REPORTING A CASE OF AXENFELD-RIEGER SYNDROME

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First described by Axenfeld in 1920, it is characterized by a posterior embryotoxon and iris strands adherent to the anteriorly displaced Schwalbe's line. In 1935 Rieger described congenital iris abnormalities including iris hypoplasia, correctopia and polycoria, now called Rieger anomaly. Aim: To diagnose as early as possible Axenfeld- Rieger syndrome and treat if necessary. Method: A 10 year old girl was brought by her mother with complaints of low vision and funny looking eyes. Results: On examination the patient had an umbilical skin tag, a cardiac murmur diagnosed as Atrial septic defect. She also had dental abnormalities which included microdontia and adontia. Her BCVA was 6/6 in both eyes and she had compound myopic astigmatism. Her anterior segment evaluation revealed iris stromal atrophy in the right eye with a posterior embryotoxon. The left eye showed polycoria and correctopia with a posterior embryotoxon. IOP at the time was 12mm of hg OD and 13 mm of hg OS. Gonioscopy revealed peripheral anterior synechiae involving less than 180 degrees in both eyes. Fundus examination was normal in both eyes. She was diagnosed as case of AXENFELD RIEGER syndrome and is being followed up once every 6 months for any glaucomatous changes that might emerge. She was referred to a cardiologist and a dentist for appropriate care. Conclusion: To diagnose AXENFELD RIEGER syndrome, of relevant importance are ophthalmic examination, dental and cardiac examination, general physical examination. Early detection of eye problems to reduce effects of glaucoma and referrals for cardiac management.