Seeing again: treatment of functional visual loss

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TITLE PAGE

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SUMMARY

There is very little published literature on treatment strategies for functional visual loss. We present two cases of long duration functional visual loss (eleven months and nine years respectively) where complete recovery was achieved with a novel combination of therapeutic approaches including: 1) transparency regarding positive signs such as optokinetic nystagmus to persuade family members of the diagnosis, 2) use of regular positive acknowledgement of everyday events indicating the presence of visual ability, 3) occipital transcranial magnetic stimulation (TMS) to artificially induce phosphenes as a temporary visual experience, and 4) hypnotherapy in promoting visual recovery. We discuss these individual therapeutic approaches in further detail including their background and rationale, and the patients' reflection on their experience of treatment.

Introduction

Functional visual loss, also called psychogenic or conversion disorder, is characterised by the presence of genuinely experienced visual impairment in the absence of a recognised pathophysiological cause. Individuals have a structurally intact visual pathway but have reduced or absent visual awareness. Literature on functional visual loss mostly explores techniques to diagnose the condition, with only a handful of studies looking at epidemiology and outcome. Virtually no literature explores what treatment should be considered, and none describes therapy specific for visual loss, as distinct for treatment principles of functional neurological disorder in general. The use of transcranial magnetic stimulation (TMS) has been described for functional motor disorders. A recent trial found that both transcranial and nerve root magnetic stimulation had an equivalent positive effect on outcome suggesting that its benefit is largely through positive expectation and possibly the effect of TMS in inducing the experience of movement[1]. TMS has only once been described as a treatment for functional visual loss[2]. In that study all patients benefited but they had short duration symptoms and the context of their recovery in relation to other factors was not described.

We present two cases of functional visual loss with visual recovery demonstrating a novel combination of therapeutic approaches including: 1) transparent demonstration and discussion of optokinetic nystagmus and other positive signs of functional loss to persuade patients and family members of the diagnosis, 2) use of regular positive acknowledgement of everyday events indicating the presence of visual ability, 3) occipital TMS to artificially induce temporary visual experience, and 4) hypnotherapy in promoting visual recovery. We suggest these elements may be useful components of a formalised therapy package for functional visual loss.

Clinical history prior to treatment

Case 1 Ms A

Ms A, presented with a six month history of visual impairment and a three month history of complete visual loss. At onset she was an 18-year-old right-handed female who worked as an apprentice in an administration post with no previous significant medical history. She presented with gradual onset bilateral visual loss developing from prolonged photophobia associated with severe migraine. Her visual symptoms began with acute onset migraine, accompanied by photophobia and nausea. Variable migraine persisted, and two weeks later, vision in the right eye became increasingly blurry followed, over several days, by increasing blurred vision in her left eye. Over the subsequent six months, she continued to experience chronic daily migraine and resorted to spending most of her time in the dark. Over this time, her visual acuity gradually diminished. Six months after onset, she woke up with profound visual loss. She was only able to perceive a faint glow related to a light source in her room with blackness in the rest of her vision. This persisted throughout her presentations to ophthalmologists and into her presentation to specialist neurology services three months later. She had been holding on to furniture and step counting to navigate around her home and had been unable to work or drive. There were no experiences of positive visual phenomena such as illusions or hallucinations. Over repeated assessments, no history of stressful or traumatic events in childhood or adulthood emerged. She had no prior history of head injuries, or other psychiatric disorder. She did not present as distressed and there were no features of anxiety or depression. She was defensive about any suggestion of a psychological contribution and described unhappiness at previous suggestions that her blindness was psychogenic or related to an undiscovered psychological trauma. Her mother agreed that at home she had proven to be surprisingly resilient to her new circumstances. Neither the patient nor her family had been persuaded that the diagnosis of functional blindness was correct by previous ophthalmologists, neurologist or psychiatrist.

She had normal eye gaze, pupillary responses, ocular movements, and an optokinetic nystagmus reflex indicating preservation of visual pathways. She demonstrated an ability to sign her name and was able to hold two fingers in front of her face, tasks that patients blind for ocular reasons can usually also do but which are sometimes abnormal in functional blindness. The rest of her neurological examination was normal. Investigations including visual evoked potentials, optical coherence tomography and magnetic resonance imaging (MRI) of the brain were normal.

Case 2 Mr B

Mr B, a 43-year-old right-handed network engineer, presented with a nine-year history of complete visual loss. He had a fifteen year history of chronic back pain following a traumatic fracture of T11 from a motorcycle accident. There was an eleven year history of functional right hemiparesis for which he had undergone an unsuccessful two-month period of inpatient physiotherapy and rehabilitation. During the weekend of a planned discharge date, he experienced a typical dissociative (non-epileptic) attack. When he regained awareness, there was complete blindness in both eyes with no light perception which had persisted for nine years.

He was accepting of the diagnosis of functional blindness from the outset. Over the next three years, he had monthly sessions of hypnotherapy for his functional right-sided weakness, visual loss and dissociative attacks with no substantial symptomatic change. He continued to employ self-hypnosis which helped him to relax. Three years after onset of his right hemiparesis, he had therapeutic sedation with propofol during which he regained full power in his right arm and leg, with subsequent sustained improvement in his right arm but with a return in his right leg weakness after half an hour. His case was included in a publication on that technique[3]. His vision remained unchanged with no light perception. Neuropsychiatric assessment and treatment did not lead to any symptomatic change. On examination, he had normal eye gaze, pupillary responses, optokinetic nystagmus reflex, and catch-up saccades on head turning. Investigations including electroencephalogram and MRI brain were normal. A diagnosis of functional visual loss was agreed by a consultant neuro-ophthalmologist.

Finding an Explanatory model and Demonstrating Reversibility

For Ms A, the initial challenge was persuading her and her family of the diagnosis of functional visual loss. In her case we used a video on YouTube imitating the rotating optokinetic drum (Figure 1) to show her mother how her eyes responded normally and asked them to practise this and show to other family members at home[4]. The second challenge was to alter the formulation away from a simplistic psychogenic/stressor model to one that fitted her own experience. Her visual loss was formulated and explained as arising from her persistent migraines which had caused her brain to "seek out darkness" on a regular basis as a protective mechanism to improve her migraines, a pattern of involuntarily conditioned behaviour which had now become 'stuck' in that mode. Her condition was recognised as a disorder where she has the ability to see but in which the 'idea' and expectation of blindness has become so dominant in her brain that all other information, including normal visual information, had to fit in with this fixed view. The treatment aim therefore would be to resolve the mismatch between her ability to see and her brain "telling itself that it is not seeing".

In Mr B's case there was acceptance from an early stage that this was functional visual loss in the context of a generalised functional neurological disorder. Mr B had previously gained confidence in this diagnosis through the demonstration of positive signs such as Hoover's sign and the way in which self-hypnosis could modulate his attacks.

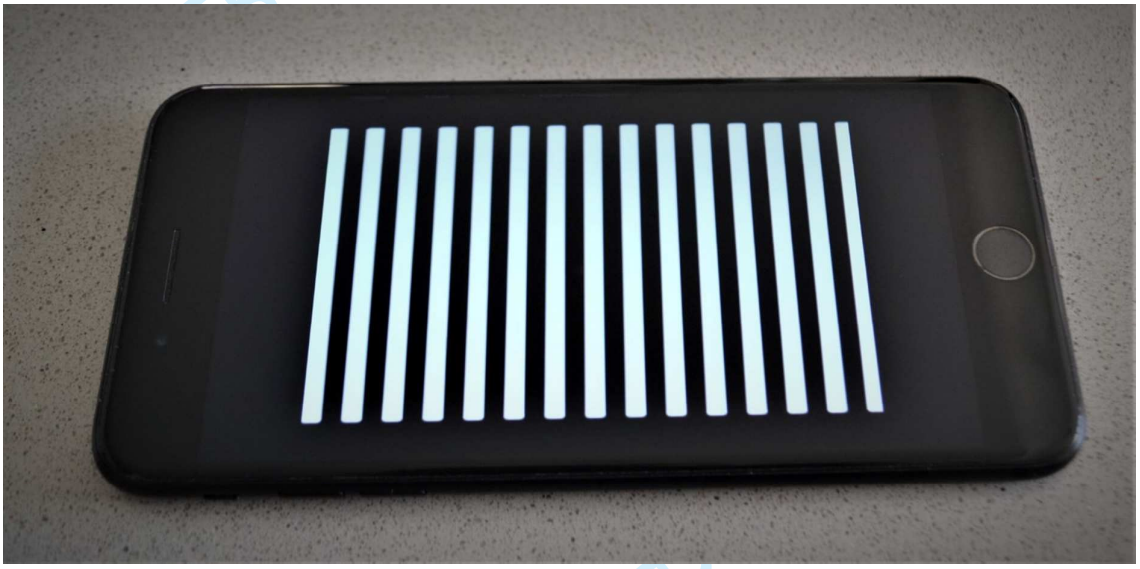

Figure 1. If you don't have an optokinetic nystagmus drum, then use a YouTube video on a mobile phone – and ask the family to practice at home with it

Therapeutic alliance and active seeking of non-experienced visual ability

Regular therapeutic consultations formed an important treatment approach. They served as opportunities for reinforcement of diagnosis using explanation and clinical examination techniques. For Ms A, during routine clinic consultations, regular indications of intact visual ability evidenced by her ability to make direct eye contact, to copy visual gestures and to point to objects in the room all help to reinforce to her that her visual pathways were preserved and therefore recovery was possible. She commented after recovery that the positive and encouraging nature of her consultations and the provision of a list of treatment options had been factors aiding her recovery. She also commented that this contrasted with her previous uncertainty about the diagnosis and the distress at the lack of treatments offered, other than exploration of psychological distress. The use of explicit acknowledgement of her ability to see objects when she was unaware of doing was

extended outside the consultation environment by encouraging her family members to specifically look for and report these occasions to the patient. This reinforced the idea that vision was occurring, but that the "brain was not expecting to see", therefore no visual phenomena was perceived.

In Mr B, bedside techniques such as inability to touch his index fingers together allowed discussions around the brain altering his experience and behaviour around that expectation. He was encouraged to discuss his vision with his partner or father and to discuss events which indicated that he had vision, for example noticing that he liked a particular T-shirt in a shop, and to think of these as opportunities for therapeutic progress. Previously his family had found it difficult to know how to discuss these events in a way which was supportive.

Occipital TMS

TMS utilises electromagnetic induction to produce an electrical current in neural tissue that depolarises cortical neurons and triggers action potential at suprathreshold stimulus intensities[5]. Occipital TMS is known to stimulate visual sensations in the occipital cortex[6]. Amassian *et al* (1989) initially showed that occipital TMS interferes with visual perception by demonstrating that single-pulse occipital TMS disrupts visual processing between 80 and 100 milliseconds after stimulus onset[7]. More recently, Mulckhuyse *et al* (2011) demonstrated the use of occipital TMS in facilitating the production of visual perception in humans by producing light flashes called phosphenes[8]. They found that single-pulse TMS applied 150 to 200 milliseconds before stimulus onset facilitates the discrimination of an orientation target in the contralateral compared with the ipsilateral visual field, suggesting that occipital stimulation enhances the excitability of the visual cortex to subsequent visual perception and thus facilitates visual perception. Occipital TMS has only once previously been described in the literature as a treatment tool for functional visual loss[2]. We considered that production of phosphenes using occipital TMS might help to demonstrate to patients their ability to have visual experiences and trigger better visual awareness.

Ms A underwent three sessions of occipital TMS over three months, six months after the onset of complete visual loss. TMS was applied in an exploratory fashion – initially with single impulses, gradually increasing the amplitude of the stimulus until phosphenes were experienced and ensuring the patients was able to tolerate the accompanying facial twitching produced by the procedure. We found that trains of rapid stimulation at 10Hz produced the most intense visual experiences so continued to use these, in repetitions of 10 blocks over a 30 minute session. During the first TMS session, central occipital stimulation with trains of ten pulses per second for ten seconds produced round phosphenes. Following this, she was able to perceive light from her mobile phone more

intensely. During the second TMS session a week later, a similar stimulation in trains of ten for ten seconds was again effective at inducing phosphenes of lines, dots and white cubes, following which perception of light intensity was transiently increased and there was a spontaneous episode of seeing a blue square a few days after this. Her third TMS session six weeks later produced visual experiences of a red squiggle and red and white dots. She reported that these experiences were associated with an ability to see coloured images for the first time and had helped her to gain confidence in the possibility of visual improvement.

Mr B's experience with occipital TMS started seven years following his initial diagnosis of functional visual loss. He received six sessions over the period of a year. During his first session of occipital TMS, he experienced transient flashes and colours which lasted for two days. A second session enabled him to see his partner's face for a few minutes and, with hindsight allowed a more frank discussion around functional blindness. A third session of stimulation to his left occipital cortex produced a right-sided monocular 'lightening' of vision and detection of his own hand coming over his eye which persisted to the end of the session. A fourth session resulted in the ability to detect the presence of bright lights for three weeks. Fifth and sixth sessions with repetitive and single-stimulus TMS produced some phosphenes and trails but no significant change in vision. He reported that TMS provided him with hope for reversibility and that the experience of transiently producing visual experiences had enabled him to have an easier discussion with his partner regarding the nature of his condition.

Hypnotherapy

Ms A found hypnotherapy, given after TMS, to be of additional benefit. She had previous experience of hypnosis under an acupuncturist for her migraines. She underwent four hypnosis sessions where she described being in a state of deep sleep but in which she was completely aware. She subsequently undertook self-hypnosis sessions where she would count backwards from ten to one, and described how she would concentrate on breathing the pain from her head and eyes to her lungs. She felt this ''stirred something'' in her brain and was a contributing factor to her improvement. Mr B found hypnotherapy helpful in learning relaxation techniques and in controlling his seizures but it had no effect on his vision.

Visual recovery

For Ms A, ten months after the onset of her visual loss, and seven months after more active treatment, she became more light-sensitive and was able to see persistent moving patterns of spots

and splodges indicative of increased visual perception. Her migraine attacks had become fewer and shorter. A month later, the pain in the middle of her eyes which she has had throughout the duration of her visual loss started to dull, and she awoke one morning with complete recovery of her vision and a resolution of the pain. She returned to work two weeks later, and at time of writing is asymptomatic twelve months following recovery. The potential for relapse was discussed to reduce the risk of anxiety or panic, which could in turn intensify symptoms, should this occur.

Mr B regained his vision after nine years of visual loss following a head injury. He had accidently fallen against his kitchen counter and had hit the back of his head on a table. Over the next few days, his vision grew brighter with shapes. Four days later when he was sitting after a shower, his vision came into focus and he was able to perceive a blurred vision of his partner. Over days, this became clearer with some dark spots and associated light sensitivity. He acknowledged that temporally the injury had triggered his recovery but still attributed his recovery to the previous therapy. After a week or so, full natural vision returned, and he has remained in remission for 15 months. He is similarly aware of the potential for relapse.

Discussion

These two cases highlight novel multimodal and ultimately successful approach to the management of functional visual loss in two patients with persistent symptoms. Management began during the consultation with a positive diagnosis incorporating explanation and clinical assessment of visual ability to demonstrate the integrity of visual pathways. We focused especially on positive diagnostic and experiential features to help patients gain confidence in the diagnosis of functional visual loss and in the potential for visual recovery. This approach was inspired by how often such transparency in explaining how the signs work appears to help patients with functional motor disorders[9]. The four methods described above all centre on the recognition and demonstration of visual ability and reversibility. TMS equipment is often available in clinical neurophysiology departments where they are already being used for diagnostic purposes. It does not take long to train an interested neurologist to use one for these, non-diagnostic purposes. If someone with hypnotherapy skills is not available then various professional organisations, eg the British Society of Academic and Clinical Hypnosis (www.bsach.com) may be able to help.

Available literature on the treatment and prognosis of functional visual loss in adults have concentrated on the use of reassurance, follow-up and psychological therapies, with variable improvement in visual function over time. Kathol *et al* in a cohort study of 42 individuals with functional visual loss and a mean age of 32 years, found that 45% regained normal visual function

whilst 55% had persistent visual dysfunction at a mean follow-up of 53 months. No difference was observed between the use of reassurance alone or in combination with psychotherapy[10]. Barris *et al* found that 78% of the 45 individuals in his cohort with a mean age of 25.9 years showed improvement or normalisation of vision during a mean follow-up of 114 days with the use of a timetable for recovery consisting of reassurance and visual exercises[11]. Sletteberg *et al* found that 51% of the 41 individuals in his group reported good visual function as opposed to 49% reporting poor visual function at a mean follow-up of 2 years[12]. The latter two studies also found that individuals younger than 16 years old were more likely to recover normal visual function compared with older individuals. None of the studies give detail about specific approaches to therapy.

We acknowledge that recovery in these cases could have occurred spontaneously, or may have been related to non-specific contact with a clinician interested in their disorder (JS), rather than the elements of treatment identified. Nonetheless both patients identified similar themes to a separate clinician (JMY) discussing their views after recovery.

Functional visual loss represents a common disorder in neuro-ophthalmological practice. In one study it represented 12% of annual new patient referrals [13], although bilateral blindness like this is much rarer. Thus far very little work has been done looking at treatment for this group of patients and we hope this study provides some useful ideas for future developments. A standardised protocol for the use of occipital TMS and hypnotherapy in the treatment of functional visual loss would be helpful in advancing the utility of these therapeutic approaches. Subsequently the efficacy of these techniques in comparison to usual care in reducing the severity and duration of functional visual loss could be evaluated in a randomised controlled trial.

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KEY POINTS

1. Functional visual loss is a treatable condition. There are therapeutic options which can be initiated and offered to patients to aid visual recovery.

2. Regular therapeutic contact with a healthcare professional providing an explanatory model including explicit discussion about visual function appears to be important.

3. Occipital TMS and hypnotherapy are adjunctive treatment options which can also demonstrate potential for recovery.

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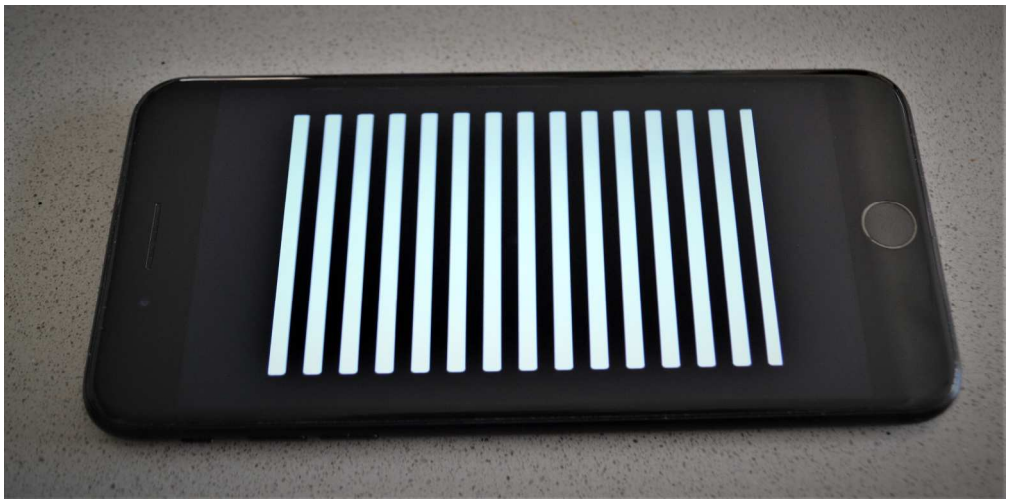

Figure 1. If you don’t have an optokinetic nystagmus drum, then use a YouTube video on a mobile phone – and ask the family to practice at home with it

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