Congenital malformations of the inner ear: a classification based on embryogenesis.

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Approximately 20% of patients with congenital sensorineural hearing loss have radiographic abnormalities of the inner ear. A broad spectrum of anomalous patterns have been described, most of which have been lumped together under the term "Mondini's dysplasia." We feel that this grouping of many dissimilar entities under a single umbrella term is unwarranted. Based on a review of 63 patients with 98 congenitally malformed ears, we have been able to recognize a number of distinct anatomic patterns from their radiographic appearance. A remarkable similarity between these morphologies and the appearance of the inner ear at various stages of embryogenesis was found. This led us to propose a classification system based upon the theory that these deformities result from an arrest of development during varying stages of inner ear organogenesis.