

Insights on Hunter's Mucopolysaccharidosis Type II

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Introduction

Mucopolysaccharidosis, or Hunter's syndrome, is a rare sex-linked hereditary condition that can range greatly in severity but is typically marked by some degree of dwarfism, mental retardation, and deafness. Only males are afflicted by the illness, which first manifests itself in the first three years of life. The child's speech and mental development are delayed, and as the illness worsens, a classic constellation of physical symptoms—protruding abdomen, claw hands, excessive hair growth, coarsening of the face, and stunted growth—becomes apparent. Iduronate sulfatase insufficiency is the disorder's primary cause [1].

Description

The body accumulates mucopolysaccharides as a result of this deficiency's poor chemical breakdown of the carbohydrates necessary for the development of connective tissues, which in turn leads to the disease's recognisable mental and physical impairments. When a child receives a defective chromosome from his or her mother, Hunter syndrome develops.

An enzyme needed to disassemble complex sugar molecules is either absent or not functional as a result of that defective chromosome. In the absence of this enzyme, enormous numbers of these complex sugar molecules build up in the cells, blood, and connective tissues, resulting in progressive and irreversible harm. The family's past. A child must inherit a damaged chromosome, which is the disease's cause, in order to have Hunter syndrome. Hunter [2].

Men are virtually usually affected with Hunter syndrome. Girls are much less prone to get this condition since they inherit two X chromosomes. The healthy X chromosome can offer a functioning gene if one of the X chromosomes is damaged. There isn't another normal X chromosome to make up for a male's damaged X chromosome, though. Depending on the type and degree of the condition, Hunter syndrome can result in a number of consequences. In addition to the brain and neural system, complications can affect the lungs, heart, joints, connective tissue, and joints [3,4].

An swollen tongue, thickened gums, and thickness of the nasal passages and windpipe make breathing difficult (trachea). Children frequently develop chronic respiratory infections, pneumonia, and ear and sinus infections. Airway constriction is a common cause of sleep apnea, a disorder in which breathing is intermittently interrupted while sleeping. The thickening of heart tissue may cause heart valves to not close properly. The heart and other organs consequently do not receive blood as effectively. As the illness worsens, these symptoms frequently develop, finally resulting in heart failure. Aortic and other

blood artery narrowing can also be brought on by tissue thickening. This can then result in excessive blood pressure and constriction of the pulmonary arteries [5].

Conclusion

Bone, joint, and ligament problems are brought on by the buildup of complex sugar molecules that have not been metabolised in connective tissues. This prevents your child from growing normally, resulting in discomfort, physical deformities, and difficulty moving. Joint stiffness is brought on by swollen connective tissues in the joints as well as abnormalities in the cartilage and bones. Your child will probably move less while he or she is in discomfort, which might result in more stiffness and pain. A collection of anomalies known as dysostosis multiplex are frequently found in the bones of persons with Hunter syndrome. These anomalies can result in children having spines, ribs, limbs, fingers, legs, and pelvises with aberrant shapes.

Acknowledgement

None.

Conflict of Interest

None.

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