SCIENTIFIC REPORT

The Newcastle Control Score: a new method of grading the severity of intermittent distance exotropia

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Aim: To describe the development and application of a novel scoring system for grading the severity of intermittent distance exotropia (IDEX) and its potential application as an intervention criterion for surgical intervention.

Methods: The Newcastle Control Score (NCS) for IDEX was developed by incorporating both subjective (home control) and objective (clinic control) criteria into a scale to grade severity. The score structure described was evaluated for interobserver and test-retest reliability. To determine an optimal score threshold for surgical intervention, 170 cases of IDEX were scored retrospectively. Cure rates for surgical and non-surgical cases were then compared according to preoperative or presenting scores.

Results: Interobserver and test-retest reliability were good ($r=0.82$ and $r=0.89$ respectively). Total cure rate with surgery was 54% and without surgery 18% ($\chi^2=23.093$, $df=1$, $p<0.001$). Significantly fewer patients with NCS ≥ 3 achieved cure without surgery than those with NCS 2 ($\chi^2=3.362$, $df=1$, $p<0.047$).

Conclusions: The NCS is a reliable method for grading the severity of IDEX and aids decisions regarding intervention. Patients with a score of 3 or more are unlikely to attain a cure without surgery.

The most common type of childhood onset exotropia is intermittent distance exotropia (IDEX)¹ also known as divergence excess exotropia,² intermittent exotropia, or X(T). Onset is usually before 18 months of age.³ The initial abnormality is outward deviation of one eye when viewing a distant object, often during periods of inattention, or when the patient is tired. This is often accompanied by monocular eye closure.⁴ Double vision is not a characteristic feature due to suppression of the image from the deviated eye.^{5,6} The initial functional impairment is reduction of the quality of binocular single vision (BSV) to distant targets. The eyes remain aligned when viewing near objects. The natural history of IDEX is not well documented: some believe it to be a progressive condition leading to irreversible loss of BSV for both distance and near fixation.⁷

IDEX that is well controlled may resolve without treatment.⁸ When therapy is required it may be conservative, in the form of prisms or occlusion^{9,10} or surgical. Both the size and the frequency of the deviation are considered to be important factors in the decision to intervene. Non-surgical treatment has been recommended if the deviation is <20 prism dioptres.⁹ In larger deviations surgery is thought to be indicated if observed by carers to be present during at least 50% of waking hours¹¹ and poorly controlled on clinical examination and at home.¹²

The indications for treatment, particularly surgery, are not well validated. Our aim was to establish evidence based

criteria for the management of IDEX by rating severity and defining a suitable threshold for surgical intervention. A rating scale was developed that differentiates between well and poorly controlled IDEX and this was used to examine surgical cure rates for IDEX of different severities.

MATERIALS AND METHODS

Score development

The Newcastle Control Score (NCS) was developed by incorporating subjective (home control) and objective (clinic control) criteria into a control rating scale. Modified descriptions of control as outlined by Rosenbaum and Santiago¹² were used to rate the frequency with which the strabismus was noticed to be present (subjective) and the ease with which binocular single vision was regained after cover test (objective).

Reliability assessment

To assess interobserver and test-retest reliability for the NCS, ten preoperative and ten postoperative IDEX case scenarios were rated by orthoptists. Case scenarios were taken verbatim from actual clinical records and covered each possible outcome score on the rating scale. Observers were asked to complete the exercise individually. The reliability assessment was completed in two stages. For interobserver evaluation (stage 1) each observer was provided with a copy of the case scenarios and a standardised form for recording scores. All record forms and copies of case scenarios were then returned to the research assistant. This process was repeated after a period of 2 weeks with the case scenarios re-ordered and observers masked to their previous scores for test-retest evaluation (stage 2).

For stages 1 and 2, scores for the case scenarios were correlated and intra-class correlation coefficients produced to indicate the level of interobserver and test-retest agreement.

Surgical threshold evaluation

To establish a suitable threshold for surgical intervention, the score was applied retrospectively to a cohort of patients with IDEX. This was defined as demonstrable BSV for near fixation (one third of a metre) with an intermittent or constant manifest divergent strabismus for distance (6 metres). The minimum distance measurement included was ≥ 20 prism dioptres. The deviation for near fixation measured at least 10 dioptres less than the deviation for distance. Patients older than 11 years or with neurological disease, ocular pathology, conservative treatment for the strabismus, or less than 6 months follow up were excluded.

Scores were allocated for surgical cases based on the last preoperative visit and for non-surgical cases based on the

Abbreviations: BSV, binocular single vision; IDEX, intermittent distance exotropia; NCS, Newcastle Control Score

Table 1 Breakdown showing components of Newcastle Control Score

Score	Component
Home control	
0	Squint/monocular eye closure never noticed
1	Squint/monocular eye closure seen occasionally (<50% of time child observed) for distance
2	Squint/monocular eye closure seen frequently (>50% of time child observed) for distance
3	Squint/monocular eye closure seen for distance & near fixation
+	
Clinic control near	
0	Manifest only after cover test and resumes fusion without need for blink or refixation
1	Blink or refixate to control after CT
2	Manifest spontaneously or with any form of fusion disruption without recovery
+	
Clinic control distance	
0	Manifest only after cover test and resumes fusion without need for blink or refixation
1	Blink or refixate to control after CT
2	Manifest spontaneously or with any form of fusion disruption without recovery

NCS total = Home + Clinic near + Clinic distance.

presenting severity. Outcome was assessed on the last recorded visit and cure rates were analysed for each preoperative or presenting score.

RESULTS

Score development

The components chosen to make up the NCS (see table 1) were selected as they describe the whole range of likely clinical presentations while still being easy to categorise. Each component was weighted to reflect increasing severity resulting in the 8 point scale ranging from 0 to 7. Cure was defined as NCS 0 or 1.

Reliability assessment

Twenty five orthoptists, from eight orthoptic departments across the UK rated the 10 surgical case scenarios. The mean period between test and retest data was 2 weeks (range 1–3 weeks). Interobserver reliability was good ($r = 0.82$, 95% CI 0.71 to 0.91, $n = 25$). Test-retest agreement was also very high ($r = 0.89$, 95% CI 0.85 to 0.92, $n = 12$).

Threshold evaluation

The NCS was applied retrospectively at seven UK centres to a cohort of 67 patients who had received surgery for IDEX and 103 who had received no active intervention; all fulfilled the specified inclusion criteria. Cure rates for each preoperative or presenting score for both surgical and non-surgical cohorts are shown in table 2: in the non-surgical group the overall cure rate was 18% and in the surgical group 54% ($\chi^2 = 23.093$, $df = 1$, $p < 0.001$).

Analysis by presenting score shows that for those with NCS 2 the difference between surgical and non-surgical groups is

not statistically significant (Fisher's exact test, $p = 0.200$) despite both surgical cases achieving a cure.

For those with NCS ≥ 3 significantly higher cure rates were achieved with surgery than without (NCS 3: $\chi^2 = 8.036$, $df = 1$, $p = 0.005$; NCS 4: $\chi^2 = 18.666$, $df = 1$, $p < 0.001$; NCS 4–7 inclusive: $\chi^2 = 15.413$, $df = 1$, $p < 0.001$). In addition significantly fewer patients with NCS ≥ 3 achieved cure without surgery than those with NCS 2 ($\chi^2 = 3.362$, $df = 1$, $p = 0.047$).

DISCUSSION

Management decisions in IDEX present a challenge to the clinician that is not encountered for most other types of childhood onset strabismus. Any intervention must endeavour to improve unacceptable misalignment and poor BSV for distance fixation whilst preserving or improving BSV demonstrable for near fixation. Decisions to intervene are currently guided by both the size and control of the deviation. Where the distance angle is less than 20 prism dioptres, conservative treatment is thought to be more appropriate and surgical intervention not indicated.^{11–14} In IDEX where the distance angle is greater than or equal to 20 prism dioptres, the control of the deviation is considered the important factor in determining whether or not to intervene.

Control is assessed by noting how frequently the strabismus is observed at home and in the clinic and the ease with which it is controlled on cover testing.¹² Observation of the strabismus for >50% of waking hours and poor control on cover test is thought to suggest the need for surgical correction¹¹ but despite this, most clinicians remain unclear as to when intervention is appropriate. Measures of control have not previously been standardised and this may be in part due to the difficulties inherent in relying on parental reports of the frequency of the strabismus.

The NCS incorporates both objective and subjective measures of control into a simple grading system that differentiates and quantifies the various levels of severity in IDEX. The measures of reliability and repeatability found in this study indicate that the NCS is a consistent and robust method of rating severity that can be used accurately in clinical practice. It provides a classification of cure (NCS 0 or 1) and also enables any change over time to be more easily monitored.

We used the NCS to try and establish an appropriate threshold for surgical intervention and thus address the current uncertainties surrounding this issue. There does not seem to be a significant management dilemma for those patients at each end of the scale. For those with well controlled IDEX (NCS 2) surgical intervention may be difficult to justify. We found a significant spontaneous cure rate (39%) in this subgroup and generally surgery was not undertaken. On the other hand, in very poorly controlled IDEX (NCS 7) the clinical picture is one where surgery would generally be indicated. However the majority (86%) of patients were moderately affected with NCS 3–6. Of these only 16% spontaneously cured compared with 53% who were cured following surgery. This suggests that surgical intervention is necessary to achieve a cure in this subgroup. The

Table 2 Control scores v cure rates for surgical and non-surgical cases of IDEX using the Newcastle Control Score

NCS	2	3	4	5	6	7	Overall
Surgical	(n = 2)	(n = 12)	(n = 26)	(n = 16)	(n = 6)	(n = 5)	(n = 67)
Cure	2 (100%)	7 (58%)	17 (65%)	8 (50%)	0	2 (40%)	36 (54%)
Non-surgical	(n = 13)	(n = 45)	(n = 38)	(n = 5)	(n = 1)	(n = 1)	(n = 103)
Cure	5 (39%)	8 (18%)	5 (13%)	1 (20%)	0	0	19 (18%)

spontaneous cure rate is significantly lower for NCS ≥ 3 than for NCS 2 (χ^2 , $p=0.047$) suggesting that NCS 3 is an appropriate threshold for surgery. The cure rate from surgery in the moderately affected IDEX group (NCS 3–6) of 53% is comparable with that reported in other studies.^{15–17} In a condition that is generally asymptomatic with normal BSV for near fixation, this may portray surgery as being of dubious benefit, especially in view of the risk of over-correction with loss of pre-existing binocular function. Surgery however appears to be driven mainly by parental and peer awareness of the strabismus rather than clinician concern for function, and it may be that improvement in alignment without achieving a “cure” is an acceptable outcome. Information obtained from the use of the NCS can be used to counsel patients and parents preoperatively about the likely outcome following surgery.

There is clearly a need for a well planned, prospective clinical trial of surgical intervention in this group of patients to address questions regarding the nature of the condition itself, the most appropriate management for different severities of IDEX, and realistic goals of treatment.

CONCLUSION

The Newcastle Control Score is a reliable, clinically sensitive method for grading the severity of IDEX. As each level of severity is quantified on a linear scale, it is a simple tool with which to measure any change over time and is suitable for use both clinically and in research. Its implementation in clinical practice will help standardise the management of this condition, enable a suitable threshold for surgical intervention to be set, and allow a more accurate assessment of outcome.

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