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“Glut 1 DS Characteristics of Cognition and Behavior”

First:

Please remember that these findings are from GROUP data. The data show what children are “at risk for” if they have the Glut 1 DS diagnosis, but not all children will have every characteristic.

Every individual is unique! Each child needs a thorough clinical neuropsychological evaluation to determine his or her own individual strengths and weaknesses and remediation strategies best suited for his/her profile.

Keeping that in mind, here are 10 Take Home Points & Implications:

1. Across children diagnosed with Glut 1 DS there is a wide range of cognitive function. As a group, scores appear to be normally distributed, but are shifted down from general population about 1 1/2 to 2 standard deviations.

- Some children’s IQ will be in the “normal” range, but most will have scores significantly lower than expected for general population.

2. Among children with Glut1 DS, receptive language skills are stronger than expressive language skills (regardless of overall level of IQ)

- Speech therapy is recommended!
- Because speech in individuals with GLUT 1 DS tends to be dysfluent and poorly articulated, children may appear to be more impaired

than they are. Speech therapy can help children express themselves with more ease.

- Children may be frustrated at times by the difficulties they face getting understood by those who don't know them well
- For some children augmentative communication devices may be helpful

3. Among children with Glut1 DS, visual attention to details tends to be weak

- These are areas emphasized in academic school work and can be trained and improved
- Visual search puzzle-games like “spot the difference” in pictures, eye-spy, word search and “Where’s Waldo” help train this ability
- Help go over school assignments carefully, step-by-step to help teach your child to focus on details
- This may be variable throughout the day; there may be “spells” of inattention

4. Among children with Glut1 DS, fine motor skills are weak

- This is another areas emphasized in academic school work - writing and copying
- Physical therapy and occupation therapy can help with this
- Encouraging play with blocks and Legos and small items as well as encouraging drawing is recommended
- For some, use of an assistive keyboard may be essential

5. There is a definite bias in cognitive processing style. Children use a sequential processing approach preferentially. Use this to your advantage!

- They are better at seeing the trees, than the forest
- Use teaching strategies that rely on sequential or step-wise approaches to material.
- Teach one thing, then another and another
- Rely on repetition and rote memorization
- Break large assignments into manageable steps
- Consider using organizational strategies similar to those recommended for children with attention deficit disorder
- Explain clearly what the general point or “take home” message is

6. Children with Glut1 DS have difficulty using simultaneous or “whole picture” processing approaches. Be aware of limitations

- They have difficulty seeing the forest, and tend to focus on the individual trees
- Give extra help when trying to pull together the whole picture
- Talk about stories and what the point is (fables and folktales work well for this!)
- Play with abstract puzzles where the whole is greater than the sum of the parts -work on integrating visual information

7. Adaptive behavior scores are comparable to scores on cognitive tests

- There is a wide range of adaptive behavior performance
- Although for some children adaptive scores may fall in the “normal range,” the majority of children with Glut1 DS perform at levels below those expected for their age
- Compared to Daily Living, Communication and Motor Skills, Socialization skills are strengths (across all levels of function)
- Adaptive behavior skills are crucial areas to learn
- Some parents may be reticent to give their children age-appropriate responsibilities in caring for themselves and their home, but having children participate in day-to-day activities and helping them become proficient in self care and household skills will likely increase the child’s sense of family and self worth
- Helping children be more independent in caring for themselves will instill greater self confidence and help them achieve more

8. Social skills are strengths!!

- This is not easy to measure, but readily apparent!
- Children with Glut1DS stand out in their “winning personalities”
- Children with Glut1 DS present as delightful, full of charm, empathetic, socially outgoing and have a playful sense of humor.

- Their ability to make friends will serve them well throughout life
- Children with Glut 1 DS should be encouraged to be active in school groups and all social settings, giving them ample opportunities to develop and enjoy warm relationships with others
- This is a remarkable strength and one that is not emphasized as much academically as other skills, yet may well help them in all aspects of their lives

9. There are no known aberrant behaviors associated with Glut 1-DS.

- Although some children may have problem behaviors, there is no consistency observed across the group. There is no evidence of any significant psychopathology associated with the diagnosis.
- Many children with Glut1 DS may have attention difficulties, and these may fluctuate during the course of the day.
- As children mature and gain more insight, some may have adjustment concerns related to living with a developmental disability. These adjustment problems seem similar to those observed in adolescents living with any type of disability and may be responsive to intervention with supportive psychotherapy
- Again, the predominant behavior characteristic associated with Glut1 DS is how cued into others the children are.
- Thus, praise and encouragement are likely the most useful tools in teaching children with Glut-1 DS! They respond very well to positive feedback.

10. For those children on the diet, normal developmental gains are made over time. There is no evidence of decline!

- Even though the ketogenic diet may not be a “cure,” it does help keep children developing and there is no evidence of progressive worsening over time
- Attention is better maintained, allowing for the child to learn more
- Stay in good ketosis!!!

Lecture notes prepared by Veronica Hinton