

# AMERICAN ACADEMY OF PEDIATRICS

Committee on Substance Abuse and Committee on Children With Disabilities

## Fetal Alcohol Syndrome and Alcohol-Related Neurodevelopmental Disorders

**ABSTRACT.** Prenatal exposure to alcohol is one of the leading preventable causes of birth defects, mental retardation, and neurodevelopmental disorders. In 1973, a cluster of birth defects resulting from prenatal alcohol exposure was recognized as a clinical entity called *fetal alcohol syndrome*. More recently, alcohol exposure in utero has been linked to a variety of other neurodevelopmental problems, and the terms *alcohol-related neurodevelopmental disorder* and *alcohol-related birth defects* have been proposed to identify infants so affected. This statement is an update of a previous statement by the American Academy of Pediatrics and reflects the current thinking about alcohol exposure in utero and the revised nosology.

ABBREVIATIONS. FAS, fetal alcohol syndrome; ARND, alcohol-related neurodevelopmental disorder; ARBD, alcohol-related birth defects.

The term *fetal alcohol syndrome* (FAS) refers to a constellation of physical, behavioral, and cognitive abnormalities.<sup>1</sup> In addition to the classic dysmorphic facial features, prenatal and postnatal growth abnormalities, and mental retardation that define the condition, approximately 80% of children with FAS have microcephaly and behavioral abnormalities.<sup>2</sup> As many as 50% of affected children also exhibit poor coordination, hypotonia, attention-deficit hyperactivity disorder, decreased adipose tissue, and identifiable facial anomalies, such as maxillary hypoplasia, cleft palate, and micrognathia. Cardiac defects, hemangiomas, and eye or ear abnormalities are also common.<sup>3-5</sup>

The term *fetal alcohol effects* was developed originally to describe abnormalities observed in animal studies, but it was adopted quickly by clinicians to describe children with a variety of problems, including growth deficiency, behavioral mannerisms, and delays in motor and speech performance, who lacked the full complement of FAS diagnostic criteria. The lack of specificity and absence of definitive diagnostic criteria have made research and classification difficult, and a 1980 report from the Research Society on Alcohol suggested that fetal alcohol effects encompassed "any condition thought to be secondary to alcohol exposure in utero."<sup>6</sup> Clearly, such a definition was cumbersome and allowed for wide diver-

gence in interpretation.<sup>6</sup> The Institute of Medicine in 1996 issued a report proposing the terms *alcohol-related neurodevelopmental disorder* (ARND) and *alcohol-related birth defects* (ARBD) to describe conditions in which there is a history of maternal alcohol exposure (defined as substantial regular intake or heavy episodic drinking) and an outcome validated by clinical or animal research to be associated with that exposure.<sup>7</sup> This new terminology uses a pathophysiologic basis for the diagnostic categories to describe conditions resulting from prenatal alcohol exposure (Fig 1).

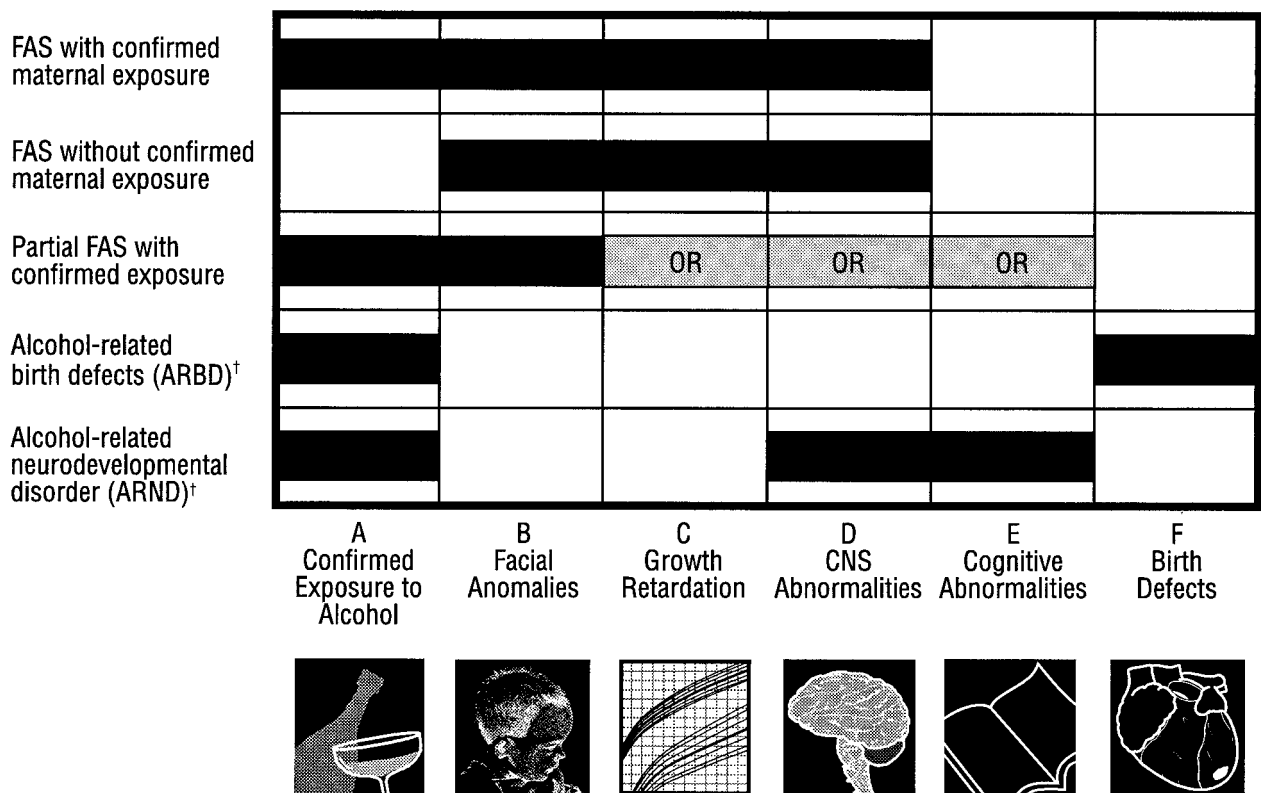
Children with FAS, ARND, and ARBD may manifest cognitive, behavioral, and psychosocial problems that cause lifelong disabilities, although the resulting manifestations may vary with age and circumstances. Streissguth et al<sup>5,8</sup> traced the natural history of alcohol-affected children into adulthood and demonstrated the profound, pervasive, and persistent nature of the disorder. Abnormal cognitive functioning manifested itself in many domains, including specific mathematical deficiency, difficulty with abstraction (eg, time and space, cause-and-effect), and problems with generalizing from one situation to another. The affected persons also demonstrated poor attention and concentration skills, memory deficits, and impaired judgment, comprehension, and abstract reasoning. Behavioral issues, such as hyperactivity and impulsivity, and conduct problems, such as lying, stealing, stubbornness, and oppositional behavior, were common and were quantitatively and qualitatively different from those found in other forms of mental retardation.

None of the persons in the aforementioned study had achieved age-appropriate socialization or communication skills. Maladaptive social functioning was evidenced by their failure to consider consequences for their actions, lack of response to appropriate social cues, lack of reciprocal friendships, social withdrawal, sullenness, mood lability, teasing and bullying behavior, and periods of high anxiety and excessive unhappiness. Secondary disabilities, such as mental health problems, chemical dependency, failure to develop appropriate sexual behavior, and consequent legal problems, were also common in adults diagnosed with FAS. Current evidence suggests that while IQ scores <70 in this population increase the likelihood of such outcomes, early diagnosis and intervention may reduce the occurrence of secondary disabilities.<sup>9</sup>

As one of the most commonly identifiable causes of mental retardation, FAS is estimated to occur at

The recommendations in this statement do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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\*Adapted from *Fetal Alcohol Syndrome: Diagnosis, Epidemiology, Prevention, and Treatment*. 1996;4–5. Letter designations in the figure indicate the following:

- A. Confirmed maternal alcohol exposure indicates a pattern of excessive intake characterized by substantial, regular intake or heavy episodic drinking. Evidence of this pattern may include frequent episodes of intoxication, development of tolerance or withdrawal, social problems related to drinking, legal problems related to drinking, engaging in physically hazardous behavior while drinking, or alcohol-related medical problems such as hepatic disease.
- B. Evidence of a characteristic pattern of facial anomalies that includes features such as short palpebral fissures and abnormalities in the premaxillary zone (eg, flat upper lip, flattened philtrum, and flat midface).
- C. Evidence of growth retardation, including at least one of the following:
- low birth weight for gestational age
  - decelerating weight over time not caused by nutrition
  - disproportional low weight to height
- D. Evidence of CNS neurodevelopmental abnormalities, including at least one of the following:
- decreased cranial size at birth
  - structural brain abnormalities (eg, microcephaly, partial or complete agenesis of the corpus callosum, cerebellar hypoplasia)
  - neurological hard or soft signs (as age appropriate), such as impaired fine motor skills, neurosensory hearing loss, poor tandem gait, poor eye-hand coordination
- E. Evidence of a complex pattern of behavior or cognitive abnormalities that are inconsistent with developmental level and cannot be explained by familial background or environment alone, such as learning difficulties; deficits in school performance; poor impulse control; problems in social perception; deficits in higher level receptive and expressive language; poor capacity for abstraction or metacognition; specific deficits in mathematical skills; or problems in memory, attention, or judgment.

F. Birth defects associated with alcohol exposure include:

|          |  |   |
|----------|--|---|
| Cardiac  | Atrial septal defects<br>Ventricular septal defects  | Aberrant great vessels<br>Tetralogy of Fallot   |
| Skeletal | Hypoplastic nails<br>Shortened fifth digits<br>Radioulnar synostosis<br>Flexion contractures<br>Camptodactyly  | Clinodactyly<br>Pectus excavatum and carinatum<br>Klippel-Feil syndrome<br>Hemivertebrae<br>Scoliosis |
| Renal    | Aplastic, dysplastic, hypoplastic kidneys<br>Horseshoe kidneys   | Ureteral duplications<br>Hydronephrosis   |
| Ocular   | Strabismus<br>Retinal vascular anomalies   | Refractive problems secondary to small globes   |
| Auditory | Conductive hearing loss  | Neurosensory hearing loss   |
| Other    | Virtually every malformation has been described in some patient with FAS. The etiologic specificity of most of these anomalies to alcohol teratogenesis remains uncertain. |   |

<sup>†</sup>Alcohol-related effects indicate clinical conditions in which there is a history of maternal alcohol exposure, and where clinical or animal research has linked maternal alcohol ingestion to an observed outcome. There are two categories, alcohol-related neurodevelopmental disorder and alcohol-related birth defects, which may co-occur. If both diagnoses are present, then both diagnoses should be rendered.

Fig 1. Diagnostic classification of fetal alcohol syndrome (FAS) and alcohol-related effects.\*

the rate of 5.2/10 000 live births in the United States.<sup>10–12</sup> Higher rates are reported among selected subgroups (eg, 30/10 000 among Native Americans).<sup>11–13</sup> There seems to be a number of factors that determine the outcome of a pregnancy during which the mother consumes alcohol. Mills et al<sup>14</sup> prospectively studied approximately 31 000 pregnancies in an attempt to determine how much alcohol pregnant women can consume safely. The consumption of 1 or more drinks (a *drink* is defined as 1.5 oz distilled spirits, 5 oz of wine, or 12 oz of beer) per day was associated with increased risk of giving birth to an infant with growth retardation. Although maternal age, parity, and health as well as specific fetal susceptibility may contribute to the infant's outcome, the potential for harm to the fetus is much stronger with large amounts of maternal alcohol consumption than with smaller amounts.<sup>15</sup> Nevertheless, current data do not support the concept of a "safe level" of alcohol consumption by pregnant women below which no damage to a fetus will occur.

The economic effects of FAS, ARND, and ARBD based on the medical, surgical, behavioral, custodial, and judicial services required takes its toll on the individual, the family, and society. Annual cost estimates for the United States range from \$75 million to \$9.7 billion.<sup>16</sup> The total lifetime cost of caring for a typical child with FAS may be as high as \$1.4 million.<sup>5</sup> The mental retardation related to FAS has by itself been estimated to account for as much as 11% of the annual cost of caring for all mentally retarded institutionalized residents of the United States and may account for up to 5% of all congenital anomalies.<sup>17,18</sup> The nonfiscal costs to families and affected children in terms of emotional and social effects are enormous.

### RECOMMENDATIONS

1. Because there is no known safe amount of alcohol consumption during pregnancy, the Academy recommends abstinence from alcohol for women who are pregnant or who are planning a pregnancy.
2. Major efforts should be made at all levels of society to develop high-quality educational programs about the deleterious consequences of alcohol for the unborn child. This information should be integrated into a comprehensive drug prevention education curriculum for all elementary, junior high, and high school students. It also should be a part of similar education efforts in all postsecondary and adult centers of learning.
3. Pediatricians and other health care professionals who provide care for women and their newborns should increase their own awareness and that of their patients about FAS, ARND, and ARBD and their prevention. Pediatricians should increase their awareness of the prevalence of alcohol use by pregnant women in their communities and advocate for programs that identify the users and offer them treatment. When a child with problems related to maternal alcohol consumption is identified, alcohol treatment and prevention resources should be offered to the family and affected child.

4. Infants and children with a suspected diagnosis of FAS, ARND, or ARBD should be evaluated by a pediatrician who is knowledgeable and competent in the evaluation of neurodevelopmental and psychosocial problems associated with the diagnoses. The need for a skilled evaluation at an early age necessitates referral to a pediatric medical specialist as well as referral to early intervention and education agencies providing services under the provisions of the Individuals With Disabilities Education Act.
5. Parents of children given a diagnosis of FAS, ARBD, or ARND should receive appropriate support services for themselves and their child, including careful anticipatory guidance directed toward preventing similar problems in the future.
6. The Academy supports federal legislation that would require the inclusion of health and safety messages in all print and broadcast alcohol advertisements based on the US Surgeon General's warning: "Drinking during pregnancy may cause mental retardation and other birth defects. Avoid alcohol during pregnancy."
7. The Academy supports the development of state legislation that makes information about FAS, ARND, and ARBD available at marriage-licensing bureaus and other appropriate public places, including points of alcohol sale.
8. Pediatricians are encouraged to assume a leadership role in public education campaigns aimed at decreasing the incidence of FAS through reduction in alcohol use by pregnant women.

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