INTRODUCTION Congenital esotropia is a convergent deviation of the eyes when fixating on an object. It appears during the first six months of life and affects muscle structure and physiology, as well as the relation of the eyes with the environment, retinal relations and neural integration phenomena. Amblyopia develops in 30–50% of those affected. Timely surgery is effective in most cases.


METHODS A descriptive, retrospective study was carried out through review of 127 cases (aged 1–18 years, 55 male and 72 female) operated on for congenital esotropia between January 2008 and May 2009. Variables used were: age at surgery, degree of preoperative ambyloida, type of fixation, refractive error, associated conditions, type of surgery performed and pre- and postoperative deviation angles.

RESULTS The largest number of congenital esotropia cases were found in children <2 years old (38.6%). Before surgery, 37% of cases studied showed mild amblyopia; 22.8% moderate and 12.6% severe; 27.6% did not cooperate with visual acuity testing. With respect to fixation: 91.3% showed central fixation and 8.7% eccentric. Refractive errors found were: mild hyperopia (65.4%), moderate hyperopia (29.1%), and myopia (5.5%). Associated conditions were: overaction of the inferior oblique muscles (48%), latent nystagmus (16.5%), and dissociated vertical deviation (8.7%); no alterations were found in 26.8% of cases. The most frequently used surgical treatment was bilateral medial rectus muscle recession (92.1%). The most common preoperative deviation angle range was 31–40 prism diopters (PD) in 51.9% of patients; the postoperative deviation angle most commonly found after 12 months was <10 PD in 64.6% (orthotropia).

CONCLUSIONS Surgery, most commonly with medial rectus muscle resection, was effective in correcting congenital esotropia.

KEYWORDS Esotropia/congenital; esotropia therapy; oculomotor muscles/surgery; strabismus; squint; amblyopia; vision, binocular; Cuba

Strabismus is an ophthalmological disorder in which the eyes are misaligned; it affects 2–4% of children.[1] About half of these disorders are esodeviations, whose causes are anatomical, neurological, mechanical, refractive, genetic and accommodative.[2,3] Congenital esotropia is a convergent deviation of the visual axes when fixating on an object. It appears in the first six months of life, generally between two and four months. While timing of its first clinical signs varies, it is the result of a congenital failure to develop normal binocular vision, probably genetically-determined development in normal binocular vision.[4,5] In general, it occurs in children who show orthotropia or exodeviations at birth. Congenital esotropia should not be confused with other true congenital deviations (occasionally fixed esodeviations), which can be detected from the first day of life and are clinically well defined.

Reported prevalence of the condition varies. Some authors say it affects some 0.1% of the population;[6] Mohney et al. reported a 9–year period prevalence of 0.27%. [1] Prevalences between 0.67 and 2% have been found among ophthalmological patients;[7,8] and a study of strabismus patients in Mexico found congenital esotropia to be the most frequent type (28.46%), with similar rates for both sexes. [9–11] There are no prevalence data on this condition in Cuba.

In most cases, congenital esotropia appears to be caused by defective development of fusion (cerebral integration of images perceived by both eyes). This hypothesis is supported by some patients’ failure to achieve normal binocularity even with very early alignment of the eyes. However, environmental factors cannot be discounted, since a high frequency of strabismus and amblyopia is notable among low birth-weight and premature infants, and those suffering perinatal hypoxia. Smoking, drugs and alcohol abuse during pregnancy also disrupt brain development and are closely associated with amblyopia and strabismus.[12] Additionally, inheritance may play a role in the pathogenesis of congenital esotropia; some authors report binocular vision anomalies in 16% of parents of children with congenital esotropia.[12,13]

Congenital esotropia may be accompanied by amblyopia (reduction in uni- or bilateral best corrected visual acuity not attributable to structural abnormality of the eye or posterior visual pathway), thus establishing the sensory nature of the impairment.[14] Ocular misalignment is in fact the main cause of amblyopia. Constant esotropic deviations, rather than alternating ones, are the fundamental cause of strabismic amblyopia.[15] Amblyopia is classified according to best corrected visual acuity (VA): mild (0.9–0.6), moderate (0.5–0.3) and severe (0.2–0.05).[15] Some authors do not explicitly define mild amblyopia and others ignore it, referring only to moderate and severe amblyopia.[15]

Limitations in social and psychological development resulting from amblyopia make effective and early correction imperative. Delay in surgical alignment of congenital esotropia is an important risk factor for postoperative amblyopia.[5,12] However, criteria vary regarding the optimal timing of surgery. Costenbader—a recognized authority in strabismus management—suggested surgery was needed at 12 months to expect functional cure.[4] Most ophthalmologists agree that surgery should be done early, aligning the eyes before 24 months of age to attain binocular vision. Some hold that surgery may be performed in healthy children as early as 4–6 months of age to maximize binocular function and improve stereopsis.[11,12]

Significant refractive errors should be corrected and treatment of amblyopia undertaken before surgery, creating better visual conditions, and mitigating accommodation or anisometric defects. In very farsighted children with a small deviation angle <20 prism diopters (PD), correcting the refractive error may avoid surgery. When amblyopia is suspected, occlusion therapy is indicated—
daily patching time dependent on the child’s age—to force the weaker eye to fixate.[12] Patching is not just ideal to prevent or treat amblyopia but also to improve esotropia as such; by reducing hypertonicity in the medial rectus muscle and stimulating abduction of the non-occluded eye, it enhances binocular vision from a sensory perspective.[13]

Conventional operations routinely used for other clinical forms of esotropia are often inadequate for alignment in cases of congenital esotropia, the latter sometimes involving large deviation angles; nor are these surgeries generally effective for correcting torticollis.[12] Hence specific surgical procedures are needed.

Bilateral recession of the medial rectus muscles was already being done in the 1950s. Costenbader postulated that congenital esotropia treatment should be based on weakening the medial rectus muscles to prevent eventual loss of cortical inhibition of hyperactive convergence.[4,5,12,14] This is the rationale for standard treatment: bilateral recession of the medial rectus muscles. The extent of recession is generally based on the degree of deviation with near fixation, since in infants it is difficult to obtain exact measurements with distant fixation. In older patients with significant irreversible amblyopia, surgery should be limited to the amblyopic eye, with recession of the medial rectus muscle and resection of the lateral rectus muscle.[4,5,12,14]

In Cuba’s national health system, primary health care is key to identifying children with congenital esotropia. They are seen in well-baby visits involving a neighborhood family doctor and pediatrician, and later referred to ophthalmology and strabismus services in provincial pediatric hospitals throughout the country, including six in Havana plus the Ramón Pando Ferrer Ophthalmology Institute (ICO, its Spanish acronym).

School-age children who have not received early surgery are enrolled in public special education schools, where the goal is to develop each child’s visual function as much as possible, optically and perceptually, through a comprehensive system of education and rehabilitation. This involves methodologies based on physiological, ophthalmological, psychological and pedagogical principles to foster greater independence and active social participation.[16] In the case of amblyopic children, they are able to follow their daily patching routine with fewer psychological implications, especially as they reach ages when appearance becomes more important. Parents receive guidance on how to manage these conditions and on the rehabilitation necessary for their child’s eventual integration into the mainstream educational system.[16]

The ICO is Cuba’s national ophthalmology reference center.[17] The ICO Pediatric and Strabismus Service receives patients referred from Havana and across the country for treatment intended to achieve ocular alignment, correct torticollis, and prevent amblyopia, with a minimum of surgical interventions. It also provides psychological support for the children and their families. In addition, the Service receives those congenital esotropia patients whose comorbidities (dissociated vertical deviations, overaction of the oblique muscles, vertical anisometropia) complicate their surgical management by other hospitals around the country.

Our prophylactic or therapeutic practice in suspected amblyopia is alternating patching before age one, with the regimen determined by the child’s age. Refractive errors are corrected to improve vision as much as possible and to mitigate associated accommodation factors or anisometropia. When a patient has alternating esotropia from onset, with a deviation angle >20 PD and without significant neurological damage, surgery is performed prior to 24 months to ensure the best chance for normal binocular vision development.

The objective of the present study is to describe surgical results in congenital esotropia cases treated at the Ramon Pando Ferrer Ophthalmology Institute in Havana between January 2008 and May 2009.

**METHODS**

A descriptive retrospective study was designed, based on review of clinical records, to evaluate results of surgical treatment of congenital esotropia in cases referred to the ICO Pediatric and Strabismus Service from January 2008 through May 2009.

Of 149 records reviewed of patients aged <18 years, 127 met inclusion criteria—deviation angle >20 PD and no significant neurological damage. Of those, 55 were male and 72 female. Excluded were those whose congenital esotropia did not fulfill surgical criteria (i.e. deviation angle ≤20 PD or significant neurological damage), or whose clinical records were incomplete.

Variables studied were: age at surgery, sex, degree of preoperative amblyopia, type of fixation, refractive error, comorbidities, type of surgery performed and pre- and postoperative deviation angles (Table 1).

<table>
<thead>
<tr>
<th>Variable</th>
<th>Parameters</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at surgery (years)</td>
<td>&lt;2, 2–4, 5–7, 8–10, &gt;10</td>
</tr>
<tr>
<td>Degree of amblyopia (visual acuity, VA)</td>
<td>Mild, Moderate (0.6–0.9), Severe (0.05–0.2)</td>
</tr>
<tr>
<td>Type of fixation</td>
<td>Central, Eccentric</td>
</tr>
<tr>
<td>Refractive error (diopters, D)</td>
<td>Mild hyperopia &lt;3.00, Moderate hyperopia 3–6, High hyperopia &gt;6, Myopia (same as for hyperopia), Astigmatism</td>
</tr>
<tr>
<td>Associated conditions</td>
<td>Overaction of inferior oblique muscles, Dissociated vertical deviation, Torticollis, Nistagmus</td>
</tr>
<tr>
<td>Deviation angle (prism dipters, PD)</td>
<td>&lt;10, 10–20, 21–30, 31–40, 41–50, &gt;50</td>
</tr>
<tr>
<td>Type of surgery</td>
<td>Recession of both medial rectus muscles, Monocular surgery (recession medial rectus muscle, resection lateral rectus muscle)</td>
</tr>
</tbody>
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**Table 1: Definition of variables**
Techniques and procedures Each case’s clinical record was reviewed to confirm that the following procedures had been carried out:

- General ophthalmological examination: date, duration of deviation, age at onset, associated symptoms and ocular examination of both eyes; slit-lamp examination of the anterior segment (cornea, iris, pupil and lens);
- Ocular motility examination: deviation angle in primary-gaze position with Hirschberg method; cover test; ocular movements (ductions and versions); uncorrected and best corrected VA; ocular dominance test in preverbal children aged <2 years; Kay Picture test for children 2–5 years; and Snellen E test and letter and number charts for those >5 years;[18]
- Motor function study, determining deviation angle with distance and near prisms, using the Krimsky method if VA by prism cover test ≥0.4; or, modified Krimsky if VA<0.4 or children did not cooperate during the exam;[18]
- Study of sensory function based on uncorrected and best corrected VA;
- Dilation of both pupils three times with 1% cyclopentolate, at 5-minute intervals; after 60 minutes, with paralyzed accommodation to define type of fixation and inspect fundus with binocular indirect ophthalmoscope; and
- Surgical treatment for bilateral symmetric cases according to the American Academy of Ophthalmology guidelines, which describe extent of medial rectus recession for given deviation angles; for unilateral cases, monocular recession-resection, selecting appropriate muscle length.[14]

Ethical considerations The study was approved by the ICO Ethics Committee. Data were obtained exclusively from clinical records, maintaining patient anonymity.

Data analysis Primary data were processed using Excel 2003. Qualitative variables were statistically described using absolute and relative frequencies, percentages calculated for the latter. Data were organized in tables and figures.

RESULTS

The largest group of congenital esotropia cases referred to the ICO for evaluation consisted of children aged <2 years (38.6% of total) (Figure 1).

Before surgery, 37% of cases presented mild amblyopia; 22.8% moderate; 12.6 % severe; and 27.6% did not cooperate in VA testing (children <3 years old). By far the most common type of fixation was central (91.3%); 8.7% had eccentric fixation. The most frequent refractive error was mild hyperopia (65.4%), followed by moderate hyperopia (29.1%) and myopia (5.5%).

The most common comorbidity was overaction of the inferior oblique muscles (48%); the least frequent was dissociated vertical deviation (8.7%). No comorbidities were found in 26.8% of cases (Figure 2).

DISCUSSION

Clinical records showed most patients received early medical attention, providing opportunity for appropriate management. Hence, the majority of cases in the present study were under 2 years old. Beginning occlusive treatment at 6 months of age and correcting refractive errors promotes the best possible visual conditions for surgery before 24 months.

While authors still disagree on optimal age for surgery, Prieto Díaz and others suggest that the earlier ocular alignment is achieved, the better the functional result attained.[12] He studied 171 surgically-aligned cases aged >4 years. Of these, 123 were selected because they fully cooperated: 108 (87.8%) showed fusion (assessed using Bagolini striated lenses); and 92 (74.8%) a degree of stereopsis (assessed by Titmus test). He observed close association between fusion and stereopsis and age at alignment: all 76 cases that were aligned between 8 and 17 months of age showed fusion and stereopsis, while of the 16 aligned after 36 months of age, only 8 showed fusion and none showed stereopsis.

There is a period of sensory plasticity in the first 18 months of life during which subnormal or anomalous binocular vision may still develop fully, the potential gradually decreasing thereafter until around 36 months.[12] Helveston held that 4 months old
was the earliest age for congenital esotropia surgery, if diagnostic certainty was possible.[11] Yet, when he presented 10-year follow-up results on 12 cases operated on at about that age, he found no better results than for those who received surgery at 12 months of age. Early alignment produces postoperative esotropia with a small deviation angle (ortho ± 10 PD), or, exceptionally, orthotropia.[12] In the present study, surgical alignment was satisfactory in a high percentage of children aged <2 years.

Preoperative, most cases presented mild amblyopia, consistent with other reports of amblyopia in approximately 40–50% of children with congenital esotropia.[19,20] This is likely due to the presence of fixation micro-nystagmus causing visual maturation delay, because the fovea does not receive stable images. However, this deficit usually improves gradually over time.[19]

The majority of cases (91.3%) had central or foveal fixation, corresponding to the mild degree of amblyopia found. In congenital esotropia, patients may alternate fixation or show a fixation preference for one eye. A strong fixation preference indicates significant amblyopia and should be treated by patching the dominant eye before strabismus surgery, as done at ICO.

Mild hyperopia was the most frequent refractive error, consistent with the literature.[12,13,19,20] If congenital esotropia patients have hyperopia, it tends to be moderate (around +2.50 D) and does not influence deviation angle. According to Prieto Diaz, in a study of 256 congenital esotropia cases, only 14.4% (37 cases) showed ≥2.50 D spherical hyperopia.[12] Other studies reveal that these patients' refractive errors are similar to those in the general population and suggest that in cases where hyperopia is >3.00 D, it should be carefully assessed, since it could result in overcorrection after esotropia surgery.[1,3]

In this study, the ocular disorder most commonly associated with congenital esotropia was overaction of inferior oblique muscles, consistent with standard texts indicating that this is more frequent in congenital cases than in those of later onset (acquired comitant esotropia).[12,13] In a study of 120 consecutive cases of congenital esotropia, 70% showed overaction of the oblique muscles, 46.7% of the inferior obliques. Primary overaction of the inferior oblique muscle was found in 72% of patients, sometimes upon first examination and sometimes later. Overaction of the superior oblique muscle is less common.[12]

More recently it has been stated that in children with esotropia and loss of fusion, normal vergence control mechanisms are lost. The neural impulse favors extorsion over intorsion, leading to progressively increased tone in the inferior oblique muscle with secondary shortening and reduction in tone of the superior oblique with secondary elongation, similar to what happens in sensory esotropia.[5,21]

In the present study, the most common surgical technique was recession of both medial rectus muscles, in line with other authors.[5,12–14,19,21,22] This is the most frequently used technique, with the extent of recession determined by the deviation angle.[5,12,14] We prefer this technique to recession-resection because its symmetry allows angle correction without causing inconstancy on lateroversion and because it simultaneously corrects the bilateral motor abduction imbalance characteristic of these children. All clinical phenomena (crossed fixation, abduction limitation, optokinetic nystagmus) were symmetrical and bilateral, and in a high percentage preoperative examinations are also symmetrical (alignment test under general anesthesia 74%; passive duction test 71%; test for muscle elongation 79%), so we prefer to use symmetrical rather than asymmetrical surgical correction.[5,19,21]

In this study, preoperative deviation angle in most cases was in 31–40 PD range, consistent with other reports.[12,19–22] One year after surgery, 64.8% of cases had achieved alignment, similar to results obtained by Helveston.[23] Cases that maintained deviation angles of 10–20 PD were considered functionally and aesthetically acceptable, not requiring a second surgery. These showed continued spontaneous improvement in deviation angle with corrective lenses for moderate hyperopia. All this is consistent with results of other studies assessing evolution at 6 months to one year, reporting success rates of 70–80%.[20,22–24]

A pharmacological alternative to surgery now exists for congenital esotropia, used as first line treatment in children aged <18 months who do not have important vertical deviations. It involves injecting the medial rectus muscles with botulinum toxin (Botox)
to reduce deviation angle. This is done in several sessions, generally once every three months. According to studies reviewed, the treatment has obtained very good horizontal results.[12,13,25] Other authors such as Helveston consider that, if general anesthesia is administered to expose the muscle for Botulin injection, the muscle could well be weakened the traditional way with only a few more minutes of surgery. Helveston contends that this drug has an important but limited role, given that most studies show the need for multiple injections to maintain the effect.[23]

We have no direct experience with botulinum toxin for congenital esotropia. At this point, we share Helveston’s opinion and consider that the treatment of choice is still conventional surgery, which results in more stable alignment in most cases and avoids repeated invasive procedures, with the attendant risks of general anesthesia.

A limitation of this study is that it was too short to permit assessment of visual outcome—VA, fusion, and stereopsis—in relation to age at alignment. This exploration of binocularity could be the objective of further research including more cases from the country’s pediatric hospitals and special schools.[26] Ideally, these institutions should participate in determining prevalence of congenital esotropia in Cuba and assessing treatment results.

CONCLUSIONS

This study of surgical treatment of congenital esotropia revealed success rates in Cuba similar to those seen internationally, with satisfactory rates of orthotropia one year postoperatively. Further study involving follow-up of these and additional cases is recommended to verify the relation between age at alignment and sensory results obtained.

REFERENCES

Erratum

Page 18, Abstract, Methods, first sentence should read: "A descriptive, retrospective study was carried out through review of 127 cases (aged 1–18 years, 55 male and 72 female) operated on for congenital esotropia between January 2008 and May 2009."