

# Ocular motor outcomes after bilateral and unilateral infantile cataracts

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Received 18 July 2005; received in revised form 27 September 2005

## Abstract

We wished to study how the severity and duration of early onset visual deprivation affects eye alignment and ocular stability. Thirty-three patients (aged 1 week to 12.8 years) with infantile cataracts (16 bilateral, 17 unilateral) were examined for periods up to 61 months. Twenty-three patients were considered to have cataracts, which were a major obstacle to vision (major form deprivation), 9 of whom underwent surgery within 8 weeks of birth (mean and SD =  $5.2 \pm 2.3$  weeks) and 10 after 8 weeks (mean and SD =  $33.9 \pm 29.7$  months). Eye alignment and fixation stability was measured using infrared recording systems and video. Visual acuity was assessed using forced-choice preferential looking techniques in the neonates and infants and with optotypes in the children. Fifteen of the 23 (65%) patients who experienced major form deprivation exhibited a nystagmus, of which 11 (73%) were manifest latent nystagmus (MLN). Nineteen of the 23 (85%) had strabismus. Of the nine patients who underwent early surgery ( $\leq 8$  weeks), two displayed a preoperative nystagmus whilst between 10 and 39 months post-operatively 8 (89%) exhibited a nystagmus. Of the group of 10 patients with minor cataracts only 2 (1 late surgery, 1 no surgery) had nystagmus and 2 strabismus. We conclude that following optimal post-operative management of infantile cataracts a sustained nystagmus—typically an MLN—is the most likely ocular motor outcome, even when the period of deprivation is as short as 3 weeks.

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**Keywords:** Infantile cataracts; Visual deprivation; Nystagmus; Manifest latent nystagmus; Strabismus

## 1. Introduction

The prevalence of infantile cataracts is approximately 3 in 10,000 live births (James, McClearn, & Waters, 1993; Stayte, Reeves, & Wortham, 1993; Rahi, Dezateux, & the British Cataract Interest Group, 2000), with bilateral opacities being more common than unilateral (Lambert & Drack, 1996; Stayte et al., 1993; Taylor, 1998). Cataracts vary greatly in their morphology and position, with some progressing with time (Lambert & Drack, 1996; Taylor, 1998). Consequently a decision must be made as to whether the cataract is significantly interfering with visual function. Small, mild cataracts or those that are eccentrically placed with respect to the visual axis are frequently managed by

observation. Dense and large cataracts necessitate immediate management (Lambert & Drack, 1996; Taylor, 1998).

Past studies on the visual outcomes have tended to concentrate on visual acuity (VA) measures (Birch & Stager, 1998; Kushner, 1995; Maurer & Lewis, 1993; Pratt-Johnson & Tillson, 1981), stressing the VA benefits of early surgery (i.e., within the first 8 weeks), particularly when linked to appropriate optical correction and full compliance with occlusion.

To date, the literature describing primary gaze fixation behaviour (i.e., eye alignment and ocular stability) of patients with infantile cataracts has been, in the main, qualitative (Birch & Stager, 1998; Kushner, 1995; Maurer & Lewis, 1993; Parks, Johnson, & Reed, 1993; Pratt-Johnson & Tillson, 1981). These reports frequently documented the presence of a “nystagmus” but did not quantify the amplitude, frequency, waveform and beat direction of the oscillations. Generally, nystagmus tends to develop as a

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consequence of the cataract type. Parks and his colleagues (1993) reported 100% of their infants with a total cataract, and 43% of those with a nuclear cataract developed a nystagmus. Nystagmus has also been reported to be more frequent in bilateral rather than unilateral infantile cataract (Lambert & Drack, 1996; Maurer, Lewis, & Brent, 1983; Wright, Christensen, & Noguchi, 1992); its overall prevalence depending on the age of the child and the stage in the management programme (Bradford, Keech, & Scott, 1994; Parks et al., 1993; Spierer, Desatnik, Rosner, & Blumenthal, 1998). Thus there appears to be a strong relationship between early visual form deprivation with an absence of binocularity and/or the presence of a nystagmus.

The timing of surgery, and therefore the duration of deprivation, has been considered an important factor in the development of nystagmus (Lambert & Drack, 1996; Maurer & Lewis, 1993; Taylor, 1998). Past studies have documented the presence of a pendular nystagmus after 10–12 weeks of life in untreated, total cataracts (Bradford et al., 1994) and, if surgery is left until 6 months post-natally, past reports suggest that a nystagmus will invariably occur (Gage, Abadi, Lloyd, & Thompson, 2001; Lloyd, Kriss, Taylor, & Russell-Eggitt, 1994; Popovic, Thaug, & Abrahamsson, 1997; Rogers, Tishler, Tsou, Hertle, & Fellows, 1981). On the other hand, Wright and his colleagues (1992) reported that when surgery was undertaken after 10 months of age, in a group of 29 patients with bilateral major cataracts (e.g., total and nuclear), only 20% developed nystagmus post-operatively. Such disparities in the reported incidence of nystagmus often reflect the differences in sample size and sample homogeneity as well as methods of recording fixation stability. As non of these studies used eye movement recording systems to assess the presence or nature of the involuntary ocular oscillations, the precise prevalence is unknown.

In this present study, we wished to determine longitudinally the ocular motor outcomes of 23 neonates and 10 infants and children with infantile cataracts. In particular, we wished to establish fixation stability and eye alignment during primary gaze. Twenty-three of the 33 were classed as having cataracts which were a major obstacle to vision (major form deprivation) and nine underwent surgery within 8 weeks of birth. Our results will show that in spite of early surgical intervention and optimal post-operative management, the large majority of patients in due course exhibited a nystagmus (65%) and a strabismus (85%). On the other hand, only 20% of the patients who experienced a minor visual deprivation exhibited either a nystagmus and/or a strabismus. These findings support the view that early pattern vision is a primary factor in ensuring normal ocular alignment and stable primary gaze holding.

## 2. Materials and methods

### 2.1. Subjects

Thirty-three patients (aged 1 week to 12.8 years) with infantile cataracts took part in this study, 12 of whom were

male and 21 female. Sixteen of the patients had bilateral and 17 unilateral infantile cataracts. Twenty patients (61%) presented with their cataracts between 1 and 24 weeks with the remaining 13 (39%) presenting between 26 weeks and 12.8 years of age. A total of 19 patients underwent surgery. Early surgery (i.e.,  $\leq 8$  weeks; mean and SD =  $5.2 \pm 2.3$  weeks) was carried out on 9 patients and late surgery (i.e.,  $\geq 8$  weeks; mean and SD =  $33.9 \pm 29.7$  months) was undertaken on 10 patients. None of the patients had any associated ocular or systemic abnormalities, and none of the cohort developed post-operative glaucoma, retinal detachments, etc. Informed consent was obtained according to tenets of the Declaration of Helsinki.

All patients underwent a full ophthalmological examination before and after cycloplegic dilation by one of the authors (ICL).

### 2.2. Grading the cataracts

Cataract morphology, density and position were evaluated with a slit lamp and ophthalmoscope when the patients were both undilated and dilated. Transmission or absorption characteristics of some of the cataracts were also computed (see Forster, Abadi, Muldoon, & Lloyd, *in press*; Gage et al., 2001, for further details). An 11 point ordinal grading scale (0–11) was assigned to the range of infantile cataracts and related to the severity of the cataract. The cataracts were divided into those which cause minor forms of visual deprivation and best left in situ and regularly observed (i.e., grades  $\leq 4.0$ ) and those which were considered to constitute a major visual deprivation and require (subject to parental permission) surgical removal (grades  $\geq 5.0$ ). Included in the first group were lamellar, sutural, sectoral and posterior polar opacities and in the second group were total, cortical, posterior polar, nuclear, posterior lenticonus and persistent hyperplastic primary vitreous cataracts. Cataracts were surgically removed in 19 out of 23 cases to avoid major form deprivation (Tables 1 and 2) and in 2 out of 10 cases in patients with minor opacities (Tables 3 and 4).

### 2.3. Eye movement recording

Binocular and monocular eye alignment and fixation stability were assessed during primary gaze using one of two infrared limbal reflection recording systems (ACS Applied Research Developments, Manchester, UK and Skalar Medical, Delft, The Netherlands). Eye movement signals from both eyes were sent either directly to a 4-channel chart recorder or relayed to a computer for off-line analysis.

For the younger patients ( $< 5.0$  years of age), the ACS sensor units were mounted in a pair of goggles (Fig. 1) and the patient's head was stabilised by either the parents or one of the authors. The system was linear to  $\pm 20^\circ$  and resolution was in the order of 5–10 min of arc. Binocular and monocular recordings were principally made in the primary

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