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Editorial

Editorial Note on Progeria

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DESCRIPTION

Progeria is really rare, fatal genetic condition that affects children and gives them associate look of accelerated aging. The word Progeria comes from the Greek rogeros which means 'prematurely old'. It was 1st represented in an educational journal by Dr. Jonathan Hutchinson. The condition was later named Hutchinson-Gilford Progeria Syndrome (HGPS). Children with Progeria generally seem normal at birth. During the first year, signs and symptoms, like slow growth and hair loss, begin to seem. Single mistake in certain gene sequence causes it to form an abnormal protein. Once cells use this protein, referred to as progerin, they break down very simply. This leads kids with Progeria to age rapidly.

SYMPTOMS

Although they're born looking healthy, children with Progeria begin to show several characteristics of accelerated aging at around 18-24 months of age. The kids have a remarkably similar look, despite differing ethnic backgrounds. Most of the followiong symptoms are manifested after the age of 3 years in children with Hutchinson-Gilford Progeria Syndrome:

- Baldness
- Small, wrinkled face
- Head is large according to the size of the face
- Loss of eyebrows and eyelashes
- Prominent scalp veins
- Delay in tooth formation
- Loss of muscles and body fat
- Bulging eyes
- Wrinkled, scaly, dry skin
- High pitched voice
- Short stature
- Stiffness in joints
- Progressive cardiovascular diseases
- Progressive atherosclerosis

TYPES OF PROGERIA

- Classical HGPS: Classically affected patients powerfully tally each other
- Non classical (Atypcal) HGPS: A bunch of patients with Progeria that show a precise overlap with patients with other syndromes “e.g. Mandibulo-Acral Dysostosis (MAD)”.

CAUSES

Progeria is caused by a mutation (change) within the lamin A (LMNA) gene. This gene makes a protein that holds the nucleus of a cell together. Due to the modification within the gene, the protein becomes defective. This makes the nucleus unstable and this is believed to cause the premature aging method. The mutation of the LMNA gene doesn't run in families. In fact, parents and siblings of children with Progeria are seldom affected.

DIAGNOSIS

The symptoms are clear and visible. If changes are noticed in kids that appear like symptoms of Progeria, then appointment should to be taken with pediatrist or family doctor. Doctor will do a physical examination, check hearing and vision, measure pulse and blood pressure, and compare child's height and weight to other children of same age. If pediatrician is concerned, it should get to see a specialist in medical genetic science that will ensure the identification with a blood test. Before the genetic blood tests were availables, doctors could only diagnose Progeria with X-rays and observation.

TREATMENT

There's no cure for Progeria. Consistent monitoring for cardiovascular disease might benefit with treatment of the child's condition. Some children undergo coronary artery bypass surgery or dilation of cardiac arteries (angioplasty) to slow the progression of cardiovascular disease. Certain therapies might ease or delay some signs and symptoms. They include:

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- Low-dose aspirin- A daily dose might prevent heart attacks and stroke.
- Other medications- According to the child's condition, doctor might prescribe other medications, anticoagulants to prevent blood clots. The usage of growth hormone might aid increase height and weight.
- Physical and occupational therapy- These might help with joint stiffness and hip issues and may allows the children to stay active.
- Extraction of primary teeth-Extraction might prevent issues related to the delayed loss of baby teeth.