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FETAL ALCOHOL SYNDROME

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From THE HANDBOOK OF INTERNATIONAL ADOPTION MEDICINE by Laurie C. Miller.

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FETAL ALCOHOL SYNDROME

INTRODUCTION

Families adopting internationally have usually heard of fetal alcohol syndrome (FAS) as a risk for their children. Few realize however, the spectrum of problems that occur after prenatal alcohol exposure, the lifelong disabilities these children experience, and the difficulty establishing this diagnosis in the absence of reliable maternal history.

Fetal alcohol syndrome has been identified among internationally adopted children from virtually every sending country. However, FAS is much more frequent in children from Russia, Ukraine, and other countries of the former Soviet Union. Adoptees from other Eastern European countries such as Romania have an intermediate incidence of FAS. FAS is uncommon in children from Korea, Guatemala and China. The incidence of FAS in international adoptees parallels the incidence of alcoholism in each sending country. The World Health Organization's Global Alcohol Database provides comparable country-specific statistics: http://www.who.int/substance_abuse/publications/globalstatusreportalcoholprofiles/en/index.html.

DEFINITIONS

Fetal alcohol syndrome is a constellation of physical and neurobehavioral abnormalities resulting from maternal ingestion of alcohol during pregnancy.

The characteristics of FAS include:
1) Pre- or postnatal growth retardation (below the 10^{th} percentile)
2) Facial dysmorphology (structural defects) such as midline facial defects, micro-ophthalmia (an abnormally small eye, a congenital malformation of the globe) and/or short palpebral fissures (short eye openings), poorly developed philtrum (area above the upper lip), thin upper lip and absent cupid's bow, flattened maxillary area (flat cheekbones) and low-set ears
3) Central nervous system anomalies such as developmental delay, intellectual impairment, behavioral disturbances and microcephaly (less than third percentile)

Fetal alcohol effects (FAE) is sometimes used to describe prenatally exposed children who lack the characteristic facial appearance of FAS. These children often have growth and developmental delays and neurobehavioral abnormalities. However FAE lacks specific diagnostic criteria and is not a mild form of FAS.

In 1996, the Institute of Medicine proposed the replacement of FAE with the terms alcohol-related neurodevelopmental disorder (ARND) and alcohol-related birth defects (ARBD). Others suggest use of the term fetal alcohol spectrum disorders. These new terms reflect the reality of medical and developmental problems that occur after substantial regular intake or heavy episodic prenatal exposure to alcohol, regardless of the phenotypic (observable physical characteristics) appearance of the child.

The use of these definitions for international adoptees is questionable. Many internationally adopted children have multiple, complex reasons for pre- or postnatal growth retardation. Likewise, other exposures such as malnutrition, micronutrient deficiencies, institutionalization, stress, prenatal drugs or tobacco may result in neurobehavioral problems and cognitive delays. Furthermore, maternal history of alcohol ingestion is rarely available. Nonetheless, FAS and ARND/ARBD may be identified with certainty in some internationally adopted children.

PHYSICAL FEATURES OF FETAL ALCOHOL SYNDROME AND ALCOHOL-RELATED BIRTH DEFECTS AND NEURODEVELOPMENTAL DISORDERS

Children with prenatal alcohol exposure have a variety of abnormal physical features. The characteristic facial appearance is usually under recognized in the newborn period but becomes more apparent during late infancy and early childhood. Many of the facial features become less prominent as children enter adolescence.

Growth delays are characteristic of FAS and ARBD. Nearly 80% of children have height, weight and/or head circumference below the fifth percentile at birth, throughout infancy and beyond. The microcephaly results from poor brain growth.

SPECIFIC PHYSICAL FEATURES OF FAS BY TYPE [1]

Skeletal

- Clinodactyly - curving of the fifth finger (the little finger) toward the fourth finger (the ring finger)
- Camptodactyly - inability to fully extend the fingers
- Radioulnar synostosis - condition where the 2 bones in the forearm are fused together
- Flexion contractures - inability to fully straighten or extend at a joint
- Altered creases on the palm of the hand
- Small distal phalanges - last bones of the fingers
- Short fourth and fifth metacarpals - cylindrical bones extending from the wrist to the fingers
- Small fifth fingernails
- Short neck cervical vertebral malformations
- Hemivertebrae - Congenital malformation of the spine in which only half of a vertebral body develops
- Pectus excavatum - funnel chest, "caved-in" chest
- Pectus carinatum - pigeon-breasted
- Rib anomalies
- Myelomeningocele - congenital disorder where the backbone and spinal canal do not close before birth. Also known as spina bifida.
- Hydrocephalus - abnormal accumulation of cerebrospinal fluid (CSF) in the brain
- Maxillary hypoplasia and micrognthia - flat cheekbones

Cardiac

- VSD - ventricular septal defect - the heart has a hole in the wall (the septum) between its two lower chambers (the ventricles)
- ASD - atrial septal defect **-** a hole in the septum, the wall, between the atria, the upper chambers of the heart
- Tetralogy of Fallot - A combination of four heart defects that are present together at birth: ventricular septal defect, pulmonary stenosis, right ventricular hypertrophy and overriding aorta
- Coarctation of the aorta - a congenital constriction of the aorta, impeding the flow of blood below the level of the constriction and increasing blood pressure above the constriction
- Aberrant great vessels

Craniofacial

- Microcephaly - an abnormally small head due to failure of brain growth
- Short palpebral fissures - short eye openings
- Epicanthal folds - inner eye folds
- Ptosis - drooping of the eyelids, may be asymmetric
- Myopia - nearsightedness
- Micro-ophthalmia - An abnormally small eye, a congenital malformation of the globe
- Increased retinal vessel tortuosity - curving of blood vessels in the back of the eye unrelated to surgery
- Optic nerve hypoplasia – underdeveloped optic nerve
- Strabismus - crossed eyes
- Cleft lip and/or palate
- Other orofacial clefts
- Maxillary hypoplasia - flat cheekbones
- Hypoplastic nasal bridge – underdeveloped nasal bridge
- Short nose
- Anteverted nostrils - short, upturned nose
- Smooth philtrum (area above the upper lip) with thin upper lip,
- Absent cupid's bow – upper lip has no shape
- Protruding ears
- Low-set and posteriorly rotated ears

Renal

- Aplastic, dysplastic, hypoplastic kidneys – missing or development is abnormal or incomplete
- Horseshoe kidneys – normal kidneys are bean shaped
- Hydronephrosis (although some authors disagree) - distention of the kidney with urine

Other

- Hypoplastic labia majora - abnormal development of the two outer folds of the vulva
- Strawberry hemangiomata - congenital bright red superficial vascular tumor resembling a strawberry
- Accessory nipple
- Single umbilical artery

NEUROBEHAVIORAL FEATURES OF FETAL ALCOHOL SYNDROME AND ALCOHOL-RELATED BIRTH DEFECTS AND NEURODEVELOPMENTAL DISORDERS

Impaired cognitive and psychosocial function are the most disabling features of FAS and ARBD. Children with FAS have a broad range of IQs with an average IQ of about 70. Cognitive deficits are common in children after heavy prenatal alcohol exposure with or without the physical features of FAS.

About 80% of children with FAS have obvious behavioral abnormalities by middle-school age. About 50% of children affected with FAS have some combination of poor coordination, hypotonia (decreased muscle tone) or attention-deficit hyperactivity disorder (ADHD) and frequently have poor interpersonal skills. Even in the absence or mental retardation some children have poor executive function, verbal learning and memory deficits, especially spatial memory. While some children have intelligence in the normal range, many experience academic failure due to problems of activity and attention regulation (especially visual and auditory), severe learning disabilities, behavior disorder, delayed motor development, poor balance and marked instability.

Many children also exhibit sleep disorder, abnormal habits and stereotypy. Auditory problems that are also common include a developmental delay in auditory maturation, sensorineural hearing loss, intermittent conductive hearing loss due to recurrent serious otitis media and central hearing loss.

DIAGNOSIS

The diagnosis of FAS is based on clinical finding and maternal history. But in the absence of maternal history FAE or ARDD/ARND can not be determined if diagnostic criteria are strictly applied. However, the combination of facial features, growth delays (especially microcephaly), and neurobehavioral abnormalities strongly suggest this diagnosis in international adoptees, especially in those from high-risk areas.

OUTCOME OF CHILDREN WITH FETAL ALCOHOL SYNDROME

Multiple pre- and postnatal factors can complicate the outcome of children with FAS. Birth weight, microcephaly, gestational age and prenatal exposure to drugs and tobacco may each independently influence outcome. Isolation of prenatal alcohol exposure as an independent variable in the assessment of outcome is difficult to achieve. Also, referral and case ascertainment bias may select for children with more severe outcomes in some studies.

What is relatively straightforward is the outcome of growth deficits in children with FAS. Height, weight and head circumference deficits gradually improve during childhood. In boys, short stature and underweight persist while girls generally achieve a normal weight by adolescence. By 15 years of age microcephaly persists in about 65% of children but improves in the remainder.

No studies to date specifically address the outcome of children with intrauterine alcohol exposure placed as newborns or infants in stable foster care or adoptive homes.

HERITABILITY OF ALCOHOLISM

Many adoptive and foster parents of children with FAS wonder if their child is at increased risk for development of alcoholism. While there is biochemical and molecular evidence that alcoholism may be inherited, environmental factors play a substantial role in determining the development and expression of alcoholism. Conflict or psychopathology in the adoptive family will increase the risk of adopted children of alcoholic birth parents will themselves become alcoholics.

Early, concrete and continuous education about alcohol use and decision making is recommended for children with FAS.

PREVENTION OF SECONDARY DISABILITIES

A child with fetal alcohol exposure presents many challenges. Parenting concerns in raising a child with FAS include:

1. Sleep disturbances – difficulty falling asleep, frequent wakening during the night
2. Poor appetite, difficulties coordinating sucking and swallowing
3. Developmental delays
4. Speech and language delays – expressive language usually better than receptive language
5. Frequent ear infections, dental problems, and upper respiratory infections
6. Sensory integration disorder
7. Hyperactivity, poor attention span
8. Learning disabilities, cognitive delays, often scattered
9. Inappropriate social behaviors, unresponsiveness to social cures, poor judgment
10. Problems making or keeping friends
11. Parenting stress

Early intervention may provide ongoing physical, occupational and speech therapy. Comprehensive educational support in the school should be instituted early and changing needs should be addressed. As different challenges emerge, the pediatrician should expect to be involved with a team of specialists to provide ongoing support to the child with FAS and ARBD and the family.

A number of positive characteristics have been identified in some children with FAS/FAE:
1. Cuddly, cheerful, tactile
2. Friendly and happy
3. Caring, kind, loyal, nurturing, compassionate
4. Trusting and loving
5. Determined, committed, persistent
6. Curious, involved
7. Energetic, hard-working, athletic
8. Artistic, musical creatively intelligent
9. Fair, cooperative
10. Highly verbal
11. Fair with younger children and animals
12. Able to have long-term visual memory
13. Able to participate in problem solving

KEY POINTS FOR INTERNATIONALLY ADOPTED CHILDREN

1. Alcohol exposure history is usually unknown for internationally adopted children.
2. Diagnosis is therefore based on characteristic facial appearance, growth delays and neurobehavioral difficulties.
3. Factors other than alcohol exposure can account for abnormal growth or neurobehavioral findings.

RESOURCES

Centers for Disease Control and Prevention www.cdc.gov/ncbddd/fas/

National Organization on Fetal Alcohol Syndrome www.nofas.org/

The Arc of the United States www.thearc.org

Fetal Alcohol Syndrome Family Resource Institute www.fetalalcoholsyndrome.org/

University of Washington (Seattle) Fetal Alcohol and Drug Unit depts.washington.edu/fadu/

Notes

[1] Most definitions were obtained from the website MedicineNet.com
http://www.medicinenet.com/diseases_and_conditions/article.htm