# Dental Observations of Hyper IgE Disorder

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*Recognition of "Jobs Syndrome" disorder may lead to better care. One of the dental manifestations reported is a high arched palate. Measurements of the maxillary arch were made to compare affected Job's patients, unaffected family members, and controls. Measurements of the palate found little difference between the three groups. DMFS, periodontal disease, and oral hygiene were evaluated with little difference between groups.*

***Key words:*** *Jobs syndrome, hyper IgE disorder, maxillary arch*
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## INTRODUCTION

In this era of sophisticated medicine, it behooves all clinicians to be aware of conditions that may affect treatment and well being in order to provide the best care for patients. One of the recently identified conditions, particularly in children with dental implications, is Job's syndrome. Job's syndrome was the name given to the condition of recurring skin abscesses first described by Davis in 1966.[1] The syndrome is a rare immunodeficiency familial disorder. Buckley subsequently related the condition to elevated levels of IgE, which resulted in the additional identification of the condition as Buckley's syndrome.[2] This condition is now recognized as a multisystem disorder with manifestations of abnormalities in the skeleton, head and neck features, as well as dental and immunological deficiencies.[3] Recurrent pulmonary infections in addition to the skin infections are common.[4] Reports of facial and dental findings are frequent.[5,6,7,8,9,10] Dental abnormalities are variable and include retention of primary teeth which sometimes results in double rows of teeth upon the eruption of the permanent teeth, and high arched palates.[11] Additonally, Charon and others reported a higher incidence of gingivitis, thrush, and plaque in a group of patients with Job's syndrome.[12,13]

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It has recently been reported that there are divisions or various types of immunoglobulin E entities. Remmer, *et al*., report that individuals with the autosomal dominant form of hyperimmunoglobulin E syndrome have skeletal and dental abnormalities, whereas, the recessive form does not.[14]

O'Connell and others stress that oral health maintenance and prompt treatment of dental infections are extremely important in patients with Hyper IgE syndrome because significant morbidity can result from infections of odontogenic origin.[15,16] O'Connell reported 44% of individuals with the syndrome had elevated palates,[17] while Grimbacher reported that palates were higher in 71% of the individuals in one paper and 65% in another paper.[18] While retention of primary teeth is obvious, the appearance of a high palate is more subjective. We were concerned about the possible misinterpretation of high palates, which were identified through visual observation only. This paper appears to be the first report involving actual measurements of palate height and of the incidence of candida, caries, and oral hygiene.

## METHOD

Dental examinations were completed on 39 individuals as part of a Hyper IgE study at the University of Utah Medical Center on confirmed hyper-IgE patients with manifestations and their families. This study included eight individuals affected with Hyper IgE and 31 members of affected families. Affected individuals had IgE levels of < 2000u, chronic skin eruptions beginning before twelve months of age, and recurrent sinopulmonary infections. The study complied with and had the approval of the Institutional Review Board. The dental examination recorded decayed, missing, and filled surfaces (DMFS), gingivitis evaluation, and oral hygiene evaluations. Alginate impressions of the maxillary arch were made on all members of the Job's syndrome families. A decision to confirm the observation of higher palates on Job's syndrome individuals was made and done by measurements of the dental models.

In order to determine the height of the palatal vault, it was

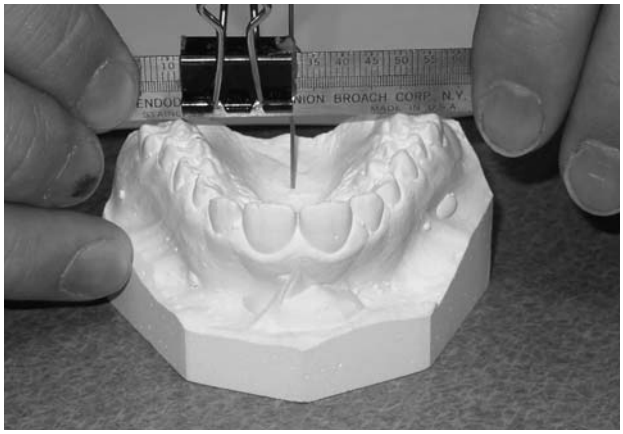

**Figure 1.** Photograph of the device to measure the height of palates. The binder clip holds a tube through which is inserted a piece of orthodontic wire. The wire is measured from the level of the ruler to the palate.

decided to make measurements of the width of the arch between the lingual cusps in two locations. The two locations were the lingual cusps of the first primary molars or the first premolar and the mesial-lingual cusp of the first permanent molars. The elevation of the palate was measured using a binder clip to which a piece of orthodontic tube was attached (Figure 1). The binder clip was placed on a millimeter ruler and a slightly bent piece of orthodontic wire was inserted through the tube. This allowed easy movement of the wire in the tube without being displaced. The ruler was placed on the cusps of the teeth previously measured to record the width of the arch. The height of the palate was determined by measuring the length of wire from the palate to the base of the ruler with a second ruler.

It was assumed that a larger individual would have a higher palatal vault than a smaller individual. To attempt to minimize the effect of an individual's size, a ratio of the width divided by the height of the palate was calculated. A larger the ratio would indicate a lower palate. Taking measurements at two locations provided a better representation of the entire palatal vault. For control, similar measurements were made from 28 randomly selected casts made for subjects age 20 years and older from another general population study. Control measurements for patients from age 10 to 19 years of age were made from orthodontic casts selected at random from a private orthodontic practice. The cases from the orthodontic practice did not have DFMS, oral hygiene, or gingivitis data. Each of the casts used in the study were measured by each of the three authors with an average taken for each measurement to be included in the results.

DMFS index and the gingival index were reported by a method established by the National Institute of Dental Research.[19] Each surface of the tooth that was missing, filled, or decayed was listed. The gingival index was reported by the degree of inflammation present. A Class "0" indicated no gingival inflammation. Class "1"evidenced mild or moderate inflammation without bleeding on less than half of the teeth, and Class "2" mild to moderate inflammation without bleeding on more than half of the teeth. Class "3" indicates inflammation with bleeding when the gingival is pressed with the mouth mirror, while Class "4" indicates an obvious loss of attachment. The plaque index was determined by observing the condition of the coronal teeth. Plaque was classified between 0 and 4 with 0 being an absence of plaque and 4 being heavy accumulation of plaque on all of the teeth. Scores of 1 through 3 indicated an increasing accumulation of plaque. This was a modification of Green and Vermillion.[20]

## RESULTS

Measurements were made for seven individuals, 10 to 19 years of age, who were affected by Job's syndrome. These measurements were compared with individuals of the same age range in Job's syndrome families who were not affected. Then, these measurements were compared to a control group of similar ages. (Tables 1, 2, and 3)

The ratios for the premolar area were 2.11 for the affected individuals, 2.08 for unaffected family members, and 2.13 for the control group. For the molar region, the ratio was 2.24 for affected individuals, 2.13 for unaffected family members and 2.24 for the control group. The measurement of the palate found little difference between affected Job's individuals and unaffected family members and the controls. There are no marked differences between the groups. The DMFS for the affected individuals were greater than for the unaffected individuals. Gingival index findings are similar for both groups and plaque scores are slightly higher for the unaffected individuals.

Only one affected individual over 20 years of age (Table 1) was seen. This individual was compared to unaffected family members (Table 2) and control individuals (Table 3). The ratio for the one affected adult premolar area was 1.88,

**Table 1.** Affected Job's Family Members

| Age | N= | Premolar Ratio | | Molar Ratio | |
|---|---|---|---|---|---|
| 10-14 years of age | 7 | 2.11<br>σ= 0.354 | | 2.24<br>σ= 0.318 | |
| | Female 3<br>Male 4 | Average age 13 | Average DMFS 15<br>Class(Number)<br>0 (4)<br>1 (3)<br>2 (0)<br>3 (0) | Gingivitis Index | Plaque<br>Class(Number)<br>0 (2)<br>1 (2)<br>2 (1)<br>3 (2) |
| Candida neg. | | | | | |
| **20+ years of age** | **28** | 1.88 | | 2.64 | |
| | Female 1 | Age 33 | DMFS 35 | Gingivitis Index<br>Class(Number)<br>0 (1) | Plaque Index<br>Class(Number)<br>0 (1) |
| Candida pos. | | | | | |

**Table 2.** Unaffected Job's Family Members

| Age | N= | Premolar Ratio | | Molar Ratio | |
|---|---|---|---|---|---|
| 10-19 years of age | 7 | 1.89<br>σ = 0.250 | | 2.04<br>σ = 0.295 | |
| | Female 5<br>Male 2 | Average age 14.2 | Average DMFS 4.6 | Gingivitis Index<br>Class(Number)<br>0 (3)<br>1 (2)<br>2 (2)<br>3 (0) | Plaque Index<br>Class(Number)<br>0 (2)<br>1 (2)<br>2 (0)<br>3 (3) |
| Candida neg. | | | | | |
| 20+ years of age | 24 | 1.89<br>σ = 02.250 | | 2.04<br>σ = 0295 | |
| | Female 12<br>Male 12 | Average age 42 | Average DMFS 41 | Gingivitis Index<br>Class(number)<br>0 (12)<br>1 (6)<br>2 (5)<br>3 (0) | Plaque Index<br>Class(Number)<br>0 (11)<br>1 (6)<br>2 (6)<br>3 (1) |
| Candida neg. | | | | | |

**Table 3.** Control

| Age | N= | Premolar Ratio | | Molar Ratio | |
|---|---|---|---|---|---|
| 10-19 years of age | 15 | 2.13<br>σ = 0.670 | | 2.24<br>σ = 0.335 | |
| Candida n/a | | | | | |
| 20+ years of age | 28 | 1.79<br>σ = 0.265 | | 1.87<br>σ = 02.66 | |
| | Female 15<br>Male 13 | Average age 34 | Average DMFS 28 | Gingivitis Index<br>Class(Number)<br>0 (7)<br>1 (16)<br>2 (5)<br>3 (0) | Plaque Index<br>Class(Number)<br>n/a |

for the unaffected family members 1.89, and for the control individuals of 1.79. The molar ratio was 2.64 for the affected individual, 2.04 for unaffected family members, and 1.87 for the control individuals. The difference in ratio from the premolar to the molar area may indicate a different slope for the affected individual but not necessarily a higher palatal vault for all Job's syndrome individuals. The sample of one does not provide reliability for a definite conclusion. Because of the small sample size, no statistical analysis was attempted; however, there seems to be little difference between the three groups of individuals.

The one affected individual had fewer DMFS than the unaffected group average, but more DMFS than the control. The unaffected individuals had a greater number of DMFS than the control group, but the unaffected group had an average age greater than the control group. The differences in DMFS may be related to the age differences. The affected individual had no gingivitis while there were varying degrees of gingivitis in the unaffected and control groups. The comparison of the plaque index indicates better hygiene for the affected individual than for the unaffected group. No plaque index was available for the control group because there was no record of it for the orthodontic models. No Candida infections were observed for any of the study individuals except the affected individual over twenty. The incidence of candida infection was less than other reports reviewed.

## DISCUSSION

Both Grimbacher *et al*[21] and O'Connell[22] describe one of the oral manifestations of hyper IgE syndrome as a high-arched or elevated palate. This conclusion was derived from observation only and not from objective measurements.[10] Orthodontists are concerned with the width of the arch, but are not generally concerned with the height of the palate. Previously, little information was available on measurements or comparisons of palatal heights. The affected individuals, ages 10-19, had essentially the same width to height ratio as the unaffected Job's family, 2.11 and 2.08, respectfully. The molar measurements were 2.24 for the affected family verses 2.13 for the non-affected family indicating, on average, a lower palate for the affected.

For the affected individuals age 20 and over, the premolar measurement was essentially the same as for unaffected family members, 1.88 verses 1.89, while the average for the control was 1.79. The molar measurement for the affected individual was 2.04 while the molar average measurement for the control was 1.87. For the only affected individual, measurements at the premolar and molar sites indicated a lower palate than unaffected individuals.

With hyper IgE individuals, it might be expected to see an increased risk for infectious disease, such as dental caries and periodontal disease, represented by increases in DMFS, gingival index, and plaque index. In this report there was a larger number of DMFS for affected individuals in the 10 to 19 year age group as compared to matched unaffected family members. There were, however, similar gingival and plaque indexes for both the affected and unaffected family members between the ages of 10 to 19. The control group did not have reported DMFS, gingival, or plaque indexes. The lone affected individual in the 20+ age group had a low DMFS, gingival, and plaque index. The unaffected family members had a lower DMFS than the control, but this may be attributed to the younger average age and to shorter exposure to the oral environment.

## CONCLUSION

The objective measurements of the palate conducted in this study indicate that affected Job's syndrome individuals'

palates are not markedly different from their family members and controls. Except for delayed exfoliation of primary teeth, this study indicated Job's syndrome individuals essentially present with normal dentitions.

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