Hunter Syndrome

Hunter’s disease is a mucopolysaccharidosis lysosomal storage disease which results in the abnormal accumulation of glycosaminoglycans due to a defect in the enzyme iduronate sulfatase. Without the enzyme, cells are less effective at breaking down carbohydrates within the lysosome resulting in the accumulation of heparan sulfate and dermatan sulfate. Hunter’s disease is inherited in an x-linked recessive pattern which results in a predominantly male disease. It often presents as a milder form of Hurlers disease without corneal clouding but with the addition of aggressive behavior.



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Pathophysiology

X-linked Recessive
X-suit with Recessive-chocolate
This disease is inherited in an x-linked recessive manner which results in a predominantly male disease.

Mucopolysaccharidoses
Mucous-poly-sack
Hunter syndrome is a mucopolysaccharidosis, meaning it is a disorder caused by the absence or malfunctioning of lysosomal enzymes, leading to an inability to break down glycosaminoglycans. Hunter syndrome is also referred to as Mucopolysaccharidosis II (MPS II).

Iduronate Sulfatase Deficiency
Iditarod of Sulfur-matches Broken
Iduronate sulfatase is the enzyme deficient in Hunter's disease, which results in abnormal carbohydrate breakdown in the lysosome.

Increased Dermatan Sulfate
Up-arrow Deer-man-tan with Sulfur-match
This is a glycosaminoglycan which accumulates in Hunter's disease due to the defect in iduronate sulfatase.

Increased Heparan Sulfate
Up-arrow Hippie-run with Sulfur-match
This is another glycosaminoglycan which accumulates in Hunter's disease due to the defect in iduronate sulfatase.

Signs and Symptoms

No Corneal Clouding
Anti-sign on Corn Cloud
Unlike hurler's disease, hunter's disease does not have accumulation of glycosaminoglycans in the cornea and does not have corneal clouding.

Aggressive Behavior
Aggressively Shaking Fist
Those with Hunter's syndrome are characterized by having aggressive behavior, hyperactivity, restlessness and other behavioral issues.

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