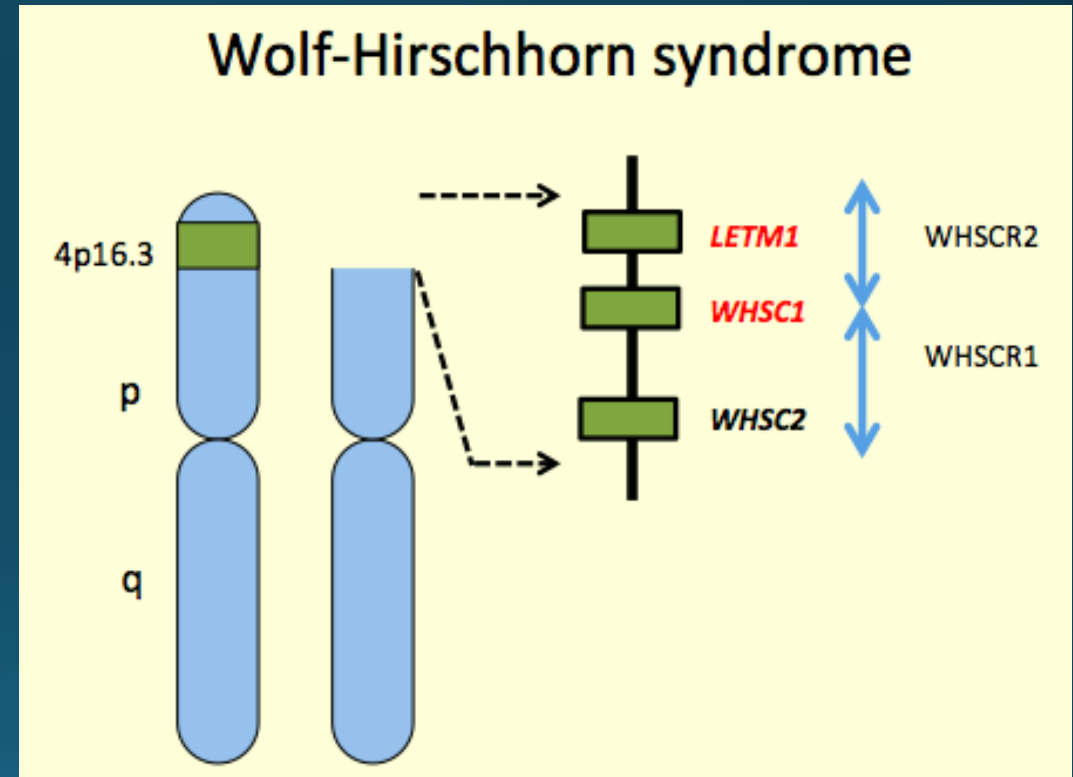


By: Thea Erickson

Wolf-Hirschhorn Syndrome

Etiology

- WHS is a genetic disorder that is caused by deletions in the short arm of chromosome 4
- The degree of the deletion in the chromosome and the other genetic rearrangements create a large range of clinical manifestations



Genetics



- Wolf-Hirschhorn syndrome is not an inherited disorder
- It is a mutation in a person's DNA that happens spontaneously

Symptoms

- WHS is described by pre and postnatal delay in growth, microcephaly (head is smaller than normal), seizures, hypotonia (low muscle tone and strength), developmental delay, congenital abnormalities and a recognizable facial appearance.
- The facial appearances can include: hypertelorism (increased distance between two body parts, commonly the eyes), protruding eyes, epicanthus (small fold of skin on the upper eye), arched eyebrows, prominent nasal bridge, downturned corners of the mouth, micrognathia (jaw is smaller than normal), and a short philtrum (space between bottom of nose and top of mouth)
- As the person gets older their nose will be more prominent
- About half of patients have dental abnormalities such as retained primary teeth, and peg-shaped teeth



Prognosis



- The prognosis of a person with Wolf-Hirschhorn syndrome depends on which of the specific features that person has and how bad it is
- The average life expectancy of WHS is unknown
- The muscle weakness may increase the risk of having chest infections and could possibly lower the life expectancy for this condition

Diagnosis

- The diagnosis of WHS is to notice the facial abnormalities, and there can also be genetic testing to see if that person has the deletion in that part of the chromosome



Treatment

- Some treatments to help improve WHS include: surgery to help repair defects, occupational or physical therapy, genetic counseling, support from social services, control of seizures, special education, and drug therapy



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