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

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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

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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 post-operative 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) pre-operatively 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 present significant management challenges. The shallow anterior chamber in a phakic eye increases the risk of choroidal 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

Table 1
Patient baseline characteristics and concurrent intraocular procedures.

Demographics						Preoperative					Intraoperative				Postoperative				
Patient Number	Sex	Age onset	Age surgery	R/L	ROP laser	VA	Tmax	IOP	Drops	Oral CAI	IOL	Vit	GSL	ECP	3 mo IOP	6 mo IOP	Final IOP	Final VA	Final meds
1	F	10	24	R	Yes	20/80	50	8	3	Yes	Yes	No	Yes	No	11	11	11	20/70	0
2	F	36	37	R	Yes	HM	62	17	0	No	No	Yes	Yes	No	NA	NA	8	CF	0
2	F	36	37	L	Yes	20/200	55	10	4	Yes	No	Yes	Yes	No	10	12	12	20/70	0
3	F	10	11	R	Yes	20/150	30	24	2	No	No	Yes	Yes	Yes	10	NA	17	20/200	0
3	F	10	10	L	Yes	CF	29	21	2	No	No	Yes	Yes	Yes	20	10	21	20/350	0
4	F	17	17	R	Yes	20/800	31	31	4	Yes	Yes	No	Yes	No	15	NA	15	20/800	0
5	F	25	26	R	Yes	20/60	34	15	2	No	Yes	No	Yes	Yes	14	11	5	20/30	0
5	F	25	27	L	Yes	20/50	44	21	2	No	Yes	No	Yes	Yes	11	11	5	20/30	0
6	M	16	16	L	No	20/40	28	16	2	No	Yes	Yes	Yes	No	16	11	13	20/25	0
6	M	16	17	R	No	20/25	29	16	1	No	Yes	Yes	Yes	No	12	12	11	20/20	0
7	F	8	11	L	No	20/20	30	21	1	No	Yes	No	Yes	No	16	12	12	20/25	0

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), M (male), mo (months), meds (total number of topical and oral pressure-lowering agents), NA (not available).
R/L (right versus left eye; R right, L left), ROP (retinopathy of prematurity), Tmax (maximum intraocular pressure during follow up period, in mmHg), VA (visual acuity), Vit (vitrectomy).

Table 2
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*
	Pre-operative	Post-operative		
VA (logMAR)	0.88 (0.77)[a]	0.68 (0.68)[b]	−0.20 (−0.25 to −0.15)	0.029
IOP (mmHg)[c]	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)	0 (0)	−2.4 (−2.7 to −2.0)	<0.001
Topical medications	2.1 (1.2)	0 (0)	−2.1	
Systemic CAI	0.3 (0.5)	0 (0)	−0.3	
Mean follow up duration (months)		13.7 (11.5)		

[a] Snellen equivalence of 20/150.
[b] Snellen equivalence of 20/100.
[c] 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).

patients achieved significant improvements in IOP and VA without requiring a bleb-forming glaucoma procedure.

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 "younger age is a risk factor for surgical failure."[5] The authors implicated a more robust post-operative wound healing response and inflammatory reaction as potential attributing factors to the increased risk of failure, defined as (1) persistent IOP≥21 mmHg, (2) IOP values of ≥18 mmHg, with or without topical ocular medication, at two consecutive follow-up visits, or (3) additional operations needed. However, the lowest age within the range of this study cohort was 47 years (mean age 70±9.4). Therefore, additional studies will be required to support this claim within the younger patient population.

Furthermore, the pathophysiology of AC in juvenile patients may vary quite dramatically from that of older individuals, which typically results from relative pupillary block (RPB). While RPB still occurs in the juvenile population, the angle closure is often attributed to congenital structural abnormalities rather than age-related phacomorphic lens enlargement. For example, nanophthalmos, which is common in congenital ocular syndromes, presents with short axial length, hyperopia, narrowed angles, and an enlarged lens-to-ocular volume ratio leading to pupillary block.[1] Similarly, anterior segment under-development following infantile retinal ablation for retinopathy of prematurity (8 out of 11 patients in our case series) can cause anterior displacement of the lens-iris diaphragm and result in AC that is refractory to LPI. This proposed pathophysiology suggest that the removal of lens followed by goniosynechialysis has reasonable prospect for restoring the trabecular outflow and controlling IOP, and our findings support this notion.

Limitations of this study include retrospective design, modest sample size, and variable lengths of follow-up. Furthermore, the true efficacy of LE-GSL could not be assessed since the majority of our cohort also had concurrent PPV and/or ECP. However, given the low incidence of juvenile angle closure, designing and implementing a prospective study would be challenging. This study also has weaknesses inherent to all retrospective clinical studies.

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.

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Authorship

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Declaration of competing interest

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CRediT authorship contribution statement

John Y. Lee: Conceptualization, Data curation, Formal analysis, Writing - original draft. **Audina M. Berrocal:** Conceptualization, Resources. **Alana L. Grajewski:** Conceptualization, Resources. **Ta Chen Chang:** Conceptualization, Resources, Formal analysis, Writing - review & editing, Project administration, Funding acquisition.

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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*. 2013:155–177. Weinreb 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. Azuara-Blanco A, Burr 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.