Familial Occurrence of Iris Atrophy and Cataract

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A description is given of a syndrome occurring at an early age, comprising atrophy of the posterior layer of the iris and cataract, which was found in five families. Heredity would appear to be autosomal and dominant.

Discussion

Worst: The dark ring behind the iris is considered by you to be the projection of a ring of pigment on the equator of the lens. This pigment can be seen in cases of coloboma. Nevertheless it is possible that this may be an optical phenomenon, the visible ring of pigment at the equator may not produce a dark ring but may be overilluminated. In preparations photographed by the method you have used, the equator sometimes appears as a dark ring even after the iris has been removed. The ring can naturally be the result of both an optical and an anatomical effect. Do you consider that the optical effect (total reflection from the sharp curve of the equator) or the pigmentary concentration is more important?

Breebaart-de Miranda: It is quite possible than an optical effect is involved in addition to the pigmentation, but in our patients at least a pigment ring could definitely be demonstrated.

Waardenburg: Do you understand why these irides react so poorly to mydriatics? Are the pupillary reaction normal without anomalies of dilatation? Were the atrophic areas of the pigment epithelium very variable in size? It seems to me to be an unknown combination.

Breebaart-de Miranda: We do not know reason for the poor mydriasis. The pupillary reactions were normal in all cases. The atrophy of the posterior layer of the iris was sometimes patchy and the patches were of very variable size.