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Steep corneal curvature: A fetal alcohol syndrome landmark

JOHN M. GARBER, O.D.

ABSTRACT—Keratometry was performed on 30 fetal alcohol syndrome (FAS) and fetal alcohol effect (FAE) children. Horizontal keratometric measurements that are 46.00 diopters or greater appear to be an objective landmark of FAS. Keratometry is recommended as a routine test in evaluating suspected FAS children and appears to have value in screening the general school population for FAS.

KEY WORDS—astigmatism, Indian, fetal alcohol syndrome, fetal alcohol effect, keratometry

"In the book of Judges, 13:7, it says, 'Behold, thou shalt conceive and bear a son, and now drink no wine or strong drink.' This, it turned out, is better medical advice than physicians have been giving since."—Dr. Frank Iber, M.D., in *Nutrition Today.*[1]

In all populations fetal alcohol syndrome (FAS) is an unnecessary devastating form of prenatal child abuse that permanently cripples the physical and mental state of its victim. FAS is caused by the direct teratogenic effects of consumed maternal alcohol on the human fetus.[2] Even though a mother would never consider giving an alcoholic beverage to her newborn, let alone inducing intoxication, this disturbing scenario occurs when a pregnant woman drinks alcohol. The greater the prenatal alcohol consumption, the greater the chance for more severe manifestations of the syndrome. In one study, a single cocktail eliminated the unborn child's breathing movements for half an hour or longer when consumed by pregnant women at gestation of 37 to 39 weeks.[3]

Other concerns in prenatal alcohol consumption are the teratogenic effects of binge drinking,[4] the higher incidence of stillbirth and spontaneous abortion with alcoholic mothers,[4] the synergistic effects of other teratogens such as cigarettes,[5] and the fact that animal studies show learning impairment with lesser alcohol intake.[6] These facts along with others explain the reasons that no safe level of alcohol consumption during pregnancy has been established. Through the efforts of Drs. Smith and Jones of the University of Washington, FAS has been brought to light in the past decade and continues to gain the attention of the health care community. In 1982 the Department of Health and Human Services officially recommended that women abstain from alcohol during pregnancy.[4]

The full syndrome and its lesser forms are still being defined and debated. Universally accepted FAS landmarks are low weight, small stature, and decreased head circumference (all two standard deviations below the norm).[2] Abnormal facial features include small palpebral fissures, flat nasal bridge, thin vermillion, and a hypoplastic philtrum. Mental retardation usually occurs, with an average IQ of 68.[5] Fetal alcohol effect (FAE) is a term used by some in reference to children who do not manifest the complete morphology of FAS but do have obvious teratogenic alcohol effects, simply to a lesser degree.[7] By its nature FAE is a gray zone that awaits further clarification. Nevertheless, when the total teratogenic effects of alcohol are considered, it is the primary cause of mental impairment in modern nations.[1,5]

I initiated this investigation to evaluate the relationship between FAS and high corneal astigmatism which a previous paper defined as two diopters or greater in at least one eye.[8] In my preliminary findings I observed a high incidence of steeper corneal curvature in the FAS children. In this paper guidelines for keratometric screening for FAS and FAE are discussed.

Population

Drs. Garber, Goodwin and Hughes examined 19 FAS children: 11 at Fort Defiance, 6 at Zuni, and 2 at Towac eye clinics. The majority were identified by the IHS Pediatric Department at the Fort Defiance Indian Hospital and verified by university dysmorphologists, specialists who evaluate different morphologic presentations in human anomalies and disease. Keratometry was obtained from the preliminary report on four FAS children who had been previously unmeasurable due to inadequate cooperation. One FAS child from the initial study had to be excluded due to poor cooperation. Two children had unilateral measurements due to corneal distortions from scarring.

Another eleven children were examined who had been classified as FAE. Seven were from Towac and four from Fort Defiance.

The entire study consisted of 30 children, 15 males and 15 females. Ages at evaluation ranged between 1.5 and 19 years, with a mean of 6.2 years for FAS and 9.3 years for FAE.

When determining FAS predictive values, the two Towac FAS children were excluded because they did not appear to manifest the complete syndrome in the opinion of the investigators in the keratometric clinic prior to the testing. These two children were enrolled in the general public school system unlike the other institutionalized or special educated FAS children.

Results

Astigmatism of two diopters or greater in at least one eye occurred in 42.1% of the FAS children and 27.3% of the FAE children. None of these percentages are significantly different from the 26.3% observed in the Navajo school children in a previous study.[9]

Corneal curvature of 46.00 diopters or more occurred in 29 of 32 FAS eyes (90.6%). Of the three eyes in which it did not, the K-readings exceeded 45.00 diopters. If the two Towac FAS children were included, the corneal curvature of 46.00 diopters or more occurred in 29 of 36 eyes (80.6%).

In the FAE children none of the horizontal powers exceeded 46 diopters. The distribution of the horizontal corneal curvature in the FAS, FAE, and Navajo school children for individual eyes is shown in Table 1.

From a previous study the average curvature for the Navajo school children was a horizontal of 42.11 standard deviation of 1.41; and a vertical power of 43.51, standard deviation of 1.43. For all of the FAS children the horizontal mean is 47.06 and the vertical mean is 48.78, and both are significantly different from that of the Navajo school children. The FAE children have a horizontal mean of 43.75 and a vertical mean of 44.82 and are significantly different from the Navajo school children (see Table 2).

Sensitivity is the ability to correctly identify those who have a disease, and specificity is the ability to identify those who do not.[12] Predictive values are the percentages of positive or negative results

TABLE 1

Distribution of Horizontal Corneal Curvature

Curvature	FAS Eyes	FAE Eyes	Navajo Children's Eyes
Less than 41.00	0	0	318 (19.7%)
41.00–41.87	2 (5.6%)	2 (9.1%)	394 (24.4%)
42.00–42.87	0	2 (9.1%)	462 (28.6%)
43.00–43.87	0	7 (31.8%)	310 (19.2%)
44.00–44.87	2 (5.6%)	8 (36.4%)	96 (5.9%)
45.00–45.87	3 (8.3%)	3 (13.6%)	30 (1.9%)
46.00–46.87	7 (19.4%)	0	4 (0.3%)
47.00–47.87	5 (13.9%)	0	4 (0.3%)
48.00–48.87	11 (30.6%)	0	0
49.00–49.87	6 (16.7%)	0	0
Total	36	22	1,618

TABLE 2

Mean values of Principal Meridians of FAS and FAE in Navajo School Children*

	FAS	Group FAE	FAS (excluding Towac FAS)
Horizontal:			
Sample size	36	22	32
Mean	47.06	43.75	47.61
Standard deviation	2.07	1.01	1.30
t-test	20.58	5.44	21.88
p-value	<.001	<.001	<.001
Vertical:			
Sample size	36	22	32
Mean	48.78	44.82	49.23
Standard deviation	1.59	1.46	1.59
t-test	21.82	4.27	22.36
p-value	<.001	<.001	<.001

* All values are significantly different from those of the general Navajo children population.

TABLE 3

Sensitivity and Specificity Using 46.00 Diopter Horizontal Meridian in Predicting FAS*

Average Horizontal	FAS (Excluding Towac)	Navajo Children (Gen. Pop.)
46 or more	16	4
Less than 46	1	805

* Sensitivity = 16/17 = 94.1%; specificity = 805/809 = 99.5%.

TABLE 4

Predictive Value of 46.00 Diopter Horizontal Meridian as FAS Standard (Prevalence 1/690, Projected Population 100,000)*

	FAS (excluding Towac)	Normal	Total
46.00 or more	136	494	630
Less than 46	9	99,361	99,370
Total	145	99,855	100,000

* Predictive value of a positive test = 136/630 = 21.6%; predictive value of a negative test = 99,361/99,370 = 99.9%.

that are true positives or true negatives when a test is applied to the total population consisting of healthy and diseased individuals.[11]

Using 46.00 diopters of curvature as a criterion, the sensitivity is 93.3% and the specificity is 99.5%. Using the 1/690 prevalence of FAS in the Navajo population[12] and the fact that four of the 809 Navajo school children had 46.00 diopters or greater curvature, the predictive value of a positive test and negative test can be determined supposing that 100,000 children are screened with the 46.00 diopter cut-off. The predictive value of a positive test is 21.6%, and the predictive value for a negative test is 99.9%. These findings imply that in a general screening if a child has a horizontal curvature of 46.00 diopters or greater, there is a one out of five chance that the child has FAS. If the child's horizontal curvature is less than 46.00 diopters, there is a 99.9% chance that the child does not have FAS (see Tables 3 and 4).

Considering the possibility that the four children in the Navajo school group might have undiagnosed FAS, Dr. Nasenbeny, chief of pediatrics at FDIH, reviewed these children's records. Two of the children were suspect with physical traits in the lower third percentile; one of them had been placed in a special education school. If these two children's readings were discarded, the predictive value of a positive test would be 35.5%.

Discussion

In this study the FAS children have a greater prevalence of high astigmatism (42.1%) as contrasted to the general Navajo school population (26.3%), but these differences are not statistically significant. However, if high astigmatism is related to a change in diet that occurred in the early 1940s[13,14] and it is assumed that the FAS child is reared in a poorer nutritional environment until diagnosis, then an increased prevalence of high astigmatism in the FAS Indian population might be expected.

The corneal curvature of FAS is significantly different from that of the general Navajo school population. If an Indian child is suspect for FAS and the keratometry readings are not 46.00 diopters or greater, the diagnosis may be questionable.

With a predictive value of at least 21.6%, keratometry appears to be one objective screening device for the general Indian population. If a patient's keratometry readings are 46.00 diopters or greater, then there is at least a one out of five chance of that person having FAS.

Due to the overall retarded physical and neurological development in FAS, it is not surprising that the eye should be microphthalmic in association with FAS. The smaller the eye, the shorter the radius, and the steeper the curvature. Gross microphthalmia has been noted in some FAS patients. However, this consistent keratometric measurement suggests that the majority have smaller eyes than the general Navajo population. The small palpebral fissures of FAS probably reflect the inability of the smaller eye to distend the adnexa, as has been suggested by Clarren and Smith.[2] Keratometry for non-Indian FAS children needs to be evaluated.

FAE is more difficult to discuss because its true definition has not been established in regard to rigid objective criteria. However, the FAE children evaluated in this study have a significantly steeper cornea (horizontal mean of 43.75) than the general Navajo school children (horizontal mean of 42.11). Perhaps with more subjects a keratometric gradient for FAE can be established.

Horizontal keratometric measurements that are 46.00 diopters or greater appear to be a guideline for screening FAS. Corneal curvature measurements should be a routine procedure in evaluating suspected FAS children.

In a previous study[2] I recommended that keratometry be used in the routine school health screenings of Indian children to detect high astigmatism that had a 33% chance of being undiagnosed by conventional screening tests other than retinoscopy. This study demonstrates that keratometry can also be applied as a screening device to assist in the detection of

FAS children who have been previously undiagnosed.

If keratometric guidelines for FAS could be applied to infants, an objective test for the early detection of FAS could be within our grasp. This prompt recognition would hopefully accelerate the special care that the FAS child needs and would assist the health care community in providing the prompt family counseling needed to avert a repetition of this needless disease. **AOA**

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