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Cleidocranial dysostosis

Cleidocranial dysostosis is a disorder involving the abnormal development of bones in the skull and collar (clavicle) area.

Causes

Cleidocranial dysostosis is caused by an abnormal gene. It is passed down through families as an autosomal dominant trait. That means you only need to get the abnormal gene from one parent in order for you to inherit the disease.

Cleidocranial dysostosis is a congenital condition, which means it is present from before birth. The condition affects girls and boys equally.

Symptoms

People with cleidocranial dysostosis have a jaw and brow area that sticks out. The middle of their nose (nasal bridge) is wide.

The collar bones may be missing or abnormally developed. This pushes the shoulders together in front of the body.

Primary teeth do not fall out at the expected time. Adult teeth may develop later than normal and an extra set of adult teeth grow in. This causes the teeth to become crooked.

Intelligence level is most often normal.

Other symptoms include:

- Ability to touch shoulders together in front of body
- Delayed closure of fontanelles ("soft spots")
- Loose joints
- Prominent forehead (frontal bossing)
- Short forearms
- Short fingers
- Short stature
- Increased risk of getting flat foot, abnormal curvature of spine (scoliosis) and knee deformities

- High risk of hearing loss due to infections
- Increased risk of fracture due to decreased bone density

Exams and Tests

The health care provider will take your family history. The provider will do a physical examination and may do series of x-rays to check for:

- Undergrowth of the collarbone
- Undergrowth of the shoulder blade
- Failure of the area in the front of the pelvis bone to close

Treatment

There is no specific treatment for it and management depends on each person's symptoms. Most people with the disease need:

- Regular dental care
- Head gear to protect skull bones until they close
- Ear tubes for frequent ear infections
- Surgery to correct any bone abnormalities

Support Groups

More information and support for people with cleidocranial dysostosis and their families can be found at:

- Little People of America -- www.lpaonline.org/about-lpa [https://www.lpaonline.org/about-lpa]
- FACES: The National Craniofacial Association -- www.faces-cranio.org/ [https://www.faces-cranio.org/]
- Children's Craniofacial Association -- ccakids.org/ [https://ccakids.org/]

Outlook (Prognosis)

In most cases, the bone symptoms cause few problems. Appropriate dental care is important.

Possible Complications

Complications include dental problems and shoulder dislocations.

When to Contact a Medical Professional

Contact your provider if you have a:

- Family history of cleidocranial dysostosis and are planning to have a child.
- Child with similar symptoms.

Prevention

Genetic counseling is appropriate if a person with a family or personal history of cleidocranial dysostosis is planning to have children. The disease may be diagnosed during pregnancy.

Alternative Names

Cleidocranial dysplasia; Dento-osseous dysplasia; Marie-Sainton syndrome; CLCD; Dysplasia cleidocranial; Osteodental dysplasia

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