Incidence of Cleft Lip and Palate in British Columbia Indians

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It is generally recognized that there is a genetic component in the cleft lip and cleft palate malformation in man. There is good evidence that this malformation is heterogeneous, and that parts of it, for example isolated cleft palate compared with cleft lip with or without cleft palate, may have different genetic origins. The types of genetic mechanisms postulated for these defects have included both major gene (autosomal dominant and recessive) and polygenic effects (Carter, 1965). Empirical risk figures derived from family studies are in reasonably good agreement with a polygenic genetic system. These empirical risk figures have a practical value in the provision of genetic counselling advice to families in which these defects have occurred. To date, family studies have been confined almost exclusively to Caucasian populations, and hence the recurrence risks derived from such studies are not applicable directly to non-Caucasian populations. As part of a larger study involving the determination of recurrence risks in the North American Indian population of the Province of British Columbia, estimates of incidence rate were obtained, and this report is concerned with these rates.

Population

The Indian population comprises about 44,000 persons, out of a total population of two million in British Columbia. The classification of 'Indian' is not easy to make in many instances. Our studies involve subjects who are registered with a particular Indian band under the Canadian Indian Act of 1879. There are many Indians who no longer live in bands, and who are counted, for vital statistics purposes, with the larger white community. Many of them are no less Indian in terms of their genotype than their confrères on the reservations. In

addition, it is clear, from family studies which one of us is in the process of carrying out and from previous studies, that there is an admixture of white genes in the entire Indian population (Thomas *et al.*, 1964).

Sources of Data

The British Columbia Registry for Handicapped Children and Adults has been the primary source of data. This Registry has multiple sources for obtaining information: physician's notice of birth, public health units, Indian health services (Department of National Health and Welfare), private physicians, children's hospitals, and specialized treatment centres (e.g. the Cleft Palate Programme at The Health Centre for Children, Vancouver), and it is believed that ascertainment of cases relevant to this study is almost complete.

Results

Tables I and II show the number of cases, percentage distribution, and the rates per 1000 live births of Indian and non-Indian cleft lip and palate cases in British Columbia over the years 1952-1964. These are based on the Registry case load, both live

TABLE I

PERCENTAGE DISTRIBUTION BY TYPE AND NUMBER OF CASES OF CLEFT LIP AND/OR PALATE PER 1000 LIVE BIRTHS IN BRITISH COLUMBIA (LIVE AND DEAD REGISTRY CASE LOAD OF CHILDREN BORN IN BRITISH COLUMBIA 1952-1964 INCLUSIVE)

Type of Cleft Lip and/or Palate	Indian		Non-Indian		Total	
	No.	%	No.	%	No.	%
Cleft lip only	5	7.8	172	23.3	177	22.1
Cleft palate only	9	14.1	244	33.1	253	31.6
Cleft lip and palate	50	78.1	321	43.6	371	46.3
All forms of cleft lip and/or palate	64	100	737	100	801	100

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TABLE II
RATES PER 1000 LIVE BIRTHS IN BRITISH COLUMBIA
IN 1952-1964 (RACE SPECIFIC)

Type of Cleft Lip and/or Palate	Indian	Non-Indian	Total
Cleft lip only	0.25	0.38	0.37
Cleft palate only	0.44	0.54	0.54
Cleft lip and palate	2.48	0.71	0.78
All forms of cleft lip and/or palate	3.17	1.63	1.69

and dead, at the end of 1965. Our rate of 1.63 for non-Indian cases is similar to the Caucasian rate found in many parts of the world (Moller, 1965). Not only do Indians have a strikingly higher rate (3.17 as against 1.63), but the proportions of each malformation are quite different: 78% of the Indians had the cleft lip and palate combination compared with 43% in the non-Indian population. An additional 8% of Indians had isolated cleft lip, whereas 23% of the non-Indian had this defect. Isolated cleft palate which is recognized to be a different genetic entity comprises only 14% of the Indian and 33% of the non-Indian totals.

A breakdown of the Agencies of these Indian cases discloses that the cases of cleft lip and palate are fairly evenly spread over the total Indian case load, though there are a few Agencies which have no reported cases (Table III). It is unlikely that cases of cleft lip and palate would be overlooked in one area and not in another. The population in the areas without any reported cleft lip and palate cases is not large, though the Kamloops Agency (popula-

TABLE III
POPULATION OF INDIANS BY AGENCY WITH
NUMBER OF CASES REGISTERED WITH REGISTRY
FOR HANDICAPPED CHILDREN (TOTAL AND
CLEFT LIP AND PALATE CASES)

Agency	Population 1967	Total Cases Registered	Cleft Lip and/or Palate
Babine	2798	70	4
Bella Coola	2029	70	7
Cowichan	4157	134	5
Kamloops	1963	44	—
Kootenay	568	20	—
Kwawkwalth	2683	80	5
Lytton	2517	54	5
Nicola	1315	37	—
Okanagan	1804	43	4
Queen Charlotte Islands	1319	45	—
Skeena River	2921	87	5
Stuart	2445	56	2
Fraser	6443	276	10
West Coast	3005	110	2
Williams Lake	2978	109	7
Fort St. John	747	17	2
Terrace	2380	77	1
Burns Lake	1094	24	2
Total cleft lip and/or palate			61*

* Three of the 64 Indians in Table II were no longer living on reservations, and had, therefore, lost their Indian status.

tion 2000) might be expected to have several cases. This Agency has been studied in some detail because of the occurrence of hyperprolinaemia (Perry *et al.*, 1968). This would be an additional reason to suppose that cases had not been missed.

The percentage of Indian births taking place in hospital increased over the period studied from about 70% in the earlier years to about 90% in recent years. This means that the physician's notice of birth, a mandatory form which asks the physician to record the presence and type of any congenital malformations, will be filled in on 90% of Indian births at the very minimum. It is recognized that physician's notice of birth does not provide full ascertainment of malformations (Milham, 1963). Cleft palate, particularly the submucous variety, is frequently missed, but it is less likely that cleft lip will be missed, and this accounts for 86% of Indian cases.

There is one family group in which submucous cleft palate appears to be the result of a single X-linked gene (Lowry, 1968). In this family the cleft of the secondary palate is never complete, but involves the soft palate only.

Discussion

Tretsven (1963) pointed out the high frequency of facial clefts in Montana Indians. Miller (1964) published earlier incidence rates based on the same population reported here, and compared these with those for the Japanese reported by Neel (1958). Other Japanese figures have been published (Kurozumi *et al.*, 1963) indicating a frequency, in four studies, of just over 2 per 1000, and in one study of just less than this. Tanaka (1963) showed that the sex distribution in the Japanese was due to a larger number of females with isolated cleft lip. Among the British Columbia Indians isolated cleft lip is an uncommon entity, and the majority of those affected show the double combination of cleft lip and cleft palate. Furthermore, the British Columbia Indians continue to display the male preponderance in this malformation which is found in Caucasian populations (see Table IV). Though cleft lip, plus or minus cleft palate, was more frequently found in males than in females, the difference among Indians was not statistically significant. Conclusions about the sex ratio in isolated cleft palate cannot be drawn since the number of cases is small and the sample is biased by one particular kindred who have an X-linked recessive gene for submucous cleft palate.

Other Asiatic races have a higher incidence of cleft lip and palate, for example the Malays in

TABLE IV

INDIAN AND NON-INDIAN SEX SPECIFIC
RATES OF CLEFT LIP AND PALATE
PER 1000 LIVE BIRTHS (BASED ON 1965
LIVE AND DEAD CHILDHOOD CASES
BORN IN BRITISH COLUMBIA IN 1952-
1964 AND CORRESPONDING LIVE BIRTHS
IN BRITISH COLUMBIA)

	Indian		Non-Indian	
	No.	Rate*	No.	Rate†
Cleft palate only				
Male	5	0.49	120	0.52
Female	4	0.40	124	0.56
Cleft lip ± cleft palate				
Male	34	3.33	322	1.39
Female	21	2.10	171	0.77
Total cleft lip and palate complex				
Male	39	3.82	422	1.91
Female	25	2.50	295	1.33

* Male births, 10,208; female births, 9986.

† Male births, 231,988; female births, 221,218.

Singapore and Kuala Lumpur had an incidence of 2.06 and 2.32 per 1000 total births, respectively (Stevenson *et al.*, 1966). Recently Niswander and Adams (1967) reported a rate of only 1.97 per thousand live births in American Indians. Their study, however, covered a short period of three years and had only one source of ascertainment. They found isolated cleft palate to be about twice the Caucasian rate, whereas we have found them to be more nearly equal. They suggested that the unusual frequency of oral clefts in Montana Indians found by Trettsven may have been due to the presence of an unusually rare major gene at high frequency among the Flathead Tribe. Our studies do not suggest that there is a very high frequency in one particular band in the British Columbia Indians which would bias our statistics, since there is a fairly even distribution of cases throughout the Province.

The higher incidence rates in British Columbia Indians could perhaps be ascribed to increased consanguinity, and this point awaits further study. Of 16 families who have been personally studied, there are 2 instances where the parents are related and two other possible instances.

Summary

Data have been collected on the cleft lip and cleft palate malformation in the Indians of British Columbia, using the Registry for Handicapped Children and its multiple sources of ascertainment. These studies show that the British Columbia Indian has a high frequency of oral clefts. This agrees better with Trettsven's findings, than does that (Niswander and Adams, 1967) for the other American Indian groups.

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