Neurodevelopmental Conditions in the Classroom

How to support students with neurodevelopmental genetic conditions – Fragile X as example

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We acknowledge and pay tribute to the traditional territories of the peoples of Treaty 7, which includes the Blackfoot Confederacy (comprised of the Siksika, the Piikani, and the Kainai First Nations), the Tsuut’ina First Nation, and the Iethka Stoney Nakoda (including Chiniki, Bearspaw, and Goodstoney First Nations). The City of Calgary is also home to the Métis Nation of Alberta Region 3.
Outline

• Introductions
• Introduction of genetic neurodevelopmental disorders – example Fragile X - Billie Au
• Particular challenges for unique kids - fragile X syndrome as example – Kara Murias
• Support for inclusive environment in education – Gabrielle Wilcox
• Successful moments and joys – Karen Kelm
• Discussion - All
Neurodevelopmental conditions (disorders)

• Some conditions are characterized by clinical characteristics: e.g. cerebral palsy, autism spectrum
• Some conditions are characterized by a physical medical difference: e.g. traumatic brain injury, Fragile X syndrome.
• In both categories children are at risk of disability and for other co-occurring conditions that may increase or complicate impairment e.g. anxiety.
• In each category: each child may have overlapping and unique strengths and weakness
Definitions

• Neurodevelopmental disorders (NDD): a group of heterogeneous conditions characterized by impairment in “personal, social, academic, or occupational functioning” with onset early in development (i.e. ASD, ID, GDD, ADHD)

• Neurodevelopmental genetic conditions: heterogeneous conditions characterized by brain dysfunction (epilepsy, movement disorders, cognitive, developmental, behavioural, mental health disorders) due to predominantly underlying genetic cause
Genetic variants (mutations)

- Variants can be *de novo* (new in an individual) or inherited in different patterns depending on the condition.
- Different variants infer different risk, but in all cases there is a range of presentation.
- In most cases NDD are the result of polygenetic inheritance – multiple genes working together with environmental risk.
- In some cases NDD is due to a single variant (genetic change) – These syndromes can have unique medical and developmental/behavioural features.
Fragile X syndrome is caused by a repeat expansion affecting the gene FMR-1, that results in insufficient FMRP protein in the brain.
This is a genetic condition that can run in families.

There can be more than one affected child in the family.

Carriers have health concerns too.

Figure 2. Sample pedigree of a family with FXTAS and FXS.
What does FMRP do?

- Present in many tissues, especially brain, at all stages of brain development
- Important for synapse development and implicated in various neurotransmitter signaling pathways (mGluR, GABA, BMPR2 etc)
FRAGILE X Syndrome – sources of variability

- Female vs male
- Mosaicism
- Contribution from other genetic factors
- Contribution from environmental factors
• There are thousands of other genetic variants contributing to differences in brain function

• Never underestimate the importance of environment

Like any individual - we cannot fully define or predict outcome for any individual based on a single genetic factor (even if it is a powerful one like Fragile X)
Example - Learning and behaviour profiles in Fragile X
Challenges in Fragile X

- Medical concerns
  - Motor impairment and hypotonia
  - Differences in appearance
  - Seizures
  - Reflux
  - Sleep disruption including restless leg syndrome, sleep apnea
  - Cardiac concerns
  - Pain syndromes such as migraine, central pain sensitivity
Strengths in Fragile X

- Energetic
- Empathetic
- Motivation to be helpful and be included
- Often good at imitation
- Often funny with a good sense of humor
- Good visual memory
Challenges in Fragile X

• Developmental delay – reach early milestones later than other children
• Intellectual disability - HIGHLY VARIABLE
  • typically moderate range for affected males (FSIQ 40s)
  • Can range from FSIQ10 to NORMAL range!
• Variable cognitive profiles – bigger gaps between strengths and weaknesses
• ADHD
• Autism or autistic traits – about 50%
• Behaviour differences (doesn’t always mean autism)
  • Sensory sensitivities or tactile defensiveness
  • Irritability
  • Aggression
  • Stereotypies like hand flapping
Anxiety and Hyperarousal

- Leads to:
  - Avoidance
  - Fatigue
  - Increased distractibility
  - Dysregulation
  - Outbursts
  - Self-harm
  - Aggression (usually less severe than other populations with intellectual disability)
Behavioural expression

- Increased symptoms of anxiety are associated with more behavioural concerns
- Social anxiety is prominent and impairs social interactions (with and without Autism)
- Prevalence of behavioural difficulties (but not self harm) may increase with age.
Supporting Students with Fragile X in School
General Considerations

• Some recommendations are related to challenges that are related to comorbidities or are common across disorders
• Common comorbidities include intellectual disability, autism, and ADHD
• Sex is a factor: boys generally have more cognitive and academic challenges
• Every child is unique
  • It is important to have an evaluation to identify strengths and weaknesses
  • Individualize interventions and accommodations to meet the individual needs
  • Monitor progress to inform whether any changes are needed to fully support the child

Nash et al., 2019
Common Challenges

Cognition
- Language difficulties make this harder to assess
- Intellectual disability is common (especially in boys)
- Cognitive declines are common in middle childhood

Language
- Expressive language weaker
- Stronger vocabulary
- Comprehension difficulties

Learning and Memory
- Strength in verbal short term memory and memory for meaningful information
- Weakness in memory for abstract information

Attention and Executive Functioning
- Weakness in selective attention and attentional control
- Comorbid ADHD is common
- Weakness in executive functioning broadly

Academics
- Reading performance variable
- Weakness in math

Emotional/Behavioural
- Symptoms of ADHD and autism
- Common: social phobia, depression, obsessive compulsive disorder, panic disorder, mood dysregulation

Murphy, 2009; Riccio et al., 2010
Interventions/Remediation

Teaching Skills
General

• Students with Fragile X often have inconsistent performance

• They gain academic skills more slowly than peers

• Verbal expression is hard: allow non-verbal expression too (pointing, gesturing)

• They often need more repetitions and concrete learning opportunities to learn new material
Reading

• Difficulty with phonological awareness makes learning to read difficult

• Little research to guide decisions about reading intervention for students with Fragile X (Randel et al., 2015)

• There is emerging evidence that reading intervention that first address phonemic awareness can support reading development in individuals with Fragile X (Adlof et al., 2018)

• Edmark is a sight word based program that can be helpful for students who are not able to gain the phonemic awareness skills to learn decoding
Math

- Difficulties with attention, executive functioning, language, and visual spatial processing contribute to math difficulties

- Relative strengths: rote counting, reading/writing numerals

- Relative weaknesses: applied counting (moving objects does not change the number of objects)

- Touch Math is a tool support math skill acquisition, making the process more concrete

Can Calik & Kargin, 2010; Murphy, 2009
Writing

• Students with Fragile X often have fine motor difficulties

• Handwriting without Tears is a program that can support writing development

• Some students may benefit from speech to text software if they do not have expressive language difficulties
Social/Emotional

• Hyperarousal and anxiety
  • Teach students to be know when they are hyper aroused or anxious
  • Teach calming strategies (breaks, breathing techniques, progressive muscle relaxation)

• For challenging behaviours, request a functional behaviour assessment (FBA)
  • This will help to determine underlying causes, the need they are trying to meet, and missing skills
  • It will inform a positive behaviour support plan (PBSP) that can support the student in gaining ways to meet needs more prosocially
Transitions

Teach transition routines to the whole class

Visual timer can help with transitions

Visual schedule
  - Helps with transitions
Accommodations

Additional Supports (technology [calculator, speech to text, etc.], seating, breaks)
Individual Program Plans

Goal should be related to assessment findings

They should be observable and measurable

It should be clear who is providing what supports when

It should be clear who is measuring progress, how often, and which tools

They should be reviewed regularly to determine if they are working or not and modification should be made if they are not working
Resources

• https://www.learnalberta.ca/content/inmdict/html/fragile_x.html
• https://fragilex.org/our-research/treatment-recommendations/general-educational-guidelines/
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Discussion

https://research4kids.ucalgary.ca/owerko

• Sharing of successes and challenges
• Questions for discussion
• Comments on areas for future learning