CLINICAL SCIENCE

Changes in visual acuity from 4 to 12 years of age in children operated for bilateral congenital cataracts

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Aims: To investigate the long term effects of age at surgery on the development of visual acuity (VA) by measuring VA from preschool age to puberty. Furthermore, to report the VA levels at 12 years of age in a geographically based cohort of operated congenital bilateral cataracts.

Methods: All children born in four western counties of Sweden between January 1980 and December 1993 who were diagnosed with congenital cataracts were included in a longitudinal prospective study. The monocular VA of the better eye in 38 subjects was analysed at 4, 7, 10, and 12 years of age, with 20 total and 18 partial cataracts. The mean follow up time was 9.3 years after surgery.

Results: The final value of VA was 0.4 or above for approximately 50% of the subjects at 12 years of age. Visual acuity improved to a considerable extent after school age, especially in children who underwent surgery between the ages of 7 weeks and 1 year. Results for partial cataracts were favourable compared to those for total cataracts, reaching a mean of approximately 0.5 at age 12. The mean VA in the group of total congenital cataracts operated on before 7 weeks of age achieved higher values of VA at 4 years of age compared to children with total cataracts operated on between 7 weeks and 1 year of age. However, no statistically significant difference in VA results among these groups could be proved.

Conclusion: Visual acuity improves to a considerable extent after school age in children with delayed visual development caused by congenital cataracts. Surgery within 7 weeks results in a more rapid development of VA, initially.

Since the 1970s, when surgical techniques were improved and early surgery was introduced, management of congenital cataracts has resulted in normal or near normal visual development.¹⁻⁸ Despite the increased chance of restored visual function after early surgery, in reality a complete visual rehabilitation is rarely obtained.⁹⁻¹⁷ Several studies on monocular cataracts prove a association between early surgery and normalised visual function,^{8 18 19} which implies reduced risk of unequal competition,²⁰ and a critical period for surgical treatment within the first 6 weeks of life was proposed in 1996 by Birch and Stager.²¹ However, the literature is not as conclusive regarding the critical period in

cases of bilateral cataracts and the relation between age at surgery and visual outcome (Table 1).

Normally, visual acuity improves rapidly in the first 6 months after birth and more gradually thereafter, reaching adult values at 4-6 years of age.²²⁻²⁴ Owing to visual deprivation in early infancy normal visual acuity (VA) development in children with congenital bilateral cataracts is disturbed. In order to improve management of congenital cataracts, as well as to understand the maturing process of the human visual system,²⁵ evaluation of this disrupted or delayed visual development has been a subject of interest in the literature.^{1-6 9 11-17} However, it is difficult to interpret and compare these studies. Firstly, they do not provide a conclusive

Table 1 Summary of literature reports on congenital bilateral cataracts regarding relation between age at surgery and visual outcome

Study	Preferable age at surgery	Comments on visual acuity (VA)	No	Type of cataract investigated	Population
Rogers <i>et al</i> ⁶	<8 weeks	Surgery after the age of 8 weeks resulted in a substantial visual lag within the first 1.5 years, compared to surgery before 8 weeks.	7	Total	Hospital based
Gelbart <i>et al</i> ⁶	<8 weeks	Children with the best visual outcome (17 individuals) were operated on within the first 8 weeks of life, exhibiting a mean postoperative VA of 0.5 Those who underwent surgery between 8 and 41 weeks achieved a mean postoperative VA of 0.06 (7 individuals).	24	Total and partial	Hospital based
Schulz ¹⁰	<6 months	VA increased up to school age (retarded development) when operation occurred before the age of 6 months.	8	Total	Hospital based
Lorenz and Wörle ⁷	<1 year	Surgery before the age of 1 year resulted in normal VA development until 18-24 months of age.	29	Total	Hospital based
Kugelberg ⁴	<8 weeks	Surgery after the age of 3 months resulted in subnormal VA (7 individuals). Children who underwent surgery before 2 months of age (7 patients) achieved almost normal VA development.	14	Total	Hospital based
Wright <i>et al</i> ¹²	Not stated	Surgery after the age of 10 months can result in VA as good as 0.7 at the age of 4 years (total cataracts) and 1.0 for partial cataracts.	47	Total and partial	Hospital based

answer as to whether or not VA development is generally to be regarded as final at a specific age for patients with delayed visual development. This ambiguity results in the use of a final VA concept at various ages in different studies without taking into account the VA variability depending on age. Secondly, age at surgery is not the only important factor affecting VA development. Several studies contain a number of biases, such as the inclusion of various ages at onset of the visual defect, varying cataract morphology, and differing surgical techniques and ages at surgery.²⁶

In many long term follow ups of postoperative VA results presented earlier in the literature, measurements have been reported from one separate occasion.^{6 13 27 28} Our study design permits evaluation of an individual's continuous VA developmental curve related to the ages at which the measurements were taken. To our knowledge, this type of VA analysis has been performed only once before by Schulz in 1990 on seven cases of total bilateral cataracts.¹⁰

The aim of this study was to investigate the long term effects of age at surgery on visual acuity development by measuring visual acuity between preschool age and puberty. Furthermore, it aimed to report the VA levels at 12 years of age in a geographically based cohort of operated congenital bilateral cataracts.

METHODS

All children born in four western counties in Sweden between January 1980 and December 1993 who were diagnosed with congenital bilateral cataracts were included in the study. The cataracts were presumed congenital when arising early with no distinct postnatal origin defined, taking a conservative view. The cataracts were divided into two subgroups, total and partial. The diagnosis of total required that the pupil area had to be totally covered by the cataract, without being dilated, and that no red reflex was obtained. In cases of one partial and one total cataract on each eye, the patient was defined as having partial cataract. Most of those needing surgery were operated on in the department of ophthalmology, Sahlgrenska University Hospital, and followed up by the senior author (JS) regarding visual outcome and postoperative complications. The mean follow up time was 9.3 years following surgery. In all cases of total cataract the surgery was performed within 3 days of the definitive diagnosis.

During the 1980s, surgery was mainly performed using aspiration techniques and vitreous cutting instruments. From approximately 1985, the majority of lensectomies performed during the first 12 months of age included a shallow anterior vitrectomy. During the early 1990s, the technique changed to phaco enhanced aspiration, and a more extensive anterior vitrectomy is now performed. Techniques for handling the posterior capsule also evolved over this period. During the early 1980s, when aspiration with a needle was used, the posterior capsule was left intact if it was perfectly clear; otherwise, an incision was made. When the vitreous cutter was used, the visible part of the posterior capsule was removed before the anterior vitrectomy was performed in all infants. During the 1990s, a technique using curvilinear capsulorhexis was introduced, and the anterior vitrectomy was performed thereafter in all cases, regardless of age. An intraocular lens was implanted at the time of cataract extraction in one case only. Secondary implantation was performed in eight children with partial cataracts. Most of the children with secondary implants underwent surgery after 1 year of age (7/8). The impact of secondary implantations on visual acuity was therefore presumed to be limited.

Data were recorded mainly by prospective protocols created and used continuously from the beginning of the study. As a supplement to the protocols, in a minority of cases, an active retrospective search had to be performed and the medical records of these patients were reviewed. An HVOT letter

matching chart at a distance of 3 metres was used, with a 75% criterion for testing visual acuity. A shorter distance was used if VA was 0.3 or less in order to have at least eight optotypes to identify in each row. Optical correction with spectacles and contact lenses for distance was used during testing. The visual acuity of the better eye was recorded and geometric mean values were calculated. The cohort was compared with a national register recorded by low vision clinics in order to minimise the number of missed cases.

The cohort consisted of 71 children with bilateral congenital cataracts. Two children died because of multiple malformations, five children could not cooperate during visual acuity examination because of severe mental retardation, one child could not be contacted for follow up because of emigration, and in two cases the medical recordings were missing. A total of 39 children (21 boys and 18 girls) underwent cataract surgery before age 12. Three children were operated on after age 12 and were consequently excluded from the study. Of the operated patients a total of 20 children suffered from total bilateral congenital cataracts and 19 from partial cataracts. One patient with partial cataracts who underwent monocular surgery was excluded from the study because the surgery resulted in amaurosis, and the number of children analysed was consequently 38. Another 19 children (11 boys and eight girls) with partial cataracts did not qualify for surgery and were therefore not operated on at all.

Patient age at the time of cataract surgery varied from 2 days to 9 years, with a median age of 119 days. The percentage of eyes operated on at the age of 100 days or before was 46%. Twenty eight per cent were operated on after 1 year of age.

The number and types of coexisting systemic abnormalities in the cohort are shown in Table 2. Factors that may influence VA are listed in Table 3. (It should be noted that the eyes with coexisting ocular abnormalities in no cases represented the better eyes.)

The visual results from children operated on before 7 weeks were statistically compared with patients operated on between 7 weeks and 1 year, using parametric as well as non-parametric methods.

RESULTS

Almost half of the children studied (47%) demonstrated a monocular VA of 0.4 or higher during the study period and 16% (6/38) achieved a VA of 1.0 or better. Only one patient remained legally blind (VA <0.05) at 10 years of age after surgery.

Table 2 Coexisting systemic abnormalities (number of cases defined as partial cataracts appear in parentheses)

Abnormalities	No	%
Systemic:		
Trisomy 21	3 (1)	
Congenital hypothyroidism	1 (1)	
Lowe's syndrome	2	
Subclinical rubella	1	
Undefined retarded development	2 (2)	
Diabetes mellitus	1 (1)	
Total		21 (13)
Other:		
DAMP, autism	1 (1)	
Cleft palate	2 (2)	
Cerebral palsy	1 (1)	
Tourette's syndrome	1 (1)	
Epilepsy	1	
Rheumatoid arthritis*	1 (1)	
Asberger's syndrome	1 (1)	
Coeliac disease	1	
Total		24 (18)

*Coexisting with the case of congenital hypothyroidism.

Table 3 Potential factors affecting visual acuity in relation to age at surgery for type of cataract (number of cases defined as partial cataracts appear in parentheses)

	Glaucoma	PCO	Nystagmus	Secondary IOL	Ocular abnormality	Systemic disease	Hereditary factors	Subjects
Age at surgery	No	No	No	No	No	No	No	No
<7 weeks	2 (1)	10 (2)	10 (2)	0	1	4	11 (2)	15 (2)
7 weeks–1 year	4 (1)	8 (4)	9 (3)	1 (1)	1 (1)	1	4 (2)	13 (6)
>1 year	0	6 (6)	1 (1)	7 (7)	1 (1)	5 (5)	4 (4)	10 (10)
Total	6 (2)	24 (12)	20 (6)	8 (8)	3 (2)	10 (5)	19 (8)	38 (18)

Figure 1 Individual visual acuity development in the better eye in relation to age at surgery and type of cataract. Total cataracts are marked with a continuous black line and partial with a broken line. The VA development curves are discontinuous in a small number of cases because of lack of cooperation during the examination. Three patients operated on <7 weeks whose first examined VA was below 0.1 at the age of 7 years all suffered from systemic abnormalities: Lowe's syndrome, undefined retarded development and trisomy 21. **Figure 2** Stabilisation in VA development in relation to age at surgery. If no improvement in VA level was shown between the ages at which the measurements were taken, or if VA remained at the same level or below, development was defined as stabilised.

Individual visual acuity development in relation to age at surgery and type of cataract is shown in Figure 1. The children operated on after 1 year of age all had partial cataracts and exhibited higher levels of VA in a majority of cases.

VA developmental curves were analysed with the purpose of investigating the relation between age at surgery and stabilisation of VA development (Fig 2). A disparity between the two earliest operated categories was found. In the earliest group (<7 weeks) 64% stabilised after 7 years of age. Corresponding analysis of the patients who had undergone surgery between 7 weeks and 1 year of age resulted in 40%. After 10 years of age 75% in the earliest operated group had ceased to improve, whereas only 46% of patients operated on between 7 weeks and 1 year had stabilised.

The mean VA at 4 and 12 years in relation to age at surgery and type of cataract is shown in Table 4. No total cataracts were operated on after 1 year of age. On the other hand, only two cases of partial cataracts underwent surgery before 7 weeks of age and therefore no mean values were calculated. A difference in mean VA of total cataracts at 4 years of age was shown when comparing surgery within the first 7 weeks of life with surgery between 7 weeks and 1 year of age, VA 0.15 versus 0.08. Despite this finding, all children with total cataracts reached an equal mean VA level at 12 years of age independent of age at surgery. However, no statistically significant difference between the group of children operated on before 7 weeks and the group operated on between 7 weeks and 1 year could be proved.

The majority of the patients with partial cataracts had a higher level of VA at the ages of 4 and 12 compared to the group with total cataracts. The mean VA calculations in the group with partial cataracts resulted in values up to 0.52 at 12 years of age.

Table 4 Mean VA calculated for the better eye at 4 and 12 years of age in relation to age at surgery and type of cataract (number of cases defined as partial cataracts appear in parentheses)

Age at surgery	Partial		Total		Subjects (n)
	Mean VA		Mean VA		
	4 years	12 years	4 years	12 years	
<7 weeks	–	–	0.15	0.27	15 (2)
7 weeks–1 year	0.21	0.44	0.08	0.26	13 (6)
>1 year	0.26	0.52	–	–	10 (10)
Total					38 (18)

DISCUSSION

In a study of continuous VA development by Schulz, all subjects were operated on before 6 months of age and the author concluded that VA development increased up to school age.¹⁰ This finding correlates with the group of children operated on before 7 weeks in our study although we noted that 25% were still improving after age 10.²⁹ That 54% of the children operated on between 7 weeks and 1 year were also still improving in VA after age 10 was unexpected. However, a study of a normal population in Sweden by Frisén and Frisén in 1981 showed continuous development of VA until 25 years of age and a gradual decline thereafter.³⁰

Apart from age several other important covarying factors and conditions affect visual outcome, such as correction of refraction, presence of intraocular lenses, postoperative complications, nystagmus, and coexisting ocular as well as systemic abnormalities. Their impact is difficult to estimate in the present study. Compliance with amblyopia therapy probably has less effect on the results than other factors since the VA values represent the better eye. Table 3 shows that in the group of children operated on within 7 weeks, systemic diseases and hereditary factors are more common than in the other groups. These factors imply that state of health is lower in this group, which can lead to an underestimation of the gains of early surgery. The remaining covarying factors in Table 3 are otherwise represented to the same extent in the two categories of children operated on within the first year of life.

Surgical intervention before 7 weeks on total cataracts implied no improvement in VA at 12 years of age, which was unexpected but in accordance with a study by Bradford *et al* in 1994.²⁷ One possible explanation could be that some of the total cataracts discovered after 8 weeks of age were actually initial partial and progressive cataracts. Another explanation could be that visual rehabilitation after surgery was perhaps less successful in our cohort and consequently concealed the positive effects of early surgery. However, if this was the case our study would have most likely resulted in overall lower VA levels compared to those reported by Gelbart *et al*,⁶ which indicated a relation between surgery before 8 weeks and good visual outcome, but the VA levels in our study were similar. Despite this finding, the mean VA level at age 4 was higher for those who underwent surgery before 7 weeks of age.

Early surgery within the first weeks of life is on one hand described as preferable,^{4–6 8 18 19 21} yet has on the other hand been proposed to be a risk factor for postoperative complications such as glaucoma.^{31 32} The reported incidence of postoperative glaucoma after the 1970s is higher than before this decade.^{33 34} However, the literature does not provide a conclusive explanation for the seeming increase in aphakic glaucoma in these children.³⁵

The VA measured at 12 years of age in this study corresponds to the results of several studies on congenital and infantile bilateral cataracts performed during the past two

decades. They report a VA of 0.5 or higher for over 40% of the subjects studied.^{3 6 7 10 12 13 27 36–38} Two cases of total cataracts achieved a VA of 1.0 and 0.8, respectively, and both underwent surgery before 4 months of age. The range of age at surgery regarding total cataracts varied from 2 days to 7.5 months. One subject with total cataracts remained legally blind at age 10. This patient suffered from a systemic abnormality (Lowe's syndrome). In the present study 21% of the patients achieved a VA of ≥ 1.0 on the better eye at either 10 or 12 years of age, as compared to 98 % in a normal population.³⁹ The visual results have been calculated on geometric means⁴⁰ of VA of the better eye, which implies that the reported VA should be regarded as a minimum level of achievable outcome.

In this study we have not focused on the treatment after surgery as much as on the actual timing of the surgical intervention in an attempt to understand the significance of early surgery for bilateral cataracts. Despite the fact that the present study is unique regarding length of follow up and the geographical basis of the cohort, the combination of low patient number and numerous covarying factors made it impossible to statistically prove the favourable effect of young age at surgery (<7 weeks). Like Bradford *et al* in 1994,²⁷ we failed to prove that surgery during the very first weeks of life affects VA on a long term basis. On the other hand, there is a possibility that the influence of age at surgery in bilateral cataracts is limited before the age of 3 or 4 months. The characteristics of the data in our cohort did not allow further analysis regarding outcome according to this age limit. Increased knowledge about the detrimental effects of competitive inhibition⁴¹ makes it natural to consider that there may be a difference between unilateral and bilateral cataracts in the length of the latent period of visual development. If this is the case, there is a possibility that the length of the critical period also differs.

In conclusion, approximately 50% of the subjects received a VA rating of 0.4 or higher, corresponding to previous studies conducted during the past two decades. Visual acuity improves greatly after school age, especially for those who undergo surgery between the ages of 7 weeks and 1 year. VA prognoses for partial cataracts are positive compared to those for total, reaching a mean of approximately 0.5 at 12 years of age. Surgery within the first 7 weeks of life initially results in a more rapid development of VA compared with the group operated on between 7 weeks and 1 year. However, no statistically significant difference in VA results among these groups could be proved.

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