

# Effect of lens surgery on health-related quality of life in preschool children with congenital ectopia lentis

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## Abstract

• **AIM:** To evaluate the effect of lens surgery on health-related quality of life (HRQoL) of preschool children with congenital ectopia lentis (CEL).

• **METHODS:** A prospective self-controlled study was conducted in Zhongshan Ophthalmic Center. Children aged from 5 to 7y whom were diagnosed with CEL and underwent phacoemulsification with scleral-fixated posterior chamber intraocular lens implantation and their parents were enrolled in this study. All of them completed the child and proxy (parental) PedsQL™ 4.0 before and after the surgery. Their preoperative scores were compared to their postoperative ones. Subgroup analyses were performed based on gender and preoperative bilateral presenting visual acuity of the children.

• **RESULTS:** Thirty-two children with CEL successfully underwent surgery without any complications, among whom 8 had monocular surgery and 24 had binocular surgery. Preoperative and postoperative questionnaires were completed by 32 child-parent pairs. Surgical intervention could significantly improve the vision of affected children ( $P<0.001$ ). The medians of physical, psychosocial and total health scores self-reported by the children were 68.75 (62.50, 81.25), 65.00 (60.00, 80.00) and 67.39 (60.87, 78.26) preoperatively and were 93.75 (87.50, 100.00), 90.00 (83.33, 96.67) and 89.13 (85.32, 95.65)

postoperatively. The preoperative scores of the affected children were significantly lower in all scales than age-matched healthy children ( $P<0.001$ ). All the postoperative scores were significantly higher than the preoperative scores in affected children and their parents ( $P<0.001$ ). In the physical functioning evaluation, the preoperative score reported by parents of girls was higher than parents of boys ( $P=0.041$ ), and the postoperative score of girls was higher than that of boys ( $P=0.036$ ).

• **CONCLUSION:** CEL is associated with significantly worse quality of life in preschool children. Surgical intervention can significantly improve the HRQoL in affected children from both personal and family perspective.

• **KEYWORDS:** congenital ectopia lentis; surgical intervention; health-related quality of life; preschool children

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## INTRODUCTION

Congenital ectopia lentis (CEL) is a rare condition related to subluxation of the natural crystalline lens and it develops progressively with age<sup>[1]</sup>. Dislocation of the lens may cause refractive changes such as high myopia, irregular astigmatism, or aphakic hyperopia that will affect visual acuity. It may also bring about complications including amblyopia, angle-closure glaucoma and retinal detachment, and even blindness<sup>[2]</sup>. If ectopia lentis happens during childhood, which is the crucial period of visual development, it could damage visual function seriously<sup>[3]</sup>.

Visual impairment may influence the general health-related quality of life (HRQoL) of affected children<sup>[4]</sup>. Many studies on the HRQoL in preschool children indicated that congenital eye diseases, such as glaucoma, congenital cataract, hereditary retina disorders, strabismus and amblyopia, may significantly impact the affected children and their families<sup>[5-10]</sup>. Children with these chronic ophthalmic disorders might experience multiple visits and surgeries that might affect their education and social life<sup>[11-12]</sup>. However, whether the quality of life in

paediatric children with CEL is affected remains unknown. The surgical treatment for CEL in childhood aims to change their poor refractive status and alleviate complications caused by dislocation of the lens. Objective measures, such as visual acuity, are usually used to evaluate the effectiveness of the treatment. However, these indicators only evaluate the treatment from the perspective of visual function. Recently, in health care systems, there has been an increasing emphasis on evaluating the impact of disease and treatment on the quality of life from the perspective of patients and their family<sup>[13]</sup>. Thus, the aim of this study was to evaluate the effect of lens surgery on HRQoL of preschool children with CEL in order to provide further insight into the broader impact of this disease and its individualized comprehensive treatment.

## SUBJECTS AND METHODS

**Ethical Approval** The study was approved by the Ethical Committee of Zhongshan Ophthalmic Center (2019KYPJ183) and all the procedures complied with the tenets of the Declaration of Helsinki. Informed consent was given by the parents/legal guardians of the child prior to enrolment in the study.

Between January 2019 and January 2021, a total of 32 paediatric patients aged between 5 to 7 years old were included in this prospective self-controlled study. All patients were seen at the Zhongshan Ophthalmic Center and were diagnosed with CEL after excluding any history of trauma and other ophthalmic complications. Children with dysgnosia or some systemic diseases that can severely impact daily life were also excluded. For example, cardiovascular manifestations of Marfan syndrome such as aortic root dilation or aortic dissection may threaten their lives. All surgeries were performed by the same surgeon and conducted using phacoemulsification, which presented better clinical outcomes and fewer complications compared to traditional intracapsular and extracapsular cataract extraction<sup>[14]</sup>, and implantation of a sclera-fixated intraocular lens (IOL)<sup>[15]</sup>. All eyes underwent bilateral presenting vision test, best-corrected visual acuity (BCVA) test and slit-lamp examination before and after operation.

**Assessment of Health-Related Quality of Life** The patient's quality of life was assessed using the PedsQL 4.0™ instrument<sup>[16]</sup> before and 3mo after the operation. It could be applied to compare not only the responses of an age-matched healthy group but also with other diseases. The questionnaire of the 5-7's version for proxy or parent comprises an eight-item physical health scale evaluating physical function and a 15-item psychosocial health scale with school, social and emotional functioning sub-scales. Each question is scored from 0 (never a problem) to 4 (almost always a problem), which can be transformed into a score ranging from 0 to 100, with higher

scores indicating a better quality of life. Finally, the physical, psychosocial and total scores were produced. The physical health summary score is the same as the physical functioning sub-scale. The psychosocial health summary score is the sum of the 15 items answered on the emotional, social, and school functioning sub-scales. The total score consists of all 23 items. Thus, the questionnaire of 5-7's version for children is a little different from the parents'. Each question is scored at 0 (never a problem), 2 (sometimes a problem) and 4 (almost always a problem), which can be transformed into scores of 100, 50, and 0. Due to their difficulties in reading the content of the questionnaire, all of the questions are asked by the same principal investigator. The questionnaire has excellent internal consistency and clinical validity, as proven by previous studies in China<sup>[17-19]</sup>.

**Statistical Analysis** Bilateral presenting vision and BCVA were converted into logarithm of minimum angle of resolution (logMAR) for statistical analysis. Self- and proxy-reported physical, psychosocial and total scores were generated. Descriptive analysis was undertaken according to the clinical variables. Furthermore, *t*-tests and Wilcoxon-test scores were used to compare the quality of life preoperation with postoperation, as well as with healthy children and subgroup analysis. Internal consistency was assessed with Cronbach's  $\alpha$  coefficient<sup>[20]</sup>. Agreement between the child's own assessment and the parental assessment of their child's quality of life was evaluated using the Bland-Altman measure<sup>[21]</sup>.

## RESULTS

**Participation** Totally 32 parent-child pairs were invited to participate in our study. All participants completed the preoperative and postoperative questionnaires at hospital independently. Totally 21 of the 32 affected children (65.6%) were boys, 11 (34.4%) were girls. The mean age of the children was 6y. Eight children had monocular surgery and 24 children had binocular surgery (Table 1). None of them had postoperative complications, including intraocular pressure elevation, IOL dislocation/subluxation, choroidal detachment, haemorrhage and endophthalmitis. The median of their preoperative and postoperative bilateral presenting vision was 1.00 logMAR and 0.26 logMAR respectively. The preoperative and postoperative BCVA was 0.70 logMAR and 0.15 logMAR. Of the 32 affected children, 17 (53.1%) had moderate visual impairment (bilateral presenting vision between 0.52 logMAR and 1.00 logMAR), and 15 (46.9%) had severe visual impairment (bilateral presenting vision more than 1.00 logMAR) before surgery. Surgical intervention could significantly improve the vision of affected children ( $P < 0.001$ ). A high degree of internal consistency was seen in all completed questionnaires, with Cronbach's  $\alpha$  correlation co-efficient  $> 0.56$  (Table 2).

**Table 1 Basic information of congenital ectopia lentis patients**

Basic information	Number (n=32), n (%)
Male	21 (65.6)
Age (y), median (quartile)	6 (5,7)
Visual impairment	
Moderate (bilateral presenting vision $\leq 1.00$ logMAR)	17 (53.1)
Severe (bilateral presenting vision $> 1.00$ logMAR)	15 (46.9)
Surgery	
Monocular surgery	8 (25)
Binocular surgery	24 (75)

logMAR: Logarithm of minimum angle of resolution.

**Table 2 Reliability indices of health-related quality of life (PedsQL™ 4.0)**

Cronbach's $\alpha$ correlation coefficient	Child-reported	Parent-reported
The total scale	0.865	0.876
Physical	0.561	0.761
Psychosocial	0.854	0.834

**Table 3 Pediatric quality of life inventory (PedsQL™ 4.0) scores for total, summary and sub-scales stratified by congenital ectopia lentis**

Item (n=32)	median (quartile)		
	Preoperative	Postoperative	P
<b>Children</b>			
Total	67.39 (60.87, 78.26)	89.13 (85.32, 95.65)	<0.001
Physical	68.75 (62.50, 81.25)	93.75 (87.50, 100.00)	<0.001
Psychosocial	65.00 (60.00, 80.00)	90.00 (83.33, 96.67)	<0.001
Emotional	65.00 (60.00, 80.00)	90.00 (82.50, 100.00)	<0.001
Social	75.00 (60.00, 80.00)	100.00 (90.00, 100.00)	<0.001
School	65.00 (50.00, 80.00)	80.00 (70.00, 90.00)	<0.001
<b>Parent</b>			
Total	71.19 (59.78, 82.33)	89.13 (85.32, 95.65)	<0.001
Physical	71.88 (62.50, 84.38)	87.50 (81.25, 96.88)	<0.001
Psychosocial	69.17 (62.08, 83.33)	85.83 (74.16, 91.67)	<0.001
Emotional	72.50 (61.25, 80.00)	85.00 (76.25, 93.75)	<0.001
Social	75.00 (65.00, 90.00)	90.00 (75.00, 98.75)	<0.001
School	70.00 (56.25, 80.00)	82.50 (71.25, 88.75)	<0.001

**Preoperative HRQoL of Preschool Children with Ectopia**

**Lentis** The median and quartile of the PedsQL™ sub-scale and total scale scores are shown in Table 3. The preoperative physical, psychosocial and total health scores self-reported by the affected children were 68.75 (62.50, 81.25), 65.00 (60.00, 80.00) and 67.39 (60.87, 78.26). The score of their school sub-scale was the lowest of all. For instance, more than half of affected children found it hard to pay attention in class and had trouble keeping up with their schoolwork. There was no significant difference of the HRQoL scores before surgery between girls and boys. Similarly, no significant difference was found between children with moderate visual impairment and severe visual impairment. As expected, children with CEL had lower HRQoL scores than those previously reported by Chinese healthy children (all  $P < 0.001$ )<sup>[18]</sup>.

**Postoperative HRQoL of Preschool Children with Ectopia**

**Lentis** The postoperative physical, psychosocial and total health scores self-reported by the children were 93.75 (87.50, 100.00), 90.00 (83.33, 96.67) and 89.13 (85.32, 95.65). The postoperative physical, psychosocial, and total scores were significantly higher than the preoperative scores (all  $P < 0.001$ ; Table 3). There was no significant difference between the postoperative psychosocial score and the score of age-matched healthy Chinese children ( $P = 0.51$ )<sup>[18]</sup>. Intriguingly, the postoperative physical score of girls was higher than that of boys ( $P = 0.036$ ).

**Evaluation of the HRQoL on Affected Children by**

**Parents** The physical, psychosocial and total health scores reported by parents were 71.88 (62.50, 84.38), 69.17 (62.08, 83.33) and 71.19 (59.78, 82.33) preoperatively and were 87.50 (81.25, 96.88), 85.83 (74.16, 91.67) and 88.04 (78.26, 93.20) postoperatively. Similarly, the postoperative physical, psychosocial, and total scores were significantly higher than the preoperative scores (all  $P < 0.001$ ), and the preoperative physical score reported by parents of girls was higher than parents of boys ( $P = 0.041$ ; Table 3). There was a wide range of agreement<sup>[21]</sup> between child-parent pairs in all assessments (Figures 1-4).

**DISCUSSION**

Our findings showed that the HRQoL in preschool children with CEL was significantly affected, including physical, emotional, social, and school functioning, as measured by the PedsQL™, especially in school behaviour. The postoperative physical, psychosocial, and total scores were significantly higher than the preoperative scores, which indicating that surgical intervention is more than a good approach to improve the visual function, but is also of great significance to improve the quality of life of these affected children.

To the best of our knowledge, this is the first study that focus on the HRQoL of preschool children with CEL. Previous studies have shown that CEL and its complications severely impaired visual function<sup>[3,22]</sup>. Surgical intervention could significantly improve the visual function of these affected children and is vital for their visual rehabilitation<sup>[23-25]</sup>. It is well-known that childhood is not only a critical period of visual development but also a key stage for psychological and emotional development. As there is an increasing emphasis in health care systems on evaluating the quality of life, evaluation of the effectiveness of therapies requires us to measure not only their visual function but also its impact on the quality of life of affected children, as well as any improvement resulting from such treatments. However, there is no previous research that focus on the function of social psychology among children with CEL<sup>[5-7,13]</sup>. There are three sub-scales in the assessment of psychological functioning, including emotional, social

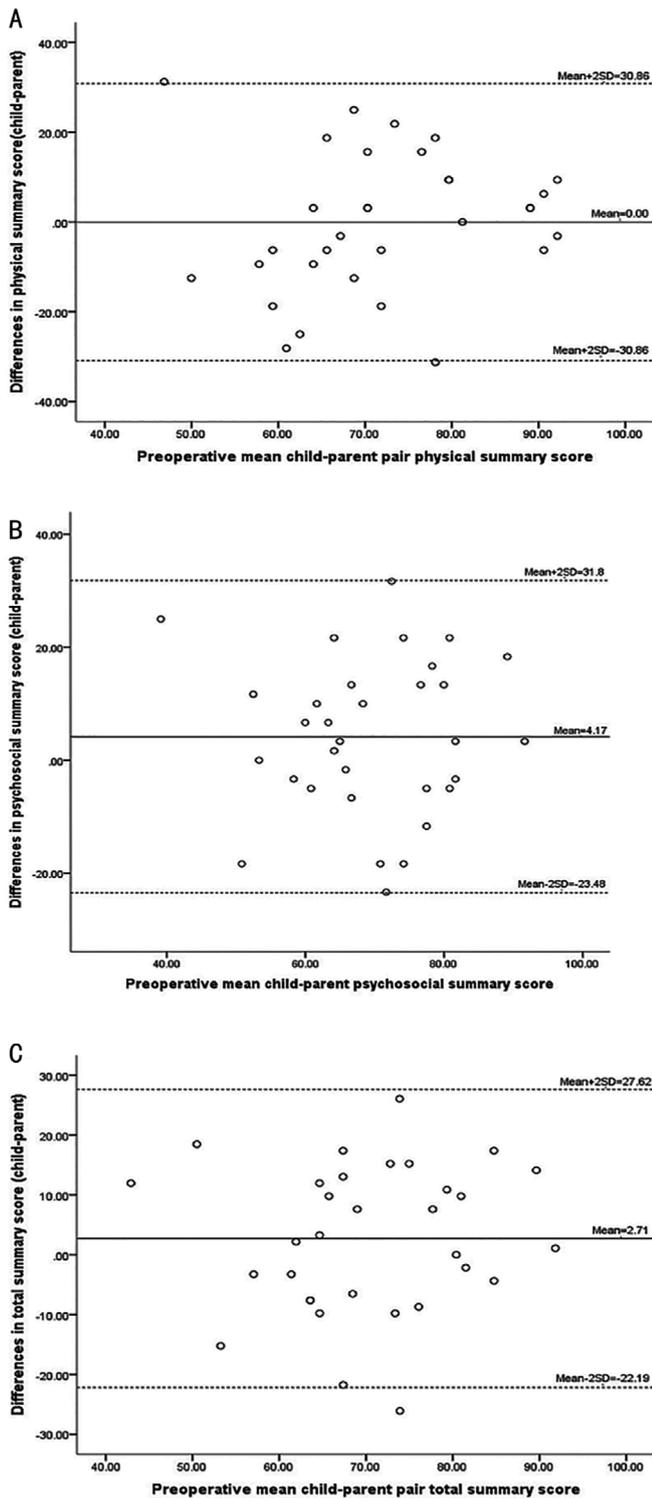


Figure 1 Bland-Altman plots of physical scores (A) psychosocial health scores (B), and total scores for PedsQL scores (C) for preoperative child-parent pairs.

and school functioning. Children with CEL had significantly lower psychological score than age-matched healthy Chinese children, especially their school sub-scale<sup>[18]</sup>. These results indicated that ectopia lentis affected the psychological function of children, especially behaviour in school. Fortunately, the postoperative median subscores of emotional, social and school functioning were significantly higher than those of the

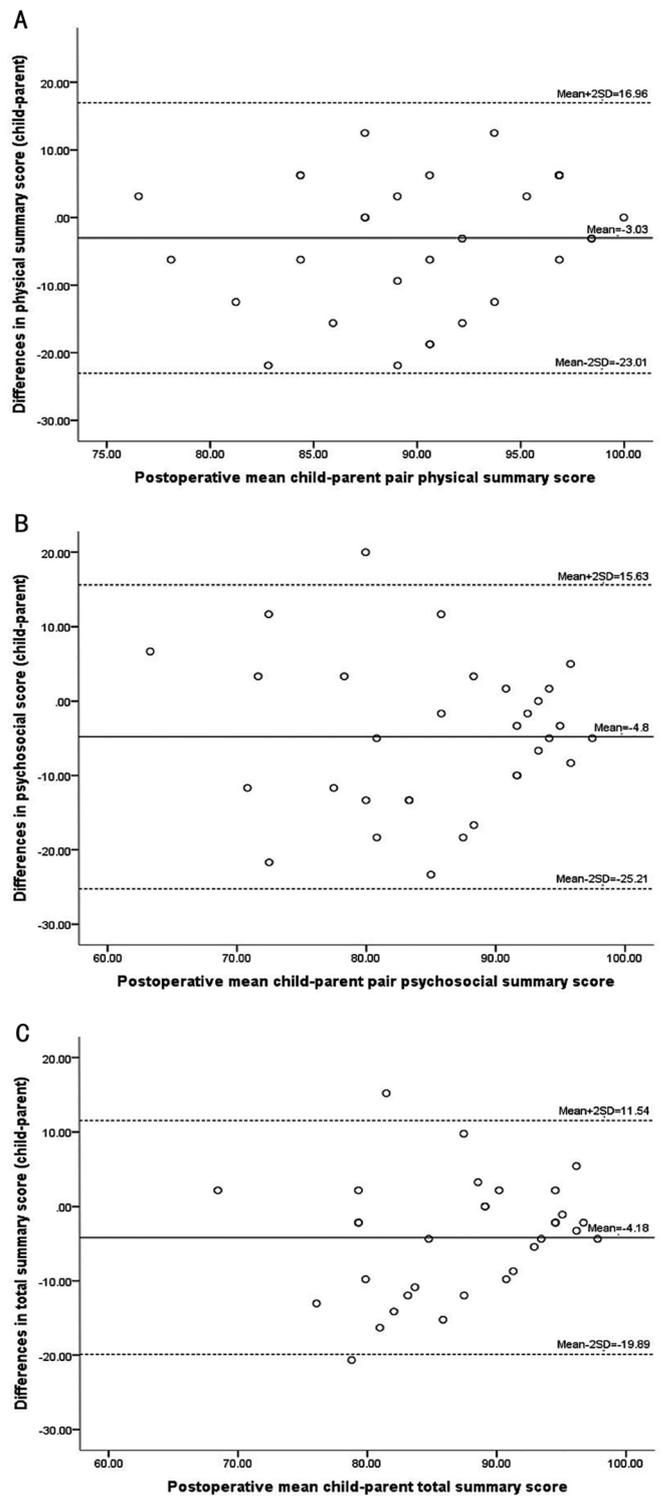
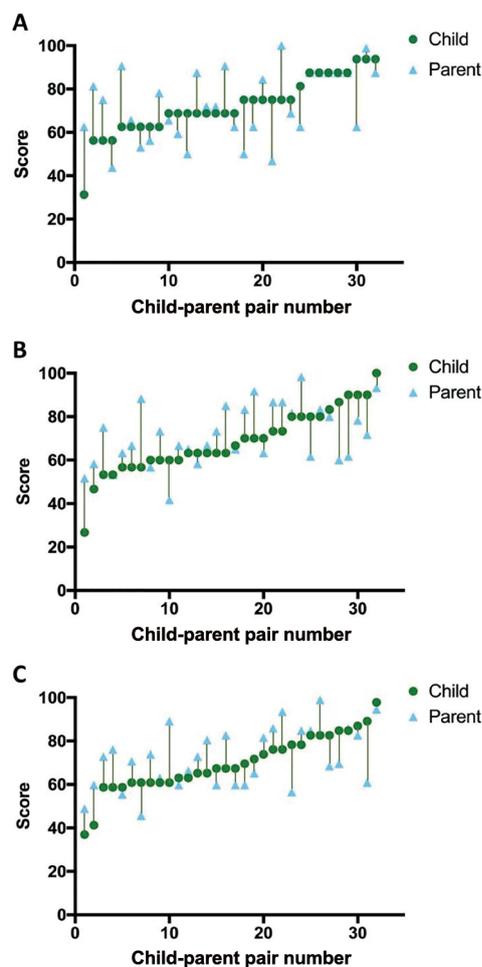


Figure 2 Bland-Altman plots of physical scores (A) psychosocial health scores (B), and total scores for PedsQL scores (C) for postoperative child-parent pairs.

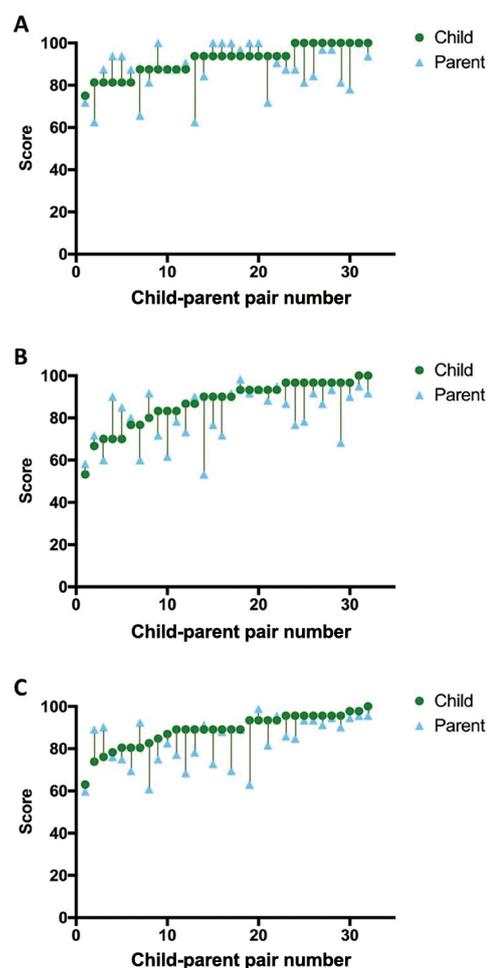
preoperative. Our research showed that surgical intervention is meaningful for the psychosocial and emotional development of affected children. We hope that this research on the quality of life in preschool children with CEL can be an effective way to provide a proper time of surgery, which will facilitate their development of physical and mental health. The PedsQL<sup>TM</sup> 4.0 is an acceptable instrument for assessing



**Figure 3** Distribution of child-parent proxy pair preoperative scores for physical health scores (A), psychosocial health scores (B), and overall scores (C).

the HRQoL in children with ophthalmic disorders, including glaucoma, strabismus, congenital cataract and hereditary retinal disorders<sup>[6-10]</sup>. It has four versions corresponding to different age groups. Children in different age groups have different comprehensions, live in different social environments, and finish different tasks. Each question is set up according to the characteristics of children at different ages. In addition, it allows for both self- and proxy-reporting of HRQoL to objectively assess their quality of life. That will avoid the arbitrary answers that might be obtained from child self-reporting. A recent study on the HRQoL of pediatric patients with allergic conjunctivitis has verified the reliability and validity of the Chinese version of PedsQL<sup>[13]</sup>. There is a high degree of internal consistency was seen in all completed questionnaires in our study.

Of interest, parents assessed the HRQoL of their child with CEL as better than the self-evaluated level by the child before the operation. This may be attributed by that affected children just have blurred vision at the beginning of the disease, like myopia<sup>[26]</sup>. As the weakness or dysfunction of their zonular fibres progresses, they have worse distant vision or present



**Figure 4** Distribution of child-parent proxy pair postoperative scores for physical health scores (A), psychosocial health scores (B), and overall scores (C).

a compensatory head posture so that their parents find their behaviour to be abnormal. This difference in cognition further prompts parents to pay more attention to children's vision abnormalities in the early stage. Intriguingly, the preoperative physical score reported by parents of girls was higher than parents of boys. The possible reason is that boys are more active than girls in daily life, so the visual impairment has a greater impact on the physical function of boys.

The management of CEL in childhood is still challenging currently<sup>[23,27]</sup>. There are no widely accepted criteria for surgical intervention all around the world. Many ophthalmologists decide the time for surgical intervention based on visual acuity. Our study suggests that the evaluation of the quality of life of affected children can also provide a helpful complement to the timing of surgical intervention. In addition, it may be an indication for the evaluation of treatment effect.

In conclusion, our study demonstrated the considerable influence on life quality that CEL can have on affected children. An appropriate surgical intervention can improve not only their visual function but also their HRQoL. We hope that there will be more paediatric vision-specific instruments

developed in the future to offer the prospect of a more detailed evaluation of vision-specific aspects of visual function and quality of life. Their use in future research might help us to deepen our understanding of the real-life impact of paediatric ophthalmic disorders. Moreover, we suggest that the use of generic and vision-specific instruments to assess the child's quality of life should be one of the key outcome measures in future clinical practice.

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**Authors' contributions:** Huang YQ and Cao QZ conceived and designed the study. Huang YQ wrote the main manuscript and prepared all figures. Wang YY analyzed the data, Zhou YJ conducted the questionnaire. Zheng DY oversaw the project and assisted with the writing of the manuscript. All authors reviewed the manuscript.

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