

Complete Information on Aase Smith syndrome with Treatment and Prevention

Aase syndrome or Aase-Smith syndrome is a rare inherited disorder. A familial deformity syndrome of variable expressivity, characterised by congenital hypoplastic anaemia and congenital triphalangy of the thumbs. Other features may be hydrocephalus with Dandy-Walker anomaly, cleft palate, and multiple contractures of the joints, narrow shoulders. The genetic basis of the disease is not known. The anemia is caused by underdevelopment of the bone marrow, which is where blood cells are formed. The disorder is primarily characterized by the presence of three bones within the thumbs rather than the normal two and abnormally reduced production of red blood cells. In some instances, additional abnormalities may be present.

Heart problems can lead to a kind of complications, which stem from the particular flaw. Most cases of Aase syndrome happen without a known cause and are not passed down through families. However, some cases have been shown to be inherited as an autosomal dominant and autosomal recessive trait. The two symptoms that must be existing to regard the diagnosis of Aase syndrome are CHA and TPT. CHA is a substantial decrease from birth in the amount of red blood cells in the blood. TPT means that one or both thumbs have three bones. As with most hereditary diseases there is no way to prevent the whole disease. With prompt acknowledgement and handling of infections in childhood, the complications of reduced light-colored blood cell counts may be limited. Genetic guidance is recommended if there is a family history of Aase syndrome.

Aase syndrome is sometimes also called Aase-Smith syndrome, or Congenital Anemia-Triphalangeal Thumb syndrome. It is a really uncommon genetic syndrome involving dual birth defects. Complications related to anemia include fatigue, tiredness, and decreased oxygenation of the blood. Heart problems can lead to a kind of complications, which stem from the particular flaw. Severe cases of Aase syndrome have been associated with early birth or earlier death. Frequent blood transfusions are given in the best years of living to handle anemia. Prednisone may be given, although this should be avoided in infancy because of its effects on bone and brain growth. An bone marrow transplantation may be needed if new treatment fails.

About the Author

Juliet Cohen writes articles for [health problems](#). She also writes articles for [updo hairstyles](#).

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