Practical Tips for Parents of Children with MECP2 Duplication Syndrome

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Learner’s Objectives

• Utilize practical interventions for sleep and feeding problems in children with MECP2 Dup

• Know an approach to teach child with MECP2 Dup

• Identify appropriate strategies for reducing problem behaviors in a child with MECP2 Dup

• Discuss appropriate lifetime planning for a child with MECP2 Dup
Tip # 1

• Take joy in the child you have
  - Do this daily
  - Don’t miss the fun while wishing for something else
Tip # 2

• Remember that successful parenting is raising a child who is
  
  - happy, enjoying life
  
  - as healthy as possible
  
  - at his/her optimal ability level no matter the plateau
Tip # 3

• Celebrate small gains
  - Do this daily
  - Don’t compare to others
Tip # 4

• Take care of yourself and your relationships
  - Use community resources/extended family/other parents with children like your child for giving you respite time for yourself and your relationships
  - Use trusted counselors as needed
  - Your child’s success depends on happy, healthy parents and siblings
Tip # 5

• Avoid making MECP2 Dup the center of your life and your family’s life

- It is okay to make MECP2 Dup a part of your life/your family’s life just like another child’s Asthma is a part of your life/your family’s life when making activity plans
Tip # 5

-It is okay to involve the whole family in shared responsibilities to help with a child with MECP2 Dup as long as the child with MECP2 Dup is involved in activities centering on the other children also

-Take joy in EACH family member for his/her unique characteristics, not just the one with MECP2 Dup
Tip # 6

• Remember that your child is more typical than not and has
  - the same need for love, dignity, value as others
  - the same health risks as others
Tip # 6 cont’d

-the same needs for medical monitoring and prevention as others

-the same joy over success as others

-the same capacity as others to bring joy and love to others
Tip # 7

• With regard to helping children learn, behave, eat, sleep, and have appropriate bowel movements: the old methods work best most of the time.
Tip # 8

• The intervention that brings the most improvement, especially for the non-verbal or barely verbal children, continues to be the combination of speech/language therapy and applied behavioral analysis/ABA therapy. (This is true for children with or without autism.)
Tip # 8 cont’d

*ABA Therapy=Applied Behavioral Analysis Therapy

-Identifies the maladaptive behavior and its trigger, then gives a trial of what could entice the person with the maladaptive behavior to cooperate or switch to an appropriate behavior=great for reducing self-injury/aggression and for enticing cooperation with instructor

-Works for all ages/ability levels & extremely useful for those who are non-verbal/barely verbal with profound intellectual disabilities or totally deaf/blind
Tip # 8 cont’d

-Use of ABA methods to entice the child to cooperate with the speech/language therapist brings optimal results if

-the speech/language therapist aims the therapy at the child’s baseline level, helping that child with understanding language and communicating using pictures and/or signs (if not speaking yet) while the therapist says the word with the picture/sign (or using a communication APP) so the child will hear the word and eventually say it
Tip # 8 cont’d

AND if

-the parents and other caretakers use the same ABA methods to entice the child to use what that child has learned in speech/language therapy

-Once the child is in a classroom setting, continued use of ABA methods to entice the child to cooperate with appropriately-aimed instruction practice continues to bring optimal results
Tip # 8

• ABA Therapy can be expensive but some states’ law requires that some medical insurance types cover this (unfortunately, not all, so an aim of a local parent support group should be to have on the group’s website guidelines for which insurance types cover ABA Therapy/how to get insurance coverage for ABA Therapy/where to get ABA Therapy under scholarships/grants or at university behavioral psychology or education training programs/where to obtain parent training in ABA methods)
Tip #8 cont’d

• See http://media.mindinstitute.org/education/ADEPT/Module1Menu.html and http://media.mindinstitute.org/education/ADEPT2/Module2Menu.html for parent training in ABA methods taught by experts from The MIND Institute in California.
Tip # 8 cont’d

• Speech/Language Therapy is somewhat easier to obtain through IDEA part C for 0-3 year olds and through the public schools from 3 years old forward (through IDEA part B) or privately.

- Get speech/language therapy even if not coupled with ABA therapy if you can’t get both

- If child is non-verbal/barely verbal, don’t wait to get speech/language therapy
Tip # 8 cont’d

- If child is non-verbal/barely verbal and you can get ABA without an autism diagnosis (or you can get parent training in ABA without an autism diagnosis), don’t wait for a diagnosis since ABA is useful for enticing non-verbal/barely verbal children without autism to cooperate with using what the speech/language therapist is teaching them.
Tip # 9

• Don’t forget the “Do No Harm” and “If Not Harmful, Avoid Expensive Treatment for Possibly Low or No Benefit” Rules when considering an intervention
Tip # 9 cont’d

- Ask your child’s physicians about safety and effectiveness of treatment issues


- Use [http://clinicaltrials.gov/](http://clinicaltrials.gov/) to look for safety-monitored, free clinical trials of promising but expensive not definitely proved interventions of which you hear or see on the internet
Tip # 10

• Never try an elimination diet without working with a licensed dietician
  - to gain advice on how to make sure child receives needed nutrients eliminated in diet
  - to gain advice on what to eliminate
Tip # 10 cont’d

- General rule:

- Eliminate one food at a time

- Do total elimination for 2 months and if no improvement, stop
Tip # 11

• Most sleep problems are behavioral and can be solved with behavioral methods

  - Observe to rule out physical causes like sleep apnea & seizures & restless leg syndrome

  - Talk to child’s pediatrician about signs of physical causes which can be investigated through a 4-hour sleep lab
Tip # 11 cont’d

- Practical Treatment for Behavioral Sleep Problems
  - Have same nightly bedtime
  - Have a bedtime ritual to include no electronics in the hour before bed plus a warm bath followed by a bedtime story in the hour before bedtime
  - Have bedroom at a cool temperature and with total darkness except dim nightlight when “lights out”
Tip # 11 cont’d

• Starting the routine may be rough for 2 months but routine can be established if parents do not “give in”

• Try to avoid medication for sleep
Tip # 11 cont’d

• If the bedtime routine is firmly established and the individual still has problems:
  Call Dr. Daniel Glaze who gave the sleep presentation earlier today as he truly is an expert sleep neurologist for children with genetic differences who have neurodevelopmental disabilities.
**Tip # 12**

- Use the following guide to address feeding problems

  - Observe to rule out physical causes but also look for behavioral causes

  - Enlist help of speech & occupational therapists to look at & address problems with oromotor coordination & abnormal swallow studies + the help of GI specialists to look at & address gastroesophageal reflux

  - Address “the trigger” of the behavioral feeding problem
Tip # 12 cont’d

• Identify the trigger

• Eliminate the trigger or help the person with the Developmental Disability adapt to the trigger

• Work with behavioral therapist (ex: when child with resolved reflux no longer has reflux but still fears pain after eating so won’t eat or when child avoids food textures/smells due to preference and not due to physical problems or fears based on resolved physical problems)
Tip # 12 cont’d

• Use your child’s preferences/atypicalities to entice eating

- identify textures/tastes preferred by child and prepare foods that mimic these textures/tastes just like Oprah’s cook did in the early 1990’s

- make use of child’s desire for sameness and make certain the food shape is uniform

- make use of child’s obsessions and match foods to these/have person who is higher-functioning go to exotic vegetable aisle and learn everything about a certain vegetable which he/she will learn to prepare and eat
Tip # 13

• A routine exercise regimen can solve a lot of problems
  - burn off energy
  - keep in shape
Tip # 13 cont’d

-per national leader regarding mitochondrial diseases, Bruce Cohen (MD), says that even for his patients known to have serious mitochondrial disease, daily exercise enforced (having child quickly pace back/forth, run around track, pedal stationary bike, etc) does better than 3000 mg/day of Coenzyme Q 10

-work with physical therapist to devise ways to help non-ambulatory children with profound ID exercise
Tip # 14

• To reduce/eliminate a behavior problem
  - Identify the behavior
    - Attention problem
    - Social isolation
    - Non-cooperation
    - Compulsion
Tip # 14 cont’d

• Ask “Is the cause medical, environmental, or both?”

- Ex: Getting out of seat to squat could be subconscious need to increase blood pressure

- Ex: Inattention or social isolation could be due to not understanding instead of ADHD or Autism

- Ex: Non-cooperation or compulsion could be a reaction to not being able to do a skill/hearing noise
Tip # 14 cont’d

- Ex: Self-injury could be due to
  - having nothing else to do
  - pain, not necessarily where the person is aiming the self-injury
  - anxiety over new situation/new environment
Tip # 14 cont’d

-Ex: Constant screaming could be due to

- having nothing else to do

- pain

- anxiety over new situation/new environment
Tip #14 cont’d

• Eliminate cause if possible
  
  - Ex: Give child plenty to do—for non-verbal, use switch toys with lights/music

  - Ex: Treat medical problem causing pain or high blood pressure or etc.

  - Ex: Stop noise or move bothersome classmate or etc.
Tip # 14 cont’d

• Choose management for the behavior if cause can’t be eliminated

  - Medication (Ex: for ADHD or Anxiety)

  - Therapy (Ex: ABA or Cognitive)

  - Environmental Change (Ex: Change to classroom that is routine-driven, non-chaotic or that aims toward student’s abilities instead of higher)

  - All of the Above
Tip # 15

• New behavior problems usually signal one of the following:
  
  - a new medical problem
  
  - a situation/environmental change
  
  - new learning expectations/approaching at too high of a level for child’s ability
Tip # 15 cont’d

• Look for the reason for behavioral changes before considering a behavioral medication or a change in behavioral medication
Tip #16

• Everyone does better in an organized, non-chaotic, routine-driven setting with few surprises when not hungry or tired

  - Have regular mealtimes, bedtimes, rules with consistent enforcement

  - Use picture schedules

  - Warn about upcoming routine changes/needs for transition in enough time to help (ex: have pictures to review several days before routine change/use kitchen timer with warning of when it will go off to indicate need for activity change)
Tip # 17

• Use your pediatrician’s office for a medical home and case management

  - allows primary physician to give continuity care which allows that physician to see your child’s baselines, progress, trajectory

  - allows early identification of true regression, new problems instead of saying “that’s just how that child is”

  - allows for primary physician to know what all the subspecialists have done/recommended for child
Tip # 17 cont’d

- allows for parent partnership with long-time provider which mutual trust to allow for parent to ask questions/share new information without feeling threatened/sounding threatening

- allows primary care office to “learn along with parent” regarding updated findings, recommendations for management, research studies, resources for those with autism

- allows for help with insurance/medicaid papers and approvals and subspecialty referrals
Tip # 18

• When a child loses focus, check for understanding and if the child does not understand:

  - show child as well as tell child at his solid language understanding level
  
  - within the attention span expected for someone at the child’s language understanding level (1 min/year of language understanding age)
Tip # 18, cont’d

-then give supervised practice to re-instruct, redirect, reinforce until the child demonstrates independent understanding/independent application of what he has been taught
Tip # 19

- Parents are the most powerful and most successful teachers of their children if the parents
  - use common sense
  - learn from their child’s teachers & therapists what to practice with their child daily at home in short, practical ways applied to daily life instead of worksheets
Tip # 19 cont’d

-remember that everyone, even individuals with profound intellectual disability, can learn new things at their learning ability level

-remember that a learning problem that is a disability is just like the parent’s own learning weakness but just worse

-remember how they learn something in their weak area and teach they way they learn best in their weak area
Tip # 19 cont’d

-remember that people learn to get better in their weak area by learning one step at a time with much practice before going on to the next step and do this with their child.

-remember that people do best when taught to identify their strong areas and use those areas to help them learn in their weak areas.

-remember that people pay better attention when they understand something completely.
Tip #19 cont’d

-Aim instruction at child’s independent receptive language level

-Show as well as tell child using concrete language

-Break instruction down one step at a time

-Help child connect new learning to what he/she already knows
Tip # 19 cont’d

- Allow for supervised practice to reinforce/reinstruct/redirect until child can do the skill independently

- Remember to limit instruction/practice before having to redirect to the attention span expected for someone at the chronological age of their child’s language understanding age (attention span = 1 min/year…so a 10 y/o with language age of 1 y/o can only attend for 1 min before redirection)
Tip # 19 cont’d

-remember it takes 28 days to make a habit, so to help their child remember something, it needs to be reviewed once/day for at least 28 straight days or even longer

-remember if person plateaus and can’t go higher to next level, person can still learn many more skills/do many more activities at the level he/she has plateaued (GO HORIZONTAL ONCE CHILD CAN’T GO VERTICAL IN LEARNING)
Tip # 19 cont’d

--Use strategies like tracing of letter character or sight/spelling word or number character or quick math fact printed in black marker on sandpaper square

-Trace and simultaneously say the letters then letter sound/word/numbers and the answer

-Do 5 times the first day then do the same tracing in air with eyes closed to imagine letter/word/number/problem while saying what they are tracing
Tip # 19 cont’d

- review once/day as above for 28 days such that every day the child is doing the routine once for every item he/she has learned the previous 28 days while introducing a new item to learn by following the tracing 5x routine

- remember that 5 min/day of practice every day is better than an hour of practice once/week

- remember to give supervised practice to allow for prompting, correcting, reinstructing so child will not practice something wrongly
Tip # 20

• Parents can be their child’s most effective school advocate if they
  
  - know state/federal education laws for special services (KNOWLEDGE IS POWER)

  - approach the school officials in a respectful, non-adversarial way using just the facts in a non-accusing manner
Tip # 20 cont’d

-work as equal partners with the school officials/teachers/therapists to make certain that instruction methods and classroom settings are those geared for the way their child learns best
Tip #21

• For Problems with fine motor

- Use occupational therapy along with occupational therapy consultation to the teacher/parent

- Modify expectations (truly, in 20 years everyone will use voice dictation instead of writing, so children need to spend more time learning oromotor and touch screen skills instead of “perfect handwriting”)
Tip # 21 cont’d

- Use accommodations like assistive technology
- Use less writing/more oral testing at school
- Separate grades for writing neatness and content at school
Tip #22

• Use a Pediatric Physical Medicine/Rehabilitation Physician if your child has hypotonia, especially if your child is non-ambulatory

  - This person can advise regarding therapy
  - This person can advise regarding equipment including wheelchairs, orthotics, prone standers, etc.
Tip # 22 cont’d

-This person can advise regarding prevention of future problems due to maladaptive positioning, maladaptive movement, lack of weight bearing, etc.
Tip # 23

• Prepare for your child’s adulthood from Day 1

  - Prepare for the worst but work and pray for the best

  - Make will

  - Put child on adult waiver lists
Tip # 23 cont’d

- Establish Special Needs Trust but still put money into post-secondary training fund

- Update plans as you see needs

  - routinely update will

  - routinely review waiver lists
Tip # 23 cont’d

- routinely review trust/education funds

- keep up with community resources

- visit places offering adult day habilitation and residential care to determine appropriate places (or plan such a place with other parents) in case these are needed
Tip # 24 cont’d

• How to Anticipate Child’s Future Ability

- >30% delay in any area is significant

- If a 3 y/o is like a 2 y/o despite good therapy, this child has a significant delay

- That might mean intellectual disability (IDD) as an adult if the delay is in language & non-verbal problem solving
Tip # 24 cont’d

• Anticipated Adult Skill Levels

- Mild IDD (IQ=55-69) = 8 - 11 year old

- Moderate IDD (IQ=40-54) = 5 - 8 year old

- Severe IDD (IQ=25-39) = 3 - 5 year old

- Profound IDD (IQ<25) = <3 year old
Thank you!

- Meyer Center for Developmental Pediatrics at Texas Children’s Hospital: 832-822-3400

- Texas Children’s Hospital’s Autism Center: 832-822-3400

- Texas Children’s Hospital’s Fragile X Clinic: 832-822-3400
Thank you!

• E-mail any questions and feel free to give out my e-mail to any high school or college student interested in a career in Neurodevelopmental Disability Services as a physician, psychologist, special educator, or therapist

• ssvinson@texaschildrens.org and if no answer within 48 hours, call my secretary at 832-822-3425