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Ataxia–telangiectasia

What is ataxia–telangiectasia?

Ataxia–telangiectasia (A–T) is a rare inherited disorder that is characterised by ataxia (lack of co–ordination) and telangiectases (tiny red spider–like veins) on the skin and eyes. Recurrent respiratory infections are another common feature of the disease.

What are the signs and symptoms?

Children with A–T appear normal at birth and it is not until they learn to walk that it becomes apparent there is a problem. Ataxia results from the progressive degeneration of a part of the brain called the cerebellum. Signs and symptoms of progressive neurological impairment include:

- An unsteady walk
- Abnormal, jerking movements
- Limb co–ordination becomes difficult
- Speech becomes slow and slurred
- Dull, sad, inattentive facial features
- By age 10 or 11 years most patients will need a wheelchair
- Mental deterioration resulting in slowing down of thinking speed

The second major clinical manifestation of A–T is the development of telangiectases. These usually do not occur until the child is between 3 and 5 years and may not even be apparent until adolescence. Tiny red spider–like veins first appear around the corners of eyes and then spread to the ears and cheeks.

Patients with A–T also have a weakened immune system, which makes them vulnerable to recurrent respiratory infections. They are also at greater risk of getting cancers.

How do you get A–T?

A–T is an autosomal recessive inherited disease. This means that you have inherited two A–T genes (one from each parent). If your parents are only carriers of A–T (each have one A–T gene and one normal gene), they will not show any signs of the disease themselves. The responsible gene has been mapped to chromosome band 11q22–23.

A–T may occur in any race or sex. Reported rates of incidence range from 1 in 40,000 to 1 in 100,000 births.

Is there a cure and what is the long–term outcome?

There is no cure for A–T. Currently no treatment exists to slow down the progression of the disease. The management of A–T is aimed at treating symptoms as they arise, preventing complications and most importantly providing support and education to both patients and their families. Patients with A–T usually die in their teenage years or early adulthood as a result of respiratory complications, cancer, or both.

Related information

References:

Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Vascular skin problems](#)

Other websites:

- [Ataxia–Telangiectasia Society](#)
- [Ataxia–Telangiectasia Children's Project](#)
- [Ataxia–telangiectasia](#) – emedicine dermatology, the online textbook

Books:

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If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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