

Ear Defects

BIRTH DEFECT RESEARCH FOR CHILDREN



What is Microtia-Atresia?

Microtia means that the external part of the ear is small, absent (anotia) or abnormally shaped. 90% of children with Microtia have only one ear (unilateral) affected and 10% have Microtia of both ears (bilateral). More children have Microtia of the right ear than the left.

Atresia means that the ear canal is absent or severely narrowed. Atresia usually accompanies Microtia, but not in all cases. Atresia impairs the hearing of the child only in the affected side.



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How often do these disorders occur?

Approximately one baby in every 7,000 to 10,000 is born with an ear deformity. Some Native Americans populations have a greater incidence (1 in 1,200 births). Males have a slightly greater incidence than females.

When are these conditions detected?

Many ear malformations are usually not detected until birth, but a prenatal ultrasound might identify total absence of the ear (anotia). Some doctors suggest a high-resolution 3-D CT scan of the temporal bones to aid in diagnosis and treatment.

What causes Microtia-Atresia?

Drugs such as thalidomide and Accutane (a vitamin A derivative), the rubella virus and other intrauterine infections have been implicated as factors in some cases of Microtia-Atresia. One study also indicated that mothers with maternal insulin dependent diabetes were at significantly higher risk for having a child with an ear defect.

In a minority of cases, ear malformations are autosomal dominant traits so genetic counseling is suggested.

What treatment exists for ear malformations?

There are surgical procedures that can reconstruct the external ear, ear canal and middle ear. The amount of cosmetic and/or functional improvement depends on the severity of the ear defect(s). Improved hearing may result from these reconstructive procedures. In bilateral cases, hearing aids should be fitted until the hearing mechanism can be reconstructed. Plastic surgeons usually work closely with an otologist to restore not only the auditory function but also the form of the ear.

Until recently, surgical reconstruction was suggested beginning at the age of six years. With improved reconstructive procedures, however, many surgeons

recommend waiting until the child has reached the age of ten years when more detailed, better quality reconstruction is possible.

Three-dimensional computed Tomography (3DCT) imaging methods also offer new diagnostic possibilities for the reconstruction of the external ear.

What other conditions may accompany ear malformations?

In most cases, these conditions occur as isolated incidents. In some patients, however, ear malformations may be part of a syndrome that involves other craniofacial, skeletal, spinal, and/or internal abnormalities.

What is the prognosis for affected children?

Restoration of an acceptable level of hearing occurs in 50-67% of children who have had surgical reconstruction. Surgery does, however, carry some risks of complications such as permanent hearing loss or damage to facial nerves.

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