

## Clinical Reports

# Early diagnosis of Graves' optic neuropathy using visual evoked responses

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**Summary:** A 27 year old woman with Graves' disease developed progressive ophthalmopathy and was noted to have abnormal visual evoked responses (VER). She was treated with high dose prednisone with clinical improvement and return of the visual evoked responses to normal. On withdrawal of steroids symptoms recurred and VER again were abnormal. Orbital irradiation was given with improvement in the ophthalmopathy and VER again normalized. To our knowledge this report represents the first demonstration of improvement in VER with treatment in Graves' optic neuropathy.

### Introduction

Optic neuropathy in Graves' disease is an uncommon but treatable cause of visual loss and may occur in up to 5% of patients.<sup>1</sup> Optic nerve damage is thought to be secondary to compression by swollen extraocular muscles at the apex of the orbit. Congestive symptoms of Graves' ophthalmopathy commonly precede optic neuropathy and visual loss, which is usually bilateral, symmetrical and gradual in onset. Assessment of the severity of optic neuropathy has previously relied on examination of the optic disc, decreased visual acuity, loss of colour vision and visual field defects. Central scotomas are most commonly reported.<sup>2</sup> However, perimetric follow-up<sup>3</sup> reveals diffuse and peripheral defects. Computerized tomographic (CT) scanning of the orbits has also been used to diagnose and monitor optic nerve compression by enlarged muscles.<sup>4</sup> Visual evoked responses (VER) have not been widely used in the follow-up of Graves' optic neuropathy<sup>5</sup> but may be of considerable use in the early diagnosis of optic nerve compression.

This report describes VER findings in a patient with progressive Graves' ophthalmopathy.

### Case report

A 27 year old woman presented with Graves' disease. Thyroid function tests showed thyroxine 228 nmol/l (normal 60–150), T<sub>3</sub> resin uptake (T<sub>3</sub>RU) 38% (27–43) and free thyroxine index (FTI) 88 (17–59). She remained thyrotoxic on carbimazole 30 mg/day although compliance was a problem. She underwent subtotal thyroidectomy.

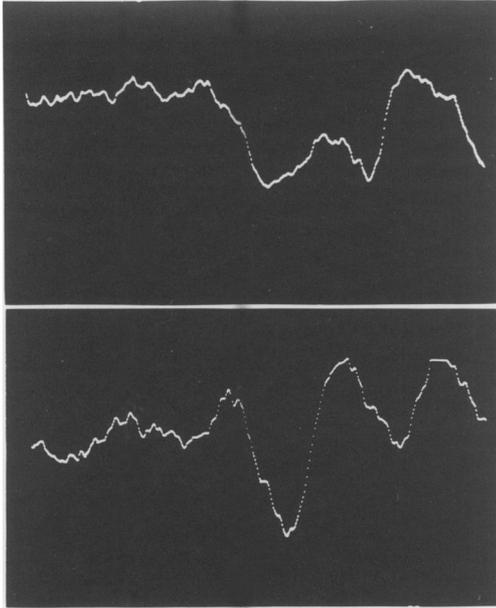
At this time she had developed pretibial myxoedema, proptosis and some limitation of upward gaze. Computed tomographic (CT) scan of the orbits revealed enlargement of the medial and lateral rectus muscles with bilateral proptosis. Visual evoked responses were normal as measured by Medlex Sensor.

Following surgery the ophthalmopathy progressed despite the patient being euthyroid. She had a visual acuity of 6/6 in both eyes. Hertel measurements showed, Right: 25; Left: 27, with a base of 114. Optic fundi were normal. Repeat CT scan showed compression of both optic nerves at the optic foramina by swollen extraocular muscles. The VER showed dispersed wave forms (Figure 1). There was no evidence of conduction delay in the P100 response but the dispersal of the wave form was consistent with compressive optic neuropathy. Prednisone 100 mg daily was commenced with clinical improvement of the ophthalmopathy. Ten days later VER were normal.

The ophthalmopathy remained stable for several months, however symptoms recurred. Although there was no change in proptosis or visual acuity repeat VER were again abnormal. Radiotherapy with 2,000 rads was given to posterior orbits over 2 weeks. Two weeks following completion of radiotherapy Hertel measurements were Right: 25; Left: 23, with base 114 and visual evoked responses had returned to normal.

### Discussion

Visual evoked responses have been used in a variety of neurological disorders including optic neuritis, multiple sclerosis, papilloedema and tumour com-



**Figure 1** Above – abnormal VER in Graves' optic neuropathy; Below – Normal VER same eye 10 days after prednisone therapy.

pression of the anterior visual pathway.<sup>8</sup> The clinically interpreted visual evoked response is a single wave often called P100, generated in the striate and parastriate visual cortex. The preferred stimulus for clinical investigation of the visual pathways is a shift (reversal) of a checkerboard pattern. The measurements used in the interpretation of pattern shift visual evoked potentials are absolute (stimulus to peak) latency of P100, and P100 latency and amplitude differences between the two eyes; amplitude is much less reliable than latency.<sup>9</sup> Visual evoked responses in our case showed dispersal of the waveform consistent with optic nerve compression.

Our case illustrates that VER are a sensitive method of early detection and monitoring of optic neuropathy in Graves' ophthalmopathy. Insidious visual loss in optic neuropathy may be a slow progression of bilateral or asymmetrical visual loss over several months with patients unaware of the

loss until late in the clinical course.<sup>6</sup> Although most patients will have mild to moderate local congestive symptoms of conjunctival and ocular irritation, no direct relationship exists between the degree of congestive symptoms or proptosis and the optic neuropathy.

Untreated Graves' ophthalmopathy is usually self-limiting, but can be unpredictable. Spontaneous recovery can occur within 3 months;<sup>6</sup> however, in a combined series of 32 untreated eyes, seven were left with 20/100 acuity or less.<sup>7</sup> Early treatment of optic neuropathy is more likely to be more successful.<sup>6</sup> Therapy may reverse inflammatory infiltration in the retro-orbital tissue but is not effective when the infiltrate has been replaced by fibrotic tissue.<sup>8</sup>

One third of patients with optic neuropathy demonstrate mild to moderate swelling of the optic disc.<sup>6</sup> Characteristically, such patients have decreased visual acuity, colour vision loss, afferent pupillary defect and visual field defects. On fundoscopy, the optic disc may be normal, may show signs of oedema with haemorrhage or may be pale and atrophic. CT scanning of the orbits demonstrates optic nerve compression at the apex of the orbit due to ocular muscle inflammation.<sup>4</sup> Our case illustrates early VER detection of optic neuropathy with Graves' congestive ophthalmopathy prior to any other objective signs of optic neuropathy or evidence of visual loss.

In conclusion, our case demonstrates the usefulness of VER in early diagnosis and monitoring of Graves' optic neuropathy. Treatment was commenced before any visual loss, visual field change or optic disc abnormalities appeared and appears to have been successful in improving and stabilizing the ophthalmopathy and returning the VER to normal. We recommend measurement of VER in patients with significant Graves' ophthalmopathy so that early optic nerve compression can be diagnosed and treatment instituted before irreversible optic nerve damage occurs.

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