

A study of elbow crease and some dental anomalies in patients with downs' syndrome

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Abstract

Backgrounds:

Downs' syndrome is a genetic disease resulting from trisomy 21 chromosome. It was the first chromosomal abnormality discovered in humans and has also been observed in other primate.

Patient and methods:

The elbow crease associated with enamel anomalies in teeth was studied in 44 males and 16 Females patients with Down syndrome in mentally retarded institutions aged from 8-10 years in Baghdad city.

Aim of study:

The aim of the present study is to know the frequency of the presence or absence forms of this crease: in addition to know the relation between elbow crease and enamel anomalies in primary and permanent teeth from the genetic side.

Result:

Shows difference was statistically highly enamel anomalies teeth compared with control group, therefore, the result demonstrate that both age groups patient male & female enamel anomalies teeth compared with absence of elbow crease a higher significant differences.

Conclusion:

Absence of elbow crease was predict concerning enamel anomalies teeth in patients with DS.

Keywords: elbow crease, dental anomalies. Down's syndrome.

Introduction

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Down syndrome, also denominated as trisomy 21, is a genetic alternation in which the affected individuals carry an extra chromosome 21 ^[1].

It was the first chromosomal abnormality discovered in humans and has also been observed in other primate species, including the chimpanzee. It occurs in 0.5% of all conceptions and one in 900 live births ^[2]. The cranio facial and oral feature involved in down syndrome include brachycephaly (condition where the head is disproportionately wide), usually small nose associated with alow nasal bridge, small maxilla ,givalpalate and tongue with fissures and papillary hypertrophy ^[1]. Children with down syndrome have smaller brain volume than other children might be responsible for the particular features of mental retardation that in some way results from trisomy 21 ^[3]. The hands of down syndrome patients are typically broad and short with thick, stubby fingers, The ridges of their hands are ill-formed and dotted.

Etiology of down's syndrome

The causes of down's syndrome are a variety of genetic and environmental factors have been proposed, including radiation, viral infection, hormonal levels and gentic predisposition, to date the only factor clearly related to autosomal aneuploidy is advanced maternal age. In fact, a relationship between maternal age and down's syndrome was well established 25 year before the chromosomal basis for the condition was discovered. ^[4]The incidence of trisomy 21 correlates strongly with increasing age ,that is young mothers have allow probability of having trisomy 21 children.

Diagnosis of down syndrome

The diagnosis of down's syndrome is ordinarily made at the time of birth by observing the flaccid state and characteristic physical appearance of the infant. The most clinical findings of D.S as seen: ^[4]

Mental retardation	Folded ear
Protruding tongue	Short neck
Abnormal teeth	Short, broad hand
Epicanthic folds of the eyes	Short fifth finger
Congenital heart disease	Incurved fifth finger
Brushfield spots	Open mouth

Dental

Dental anomalies are very common, both in the primary and permanent teeth, and in the patients with D.S, dental anomalies occur with an incidence five times greater than in the normal population. ^[5]

In the primary dentition, the most commonly absent teeth are lateral incisors, while in the permanent dentition, third molars, second premolars and lateral incisor in this sequence are the most frequently missing teeth. ^[6]

According to ^[7] the most common dental associated with DS are variations in tooth number and morphology.

Tooth eruption may be delayed, may occur in an unusual order and can be 2 to 3 years behind a child's normal eruption pattern, over retained primary teeth are also common.

There is a high incidence of impacted teeth and hypodontia is a frequent finding, peglaterals, shovel-shaped incisors and taurodontic teeth *are often observed*.

Enamel defect among down's syndrome

Enamel as other structure in the body can be affected by the developmental anomalies which indicate there is metabolic stress during growth and development. ^[8]

Enamel formation can be divided into two stages:

Deposition of matrix and maturation, if the matrix formation is affected enamel hypoplasia will ensure, if maturation is lacking hypocalcification of enamel results in which a deficiency in the mineral content of enamel is found.

Multiple hypoplasia developed if enamel formation is interrupted on more than one occasion.

Hypoplasia affected teeth in human being in varying degree of intensity ranging from an almost scopic rough surface to deep pits which may penetrate to dentin, wavy surface of the enamel or deep funnel like depression of missing enamel.

Opacity is qualitative defect of enamel visually as an oval in form, abnormal translucency of enamel represented by white or discolored the enamel surface is smooth and the thickness of it is normal.

Many etiological agents have been reported to cause developmental defects of enamel, those which are causing localized defects are infection, trauma while the systemic factors are nutritional deficiency, neurological disturbances, endocrinopathies, intoxication, radiation. ^[9]

Development enamel defects in primary teeth have been found at least twice as frequently in children with mental retardation as in control children. Concerning the enamel defect among D.S, almost 50% of persons with D.S. exhibit three or more dental anomalies. ^[10]

Elbow crease in down's syndrome patients

We can noticed one or two elbow crease at distance (5 cm) from the elbow in one arm or the two arms in some people, so elbow line define as a white line appearance clearly in the skin and it was form transverse (cross) line.

The presence or absence forms of this crease is not occurred randomly, but controlled by a certain gene component, probably more than one pair of genes. These genes may be interacted in order to produce the expressed form of this crease. ^[11]

Material & Methods

The patients subjected to this investigation were 60 with down syndrome, their age was (8-10) years, 44 patients was male and 16 were female, information were collected from histories, reviewed records, concerning medical condition and dental examination with diagnosis by dental specialists.

This study conducted at Al-Rajaa Institutions for mentally retarded in Baghdad city (Al-Karrada), from October 2010 to April 2011, and the control group were chosen randomly in the same geographical area.

2.1. Enamel anomalies : The diagnosis was by dental specialists worked in Al-Rajaa Institutions for mentally retarded therefore, the codes and criteria off W.H.O. (1997) were followed. ^[12]

2.2. Elbow crease : Whole study elbow crease in both sample male and female patients with DS. And compared with control group, while formed for this purpose application to be sure of presence or absence elbow crease in the study sample appendix (1). Might full to seek the help of method ^[11] to diagnosis elbow crease and demonstrated Figure (1-1)



Figure (1-1) position of elbow cease in left hand

Result

Table (3-1) demonstrates numbers and percentages for presence and absence of elbow crease in two hands among male patients and control group, results showed that the percentage of presence of elbow crease was higher in the control group than that seen in the patients group, difference were statistically not significant between two samples $X^2 = 0.569$, $P = 0.903$. $P > 0.05$ while results revealed that the difference were statistically significant between female patients with D.S. and control group for presence and absence of elbow crease $X^2 = 4.824$, $P = 0.046$, $P < 0.05$, and the percentage of absence of elbow crease in left hand in female patients were higher compared with control group, table (3-2).

Data analysis considered the distribution of age patients with DS and association of dental anomalies compared with control group. Table (3-3) shows that males patients with enamel anomalies teeth in age (10) demonstrated higher percentage compared to males in the control group, differences were statistically highly significant between patients with DS concerning enamel anomalies teeth and control group, $X^2 = 4.736$, $P = 0.023$, $P < 0.05$. however, the result showed statistically significant in age (10) years between males and females compared to control group, $X^2 = 2.202$, $P = 0.049$, $P < 0.05$, while the result showed statistically no significant in age (8) years patients with DS compared to control group.

Table (3-4) shows the distribution of patients concerning enamel anomalies primary and permanent teeth and control groups. Demarcated opacities and diffuse opacities were the most

A study of elbow crease and some dental anomalies in patients with downs' syndromeRasha Abbas Aziz

distributed type in permanent teeth among patients with DS compared to control group, therefore the percentage of these type were higher among patients with DS compared to control group. Hypoplasia was recorded in primary teeth of the patients with DS.

The results demonstrate in table (3-5) patients with DS concerning elbow crease and enamel anomalies, total males recorded a higher number of teeth with enamel anomalies among ages (8), (10) years compared with females patients at both age group statistical higher significant differences were noticed in primary and permanent teeth with enamel anomalies for males and females compared with absence of elbow crease, $X^2 = 9.369$, $P = 0.025$ $P < 0.05$ while results recorded not significant differences between presence of elbow crease and teeth with enamel anomalies in patients with DS, therefore, both right and left hand shows statistical significant differences among absence of elbow crease in patients with DS. Compared with presence of elbow crease. X^2 of RH= 2.492, $P < 0.05$ and X^2 of LH= 6.875, $P < 0.05$.

Table (3-1)

Presence and absence of elbow crease among male patients with DS. And control group.

	Sample of male patient (n= 44)				Control group (n= 40)			
Hand	Presence		Absence		Presence		Absence	
	Number	%	Number	%	Number	%	Number	%
Right	21	48	23	45	29	72	11	28
Left	22	50	22	47	26	65	14	35

$X^2 = 0.569$, $P = 0.903$, $P > 0.05$

Table (3-2)

Presence and absence of elbow crease among female patients with DS and control group.

	Female patients (n= 16)				Control group (n= 20)			
Hand	Presence		Absence		Presence		Absence	
	Number	%	Number	%	Number	%	Number	%
Right	4	25	12	75	14	70	6	30
Left	2	12	14	88	14	70	6	30

$X^2 = 4.824$, $P = 0.046$, $P < 0.05$

A study of elbow crease and some dental anomalies in patients with downs' syndromeRasha Abbas Aziz

Table (3-3)

Distribution of patients with DS .concerning enamel Anomalies and age compared to control group.

Age (years)	Patients with DS	D.S. with enamel anomalies *			Control group		
		Total No.	No.	%	Total No.	No.	%
8	Male	17	3	18	20	4	20
	Female	7	1	14	10	1	10
10**	Male	27	9	33	20	1	5
	Female	9	1	11	10	1	10
Total		60	14	23	60	7	12

* $X^2 = 4.736$, $P < 0.05$

** $X^2 = 2.202$, $P > 0.05$

Table (3-4)

Number of primary and permanent teeth with enamel anomatics types among patient with D.S. and control groups.

Type of enamel anomalies	Patients with DS (N= 60)				Control group (N= 60)			
	Primary teeth		Permanent teeth		Primary teeth		Permanent teeth	
	No.	%	No.	%	No.	%	No.	%
Demarcated opacities	3	5	6	10	2	3	2	3
Diffuse opacities	0	0	3	5	1	2	2	3
Hypoplasia	2	3	0	0	0	0	0	0

Table (3-5)

Patient with D.S. concerning elbow crease and teeth with enamel anomalies.

Patients with DS	Presence of elbow crease		Absence of elbow crease		Teeth with enamel anomalies	
	RH	LH	**RH	***LH	Years * 8	Years * 10
Male	21	22	23	22	3	9
Female	4	2	12	14	1	1

* $X^2 = 9.369$, ** $X^2 = 2.492$, $P < 0.05$

*** $X^2 = 6.875$, $P < 0.05$

Discussion

The presence or absence form of this crease is not occurred randomly, but controlled by a certain genetic component, probably more than one pair of genes.

In our study the results, showed that the percentage of presence elbow crease was higher (72%), (65%), in the control group compared with patients male groups, similar result had been observed in other study (Al-Najar, 2002).^[13]

In our study, results recorded that the difference were statistically significant between female patients with DS and control group for presence and absence elbow crease, these result were agreement with (Al-Najar, 2002) ^[13] which showed a significant decrease ($P < 0.001$) between female patients in the elbow crease variable as compared with the control.

In our study showed that enamel anomalies in both primary and permanent teeth were higher in down syndrome.

Compared to the control group, this result was agreement with studies (Regezi et al, 2003 ^[10]; Radhi, 2009^[14]), this may be attributed to genetic disorders. Regulation of tooth size and shape has long been known to be polygenic, and their absence or duplication grossly affected tooth size and enamel thickness. The X chromosome has a role in the determination of tooth shape and enamel apposition and the Y chromosome has been related to larger tooth size in males and especially in the canine tooth. ^[15]

A number of studies have shown that the later developing teeth in DS are the most severely affected, in keeping with the general deceleration in growth and development. ^[15]

In the present study the relation between elbow crease and enamel anomalies in primary and permanent teeth in patients with DS were noticed a higher significant correlation, this may be related to their chromosomal abnormalities.

All the Iraqi study has been conducted among down's syndrome to allow comparison the results of current study with.

Conclusion

1. The percentage of absence elbow crease in left hand were higher (88%) in female patients with DS.
2. Patients with DS concerning enamel anomalies shows statistically highly significant.

A study of elbow crease and some dental anomalies in patients with downs' syndromeRasha Abbas Aziz

3. Down syndrome in age (10) years recorded highly percentage of concerning enamel anomalies close to (33%) in primary & permanent teeth.
4. Absence of elbow crease was predict concerning enamel anomalies teeth in patient with Ds.
5. A study of dermatoglyphics were important for diagnosis the genetic diseases.

References

1. Ficher-Brandies, H; Schmid, R.G. and Fischer-Brandies, E. (1986). Cranio-facial development in patients with down's syndrome from birth to 14 years of age. European, J. Orthod.; 8: 35-42.
2. Behrman, R.E. and Kliegman, R.M. (2002). Nelson essentials of pediatrics. 4th ed., W.B. Saunders, U.S.A.
3. Skrinjaric, T.; Glavina, D. and Jukic, J. (2004). Palatal and dental arch morphology in down syndrome. Coll. Anthropol.; 28: 841-847.
4. Cummings, M.R. (2003). Human heredity, principles and issus. 6th ed., Thomson, Canada. PP :155-157.
5. Ingalls, T.H. and Butler, R.L. (1953). Mongolism, Implication of dental anomalies. N. Engl. J. Med.; 19: 511-512.
6. Seagriff-Curtin, P.; Pugliese, S. and Romer, M. (2006). Dental considerations for individuals with down syndrome. NY. State Dent. J.; 72: 33-35.
7. Murray, J.J.; Nunn, J.H. and Steele, J.G. (2003). The prevention of oral disease. 4th ed. Oxford University Press, Italy.
8. Guatelli-Steinberg, D. (2000). Linear enamel hypoplasia in gibbons (HylobaterCarpeteri). Am. J. Phys. Anthropol., 112: 395-410.
9. Cameron, A. and Widmer, A. (1998). Hand book of pediatric dentistry. Mosby, Spain.
10. Regezi, J.; Sciubba, J. and Jordan, R. (2003). Oral pathology. 4th ed. Saunders, U.S.A.: 358-363.
11. Abdullah, N.F. and Mohaissin, A.H. (1998). Inheritance of elbow crease in iraqi families. J. Ibn Al-Haitham, Pure & App. Sci., 9(2): 19-28. (In Arabic).
12. WHO. (1997): oral health surgery basic methods 4th ed. World Health Organization. Geneva, Switzerland.
13. Al-Najar, N.A.A. (2002). A study of dermatoglyphics traits and sex chromatin in down's syndrome children and their parents in the population of northern Iraq. PH. Thesis, College of Education/ Ibn-Al-Haitham, University of Baghdad. (In Arabic).
14. Rahi, N.J.M. (2009). Oral health status in relation to nutritional analysis and salivary constituents among a group of children with downs syndrome, in comparison to normal children. Ph. Thesis, College of Dentistry, University of Baghdad.
15. Zilberman, U.; Patricia, S.; Kupietzky, A. and Mass, E. (2004). The effect of hereditary disorders on tooth components: adiographic morpho metric study of two syndromes. J. Arch. Orl. Bio., 49: 621-629.

Appendix (1) Patients information

استمارة المعلومات للمريض

الاسم:

مكان وتاريخ الولادة:

العنوان الحالي:

الجنس:

عدد الاخوة والاخوات:

تسلسله بين اخوانه:

علاقة الاب بالام:

الامراض في العائلة:

هل في العائلة افراد معوقين:

هل للمريض توائم:

وجود خط الساعد

اليمنى

اليمنى

Enamel defect

Normal

Demarcated opacities

Diffuse opacities

Hypoplasia

**دراسة خط الساعد وبعض انواع عيوب الميناء في الاسنان اللبنية
والدائمة في مرضى متلازمة داون**

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ماجستير علوم الحياة

فرع العلوم الاساسية

كلية طب الاسنان / جامعة بغداد

الخلاصة

متلازمة داون مرض وراثي ناتج من زيادة عدد الكروموسومات المتمثلة بثلاثية الكروموسوم (21). تمت دراسة خط الساعد المتصاحب مع وجود عيوب الميناء في 44 ذكر و 16 أنثى مصابين بمتلازمة داون في معهد الرجاء للعوق العملي في بغداد وتتراوح اعمارهم بين (8-10) سنة ومقارنتهم بافراد العينة القياسية في 40 ذكر و 20 أنثى .

وان هدف هذه الدراسة هو معرفة تكرار وجود او غياب خط الساعد بالإضافة الى معرفة الجانب الوراثي لوجود او غياب خط الساعد وعلاقته بعيوب ميناء الاسنان اللبنية والدائمة في مرضى متلازمة داون .

اوضحت النتائج وجود فروق معنوية مرتفعة لمرضى متلازمة داون المصابين بعيوب ميناء الاسنان مقارنة بالعينة القياسية، كذلك بينت النتائج ان كلا العينتين المرضية المدروسة للذكور والاناث وجود فروق معنوية عالية بالنسبة لغياب خط الساعد وعلاقته بعيوب ميناء الاسنان اللبنية والدائمة .

استنتجت من الدراسة الحالية ان غياب خط الساعد يعتبر مؤشر لوجود عيوب ميناء الاسنان اللبنية والدائمة في مرضى متلازمة داون .