

Cleft Palate, Cleft Lip, and Cleft Uvula in Navajo Indians: Incidence and Otorhinolaryngologic Problems

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The purpose of this study is to provide information about the high incidence of cleft lip and palate in the Navajo Indians, along with their otorhinolaryngologic problems.

A Cleft Palate Clinic was established at the PHS Indian Hospital in Gallup, New Mexico, which served as the primary referral center for cleft palate patients on the Navajo reservation. 115,000 Navajos, comprising the largest tribe of Indians in the U.S., are located on their reservation in the southwestern part of the country. They are sparsely settled in a large part of Arizona, New Mexico, and Utah. They are a poor people and their traditional life style has resulted in a high incidence of infectious diseases, widespread malnutrition, plus considerable tribal inbreeding.

Incidence of Cleft Lip and Cleft Palate in the Navajo

All hospitals on the Navajo Reservation were contacted to obtain the name of any patient born with a cleft lip and/or cleft palate between January 1, 1961, and December 31, 1967. Prior to 1961, the records were incomplete. In addition, the records from the Crippled Children's Service office, with diagnoses made only by physicians and located at the Gallup PHS Indian Hospital, were reviewed to double-check these reports and to identify additional cases during the same seven years.

In these seven years, there were approximately 28,000 Navajo births, and 70 had a cleft palate and/or lip. Of these, 15 (22%) had cleft lip, 10 (14%) had cleft palate and 45 (64%) had cleft lip and cleft palate (Table 1). Of these patients, 3 had bilateral cleft lips and bilateral cleft palates, and 2 had a unilateral cleft lip with a bilateral cleft palate. Associated anomalies were not tabulated in this study.

Table 2 relates the incidence of cleft lip and/or cleft palate in dif-

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TABLE 1. Incidence of cleft lip, cleft palate, and cleft lip and cleft palate in Navajos born between 1961 and 1967. (Information regarding live births was obtained from Frank Baty, Program Analyst, Division of Indian Health, Albuquerque, New Mexico.)

<i>year</i>	<i>live births per year*</i>	<i>cleft lip</i>	<i>cleft palate</i>	<i>cleft lip and palate</i>	<i>total</i>
1961	3,680	9	1	4	14
1962	3,887	0	1	4	5
1963	4,142	2	1	9	12
1964	4,439	0	1	9	10
1965	3,976	2	2	5	9
1966	3,900 (est.)	0	4	3	7
1967	3,900 (est.)	2	0	11	13
total	28,000	15	10	45	70

TABLE 2. Comparative incidence in ethnic groups of cleft lip with or without cleft palate and of cleft palate.

	<i>total births during study</i>	<i>cleft lip with or without cleft palate</i>	<i>cleft palate</i>
Navajo Indians (this survey).....	28,000	60 (1:467)	10 (1:2800)
American Indians*, all tribes.....	25,341	35 (1:724)	15 (1:1689)
Montana Indians*.....	7,461	18 (1:415)	9 (1:829)
Caucasian*, total of 35 studies....	204,341	245 (1:834)	68 (1:3005)
Negro*, total of 2 studies.....	96,801	26 (1:3723)	24 (1:4033)
Japanese*.....	63,796	136 (1:469)	35 (1:1823)

* Data obtained from American Indian Congenital Malformation Study (5).

ferent population groups. The incidence of all types of clefts in the Navajo is 1:400 live births, or twice the incidence of the Caucasian population, in which the incidence is 1:834.

The incidence of isolated cleft palate is low and approximates that of the Caucasian group.

Incidence of Cleft Uvula

A survey of 944 Navajo school children was conducted in September 1966 at Fort Wingate Boarding School at Fort Wingate, New Mexico, by one of the authors (B.F.J.). Clefts of the uvula were divided into 4 groups depending on the percentage of the uvula that was cleft (Figure 1). The rest of the soft palate and hard palate was normal by inspection and palpation. Specifically, no notching of the hard palate was present as in submucosal clefts. Of these children, 11% (106/944) had a cleft uvula. The distribution into severity of cleft uvula was as fol-

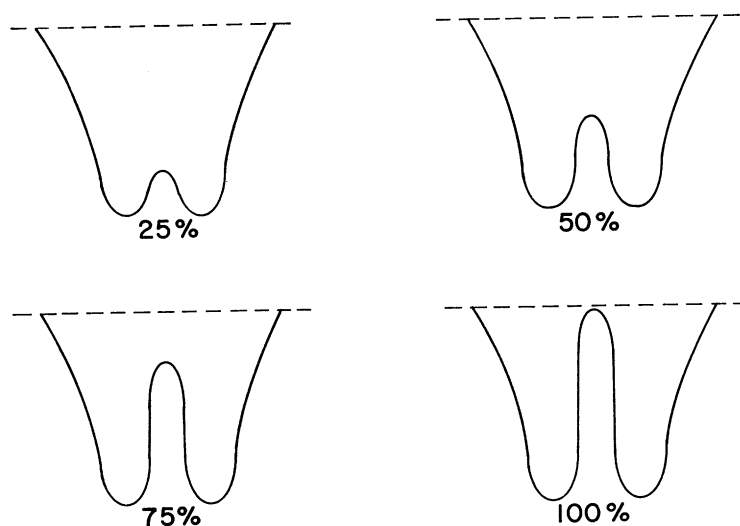


FIGURE 1. Degree of cleft in uvula.

lows: 65 had $\frac{1}{4}$ clefts; 25 had $\frac{1}{2}$; 10 had $\frac{3}{4}$, and 6 had total clefts of the uvula.

In comparison, the Caucasian population in the U.S.A. has a reported incidence of 1.47% (3), about $\frac{1}{10}$ th as common as in the Navajo.

The sex incidence revealed 13% (57/455) males and 10% (49/489) females. Meskin and associates (3) also reported a male predominance in their series with 1.53% males and 1.38% females.

Cleft Uvula Related to Ear Disease

Otoscopic examinations were performed in the same 944 Navajo children to correlate the relationship between middle ear disease and the cleft uvula. The middle ear diseases that were tabulated included chronic suppurative otitis media with a perforated tympanic membrane, a healed central perforation of the tympanic membrane, and a "dry" atelectatic middle ear (retracted tympanic membrane) or "wet" atelectatic middle ear (serous otitis media).

When a major ($\frac{3}{4}$ or total) cleft of the uvula was present, the incidence of middle ear disease was 36%. This was almost twice the rate of 20% with middle ear disease seen in the 838 Navajo children with a normal uvula.

An increased incidence of ear disease is common in patients with cleft palates, and is usually explained by abnormal musculature of both the soft palate and the eustachian tube. The cleft uvula is a minor form of the cleft palate (4). Perhaps the same mechanisms are responsible for the higher incidence of middle ear disease in patients with major clefts of the uvula as in the cleft palate patients. No previous reports have correlated cleft uvula to middle ear disease.

Cleft Palate Related to Sinusitis

A mucopurulent discharge from the nose was frequently noticed during routine examination of our cleft lip and palate patients, most of whom had had surgical repair of the defects. The possibility of sinusitis was suggested and a pilot project begun (2). Twenty-one control and 21 cleft palate patients had paranasal sinus X rays regardless of the clinical findings during anterior rhinoscopy.

In the 21 Navajo control children, maxillary sinusitis was found radiologically in 14% (3/21). In 21 Navajo cleft palate children, maxillary sinusitis was found radiologically in 62% (13/21). Of these 13 cleft palate patients, 8 had air-fluid levels in the maxillary sinus and 5 had mucosal membrane thickening. There was close correlation between mucopus in the nose and the radiologic evidence of sinusitis. Treatment consisted of oral penicillin or ampicillin for 10–21 days and neosynephrine nose drops. When the sinusitis persisted antral irrigations were performed revealing pus in 2 patients and mucus in 1 patient. Culture revealed diplococcus pneumonia in 1 patient and non-hemolytic staph in the other. Intractable cases were treated surgically by indwelling maxillary sinus tubes or naso-antral windows.

Middle ear disease was then correlated to the status of the maxillary sinuses. Middle ear infections were present in 83% (18/24) of the ears when sinusitis was present, but only 42% (6/14) of the ears when sinusitis was absent.

Cleft Palate Related to Ectopic Teeth

An ectopic tooth in the floor of the nose can act as a foreign body creating nasal discharge. One cleft palate child was referred to our clinic for a chronic unilateral nasal discharge, and a malformed loose tooth was found in the anterior floor of the nostril. The ectopic tooth was removed, and postoperatively the nasal drainage ceased.

Another patient with nasal discharge had a tooth bud covered by granulation tissue in the floor of the nose. The ectopic tooth was removed during the primary repair of the cleft lip.

Discussion

This study documents a high incidence of cleft lip, cleft palate, and cleft uvula in the Navajo Indians. Physicians caring for the Navajo recognize other congenital anomalies as also having a higher incidence in the Navajo, such as congenital heart disease, congenital dislocation of the hip, congenital skeletal anomalies and congenital aural atresia (1).

Inbreeding, infections, and malnutrition have each been indicated as factors contributing to congenital anomalies. It is probable that inbreeding occurred in the Navajo, since marriage outside the tribe is discouraged. The population has increased from 8,000 to 115,000 in only one hundred years. Infectious diseases are the most prevalent medical

problems on the Navajo reservation in all age groups. Chronic infections during pregnancy are highly probable, and may contribute to the production of congenital anomalies. Malnutrition, which is widespread, may be a factor in producing a cleft palate. Even kwashiorkor in Navajo infants is occasionally seen. The malnutrition arises from the basic inadequate diet of the traditional Navajo: mutton stew, cornbread, and coffee.

The high incidence of cleft lip and palate in the Navajo Indians is in agreement with the findings of Tretsvén, who noted a high incidence of cleft lip and palate in 11 Indian tribes in Montana (7). It is yet to be shown whether all U.S. Indians will share this trait.

It is known that otitis media is common in cleft palate children, and is thought to be related to abnormal structure and/or abnormal function of the eustachian tube. The study identifies another etiologic factor: sinusitis. Perhaps the high incidence of middle ear disease is also due to the mucopurulent discharge from the sinus bathing the eustachian tube orifice with bacteria. No previous reports have been found to show any relationship between cleft palate patients and sinusitis.

Nasal discharge is common in cleft palate children and this study stresses two etiologies which should be sought for specifically in the cleft palate patient: paranasal sinusitis, and ectopic teeth in the floor of the nostril with associated granulation tissue and infection. The persistence of chronic nasal infection could lead to otitis media as mentioned above. The role of sinusitis in producing palatal sutureline breakdown is now being studied.

A cleft uvula is considered to be a minor form of a cleft palate. Supernumerary teeth are more common in cleft palate patients, and Pöler (6) reported that supernumerary teeth are also more common in the Navajo with cleft uvula compared to the Navajo with a normal uvula. Our study reveals another similarity between the cleft uvula patient and the cleft palate patient, that is, a higher incidence of middle ear disease. Abnormal eustachian tube function is suspected in both situations.

Summary

The incidence of cleft lip and/or cleft palate is 1:400, or almost twice as common as in the Caucasian population. Heredity, chronic infections, and malnutrition in the Navajo may contribute to this high incidence. Cleft uvulas are 10 times as common in the Navajo as in the Caucasian population, and occurred in 11% of the Navajos. A major cleft of the uvula was associated with twice as much middle ear disease as was found in the normal Navajo population. A cleft palate was associated with maxillary sinusitis in 62% (13/21) of the patients. This may be an important factor in explaining the high incidence of middle ear disease found in patients with cleft palates. A cleft palate is sometimes

associated with ectopic teeth in the floor of the nostril, which may produce a chronic nasal discharge. Intranasal extraction is advised.

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