Improved Visual Acuity in Patients with Congenital Nystagmus Following Anderson-Kestenbaum Procedures

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ABSTRACT

This retrospective study conducted within a large paediatric clinical practice identified eleven patients who were operated on during the period of 1996-2011, using the Anderson-Kestenbaum procedure. These patients, aged 4 to 17 years, were operated on by the same surgeon. Visual acuity was measured using age-appropriate linear or single optotypes and results were compared pre- and post-operatively. Observations of head posture by ophthalmologist, orthoptist and parent were recorded pre- and post-operatively. Results showed the Anderson-Kestenbaum procedure reduced the compensatory head posture and improved visual acuity in 75% of patients, with a mean improvement in visual acuity of 3.75 letters. Patients showed minimal residual head posture. This study is limited by its retrospective nature and small subject numbers.

Keywords: nystagmus, compensatory head posture, visual acuity

INTRODUCTION

Anderson and Kestenbaum were the first ophthalmic surgeons to describe the use of bilateral extraocular muscle procedures to reduce the size of the compensatory head posture and improve function in patients with congenital nystagmus.[1,2,3,4] While their procedures had a common aim, Anderson operated on only two muscles whereas Kestenbaum operated on all four horizontal extraocular muscles.[1,4,5] The aim of the procedure is to displace an eccentric null zone towards the primary position using recession and resection of the horizontal recti muscles so that the patient is able to achieve a zone of minimal nystagmus without having to adopt a large compensatory head posture.[1,2,3,4] The procedure involves recessing the pair of extraocular muscles opposite to the direction of intended gaze and resecting the pair of agonists contracting in the intended direction of gaze.[1,2,4] Taylor and Jesse supported Anderson's proposal that through the dampening of the nystagmus, patients' visual acuities would also increase.[1,2] This is consistent with the rationale that the lower the intensity of the nystagmus, the greater the improvement in visual acuity.[1,5,6] The Anderson-Kestenbaum procedure is not widely used as there are a limited number of patients for which the procedure is suitable. Previous studies have been limited by these constraints. The majority of patients who have congenital nystagmus have a null point, a direction of gaze in which the amplitude and/or frequency of the nystagmus is reduced thereby allowing best potential visual acuity to be realised.[1,4,6] Patients, who adopt a compensatory head posture to utilise their null point, can often encounter many problems. These include changes in musculo-skeletal neck and spinal structures in the developing child, poor use of optics in corrective lenses, social/psychological aspects of a large face turn and in older patients, include the acquisition of driving skills, sporting and other physical abilities. It is for these reasons that nystagmus surgery for a compensatory head posture is often proposed.[4,6] The question has been posed as to whether visual acuity is also significantly improved. The aim of this retrospective study is to compare the pre- and post-operative visual acuity results of patients with congenital nystagmus who had the Anderson-Kestenbaum procedure performed by the one surgeon, with the purpose of reducing the size of the compensatory head posture.

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METHODS

The Anderson-Kestenbaum (AK) procedure had been performed on a total of eleven patients who were identified by researching the clinical records of the largest private paediatric ophthalmology clinic in Victoria, Australia. The years 1996-2011 were chosen, this being the period in which the surgeon had been operating continuously. No AK procedures have been performed since 2011. The patients suitable for analysis had been listed in the surgical records and data was manually collected. Of the eleven patients six wore no refractive correction, four wore full-time hypermetropic correction and one patient had a low amount of hypermetropic correction used only for near work (therefore not worn in the measurements used in this study). Although only the one surgeon operated in this study, several orthoptists provided clinical measurements pre- and post-operatively as part of normal clinical practice.

The visual acuity (VA) of the eleven patients, aged 4 to 17 years, had been measured both pre- and post-operatively as part of a full clinical assessment. The age-appropriate VA optotypes used were the 3 metre LogMAR illuminated chart and the 3 metre Kay picture recognition test. VA was analysed to determine whether there had been an improvement in visual acuity with the reduction of the CHP postoperatively. The VA measurements that have been analysed are those taken with both eyes open using the CHP, as this was the measurement integrating visual acuity and compensatory head posture. Subjective observations of the CHP were recorded pre- and post-operatively by ophthalmologist, orthoptist and parent. Geometric measures of the CHP were not routinely employed in the clinical setting. The ophthalmologist based his surgical decision on a combination of both Anderson and Kestenbaum procedures as well as the Parks method, basing the amount of recession and resection on all four extraocular muscles on the type and severity of head posture and any accompanying strabismus.

RESULTS

Results can be viewed in Tables 1 - 6 and Figure 1. Tables 1 and 2 show the direction of the CHP including face turn, head tilt and chin elevation or depression, the presence of strabismus and the AK surgery. Table 1 describes patients in the linear optotype group (patients 1 to 8) and Table 2 shows the single optotype group (patients 9 to 11). Seven patients had CHP comprising face turns to the left, three patients had CHP of face turns to the right and one patient had an alternating head turn. Five patients had strabismus preoperatively with three showing reduced horizontal angles postoperatively. Due to age, VA measurements were not obtained by using the same optotype on all patients. Best recorded VA preoperatively and the best recorded

Table 1. Characteristics of patients in the linear optotypes group

Patient	Age at surgery (years)	CHP pre-op	CHP tilt pre-op	CHP post-op	Strabismus pre-op	Strabismus post-op	Procedure
1	10.1	Left	No	No	No	No	RLR and LMR recess RMR and LLR resect
2	17.1	Right	Right tilt	Right tilt Chin-up	RET +10pd	Flick RET	RMR and LLR recess RLR and LMR resect
3	6.7	Left	No	No	No	No	RLR and LMR recess RMR and LLR resect
4	8.9	Left	No	No	No	No	RLR and LMR recess RMR and LLR resect
5	11.6	Right	No	No	No	No	RMR and LLR recess RLR and LMR resect
6	10.4	Left	No	No	No	No	RLR and LMR recess RMR and LLR resect
7	6.2	Left 20°	No	No	LET +10 pd	LET +10 pd	RLR and LMR recess RMR and LLR recess
8	6.9	Alternating	No	Small alternating	LET +10 pd, R/L 14 pd	Exophoria -6 pd, R/L 2 pd	RMR and LLR recess RLR and LMR resect

Table 2. Characteristics of patients in the single optotypes group

Patient	Age at surgery (years)	CHP turn pre-op	CHP tilt pre-op	CHP post-op	Strabismus pre-op	Strabismus post-op	Procedure
9	5.4	Right turn	No	Small right turn	RET +18 pd	RET +6 pd	RMR and LLR recess RLR and LMR resect
10	4.0	Left turn	Left tilt	No turn Small left tilt	LET +35 pd, L/R 12	L/R 12 pd	RLR and LMR recess RMR and LLR resect
11	5.7	Left turn	No	Small left turn	No	No	RLR and LMR recess RMR and LLR resect

VA postoperatively are described in Table 3, using the total number of letters seen, for patients in the linear optotype group. The VA improved in six patients, remained unchanged in one patient and was reduced by five letters in one patient.

Table 3. Linear optotypes group, visual acuity pre- and post-operatively (number of letters identified on LogMAR chart)

Patient	VA BEO pre-op (using CHP)	VA BEO one month post-op	Best recorded VA BEO post-op	Change
1	50	50	55	10%
2	55	50	50	-10%
3	38	35	45	16%
4	45	42	55	18%
5	40	40	40	0%
6	35	35	38	8%
7	30	35	35	14%
8	45	45	50	10%

Figure 1 shows the improvement in VA in the linear group. This is based on the number of letters gained/lost following surgery. The range of letters seen by patients preoperatively was between 30 and 55 letters and postoperatively between 35 and 55, indicating that none of the patients had VA less than 3/9. Postoperatively, 75% of the linear optotype group had some level of VA improvement. The most significant improvement was noted in patient 4 with an 18% (a gain of 10 letters) improvement in VA recorded postoperatively. Mean VA increase postoperatively was 3.75 letters. Figure 1 demonstrates a decrease of 5 letters recorded by patient 2. Further investigation found that this patient preferred not to wear his refractive correction, thereby reducing the VA from 3/3 (55 letters) to 3/3.8 (50 letters) postoperatively. The mean VA improvement in the linear group was 6.81% (3.75 letters) when patient 2 was included or 9.09% (5 letters) when patient 2 was excluded.

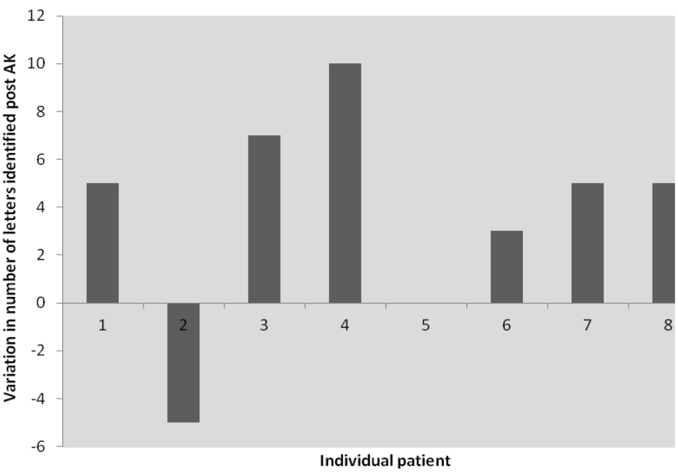

Figure 1. Visual acuity change post Anderson-Kestenbaum procedure (linear optotypes group).

Table 4 shows the VA of patients in the singles optotype group. VA improved postoperatively in two patients (66.7%) and remained unchanged in one patient. This VA cannot be described by the number of letters but only by the Snellen's equivalent value of size of picture recognition.

Table 4. Single optotypes group, visual acuity pre- and post-operatively

Patient	VA BEO pre-op	VA BEO at one month post-op	Best recorded VA BEO post-op
9	3/6	3/6	3/4.8 (45 letters)
10	3/6	3/6	3/6
11	3/9	3/6	3/6

Table 5 shows the amount of muscle recession or resection in each case. It can be seen that the amount of recession/resection in all patients was between 5 and 11 mm, with the largest amount involving the lateral rectus. Patients 2, 7, 8, 9 and 10 had strabismus recorded pre- and post-operatively. The amounts of extraocular muscle recession and resection included the consideration of the correction of the strabismus. The emphasis was on greater amounts of recession/resection of the lateral rectus in these cases. Four of the five patients with strabismus had a reduction in the size of their esotropia. The prism cover test results are listed in Tables 1 and 2. There were no instances of limited adduction or abduction on horizontal gaze as recorded on ocular movement testing. No cases of iatrogenic strabismus were induced and no consecutive exotropia occurred.

Table 5. Amount of extraocular muscle recession/resection (mm)

Patient	RLR	LMR	RMR	LLR
1	unknown	unknown	unknown	unknown
2	unknown	unknown	unknown	unknown
3	10 recess	7 recess	7 resect	9 resect
4	9 recess	6.5 recess	8 resect	10 resect
5	9 resect	7 resect	6.5 recess	9 recess
6	9 recess	6.5 recess	8 resect	10 resect
7	7 recess	5 recess	5 resect	7 resect
8	9 resect	6 resect	6.5 recess	8 recess
9	11 resect	7 resect	7 recess	10 recess
10	10 recess	8 recess	7.5 resect	10 resect
11	7 recess	8 recess	6 resect	5 resect

Table 6 lists the stereopsis results for all patients. As can be seen, no procedure resulted in compromised binocular functions. In patients 1, 3, 4, and 5 the level of stereopsis on the Lang II test either remained stable or improved, patient 6 was never tested, and patients 2, 7, 8, 9, 10, and 11 did not show the presence of stereoacuity pre- or post-operatively.

Table 6. Lang II stereopsis (seconds of arc)

Patient	Stereopsis pre-op	Stereopsis post-op
1	200	200
2	No SV	No SV
3	400	200
4	1200	600
5	1200	1200
6	Test not completed	Test not completed
7	No SV	No SV
8	No SV	No SV
9	No SV	No SV
10	No SV	No SV
11	No SV	No SV

Patients in both single and linear optotype groups were described as having an improvement in VA, but with minimal residual CHP remaining following the procedure. Qualitative description was required as no numerical value was provided for pre- and post-operative CHP and was based on observation of the ophthalmologist, orthoptist and parent. Patient 4 CHP pre- and post-operatively can be seen in Figures 2A and 2B. Head postures were not reversed in any instance. Analyses of all results in relation to statistical significance are not given due to the limited subject numbers.

Figure 2A. Patient 4 preoperative CHP.

Figure 2B. Patient 4 postoperative CHP.

DISCUSSION

The first premise that this study supports is that the VA measured with both eyes open for patients with congenital nystagmus and a CHP improves with a reduction in the eccentricity of the null zone. The closer the null zone approximates the primary position, the better the ability of the patient to maximally utilise their VA. Refractive corrections can be optimally used, and increased retinal image stabilisation probably occurs with a reduction in the nystagmus amplitude and frequency.[6]

In this retrospective study, eight out of the eleven patients had improved VA following AK surgical correction of their CHP associated with their congenital nystagmus. It must be noted that the pre- and post-operative measurements were performed by several orthoptists over time at this large paediatric clinic.

Previous studies have quantified the CHP pre- and post-operatively. Wang et al[7] propose that the measurement in degrees of the patient's CHP is the deciding factor in the required amount of surgery. In patients with CHPs less than 15 degrees, the surgeon used Anderson's two muscle approach. If the CHP was 15 to 25 degrees the surgical decision was based upon Kestenbaum's four muscle approach and in patients with CHP that was greater than 25 degrees, the surgery followed the Parks method.[4] Wang et al's results were positive, showing that 72.27% of patients had less than 8 degrees of CHP postoperatively. While they described the surgical decisions based primarily on the size of the CHP, their retrospective study showed that the main deciding factor was the direction rather than the size of CHP. This raises the question as to the importance of categorising patients into degrees of CHP or not. Wang et al also reported that 79.55% (35 out of 44) of their patients had an increase in VA of 10 letters postoperatively. Our retrospective study compares well, with 75% of patients demonstrating an increase in VA, however the magnitude of the change (3.75 letters) was less than that reported by Wang et al.

Based on previous findings,[8] the surgeon in this study chose to surgically adjust the horizontal component alone of the CHP and not actively intervene in the vertical or torsional components. Similar studies with patients who had both head tilts as well as head turns, showed that surgery exclusively on the horizontal muscles was able to provide significant improvement to the VA and CHP post operatively.[8]

The benefit of limiting the recession of the medial rectus is a common discussion among surgeons. In many studies it is suggested that this muscle should be recessed no more than 5 mm in order to avoid reduced adduction.[9-11] In our study, the surgeon chose to recess the appropriate rectus muscle between 5 and 10 mm depending on the combined CHP and strabismus. While specific adduction measurements were not made postoperatively due to the retrospective nature of the study, there was no symptomatic loss of function. A small amount of symmetrical duction restriction may in fact be desirable postoperatively in the direction of gaze induced by the original CHP. It can be argued that the benefits of the reduction in the patient's CHP outweigh the risk of over-recessing the medial rectus muscle. Sternberg[12] completed a similar evaluation of ten patients who received operations based on Anderson-Kestenbaum procedures. He recessed the medial recti muscles only up to 3 mm, with the rationale of preventing convergence insufficiency. Results however, showed that VA remained the same in eight of the ten patients, and improved in only two. It seems therefore that a greater recession of the medial rectus aids significantly in the reduction of the CHP without adduction deficit risk and therefore allowing improved VA.[3]

Von Noorden[4] discusses the use of the Parks method, which is similar to the method employed in this study. Parks recommended 5 mm recession of medial recti, 6 mm resection of medial recti, 7 mm recession of lateral recti, and 8 mm resection of lateral recti. These contrast with the measurements in the current study which are larger, the greatest being those with strabismus. Parks further recommended adding 1 – 2 mm to the recession/resection in patients with larger head turns.[4] Results in 18 patients whose surgery followed the Parks method, show the elimination or reduction of the CHP to less than 5 degrees in 77% of patients, indicating the success of this method.[4]

The level of data collected for this retrospective study was limited; the main reason being that the Kestenbaum operation is not a common surgical procedure. Similar prospective studies were able to compare larger numbers, with surgery on 43 patients over a period of 15 years.[1] Statistical analyses of those results could therefore be conducted. In our study, the patients were divided into the single and linear opotype groups, required due to the age-related VA testing performed at the time of surgery. Three patients in the single optotype group were unable to complete all or any of their testing with a linear VA chart due to age and maturity level. In a prospective study completed by El Kamshoushy et al,[13] a protocol was completed prior to testing, ensuring that all patients were aged over 5 years. Testing was conducted with one common method and the results were repeated on separate days to establish consistency. The retrospective nature of our study meant there were limitations to these VA protocol settings. El Kamshoushy et al aimed to demonstrate that improving the CHP also improved the recognition time, not just the VA level. The results were unable to be statistically analysed due to the small size of the population. It is useful to note that our study compared favourably with El Kamshoushy et al's, in that there was no evidence of uncosmetic CHP or poor muscle function following the AK procedure.

Surgical results in all patients were successful, including those with strabismus preoperatively. Von Noorden discusses the risk of operating using the Anderson-Kestenbaum procedure on patients with strabismus, as results are often found to be less predictable than strabismus surgery uncomplicated by nystagmus null zones.[4] The four strabismus cases in our study all had unilateral esotropia. In three of the four, the angle of esotropia significantly reduced following the AK procedure, one was unchanged and there were no overcorrections or reversals.

Binocular functions are often useful to include within studies on extraocular muscle surgical treatments. Von Noorden states that in using Parks' version of the Anderson-Kestenbaum procedure, binocular function should not be compromised. The exception is possible diplopia postoperatively which should subside quickly.[4] As shown in the patients within our study, none of the 11 cases who were able to demonstrate stereopsis preoperatively had their stereopsis compromised postoperatively. This supports von Noorden's proposal for the preservation of binocularity when following the Parks method of the AK procedure.

CONCLUSION

This study concludes that using the Anderson-Kestenbaum surgical procedure to improve CHP and VA is successful for the majority of patients with congenital nystagmus. These results support the findings of similar studies. Although the numbers in this data collection were limited, the number of patients gaining improvement in their linear VA is similar, at 75%, to the work of previous authors.

The ideal prospective study would be designed where the patients presented with congenital nystagmus, an established CHP, no refractive error, and were able to reliably perform a linear optotype visual acuity test. VA would be tested both eyes open with and without the CHP preoperatively to establish that the CHP was indeed being utilised to maximise VA. Electronystagmography measures pre- and post-operatively, would provide evidence for changes in amplitude, frequency and latencies of the nystagmus and provide clues for mechanism changes. Clinical data would be collected periodically by the same orthoptist and the operations performed by the same ophthalmologist. The CHP would be quantitatively recorded and qualitative assessment would also be made. The project would be ongoing until sufficient numbers were obtained so that statistical significance could be determined.

This study confirms that the Anderson-Kestenbaum procedure continues to be a useful and successful tool in the management of patients with congenital nystagmus and a compensatory head posture. This procedure reduces the head posture and improves the visual acuity in the majority of patients.

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