

# Endoscopy-guided in vivo evaluation of ciliary sulcus location in children with ectopia lentis

Jing Yang\* , Zhirong Wang\*, Qianzhong Cao, Yiyao Wang, Jieyi Wu, Zhangkai Lian and Danying Zheng

## Abstract

**Objective:** To assess a new method to measure the distance of the needle passage from the ciliary sulcus to the corneal limbus anterior border (CTC) in eyes with ectopia lentis directly in vivo via endoscopy and to further evaluate the correlations among the CTC, age, automated horizontal white-to-white distance (WTW), and ocular axial length (AL).

**Methods:** The WTW and AL were measured using an optical biometer. An intraocular endoscope was used during transscleral suture fixation of posterior chamber intraocular lenses to identify the true location of the ciliary sulcus. Linear regression analysis was used to assess the correlation between the CTC and other ocular biological parameters, including age, WTW, and AL.

**Results:** Thirty eyes of 30 children with ectopia lentis were evaluated. A statistically significant correlation was found between age and the CTC. The CTC could be predicted by the equation  $CTC = 0.1313 \times \text{Age} + 0.9666$ . No statistically significant correlations were found between CTC and WTW, CTC and AL, WTW and AL, or WTW and age.

**Conclusion:** Endoscopy is useful for precisely suturing intraocular lens haptics in the real ciliary sulcus. Age can be used as an equivalent parameter for prediction of the true ciliary sulcus location.

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**Keywords**

Ciliary sulcus location, endoscopy, ciliary sulcus to cornea limbus anterior border distance, cornea white-to-white distance, ocular axial length, ectopia lentis

Date received: 1 April 2021; accepted: 13 October 2021

**Introduction**

Ectopia lentis occurs as an isolated abnormality or as an ocular sign of a systemic disorder such as Marfan syndrome, homocystinuria, or Weill–Marchesani syndrome. Progressive displacement of the lens may induce high refractive error and impair visual function, especially during sensitive periods of visual development. Therefore, prompt and safe treatment is of paramount importance for children with ectopia lentis. When the optimal correction fails, surgical intervention must be considered. Among all surgical strategies, transscleral fixation of a posterior chamber intraocular lens (IOL) is most commonly performed.

IOLs have gained popularity because of their advantages of fixation close to the natural crystalline lens position and minimal damage to ocular structures.<sup>1–4</sup> Unfortunately, however, a significant disadvantage of IOL implantation is that it is difficult for surgeons to precisely suture the IOL in the sulcus. Postoperative ultrasound biomicroscopy (UBM) examinations have revealed that only 30% to 50% of IOL haptics are sutured in the ciliary sulcus.<sup>5,6</sup> Without direct visualization, surgeons choose the penetrating site based on their own experience, and such an arbitrary technique can result in multiple complications. If haptics are located anterior to the ciliary sulcus, IOL–iris contact, pigment dispersion, and ocular bleeding may occur; if haptics are located posterior to the ciliary sulcus, high aqueous flare and vitreous incarceration may occur.<sup>7</sup>

Eyeball development in children is basically completed by the age of 2 years. Development then enters a slow stage and finally reaches the adult level at approximately 10 to 15 years of age.<sup>8,9</sup> Few studies to date have reported the distance from the corneal limbus to the ciliary sulcus (CTC)<sup>6</sup>; moreover, these studies used adult cadaver eyes as a study model, which may be unreliable because of dehydration. Additionally, no reports have described the CTC in children, let alone children with Marfan syndrome. Therefore, the issue of precise and objective estimation of the ciliary sulcus region in children should be addressed to achieve a better anatomic prognosis and visual outcome. The corneal limbus is commonly defined as the edge of the cornea, and it may be defined in different ways according to the way in which it is clinically identified.<sup>10</sup> From a microscopic approach, the corneal limbus is the junction between the cornea and the sclera; in terms of transparency, however, it is the transition border between the transparent cornea and the opaque sclera.<sup>11</sup> To facilitate measurement and surgical application in the present study, we defined the corneal limbus anterior border as the edge of the transparent cornea.

Intraocular endoscopy has been successfully applied to lens dislocation surgery, and surgeons can precisely implant both IOL haptics in the ciliary sulcus.<sup>12–14</sup> We used this advanced technique to objectively measure the location of the ciliary sulcus in vivo in children with ectopia lentis.

Additionally, previous reports have revealed correlations among the sulcus diameter, horizontal white-to-white distance (WTW), and ocular axial length (AL).<sup>15</sup> Based on these studies, it is reasonable to suspect that these values may be clinically correlated with the distance of the corneal limbus anterior border to the sulcus.

The present study was performed to measure the CTC in vivo in pediatric eyes with ectopia lentis and investigate its relationship with age, WTW, and AL.

## Patients and methods

### Patients

This retrospective study included consecutive pediatric patients who had undergone endoscopy-guided transscleral fixation of a posterior chamber IOL for ectopia lentis from July 2013 to December 2017. Thirty eyes of 30 patients were evaluated (mean age, 6.5 years; range, 4–10 years). Fifteen patients were female and 15 patients were male. If the child underwent surgery on both eyes, we randomly selected the right or left eye for enrollment. However, most of the patients underwent surgery on only one eye. Among the 30 patients, 5 were diagnosed with Marfan syndrome and the remaining 25 had idiopathic dislocation of the lens and could not be diagnosed with Marfan syndrome, homocystinuria, or trauma.

### Biological data measurement

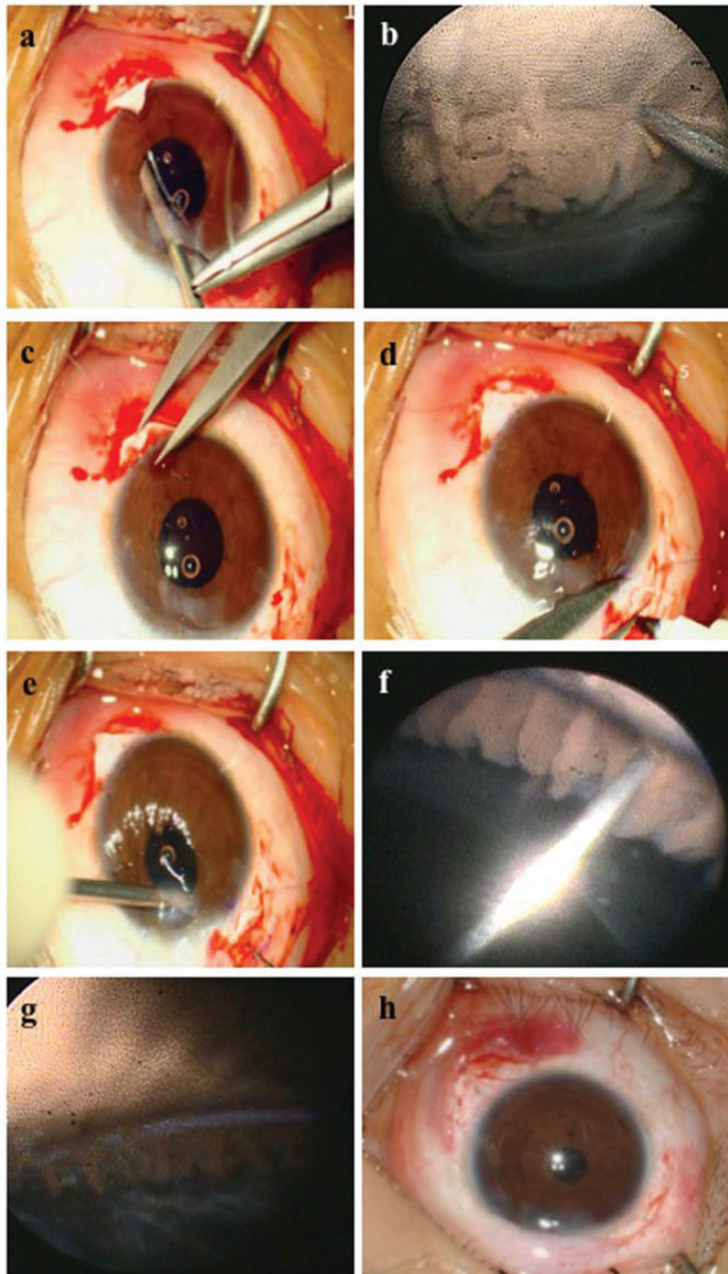
The AL and WTW were measured using an IOLMaster (Carl Zeiss Meditec, Jena, Germany), which is a partial coherence interferometer used for anterior segment measurements and has been proven to be precise for measuring AL and WTW in children.<sup>16</sup>

The CTC was defined as the distance from the ciliary sulcus to the edge of the transparent cornea and was measured with a caliper during the surgery (distance from needle penetration point to edge of transparent cornea).

### Surgical intervention

The surgical intervention is depicted in Figure 1. All operations were performed by the same surgeon (Z.D.Y.). General anesthesia was used in all patients. Endoscopy was performed using an Endo Optiks E2 system (BVI Medical, Waltham, MA, USA). The posterior chamber IOL that was implanted in this study was an AR40e (Abbott Medical Optics, Santa Ana, CA, USA), which is made of methyl methacrylate and has a 6.0-mm optic and overall length of 13 mm. Preoperatively, 0.5% (wt/vol) tropicamide and 0.5% (wt/vol) phenylephrine hydrochloride were used to dilate the pupil. The operation was then performed in the following steps.

1. A 3.2-mm corneal scleral wound was made at the 12-o'clock position.
2. A partial-thickness limbus-based triangular scleral flap 3.0 mm high and 2.0 mm wide was produced at the 3- and 9-o'clock positions.
3. After three ports were made and an infusion cannula was set, the luxated lens was removed.
4. A straight needle carrying 10-0 polypropylene was used to penetrate the 9-o'clock scleral bed parallel to the iris, and the procedure was precisely observed intraocularly by insertion of the endoscope through the 2-o'clock positioned vitreous side port. The 9-o'clock suture was placed *ab inferno* (from the inside), and the 3-o'clock suture was then placed *ab externe* (from outside and in). The needle was



**Figure 1.** Surgical intervention under endoscopy. (a) A needle carrying 10-0 polypropylene was used to penetrate the sclera outward from inside through the ciliary sulcus with the help of endoscopy. (b) Endoscopic image showing the needle penetrating the sclera from inside. (c) Measurement of the distance from the corneal limbus to the ciliary sulcus (CTC) from the penetrating point to the corneal limbus. (d) Marking of the CTC on the other side. (e) Another needle carrying 10-0 polypropylene was used to penetrate the sclera inward from the marked point shown in (d). (f) Image showing the needle penetrating the sclera from outside, with penetration through the ciliary sulcus. (g) Image showing the haptic of the intraocular lens, which was located in the ciliary sulcus. (h) Postoperative image of the eye.

repenetrated when necessary if the penetration site was adjacent to the ciliary sulcus.

5. A needle entered the eye through the 3-o'clock scleral bed. Intraocular observation of the penetration site was performed by insertion of the endoscope through the 10-o'clock side port.
6. A loop of the suture was withdrawn through the corneal sclera wound using a hook. The loop was cut and tied to the haptics of the IOL.
7. The IOL was inserted through the corneal scleral wound and fixated to the ciliary sulcus by tying polypropylene sutures to the scleral bed. Before closing the wound, fixation of the posterior chamber IOL to the ciliary sulcus was confirmed with endoscopy.
8. The three ports and the wound were closed.

### *Follow-up*

The patients were required to visit the doctor 1, 3, 6, and 12 months after surgery, and slit lamp examination, intraocular pressure measurement, and subjective refraction were performed at each visit. The fundus was carefully evaluated after pupil dilation with 0.5% tropicamide (Mydriaticum; Sinqi Pharmaceutical Company, Shenyang, China). The presence of complications, including endophthalmitis, intraocular bleeding, glaucoma, iritis, IOL dislocation, and retinal detachment, was documented. The IOL position was observed by slit lamp examination and Pentacam (Oculus Optikgeräte, Wetzlar, Germany) evaluation.

### *Statistical analysis*

The data are presented as mean  $\pm$  standard deviation. Correlation analysis was performed between the CTC and age, CTC and AL, and CTC and WTW.

Linear regression analysis was performed between the CTC and age, CTC and AL, and CTC and WTW measurements. The regression equation and Pearson's correlation coefficient were calculated for each pair of parameters. Residual analysis was performed for each pair of parameters to represent the potential error in using the CTC and age, CTC and AL, and CTC and WTW to predict the CTC.

Residuals were calculated as the difference between the measured value and the value predicted using the corresponding linear regression equation. The standard deviation and 95% confidence interval of the residuals were calculated for each pair of parameters.

Statistical analysis was performed using Prism 7 statistical software (GraphPad Software, San Diego, CA, USA). A P value of  $<0.05$  was considered statistically significant.

### *Ethics statements*

This study was approved by the ZOC Clinic Institutional Review Board and adhered to the tenets of the Declaration of Helsinki. Verbal informed consent was obtained before patient enrollment. The reporting of this study conforms to the STROBE guidelines.<sup>17</sup>

## **Results**

### *Demographic characterization*

At the first visit, the patients underwent an ophthalmologic examination and were then referred to cardiologists for a cardiovascular evaluation. Information including age, sex, systemic disease, family history, intraocular pressure, best-corrected visual acuity, refraction error, and complete ocular examination data were collected before surgery. The mean age of the 30 patients was  $6.5 \pm 1.7$  years (range, 4–10 years), and they



comprised 15 boys and 15 girls. Their mean best-corrected visual acuity was  $0.6 \pm 0.3$  LogMAR, and their mean intraocular pressure was  $12.4 \pm 1.8$  mmHg.

### Surgical complications

No intraocular bleeding or other intraoperative complications occurred. The only postoperative complication was transient IOL pupillary capture, which occurred in three eyes, and all cases resolved spontaneously. No IOL tilt and decentration occurred. The mean postoperative best-corrected visual acuity was  $0.6 \pm 0.2$  LogMAR.

### Correlation analysis between CTC and age, CTC and AL, and CTC and WTW

The mean scleral exit point for true ciliary sulcus fixation was  $1.9 \pm 0.3$  mm (range, 1.4–2.5 mm). The mean AL was  $24.8 \pm 2.9$  mm, and the mean WTW was  $12.2 \pm 0.4$  mm.

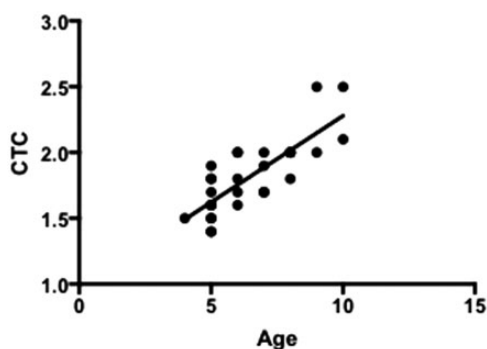
Linear trend correlations were found from the scatter plots only between the CTC and age ( $r = 0.8013$ ,  $R^2 = 0.642$ ,  $P < 0.0001$ , 95% confidence interval = 0.62–0.9013) (Figure 2). The results indicated that the CTC of children with ectopia lentis was positively correlated with their age. No significant correlations were found from the scatter plots between the CTC and WTW or AL (Figure 3). A linear regression model was developed to describe the linear association between the CTC and age. The multivariate linear regression equation was as follows:

$$\text{CTC} = 0.1313 \times \text{Age} + 0.9666$$

The results of the correlation analysis are summarized in Table 1.

### Discussion

The results of this study showed that the traditional estimation of distance may be



**Figure 2.** Scatter plots showing linear trend correlations between CTC and age. Linear trend correlations were found from the scatter plots only between the CTC and age ( $r = 0.8013$ ,  $R^2 = 0.642$ ,  $P < 0.0001$ , 95% confidence interval = 0.62–0.9013). CTC, distance from the corneal limbus to the ciliary sulcus.

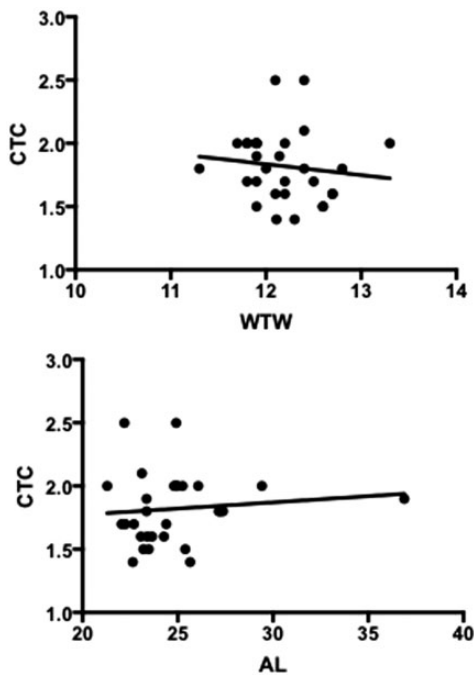
inaccurate. The first anatomic study of the ciliary sulcus was carried out by Duffey et al.<sup>18</sup> in 1989. They used 21 cadaver eyes to study the anatomic structure of ciliary sulcus-fixated IOL implantation and suggested that transscleral sutures should exit the sclera less than 1 mm posterior to the corneal limbus for true ciliary sulcus fixation. However, this experiment was performed in adult cadaveric eyes, not pediatric eyes, and the cadaveric eyes were dehydrated, which may have resulted in inaccuracies. In another study, intraocular bleeding was reported in 13 (52%) of 25 eyes in which the suture was placed 1.0 mm posterior to the limbus.<sup>19</sup> In reports regarding scleral-fixated IOLs in pediatric patients, surgeons tend to place the suture 0.5 to 2.0 mm posterior to the limbus.<sup>1–3,20,21</sup> However, studies of postsurgical UBM evaluation of haptic locations have revealed the difficulty of reliably placing haptics in the ciliary sulcus; the reported success rate ranges from 33% to 55%.<sup>7,19,22</sup> In a further study, Kumar et al.<sup>23</sup> used UBM to guide surgeons to transsclerally fix posterior chamber IOLs

in eyes with inadequate capsules. However, the patients in their study were not merely children with ectopia lentis, and it has been proven that the ciliary sulcus is a type of dysontogenesis that may not be easily detectable on UBM images. In addition, according to our experience, compliance in children aged <7 years is poor during UBM

examination, and additional anesthesia could lead to higher risks in children. Furthermore, these machines are expensive and not available in many hospitals.

In an attempt to overcome these problems, we applied ocular endoscopy for precise suturing of the haptics in the sulcus, measured the true location of the ciliary sulcus, and explored a safe zone for surgeons to choose penetration. Intraocular endoscopy has been applied to ocular surgery for many years, and the development of equipment and techniques has led to its widespread use in lacrimal passage, glaucoma, cataract, and vitreous surgeries. This is the only technique that allows the surgeon to directly observe the structures behind the iris, accurately pass the needle through the ciliary sulcus, and confirm the relation between the haptics and the ciliary sulcus before closing the wound. The results of the present study indicate that it is a safe and efficient method for transscleral fixation of posterior chamber IOLs in eyes with ectopia lentis.

The main objectives of this study were to measure the real distance of the ciliary sulcus posterior to the cornea limbus anterior border in pediatric eyes with ectopia lentis and investigate the relationship of the sulcus-to-cornea limbus anterior border distance with other anterior segment anatomical parameters and age. In our study, the mean location of the ciliary sulcus posterior to the limbus anterior border was  $1.9 \pm 0.3$  mm (range, 1.4–2.5 mm) in pediatric eyes with ectopia



**Figure 3.** Scatter plots between the CTC and WTW or AL. The scatter plots showed no significant correlations between the CTC and WTW or between the CTC and AL. CTC, distance from the corneal limbus to the ciliary sulcus; WTW, white-to-white distance; AL, ocular axial length.

**Table 1.** Correlation analysis.

	r	R <sup>2</sup>	P	95% CI	Equation
Age vs. CTC	0.8013	0.642	<0.0001	0.62 to 0.9013	$CTC = 0.1313 \times Age + 0.9666$
WTW vs. CTC	-0.1781	0.3172	0.3295	-0.496 to 0.182	/
AL vs. CTC	0.1134	0.0129	0.5367	-0.245 to 0.445	/

CI, confidence interval; CTC, distance from the corneal limbus to the ciliary sulcus; WTW, white-to-white distance; AL, ocular axial length.

lentis. Based on our experience, the suitable and safe suture zone may be 1.5 to 2.0 mm posterior to the corneal limbus anterior border in children with ectopia lentis to reduce the incidence of pupillary block. The results of our study showed that there is variability in the CTC among individual patients and that there is a poor relation between the CTC and WTW and between the CTC and AL but a significant correlation between age and the CTC. Age can be a predictor of the ciliary sulcus location for surgeons to determine the site of needle penetration.

The absence of intraocular bleeding in this study, which we have observed more frequently as a postoperative complication among patients undergoing scleral fixation, may have been related to the use of endoscopy. Although the incidence of IOL pupillary capture was high among our cases, all of the cases were transiently captured and resolved spontaneously or vanished as soon as the light shone on the iris. Neither iris synechiae nor other ocular disorders were detected. We considered that the haptics residing in the sulcus may have been slightly close to the iris. A possible explanation for this phenomenon could be that pupil retraction results in a change in the difference between the anterior chamber and posterior chamber, and the pressure pushes the iris backward until it touches the IOL optic.<sup>24</sup> In 2011, Ma et al.<sup>6</sup> suggested that the pars plana location for posterior chamber IOL transscleral fixation was as safe and effective as the ciliary sulcus location and that suturing the haptics in the pars plana can reduce IOL dislocation and pupillary capture of the IOL optic without increasing the complication rate. Another reason for transient IOL capture might be iris laxity, especially in patients with Marfan syndrome.<sup>25,26</sup> According to our experience with this kind of surgery, we suggest that the optimal position of the IOL in the ciliary sulcus is close to the posterior margin of

the sulcus to reduce the incidence of IOL pupillary capture. The suture can be easily and precisely controlled with the assistance of endoscopy to avoid ciliary body injury and intraocular bleeding.

Moreover, in our series, our findings during direct observation of the ciliary sulcus with the assistance of endoscopy were consistent with the findings reported by Pavlin et al.,<sup>5</sup> who suggested that eyes without an intact lens capsule and zonular tension had significantly shorter ciliary processes and a shallower ciliary sulcus than normal eyes with an intact lens and zonular complex.<sup>5,7</sup>

The initial reason that patients presented to our clinic was failed medical examinations for school enrollment, such as poor vision outcomes or high refractive error. This is attributed to most of the patients being younger than 7 years. To better explore the relationship between the CTC and age and whether there is an obvious difference between adults and children, further studies should enroll more patients older than those in the present study. Additionally, this study involved only Chinese children; whether the position of the ciliary sulcus can be expected to be located at the same place in children of other ethnicities requires further verification.

In conclusion, endoscopy is useful for precisely suturing IOL haptics in the real ciliary sulcus. Based on our experience, transscleral suture exit points from 1.5 to 2.0 mm posterior to the corneal limbus anterior border in children with ectopia lentis are recommended to reduce the incidence of pupillary block. Age can be used as an equivalent parameter for prediction of the true ciliary sulcus location.

### **Authors' contributions**

JY performed the statistical analysis and drafted the manuscript. ZW helped to collect the data and participated in drafting the manuscript. QC, YW, JW, and ZL performed the data



collection and statistical analysis. DZ helped to design the study and revise the manuscript. The first two authors (JY and ZW) contributed equally to this work and should therefore be considered co-first authors.


### Declaration of conflicting interest

The authors declare that there is no conflict of interest.

### Funding

This study was supported by a grant from the National Natural Science Foundation of China (grant No. 81873673) to DZ and a grant from the Natural Science Research Foundation of Guangdong Province of China (grant No. 2019A1515010940) to JY.

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