

Retinitis pigmentosa (RP) comprises a large group of inherited vision disorders that cause progressive degeneration of the retina, the light sensitive membrane that coats the inside of the eyes. Peripheral (or side) vision gradually decreases and eventually is lost in most cases. Central vision is usually preserved until late in these conditions. Some forms of RP can be associated with deafness, obesity, kidney disease, and various other general health problems, including central nervous system and metabolic disorders, and occasionally chromosomal abnormalities. RP as a group of vision disorders affects about 1 in 3,000 to 1 in 4,000 people in the world. This means that, with a population of about 324 million in the United States in mid-July 2017 (see < <http://www.census.gov/>> for continuous updates), about 81,000 to 108,000 people in the United States have RP or a related disorder. With a worldwide population presently estimated at over 7.05 billion, it can be estimated that approximately 1.77 to 2.35 million people around the world have one of these disorders. Excluding age-related macular degeneration and glaucoma, the genetic causes of which are complex and linked simultaneously to more than one gene (so called “polygenic” disorders), RP is the most common cause of inherited visual loss. RP is diagnosed by electroretinography (ERG) showing progressive loss in photoreceptor function, visual field testing, and retinal imaging [mainly by optical coherence tomography (OCT) and fundus auto-fluorescence (FAF) that show detailed microanatomical features that cannot be resolved by naked eye]. Molecular genetic testing for mutations in many of the genes associated with RP is available to confirm the diagnosis.