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An Editorial on Babinski-Froelich Syndrome

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Editorial

Froehlich syndrome is characterized by increased or excessive eating that leads to obesity, small testes, and a delay in the onset of puberty. It is also common for children with Froehlich syndrome to experience the delay in physical growth and the development of secondary sexual characteristics. Froehlich syndrome, also known as adiposogenital dystrophy, is a constellation of endocrine abnormalities believed to result from damage to the hypothalamus, a part of the brain that links the nervous system to the endocrine system via the pituitary gland. The hypothalamus regulates sleep cycles and body temperature and composition while stimulating the pituitary gland to release a variety of hormones that control growth, metabolism, and body development. Thus, numerous pituitary gland hormones could be indirectly disrupted by damage to the hypothalamus. Unlike similar diseases such as Prader-Willi syndrome, Froehlich syndrome is acquired, not inherited, and is associated with tumors of the hypothalamus area or their surgical treatment, causing increased appetite and depressed secretion of gonadotropin. This syndrome affects males more often than females.

The more obvious and frequently encountered characteristics include delayed onset of puberty, short stature, small testes, and obesity. Teenage boys with this disorder must be distinguished from those who have inherited growth delay disorders or Prader-Willi syndrome. Froehlich syndrome results from an injury of a part of the hypothalamus. The hypothalamic gland is the endocrine gland that produces substances that stimulate the pituitary and regulate appetite. In Froehlich syndrome, the front portion of the pituitary gland fails to secrete the hormones that are necessary for the onset of normal puberty. The most frequent cause of Froehlich syndrome is a specific tumor of the pituitary-hypothalamus area, an expanding hollow lesion or its surgical treatment.

Prader Willi syndrome is a complex disorder affecting many systems in the body. It is diagnosed more often in males born after a prolonged, delayed birth often in the breech position and is characterized by muscular weakness and failure to thrive during infancy. As the child grows there is a decrease in the function of the testes or ovaries, short stature, and impaired intellectual capabilities. The need to eat an extraordinary amount of food usually develops between 1 and 3 years of age. If left uncontrolled, the obesity of Prader Willi syndrome can lead to life- threatening heart and lung complications. Bardet-Biedl syndrome is a rare disorder affecting many systems in the body. It is inherited as an autosomal recessive genetic trait. The major symptoms of this disorder may include intellectual disability, obesity, delayed sexual development, underdeveloped reproductive organs, and degeneration of the retina of the eyes, kidney abnormalities, and/or abnormal fingers or toes. This disorder affects males and females in equal numbers.

If infection is suspected, blood cultures may be taken. In the event of a suspected legion or abnormal growth, imaging studies of the brain are conducted. These may include x-ray of the skull, CT scan of head and neck region, or MRI scan of the brain. Additional tests such as methylation analysis are needed to rule out Prader-Willi syndrome before a definite diagnosis of Froehlich syndrome can be made.

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