

The Centre For FASD



Managing Foetal Alcohol Spectrum Disorder

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The Neuropsychology of FASD





Agenda

- Overview of FASD
 - What alcohol exposure does to the brain
 - Prognosis factors
 - Why getting the right diagnosis is important
- 



'Foetal Alcohol Spectrum Disorder is an umbrella term

- ▶ 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 common, and also easiest to miss/mis-diagnose*

FASD: Typical Facial Features

- Microcephaly
- Flat mid-face
- Smooth philtrum
- Thin upper lip
- 'Train track' ears
- Epicanthal folds



Facial Features (and brain abnormalities) are not species specific

- Reconstructed mouse embryos at 14 days' gestation.
- Figures 'b' and 'd' were exposed to alcohol at day 7





Alcohol is a Teratogen

- Disordered or halted neural crest migration
 - Delayed myelination
 - Characteristic facial features
 - Growth problems
 - Characteristic neurocognitive profile
 - Behavioural challenges
 - Speech and language problems
 - Sensory integration problems
 - Physical problems (heart, kidney, eye and ear most common)
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MRI Findings

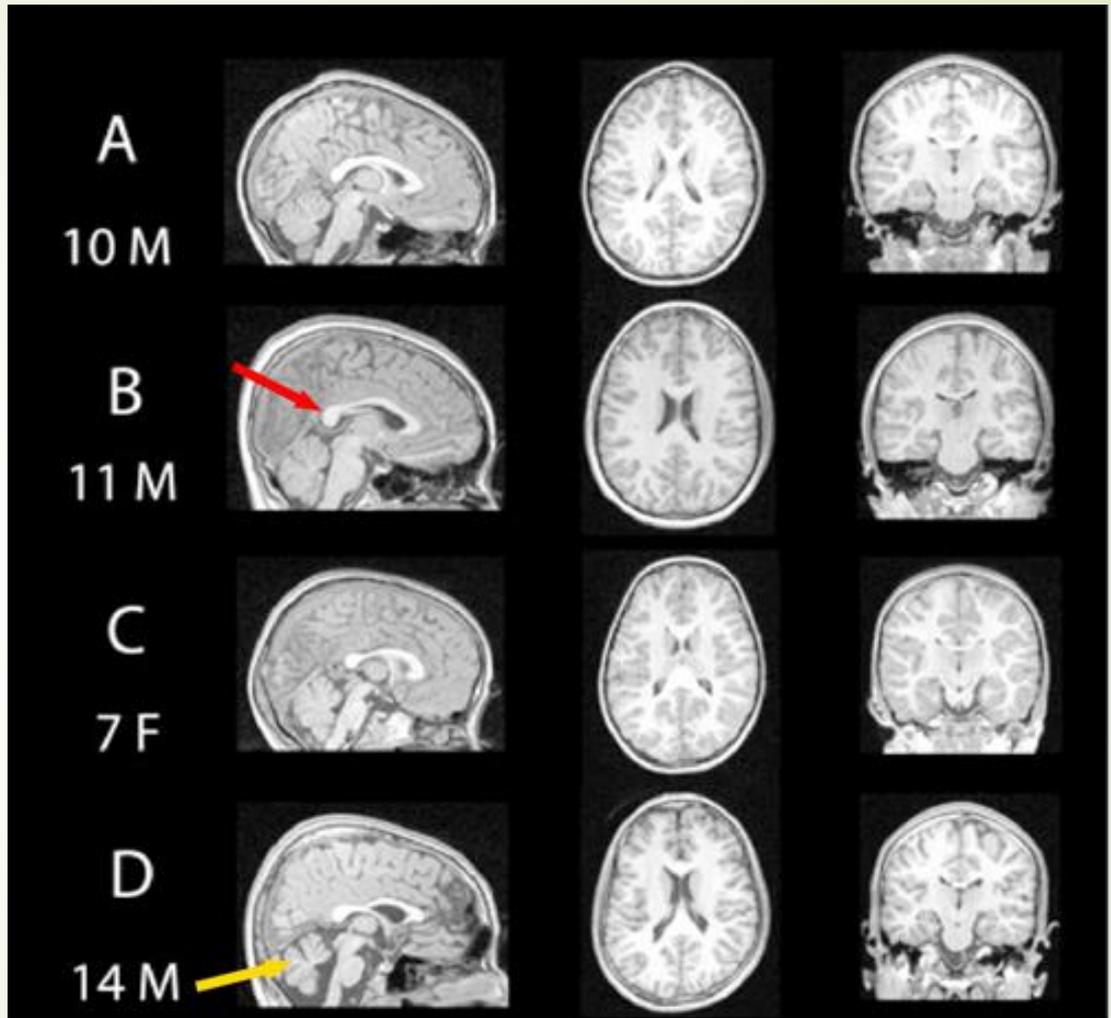
- ▶ **Most common findings on MRI:**
 - ▶ Volume loss of corpus callosum
 - ▶ Frontal and parietal lobes most affected: EF and dorsal stream defect ('where').. Ventral stream usually less impaired ('what')
 - ▶ Hippocampus volume reduction
 - ▶ Basal ganglia volume reduction
 - ▶ Overall loss in brain volume/size
- ▶ **Most common SEVERE effects:**
 - ▶ Complete agenesis of corpus callosum
 - ▶ Schizencephaly / lissencephaly
 - ▶ Hypoplasia of cerebellum
- ▶ **Many MRIs are reported as normal.. But..**
 - ▶ Structural MRI does not show up loss white matter integrity and/or regional connectivity
 - ▶ Normal MRI does not mean 'no damage'
 - ▶ MRI is not necessary or useful for diagnosis of FASD

MRI examples

- A = unaffected 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 across children
- Changes in volume and shape of the corpus callosum - affects hemispherical connectivity (red arrow)
- Changes in volume and shape of the cerebellum (yellow arrow)
- General reduction in brain volume/underdevelopment of cortical structures



Sowell, E.; Nunez, S.; Roussotte, F. Structural and functional brain abnormalities in fetal alcohol spectrum disorders, *Alcohol Research & Health*, in press.

Dose Dependent Relationship

- High levels = high risk
- Low levels = lower risk
- NO ALCOHOL = NO RISK

- Problem... define 'low'..

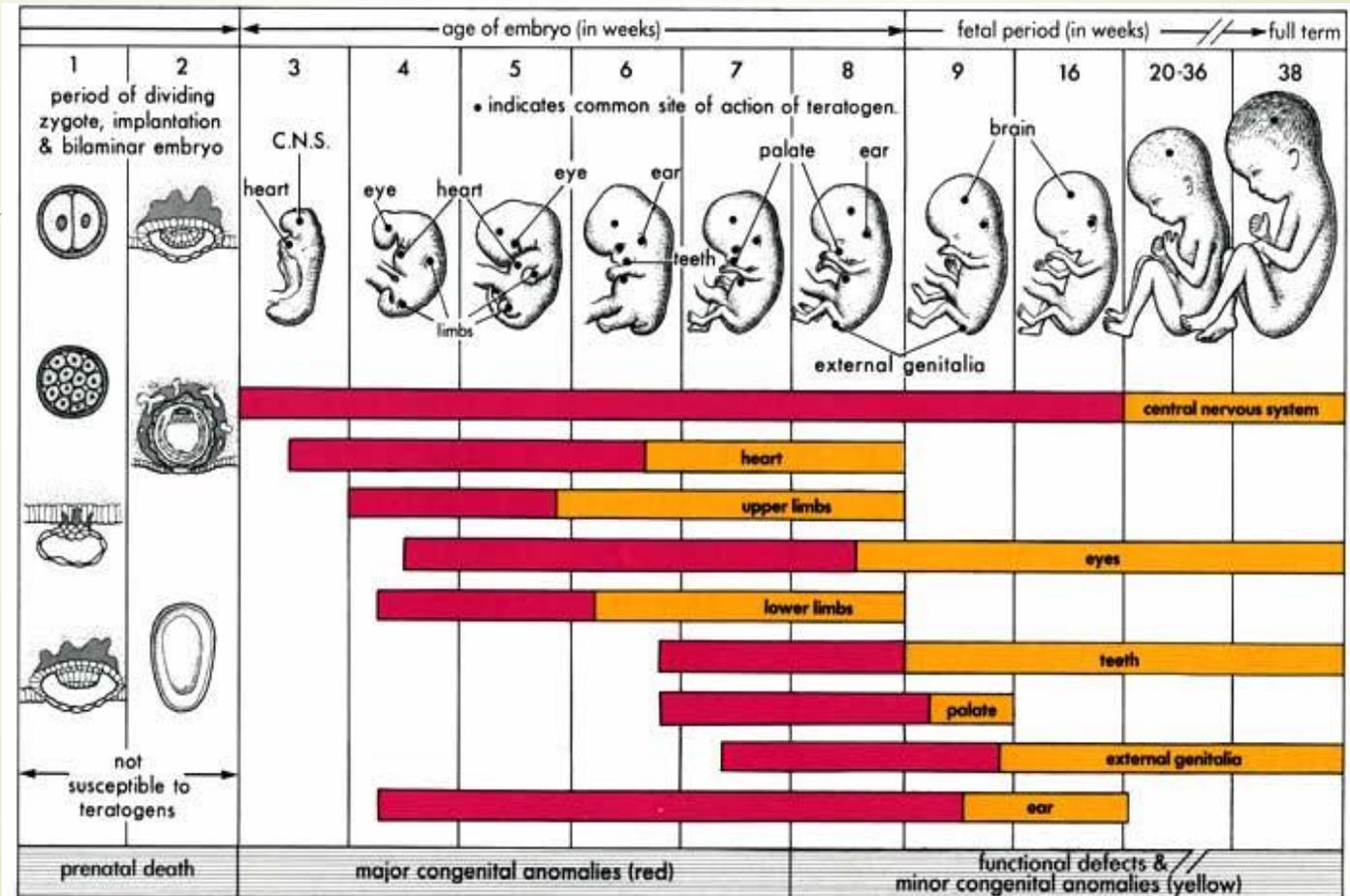




Alcohol affects every foetus differently

- ▶ Hemingway et al (2019)
- ▶ The effects of teratogens like alcohol can be modified by genetic differences in foetal susceptibility and resistance
- ▶ Twin study – twins with identical alcohol exposure present with FASD conditions at opposite ends of the spectrum (even facial features and growth factors)
- ▶ More likely to see differences in presentation in non-identical twins. Presentation more similar in identical twins.

There is no good time to drink during pregnancy





The Importance of Neuropsychological Assessment

- Used in conjunction with medical assessment for diagnosis
- The neuro-cognitive impairment in FASD is one of the most significant aspects of the diagnosis
- Neuropsychological assessment provides prognostic indicators when considered *alongside* knowledge general adaptive functioning
- Vital for planning future needs/support



'Textbook' Neurocognitive Profile in FASD

- ▶ Verbal IQ is frequently lower than nonverbal IQ
 - ▶ Inference, abstract reasoning, receptive comprehension
- ▶ IQ, while variable, most typically:
 - ▶ ARND = 80s
 - ▶ FAS = 70s
 - ▶ Essentially borderline, and therefore falling between gaps in educational provision
- ▶ Working memory weakness or impairment
- ▶ Episodic memory often more spared
- ▶ Significant executive Functioning Impairment demonstrated in:
 - ▶ Neuropsychological assessment
 - ▶ Reports by parents and teachers
 - ▶ Difficulties with peer relationships
 - ▶ General adaptive functioning for chronological age



Executive Functioning:

The managing director of the brain

Hot Executive Functioning Problems

- Social immaturity
- Poor peer relations
- Naivety/vulnerability
- Hyperactivity
- Poor impulse control
- Poor emotion regulation
- Lack of insight into effect of behaviour
- Poor learning from past mistakes

Cold Executive Functioning Problems

- Poor working memory
- Poor understanding of abstract concepts
- Poor planning and organisation
- Difficulty sequencing
- Perseveration
- Poor sustained attention/concentration
- Inability to work independently



Many children with FASD have also been exposed to other substances in the womb, which cause additional problems

- ▶ Smoking (oxygen deprivation, small size, dopamine circuits, ADHD)
- ▶ Opiates (ACH and opiate receptors within limbic system)
- ▶ Cocaine (frontal dopamine circuits)
- ▶ Neglect and emotional abuse (attachment and trauma factors) – recent paper, Mukherjee et al (2018) – ‘postnatal neglect in children with FASD did not make the developmental outcome any worse, suggesting that prenatal alcohol influences these outcomes independently.’

Often the difficulties a child with FASD has, are the result of a compound effect of several, or all of the above.



FASD: An Invisible but Significant Disability

- ▶ Only 1 in 9 children with FASD have facial features
- ▶ Often have good expressive language with a (near) normal IQ
- ▶ However their brains struggle to process information normally
- ▶ They have executive function impairment in varying degrees
- ▶ Because of these issues, a child with FASD often does not respond well to 'traditional' behavioural management strategies.
- ▶ "Like trying to navigate round London with a map of Birmingham!"
(parent of a child with FASD)



Even with a reasonably 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
- Planning
- Organising their time/belongings/school projects
- Regulating responses to frustration
- Managing peer relationships

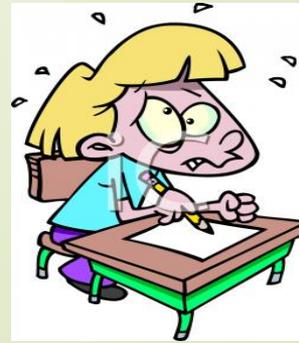


'If s/he would just try harder and listen more, s/he could do it..'

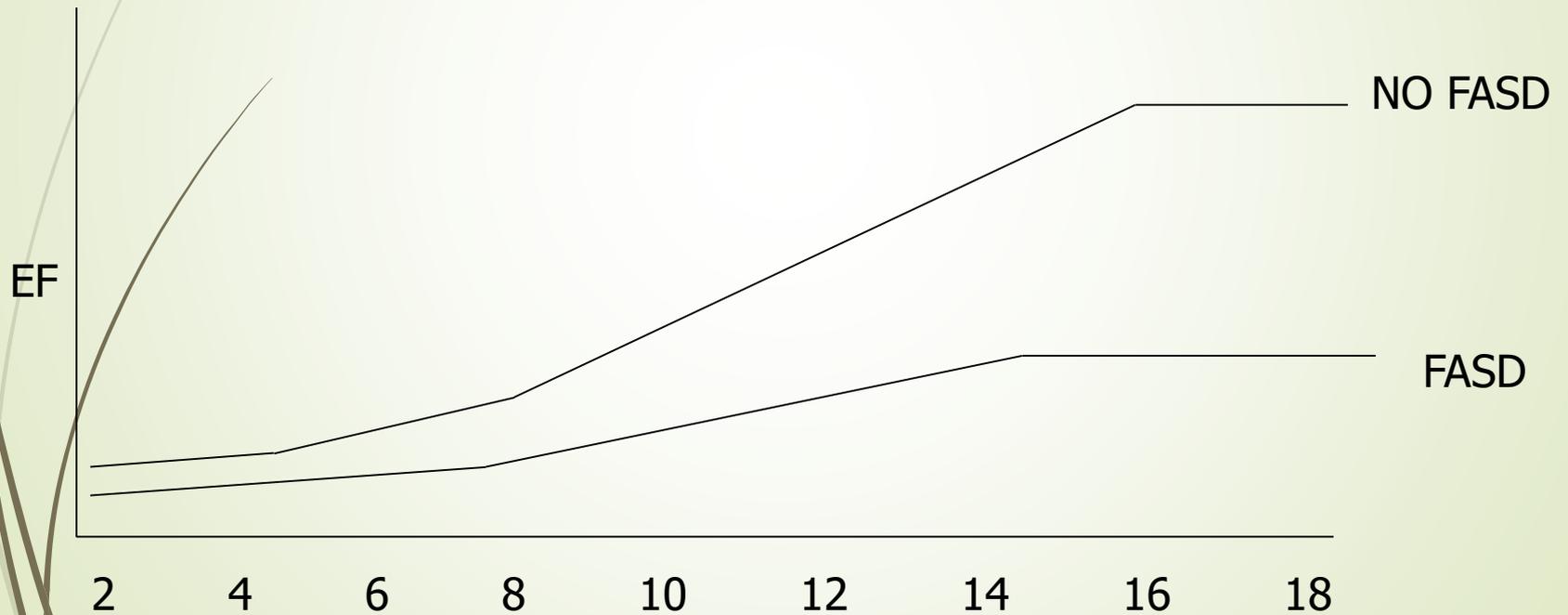
- ▶ Performance always fluctuates from day to day
 - ▶ Many children with FASD are trying to the point of exhaustion
 - ▶ Constant hyper arousal... meltdowns... more negative labels... further knock-on effect on self esteem.. Vicious cycle
- 

If Executive Functioning Impairment associated with FASD is not understood and recognised (particularly in school):

- A child can be incorrectly labelled as:
 - Lazy
 - Unmotivated
 - Disorganised
 - Oppositional
 - Wilful
 - Non-compliant
 - Poorly parented
 - Generally naughty



Executive Functioning: a Developmental Trajectory





Problems associated with EF Impairment often begin in school from year one onwards:

- 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)
- Inconsistent learning profile and poor working memory – ‘forgetting’ learning and requiring continual repetition
- Difficulty separating fact from fiction
- Temper tantrums, stealing, lying, disobedience, defiance of authority
- Poor understanding of social rules



Cluster of Neurocognitive Symptoms

- Working memory deficit
- Poor planning / sequencing
- Obsessive/perseverative presentation
- Slow processing speed
- Receptive language deficits
- Expressive language deficit (less common)
- Social immaturity
- Poor social understanding
- Hyperactivity
- Cognitive inflexibility
- Impulsivity
- Confabulation
- Poor imagination
- Tics
- Sensory integration issues
- Inattention and poor concentration



Multiple diagnoses are common: FASD is a great 'mimicker'

- Shared executive functioning deficits
 - Some shared social communication difficulties
 - Shared hyperactivity and inattention
 - ASD (often called 'atypical')
 - ADHD
 - Conduct Disorder
 - Dyspraxia
 - Learning disability
 - Attachment disorder
 - Environmental (neglect)
 - Borderline personality disorder
 - Sensory processing disorder
- 



Social Functioning Difficulties

- ▶ Normal nonverbal communication, but:
 - ▶ Difficulty providing the right information at the right time
 - ▶ Difficulty adapting social skills with increasing age
 - ▶ Problems understanding other's behaviours and motivations
 - ▶ Difficulty learning from previous experience
 - ▶ Problems recalling consequences of past actions
 - ▶ Often misdiagnosed as atypical autism (can be hard to differentiate)

ASD vs. FASD

ASD

- ▶ Abnormal eye contact
- ▶ Limited facial expression/face processing
- ▶ Often disinterest in peers
- ▶ EF deficit (usually milder)
- ▶ Difficulty initiating social interaction (and poor quality when accomplished)
- ▶ Sharing affect impaired
- ▶ Impaired non-verbal communication

FASD

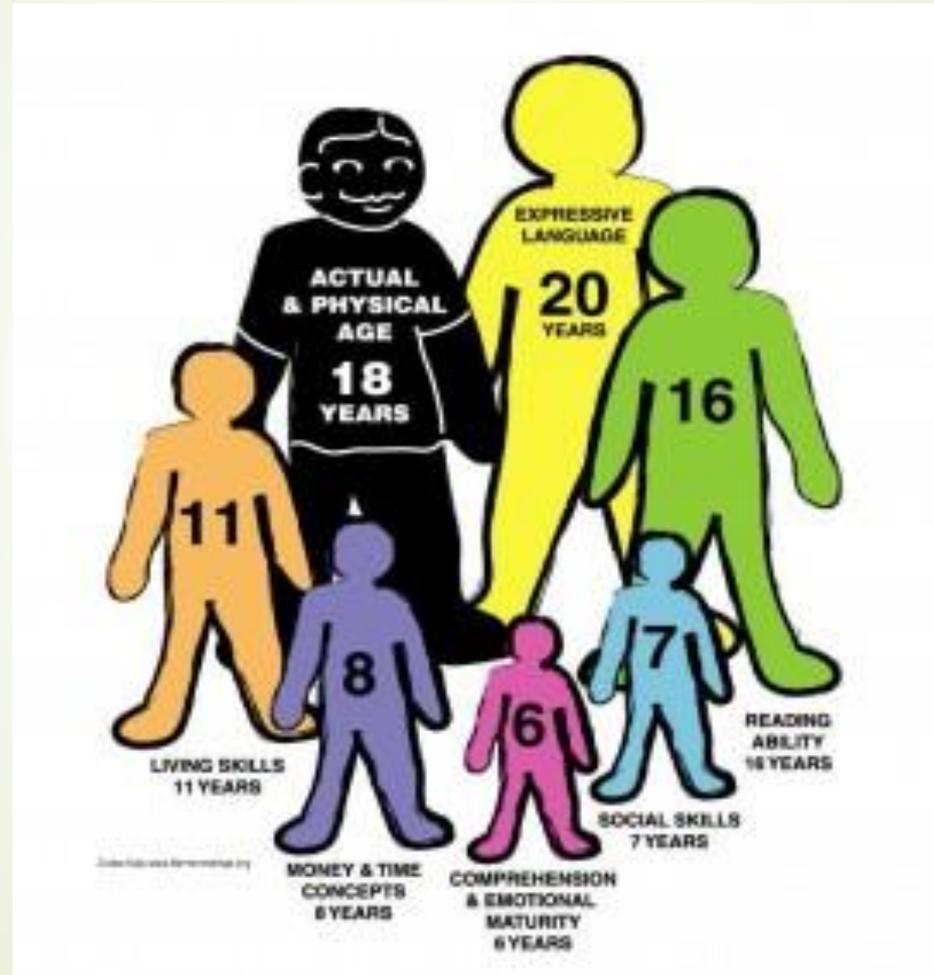
- ▶ Normal eye contact
- ▶ Normal facial expression/face processing (though can still struggle with reading emotions)
- ▶ Normal interest in peers
- ▶ EF deficit (usually more sig.)
- ▶ No difficulty initiating social interaction (although quality may be poor. Often socially disinhibited/'over-friendly')
- ▶ No impairment in sharing affect
- ▶ Normal non-verbal communication



ADHD and FASD

- ADHD is a common *symptom* of FASD
- 74% of FASD children meet the criteria for ADHD (Roozen et al, 2016)
- ADHD is itself a disorder of EF
- 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

An 18-year-old with FASD





Streissguth et al (2000) – Predictors of best prognosis for children with FASD:

- ▶ Diagnosis by age 8yrs
- ▶ Stable and nurturing home for >72% of life
- ▶ Stability between ages 8-12 critical (when educational difficulties tend to become most explicit)
- ▶ Receiving appropriate early education
- ▶ Eligibility for SEN services
- ▶ Prognosis is inversely related to number of placement moves for those in care – attachment disorder behaviours and FASD



Lynch and Kable (2016) - Predictors of best prognosis in FASD:

- Full term birth
- Being female
- Fewer dysmorphic facial features (the face as a 'window' to the brain)
- Higher FSIQ
- Fewer aversive life events in childhood
- Parental warmth
- Fewer siblings

The Importance of Diagnosis

- ▶ **No diagnosis means:**
 - ▶ Significant social vulnerability and no understanding or protections
 - ▶ Increased risk of mental health problems
 - ▶ Increased risk of offending behaviour
 - ▶ Increased risk of drug and alcohol abuse
 - ▶ Poor educational attainment and poor employment rates (due to misunderstanding and lack of support)
- ▶ **Timely diagnosis means:**
 - ▶ Prevention of many of above factors
 - ▶ Eligibility for SEN services into adulthood
 - ▶ Supervision and support

Any Questions?

