

Education of children with Sanfilippo syndrome: A guide for educators

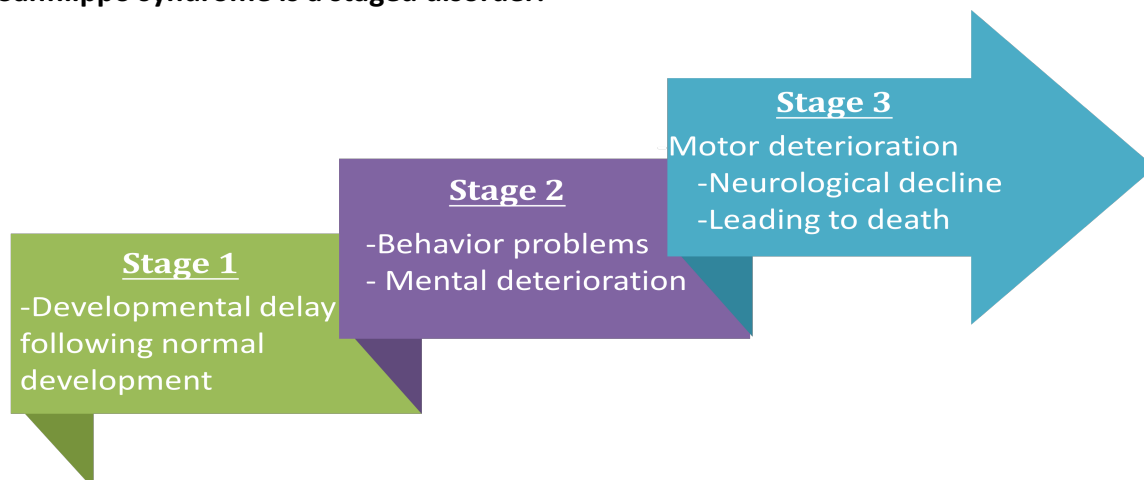
What is Sanfilippo syndrome?

Mucopolysaccharidoses type III (MPS III) is a type of metabolic disorder known as Sanfilippo syndrome. Dr. Sylvester Sanfilippo was one of the doctors in the United States who described the condition in 1963. To date, four different enzyme deficiencies have been found to cause MPS III syndrome, and therefore the condition is described as type A, B, C, or D. There is usually very little difference between the four types of the disease, but the cognitive decline in types B, C, and D tends to be later than in type A.

Background

- Sanfilippo syndrome is also called MPS III (Mucopolysaccharidoses type III).
- Sanfilippo syndrome is a progressive, life-limiting, inherited metabolic disease.
- Children with Sanfilippo syndrome also have medical issues, including hearing loss, poor visual acuity, and skeletal abnormalities.
- It is rare, affecting approximately 1 in 70,000 people.
- The most striking feature of Sanfilippo syndrome is developmental decline, leading to the behavioral problems including hyperactivity, impulsivity, aggression, and poor sleep.
- At this time, there are no approved treatments available in the US. There is no cure for Sanfilippo syndrome.

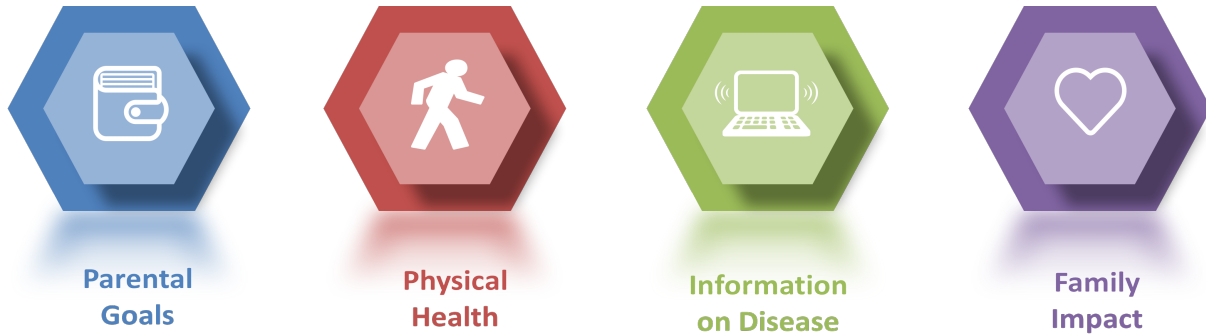
Sanfilippo syndrome is a staged disorder:



- It is vital to note that the progression of this disease causes regression. The natural course of the disease causes children to lose skills, so the most important thing we can do is help these children maintain the skills that they have.

- Educators should never give up on a child due to a diagnosis of Sanfilippo syndrome.

What should a child with Sanfilippo syndrome's school plan address?



Inclusion

- This is a choice that inevitably should be made by the parent. Keep in mind that as the disease progresses their feelings about the importance of inclusion may change.
- Some parents find solace in the fact that the children who attend school with their child remember them talking and walking independently, even though she has since lost those skills.
- Other families feel the need of inclusion can be found through other avenues (sibling interaction, social groups) and an educational program that pushes their child is more important. This may mean that the child is in a special type of school or classroom, because their needs cannot be met in an inclusion setting. These feelings can change over time.

Physical Ability

- The physical ability of a child with Sanfilippo is going to be very specific to where in the progression of the disease the child is. Some children may be walking, some may need assistance, some may be in a wheelchair for part of the day while still working on maintaining strength, and some may need more constant aid.
- Physical therapy is very important. A mother of a daughter with Sanfilippo syndrome said, *"...the physical therapy is- I feel like is the most important...She has completely lost the ability to walk independently... but the fact that she's still up everyday walking in the walker and in the stander, I think that has really been good for her physical health..."*
- Some parents feel that maintaining core strength helps to slow the progression of disease.

Communication

- Some children with Sanfilippo syndrome do not communicate verbally. Many of the children who are verbal can be difficult to understand.
- Parents typically use emotional cues, gestures, and facial expressions to understand how their child is feeling or what their needs are. This can be difficult for educators who are not as familiar with the child.
- Some children with Sanfilippo syndrome have done very well with assisted technology devices to communicate (such as an iPad).
- It is very important to note that any time a child cannot communicate as effectively as they would like to it is going to affect their behavior. One parent notes, *"if she needed to express something and that was the fastest way to do it [hitting someone]-... to get attention she would do that."*

Behavior Concerns

- As with other aspects of the disease, the behavior of a child with Sanfilippo syndrome can change dramatically over time. Erratic or difficult behavior can be present in the early stages, and then can progress to more docile behavior in the later stages.
- Behaviors are typically the most challenging during stage two of the disease.
- A mother of a son with Sanfilippo syndrome said, *"On a bad day-biting, hitting, pinching, temper tantrums. That ... you just have to let him get it out of his system..."*
- Each child will be completely unique.

Positive School Interventions

- Many schools have done an amazing job discovering alternative therapies that allow a child with Sanfilippo syndrome to thrive. These include:
 - Adaptive physical education
 - Music therapy
 - Swimming
 - Art
 - A sensory room
 - Alternative breaks such as a walk in the hall (as needed)
- Alternative therapies allow the child to work on maintaining their skills while also enjoying their time while they are doing it. These children have a shortened life span and educators need to balance the goal of learning with the child's happiness.

Individualized Education Plans (IEPs)

- IEPs are traditionally written for a child who will continually make progress. Educators set a goal, the child achieves it, and a new goal is set. This convention does not work with children with Sanfilippo syndrome in the later stages of the disease.
- A mother of a daughter with Sanfilippo syndrome, *"... when you formulate an IEP you are constantly wanting a child to learn new things ... it's difficult for most educators to shift gears and to understand Sanfilippo but also not be too scared by the diagnosis."*
- Formulate an IEP that is designed uniquely for a child with developmental regression. Keep in mind the current skills of the child, the parent's goals for their child, knowledge about the disease, and the use of alternative therapies.

- Setting appropriate IEPs that are realistic help the family and the education team have one central goal.

Support

- Children with Sanfilippo syndrome bring an amazing gift to those who know them, but it can be a very difficult diagnosis. Parents note that having community support help them cope.
- There are many great MPS III groups on social media that parents/family/friends can join for additional support.
- The National MPS Society can connect families and have an array of family support programs (<http://mpssociety.org/>).

Conclusions

- Children with Sanfilippo syndrome can thrive in a variety of school settings, and the ideal placement ultimately depends on the parent's personal goals for their child
- Difficulties with communication, mobility, and behavior affect how children with Sanfilippo syndrome experience education.
- IEPs should accommodate the skills of the child, knowledge of disease, and goals of the parents.
- Life-limiting conditions are chronically difficult to manage for not only the parents but also the entire family.

How can I learn more about this condition?

- Please ask the parents! This is an extremely rare condition and therefore most people have never heard of it before. Remember that parents live with this diagnosis and know their child best.
- Ask for documentation from their medical care team. These children have dedicated doctors whose notes may be helpful to you.
- The National MPS Society: <http://mpssociety.org/>

This handout is based on upon an IRB approved research protocol. (Education of children with Sanfilippo syndrome: Identification of needs, challenges, and services required for children with Sanfilippo syndrome by their parents by Lauren Gotsell, MS, CGC, Stephanie Harris, MS, CGC, MaryAnn Campion, EdD, MS, CGC, Laurel Calderwood, MS, CGC, at Boston University School of Medicine)