Repair of Strabismus and Binocular Fusion in Children with Cerebral Palsy: Gross Motor Function Classification Scale

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PURPOSE. Children with cerebral palsy (CP) tend to be either excluded from studies of strabismus repair or pooled with children who have other neurologic disorders. The authors limited this study to children with defined CP to determine the success or failure of restoring eye alignment and fusion.

METHODS. An observational, cross-sectional, prospective study was conducted on a representative cohort of 50 children. CP severity ranged from Gross Motor Function Classification System (GMFCS) level 1 (least severe) to 5 (most severe). Mean age at entrance and surgery was 3.5 years, and mean follow-up was 4.1 years (minimum 1 year).

RESULTS. The predominant form of strabismus was infantile-onset: esotropia in 54%, exotropia in 26%, and dyskinesia in 10%. Sixty-six percent of esotropic children and 61% of exotropic children achieved optimal (microtropic) alignment after an average of 2 and 1.8 surgical procedures, respectively. The likelihood of optimal alignment was similar in children with mild (GMFCS level 1–2) versus severe (GMFCS level 3–5) CP ($P = 0.7; \chi^2$). Irrespective of GMFCS severity, 46% of children gained binocular fusion/stereopsis, but the quality of fusion gained was greater in children with mild CP ($P < 0.05$). Earlier surgery was more likely to be successful ($P < 0.05$).

CONCLUSIONS. Restoration of binocular alignment and a degree of fusion is a realistic goal in the majority of strabismic CP children. Repair may be achieved in children at both the mild and the severe ends of the GMFCS spectrum, without undue concern about treatment futility or excessive reoperation. (Invest Ophthalmol Vis Sci. 2011;52:7664–7671) DOI:10.1167/iovs.10-6906

Children with neurologic disabilities tend to be excluded from studies of strabismus repair. The exclusion stems from the belief that neurologically impaired children have poorer outcomes, with variable angles of strabismus and lack of capacity for fusion. Children with neurologic impairment tend also to be pooled together in strabismus studies. The pooling makes it difficult to tease out how strabismus differs in children with various types of neurologic dysfunction. Pooling also obscures how children with mild versus severe dysfunction fare with attempted repair. Our purpose was to study a defined severity population of children with cerebral palsy (CP).

CP is a permanent, nonprogressive disorder of movement and posture caused by a lesion of the fetal or infant brain. It is the most common cause of physical disability in childhood, affecting approximately 3 infants per 1000 live births. To establish a standardized scale for grading CP severity, Palisano et al. introduced the Gross Motor Function Classification System (GMFCS). Children at GMFCS level 1 (mildest) can walk and perform all the activities of age-matched peers, albeit with limitations of speed, balance, and coordination. Children at level 5 (most severe) must be transported; they have extreme difficulty with trunk posture and have little voluntary control of limb movement. GMFCS grading has been shown to be reliable across observers and invariant with increasing age.

Children with CP have strabismus at rates exceeding those detected in neurologically normal (NN) children. The most common form is infantile-onset, which is prevalent at each level of GMFCS severity. The first goal of our study was to determine whether CP children, at different levels of GMFCS, differ in their motor outcomes or need for reoperation after attempted strabismus repair. The second goal was to determine whether repair of sensorial binocular fusion is achievable.

METHODS
An observational, cross-sectional, prospective study was conducted by performing ophthalmic/orthoptic and neurologic measurements during the same interval (within 2 weeks, often on the same day) on patients referred to the Cerebral Palsy Center, St. Louis Children’s Hospital at Washington University Medical Center, from 2000 to 2006. The participants were enrolled, and informed consent was obtained from patients or their guardians in accordance with the guidelines of the Declaration of Helsinki. Examiners were highly experienced in the care and assessment of children with CP. Family history and prenatal, pregnancy, and birth information were collected, and obstetric and neonatal records were reviewed. The results of genetic, metabolic, and neuroimaging investigations were recorded to help confirm the diagnosis of CP and to verify the absence of progressive/degenerative neurologic disease. In total, 200 items of information were reviewed for each child. For a detailed description and glossary of the standardized visuomotor testing procedures and terms used, see Ghasia et al.

The representative cohort of 50 children and adolescents reported in this study (hereinafter referred to as children) had a mean age at entrance and surgery of 3.5 years (range, 6 months-14 years), with a mean follow up of 4.1 years (range, 1.1–8.5 years). The mean gestational age was 31.6 weeks (range, 24–38 weeks).
Preoperative and Postoperative Assessment and Inclusion/Exclusion Criteria

Measurement of angle of deviation (in prism diopters [PD]) at near (0.33 m) and distance (6 m) fixation was achieved by prism and cover testing or, when necessary, by the modified Krimsky method. Cycloplegic refraction was performed at the preoperative assessments and in follow-up at least annually. Refractive errors were reported as spherical equivalent (SE, calculated as spherical error + one-half cylindrical error) and were graded as follows, in accordance with the definitions used in our previous studies:21–26: high myopia, greater than −6 D; low-moderate myopia, −4 to −6 D; emmetropia, −0.5 to −1 D; low-moderate hyperopia, greater than +1 to +4 D; high hyperopia, greater than +4 D; anisometropia, greater than ±1.5 D difference between the eyes for SE ≥6 D or 25% difference for SE >6 D.

Visual acuity (best corrected with prescribed glasses27–28 or trial frame lenses) was quantified by using optotype (Snellen letter in 36%, HOTV in 8%, or Allen figures in 6%) testing, preferential-looking (Cardiff Acuity Test29 in 8%), or spatial-sweep visually evoked potentials (SSVEPs30 in 52%). Assessment of fixation preference was used to detect amblyopia in the remaining 10% of children who would not cooperate for these acuity tests. Amblyopia was defined as an interocular acuity difference of 0.2 logMAR (approximately two chart lines) or a failure to maintain fixation through a blink. Amblyopia was treated throughout the duration of the study, using 2 to 4 hours per day of occlusion therapy, pharmacologic penalization (atropine 1% 1 drop each morning) of the dominant eye, or both.31

Inclusion criteria were no previous strabismus surgery; heterotropia (at distance fixation, near fixation, or both) of ≥12 PD horizontally or 6 PD vertically, present constantly, or present intermittently during 50% of clinical observation time; a minimum of two sets of preoperative and postoperative measurements wearing correction for any cycloplegic refractive error exceeding ≥2.0 D spherical or 1.5 D astigmatic or anisometropic; amblyopia less than an interocular acuity difference of 0.5 log (approximately five chart lines); and follow-up of at least 1 year. Children were excluded if they did not meet these criteria or if they achieved preoperative alignment to within the range defined with spectacle or bifocal correction alone (e.g., refractive/accommodative esotropes).

Binocular vision was graded as absent to most robust using the following scale: no binocular fusion, absence of sensory or motor fusion; motor fusion, fusional vergence elicited by prism-induced binocular disparity (i.e., convergence evoked by placing a 20-D base-out prism before one eye, followed by divergence when the prism was removed); sensory fusion, report of ‘four dots’ using red-green or Polaroid glasses and viewing the Worth or Polaroid four-dot target, at near or distance; and stereopsis, report of detection and elevation of the salient figure above the background (measured in arc seconds) using Polaroid glasses to view figures of the Stereo Fly or Randot preschool stereocuity tests (Stereo Optical Inc., Chicago, IL) at 0.33 m. Both Titmus Fly and Preschool Randot testing was attempted in all patients 3 years or older; 30% (15 children) in the study could cooperate for stereopsis testing.

Strabismus Surgery

All surgeries were conducted as outpatient procedures, under general anesthesia, following routine pediatric techniques. Bilateral medial rectus recession was performed for esotropia and bilateral lateral rectus recession for exotropia, as the first surgery, using standard surgical dosage tables without modification.32 The horizontal recti were transposed vertically for A or V patterns exceeding 10 PD in up or down gaze. Recession or anterior transposition of the inferior oblique muscle was carried out to correct elevation-induction or dissociated vertical deviation. Stable and variable angle strabismus (stable, variable, and dyskinetic are defined in Statistical Analysis and Analysis Terminology) were treated for the mean angle, pooling all measurements. Dyskinetic strabismus was treated for the mean angle in the predominant direction of deviation. The informed consent discussion included presumed higher probability of suboptimal outcome and need for reoperation in children with CP. Postoperative measurement of motor alignment was recorded at 1 week, 3 ± 1 months, 6 ± 2 months, 1 year ± 3 months, and annually thereafter. Children who had undercorrection or overcorrection (i.e., residual or consecutive deviations) were seen at more frequent intervals when deemed appropriate. Binocular fusion was tested postoperatively at 3 months and each visit thereafter. Reoperation was performed 3 to 6 months after the first surgery for undercorrection or overcorrections of sufficient magnitude, defined as horizontal heterotropia ≥12 PD or vertical ≥6 PD. Reoperations were tailored to address the amount of overcorrection or undercorrection and the magnitude of muscle correction performed at the previous surgery (e.g., re-recession of a previously recessed muscle or resection of opponent muscles).

Statistical Analysis and Analysis Terminology

Eye alignment outcomes in the results were analyzed for preoperative direction of deviation and stability of misalignment angle. Preoperative test-retest variability over a mean 14 weeks was within the range reported in NN children (i.e., ≥60% of initial deviation, excluding dyskinetic), defined as follows: stable, angles of heterotropia varying by <10 PD, comparing sets of measurements obtained on different examination days; variable, angles varying by ≥10 PD; dyskinetic, angles that inverted direction (e.g., from esotropia to exotropia) during the same examination.33 Postoperative eye alignment (motor) outcomes were divided into optimal, better, or worse. Optimal was defined as horizontal heterotropia ≤8 PD and vertical as ≤4 PD with follow-up of at least 1 year. Better was defined residual heterotropia greater than optimal but less than 50% of the strabismus angle before any surgery (undercorrection). Worse was defined as heterotropia greater than optimal and either opposite in direction to the angle before any surgery (major overcorrection) or 50% of the angle before any surgery (major undercorrection). We designated major undercorrection within the category of worse outcomes because the child and family undertook the burdens and risks of surgery but received no substantial benefit.

Postoperative binocular (sensory) outcomes were analyzed as gained, same, or lost binocular fusion. Gained indicated an improvement of at least one grade (e.g., an improvement from sensory fusion to stereopsis), same indicated no change in fusion grade, and lost indicated any decrement in grade.

All the children had strabismus but had not previously undergone surgery at the entry examination. Therefore, the date of onset of strabismus was estimated in a combination of facial photographs, parental report, and review of any previous eye examination records. Strabismus onset in the first 6 months of life is known to be accompanied by a constellation of permanent ocular motor signs, such as latent nystagmus, pursuit/optokinetic nystagmus (OKN) asymmetries, and dissociated vertical deviation (DVD; i.e., fusion maldevelopment/infantile strabismus syndrome).34–36 Later-onset strabismus lacks these signs. In children in whom the first (outside) ophthalmologic examination occurred after age 6 months but in whom the signs of the infantile strabismus syndrome were manifested on entry into our study, onset was set to a default value of age 6 months.

The ophthalmologic examiners were masked to the GMFCS and to the physiological and anatomic CP classifications. Patient data were collated for analysis by unmasking for GMFCS; the database was then scanned and sorted in a consecutive, rolling fashion for GMFCS levels 1 to 5, until 10 patients in each class were identified. The strategy ensured random assignment of patients to GMFCS bins of equal size, providing a representative study population of 50 patients. Gestational age and age at examination were compared across GMFCS levels by one-tailed analysis-of-variance (ANOVA). The $\chi^2$ square test was used to assess differences between motor and sensory outcomes in GMFCS levels and types of preoperative strabismus. For this analysis, GMFCS levels were collapsed into mild (levels 1–2) versus severe (levels 3–5). This convention was used because it has been used in previous studies.
of CP severity and logically divides children into those who retain the capacity to walk unassisted (Table 1, GMFCS levels 1–2) and those who require a supportive walking device or a transporter (GMFCS levels 3–5). The t-test was used to compare durations of strabismus between groups. Significance was defined as P < 0.05.

RESULTS

Preoperative Strabismus Characteristics and Severity of CP

Eighty-nine percent of the children, across all levels of GMFCS, had infantile-onset strabismus, as evident by markers for fusion maldevelopment in the first months of life: latent nystagmus, pursuit/OKN asymmetry, and DVD. The other 11% had maldevelopment in the first months of life: latent nystagmus, had infantile-onset strabismus, as evident by markers for fusion. Eighty-nine percent of the children, across all levels of GMFCS, were detected in 52% of children. The children at entrance into the study (ANOVA; respectively). Table 2 shows the frequency and type of ametropia, amblyopia, and gaze-maldevelopment gaze disorder in the subsets of children with esotropic, exotropic, or dyskinetic strabismus.

Postoperative Alignment (Motor) Outcome, Rates of Reoperation, and Severity of CP

The first goal of the study was to determine whether the severity of CP, as graded by GMFCS level, was predictive of successful surgical realignment. The average preoperative horizontal strabismus angle was similar (P = 0.31) in children with mild versus severe CP: 27 ± 6 PD for children with levels 1 to 2 versus 31 ± 7 PD for levels 3 to 5. Motor outcomes after one or more surgeries were divided into three groups: optimal (≤8 PD) alignment, suboptimal but better, or worse than angle of strabismus at entry into the study.

Optimal motor outcomes were achieved in 58% of children overall (n = 29/50). As shown in Figure 1 (top), the likelihood of an optimal outcome was similar in children with mild versus severe CP (56% vs. 60%; P = 0.7; χ²). The number of surgeries performed in children with mild versus severe CP was also similar (1.9 vs. 2.06). The categories optimal and better can be combined to answer the question: What was the chance that strabismus surgery would, at the very least, improve alignment rather than making it worse? The chance of improvement was 86% in children with mild CP versus 69% in children with severe CP. Inverting the question, the chance of worse alignment after surgery was twice as great in children with severe CP (31% in severe CP vs. 14% in mild CP).

Binocular Fusion (Sensory) Outcomes and Severity of CP

The second goal of the study was to determine whether strabismus surgery improved binocular fusion. Figure 1 (bottom) shows that, irrespective of CP severity, 46% of children (n = 23) gained at least one grade of binocularity after one or more strabismus surgeries. A gain was measured in 52% of children with mild CP versus 40% with severe CP (P = 0.15). The risk

<table>
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<tr>
<th>Level</th>
<th>Functional Capability/Limitation</th>
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<tbody>
<tr>
<td>1</td>
<td>Walks without assistance; limited advanced motor skills</td>
</tr>
<tr>
<td>2</td>
<td>Walks without assistance; limited advanced walking</td>
</tr>
<tr>
<td>3</td>
<td>Walks with assistive walking device</td>
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<tr>
<td>4</td>
<td>Self-mobility with a transporter</td>
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<tr>
<td>5</td>
<td>Self-mobility with a transporter; limited</td>
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<td>2</td>
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Table 2. Type of Ametropia, Amblyopia, and Gaze Disorder in the Esotropic, Exotropic or Dyskinetic-Strabismus Children

<table>
<thead>
<tr>
<th></th>
<th>Refractive Errors</th>
<th></th>
<th>Amblyopia</th>
<th></th>
<th>Gaze Disorders</th>
</tr>
</thead>
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<tr>
<td></td>
<td>Mild CP % (n)</td>
<td>Severe CP % (n)</td>
<td>Mild CP % (n)</td>
<td>Severe CP % (n)</td>
<td>Mild CP % (n)</td>
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<tr>
<td>Low/moderate hyperopia</td>
<td>18 (2)</td>
<td>31 (4)</td>
<td>25 (2)</td>
<td>46 (6)</td>
<td>20 (1)</td>
</tr>
<tr>
<td>Low/moderate myopia</td>
<td>18 (2)</td>
<td>15 (2)</td>
<td>12.5 (1)</td>
<td>8 (1)</td>
<td>0</td>
</tr>
<tr>
<td>High myopia &gt; 4 to 0.5</td>
<td>0</td>
<td>8 (1)</td>
<td>0</td>
<td>23 (3)</td>
<td>0</td>
</tr>
<tr>
<td>High hyperopia &gt; 4</td>
<td>27 (3)</td>
<td>15 (2)</td>
<td>0</td>
<td>0</td>
<td>40 (2)</td>
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<tr>
<td>Anisometropia &gt;1.5</td>
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<td>8 (1)</td>
<td>12.5 (1)</td>
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<td>20 (1)</td>
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<tr>
<td>Emmetropia &lt; +1 to −0.5</td>
<td>36 (4)</td>
<td>23 (3)</td>
<td>50 (4)</td>
<td>23 (3)</td>
<td>20 (1)</td>
</tr>
<tr>
<td>Strabismic</td>
<td>64 (7)</td>
<td>38 (5)</td>
<td>25 (2)</td>
<td>31 (4)</td>
<td>20 (1)</td>
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<tr>
<td>Anisometropic + strabismic</td>
<td>9 (1)</td>
<td>0</td>
<td>12.5 (1)</td>
<td>8 (1)</td>
<td>0</td>
</tr>
<tr>
<td>None</td>
<td>27 (3)</td>
<td>62 (8)</td>
<td>62.5 (5)</td>
<td>62 (8)</td>
<td>80 (4)</td>
</tr>
<tr>
<td>LN/MLN</td>
<td>64 (7)</td>
<td>77 (10)</td>
<td>62.5 (5)</td>
<td>92 (12)</td>
<td>100 (5)</td>
</tr>
<tr>
<td>Pursuit/OKN asymmetry</td>
<td>45 (5)</td>
<td>31 (4)</td>
<td>50 (4)</td>
<td>31 (4)</td>
<td>60 (3)</td>
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<tr>
<td>DVD</td>
<td>45 (5)</td>
<td>38 (5)</td>
<td>50 (4)</td>
<td>46 (6)</td>
<td>80 (4)</td>
</tr>
<tr>
<td>None</td>
<td>27 (3)</td>
<td>8 (6)</td>
<td>0</td>
<td>8 (1)</td>
<td>0</td>
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</tbody>
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LN, latent nystagmus; MLN, manifest latent nystagmus; OKN, optokinetic nystagmus; DVD, dissociated vertical deviation.
that surgery would cause any loss of fusion was low. Loss after surgery occurred in 6% of children with mild CP and 2% in children with severe CP. No child reported persistent postoperative diplopia or visual confusion.

All children who gained binocularity had optimal motor outcomes (i.e., alignment to within ±8 PD of orthotropia). Viewed from this perspective, how many children who achieved an optimal motor outcome failed to achieve any gain in level of fusion? Four percent of children with mild CP achieved an optimal motor outcome but no gain in fusion compared with 20% of children with severe CP ($P = 0.13$; $\chi^2$).

Among the group of children who gained binocularity, differences were observed in repair of the quality of fusion. Fifty percent of children with mild CP who had improved binocularity gained sensory fusion (i.e., Worth or Polaroid four-dot) versus 19% with severe CP. A gain to some level of stereopsis was achieved in 20% of children with ET versus 18% with XT. Change in grade of binocularity was similar ($P = 0.56$) in children who had stable angles and in those who had variable angles of strabismus before surgery (Fig. 2).

**Alignment after One or More Surgeries**

Figure 3 shows the angle of strabismus at entry into the study and the angle after one or more surgeries, up to a total of three. Measurement intervals between Post1 and Pre2, as an example, ranged from 3 to 6 months. Figure 3 (top) displays primary ET children and primary XT children who achieved optimal alignment for a period exceeding 1 year (median follow-up, 3.1 years). To achieve optimal alignment, 50% of children with primary ET required one surgery, 37% required two surgeries, and 13% required three surgeries. Most children (88%) with primary ET who did not achieve optimal alignment after one surgery required reoperation for secondary XT; a minority (12%) required reoperation for a residual ET (12%).

For primary XT children, 70% required one surgery, 15% required two surgeries, and 15% required three surgeries to achieve optimal alignment. The majority (62%) of primary XT children who did not achieve optimal alignment after one surgery underwent reoperation for a residual XT; the minority (38%) needed reoperation for a secondary ET. Combining ET

**Alignment and Fusion Outcomes as a Function of Preoperative Strabismus Subtype**

Motor outcomes for the different subtypes of strabismus are shown in Figure 2 (top). Motor outcomes were similar for children with ET and for XT; 66% of ET children and 61% of XT children had optimal outcomes ($P = 0.70$; $\chi^2$). No child with dyskinetic strabismus had an optimal outcome. The stability of the preoperative angle of strabismus was not predictive of outcome (Fig. 2). The motor outcome was optimal in 67% of children with stable angles compared with 59% of those with variable angles ($P = 0.65$; as noted in Methods, dyskinetic strabismus was considered a distinct subtype and was not classified as variable angle). To quantify the chance of improving alignment, the categories “optimal” and “better” can be combined. The chance of improvement in alignment was 71% in children with ET versus 81% in children with XT ($P = 0.70$); the chance of improvement was 81% for optimal fusion compared with 58% for variable ($P = 0.13$; Fisher exact test).

Figure 2 (bottom) plots postoperative gains in grade of fusion for the different strabismus subtypes. Binocular fusion improved in 63% of ET children and 37% of XT children ($P = 0.13$; $\chi^2$). Fusion did not improve in any child with dyskinetic strabismus. The quality of fusion repair tended to be better in children with primary ET; 50% of children with ET who had improved binocularity gained sensory fusion compared with 25% with XT. A gain to some level of stereopsis was achieved in 20% of children with ET versus 18% with XT. Change in grade of binocularity was similar ($P = 0.56$) in children who had stable angles and in those who had variable angles of strabismus before surgery (Fig. 2).
those who did not was 2.6. Shorter durations were associated with better outcomes. The duration before the first surgery was analyzed to determine the effect on alignment and fusion outcomes.

FIGURE 3. Alignment before and after the first and any subsequent strabismus surgeries, up to a total of three operations. Endpoint alignment is that at last follow-up. Horizontal dashed lines: range of ±8 PD or microtropia/optimal alignment. Optimal alignment (top) plots children who achieved mirotopia at last follow-up. Persistent misalignment (bottom) plots those who did not achieve mirotropia. Measurement intervals: Pre1/Post1, before and after first surgery; Pre2/Post2, before and after second surgery; Pre3/Post3, before and after third surgery.

and XT, more than half (~60%) of children who achieved optimal alignment required one surgery. The average number of surgeries performed was 1.8 for primary XT, 2.0 for primary ET, and 2.6 for dyskinetic strabismus.

Persistent misalignment was defined as failure to achieve optimal alignment after three surgeries (Fig. 3, bottom). The primary ET children in this category tended to be overcorrected (persistent secondary XT, 72%), whereas the primary XT children tended to be undercorrected (persistent residual XT, 71%). Combining the data in Figure 3, overcorrection was approximately twice common for primary ET than for primary XT. Figure 4 plots alignment for children who had dyskinetic strabismus at entry into the study (data points = mean angle of predominant deviation). All these children had persistent misalignment.

Duration on Misalignment and Outcome

Duration of strabismus before the first surgery was analyzed to determine the effect on alignment and fusion outcomes. Shorter durations were associated with better outcomes. The duration in children who achieved optimal alignment and in those who did not was 2.6 ± 1.2 years and 4.8 ± 2.6 years, respectively (P < 0.05; t-test). The duration in children who gained versus those who did not gain a grade of fusion was 2.2 ± 0.9 years vs. 5.1 ± 3.7 years (P = 0.02). Duration of strabismus was comparable in children with mild versus severe CP (4.3 ± 3.0 years vs. 3.2 ± 1.9 years; P = 0.31). Although shorter duration was associated with better final sensory and motor outcomes, shorter duration before the first surgery was not related to the need for more than one surgery (P = 0.47).

FIGURE 4. Alignment before and after the first and any subsequent strabismus surgeries, up to a total of three operations. Measurement intervals: Pre1/Post1, before and after first surgery; Pre2/Post2, before and after second surgery; Pre3/Post3, before and after third surgery.

Ametropia and Amblyopia

Most of the children (93%), across all levels of GMFCS, had low to moderate degrees of ametropia, with hyperopes exceeding myopes by a ratio of 2.5:1 (Table 2). The most common type of ametropia was low to moderate hyperopia, found in 48% of children. Anisometropia was detected in 13%, high hyperopia in 11%, and high myopia in 9% of children. High myopia occurred with greater frequency in children with severe CP (levels 3–5; P < 0.05), as has been reported. Strabismic amblyopia was present in 44%, anisometropic amblyopia in 11%, and combined strabismic-anisometropic in 6% of children. Amblyopia was mild. Amblyopia worse than 0.2 logMAR difference between the eyes was detected in only one patient. Absence of amblyopia was not associated with better alignment or fusion outcome (P = 0.40; Fisher exact test).

DISCUSSION

The first goal of our study was to determine whether children with CP, at different levels of GMFCS, differ in their motor outcomes or need for reoperation after attempted strabismus repair. The answer for both questions is no. Independent of CP severity, more than half (58%) of the children achieved good alignment. This was accomplished in both the mild and the severe CP groups after an average of two surgeries. The second goal was to determine whether repair of sensorial binocular fusion differed as a function of GMFCS. The answer is yes. Children with mild CP had twofold to threefold greater chance of gaining Worth/Polaroid four-dot fusion or stereopsis. That is, children with more severe CP had less chance of gaining fusion even when their eyes were optimally realigned.

Strabismus Characteristics in CP versus NN Children

Compared with NN children, children with CP have a high prevalence of strabismus. The prevalence in CP children is 50% to 90% across all levels of GMFCS compared with 3% to 5% in NN children. The strabismus in CP tends also to be disproportionately early onset. Infantile-onset of the stra-
Strabismus occurs in approximately 90% of CP children25,25 compared with approximately 20% of NN children.10,43,44 For this reason, we confine the discussion here to comparisons between our data and outcomes in NN children with infantile strabismus.

We defined stable and unstable angles of strabismus as less than or greater than a 10 PD variation of the esotropia or exotropia from one set of measurements to another. Using these criteria, 76% of the CP children in our study had stable angles and 14% had unstable angles (the remaining 10% were dyskinetic and thus excluded from this analysis). Exclusion of unstable angles and the rates of amblyopia (~50%) are comparable.

Strabismus Repair Outcomes in CP versus NN Children

The CP children in our study who had infantile esotropia achieved optimal (microtropia) alignment outcomes in 66% of cases, after an average follow-up of 4 years. This outcome was within the range reported in several studies of NN children: 63% to 83%, with follow-up varying from 6 months to 8 years.2,5,45 Optimal alignment was achieved in 61% of CP children with infantile esotropia. This outcome was also within the range reported in NN children (50%–82% with follow-up from 4 to 8 years).1,3 Achieving optimal alignment in the CP children required, on average, two surgeries, similar to that required in the NN children (1.8–2.0). We did find a higher rate of secondary, exotropic overcorrection in exotropic CP children after one surgery (50%). The rates reported in NN children with infantile esotropia are 18% to 30%.2,5,45

Fusion and stereopsis outcomes in the CP children reported here were lower than those that have been reported in NN children. Postoperative fusion was restored in 50% of CP children with infantile esotropia and 25% of children with exotropia. The comparable percentages in NN children with infantile esotropia are 69% to 90%,2,4 and in NN exotropes they are 50% to 70%.1,3 Stereopsis was restored in 20% of the CP esotropes and 18% of the exotropes. The comparable percentages are 27% to 40% for NN infantile esotropes2,4 and 20% for NN infantile exotropes.1,3

The discrepancy in fusion and stereopsis outcomes between CP and NN children might not have been due entirely to the presence of neurologic disease. Part of the discrepancy might have been due to delayed referral and treatment. Because the CP children in our study were not enrolled until they presented to the CP unit, the average age at first surgery was 45 months in esotropes and 78 months in exotropes. In studies of NN children,1,3,11 however, the first surgery was performed at an average age of 5.8 months in esotropes and 15 months in exotropes. When surgery for infantile strabismus is delayed in NN children for several years, the fusion and stereopsis outcomes do not differ substantially from those of our study group.46–48 Despite the delayed treatment, we found a significantly higher rate of restoring alignment, fusion, and stereopsis in CP children with shorter durations of strabismus before the first surgery.

Opponents of early strabismus surgery for children with CP claim that the angles of misalignment are more unstable and that this factor will lead to poorer outcomes.7,10,22,49 Excluding dyskinetic strabismus, the results of our study, and those in NN children with stable versus unstable angles, do not support this argument. First, instability of alignment in the CP children was close to that reported in NN strabismic children.44 Second, outcomes of strabismus surgery, both in the CP children of the present study and in previous reports of NN children, indicate that success does not differ in children with stable versus unstable angles.6,45,50 Favorable outcomes, in both CP and NN children, are linked to shorter durations of misalignment (earlier surgical treatment), not to angle stability.4,48,51

Comparison with Previous Studies of Strabismus Repair in CP

A prime motivation for this study was the paucity of reports in the literature quantifying strabismus repair in children with CP. As noted, a limitation of our study was lack of an age-matched treatment group of NN children with infantile strabismus. The major strengths of our study were prospective treatment of a well-defined CP population, GMFCS specification of CP severity, and documented sensorimotor outcomes.

In 1975, Hiles et al.9 reported the results of strabismus surgery in a large series of CP children (retrospective, categorized as spastic mild, moderate, or severe; ataxic and athetoid). For reasons that are not clear to us, his results are more favorable than ours and are equal to or better than those reported in NN children. Ninety children with nonaccommodative esotropia and 36 children with exotropia underwent strabismus surgery at an average age of 5 years. Seventy-seven percent of esotropes achieved micro tropia or orthophoria after an average of 1.1 operations; only 5.5% developed secondary exotropia. Eight-nine percent of exotropes achieved micro tropia or orthophoria; only 3% developed secondary exotropia. No reoperations in exotropes were performed. Similar levels of success were noted for children with mild to severe CP over an average follow-up of 4.5 years (the authors did not disclose age of onset, stability of angle, overcorrection angles, or preoperative or postoperative fusion). The discrepancy between our success rates and those of Hiles et al.9 cannot be ascribed to a difference in magnitude of muscle recession or surgical technique. They used bilateral medial rectus recession or lateral rectus recession and adhered to standard surgical tables developed for surgery in NN children, as did we.

Seaber and Chandler34 reported a retrospective study of strabismus surgery in 12 CP children, 58% with esotropia and 42% with exotropia (not further defined). For the esotropes, 86% achieved micro tropia after one surgery compared with 60% of exotropes. None of the esotropic patients in their series had secondary exotropia (the authors did not disclose CP severity, age of onset, stability, surgical methods, preoperative or postoperative fusion, or follow-up interval).

A subset of CP children have strabismus that changes from esotropia to exotropia during the same examination interval, unrelated to accommodation effort or attention. The first noteworthy report of this bidirectional strabismus was that of Seaber and Chandler.34 They noted its linkage with athetoid (i.e., severe) CP and stated that it is not observed in NN strabismic children. The term “dyskinetic strabismus” was coined to describe this in a subsequent paper by Buckley and Seaber,35 who reported its occurrence in 84% of children with athetoid CP and 32% of children with severe spastic CP. Although only 10% of the CP children in our study had dyskinetic strabismus, all who had dyskinetic strabismus had severe CP. Buckley and Seaber35 commented that, “unlike other forms of strabismus, dyskinetic strabismus does not respond to surgery.” We concur. None of the children on whom we performed surgery for dyskinetic strabismus responded favorably.
Reports of Strabismus Repair in Children with Diverse Types of Neurologic Impairment

Several retrospective case series devoted to the repair of infantile esotropia have lumped CP children and non-CP neurologically impaired (NI) children.5,11–14,52–54 The non-CP diagnoses span diverse types and severities of neurologic impairment, such as hydrocephalus, mental retardation/developmental delay, Down syndrome, meningomyeleocele, intraventricular hemorrhage, seizure disorders, neurofibromatosis, and hyperactivity. The outcomes of children with CP in these series were not parsed in the analysis of results. Age at surgery and follow-up intervals varied, choice of surgical technique differed, and fusion outcomes were not reported, making comparisons with our results tenuous. Acknowledging the pitfalls, the outcomes of these series appear nonetheless to be in rough agreement with ours. Optimal/microtropic alignment is achievable in 55% to 66% of esotropic NI children after one surgery.11–14,52–54 The rate of exotropic overcorrection, using standard surgical amounts, is more common in esotropic NI than in NN children,52 but decreasing the dosage of bilateral muscle recession results in a higher rate of undercorrection.55

Implications of the Current Study

Strabismus and amblyopia deprive children of the benefits of normal eye alignment and binocular fusion.55 In NN children, subnormal binocularity impedes both eye-hand coordination and the development of gross motor skills, such as crawling and walking.56,57 These skills of coordination and balance are impaired to a greater or lesser degree in CP, independent of strabismus. The child with CP and unrepaird strabismus experiences a doubly negative effect; lack of normal binocular cues exacerbates the baseline dysfunctions of limb movement and posture. Early repair of strabismus is known to boost eye-hand coordination and acquisition of gross motor skills in NN children,57 and would be expected to have analogous beneficial effects in children with CP. Beyond the functional impairments, unrepaird strabismus harms self-image, confidence in social interactions, and success in future employ-ment.58,61 The psychosocial harm would be expected to be heightened in CP children, who may be ridiculed or ostracized both for their motor handicaps and for strabismus.

Our results indicate that the restoration of binocular alignment and a degree of fusion is a realistic goal in the majority of strabismic CP children. Earlier surgery yielded greater rates of success. Repair may be achieved in children at both the mild and the severe ends of the GMFCS spectrum without undue concern about treatment futility or excessive reoperation.

References


