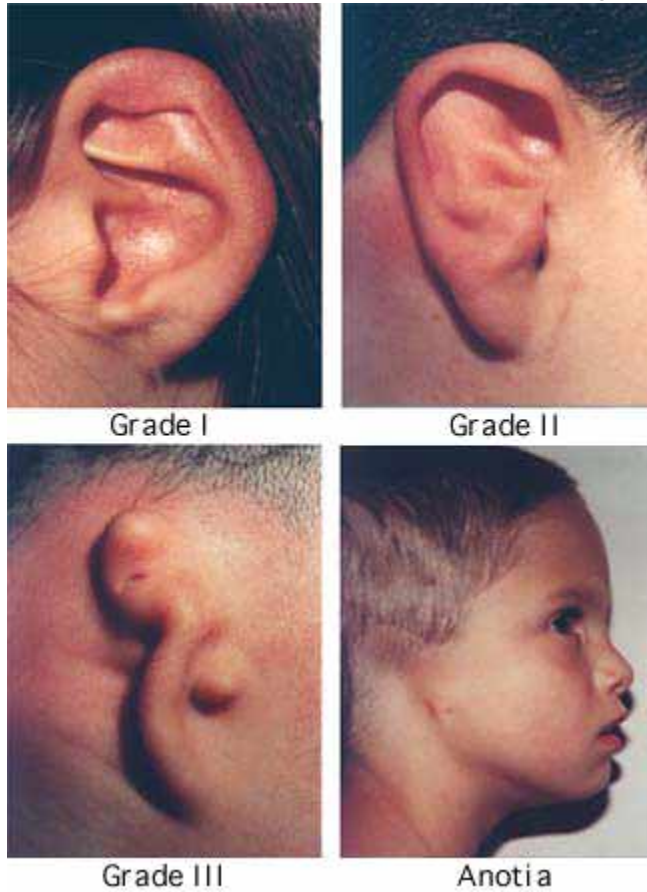


**Head: Microtia**  
**Sub: by Burt Brent, M.D.**

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Microtia varies from the complete absence of auricular (outer ear) tissues (anotia) to a somewhat normal but small ear with a narrowed canal. Between these extremes, one finds an endless variety of vestiges, the most common being a vertically-oriented sausage-shaped nubbin (figure 5). Microtia is nearly twice as frequent in males as in females, and the right-left-bilateral ratio is roughly 6:3:1.

**Figure 5 - Variance of the microtia deformity.**



*Grade I, II, III microtia and anotia (complete absence of any ear remnant).*

In my experience, children will discover that they are different at around age three to three and a half. Classically, the parents find their child comparing sides in front of the mirror. They begin to refer to the microtic ear as their "little ear" or "closed ear." It is best to agree with the child that they were born with one big ear and one little one and, when they are older, the little one can be made larger to match the other. Then they should be treated absolutely normally, without making a fuss about the deformity.

The first big psychological trial manifests in about the first grade in school, when children are continually exposed to a large group of their peers for the first time. Self-awareness is heightened as they begin comparing each other and forming a real

concept of body image. This is when name-calling and teasing begins, and the microtic patient learns what it means to be "different."

The "second round" comes with adolescence, when everyone is conforming to peer pressure to be accepted. This is the era of our lives when looks become very important and everyone wants to "fit in." Teenagers with microtia are very self-conscious about being different and are particularly motivated to having the ear repaired. They are also very critical and may have unrealistic expectations of what can be surgically produced.

In addition to holding up our eyeglasses and funneling sounds towards our ear drums to improve hearing, ears make us look better and feel much better about ourselves as a *whole* person. This is the driving reason for surgical creation of the outer ear. It is a psycho-emotional aesthetic endeavor to restore self-esteem through restoring a symmetrical, normal self-image. Far from being cosmetic surgery, repairing a congenital deformity permits a person to have a normal self-image, a normal life and to be a normal, productive member of society.

The age at which an auricular construction should begin is governed by both psychological and physical considerations. Since the body image concept usually begins forming around the age of four or five years, it would be ideal to begin construction before children enter school and before they are psychologically traumatized by their peers' cruel ridicule. However, surgery should be postponed until rib growth provides substantial cartilage to permit a quality framework fabrication, which is rarely before the age of six.

In my experience with interviewing and evaluating more than 1,500 microtia patients from age one month to 62 years, the patients and/or their families consistently stated that their psychological disturbances rarely began before age seven, and usually became overt from ages seven to 10. Hence, in general, I prefer to delay the initial cartilage graft until the patient is six years old, when there is usually sufficient rib cartilage for the repair. If patients are small for their age and/or the opposite, normal ear is large, then I find it prudent to postpone the surgery for several years.

### **Psychological and emotional benefit**

The impact of microtic deformity on patient and family sequentially becomes greater as the child enters school, approaches adolescence and reaches adulthood.

#### **Emotional Impact of Unrepaired Microtia** Rated by Patient and Family. Author's Series of 1,000 Cases.

<b>Age (Years)</b>	<b>Severe</b>	<b>Moderate</b>	<b>Mild</b>
5-10	18.6 %	46.2 %	35.2 %
11-14	16.3 %	61.2 %	22.5 %
15-20	26.7 %	60.0 %	13.3 %
21-62	44.1 %	41.2 %	14.7 %

Before 10 years of age, 64.8 percent of families rated the deformity's impact as "moderate" to "severe," whereas this figure jumped to 77.6 percent in ages 10 to 14 and 86.7 percent in patients older than 15. In ages six to 10, where 35.2 percent of families rated the impact as being only "mild," these same families often stated that the full impact had not yet been realized and that they sought surgery "not to solve

a current problem but to prevent a future, anticipated crisis" as their child grew older.

### Facial Deficiencies

Because the auricle develops from tissues of the branchial arches, it is not surprising that a significant percentage of microtia patients exhibit deficient facial components which originate from these embryological building blocks. Appearing as a flattened side of the face, this condition is known as hemifacial microsomia, and is basically an underdevelopment of the bony jaws and overlying soft tissues. The most complete genetic expression of this condition includes defects of the external and middle ear; hypoplasia of the mandible, maxilla, zygomatic and temporal bones; macrostomia and lateral facial clefts; paresis of the facial nerve; and atrophy of facial muscles and parotid gland, where even the palatal muscles are weak on the involved side.

In analyzing my own series of 1,000 microtia patients, I found that associated branchial arch deformities were common: 36.5 percent of my patients had obvious bony and/or soft tissue deficiencies, making the side of the face appear flattened or distorted. Of these, the family perceived the facial deformity as "significant" in 49.4 percent of the cases. This obvious facial asymmetry is usually dealt with *after* the auricle is repaired.

From my years of long-term observation of many patients, this asymmetry doesn't seem to worsen as the child grows but instead grows proportionally in its relation with existing facial features. This has been borne out by a recent growth analysis investigation.

Overt facial nerve weakness was found in 15.2 percent of my 1,000 microtia patients, causing a weak smile or inability to close one eye completely. Of these, 42.6 percent had involvement of more than one nerve branch (Table 2).

**Table 2 - Associated Deformities**  
Author's Series of 1,000 Microtia Patients

#### Branchial Arch Deformities

A. Obvious Bony and Soft Tissue Deficit	36.5 %
Family perceives it as "significant"	49.4 %
B. Overt Facial Nerve Weakness	15.2 %
Of these, more than one branch involved	42.6 %

Macrostomia 2.5 %

Cleft Lip and/or Palate 4.3 %

Urogenital Defects 4.0 %

Cardiovascular Malformations 2.5 %

Misc. Deformities 1.7 %

### Kidneys and Urinary Tract

Urogenital tract abnormalities increase in the presence of auricular deformities [104], particularly when the patient is afflicted with other manifestations of facial underdevelopment [103]. Several of my patients have hypospadias or vaginal agenesis, and 4 percent have proven abnormalities of their collection systems, which

include horseshoe kidney, ureteral duplication, unilateral renal agenesis and pelvic kidney.

However, because the body has enough "reserve" to easily live with just one kidney, I have yet to see any microtia patient suffer any life-threatening renal consequences from these system abnormalities. A routine screening of the urine may detect silent hematuria or proteinuria, but as likely will show nothing. Recurring urinary tract infections in microtia patients prompt renal function studies, and one should start with a renal ultrasound before considering more invasive technique to detect these deformities. Because of the increased incidence of urogenital abnormalities in microtia patients, one might consider screening all of them with ultrasound.

### **Cervical (Neck) Spine**

Cervical spine anomalies are more common in microtic patients if other "midline defects" exist, such as cardiac or renal disorders or cleft lip and palate. Because associated neurologic symptoms are rare, the frequency of these vertebral abnormalities are probably underestimated in patients with microtia. Goldenhar Syndrome (oculoauriculovertebral dysplasia) is a condition in which the microtia patient has an ocular dermoid and usually has cervical vertebral abnormalities [30].

When one notes an ocular dermoid or detects limited range of neck motion in any microtia patient, one should investigate the renal function and evaluate the cervical spine as well. Regarding the latter, CAT scans will provide useful information for any physician caring for these patients. It is particularly wise to bring this condition to the anesthesiologist's attention during *any* kind of surgery, so that unusual neck manipulation is avoided during induction of anesthesia; the child's head should not be forcibly turned, but instead, the head, neck and body should be "log-rolled" as a unit for any turning maneuvers during surgery.

If there is any suspicion of neurological disturbances seen in these children, they should undergo CT scan, MRI and appropriate neurologic examination.

### **Other Associated Problems: Cleft Lip/Palate and Heart**

Cleft lip and/or palate is seen in 4.3 percent of my patients, and 2.5 percent have cardiovascular malformations. The latter have included atrial and ventricular septal heart defects, dextrocardia, transposition of the great vessels, three-chambered heart and patent ductus. If any signs or symptoms of these heart problems are noted, the pediatrician should consult with a cardiologist for appropriate steps in management.