NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

INTERVENTIONAL PROCEDURES PROGRAMME

Interventional procedure overview of tenotomy of four horizontal eye muscles for nystagmus

Nystagmus is the involuntary movement of the eyes (most commonly from side-to-side) usually associated with impaired vision. In this procedure, the horizontal muscles of the eye (which move the eye side-to-side) are cut and either re-attached at the same place or re-attached in a position further back on the eyeball to address squint or abnormal head posture. The aim of the procedure is slow the involuntary eye movements and improve vision.

Introduction

This overview has been prepared to assist members of the Interventional Procedures Advisory Committee (IPAC) in making recommendations about the safety and efficacy of an interventional procedure. It is based on a rapid review of the medical literature and specialist opinion. It should not be regarded as a definitive assessment of the procedure.

Date prepared

This overview was prepared in June 2008.

Procedure name

- Tenotomy of horizontal eye muscles for nystagmus.

Specialty societies

- Royal College of Ophthalmologists.

Description

Indications and current treatment

Nystagmus is an involuntary oscillatory movement of the eyes, usually from side to side, but sometimes the eyes move up and down or in a circular motion.

Most people with nystagmus have vision which is much worse than average.

Nystagmus may be associated with an abnormal head turn because most patients with nystagmus have a position of gaze where the nystagmus is least and therefore vision is best (known as a null zone). For example, if the null zone is when the patient gazes to the right, then they may turn their face to the left to place the eyes in the position of right gaze. Patients with nystagmus may also have a condition called strabismus (also known as squint or heterotropia). These terms describe misalignment of the eye caused by an imbalance in the ocular muscles.

There are a variety of types of nystagmus and no consistent classification system to describe them. Nystagmus that is present at birth or which develops in the first 6 months is usually called congenital or infantile nystagmus. This may be caused by a defect in the eye or the visual pathway from the eye to the brain. Congenital/infantile nystagmus occurs in a wide range of eye disorders of childhood such as cataract, glaucoma, some disorders of the retina, and albinism. It may also be found in children who have multiple disabilities, such as Down's syndrome. Many children with nystagmus have no eye, brain or other health problems. In this case the condition is called 'congenital idiopathic nystagmus' or 'idiopathic infantile nystagmus'.

Acquired nystagmus, which develops later in life, may be a symptom of a variety of other conditions including stroke, multiple sclerosis or head injury.

There is currently no curative treatment for nystagmus. Treatments that have been tried include drugs such as anti-epileptics and muscle relaxants, injections of botulinum toxin and biofeedback techniques (making the patient aware of the eye movement using visual and audio signals and encouraging them to control it). Lenses may be worn to improve visual acuity but these do not correct the nystagmus.

What the procedure involves

Tenotomy for nystagmus is carried out with the patient under general anaesthesia, and involves the two horizontal rectus muscles (lateral and medial) of each eye. A limbal incision is made in the conjunctiva and the muscle is detached from the sclera. A small piece of the muscle may be removed. The muscle is either reattached at its original place of insertion or to a position posterior to the equator of the globe (retroequatorial recession).

The exact surgical technique, particularly the extent of recession (if any), depends on ocular alignment and the presence or absence of abnormal head posture and strabismus. For example, in a patient with a left gaze null point and compensatory head turn to the right, the aim of surgery is to move the eyes to the right. This would be achieved by recessing the left lateral rectus to a greater extent than the right medial rectus.

The aims of the procedure are to reduce the frequency (how often the eyes oscillate) and amplitude (how far the eyes move) of nystagmus by weakening

the appropriate eye muscles, to improve visual acuity and to correct abnormal compensatory head posture if present.

Efficacy

This overview is based on approximately 180 patients from eight case series.

Other studies that were considered to be relevant to the procedure but were not included in the main extraction table (table 2) have been listed in appendix A.

Head posture

In a case series of 21 patients who underwent recession of horizontal eye muscles, 95% (20/21) had both an improvement in abnormal head turn and abnormal head tilt (to the left or right shoulder) at follow-up (mean: 19 months); however, 5 patients (24%) had a small overcorrection in head turn. All patients had improved head tilt (to the left or right shoulder) [1].

Two case series of 20 and 12 patients reported improved head posture in all patients who had abnormal head posture pre-operatively (n = 7 and 4 respectively) [2,3]. In another case series of 12 patients, mean head turn improved from 33° pre-operatively to 7° after 1 week (p = 0.001) and 5° after 3 months (p-value not stated). In this study, 6 patients had no residual head turn and 1 patient had more than 15° of residual head turn after the procedure [4].

A third case series of 12 patients reported that three patients had an improvement in the amount of head turn postoperatively, two patients had no change, and one patient acquired abnormal head posture after the procedure that was not present preoperatively (data for the remaining patients were not reported) [5].

In a case series of 10 patients, in which 5 patients had abnormal head posture pre-operatively, 3 patients improved and 2 had no change in head posture after the procedure [6].

Visual acuity (assessed by Snellen chart)

In the case series of 20 patients, 2 had a 3-line increase in visual acuity, 8 had a 2-line increase, 3 had a 1-line increase, 2 had no change in visual acuity and 2 patients had a decrease (mean follow-up: 12 months). Three patients were too young to be assessed with a Snellen chart [2].

A case series of 18 patients reported that 50% (9/18) had a 1-line increase in visual acuity after the procedure (follow-up assessments at 2 weeks and 3 months). One patient in this study had a deterioration of both near and distance visual acuity, but after 2 years this returned to pre-operative levels [7].

A case series of 12 patients reported that 58% (7/12) had at least a 2-line increase and 3 had a 1-line increase in visual acuity at follow-up (mean: 13 months). One patient in this study lost 1 line of visual acuity [3].

In another case series of 12 patients, 75% (9/12) had an improvement (1 to 3 lines) in visual acuity at follow-up (mean: 16 months) [5].

Another case series of 10 patients reported that 87% (7/8) of patients who were assessed had an average improvement in visual acuity of 1 line. Two patients were too young to be assessed with a Snellen chart [6].

In a further case series of 12 patients, mean binocular decimal visual acuity (where a value of 1.0 is equal to 20/20 vision or 8 Snellen lines) in the primary position improved from 0.30 at baseline to 0.47 after 1 week ($p = 0.011$) and 0.46 after 3 months (p-value not stated) [4].

Visual acuity (assessed by Early Treatment Diabetics Retinopathy Study [ETDRS] chart)

A case series of 75 patients reported a significant improvement in visual acuity from 0.6 pre-operatively to 0.4 post-operatively ($p = 0.001$). In the same study, all patients showed increased gaze-dependent visual acuity (assessed with the patient's head fixed at different angles horizontally) ($p < 0.001$) [8].

Null zone width

The case series of 75 patients reported that the mean largest width of the null zone (position of gaze where nystagmus is least) improved from 6.9 ± 2.6° before the procedure to 23.0 ± 7.0° post-operatively ($p < 0.001$) [8].

Patient satisfaction

Two studies assessed levels of patient satisfaction with the procedure. In one, 78% (14/18) of patients replied to a mailed questionnaire: 8 were pleased with the procedure, 4 were indifferent and 2 were displeased with it. Seven patients thought there was no change in their sight, 3 thought it was slightly better and 1 thought it was worse. Eight patients stated that the procedure had not changed their daily life and 4 reported some improvement in daily activities [7]. In a case series of 12 patients, 83% (10/12) reported that they had better control of nystagmus postoperatively [5].

Safety

Six case series of 75, 21, 20, 12, 12 and 10 patients reported that no complications occurred [1,2,3,5,6,8].

In the case series of 18 patients, there were four complications. Two patients developed exotropia (divergent strabismus), one patient had asthenopic symptoms (eye strain, fatigue and redness) (this patient also had some deterioration of visual acuity after the procedure) and there was one case of scleral perforation during the procedure (with no retinal perforation, and no

treatment was required). All 18 patients reported severe pain in the first 24 hours after the procedure and still had discomfort at the 2-week follow-up [7].

One case series reported that all 12 patients had mild limitation of horizontal eye movement in the direction of muscle recession 1 week after the procedure. After 3 months, 8 patients still had some limitation and 4 had no or minimal limitation [4].

One patient in a case series of 12 acquired abnormal head posture after surgery that had not been present pre-operatively [5].

Literature review

Rapid review of literature

The medical literature was searched to identify studies and reviews relevant to tenotomy of horizontal eye muscles for nystagmus. Searches were conducted of the following databases, covering the period from their commencement to 14/05/08: MEDLINE, PREMEDLINE, EMBASE, Cochrane Library and other databases. Trial registries and the Internet were also searched. No language restriction was applied to the searches (see appendix C for details of search strategy).

The following selection criteria (table 1) were applied to the abstracts identified by the literature search. Where selection criteria could not be determined from the abstracts the full paper was retrieved.

Table 1 Inclusion criteria for identification of relevant studies

Characteristic	Criteria
Publication type	Clinical studies were included. Emphasis was placed on identifying good quality studies. Abstracts were excluded where no clinical outcomes were reported, or where the paper was a review, editorial, or a laboratory or animal study. Conference abstracts were also excluded because of the difficulty of appraising study methodology, unless they reported specific adverse events that were not available in the published literature.
Patient	Patients with nystagmus.
Intervention/test	Tenotomy of horizontal eye muscles.
Outcome	Articles were retrieved if the abstract contained information relevant to the safety and/or efficacy.
Language	Non-English-language articles were excluded unless they were thought to add substantively to the English-language evidence base.

Existing assessments of this procedure

There were no published assessments from other organisations identified at the time of the literature search.

Related NICE guidance

There is currently no NICE guidance related to this procedure.

Table 2 Summary of key efficacy and safety findings on tenotomy of four horizontal eye muscles for nystagmus

Abbreviations used: ETDRS; Early Treatment Diabetic Retinopathy Study, LogMAR; Log of the Minimal Angle of Resolution

Study details	Key efficacy findings	Key safety findings	Comments
Hertle et al (2006) [8] Study type: **prospective case series** Country: USA Study period: not stated Study population: patients with infantile nystagmus syndrome. Some patients had coexisting strabismus (68%), clinically significant abnormal head posture (48%) and associated sensory system deficit in addition to ocular motor disorder of infantile nystagmus syndrome (56%). n = **75** Age: 36 years (mean) Sex: 70% male Inclusion criteria: • clinical and electrophysiological diagnosis of infantile nystagmus syndrome • best-corrected binocular visual acuity ≥ 20/200 Exclusion criteria: • prior extraocular muscle surgery performed by the authors Technique: surgery on four horizontal rectus muscles. Patients either had surgery for strabismus alone (n = 14), eccentric null position alone (n = 33), or strabismus plus eccentric null position (n = 15). Variable amounts of recession, resection and transposition were performed in the above patients. Patients with nystagmus alone (n = 13) had tenotomy at tendon-scleral junction and immediate reattachment at the original insertion. Follow-up: **18 months (mean)** Conflict of interest: none stated	***Null zone visual acuity*** *- assessed by ETDRS chart binocularly and monocularly (reported as LogMAR)* • Mean pre-operative visual acuity: 0.6 • Mean post-operative visual acuity: 0.4 (p = 0.001) • Mean post-operative increase in visual acuity was 1 Snellen line equivalent for patients older than 8 years (n = 45) and 2 Snellen lines equivalent for those 8 years old and under (n = 30) ***Gaze dependent visual acuity*** *- assessed by ETDRS chart with patient's head fixed at different angles horizontally* • All patients showed significantly increased visual acuity (decreased LogMAR scores) (p < 0.001) ***Null zone width*** *- defined oculographically (under binocular conditions and using data from the preferred eye) as the expanse of gaze (in degrees) where the nystagmus intensity and foveation characteristics were within 20% of the primary null zone characteristics* • Mean largest pre-operative null zone width: 6.9 ± 2.6° • Mean largest post-operative null zone width: 23 ± 7° (p < 0.001) • There were no statistically significant differences between the four surgical groups.	No safety outcomes were reported.	All patients had infantile nystagmus syndrome associated with motor deficit. Half of patients also had sensory deficit. Nearly half of patients had abnormal head posture. Some patients had simple tenotomy of muscles with reattachment at the original insertion and others had recession. The Early Treatment Diabetic Retinopathy Study (ETDRS) chart is similar to the Snellen chart but has 5 letters in each row. The traditional Snellen chart has 1 letter in the first row and increasing numbers of letters in subsequent rows. Visual acuity (measured by Snellen or EDTRS charts) is often reported as the Log of the Minimum Angle of Resolution (LogMAR) for the purposes of statistical analysis.

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Arroyo-Yllanes et al (2002) [1] Study type: **case series** Country: Mexico Study period: Sept 1990 – July 1999 Study population: patients with horizontal nystagmus with a neutral zone and abnormal head posture in all 3 axes with, or without, strabismus. n = **21** Age: 18 years (median) Sex: 81% male Inclusion criteria: • neutral zone and abnormal head position in all three axes with predominance of head turn in the vertical axis Exclusion criteria: • congenital pendular nystagmus • nystagmus secondary to organic lesions such as albinism, macular scars neurological disorders and vertical-associated strabismus Technique: horizontal surgery using a modified Anderson procedure: 2mm retroequatorial recession of the horizontal yoke rectus muscles responsible for the compensatory head position. Additional strabismus surgery was performed where required. Follow-up: **19 months (mean)** Conflict of interest: none stated	***Head posture*** *- positioning of head turn to the left or right, head tilt to the left or right shoulder and chin elevation or depression measured in degrees* *- assessed at 1 week, 1 month and every 6 months.* • Improved head turn: 95% (20/21). One patient had no improvement in head turn which was the result of paralysis of a cranial nerve that had not been detected at pre-operative evaluation • 5 patients (24%) had overcorrection of head turn (although not sufficient to warrant further surgery) • Improved head tilt: 100% • Improved chin elevation/depression: 95% (20/21) (one patient had no change post-operatively)	No safety outcomes were reported.	The authors do not specify what types of nystagmus patients had (for example, idiopathic motor or sensory). All patients had abnormal head posture. All patients had complete follow-up to at least 6 months.

Abbreviations used: LogMAR; Log of the Minimal Angle of Resolution			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Bagheri et al (2005) [2] Study type: **retrospective case series** Country: Iran Study period: Jan 1994 – Dec 1999 Study population: patients with congenital nystagmus associated with low visual acuity due to sensory deficit (80%) or motor deficit (20%) with or without strabismus or abnormal head posture. Diagnosis was established by clinical examination and electroretinography if required. n = **20** Age: 15 years (mean) Sex: 45% male Exclusion criteria: • previous strabismus surgery Technique: recession of 4 horizontal rectus muscles (9–21mm). At least 2 muscles were placed posterior to the equator. Follow-up: **12 months (mean)** Conflict of interest: none stated	***Head posture*** • 39% (7/18) of patients had abnormal head posture pre-operatively (not recorded in 2 patients) and all patients improved after the procedure (4 patients had complete improvement and 3 had partial improvement) ***Visual acuity*** *- assessed by Snellen chart* • 3-line increase in visual acuity: 12% (2/17). • 2-line increase in visual acuity: 47% (8/17). • 1-line increase in visual acuity: 18% (3/17). • No change in visual acuity: 12% (2/17). • Decrease in visual acuity: 12% (2/17). *Data do not include 3 young patients in whom visual acuity was estimated by an eye fixation pattern instead of using a Snellen chart.* • Binocular distance visual acuity (LogMAR) improved from 0.73 ± 0.26 at baseline to 0.62 ± 0.32 post-operatively (p = 0.02). • The greatest improvement was in patients with congenital motor nystagmus (all 4 patients had at least a 2-line increase in visual acuity). • 75% of patients reported subjective improvement in vision after surgery.	No safety outcomes were reported.	All patients had congenital nystagmus (most had sensory deficit; fewer had a motor deficit). Seven patients had abnormal head posture. 24 patients underwent the procedure however 4 were excluded from the study because of incomplete follow-up.

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Boyle et al (2006) [7] Study type: **retrospective case series** Country: UK Study period: 1997 – 2002 Study population: patients with congenital idiopathic nystagmus. Three patients had additional strabismus. n = **18** Age: 32 years (mean) Sex: 67% male Exclusion criteria: • nystagmus associated with non-idiopathic pathology Technique: recession of 4 horizontal rectus muscles (8–12mm) and placed posterior to the equator. Three patients had asymmetrical recession to correct strabismus. Follow-up: **not stated (postoperative assessment at 2 weeks and 3 months)** Conflict of interest: none stated	***Head posture*** • no patients showed significant abnormal head posture pre-operatively by direct observation ***Distance visual acuity*** *- assessed by Snellen chart* • 50% (9/18) had a 1-line increase in visual acuity ***Near visual acuity*** *- assessed by N series measurements (no further information about this method of measurement was stated)* • No objective change in near visual acuity ***Ocular movements*** • All patients had iatrogenic limitation of each eye in adduction • 16 patients had slight limitation in abduction ***Patient satisfaction*** *- assessed by mailed questionnaire (78% response rate)* • 8 patients were pleased with the procedure, 4 were indifferent and 2 were displeased with the procedure • 7 patients thought there was no change in their sight, 1 thought their sight was slightly worse • 2 patients felt their nystagmus was slightly calmer, 3 described a minor improvement in visual acuity • 8 patients said the operation had not changed their daily life,1 reported a slight improvement and 1 felt that looking at a computer screen was easier, 2 patients reported an increase in confidence and in maintaining eye contact	***Complications*** • 1 patient had scleral perforation during the procedure (with no retinal perforation). No treatment was required. The patient also decompensated to a manifest divergent squint and continued to undergo regular botulinum toxin injections to maintain alignment. • 2 patients developed exotropia (divergent strabismus) • 1 patient had asthenopic symptoms (this patient also had some deterioration of visual acuity after the procedure). • All patients complained of severe pain in the first 24 hours and were still complaining of discomfort at the 2-week follow-up. • 1 patient had a deterioration in distance visual acuity from 6/24 to 6/36 and in near visual acuity from N10 to N14 but after 2 years this settled back to preoperative levels	All patients had idiopathic nystagmus. No patients had abnormal head posture.

Abbreviations used: ETDRS; Early Treatment Diabetic Retinopathy Study

<table>
<tr><th>Study details</th><th>Key efficacy findings</th><th>Key safety findings</th><th>Comments</th></tr>
<tr>
<td>Gupta et al (2006) [4]
Study type: prospective case series
Country: India
Study period: not stated
Study population: patients with idiopathic infantile nystagmus.
n = 12
Age: 11 years (mean)
Sex: 75% male
Inclusion criteria:
• older than 5 years
• eccentric null position with anomalous head position > 15°

Exclusion criteria:
• previous extraocular muscle surgery or strabismus
• other causes of nystagmus

Technique: modified Anderson procedure - recession of the horizontal rectus yoke muscles (9mm recession of medial rectus muscle or 12 mm recession of lateral rectus muscle depending on direction of head turn.

Follow-up: at least 3 months

Conflict of interest: none stated</td>
<td>Head posture
<table>
<tr><th></th><th>Baseline</th><th>1 week</th><th>1 month</th><th>3 months</th></tr>
<tr><td>Mean head turn (degrees)</td><td>32.5 (± 5.8)</td><td>7.0 (± 6.8) *</td><td>7.0 (± 6.8)</td><td>5.0 (± 8.7)</td></tr>
</table>
* p = 0.0001 compared to baseline
• 6 patients had no residual head turn
• 1 patient had over 15° of residual head turn.

Visual acuity
- assessed by Snellen and ETDRS charts
<table>
<tr><th>Mean values</th><th>Baseline</th><th>1 week</th><th>1 month</th><th>3 months</th></tr>
<tr><td>Mean binocular decimal visual acuity in the primary position (Snellen)</td><td>0.30 (± 0.18)</td><td>0.47 (± 0.30) [1]</td><td>0.47 (± 0.30)</td><td>0.46 (± 0.30)</td></tr>
<tr><td>Mean binocular letter visual acuity in the primary position (ETDRS)</td><td>50.9 (± 16.4)</td><td>60.0 (± 15.9) [2]</td><td>59.8 (± 15.1)</td><td>60.2 (± 15.8)</td></tr>
</table>
[1] p = 0.011 compared to baseline
[2] p = 0.007 compared to baseline

Eye movement
- assessed by electronystagmography
<table>
<tr><th></th><th>Baseline</th><th>3 months</th><th>p-value</th></tr>
<tr><td>Mean nystagmus amplitude (deg)</td><td>6.3 (± 3.3)</td><td>3.9 (± 3.0)</td><td>0.022</td></tr>
<tr><td>Mean nystagmus frequency (cycles/sec)</td><td>3.4 (± 0.8)</td><td>2.1 (± 1.3)</td><td>0.004</td></tr>
<tr><td>Mean nystagmus intensity (degree cycles/sec)</td><td>22.0 (± 15.9)</td><td>10.6 (± 10.2)</td><td>0.006</td></tr>
</table>
</td>
<td>• All patients had mild limitation of horizontal eye movement in the direction of muscle recession 1 week postoperatively. At 3 months, 8 patients still had mild limitation and 4 had no/minimal limitation.</td>
<td>All patients had idiopathic congenital nystagmus.
All patients had abnormal head posture.</td>
</tr>
</table>

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Davis et al (1997) [3] Study type: **retrospective case series** Country: USA Study period: not stated Study population: patients with ocular or oculocutaneous albinism demonstrating nystagmus. Three patients had additional strabismus. n = **12** Age: 20 years (mean) Sex: 83% male Technique: large retroequatorial recessions of 4 horizontal rectus muscles. 9 patients had symmetrical 4-muscle surgery and 3 patients with strabismus required additional compensatory adjustment for strabismus. Follow-up: **13 months (mean)** Conflict of interest: none stated	***Head posture*** • Of the 4 patients who had a head turn pre-operatively, 3 had none post-operatively and 1 had an improved head turn post-operatively. ***Best-corrected visual acuity*** *- assessed by Snellen chart* • > 1-line increase in monocular or binocular visual acuity: 83% (10/12). • > 2-line increase in monocular visual acuity: 58% (7/12).	• 1 patient lost 1 line of visual acuity. • 3 patients required further surgery for postoperative strabismus during follow-up.	All patients had nystagmus secondary to ocular albinism. Four patients had abnormal head posture.

Abbreviations used:

<table>
<tr><th>Study details</th><th>Key efficacy findings</th><th>Key safety findings</th><th>Comments</th></tr>
<tr>
<td>Atilla et al (1999) [5]
Study type: prospective case series
Country: Turkey
Study period: not stated
Study population: patients with sensory nystagmus secondary to albinism, dyschromatopsia and degenerative myopia (n = 8) or congenital motor nystagmus (n = 4). Four patients had additional strabismus.
n = 12
Age: 8 years (mean)
Sex: 75% male

Technique: recession surgery (7–10mm) on all four horizontal rectus muscles. Patients with strabismus had asymmetrical muscle recessions.

Follow-up: 16 months (mean)

Conflict of interest: none stated</td>
<td>Head posture
- patients fixated on a target and the angle between the target and head deviation was measured with an orthopaedic ruler
• 3 patients had an improvement in the amount of head turn post-operatively.
• 2 patients had no change in head posture post-operatively.
No further information about head posture is provided. It is not clear whether all 12 patients had abnormal head posture preoperatively or just these five.. .

Visual acuity
- assessed by Snellen chart
• binocular visual acuity improved post-operatively in 9 patients, did not change in 2 patients and decreased in 1 patient.

<table>
<tr><th></th><th>Baseline</th><th>Postoperative</th><th>p-value</th></tr>
<tr><td>Congenital nystagmus (n = 4)</td><td>0.44 (± 0.09)</td><td>0.66 (± 0.11)</td><td>p < 0.001</td></tr>
<tr><td>Sensory (secondary) nystagmus (n = 8)</td><td>0.16 (± 0.09)</td><td>0.22 (± 0.14)</td><td>p < 0.01</td></tr>
</table>

Eye movement
- assessed by electronystagmography
<table>
<tr><th></th><th>Baseline</th><th>Postoperative</th><th>p-value</th></tr>
<tr><td>Mean nystagmus amplitude (deg)</td><td>7.8 (± 2.9)</td><td>3.9 (± 2.4)</td><td><0.01</td></tr>
<tr><td>Mean nystagmus frequency (cycles/sec)</td><td>3.9 (± 1.5)</td><td>3.5 (± 1.3)</td><td><0.05</td></tr>
<tr><td>Mean nystagmus intensity (degree cycles/sec)</td><td>28.9 (± 11.3)</td><td>12.7 (± 6.3)</td><td><0.01</td></tr>
</table>

Patient satisfaction
83% (10/12) of patients reported that they had better control of nystagmus after the procedure.</td>
<td>Complications
• 1 patient acquired head posture after surgery that was not present pre-operatively.</td>
<td>Patients had various nystagmus pathologies (most had sensory nystagmus secondary to ocular disorders).</td>
</tr>
</table>

Abbreviations used:			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Helveston et al (1991) [6] Study type: **case series** Country: USA Study period: Feb 1988 – May 1990 Study population: patients with congenital motor nystagmus (n = 5), oculocutaneous albinism (n = 2), and optic nerve hypoplasia (n = 2). Six patients had additional strabismus. n = **10** Age: 18 years (mean) Sex: not stated Inclusion criteria: not stated Technique: recession of four horizontal rectus muscles (11–13mm). Muscles were placed at or just behind the equator. Three patients with strabismus had asymmetrical muscle recessions. Follow-up: **11 months** Conflict of interest: none stated	***Head posture*** • Of 5 patients who had an abnormal head posture preoperatively, 3 improved after the procedure and 2 were unchanged. ***Visual acuity*** *- assessed by Snellen chart* • Improved visual acuity: 7/8 patients (data do not include 2 young patients who could not be assessed using a Snellen chart). • Average improvement in visual acuity was 1 line (near and/or distance vision) Eye movement • Improved nystagmus amplitude: 8/10 patients	***Complications*** No safety outcomes were reported.	Patients had a mix of nystagmus pathologies (most had congenital motor nystagmus). Five patients had abnormal head posture. One patient was lost to follow-up.

Validity and generalisability of the studies

Indications

- Patients included in the studies had a range of types of nystagmus. Some studies included patients with idiopathic congenital nystagmus and others included patients with nystagmus secondary to ocular albinism.
- Some patients had nystagmus only and others had both nystagmus and strabismus.
- Abnormal head posture was an inclusion criterion in some studies and in others no patients had abnormal head posture.
- Some authors suggested that the procedure should be restricted to patients with congenital motor nystagmus however the studies report on patients with both sensory and motor congenital nystagmus.
- There was variation in the age of patients (mean age ranged from 10 to 32 years).

Procedure

- The procedures varied. All but one study (Hertle and Yang 2006) performed recession of the muscles (with varying amounts of recession 2–21mm). Thirteen patients in Hertle and Yang (2006) underwent simple tenotomy with reattachment at the original insertion point (that is, no recession).

Outcomes

- The primary outcomes varied: some studies focused on visual acuity, others on abnormal head posture and some on eye movement recordings.
- There is potential for bias in results where clinician assessment of visual acuity testing is done (most of the studies).
- Sparse details of patient reported outcomes are provided.
- A few studies of tenotomy of eye muscles for acquired nystagmus and of an equivalent procedure (on vertical muscles) for vertical nystagmus were identified in the literature search however these were not included.
- Four studies did not report safety outcomes whilst three only reported those relating to efficacy outcomes, it is not clear that this is because no safety issues were raised by the studies.

Specialist Advisers' opinions

Specialist advice was sought from consultants who have been nominated or ratified by their Specialist Society or Royal College. The advice received is their individual opinion and does not represent the view of the society.

Irene Gottlob, John Lee, Isabelle Russell-Eggit, Robert H Taylor (Royal College of Ophthalmologists).

- One Specialist Adviser had performed this procedure at least once and three had not performed it (one sees patients who would potentially benefit and another intends to start performing it this year).
- Two Specialist Advisers thought it was novel (however one stated that the procedure was established but that this was a different concept) and two thought it was a minor variation of an established technique (similar to strabismus surgery but for a new indication).
- Three Advisers stated that there is no direct comparator.

Safety

- Theoretical adverse events included: loss of vision, retinal damage, need for revision surgery, damage to the eye, redness, swelling, diplopia, infection, perforation of globe, slipped muscles, worse eye alignment, induced strabismus and loss of vision.
- One Adviser stated that development of a conjunctival cyst had been reported in the literature.
- Three Specialist Advisers stated that there are no uncertainties about the safety of the procedure.

Efficacy

- Efficacy outcomes included: best corrected binocular visual acuity under varying gaze angles, null point width, stereoacuity, ocular movement recordings, nsystagmus, visual function in day-to-day life, quality of life, cosmesis and head posture.
- One Specialist Adviser thought that the main uncertainty with the procedure was around the difficulty in measuring improvement and standardising viewing angles. Another stated that the procedure produces a small improvement in a measurable feature of ocular movement (foveation time) but there is no consistent improvement in visual acuity assessed by Snellen charts. One Adviser thought that there is uncertainty related to the long-term benefit of the procedure.

Other

- Three Specialist Advisers thought that clinicians should have experience in performing strabismus/eye muscle surgery.
- Three Specialist Advisers thought that the procedure would potentially have a minor impact on the NHS and one thought it would have a moderate impact.
- A few studies reported on vertical eye muscle surgery for vertical nystagmus. These were not included in the overview.

Issues for consideration by IPAC

- Consider alternative titles:
  - Tenotomy of horizontal eye muscles with or without recession
  - Four muscle surgery
  - Horizontal rectus tenotomy with or without recession.
- Few safety concerns were reported.
- Small patient numbers in the evidence base.

References

1. Arroyo-Yllanes ME, Fonte-Vazquez A, Perez-Perez JF (2002) Modified Anderson procedure for correcting abnormal mixed head position in nystagmus. British Journal of Ophthalmology 86:267-269.

2. Bagheri A, Farahi A, Yazdani S (2005) The effect of bilateral horizontal rectus recession on visual acuity, ocular deviation or head posture in patients with nystagmus. Journal of AAPOS: American Association for Pediatric Ophthalmology and Strabismus 9:433-437.

3. Davis PL, Baker RS, Piccione RJ (1997) Large recession nystagmus surgery in albinos: effect on acuity. Journal of Pediatric Ophthalmology and Strabismus 34:279-283.

4. Gupta R, Sharma P, Menon V (2006) A prospective clinical evaluation of augmented Anderson procedure for idiopathic infantile nystagmus. Journal of AAPOS: American Association for Pediatric Ophthalmology and Strabismus 10:312-317.

5. Atilla H, Erkam N, Isikcelik Y (1999) Surgical treatment in nystagmus. Eye 13:11-15.

6. Helveston EM, Ellis FD, Plager DA (1991) Large recession of the horizontal recti for treatment of nystagmus. Ophthalmology 98:1302-1305.

7. Boyle NJ, Dawson EL, Lee JP (2006) Benefits of retroequatorial four horizontal muscle recession surgery in congenital idiopathic nystagmus in adults. Journal of AAPOS: American Association for Pediatric Ophthalmology and Strabismus 10:404-408.

8. Hertle RW and Yang D (2006) Clinical and electrophysiological effects of extraocular muscle surgery on patients with Infantile Nystagmus Syndrome (INS). Seminars in Ophthalmology 21 (2) 103-110.

Appendix A: Additional papers on tenotomy of four horizontal eye muscles for nystagmus

The following table outlines the studies that are considered potentially relevant to the overview but were not included in the main data extraction table (table 2). It is by no means an exhaustive list of potentially relevant studies.

Article	Number of patients/follow-up	Direction of conclusions	Reasons for non-inclusion in table 2
Bagheri A. Farahi A, and Guyton DL (2003) Astigmatism induced by simultaneous recession of both horizontal rectus muscles. Journal of Aapos: American Association for Pediatric Ophthalmology & Strabismus 7 (1) 42-46.	n = 13 Follow-up: not stated	Recession resulted in a statistically significant change in astigmatism in the with-the-rule direction.	Newer study reporting on the same patients included in table 2.
Brodsky MC, Wright KW. (2007) Infantile esotropia with nystagmus: a treatable cause of oscillatory head movements in children. Archives of Ophthalmology 125 (8): 1079.	n = 3 Follow-up: not stated	Ocular alignment was restored Head shaking resolved in all patients. In 1 patient, head shaking accompanied recurrence of the esotropia and again resolved following surgical realignment of the eyes.	Larger studies were included in table 2.
Calhoun JH and Harley RD. (1984) Surgery for abnormal head position in congenital nystagmus. Transactions of the American Ophthalmological Society 71 70-83.	n = 19 Follow-up: not stated	6/19 (32%) patients had no turn while attaining maximum visual acuity (mean follow-up: 5 months). 10/19 (53%) patients showed varying degrees of improvement (some patients had no head turn immediately postoperatively but reverted during follow-up). 2/19 (11%) patients showed no improvement in head turn.	More recent studies were included in table 2.
Datta H and Prasad S. (1994) Postequatorial horizontal rectus recession in the management of congenital nystagmus. Indian Journal of Ophthalmology 42 (4) 203-206.	n = 9 Follow-up: 6–15 months	15/18 eyes showed decreased amplitude of nystagmus. 12/18 eyes showed an increase in visual acuity.	Larger studies were included in table 2.
Erbagci I, Gungor K, and Bekir NA. (2004) Effectiveness of retroequatorial recession surgery in congenital	n = 7 Follow-up: 8 months	5/7 patients had improved visual acuity. All 5 patients who had abnormal head position preoperatively, had decreased head position and	Larger studies were included in table 2.

nystagmus. Strabismus 12 (1) 35-40.		nystagmus intensity.	
Girard LJ, Ghuman T, Donaldson K. (1996) Nystagmus reduction by horizontal rectus retroplacements. Ophthalmic Surgery & Lasers 27 (11): 958.	n = 7 Follow-up: not stated	Nystagmus improved in all seven patients. All patients improved at least 1 line for distance and from 3 to 5 lines for near	Larger studies were included in table 2.
Gradstein L, Reinecke RD, Wizov SS, et al. (1997) Congenital periodic alternating nystagmus. Diagnosis and Management. Ophthalmology 104 (6): 918.	n = 7 Follow-up: not stated	Visual acuity and abnormal head posture improved, at least for several years. Nystagmus parameters (cycle, waveform, frequency, amplitude and velocity) improved in all patients. Mean foveation fractions increased significantly after surgery.	Larger studies were included in table 2.
Hertle RW, Dell'Osso LF, FitzGibbon EJ, et al. (2003) Horizontal rectus tenotomy in patients with congenital nystagmus: results in 10 adults. Ophthalmology 110 (11) 2097-2105.	n = 10 Follow-up: 1 year	9/10 patients had persistent, significant postoperative increases in the expanded nystagmus acuity function of their fixing (preferred). Binocular visual acuity measured with the ETDRS chart increased in 5 patients and was unaffected in 5.	Newer study reporting on the same patients included in table 2.
Hertle RW, Anninger W, Yang D, et al. (2004) Effects of extraocular muscle surgery on 15 patients with oculo-cutaneous albinism (OCA) and infantile nystagmus syndrome (INS). American Journal of Ophthalmology 138 (6): 978-987.	n = 15 (3 for strabismus alone, 3 for nystagmus alone, 5 for eccentric gaze null zone alone, and 4 for eccentric gaze null zone plus strabismus Follow-up: not stated	14/15 patients had improvement in best optically corrected acuity of ≥ 0.1 LogMar. Patients with abnormal head posture improved significantly postoperatively ($p < 0.01$). The expanded nystagmus acuity function, null zone position, null zone width and foveation time showed persistent, significant increases in all patients ($p < 0.01$).	Larger studies were included in table 2.
Hertle RW, Dell'Osso LF, FitzGibbon EJ et al. (2004) Horizontal rectus muscle tenotomy in children with infantile nystagmus syndrome: a pilot study. Journal of AAPOS: American Association for Pediatric Ophthalmology and Strabismus 8 (6) 539-548.	n = 5 Follow-up: 1 year	2 of the 3 patients (for whom this was measured) had persistent, significant postoperative increases in expanded nystagmus acuity function of their fixating eye. Measured binocular visual acuity increased in four patients; the remaining patient had retinal dystrophy.	Newer study reporting on the same patients included in table 2.
Kushner B, Coats D, Kodsi, S et al (2002) A case of	n = 1	Patient had recession surgery for nystagmus and developed	Larger studies were included in table 2.

consecutive exotropia after recession of all four horizontal rectus muscles for the treatment of nystagmus. Binocular Vision and Strabismus Quarterly 17 (4) 304-311.	Follow-up: not stated	exotropia postoperatively. Upon reoperation 'the left medial rectus muscle was found to be essentially lost'. After a further procedure on vertical muscles the condition was greatly improved.	
von Noorden GK, Sprunger DT. (1991) Large rectus muscle recessions for the treatment of congenital nystagmus. Archives of Ophthalmology 109 (2): 221.	n = 3 Follow-up: not stated	2 patients had modest improvement in visual acuity. In 1 patient, a compensatory head turn was eliminated by shifting the neutral zone of the nystagmus to the primary position of gaze.	Larger studies were included in table 2.
Wang Z, Dell'Osso LF, Jacobs J et al. (2006) Effects of tenotomy on patients with infantile nystagmus syndrome: foveation improvement over a broadened visual field. Journal of Aapos: American Association for Pediatric Ophthalmology & Strabismus 10 (6) 552-560.	n = 7 Follow-up: not stated	3/7 patients had a moderate improvement in expanded nystagmus acuity function (13.9-32.6%), 5 had a large improvement (39.9-162.4%), 1 had no change. All patients had reductions in nystagmus amplitudes ranging from 14.6 to 37%.	Larger studies were included in table 2.
Yang MB, Pou-Vendrell CR, Archer SM, et al. (2004) Vertical rectus muscle surgery for nystagmus patients with vertical abnormal head posture. Journal of Aapos: American Association for Pediatric Ophthalmology & Strabismus 8 (4): 299-309.	n = 20 Follow-up: 49 months	Preoperative vertical abnormal head posture ranged from 10 – 45°. After recession alone, 5/11 patients had residual head posture of ≤ 5° and 6/11 had > 10°. After combined recession/resection, 6/9 patients had residual head posture ≤ 5° and 3/9 had < 10°.	Larger studies were included in table 2.

Appendix B: Related NICE guidance for four tenotomy of horizontal eye muscles for nystagmus

There is currently no NICE guidance related to this procedure.

Appendix C: Literature search for tenotomy of four horizontal eye muscles for nystagmus

Database	Date searched	Version/files
Cochrane Database of Systematic Reviews – CDSR (Cochrane Library)	13/5/08	Issue 2, 2008
Database of Abstracts of Reviews of Effects – DARE (CRD website)	14/5/08	N/A
HTA database (CRD website)	14/5/08	N/A
Cochrane Central Database of Controlled Trials – CENTRAL (Cochrane Library)	13/5/08	Issue 2, 2008
MEDLINE (Ovid)	13/5/08	1950 to April Week 3 2008
MEDLINE In-Process (Ovid)	13/5/08	April 29, 2008
EMBASE (Ovid)	13/5/0/8	1980 to 2008 Week 17
CINAHL (Search 2.0, NLH)	13/5/08	1982 to date (via Dialog)
BLIC (Dialog DataStar)	14/5/08	1993 to date
National Research Register (NRR) Archive	12/5/08	N/A
UK Clinical Research Network (UKCRN) Portfolio Database	12/5/08	N/A
Current Controlled Trials metaRegister of Controlled Trials - mRCT	12/5/08	N/A
Clinicaltrials.gov	12/5/08	N/A

The following search strategy was used to identify papers in MEDLINE. A similar strategy was used to identify papers in other databases.

1	Tendon/su
2	Tendon Transfer/
3	Tenotom$.tw.
4	Tendotom$.tw.
5	(Tendon$ adj3 (surg$ or cut$ or reattach$ or re-attach$ or transfer$)).tw.
6	Oculomotor Muscles/su
7	(Oculomotor$ adj3 muscle$ adj3 surg$).tw.
8	(Extraocular$ adj3 muscle$ adj3 surg$).tw.
9	(Eye$ adj3 muscle$ adj3 surg$).tw.

10	Ophthalmologic Surgical Procedures/
11	(Ophthalmolog$ adj3 Surg$ adj3 Procedure$).tw.
12	or/1-11
13	Nystagmus, Optokinetic/
14	Nystagmus, Physiologic/
15	Nystagmus, Congenital/
16	Nystagmus, Pathologic/
17	Nystagmu$.tw.
18	(Involuntar$ adj3 eye$ adj3 movement$).tw.
19	or/13-18
20	12 and 19
21	Animals/
22	Humans/
23	21 not (21 and 22)
24	20 not 23