

a guide to understanding
**frontonasal
dysplasia**

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a guide to understanding frontonasal dysplasia

this parent's guide to frontonasal dysplasia is designed to answer questions that are frequently asked by parents of a child with a cleft lip and palate. It is intended to provide a clearer understanding of the condition for patients, parents and others.

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This booklet is intended for information purposes only. It is not a recommendation for treatment. Decisions for treatment should be based on mutual agreement with the craniofacial team. Possible complications should be discussed with the physician prior to and throughout treatment.

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frontonasal dysplasia (median cleft syndrome)

frontonasal dysplasia (FND) is a rare disorder characterized by mild or severe abnormalities of the head and face, especially the forehead, nose and central portion of the upper lip. Occasionally, there are also abnormalities found in the brain, heart and some of the bones. In severe cases, mild to moderate retardation may be present.

signs and symptoms

Head and Face

Children with frontonasal dysplasia typically have a cleft (groove), which can extend from the middle of the upper lip to the forehead. Affected individuals may also have widely spaced eyes (hypertelorism), hearing loss, a hair line which comes to a peak in the central forehead and a broad, flat nose which may be divided in half by the cleft. In some cases, the nasal tip may be missing. A cleft of the palate may also be present.

Central Nervous System

Brain abnormalities can include an absence of the connection (corpus callosum) between the two halves of the brain and an encephalocele, which is protrusion of brain tissue through a

opening in the base (floor) of the skull. Varying degrees of mental retardation or developmental delay may be present when severe brain abnormalities occur, although most people with FND are of normal intelligence.

Heart

Rare cases of frontonasal dysplasia may be associated with congenital heart abnormalities, particularly, Tetralogy of Fallot. Tetralogy of Fallot is a combination of four heart defects which are present at birth and usually require surgical correction before the age of two years.

Skeleton

Reported skeletal abnormalities include the presence of an extra great toe (polydactyly) and an underdeveloped shin (tibia) bone. The tibia bears much of our weight when we stand. Orthopedic surgery to remove the extra toe and correct the underdeveloped shin bone may also be required to aid with walking.

etiology

most cases of frontonasal dysplasia occur sporadically meaning they show no genetic linkage. The condition is rare enough that the exact incidence is unknown.

diagnosis

the diagnosis of frontonasal dysplasia is made at birth or by prenatal ultrasound and is based on the observed facial abnormalities. As there is usually no known genetic cause of FND, there is no blood test to aid in the diagnosis. A head and face CT scan and an MRI of the brain will likely be performed early on for further evaluation and eventual surgical planning.

treatment

Complex surgery to correct the position of the eye sockets (orbits), reconstruct the nose and repair the cleft are usually recommended during childhood. Many people will require secondary surgery later in life to further correct the position of the eyelids and appearance of the nose. Evaluation and treatment by an experienced craniofacial team is highly recommended.

prognosis

many people with FND have normal intelligence and can expect a normal lifespan.

how can children's craniofacial association (cca) benefit my family?

CCA understands that when one family member has a craniofacial condition, each person in the family is affected. We provide programs and services designed to address these needs. A detailed list of CCA's programs and services may be found on our website at www.ccakids.org or call us at 800.535.3643.



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