A Child with Myasthenia Gravis and Defective Accommodation: A Case Study

Karen Pedemont, DipAppSc(Orth) DOBA[1,2]
Neryla Jolly, DOBA(T) MA[2]
Loreto Rose, BSc(Hons) MBBS(Hons) FRANZCO[3]

[1]Department of Orthoptics, Bankstown-Lidcombe Hospital, Sydney South West Area Health Service, Sydney, Australia
[2]The University of Sydney, Faculty of Health Sciences, Sydney, Australia
[3]Department of Ophthalmology, Bankstown-Lidcombe Hospital, Sydney South West Area Health Service, Sydney, Australia

ABSTRACT

Background: This case details a 14-year old female with myasthenia gravis (MG) who on presentation had ocular symptoms, which included bilateral ptosis and vertical diplopia. Four weeks after the onset of the MG she developed accommodative insufficiency. This condition has not been reported before in childhood MG, but has been documented in adult onset MG. The onset in this case was later in the course of the disease, not initially, as found in the adult cases reviewed in the literature.

Method: Measurements were taken at different stages over an 18-month period to determine the impact of medication and fatigue. Tests for near vision, accommodation, convergence, bar reading and near deviation were performed.

Results: All measurements were reduced and further affected by fatigue with the exception of the size of the near deviation. The patient was symptom-free by 15 weeks post onset. Eighteen months later the patient remained symptom-free with all measurements normal with the exception of accommodation, which remained below normal and affected by fatigue after reading.

Conclusion: This single case highlights the occurrence of smooth muscle involvement in MG and its debilitating effect. It is recommended that testing of accommodation function becomes standard practice in patients with MG and the use of additional plus lenses considered if required.

Keywords: Myasthenia gravis, accommodative insufficiency, child

INTRODUCTION

Myasthenia Gravis (MG) is a disorder of neuromuscular transmission where antibodies that work against the acetylcholine receptors are produced. MG characteristically affects striate or skeletal muscles resulting in weakness and fatigue with ophthalmic involvement of the extraocular muscles.[1] Commonly affecting adults, MG is not often seen in children.[2]

Ocular characteristics seen in patients with MG are diplopia, ptosis and lid closure problems. Diplopia is the most common symptom, with the medial recti, inferior recti and superior oblique most frequently affected.[3,4] A less commonly reported ocular feature of myasthenia gravis is the effect on accommodation. The ciliary nerve is involved in accommodation and innervates the smooth muscle via muscarinic acetylcholine receptors,[5] therefore in MG accommodation should remain unaffected.

Correspondence: **Karen Pedemont**
Department of Orthoptics, Bankstown-Lidcombe Hospital
Locked Bag 1600, Bankstown, NSW 2200, Australia
Email: karen.pedemont@sswahs.nsw.gov.au

Reduced accommodation has been reported in single case studies of adults[1,6] and in eight of nine cases in a series of adults with myasthenia gravis.[7] In childhood MG, the reported ocular presenting characteristics to date include unilateral or bilateral ptosis, strabismus, limitation of ductions and Cogan's lid twitch.[2,8-15]

There have been no reports of accommodative involvement in childhood MG. This report details the ocular findings in a child with MG who had affected accommodation. The results of other near functions are also described.

CASE REPORT

A previously well 14-year old female presented to the emergency department where her results were recorded as bilateral ptosis, which was more marked for the right eye, plus a head tilt to the right. Straightening of the head revealed vertical diplopia. Other symptoms documented included dysarthria, weakness when walking and an increased physical effort required when writing. The medical notes further reported that the pupillary reflexes and accommodation

were normal as was the visual field, visual acuity and ocular fundi. A clinical diagnosis of myasthenia gravis was made based on ocular findings and the impact of fatigue. The acetylcholine receptor antibody result was 0.01 nmol/L (0 - 0.25 nmol/L = negative) referred to as seronegative MG. Prednisolone and pyridostigmine were commenced and a single dose of intravenous immunoglobulin (Intragam) was given. This resulted in a symptomatic improvement, with a reduction in the ptosis and fatigability within 24 hours. The patient was discharged three days later, with a plan for continued current medications plus monthly Intragam infusions for three months.

Four weeks after the diagnosis, the patient was referred for an orthoptic assessment, with the referral noting "complaining of difficulty reading". The assessment revealed the symptom of blurry vision when reading, no previous history of eye problems, no diplopia or ptosis, visual acuity unaided right and left 6/6, N8 both eyes open, a near exophoria of 2 prism dioptres, full ocular motility, reduced binocular accommodation of 5.50 dioptres (D) (40 years of age equivalent), reduced monocular accommodation right eye 4.75 D and left eye 4.50 D and a convergence near point (CNP) of 5 cms. The diplopia and ptosis, as reported at the initial examination, had resolved. At this point a diagnosis of accommodative insufficiency was made.

Additional plus lenses were recommended for near as required, to relieve the ocular symptoms. The choice of +1.00 D or +1.50 D lenses was offered, based on the presbyopic correction that correlated with the accommodation level measured. The patient preferred the +1.50 D lenses over the +1.00 D and was asymptomatic. With the plus +1.50 D lenses, the near vision improved to N5 and the binocular accommodation to 11.00 D. The patient was not given orthoptic exercises at this stage as a gradual improvement in accommodation was expected in line with the improvement of the patient's other general symptoms.

Further orthoptic assessments were conducted over a 3-month period. An additional follow-up assessment was conducted at 18 months after diagnosis to determine the longer-term outcome for this patient. The assessments included near vision with both eyes open, accommodation and convergence near points using a RAF rule, motor control of the near deviation by testing bar reading using the Merrick Children's Bar Reading Book, and measurement of the near deviation using a subjective test, the Maddox Wing. Both the monocular and binocular accommodation were measured at four weeks and nine weeks after diagnosis, and both were found to be reduced. At subsequent visits the accommodation was measured binocularly only to avoid fatigue (Table 1). Overall there was a gradual improvement in near function (near vision, accommodation, convergence and bar reading) from the first orthoptic assessment to the visit at 18 months after diagnosis and apart from the accommodation all measurements attained a normal result. Interestingly though, at nine weeks after diagnosis, the convergence near point showed a large reduction to 17 cms, with diplopia reported on convergence break point. Convergence exercises were introduced at 15 weeks after diagnosis when the gradual improvement in convergence near point reached a plateau.

Table 1. Effect of time over an 18-month period

Time post-diagnosis	Near vision	Accommodation (14 years age-normal = 13 D)	CNP	Bar reading	Near deviation
4 weeks	N8	RE 4.75 D LE 4.50 D Binoc 5.50 D	5 cm	*	Exophoria 2Δ
9 weeks	N8	RE 4.75 D LE 4.50 D Binoc 5.25 D	17 cm	N8	Exophoria 2Δ
12 weeks	N8	Binoc 5.50 D	11 cm	N8	Exophoria 2Δ
15 weeks	N8	Binoc 7.50 D	11 cm	N8	Exophoria 2Δ
18 months	N4.5	Binoc 10.00 D	5 cm	N5	Exophoria 3Δ

* Not assessed at this visit

The influence of medication was also noted on the ocular posture. Measurements were taken four days prior to the use of Intragam when the ocular function was at its worst, then three days after the Intragam administration when the impact should be at its most effective. No short-term improvement was seen. In fact, there was a decrease in all measurements except the near deviation post the drug administration (Table 2).

Table 2. Short-term effect of Intragam infusion medication

9 weeks post-diagnosis	Near vision	Binocular accommodation (14 years age-normal = 13 D)	CNP	Bar reading	Near deviation
4 days before medication @ 1800	N8	4.50 D	21 cm	N10	Exophoria 2Δ
3 days after medication @ 1900	N10	3.75 D	23 cm	N12	Exophoria 2Δ

Between nine weeks and 18 months after diagnosis, both the accommodation and convergence showed improvement. However, it was found that at each session when the patient was asked to read for 10 minutes, both the accommodation and the convergence decreased, indicating an additional impact of fatigue on the accommodative insufficiency.

Twelve weeks after diagnosis, the convergence near point showed a greater effect by fatigue than on other visits, however there was no change in near control as assessed by bar reading. A change in bar reading/near control was only observed at week nine after the same reading period. Throughout all visits the deviation size remained constant (Table 3). Although the accommodation improved overall, the fatigue factor was consistent at each visit (Figure 1).

Fatigue from morning to night affected the convergence near point and bar reading at all visits. The near vision remained unaffected by fatigue until 18 months, where the response is seen to be slightly reduced in the evening compared with the morning (Table 4). Binocular accommodation was consistently affected until 18 months where the response improved (Figure 2). The near deviation was not affected.

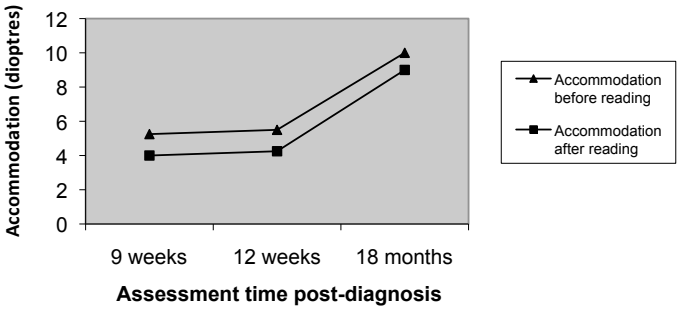

Figure 1. Effect over time on accommodation from 10 minutes of reading.

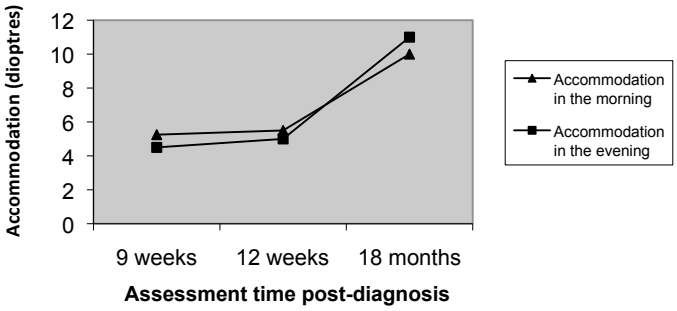

Figure 2. Effect of fatigue from morning to night (8.5hrs).

Table 3. Impact of fatigue from 10 minutes of reading

Time post- diagnosis	Before and after reading	Binocular accommodation (14 years age-normal = 13 D)	CNP	Bar reading	Near deviation
9 weeks	Before reading	5.25 D	17 cm	N8	Exophoria 2Δ
	After reading	4.00 D	19 cm	N12	Exophoria 2Δ
	Near function change	*-1.25 D*	*-2 cm*		
12 weeks	Before reading	5.50 D	11 cm	N8	Exophoria 2Δ
	After reading	4.25 D	17 cm	N8	Exophoria 2Δ
	Near function change	*-1.25 D*	*-6 cm*		
18 months	Before reading	10.00 D	5 cm	N5	Exophoria 3Δ
	After reading	9.00 D	7 cm	N5	Exophoria 2Δ
	Near function change	*-1.00 D*	*-2 cm*		

Table 4. Impact of fatigue from morning to night (8.5 hours)

Time post-diagnosis	Time	Near vision	Binocular accommodation (14 years age-normal = 13 D)	CNP	Bar reading	Near deviation
9 weeks	0930	N8	5.25 D	17 cm	N8	Exophoria 2Δ
	1800	N8	4.50 D	21 cm	N10	Exophoria 2Δ
	Near function change		*-0.75 D*	*-4 cm*		
12 weeks	0930	N8	5.50 D	11 cm	N8	Exophoria 2Δ
	1800	N8	5.00 D	18 cm	N10	Exophoria 2Δ
	Near function change		*-0.50 D*	*-7 cm*		
18 months	0930	N4.5	10.00 D	5 cm	N5	Exophoria 3Δ
	1800	N5	11.00 D	9 cm	N6	Exophoria 2Δ
	Near function change		*+1.00 D*	*-4 cm*		

DISCUSSION

The initial presentation of vertical diplopia, along with the improvement gained with medication is consistent with findings in the literature on childhood MG.[2] The development of near problems is not. The presence of reduced accommodation in this case is consistent with the findings of other authors of adult MG cases.[1,6,7] However in the adult cases detailed by Cooper et al (2000) and Matsui et al (1995), the accommodation defect was present on diagnosis of the illness.

Another possibility is that the patient had Miller Fisher syndrome and not MG. This can also cause external and internal ophthalmoplegia with abnormalities in pupils and accommodation. However, our patient did not have systemic manifestations such as ataxia and areflexia, and did not have abnormal pupils recorded. Our patient was not tested for anti-GQ1b antibody, a very sensitive test for Miller Fisher syndrome and the treatment of immunosuppression does overlap for the two diseases.[16]

In our patient, responses showed that along with the decrease in accommodation, near vision and near control were also decreased. Neither near vision nor bar reading skills have been reported by other authors, but provide added insight into the impact of the MG. The convergence near point remained normal for several weeks after the reduction in accommodation was documented. The convergence was at maximal level so there was no link between decreased convergence and defective binocular accommodation. The presence of reduced monocular and binocular accommodation confirms the diagnosis of accommodative insufficiency. Cooper et al (2000) also reported the delayed onset of convergence near point involvement. Reflexes of accommodation and convergence occur simultaneously and can function individually, so it may be possible that they can also be affected individually. At 12 weeks after diagnosis, the convergence near point was shown to be more affected by fatigue than on other visits. This could be explained by the fact that the patient had received an Intragam infusion earlier that week. Table 2 showed that the short-term effect of the Intragam was in fact a reduction in all measurements except for the near deviation.

The near deviation remained unaffected by both short-term and long-term fatigue, which is surprising as striate muscles are responsible for maintaining ocular balance. This finding is inconsistent with Cooper et al (2000), who noted an increase in the heterophoria throughout the day, typical of MG fatigue. Our use of subjective measurements instead of objective measurements[1] may explain this.

Bilateral ptosis was present at the initial visit along with vertical diplopia, which was managed by adopting a head tilt. Both the ptosis and the diplopia responded to medication and had resolved at four weeks post diagnosis, with no recurrence during the 18-month period. Unlike the accommodative fatigue, the diplopia was not debilitating as it was managed by a slight head tilt. Manson (1965) suggested that the presence of diplopia in his subjects may have prevented them noticing the near vision problems.[7]

Consistent with other studies, the ptosis and vertical imbalance responded well to treatment, with no permanent harm to these muscles.[8,10,14,17] Unlike the ptosis and vertical imbalance, the other near skills affected showed no benefit from Intragam infusion. This response to medication is inconsistent with reports from other authors where their cases all showed an improvement in near function with medication.[1,6,7] Ocular motility assessment is usually limited to the striate muscles in patients with MG. Manson and Stern (1965) and Matsui et al (1995) focused only on measuring accommodation, whereas Cooper et al (2000) measured accommodation, pursuits, saccades and near heterophoria. In our case report near vision, accommodation, convergence near point and bar reading were measured, with accommodation and convergence fatiguing the most, which is consistent with the findings of Cooper et al (2000).[1] To validate which additional ocular tests are essential in managing ocular symptoms of MG a larger sample would be required.

In this case report a subjective measure of accommodative amplitude was used, the RAF rule. Matsui (1995) and Manson (1965) also used subjective measurements, whereas Cooper et al (2000) used objective measurements. Use of subjective measures can be limiting but in this case, the patient was consistent with her responses. In all reported studies, contrary to our case, the subjects had an improvement in near function from medication.[1,6,7] Similarly, Cooper et al (2000) reported that the use of additional lenses assisted in eliminating symptoms of near vision problems.

CONCLUSION

This case study demonstrates that in childhood myasthenia gravis, accommodation, near vision, convergence near point and bar reading can all be affected, but the near deviation may remain unchanged. This case along with others reported in the literature demonstrates the occurrence of smooth muscle as well as striate muscle involvement in MG and its debilitating effect. This case report highlights the need to assess accommodative function in all patients with myasthenia gravis, not only on diagnosis but also later on, and in cases where medication may not assist with near function problems, that management with additional plus lenses be considered.

REFERENCES

1. Cooper J, Pollak GJ, Ciuffreda KJ, et al. Accommodative and vergence findings in ocular myasthenia: a case analysis. J Neuroophthalmol 2000;20(1):5-11.
2. Gamio S, Garcia-Erro M, Vaccarezza M, Minela JA. Myasthenia gravis in childhood. Binocul Vis Strabismus Q 2004;19(4):223-231.
3. Barton J, Fouladvand M. Ocular aspects of myasthenia gravis. Sem Neurol 2000;20(1):7-20.
4. Awwad S. Ophthalmologic manifestations of myasthenia gravis; 2007 [Updated Jul 2011, cited 2011 7th Sep] Available from: http://www.emedicine.com/oph/topic263.htm.
5. Hardman JG, Limbird LE, Gilman AG, editors. Goodman & Gilman's The Pharmacological Basis of Therapeutics. 10th Ed. New York: McGraw-Hill; 2001.
6 Matsui M, Enoki M, Matsui Y, et al. Seronegative myasthenia gravis associated with atonic urinary bladder and accommodative insufficiency. J Neurol Sci 1995;133(1-2):197-199.
7 Manson N, Stern G. Defects of near vision in myasthenia gravis. Lancet 1965;1(7392):935-937.
8 Mullaney P, Vajsar J, Smith R, et al. The natural history and ophthalmic involvement in childhood myasthenia gravis at the Hospital for Sick Children. Ophthalmology 2000;107(3):504-510.
9. Anlar B. Juvenile myasthenia: diagnosis and treatment. Paediatr Drugs 2000;2(3):161-169.
10. McCreery K, Hussein M, Lee A, et al. Major review: the clinical spectrum of pediatric myasthenia gravis: blepharoptosis, ophthalmoplegia and strabismus. A report of 14 cases. Binocul Vis Strabismus Q 2002;8(3):181-186.
11. Vishwanath M, Nischal K, Carr L. Juvenile myasthenia gravis mimicking recurrent VI nerve palsy of childhood. Arch Dis Child 2004;89(1):90.
12. Chaudurhi Z, Pandey PK, Bhomaj S, et al. Childhood myasthenia gravis in an infant. Br J Ophthalmol 2002;86(6):704-705.
13. Kim JH, Hwang JM, Hwang YS, et al. Childhood ocular myasthenia gravis. Ophthalmology 2003;110(7):1458-1462.
14. Ortiz S, Borchert M. Long-term outcomes of pediatric ocular myasthenia gravis. Ophthalmology 2008;115(7)1245-1248.
15. Parr JR, Jayawant S. Childhood myasthenia: clinical subtypes and practical management. Dev Med Child Neurol 2007;49(8):629-635.
16. Caccavale A, Mignemi L. Acute onset of a bilateral areflexical mydriasis in Miller-Fisher syndrome: a rare neuro-ophthalmologic disease. J Neuroophthalmol 2000;20(1):61-62.
17. Ashraf VV, Taly AB, Veerendrakumar M, Rao S. Myasthenia gravis in children: a longitudinal study. Acta Neurol Scand 2006;114(2):119-123.