Fetal Alcohol Syndrome: Two Reports

I. Natural History: A Ten-Year Follow-up of 11 Patients

In 1973, Jones et al described 11 children with a common pattern of altered morphogenesis and central nervous system dysfunction. Their mothers were chronic alcoholics who continued to drink heavily during pregnancy. Since then, fetal alcohol syndrome has been identified in children from every racial group and in many countries. The teratogenicity of alcohol has been confirmed in laboratory studies involving many different species of animals, and a dose-response curve for prenatal alcohol exposure has been established.

In this report, the authors describe how the 11 children have developed physically and mentally over the past ten years. Two are now dead, one is lost to follow-up, and the remaining eight continue to be growth deficient (with respect to height, weight, and head circumference) and dysmorphic. Although most showed some catch-up linear growth during the first year of life, weight and head circumference decreased relative to the norms during this time in most of the children. Thereafter, length and head circumference remained relatively constant with respect to the norms, whereas there was some catch-up in weight with increasing age. There was relatively slow growth of the head after delivery. During childhood, the children were all strikingly underweight for height.

The major craniofacial features—especially the short palpebral fissures, hypoplastic philtrum, thin vermilion border of the upper lip, and flat midface—did not change during the ten years of follow-up. However, their noses changed, with more prominent growth of the nasal bridge. Cardiac anomalies, which consisted of an atrial septal defect in one patient, patent ductus arteriosus in another, and a grade 3/4 systolic murmur interpreted as a ventricular septal defect in six, have all resolved spontaneously or have become insignificant. Orthopedic complications were managed successfully in almost all patients by casting or splinting.

The short palpebral fissures are thought to be secondary to the decreased growth of the eye. Frank microphthalmia was observed at necropsy in one of the patients. Chronic serous otitis media, probably secondary to eustachian tube dysfunction associated with maxillary hypoplasia, required medical and surgical procedures in four of the children.

None of the eight children followed had normal intellectual development. Four were mildly and four were seriously retarded. The degree to which postnatal environmental factors influenced the development of these children is difficult to assess. Mothers of three of the four seriously retarded children were so severely alcoholic that they died of alcohol-related causes within six years of giving birth.

The two major predictive factors concerning prognosis were the severity of the maternal alcoholism and the extent and severity of the initial pattern of malformation. The four children with the most striking craniofacial abnormalities had the most severe degree of microcephaly, the shortest stature, and the lowest intellectual function. The severity of maternal alcoholism appeared to be the most predictive factor in the backgrounds of the four most severely retarded children.

Streissguth AP, Clarren SK, Jones KL: *Lancet* 1985;2:85-91.

II. Prospective Study of Children Exposed to Variable Amounts of Alcohol in Utero

Although it is well known that offspring of mothers who consume large quantities of alcohol during pregnancy are at high risk for physical and mental deficiencies, few prospective studies have dealt with the fetal effects of interrupted alcohol consumption during pregnancy as the result of an intervention program. The authors describe a Swedish antenatal program that was started to help pregnant women stop alcohol abuse with the hope of reducing the adverse effects of alcohol on the fetus.

A total of 40 children born to alcoholic women (Groups 2 and 3) and 40 children born to nonalcoholic women (Group 1) attending the same local maternity health clinics for antenatal care were studied between the ages of 18 and 27 months. The mothers in Group 1 drank less than 30 g of pure alcohol prior to the first prenatal visit and abstained or minimized their consumption thereafter. Group 2 consisted of 25 children born to women who were classified as excessive drinkers and had an average consumption of 30 to 150 g of pure alcohol per day during the month before their first visit to the clinic. All mothers in this group markedly reduced their alcoholic consumption after their first visit and 19 abstained completely. Group 3 consisted of 15 children of alcoholic mothers who had an average consumption of more than 125 g of pure alcohol per day during the month before the first visit to the clinic. Nine mothers in this group stopped drinking alcohol during the first or second trimester, but the remaining six continued drinking throughout the pregnancy.

A statistically significant reduction in weight, height, and head circumference was seen in Group 3 children when compared with Group 1 children. Six of the 15 alcoholic women (Group 3) continued to abuse alcohol throughout pregnancy. Three of these women gave birth to children with abnormalities characteristic of fetal alcohol exposure; one child had the complete fetal alcohol syndrome. Only one child in Group 3 was normally developed in all physiological parameters and had normal behavior.

No fetal growth retardation was found among the children in Group 2, where the mothers reduced or ceased alcohol consumption after their first prenatal visit. Neither did these children show any other physical or physiological characteristic of fetal alcohol syndrome. About half of them, however, had retarded speech that the authors attributed to postnatal environmental influences. Indeed, signs of social instability, such as frequent separations between the parents and frequent registrations with the social welfare department, were seen in Group 2.

The authors suggest that fetal exposure to alcohol has a severe adverse effect on development, but

this can be significantly reversed by abstaining from alcohol in the first trimester of pregnancy. Cessation of alcohol abuse after the first trimester cannot reduce the documented increased risk of congenital malformations.

Larsson G, Bohlin AB, Tunell R:
Arch Dis Child 1985;60:316-321.

Editor's comment—*Fetal alcohol syndrome has been well established as an important cause of congenital malformations, mental retardation, and prenatal and postnatal growth retardation. This syndrome is being diagnosed more frequently now that physicians are specifically questioning mothers-to-be about teratogenetic exposure and, especially, alcohol exposure. The study by Streissguth et al indicates the relationship between the severity of the maternal alcoholism and the severity of the physical and mental handicap. It also points out that the various features of this syndrome tend to be correlated to severity: degree of mental retardation, growth deficiency, and intellectual impairment. This study also points out the difficulty in diagnosing this syndrome after mid-childhood, since some of the typical facial characteristics change, specifically, the structure of the nose, while the markedly underweight appearance of the female children who had reached puberty disappeared. It is unknown whether this applies to the weight of males of pubertal age, since none of the boys had reached puberty. Moreover, it becomes increasingly difficult to obtain a documented history of maternal alcohol abuse as the children grow older.*

The Swedish study documents the importance of discontinuing alcohol before pregnancy or at least during the first few weeks of pregnancy. It is interesting that those women who drank fairly heavily during the first month or so of pregnancy and then stopped, or markedly reduced their intake, had no physical abnormalities in their children. Those women who did not stop or reduce their drinking until the second or third trimester, however, continued to have children with abnormalities characteristic of fetal alcohol exposure. It is hoped that with the increased emphasis and warnings concerning alcohol exposure in utero, the incidence of this devastating syndrome will be reduced in coming years.