Juvenile angle closure management: The role of lens extraction and goniosynechialysis

John Y. Lee, Audina M. Berrocal, Alana L. Grajewski, Ta Chen Chang*
Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, 900 NW 17th St, Miami, FL, United States

ARTICLE INFO
Keywords:
Angle closure
Juvenile
Lens extraction
Goniosynechialysis
Retinopathy of prematurity

ABSTRACT

Purpose: Angle closure (AC) is a rare condition in young people. In adults with AC, lens extraction and goniosynechialysis (LE-GSL) are effective in restoring angle anatomy and function. However, the efficacy of LE-GSL is poorly understood in the juvenile population. In this study, we report the efficacy and safety of LE-GSL in a series of young patients with AC.

Methods: We reviewed the medical records of consecutive patients with AC. Eyes were included if aged <40 years at the time of diagnosis, had angle closure, at least one month of postoperative follow-up, and had undergone LE-GSL between January 1, 2015 and June 30, 2019. Eyes were excluded if they had undergone prior incisional glaucoma surgery. AC was defined as elevated untreated intraocular pressure (IOP) > 24 mmHg and (1) less than 180° of visible trabecular meshwork, or (2) any peripheral anterior synechiae noted on gonioscopy, or (3) iridocorneal apposition prior to dilation > 180° identified on anterior segment imaging.

Results: A total of 11 eyes (7 patients) were included. The mean ages of diagnosis and LE-GSL were 19.0 and 21.2 years, respectively. Of the 11 eyes, 8 had a history of laser retinal ablation for the treatment of retinopathy of prematurity (ROP, 72.7%). Intraoperatively, 7 eyes received intraocular lens (63.3%), 6 had concurrent vitrectomy (54.5%), and 4 had concurrent endocyclophotocoagulation (36.4%). Following LE-GSL, visual acuity (VA) improved from a mean preoperative LogMAR of 0.88 (20/150 Snellen equivalence) to a mean LogMAR of 0.68 (20/100 Snellen equivalence, P = 0.029). IOP significantly decreased from 18.2 mmHg (Tmax 38.4 ± 12.2 mmHg) preoperatively to a mean of 11.3 mmHg postoperatively (P = 0.009). The number of glaucoma medications was significantly reduced from a mean of 2.4 preoperatively to no medications at final follow-up (mean duration of 13.7 months, P < 0.001). There were no significant intra- or postoperative complications in any eyes.

Conclusions and importance: In our cohort, LE-GSL significantly lowered IOP, improved visual outcomes, and decreased medication burden in young patients with AC, many of which had infantile retinal ablation for ROP. LE-GSL may be considered an effective intervention in young patients with AC.

1. Introduction

With peak incidence occurring between 55 and 70 years of age, angle closure is a rare finding in children and young adults, which comprises less than 2.5% of the population with angle closure.1 Etiologies of angle closure in young patients may include plateau iris syndrome, iridociliary cysts, and retinopathy of prematurity (ROP) rather than primary relative pupillary block, which is more commonly seen in older individuals. Currently, there is no consensus among pediatric glaucoma specialists on the optimal surgical approach for juvenile angle closure, and the outcomes of surgical management remain uncertain.2 In this study, we examined the safety and efficacy of lens extraction/goniosynechialysis (LE-GSL) in the management of juvenile angle closure.

2. Materials and methods

We performed a retrospective review of medical records in consecutive young patients with angle closure. Patient were identified using financial claims data, and were included for analysis if they were younger than 40 years of age at the time of diagnosis, had angle closure, had at least one month of post-operative follow up, and had undergone LE-GSL between January 1, 2015 and June 30, 2019. Patients whose records lacked sufficient detail in pre-operative angle anatomy and/or surgical techniques were excluded. Angle closure was defined as
elevated untreated intraocular pressure (IOP) ≥ 24 mmHg and (1) less than 180° of visible trabecular meshwork, or (2) any peripheral anterior synechiae noted on gonioscopy, or (3) iridocorneal apposition prior to dilation of 180° or greater on anterior segment imaging. Glaucoma is defined as the presence of optic nerve damage based on structural/functional testing; those without glaucoma are considered glaucoma suspects.

The study was approved by the University of Miami Miller School of Medicine Institutional Review Board. It was fully compliant with the requirements of the United States Health Insurance Portability and Accountability Act and adherent to the tenets of the Declaration of Helsinki.

The following historical information was extracted from the review of medical records: sex, laterality, age of diagnosis, age at the time of surgery, number of pre-operative glaucoma medications, whether the patient had undergone retinal ablation for retinopathy of prematurity, pre-operative visual acuity (VA) and intraocular pressure (IOP), and maximum IOP (Tmax). The following intraoperative information was extracted: presence or absence of concurrent intraocular lens (IOL) implantation, vitrectomy, and endoscopic cyclophotocoagulation (ECP). Post-operatively, IOP at 3 and 6 months, final VA, final IOP, final number of glaucoma medications, and duration of follow up were recorded. While not always explicitly stated in the records, the decision to perform concurrent vitrectomy is usually based upon suspect vitreoretinal traction as prophylaxis against postsurgical retinal detachment, while ECP is offered if significant anterior rotation of the ciliary body is observed in order to rotate the ciliary body posteriorly. VA was converted to logarithmic minimum angle of resolution (log MAR) for statistical analysis, and paired, 2-tailed student t-tests with 95% confidence interval (P < 0.05) were performed to note statistical significance between corresponding pre- and post-operative findings.

3. Results

A total of seven patients (eleven eyes) were included in this series, including 8 eyes diagnosed with glaucoma and 3 eyes with suspected glaucoma. Patient baseline characteristics and preoperative, intraoperative and postoperative details are outlined in Table 1. More than two-thirds of the eyes had a history of retinal ablation for retinopathy of prematurity, and over half of the eyes received concurrent IOL implantation and vitrectomy. None of the patients had anterior segment neovascularization.

A comparison of visual acuity, IOP, and glaucoma medication burden before and after LE/GSL is outlined in Table 2. Patients were followed-up for a mean (SD) length of 13.7 months (11.5) with no intra- or postoperative complications in any eyes. Following the surgery, there was a statistically significant improvement in VA and a statistically significant decrease in IOP (35.2% reduction; P = 0.009) after a mean follow up of 13.7±11.5 months. The number of glaucoma medications (topical and systemic) significantly reduced from a mean (SD) of 2.4 (1.6) preoperatively to no medications at final follow-up (difference = −2.4; 95% CI, −2.7 to −2.0; P < 0.001). Excluding the four eyes that had undergone ECP, there is a 31.1% reduction in IOP (preoperative IOP 17.0 mmHg preoperatively and 11.7 mmHg postoperatively), although this comparison failed to reach statistical significance (P = 0.10) due to the small sample size. There is a significant reduction in the number of medications (from mean of 2.14 preoperatively to zero postoperative).

4. Discussion

Angle closure in young patients presents significant management challenges. The shallow anterior chamber in a phakic eye increases the risk of choioidal effusion and corneal decompensation, while the patient’s young age increases the cumulative risk of bleb-forming procedures.3 The role of lens extraction (with or without concurrent goniosynechialysis) in adult-onset angle closure is well validated.4 However, to the authors’ knowledge, this is the first study to evaluate the safety and efficacy of LE-GSL in the treatment of angle closure in patients younger than 40 years of age. In our cohort, all patients were able to discontinue glaucoma medications, and the large majority of

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**Table 1**

| Patient Number | Sex | Age (mo) | Laterality | Age of diagnosis (mo) | IOL | GSL | ECP | Preoperative | Postoperative |
|----------------|-----|----------|------------|----------------------|-----|-----|-----|--------------|---------------|
| 1              | F   | 10       | R          | 24                   | Yes | No  | No  | Yes          | No            |
| 2              | F   | 36       | R          | 37                   | Yes | No  | No  | Yes          | No            |
| 3              | F   | 10       | R          | 11                   | Yes | No  | No  | No           | No            |
| 4              | F   | 17       | R          | 17                   | Yes | No  | No  | Yes          | No            |
| 5              | F   | 25       | R          | 26                   | Yes | No  | Yes | No           | Yes           |
| 6              | M   | 16       | L          | 16                   | No  | No  | No  | Yes          | No            |
| 7              | F   | 8        | L          | 11                   | No  | No  | No  | Yes          | No            |

**Demographics**

- **Sex**: F (female), M (male)
- **Age**: in months
- **Laterality**: R/L (right versus left eye, R right, L left)
- **Age of diagnosis**: in months
- **IOL**: intraocular lens implantation
- **GSL**: goniosynechialysis
- **ECP**: endocyclophotocoagulation

**Preoperative**

- **VA**: visual acuity (in Snellen fraction)
- **Tmax**: maximum intraocular pressure during follow up period (in mmHg)
- **IOP**: intraocular pressure (in mmHg)
- **Drops**: number of topical pressure-lowering agents
- **Oral CAI**: carbonic anhydrase inhibitor

**Intraoperative**

- **IOL**: intraocular lens implantation
- **Vit**: vitrectomy
- **GSL**: goniosynechialysis
- **ECP**: endocyclophotocoagulation

**Postoperative**

- **3 mo**: intraocular pressure at 3 months
- **6 mo**: intraocular pressure at 6 months
- **Final IOP**: final intraocular pressure
- **Final VA**: final visual acuity
- **Final meds**: total number of topical and oral pressure-lowering agents

**Key**

- CAI: carbonic anhydrase inhibitor
- CF: count finger
- Drops: number of topical pressure-lowering agents
- ECP: endocyclophotocoagulation
- F: female
- GSL: goniosynechialysis
- HM: hand-motion
- IOL: intraocular lens implantation
- IOP: intraocular pressure
- L: left
- LogMAR: logarithmic minimum angle of resolution
- M: male
- NA: not available
- R: right
- Tmax: maximum intraocular pressure during follow up period
- VA: visual acuity
- mo: months
- yrs: years

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40 years). A retrospective study by Kameda et al. indicates that for 109 patients treated with phacoemulsification and GSL patients achieved significant improvements in IOP and VA without requiring a bleb-forming glaucoma procedure.

Comparison of visual acuity, intraocular pressure and glaucoma medication burden before and after lens extraction-goniosynechialysis.

| Characteristic                        | Mean (SD) | Mean difference (95% CI) | P-value |
|---------------------------------------|-----------|--------------------------|---------|
| VA (logMAR)                           |           |                          |         |
| Pre-operative                         | 0.88 (0.77) | –0.20 (–0.25 to –0.15) | 0.029   |
| Post-operative                        | 0.68 (0.68) |                          |         |
| IOP (mmHg)                           | 18.2 (6.4) |                          |         |
| 3 months (N – 10)                     | 13.5 (3.3) | –5.9 (–7.4 to –4.5)      | 0.043   |
| 6 months (N – 8)                      | 11.3 (0.7) | –4.8 (–6.0 to –3.5)      | 0.037   |
| Final follow up (N – 11)              | 11.8 (4.8) | –6.4 (–7.7 to –5.0)      | 0.009   |
| Number of glaucoma medications (N)    | 2.4 (1.6)  | –2.4 (–2.7 to –2.0)      | <0.001  |
| Topical medications                  | 2.1 (1.2)  | –2.1                     |         |
| Systemic CAI                         | 0.3 (0.5)  | –0.3                     |         |
| Mean follow up duration (months)      | 13.7 (11.5)|                          |         |

Table 2

* Snellen equivalence of 20/150.
1 Snellen equivalence of 20/100.
2 Mean maximum pre-operative IOP during study period = 38.4 ± 12.2 mmHg; Abbreviations: SD (standard deviation), CI (confidence interval), logMAR (logarithmic minimum angle of resolution), CAI (carbonic anhydrase inhibitor).

Since angle closure is an uncommon finding in young patients, there are few reports on the efficacy of GSL in this patient population (age < 40 years). A retrospective study by Kameda et al. indicates that for 109 patients treated with phacoemulsification and GSL patients achieved significant improvements in IOP and VA without requiring a bleb-forming glaucoma procedure.

In conclusion, our case series demonstrated that LE-GSL is a safe and efficacious surgical option that can significantly lower IOP, improve visual outcomes, and alleviate medication burden in young angle closure patients.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Funding

NH Center Core Grant P30EY014801, Research to Prevent Blindness Unrestricted Grant, The University of Miami Institute for Advanced Study of the Americas 2019 Pilot Grant, Research to Prevent Blindness Medical Student Fellowship.

Acknowledgements

The authors thank Mr. and Mrs. Theofanis and Wendy Kolokotrones for their generous philanthropic support of The Samuel & Ethel Balkan International Pediatric Glaucoma Center’s research efforts.

References

1. Ritch R, Chang BM, Liebmann JM. Angle closure in younger patients. Ophthalmology. 2003;110(10):1880–1889.
2. Banitt MCJ, Cvenkel B, Ramulu P, et al. Section 7: glaucoma associated with non-acquired ocular anomalies. In: World Glaucoma Association Consensus Series – 9: Childhood Glaucoma. 2012:155–177. Weissreb RN, Grajewski A, Papadopoulos M, Grigg J Freedman S.
3. Gedde SJ, Herndon LW, Brandt JD, Budenz DL, Feuer WJ, Schiffman JC. Postoperative complications in the Tube versus Trabeculectomy (TVT) study during five years of follow-up. Am J Ophthalmol. 2012;153(5):804–814. e801.
4. Azaiza-Blanco A, Burt J, Ramsay C, et al. Effectiveness of early lens extraction for the treatment of primary angle-closure glaucoma (EAGLE): a randomised controlled trial. Lancet. 2016;388(10052):1389–1397.
5. Kameda T, Inoue T, Inatani M, Tanihara H. Long-term efficacy of goniosynechialysis combined with phacoemulsification for primary angle closure. Graefes Arch Clin Exp Ophthalmol. 2013;251(3):825–830.