurence-Moon-Bardet-Biedl syndrome (LM/BBS) [1,2]. It is now known that Bardet-Biedl syndrome (BBS; MIM 209900) is a distinct clinical entity similar to, but different from Laurence-Moon syndrome (LMS; MIM 245800), mainly by the absence of spastic paraplegia and presence of polydactyly [2–4]. BBS is typified by developmental and progressive degenerative defects characterised by retinal dystrophy, obesity, polydactyly, kidney dysfunction, hypogonadism and cognitive impairment [4–6]. BBS is a rare but severe syndrome that is often mis- or undiagnosed [3–6]. There is no definite treatment, but early diagnosis may reduce the disability.

Dental problems such as delayed eruption, hypodontia, small teeth, enamel hypoplasia, short roots, mild micrognathia and a thickened mandibular body appear to be common in individuals diagnosed with LM/BBS [5–8].

Taurodontism

Taurodontism is a variation in tooth form characterized by a vertically elongated pulp chamber, increased crown-to-root ratio and a lack of constriction at the tooth-cervical margin [8–10]. This condition occurs as a consequence of disturbances of the developmental processes of the root and an unsuccessful invagination of Hertwig's epithelial root sheath [10,11] (Figure 1). Taurodontism may affect both the primary and permanent dentition [12]. Taurodont teeth are found in diverse populations around the world with a prevalence of 0.3–11.3% depending on the definition of taurodontism [13]. Permanent second and third molars are more commonly affected than the first molar [14]. The aetiology of taurodontism is unclear. It is thought to be multifactorial and includes genes (ALPL and DLX3), environmental factors such as infection (osteomyelitis), high-dose chemotherapy or a history of bone marrow transplantation and their interaction [11,15]. Taurodontism occurs most frequently as an isolated anomaly or in association with several X-linked conditions [16]. such as amelogenesis imperfecta [11] and hypodontia [17].

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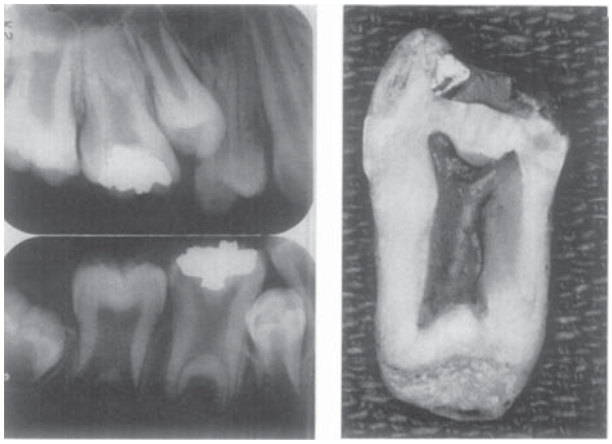

Figure 1. Left: Maxillary and mandibular x-rays showing excessive roominess of pulp chambers, characteristic of taurodontism. Right: Gross view of mandibular right first molar from the same patient [14].

Taurodontism has been reported in individuals with cleft lip and palate and in individuals with multiple-system malformations such as Down syndrome, ectodermal dysplasia, Klinefelter syndrome, oro-facio- digital syndrome, tricho-dento-osseous syndrome and van der Woude syndrome [18–23]. Taurodontism has also been described together with hypodontia in many syndromes, such as Smith-Magenis, Williams, McCune-Albright and Ellis-van Creveld syndromes [20].

Methods and participants

The present study is a retrospective record review study which was carried out at the TAKO-Centre, National Resource Centre for Oral Health in Rare Medical Conditions, Lovisenberg Diakonale Hospital, Oslo, Norway. Thirty-nine participants constituted the whole population of known LM/BBS individuals in Norway at the time of examination (1998–2007). Taurodontism was assessed using panoramic radiographs (OPG), which were available for 35 individuals (18 males, 17 females) with LM/BBS. The mean age was 25.9 years with an age range of 8.0–61.2 years.

Taurodontism of the mandibular 1st and 2nd permanent mandibular molars was assessed by measuring and calculating the CB/R ratio (crown-body/root ratio) on OPGs according to Seow and Lai [17] (Figure 2). The limit values for diagnosing taurodont teeth of CB/R was ≥ 1.10. Previous studies have shown that there is minimal distortion of radiographic images of the permanent mandibular molar on OPGs [17,22,24].

Criteria for exclusion from this study

Four individuals with all mandibular molar missing were excluded from the study.

Results

The prevalence of taurodontism was calculated as 82.9% among the 35 individuals with LM/BBS. The mandibular molar most affected was the second molar on the left side (78.3%). The second molar was affected more often (72.3%) than the first molar (58.2%) (Figure 3). Of the 102 molars in the mandible examined 66 teeth appeared as taurodont teeth giving the LM/BBS population at tooth level a prevalence of 64.7%.

Discussion

The estimated prevalence of taurodontism of 82.9% in this study is much higher than the prevalence of taurodontism of 0.3–11.3% in the general population [13]. The second molar was affected more often (72.3%) than the first molar. This is in agrement with Shaw [19], who suggested that the second molar should be considered as the 'standard tooth' for determining taurodontism. We chose to use CB/R ratio instead of tooth length linear measurement because in a radiographic study alteration in tooth angulation is known to affect the radiographic tooth length, but not the ratio [25].

Magnification may vary between OPGs taken using different machines, and also between different regions on the same radiographs. However, Stramotas et al. [24] stated that the R/C ratio can be measured accurately from OPGs and is reproducible when a patient is correctly positioned. CB/R ratio of permanent teeth can be measured from OPGs with acceptable reproducibility. Therefore, the results from this study can be used as a baseline for taurodontism in individuals diagnosed with LM/BBS.

Since none of the participants had undergone a genetic screening, we could not determine whether the patients had Bardet-Biedl or Laurence-Moon

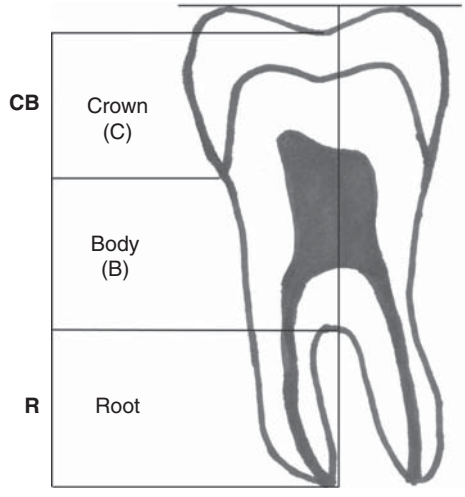

Figure 2. Assessment of the tooth morphology was based on measurements of Crown (C), Body (B) and Root (R) [21].

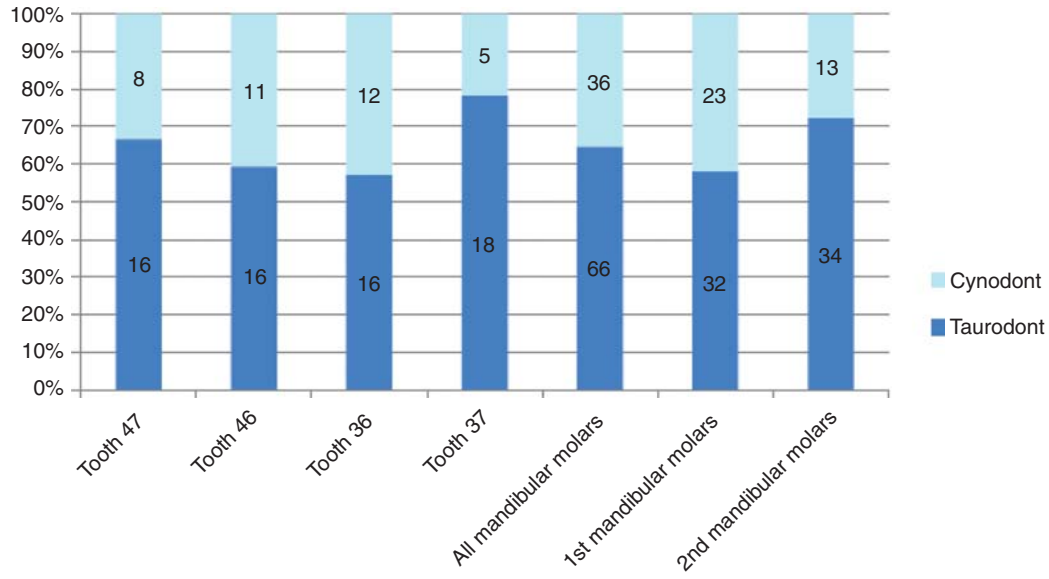

Figure 3. The findings of taurodont teeth of the mandibular molars from the radiographic study. Molars missing in the mandible were excluded from the statistics, since they could not be diagnosed.

syndrome. However, all were diagnosed by an experienced clinical geneticist based on accepted diagnostic criteria at that time [2,26].

It is highly likely that taurodontism is a trait that should be considered, amongst other dental anomalies, to assist with the diagnosis in patients with suspected LM/BBS.

Conclusion

We suggest that taurodontism should be included as a minor diagnostic criterion for LM/BBS.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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