Name	How is it inherited?	Type & severity of hearing loss	Other features of condition
Alport syndrome	X-linked (most common)	Sensorineural	**Renal (kidney) problems**
	Autosomal dominant	Usually progressive	**Ocular (eye) problems**
	Autosomal recessive	Mild – profound	
		Hearing loss usually not seen before 10 years of age	
Biotinidase deficiency(included in Indiana's newborn screen)	Autosomal recessive	Hearing loss occurs in 75% of children with untreated biotinidase deficiency	**Seizures**
		Sensorineural	**Hypotonia (low or "floppy" muscle tone)**
			Developmental delay
			Ataxia (clumsiness)
			Hair loss
			Eye problems
			Skin rashes
Branchio-oto-renal syndrome	Autosomal dominant	Conductive	**Abnormal formation of outer, inner, or middle ear**
		Sensorineural	**Renal (kidney) problems**
		Mixed	**Branchial cysts(growths) or tracts(openings) along neck muscles**
CHARGE syndrome	Autosomal dominant	Sensorineural	**Colobomas (keyhole openings in iris, or colored part of eye)**
	Most people with CHARGE syndrome do not have other family members with CHARGE syndrome	Mixed	**Heart problems**
		Growth and developmental delays	**Choanal atresia(narrowing of nasal passages)**
		Genital abnormalities	**Ear abnormalities**
		Eye abnormalities	

Jervell and Lange-Nielsen syndrome	Autosomal recessive	Sensorineural	**Cardiac arrhythmias(abnormalities of heartbeat)**
		Profound	**Syncope (fainting)**
		Usually bilateral	
		Usually present at birth	
Kearns-Sayre syndrome	Mitochondrial	Pigmentary retinopathy("salt and pepper" appearance to retina, or back of eye)	**Symptoms usually appear before the age of 20**
		Heart rhythm abnormalities	**Progressive external ophthalmoplegia (drooping of eyelids and paralysis of eye muscles)**
			Cerebellar ataxia(uncoordinated muscle movement or clumsiness)
		Sensorineural hearing loss is common in patients with MELAS-Suzanne	**Onset of symptoms usually in childhood (2 – 10 years)**
			Short stature
Mitochondrial Encephalopathy, Lactic Acidosis, and Stroke-like Episodes (MELAS)	Mitochondrial		**Seizures**
			Recurrent headaches
			Recurrent vomiting
			Limb (arm/leg)weakness
			Short stroke-like episodes of paralysis or blindness
		Sensorineural	**Goiter (growth on/near thyroid)**
		Severe to profound hearing loss	**Abnormalities of cochlea**
Pendred syndrome	Autosomal recessive	Usually bilateral	
		Usually prelingual	
		Nonprogressive	
		Conductive	**Myopia (vision problems)**
		Sensorineural	**Cataracts (cloudiness of eye)**

Stickler syndrome	Autosomal dominant		**Cleft palate (opening in roof of mouth)**
			Micrognathia (small chin)
			Bone abnormalities
			Features can vary between family members with Stickler syndrome
		Sensorineural	**Retinitis pigmentosa(progressive loss of function of the retina, or back of the eye)**
		Profound	**Vision problems**
Usher syndrome type I	Autosomal recessive	Bilateral	
		Usually present at birth	
		Sensorineural	**Retinitis pigmentosa(progressive loss of function of the retina, or back of the eye)**
		Mild to moderate (low frequencies), severe to profound (high frequencies)	
Usher syndrome type II	Autosomal recessive	Bilateral	
		Usually present at birth	
		Sensorineural	**Pigmentary changes of iris (colored part of eye),hair, skin**
		Usually profound	**Most patients with Waardenburg syndrome have white forelock (front part of hair) or have gray hair by the age of 30**
Waardenburg syndrome	Autosomal dominant (most common type of autosomal dominant hearing loss)	Unilateral or bilateral	
		Usually nonprogressive	
		Usually present at birth	