Hurler Syndrome

Hurler Syndrome, formerly called mucopolysaccharidosis type I (MPS I), is a rare, inherited disease in which a person has a lysosomal enzyme deficiency that is needed to break down long chains of sugar molecules called glycosaminoglycans. An overabundance of these molecules within cells of various organs compromises their function. This disease is inherited in an autosomal recessive manner, which means the child must inherit one copy of the gene from each parent in order to develop the disease. There are no exact figures, but Hurler Syndrome has a prevalence rate of about one case per 100,000 births.

Hurler Syndrome is just one of the syndromes of a group of genetic disorders called mucopolysaccharidoses (MPS). Of the MPSs, Hurler Syndrome is the most common. Hurler Syndrome can be broke down into three separate syndromes: Hurler syndrome, Hurler-Scheie syndrome and Scheie syndrome. Hurler syndrome is the most severe of the three. People with Scheie syndrome, the mildest of the three disorders, tend to have the longest lifespan. Hurler-Scheie syndrome is a moderately severe disorder in which a person has one Hurler mutation and one Scheie mutation.
At birth, a baby may not show any symptoms of Hurler Syndrome, but symptoms will generally begin within a few months. Most symptoms appear between ages 3 and 8. Some symptoms include stunted growth, joint disease and stiffness, abnormal bones in the spine, claw hands, thick facial features with a low nasal bridge, and cloudy corneas. Children with Hurler syndrome may also experience deafness, heart valve problems, and intellectual disabilities that continually get worse. Generally, children with Hurler syndrome will have a shortened life span. Children with Scheie syndrome, however, may experience normal intelligence and possibly live to adulthood. Children with Hurler-Scheie syndrome may also have normal intelligence and experience mild to severe physical symptoms.
As of yet, there is no cure for Hurler Syndrome. There are, however, treatments to help relieve symptoms. Aldurazyme is a treatment that replaces whichever enzyme is deficient. This treatment is given by an infusion once a week for the duration of a person’s life. Another possible treatment for Hurler syndrome is bone marrow transplant. This treatment puts normal cells in the body that will produce the missing enzyme. However, bone marrow transplant is not possible for everyone with Hurler Syndrome because it requires chemotherapy. For those children with heart disease, chemotherapy is not an option. Genetic counselors are also able to talk to families about their specific risks of passing on the syndrome.

Teaching Strategies for Hurler Syndrome

- Direct, specific instruction.
- Depending upon the extent of impairments, the student may need auditory or visual accommodations or modifications.
- Instruction should focus on teaching the students functional skills that will assist them to live as independently as possible.
- Connect instruction to specific interests and abilities of the particular student.
- Instruction should occur in small groups or in a one-on-one format.
- Use physical and verbal prompting as well as verbal praise to reinforce
Definition of Hurler Syndrome. (2011, April 27). Retrieved from MedicineNet:

Genetics of Mucopolysaccharidosis Type I. (2012, January 26). Retrieved from Medscape Reference:
http://emedicine.medscape.com/article/1599374-overview#showall