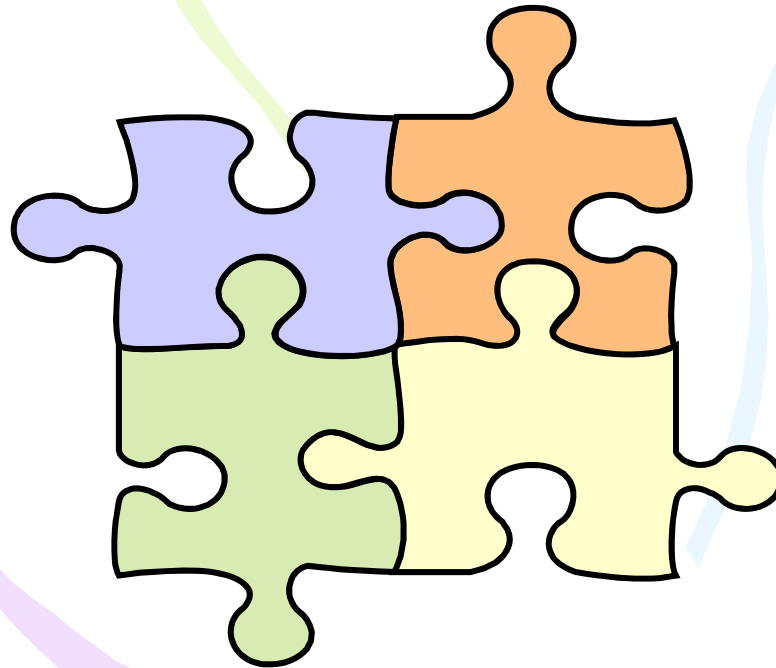

Children with Mucopolysaccharidosis Type 2 (ML2) or I-Cell

Feeding and Nutrition





Introduction

- Receiving the diagnosis of I-Cell Disease could be devastating for a family. A group of families gathered to compile questions that newly diagnosed families may have about this disease and the prognosis. The objective of this document is to help other families find the best quality of life for their child.
- This presentation provides information and tips to help families with feeding and nutrition issues/challenges their children may face. Information was collected from surveys sent to parents of children with I-Cell Disease and is based on their experience.
- The objective is to describe the spectrum of issues and give parent-to-parent recommendations from the families surveyed.
- *Tip - It is important to work with your medical team to learn about the disease, but please remember that the range of symptoms can vary widely from one child to another. In addition, a child's symptoms may be severe in one area, yet mild in another.*



Survey results: Birth-infant sucking issues

- At birth, sucking (bottle or breast) was an issue for a majority of families.
- Within a few months, many infants learned to suck from a bottle; however, some tired easily due to underlying heart conditions or inadequate strength.
- On average, babies settled at consuming four ounces of milk by the time they were six months old.
- *Tip - Try not to prolong feeding beyond 30 minutes. Any longer than that, your child may be burning more calories than they are consuming.*
- *Tip - Dr. Browns bottles are vented and keep air from getting to the baby, which helps reduce reflux. [\[Link to website\]](#)*
- *Tip - Some families introduced slow-release sippy cups at an early age when sucking remained a challenge. These cups release some fluid without sucking, but the more the child sucks, the more fluid is released. [\[Link to website\]](#)*

Survey results:

Toddler-childhood solid food issues

- Many children transitioned from bottle feeding to solid food.
- Some families found bottle feeding was the most successful method for their child to reach their daily nutritional needs.
- Some families found food with strong flavors, tough textures and/or cold temperatures are beneficial for pleasure tasting due to challenges with diminished oral sensory.
- Some children find it challenging to chew and swallow due to small airways and large gums which prevents optimal breathing.
- When transitioning to solid foods a majority of families reported the most successful tastes and textures were:
 - *Mashed potato and Infant cereals (e.g. rice cereal)*
 - *Puddings (e.g. Boost [\[Link to website\]](#))*
 - *Yogurt*
 - *Soup broths and water*
 - *Gravy*
 - *Stage 2 baby foods with or without cereal added to it.*
 - *Blended/pureed foods with a slightly runny consistency and texture of yogurt.*
 - *Soft ice-cream or homemade milkshakes with a scoop of peanut butter.*

Survey results:

Toddler-childhood solid food issues (continued)

- When children were not receiving an adequate amount of calories, some parents supplemented solid foods with Pediasure. [\[Link to website\]](#)
- Some families also allowed their child to suck on the below snacks to stimulate their taste buds.
 - *Lollipops (sometimes dipped in water)*
 - *Infant cookies, biscuits or crackers*
 - *Pretzels or Fritos*
 - *Ice-pops (frozen soup broth or Pedialyte for a salty taste)*
 - *Smoked meats*
- *Tip – Some children are seen regularly by an Ears, Nose and Throat doctor (ENT) for recommendations on food restrictions due to upper airway narrowing.*

Survey results:

Toddler-childhood solid food issues (continued)

- *Tip – One family's recipe focused on adding protein and fat to their child's diet.*

1 Egg

1 tbs. Milk

2 tbs. Heavy Cream

2 tsp. Butter

Beat the egg with the milk. Melt butter in a frying pan and turn heat to medium. Scramble the egg/milk combination very lightly, until just cooked through keeping heat on medium to low. Pour heavy cream in a small microwave safe bowl and heat for 12 seconds. Combine the lightly cooked eggs and warm heavy cream into a small blender. Blend for 10-15 seconds. The results are eggs that are totally pureed with a whipped texture (similar to yogurt).



Survey results: Gastrointestinal (GI) Considerations

- It is important to consult with a Gastroenterologist (GI doctor) as the majority of families struggled with GI related issues.
- The majority of children had a radiological/barium swallow study, which was referred by a GI doctor, to better understand the strength and function of their swallow.
- A GI doctor may also recommend a gastric empty study to better understand the time it takes food to travel from the stomach to the intestines.
- Some children had symptoms of either GERD/reflux (Gastroesophageal Reflux Disease, acid reflux, heartburn) or aspiration/leakage in the trachea.
- Some children received a surgical procedure called a fundoplication (aka Nissen) to reduce vomiting or the symptoms of GERD.

Survey results:

Gastrointestinal (GI) Considerations (continued)

- *Tip - The following medications (or thickening the formula) may help control the symptoms of reflux:*
 - Zantac
 - Nexium
 - Prevacid
 - Protonix
 - Mylicon
 - Carafate
- *Tip - Reglan is a medication used by some families to help speed up digestion. When a child has a delayed gastric empty of stomach contents to the intestinal track this medication can increase the motility.*
- *Tip - For aspiration/leakage in lungs/trachea, discuss the best path forward with your GI specialist. Some families did continue to feed by mouth for pleasure tasting instead of nutrition.*
- *Tip - If constipation is causing a decrease in appetite some families found these medications to helpful:*
 - MiraLax
 - GlycoLax

Survey results: Infant Age Oral Stimulation

- Several children enjoyed teething toys or objects that greatly reduced oral hypersensitivity. [\[Link to website\]](#)
- Some tools used to provide oral stimulation and sensory input that kids love are:
 - Z-Vibe [\[Link to website\]](#)
 - Nuk Brush [\[Link to website\]](#)
- *Tools that worked to clean the gums and introduce tastes:*
 - Finger brush for gums (with or without infant toothpaste) [\[Link to website\]](#)
 - Flavored toothettes [\[Link to website\]](#)
- Many families worked with their children on tongue movements below to strengthen their entire mouth while having fun:
 - *Blowing raspberries*
 - *Sticking tongue out*
 - *Clicking tongue on roof of the mouth or side of the cheek*
- Feeding therapy was extremely helpful in strengthening the child's mouth, tongue and swallow. It also helped some families with the gag reflex and improved the sucking strength/motion which is needed at a young age.



Survey results: Pediatric Age Feeding Therapy

- Continue infant age techniques with graduated goals such as sound making with proper lip closure.
- Introduce textures and tastes to improve oral stimulation and sensory input.
- Speech/Feeding therapists used special spoons that allowed the child's mouth to form better over the spoon, stretching out the child's mouth and closing the lips together to take food off.
- Some families used speech therapy primarily for speech purposes only and did not incorporate feeding therapy.
- Speech/Feeding therapy was extremely helpful in strengthening the child's mouth, tongue and swallow. Speech therapy seemed to help distinguish between behavioral and technical challenges.
- Tip – Keep feeding enjoyable and try not to force it or get frustrated. Your child may respond better with small amounts of different things to taste while receiving nutrition via a bottle or tube feeding.
- *Tip – Allow your child to suck the food off the spoon. Don't remove the spoon too early.*
- *Tip – Some children advanced to sucking liquid from a straw.*



Survey results: Feeding tubes

- When a child with I-Cell is not getting enough nutrients, such as during an illness, a temporary feeding tube may be necessary.
- Some children progressed from a temporary to a permanent feeding tube.
- Tube feeding can be delivered in different methods (bolus or continuous drip) or speeds depending on the tolerance and needs of your child.
- Tube fed children need periodic release (venting) of stomach gases to prevent aspiration and discomfort of gas build-up. Venting can occur manually using a large syringe or during feeding using a Farrell Bag. [\[Link to website\]](#)
- The permanent feeding tube brand used by the majority of families is the Mic-Key Button. [\[Link to website\]](#)

Survey results: Feeding tubes (continued)

- Tip - Consult with a doctor to determine feeding tube options for your child. Many families found it most tolerable for their child to start enteral pump tube feeding slowly and increase the pump rate gradually.
- Tip – Although it is always preferable to have your child take food by mouth, the point may be reached where a permanent tube is the most practical and beneficial approach to making sure your child is properly nourished.
- *Tip – Prolonged periods of not taking food by mouth can cause oral aversion, while nasogastric (NG) feeding tubes can decrease a child's suck/swallow mechanism. Some families found it beneficial to continue short periodic spurts of bottle feeding as practice and strength building for their child.*
- *Tip – If faced with irritation or infection of the skin around the gastrostomy tube site some families found the below to work:*
 - Clean the site twice a day and dry thoroughly.
 - Use a topical cream such as Silvadene [\[Link to website\]](#) or Triamcinolone [\[Link to website\]](#)
 - Cover the site with a gauze pad.
- *Tip – Nasogastric feeding tubes may cause irritation and breakdown of the facial skin, so it is important to keep the skin clean and dry. And find a tape that does not irritate your child's skin.*



Types of Feeding Tubes

- NG - Nasogastric Tube Feeding (tube enters from the nose & ends in the stomach) - Used for temporary or short time tube feeding, although some families have used it more permanent. These tubes do not require surgical placement.
- NJ - Nasogastric Jujunal Tube Feeding - (tube enters the nose & passes through the stomach and ends in the Jujunal (middle of the small intestines) - Used for temporary or short time tube feeding when a child's stomach can not tolerate a NG tube feeding. Some reasons for not tolerating NG feeding are severe gastric reflux, inadequate gastric motility and high risk of aspiration. These tubes require interventional radiological placement, but do not require surgery.
- G - Gastrostomy Tube Feeding - Used for long term enteral feeding and avoid irritating the nasal passage, esophagus, trachea and do not interfere with breathing or cause skin irritation. These tubes are surgically placed directly into the stomach with a button at skin level.
- GJ - Gastrostomy Jujunal Tube Feeding - Used for long term tube feeding when a child's stomach can not tolerate G tube feeding. Some reasons for not tolerating G tube feeding are severe gastric reflux, inadequate gastric motility and high risk of aspiration. These tubes require interventional radiological placement, but do not require surgery.



Survey results: Final thoughts on feeding

- Being underweight or overweight are common issues in children with I-Cell. It is important that they eat according to their height/growth pattern and not in an effort to increase growth.
 - Being overweight can affect breathing and mobility for children with I-Cell.
 - Due to caloric storage, they have an extremely hard time losing weight.
- Families should talk to their physicians and or nutritionist about daily caloric needs and recommendations to increase/decrease weight.
- The majority of families used Pediasure (with or without fiber). Other formulas used:
 - Bright Beginnings Soy Beverage
 - IsoMil Soy
 - Enfacare by Enfamil
 - Nutramigen infant formula
 - Peptamin Jr. Pediatric



Survey results: Final thoughts on feeding (continued)

- Some families used Pedialyte for a separate feeding or mixed it with formula to reduce caloric intake while still hydrating their child.
- Work with a therapist (speech, physical or occupational) to find appropriate positioning for your child while eating. Positioning needs may change based on age and strength so you should consult with a therapist periodically for help.
- *Tip - Children enjoy being part of the family and eating at the dinner table.*
- *Tip - Don't stress out over feeding.*
 - *"As parents, it was tough to give up that battle. But it is important to understand that there are so many battles to fight with this disease that it is easy to become overwhelmed with the various details. If there is a simpler solution, like bottle feeding with an adequate dietary supplement and your child is happy and maintaining weight, then go with that. Our family life drastically improved when we stopped trying to force solid foods."*
 - *"Don't force it and let them do it on their own. Try to make it "a game" or fun and the children will not realize they are eating and stress over it!!"*



Website Addresses

Below are helpful links and website addresses of products discussed throughout the slides.

[Dr Browns Bottles](#)

<http://www.handi-craft.com/products/bottles.htm>

[Slow Release Sippy Cups](#)

<http://www.babyearth.com/the-first-years-take-toss-spill-proof-cups-7oz-6pk.html>

[Boost Pudding](#)

<http://www.nestle-nutrition.com/products/Product.aspx?ProductId=d01ab52e-5352-4f05-8a0a-5501410044ea>

[Pediasure](#)

<http://pediasure.com/>

[Teething toy sample](#)

<http://www.beyondplay.com/ITEMS/T339.HTM>

[Z-Vibe](#)

<http://www.beyondplay.com/ITEMS/T928.HTM>

Website Addresses (continued)

Nuk Brush

<http://www.superduperinc.com/products/view.aspx?pid=om320>

Gum brush

<http://www.babyearth.com/gerber-tooth-gum-cleanser-infant.html>

Toothette

<http://www.allegromedical.com/personal-care-c532/toothette>

Farrell Bag

http://www.medplususa.com/list-product_info-p-Corpak Farrell Valve Bag Pressure Relief System 30 Case-pid-11729.html

Mic-Key Button

<http://www.mic-key.com/index.asp?page=product>

Silvadene

<http://www.drugs.com/cons/silvadene-topical.html>

Triamcinolone

<http://www.drugs.com/triamcinolone.html>



Glossary

- **Aspiration**

- Aspiration is the sucking in of food particles or fluids into the lungs (food or fluid going down the breathing tube instead of the food tube) usually due to reflux or vomiting. Aspiration pneumonia is the infection of the lungs due to aspiration.

- **Barium swallow study**

- Barium swallow, or “upper GI series” is an x-ray test, where the patient or child drinks a barium solution (often mixed in milk for children) and then X-rays of the upper digestive system are taken. The barium helps visualize the passage of the liquid or food from the mouth to the stomach by X-rays and is used to diagnose reflux.

- **Bolus Feeding**

- A method of tube feeding when food is delivered in shorter feeding intervals several times a day, but vary by child. The purpose of bolus feeding is to keep your child on a normal feeding pattern. Bolus feeding can be performed by syringe or gravity drip method.

- **Carafate**

- A medication used to treat or prevent ulcers in the intestines.

- **Constipation**

- Constipation is defined medically as fewer than three stools per week and severe constipation as less than one stool per week.

- **Continuous Drip Feeding**

- A method of feeding when food is delivered for unlimited periods of time each day. Feeding schedules differ based on needs of a child. Feeding is typically performed by enteral pump.



Glossary (continued)

- **Esophagus**
 - The food tube that connects the pharynx (throat) with the stomach. When a person swallows, the muscular walls of the esophagus contract to push food down into the stomach.
- **Fundoplication (nissen) surgery**
 - A surgery used for GERD to prevent the acid from going back into the esophagus and allows the esophagus to heal from acid damage. During fundoplication surgery, the upper curve of the stomach (the fundus) is wrapped around the esophagus and sewn into place so that the lower portion of the esophagus passes through a small tunnel of stomach muscle.
- **Gagging**
 - A throat spasm that makes swallowing or breathing difficult.
- **GERD**
 - Gastro esophageal reflux disease (GERD) or commonly known as acid reflux, is a condition in which the liquid content of the stomach regurgitates (backs up or refluxes) into the esophagus. The regurgitated liquid usually contains acid produced by the stomach and can inflame and damage the lining (cause, cause esophagitis) of the esophagus.
- **Gastroenterologist**
 - A physician who specializes in the diagnosis and treatment of disorders of the gastrointestinal tract, including the esophagus, stomach, small intestine, large intestine, pancreas.



Glossary (continued)

- **Gastric Emptying Study**
 - A gastric emptying study evaluates the emptying of food from the stomach. For a gastric emptying study, a patient/child eats a meal in which the solid food, liquid food or both are mixed with a small amount of radioactive material. A scanner is then placed over the stomach to monitor the amount of radioactivity in the stomach for several hours after the meal. In patients with abnormal emptying of the stomach, the food and radioactive material stay in the stomach longer than normal before emptying into the small intestine.
- **H2 Antagonists**
 - A group of drugs that reduce the production of stomach acid and are often used to treat GERD or acid reflux. Examples include Zantac, Tagamet or Pepcid that are available in liquid form.
- **Interventional radiology**
 - Continuous X-Ray machine. Allows a technician to watch the movement of a procedure on an X-Ray machine.
- **Oral Aversion**
 - Reluctance or refusal to feed or eat. A familiar example of oral aversion is a baby's refusal to breastfeed. Prolonged intravenous or nasogastric feeding may contribute to the development of oral aversion.
- **Otolaryngology**
 - A physician specialized in diagnosing and treating diseases of the head and neck, especially those involving the ears, nose, and throat (ENT). Also called an ENT, ENT doctor, or ENT physician



Glossary (continued)

- PPI's (Proton pump Inhibitors)
 - Proton pump inhibitors reduce the production of acid by blocking the enzyme in the wall of the stomach that produces acid. These group of drugs are also often used to treat GERD, acid reflux or stomach ulcers. Examples include Prilosec, Prevacid, Aciphex, Protonix, Nexium and Zegarid. Some of these formulations may be available in liquid formulation whereas others are available as granules or tablets to be given with applesauce.
- Reflux
 - See GERD
- Reglan
 - Reglan or Metoclopramide is a medication used to promote or speed emptying of food from the stomach and facilitate the passage of food through the gastrointestinal tract. This medication is available in liquid formulation.
- Speech therapy
 - The treatment of speech and communication disorders. The approach used depends on the disorder and may include oral (mouth/tongue) exercises to strengthen the muscles used in speech and swallowing. Speech therapy is conducted by a specialist known as a speech therapist.
- Trachea
 - Also known as the windpipe or breathing tube that connects with the lungs. The openings of the esophagus and the larynx (voice box) are very close together in the throat. When we swallow, a flap called the epiglottis moves down over the larynx to keep food out of the windpipe.