

2023 OMS Conference

Keynote: Alicia Mantz

***WE ARE
RARE***



BUT STRONG

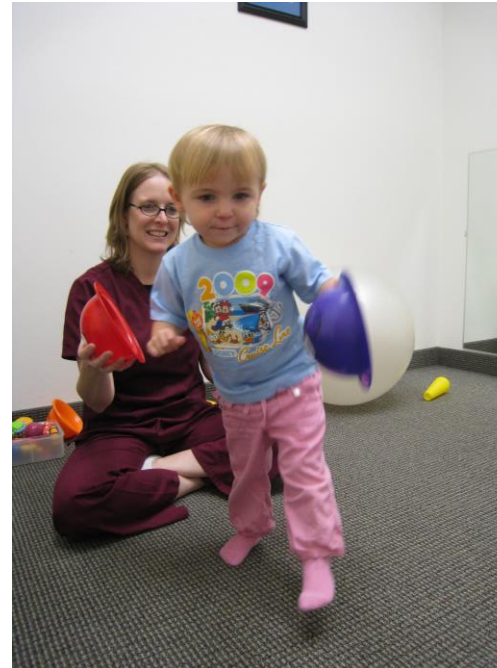
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February 2009

- ▶ Differential Diagnosis of acute post-infectious cerebellar ataxia/cerebellitis
- ▶ Treated with 3 days of IV steroids
- ▶ EEG due to eye movements-placed on Keppra
- ▶ Regressed to 6-7 months old physically
- ▶ PT, 1st steps for the 2nd time

April 2009

- ▶ Symptoms relapse, hospitalized a second time, 3 days steroids
- ▶ All tests normal, waiting on one



May 2009

- ▶ Symptoms relapse again, told to give it time; called for test result
- ▶ May 28- neuro called to tell me her urine was elevated for HVA/VMA, and she might have neuroblastoma
- ▶ CT and MIBG done, nothing found; told to wait and see

June 2009

- ▶ Researched hospitals for oncology and decided to get a second opinion for neurology while there
- ▶ Decided on Boston Children's Hospital
- ▶ Gather all testing and made an illness timeline



Symptoms:

- ▶ Ataxia, falling
- ▶ no longer able to walk
- ▶ tremors
- ▶ eye movements/opsoclonus
- ▶ sleep disturbances
- ▶ rages/outbursts not typical of a 2-year-old; older-emotional swings (Decreased ability to reason); OCD got worse
- ▶ Loudness regulation (too quiet or loud)
- ▶ ataxic dysarthria (motor speech disorder; words sound slurred)
- ▶ Difficulty getting words out/expressive language
- ▶ Headaches
- ▶ Decreased processing and speed
- ▶ Increased fatigue
- ▶ Staring into space, “in her own world”
- ▶ “not her usual self”



July 14, 2009

- ▶ Met Dr. Gorman and returned home with a diagnosis of moderate to severe opsoclonus-myoclonus syndrome (OMS)

Fun fact: Lauren was his 11th patient

- ▶ Began high doses of daily oral steroids (prednisolone) and IVIg every 4 weeks then every 3
- ▶ Opsoclonus originally lasted 30-45 minutes; just sit and hold her while she whined and cried
- ▶ Did video EEG and second opinion of original EEG indicated no seizure activity



May 2010

- ▶ OMS Symptoms: Front tooth from fall 2009 abscessed and removed

June 2010

- ▶ 1st round Rituxan

February 2011

- ▶ 2nd round Rituxan

March 2011

- ▶ 1st Normal HVA/VMA in urine since onset! (dispel? Rituxan changed cells?)

November 2, 2011

- ▶ Down to .5mL once a day but still having symptoms if taper so brainstorming what cause could be. I asked for scan other than MIBG



November 3, 2011

- ▶ Guess what they found on MRI?

A tumor! (upper abdominal cavity near the aorta and vena cava; able to see on 2009 scans once knew where to look)

November 21, 2011

- ▶ Tumor removed and determined to be a ganglioneuroma
- ▶ Complications after tumor removal included pancreatitis and pseudocyst
- ▶ Pancreatitis is not very common in children
- ▶ Spent 6 months inpatient and NPO, moved to Boston after a month



2012

- ▶ ERCP attempted to fix pancreas complications, but 2 attempts were unsuccessful
- ▶ Led to major surgery in March 2012 to bypass to the intestine; gall bladder and appendix also removed
- ▶ Relearn to eat and drink



February 2013

- ▶ Got 1st port

April 2013

- ▶ Ruptured intestines: bike accident; stress dose steroids, No relapse with steroids continued

May 2013

- ▶ 3rd round of Rituxan

November 12, 2013

- ▶ Last dose of prednisolone!



- ▶ Relapse of symptoms in Jan/Feb 2014; did a 3 day course of Dex and began Cell-Cept in March 2014
- ▶ Cell-Cept allowed us to finally wean IVIg 1 week each time over a couple years and we made it to every 6-7 weeks

April 2018

- ▶ Port replaced—OMS relapse due to infection, allergic reaction

July 2019

- ▶ Discontinued Cell-Cept as no longer getting any benefits
- ▶ Had to switch facilities for IVIg due to shortage; denied where had been since 2009

March 2020

- ▶ Relapse as missed IVIg due to insurance saying “experimental or investigational” and no authorization- 3rd party review



December 2020

- ▶ Found a hole in her heart: Atrial Septal Defect (ASD)

2021

Jan-Hospitalized with severe pancreatitis

Feb-ASD repaired, ERCP attempted to fix pancreas-unsuccessful

- ▶ Continued with 1-2 cases pancreatitis every month, watch and see

Nov-Diagnosed Gammaglobulinemia

January 2022

- ▶ ERCP in FL with serotonin-investigative, unsuccessful; determined to need Puestow surgery, planned for summer 2023

September 2022

- ▶ Monthly cases of pancreatitis increased eventually getting to 1-2 cases a week



November 2022

- ▶ OMS Relapse, worst in a long time

December 2022

- ▶ Hospitalized with pancreatitis

February 13, 2023

- ▶ Puestow surgery done sooner due to atrophy and successful
- ▶ Complications included allergic reactions to meds on allergy list

April 2023

- ▶ Port replaced
- ▶ Anaphylaxis in OR, had “rebound effect” (2%)

“Rare”

- ▶ Uh...we’re living it



Some Take-aways:

- ▶ Every OMS Warrior may have a similar but individual path

-Advocate

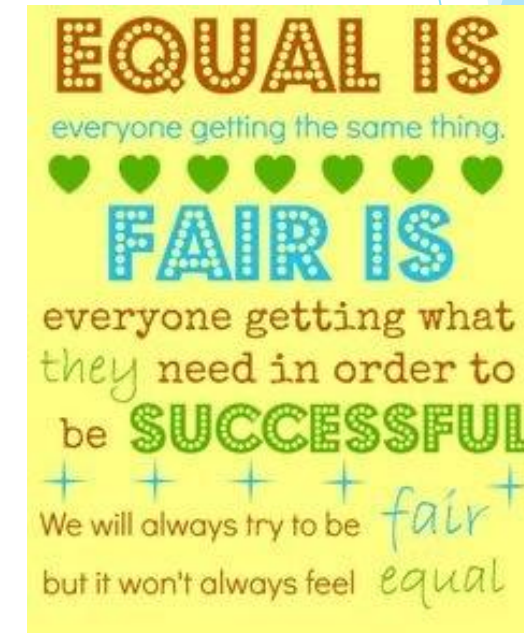
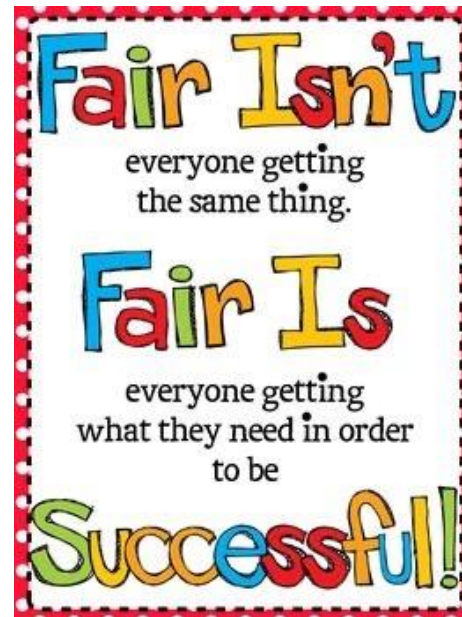
- ▶ Know your child best
- ▶ Be realistic, Be Kind
- ▶ Medical Errors, Allergies
- ▶ Speak to a supervisor; Hospital Advocate

-Mental Health

- ▶ Patient, Parent/Caregiver, Siblings

-Funding

- ▶ State Scholarships-Family Empowerment Scholarship for Students with Unique Abilities (FES-UA)



Academics

- ▶ Lauren repeated K-4; teacher felt she was good academically but socially struggled/possible OMS learning difficulties
- ▶ Abilities appropriate for the grade she is in
- ▶ Meet with teacher(s) before school started/list what to expect
- ▶ Difficulty with handwriting, slower to produce, decreased visual/motor skills (note-taking)
- ▶ 2 years Occupational Therapy (OT)
- ▶ Accommodations were needed more as she got older, and demand became more in school
- ▶ Often takes her 2-3 times longer to do work



▶ Accommodations-- 504 vs IEP

▶ Examples:

1. Preferential seating to avoid auditory and visual distractions.
2. Extra time on assignments and tests, up to 1.5x, and advanced notice time on all assignments.
3. Provide a quiet distraction-reduced place and breaks during testing as needed.
4. When absent, provide copies of classroom notes or lectures, assign student if needed.
5. Allow use of laptop to type notes or written assignments, as needed.
6. Large projects or tasks may need to be broken down into smaller steps.
7. Adjust written assignments and/or allow extra time for completion to demonstrate quality of work over quantity.
8. Allow her to remove herself from class to the nurse for medical attention needs or the restroom as frequently as needed.
9. Advance notice of all exams/tested assignments for academic credit (no pop quizzes).
10. Placement with instructional teachers who are supportive and understanding of her diagnosis and comfortable implementing accommodations on a daily basis.

OMS Awareness and Research

- ▶ The Lauren Mantz Fund for OMS Research
- ▶ Toy/Bear Drive
- ▶ OMSLife Foundation
- ▶ Mother-Son Dance
- ▶ The Lauren Mantz OMS Fellowship Fund



Lauren's Journey

