

Nutrition Management of Children with Rett Syndrome

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Nutrition Goals

- Well Balanced
- Texture Modified
- Adequate Energy, protein, key nutrients



Nutrition Goals

- Maintain Acceptable Weight, BMI



Nutrition Goals

Identify and initiate appropriate feeding methods



Nutrition Goals

- Manage Medical conditions related to nutrition such as:
 - Poor growth
 - Low bone density
 - Swallowing Problems
 - Constipation or Poor hydration
 - Seizures, anticonvulsant medications



Nutrition Interventions

- Poor Growth
 - Increase calories and protein by using
 - Calorie dense foods
 - Frequent feedings
 - Nutrition supplements
 - Feeding Tube



Nutrition Interventions

- Low Bone Mineral Density
 - Increase Calcium and Vitamin D
 - Dairy products
 - Supplements
 - Sunshine!



Nutrition Interventions

- Constipation
 - Increase hydration
 - Increase fiber
 - Laxative foods
 - Medication may be necessary



Nutrition Interventions

- Seizures
 - Anticonvulsant medications
 - Interfere with calcium absorption
 - Altered appetite



Nutrition Interventions

- Seizures
 - Ketogenic Diet



Nutrition Tests

- Blood work
 - Vitamin D (OH-25)
 - CBC
 - Zinc
 - Folate



Nutrition Tests

- Dexa
- Videofloroscopy



G-Tube

- Indications
 - Weight
 - Hydration
 - Safety
 - Quality of Life
- Supplemental



Nutrition Care Plan

- Individualized
- Monitored over time
 - Childhood
 - Adolescence
 - Adulthood



Feeding /Swallowing problems associated with Rett syndrome

- Extended meal length- 30-60 minutes
- Inability to self feed
- Poor lip closure on spoon
- Weak and inefficient chewing
- Poor tongue mobility
- Extraneous movements interfering with feeding/eating
- Delayed oral transit

- Motil, K.,; Schultz, R.; browning, K., Trautwein, Lynn. Gastroesophageal dysmotility are present in girls and women with rett syndrome. Journal of pediatric Gastroenterology and Nutrition. (1999). 29 (1), 31-37.



Common Swallowing Difficulties

- Nasopharyngeal reflux
- Poor bolus formation secondary to poor tongue mobility
- Weak base of tongue retraction
- Falling of liquids and solids over the base of the tongue before swallow initiation
- Pooling of liquids and solids in the valleculae and pyriform sinuses and laryngeal penetration- (all increase the risk of aspiration).

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Signs of swallowing dysfunction

- Coughing, choking, gagging, watery eyes with eating.
- Frequent upper or lower respiratory infections



Addressing the issue

- Diet history-include texture, amounts, and time it takes to consume foods.
- Occupational therapy evaluation to address oral motor function and proper positioning for safety
- Speech therapy/occupational therapy evaluation of swallow function- may have to do these a few times as your child progresses through the stages.



Modifications to support successful mealtimes

- Modify textures to match the child's oral motor skills. -this may include thickening liquids, pureeing foods, mechanical soft foods.
- Begin meal with face washing, vibration or simple facial massage to wake up the mouth
- For children that are low tone and have poor oral awareness punch up the flavor in their foods to give them more sensory awareness, change temperature qualities-give foods that are colder at the beginning of the meal, crunchy foods (within the child's skill level-melttable solids).
- If working on advancing texture give the foods that are a higher texture at the beginning of the meal
- Meals should last around 30 minutes- punch up foods according to nutritionist to decrease volume, and decrease amount of time a meal takes.



Case Study

- T.M.- Four year 7 month old with Rett syndrome
- History of nasopharyngeal regurgitation with purees and soft solids (normal swallow with thin liquids), seizures, poor weight gain, and immature oral motor skills
- Goal to support adequate nutrition for growth and development.
- Diet History
- Changes made



Case Study

- 7/09 27.3 lbs
- 1/10 27.4 lbs
- Nutrition and Feeding Intervention Started
- 1/10 27.4 lbs
- 4/10 29.5 lbs



Case Study

- Calorie Count
- 1/10 1475 Calories per day
- 5/10 1717 Calories per day
- Diet changes made
 - Soy milk changed to Bright Beginnings Soy Pediatric Beverage
 - Hot dog and vegies dipped in ranch dressing

