Fetal Alcohol Spectrum Disorders and the Foster Care System

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Disclosure: Julia Bledsoe, MD
I have no relevant financial relationships with the manufacturer(s) of any commercial product(s) and/or provider of commercial services discussed in this CME activity.
I do not intend to discuss an unapproved/investigative use of any commercial product/device in my presentation.

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Overview
• Define Fetal Alcohol Spectrum Disorders
• Approaches to diagnosis
• How the brain is impacted by alcohol
• Intervention strategies

What Is FAS?
• Permanent birth defect syndrome caused by maternal alcohol consumption during pregnancy
• An FAS diagnosis requires:
  - Pre and/or postnatal growth deficiency
  - Cluster of minor facial anomalies
  - Brain dysfunction
  - Prenatal alcohol exposure

A Brief History of FAS
• Christy Ulleland - UW Pedst Resident! HMC FTT study ->
• Named in 1973 by our own Jones and Smith
• But the French got there first (Lemoine, 1968)
• Confirmation, widening, diffusion of the diagnosis
• Attempts to standardize
• What’s in a name?
What about PFAS, AFAS, FAE, ARBD, ARND, etc etc etc?

- Partial FAS, atypical FAS, fetal alcohol effect, alcohol-related birth defects, alcohol-related neurodevelopmental disorder, and all of the above without confirmed maternal alcohol exposure...
- Patients missing one or more of the four FAS criteria
- "FAE" has been retired: the alternatives aren't much better
- Fetal Alcohol Spectrum Disorders - an umbrella term, not a diagnosis
  - We have a marker for FAS: "the face"
  - We lack a specific neurobehavioral phenotype for FASD

The Current Problem

- Leading known cause of mental retardation
  - As common as Down Syndrome and spina bifida
- Incidence of FAS in general population from 1-3/1,000 live births, similar in Europe
  - Certain Native American groups: 10/1,000
  - King County foster care: 10-15/1,000
  - South African study: 45/1,000
  - Russian specialized orphanage: 14%
- $17,000/yr in medical costs (9x those without)
- Alcohol effects estimated 5-10x FAS

Who is at Highest Risk to Have Children with FASD?

- Characteristics of 80 FAS Mothers (Astley, '00)
  - Range of SES, but definitely skews lower
    - 61% didn't complete high school, 25% some college
    - 78% had household income <$10,000 at index birth
  - Race similar to WA state
  - Majority had multiple mental health disorders
  - ~50% reported physical/sexual abuse as children
  - 85% reported physical abuse as adults
  - Over 10% were dead at time of attempted contact

FAS Mothers, continued

- Mean age at birth of index child: 27 (18-41)
- Gravidity of index child: 3.3 (SD 1.9)
- % births unplanned: 73% (SD 33)
- % pregnancies exposed to alcohol: 76% (SD 30)
- Alcohol just before index pregnancy
  - Drinks per "drinking occasion": 18.5 (0-104)
  - Frequency: 44% daily, 30% one to sev x/wk
- Other substances around FAS birth
  - Tobacco 84%, Marijuana 17%, Cocaine/crack 12%

Alcohol Use in Pregnancy & FAS

- Alcohol is a teratogen
- Timing of use
- Dose of alcohol
- Pattern of use
- Individual risk/protective factors
Individual Factors

- Poorly understood
- Nutritional status, smoking, other drugs affect risk
- Only 10-40% births to chronic alcoholics have full FAS
- Discordant twin outcomes
- Maternal and fetal genetic factors

One Drink with Dinner?

- Alcohol is a known teratogen - “No safe amount of alcohol”
- No clear evidence that 1 drink or less per day has caused damage either, unless using averages
- Europe more permissive, up to 1 drink/day
- Transparency vs consistency of message, public vs private discussions

Fetal Alcohol Powers?

- Kelly et al. report that “light-moderate” prenatal alcohol exposure is not assoc with behavioral/cognitive deficits at 3 yo, 5 yo
- Actually, they do better than non-exposed kids?!?
- Don’t believe the media hype
- Major methodologic concerns

Diagnosing FAS ...

What EVERYONE Agrees On:
FASD involves ...

- Growth deficiency
- Facial anomalies
- Organic brain damage
- Alcohol exposure during gestation

What Everyone DISAGREES on in FASD
The DEFINITION of:

- Growth deficiency
- Facial anomalies
- Organic brain damage
- Alcohol exposure during gestation
**Growth Deficiency**

- Height deficiency (birth or since)*
- Weight deficiency (birth or since)*

*Not better explained by other influences on growth (chronic illness, severe malnutrition, etc)

**Facial Anomalies in FAS**

1. Small palpebral fissures
2. Smooth philtrum
3. Thin upper lip

Others are inconsistent and change with age

These probably don’t

**Facial Features Over Time**

<table>
<thead>
<tr>
<th>Birth</th>
<th>9 months</th>
<th>5 years</th>
<th>15 years</th>
</tr>
</thead>
</table>

**Sentinel Facial Features**

Palpebral fissure length

**Measuring Palpebral Fissure Length in Kids With Epicanthal Folds**

**Use of image analysis software**

Palpebral fissure length

Formula to compute true palpebral fissure length (PFL):

\[
\text{True PFL} = \left( \frac{\text{true pixel size}}{55.6 \text{ pixels}} \right) \times (\text{pixels in photograph})
\]

\[
\text{True PFL} = 2 \text{ cm} / 55.6 \text{ pixels} = 2 \times 1.07 = 2.5 \text{ cm}
\]
**Sentinel Facial Features**

**Philtrum & Lip**

No Smiling!

**Evidence of Brain Damage**

- Microcephaly
- "Hard" Neurologic signs, e.g. seizures
- Functional Evidence of Brain Damage

**An Ideal Photograph**

- Standard head position
- Reference measurement scale
- Neutral facial expression

**Use of image analysis software**

Upper lip circularity score
Other Organ Systems: ARBDs

- Eye: Myopia, strabismus, ptosis, optic nerve hypoplasia
- ENT: Hearing impairments, clefts, micrognathia, external ear anomalies, recurrent/chronic ear infections
- Cardiac: ASD/VSD, PS, PDA, AS, Tet, etc. Septal defects make up most of ARBDs in a recent study
- Renal: hydronephrosis, dys/hypoplastic kidneys
- Skeletal: clinodactyly, limited ROM, pectus, scoliosis, etc
- BUT - inconsistent definitions of FAS, no controlling for other risks, many not found in recent survey (overall, ARBDs 4x more likely in heavy 1st trimester PAE)

IOM (1996)
4-Digit Code (2004)
CDC (2004)
Canadian (2005)
Hoyme (2005)

Growth
- At least one.
  - Low weight for height
  - Low birth weight
  - Decelerating weight

4-Digit Code (2004)
- Pre- or postnatal height or weight \( \leq 10\% \)
  - Growth Rank 2, 3, or 4

CDC (2004)
- Pre- or postnatal height or weight \( \leq 10\% \)
  - Growth Rank 1

Canadian (2005)
- Pre- or postnatal height or weight \( \leq 10\% \)
  - Growth Rank 1

Hoyme (2005)
- Pre- or postnatal height or weight \( \leq 10\% \)
  - Growth Rank 1

Face
- Characteristic pattern that includes features such as:
  - Short PFL
  - Flat upper lip
  - Flattened philtrum
  - Flat midface

4-Digit Code (2004)
- All 3 required
  - (Face Rank 4)
    - PFL \( < 3\% \)
    - Philtrum (Rank 4 or 5)
    - Lip (Rank 4 or 5)

Canadian (2005)
- All 3 required
  - (Face Rank 4)
    - PFL \( < 3\% \)
    - Philtrum (Rank 4 or 5)
    - Lip (Rank 4 or 5)

Hoyme (2005)
- All 3 required
  - (Face Rank 4)
    - PFL \( < 3\% \)
    - Philtrum (Rank 4 or 5)
    - Lip (Rank 4 or 5)

CNS
- At least one bulleted feature:
  - Structural
    - OFC \( < 3\% \) (microcephaly)
  - Abnormal structure and/or neurological
  - Hard/soft signs

4-Digit Code (2004)
- All 3 required
  - Structural (Brain 4)
    - OFC \( < 3\% \)

Canadian (2005)
- All 3 required
  - Structural (Brain 4)
    - OFC \( < 3\% \)

Hoyme (2005)
- All 3 required
  - Structural (Brain 4)
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    - OFC \( < 3\% \)
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    - Hard/soft signs

4-Digit Code (2004)
- All 3 required
  - Structural (Brain 4)
    - OFC \( < 3\% \)
    - Abnormal structure

Canadian (2005)
- All 3 required
  - Structural (Brain 4)
    - OFC \( < 3\% \)
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Foster Care Screening (Astley, '02)
- 600 kids entering foster care in King County screened with photos, OFC
  - 10 screened positive->clinic for confirmation
- Nested into preexisting system (Health and Education "passport")
- Even screen-negative subjects assessed
- Photo analysis was amazingly accurate - 100% sensitive, 99.8% specific
- Children and families benefited
FASD and the Brain

We're poised to better understand the cognitive and behavioral features of fetal alcohol spectrum disorders.

As pediatricians (and parents!) we're all searching for ways to understand...

what's "normal" and what's not
what's FAS and what's something else
what's "WONT" and what's "CAN'T"

The brain injuries in FASDs often go undiagnosed and unserved

It's not the face that needs services
FASD is often an "invisible" disability

Children often fail to qualify for services until later school years - too late!

Variability is the rule - for the fetal alcohol spectrum and for the child.

University of Washington FASD MRI/S fMRI Study

Primary Hypothesis:
- The following will differ between children with FAS/D and typically developing children:
  - neuro-structure (size of specific brain structures)
  - neuro-function (as measured by psychological, psychiatric, and fMRI assessment)
  - neuro-chemistry (neurometabolites: choline and n-acetyl-aspartate)
- Are the neuro-structural, neuro-functional and neuro-chemical impairments inter-correlated?

Long-range Question:
Can MRI, MRS, and/or fMRI improve the accuracy of an FASD diagnostic evaluation?

Variability: Intra-Child "Scatter"

Variability is the rule - for the fetal alcohol spectrum and for the child.

Typical school IQ and achievement testing would miss this ...

Our Study Population

Four groups of school-age children (8-15 years old):
1. FAS / Partial FAS (n = 20)
2. Static Encephalopathy / Alcohol Exposed (n = 24) - "Severe ARND"
   Brain 3 or 4 - brains as affected, but didn't have the face
3. Neurobehavioral Disorder / Alcohol Exposed (n = 21) - "Mild ARND"
   Brain 2 - alcohol exposed, but brains much less affected
4. Controls - normal to high-functioning, no/minimal alcohol exposure (n = 16)

Strengths:
- strictly defined diagnoses
- covers the spectrum of alcohol impacts
- large number of subjects
- very thorough imaging AND neuropsych testing
Key Structural Findings - Overall

- All brain regions become progressively smaller (absolute size) as you advance across the 4 Diagnostic Groups from Group 4 (Controls) to Group 1 (FAS / PFAS).

The more severe the facial features, the more severe the impairment in brain function.

Some functional impairments correlate with reduction in specific brain regions.

Frontal Lobes - The Brain CEO

- Higher cognitive, executive functions
- Social/emotional regulation
- Reduced frontal lobe growth in FASDs
- Stress/trauma/neglect all can impair the development of the frontal lobes as well

Key Structural Findings - Frontal Lobes

- The frontal lobe is significantly smaller in FAS/PFAS relative to "Severe ARND", "Mild ARND", and Controls, even after adjustment for overall brain size.

Executive Function - The Brain CEO

- Executive function (EF) underlies many realms of adaptive behavior
- Independent of overall intelligence
- EF develops later, and continues to mature into early adulthood ...

This is a good and a bad thing
Caudate - The Executive Assistant

• Has lots of connections to the frontal lobes, especially executive function areas
• Acts as the secretary to the CEO, passing info to the frontal lobes
• Seems especially sensitive to alcohol
• Relatively smaller in many FAS imaging studies, including ours …

Hippocampus - The File Clerk

• Helps integrate new explicit memories
• Innervated by cholinergic neurons from the caudate
• Absolute but not relative volume decreased across our groups
• Perhaps more vulnerable to later alcohol consumption
• Also impacted by prolonged stress and trauma

Corpus Callosum - Middle Management

• Low-hanging fruit of FAS brain imaging
• Complex shape and size differences reported in FAS, but …
• Our MRI data doesn’t support this (relatively larger in FAS)
• Absent corpus callosum rare in our clinic population

Key Structural Findings - Corpus Callosum

• The corpus callosum becomes progressively larger, after adjustment for overall brain size, as one advances from the Control to the FAS/PFAS group. Hmm …

Midline Cerebellum

• Balance and coordination
• Also may influence learning, attention, mental health
• Calms the limbic system?
• Anterior midline vermis especially affected by prenatal alcohol
• Sleep architecture

Sleep and FASDs

• 30-50% of our patients have problems with sleep
• Rate of sleep disorders goes up with alcohol exposure
• Alcohol affects circadian rhythms, the body clock
• Facial anomalies and low tone make them high risk for obstructive sleep apnea
• The midline cerebellum controls response to cardiorespiratory stressors
• We see disturbed sleep architecture, less REM sleep
• Poor sleep can mimic or worsen ADHD and other daytime cognitive and behavioral issues
• Have a low threshold for sleep clinic referral
Stress, Neglect & Maltreatment

- Before we blame it all on drink & drugs... don't forget other influences, like "complex trauma"
- Prenatal stress and anxiety may also affect postnatal stress response, IQ, ADHD, sleep
- Early childhood maltreatment can impact amygdala, hippocampus, corpus callosum, frontal lobes, cerebellar vermis
- Sound familiar?

Summary - What are we finding?
So far we've seen:
- Significant neuropsychometric contrasts between all 4 study groups
- Those with the face of FAS are indeed more impaired
- The entire brain is smaller in affected kids
- But certain key structures are even smaller...
- The brain structural, chemical, and functional analyses fit together, and match the emerging literature on alcohol damage and brain development
- Stay tuned for lots more analysis...

Diagnosis may get more accurate
- Neurology guidelines for global development recommending MRI
- Neuroimaging will likely serve as a more sophisticated "head circumference"
- It may also suggest specific testing and interventions
- New standards are being published for normal pediatric MRI/S values
- But MRI alone cannot diagnose FASD (non-specific)

Our findings fit recognized cognitive and behavioral profiles of FASDs
- Overall Cognition
- Memory
- Language
- Visual Motor Skills
- Academics
- Executive function
- ADHD
- Sensory integration
- Motor skills
- Behavior
- Social Skills
- Adaptive Functioning
- Sleep

A hallmark of the fetal alcohol spectrum is variability, but the next slides contain some generalizations about potential strengths and weaknesses

Newborns with FAS
- Small, microcephalic?
- Facial features present but trickier to assess
- Often with poor state regulation, irritability, disturbed sleep, feeding difficulties, disorganized attachment
- Can be a terrible "fit" - impaired parent with a very difficult infant
FASD in Preschool Years

- Often missed, unserved at this stage
- Language, adaptive, gross and fine motor delays
- Difficulty regulating emotions and behavior; extreme tantrums
- Overactive, impulsive, inattentive
- Slow to learn social boundaries
- Tools of the Mind program might be helpful

Overall Cognition

- Strengths
  - Tested intelligence is usually not in retarded range
    - This can also be a liability, services-wise
- Weaknesses
  - IQ scores lower than expected based on genetic potential
  - FAS mean IQ ~ 66 to 80 in various studies
  - FASD more variable
  - Lots of sub-test variability, verbal IQ vs performance IQ “splits”
  - Slower information processing
  - Nonverbal abstract reasoning

Types of Memory

- Short-term memory (inability to recall series of directions)
- Long-term memory (inconsistent retrieval of learned information and skills)
- Source memory deficit (confuse what they were asked to recall with random info)
- Explicit (conscious) memory affected, implicit (unconscious) memory tends to work

Memory

- Strengths
  - Recall of single word vocabulary and categorical labels
  - Recall of visual and kinesthetic patterns
  - Often capable of retaining verbal information (especially if words are in a rhythm or song)
- Weaknesses
  - Impaired verbal learning
  - Auditory sequential memory
  - Limited working memory span
  - Integration and retrieval of information and concepts

Language

- Strengths
  - Superficial conversational speech - talkative and fluent
  - Ability to learn vocabulary and comprehend single words
- Weaknesses
  - Comprehension in complicated discussions and explanations (especially out of context)
  - Language less complex, more superficial, more literal than peers
  - Comprehension scores less than expressive
    - “They talk better than they understand”
    - Understanding directions
  - Social communication deficits

Visual Motor Skills

- Strengths
  - Use of color
  - Sculptural abilities
  - Ability to make direct copies - especially of simple geometric forms
- Weaknesses
  - Visual spatial organization
  - Making creative, complex drawings
  - Handwriting
Academic Skills

- **Strengths**
  - Decoding words and oral reading
  - Spelling skills
- **Weaknesses**
  - Reading comprehension
  - Story, essay and report writing
  - Arithmetic skills
  - Math reasoning
  - Organization and study skills
  - Academic achievement lower than IQ would predict

FASD Executive Functioning Deficits

- **SELF-REGULATION**
  - The ability to stay in control of emotions ("hot EF"); awareness of how others perceive you; use of self-talk strategies to monitor self and behavior
- **SEQUENCING OF BEHAVIOR**
  - Knowing when and how to start an activity, keeping track of what to do next, initiating tasks
- **FLEXIBILITY**
  - The ability to shift tasks smoothly, accept change, deal with transitions appropriately, absence of rigidity

Executive Functioning Deficits (continued)

- **RESPONSE INHIBITION**
  - Lack of impulsivity, ability to inhibit first "knee-jerk" response to difficult situations and think before acting
- **PLANNING**
  - The ability to use mental and action steps to complete tasks, to anticipate what is needed to complete tasks, related to sequencing of behavior
- **ORGANIZATION**
  - The ability to keep one's self and materials organized, in order, predictable, etc.

Attention and Behavior

- Behavior regulation/mood swings
- Easily overwhelmed by stimulation
- Obsessive and perseverative features
- Problems with visual and auditory attention
- Variations on ADHD - many will receive the diagnosis, but response to meds is variable
- Risk of ADHD goes up with increasing alcohol exposure (50% if Rank 4, 30% Rank 3, 15% rank 2 in several FAS clinics ... Bhatara et al.)

Sensory and Motor Issues

- Poor balance and coordination
- Other "soft neurologic signs"
- Visual-spatial motor skill difficulties
- Sensory over-sensitivities and sensation-seeking

Athletic Skills

- **Strengths**
  - Individual sports requiring strength and endurance
- **Weaknesses**
  - Team sports with demands to listen, follow directions, understand rules, and sequences and memorize procedures
Social and Behavioral Skills

- **Strengths**
  - Likable, friendly, engaging and often kind
  - Not necessarily “syndromic” in appearance

- **Weaknesses**
  - Poor impulse control
  - Emotionally labile
  - Lack of understanding of personal boundaries
  - Naïve, gullible—often become a “victim”

Social/Adaptive Functioning

- Social and adaptive skills often delayed, and may be half their chronologic age
- This gap WIDENS as they grow older
- Perform better in small, highly structured environments, with range of ages
- Big public high schools - OY
- SAFETY! Easily victimized ...

SECONDARY DISABILITIES

Are the consequences of the primary disabilities, and arise from a gap between expectations and abilities

- Disrupted school experiences
- Trouble with the law
- Mental health problems
- Alcohol & drug abuse
- Being homeless
- Having children you can’t care for

These are what we hope to prevent

FAS in Adulthood

![FAS in Adulthood Graph]

Secondary Disabilities from Streissguth et al, 1996

Interventions for FASD

- Early diagnosis and intervention
- A caregiving environment (in middle childhood) that is:
  - Nurturing, stable
  - Appropriately structured & stimulating
  - Geared to the child’s developmental needs
  - Free from caregiver substance abuse
  - Safe from violence
- Appropriate social services

Protective Factors Against Development of Secondary Disabilities

- Early diagnosis and intervention
- A caregiving environment (in middle childhood) that is:
  - Nurturing, stable
  - Appropriately structured & stimulating
  - Geared to the child’s developmental needs
  - Free from caregiver substance abuse
  - Safe from violence
- Appropriate social services

(Adapted from Streissguth et al., 1996)
### Intervention Research - Bertrand '09

- **Self-Regulation & Sensory Strategies**
  - The Alert Program (Children's Research Triangle)
- **Learning How to Learn - "Cognitive Habilitation"**
  - Math Interactive Learning Program (Marcus Institute)
- **Social Skills Interventions**
  - Children's Friendship Training (UCLA)
- **Behavioral Support**
  - Families Moving Forward (UW research)
- **PCIT vs Parent Support and Management**
  - University of Oklahoma

### Key Points for Caregivers and Professionals

- FASDs are too often an "invisible disability"
- Refer alcohol-exposed kids for early evaluation
- Thorough testing is so important
- Individualized, longterm interventions
  - Reframe behaviors, adjust expectations and the child's environment
  - Behavioral consultation, self-regulation, social, learning to learn
  - Targeted medication evaluations
  - Anticipate adolescent and adult transitions
- For caregivers
  - Education, support groups, linkage, school advocacy, respite

### Online FASD Resources

- **UW Publications, Diagnostic Tools, Guides and Training Programs**
  - [www.fasdpn.org](http://www.fasdpn.org) (including an online course in 4-Digit Code)
  - [depts.washington.edu/fasdpn/htmls/literature.htm](http://depts.washington.edu/fasdpn/htmls/literature.htm)
  - [www.adoptmed.org](http://www.adoptmed.org)
- **Other Online Resources**
  - [www.cdc.gov/fasd/](http://www.cdc.gov/fasd/)
  - [www.fascenter.samhsa.gov/](http://www.fascenter.samhsa.gov/)
  - [www.nofas.org/](http://www.nofas.org/) (with national resource directory)
- **Teaching Students with FASD**
  - [www.education.gov.ab.ca/k_12/specialneeds/fasd.asp](http://www.education.gov.ab.ca/k_12/specialneeds/fasd.asp)
- **FAS - A Guide for Living: Parenting Children with FASD**

### Thank Yous

- Susan Astley, PhD
- Julia Bledsoe, MD
- Julie Gelo
- Heather Carmichael Olson, PhD
- Allison Brooks, PhD
- The FAS Clinic Team
- The FAS MRI/MRS/fMRI Study Team
- The Families Moving Forward Study Team
- Our Clinic and Study Families