



TABLE 12.2 Disorders Associated With Hearing Loss in Children

Disorder	Description
Cytomegalovirus (CMV)	<ul style="list-style-type: none"><li>• Most common congenital viral infection causing hearing loss today, occurring in 1 in 1,000 live births</li><li>• Contracted in utero or postnatally from the mother</li><li>• Can result in sensorineural hearing loss as well as central nervous system, cardiac, optic, and growth abnormalities</li><li>• Symptoms may not be apparent at birth, with onset about 18 months of age</li><li>• Progresses rapidly during the first year</li></ul>
Down Syndrome	<ul style="list-style-type: none"><li>• Congenital chromosomal abnormality (trisomy 21)</li><li>• Individuals frequently have low-set small ears, external canal stenosis, middle ear deformities, and facial nerve abnormalities</li><li>• 30% of these children have sensorineural hearing loss</li><li>• Most have poor Eustachian tube function, resulting in chronic middle ear disease with associated conductive hearing loss</li></ul>
Meningitis	<ul style="list-style-type: none"><li>• Neonatal infection, can be viral or bacterial</li><li>• Most common cause of acquired sensorineural hearing loss</li><li>• Hearing loss can range from mild to profound, and may be progressive</li><li>• Symptoms may include headache, neck stiffness, photophobia, and suppurative otitis media</li></ul>
Ototoxicity	<ul style="list-style-type: none"><li>• Can be caused by a wide variety of strong antibiotics, chemotherapeutic agents, or loop diuretics</li><li>• Can also result from exposure to various chemical agents in the environment</li><li>• Characterized by a progressive high-frequency sensorineural hearing loss following such exposure</li></ul>
Usher Syndrome	<ul style="list-style-type: none"><li>• Autosomal recessive</li><li>• Occurs in 6%–12% of congenitally deaf children, and 3 in 100,000 of the general population</li><li>• Involves retinitis pigmentosa and progressive moderate to severe sensorineural hearing loss</li><li>• Can vary greatly in age of onset, severity, and progression</li></ul>
Waardenburg Syndrome	<ul style="list-style-type: none"><li>• Autosomal dominant</li><li>• 20% have white forelock, 99% have increased distance between the eyes, 45% have two differently colored eyes (typically one brown and one blue)</li><li>• Depigmentation of the skin and eyebrows that meet over the bridge of the nose</li><li>• 50% have mild to severe sensorineural hearing loss, which can be unilateral or bilateral and is progressive</li></ul>