Foetal Alcohol Spectrum Disorder: Living in the here and now

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Agenda

• Review of general FASD presentation
• Organic brain damage in FASD
• Assessing and diagnosing: the neuropsychological profile in FASD
• The importance of timely diagnosis
• Behavioural management
Foetal Alcohol Spectrum Disorder

- **Foetal Alcohol Syndrome (FAS):**
  - Confirmed (strongly suspected) alcohol usage in pregnancy
  - Associated physical/facial features
  - Associated neurocognitive effects

- **Partial Foetal Alcohol Syndrome (PFAS):**
  - Confirmed (strongly suspected) alcohol usage in pregnancy
  - *Some* associated physical/facial features
  - *Some* associated neurocognitive effects

- **Alcohol Related Neurodevelopmental Disorder (ARND):**
  - Confirmed (strongly suspected) alcohol usage in pregnancy
  - Absence of associated physical/facial features
  - Associated neurocognitive effects
  - *This diagnosis is most commonly present, and also most commonly missed*
Typical Facial Features

- Microcephaly
- Flat mid-face
- Smooth philtrum
- Thin upper lip
- ‘Train track’ ears
- Short palpebral fissures
- Sometimes small nose

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Alcohol is a Teratogen:

- Halts neuron development and disorganises neural crest migration in the developing foetus
- Characteristic dysmorphic facial features (timing effect is literally days; seen also in mouse studies)
- Growth problems
- Behaviour problems
- Learning problems (specific cognitive profile)
- Speech and language problems
- Sensory integration problems
- Physical problems (heart, kidney, eyes, hearing etc.)
Neural crest migration in the embryo with increasing concentrations of alcohol exposure: as concentration increases, migration is halted; other cells migrate in a disorganised fashion and to the ‘wrong places’

Alcohol exposure disrupts neural crest migration. (A) Both slug and twist1 in situ hybridization, performing along with gradient alcohol treatment, displays neural crest migration is accordingly blocked by the increasing concentration of alcohol. (B,G) indicates 0.5% alcohol treatment still allows fair migration of neural crest versus control embryos (A,F). 1.0% alcohol treatment deteriorate the egressing process of neural crest, however, there is perceivable migration occurring (C,H). Finally, both 1.5% and 2.0% appears completely freezing the migration (D,E,I,J). Shi et al. Molecular Brain 2014 7:67 doi:10.1186/s13041-014-0067-9
Reconstructed Mouse embryos at 14 days gestation

Figures ‘b’ and ‘d’ were exposed to alcohol at day 7
Alcohol and Myelin

• Clinically delayed myelination has been observed
  – Caused by delayed expression of myelin basic protein (MBP) and transferrin

• Effect is decreased speed of neural processing
  – Which in turn leads to slow cognitive processing
Prerequisites for Diagnosis

1. More than minimum alcohol exposure
2. Impaired neurocognitive functioning
3. Impaired self regulation
4. Impaired adaptive functioning
5. Childhood onset of problems
6. Not better explained by any other condition

FASD is a diagnosis of exclusion as well as inclusion

(Systematic review found 298 conditions associated with pre-natal alcohol exposure - Mukherjee et al)
Dose Dependent Relationship

- High levels = high risk
- Low levels = lower risk
- NO ALCOHOL = NO RISK
- Problem... define ‘low’..
null
Other factors in the pregnancies of many children with FASD

• Smoking *(size, brain stem, dopamine and other neurotransmitter circuits, ADHD)*
• Opiates *(dopamine and ACH in limbic system)*
• Cocaine *(frontal dopamine circuits)*
• Cannabis

*Compound effects:*
Alcohol + smoking + cocaine = ?? FASD ++
The added factor of neglect in looked after/adopted children

• Many FASD children who come into care, have had a neglectful early start in life
• Neglect is associated with chronic difficulties in behaviour, emotional issues, cognitive problems..

*Compound effect:*

FASD + other substances + neglect = Your child
FASD: An invisible but significant Disability

• Many children with FASD do not have the associated facial features
• Often very verbal with a normal IQ
• But they have **Organic Brain Damage:**
  – They cannot process information normally
  – Their damaged frontal lobes mean they have an executive functioning disorder
  – Therefore using traditional behaviour management techniques aimed at neuro-typical children rarely works
  – Children and their families are so often set up to fail through being sent on inappropriate parenting courses by well-meaning professionals
Variability in visible brain damage

- Most MRI scans will ‘appear’ normal
- MRI is not a useful means of diagnosis
- **Most common findings on MRI:**
  - Volume loss of corpus callosum
  - Frontal and parietal lobes most affected: EF and dorsal stream defect (‘where’). Ventral stream usually less impaired (‘what’)
  - Hippocampus volume reduction
  - Basal ganglia volume reduction
  - Overall loss in brain volume/size
- **Most common SEVERE effects:**
  - Complete agenesis of corpus callosum
  - Schizencephaly / lissencephaly
  - Hypoplasia of cerebellum

Structural MRI does not measure white matter integrity / regional connectivity
Normal MRI does not always mean no significant damage in FASD
MRI Scans of four children

A = typically developing 10 year old boy
B = 11 year old boy with FASD
C = 7 year old girl with FASD
D = 14 year old boy with FASD

NOTE:
- The variability in brain structures in children with FASD
- Changes in volume and shape of the corpus callosum which affects hemispherical connectivity and integrity (red arrow)
- Changes in volume and shape of the cerebellum (yellow arrow)
- General reduction in brain volume/underdevelopment of cortical structures
- Reduced white matter integrity not visible

A typical FASD cognitive profile

- Often verbal ability is lower than non-verbal ability
  - (Expressive language usually higher than receptive language)
- IQ can be normal, or there can be general learning disability (most common = IQ 70’s /80’s)
- Executive functioning is always impaired:
  - Hot / cold executive functions
  - Working memory
  - Attention skills
  - ADHD symptoms of hyperactivity, impulsivity, inattention and sensory-seeking behaviour
  - Impaired social functioning > vulnerability
Executive Functioning: A Developmental Trajectory

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If Executive Functioning Disorder is not recognised and understood – particularly in school...

A child can be incorrectly labelled as:

- Lazy
- Unmotivated
- Disorganised
- Oppositional
- Wilful
- Non-compliant
- Poorly parented
- Generally naughty
Problems associated with EF impairment often begin in the first couple of years of school

- Easily influenced by others
- Difficulty understanding cause and effect and predicting consequences
- Difficulty learning from past mistakes
- Appearing capable but with less actual ability behind this (confuses and frustrates teachers)
- Difficulty separating fact from fiction
- Temper tantrums, stealing, lying, disobedience, defiance of authority
- Poor understanding of social rules
Even with a normal IQ, children with FASD often lack the core skills required to learn and succeed:

- Sitting still
- Listening without distraction
- Sustaining attention
- Understanding cause and effect
- Following complex verbal instructions (due to receptive language difficulties and EF)
- Planning
- Organising their time/belongings/school projects
‘If he would just try harder, he could do it!’

• Performance always fluctuates
• Many children with FASD are trying to the point of exhaustion
• Constant hyper arousal... meltdowns... more negative labels... further knock-on effect on self esteem
Social Functioning Impairment

Normal facial expressions, good eye contact, non-verbal communication in tact, but..

- Difficulty providing the right information to communicate
- Difficulty adapting social skills with increasing age
- Problems understanding other’s behaviours and motivations (which are often not what they outwardly portray)
- Difficulty learning from previous experience
- Problems recalling consequences of past actions
- Extremely socially vulnerable
- Often misdiagnosed as atypical autism (can be hard to differentiate)
An 18 year old with FASD
Multiple diagnoses are common

- FASD is a great ‘mimicker’
- Shared executive functioning deficits
- Shared social issues
- Shared hyperactivity and inattention
  - ASD
  - ADHD
  - Conduct Disorder
  - Dyspraxia
  - General learning disability
  - Attachment disorder
  - Environmental (neglect)
  - Borderline personality disorder
  - Sensory processing disorder
## ASD vs. (‘pure’) FASD

<table>
<thead>
<tr>
<th>ASD</th>
<th>FASD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal eye contact</td>
<td>Normal eye contact</td>
</tr>
<tr>
<td>Limited facial expression/limited face processing</td>
<td>Normal facial expression/limited face processing</td>
</tr>
<tr>
<td>Often disinterest in peers</td>
<td>Normal interest in peers</td>
</tr>
<tr>
<td>EF deficit (usually milder)</td>
<td>EF deficit (more severe)</td>
</tr>
<tr>
<td>Difficulty initiating social interaction (and poor quality if accomplished)</td>
<td>No difficulty initiating social interaction (although quality often poor)</td>
</tr>
<tr>
<td>Sharing affect impairment</td>
<td>No impairment sharing affect</td>
</tr>
<tr>
<td>Impaired general non-verbal communication</td>
<td>Normal non-verbal communication</td>
</tr>
</tbody>
</table>

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FASD with associated ADHD

• 74% of FASD children meet the criteria for ADHD (Roozen et al, 2016)
• ADHD is itself a disorder of executive functioning
• Individualised approach to treatment:
  – Systemic formulation required
  – Pharmacological treatment effective for FASD-associated ADHD (Young et al, 2016)
  – In conjunction with appropriate behaviour management strategies
  – Sensory integration therapy can also be beneficial
  – Sleep onset difficulties common: melatonin
• The diagnosis should not be ‘ADHD AND FASD’ - it is ‘FASD with associated ADHD’
Diagnosis changes prognosis

- Accurate diagnosis by age 8
- Stable and nurturing home for >72% of life
- Stability between ages 8-12 critical (when educational difficulties really kick in)
- Receiving appropriate early education
- Being eligible for SN services
- (Prognosis is inversely related to number of placement moves for those in care)
Other predictors of best prognosis:

- Lynch and Kable (2016):
  - Full term birth
  - Being female
  - Fewer dysmorphic facial features
  - Higher IQ
  - Less aversive life events in childhood
  - Parental warmth
  - Fewer siblings
• Children with FASD often do not respond like a typically developing child

• “Like trying to navigate round London with a map of Birmingham” (*parent of child with FASD*)
Traditional Behaviour Management

• For these techniques to work, a child must:
  – Understand the concept of ‘future earning’ / deferred gratification
  – Have the impulse control to change or inhibit behaviour in future situations
  – Understand cause and effect
  – Have some understanding of their impact on others
  – Be able to monitor and regulate emotional response
  – Have some concept of time

• The typical child with FASD often does not have these skills
So the following are rarely successful:

- Tokens
- Stickers
- Star charts
- Money
- Removing possessions
- Cancelling trips and treats
- Time out
Prevention is key..

- Children with FASD are easily over-whelmed by their environment (sensory integration)
- Often it is environmental and external factors that trigger meltdowns
- Pre-planning and analysis of triggers – hard work but effective
- Preparation: photographs of people and places to prepare child for upcoming events
- Consistency and routine
- Reassurance and repetition ++
MELTDOWNS: Reducing risk of sensory overload

- Sensory overload is a key trigger for FASD children
- Difficulty filtering and screening out background interference
- Avoiding busy and loud areas where possible
- Teaching self soothing techniques
- Factoring ‘explosion’ times – during/after school ++
- Building in movement breaks at school and home
- Distraction breaks in the classroom – can be masked by ‘doing a little job for the teacher’
Too many choices

• To make a choice one needs to hold two or more things in mind at the same time and compare them. This requires working memory. Working memory is generally impaired in FASD.

• Closed and limited choices from two physically available objects (as opposed to verbal/abstract concepts)
Transitions

• Common difficulty
• Can become stuck on one activity and find it hard to move to another (part of executive functioning deficit)
• Visual schedules can be helpful
• Sand timers for screen time, bath times etc. for younger children
‘Quiet time’ – not ‘time out’

• Time out will not help a child with FASD learn to self regulate. It can exacerbate the situation as they are often unable to calm down by themselves.

• Find a calm cosy place for ‘quiet time’ where a child can go when becoming overwhelmed. Do not make it feel like a punishment.

• Expressing anger in a safe way is ok (bouncing on trampoline, throwing a bean bag, hitting a pillow)

• This strategy may over time help a child to learn to better manage their emotions.

• No matter how frustrating the situation is... shouting and losing the plot will only prolong it.. !

• As parents and teachers, we need to be Bigger.. Stronger.. Calmer and Containing. (Even when we don’t feel any of those things!)
CIRCLE OF SECURITY
PARENT ATTENDING TO THE CHILD'S NEEDS

I need you to...

Watch over me
- Delight in me
- Help me
- Enjoy with me

Support My Exploration

Always: be BIGGER, STRONGER, WISER, and KIND.
Whenever possible: follow my child’s need.
Whenever necessary: take charge.

SECURE BASE

SAFE HAVEN

- Protect me
- Comfort me
- Delight in me
- Organize my feelings

Welcome My Coming To You

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When prevention didn’t work..

During a meltdown..

• Get down on the floor/under the bed/under the table.. Calmly and firmly label the emotion: ‘It looks like you are feeling cross/angry/scared..’ (this is vital in general)
• Make sure you know what you are going to do next to sooth them, which is different for all children: firm hold, light touch, singing, or no contact at all
• When calm, cuddle them and then distract
• Remember that learning from past mistakes is a challenge for children with FASD (frontal lobe damage)
• Reinforce positive behaviour through repetition and praise
Communication

• Many children with FASD can ‘talk the talk’. Which masks their underlying difficulties with receptive language

• Imagine being at a party where you cannot filter out anyone’s conversation at any point..

• At school: what is often reported as ‘won’t follow instructions’ is more likely to be ‘can’t’. (language comprehension, working memory, executive functioning).
School: Learning Needs:  
*Strategies not Solutions Document*

- **Hyperactivity**: calmer learning environment, lighting, reduction of clutter
- **Short attention span**: short key information, carrying word instructions, language clear and concise
- **Erratic moods**: personal space, support, praise, work on feelings (nurture groups)
- **Poor working memory**: visual schedules and timetables, repetition
- **Poor social skills**: scripting, social stories, role play
- **Sensory integration difficulties**: movement breaks, bear caves and rabbit holes
- **Poor instruction retention/poor planning**: task breakdown, visuals, hooks for previous learning, repetition, time.
Friendships and Social Life

• A child with FASD can struggle with maintaining friendships, and may need support with this
• Due to difficulties initiating suitable interaction, encourage common topics for discussion and sharing
• Support sustaining a friendship by sharing hobbies
• The child with FASD is likely to need continual rehearsal for new situations for socially acceptable behaviours, so treat each new set of circumstances as unknown, e.g. school trips, birthday parties and holidays.
Social Immaturity and Vulnerability

• Remember to halve an affected child’s chronological age: this is where they are functioning socially and emotionally
  – Pitch your expectations at this level

• Social vulnerability with peers > need for supervision
• Role playing
• Ownership and stealing
• Lying and fabrication
As carers/teachers of an FASD child it is important to..

- Be their ‘external brain’
- Remember your child will not fully understand the link between consequences and behaviour, even if they outwardly appear to
- Remember that repetition is key to learning
- Understand they will struggle to transfer learning a skill from one environment to another... hence the repetition
- Supervise to the level of half their age
- Accept that there are some skills they may never learn and may always need support with.
The Future

- FASD is a spectrum
- Independent vs. interdependent living
- EF and IQ as predictors
- EHCP / adult services
Thank you for listening

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