Epidemiology of Fetal Alcohol Syndrome in American Indians, Alaskan Natives, and Canadian Aboriginal Peoples: a Review of the Literature

LARRY BURD, MS
MICHAEL E. K. MOFFATT, MD

Mr. Burd is Assistant Professor, Department of Pediatrics and Department of Neuroscience, and Child Evaluation and Treatment Program, University of North Dakota, School of Medicine, Grand Forks. Dr. Moffatt is Associate Professor, Department of Community Health Sciences and Department of Pediatrics and Child Health, University of Manitoba, Winnipeg, Canada.

This work was supported by the Department of Community Health Sciences, University of Manitoba, and the Department of Pediatrics, Department of Neuroscience, and the Child Evaluation and Treatment Program, University of North Dakota School of Medicine.

Tearsheet requests to Mr. Larry Burd, Child Health and Treatment Program, 1300 South Columbia Rd., Grand Forks, ND 58202, telephone 701–777–3065, FAX 701–777–3894.

Synopsis

A critical review of available reports on the epidemiology of fetal alcohol syndrome among American Indians, Alaskan Natives, and Aboriginal peoples of Canada was completed. A search of Medline, the National Institute on Alcohol Abuse and Alcoholism Database, and other relevant data bases was conducted. The reference lists of several publications on fetal alcohol syndrome were reviewed, and four prominent researchers and four government agencies were contacted to identify unpublished articles.

This search identified 10 studies, 8 of them cross-sectional. Four of these studies used primary data from the authors' evaluations of children suspected of having fetal alcohol syndrome; the other six used secondary data. The prevalence of fetal alcohol syndrome in the American Indians of the United States and Aboriginal peoples of Canada was consistently high across the 10 studies. These studies have significant restrictions which limit both the confidence in the rates reported and the generalizability of the results.

Three studies used data from the province of British Columbia. No study evaluated all children in the study area. Only two studies reviewed death certificates. In only one study were examiners blinded to maternal alcohol use, and no study presented evidence on the sensitivity and specificity of either the screening efforts or diagnostic criteria. Such evidence is especially important in studies of secondary data and in studies that report rates for newborn populations.

Studies of the sensitivity and specificity of both the screening and diagnostic criteria for fetal alcohol syndrome would be useful areas for further study. Other study designs, including longitudinal cohort studies, are needed. Additional studies of populations of the American Indians, Alaskan Natives, and Aboriginal peoples of Canada, where low rates of fetal alcohol syndrome are suspected, should be completed. Reviews of death certificates may also be a potentially important source of cases.

PUBLISHED PREVALENCE estimates for fetal alcohol syndrome (FAS) vary more than a hundredfold (*1–7*). Higher rates are found in black, Alaskan Native, Aboriginal peoples of Canada, and American Indian populations (*5–7*). FAS is now reported to be the leading identifiable cause of mental retardation in the United States and western Europe (*1*). Between 5 and 10 percent of children with FAS have been reported to die before 12 years of age, and one study reports that up to 18 percent may die before adulthood (*8*).

American Indians and Aboriginal peoples of Canada are reported to be at very high risk for FAS (*4–6,8*). The toll of FAS on Indian people is graphically portrayed in "The Broken Cord" by Michael Dorris, which describes the anguish and suffering of one FAS child and his family (*9*). He suggests that FAS in the less severe form or the disorder fetal alcohol effect (FAE) is largely unrecognized (*9*).

Due to the potential importance of FAS as a cause of childhood morbidity and mortality, we elected to review available information on the epidemiology of

FAS in Indian, Eskimo, and Aboriginal populations in the United States and Canada. The purpose of this review is to address the following questions:

1. What are the most common epidemiologic designs used in prevalence and incidence studies of FAS?
2. What do these reports have in common that would increase our understanding of FAS?
3. Are there consistent methodological problems in this literature that a systematic review could identify?
4. If consistent methodological problems are identified, what strategies could be suggested to resolve these problems?

Methods

In November 1991, Medline searches were conducted using the terms fetal alcohol syndrome, fetal alcohol effects, alcohol-related birth defects, epidemiology, prevalence, incidence, English only, Indian, Native, Eskimo, Aboriginal, and Canada. For articles published before October 1983, the literature search from the National Library of Medicine No. 83–22 was used (*10*). An additional search was conducted through the Psych Abstracts Database and Social Sciences Citation Index. The search covered literature published from 1979 through 1991, and it yielded 48 articles. A second search of the National Institute on Alcohol Abuse and Alcoholism Database identified no articles that had not been previously identified.

The Addiction Research Foundation's national office in Ottawa, Canada, the Centers for Disease Control in Atlanta, GA, personnel of Indian Health Service, the National Institute on Alcohol Abuse and Alcoholism, and four researchers in the United States and Canada were contacted to identify other unpublished sources. The reference lists of published papers on the epidemiology of FAS and from an FAS text (*7*) were screened to locate other reports. This procedure identified two additional studies. The papers were then screened to identify

- those which did not provide data on the topic of the review,
- those that were general program descriptions, and
- those that, although relevant to the review topic, have not yet collected data that could be used to develop either prevalence or incidence rates after the screening process.

These papers were then omitted from the review. Two papers published after the literature search was completed were included at the suggestion of the journal reviewers of this paper.

Results

We were then left with six published papers which provided information on the epidemiology of FAS in our target population (*4,5,11–14*). Four unpublished reports were also identified which may be obtained by contacting the authors:

"Survey of Children with Chronic Handicaps and Fetal Alcohol Syndrome in the Yukon and British Columbia," 1985, Kwadwo Asante, MD, Clinical Assistant Professor of Pediatrics, University of British Columbia, Suite #103, 12195 Harris Road, Pitt Meadows, British Columbia V3Y 2E9

"Fetal Alcohol Syndrome in British Columbia," 1983, Nancy Wong, British Columbia Surveillance Registry, Ministry of Health, Vancouver, British Columbia

"Fetal Alcohol Syndrome in North Dakota," 1991, Larry Burd, Director of the North Dakota Fetal Alcohol Syndrome Center, Medical Center Rehabilitation Hospital, 1300 S. Columbia Road, Grand Forks, ND 58201

"FAS/FAE in the State of Washington," 1991, Civillia Winslow-Hill, Head, Birth Defects Registry, Washington State Department of Health, Olympia, WA 98504–7811.

A summary of the papers and available incidence and prevalence data are presented in the box. Of the 10 papers, 8 were cross-sectional prevalence studies. In four, the authors examined the individual patients and made the diagnosis (*4,5,13,* and Asante and coworkers). The age ranges in the studies using primary data were from 0–19 years. In these four studies, 774 children were evaluated and 307 (39.6 percent) were found to have FAS.

At least three different sets of diagnostic criteria were used for case definition in these four studies (*4,5,13,* and Asante and coworkers)—FAS criteria from the Research Society on Alcoholism, criteria developed by an expert panel of dysmorphologists; the International Classification of Diseases, ninth revision; and the criteria from Sokol and Clarren (*15*). From an operational standpoint, it is unlikely there are significant differences between these criteria as they all appear to use the major features of central nervous system dysfunction, growth impairment,

Summary of Data from 10 Studies on the Epidemiology of Fetal Alcohol Syndrome (FAS) and Fetal Alcohol Effect (FAE) in Indian or Native Populations in the United States and Canada

1. May, Hymbaugh, Aase, and Samet, 1983 (Reference 5) [1].
Location: 16 sites in southwest United States
Study design and criteria: cross-sectional, developed for study by expert panel
Ascertainment strategy: referrals to study after professional education to increase awareness
Denominator population and age range: 22,963 children (1979 census); 0–14 years
Total FAS and FAE: 115 of 243 in study (47.3 percent)
FAS: 41 males, 35 females
FAE: 26 males, 13 females
Rate: FAS prevalence—mean of 2.0 per 1,000; FAS incidence—range 1.9 to 18.3, mean 2.8 [2]
Comments: Dysmorphologist evaluated each child. Maternal alcohol use known.

2. Robinson, Conry, and Conry, 1987 (Reference 4)
Location: Canim Lake, British Columbia
Study design and criteria: cross-sectional, Research Society on Alcoholism
Ascertainment strategy: evaluated 116 of 123 children living on reservation
Denominator population and age range: 123; 0–18 years
Total FAS and FAE: 22 of 116 in study (18.9 percent)
FAS: 13 males, 9 females
FAE: not available
Rate: prevalence, FAS 190 per 1,000; incidence not available
Comments: Diagnosis made before knowledge of maternal alcohol use.

3. Asante et al., 1985
Location: Yukon and northwest British Columbia, 36 communities
Study design and criteria: cross-sectional, Research Society on Alcoholism
Ascertainment strategy: referrals after extensive public, government, and professional awareness programs
Denominator population and age range: 586 Natives, 195 non-Natives; 0–16 years
Total FAS and FAE: FAS—166 of 391 native children (42.5 percent of native children)
FAS: 113 males, 63 females
FAE: not available
Rate: prevalence, FAS and FAE, 46 per 1,000 Yukon Natives; 25 per 1,000 NW British Columbia Natives; incidence not available
Comments: visited each community, evaluated each child. Maternal alcohol use known.

4. Christensen, 1990 (Reference 11)
Location: Alaska
Study design: cross-sectional, criteria not available
Ascertainment strategy: medical records data
Denominator population and age range: not available
Total FAS and FAE: not available
FAS: not available
FAE: not available
Rate: prevalence—from January 1981 to May 1986, FAS 5.1 per 1,000, FAE 1.7 per 1,000; June 1986 to December 1988, FAS 2.7 per 1,000, FAE 1.7 per 1,000

5. Chávez, Cordero, and Becerrant, 1988 (Reference 12)
Location: United States
Study design and criteria: surveillance, cross-sectional; International Classification of Diseases, ninth edition
Ascertainment strategy: surveillance from medical charts of newborns. Results sent to CDC.
Denominator population and age range: 19,412 births or 9.3 percent of Indian births in United States during 6-year period, average of 3.2 percent births per year.
Total FAS and FAE: not available
Rate: prevalence not available; incidence, 2.93 per 1,000
Comments: Sixty-nine percent of reporting hospitals have fewer than 1,000 births per year.

6. Burd, et al., 1991 [3]
Location: North Dakota
Study design and criteria: cross-sectional, Research Society on Alcoholism
Ascertainment strategy: Questionnaires sent to physicians, developmental clinics, geneticists, and Indian Health Service clinics.
Denominator population and age range: 15,531 children ages 0–18 from North Dakota census data
Total FAS and FAE: not available
FAS: 18 males, 9 females
FAE: not available
Rate: prevalence for FAS, 3.1 per 1,000; incidence, not available
Comments: Criteria for FAS listed on questionnaire.

7. Wong, 1983
Location: British Columbia
Study design and criteria: long-term followup, surveillance, International Classification of Diseases, ninth edition
Ascertainment strategy: surveillance from British Columbia Birth Defects Registry
Denominator population and age range: 11,226 births, 1972–80
Total FAS and FAE: not available
FAS: 54 males, 44 females
FAE: not available
Rate: prevalence, not available; incidence, 6.6 per 1,000

Comments: If mother is alcoholic, records reviewed for possible FAS.

8. Winslow-Hill, 1990
Location: Washington State
Study design and criteria: surveillance—modified cohort; rate includes FAS-FAE combined.
Ascertainment strategy: surveillance from Washington State Birth Defects Registry
Denominator population and age range: 11,226 births, 1972–80
Total FAS and FAE in study population: not available
FAS: not available
FAE: not available
Rate: FAS and FAE: 1987, 0.53; 1988, 0.97; 1989, 2.73; incidence, not available
Comments: If mother is alcoholic, records reviewed for possible FAS.

9. Duimstra, et al., 1993 (Reference 13)
Location: 4 different reservation sites in South Dakota
Study design and criteria: surveillance, cross-sectional, Research Society on Alcoholism
Ascertainment strategy: live births weighing less than 3,000 grams
Denominator population and age range: 1,022 live births in 1 of the 4 Indian Health Service clinics during a 1-year period; age range, less than 2 years
Total FAS and FAE: 4 per 24
FAS: not available
FAE: not available
Percent affected: 17
Rate: prevalence, not stated; incidence, actual FAS 3.9 percent, best estimate 8.5 percent
Comments: No data on children born outside of Indian Health Service clinics. Less than 50 percent of eligible children completed entire process.

10. Bergeson, et al., 1993 (Reference 14)
Location: Alaska
Study design and criteria: cross-sectional, Research Society on Alcoholism
Ascertainment strategy: birth and death certificates, Indian Health Service case files, medical claims from a private practice of pediatricians
Denominator population and age range: 32,932 live births from 1978–91; 0–19 years
Total FAS and FAE: 83 of 348 in study (27 percent)
FAS: 46 males, 37 females
FAE: not available
Rate: prevalence, 2.1 per 1,000 for 1978–91; incidence, not available
Comments: Data available from only 1 pediatric practice; medical records were not always available.

[1] Adjusted by proportion of each cultural population in total area.
[2] Birth incidence.
[3] For 34 children data on sex were not available.

presence of facial features, and maternal alcohol ingestion.

The other six studies all used secondary data. Four studies used surveillance data on newborns (*12,14,* Wong and Winslow-Hill). The prevalence rates for three of these studies include children found to have FAS after the newborn period (Wong, Winslow-Hill, *14*). In two studies, the records of all reported cases of FAS were reviewed by consultant dysmorphologists to confirm the diagnosis (Wong, Winslow-Hill). Two additional papers were also cross-sectional studies which used secondary data sources to identify cases of FAS (*11,* and Burd). One study did determine the differences in prevalence rates among different tribes and demonstrated rates that varied across reservations by a factor of 10 using a common method of case identification (*5,16*).

Discussion

The papers included in this review suggest that the rate of FAS among Indian or Aboriginal peoples in the United States and Canada may be greatly increased compared with rates for whites. The ethnic differences between prevalence rates presented in the box are consistent across the studies and, even with different methods of ascertainment, the high rates among Canadian Aboriginals and American Indians are consistent.

Although the elevated rates of FAS in these populations appear to be consistent, wide gaps in the science base of existing epidemiologic data in this area are apparent. Three of the studies use data from a single geographic area, British Columbia; as a result, the generalizability of these data are limited (*4,* Wong, and Asante). However, these data may provide support for the elevated prevalence rates of FAS in this geographic area and among the populations studied.

The rates of FAS reported will represent an underestimate of FAS rates in these populations. No study evaluated all children who may have had FAS. Robinson and coworkers (*4*) and Asante and coworkers evaluated nearly all eligible children, but both relied on referral sources and may have missed either very severely affected or mildly affected children. In only two studies were death certificates used to estimate the number of children with FAS who had died (*14* and Wong).

In the study by Wong, 12 children (5.6 percent) with FAS had died. Of these deaths there was one stillbirth, nine children died before 1 year of age, one at age 10, and one at age 12. In the study by Bergeson and colleagues, 1 out of 83 affected children were identified from death certificates (*14*). In one related study, 18 percent of children with FAS had died during a 10-year followup period (*8*). These studies suggest that death certificates may be an important source of cases (*14,* Wong and Winslow-Hill). However, death certificates would not be a highly reliable source of information since they often contain inaccurate data.

Only one study provided a modest degree of blinding in the diagnostic process to maternal alcohol use during pregnancy (*4*). In this study, histories of maternal use of alcohol were not known until after the diagnosis of FAS was decided. This is a difficult problem since, in some diagnostic schemas, confirmation of maternal alcohol use is essential for a diagnosis of FAS.

In a review of prevalence studies of FAS, Bray and coworkers observed that caution should be used before one concludes that the Aboriginal population of Canada has elevated rates of FAS (*17*). We would concur with their observation and extend it to the data from the United States as well. We do not suggest that in the communities surveyed FAS is not a major public health problem. However, none of the studies based on primary data compared rates of FAS in other ethnic groups using similar case finding methods and diagnostic criteria.

It appears that some of the communities surveyed were selected because FAS was thought to be a major public health problem. This is likely since 39.6 percent of the children evaluated in the four studies using primary data were found to have FAS (*4,5,13,* and Asante). Comparable studies of Indian or Aboriginal communities selected because FAS rates are presumed to be low have not yet been done. The results reported by Chávez and coworkers are based on surveillance of more than 9 percent of all Indian births in the United States and are consistent with the other studies, which suggest that FAS rates may be increased in this population (*12*). However, clinicians may be more likely to diagnose FAS in the newborn period in American Indians than in other ethnic groups. Thus, these rates may not be comparable due to relative underascertainment in other ethnic groups.

Suggestions for Future Research

Studies are needed to determine sensitivity and specificity of clinical diagnosis by experts of FAS at birth, at 4 or 5 years of age, and in adulthood to determine the confidence one can place in current diagnostic criteria. A major obstacle for this endeavor is the lack of a "gold standard" to compare with clinicians' diagnosis. The developmental course of FAS results in less distinctive clinical features in adults (*18*).

The ability to diagnose FAS at birth is widely debated with some studies suggesting that when the normal process of differential diagnosis is used, clinicians have considerable difficulty identifying affected newborns (*3*). Identification of newborns is quite problematic, since the syndrome is not as distinct at that period as, for example, at 3 or 4 years of age, and it is likely that a proportion of the diagnoses during infancy is made based on a history of maternal alcohol ingestion. Given the potential for misdiagnosis of FAS and the very adverse social ramifications for the parent, family, and child, this issue requires considerably more study in the future.

Future studies should include four features: (*a*) the cohorts should include people with both FAS and developmental disorders other than FAS, (*b*) the cohorts should be stratified by ethnic status, (*c*) the blinding of diagnosticians to the history of maternal alcohol use during pregnancy, and (*d*) the expansion of study designs to allow for identification of sensitivity and specificity of both screening methods and diagnostic criteria.

Future studies need not employ all four of these criteria. Blinding diagnosticians to maternal alcohol use should be a feature of some studies but is not recommended for routine use in clinical practice. Two additional factors should be considered in the design phase of future studies. One is the problem of ascertainment bias if the study base is inappropriately selected; the next most obvious factor leading to either over or under ascertainment would be misdiagnosis.

The potential effects of misdiagnosis could be reduced by linking two diagnostic systems or results from two diagnosticians. A second issue to be considered would be the effect of preconceived ideas about the occurrence rate of FAS in native populations, especially when surveillance systems are used. It would be important to know if a minority of participating physicians account for the majority of cases of FAS in these systems.

Needed are cohort studies to identify a population of children at birth and follow them over time to determine the accuracy of making a diagnosis of FAS in individual children at several ages. Such studies are underway and will be helpful in providing additional information about the natural history of the

disorder; for example, why are so few adult females diagnosed? The results of recent research on adolescents and adults with FAS suggest this area will require considerable effort in the future (*18*).

The problem of establishing comparable FAS surveillance systems and FAS rates is formidable. Further research to determine how the diagnostic criteria for FAS are applied to different ethnic and socioeconomic groups will be important in this effort. The variability in clinicians' willingness to label children with FAS will also have an important influence on rates. The effects of these problems on FAS rates in any individual study will be difficult to determine if these factors are not considered during the design phase of future studies.

The effects of these variables on FAE rates may profoundly affect on the rates of children who are identified and diagnosed with the less distinctive features of the syndrome. The suggestion that studies of longitudinal cohorts are important must be tempered by the fact that very large scale studies would be needed to detect a doubling of risk. The problem of clearly defining exposure status at entry into such a study would also be very difficult. If a study stratified children by race or ethnic status, or both, the population required would rise very rapidly, be difficult to manage, and require a large funding commitment. It seems likely that a series of smaller, carefully coordinated studies may be the most prudent course at this time.

The development of registries for developmental disorders in general and FAS specifically would be very helpful in future surveillance efforts. Service systems like the Indian Health Service would be one logical location of such a data base; it could provide information on developmental disabilities for one-half of the Indian people in the United States. However, the differences in health care delivery systems serving American Indians will make it very difficult to conduct FAS incidence and prevalence studies that will use comparable methods for ascertaining cases in native and non-native populations. The current effort by the Centers for Disease Control and Prevention to support a variety of approaches to FAS surveillance in various population groups throughout the country may improve the science base on FAS surveillance techniques over the next several years.

Consideration of these issues could yield additional data to compare with the existing data on rates of FAS in American Indian, Alaskan Native, and Canadian Aboriginal populations. This information will be crucial in prioritizing funding for intervention and prevention efforts in the future.

References

1. Abel, E. L., and Sokol, R. J.: Fetal alcohol syndrome is now leading cause of mental retardation. Lancet 8517: 1222, Nov. 22, 1986.
2. Abel, E. L., and Sokol, R. J.: A revised conservative estimate of the incidence of FAS and its economic impact. Alcohol Clin Exp Res 15: 514–524 (1991).
3. Little, B. B., et al.: Failure to recognize fetal alcohol syndrome in newborn infants. Am J Dis Child 144: 1142–1146 (1990).
4. Robinson, C. G., Conry, J. L., and Conry, R. F.: Prevalence of fetal alcohol syndrome in an isolated community in British Columbia. Can Med Assoc J 137: 203–207 (1987).
5. May, P., Hymbaugh, K. J., Aase, J. M., and Samet, J. M.: Epidemiology of fetal alcohol syndrome among American Indians of the Southwest. Soc Biol 30: 374–387 (1983).
6. Aase, J. M.: The fetal alcohol syndrome in American Indians: a high risk group. Neurobehav Toxicol Teratol 3: 153–156 (1981).
7. Abel, E. L.: Fetal alcohol syndrome. Medical Economics Books, Oradell, NJ, 1991.
8. Streissguth, A. P., Clarren, S. K., and Jones, K. L.: Natural history of the fetal alcohol syndrome: a 10 year follow-up of eleven patients. Lancet 8446: 85–92, July 13, 1985.
9. Dorris, M.: The broken cord. Harper and Row, New York, NY, 1989.
10. Maternal alcohol consumption and fetal and newborn effects including the fetal alcohol syndrome (FAS). National Library of Medicine Literature Search, No. 83–22, Public Health Service, National Institutes of Health, 1983.
11. Christensen, R.: Health problems among Alaskan Eskimo infants and young children. Arctic Med Res 49: 63–67 (1990).
12. Chávez, G. F., Cordero, J. F., and Becerrant, J. E.: Leading major congenital malformations among minority groups in the United States 1981-1986. MMWR Morb Mortal Wkly Rep 37 (SS-3): 17–24, July 1988.
13. Duimstra, C., et al.: A fetal alcohol syndrome surveillance pilot project in American Indian communities in the Northern Plains. Public Health Rep 108: 225–229, March–April 1993.
14. Bergeson, M., et al.: Linking multiple data sources in fetal alcohol syndrome surveillance-Alaska. MMWR Morb Mortal Wkly Rep 42: 312–314, Apr. 30, 1993.
15. Sokol, R. J., and Clarren, S. K.: Guidelines for use of terminology describing the impact of prenatal alcohol on the offspring. Alcohol Clin Exp Res 13: 597–598 (1989).
16. May, P. A., and Hymbaugh, K. J:. A pilot project on fetal alcohol syndrome among American Indians. Alcohol Health Res World 7: 3–9 (1982).
17. Bray, D. L., and Anderson, P. D.: Appraisal of the epidemiology of fetal alcohol syndrome among Canadian Native peoples. Can J Public Health 80: 42–45 (1989).
18. Streissguth, A. P., et al.: Fetal alcohol syndrome in adolescents and adults. JAMA 265: 1961–1967, Apr. 17, 1991.