Peters Anomaly

Peters anomaly is a rare form of anterior segment dysgenesis in which abnormal cleavage of the anterior chamber occurs or a congenital absence of two layers of the cornea. Involving the central or entire cornea, Peters anomaly is divided into 2 types depending on whether or not the lens is abnormal. Type 1 is unilateral, characterized by a central or paracentral corneal opacity with iris strands that arise from the iris collarette and attach to the cornea. Type 2 is bilateral in 60% of the cases and shows lens adherence to the posterior cornea due to lack of separation from the lens. This finding is associated with cataract. Peters anomaly may have an inherited pattern.

Isolated Peters anomaly usually occurs in an autosomal recessive pattern but autosomal dominant patterns have been reported as well. The recessive disorder may be caused by a mutation in several genes, notably PAX6, PITX2, CYP1B1 and FOXC1. It is likely the result of a disruption in some common pathway or pathways.

The anomaly has been reported as a feature of fetal alcohol syndrome.

Treatments:

Treatment of Peter's anomaly depends on the severity of the opacity. If the opacity is bilateral and blocks the visual axis, nystagmus and dense bilateral amblyopia are common. Under these conditions, corneal transplantation is advised. The prognosis for corneal transplantation is guarded because of the complications of graft rejection and dense amblyopia. Because of the poor results with corneal transplantation, patients with unilateral Peter's anomaly are often treated conservatively without surgery if at all possible. Even so, some unilateral cases require early intervention. Because of the high incidence of glaucoma associated with Peter's anomaly, these children require constant follow-up to make sure the pressures are under control. Vision in the affected eye varies, but is usually poor secondary to the glaucoma and dense amblyopia.

Abnormalities:

Short stature, mental retardation, abnormal ears, cleft lip and/or palate, defects of extremities, genitourinary system defects, cardiovascular anomalies, and gastrointestinal defects are systemic abnormalities that effect people with Peters Anomaly. Ocular effects are congenital glaucoma, Microphthalmos, iris hypoplasia, aniridia, persistent fetal vasculature, colobomas, and optic disc hypoplasia.
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Developed by Samantha Pipes