Clinical profile and prevalence of fetal alcohol syndrome in an isolated community in British Columbia

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The authors were invited by the band council to carry out a study to determine the prevalence of alcohol embryopathy among children in a native Indian community in British Columbia. The mothers of the 123 children aged 18 years or less who lived in the community were interviewed. In addition, educational screening was carried out for children in grades 1 through 12, and 116 of the children underwent medical examination. A diagnosis of fetal alcohol syndrome or fetal alcohol effects (FAS/FAE) was made in 22 children aged 3 to 18 years. Each of these children was matched for age and sex with an unaffected child in the same community, and both groups underwent psychoeducational testing. The children with FAS/FAE showed a generalized depressed level of functioning compared with the unaffected children. The finding that two thirds of the children with FAS/FAE were mentally retarded points to a major health and education problem.

À la demande du conseil de bande, nous avons entrepris d'établir la fréquence de l'embryopathie alcoolique chez les enfants d'un village amérindien de Colombie britannique. Entrevue des mères de 123 sujets âgés de moins de 19 ans. Étude de la scolarisation des enfants de la 1e à la 12e année. Examen médical de 116 enfants. Nous posons un diagnostic d'embryopathie alcoolique caractérisée ou soupçonnée (EAC/EAS) chez 22 enfants âgés de 3 à 18 ans. Chacun de ceux-ci ayant été apparié pour le sexe et l'âge à un enfant indemne vivant dans le même village, on procède à des tests psychologiques et scolaires sur tous ces sujets. Les porteurs d'EAC/EAS manifestent une infériorité générale en regard des témoins. Les deux tiers d'entre eux sont débiles mentaux. Il s'agit donc d'un grave problème sanitaire et éducatif.

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Studies of the prevalence of clinical problems provide baseline data for preventive and therapeutic measures, but the difficulties in conducting them are formidable. An invitation to survey a population of children in an isolated community in British Columbia for signs of alcohol embryopathy permitted us an unusual opportunity to conduct a prevalence study. In this paper we report the prevalence of fetal alcohol syndrome and fetal alcohol effects (FAS/FAE) in this group as well as the results of psychoeducational studies in the affected children and in a comparison group from the same community.

Methods

During the winter of 1984 and the spring of

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1985 there were approximately 350 registered members in the native Indian community, including 155 children aged 18 years or less, 123 of whom were living on the reserve.

The minimal criteria for diagnosis used were those recommended by the Fetal Alcohol Study Group of the Research Society on Alcoholism.[1] The diagnosis of FAS was made if there was a history of maternal alcohol abuse or FAS in a sibling, and the child had prenatal or postnatal growth retardation (height and weight below the 10th percentile when corrected for gestational age), central nervous system (CNS) dysfunction (including neurologic abnormalities, developmental delay and intellectual impairment) and characteristic craniofacial abnormalities, including at least two of the following three: microcephaly (head circumference below the third percentile), microphthalmia or short palpebral fissures, and poorly developed philtrum, thin upper lip and flattening of the maxillary area.

Clinical experience indicates a wide range of effects of alcohol on the developing fetus, with FAS at the severe end of the spectrum. When the child shows only one or two of growth retardation, CNS dysfunction and craniofacial abnormalities the term FAE is used.[2]

Three procedures — an interview with the mothers, educational screening and a medical examination — were used to identify the affected children. All the children identified as affected were then assessed by means of a battery of psychoeducational tests to measure the degree of intellectual impairment. To exclude the possibility that cognitive deficits were associated with environmental deprivation a comparison group of unaffected children from the same community matched for age and sex underwent the same assessment. Comparison of the data for the two groups permitted estimation of the magnitude of cognitive deficits apparently due to FAS/FAE.

For the interview with the mothers, all women with children aged up to 18 years were interviewed. A community health nurse visited each woman to explain the purpose of the survey and to have consent forms signed. Each child's date and place of birth and birth weight were confirmed, and the mother's impressions of the child's growth and development since birth were obtained. The mother's background and health history were briefly reviewed, and an appointment was made for completing the interview. At the second interview the following information was sought for each child: prenatal and perinatal history; the mother's health-related behaviour, including the extent of alcohol use, before and during the pregnancy; the child's current behaviour characteristics, including sleeping, eating and activity patterns; details of the child's growth and development and medical history; and the background of the child's father. Although it was possible to determine whether alcohol had been used during the pregnancy, the women often could not remember the amount consumed.

For educational screening a school learning profile was completed for each of the 82 children in grades 1 to 12 in the community school by the teacher(s) who best knew the details of each child's academic performance. The form was not completed for the 20 children in kindergarten or for students over 18 years of age. The information obtained included a rating of the child's general learning ability, ratings of ability in five specific subject areas, ratings of the child's activity level (e.g., hyperactive) and aggressiveness, and an indication of whether school personnel wished to refer the child for in-depth psychoeducational assessment. If a referral was indicated on this form, an additional sheet was completed, providing background details for the examiner's information.

During the medical examination measurements of height, weight and head circumference were obtained, and hearing and vision were assessed with standard screening tests (Zenith Pure Tone Audiometer, Zenith Controls, Inc., Chicago, and Keystone Telebinocular, Mast Development Co., Davenport, Iowa). Photographs of the head (anteroposterior and lateral views) were obtained by a professional photographer. Measurements of facial traits were made by means of procedures described by Vitéz and colleagues.[3] The photographs were of value in reviewing the diagnoses, but the presence of epicanthic folds and other anthropomorphic features of native Indians limited the comparability of these measures to standards reported from other research. Physical examination was performed by one of two physicians, who were unaware of the maternal history of alcohol use during the pregnancy. The clinical findings were recorded on a medical form that itemized the various neurologic, craniofacial and other abnormalities.

Using the information obtained from these three procedures we made a provisional diagnosis of FAS/FAE. Each affected child was then matched with another child who was of the same sex, was not affected (on the basis of the results of the medical examination), was not a sibling of a child with FAS and had the closest possible date of birth. The comparison children were chosen without regard to the results of the interview with the mother or of the educational screening.

A provisional diagnosis of FAS/FAE was made in 22 children. When the results of the psychoeducational assessment and examination of the photographs, reflecting CNS dysfunction and craniofacial abnormalities respectively, were available, we reviewed all the data and made a final diagnosis of FAS or FAE. One child was removed from and one added to the FAS/FAE group. Thus, there were 22 children in the FAS/FAE group but only 21 in the comparison group owing to the removal of one child matched with the removed affected child.

The battery of psychoeducational tests was designed to provide an assessment of a broad range of abilities and learning styles, particularly

those sensitive to CNS insult due to maternal alcohol use. The results of intelligence tests that reflect CNS dysfunction are described in this paper; details of the results of the other psychoeducational measures are available from the authors on request.

Depending on the child's age, the appropriate Wechsler Intelligence Scale or, if preschool age, the McCarthy Scale of Children's Abilities was administered. The subtests on the Wechsler Intelligence Scale establish a verbal intelligence quotient (IQ) (ability to acquire information, use words and understand social situations), a performance IQ (ability to solve problems requiring the manipulation of objects, often labelled perceptual organization) and a composite full-scale IQ. Additionally, these tests demand various degrees of acquired knowledge, memory, and fine motor and other skills that are complemented by other tests in the battery. The McCarthy Scale of Children's Abilities provides a general cognitive index and includes separate verbal, perceptual, motor, memory and quantitative subtests, thus paralleling the tests administered to the older children.

Results

Consent to participate was obtained from the parents or guardians of all 123 children (61 boys and 62 girls) who were living in the community. Seven of the 123 were excluded: 1 had an acute illness, 2 declined to participate, and 4 either attended another school or were away from home (hunting) during the survey. Thus, screening was completed for 116 children of 45 mothers. Fourteen women had 22 children with FAS/FAE; 5 of these women accounted for 12 (54%) of the affected children.

The number of affected children, by use of alcohol during the pregnancy, is shown in Fig. 1. Alcohol use during the pregnancy was confirmed for all 22 children with a diagnosis of FAS/FAE.

Of the 22 children 14 had FAS and 8 had FAE (Table I). The prevalence rate of FAS/FAE was 190 per 1000 children aged 18 years or less.

The year of birth and sex of the 123 children and the number of children with FAS/FAE in each period are shown in Fig. 2. There were no affected children born in the 2 years immediately preceding the study. Of the 22 children 13 were boys and 9 were girls. The mean age (and standard deviation) of the FAS/FAE group was 9.7 years (4.3) and of the comparison group 9.2 years (4.3).

The mean full-scale, verbal and performance IQs on the Wechsler Intelligence Scale for 21 of the affected children and the 21 matched controls are shown in Fig. 3. The means are composite means based on three age-appropriate scales. The mean verbal and performance IQs were significantly lower for the children with FAS/FAE than for the comparison group ($p < 0.001$). However, two children with FAS/FAE had IQs in the same range as the comparison group.

It has frequently been found that performance IQ is significantly higher than verbal IQ among native Indian children and that these children appear to have relative strength in tasks requiring a holistic visual/spatial problem-solving strategy.[3] This pattern was found among the children in the comparison group, whose mean verbal IQ was low-average and whose performance IQ was average. However, among the children with FAS/FAE, both verbal and performance abilities were at a borderline retarded level, with no difference between the two types of ability. The effect of alcohol is apparently greater on performance IQ than on verbal IQ, causing a generalized depression of intellectual performance. Fourteen of the 22 children had IQs in the retarded range (less than 70), and 4 had an IQ of less than 55.

It must be emphasized that these results reflect the average differences between the two groups. A small number of children with FAS/FAE who showed only minor effects had scores similar to those of the comparison group. However, as a

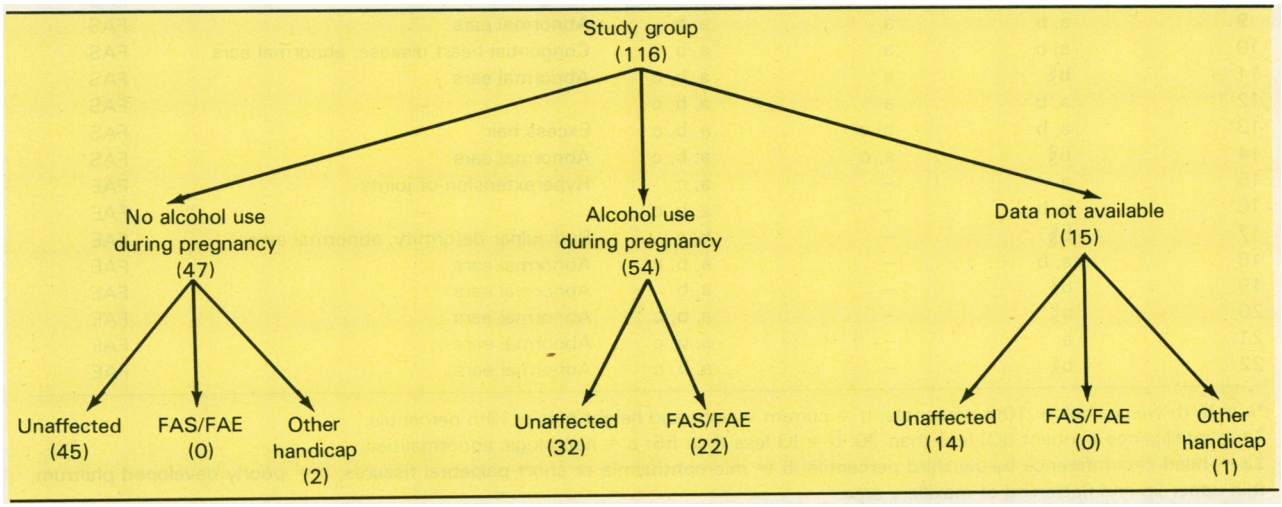

Fig. 1 — Number of children with fetal alcohol syndrome or fetal alcohol effects (FAS/FAE), by maternal alcohol use during pregnancy, among 116 native Indian children in British Columbia.

group the children with FAS/FAE had a generalized depression of neurodevelopmental performance. There are indications in the data of a cumulative deficit: the older children appeared relatively worse off compared with their peers than the younger children; that is, as the children grow older they lag further and further behind.

There was no evidence of increased severity of FAS in subsequent children, as measured by the IQ of the second or third child of the same woman.

Discussion

The frequency of maternal alcohol abuse and the resultant effect on the children in a community are difficult to determine for several reasons. Maternal alcohol abuse is often denied, and the diagnosis of FAS/FAE is not easy to establish. There is no single diagnostic test, and the clinical findings range from overt FAS to one or more suggestive signs. While it is possible to document growth retardation and CNS dysfunction, recognition of some of the craniofacial characteristics is subjective. Measurements of facial traits, including palpebral fissures and nose and lip anomalies, have been described as objective signs,[4] but the anthropomorphic characteristics of Indian children reduced the value of these measures in our study. As a result, underdiagnosis probably occurred. Streissguth[5] summarized the available data on the birth prevalence of FAS and noted that they derived from local studies of available samples and were not designed for broad generalization.

Three studies in northern France,[6] Sweden[7] and the United States[8] showed prevalence rates of FAS in the range of 1 per 750 births. May and colleagues[9] reported higher rates, ranging from 1 per 100 to 1 per 750 live births, among various Indian tribes in the southwestern United States. Studies in British Columbia have indicated prevalence rates ranging from 1 per 2717 live births for the general population to 1 per 150 live births for native Indians (BC Health Surveillance Registry: personal communication, 1983).

Obtaining an accurate measurement of the prevalence of FAS/FAE in a given region involves the study of every family and child; this is time consuming, costly and rarely possible. In our study, however, we had this opportunity, and each child was carefully studied, with particular attention to maternal alcohol use during pregnancy. The prevalence rate of FAS/FAE of 190/1000 children aged up to 18 years was unexpectedly high. In a recent study of handicapped children aged 16 years or less in 23 communities in northwestern British Columbia and 14 communities in the Yukon Territory, prevalence rates of FAS/FAE of

Table I — Clinical findings in 22 children with fetal alcohol syndrome or fetal alcohol effects (FAS/FAE)

Patient no.	Finding: Growth retardation*	Central nervous system dysfunction†	Craniofacial abnormalities‡	Other	Diagnosis
1	a, b	a	a, b, c	Body hair, abnormal ears	FAS
2	b§	a	a, c	Abnormal ears	FAS
3	b	a	a, b, c	–	FAS
4	a, b	a, c	a, c	Excess hair	FAS
5	b§	a, c	a, b, c	–	FAS
6	a, b	b, c	a, b, c	–	FAS
7	b§	b	b, c	Ptosis, abnormal ears	FAS
8	a, b	b, c	a, b, c	Congenital heart disease	FAS
9	a, b	a	a, b	Abnormal ears	FAS
10	a, b	a	a, b, c	Congential heart disease, abnormal ears	FAS
11	b§	a	a, b, c	Abnormal ears	FAS
12	a, b	a	a, b, c	–	FAS
13	a, b	b, c	a, b, c	Excess hair	FAS
14	b§	a, c	a, b, c	Abnormal ears	FAS
15	a	–	a, c	Hyperextension of joints	FAE
16	a, b	–	a, b, c	–	FAE
17	b§	–	b, c	Radioulnar deformity, abnormal ears	FAE
18	a, b	–	a, b, c	Abnormal ears	FAE
19	b§	–	a, b	Abnormal ears	FAE
20	b§	–	a, b, c	Abnormal ears	FAE
21	a	–	a, b, c	Abnormal ears	FAE
22	b§	–	a, b, c	Abnormal ears	FAE

*a = birth weight below 10th percentile; b = current weight and height below 10th percentile.
†a = intelligence quotient (IQ) less than 70; b = IQ less than 55; c = neurologic abnormalities.
‡a = head circumference below third percentile; b = microphthalmia or short palpebral fissures; c = poorly developed philtrum, thin upper lip and flattening of maxillary area.
§Birth weight not available.

26 and 46 per 1000 Indian children were found (Kwadwo Asante and Joyce Nelms-Matzke: unpublished data, 1985). Higher rates would probably have been found in severely affected communities in these two regions if it had been practical to use the methods of our study. Furthermore, the elevated death rate among infants with FAS/FAE reduces the number of live affected children; hence, the prevalence rate may be an underestimate.

In our study most of the children with developmental delay, as observable on clinical examination, were assigned to the affected group. Thus, it is not surprising that the psychoeducational studies showed that the average IQ was significantly lower in the affected group than in the unaffected group.

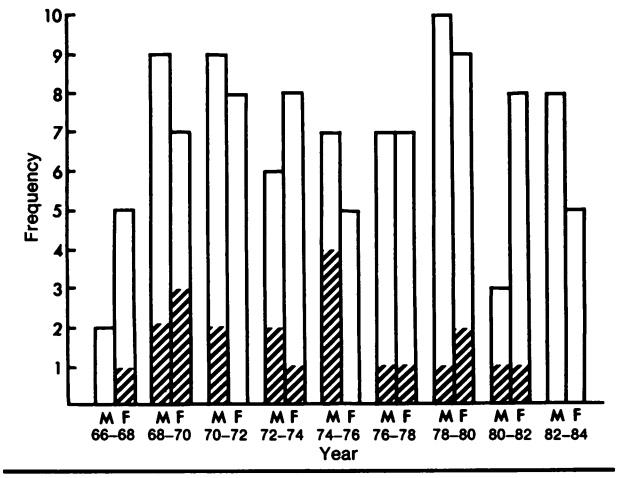

Fig. 2 — Year of birth and sex of study population (n = 123). Striped areas of bars represent children with FAS/FAE; M = male; F = female. Each 2-year period begins on Oct. 1 and ends on Sept. 30.

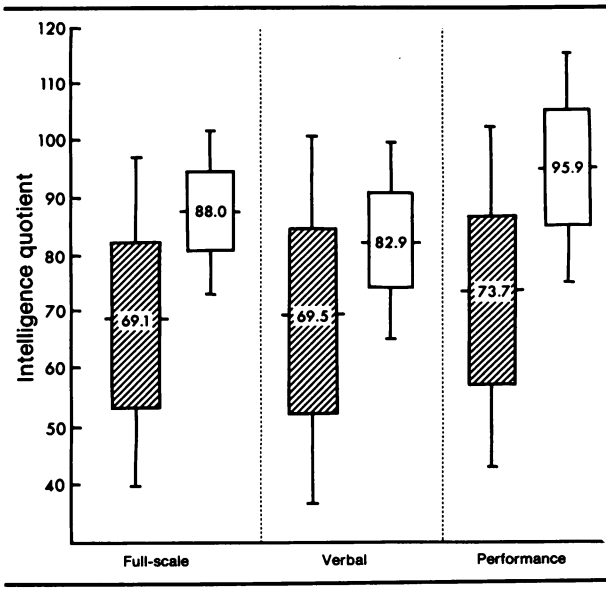

Fig. 3 — Mean full-scale intelligence quotient (IQ), verbal IQ and performance IQ for 21 children with FAS/FAE (striped boxes) and 21 control children (white boxes). Boxes extend to ± 1 standard deviation (SD), vertical lines to ± 2 SD.

However, the severity of the intellectual deficit was unexpected. Of the 22 children with FAS/FAE 64% were mentally retarded. Thus, in addition to the psychologic correlates of environmental deprivation and social disadvantage that may occur in isolated communities, children in such communities whose mothers used alcohol during the pregnancy exhibit other effects.

The finding that two-thirds of the children with FAS/FAE in this community are mentally retarded highlights a major health and education problem. FAS is obviously the most common cause of mental retardation in this community. Recent studies indicate that significant behavioural difficulties continue into adolescence and adulthood,[10,11] and this will surely apply in this community as well. The tragedy is that these disabilities are preventable.

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