Cystinosis in the Classroom

A learning resource to best support children with Cystinosis to thrive in school

What is Cystinosis?

Cystinosis is a rare metabolic disease characterized by the accumulation of the amino acid cystine in organs and tissues, leading to severe organ dysfunction over time. The transporter for cystine is defective in children with cystinosis, which causes the cystine to crystallize within tissues. All organs and muscles are affected in cystinosis but the kidney is the first. Children with cystinosis will have some form of kidney damage depending on when they were diagnosed and how well they are able to take the treatment.

The current primary treatments for cystinosis are Cystagon™ and Procysbi™. These slow the progression of the disease by removing cystine from the cells. It is essential that this drug is taken every six hours to maintain drug efficacy as the cystine builds up very quickly and causes damage. Many children with cystinosis also develop crystals in their eyes. Cysteamine eye drops help to dissolve cystine crystals on the cornea as the oral medication doesn't reach the cornea. To be most effective, using these drops every hour while awake can remove the cystine crystals from the cornea and reduce pain and photophobia. A newer formulation of the eyedrops require only 4-times daily administration.

It is estimated that at least 2,000 individuals worldwide have cystinosis, though exact numbers are difficult to obtain because the disease is often undiagnosed and/or misdiagnosed.

How does Cystinosis affect school aged children?

Dr. Doris Trauner is a leading expert when it comes to cystinosis and the brain, especially with regard to children and youth. Below is a summary of some of her findings about some children with cystinosis:

Common neurological issues children with cystinosis may experience:
- Fine motor skills - including drawing, writing, etc. (messy handwriting)
- Visual/motor skills (hand-eye coordination)

Common cognitive functioning issues children with cystinosis may experience:
- Visual perception, Visual spatial function, and visual memory - being able to remember where things are located, determine direction, etc. can sometimes have an impact on maths processing.
  - Arithmetic and spelling
  - Spatial awareness

Some children with cystinosis experience bone issues. Many would have been diagnosed with rickets at a young age and may be at various stages of healing. For most children, they can participate in all school activities, including sport, but talk to the parents in this regard.

How is Cystinosis Treated?

Although there is no cure (yet) for cystinosis, a carefully timed medication routine is crucial for cystinosis patients manage this chronic illness. Many children are on a strict 6 hour medication schedule, meaning that at least one of their four daily doses will take place at school. Children may have multiple medication times throughout the school day depending on their individual treatment orders.

Medication:

Having a child with such unique medical needs in the classroom can be a bit overwhelming at first. For many teachers and SNAs, learning about the medication, what it does, administration, and proper storage are the greatest areas of unease. This simple breakdown will help to get a better grasp of how to manage medication in the school/classroom:

The Main Meds and What They Do:

Although every child with cystinosis is on a different regime of prescriptions, there are a few common medications that your student may be taking. (Parents will inform you of their own child's regime):

1. **Cystagon –** (Cysteamine Bitartrate) is the main medication for the management of cystinosis. Cystagon helps the body to deplete cystine from forming crystals, which damage soft tissue and organs in the body. This drug must be taken every 6 hours.
2. **Procysbi –** is a slow release form of Cysteamine Bitartrate.
   Note: Cysteamine Bitartrate *is a sulphur compound, and can give off a fishy or rotten-egg smell from someone with cystinosis. It is important that teachers and staff are aware of this and inform parents if it seems to become an issue at school or if other children comment.*
3. **Cystadrops** – Eye drops (gel formulation) to reduce crystals from forming in the eyes. Usually administered 4-times daily. If the child is using the non-gel formulation, hourly administration may be required. These drops require refrigeration.

Other common medications are potassium, sodium bicarbonate, calcium, one-alpha (vitamin D), carnitine.

Many children also require supplementary feeding with high-calorie feeds. Again, parents will talk to you about this if required during the day.

Administration:

Depending on the child, medication (and supplementary feed) will be administered two ways: orally, or via 'g-tube'. Using the g-tube is easily learned.

1. Orally – Medicine can come in both liquid and pill format and is usually taken with water to help flush medication
2. G-Tube – aka, a gastrostomy tube is a tube in place through previous surgery into the abdomen that delivers medication directly to the stomach. A hose is attached to the 'button' on the G-Tube, and medication is pushed into the stomach via non-needle syringe

Where Should Medication be Stored?

Some medications may need to be refrigerated and/or kept out of sunlight. Facilities may be needed to be set up in advance to ensure fridge space is available from the first day of school.

Time Management:

As mentioned above, 'med time' will happen at some point during the day. It is more than likely that the time medication needs to be taken will happen during class time. Depending on the age and independence of the child, it should take no longer than 10-15 minutes for all meds to be administered. In some instances, a family may coordinate their child's medication time to land during a break between classes. It ***incredibly important*** for medication to be taken at the time it is scheduled.

As a teacher, it may be frustrating to have a student leave in the middle of class to administer meds. Please remember that although a medication time may be inconvenient for school, it is timed in a way to suit the best interests and quality of life of the child and family. As some meds are administered every six hours, a parent or older child will be required to interrupt their sleep to keep up with doses. Living life in a 6 hour cycle can be incredibly hard for families and children with cystinosis. Parents/guardians will greatly appreciate your support in helping to maintain the health of their child, even if it comes at a slight inconvenience.

Recommendations to make things easier for both teachers and children with cystinosis:

1. **Work with the family to ensure their child keeps up with class work**
   - A survey 'Cystinosis in the Classroom' found that the number one growing concern of parents in respect to their child's education is the effects of missing class for specialist appointments. Communicating with the parents to understand when appointments take place, and what will be taught when the child is not in class is one way to reduce this strain. Sending home any homework in advance of appointments is a cystinosis-parent-approved way to make sure no one falls behind.

2. **Work with the family to understand med time**
   - The second greatest concern of parents is that they fear their child school does not understand the importance of the med schedule. Understanding that med time is the number one priority for keeping their child healthy is very important.
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3. **Understand the importance of hydration**
   - Children with cystinosis can become very easily dehydrated which can quickly affect their health. It is crucial that children have access to water whenever they may need it. Lots of water = lots of bathroom breaks. Having to wait to use the bathroom can negatively affect the kidneys, so children with cystinosis should be allowed to use the bathroom whenever they need. Asking to use the bathroom multiple times a day can also be embarrassing for children. Creating a system between the student and teacher that communicates they are going to the bathroom without drawing the attention of the class is an effective tool to implement.

4. **Offer flexible solutions to organization**

   Simple tasks like organizing a locker, desk, or paperwork is a task that many children with cystinosis find difficult to deal with. Being flexible in how a child is organized will help reduce classroom stress. For example, allowing a student to place all of their schoolwork into one binder as opposed to separate binders for each class.

5. **Adaptive learning**

   Depending on the severity of potential learning disabilities, it may be both in the school and child's best interest to request a formal learning assessment to create an effective IEP (Individual education plan). Assessments must be requested by the principal of the school to the NEPS. Getting an IEP in place at the beginning of the school year will help make the teachers workload lighter, as well as support the child with keeping up with their peers.

If an assessment is not something that can be completed, there are other ways to help children with cystinosis thrive in the classroom:

- Dr. Trauner's research showed that may children with cystinosis find it easier to grasp information when it is done verbally, as opposed to visually. Reading questions out loud to them can help relieve anxiety around learning
- Create homework reminders for your student. As mentioned above, children with cystinosis often struggle both with memory, and organization.

6. **Cystinosis is tough**
    - Living with cystinosis can be incredibly stressful for children at times. Disruption to sleep, feeling ill or lethargic, and having days where they are sad or frustrated are not uncommon for children with cystinosis. Practicing empathy goes a long way in helping children with cystinosis thrive. For example, if a child is tired and unwilling to cooperate in gym class, give them the option to sit out, read, or work on homework.
    However, we know that children don't want to stand out among their peers. Most children with cystinosis can participate in all aspects of school life and should be treated equally and expected to participate. Once the most important aspects are in in place, life should be as normal for all children.

    The most important things to remember are:
    1. **Correct timing for medications.**
    2. **Free access to drinking water. Bring water when playing sports.**
    3. **Free access to the toilet.**
    4. **Bring water on any away/school trips. Children get thirsty quickly.**

Resources:

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