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Research Article

Epidemiological Aspects of Dental Anomalies in Moroccan Patients with Rare Diseases.

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Summary

Children with rare diseases present various clinical manifestations affecting different structures and viscera. The oral cavity is no exception and shows several dental anomalies. These anomalies are very frequent but not well studied in our population. Thereby, the present study was conducted to evaluate the epidemiological and clinical features of dental anomalies in Moroccan patients with rare diseases.

Material and methods: A descriptive study was carried out on a sample of children and adolescents with rare disease recruited from the Pedodontics and the Medical Genetics Departments of ibn Rochd University Hospital of Casablanca. Each patient underwent clinical and radiographic examinations. Sociodemographic, clinical and para-clinical data were collected.

Results: A total of 65 patients with rare diseases were included in the study. 44.6% of the cases had a labeled rare disease while 55.4% of the patients were awaiting diagnosis. Dental anomalies were present in 76.9% patients including developmental anomaly, structural anomaly and eruption anomaly with frequencies of 45.5%, 21.1% and 33.3% respectively.

Conclusion: The prevalence of dental anomalies in the present study underlines the importance of a systematic dental examination in patients with rare diseases. The objective of this examination is to detect the pathognomonic oral signs of certain rare diseases which could contribute to the acceleration of the diagnostic process and to take care of these dental disorders at an early stage.

Introduction

The qualification of a disease as rare results from the observation of the disease below a given threshold. For the European organization for rare diseases (EURORDIS), this threshold is set at 1/2000. This threshold has been adopted by several European countries, but not all of them. Indeed, lower thresholds of 1/10000 and 1/50000 have been adopted by Denmark Sweden, and United Kingdom respectively. However, in the United States the threshold is set at 1/1000. In Morocco, these pathologies affect approximately 1.5 million people (Alliance Maladies Rares Maroc) [1,2]. Today, the number of rare diseases is estimated at between 6000 and 8000. These diseases are characterized by a great disparity in terms of frequency, i.e., within this group of diseases, some are even rarer than others according to a gradient of rarity [1,3]. Consequently, the diagnosis of these diseases takes far too long. Moreover, these patients generally receive symptomatic treatments before the discovery of the disease. Basic treatment is limited to certain less rare pathologies, because the fewer people affected, the less likely it is to be treated adequately [1,3]. On the other hand, the etiology of many rare diseases remains unknown. Still, genetic origin explains a large number of these diseases. Other etiologies may be responsible, such as infectious, tumoral or other origins.

These rare diseases are very expressive in terms of clinical manifestations, affecting different structures and viscera. The oral cavity is no exception and shows several dental anomalies [1,4]. Indeed, these rare pathologies disturb the fundamental factors of odontogenesis and lead to the appearance of one or several dental anomalies. This can be an alteration of the external appearance, internal structure or topography of one or more teeth. These developmental disorders may be manifested by variations in the number, position, size, shape, eruption or structure of the teeth [5]. These dental changes have been reported to be important components in many rare syndromic diseases. Their presence can be a useful diagnostic clue for rare diseases [5]. According to the literature, among more than 5000 known syndromes, more than 900 have dento-orofacial signs and 750 are associated with orofacial clefts[4]. Despite the diversity of dental manifestaions in rare diseases, very few studies have addressed this topic. Generally, studies have evaluated the prevalence of dental anomalies in some specific patients not affected by rare diseases such as patients undergoing orthodontic treatment and, patients with non-syndromic cleft lip and palate[6-12]. The aim of the present study was to analyze the epidemiological and clinical features of dental manifestations in Moroccan patients with rare diseases observed in the Pedodontics Department (CCTD of Casablanca) and the Medical Genetics Department (Ibn Rochd University Hospital of Casablanca).

Material and methods

Study Population and Data Collection

A descriptive cross-sectional study was conducted in two departments of Ibn Rochd University Hospital namely the Pedodontics departments at the Casablanca Dental Consultation and Treatment Center and the Molecular Genetics and Medical Biology Department. The survey was conducted for one year (January 2019-December 2019).

All patients meeting the criteria below, were included in this study:

a) Patient with an identified or pending rare disease.

b) Age between 3 and 19 years

c) Dental status allowing the examination of a possible dental anomaly (Absence of severe EPC,

Absence of iatrogenic traumatic lesions, Absence of extensive and multiple coronal fillings).

a. Optimal cooperation in the management of the panoramic radiograph.

b. Informed consent of the parents.

Data were collected using a form, designed by the Pedodontics Department, with 3 sections:

a) Patient identification: sex, age, city, department assigned and reason for consultation.

b) General condition: consanguinity, rare disease, family history, affected ectodermal derivatives, craniofacial syndrome, affected craniofacial structure and general manifestations.

c) Dental status: dentition and dental anomaly.

All patients benefited from a general examination, a dysmorphic examination and a radiographic examination (a panoramic image). Statistical analysis was performed using the SPSS software in the laboratory of epidemiology and bio-statistics of the Faculty of Dentistry of Casablanca

Results

The study involved 83 patients with an identified or pending rare disease. 18 patients were excluded from the study because of lack of cooperation. The general characteristics of the patients (age, sex, reason for consultation) are presented in (Table 1). Consanguinity was observed in 38 patients (58.5%). Most of them (71.1%) had 1st degree consanguinity. Eight patients (12.3%) have reported a positive family history especially in female siblings (75%). In 29 patients (44.6%), the rare disease was diagnosed, while the diagnosis is in progress in 36 patients (55.4%). The study revealed 22 labeled rare diseases with various clinical manifestations (Table 2). Moreover, the results showed that 50 patients (79.9%) had at least one dental anomaly (Table 3). Different types of dental anomalies were observed including developmental anomalies (45.5%), structural anomalies (21.1%) and eruption anomalies (33.3%). Among patients with dental anomalies, 31 cases (62%) were born to consanguineous marriage. Concerning developmental anomalies, the study showed the presence of 64 teeth with some types of developmental anomalies. Indeed, 12 teeth had a shape anomaly (18.8%), 12 teeth had a size anomaly (18.8%), 18 teeth had a number anomaly (28.1%) and 14 teeth had a position anomaly (21.9%) (Table 4). Furthermore, structural anomalies were very varied and have affected 14 patients (Table 5). Eruption anomalies were observed in 22 patients (33.3%) affecting 24 teeth (Table 6). Dental anomalies were also studied according to the identified rare diseases and those in the process of being identified. Among all the patients diagnosed with dental anomaly, 21 cases (42%) had an identified rare disease, 29 cases (58%) had a rare disease under identification. Table 7 shows the different dental anomalies observed for each identified rare disease.

Table 1: General characteristics of the patients.

Variables	N	%
Age		
· [2-5]	7	10,7

· [6-10]	27	41,6
· [11-19]	31	47,7
Gender		
· Male	34	52,31
· Female	31	47,7
Consultation department		
Pedodontics-Prevention:	42	64,6
Medical genetics and molecular biology:	23	35,4

Table 2: Rare disease and its manifestations.

Variables	N	%
Rare disease identified		
Yes	29	44,6
No	36	55,4
Ectodermal derivatives affected		
Yes	57	87,7
No	8	12,3
General attack		
Yes	64	98,5
No	1	1,5

Table 3: Distribution of the sample according to the presence of the dental anomaly.

Variables	N	%
Child with dental anomaly		
Yes	50	76,9
No	15	23,1

Table 4: Distribution of developmental anomalies.

Developmental abnormalities	N	%
Anomalies type and development		
Shape	12	18,8
Size	12	18,8
Numbers	18	28,1
By position	14	21,9
Shape anomalies		
Gemination	1	8,3
Fusion	1	8,3
Invagination	0	0
Taurodontism	2	16,7
Others	8	66,7
Size anomalies		
Localized macrodontia	3	15
Generalized macrodontia	2	10
Localized microdontia	1	5

Generalized microdontia	2	10
normally affecting the length of the roots	4	20
Number anomalies		
Agenesis	18	100
Supernumerary tooth	0	0
Positional anomalies		
Transposition	1	7,1
Rotation	4	28,6
Ectopy	0	0
Anastrophy	8	57,1
Malposition		

Table 5: Distribution of structural anomalies by type.

Variables	N	%
Structural anomalies		
Enamel anomaly	15	83,3
Dentine anomaly	2	11,1
Enamel and dentin abnormality.	1	5,6
Hereditary structural anomalies of the enamel		
Hypoplastic amelogenesis imperfecta		
Structural abnormalities of the acquired enamel	13	86,7
Moderate Fluorosis	2	15,4
training due to tetracyclines	1	7,7
MIH	10	77
Light	5	50
Moderate	3	30
evere	2	20
Dentin anomalies		
· Dentinogenesis imperfecta Type 1:	1	50
· Dentin Dysplasia Type 1:	1	50
Enamel and dentin anomalies		
Generalized Odontodysplasia	1	100

Table 6: Distribution of rash anomalies by type.

Eruption anomalies	N	%
Late eruption	3	12,5
Delayed eruption	18	75
Others	3	12,5
Total	24	100

Table 7: Main dental anomalies observed in each rare disease identified.

Rare disease and definition	Clinical description of dental anomalies	Radiological description of dental anomalies
Bekwith-wiedman : genetic disease characterized by overgrowth, tumor predisposition and congenital malformations		-

Malformative syndrome of Cornelia: syndrome of variable expression characterized by a very recognizable facial dysmorphia accompanied by a deficit of severity, an important growth retardation at ante-natal onset (second trimester), anomalies of the extremities and others.	- Localized Macrodontia	- Enlarged roots in the mesio-distal and cervical-apical direction with a voluminous pulp cavity.
Cri-de-chat: Chromosomal abnormality resulting from a deletion of variable size of the end of the short arm of chromosome 5. The clinical manifestations are: an acute monochromatic cry, microcephaly, a characteristic craniofacial dysmorphism, a significant mental and psychomotor delay.		-
Crouzon: It is a craniofacial dysostosis of the acrobrachycephaly or oxycephaly type, often of hereditary origin, but there are sporadic cases.		-
Dyggvemechiorclausen : disease of genetic origin, belonging to the group of spondyloepithelial Dysplasias, characterized by a severe and progressive statural deficit at the expense of the trocar, a thorax, shield, microcephaly, intellectual deficiency and a pathognomonic radiology.	- Agenesis	Absence of germs
Ectodermic Dysplasia: genetic disease characterized by the damage of at least two derivatives of the ectoderm (polish, hair, teeth, sweat glands, nails).	- Multiple agenesis	- Absence of tooth germs
	-	- Retention of permanent teeth due to persistence of temporary canines and molars
	- Oligodontia	- Short roots
	-	- Root dysmorphia
	- Delayed eruption	- Mesial/distal tilt of some retained permanent teeth
	-	
	- Anomaly of shape	
	-	
	- Anomaly of position	
Goldenhar: oculo-auriculo-vertebral dysplasia. It is a malformative syndrome related to developmental anomalies of the first branchial arch	- Malposition	- Delayed maturation
	- Delayed eruption	
Hallerman-Streif:	- Agenesis	Amelo-dentinal hypoplasia
Syndrome characterized by a typical face with beak nose and hypoplasia of the mandible, small harmonious size, hypotrichosis, microphthalmia with congenital cataract, hypoplasia of the clavicles and ribs.	-	
	- Generalized odontodysplasia	Large pulp volume, apical displacement of the furcation area
	-	
	- Anomaly of shape	Short roots, gaping apices
		Taurodontism
Hurler: It is a severe form of mucopolysaccharidosis type 1. These degenerative genetic diseases, transmitted in an autosomal recessive mode, are due to the deficiency of an IDUA enzyme	- Ectopy	- Malpositioned teeth, far from the normal site
	-	-
	- Mild MIH	- Reduced contrast enamel
Ichthyosis: is a scaling and exfoliation of the skin that ranges from mild but embarrassing dryness to severe disfiguring disease.	- Hypoplastic amelogenesis imperfecta	Reduced enamel thickness
	- Delayed eruption of premolars	
		-Delayed root maturation in permanent molars

Kabuki: it is a rare congenital malformative syndrome characterized by pre- and post-natal growth retardation, intellectual deficit, facial morphological variations and various malformations affecting various viscera.	- Delayed eruption	- Short roots, wide canals, - - Tooth retention, delayed root maturation - - Anomaly affecting the length of the roots.
Krabbe disease: an autosomal recessive disorder caused by a deficiency of galactocerebrosidase, a lysosomal enzyme involved in the catabolism of a major lipid component of myelin.		-
Langer Giedion: syndrome characterized by the combination of an intellectual deficit and numerous abnormalities including excess skin, a characteristic face and cone-shaped epiphyses in the phalanges.	- Merger of the 83 and 82 - - Generalized macrodontia - - Agenesis of 15, 25 and 32 - - Delayed eruption	Delayed maturation of the second premolars
Marfan: is a rare genetic disease. It is characterized by damage to one or more organs and can result in skeletal, ophthalmological and cardiac disorders	- Agenesis of premolars	Temporary molar roots are thin, tapered and long Absence of the germs of the premolars Root length abnormality Delayed radioulnar maturation in the six-year-old teeth.
Noonan: genetic syndrome manifested by a particular aspect of the facial features, malformations of the heart and a small size	- Hypoplastic amelogenesis imperfecta	Reduction of enamel thickness, also visible in pre-eruptive teeth
Pentasomy X: anomaly of the X sex chromosome, due to the presence of three supernumerary X chromosomes in women (49, XXXXXX instead of 46, XX)	- Generalized macrodontia	Mesio-distal and cervico-apical dimensions are important
Praderwili : genetic disease caused by an abnormality of chromosome 15, it occurs accidentally during conception.		-
Pudlack-Hermansky : It is a multisystem disorder characterized by oculocutaneous albinism, hemorrhagic diathesis and in some cases neutropenia, pulmonary fibrosis or granulomatous colitis. 		- Absence of the germ of the 48 - - Taurodontism on 47 and 37
Mosaic trisomy 21: it is a disease characterized by the presence of both normal cells and trisomic cells		-
West: It is a severe form of epilepsy that manifests itself by the occurrence of a series of involuntary muscle contractions in infants.		-
Wiliams: genetic disease that manifests itself by cardiac malformations, infantile hypercalcemia, mild to moderate intellectual delay, physical and behavioral characteristics, hypersensitivity to noise	- Hypo-mineralization of fluorotic appearance - - Retention of 25, 35 and 45	Delayed root maturation in permanent molars
X fragile: a genetic disease that most often results in intellectual deficits, behavioral problems and physical abnormalities that vary from person to person .	-	-

Discussion

The present study has enrolled 65 patients with a rare disease. Our sample size is quite large compared to previous studies such as the study of Hubert Désiré Mbassi Awa et al. [13], which analyzed 37 patients with rare diseases. This final sample size was achieved because the recruitment was conducted in two departments potentially frequented by patients with rare diseases. Nearly half of our patients were male aged between 11 and 19 years. Rare disease was not yet labeled in over half of the cases. Therefore, these patients did not benefit from etiological treatment, their management was limited to symptomatic treatments. In addition, the present study found parental consanguinity in 58.5% of all patients especially the 1st degree consanguinity. As previously reported, the relationship between consanguinity and genetic abnormalities has been confirmed [14]. On the other side, most of the patients had consulted a pedodontics. This has facilitated the investigation as these patients have been treated in the department and were familiar with the dental environment. Indeed, the prevalence of dental anomalies in this series was higher compared to healthy patients, patients receiving orthodontic treatments, and patients with non-syndromic cleft lip and palate [4,6,15-19].

The results of the major studies that addressed this topic are summarized in Table 8. It should be noted that dental anomalies have rarely been evaluated in patients with rare diseases. Majority of studies are descriptive. For example, the study conducted by Hubert Désoré Mbassi Awa et al. [13] , was interested in all oral manifestations including dental anomalies. This study reported a high prevalence of 97.2%. However, this result does not provide information on the prevalence of dental anomalies since the analysis also includes dental caries and MDD. Given the scarcity of surveys on rare diseases and dental anomalies, the distribution of the latter was compared to the results reported in healthy patients. Our study showed that 62% of patients with dental anomalies were born to consanguineous marriage. This is consistent with the results of Saima Y Khan et al, and Umamaheswari T N et al. These findings highlight the implication of genetics in tooth development and formation [20,21].

In our study, developmental anomalies were the most common, followed by eruption anomalies and structural anomalies. Regarding dental anomalies, the majority of patients with a rare disease had at least one dental anomaly. This finding suggest a possible causal link between rare diseases and dental anomalies. Other studies have shown similar prevalence of dental anomalies [22-26]. Furthermore, the obtained results are in line with the commonly accepted finding that developmental anomalies are more common than structural anomalies [27]. With regard to developmental anomalies, numerical abnormalities were manifested mainly by dental agenesis. Several studies carried out in different countries have demonstrated the important prevalence of dental agenesis compared to other types of numerical dental anomalies. The analysis of the characteristics of eruption anomalies shows that delayed eruption was the most frequent form and represented 75% of all eruption anomalies. Structural anomalies are the least common and are characterized mainly by acquired enamel anomalies. Several studies have reported the rarity of structural anomalies affecting enamel, dentin or both [28,29]. (Table 9) The comparison of dental anomalies observed in rare diseases with the literature enriched the list of possible dental anomalies observed in these diseases by adding to the existing anomalies new types that have not been previously reported [30-50]. Furthermore, the present study have identified for the first-time some dental anomalies associated with some rare diseases such as Langer Giedon's disease, for which our results showed the presence of dental fusion, agenesis and macrodontia.

Table 8: Prevalence of dental anomalies in different populations.

Country	years	Study Population	Sample size	Examination used	Prevalence of DAs*.
Iran (15)	2007	Healthy patients	480	Panoramic X-ray	40,8%
Arabie Saudi (4)	2012	Healthy patients	878	Panoramic X-ray	45,5%
Norway (6)	2013	Healthy patients	500	Panoramic X-ray	28,2%
France (16)	2018	Patients receiving orthodontic treatment	551	Intra-oral photography and pan-oramic X-ray	45,74%
New York (17)	2013	Patients with Down syndrome	496	Clinical examination and panoramic X-ray	20,4%
Iran (30)	2013	Healthy patients	1224	Clinical examination and panoramic X-ray	32,4%
Brazil (18)	2013	Patients with cleft lip and palate	296	Clinical examination and panoramic X-ray	39,9%
Brazil (19)	2016	Patients with Down syndrome	105	Panoramic X-ray	50,47%
Arabie Saudi (26)	2016	Healthy patients	1000	Panoramic X-ray	37,8%

Yemen (7)	2019	Healthy patients	1675	Panoramic X-ray	22,96%
Notre etude	2020	Patients with rare diseases	65	Clinical examination and panoramic X-ray	76,9%

Table 9: Comparison between the dental anomalies identified in our study and those reported in the literature.

Syndrome	Dental anomalies found in our study	Dental anomalies found in the literature
Goldenhar (31, 32)	Eruption delayed	Supernumerary tooth
		Structural abnormality of enamel and dentin
		Delayed tooth eruption
Marfan (33, 34)	- Root length anomaly	Concurrent presence of supernumerary teeth and dental agenesis
	-	
	- Agenesis	Abnormalities in root length
		Dentinogenesis imperfecta
		Enamel hypoplasia at the PM
Pudlack-Hermansky (35)	Agenesis	-
Ichthyosis (36)	Hypoplastic amelogenesis imperfecta	Structural abnormality
	Delayed eruption, persistence of temporary teeth	Delayed eruption
		Anomaly of position
Hallerman-Streif (37)	- Agenesis	Structural abnormality
	-	
	- Generalized odontodysplasia	Anomaly of eruption, shape and number
	-	
	- Anomaly of shape	
Ectodermic Dysplasia (38)	Agenesis	- Agenesis
		-
	Oligodontia	- Oligodontia
		-
	Delayed eruption	- Delayed eruption
		-
	Anomaly of shape	- Anomaly of shape
Kabuki (39)	**- Root length abnormality**	Hypodontia
	-	
	- Delayed eruption	Agenesis
		Fusion, Gemination

Langer Giedion (40)	Fusion	Supernumerary tooth
	Generalized macrodontia	Delayed eruption
	Agenesis	Dental retention
	Delayed eruption	
Crouzon (41)	-	Supernumerary tooth, Mesiodens
		Dental agenesis
		Delayed eruption
Wiliams (42)	Moderate Fluorosis	Shape anomaly
		Dental agenesis
		Microdontia
Noonan (43)	Hypoplastic amelogenesis imperfecta	Supernumerary tooth
		Anomaly affecting the root shape of molars
		Dental retention
Hurler (44)	- Ectopy	Hypodontia, microdontia
	- MIH light	
		Anomaly of enamel structure
		Anomaly of shape
		Taurodontism
Pentasomy X	Generalized macrodontia	-
Beckwith Wiedman (45)	-	Anomaly of shape, size, number and structure
Praderwili (46)	-	Delayed tooth eruption
		Enamel abnormality
		Enamel hypoplasia
		Microdontia
Mosaic trisomic	-	-
West (47)	-	- Anomaly of shape and position
		-
		- Anomaly of eruption
		-
		- Anomaly of number
Krabbe	-	- -

X fragile	-	Anomaly of position
		Anomaly of shape
		Supernumerary teeth
Cornelia (48)	Localized macrodontia	Microdontia
		Delayed eruption
		Anodontia
Dyggve Mechior Clausen	- Agenesis	-
Cri de chat (49, 50)	-	Enamel hypoplasia

Conclusion

The results of the present study showed that rare diseases are prone to complex and diverse dental anomalies. The diagnosis of these dental manifestations is important since it will allow us to collect data that can be used to detect the common characteristics of rare diseases. This will enrich the clinical picture of these diseases and contribute to their diagnosis. In addition, the early management of dental anomalies will promote the oral health of patients with rare diseases, thus improving their quality of life.

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