

Usher syndrome affects approximately three to ten in 100,000 people worldwide. Higher than average numbers of people with Usher syndrome have been found among Jewish people in Israel, Berlin, Germany; French Canadians of Louisiana; Argentineans of Spanish descent; and Nigerian Africans. USH3, the rarest form in most populations, comprises about 40% of Usher patients in Finland. Usher syndrome is the most common genetic disorder involving both hearing and vision abnormalities. Usher syndrome types 1 and 2 account for approximately 10 percent of all cases of moderate to profound deafness in children. Usher syndrome is diagnosed by hearing, balance and vision examinations. A hearing (audiologic) exam measures the frequency and loudness of sounds that a person can hear. An electroretinogram measures the electrical response to the light-sensitive cells in the retina of the eyes. A retinal exam is done to observe the retina and other structures in the back of the eye. Vestibular (balance) function can be assessed by a variety of tests that evaluate different parts of the balance system. Genetic testing is clinically available for most of the genes associated with Usher syndrome.