

## Juvenile onset primary open angle glaucoma in members of a single Indian family-A case series

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**Abstract:** We report a rare case series of bilateral juvenile open-angle glaucoma (JOAG), in three young girls of a same family with discussion of current understanding of its pathogenesis, differential diagnosis, and management. Two sisters named A and B aged 9yrs and 16yrs respectively presented in eye opd chiefly for blurring of vision and were found to have best corrected visual acuity(BCVA)of 6/18both eyes(BE) for A and 6/12,6/24 in right eye(RE) and left eye(LE) respectively for B with normal anterior segment examination. There was an incidental finding of high intra ocular pressure(IOP) in both eyes for both the girls on routine tonometry. Dilated fundus examination revealed normal optic disc and cup disc (CD) ratios with subsequent automated perimetry showing visual field defects in eyes of both the girls.Eldest sister aged 22years was having very poor visual acuity with only hand movement appreciation, IOP was 25.8 mm of hg(BE) with CD ratios of 0.9(RE) and 1(LE).She was on anti glaucoma medications since 1 month and already operated withtrabeculectomy(BE).There was wide open angle on gonioscopy for all the three sisters.

**Keywords:** JOAG,POAG,Juvenile, Adolescent.

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### I. Introduction

Juvenile open-angle glaucoma (JOAG) is a rare form of glaucoma that differs from adult-onset primary open angle glaucoma (POAG) in its age of onset and often in the magnitude of IOP elevation. By definition JOAG has its onset between 3 and 40 years of age, while POAG has its onset after the age of 40. Also, patients with JOAG often have extremely high IOP, sometimes greater than 50 mm Hg. The genetic basis of JOAG is much more obvious than that of POAG.The purpose of reporting this case series is to emphasize the importance of tonometry and ocular examination regardless of patient's age.

### II. Case History

Two sisters named A and B aged 9years and 16years presented in eye opd chiefly for blurring of vision and were found to have BCVA 6/18(both eyes)for A and 6/12(RE) ,6/24(LE) for B and normal anterior segment examination. Non contact tonometry was done with dilated fundus examination and subsequent reliable and reproducible visual field tests using standard 30-2 Humphrey automated perimetry were analysed. Gonioscopy was done to rule out angle closure glaucoma and other angle anomalies.

On non contact tonometry high IOP was detected incidentally with around 50mm of hg value for both the girls in both the eyes which were reproducible on the second visit also. None of them complained of eye pain in spite of high IOP. Dilated fundus examination revealed normal optic disc and CD ratios for both of them. Automated perimetry showed visual field defects in eyes of both the girls. Eldest sister aged 22years was having BCVA HM+ve(BE),IOP 25.8 mm of hg(BE),CD ratios of 0.9(RE) and 1(LE). She was on anti glaucoma medications since 1 month and already operated for trabeculectomy(BE).Gonioscopy revealed wide open angle for all the three sisters.

Corneal diameters, keratometry and axial lengths were normal for their ages.After ruling out all other causes of childhood glaucomas, a diagnosis of juvenile onset primary open angle glaucoma was made.

### III. Management

Both the younger siblings were started on systemic acetazolamide from the day of their diagnosis to control IOP and were also advised trabeculectomy for further management.

### IV. Discussion

Juvenile glaucoma is a rare juvenile-onset open-angle glaucoma (JOAG).This entity does not include other childhood glaucomas.JOAG is differentiated from late congenital glaucoma and other childhood glaucoma by the absence of buphthalmos, megalocornea, Descemet's breaks (Haab'sstriae), or anterior segment dysgenesis.

Understanding of juvenile onset primary open angle glaucoma is important because of it's significant difference with adult forms of open angle glaucoma especially with regard to inheritance, prevalence, severity and age of

onset. While the adult form of the disease is likely to be inherited as a complex trait, without an obvious pattern, JOAG is inherited as an autosomal dominant Mendelian trait with high penetrance.<sup>1</sup>

Similar three separate cases of JPOAG were reported by Bachman J A at Illinois Institute(Chicago).Age of the cases were 16 years,9 years and 28 years at diagnosis.<sup>2</sup> Larry Hou-YanNg also reported two cases of JPOAG in two Chinese children of age 4 years and 12 years at Hong Kong polytechnic university in the year 2008.Although our case series does not demonstrate association of JOAG with myopia or keratoconus but Some of the case series in the literature shows association of JOAG with keratoconus<sup>3</sup> and myopia pointing out the importance of proper ophthalmological work up of a patient with JOAG.

## **V. Conclusion**

Early screening of other members of the family (irrespective of age) with a known glaucoma patient can help to diagnose JOAG at an earlier stage and could probably postpone the final visual outcome if not totally prevent it. Surgical management should be undertaken without any further delay because of the severity of the disease and its early age of onset.

## **References**

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