Abnormal Morphology of the Soft Palate: I. The Prevalence of Cleft Uvula

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Isolated clefts of the secondary palate, without involvement of the lip or primary palate, have been reported to occur with a frequency of once in approximately every 3,000 total births in a Caucasian population (3, 4, 5, 10, 12). Vital statistics such as this have been useful in studies of the etiology of this congenital malformation and in genetic counseling. These reports, however, have been based upon the presence of readily recognizable clefts of either or both the hard and soft palates. The reports have commonly not included the frequency of cleft uvula since this trait is easily overlooked and, if present, is of apparently little functional significance to the patient (7).

On the other hand, cleft uvula has been reported to represent the simplest manifestation of cleft palate (1, 11). Based on this hypothesis, it is apparent that information regarding the incidence of cleft uvula could potentially be highly significant in studies of cleft palate etiology and in genetic counseling. Unfortunately, however, reports of the prevalence of cleft uvula and its relation to more gross clefts of the secondary palate are sparse and conflicting. Therefore, this study was undertaken to investigate the prevalence of cleft uvula in a large population and to discover the relation of cleft uvula to the more readily apparent clefts of the secondary palate.

Methods and Materials

A total of 9,701 individuals were examined in two separate studies. The first study included 1,864 Caucasian dental clinic patients at the University of Minnesota School of Dentistry. These individuals were examined for cleft uvula between March and June of 1962. The group was composed of 810 males and 1,054 females and encompassed all ages.

In the second study, 7,837 students who were entering the University of Minnesota were examined for cleft uvula in September of 1962. This group included 4,713 males and 3,124 females.

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FIGURE 1. Upper left, normal uvula; upper right, uvula bifurcated up to one fourth of its total length; lower left, uvula bifurcated from one fourth to three fourths of its length; and lower right, uvula bifurcated from three fourths to its total length.

The uvulae of all individuals were examined in a similar manner. In every instance a pen-type flashlight and tongue blade were used for direct intraoral uvular examination. In some instances the tongue blade was used to separate what apparently was only a furrow in the uvula. In other instances it was necessary to require the subject to trill the letter r in order to ascertain the uvular morphology. Whenever a mucous coating was present the subject was requested to rinse with mouth wash until the uvula could be readily observed.

The uvulae were classified into one of four arbitrary categories on the basis of their morphology (Figure 1): Type A: normal uvula, Type B: uvula bifurcated up to one fourth of its total length, Type C: uvula bifurcated from one fourth to three fourths of its length, and Type D: uvula bifurcated from three fourths to its total length.

Results

A total of 25 of the 1,864 dental clinic patients in the first study demonstrated some degree of uvula bifurcation. Thus, the frequency of this trait in that population was about one in every 76 individuals (1.34%). When the prevalence of cleft uvula was divided according to the extent of the uvular bifurcation present, the frequency of Type B was 1.18%

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Type of Cleft		N = 810	$\begin{array}{c} Female \\ N = 1,054 \end{array}$	$ \begin{array}{c} Total \\ N = 1,864 \end{array} $
Type B	N	11	11	22
	%	1.36	1.04	1.18
Type C	N	2	0	2
	%	.25	.00	.11
Type D	N	0	1	1
	%	.00	. 09	.05
All types	N	13	12	25
	%	1.61	1.13	1.34

TABLE 1. Prevalence of cleft uvula for 1,864 dental clinic patients according to type of cleft and sex.

TABLE 2. Prevalence of cleft uvula for 7,837 university students according to type of cleft and sex.

Type of Cleft		Male N = 4,713	Female N = 3,124	$\begin{array}{c c} Total \\ N = 7,837 \end{array}$
Type B	N	57	37	94
	%	1.21	1.18	1.20
Type C	N	10	5	15
	%	.21	.16	.19
Type D	N	5	1	6
	%	.11	. 03	.08
All types	N	72	43	115
	%	1.53	1.38	1.47

while Types C and D accounted for the remaining 0.16%. Further tabulation according to sex indicated that an apparently higher prevalence of cleft uvulae were observed in the males (1.60%) than in the females (1.13%) as shown in Table 1.

In the second study involving 7,837 entering college students, a total of 115 bifurcated uvulae were found. Thus, in this group cleft uvula was present in an average of one out of every 68 individuals giving a prevalence of 1.47%. Subdividing these cleft uvulae according to the extent of uvular bifurcation showed that the prevalence of Type B clefts was 1.20% with the C and D types of bifurcation representing 0.19 and 0.08%, respectively. Analysis of the data according to sex revealed that the cleft uvulae were again slightly more common in the males (1.53%) than in the females (1.38%) as shown in Table 2.

Discussion

It has been reported that cleft uvula is a minor manifestation of severe cleft palate (1, 11). Based on this hypothesis and based on the frequency of cleft uvula found in this study, it would appear that isolated palatal clefts of all degrees of severity are considerably more common than the

usual one in 3,000 frequency noted in the literature (3, 4, 12). On the other hand, since severe cleft palate has been noted to be twice as common in females than males (4), why was no such female sex predilection for uvular clefts noted in this study?

One hypothesis, based on the work of Fogh-Anderson (4), may be formulated in answer to this question. He analyzed a group of cleft palate patients according to the subdivisions of clefts of the hard and soft palates and clefts of the soft palate alone. He reported that as the clefts became less severe, the sex affinity for the female also decreased. Similar findings have recently been reported by Knox and Braithwaite (8). Consequently, it may be suggested that if this trend continued, the sex affinity for the female could disappear completely or even become reversed. Such a phenomenon could explain the apparent slightly higher prevalence of cleft uvulae observed in the males of this study.

Unfortunately, it is difficult to verify this hypothesis or even to verify the prevalence of cleft uvula observed in this study due to a lack of comparable studies of this nature, the adoption of arbitrary classification systems, the selectivity of the populations studied, the amount of disagreement concerning the constitution of normal uvular morphology, unknown environmental effects (one case of a surgically produced cleft uvula was found in this study), and the problem of adequately visualizing uvular morphology.

The problem of adequate visualization of the uvula has been perhaps most troublesome. The presence of enlarged tonsillar tissue, commonly present in children, plus a small oral pharyngeal area make difficult a thorough examination of the uvula. Considering that examinations for congenital anomalies are commonly done on young infants, it is not surprising that few investigations of the frequency of cleft uvula have been reported. In substantiation of this premise, an investigation of the records of the Mayo Memorial Hospital in Minneapolis since 1953 to the present indicated only two listings of cleft uvula, the congenital anomaly designated as Number 755 by the International Classification of Disease (6).

Two studies have been reported previously concerning the prevalence of cleft uvula. Berans (2) divided the abnormal uvula into three classifications (fishtail, deep clefts, and complete separation). These categories would apparently correspond closely to Types B, C, and D, respectively, described in this study. He examined 3,000 individuals and reported the frequency of fishtail uvulae as 1.30%, deep cleft uvulae, 0.45%, and completely separated uvulae, 0.06%. The total sample prevalence was 1.82% which is only slightly higher than the 1.47% prevalence noted in this study. Considering the possible variation in classifications the findings of these two studies are in good agreement.

McIntosh (9) examined 6,053 young children and infants for a variety of congenital anomalies including cleft uvula. He noted 11 cleft

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uvulae among his subjects, a markedly lower frequency than that found by Berans (2) and that reported in the present study. However, the previously noted difficulties associated with uvular examinations in infants and young children very possibly could mean that the clefts noted by McIntosh were severe (Types C or D) and suggests that his findings cannot be compared to Berans' (2) study or those reported here.

Summary

The prevalence of cleft uvula has been studied in two independent populations. In 1,864 University of Minnesota dental clinic patients a 1.34% prevalence of cleft uvula was discovered. In 7,837 entering University students a 1.47% prevalence of cleft uvula was noted. Thus, the prevalence of cleft uvula in these studies combined was 1.44% in a total of 9,701 individuals. In each study males were slightly more affected than females. Based on the hypothesis that cleft uvula is a minor manifestation of a more severe cleft of the palate and considering the frequency of cleft uvula demonstrated in this study, it would appear that palatal clefts of all forms of severity are considerably more common than the prevalence commonly reported in the literature.

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References

- 1. ALLAN, F. D., Essentials of Human Embryology. New York: Oxford University Press, 1960.
- 2. BERANS, C., Anomalies of the uvula. Philadelphia med. Bull., 15, 177-179, 1893.
- 3. Book, J. A., The incidence of congenital diseases and defects in a south Swedish population. Acta Genet., 2, 289-311, 1951.
- 4. FOGH-ANDERSON, P., Inheritance of Harelip and Cleft Palate. Kobenhavn: Ejnar Munksgaard, 1943.
- 5. HARRIS, L. E., and STEINBERG, A. G., Abnormalities observed during the first six days of life in 8,716 live born infants. *Pediatrics*, 14, 314-326, 1954.
- International Classification of Diseases (1955 Revision). Geneva: World Health Organization, Vol. 1-2, 1957.
- JARVIS, J. F., and MWATHI, S. N., Uvulotomy among East African Tribes. J. Laryng. Otol., 73, 436-438, 1959.
- 8. KNOX, G., and BRAITHWAITE, F., Cleft lips and palates in Northumberland and Durham. Arch. dis. Child., 38, 66-70, 1963.
- MCINTOSH, R., MERRITT, K. K., RICHARDS, M. R., SAMUELS, M. H., and BEL-LOWS, M. T., The incidence of congenital malformations: a study of 5,964 pregnancies. *Pediatrics*, 14, 505-522, 1954.
- NEEL, J. V., A study of major congenital defects in Japanese infants. Amer. J. hum. Genet., 10, 398-445, 1958.
- 11. STARK, R. B., Embryology, pathogensis and classification of cleft lip and cleft palate. In S. Pruzansky (Ed.), p. 66-85, Congenital Anomalies of the Face and Associated Structures. Springfield, Illinois: Charles C Thomas, 1961.
- STEVENSON, S., WORCESTER, J., and RICE, R. G., Six hundred and seventy-seven congenitally malformed infants and associated gestational characteristics. *Pediatrics*, 6, 37-50, 1950.