
The goal of this study was to establish how common certain otologic (relating to the ear or hearing) issues are in individuals with distal 18q-. We reported on 113 people with 18q-. As expected, a relatively high percentage had some degree of aural stenosis or atresia (narrow or closed ear canals). No one had any malformations of the outer ear. Importantly, this study was able to follow several patients over the course of several years to determine whether the diameter of the ear canal widened or narrowed with age. In 50% of the ears with narrow ear canals, the ear canal diameter widened with time. Hearing loss was identified in a number of individuals. About 50% of the hearing loss was conductive, while 28% was sensorineural (related to the function of the nerves involved with hearing). Additionally, eustachian tube dysfunction was quite common. This occurs when the ear drum is not able to vibrate appropriately, often due to a cold or fluid in the ears. Almost 80% of ears examined had some degree of Eustachian tube dysfunction. The paper ended with some recommendations regarding the management of ear/hearing issues in people with 18q-. In those with exceptionally narrow ear canals, bone conduction hearing aids are important. Those with larger ear canals may benefit more from conventional hearing aids. In addition, to ensure that there are no problems with hearing, frequent hearing tests are recommended, as well as routine monitoring of eustachian tube function.