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Title: Quality of Life and Functional Vision in children treated for cataract - a cross-sectional study

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Contribution of authors: ADN and MP developed the study protocol. VT enrolled participants, collected data and entered data onto the electronic database which ADN had developed. ADN and CB conducted data analysis. All authors reviewed and discussed and interpreted the data acquired. ADN drafted the manuscript, which was then critically reviewed and modified by all authors.

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Abstract

Aims: Children with cataract and their families face intensive medical and surgical management, with numerous hospital attendances, topical medications and surgical procedures, as well as uncertainty about the child's future visual ability, education and independence. Little is known about the impact on functional visual ability, vision- and health-related quality of life (VR-, HR-QoL).

Methods: 72 children age 2 to 16 years (mean 8.45, standard deviation SD 4.1) treated for developmental or secondary cataract and their parents/carers completed three validated instruments measuring functional visual ability, VR- and HR-QoL: the Cardiff Visual Ability

Questionnaire for Children (CVAQC), Impact of Vision Impairment for Children (IVI-C) and PedsQL™ V 4.0.

Results: All scores are markedly reduced: median (interquartile range IQR) CVAQC score -1.42 (-2.28 to -0.03), mean (SD) IVI-C score 65.67 (16.91), median (IQR) PedsQL™ family impact score 75 (56.94-88.19), parent report 71.74 (51.98-88.5), self-report 76.09 (61.96-89.13). Psychosocial PedsQL™ subscores are lower than physical subscores. Parent-completed tools (PedsQL™ family and parent report) state greater impact on HR-QoL than tools completed by children/young people, particularly in teenagers. Older children/young people have higher functional visual ability scores than younger children.

Conclusions: Cataract has a marked and long-term impact on functional visual ability and quality of life of children and young people, with HR-QoL affected to degrees reported in children with severe congenital cardiac defects or liver transplants.

66 **Main Text**

67 **Introduction**

68 With an incidence of 1-3 per 10,000 live births, congenital/infantile cataract is a rare, but
69 sight-threatening condition [1]. Congenital/infantile cataract poses significant management
70 challenges. The risk of developing glaucoma is high, with 31% of children either having
71 glaucoma or being monitored as "glaucoma suspect" at five years after cataract surgery [2],
72 and an overall annual incidence of postoperative glaucoma of 5.25 per 100 cataract
73 operations [3 4]. Visual outcomes are often disappointing. Younger age at surgery is associated
74 with better visual outcomes [5, 6]. Unilateral cases have a worse visual prognosis in the
75 affected eye than bilateral: at six years after surgery, a median visual acuity 0.48 in bilateral
76 and 1.0 logMAR (affected eye) in unilateral cases has been reported [3]. Amblyopia and its
77 management with hours of occlusion or blurring of the better seeing eye may affect the
78 child's motor co-ordination, functioning and emotional state [7, 8].

79 Children with cataract require frequent hospital attendances, which means that children
80 miss school and parents lose time from work. If glaucoma develops, additional treatment in
81 the form of eye drops (95%)[2] or glaucoma surgery (up to 40% at 5 years of follow-up)[2] is
82 required, carrying an additional burden of anxiety for the child and the family. Yet little is
83 known about the impact of cataract and its management on children and their families.
84
85 Studies using parental proxy VR-QoL-tools such as the Children's Visual Function
86 Questionnaire found that family impact and treatment difficulty scores are worse in families
87 of children with unilateral compared with those with bilateral cataracts, and in those
88 treated with an aphakic contact lens compared with those treated with an intraocular lens
89 implant [9]. Bilateral congenital/infantile cataract is associated with reduced VR-QoL scores

compared to healthy controls [10]. A study in China using a specifically developed self-report tool showed a correlation of QoL scores with visual acuity, surgery and density of cataract [11] [12].

Over recent years, self-report instruments have been developed to measure health-related quality of life (HR-QoL) in children with various conditions, and vision-related quality of life (VR-QoL) and functional visual ability (FVA) in children with sight impairment. For example, the Impact of Vision Impairment for Children (IVI-C) instrument [13] can be used to assess the impact of visual impairment on VR-QoL, and the Cardiff Visual Ability Questionnaire for Children (CVAQC) [14] and the Functional vision questionnaire for children and young people with visual impairment [15], to assess the impact on activities of daily living (functional visual ability).

Whilst none of these has been used specifically in children with cataract, previous studies in this population have applied a generic HR-QoL tool for children and families, the PedsQL™ Inventory (www.pedsql.org) [16] [17]. The PedsQL™ covers different domains (physical and psychosocial - emotional, social, school) and contains age-specific versions as well as separate questionnaires for parents/ carers, one as a report about their child, and another to evaluate the impact of the child's condition on the family. Using the PedsQL™, it has been reported that six years after treatment for congenital/infantile cataract, parents and children report lower HR-QoL scores than normative controls [18].

However, none of the previous studies included children/young people with secondary cataract, and none offers data on long-term impact of cataract treatment in early childhood.

The main objective of the present work was to explore FVA, VR-QoL and HR-QoL in children treated or monitored for developmental and secondary cataract and their parents/carers, using CVAQC, IVI-C and PedsQL™. We included young people up to the age of 16 years to explore long-term impact of childhood cataract and its management.

Materials and methods

This work is part of a larger cross-sectional observational study, approved by the National Research Ethics Committee South Central – Oxford A (14/SC/1052). Between 25/6/2014 and 03/06/2015 we enrolled children and young people age 2-16 years who attended clinics at Moorfields Eye Hospital, London, UK. In the present study, we included children with congenital/infantile and cataract secondary to uveitis. At enrolment, exclusion criteria were: visually not significant cataract, inability to communicate in English, surgical intervention (incisional or laser) within one month of date of completing questionnaires (before or after). We enrolled consecutive patients, noting reasons for not wanting to take part. We gave parents/carers and children age-appropriate information material about the study and addressed any questions. Parents/carers gave written consent, and children could sign an assent form.

We recorded age at study participation, gender and ethnic background. From the medical notes, we recorded ocular and systemic diagnoses, age at diagnosis of the eye condition and best corrected visual acuity (BCVA) with both eyes open in logMAR on the day of study participation. Where visual acuity was recorded as “counting fingers”, we noted a BCVA of 1.3 logMAR, for “hand movements only” we noted 1.6 logMAR, for “perception of light” 2.7 logMAR, and for “no perception of light” or “ocular prosthesis/artificial eye”, 3 logMAR [19].

We also recorded details of previous and current treatment, such as number of previous surgical interventions (sum of interventions right and left eye, including incisional surgery, laser treatment, removal of sutures, injections, excluding examinations under anaesthesia), number of general anaesthetics including examinations under anaesthesia and number of current topical medications (sum of eyedrop applications per day right and left eye).

Main outcome measures

Functional visual ability was assessed with the CVAQC [14]. The 25 questions with answers selected on a four-point scale were completed by children from the age of 5 and covered areas such as education, near and distance vision, getting around, social interaction, entertainment and sports. We transformed the raw CVAQC scores into logarithmic scores [14]. The resulting scores range from -3.00 (normal visual ability) to +2.80 (severe visual impairment).

To evaluate VR-QoL, children aged 8 years or older completed the IVI-C questionnaire regarding mobility, interaction, school and emotional state. Due to delays in agreements and permissions we started using the IVI-C [13] five weeks after the start of the study (01/08/2014). We scored responses as recommended by the developers, allocating values between 0 and 4 to the responses from “never” to “always”. We did not allocate a score when the response “no, for other reasons” was selected. As the tool comprises 24 items, the resulting raw scores range from 0 to 96, with 96 indicating normal VR-QoL.

To assess HR-QoL age specific versions of the PedsQL™ Inventory were completed by children between the ages of 5-16 years and their parents/carers. The questionnaire for

children age 8-12 years consists of 23 questions in the domains of “my health and activities”, “my feelings”, “how I get along with others” and “about school”; answers are given on a 5-point Likert scale from 0 (never a problem) to 4 (always a problem). The set for young children (5-7) and teenagers aged 13-18 years comprises of 23 questions about physical, emotional, social, and school functioning; answers are on a 5-point Likert scale. We asked parents to complete two questionnaires, the PedsQL™ Parental report about the child/young person, and the PedsQL™ Family report, which measures the impact of a child’s condition on the family. The parental report on: children aged 5 years and older; children aged 8-12 years and that for teenagers aged 13-18 years consisted of 23 questions with answers on a 5-point Likert scale The parental report on children age 2-4 years consisted of 21 questions. The family impact questionnaire includes “physical”, “emotional”, “social”, “cognitive” functioning of the parent, “communication”, “worry”, “daily activities”, and “family relationships”. This questionnaire contains 36 questions with answers on a 5-point Likert scale. Parents and children from the age of 5 can self-administer the questionnaire (PedsQL™ administration guidelines). We calculated the PedsQL™ scores as detailed in the scoring instructions. If items were left blank, we adjusted the denominator, using the number of completed items instead of the number of total items. It is recommended to remove questionnaires from the analysis if 50% or more of the items have been left blank; this did not occur in our sample. In summary, self, parent and family overall total, physical and psychosocial HR-QoL scores are generated with PedsQL™ scores ranging from 0 to 100, 100 indicating normal health-related quality of life.

All questionnaires were self-administered and completed on the same day, during a regular

clinic appointment. When children needed help completing the questionnaires, they were assisted by a member of the research team or play leaders, but not by family members.

Statistics

We aimed for an overall sample size of between 50-100 children with cataract, as recommended for Bland Altman limits of agreement analysis. Demographic and clinical data and the CVAQC, IVI-C and PedsQL^{TM} scores were transferred to a dedicated database in Microsoft Office Excel by a member of the research team. Calculation of scores and data transfer were double-checked by a second member of the team.

Analysis was carried out in Microsoft Office Excel, SPSS v23 (IBM) and Stata (V14). Where data were missing, datasets were excluded from the relevant analyses. We applied descriptive statistics throughout, reporting means and standard deviations or median and interquartile range (IQR) as appropriate.

Where data were missing for individual items in the PedsQL^{TM} and IVI-C, we adjusted the denominator accordingly. The conversion from raw to logarithmic CVAQC scores takes into account missing data [14].

We assessed relationships between age and quality of life scores using Spearman rank correlation and assessed whether differences observed between age groups were statistically significant using the Rank Sum test or independent t-test. Agreement between adult and child PedsQL scores was assessed using Bland-Altman techniques.

Statistical significance was set at the 5 % level and all tests conducted were two-tailed.

Results

Enrolment

We screened approximately 3,800 sets of medical notes to identify children eligible to take part in the wider study. We approached 214 children and their families; 13 declined taking part, all because of a perceived lack of time to complete the questionnaires. We enrolled 201 children and young people (Fig 1). As this report focuses on childhood cataract (ChC), we excluded 124 datasets of children who did not have cataract or a history of cataract surgery. We removed two datasets as children had undergone incisional surgery or laser treatment within four weeks of study participation and had been enrolled in error. In addition, we removed two datasets of children with traumatic cataract, which was not included in our inclusion criteria, and one dataset, as neither parents nor child completed the questionnaires after having given consent. The statistical analysis was carried out on the remaining 72 datasets (Fig 1).

Missing data

The proportion of missing data was low. No data were missing for age, gender, diagnoses, laterality, BCVA and number of daily eye drops. Ethnicity was unknown in 8 participants (11.11%). Age at diagnosis of the eye condition could not be determined exactly in 3 children (4.17%). Two children had previous surgical interventions at other centres, and information about previous number of operations and general anaesthetics was incomplete (2.78%). For all questionnaires administered, response and completion rates were high. The response rate for the PedsQL^{TM} family report was 95.83%, for the parent report 97.22%, for the self-report 90.74% (Supplementary Material). The proportions of fully completed

questionnaires were 88.41, 90.00 and 97.96%, respectively. CVAQC and IVI-C response rates were 90.74% and 77.78%, respectively. CVAQC and IVI-C scores both contain a “for other reasons” category; selection of this category is taken into account during calculation of the scores.

Participants

The mean (SD) age of participants was 8.45 (standard deviation SD 4.10) years. Eighteen children were 2-4 years old, 16 were 5-7 years old, 25 were 8-12 years old and 13 were 13-16 years old (Table 1). Thirty-eight participants (52.78%) were female. Sixty-eight percent of participants were White, 15.28% Asian or Asian British, 4.17% Black or Black British, 1.39% other; ethnicity was unknown in 11.11%.

Clinical details

Sixty-nine participants (95.83%) had congenital/infantile cataract, of these, 31 (43.06%) had developed glaucoma after cataract surgery. Eight also had microphthalmia; another had microphthalmia, cataract surgery and secondary glaucoma. Two had cataract associated with either aniridia or primary congenital glaucoma. In total, 37 children and young people (51.39%) had glaucoma.

Lens status

Over a third of participants were aphakic in both eyes (n=28, 38.89%) (Table 1). Sixteen (22.22%) had aphakia in one eye, whilst the other eye had not undergone surgery. In one case, one eye was aphakic, the other replaced by an ocular prosthesis. Seven participants (9.72%) had lens implants in both eyes, and another seven had not undergone surgery.

The condition was bilateral in 43 cases (59.72%), and the mean age (SD) at diagnosis was 1.25 years (SD 2.84) (Table 1). Median best corrected visual acuity in the better seeing eye or with both eyes open was 0.18 logMAR (IQR 0.02-0.43). Children applied a median of 0 (0-2.25) eye drops each day, with a maximum of 16 daily drops. The median number of eye operations (incisional or laser) performed on both eyes combined was 3 (2-5, maximum 35), and the median number of general anaesthetics including examinations under anaesthesia the children/young people had undergone was 3 (2-5.75, maximum 26).

Functional visual ability

Forty-nine of 54 children and young people age 5-16 years completed the CVAQC (90.74%). The median score was -1.42 (IQR -2.28 to -0.03), indicating moderate impairment of FVA (Table 2). Scores were better in older children than in the younger age group: -1.91 (-2.30 to-1.36) in 13-16 year olds, -1.42 (-2.37 to 0.06) in 8-12 year olds, and -0.36 (-1.94 to 0.37) in 5-7 year olds (-3.00 indicating normal visual ability) (Figure 2a). There was evidence of an association between age and CVAQC score (Spearman's rho correlation coefficient SRCC -0.291, P = 0.04).

Vision-related quality of life

Twenty-one of 27 eligible children and young people age 8-16 years completed the IVI-C (77.78%). The mean score was 65.67 (SD 16.91). The mean score in the older age group (73.5, SD 9.26, in 13-16 year olds) was higher than that in the younger age group (63.82, SD 17.96, in 8-12 year olds), with 96 indicating normal VR-QoL (Figure 2b). The observed difference was not statistically significant in these data however there were just four subjects in the older age group and thus power to detect such a difference was limited.

Health-related quality of life

The median of the overall self-report completed by children/young people (n=49/54, 90.74%) was 76.09 (61.96-89.13) (Figure 2c), the median of the parent report about the child (n=70, 97.22%) was 71.74 (51.98-88.19) and the median score of the family report completed by 69/72 parents/carers (response rate 95.93%, supplementary material) was 75 (56.94-88.19, Table 2 with 100 indicating normal HR QoL. Figure 2d shows a Bland-Altman plot assessing agreement between parent and child PedsQL^{TM} scores. There is evidence of decreasing dispersion – for individuals with lower scores, there is larger disagreement between child and adult score than for individuals with higher scores. The average disagreement (ie estimated bias) between adult and child score was -4.06 (-7.94, -0.18), indicating significant evidence that parents scores are lower than their child's.

The overall self-report scores were slightly higher in the older age groups than the younger ones: median (IQR) 66.30 (57.07-73.91) in 5-7 year olds, 81.52 (65.76-96.2) in 8-12 year olds, and 80.98 (68.75-91.85) in 13-16 year olds (Table 2) and the Family impact PedsQL^{TM} scores tended to be higher in parents of children age 5-12 years than in very young children and teenagers (Table 2).

The median "physical wellbeing" PedsQL^{TM} subscores were 82.81 (64.84-94.53) in the self-report, 81.25 (50-93.75) in the parent report and 79.17 (58.33-100) in the family report. The median "psychosocial wellbeing" subscores were 73.33 (60-85.95) in the self-report, 68.33 (53.33-85.71) in the parent report and 73.33 (54.79-85.63) in the family report. Psychosocial scores appeared more affected than physical scores. Figures 2e and 2f

304 are Bland-Altman plots for physical and psychosocial scores. There was no evidence of
305 systematic bias for physical scores, but parents tended to give lower scores than their
306 children on the psychosocial score with an estimated difference of -4.19 (-8.12, -0.26).
307
308 **Associations**
309 Our study was not powered to detect statistically significant associations. However, older
310 age at study participation tended to be associated with better CVAQC and PedsQL™ self-
311 report scores. Bilaterality of cataract tended to be associated with worse PedsQL™ self-
312 report and parent scores and poorer BCVA tended to be associated with worse a lower
313 CVAQC score, IVI-C score, PedsQL™ self-report, parent and family scores.
314
315
316 **Discussion**
317 **Key results**
318 The principal aim of this study was to explore the effects of childhood cataract on functional
319 visual ability (FVA), vision-related quality of life (VR-QoL) and general health-related quality
320 of life (HR-QoL) as reported by children/young people and their parents/carers. Due to its
321 inclusive design, involving children of a broad age range and their parents, this study
322 delivers new insights into the impact of childhood cataract on families.
323
324 Childhood cataract is not only associated with sight impairment, but is often complicated by
325 a need for multiple additional operations and anaesthetics and the need for daily eye drops,
326 most commonly for secondary glaucoma. The effect on FVA, VR-QoL and HR-QoL is
327 profound. Children themselves report reduced levels of FVA and VR-QoL; this is most

pronounced in younger children. Health-related quality of life (HR-QoL), reported by the young person themselves and by parents on behalf of their child, is significantly reduced, as is the HR-QoL experienced by the family. Children and young people of all age groups report a greater impact on psychosocial than physical well-being.

Limitations

Our study design carries a number of potential sources of bias. Firstly, enrolling children attending a single site may induce selection bias. We attempted to reduced further bias by approaching consecutive patients; only a small proportion (n=13/224, 5.8%) declined to take part. However, families may have stopped attending clinics due to dissatisfaction with the services, or unwillingness or inability to comply with intense treatment regimes. On the other hand, families of children with good visual outcome may equally stop attending and be lost to follow-up. We have no data to estimate the proportion of families who stop attending, but from clinical experience consider the overwhelming majority of parents to be eager to provide the best possible healthcare for their child. Lack of a control group of healthy children may be considered a limitation. However, CVAQC and IVI-C were specifically developed for children with sight impairment; this would lead to a ceiling effect if used in healthy children, and comparison with healthy children would be misleading. For the PedsQL™, normative data are available from large numbers of healthy children, and a control group is not required**.** We limited inclusion to families able to communicate in English, which may induce selection bias. However, during enrolment we did not encounter any family who could not communicate in English.

It would be interesting to explore a possible association between the number of surgical interventions and QoL. Previous studies have used the number of surgical procedures as a

proxy of painful episodes the child had encountered as part of their eye treatment [20]. Similarly, the number of general anaesthetics (including EUAs, as these are often arranged on the understanding that should findings indicate a need for additional surgery, this will be carried out under the same anaesthetic) has been used as a proxy for episodes of emotional upset and anxiety [20]. However, our study was not powered to explore associations between QoL and putative risks factors for a reduction in QoL. As we were mindful of the need to avoid multiple significance tests and the potential for misinterpretation of non-statistically significant findings, we focused our analysis on the main objectives of the study.

Whilst logMAR visual acuity is a well-established measure of visual function, it is not always possible to use logMAR methods in children with sight impairment, and “hand movements” or “counting fingers” at a specified testing distance are still occasionally used. Complete blindness, “no perception of light”, or “artificial eye/ocular prosthesis” can also not be expressed in logMAR. In order to allow a quantitative analysis, we followed the approach of using logMAR values of of 1.3 to 3 in these cases [19]. This may have led to an under- or overestimate of logMAR acuity in some cases.

The heterogeneity of our study population, including children with secondary cataract and secondary glaucoma, may be considered problematic, but it allowed us to gather the views of older children and young people, which no previous study had explored. Another strength of our study is that children/young people completed the questionnaires by themselves, or were supported by play specialists if necessary. This eliminated parental perceptions influencing the children’s answers, though it may not have fully eliminated an adult’s bias from children’s answers.

Interpretation

The reduction in HR-QoL in children with cataract we report here is comparable to levels reported by children with severe congenital heart defects or liver transplants [21 22]; psychosocial subscores are reduced to levels comparable with children undergoing treatment for acute lymphoblastic leukaemia [23]. A previous study exploring HR-QoL in children who had undergone surgery for congenital cataract and their parents also reported reduced levels [18]; the scores we observed in the equivalent age group are even lower. In addition, the present study extends the finding of reduced HR-QoL to children who have not undergone surgery and children with secondary cataract.

A novel finding is that family and parental HR-QoL scores are higher in children age 5-12 compared with younger children and teenagers. The cause for this is unclear; possibly child and family initially adjust, but in teenagers expectations and frustrations about education and transition to independence may increase. Alternatively, as better QoL is associated with better BCVA, the observed improvement in vision over time may explain increased levels in HR-QoL.

Two of the tools we used, the CVAQC and the IVI-C, have not previously been used in children with cataract, so direct comparison with other studies is not possible.

Generalisability

Within the limits of the study design, i.e. selection bias which may have led to inclusion of a higher proportion of more treatment-adherent families and the limitation of enrolling

399 participants at a single site in one highly developed country, our findings can be generalised
400 to other children with cataract who receive care in similar settings.
401
402 **Conclusions**
403 Whilst treatment for cataract in adults is a highly successful sight-restoring procedure,
404 cataract in children can have a dramatic effect on the life of affected children/young people
405 and their families. More research is needed to evaluate the impact of multiple interventions
406 and lifelong hospital follow-up. Families and young people may benefit from support to
407 address psychosocial problems and difficulties with children's activities of daily living.
408
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417 the NIHR or the Department of Health.
418
419 **Conflicts of interests**
420 None of the authors has interests to declare with regards to this publication.
421
422

What was known before:

- Only five previous studies have investigated QoL in children with cataract, all including congenital cataract patients only. The applied tools varied between studies. In children with congenital/infantile cataract:
- Health related quality of life scores are reduced to levels similar to severe systemic chronic diseases.
- Parent- and children's self-report scores on health-related quality of life differ.
- Low visual acuity is associated with lower QoL scores, and surgery improves visual acuity and QoL.

What this study adds

- Paediatric cataracts can have a profound impact on health- and vision-related quality of life, and on functional visual ability, even many years after primary surgery.

Amongst children who have undergone cataract surgery

- Functional visual ability and vision-related quality of life scores are higher in teenagers than in younger children.
- Children/young people report higher health-related quality of life scores than their parents.
- Self-reported physical and psychosocial PedsQL^{TM} subscores are higher in older children.

References

1. Foster A, Gilbert C. Cataract in children. *Acta Paediatr* 2003; **92**(12)**:** 1376-1378.

2. Freedman SF, Lynn MJ, Beck AD, Bothun ED, Orge FH, Lambert SR *et al.* Glaucoma-Related Adverse Events in the First 5 Years After Unilateral Cataract Removal in the Infant Aphakia Treatment Study. *JAMA Ophthalmol* 2015; **133**(8)**:** 907-914.

3. Chak M, Rahi JS, British Congenital Cataract Interest G. Incidence of and factors associated with glaucoma after surgery for congenital cataract: findings from the British Congenital Cataract Study. *Ophthalmology* 2008; **115**(6)**:** 1013-1018 e1012.

4. Plager DA, Lynn MJ, Buckley EG, Wilson ME, Lambert SR, Infant Aphakia Treatment Study G. Complications in the first 5 years following cataract surgery in infants with and without intraocular lens implantation in the Infant Aphakia Treatment Study. *Am J Ophthalmol* 2014; **158**(5)**:** 892-898.

5. Hartmann EE, Lynn MJ, Lambert SR, Infant Aphakia Treatment Study G. Baseline characteristics of the infant aphakia treatment study population: predicting recognition acuity at 4.5 years of age. *Investigative ophthalmology & visual science* 2015; **56**(1)**:** 388-395.

6. Hartmann EE, Stout AU, Lynn MJ, Yen KG, Kruger SJ, Lambert SR *et al.* Stereopsis results at 4.5 years of age in the infant aphakia treatment study. *Am J Ophthalmol* 2015; **159**(1)**:** 64-70 e61-62.

7. Webber AL, Wood JM, Gole GA, Brown B. The effect of amblyopia on fine motor skills in children. *Investigative ophthalmology & visual science* 2008; **49**(2)**:** 594-603.

8. Webber AL, Wood JM, Gole GA, Brown B. Effect of amblyopia on self-esteem in children. *Optom Vis Sci* 2008; **85**(11)**:** 1074-1081.

9. Birch EE, Cheng CS, Felius J. Validity and reliability of the Children's Visual Function Questionnaire (CVFQ). *J Aapos* 2007; **11**(5)**:** 473-479.

10. Lopes M, Salomão S, Berezovsky A, Tartarella M. Assessing vision-related quality of life in children with bilateral congenital cataracts. *Arq Bras Oftalmol* 2009; **72**(4)**:** 467-480.

11. Chen W, Ye H, Deng D. Development and evaluation of the scale of quality of life for children with bilateral congenital cataract. *Zhonghua Yan Ke Za Zhi* 2007; **43**(3)**:** 239-244.

12. Ye H, Chen W, Deng D, Lin Z, Yang W. Quality of life assessment in children with congenital bilateral cataract. *Zhonghua Yan Ke Za Zhi* 2007; **43**(11)**:** 996-999.

13. Cochrane GM, Marella M, Keeffe JE, Lamoureux EL. The Impact of Vision Impairment for Children (IVI_C): validation of a vision-specific pediatric quality-of-life questionnaire using Rasch analysis. *Investigative ophthalmology & visual science* 2011; **52**(3)**:** 1632-1640.

14. Khadka J, Ryan B, Margrain TH, Court H, Woodhouse JM. Development of the 25-item Cardiff Visual Ability Questionnaire for Children (CVAQC). *Br J Ophthalmol* 2010; **94**(6)**:** 730-735.

15. Tadic V, Cooper A, Cumberland P, Lewando-Hundt G, Rahi JS, Vision-related Quality of Life G. Development of the functional vision questionnaire for children and young people with visual impairment: the FVQ_CYP. *Ophthalmology* 2013; **120**(12)**:** 2725-2732.

16. Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. *Med Care* 2001; **39**(8)**:** 800-812.

17. Varni JW, Seid M, Knight TS, Uzark K, Szer IS. The PedsQL 4.0 Generic Core Scales: sensitivity, responsiveness, and impact on clinical decision-making. *J Behav Med* 2002; **25**(2)**:** 175-193.

514 18. Chak M, Rahi J. British Congenital Cataract Interest Group. The health-related quality
515 of life of children with congenital cataract: findings of the British Congenital Cataract
516 Study. *Br J Ophthalmol* 2007; **91**(7)**:** 922-926.
517
518 19. Schulze-Bonsel K, Feltgen N, Burau H, Hansen L, Bach M. Visual acuities "hand motion"
519 and "counting fingers" can be quantified with the freiburg visual acuity test.
520 *Investigative ophthalmology & visual science* 2006; **47**(3)**:** 1236-1240.
521
522 20. Freedman B, Jones S, Lin A, Stinnett S, Muir K. Vision-Related quality of life in children
523 with glaucoma. *J AAPOS* 2014; **18**(1)**:** 95-98.
524
525 21. Knowles RL, Day T, Wade A, Bull C, Wren C, Dezateux C *et al.* Patient-reported quality
526 of life outcomes for children with serious congenital heart defects. *Arch Dis Child* 2014;
527 **99**(5)**:** 413-419.
528
529 22. Limbers CA, Neighbors K, Martz K, Bucuvalas JC, Webb T, Varni JW *et al.* Health-related
530 quality of life in pediatric liver transplant recipients compared with other chronic
531 disease groups. *Pediatr Transplant* 2011; **15**(3)**:** 245-253.
532
533 23. Sung L, Yanofsky R, Klaassen RJ, Dix D, Pritchard S, Winick N *et al.* Quality of life during
534 active treatment for pediatric acute lymphoblastic leukemia. *Int J Cancer* 2011; **128**(5)**:**
535 1213-1220.
536
537

Figure legends

Fig. 1. Enrolment, intervention and analysis flowchart (modified from CONSORT, www.consort-statement.org).

Fig. 2. Top: Box plots of median and interquartile range (IQR) Cardiff Visual Ability for Children (a), Impact of Vision Impairment for Children (b) and PedsQL™ self-report scores (c) of children with cataract. There is a trend towards less impairment with increasing age (decreasing scores on CVAQC, increasing scores on IVI-C and PedsQL™), but with large overlap of the IQR.
Bottom: Bland Altman plots of agreement between parent and child PedsQL scores; x axis: mean of parent and child scores; y axis: difference of parent and child scores. Parents report lower overall scores than their children; the lower the scores, the larger the disagreement (d). The plot for physical and psychosocial PedsQL subscores shows no systematic bias (e), but parents tended to report lower scores than their children on the psychosocial scale (f).

Table legends

Table 1. Demographic and clinical characteristics, associated conditions and lens status in study participants.

Table 2. Scores for functional visual ability (FVA), vision- and health-related quality of life (VR-QoL, HR-QoL) reported by parents/carers and children. Possible CVAQC scores (FVA) extend from -3.00 (normal visual ability) to +2.80 (severe visual impairment). Possibe IVI-C scores are between 0 and 96 (96 = normal VR-QoL); participants in this study reported markedly reduced VR-QoL. Possible PedsQLTM scores range from 0 to 100 (100 = normal HR-QoL); in this study, scores were severely reduced in all versions and subscales of the instrument (parent report, family report, self-report, physical and psychosocial subscores).

Supplementary Material: Table “Response and Completion Rates”. Parents were asked to completed two questionnaires, and children from the age of 5 years were asked to complete two or three questionnaires. Response and completion rates were high.