CASE REPORT

A Journey Towards 1st Step of Life – A Case Report on Vision Therapy for A Patient with Global Developmental Delay and Nystagmus

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ABSTRACT

Introduction

Children with Global Developmental Delay (GDD) fail to achieve developmental milestones expected for their age range as compared to their peers. They typically have visual problems in addition to poor gross and fine motor skills. Developmental delays extend to speech, language, cognition, and personal-social domains, and impact activities of daily living.

Case Summary

A 4-year-old child presented with chief concerns from his parents that he had difficulty making eye contact was unable to stand and walk. He was not making adequate progress in physiotherapy. Neuro-developmental assessment resulted in a diagnosis of GDD. Visual Evoked Potential showed decreased visual acuity, and nystagmus. Comprehensive examination showed that the patient had a face tilt and head turn. Spectacle lenses with prism was prescribed. A therapy treatment plan was recommended and it was divided into three phases, with the primary goal of improving visual functions through vision therapy. At the end of the therapy, the child developed the confidence to stand and walk without any support.

Conclusion

This case report demonstrates the importance of vision therapy in improving the patient's overall visual performance. In particular, it demonstrates the role of visual intervention in aiding motor skills to the extent that standing and walking was enabled without the need for additional support.

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INTRODUCTION

Global developmental delay (GDD) occurs in children who fail to achieve developmental milestones within their age range as compared to other children in their peer group. Developmental delays include gross or fine motor skills, speech, language, cognition, vision, personal-social and activities of daily living.[1] GDD has a prevalence ranging from 1 in 213 to 1 in 448 births depending on parental age, and globally accounts for about 400,000 affected children born every year. In developed countries the reported incidence of GDD is 1–3% of children aged 5 years or younger.[2]

There are numerous reported causes of GDD, including malnutrition, with precipitating factors occurring during the prenatal, perinatal, or postnatal periods.[3] To establish the cause a thorough history, clinical examination and investigation of both child and parents is necessary.

Children with GDD require rehabilitation for motor system development, speech, physical therapy and related therapeutic interventions.[4] However, visual problems are often overlooked which can delay the efforts required in managing the problem.

There is a limited amount of literature providing an accurate account of visual function in children with GDD, and the role that vision therapy can play in the development of these children.[5] A study in Calicut reported that 75.2% of cases showed ocular manifestations, with refractive error being the most common finding, followed by optic atrophy, strabismus and Cortical Visual Impairment. Oculomotor or accommodative dysfunctions significantly impacted the child's daily life. Vision rehabilitation or therapy helps these children to improve their daily capabilities.[6]

The following case of a child diagnosed with GDD and nystagmus documents the visual deficits present, and shows how a stepwise approach to vision therapy helped the patient in improving visual performance and motor skills.

CASE REPORT

ZM, a child aged 4 years and 6 months from Bangladesh, presented to our Caring Vision Therapy & Neuro-vision Rehabilitation Centre in Chennai in June of 2019 for a developmental vision exam, accompanied by his parents. His mother reported concerns related to poor eye contact, poor visual fixation, jerky eye movements, and a limited attention span. The child was undergoing physiotherapy, and the physiotherapist was concerned that there was not much progress in maintaining balance and standing posture. She suspected that there may be an underlying visual problem.

Birth history reported was by c-section at the 37th week, and perinatology history was normal. After the first month of life, the paediatrician was concerned about the child's lack of eye contact, and limited visual reaction persisted at 3 months of age. An MRI of the brain revealed dysgenesis of the corpus collasum, with parietal white matter volume loss. Myelination was normal, but the anterior skull base was small. Head circumference was 47.5 cm, with brachycephaly, hypotelorism, general hypotonia and articular hyperlaxity noted. Radiology of the pelvis was done to determine the reason for his inability to walk, and an x-ray showed that the acetabulum was shallow, more so on the right than left slide, with apparent subluxation of the right hip joint.

Previous psychomotor development testing and physical examination, conducted in 2018, revealed head lift ability at 12 months age without the ability to crawl. Poor expressive language development was noted, with screaming and making unintelligible sounds. The spine was aligned and no scoliosis was found. The standing position was not attainable, even with support, and was hindered by ankle valgus deformity resulting in pronation of the feet.

Neuro-developmental assessment done in January 2019 by a pediatric neurologist reported the presence of hand flapping movements, the inability to stand without support, and excessive fear. Symptoms suggestive of autism were noted, but a definitive diagnosis of autism was not established. Past ocular history showed the presence of horizontal nystagmus, and limited visual contact. Visual Evoked Potential (VEP) study revealed signs of visual pathway dysfunction with possibly reduced visual acuity. A visual problem of cerebral origin was reported, and he was prescribed spectacles of −3.00 D sphere in both eyes. ZM's final diagnosis was established as:

1. Global developmental delay

2. General retardation
3. Nystagmus with visual deficits
4. Dysgenesis of the corpus callosum

ZM was referred to our center for visual rehabilitation. His developmental vision exam revealed the ability to follow a light with both eyes, but resistance to occlusion of the right eye. Fixation was very poor and of very short duration (< 3 seconds), with significant latency of initiation and large fixation error noted. Visual fixation was not central, not steady and not maintained. The patient had little ability to control eye movements and often could not move his eyes to the desired targets, even with concentration and effort. Hirschberg testing revealed mild esotropia of the left eye with a face turn to the right. Horizontal nystagmoid movements were noted. Habitual head posture was lowered, and ZM was unable to sustain lifting his head even with visual fixation.

Ocular movements were full in all gazes, but pursuit movements were severely deficient. Pursuits were characterized by significant head and torso movement, and profound inaccuracy and inability to complete a single rotation. Saccadic eye movements were also poor. Head and torso movements were required to initiate saccades and were grossly inaccurate, with very poor ability to complete a single cycle between targets.

Manifest retinoscopy values were −4.00 D sphere in the right eye and -4.75 D sphere in the left eye. Monocular Estimate Method (MEM) showed lag of +1.00 Dsph in each eye. Cycloplegic retinoscopy values were the same as manifest, at −4.00 D sphere in the right eye and -4.75 D sphere in the left eye. Anterior segment examination with hand held slit lamp was un-remarkable. Posterior segment evaluation revealed a cup disc ratio of 0.3/0.3 in both eyes with generalized pallor. Foveal reflexes were present, and peripheral retinae were normal in both eyes. Tests of visual perception were not administered at this time. The patient was made to stand with support but was very reluctant and fearful while in this position. While standing, the patient screamed and repeatedly wanted to sit on the ground.

Test of primitive reflexes results are as follows:

1. Moro Reflex – Retained - grade 4
2. Tonic Labyrinthine Reflex – Retained – grade 3
3. Asymmetric Tonic Neck Reflex – Retained – grade 4
4. Symmetric Tonic Neck Reflex – Retained – grade 4
5. Babinski Reflex – Retained – grade 3
6. Spinal Gallant Reflex – Retained – grade 4
7. Segmental Rolling – Retained – grade 4

TREATMENT

The treatment plan for the patient was divided into three phases as follows:

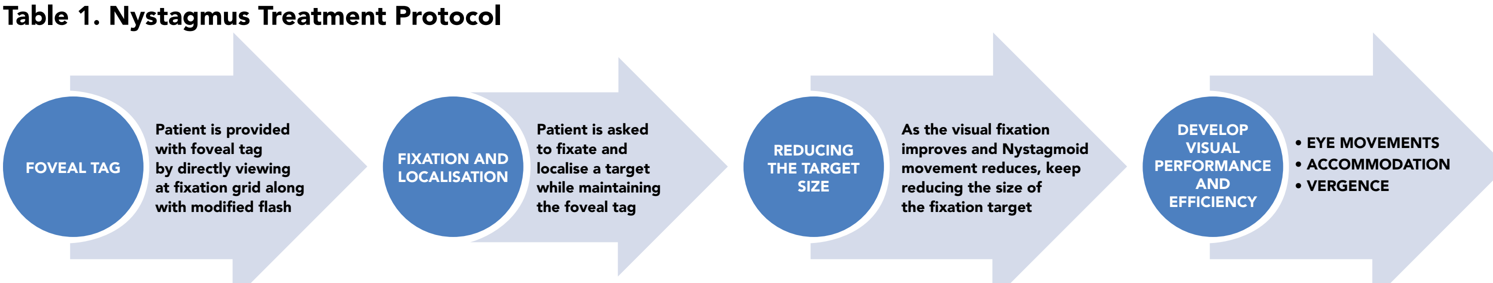
Phase 1

The first phase was to give an optimum optical correction of −4.00 Dsph in the right eye and −3.75 Dsph left eyewith 4Δ base down yoked prism along with Bi-Nasal patching.

Phase 2

The second phase of treatment was to reduce nystagmus, to improve visual fixation, and to improve oculomotor control along with

Table 1. Nystagmus Treatment Protocol



normalizing accommodation and vergence control. An approach was taken to reduce nystagmus through biofeedback mechanisms according to our nystagmus treatment protocol **(see Table 1)**. The patient is provided with a foveal tag monocularly through a modified flash **(see Figure 1)**. This is followed by looking into a direct ophthalmoscope grid **(see Figure 2)** over the period of 20 minutes during each session, for 10 sessions.

Figure 1: Modified Flash

Figure 2: Direct Ophthalmoscope Grid

After 10 sessions there was a marked reduction in nystagmus. At that point, we fine-tuned fixation by combining the visual feedback with tactile feedback. The patient was provided with a foveal tag monocularly and asked to fixate and point to the visual targets at 40 cm. This was followed by a controlled, progressive reduction in the size of the visual targets from N72 print size to N18 print size over the course of the next 20 sessions. The after-image technique allows the patient to use visual feedback to monitor changes in eye position and relative movement directly. Thus, the patient visualizes the fixation stability. After one month of rigorous training the patient was exhibiting reasonably consistent control of nystagmus. In the later sessions, therapies for stabilizing and addressing oculomotor deficits such as saccades, pursuits accommodation and vergence control were carried out. **(see Table 2)**.

Table 2. Vision therapy activities for developing visual efficiency in global developmental delay in non-verbal.

ACTIVITY	DESCRIPTION
1. PURSUITS	
Chalk Board Maze Tracking	Follow a torch light or laser pointer light with in the track on chalk board, keep changing the track pattern, as patient progresses keep narrowing the track and follow from distance of 1 meter. Monocular / Binocular
Pie pan rotation	Rolling ball in a pan and track. Monocular / binocular
Marsden ball	Tracking Marsden ball with lying down and then tracking, catching and tapping in seating position
SVI	Rotator Module, clockwise/ anticlockwise with gradually increasing speed
SVI	Tracking Module
Pegboard Rotator	Following hole in Slow rotation for 1 full rotation and then putting pegs.
2. SACCADES	
SVI	Saccades module
Saccadic Fixator	Saccadic fixator with anaglyph glasses
SVI	Eye hand coordination module monocular/ binocular
Hart chart	Hart chart with shapes and pictures
4 corner wall fixations	Flashing torch light on the wall at four corners in sequence and random flashing, monocular/binocular
3. ACCOMMODATION	
Soft Toys, picture charts	Patient has to fixate on the details of the toys and picture card from a distance of 40 cm gradually reduced to 5 cm monocularly.
4. VERGENCE	
Eye port fixation trainer	Patient has to fixate binocularly on the LED targets at different speed level and variation of programs.
Styrofoam rod	A 40 cm Styrofoam rod marked with fluorescent marker highlighted under blue light, Patient has to fix pegs at varying distances while fixating binocularly.

The third phase of treatment was to improve balance, posture, and ultimately walking, together with integrating the primitive reflexes. In order to improve balance and posture, in-office sessions of optometric vision therapy were combined with daily home therapy activities for integrating primitive reflexes to address the vestibulo-ocular system and maturation of reflexes.

At first the vision therapy sessions started with three to four activities. These were done in conjunction with 8Δ base down yoke prism while sitting on a balance board with support. The patient was instructed to fixate on a target such as cellphone videos or toys. Pursuits and saccades training were done by following and tracking the Marsden ball, while rocking on a wooden balance board in the sitting position. These activities were done for one month, while progressively increasing the levels of difficulty. ZM was scheduled for a progress check one month following the implementation of in-office and home therapy sessions. After one month the patient was confident in balancing himself on the balance board without any support, and was able to fixate, locate and track the visual target.

Given the rapid progress after the first month of therapy, visual-vestibular therapy was taken to the next level whereby ZM stood with support while performing pursuits, saccades and eye tracking activities with the Marsden ball and subsequently with the Sanet Vision Integrator. Initially ZM was reluctant and fearful to stand, and was crying a lot, but slowly over the next 15 days the fear reduced and he started enjoying the session. As he became more comfortable while standing, we were able to introduce visual-vestibular activities in the standing position. This was done in conjunction with high density foam, and then the wooden balance board, Bosu ball, Bernell balance board and therapy balls respectively.

Towards the end of the month patient had made significant progress on his balance and posture. Moro reflex also reduced to Grade 1 and he could stand without any support, he was more comfortable and calmer while standing. **(see Figure 3)** He was able to reach targets without any hesitation.

Figure 3: Standing without support at the end of 2nd month of therapy.

In the beginning of the third month we initiated therapy for integrating the patient's Tonic Labyrinthine Reflex (TLR) reflex with his visual abiltites. He was guided on walking with support while fixating on a visual target. As ZM gained confidence in taking small steps, the support was gradually reduced. Toward the end of the third month, the day arrived when ZM was able to take the first step of his life without support.

Table 3. Visuo-vestibular therapies in global developmental delay to develop balance and posture.

ACTIVITY	DESCRIPTION
Therapy Balls	Slow bouncing therapy balls while fixating on a visual fixation target e.g.: SVI – Tracking module
Swing	Slow swinging visual targets while fixating eg. rhymes on phone etc.
Balance Board Activities	First, seating on balance board and once patient improves then standing on balance board along with various visual performance task like saccades, pursuits, eye hand coordination, etc., start with bi-directional balance board (wooden one) and then gradually progress to Bernell balance boards
High Density Foam	High density foam is used before balance boards to create vestibular input while standing and balancing along with visual activities.
Bosu Ball	A dome shaped ball used for standing and sustaining balance along with various vision therapy activities
Marsden Ball	Marsden ball activities performed initially while seating on balance board followed by standing on balance boards and Bosu ball.

DISCUSSION

In general Vision Therapy has been shown to be effective for the treatment of eye movement disorders, visual dysfunctions, and a variety of conditions for a wide range of populations. VT has also been shown to improve quality of life, and is an effective form of rehabilitation for many children.[5] However, the efficacy of Vision Therapy for improving developmental delay is not well established, therefore many clinicians may not consider it as a treatment option for this population. However, as this case report clearly demonstrates, a multimodal approach to vision rehabilitation including the use of yoked prism, vision therapy, and visuo-vestibular therapy, is an efficient management strategy for a child with Global Developmental Delay (GDD) and nystagmus. With regular physiotherapy, there was not much improvement in the posture and balancing ability of the child. When the limited gains had plateaued, his physiotherapist referred the patient for visual assessment and intervention. The child's parents were concerned about his delayed milestones as well as poor eye contact.

Head posture plays a significant role in a child's overall growth. Correct posture and the ability of voluntary movements develop the environmental information for which somatosensory, vestibular and visual system form a triad guiding the posture and locomotion systems.[7,8] The visual system not only provides clarity, but processes the information from eyes to head, to the shoulders, to articulated muscle-joint system evolved for bipedal locomotion.[9] The visual system is often overlooked in the clinical setting, as it takes time and special attention to formulate appropriate treatment.

Head posture can be influenced by yoked prism. In the case of ZM, yoked prism was used to correct a face turn and elevate chin posture. A prior case report in the literature proposed the use of base down (BD) yoked prisms to aid mobility with a head tilt by modifying the line of sight.[10] A study by Errington et al, showed that base down yoked prism affects gait in young adults while walking.[11] For our patient, a significant amount of abnormal head posture was corrected by prescribing 4Δ Base down yoked prism which was used during the vision therapy. Along with base down yoked prism, binasal patching was advised to gain peripheral fusion. Fox has suggested that introduction of binasal patching is a useful adjunct to vision therapy, which results in better performance.[12]

Our treatment approach to nystagmus reduces its amplitude and frequency, thereby resulting in improved visual acuity. Although the VEP revealed visual pathway dysfunction and decreased visual acuity, there were significant amounts of head tilt and face turn due to the nystagmus. Nystagmus is caused by disorders in the vestibular system, the gaze holding mechanism, visual system and the smooth pursuit system.[13] Reduced visual acuity in patients with nystagmus is usually not due to motion-induced blur, but due to the image not being placed within the foveal fixation region for an adequate period of time.[14] Clinical management for nystagmus is aimed toward increasing foveal fixation time by reducing nystagmus intensity.[15] Vision therapy through biofeedback mechanisms incorporating after-image techniques helps to increase voluntary control over nystagmus and stabilize fixation.[16,17] A study by Mezawa et al proved that biofeedback training reduces nystagmus, and extends foveation time which increases the ability to fixate.[18] A case has previously been reported in which vision training improved visual acuity, motor alignment, and visual functions like accommodation and vergence.[19]

An approach of optometric vision therapy is also important in managing global developmental delay and should include neuro-developmental assessment of motor skills and retained primitive reflexes, balance, and bilateral co-ordination.

Primitive reflexes are the special reflexes that develops in the brain stem and executed without cortical involvement. This set of

Table 4. Influences of retained primitive reflexes on visual system.

PRIMITIVE REFLEXES	EFFECTS OF RETAINED REFLEXES ON VISUAL DEVELOPMENT
Moro Reflex	Vestibular problems, Oculomotor and Visual Perceptual Delay, acute sensitivity to sound, light, wind, temperature change, smell and tension.
Tonic Labyrinthine Reflex	Balance problem, postural instability, ocular motor dysfunction, delay in anti-gravity development, difficulty with proper development of eye-hand coordination, convergence and binocular development.
Asymmetric Tonic Neck Reflex	Poor saccades, Difficulty with bilateral integration, Poor handwriting, difficulty reading, difficulty in eye tracking without head movement.
Spinal Galant Reflex	Poor visual fixation, Poor Concentration, Auditory processing disorder
Babinski Reflex	Poor balance and Toe- walking
Symmetrical Tonic Neck Reflex	Difficulty in accommodation, poor saccades and pursuits, poor standing balance, poor seating posture, it also interferes with near point vision development and binocularity.

involuntary primitive protective reflexes helps the baby with functions such as positioning in the womb, birthing, breathing, feeding, and urination. These reflexes are necessary for the newborn's survival during the first weeks of life. They also provide essential learning experiences as foundations for future neuromuscular development. These reflexes should mature at the proper age, and be inhibited or controlled by higher brain centers to permit development of higher-level postural reflexes.[22]

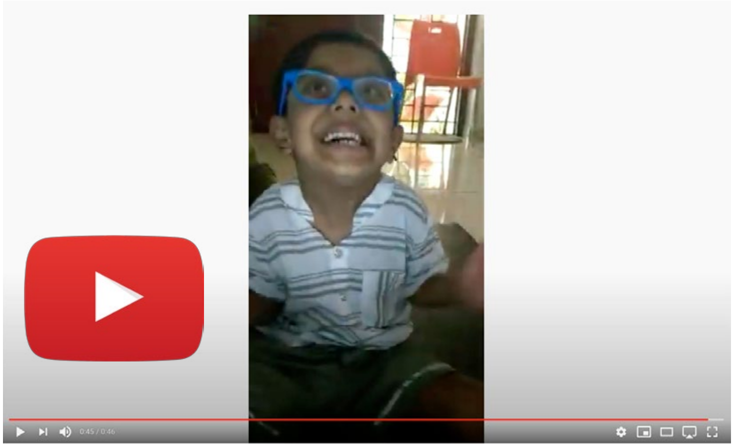
Primitive reflexes that remain after the age of 6 months may hinder normal development of the postural reflexes and ocular motility. These include the Moro Reflex (MR), Asymmetrical Tonic Neck Reflex (ATNR), Tonic Labyrinthine Reflex (TLR), and the Symmetrical Tonic Neck Reflex (STNR).[20] A study by Gonzalez et al showed a significant association between saccadic eye movement and primitive reflex especially the TLR reflex.[21] In our patient we focused on TLR training and the result obtained was significant, as not only the visual acuity was better but the motor skill improved by taking the first step in life without any support.

CONCLUSION

This case report illustrates the importance of a multimodal vision therapy approach to improve patient's visual performance, motor skills in patients with global developmental delay and nystagmus. This case also demonstrates the importance of optometric vision therapy along with integration of primitive reflexes to address the vestibulo-ocular system and maturation of reflexes that enables a child with GDD to stand and walk independently. Thus, a combined approach of managing nystagmus, abnormal head posture and integration of primitive reflexes is crucial to improve visual function and quality of life for such a child. We recommend this multimodal approach for effective management of patients with global developmental delay and nystagmus.

Please see the following supplementary videos demonstrating nystagmus pre and post VT, and walking pre and post VT.

Video 1: Pre-VT Nystagmus



Video 2: Post-VT Nystagmus

Video 3: 1st day of Standing Posture, Balance and Walking Assessment

Video 4: After 2 weeks of Visuo-Vestibular Therapies

Video 5: Independent standing on Balance Board

Video 6: 1st Step of Life

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