

Classification and Frequency of Cleft Lip and / or Palate

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Classifications or taxonomies of the cleft lip and palate syndrome have employed regional anatomy (1, 11), and embryologic and anatomic considerations (4, 5, 7, 12). The classification that has probably appeared most frequently in literature is the one presented by Veau (12) and later by Fogh-Andersen (3). This classification scheme includes only three categories (cleft lip, cleft lip and palate, and cleft palate) and has been criticized because there are some cleft types which cannot be properly classified using such a scheme.

A second classification scheme, presented by the American Cleft Palate Association (4), has the capability of describing all observable types of clefts in light of their embryonic origin and postnatal pathomorphology. It includes two primary cleft types: those clefts which are primarily the result of a failure in the growth and development progress of the frontal process (eminence) and the maxillary processes, *prepalate clefts*; and those clefts which are primarily the result of a failure in growth of the palatal shelves of the maxillae, *palatal clefts*. Thus prepalate clefts (premaxillary) may occur with or without the involvement of the palatal shelves of the maxillae and they may be either unilateral or bilateral. Palatal clefts involve only the palatal shelves of the maxillae, the vomer, the horizontal processes of the palatine bones, and the soft palate. In view of the number of independent but normally integrated structures involved, and the infinite chances for failure or variation in the normal processes of growth and development, the amount of heterogeneity encountered with clefts (prepalatal or palatal) is all but astronomical.

Apart from the matter of classifying, a second problem is estimating the frequency with which various cleft types occur. Fogh-Andersen (3) has hypothesized a frequency of occurrence of 1:2:1 (cleft lip:cleft lip and palate:cleft palate). Similar hypotheses have not been formulated for the ACPA classification scheme. Indeed, a statistical evaluation of the incidence of cleft types for any cleft classification, whether

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based upon embryonic or postnatal criteria, has not appeared in literature.

This research project was designed to evaluate the appropriateness of various frequencies of occurrence of cleft types categorized by two major classification schemes.

Materials and Methods

Population data for the incidence of clefts were assembled from various sources in the literature. The data used were the results of epidemiological surveys and were derived from the analyses of live birth data from Pennsylvania (5), California (9), Wisconsin (10), Hawaii (8), Denmark (3), and from two compilations of data from several states of the United States (2, 6). Data from clinical populations were rejected because of the bias which may be associated with the preselected nature of such populations. The majority of the data which were used were categorized initially according to the scheme presented by Veau (12) and Fogh-Andersen (3) (see Table 1). Subsequently, the data were regrouped according to the ACPA scheme (Table 2) with cleft types involving the lip and premaxilla and/or palate classified as prepalate and all others as palate. The chi-square statistic is used to evaluate the goodness of fit of the observed data to several frequencies of occurrence of two classification schemes: cleft lip-cleft lip and palate-cleft palate, 1:2:1; and prepalate-palate, 6:3, 7:2, 7:3, and 8:2.

TABLE 1. Observed and expected frequencies of occurrence of cleft lip, cleft lip and palate, and cleft palate as reported by several investigators. Expected frequencies, according to the ratio of 1:2:1, are in parentheses. Chi-square values which are asterisked are significant at the .01% level (df = 2).

<i>Source</i>	<i>Cleft lip</i>	<i>Cleft lip and palate</i>	<i>Cleft palate</i>	<i>Total</i>	χ^2
Pennsylvania (5).....	392 (361.5)	659 (723)	395 (361.5)	1446	11.34*
Wisconsin (10).....	157 (173)	336 (346)	199 (173)	692	5.67
California (10).....	622 (536.25)	934 (1092.50)	629 (546.25)	2185	46.03*
Denmark (3).....	138 (156.25)	360 (312.50)	127 (156.25)	625	16.72*
Hawaii (8).....	38 (32)	43 (64)	47 (32)	128	14.83*
29 U.S. states (6).....	1785 (1674.5)	2869 (3349)	2044 (1674.5)	6698	157.62*
4 U.S. states (2).....	588 (564)	1073 (1128)	595 (564)	2256	5.40

TABLE 2. Observed and expected frequencies of occurrence of prepalate and palate clefts as reported by several investigators. Expected values for each of four ratios are reported. Chi-square values which are asterisked are significant at the 1% level ($df = 1$)

<i>Source</i>		<i>Prepalate</i>	<i>Palate</i>	<i>Total</i>	χ^2
Denmark (3)	observed	498	127	625	
	expected				
	6:3	416.66	208.33		47.63*
	7:2	486.01	138.89		1.31
	7:3	437.5	187.50		27.88*
Pennsylvania (5)	8:2	500.0	125.00		.04
	observed	1051	395	1446	
	expected				
	6:3	964.02	482.01		47.63*
	7:2	1124.69	321.13		21.71*
Wisconsin (10)	7:3	1010.20	433.80		4.95
	8:2	1156.80	289.20		48.38*
	observed	493	199	692	
	expected				
	6:3	461.34	230.61		6.52
California (9)	7:2	518.23	153.74		17.09*
	7:3	434.40	207.60		.50
	8:2	545.60	138.40		36.16
	observed	1556	629	2185	
	expected				
Hawaii (8)	6:3	1456.68	728.34		20.32*
	7:2	1699.46	485.56		54.43*
	7:3	1529.50	655.50		1.53
	8:2	1748.00	437.00		296.21*
	observed	81	47	128	
29 states, U.S.A. (6)	expected				
	6:3	85.32	42.66		.66
	7:2	99.54	28.44		15.56*
	7:3	89.60	38.40		2.75
	8:2	102.40	25.60		22.36*
4 states, U.S.A. (2)	observed	1661	595	2256	
	expected				
	6:3	1755.46	501.50		49.16*
	7:2	1501.68	752.34		22.50*
	7:3	1579.20	676.80		14.12*
8:2	1804.80	451.20		57.28*	

Results and Discussion

The hypothesis for the 1:2:1 frequency of occurrence was tested using the Veau and Fogh-Andersen classification on population data from Pennsylvania, Hawaii, California, Wisconsin, Denmark, and compiled data from several states of the United States. As shown on Table 1, the resulting chi-square values for goodness of fit indicated that all but two of the distributions differed significantly from the expected incidence. (The fact that the Denmark data failed to fit Fogh-Andersen's hypothetical model was indeed surprising.) It was interesting to note that the data from Wisconsin (10) and the data reported by Donahue (2) did not deviate from the hypothesis in contrast to the other United States data.

Data for evaluating the four hypothesized cleft type ratios 6:3, 7:2, 7:3, and 8:2, using the dichotomous prepalate-palate scheme, are presented in Table 2 and are summarized in Table 3. In general, only the 7:3 ratio gives expected frequencies which are similar to the observed frequencies reported by the various investigators. The exceptions for the 7:3 ratio were the data from Denmark (3) and the data reported by Donahue (2). The other three ratios, 6:3, 7:2, and 8:2, give distributions of expected frequencies which are significantly different from all but three (one each) of the observed frequencies.

Apparently, then, the 7:3 ratio fits relatively well the observed frequencies of occurrence of the ACPA prepalate and palate classification scheme. In contrast, the 1:2:1 ratio (cleft lip:cleft lip and palate:cleft palate) fits the observed frequencies from only two of the seven investigations and cannot be justifiably adopted to represent the frequency of occurrences of cleft types.

Summary

This project was designed to evaluate the appropriateness of several ratios representing the frequency of occurrence of types of clefts. The chi-square statistic was used to evaluate the goodness of fit of ex-

TABLE 3. Summary of findings regarding the evaluation of data from seven studies, by chi-square, of the goodness of fit of four hypothesized ratios for frequency of occurrence of prepalate:palate cleft types. Asterisks represent significant differences between the observed and expected frequencies. Dashes represent such differences which are not significant.

<i>Ratio</i>	<i>Denmark</i>	<i>Penna.</i>	<i>Wisc.</i>	<i>Calif.</i>	<i>Hawaii</i>	<i>29 U.S. states</i>	<i>4 U.S. states</i>
6:3	*	*	*	*	—	*	*
7:2	—	*	*	*	*	*	*
7:3	*	—	—	—	—	—	*
8:2	—	*	*	*	*	*	*

pected frequency distribution from five ratios to the observed frequencies reported by seven investigators. Ratios which are tested were 1:2:1 (cleft lip:cleft lip and palate:cleft palate), 6:3, 7:2, 7:3, and 8:2 (prepalate:palate). Of the five, only the 7:3 ratio did not give expected frequency distribution significantly different from the observed frequency distribution.

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