

Prevalence of Cleft Uvula in British Columbia

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The incidence of infants born with a cleft lip, cleft lip-cleft palate or a cleft palate in the United States is approximately one in every 750 births.¹¹ Clefts contribute approximately 13 per cent of all reported birth anomalies and are the second most common congenital malformation.⁵ To aid in understanding more about clefts many studies have been undertaken including a number of epidemiological studies. These epidemiological studies have shown the incidence and prevalence of clefts vary according to sex, race and cleft type. Other variables such as birth weight, birth rank, season at the time of birth, associated malformations and length of gestation have also been examined as to their relationship to clefts.¹¹

In addition to the numerous studies showing sex and type differences, several studies have also shown variability in the incidence of clefts among different races within the same community.^{11,12} These studies showed that different racial groups exhibit different incidence rates. In general, the Mongoloids exhibit the highest incidence, the Negroids the lowest and the Caucasians intermediate.

The cleft or bifid uvula has in recent years been considered a microform of a cleft palate. Meskin *et al.*¹⁰ observed an increased frequency of cleft uvula in families of cleft probands as compared with the frequency of cleft uvula found in normal control families. They considered these data sufficient to ac-

cept the thesis that the cleft uvula is a microform of cleft palate.

Considering the thesis that the cleft uvula is a microform of cleft palate and knowing that there are different prevalence and incidence rates for palatal clefts in different racial groups, it would seem logical to also have different prevalence rates for cleft uvula in different racial groups.

METHODS AND MATERIALS

In order to test the hypothesis that there is a racial difference in the prevalence of cleft uvula, 967 children ranging in age from 13 to 17 were examined for the presence or absence of cleft uvula. Uvulae were considered to be cleft or bifid if on visual examination a cleft was observed or, as in a number of cases, after lateral displacement of the uvula with a tongue blade, a cleft was noted. The children who were examined for cleft uvula were divided into four different groups. The first group was a sample of 432 Caucasian school children from schools in Cranbrook, Kimberly, Creston, Trail, Nelson and Castlegar, British Columbia. The sampling procedure, which was under the direction of the Divisions of Vital Statistics and Preventive Dentistry of the British Columbia Provincial Government, has been previously described.² In the second group examined there were 147 children of reportedly Italian ancestry residing in Trail, British Columbia. In this group, 93 children had all eight great grandparents of Italian ancestry. In the third group examined there were 191 children of Chinese ancestry residing in Vancouver, British Columbia. Of this number, 142 had all eight great grand-

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TABLE I
Prevalence of Cleft Uvula — British Columbia

	Male		Female		Total	
	No.	No. with Cleft Uvula	No.	No. with Cleft Uvula	No.	No. with Cleft Uvula
Group I (Caucasian)	224	10	208	4	432	14
Group II (Italian-Caucasian)	74	2	73	6	147	8
Group III (Chinese-Mongoloid)	82	7	109	6	191	13
Group IV (United Kingdom-Caucasian)	81	5	116	4	197	9

parents of Chinese ancestry. In the fourth group examined there were 197 Caucasian children of primarily United Kingdom ancestry residing in Vancouver. The ancestry was determined by a personal interview of the parents by a public health nurse.

RESULTS

The prevalence of cleft uvula for the four different groups is shown in Table I. In the first group (Caucasians from Eastern British Columbia) there were 10 cleft uvulae and 214 normal uvulae observed in the males and 4 cleft uvulae and 204 normal uvulae in the females. In the second group (Italians in Trail) there were 2 of 74 males and 6 of 73 females with cleft uvulae. In the third group (Chinese in Vancouver) 7 of 82 males and 6 of 109 females had cleft uvulae. In the fourth group (Caucasians of primarily United Kingdom ancestry in Vancouver) 5 of 81 males and 4 of 116 females exhibited cleft uvulae. There were no significant age, sex, or racial differences within any one group. In the Eastern British Columbia group, the Vancouver Chinese group and the Vancouver Caucasian group, the prevalence rate

was less in the females than in the males. The reverse was observed in the Italians.

The prevalence of cleft uvula of the Mongoloid population was compared with all of the Caucasians (Table II). There was a significant difference between the two groups with the greater prevalence in the Mongoloid group. There was also a significant difference observed between the Chinese and the first group (Eastern British Columbia Caucasians).

DISCUSSION

The prevalence of cleft uvula has been previously reported by several investigators. Berans observed 3000 individuals and reported a frequency of 1.82 per cent.⁸ MacIntosh *et al.*⁷ observed 6053 infants for a variety of congenital anomalies including cleft uvula. They observed only eleven cleft uvulae. However, they were looking for a different degree or severity of clefting. Meskin *et al.*⁸ observed a frequency of 1.44 per cent in a total of 9701 individuals at the University of Minnesota. A prevalence rate of approximately 10 per cent was observed in 670 Indian children at the Red Lake Reser-

TABLE II
Prevalence of Cleft Uvula — British Columbia

	Number examined	Cleft Uvula	Per cent
Group II (Mongoloid)	191	13	6.81
Groups I, II, & IV (Caucasians)	776	31	3.99

vation in northern Minnesota by Cervenka and Shapiro.¹⁴ A prevalence rate of 11 per cent was observed by Jaffe and DeBlanc in a Navajo Indian population in New Mexico.⁴ In the same school several years later a prevalence rate approaching 20 per cent was observed by Meskin and Shapiro.¹⁴ The prevalence rate of cleft uvula has been observed by Richardson to be 0.27 per cent in a Negroid population.¹³

The prevalence rate calculated was greater in this study for Caucasians than that observed for other Caucasians studied in North America with the exception of a study in Oregon by Blakeley.¹ Blakeley observed prevalence rates of greater than 5 per cent. The percentage observed in this study is intermediate between the percentages observed by Meskin and Blakeley.

This discrepancy in prevalence rates may be explained in several ways: 1) the populations (i.e., racial and geographic variability) studied had a different gene(s) frequency for cleft palate. However, this is assuming that cleft uvula is a microform of a cleft palate. 2) Penetrance within the different populations might vary. 3) Cleft uvula relationship to cleft palate might be related as in the first example or coincidental. In other words there may be two separate genetic mechanisms for cleft uvula, one being a microform of cleft palate and the other a gene for only a cleft uvula or submucous cleft palate. Lowry⁶ recently reported on one family of Indians in British Columbia which showed a single X-linked gene for submucous cleft palate. 4) The methodology of the examinations could vary sufficiently among the several investigators so that the prevalence rates differ. This is particularly evident when comparing the two prevalence rates observed by two different investigators at Fort Wingate, New Mexico on Navajo Indians.^{4,14}

Of what value are some of the relationships that we are trying to establish? What difference does it make if a cleft uvula is associated with a cleft palate? Probably the most important reason is that this information may be used in genetic counseling. If the cleft uvula is a microform of cleft palate and is predictable both racially and geographically, this will be extremely helpful for providing more information to families desiring genetic counseling. There is also some evidence of its value in planning tonsil and adenoid removal to prevent problems with velarpharyngeal incompetency and speech defects. The prevalence of cleft uvula could also be a sensitive parameter to aid in determining secular trends in the prevalence or incidence of cleft palate if cleft uvula is a microform of cleft palate.

CONCLUSIONS

As hypothesized, there were significant differences observed between two different major racial groups in the prevalence of cleft uvula. Significant differences were observed between the Mongoloid (Chinese) group and Caucasian groups. This was certainly expected more than differences between the different Caucasian groups. There were no age or sex differences observed in the prevalence of cleft uvula. Cleft uvula is now considered by many to be a microform of a cleft palate. The condition is believed to be autosomal dominant with variable penetrance.⁹ No doubt, this does occur. However, there are data by Coccia *et al.*³ that deal with cleft lip, cleft lip-cleft palate, cleft palate and cleft uvula and data by Lowry⁶ that deal with x-linkage that require further study to aid in understanding the relationships between cleft uvula and cleft palate.

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