

a guide to understanding
**fibrous
dysplasia**

a publication of children's craniofacial association

a guide to understanding fibrous dysplasia

this parent's guide to fibrous dysplasia is designed to answer questions that are frequently asked by parents of a child with fibrous dysplasia. 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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what Is fibrous dysplasia?

fibrous dysplasia is a condition of the skeleton (bones). It is a birth defect that is a non-cancerous disease. It is not hereditary so your child did not get it from you nor will he/she pass it along to his/her children.

how do I recognize this condition in my child?

fibrous dysplasia is usually detected in early childhood as a result of swelling of the jaw. Also, in some cases it may cause the teeth to separate.

how does the disease progress?

fibrous dysplasia gets progressively worse from birth until the bones finish growing. As it progresses, normal bone is replaced by various amounts of structurally weak fibrous and osseous (bone-like) tissue. In normal bone formation, woven bone appears first and later matures into lamellar bone. In fibrous dysplasia, bone does not mature and development stops in the woven bone stage.

Fibrous dysplasia causes misshapen bones. It can occur in the bones in the front of the head and/or sphenoid bones that are situated at the base of the skull. If this happens, it can eventually lead to deformation of facial features and affect the shape of the skull.

how many types of fibrous dysplasia are there?

There are three types of fibrous dysplasia:

Monostotic disease is the most common type of fibrous dysplasia, occurring in 70% of cases. Monostotic simply means involving one bone. It most often occurs on the long bones such as the femur (thigh bone), ribs and skull.

Polyostotic disease, affects 30% of patients. Polyostotic means occurring in more than one bone. The head and neck are involved in half of these patients.

The third type is **McCune-Albright syndrome**. It only occurs in 3% of cases. It is characterized by polyostotic fibrous dysplasia (fibrous dysplasia occurring in more than one bone); skin pigmentation; and, in females, early puberty.

how often does fibrous dysplasia affect the face and head?

Skull involvement occurs in 27% of monostotic and up to 50% of polyostotic patients. Fibrous dysplasia involving the face and skull is called "Leontiasis ossea." Without treatment, one or more bones progressively increase in size, and move into the cavities of the eye, mouth, and/or the nose and its sinuses. Also, abnormal protrusion of the eyeball (exophthalmos) may develop and eventually cause complete loss of sight because it presses on the optic nerve. In addition, there may be interference of the nasal passage and with eating.

what are the effects of fibrous dysplasia of the skull base?

When fibrous dysplasia of the frontal (forehead bone) and/or sphenoid (bone at the base of the skull) bones progresses, these bones become thick and dense. This increase in size eventually causes the facial features and skull to become misshapen. In these cases more than one bone is usually involved. It can also result in cranial nerve problems. If the temporal bone is affected, the patient may suffer as much as 80% hearing loss when the inner ear canal narrows. It may also cause facial nerve paralysis or dizziness. However, any of our 12 cranial nerves can be involved with fibrous dysplasia. The more common results could include cranial nerve problems, and sight and hearing loss.

are there any other effects of fibrous dysplasia?

It is estimated that patients with fibrous dysplasia are 400 times more likely than the general population to develop a malignant bone tumor.

what is the treatment for fibrous dysplasia?

Physicians decide on treatment options after assessing a patient's symptoms. First the doctor observes the patient. Then he will consider conservative treatment such as surgically shaving or removing the fibrous tissue. In more severe cases the doctor may recommend complete removal of the bone.

Surgery is used to return the face to its normal structure and/or to relieve effects when a cranial nerve is being pinched. In these cases the abnormal bone must be completely removed. It is best to wait until adolescence for surgery. However, if the progression of the disease affects nerve function, a decompressive procedure should be considered early in childhood to keep normal function.

if surgery is recommended, how many will be necessary?

Sometimes the fibrous tissue can be completely removed successfully by a single procedure. However, most fibrous tissue can be managed through staged procedures with overall very favorable results and good long term prognosis.

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 Web site at www.ccakids.com or call us at 800.535.3643.



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