EXTENDED REPORT

The health-related quality of life of children with congenital cataract: findings of the British Congenital Cataract Study

Melanie Chak, Jugnoo Sangeeta Rahi, on behalf of the British Congenital Cataract Interest Group*

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See end of article for authors' affiliations

Correspondence to:
Dr J S Rahi, Centre for
Paediatric Epidemiology,
Institute of Child Health, 30
Guilford Street, UCL, London
WC1N 1EH, UK;
j.rahi@ich.ucl.ac.uk

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Background: There is increasing emphasis on patient-based assessment of healthcare outcomes, but evaluation of the impact of ophthalmic disorders on quality of life is undertaken infrequently.

Aim: To report on the health-related quality of life (HRQOL) of children with congenital cataract.

Methods: At least 6 years after diagnosis, 41 children (representative of a national cohort) and their parents completed the child and proxy (parental) versions of PedsQL 4.0, a generic multidimensional paediatric HRQOL instrument, independently. This provided a physical health summary score, a psychosocial health summary score and an overall score (range 0–100, higher scores indicating better HRQOL).

Results: Questionnaires were completed by 33 parent–child pairs and by 8 parents alone. The mean (SD) total score self-reported by children was 75.85 (15.56) and that reported by parents was 75.91 (16.79). The mean (SD) physical health score self-reported by children was 80.76 (8.61), which was higher than the psychosocial health score of 72.93 (16.06). There was considerable variation in agreement of scores reported by individual child–parent pairs (the largest difference was 56.3 points). Notably, the PedsQL scores of children with congenital cataract were comparable with those reported for some children with severe systemic diseases such as rheumatological disease and some cancers.

Conclusions: These findings serve to characterise the significant broader impact of congenital cataract on patients and their families. The PedsQL 4.0 proved to be an acceptable instrument for assessing generic HRQOL, permitting valuable reporting by children and their parents. However, to capture the subjective experience of ophthalmic disease in childhood fully, specific vision-related quality of life measures for children are needed.

Visual loss influences the general health, development and welfare of affected children. Children with congenital cataract might undergo numerous procedures, experience demanding amblyopia treatment and undertake multiple, frequent hospital visits over a long period of time, during which there can be uncertainty about visual prognosis. Objective measures alone, such as visual acuity, are recognised to be inadequate descriptors of functioning, and even less predictive of health-related quality of life (HRQOL). Thus, patient-based assessment of the impact of disease is now widely advocated.¹ However, there is presently no validated instrument for assessing specifically the vision-related quality of life (VRQOL) of children, although pilot work has been reported.² Thus, we have investigated the general HRQOL of a representative group of children with congenital/infantile cataract to provide further insight into the broader impact of this disease.

METHODS

All children newly diagnosed with congenital/infantile cataract in the UK over a 12-month period during 1995–6 (the British Congenital/Infantile Cataract Study)^{3–7} were eligible for the present study. We have reported previously^{3–7} the methodological details regarding original recruitment. In 2002–3, the cohort was traced through the managing ophthalmologists and information collected using questionnaires.

For validation of data, children attending the six main ophthalmic units contributing to the overall study were independently examined by the principal investigator. This direct contact allowed recruitment to the present study, after detailed individual discussions and after having received written informed consent. In all, 42 (17.8% of the total cohort) pairs of children and their parents were invited to participate.

Wherever possible questionnaires were completed at the hospital, but where space or time did not permit this they were completed at home and returned by post, with reminders as required.

As our primary aim was to assess the general HRQOL of children with cataract and compare this with the HRQOL in other diseases, we used the PedsQL 4.0 instrument.⁸ It comprises an eight-item physical health (physical functioning) and a 15-item psychosocial health (school, social and emotional functioning) questionnaire. Items are rated on a Likert scale from 0 (never a problem) to 4 (almost always a problem). Responses are transformed to a score ranging from 0 to 100, with higher scores representing a better quality of life (QOL).⁸ Thus, physical, psychosocial and total scores are generated. Although the questionnaire lacks questions specific to vision, its domains include those in which vision is important—for example, education. There are four age-appropriate versions of the instrument for self-completion by children, and a version for completion by proxy respondents, usually parents, reporting on their assessment of the affected child. The questionnaire is sensitive to cognitive development, and has excellent internal consistency and clinical validity.^{9–11} Minimum training or expertise is required, and it is simple and fast to complete.^{9–12}

Statistical analysis

Self- and proxy-reported physical, psychosocial and total HRQOL scores were generated, and descriptive analyses undertaken. Internal consistency was evaluated using Cronbach's α coefficient.¹³ Agreement between child and parental scores was assessed using the Bland–Altman¹⁴ measure.

Abbreviations: HRQOL, health-related quality of life; QOL, quality of life; VRQOL, vision-related quality of life

Table 1 Distribution of health-related quality of life (PedsQL 4.0) scores and reliability indices

	Physical health score	Psychosocial health score	Overall HRQOL score
Mean (SD): child-reported	80.76 (18.61)	72.93 (16.06)	75.85 (15.56)
Mean (SD): parent-reported	80.22 (22.01)	73.52 (16.44)	75.91 (16.79)
Ceiling effects: child-reported	0%	0%	0%
Ceiling effects: parent-reported	0%	0%	0%
Floor effects: child-reported	0%	0%	0%
Floor effects parent: reported	24.5%	5.0%	0%
Cronbach's α correlation coefficient: child-reported	0.88	0.90	0.93
Cronbach's α correlation coefficient: parent-reported	0.81	0.84	0.90

HRQOL, health-related quality of life.

RESULTS

Out of 42 invited parent–child pairs, 41 participated: in 33 (80%) pairs, both child and parent questionnaires were completed, and most (28 pairs) completed these at home. The median (range) age of children was 6.8 years (5.7–19.51), 6 (19%) children had unilateral disease, 24 (59%) were girls and 13 (32%) had additional disorders. Visual acuity (best eye) ranged from 6/4 to 6/9, median 6/6 in children with unilateral cataract, and from 6/4 to 6/72, median 6/12 in children with bilateral cataract. In all, 34 (83%) children had undergone surgery.

In all, >99%, the questionnaires were fully completed. The eight children who failed to complete the questionnaire at all tended to have poorer visual acuity and/or other medical conditions.

The internal consistency exceeded the minimum reliability standard of 0.70 for group comparisons,¹³ and exceeded or approached the minimum reliability standard of 0.90 for individual comparisons¹³ (table 1). The ceiling and floor effects (table 1) are the proportion of children scoring the maximum (100) and minimum (0) possible scores, respectively. A large proportion of floor or ceiling effects reflects the inability of the instrument to measure at the lowest or highest levels, respectively, of that domain.

The distribution of scores was normal: the mean and standard deviations of summary scores are shown in table 1. The mean psychosocial score was lower than the mean physical score.

The lowest individual physical, psychosocial and total scores, respectively, self-reported by children were 9.3, 33.3 and 25, and those reported by parents were 21.9, 38.3 and 32. The

distribution of individual child–parent pair overall summary scores is shown in fig 1.

We could find no correlations between the mean scores and age, sex, best acuity, laterality, presence of other disorders, surgery, strabismus, postoperative complications, glaucoma, posterior capsular opacity or amblyopia treatment (data not shown). However, the size of the sample is likely to have limited our ability to find true associations had they existed.

Notably, the psychosocial PedsQL scores were comparable to those reported for some children with severe systemic diseases^{15 16} (table 3).

The mean child–parent difference for each summary score was small (up to –2.0, table 2).

However, the range of child–parental pair disagreement for each scale was wide (maximum of 56.3 points), as shown in figs 2–4.

DISCUSSION

The PedsQL 4.0 proved to be an acceptable instrument for assessing the general HRQOL of children with cataract. Its use indicated that HRQOL of children with congenital cataract is variable but is comparable to that of children with some severe, systemic chronic disorders. Furthermore, there is a wide range of disagreement between children and their parents.

Our study has certain limitations. As most questionnaires were completed at home, it is impossible to know whether these were completed independently, as requested. If not, the effect would be to artificially reduce the level of child–parent disparity: despite this, disagreement was identified. In all, 8 (20%) children did not complete the questionnaire at all; incomplete participation is likely to pertain in any population of

Figure 1 Distribution of individual child–parent pairs: total scores of children for (A) bilateral cataract and (B) unilateral cataract. NK, not known.

Table 2 Agreement (Bland-Altman measure)¹⁴ between child-parent pairs on physical, psychosocial and total scale scores

	Min	Max	Mean	Lower limit of agreement	Upper limit of agreement
Difference in physical score (child-parent)	-43.8	56.3	-1.6	-44.7 (-31.2 to 58.3)	41.6 (28.0 to 55.1)
Difference in psychosocial score (child-parent)	-50.6	23.3	-2.0	-35.8 (-25.2 to 46.4)	31.8 (21.2 to 42.4)
Difference in total score (child-parent)	-46.2	32.2	-1.3	-36.7 (-25.8 to 47.6)	34.1 (23.2 to 45.0)

Max, maximum; Min, minimum.

children with eye disease which includes those with poor vision in both eyes or with cognitive impairments, or where parents have concerns either about their child self-reporting in itself or about their ability to do so effectively. Participation by children was higher when completion of HRQOL instruments is supervised,¹⁵ and this would be a better approach in future studies, rather than allowing postal response. Finally, although representative of the cohort, the sample studied was relatively small. This could have limited the ability to investigate associations of HRQOL with clinical ophthalmological factors which would be of interest. However, our primary aim was to

evaluate the general HRQOL of children with cataract and see how it differed from those with other diseases.

The functional vision status of children with ophthalmic disorders¹⁷ and the psychosocial impact of specific treatment (especially occlusion) on children and their families¹⁸⁻²⁰ have been studied. However, there has been very limited work on the distinct, although related, issue of subjective assessment of HRQOL. Thus, at present, there are no studies on the general HRQOL of children with ophthalmic disorders with which to compare our findings directly. Comparison with studies of other paediatric disorders (and using the PedsQL 4.0) suggests that children with congenital cataract experience similar HRQOL as those with severe, systemic chronic and sometimes even life-threatening diseases.^{15, 16} This important finding is unexpected, and serves to highlight the insights afforded by the use of subjective patient-based outcome measures. This finding should help inform clinicians about the broader impact of congenital cataract on their patients and their families, and be valuable to parents of newly diagnosed children. Children and parents alike reported a greater impact of congenital cataract in the psychosocial, rather than physical, domain. This might partly reflect ongoing concerns about long-term visual prognosis in a disorder in which visually impairing events can occur many years after initial treatment. This highlights the importance of psychosocial support and advice about long-term visual prognosis, even when affected children are otherwise in good health and achieve good vision.

Disagreement between children and their parents with respect to assessment of HRQOL is well recognised in paediatrics.²¹ It underlines the importance of measuring both perspectives, wherever possible, to enrich understanding of the impact on the child and the family as a whole. Self-reporting by children might not always be possible—for example, when the child is cognitively impaired, or too ill, or simply too young to participate. This is relevant to the management of many

Figure 2 Physical summary scores of child-parent pairs. **Figure 3** Psychosocial summary scores of child-parent pairs. **Figure 4** Total summary scores of child-parent pairs.

Table 3 Comparison of self-reported health-related quality of life (total, physical and psychosocial) of children with cataract and those with other systemic disorders

	Physical health mean (SD)	Psychosocial health mean (SD)	Total score mean (SD)
Congenital cataract			
Child-reported	80.76 (18.61)	72.93 (16.06)	75.85 (15.56)
Parent-reported	80.22 (22.01)	73.52 (16.44)	75.91 (16.79)
Rheumatological disorders ^{16*}			
Child-reported	68.12 (22.52)	74.23 (16.46)	72.09 (16.92)
Parent-reported	70.97 (18.49)	66.72 (24.12)	73.31 (17.62)
Paediatric cancers ^{25†}			
Child-reported	71.79 (21.80)	72.62 (16.41)	72.20 (16.38)
Parent-reported	68.75 (24.98)	70.31 (17.96)	69.70 (19.17)
Acute lymphoblastic leukaemia ¹⁵			
Child-reported	86.16 (14.72)	79.07 (13.14)	80.97 (12.57)
Parent-reported	84.73 (19.73)	73.71 (16.95)	76.50 (16.10)

*Dermatomyositis, fibromyalgia, juvenile rheumatoid arthritis, systemic lupus erythematosus, spondylarthritis.

†acute lymphocytic leukaemia, brain tumour, non-Hodgkin's lymphoma, Hodgkin's lymphoma, Wilms' tumour and other cancers.

ophthalmic disorders, where treatment needs to start very early in life, at which time it will be necessary to rely on parental reporting. The reasons for disagreement are complex and will change over time. For example, parents might have more limited experience of the impact of a disorder in certain situations, such as in school, than in others, such as at home. Equally, there will be important differences between parents and their children with respect to long-term aspirations or expectations, as well with respect to their understanding of health, the causes of illness and the benefits of treatment. In general, agreement between child and parental reports is better for physical health (observable) than for psychosocial health (less observable).⁸ Interestingly, this was not the case in our study, and it could be attributable to sighted parents (in the majority of cases) anticipating visual loss to have a greater impact on physical functioning than actually occurs in children who have always had poor sight.

Thus, eliciting disagreement should be viewed positively, contributing to a more complete picture of the child's QOL, rather than as a challenge to resolve whose view is "correct".²²

We have demonstrated the feasibility and usefulness of measuring HRQOL in children with ophthalmic disease using a generic instrument. The use of HRQOL measures in clinical practice encourages a focus on the patient rather than on the disease, alongside as increasing patient involvement in decision making and self-care and facilitating communication between patient and doctor, and between doctors in shared care situations.²³ HRQOL tools are useful in comparing the outcomes of established with emerging clinical interventions. However, we suggest that the future of assessing QOL in paediatric ophthalmology lies in the development of specific vision-related paediatric instruments which are patient-centred, and place emphasis on both the psychosocial and the physical (functional) aspects of visual loss to ensure that they evaluate QOL and not general health status.²⁴ The development of such instruments is not straightforward,²¹ and requires primary research, combining qualitative and quantitative approaches, to better understand what it means to be a visually impaired child. Such collaborative research would yield an enhanced ability to capture the subjective experiences of visually impaired children, differentiating sensitively between individuals with the same level of objectively assessed functioning, to inform better practice, policy and service provision.

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Authors' affiliations

Melanie Chak, Jugnoo Sangeeta Rahi, Centre for Paediatric Epidemiology, Institute of Child Health, UCL, London, UK

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*Appendix A provides the full list of members.

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APPENDIX A

Table A1 Members of the British Congenital Cataract Interest Group

M Abdel-Khalek	Boston	C Dodd	Manchester	J McConnell	Kent
W Acimandos	London	R Doran	Leeds	G McGinnity	Belfast
G Adams	London	G Dutton	Glasgow	B McLeod	Brighton
S Aftab	Scunthorpe	J Duval-Young	High Wycombe	A Misra	Notts
L Allen	Cambridge	C Edelsten	Ipswich	K Mohamad	Chesterfield
L A Amanat	Great Yarmouth	R Edwards	Kent	A Moore	London
S Armstrong	Chester	H El-Kasaby	Essex	B Moriarty	Cheshire
A Assaf	Milton Keynes	J Elston	Oxford	G Morrice	Stirling
NJ Astbury	Norwich	B Enoch	Barnstable	R Morris	Southampton
D Bannerjee	Wigan	A Evans	Plymouth	C Munton	Kent
A Barr	Dumferline	N Evans	Portsmouth	Mr M Neugebauer	Cheshire
L Beck	Cardiff	G Fahy	Rep of Ireland	W Newman	Liverpool
A Beckingsale	Colchester	A Fielder	London	K Nischal	London
G Bedford	Dumfries	FF Fisher	Bedford	J Nolan	Republic of Ireland
L Benjamin	Aylesbury	D Flaye	Bishop's Stortford	G O'Connor	Republic of Ireland
B Billington	Reading	B Fleck	Edinburgh	M O'Keefe	Republic of Ireland
T Blamires	Leicester	W Frank	Poole	R Ohri	London
P Bloom	London	A Gaskell	Ayr	S Perry	Kidderminster
D Boase	Portsmouth	N George	Dundee	R Phillips	Wirral
J Bolger	Hertfordshire	M Gibbens	Kent	SJ Pieris	Bedford
M Boodhoo	Chertsey	B Greaves	Kent	B Power	Dumfries
R Bowell	Dublin	P Gregory	East Sussex	N Price	Cheltenham
J Bradbury	Bradford	R Gregson	Nottingham	A Quinn	Devon
J Brazier	London	SJ Hardman	Ipswich	I Qureshi	Rochdale
A Bron	Oxford	S Haworth	Nottingham	A Rahman	Boston
D Brosnahan	Sheffield	MH Heravi	Kent	A Rennie	Aberdeen
R Brown	Shropshire	P Hodgkins	Southampton	A Ridgway	Manchester
R Brown	Stoke on Trent	R Holden	Derby	M Roper- Hall	Birmingham
S Bryan	London	R Humphry	Salisbury	E Rosen	Manchester
J Bryars	Belfast	C Hutchinson	Halifax	Russell-Eggitt II	London
R Buckley	London	J Innes	Hull	A Shun Shin	Wolverhampton
P Burgess	Swindon	I Jalili	Peterborough	A Simcock	Exeter
J Burke	Sheffield	C Jenkins	Maidstone	I Simmons	Leeds
L Butler	Birmingham	E Johnson	Gloucester	M Tappin	Surrey
D Calver	London	N Kaushik	Wrexham	R Taylor	York
A Casswell	Brighton	N Kayali	London	D Taylor	London
A Chandna	Liverpool	S Keightley	Basingstoke	V Thaller	Plymouth
W Church	Aberdeen	P Khaw	London	D Thauong	Essex
J Clarke	Middlesborough	R Kinnear	London	W Tormey	Republic of Ireland
M Clarke	Newcastle upon Tyne	S Kotta	Grimsby	S Tuft	London
M Coffey	Republic of Ireland	Y Kumar	Cornwall	M Tutton	Chester
M Cole	Devon	T Lavy	Glasgow	J Twomey	Somerset
R Condon	Chertsey	D Laws	Swansea	S Verghese	West Cumberland
P Corridan	Wolverhampton	J Leitch	Surrey	S Vickers	Brighton
M Dang	Darlington	C Liu	Brighton	A Vijaykumar	Blackburn
R Darvell	Kent	C Lloyd	Manchester	A Vivian	Bury St Edmunds
BN Das	Worcestershire	C MacEwen	Dundee	C Williams	Bristol
PD Davies	Norwich	A Macfarlane	Kent	H Willshaw	Birmingham
S Daya	West Sussex	G Mackintosh	Cheltenham	G Woodruff	Leicester
R De Cock	Margate	A Mandal	S Yorkshire	G Wright	Burnley
C Dees	Darlington	R Markham	Bristol	B Young	Republic of Ireland
				A Zaidi	S Yorkshire