

Working with Parents of Children with Developmental and Intellectual Disabilities

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Introductions

- * Kylie Gray
- * Kate Sofronoff
- * Participants

Overview

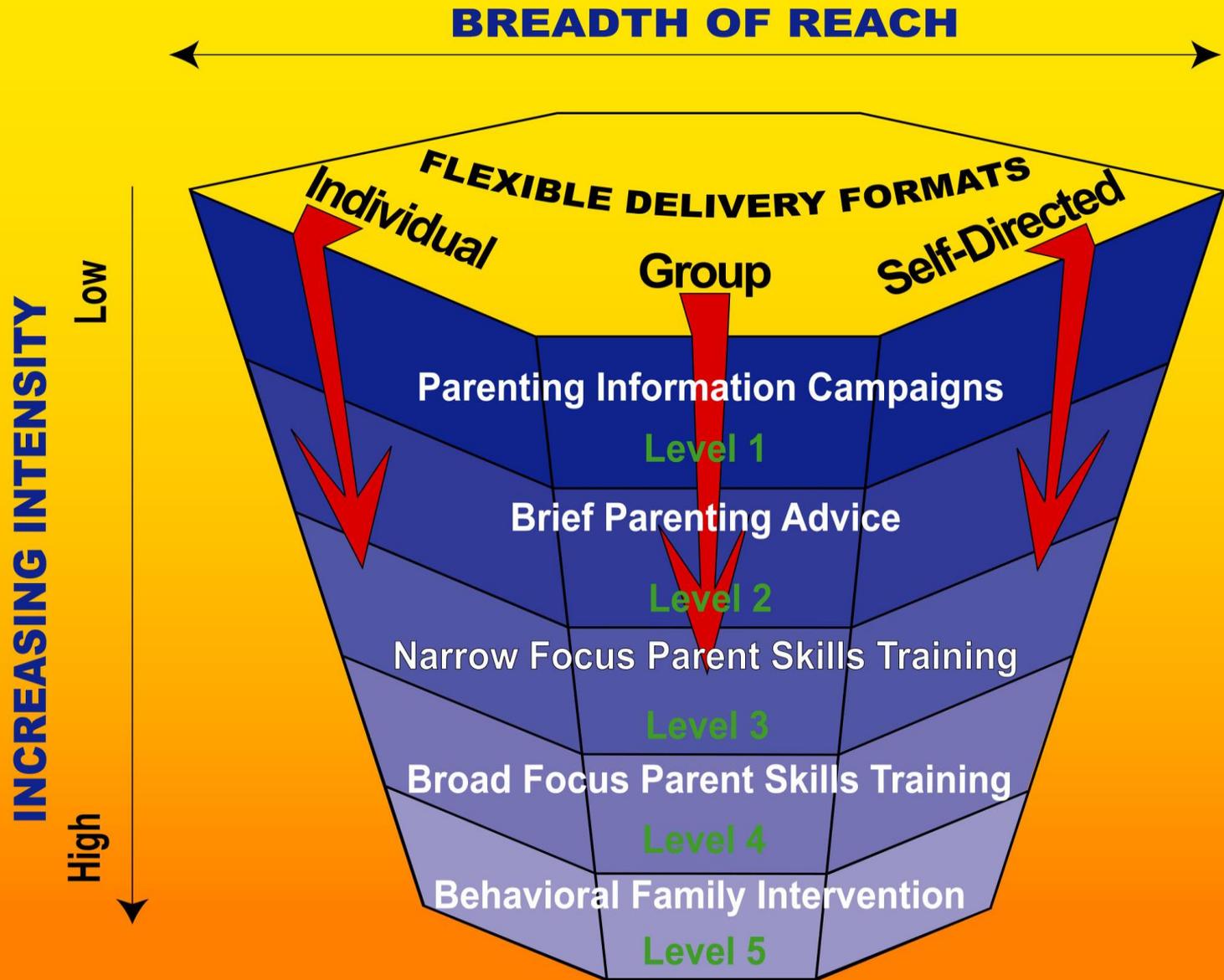
- * Stepping Stones
- * Managing different disabilities
- * Down syndrome
- * Fragile X
- * Williams syndrome
- * 22q11
- * Prader Willi syndrome
- * FASD

What is Stepping Stones Triple P

- * Forms part of the Triple P - Positive Parenting Program developed specifically for parents of a child with a disability
 - * Strategies based on ABS principles
 - * Includes functional analysis of behaviour
 - * Includes functional communication training
- * Aims to help parents promote a positive relationship with their child
- * Promotes positive strategies to increase child skill development
- * Aims to help parents deal with behaviour problems and developmental issues
- * Aims to prevent development of more severe behavioural and emotional problems by increasing parental skills and confidence

Theoretical Basis

- * Social Learning models (e.g. Patterson)
 - * Bi-directional nature of parent-child interactions
- * Social information processing (e.g. Bandura)
 - * Importance of cognitions, attributions, expectations
- * Developmental literature (e.g. Bandura)
 - * Developing competencies in real contexts
- * Child and family behaviour therapy
 - * Evidence-based strategies, functional assessment
- * Developmental psychopathology
 - * Risk and protective factors



Rationale for Development

- * **More common in children with a disability**
 - * aggression, destructiveness, noncompliance
 - * self injury, stereotyped behaviours
 - * persistent screaming, disturbed sleep
 - * Anxiety
 - * Anger issues
 - * Social and communication problems
- * **Einfeld & Tonge (1996)**
 - * 40.7% of children with Intellectual Impairment classified as showing severe emotional or behavioural disturbance
 - * 70-84% of children with developmental disabilities show mild to moderate emotional and/or behaviour problems

Common Issues for Parents

- * Managing challenging behaviours
 - * Head banging, self harm, screaming
- * Managing emotional problems
 - * Anxiety, social problems, anger, depression
- * Increasing independence
 - * What can the child do for him/herself?
- * Skill building
 - * Self care, tolerance
- * Nurturance traps
 - * It's the disorder, he/she doesn't understand

Impact of Behaviour Problems

- * Parent and sibling stress
 - * Increased when behaviour problems present
- * Need for outside assistance
 - * Increased when behaviour problems present
- * Developmental outcomes for the child
 - * Lowered when behaviour problems present
- * Interaction between parent adjustment and child adjustment

Common Beliefs

- * Some behaviours cannot be changed because they are part of the disorder so there is no point trying
- * A child cannot be taught to behave appropriately because s/he does not understand
- * Others must understand
- * A program must be developed specifically for the disorder

How Can we be Flexible Enough to Suit all Disabilities?

- * Children are children first – the disability comes way after that
- * Children with disabilities have more commonalities than differences when it comes to behaviour
- * Focus on the positive strategies first
- * Build from what the child and parent can already do
- * Assess the function of problem behaviours
- * What do we want the child **to do**
- * Remember that small changes can make a big difference to a family

Behavioural phenotype

- * A characteristic pattern of motor, cognitive, linguistic and social abnormalities which is consistently associated with a biological disorder
- * Form a basis for research into behavioural, emotional, and other aspects of biologically determined syndromes associated with intellectual disability
- * Encourage research and information sharing
- * Help to understand

Fragile X Syndrome



- Most common inherited cause of developmental difficulties
- Caused by inactivation of the FMR1 gene on X chromosome
- Prevalence estimates vary widely, but approximately twice as common in males
- Degree of ID tends to be lower in males, some females no ID but may have learning difficulties

Fragile X Syndrome

- Affects males and females differently – girls less affected as have 2 X chromosomes and only 1 is affected, males only have 1 X chromosome
- Associated with autism, ADHD (males) and ADD (females)
- ID is common, especially in males
- Delayed language development – little speech before age 3
- High aptitude for visual learning and imitative behaviours
- Sensory defensiveness – aversion to touch, loud noises, bright lights, etc.

Fragile X Syndrome

- core areas of difficulty:
 - Social anxiety, gaze avoidance, especially in new situations
 - Disturbances in speech patterns common – eg echolalia
 - Repetitive mannerisms (e.g. rocking, hand-flapping)
 - Shyness and social anxiety are common, often have difficulty making eye contact
 - Difficulty in new situations, and with changes to routines
 - Problems with co-ordination and motor skills (e.g. crawling, walking, toileting)

Williams syndrome



- Deletion on the 7q11.23 region of chromosome 7 (about 25-28 genes)
- Occurs in approximately 1 in 20,000
- Often (but not always) associated with ID, generally mild-moderate range of ID
- Diagnosis confirmed using FISH testing

Williams Syndrome

- Growth delay, failure to thrive
- Renal and cardiovascular problems are common
- Strengths in language - loquacious, auditory memory, facial recognition
- Early social-communication may be delayed
- Attention difficulties, easily distracted
- Poor response inhibition
- Difficulties in visuo-spatial processing and spatial cognition
- Often respond better to verbal teaching, rather than visual

Visuo-spatial processing deficits

- * Dissociation between language skills and spatial cognition
- * Verbal description of an elephant – 18 year old with WS

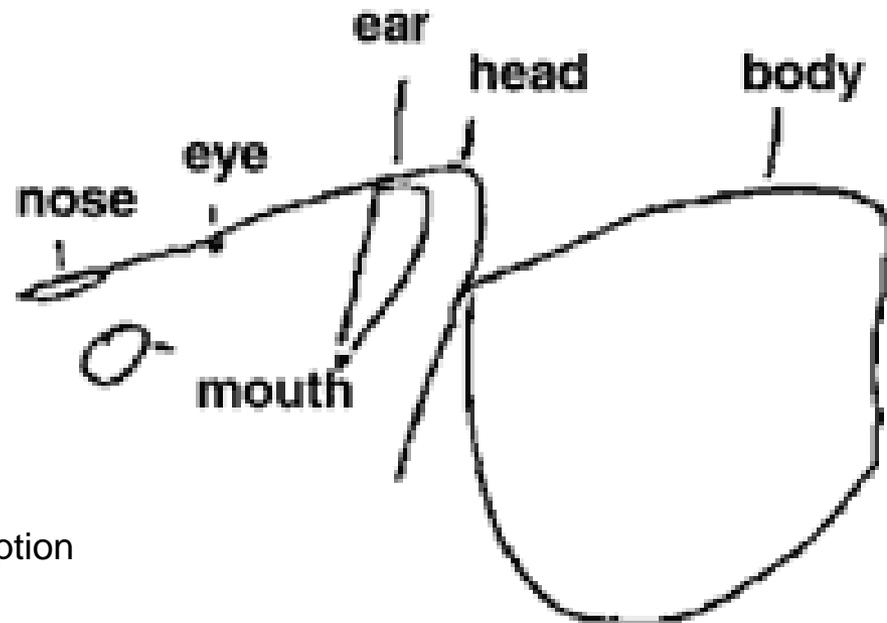
"And what an elephant is, it is one of the animals. And what the elephant does, it lives in the jungle. It can also live in the zoo. And what it has, it has long grey ears, fan ears, ears that can blow in the wind. It has a long trunk that can pick up grass, or pick up hay... If they are in a bad mood it can be terrible... If the elephant gets mad it could stomp; it could charge. Sometimes elephants can charge. They have big long tusks. They can damage a car... It could be dangerous. When they're in a pinch, when they're in a bad mood it can be terrible. You don't want an elephant as a pet. You want a cat or a dog or a bird..."

Visuo-spatial processing deficits

- * Dissociation between language skills and spatial cognition
- * Drawing of an elephant (same person, age 18, with WS)

The dissociation between language and spatial cognition in Williams syndrome is evident in this contrast between the drawing and verbal description of an elephant by an 18-year-old with Williams syndrome.

Elephant Drawing



Williams Syndrome

- Hypersocial, very friendly and outgoing, little or no fear of strangers
- Impaired social intelligence, difficulty interpreting social cues
- Difficulties with peers, with friendships
- Poor emotional modulation
- Increased rates of depression, anxiety and ADHD
- Heightened sensitivity to sounds (hyperacusis)
- Difficulties when faced with changes in routines or schedules

Prader-Willi Syndrome



- Caused by abnormality or deletion of genes on chromosome 15 (q11-q13)
- Occurs in approx. 1 in 25,000
- Mild to moderate ID common
- Increased appetite and food-seeking behaviour (hyperphagia)

Prader-Willi Syndrome

- Hyperphagia (increased seeking of food) often leads to weight issues
- Often have specific learning difficulties (e.g. reading, spelling) not related to IQ
- May have language and speech problems
- Strength in visual processing (e.g. jigsaw puzzles)
- Difficulties with motor skills
- Poor adaptive skills, despite degree of ID
- Obsessive-compulsive like symptoms

Prader-Willi Syndrome

- Obsessive thoughts
- Skin-picking, poor impulse control and temper outbursts
- Oppositional and argumentative behaviour
- Stealing, lying, possessiveness and stubbornness are common
- Mood fluctuations
- Repetitive questioning
- High rates of mental health problems – eg psychosis, depression

22q11 Deletion Syndrome (Velo-Cardio-Facial Syndrome)



- Also known as eg DiGeorge Syndrome (DGS), Shprintzen Syndrome, CATCH 22
- Caused by microdeletion on long arm of chromosome 22 (22q11)
- Occurs in (at least) 1 in 4000 to 1 in 7000 live births
- Clinical symptoms of VCFS described 50 years ago, but not until early 1990s research interest emerged
- Majority (90%) de novo mutations; neither parent affected

22q11 Deletion Syndrome

- Cardiac anomalies and immunodeficiency common
- Often (but not always) associated with mild ID, range moderate-normal
- Delayed language development, late speech onset
- Receptive language skills often better than expressive
- Some numerical difficulties
- Strengths in eg auditory/verbal rote memory, spelling, phonological processing skills
- Behavioural excitation and attentional difficulties

22q11 Deletion Syndrome

- Generally poor social interaction skills and bland/flat affect with minimal facial expression
- Difficulties with anger management
- Exaggerated response to threatening stimuli and enduring fearfulness of painful situations
- Mood disruptions, attention deficits, anxiety

22q11 Deletion Syndrome

- High rates of psychiatric disorders such as anxiety, depression, ADD / ADHD, OCD and bipolar disorder
 - ADHD rates 35-46%, compared to 3-6% typically developing children
 - schizophrenia and schizotypy – 20-30% (25 times higher than typical population)
 - Deletion of 22q.11.2 third highest risk factor for schizophrenia (after child with parents with schizophrenia, monozygotic twin of someone with schizophrenia)
- Very high rates of psychosis in adults (majority schizophrenia)
- Reports of higher rates of ASD, ranging 14-50%, conflicting reports
- OCD – rates ranging from 8-33%

Fetal Alcohol Spectrum Disorders



- Umbrella term – mild to severe impairment, hence spectrum
- Caused by maternal consumption of alcohol during pregnancy
- Wide range of disability, often in borderline to mild ID range
- 25% of people with FASD have an intellectual disability
- Learning difficulties

Fetal Alcohol Spectrum Disorders

- Attachment difficulties, poor social skills and interpersonal relatedness
- Tendency to overreact (emotionally)
- Speech onset delay, communication difficulties (especially comprehension) and poor auditory pace
- Difficulty following instructions, predicting, planning and organising
- Problems with working memory, and executive function

Fetal Alcohol Spectrum Disorders

- Problems with attention, hyperactivity, impulsivity
- Increased risk of
 - depression, anxiety
 - ADHD
 - conduct disorder and oppositional defiant disorder
- Poor coordination and fine motor skills
- Tendency towards stubbornness and tantrums
- Often lack imagination or creativity
- Difficulty understanding abstract concepts
- Tendency towards social withdrawal

Down Syndrome



- Also known as Trisomy 21
- Occurs in 1 in 1150 live births in Australia
- Most common genetic cause of ID
- Caused by an extra copy of chromosome 21 (translocation, mosaicism)

Down Syndrome

- Associated with ID (mostly mild to moderate, but can be profound)
- Difficulties with expressive language, verbal processing and motor functioning
- Increased risk of developing anxiety, depression, obsessive-compulsive disorder and ASD. Associated with early-onset dementia
- Hypothyroidism is common (can cause learning disorders, sleep problems, feeding disorders, depression, etc.)

Down Syndrome

- Congenital heart defects, immunodeficiency, and gastrointestinal problems are common
- Typically fewer emotional and behavioural problems than other children with ID, but still higher rates than typically developing children
- Common difficulties: inattention, hyperactivity, stubbornness, impulsivity, oppositional behaviour
- Pleasant and sociable personality, high levels of adaptive behaviour

Case Study

- * Amy is 4 years old and her mum complains that when she wants something she shouts and demands. This is especially difficult when there are visitors when Amy increases her volume and begins shouting about what she wants rather than saying hello to visitors.
- * Amy's mum would like her to listen to instructions, to talk in a quiet voice, and to interact more appropriately in social situations.