External Auditory Atresia  
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History
2 month old with deformed pinnae and absent external canals.

Diagnosis
Bilateral External Auditory Atresia

Discussion
External auditory canal atresia is related to anomalous development of the first and second branchial arches (mesoderm) and the first pharyngeal pouch (endoderm). Lack of first branchial arch differentiation results in malformation of the incudomalleal joint, eustachian tube, mandible and tensor tympani muscle. Disturbance of the second branchial arch affects the facial nerve canal, stapedius muscle, lower ossicular chain and styloid process. The auricle is derived from the first and second arches. The severity of external ear anomalies predicts the severity of middle ear defects. Radiographic findings in external auditory atresia can be related to the embryologic development. The external auditory canal is often stenotic and vertically oriented. The atresia plate may be membranous or bony; the bony plate may be thick or thin. The middle ear cavity is often small and poorly pneumatized (related to eustachian tube dysfunction). Ossicular anomalies are common, including fusion of the incus and malleus, hypoplasia of the manubrium of the malleus, and fixation to the lateral wall or atresia plate. The facial nerve canal may be displaced. The oval and round windows may be absent. Occasionally, labyrinthian dysplasia is present. External auditory atresia is usually unilateral, right more than left, although can be bilateral. There is a high incidence of familial occurrence of external atresias. Goldenhaar (hemifacial microsomia), Pfeiffer, Treacher Collins, and Pierre Robin syndromes may be associated with external ear anomalies.

Findings
CT-Axial and coronal images show bilateral membranous atresia. Note the small middle ear cavities with lateral ossicular fusion.

Reference
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