• Clinical Research •

Intraocular lens tilt and decentration in congenital ectopia lentis: baseline characteristics and first-year report

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Abstract

● **AIM:** To investigate the characteristics and associated factors of intraocular lens (IOL) tilt and decentration after transscleral suture-fixated IOL surgery in congenital ectopia lentis (CEL).

● **METHODS:** CEL patients undergoing transscleral suture-fixated IOL surgery were divided into two groups based on implanted IOL type (three-piece IOL or one-piece IOL). The IOL tilt and decentration at 3-month and 1-year postoperative were measured. Multivariate regression analyses were performed to identify the associated factors of IOL tilt and decentration as well as longitudinal changes.

● **RESULTS:** The 61 CEL patients (mean age 9.07±5.05y) in three-piece IOL (M/F: 14/7) group had a greater tilt than those with one-piece IOL (M/F: 28/12) group both at 3-month postoperative (horizontal: *P*=0.024; vertical: *P*=0.048) and 1-year postoperative (horizontal: *P*=0.011; vertical: *P*=0.001). Three-piece IOL group had a greater longitudinal change between 3-month postoperative to 1-year postoperative in IOL tilt (horizontal: *P*=0.028; vertical: *P*=0.026) and a greater longitudinal change in horizontal IOL decentration than one-piece IOL group (*P*<0.05). The longer axial length (AL) was associated with the longitudinal changes in IOL tilt (*P*=0.039), while the three-piece IOL was associated with the longitudinal changes in horizontal IOL decentration 1-year postoperatively (*P*=0.011).

● **CONCLUSION:** IOL tilt is greater in the three-piece IOL group than that in the one-piece IOL group 1-year postoperatively. The three-piece IOL is associated with greater longitudinal changes of IOL decentration, while longer AL is associated with longitudinal changes of IOL tilt. For CEL patients, more stable IOL type should be selected and patients with longer AL warrant closer monitoring.

● **KEYWORDS:** ectopia lentis; intraocular lens tilt; intraocular lens decentration

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INTRODUCTION

Congenital ectopia lentis (CEL) is a rare but clinically significant disorder classified within the spectrum of connective tissue diseases, characterized by abnormal development of the lens suspensory ligament. Rather than occurring in isolation, CEL frequently serves as a key ocular manifestation of several systemic or hereditary conditions. Among these, Marfan syndrome represents the most common etiological association, while other less frequent causes include homocystinuria, Weill-Marchesani syndrome, and sulfite oxidase deficiency[1-3]. CEL can often lead to severe refractive error, astigmatism, and even amblyopia[4-5]. Previously, our research has shown that CEL significantly impacts the quality of life in affected children. Appropriate ophthalmic surgical intervention can not only improve their visual function but also enhance their health-related quality of life[6]. For patients with insufficient capsular support, different types of intraocular lens (IOL) fixation techniques have been introduced, such as anterior chamber IOL, iris-claw IOL, transscleral sutured posterior chamber IOL, and sutureless intrascleral haptic fixation (Yamane technique)[7-11]. However, most of the CEL

patients are still in their developmental stages at the time of surgery, characterized by relatively soft sclera, active daily behavior, and a tendency to rub their eyes. Therefore, for these patients who meet surgical indications, traditional transscleral sutured posterior chamber IOL technique remains a common and effective option and three-piece IOL or specially designed one-piece IOL (such as Rayner) are the most commonly used IOLs[12-13].

Although this surgical treatment significantly improves visual acuity, there are more postoperative complications when compared to IOL implantation, such as IOL tilt, IOL decentration, and pupillary capture of IOL[14-15]. In a study conducted by Hayashi K, postoperative IOL position was compared across different implantation techniques. The results demonstrated that scleral suture fixation was associated with a greater degree of lens tilt and decentration than either in-bag or out-of-bag IOL implantation methods[16]. Our previous study also found that the transscleral sutured posterior-chamber IOL in CEL patients may lead to significant decentration at three months postoperatively[17]. Previous studies have shown that significant IOL tilt and decentration can lead to refractive changes, increased astigmatism and higher-order aberrations and decrease in visual quality, which can affect the visual development of pediatric patients[18-20].

However, limited research has devoted to elucidating the characteristics and the associative factors of IOL tilt and decentration in patients with CEL. This study aims to investigate the characteristics and the magnitude of longitudinal changes of IOL tilt and decentration one year postoperatively, while identifying the potential risk factors for longitudinal changes of IOL tilt and decentration at one year postoperatively.

PARTICIPANTS AND METHODS

Ethical Approval This retrospective study was carried out at the Zhongshan Ophthalmic Center. Ethical oversight was provided by the Human Research Ethics Committee of Zhongshan Ophthalmic Center (ID:2022KYPJ207), and all procedures were performed in accordance with the Declaration of Helsinki. Written informed consent was obtained from all participants prior to inclusion. In addition, all authors confirm that they fulfill the current International Committee of Medical Journal Editors (ICMJE) criteria for authorship.

Participants A total 61 CEL patients (61 eyes) who underwent scleral fixation of posterior chamber IOL surgery between April 2019 and July 2023 were enrolled. The routine follow-up data at three months (baseline) and one year postoperatively were obtained. The included CEL patients were divided into three-piece IOL (AR40e, AMO, USA) and one-piece IOL (920H/970C, Rayner, UK) groups. The inclusion criteria were as follows: 1) Diagnosis of CEL, and age ≤40y; 2) Meets the following indications for surgery with this being their first IOL procedure: 1) the best corrected visual acuity is ≤0.3 due to lens dislocation; 2) the equatorial portion of the crystalline lens is visible in the center of pupil and causes uncorrectable refractive abnormalities; 3) the crystalline lens is dislocated to the anterior chamber or vitreous cavity; 4) Those who have finished regular postoperative follow-up examinations and have the complete information for this study. The exclusion criteria were as follows: 1) patients with secondary crystalline lens dislocation caused by ocular trauma; 2) individuals with a history of ocular surgery or ocular trauma; 3) patients with Pentacam images that were unclear and unsuitable for inclusion in the study.

Axial Length Postoperative measurements of axial length (AL) and anterior chamber depth were obtained using the IOLMaster 700 (Carl Zeiss Meditec AG) at two time points: 3mo and 1y after surgery. IOL tilt and decentration were assessed with the Pentacam HR system (Oculus Inc., Wetzlar, Germany) and MATLAB image processing software, following previously reported methods[17,21-22]. The pupil center was defined as the midpoint of the line segment connecting the two ends of the iris, while the pupillary axis was determined as the line extending through the pupil center and the center of curvature of the anterior corneal surface. The IOL axis was determined by first fitting lines to the anterior and posterior surfaces of the IOL, identifying their intersection, and then drawing a line perpendicular to these surfaces at the midpoint of that intersection. IOL tilt was measured as the angle between the IOL and pupillary axes, whereas IOL decentration was quantified as the distance between these axes[23]. The vertical and horizontal meridians were designated as the 90-degree and 180-degree meridians, respectively.

Surgical Technique The transscleral suture-fixated posterior chamber IOL was implanted by a single experienced surgeon (Zheng DY) after the crystalline lens were centrally positioned with iris hooks and removed using the irrigation/aspiration (I/A) mode[24]. The IOL haptics were secured with 8-0 polypropylene sutures (Johnson & Johnson, USA) passed on a 26-G needle. The needles were introduced from the ciliary sulcus to the sclera at the 150° and 330° meridians, 2 mm posterior to the limbus. Each patient received either a three-piece or a one-piece IOL. The paired sutures were symmetrically tightened to achieve optimal IOL centration. The suture ends beneath the scleral flap were sequentially tied so that the knots were buried within the sclera, and the scleral flap was then closed with a 10-0 nylon suture. Finally, viscoelastic was injected into the space between the posterior lens capsule and the anterior vitreous membrane. The entire circumferential zonular fibers were then dissected, and the cortex and nucleus were removed using the I/A mode. Anterior vitrectomy was done if there was vitreous overflow.

Statistical Analysis Statistical analyses were conducted for all quantitative variables and differences in ocular parameters between the two groups were evaluated using t-tests. Univariate and multivariate regression models were further applied to identify potential determinants of IOL tilt and decentration, including AL, IOL type and performance of anterior vitrectomy. All statistical procedures were carried out using SPSS version 23.0 (IBM Corp., Armonk, NY, USA) and a $P<0.05$ was regarded as indicating statistical significance.

RESULTS

The demographic characteristic data and ocular biometric parameters of the patients are shown in Table 1. A total of 61 CEL patients with 61 eyes were included in this study. The mean age of CEL patients was 9.07±5.05y, and the age range of the patients was 4–37y, 42 were male and 19 subjects were female. The 21 subjects were implanted with AR40e three-piece IOL while 40 subjects were implanted with 920H/970C one-piece IOL. Among the 61 included patients, 5 underwent intraoperative anterior vitrectomy.

As shown in Table 2, the CEL patients with three-piece IOL had a greater tilt than those with one-piece IOL at 3-month postoperatively (horizontal: 2.52±2.11° *vs* 1.66±1.22°; vertical: 2.51±2.11° *vs* 1.74±1.41°, both $P<0.05$). No statistical difference was detected for IOL decentration between the two groups at 3-month postoperative. The average values of the IOL tilt and decentration in the CEL patients at one year postoperative are shown in Table 2. At 1-year postoperative, CEL patients with three-piece IOL had greater tilt in both horizontal and vertical directions than those with one-piece IOL (horizontal: 2.89±2.58° *vs* 1.63±1.58°, $P=0.011$; vertical: 3.32±2.89° *vs* 1.66±1.26°, $P=0.001$). No statistical difference was detected for IOL decentration between the two groups at 1-year postoperative.

As shown in Table 3, after adjusting for gender and age, multivariate regression analyses showed that anterior vitrectomy was identified as the associated factor for horizontal IOL decentration [$\beta=0.31$ (95%CI: 0.12, 0.90), $P=0.012$] at 3-month follow up, and the use of three-piece IOL was the main factor influencing the IOL tilt in the horizontal and vertical directions [horizontal: $\beta=0.37$ (95%CI: 0.30, 1.38), $P=0.003$; vertical: $\beta=0.33$ (95%CI: 0.27, 1.42), $P=0.004$)], but no significant factors was identified with the IOL decentration at one year follow up.

The CEL patients with three-piece IOL group had greater longitudinal change of IOL tilt than that in the one-piece IOL group in horizontal and vertical direction (horizontal: 1.80±1.59° *vs* 1.17±0.94°; vertical: 1.91±1.81° *vs* 1.19±1.07°, both $P<0.05$) and greater longitudinal change of IOL decentration in horizontal direction (0.46±0.30 *vs* 0.29±0.27 mm, $P<0.05$).

Table 1 The demographic and clinical features of CEL patients

Characteristics	One-piece (n=40)	Three-piece (n=21)
Age, y, mean (SD)	9.823 (5.28)	9.433 (5.45)
Male/female	28/12	14/7
Anterior vitrectomy, *n* (%)	4 (10)	1 (4.76)
Axial length (mm), mean (SD)	25.19 (5.192)	24.65 (5.066)
WTW (mm), mean (SD)	11.80 (0.35)	11.75 (0.4)
SE (D), mean (SD)	-1.521 (0.30)	-1.495 (0.25)
IOP (mm Hg), mean (SD)	13.83 (4.15)	13.02 (2.17)
3-month postoperatively		
Diopters sphere, mean (SD)	0.205 (1.45)	0.250 (1.51)
Astigmatism, mean (SD)	-1.622 (1.02)	-2.434 (1.50)
1-year postoperatively		
Diopters sphere, mean (SD)	0.219 (1.46)	-0.525 (2.09)
Astigmatism, mean (SD)	-1.441 (0.76)	-1.912 (1.07)

CEL: Congenital ectopia lentis; SD: Standard deviation; IOP: Intraocular pressure; WTW: White-to-white; SE: Spherical equivalent.

As shown in Table 4, three-piece IOL group had a greater longitudinal change between 3-month postoperative to 1-year postoperative in IOL tilt (horizontal: $P=0.028$; vertical: $P=0.026$) and a greater longitudinal change in horizontal IOL decentration than one-piece IOL group ($P<0.05$). After adjusting for gender and age, multivariate regression analyses showed that longer AL was identified as a significant risk factor of the longitudinal changes of horizontal IOL tilt [$\beta=0.31$ (95%CI: 0.03, 0.37), $P=0.039$], and three-piece IOL was identified as a significant risk factor for the longitudinal changes of horizontal IOL decentration in Table 5 [$\beta=0.30$ (95%CI: 0.06, 0.36), $P=0.011$].

DISCUSSION

In this study, we investigated the characteristics of IOL tilt and decentration after transscleral suture-fixated IOL surgery in congenital ectopia lentis. Scleral fixation of posterior chamber IOLs is one of the important surgical methods for IOL implantation in patients with insufficient capsule support. Yamane *et al*[25] reported an intrascleral haptic fixation of posterior chamber IOLs technique which does not require fibroin biological glue or sutures for securing the IOL haptic, thereby could avoid complications related to biological glue and sutures. However, since CEL is a progressive disease and primarily affects children during their growth and development, and the long-term efficacy and safety of the Yamane technique remains unknown, this technique has not yet been widely used in in patients with CEL. Our results showed that CEL patients with three-piece IOL had a greater tilt than those with one-piece IOL both at 3-month postoperatively and one year postoperatively. Greater longitudinal changes from 3-month postoperatively to 1-year postoperatively for IOL tilt and decentration were also detected in patients with three-

Table 2 IOL position parameters of CEL patients according to IOL types at 3-month and 1-year postoperatively follow-up

Parameters	3-month postoperatively			1-year postoperatively		
	AR40e	920H/970C	*P*	AR40e	920H/970C	*P*
Tilt (degree)						
Horizontal	2.52±2.11	1.66±1.22	0.024	2.89±2.58	1.63±1.58	0.011
Vertical	2.51±2.11	1.74±1.41	0.048	3.32±2.89	1.66±1.26	0.001
Decentration (mm)						
Horizontal	0.58±0.39	0.68±0.48	0.186	0.59±0.42	0.75±0.45	0.085
Vertical	0.55±0.34	0.65±0.51	0.205	0.52±0.28	0.63±0.51	0.180

IOL: Intraocular lens; CEL: Congenital ectopia lentis.

Table 3 Risk factors for IOL tilt and decentration at 3-month and 1-year postoperative follow-up

Variables	3-month postoperatively				1-year postoperatively			
	Univariate analysis		Multivariate analysis		Univariate analysis		Multivariate analysis	
	β (95%CI)	*P*	β (95%CI)	*P*	β (95%CI)	*P*	β (95%CI)	*P*
IOL tilt								
Horizontal								
AL	-0.00 (-0.82, 0.19)	0.579	0.02 (-0.15, 0.23)	0.697	0.14 (-0.09, 0.31)	0.268	0.23 (-0.024, 0.38)	0.084
IOL type (three-piece)	0.25 (0.00, 1.61)	0.049	0.26 (0.03, 1.74)	0.043	0.41 (0.68, 2.50)	0.001	0.44 (0.40, 1.62)	0.001
Anterior vitrectomy	0.13 (-0.18, 0.35)	0.532	-0.04 (-1.78, 1.30)	0.742	-0.01 (-1.80, 1.70)	0.954	0.01 (-1.594, 1.68)	0.958
Vertical								
AL	0.05 (-0.17, 0.24)	0.321	0.10 (-0.13, 0.28)	0.460	-0.04 (-0.26, 0.20)	0.788	0.10 (-0.126, 0.28)	0.460
IOL type (three-piece)	0.18 (-0.25, 1.61)	0.140	0.18 (-0.28, 1.58)	0.168	0.40 (0.31, 1.77)	0.001	0.28 (0.24, 1.58)	0.016
Anterior vitrectomy	-0.29 (-2.99, 0.26)	0.104	-0.21 (-0.30, 0.30)	0.107	-0.09 (-2.68, 1.26)	0.104	-0.21 (-0.30, 0.30)	0.107
IOL decentration								
Horizontal								
AL	-0.12 (-0.07, 0.03)	0.357	-0.18 (-0.08, 0.01)	0.154	-0.09 (-0.07, 0.03)	0.465	0.15 (-0.08, 0.02)	0.289
IOL type (three-piece)	-0.13 (-0.35, 0.12)	0.092	-0.12 (-0.33, 0.11)	0.327	-0.20 (-0.42, 0.05)	0.122	-0.21 (-0.44, 0.05)	0.123
Anterior vitrectomy	0.27 (0.04, 0.85)	0.031	0.31 (0.12, 0.90)	0.012	0.09 (-0.28, 0.57)	0.496	0.09 (-0.29, 0.57)	0.522
Vertical								
AL	-0.04 (-0.06, 0.05)	0.627	-0.10 (0.43, 2.72)	0.449	0.09 (-0.02, 0.05)	0.510	0.14 (-0.02, 0.06)	0.320
IOL type (three-piece)	-0.09 (-0.33, 0. 52)	0.542	-0.06 (0.15, 0.75)	0.680	-0.10 (-0.24, 0.11)	0.466	-0.07 (-0.22, 0.13)	0.615
Anterior vitrectomy	-0.05 (-0.52, 0.34)	0.611	-0.05 (-0.03, 0.02)	0.731	-0.03 (-0.34, 0.27)	0.840	-0.06 (-0.38, 0.24)	0.645

IOL: Intraocular lens; AL: Axial length; CI: Confidence interval.

Table 4 Longitudinal changes of IOL tilt and decentration from 3-month postoperatively to 1-year postoperatively

Parameters	AR40e	920H/970C	*P*
Tilt (degree)			
Horizontal	1.80±1.59	1.17±0.94	0.028
Vertical	1.91±1.81	1.19±1.07	0.026
Decentration (mm)			
Horizontal	0.46±0.30	0.29±0.27	0.014
Vertical	0.28±0.27	0.31±0.24	0.344

IOL: Intraocular lens.

piece IOL than in patients with one-piece IOL. Longer AL was identified as a risk factor for the longitudinal changes of IOL tilt, while three-piece IOL was identified as a risk factors for the longitudinal changes of IOL decentration.

In this study, at one year postoperatively, the IOL tilt and decentration were greater in patients with three-piece IOL than that in patients with one-piece IOL, and the use of three-piece IOL was a risk factor for IO L tilt in CEL patients. In a previous study, Xing *et al*[26] reported that in cataract patients, postoperative IOL tilt was significantly greater in the three-piece IOL (AR40e) group than in other aspheric IOL groups. Our findings were similar to these findings. Additionally, the longer AL, tilt and decentration of crystalline lens have been considered risk factors for postoperative IOL tilt and decentration in previous studies[27-30], whereas in our results, the use of three-piece IOL was the main factor influencing postoperative tilt and decentration at one year postoperatively. These findings may be explained by the fact that AR40e (three-piece IOL) is made of polymethyl methacrylate (PMMA), which has a high stiffness and there is a 5° angle between the loop plane and the optical surface compared to the 0° angle

Table 5 Risk factors for longitudinal changes of IOL tilt and decentration from 3-month to 1-year postoperative

Variables	Univariate analysis		Multivariate analysis	
	β (95%CI)	*P*	β (95%CI)	*P*
IOL tilt				
Horizontal				
AL	0.27 (-0.02, 0.36)	0.047	0.31 (0.03, 0.37)	0.039
IOL type (three-piece)	0.20 (0.16, 1.42)	0.328	0.22 (-0.11, 1.44)	0.089
Anterior vitrectomy	-0.21 (-2.53, 0.26)	0.107	-0.23 (-2.59, 0.13)	0.076
Vertical				
AL	-0.05 (-0.24, 0.16)	0.716	0.09 (-0.14, 0.27)	0.526
IOL type (three-piece)	0.22 (0.12, 1.80)	0.083	0.22 (-0.11, 1.81)	0.082
Anterior vitrectomy	-0.15 (-0.27, 0.70)	0.244	-0.13 (-2.56, 0.83)	0.311
IOL decentration				
Horizontal				
AL	0.15 (-0.01, 0.05)	0.261	0.17 (-0.011, 0.05)	0.201
IOL type (three-piece)	0.30 (0.04, 0.33)	0.020	0.30 (0.06, 0.36)	0.011
Anterior vitrectomy	0.16 (-0.10, 0.43)	0.209	0.18 (-0.08, 0.44)	0.162
Vertical				
AL	-0.23(-0.07, 0.00)	0.071	-0.16 (-0.06, 0.02)	0.238
IOL type (three-piece)	0.10 (-0.11, 0.24)	0.443	-0.07 (-0.13, 0.22)	0.599
Anterior vitrectomy	0.14 (-0.46, 0.14)	0.283	-0.10 (-0.42, 0.19)	0.457

IOL: Intraocular lens; AL: Axial length; CI: Confidence interval.

between the loop and the optical surface of one-piece IOL (Rayner 920H/970C), Hence, it may be more prone to angular deformation at the connection between the IOL loop and the optical part for AR40e when the IOL is pushed during surgery. Additionally, the Rayner 920H/970C (one-piece IOL) has a closed-loop haptics, a larger optical surface diameter (6.25 *vs* AR40e's 6.0 mm) and larger contact area between haptics and the optical surface, while the AR40e lacks these features. The non-closed C-loop haptics of the AR40e necessitate a gap between the suture knot and the end of the haptics to prevent dislocation, while the closed-loop design of the single-piece IOL allows for sutures to be tied at the ends of the haptics, maximizing support. These differences may contribute to greater postoperative tilt and decentration for AR40e[31-32]. Considering CEL patients usually undergo surgery at school age, when ocular development is still occurring[33-34] the selection of a more stable IOL should be one of the main considerations for surgical management in CEL patients.

As CEL is a progressive disease, we further explored the longitudinal changes in IOL tilt and decentration. Our results showed that the values of IOL tilt and decentration were increased in CEL patients at one year postoperatively compared to three months postoperatively, and patients in the three-piece IOL group had greater longitudinal change of tilt and decentration in horizontal direction than that in the one-piece IOL group. Three-piece IOL was also identified as a risk factor for longitudinal changes in IOL decentration. Our findings suggest that the use of three-piece IOL should be avoided in CEL patients with transscleral suture-fixed posterior chamber IOL implantation. In this study, AL was identified as an associated factor for longitudinal changes of IOL tilt. Wang *et al*[35] found that for cataract patients, the longer AL could affect the stability of the postoperative IOL, and Gu *et al*[19] suggested that the greater IOL decentration was dramatically associated with longer AL. Our results were similar with the above findings that the longitudinal changes of IOL tilt were greater in CEL patients with longer AL. This finding may be explained by the fact that for CEL patients, due to the lack of lens capsule, the IOL is fixed only by bilateral suture fixation, making it difficult for the surgeon to ensure that the bilateral IOL pulling force is exactly equal and to measure the IOL position status. Additionally, CEL is a progressive disease, and the postoperative IOL may become more susceptible to strain as the postoperative AL increases with disease progression. Hence, the findings also suggest that we should increase the postoperative follow-up times for CEL patients with longer AL.

There are several limitations of this study. First, this study is a single-center study of a rare disease, and the generalization of the findings needs to be supported by future multicenter, large-sample studies. Second, as this study only investigated the IOL tilt and decentration at one year postoperatively, longer follow-up data needs to be explored in the future. Third, the sample sizes of the two groups in this study are unequal, and better-matched sample sizes are needed in future studies. However, this study has the following advantages: firstly, it is

the first study focusing on longitudinal changes in IOL tilt and decentration and its influencing factors in CEL patients with a relatively large sample size. Secondly, the included patients experienced longitudinal follow-up with complete data.

In conclusion, for CEL patients enrolled in this study, IOL tilt was greater in the three-piece IOL group than in the one-piece IOL group at one year postoperatively. The use of three-piece IOL was identified as a risk factor of greater longitudinal change of IOL decentration compared with one-piece IOL, and longer AL was a significant risk factor for longitudinal changes of IOL tilt.

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