MMIHS

Newsletter (Ed. 6, February 2022 - "*Uncharted Territory*")

Greetings

I hope you are doing well and that you all have had many happy moments already in this new year of 2022! As today's world continues to challenge us in ways we never could have imagined, I am so very proud to be a part of a group that is the very meaning of tough, full of families who simply don't give up. I had the honor to sit around a table in Texas with two of these MMIHS families this past month. I felt right at home as I swapped stories with them, while watching the very sweet bond the two MMIHS girls have created with one another. Each time I am able to personally meet families like this, I am reminded just how lucky we are to have each other. I always chuckle inside as I think about everyone else's conversations around wherever we meet. Most are surely not conversing about TPN, hospitals, g-tubes, ostomy bags, central lines, etc.. These topics are uncharted territory to most, but are exactly what makes our relationships with one another so unique and special.

As I was drafting this newsletter edition, this theme of uncharted territory kept coming up. According to the dictionary, uncharted territory refers to a place not yet explored or unfamiliar situations in general. I have found myself many times throughout our journey with Everly, feeling like I am in uncharted territory, and it's terrifying. When Everly was first discharged from the hospital at three months of age, I remember her GI doctor at the time telling us that a transplant evaluation was something that needed to be considered within the year or so. I remember trying to get that topic out of my mind as quickly as it had come in. I didn't want to think about something so scary. It was again, a completely new topic for our family to uncover and explore. We were once again, in uncharted territory.

Now six years later, I can honestly say that although I have found our family in new territories more times than not, this topic of transplant is still one that makes me feel very unsettled. Maybe you feel the same?

I have come to realize that within this topic of transplant, there are various stages one can find themselves in, and that each stage comes with its own set of experiences, questions, pieces of advice, etc.. For that reason, I chose to focus this newsletter around the topic of transplant because no matter what stage you find yourself in (if you are in one), there is likely another family in that same stage or even further along in the process. My hope is that the expertise and knowledge of those who contributed their journey to this piece, makes you feel less uncharted and more familiar with the process. Although everyone's journey and experience will vary, hopefully you can find some comfort in knowing that there are others out there who are somewhat in your shoes.

Please take a little time to read about these different stages of the transplant process, along with a couple other topics related to MMIHS. A huge thank you to those of you who contributed pieces of your story to this edition, helping to show us that we are not alone! Your input is much appreciated!

The MMIHS Foundation

The MMIHS Foundation board of directors met on Sunday, February 6th to review goals and continue discussing next steps. Below is a synopsis of the conversations that took place.

MMIHS Foundation Goals:

- **Fund MMIHS Specific Grants**
  - **NORD Research Grant** - We are moving forward with the grant and currently hammering out the details of a "Memorandum of Understanding" between the foundation and NORD so that we can ensure the funds we provide will only be used for MMIHS specific research.
  - **Funds for Families Grant** - More information about the grant along with the application will be released on April 1st. The applications will be due on May 1st and reviewed by the board of directors at our next meeting on May 15th. The awardee/s will be announced shortly after!
- **Advocating for MMIHS**
  - **Rare Disease Day - "Hats Off To MMIHS"**
    - The purpose of this nationally celebrated day is to raise awareness about rare diseases and their impact on patient's lives.
    - In order to continue to raise awareness for MMIHS on this day, we will be holding a "Hats Off to MMIHS" fundraiser. Please see the following page for more details and consider participating!
- **Conference Attendance**
  - **Medical Conferences** - We are hoping to sponsor an MMIHS speaker at this year's NASPHGAN conference which will be held in Orlando in October. We are currently reaching out to prospective speakers.

Rare Disease Day, 2.28.22
"Hats Off to MMIHS"

It's important we continue to increase awareness and advocacy for MMIHS on this day, and all year long. Below are some other ways you can continue to participate!

- ☐ Print and pass out the MMIHS Brochure (found on the MMIHS.org website)
- ☐ Educate someone about MMIHS
- ☐ Wear any MMIHS apparel
- ☐ Donate your birthday on social media
- ☐ Hold a private fundraiser
- ☐ Post anything you are comfortable with on social media regarding MMIHS
- ☐ Share our MMIHS newsletter with someone
- ☐ Check in with an MMIHS Family
- ☐ Join us for the next Zoom call (see date and time at the bottom of the newsletter.

Initial Transplant Evaluation

Written by Erin Peterson (Everly's Mom)

What was the process like? - When Everly was discharged from her initial hospital stay at three months old, our GI doctor at the time had recommended getting a transplant evaluation completed around the age of two. So when Everly was nearing that age, we started this process. First, we checked with our daughter's secondary insurance (Medicaid) to see what transplant centers were covered. Omaha, Nebraska was the center they came back with. We had heard many great things about this center and knew of other MMIHS families that had completed their evaluations there, so we felt comfortable moving forward with this location. At that point, our GI doctor in Sioux Falls, SD sent a referral to Omaha for an initial transplant evaluation to take place. Many phone calls back and forth between us and Omaha took place in the months to come regarding dates, scheduling, lodging, insurance, etc.. We worked closely with our social worker to determine what things could be reimbursed. We were able to get our lodging, mileage, and meals partially covered, which helped as this evaluation was scheduled to take place over a week's time. We also needed to converse with our local pharmacy to ensure our TPN and supplies could be delivered the day before we left and that those supplies would get us through the week. We further asked for an extra bag of TPN and bags of hydration, in case something happened to any of the seven bags we were given. I have included a schedule of all Everly's appointments on the next page, just to get an idea of what took place that week. In between our appointments, we did our best to get out and do some fun things like the children's museum and zoo. However, I remember it being so physically and emotionally exhausting that oftentimes, by the end of the day, all we wanted to do was relax and sleep. Below I have compiled a couple pieces of advice I would have for anyone going through this initial stage. I hope it helps!

1. Make sure to bring enough items to occupy your child. There are many times where you want to be as present as possible with the information being provided, instead of entertaining your child.
2. Make sure to write down any questions you have ahead of time. Also, write down the topics that were discussed in each meeting. There is so much information, it's hard to take it all in at once! You can always refer back to notes.
3. Take time each night to digest the information and write any new questions you have for the following day. Also, make sure to plan something that night you can look forward to. This personally helped get us through the exhausting days. For Brody and me, it was a Netflix show and takeout each night!
4. Be prepared to be emotionally exhausted. I remember specifically at our Psychology Appointment being asked, "How do you feel about the idea of a transplant?" and doing everything in my power not to break down and cry right then and there. It's a lot, and it's ok to not feel ok. Make sure to take some time to process those feelings in whatever way you need to.

Initial Transplant Questions

Written by Dana Werner (Sam's Mom)

What are some questions to have answered during the review? - What organs need to be listed for transplant? Do you think he/she will need a liver transplant (sometimes the Dr. doesn't know until he/she "gets in there" and sees the liver)? Once my child is listed, how long will the wait be? What do we need to do to prepare for the transplant? How long will the surgery last? How long do you anticipate recovery will take? What factors will determine how long my child is in the ICU? What Rxs will my child be on following the surgery? Is this the most optimal time for the transplant? Can we wait longer until he/she is older? Are there any options other than transplant at this time?

How do the doctors decide which organs will be listed? - Likely based on the amount of motility in the stomach and small bowel; liver would depend on the amount of disease; pancreas is attached to the small bowel, so it would be transplanted; a lot depends on lab results.

How do I go about selecting a transplant center? - Ask GI and parents whose kids have had transplants for a recommendation.

What are the choices and how do I know which ones are good? - Ask for firsthand recommendations; google/yelp search to find out ratings of centers

How do I get my child listed for transplant? - Usually your GI will contact the transplant center and talk doctor to doctor (discuss medical necessity) and then a transplant coordinator will contact you.

How long will my child have to wait for the transplant? - Sam was on the list for six months. It varies with each child and can also depend on age and number of organs needed.

Initial Transplant Evaluation (Example Schedule)

Tuesday, June 20, 2017

7:00 am Central Line Draw - University Tower Treatment Center - UTTC-University Tower Treatment Center, Level 3 of Main Hospital, east past C Store toward Wittson Hall. The University Tower Treatment Center is on your right (shares space with the Geriatric Clinic)
- **Special Instructions: No food or drink after MIDNIGHT with the exception of water.**

8:00 am Chest x-ray - Diagnostic Center, Level 1 of Durham Outpatient Care Center.

8:00 am X-rays For Bone age - Radiology, Level 1 of University Hospital.

8:30 am Barium Enema - Radiology, Level 1 of University Hospital.
- **Special Instructions: No food or drink after MIDNIGHT. Follow the attached prep. Contact Nurse at 402-559-5000 with questions. - Because you may receive medication or contrast with this procedure, please call the screening nurse in advance at 402-559-2555 or 800-905-9883, Mon-Fri from 8 AM-5:30 PM with your COMPLETE med list or have your medication bottles handy for reference.**

10:30 am Social Service Consult - Mandy Pflaster - Multi Organ Clinic, Level 2 of Lied Transplant Center.

1:00 pm Pedi Gastroenterology Consult - Ruben Quiros - Multi Organ Clinic, Level 2 of Lied Transplant Center.

1:00 pm IRP Clinic Visit - Multi Organ Clinic, Level 2 of Lied Transplant Center.

1:30 pm Transplant Coordinator - Multi Organ Clinic, Level 2 of Lied Transplant Center.

2:30 pm Nutrition Consult - Angela Iverson - Multi Organ Clinic, Level 2 of Lied Transplant Center.
- **Special Instructions: For small bowel patients, please bring your most recent TPN prescription.**

Wednesday, June 21, 2017

7:00 am Central Line Draw #2 with ABO- University Tower Treatment Center - UTTC-University Tower Treatment Center, Level 3 of Main Hospital, east past C Store toward Wittson Hall. The University Tower Treatment Center is on your right (shares space with the Geriatric Clinic)

8:00 am Ultrasound-Abdomen - Radiology, Level 1 of University Hospital.
- **Special Instructions: Adults Nothing by mouth 8 hrs prior to test. Children Nothing by mouth 4 hrs prior to test if possible.**

9:30 am Echocardiogram - Heart Center, Level 2 of Durham Outpatient Care Center by Gift Shop.

10:30 am Doppler Venous Studies - Vascular Lab – The Heart Center, Level 2. Report to DOC.

1:00 pm Occupational Therapy Consult - Multi Organ Clinic, Level 2 of Lied Transplant Center.

1:30 pm Child Development Consult - Multi Organ Clinic, Level 2 of Lied Transplant Center.

Everly holding her own blood sample after one of her blood draws.

Initial Transplant Evaluation (Example Schedule)

Thursday, June 22, 2017

7:00 am Central Line Draw - University Tower Treatment Center - UTTC-University Tower Treatment Center, Level 3 of Main Hospital, east past C Store toward Wittson Hall. The University Tower Treatment Center is on your right (shares space with the Geriatric Clinic)
- **Special Instructions: No food or drink after MIDNIGHT with the exception of water.**

8:00 am History & Physical - Multi Organ Clinic, Level 2 of Lied Transplant Center.

9:00 am Financial Counselor Consult - Michelle Stilley - Multi Organ Clinic, Level 2 of Lied Transplant Center.

10:00 am Child life assessment - Multi Organ Clinic, Level 2 of Lied Transplant Center.

11:00 am Psychology Appt - SSP Building, 5th floor. Entrance to SSP is located on the 3rd floor of University Hospital between cafeteria entrance and the Lied Transplant Center.
- **This appointment will include an interview, testing, and a behavioral questionnaire. Please bring reading glasses and/or hearing aides if you require them. Family members or friends who have accompanied you to the evaluation should be present for the interview, if possible.**

1:00 pm Pharmacy Consult - Karen Bohnenkamp - Multi Organ Clinic, Level 2 of Lied Transplant Center.

Friday, June 23, 2017

7:00 am Central Line Draw - University Tower Treatment Center - UTTC-University Tower Treatment Center, Level 3 of Main Hospital, east past C Store toward Wittson Hall. The University Tower Treatment Center is on your right (shares space with the Geriatric Clinic)
- **Special Instructions: No food or drink after MIDNIGHT with the exception of water.**

8:30 am Upper GI/Small bowel series - Radiology, Level 1 of University Hospital.
- **Special Instructions: Adults Nothing by mouth 8 hrs prior to test. Children Nothing by mouth 4 hrs prior to test if possible.**

12:30 pm Transplant Surgeon Consult - Multi Organ Clinic, Level 2 of Lied Transplant Center.

Taking a much needed rest in the hospital lawn, after a day full of tests, tests and more tests!

Transplant Evaluation - Follow Ups

Written by Sarah Turman (Ava's Mom)

Once the initial transplant evaluation is completed, the transplant team at your chosen transplant center will decide whether to recommend you to be actively listed at that time, to continue to be followed and re-evaluated annually (or more often as needed), or to only return for re-evaluation and possible listing if circumstances drastically change or health declines. The transplant team at Children's Hospital of Pittsburgh (UPMC) also explained to us that it is their job to ensure that every other avenue and treatment option has been attempted prior to actively listing for transplant, as it should be a "last resort" and/or the risks of the transplant and challenging long term recovery should be outweighed by the need and overall health and quality of life of the patient. That is the view of Pittsburgh's team (as we understood it) but it seems all teams/centers have a slightly different approach. One thing I did not initially realize was that transplant is not a "you are in or you are out" or a "it's now or never" type of thing. The listing status can change and adjust based on need. For our family, our daughter has been listed for multivisceral transplant at various points in time since 2014, but currently she is not listed (due to overall positive health and the current quality of life outweighing the transplant risks). For our family, at this time with not being actively listed, our daughter is still followed by the transplant team and is one of their patients. This means we have annual appointments in Pittsburgh (pre-COVID was in person but we have done virtual appointments since COVID). The transplant team also continues to work closely with our local GI team. One of the benefits of the transplant team being involved (even though not currently listed) is having another set of medical experts looking at possible solutions or adjustments to help our daughter's day to day treatment and functioning (since we all know each person is such a unique individual and there is no "standard" way to treat/manage MMIHS). If needed, since we are still patients of the transplant team and have already completed the formal initial evaluation, if circumstances require listing in the future, she can quickly and easily have her "status" changed to an active listing.

Transplant Evaluation - Follow Ups (Example Schedule)

Wednesday, June 12, 2019

10:00 am Patient Instructions
Lab - Day 1
Special Instructions: No food or drink after MIDNIGHT with the exception of water.
Location:Buffett Cancer Center - ground floor. Proceed to the Fast Track check-in area

11:00 am Pharmacy Consult - Karen Bohnenkamp - Multi Organ Clinic, Level 3 of Lied Transplant Center.

11:15 am Financial Counselor Consult - Michelle Stilley - Multi Organ Clinic, Level 3 of Lied Transplant Center.

12:30 pm Nutrition Consult - Multi Organ Clinic, Level 3 of Lied Transplant Center.
- Special Instructions: For small bowel patients, please bring your most recent TPN prescription.

1:00 pm Transplant Coordinator - Multi Organ Clinic, Level 3 of Lied Transplant Center.

3:00 pm Pedi Gastroenterology Consult - Multi Organ Clinic, Level 3 of Lied Transplant Center.

Thursday, June 13, 2019

8:30 am Patient Instructions
Ultrasound-Abdomen
No food or drink 2-4 hours
Location:
Radiology Department on the first floor of the Buffett Cancer Center
Dept Phone: 402-559-1900

9:00 am Patient Instructions
Lab - Day 2

Special Instructions: None

Location: Buffett Cancer Center - ground floor. Proceed to the Fast Track check-in area

10:00 am Social Service Consult - Multi Organ Clinic, Level 3 of Lied Transplant Center.

11:00 am Psychology Appt - SSP Building, 5th floor. Entrance to SSP is located on the 3rd floor of University Hospital between cafeteria entrance and the Lied Transplant Center.
- This appointment will include an interview, testing, and a behavioral questionnaire. Please bring reading glasses and/or hearing aides if you require them. Family members or friends who have accompanied you to the evaluation should be present for the interview, if possible.

1:00 pm Doppler Venous Studies - Vascular Lab – The Heart Center, Level 2. Report to DOC.

2:30 pm Transplant Surgeon Consult - Alan Langnas - Multi Organ Clinic, Level 3 of Lied Transplant Center.

Sam's Transplant Story (p.1)
Written by Dana Werner (Sam's Mom)

Samuel Jon Werner was born on January 4, 2000 in Austin, TX. I started leaking amniotic fluid four days earlier, and was put on bed rest in the hospital. That was the day we first had an inkling something might be wrong with him. The doctor saw an enlarged bladder on an ultrasound. It wasn't until after he was born that we knew the bowels were involved. Shortly after his birth a geneticist evaluated him and delivered the diagnosis of MMIHS to my husband, Dennis, and me. Later, a NICU doctor told us that the syndrome was usually fatal and Sam had about six months to live — he would die of liver failure due to the TPN and not being able to process anything orally (no omegaven in those days). On day three of life, he had surgery to place an ileostomy.

My husband and I quickly decided we were not going to accept the grim prognosis, and had him transferred to Texas Children's Hospital (TCH) in Houston. Doctors there were much more positive and gave us hope. He stayed at TCH for three months. We learned during that time that he likely had little-to-no intestinal motility, as he threw up almost all the breast milk/formula he ingested. We had several close calls, as he had many line infections and became seriously ill.

TCH referred us to a wonderful GI in Austin who managed his TPN/lipids and meds. He had several surgeries to "untwist" the bowels and to remove parts of them. Again, several close calls.

When Sam was seven months old, at our GI's suggestion, we took him to Pittsburgh for a motility study. Several tests indicated that he had basically no motility in the stomach or small intestines. The doctor who performed the motility study recommended placement of a g-tube and j-tube, so at eight months of age he underwent that surgery.

He was unable to take anything by mouth the first three years of his life, 24/7 TPN. Gradually, he became sicker and sicker due to sepsis. The doctors referred to it as "bacterial translocation." He had "intestinal non-function causing bacterial overgrowth escaping the intestines." Rotating antibiotics to address the problem was not a long-term solution. So, our GI got us in touch with Pittsburgh again, and Sam was placed on the transplant list.

(P.2) Transplant was the only option to save his life. After six months on the list, we got the call. In March of 2003 (at age three), Sam underwent the 14-hour surgery. The stomach and small intestines, along with the pancreas, were transplanted. The spleen, gallbladder, and colon were removed, as they were necrotic — diseased beyond the point of function.

Sam spent two months in the NICU and two additional months in a Ronald McDonald House in Pittsburgh following the transplant. He had several occurrences of high fevers (no permanent damage). He was on a fairly high dose of prograf to prevent rejection. In all, he was on about ten different meds.

Soon after he was discharged, and we were back home in Austin, we received a call from Pittsburgh that we needed to return, because he had contracted Hepatitis C, likely through a blood transfusion. We stayed in Pittsburgh for about two weeks. The doctors there decided that our local GI could continue the treatment at home. He received treatment for one year and was cured of Hep C.

Sam had started eating and processing food while in Pittsburgh. He started on soft foods and progressed to Cheerios (one-at-a-time!) fairly quickly. He showed no oral aversion. He retained his central line for six months, and was on one bag of hydration per day. After six months, the line was pulled, and all that remained was his ileostomy bag.

Over time, Sam's meds were reduced to only the three he takes today — prograf, bactrim, and penicillin. He has been hospitalized only once since the transplant. When he was twelve, he contracted RSV, which was treated and cured after six days of hospitalization. Sam's medical routine today consists of labs every three months and a few doctor visits a year — one with his GI, one with the transplant surgeon in Seattle (his surgeon moved from Pittsburgh to Seattle Children' Hospital, and we followed him), one with his urologist, and one with an Infectious Disease doctor.

Sam's Transplant Story (p.3)

Today, Sam, at 17, is a happy, healthy, young man. He eats anything and everything, with pizza being his favorite! He takes care of his ileostomy bag himself. He self-caths three times a day. We do nothing for him medically, other than ensure that he takes his meds. He goes to school, church, the movies, and other social activities. He is currently running on his school's cross country team.

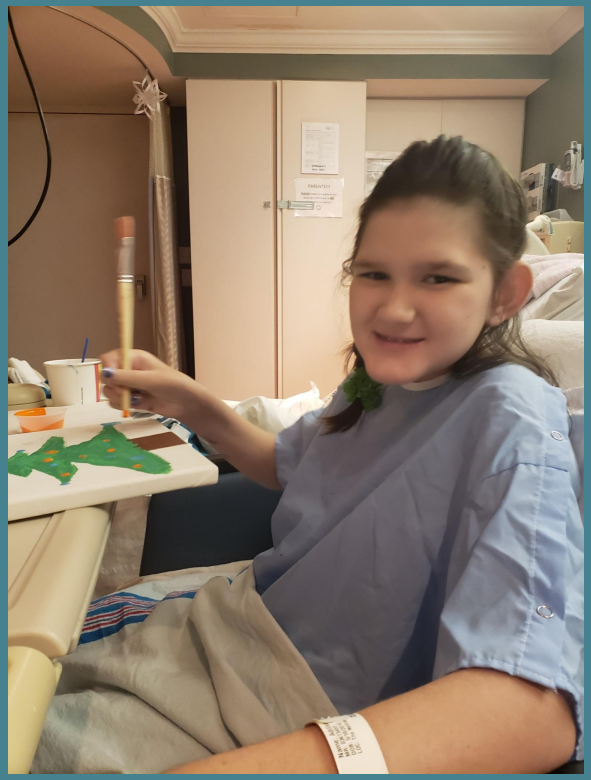
Sam's long-term prognosis is good. At one appointment (annual transplant follow-up) I told our doctor that I wanted Sam to outlive me. His response was, "That will happen." Doctors tend to emphasize that if a patient makes it to 5 years post, they are pretty much out of the woods. No doctor has ever mentioned to us the possibility of another transplant. There are possible future complications from only one med he takes — the prograf (the anti-rejection med). Since it causes him to be immunosuppressed, he is more susceptible to infections/illnesses such as viruses, bacterial infections, the flu, etc. If he catches something, it may take him longer to fight it off. He does get a flu shot every year, and has never had the flu or any bacterial infection! I think the low dose of the two antibiotics he is on (one is because he has no spleen) protects him from such things. Docs have told us that down the road he may be more prone to cancer. But, the risk is low, and I just try not to think about that.

Our family life today is so much easier and more pleasant than before the transplant. We are so glad he had the transplant, and blessed that it was successful. Our family has a strong faith. Sam was prayed for by people from all over the world. We know that God's grace helped us through the journey, and is responsible for Sam's health today. Sam is our "miracle son."

Addison's Transplant Story (p.1)

Written by Christine Martins (Addison's Mom)

After approximately 3 months on the list for a liver, pancreas, small and some large intestine, on November 3, 2020, as the United States was watching to see who was winning the election we got the call that would forever put us on a new path. As I was sitting down at my desk, my phone rang. Right when I saw the number I knew it was "The Call" especially since Omaha was 2 hours ahead and it was 10 pm there. Certainly the team wouldn't be calling at this hour to just see how we were doing. I was right. After we accepted, calls were made and then we were told to be at the airport in approximately an hour and a half. As I was rushing around getting last minute items to pack, we made phone calls and a few friends and family came to send us off.



Doing a Christmas craft in the hospital

Addison's Transplant Story (p.2)

After two flights, we arrived in Omaha from California early the next morning. We were told rounds would start around 9:30- just enough time for a nap as I was already exhausted. The loud air ambulance and the anxious excitement didn't let me nap. In the meantime, we were told an OR was booked for 10 pm, but it could be sooner. I got the idea that they don't really tell you too much because they don't want you to know where the organs are coming from and partly because they don't know themselves. The day was mostly spent waiting, making phone calls and resting up for what we knew was coming- some long days ahead.

At around 5 pm they took her down to pre-op. As we waited, my mind was going in so many places. I looked over and Addie was playing on my phone with seemingly not a worry in her mind. To her, this was just another procedure or surgery. She gives me a thumbs up, like she did on the plane ride over while strapped to the stretcher. She doesn't understand the immensity of what is about to happen and in what ways her life could change. My thought to ask how she was doing and what she thought about all this, passed. She is calm and that brings some calm to me. After meeting many people who will be with her along the journey, we meet the surgeon. He knows our general surgeon back home and helped with his training. This further calms me. I know she is in good hands and this is meant to be. After they take her back, they tell us they will call with updates so there is no need to stay at the hospital. I decided to find a hotel and wait for my husband there, where we could hopefully rest. Since there was only room on the air ambulance for Addie and I, he had to find the first morning flight. I found a hotel and got situated. He arrived soon after.

Every few hours we would get an update. Things were going great. After approximately 6 and a half hours, they called us and let us know they were finishing up and we would be able to see her soon. We headed down to the hospital to see her. She looked good despite the fact that she looked puffy. They warned us she would look this way as they purposely overloaded her with fluids to help profuse the new organs.

Addison's Transplant Story (p.3)

Addison was on a ventilator for ten days. In the past, she had difficulty coming off the ventilator after surgery so this was not unexpected. They also told us they would keep her sedated for a while as they were planning on taking her back to the OR in a few days since they did not attach the intestines to the stomach and had not given her an ileostomy or colostomy. Three days later, they connected everything and did a washout. After the connection, she had weekly biopsies for four weeks at her bedside to check for rejection. She has not had any rejection and is doing great. She was inpatient for two months, which was about a month longer than planned due to an abscess and perforation in her stomach. She left the hospital on some IV fluids and feeds. After a month outpatient, she was weaned off all fluids and her central line was pulled. We were officially ready to head home.

Today, Addie is doing great and has much more energy. She enjoys eating, although is pretty picky. She is definitely not a fan of fruit or vegetables. She does especially love hamburgers, chicken sandwiches, chicken nuggets, pizza, quesadillas and her favorite snack is croutons. She still receives overnight feeds, but that is definitely much easier than TPN!.

Sarah's Transplant Story (p.1)

It all started back when my mom was pregnant with me. Part way through, she went in for a routine ultrasound which showed that my bladder was enlarged. After further examination, doctors discovered it wasn't draining properly so I underwent two surgeries well in -utero called ("Bladder Taps "). The doctors also thought I had an obstruction of some kind and believed it was relatively a mild problem. I was then born 5 1/2 weeks early on Valentine's Day in 1997 in a small town in Ontario. I weighed 4pounds, 12 ounces. Within 24hrs, I was rushed to Sick Kids in Toronto. I wasn't even 2 days old when I had my first major surgery to attempt to repair my twisted bowel. Immediately following that surgery, my appendix ruptured and had to be removed.

After three more surgeries in two months (looking for bowel obstructions), I was diagnosed with Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome (MMIHS), which basically meant I had a large bladder and my intestine didn't work. I spent the next 5 months in Toronto and was completely being fed intravenously. I started TPN (Total Parenteral Nutrition), which kept me alive, but at a price. Within a couple months, it started to destroy my liver and my pancreas. At that time, my family already knew that I needed a bowel transplant, but due to the TPN, I also needed a new pancreas and liver. In May, my mom was told that my only hope was a multi organ transplant. I was then placed on the transplant list. Luckily on August 6th 1997, my family got the call that there was a match. At 5 months and 24 days of age, I was transported to London Children's Hospital in Western Ontario, where I received a stomach, pancreas, liver, and bowel transplant. The transplant that I had was the first pediatric multi- organ transplant in Canada and the youngest in the world. For that reason, I hold a Guinness world record! I stayed in London until December 1st. When I came home for the first time, I was 10 months old. Today I live a fairly normal life as a young adult. Thanks to my transplant, I have been able to do so many things that I know I wouldn't have been able to do: become a big sister, graduate college, travel the world, spend time with family and friends, raise awareness about organ donation, and also meet many others who have MMIHS.

Sarah's Transplant Story (p.2)

*Be prepared to advocate for your child
*Bring patience
*Keep notes
*Be positive
*Realize before /after transplant, a child's stability can change hour by hour and minute by minute
*Try not to stress too much
*Always ask questions and if you don't understand, ask again. Never say okay and be left unsure
*Ask for help or support from family and friends
*Take care of you. It's hard to remember to sleep and eat. Take a break if you need to
*You know your child best. Follow your gut feeling because it's usually right. Speak up if you need to be heard.

~ Advice from Cindy (Sarah's mom)

Camila Valentina Carrasquel

Written by Barbara Carrasquel (Camila's Mom)

February 2022 - 15 years post-transplant

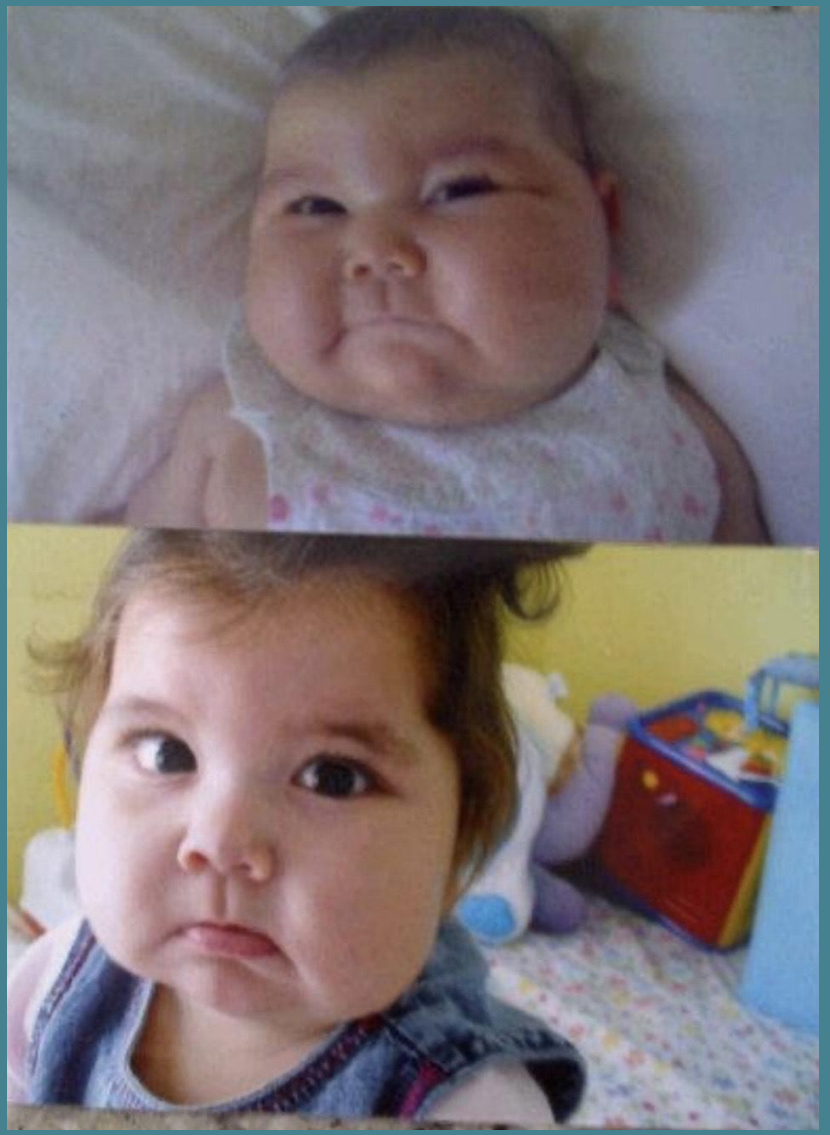
- 7.28.2006: Camila is born.
- 7.29.2006: Camila was officially diagnosed with MMIHS within 24 hours of her arrival. The surgeons at the hospital told us that they had contacted the transplant team in Miami Jackson Memorial Hospital, FL. Camila stays in the NICU until she's heavy enough to be evaluated for a multi visceral transplant (5kg).
- Mid October 2006: Camila is transferred to Jackson Memorial Hospital. We spent three weeks being evaluated. Dr. Kato came to talk to us on a Saturday, so he could sit down with us without interruptions. That meeting lasted over two hours. We had no questions; we didn't know what to ask. It was information overload. He came back the next day to answer questions.
- November 2006: Camila is released with a central line (her only surgery so far), home care, pumps, and many appointments set up for pre-transplant care.
- January 2007: Camila is admitted because her central line was blocked. She was TPN-dependent, and her liver enzymes were really concerning to the team. She looked like a glow bug.
- February 17, 2007:
    - 11am: We received the call to head to the hospital for a possible transplant.
    - Donor arrives at the hospital. Surgeon comes for a last check. He measured Camila's abdomen with his fist, eyeballing her available space. Turns out that most pediatric patients stay in the PICU for a prolonged time due to their abdomen being left open because the organs are too swollen, and they cannot close the incision at the time of the transplant. Organ harvesting is about two hours long. Then they examine the organs, if they are good enough, the transplant is a go. We wait, and wait. We have been in-patient for ten hours waiting to see if the transplant will happen.
- February 18th, 2007 (Sunday):
    - Midnight: In a hurry, the nurse comes into the room. Transplant is a go. She must start fluids with antirejection medicine at this point.
    - 2am: We walked Camila to the OR and said our good-byes. She is six and half months old, her liver is badly damaged due to her TPN and lipids. We know she does not have another chance.
    - 1pm: Main surgeon, Dr. Tomoaki Kato, comes to talk to us. He goes over the surgery. They were able to close her incision. We should be able to see her in the PICU in a couple of hours; the other surgeons were still working on Camila.
    - That same afternoon: We walked in the PICU. Behind the thirty seven different pumps, a respirator, and nurses checking everything, lays a baby girl with pink cheeks. She was not jaundiced anymore. That was our first words to each other.
- February 19th, 2007. Day 1: Camila is progressing beautifully. Surgeon talks to us again. We met Dr. Andreas Tzakis, Director of Transplant. Dr. Tzakis is also one of the first doctors to perform and research this kind of procedure. Camila's transplant was performed by the director of pediatric transplant and the director of transplant.
- February 20th, 2007: Doctors talked about starting to wean Camila off of the respirator. An hour later she's completely extubated and breathing on her own. A process that was supposed to take a couple of days.
- Day 3: Camila was moved to a step down unit because she no longer needed the care of the PICU. This was the first time for a post multi visceral transplant recipient to leave the unit so quickly. At this point, we were waiting for a bed to open on the transplant floor to move again.
- Day 8: Doctors started feeding Camila through her NG-tube.
- Day 19: Surgeon said we could move to the Ronald McDonald house. I advocate and lean on our team of home care, and even the hospital nurses, to let us go home, not the Ronald McDonald House. We lived about an hour away from the hospital. The surgeon, who was Camila's main person to check and approve anything, agreed. I commited to bring her in as many times a week as he wanted to. I just wanted to be home. Dr. Kato said that if I could get all the orders ready to go home, on a Friday, we could go then. Our team moved everything and got us out of Jackson Memorial in record time.

Day 20: We are the first family to leave the hospital, and go home, in less than three weeks post-transplant. Camila has a central line, NG-tube, ileostomy, and colostomy. Camila scar goes across her abdomen, about 72 stitches. Those stay in place for several weeks. All of the previous meds plus the many new medicines are added to our daily schedule. I keep track of all the input/output to have all the numbers ready for the doctors.

- Weekly ileostomies with biopsies, become bi-weekly.
- Week 8: Kidney stents and central line are removed and she is 100% fed via NG-tube (Neocate + pedialyte).
- 15 months post-transplant: Colostomy is reversed. G-tube is placed. Camila's biopsies are bi-monthly; non-sedated; and performed by our favorite GI. Camila refused to eat by mouth for a looong time (+/- 11 years old). That was the only reason behind the G-tube.
- 3+ years post-transplant: Biopsies are now about every six-months to a year, so the team decides it is time to close her ileostomy.

Annual endoscopies. Camila now has annual endoscopies with biopsies. Colonoscopies are rare since we have never had any rejection. Camila follows up yearly with her urologist. Every six months with her GI. Quarterly with her nephrologist. Every year she has an ultrasound of her four(!) kidneys (she has her two native kidneys plus two transplanted kidneys) and liver. A yearly DEXA scan to measure bone density. Bimonthly labs. Her GTube was closed in 2019. She started consuming 100% of her calories and hydration by mouth in early 2018.

This journey was not easy nor lineal, but after she was born, transplantation was our only hope. I always saw her transplant as the turning point. The point where we went from surviving to knowing that there was a future for her, for however long God was willing to let us have her. It felt like I was actually working towards a better life for her. Camila, as her doctors said, has been the "best case scenario". Camila has always been on the better side of every probability that was thrown at us. The time spent on the waitlist, how many times we were going to be called for a possible transplant, time spent in the ICU, step-down unit, on the floor, and/or how many episodes of rejection we would probably face post-transplant.



Top: 4 months old. 100% TPN. (2 months Pre-transplant).
Bottom: 1 year old. 6 months post-transplant. 100% formula fed.

Winter Giveaway

Since this newsletter is focused around the topic of transplant, I thought the winter giveaway could focus around this topic as well. If you would like to participate, please post what stage of the transplant journey your family is in (if you are) and one piece of advice you have for families at this particular stage. Examples of stages include: haven't started, initial evaluation, follow up evaluations, transplant, post transplant, ect…..

On **Monday, February 21st,** a message thread will be posted in the MMIHS support group asking families to make their initial post.

On **Sunday,February 27th**, a winner will randomly be selected and will receive an MMIHS Yeti mug in the mail! (see picture below)

Staying Connected

Family Outreach Coordinator: Sarah Turman is a wonderful attribute in assisting with inquiries from families and providing connections and support. Please don't hesitate to contact her, sarahannturman@gmail.com.

If you have any MMIHS related pictures or information you would like shared on our social media platforms, please feel free to message our Social Media Coordinator, Kristin Gutknecht at any time (kristin.gutknecht@gmail.com).

Zoom Date: Our next Zoom call is scheduled for **Sunday, May 15th (11:00-1:00 CST)** and will again be hosted by Brianna Larson. We will send out a reminder and link closer to the day but please mark this date. We hope you can join us for some or all of it as this is a great way to support one another!

Concluding Thoughts

Thank you for taking the time to read all things related to MMIHS. As I stated before, I truly hope that whatever stage of this journey you find yourself in, the information provided by other families helps you feel like you are a little less in uncharted territory. Know you have people to support you along every step of the way!

Much love to you all,
Erin Peterson