With the Rule Astigmatism in Congenital Horizontal Jerky Nystagmus

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Abstract

Children with congenital Nystagmus are known to have a higher incidence of astigmatism when compared to age matched controls [1]. This astigmatism is generally with the rule (WTRA) with the least corneal curvature in the horizontal meridian and the plus cylinder axis at 90 degrees (+/- 30 degrees). This study investigates the incidence of WTRA in patients with congenital Nystagmus attending National Eye Hospital, Sri Lanka. Astigmatism is common in normal neonates and reduces to adult level through the process of emmetropization. This process appears to be interrupted in infants with congenital Nystagmus [2]. A possible mechanism is discussed.

Keywords: Congenital Nystagmus; With the Rule Astigmatism; Kerato-Refractometer

Introduction

Regular or with the rule astigmatism (WTRA) is defined as astigmatism in which the least corneal curvature is in the horizontal plane and is corrected by a plus cylinder with axis vertically at 90 ± 30° or minus cylinder with axis at 180°. Duke Elder and Abrams [3] revealed that the anterior surface of the cornea is mainly responsible for the error in WTRA. Astigmatism ascribed to the posterior corneal surface tends to be of smaller magnitude and creates against the rule astigmatism (ATRA) with the more powerful plus meridian horizontally (180 ± 30°). Other possible causes of astigmatism include crystalline lens curvature and lens tilting known as residual astigmatism. In the normal population 95% have astigmatism but this is < 1.00DC in 65% and < 2.00DC in 90%. Marked Astigmatism is often found in infants and is not present at birth but increases and becomes maximum between 11 to 40 weeks [4]. Reports of the incidence of astigmatism in infants vary between studies with ATRA ranging from 40 - 90% and WTRA in 20 - 40%. Greater than 1.00DC is reported to occur in 45% of infants [4] but if the refraction was carried out with cycloplegia the incidence reduced to 19% [5]. Photorefractive measurements suggest the incidence falls rapidly during the second year of life and is down to adult levels by 18 months [6]. This process of normalization of refractive error is termed emmetropization. The exact mechanism for emmetropization is unclear but the dioptric apparatus in the eye changes to compensate for the natural change in axial length from 18 mm at birth to 23 mm by 3 years, which would produce a drift towards myopia of 15D. It is presumed that a feedback mechanism from defocussed images produces the near-emmetropic state in normal infants.

Congenital Nystagmus (CN) is defined as involuntary, bilateral, conjugate oscillations of the eyes which are present at birth or develop in the first 6 months of life [7]. It may be linked to albinism and is termed idiopathic if not caused by any other ocular pathology. The wave form of CN has a slow phase with an increasing exponential velocity but can also be associated with jerk or Pendular waveforms or a combination of the two and can be greatly modified by fixation, fatigue and attention [7].

Significant astigmatism in excess of 2.00DC has been reported in patients with congenital Nystagmus. Gamble [8] in a study of 65 children with Nystagmus found that 24% had ≥ 2.00 DC WTRA, and Norn reported > 2.0DC in 13% of 140 eyes with Nystagmus. Most measurements are taken using Keratometry which considers only a small annulus of the central cornea. Dickinson [2] recorded results from Retinoscopy, Keratometry and photoelectric keratoscopy (PEK) in patients with albino and idiopathic CN. The PEK was used in order to study how the anterior cornea contributes to the total amount of astigmatism. Topography of the whole anterior cornea provided a measure of the rate of peripheral flattening of the cornea. She calculated central curvature which is the curvature at the apex of the curve independent of shape; and shape factor which defines shape but is independent of size. Her results showed no significant difference in the shape of the periphery of the cornea in the 3 groups (albinos, idiopathic CN and age matched astigmats without Nystagmus). She found there was a significant weighting towards WTRA with 67% of albinos and 33% of idiopaths showing > 2.00DC. That the astigmatism was predominantly WTRA and corneal in origin enabled her to conclude that an individual with CN would be amenable to the same techniques of optical correction (spectacles or contact lenses) as a subject without CN.

Materials and Methods

This study was undertaken prospectively using patients with congenital Nystagmus attending the Orthoptic clinic at National Eye Hospital, Sri Lanka from January 2001 to May 2003. Those with sensory defect causes of Nystagmus for example albinism; history of ocular surgery; or contact lens wear were excluded. Full Orthoptic assessment and refraction with or without cycloplegia was carried out on all subjects. Corneal topography as performed using the Topcon KR 7100D auto kerato-refractometer (KR) (Figure 1). The patient was asked to fixate the picture target inside the KR and the automatic focus was fired when the author had a clear view of the cornea. The author took all the readings and having experience in using the KR in patients with Nystagmus and was able to ensure readings were only taken when fixation was steady on the internal target. Since there are well documented diurnal variations in KR readings [10]. Measurements were taken between 9.30 am and noon.

![Topcon Kerato-refractometer](image1.jpg)

**Figure 1: Topcon Kerato-refractometer.**

Results

KR readings (Figure 2) were taken from 50 patients (100 eyes) aged 3 - 30 years. Readings could not be obtained from one patient due to the magnitude of the Nystagmus and results were unreliable in 10 eyes. In the remaining 89 eyes, there was an incidence of WTRA of 88.75%, 52.8% were greater than 1.0DC and less than 3.0DC, the full results are shown in table 1.

![Corneal topography map showing astigmatism](image)

**Figure 2**: Corneal topography map showing WTRA.

<table>
<thead>
<tr>
<th>Refractive Error</th>
<th>Number of eyes</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>-0.25DC to -1.00DC x 180</td>
<td>25</td>
<td>28.09</td>
</tr>
<tr>
<td>-1.00DC to -3.00DC x 180</td>
<td>47</td>
<td>52.80</td>
</tr>
<tr>
<td>-3.00DC</td>
<td>07</td>
<td>7.86</td>
</tr>
<tr>
<td>Against the rule or none</td>
<td>10</td>
<td>11.23</td>
</tr>
<tr>
<td>Total included</td>
<td>89</td>
<td></td>
</tr>
<tr>
<td>Excluded</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Unable to get KR reading</td>
<td>01</td>
<td></td>
</tr>
<tr>
<td>Total recruited</td>
<td>100</td>
<td></td>
</tr>
</tbody>
</table>

**Table 1**: Results of kerato-refractometry readings.
Discussion

The incidence of 88.75% WTRA in patients with CN in this study compares favourably with other authors [1,2,8,9]. Dickinson [2] discusses the aetiology of WTR corneal astigmatism and suggests strong hereditary bias and environmental factors (visual experience and diet) could play a part. The normal emmetropization process seems to be affected by the Nystagmus. Attempts to attribute the astigmatism to the continual horizontal oscillations producing degraded vertical lines and causing more emmetropization in the horizontal meridian are unsatisfactory since they would produce ATRA rather WTRA. Dickinson suggests the band like pressure of the tarsal plate on the moldable structure of the cornea could lead to the high incidence of WTRA in infants.

This effect would be greater in eyes with low ocular rigidity; however, there was no evidence of low rigidity in her subjects. A further explanation could be that the continuous action of the Lateral and Medial Rectus muscles involved in the horizontal Nystagmus oscillations may exert tension on the globe and influence the shape of the cornea. To test this hypothesis Dickinson re-examined the data to see if there was a positive correlation between the intensity of the Nystagmus oscillations and the refractive error. No correlation was found but oscillations in CN are known to vary considerably depending on the task and direction of gaze which make the data collection unreliable [7].

Caputo [1] examined retrospectively the Cycloplegic Retinoscopy results of 128 patients (255 eyes) with CN and found an overall incidence of astigmatism of 65% (83.1% WTRA) in the Nystagmus population compared to 34% in the age matched controls. They looked at the change in astigmatism with age and found the incidence increased exponentially in the Nystagmus group whereas it remained stable in the controlled group at 30 - 40%. There was a significantly higher incidence of WTRA of 84.6% (27.1% > 2.00DC) in the CN group greater than 3 years of age. Caputo agreed with the molding mechanism suggested by Dickinson (2) and felt that the constant horizontal ocular oscillations in CN produced contraction of both Lateral and Medial Rectus muscles in the 180 meridian. The increased Rectus muscle tone flattens the anterior spherical surface of the cornea in the horizontal meridian, the axial length is reduced which produces WTRA. From their data, they also felt that the increased muscle tone had a greater effect in children less than 3 years of age during the critical period of ocular growth. This period of sensitivity was supported by evidence of corneal malleability in children under 4 years after surgery for congenital Ptosis [11]. A subset of 17 patients were followed up by Caputo [1] in a longitudinal study, only 17% had astigmatism before 1 year of age, whereas 82.4% developed astigmatism after 3 years (29.4% > 2.00DC) adding further supported to the theory of a molding mechanism increasing the incidence of WTRA in CN. The results in our paper support the findings of a higher incidence of WTRA (88.75%) in CN. The most likely explanation, in the absence of further evidence, is the molding effect on the anterior surface of the cornea produced by increased tone in the horizontal Rectus muscles in infants with CN.

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Bibliography


