Isolated Cleft Palate

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Isolated cleft palate refers to any cleft of the palate which is posterior to the anterior palatine foramen, and which does not involve the alveolar process or lip. Fogh-Andersen (*2*) has suggested that it is inherited in a relatively small number of cases and is twice as common in females as males. Snodgrasse (*11*) has indicated the need for further study of isolated cleft palate because Oldfield (*9*), in Leeds, and Curtis (*1*), in Toronto, showed that the sex frequency was equal. Later, Oldfield and Tate (*10*) and Knox and Braithwaite (*7*) showed that there was a higher proportion (3 to 2) of females with isolated cleft palate than males. Meskin, Gorlin, and Isaacson (*8*), reporting on the prevalence of cleft uvula, found a slightly higher proportion of males (3.14 to 2.51). They suggest that this may be explained on the hypothesis based on the work of Fogh-Andersen that as the isolated clefts become less severe, the sex affinity for the female also decreases.

This study, reported in greater detail elsewhere (*5*), was carried out for the purpose of obtaining more information about isolated cleft palate, with special reference to etiological association.

Material and Method

Records of 88 cases of isolated cleft palate seen at the Children's Hospital, Sheffield, were examined. In 68 cases, the parents were interviewed, and the cases were re-examined. A relatively comprehensive history was taken and, in many cases, additional relevant observations made by the surgeon were recorded. In order to compare pregnancy history, a control series of 100 cases (50 'difficult' and 50 'normal' pregnancies, equally divided for sex of the children, who were all normal) were taken at random from the files of the Jessop Hospital for women, Sheffield.

General Information. Occupation of the father, size of the families, birth rank, and month had no relation with the cleft. There was no case with any history of consanguinity.

Gestation term was calculated from the expected date of birth and actual date of birth, and divided into 'early' and 'late'. The cleft and control groups were compared. There were more 'early' cases among

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the cleft series and the difference was significant. Birth weights showed no significant differences between the cleft and control series. The average ages of the parents at marriage in the cleft series is lower than the Registrar General's figures for England and Wales. When the age of the mother at birth of the child in the cleft group (24.03) and that for the control series (26.75 years) is compared, the difference is highly significant. This partly suggests that the 'older' mothers may tend to have cleft children, but it could also be partly explained by the fact that the control series was taken from listings from a maternity hospital where possibly younger mothers were admitted for their first confinements. The cleft group showed a significantly earlier marriage compared with the England and Wales population group, but there was no significant difference in the ages of the parents at birth of the child. This suggests a longer period between age at marriage and birth of the cleft child (6.59 years for the cleft group and 3.31 years for the national group). Age of clinical detection of the cleft varied, and although the larger clefts (generally up to the anterior palatine foramen) were detected at birth, a great many of the medium-sized and smaller ones were not noticed till a week or more later. For the entire group of 68 subjects, 32% were detected at birth, 59% during the first week of life, and 9% during weeks two through eight. Generally, there is agreement with Spriestersbach (*12*) that a thorough investigation of oral structures must be done at birth.

Information regarding sex distribution is shown in Table 1. Two British studies, Oldfield (*9*) and Iyer, and the Canadian study, Curtis (*1*), show approximately equal sex distribution, while the Danish and U. S. A. studies show a very high proportion of females. Two other English studies (*7*, *10*), not shown in Table 1, give results which fall between the extremes (male:female of 2:3), indicating that there may be some geographic or ethnic variations.

TABLE 1. Sex distribution of isolated cleft palate in seven investigations.

investigator	*country*	*N*(%)		*total*
		m	*f*	
Fogh-Andersen (*2*)	Denmark	43 (33.9)	84 (66.1)	127
Oldfield (*9*)	England	81 (48.5)	86 (51.5)	167
Snodgrasse (*11*)	U.S.A.	9 (42.9)	12 (57.1)	21
Ivy (*6*)	U.S.A.	203 (39.6)	309 (60.4)	512
Curtis (*1*)	Canada	no figure given but see*		—
Spriestersbach (*12*)	U.S.A.	39 (32.5)	81 (67.5)	120
Iyer	England	44 (50.0)	44 (50.0)	88

* In reference to Fogh-Andersen's figures, Curtis states, "Some lack of agreement is seen in the sex ratios for isolated cleft palate in that the Toronto data do not show the marked increase in females."

ANATOMICAL FEATURES. No sex difference was noticed regarding judgments of the anteroposterior extent of the clefts, which does not agree with Fogh-Andersen's report (*2*). Isolated cleft palate without associated congenital deformities was more frequent (76%) than cleft palate with associated abnormalities (24%). When associated abnormalities are present, it is six times more common in males than in females. Among the anomalies shown were anomalies of the foot, hand, auricle, and eye, as well as mental deficiency and undescended testicles. Sixty-seven subjects were classified by angles classification for dental occlusion; 71% had Class I, Class II had 21%, and Class III had 8%. This distribution is markedly different from the findings of a survey on a Sheffield school sample by Gardiner (*3*), where the figures are Class I, 88.5%; Class II, 11%; and Class III, .5%. The cleft group shows a higher proportion of Class II, and a very much higher percentage of Class III. A clinical assessment of the skeletal pattern showed the following classification: normal, 63%; postnormal, 24%; and prenormal, 13%. It is suggested from these figures that there is a lack of maxillary development forwards in the isolated cleft palates.

ENVIRONMENTAL CAUSES. Regarding the parents' opinion regarding cause of cleft, the following results were obtained: do not know, 55%; inherited, 6%; and other causes (fear, stress and worry, domestic trouble, illness), 39%. Although in a retrospective study of this type, one should be wary of these 'old wives tales', one cannot ignore the apparent effect of possible 'stress' on pregnancy.

Findings from this study regarding possible environmental disturbances in the pregnancy history were compared with findings for the control series. In the control series, where half were 'difficult' and half had 'normal' deliveries, there was hardly any difference noticed in the pregnancy history. In addition, very little difference was seen between the cleft and control series. Operations and accidents appear to be significant, but, as explained above, one must view this with caution in a retrospective study. It is difficult to comment on the effect of drugs and x-ray since no comparative figures were available. The number of still births and abortions is high in the cleft palate group. Steigler (*13*) has suggested that occurrence of spontaneous abortion or still birth just before birth of the propositi tends to increase the chance of cleft palate and other structural abnormalities in the child. In the present control series, however, where there were 13 abortions and five still births, eight abortions and all five still births were immediately preceding the birth of the normal child.

GENETIC CAUSES. In 53 (78%) cases, there was no history of clefts of any type in the family. In 15 (22%) cases there was a history of some type of cleft. Further distribution of that 15 is as follows: history of only isolated cleft palate, 6 (9%); history of only harelip, 4 (6%); history of harelip and cleft palate, 3 (4.4%); and previous family history, but

siblings presented a deformity, one case with sibling having isolated cleft palate, and one case with sibling having harelip, a total of 2 (2.6%).

The 15 (22%) cases presenting some history of clefts were analyzed according to Weinberg's propositus method as done by Fogh-Andersen. Comparison was also made with the studies of Fogh-Andersen (*2*), Snodgrasse (*11*), and Fraser (*3*).

The findings are in agreement with Fogh-Andersen's, in that isolated cleft palate is inherited in a relatively fewer number of cases and, when it is inherited, it appears to be dominant. There is the possibility of genetic independence between isolated cleft palate and harelip, with or without cleft palate, as was shown by a concordant discordant analysis of the 'near' and 'distant' relatives of the propositi.

Summary

Records of 88 isolated cleft palate cases were studied and, of these, 68 cases were available for detailed interview. Occupation of father, size of family, consanguinity, birth weight and birth rank had no relationship to the cleft. Stress during pregnancy was a vague etiological factor. The gestation term of isolated cleft palate cases was slightly shorter. Although parental age did not seem significant as a factor, there was some evidence of a trend of an early marriage and a longer period between marriage and birth of the child for isolated cleft palate. Inheritance of isolated cleft palate in eight cases (11%) appeared dominant, but there was no sex influence. Equal numbers of males and females were affected, and there was also no sex difference in the severity of the defect. Associated congenital deformities were noticed in 24% cases and they were six times more frequent in males. Isolated cleft palate cases showed a very high proportion of mandibular prenormal cases and it is suggested that this is due to lack of forward development of the maxillary base. In a considerable number of cases, the cleft was not detected at birth, which suggests a lack of thorough examination of the oral structures at birth.

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