

# Australian Orthoptic Journal

2018 Volume 50

**Duane's retraction  
syndrome and synergistic  
convergence**

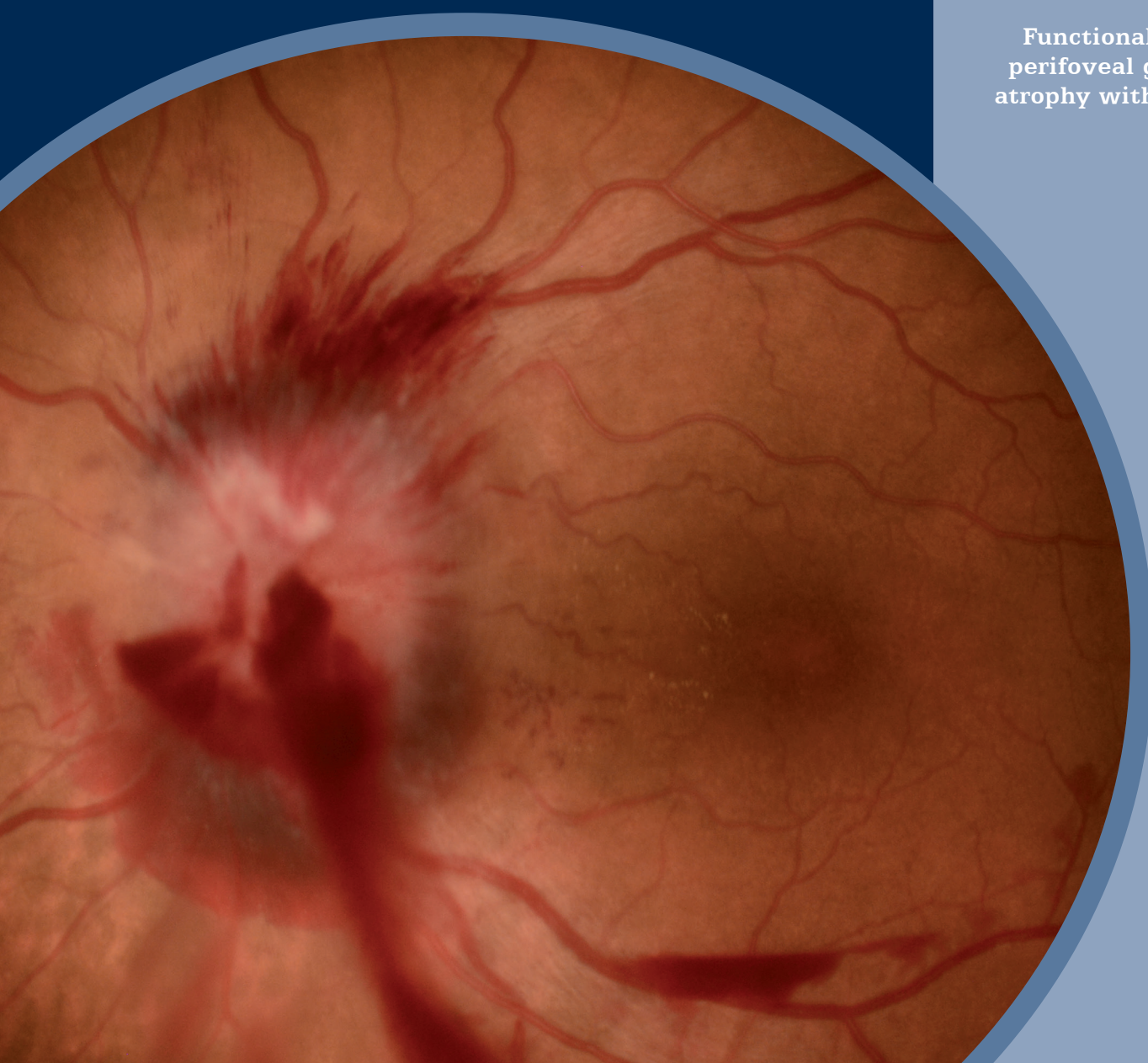
**Autoimmune retinopathy**

**A rare case of  
unexplained vision loss**

**Teaching orthoptics to  
ophthalmology residents**

**Excluding non-English  
speaking people from  
research**

**Functional impact of  
perifoveal geographic  
atrophy with dry AMD**



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# Australian Orthoptic Journal

2018 Volume 50

The official journal of Orthoptics Australia  
ISSN 2209-1262

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Published by Orthoptics Australia (Publication date: January 2019).

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## A Case of Duane's Retraction Syndrome and Synergistic Convergence

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### ABSTRACT

Duane's retraction syndrome (DRS) is a congenital eye movement disorder characterised by a limitation of horizontal gaze and narrowing of the palpebral fissure with globe retraction in adduction. Synergistic convergence, on the other hand, is a rare variant of DRS defined by simultaneous bilateral adduction on attempted lateral gaze. We report the case of an 11-year-old girl who presents with type III DRS of her right eye and synergistic convergence of her left eye.

**Keywords:** Duane's retraction syndrome, synergistic convergence

### INTRODUCTION

**D**uane's retraction syndrome (DRS), a mechanical ocular motility disorder, was classified by Huber into three categories based on the disordered horizontal motility and its corresponding electromyographic findings.<sup>1</sup> Type I is characterised by limitation of abduction, type II by limitation of adduction and type III by limitation of both abduction and adduction. All three types of DRS are associated with narrowing of the palpebral fissure and globe retraction on adduction, as well as the possibility of up and down shoots on adduction. Evidence suggests that DRS is caused by a misinnervation of the lateral rectus muscle by branches of the third cranial nerve instead of the sixth cranial nerve.<sup>1</sup> This misinnervation explains the oculomotor synkinesis noted on clinical examination.

Synergistic convergence is defined as simultaneous adduction, with an absence of pupil miosis on lateral gaze.<sup>2</sup> The condition is also associated with changes to the palpebral fissure and globe retraction. In the clinical setting, synergistic convergence can easily be misdiagnosed as DRS without careful examination of the eye movements and associated clinical signs. This paper discusses a child who

presented at the age of two with signs of bilateral ocular synkinesis who was later diagnosed at the age of eleven with DRS of one eye and synergistic convergence of the other eye.

### CASE REPORT

#### Initial presentation

A two-year-old girl presented to a tertiary hospital in Melbourne with a suspected ocular motility disorder. At this initial consultation, a bilateral limitation of abduction and adduction was noted in addition to narrowing of the palpebral fissures on adduction of either eye. On observation, it was noted that the patient had adopted a slight compensatory chin up head posture and cover testing revealed a small intermittent right esotropia in primary position at near and distance fixation. Unaided visual acuity was documented as 3/12 and 3/6 for her right and left eye respectively on Kay Picture testing.

The patient's birth and medical history indicated that she was diagnosed prenatally with oesophageal atresia with trachea-oesophageal fistula, which was surgically repaired one day post birth. She was also later diagnosed at the age of six years with congenital dextroscoliosis for which treatment was not prescribed. The family had no history of significant medical, neurologic, or ophthalmic disease.

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Accepted for publication: 26th March 2018

At this time the patient was diagnosed with a bilateral type III DRS and moderate right strabismic amblyopia. She received occlusion therapy for amblyopia from two to three years of age.

**Follow-up presentation**

At the most recent ocular examination, at age 11, the patient demonstrated best-corrected vision of right 3/3.8 and left 3/3 (-2), on the LogMAR chart. Her full refractive correction was right eye +4.50 DS and left eye +3.50 DS. A small esophoria with rapid recovery was detected in primary position, tested at both near and distance fixation, with and without correction. There was no obvious compensatory head posture when examined at the most recent consultation. The patient was able to achieve 55" stereoacuity with the Frisby Stereotest and 200" with the Lang Stereotest, both assessed with best correction.

Initially on ocular movements, limitation of both abduction and adduction was noted for both the right and left eyes. Globe retraction and narrowing of the palpebral fissure was noted on attempted abduction and adduction of either eye, left eye more than right. However, on carefully observing left gaze, simultaneous bilateral adduction was observed; that is the left eye was noted to converge on attempted abduction. Vertical gaze appeared to be intact. Figure 1 shows the eye movements in nine positions of gaze and the video (<https://www.youtube.com/watch?v=hO6OB47o3RM>) the eye movements on right and left gaze.

Pupils were equally reactive to light and to an accommodative stimulus with no evidence of pupil miosis on attempted lateral gaze. No fundus or media abnormalities were noted on ocular

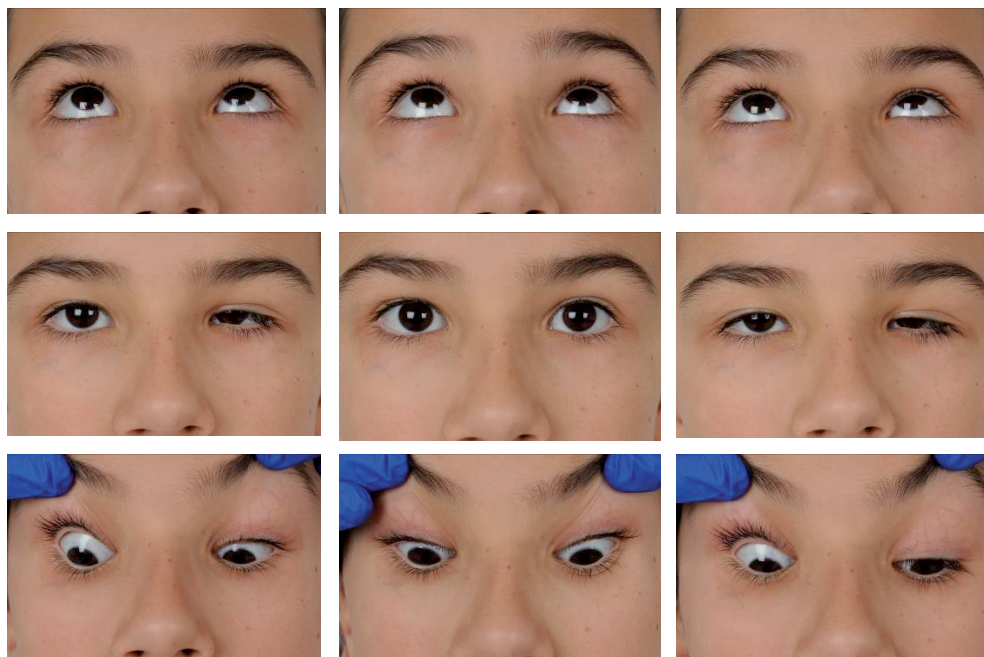
examination. On the basis of the follow-up findings, the patient's diagnosis was reclassified as a right type III Duane's retraction syndrome and left synergistic convergence. Informed consent was obtained from the patient for publication of this case report.

**DISCUSSION**

Synergistic convergence is a rare form of oculomotor synkinesis characterised by bilateral adduction on lateral gaze.<sup>3,4</sup> Whilst there are several documented cases of synergistic convergence, to the authors' knowledge this is the first patient reported to exhibit an isolated DRS of one eye and isolated synergistic convergence of the other eye.

The main characteristic which facilitated the redefinition of this patient's diagnosis from DRS to synergistic convergence of the left eye was the bilateral adduction on left gaze. It is likely that the left eye movements were misinterpreted as DRS due to the similar clinical signs noted between the two conditions and the presence of right DRS. At the follow-up visit, substituted convergence was more easily excluded as a diagnosis given the absence of pupillary changes on attempted abduction. Substituted convergence is associated with bilateral adduction on lateral gaze and is distinguished from synergistic convergence by pupil miosis on attempted lateral gaze.<sup>2</sup> It has also been linked to patients who have cerebellar, posterior fossa or pontine lesions.<sup>4</sup>

Given the lack of pupillary involvement noted in synergistic convergence, it is suggested that it is most likely caused by peripheral mechanisms.<sup>4</sup> Peripheral causes noted to be



**Figure 1.** Eye movements in nine positions of gaze.

associated with the condition include congenital fibrosis of extra-ocular muscles (CFEOM), congenital cranial dysinnervation syndrome and ocular misinnervation.<sup>5</sup> Central causes, which are rarely involved, include progressive scoliosis and brainstem dysplasia.<sup>5</sup> According to Jain et al,<sup>5</sup> horizontal gaze palsy and progressive scoliosis (HGPPS) also shares a previously recognised association. In our patient's case, the presence of scoliosis and synergistic convergence could potentially be linked to an underlying congenital disorder of the pons, specifically congenital cleavage of the pons or brainstem dysfunction.<sup>5,6</sup> However, magnetic resonance imaging (MRI) was not performed so the presence of any cortical changes cannot be confirmed.

More recent research also suggests genetic mutations linked to synergistic convergence. For instance, a homozygous mutation in the gene *ROB03* has been linked to progressive scoliosis and horizontal gaze palsies, which may suggest a similar mutation in this patient.<sup>7</sup> However, significant phenotypic variability can exist among individuals with identical genetic mutations, suggesting that other mechanisms could be involved in the clinical presentation.<sup>8</sup>

Finally, Pieh et al<sup>3</sup> have proposed that isolated synergistic convergence is associated with aberrant nerve sprouting during embryogenesis. When considering its counterpart, synergistic divergence, the suspected cause would be aberrant innervation of the oculomotor nerve fibres to the lateral rectus.<sup>9,10</sup> In this case, an assumed miswiring of the abducens motor neurons to the medial rectus muscle is more likely to be the cause. This is demonstrated by the ocular movements of the patient's left eye; adduction on attempted left lateral gaze, globe retraction and narrowing of the palpebral fissure on attempted abduction and adduction. However, as this is an exceptionally rare condition with variable associations, the current literature is unlikely to offer a definitive etiopathogenesis.<sup>2</sup> This patient would require further testing to identify the most likely cause of the ocular motility disorder, such as MRI of the brain and orbits and genetic testing. In this case, it is noteworthy that the underlying cause of this ocular motility disorder is unlikely to influence the ongoing management of this patient's ocular condition. In primary position the patient is orthotropic and demonstrates binocular single vision. She also manages the limited eye movements in lateral gaze through the use of head movement and as such maintains good cosmesis, despite the ocular motility disorder.

## CONCLUSION

This case study suggests that Duane's retraction syndrome and synergistic convergence can be diagnosed in a single patient. In the presence of DRS, it may be difficult to observe the nuanced differences between the two conditions, however careful inspection of the eye movements, the palpebral fissures, and pupillary reactions can distinguish

synergistic convergence from DRS and substituted convergence. Whilst synergistic convergence is rare, clinicians should be mindful that a patient who presents with bilateral oculomotor synkinesis can have a principal diagnosis that differs between the two eyes.

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## Autoimmune Retinopathy: Three Case Reports

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### ABSTRACT

Autoimmune retinopathy (AIR) is an uncommon condition which should be considered when a patient presents with unexplained visual loss. The three main forms are melanoma-associated retinopathy (MAR), cancer-associated retinopathy (CAR) and non-paraneoplastic autoimmune retinopathy (npAIR).

The visual loss is painless and can decline either rapidly or gradually. Symptoms may include reduced best-corrected visual acuity (BCVA), photopsias which are often described as flickering or shimmering effects, visual field loss, nyctalopia, loss of colour vision and delayed adaptation to changing light conditions. AIR is usually, but not always, bilateral and may be asymmetrical. The fundus exam and optical coherence tomography (OCT) are usually normal, however the electroretinogram (ERG) results are abnormal. The damage to retinal photoreceptors occurs when anti-retinal antibodies are created by an autoimmune reaction to retinal proteins. Serology may reveal the presence of a

range of anti-retinal antibodies but is not diagnostic without concurrent clinical manifestations. Immunosuppressive treatment may limit disease progression.

Patients with AIR are often misdiagnosed or have their diagnosis delayed. In patients with unexplained visual loss, particularly if their symptoms include photopsias, it is important to take a comprehensive medical history and undertake extensive clinical testing, including BCVA, OCT, visual field, colour vision testing, ERG and antiretinal antibody serology.

This paper presents a case each of CAR, MAR and npAIR.

**Keywords:** autoimmune retinopathy, cancer-associated retinopathy, melanoma-associated retinopathy, non-paraneoplastic retinopathy, photopsia, electroretinogram

### INTRODUCTION

**A**utoimmune retinopathy (AIR), causes visual loss when anti-retinal antibodies are created which damage the retinal photoreceptors. AIR often presents as unexplained painless visual loss, with shimmering or flickering photopsias, visual field loss, loss of colour vision, nyctalopia and photosensitivity. The fundus exam is usually normal but in some cases disc pallor, retinal vascular attenuation and retinal pigment epithelial changes may be present. The onset can be sudden or gradual, can rapidly or slowly progress, is usually but not always bilateral and may be asymmetrical. AIR is a rare condition and the prevalence is unknown.<sup>1</sup> It is often misdiagnosed, or diagnosis is delayed. There are three categories of AIR, cancer-associated retinopathy (CAR), melanoma-associated retinopathy (MAR) and non-paraneoplastic retinopathy

(npAIR).<sup>1,2</sup>

Cancer-associated retinopathy predominantly affects cone function, with symptoms including photosensitivity, decreased visual acuity, impaired colour vision and difficulty seeing in bright light. It may also affect rod function in some patients. It develops over weeks or months. Visual field testing often shows a central or para-central scotoma. It can be associated with a number of cancers including small-cell lung carcinoma, gynaecological cancers, breast cancer, colon cancer, solid tumours and blood cancers.<sup>1,3-5</sup> CAR presents before a cancer has been diagnosed in approximately half of all cases or it may indicate metastases.<sup>1,5</sup> CAR is a progressive condition that ultimately leads to loss of vision in both eyes.<sup>4</sup> CAR predominantly affects the photoreceptors and the electroretinogram (ERG) typically shows global retinal dysfunction and may be severely reduced or extinguished.<sup>5</sup>

Melanoma-associated retinopathy usually presents as nyctalopia and shimmering photopsias with a normal

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Accepted for publication: 27th March 2018



fundus exam.<sup>6</sup> Patients may also describe flickering or pulsating photopsias.<sup>7</sup> Visual field testing usually shows peripheral constriction and the ERG shows reduced or extinguished rod function and an electronegative waveform (where the b-wave is smaller than the a-wave) to a bright white flash in dark-adapted conditions. Cone function may be well preserved.<sup>1,6</sup> Visual acuity and colour vision may be unaffected.<sup>7</sup> MAR commonly presents months or years after the diagnosis of melanoma has been made, often at the stage of metastases and may stabilise rather than progress.<sup>4</sup>

The presentation of non-paraneoplastic autoimmune retinopathy is more variable than in MAR and CAR, and may include reduced BCVA, nyctalopia, visual field loss and photopsias, in the absence of malignancy, and is associated with auto-immune disease in approximately 50 percent of cases.<sup>1</sup> ERG results are also variable and can show cone and/or rod dysfunction. Immunosuppression treatment has been effective in halting progression or even improving vision in some patients.<sup>8,9,10</sup> The diagnosis of npAIR is only made after comprehensive testing for evidence of malignancy using tests such as magnetic resonance imaging (MRI), computed tomography (CT), positron emission tomography (PET) scans, ultrasounds and blood tests.

The body's autoimmune system creates autoantibodies against retinal proteins which have cytotoxic effects on the retinal cells. Several retinal proteins associated with autoimmune retinopathy have been identified including

recoverin, arrestin, transducin- $\beta$ , rhodopsin and  $\alpha$ -enolase.<sup>1,6</sup> Patients can have a wide range of anti-retinal antibodies identified, often with three to six different antibodies present.<sup>2</sup>

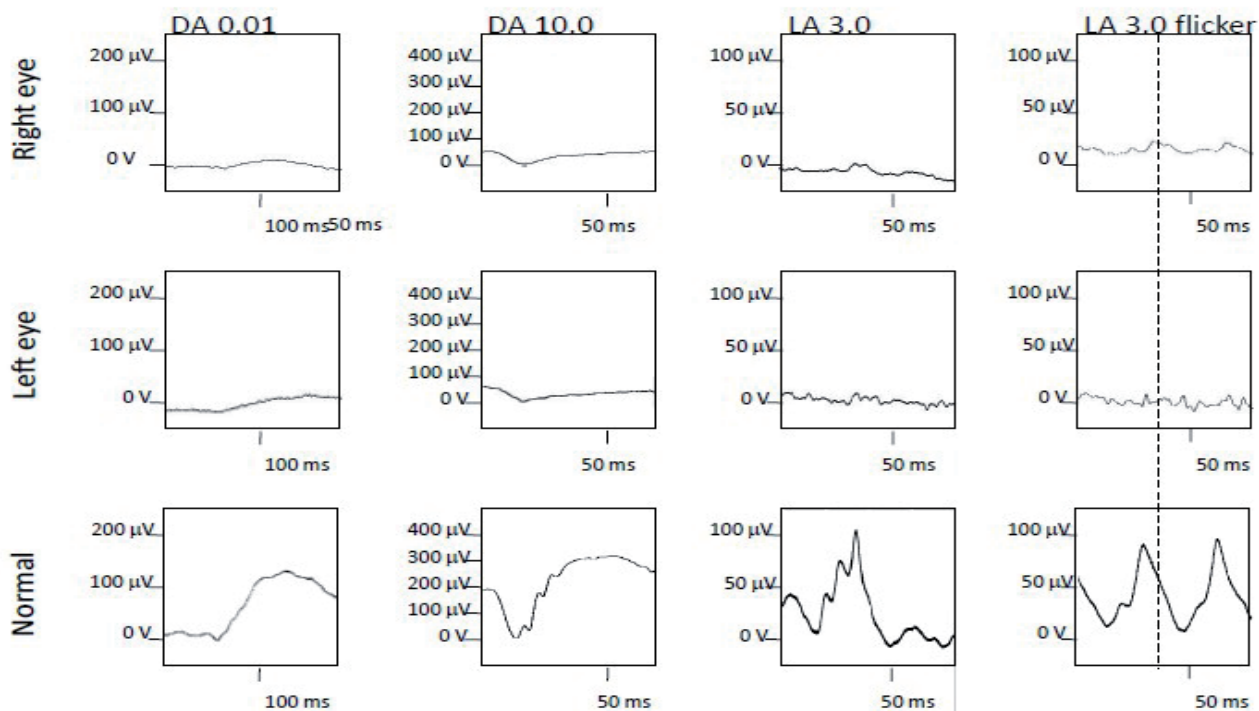
Treatment options include steroids, conventional immunosuppressives such as antimetabolites and T-cell inhibitors, monoclonal antibodies such as rituximab and intravenous immunoglobulin (IVIG),<sup>11</sup> however treatment may not prevent progression.

## CASE REPORTS

### Case Report 1

An 82-year-old man presented with a fifteen-month history of progressively declining vision not improved by cataract surgery which had been performed twelve months prior to presentation. He complained of problems with glare, difficulty identifying colours, nyctalopia and impaired ability to adapt to changes in lighting conditions. Five months after the cataract surgery he had undergone surgery to remove a gastric tumour. In this case the onset of visual loss preceded the cancer diagnosis.

His visual acuity was recorded as RE Hand Movements and LE Count Fingers. The patient's low level of vision precluded formal visual field and colour vision testing. Disc pallor was noted on his fundus exam. The ERG showed reduced and delayed responses in both dark-adapted (scotopic) and light-adapted (photopic) conditions, indicating that both



**Figure 1.** Case 1, cancer-associated retinopathy: International Society for Clinical Electrophysiology of Vision (ISCEV) ERG demonstrated reduced and delayed responses in both scotopic and photopic conditions, indicating that both rods and cones were widely affected.

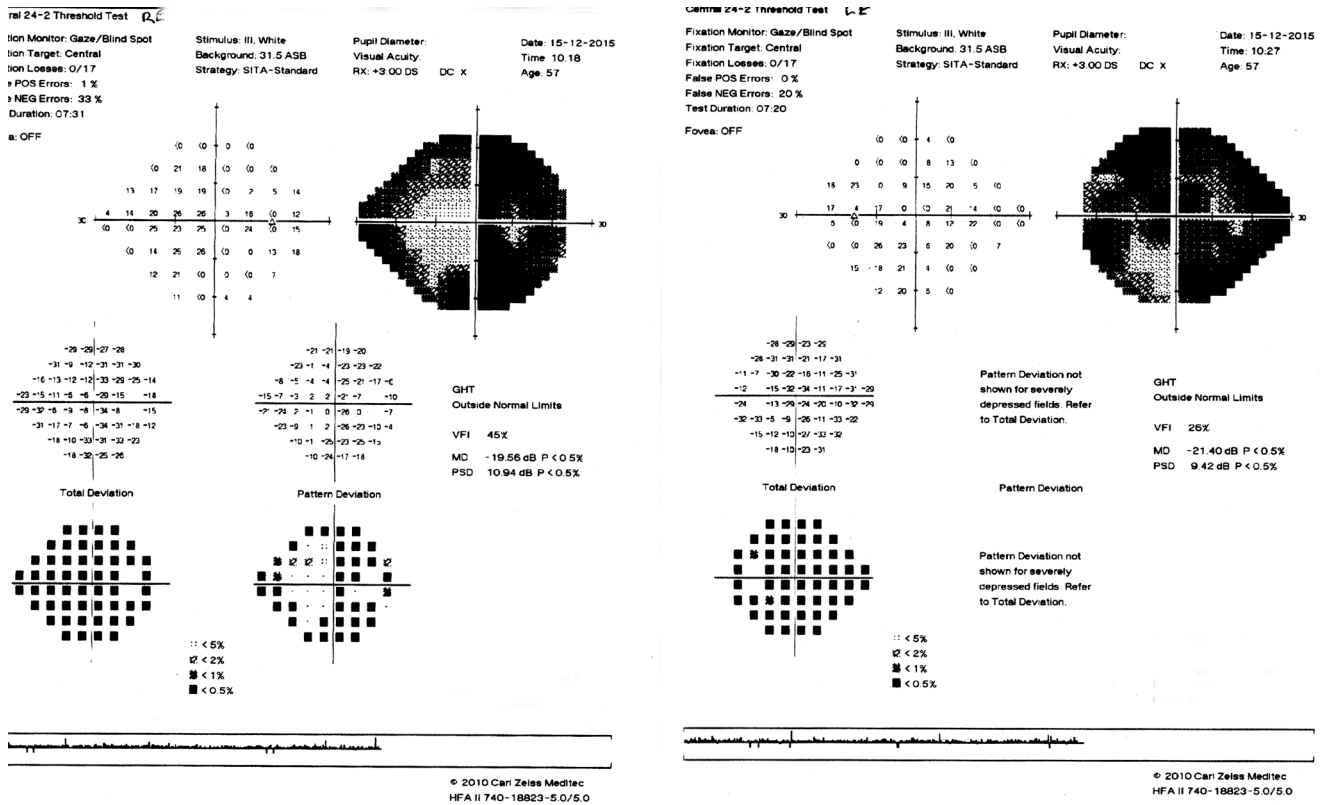


Figure 2. Case 2, 24-2 SITA Standard threshold Humphrey visual field testing demonstrated extensive visual field loss in both eyes.

rods and cones were widely affected (Figure 1).

Anti-retinal antibody serology was positive, leading to a diagnosis of CAR. The patient’s neuro-ophthalmologist noted that ‘the immune response initiated by this malignancy has resulted in collateral damage to both retinae, permanent and irreversible. Immunosuppression would probably not help but is worth a try’. A two-month course of oral prednisolone did not result in any improvement in vision. Tinted lenses were prescribed to manage glare.

**Case Report 2**

A 58-year-old male presented complaining of visual field loss, floaters, photopsias, nyctalopia and difficulty adjusting to changing light conditions over the previous two years, especially noticeable in the previous twelve months. He had decided to cease driving at night. He had had a Stage III melanoma excised four years prior. His father had died from malignant melanoma some years before.

His BCVA was RE 6/6, LE 6/7.5. Ishihara colour vision testing showed all plates correct with slower responses with the left eye and his OCT was normal. 24-2 SITA Standard threshold Humphrey visual field testing showed extensive visual field loss in both eyes (Figure 2). ERG demonstrated reduced and delayed rod responses, an electronegative waveform to a bright flash in dark-adapted conditions and cone responses within normal limits (Figure

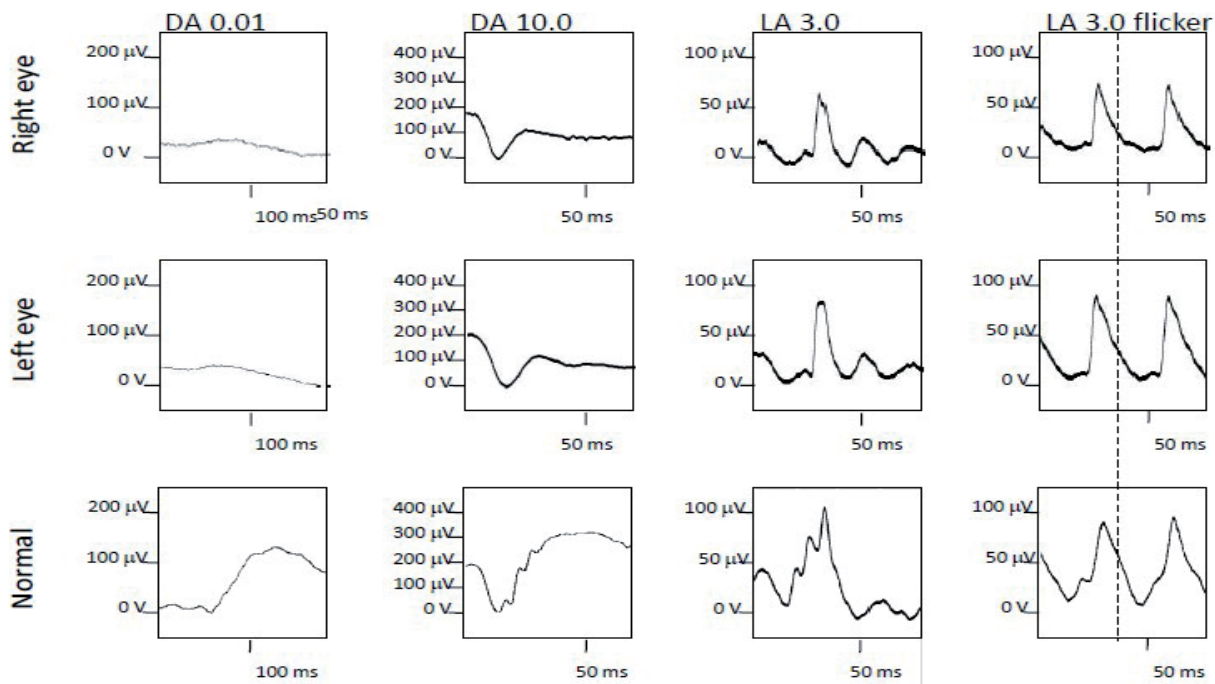
3). A diagnosis of MAR was made.

Management was commenced with three-monthly skin checks, six-monthly positron emission tomography (PET) scans and regular exams with his ophthalmologist and oncologist. A follow-up ERG twelve months later was stable. BCVA was also stable but the patient reported worsening of his photopsias and increasing glare sensitivity.

Immunosuppression was not recommended by his oncologist due to the possible destabilisation of the melanoma.

**Case Report 3**

A 62-year-old male presented with a two-month history of declining visual acuity, photopsias, photophobia, ‘patchy’ vision and shimmering effects ‘like a kaleidoscope’ which he found very disturbing. At times the photopsias were so intense it caused the patient great distress, headaches and nausea. The shimmering was present even when his eyes were closed, causing fatigue, insomnia, depression and loss of appetite. The patient’s medical history included diabetes mellitus type 1 diagnosed at age 21 years and polymyalgia. OCT and fundus examination showed evidence of previous panretinal laser treatment in the right eye, with normal discs and maculae. He had seen numerous medical specialists and undergone many investigations before he was finally referred for an ERG.



**Figure 3.** Case 2, melanoma-associated retinopathy: ISCEV ERG demonstrated reduced and delayed rod responses, an electronegative waveform to bright flash in scotopic conditions and cone responses within normal limits.

Visit 1: BCVA RE 6/15 LE 6/24. Ishihara RE 11/16 LE 10/16 plates correct.

Visit 2 (6 months later): BCVA RE 6/15 LE 6/24. Ishihara RE 1/16 LE 1/16 plates correct.

Visit 3 (9 months after Visit 2): BCVA RE 6/38 LE 6/120. Ishihara RE 0/16 LE 0/16 plates correct.

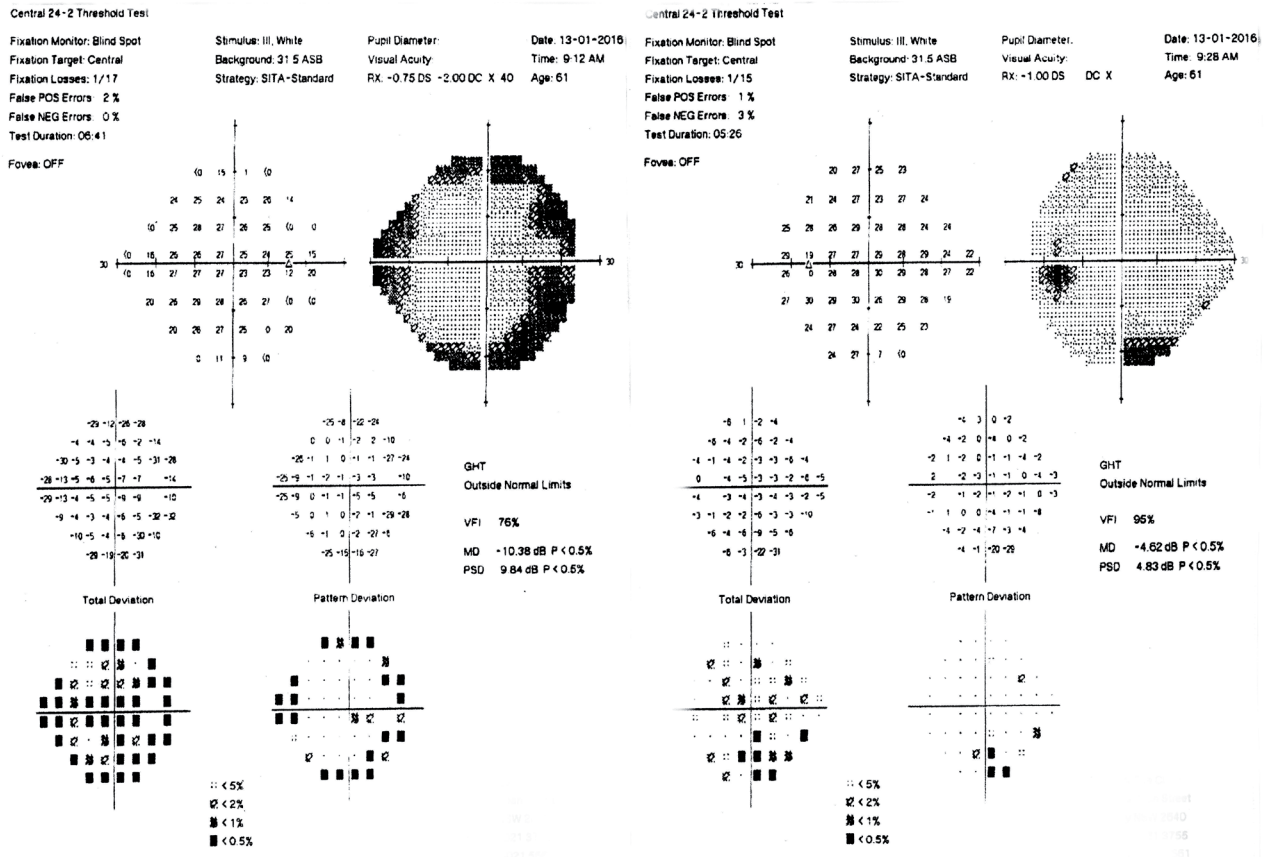
Note that at the second visit the BCVA was stable but colour vision had markedly declined. 24-2 SITA Standard threshold Humphrey visual field testing showed visual field loss in both eyes, more extensive in the right eye (Figure 4). The ERG demonstrated progressive decline in rod and cone function and delayed latencies over three visits (Figure 5).

All tests for signs of cancer including a lymph node biopsy were negative and a diagnosis of npAIR was made. Anti-retinal antibody serology was positive. Management consisted of a three-day course of IV prednisolone which did not result in any improvement in the patient's vision and caused elevated blood sugar levels. Three treatments of IV immunoglobulin (IVIg) over three months did not prevent a continuing decline in vision. The patient could not tolerate mycophenolate and was forced to give up driving and his employment due to his greatly reduced vision and his photopsias. The patient then underwent an ongoing course of plasma exchange treatment with the aim of diluting the levels of circulating anti-retinal antibodies. After eight months of treatment his haematologist reported that the patient's vision had stabilised.

## DISCUSSION

At present there is no definitive treatment of choice for AIR and evidence for therapeutic intervention is limited.<sup>12</sup> The presence of serum antiretinal autoantibodies alone does not lead to a diagnosis of AIR as they can be present in other conditions such as uveitis.<sup>13</sup> Autoimmune retinopathies including CAR, MAR and npAIR are rare and there is a need to develop diagnostic criteria and treatment protocols. In 2013 a panel of uveitis and immunology clinicians and researchers met to develop such criteria and protocols.<sup>11</sup> It was agreed that essential diagnostic criteria should include unexplained vision loss, ERG abnormality with or without visual field loss, the presence of serum anti-retinal antibodies and the absence of fundus lesions, retinal degeneration, retinal dystrophy and overt intraocular inflammation. Core tests should include malignancy evaluation, ERG and serum anti-retinal antibody testing. Follow-up testing should include ERG, visual field testing, BCVA, OCT and colour vision testing. The three case reports described here fit the criteria for cases of CAR, MAR and npAIR respectively.

Randomised placebo-controlled trials are required to evaluate the efficacy of long term immunosuppressive treatment.<sup>11</sup> In CAR, 'despite aggressive immunosuppressive therapy, the visual prognosis is poor, and rapid, relentless visual loss often occurs',<sup>5</sup> although there have been reports of some patients responding to various combinations of treatments such as corticosteroids, plasma exchange, cyclosporine, mycophenolate, IVIg and rituximab.<sup>4,5</sup>



**Figure 4.** Case 3, 24-2 SITA Standard threshold Humphrey visual field testing demonstrated visual field loss in both eyes, more extensive in the right eye.

As with CAR, treatment for MAR has been largely ineffective, but with reports of success in some individual cases.<sup>4,5</sup> Yamamoto et al<sup>14</sup> reported a patient with MAR who underwent resection of metastatic melanoma and monthly injections of interferon-β resulting in normal ERG and vision. However, it is believed that the presence of the autoimmune response may be protective, helping to eradicate melanoma cells and prevent tumour spread, which can be a reason to withhold immunosuppression in cases of MAR.<sup>5</sup>

In npAIR first-line treatment is usually local or systemic steroids and conventional immunosuppressives followed by monoclonal antibodies and IVIg.<sup>11</sup> In a case report presented in Choi et al<sup>12</sup> a patient with npAIR was treated with oral prednisolone, cyclosporine A and azathioprine, however the vision continued to worsen progressively. In some cases of npAIR visual acuity, ERG and visual fields have stabilised or improved when treated with rituximab, a chimeric monoclonal antibody, but long-term effects are not yet known.<sup>8,9,10</sup>

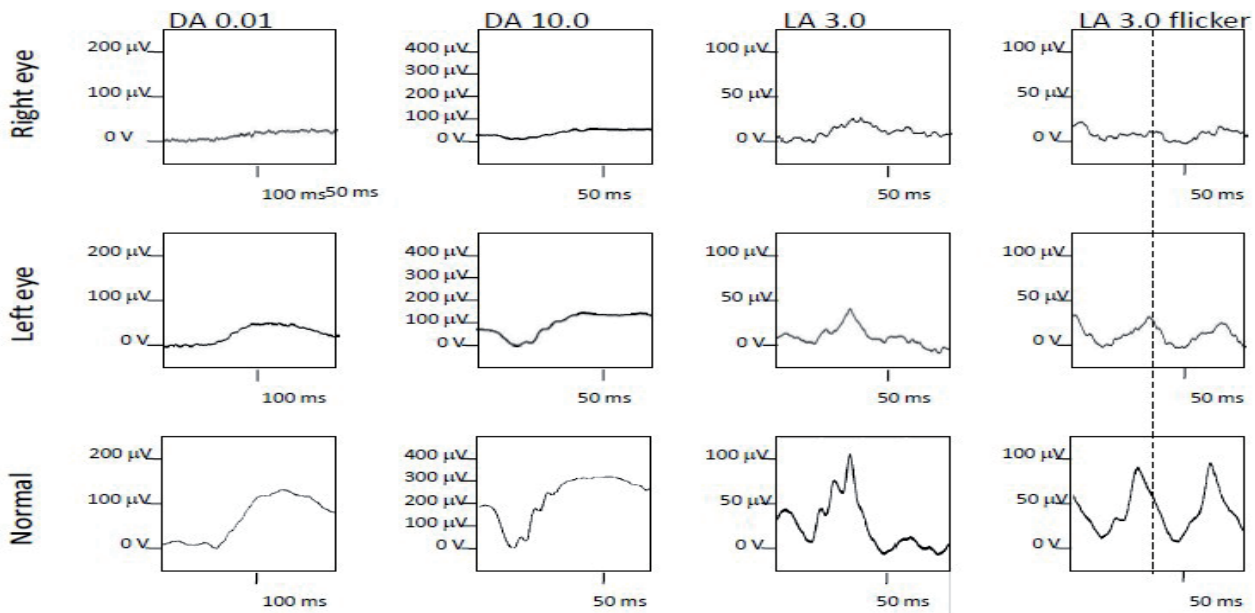
Patients with unexplained painless visual loss are referred to our clinic for an ERG. The referral from their ophthalmologist may or may not include OCT, visual field and blood test results. A comprehensive medical and family history is taken, and visual acuity and colour vision

are tested prior to the ERG. The patient then returns to their referring ophthalmologist and if treatment is required this is managed by medical specialists which may include oncologists, haematologists, rheumatologists and neuro-ophthalmologists. If treatment is undertaken the patient may return for a follow-up ERG as this is an objective way of measuring improvement, stability or progression.

**CONCLUSION**

Patients with AIR are often misdiagnosed or have their diagnosis delayed, which may lead to poor visual and health outcomes. Patients with painless subacute unexplained vision loss, particularly when complaining of photopsias, need comprehensive testing. Patients with AIR nearly always describe a shimmering effect. Testing within a general ophthalmology practice should include BCVA, colour vision testing, perimetry, OCT and careful questioning of their general health and medical history, specifically with regard to any history of cancer, melanoma or auto-immune disease. Serology to detect the presence of anti-retinal antibodies should also be undertaken. A full-field ERG is the most clinically important test in the diagnosis of AIR, MAR and





**Figure 5.** Case 3, non-paraneoplastic autoimmune retinopathy: ISCEV ERG demonstrated delayed latencies and progressive decline in both rod and cone function.

CAR, measuring the function of the retinal photoreceptors. An abnormal ERG in the presence of a normal or near-normal fundus examination is a key diagnostic criterion in the diagnosis of AIR, MAR and CAR. It is also important as a test of exclusion: a normal ERG in a patient with these symptoms would lead to further neurological testing.

## ACKNOWLEDGEMENTS

The author wishes to acknowledge and thank Dr Heather Mack for supplying the ERG slides and for sharing her expertise in the diagnosis and management of AIR, and Linda Santamaria for her outstanding editorial advice and endless patience.

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# A Rare Clinical Case of Unexplained Unilateral Vision Loss on a Background of Metastatic Breast Cancer

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## ABSTRACT

A rare clinical case of unexplained unilateral vision loss occurring in a 59-year-old Caucasian female is described. The vision loss occurred in association with an ipsilateral afferent pupil defect and on a background of primary breast carcinoma with extensive bony metastases. However, there was no clinically detectable orbital infiltration, orbital pseudo-tumour, compressive visual pathway lesion, optic neuritis or retinal pathology that could account for the ocular symptomatology. The patient was receiving palliative oral capecitabine chemotherapy at the time of presentation.

Findings on clinical examination, perimetry testing, fundus photography, optical coherence tomography and magnetic resonance imaging are presented. Other differential diagnoses underlying this case of severe, sub-acute vision loss are discussed.

**Keywords:** breast cancer, metastatic, capecitabine, chemotherapy, cytotoxicity

## INTRODUCTION

The development of vision impairment in the setting of an underlying malignancy is not uncommon and can range from mild to severe with unilateral or bilateral involvement. Vision loss is often but not always owing to the cancer itself or cancer-related treatment.<sup>1-7</sup> Specific causes of vision loss in patients with a history of cancer include metastatic involvement of the visual pathway;<sup>1-2</sup> paraneoplastic syndromes, such as cancer-associated retinopathy and melanoma-associated retinopathy;<sup>8-10</sup> ocular toxicity to chemotherapeutic agents;<sup>3,11</sup> and side effects from irradiation, particularly that applied to the head and/or orbits.<sup>5-7</sup>

Of note, the ocular side effects of both old and new-aged cancer therapies are emerging as more longitudinal data arises and the frequency of reporting increases.<sup>11</sup> In some cases, vision loss can precede the development of cancer in a patient.<sup>2,10,12</sup> When this occurs, one aetiology of consideration is non-paraneoplastic autoimmune retinopathy/optic neuropathy.<sup>2,10,12</sup> This atypical immune-mediated process occurs in the absence of any known systemic malignancy and can precede the development of cancer by months or even years.<sup>2,10,12</sup>

Some cases of vision loss occurring in the setting of cancer represent a diagnostic dilemma and the aetiology remains unclear. In such cases, a presumptive diagnosis is sometimes made by combining clinical features, examination findings and ancillary test results into a cohesive rationale. In the absence of any concrete findings, the diagnosis is reached



**Figure 1.** MRI of the brain and orbits showing bilateral orbital inflammatory process (L>R). Fusiform severe swelling of the left medial rectus, mild swelling of the left lateral and inferior recti, and slight swelling of the right medial and lateral recti.

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 Accepted for publication: 12th June 2018

by a process of exclusion rather than being definitive. Whilst no definitive diagnosis is available, these atypical cases are deserving of attention and it is equally important that they be reported such that other cases with similar phenotypical presentation may, when taken collaboratively, cast insight on the medically unknown.

We describe one patient with a background of advanced metastatic breast cancer who experienced a severe, unilateral loss of vision and had an accompanying ipsilateral afferent pupil defect that could not be explained by obvious aetiological factors. The patient was receiving palliative oral capecitabine chemotherapy at the time of presentation. Neuro-imaging studies and ophthalmic examination did not reveal any clinical evidence for the ocular symptomatology. The clinical findings are described, and several differential diagnoses are discussed. In discussing this case, we seek to highlight the complexity of aetiologies underlying vision impairment in cancer patients.

CASE REPORT

A 59-year-old Caucasian female was referred by her medical oncologist to an ophthalmology clinic complaining of a four-week history of right-sided orbital pain, especially on horizontal gaze, and a gradual worsening of vision in this eye. There was no history of floaters or photopsia, and no changes were reported in the left eye. There was no significant past ocular history and no family history of eye disease.

The patient had a past medical history of controlled hypertension and depression for which she was taking oral irbesartan and venlafaxine. In addition, she had a de novo presentation of Stage IV, oestrogen-receptor-positive, human epidermal growth factor receptor 2 (HER2)-negative breast carcinoma with widespread bony metastases but no visceral disease. Following her initial diagnosis in March 2015, the patient commenced palliative treatment with letrozole and denosumab, as well as radiotherapy to the cervical spine.

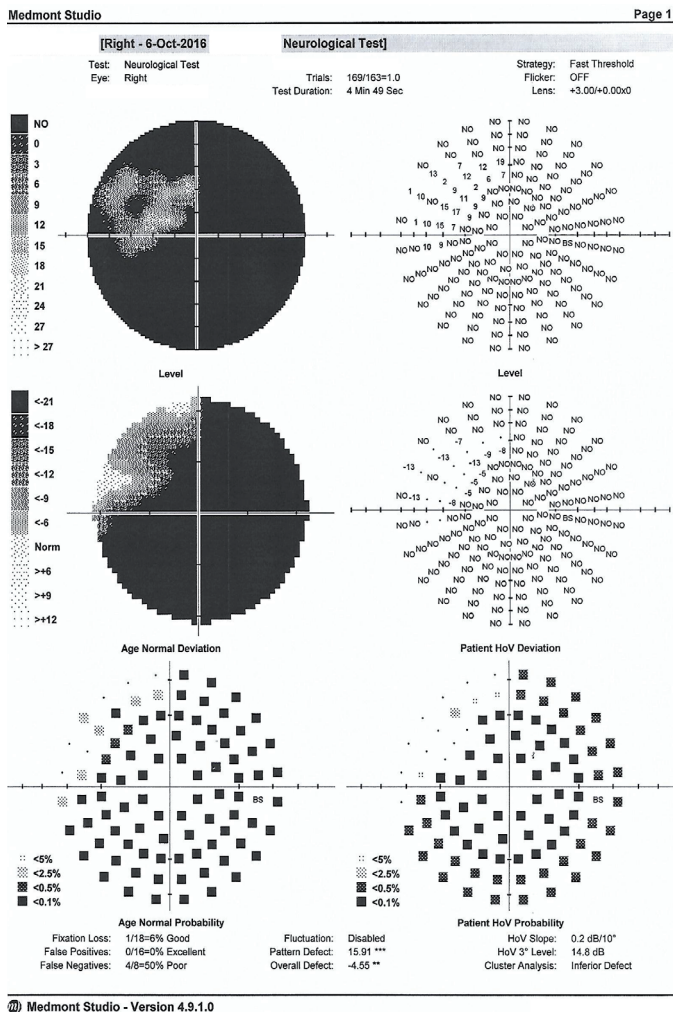


Figure 2. Medmont neurological visual field of the right eye at initial presentation showing severe widespread field loss with intact area superonasally.

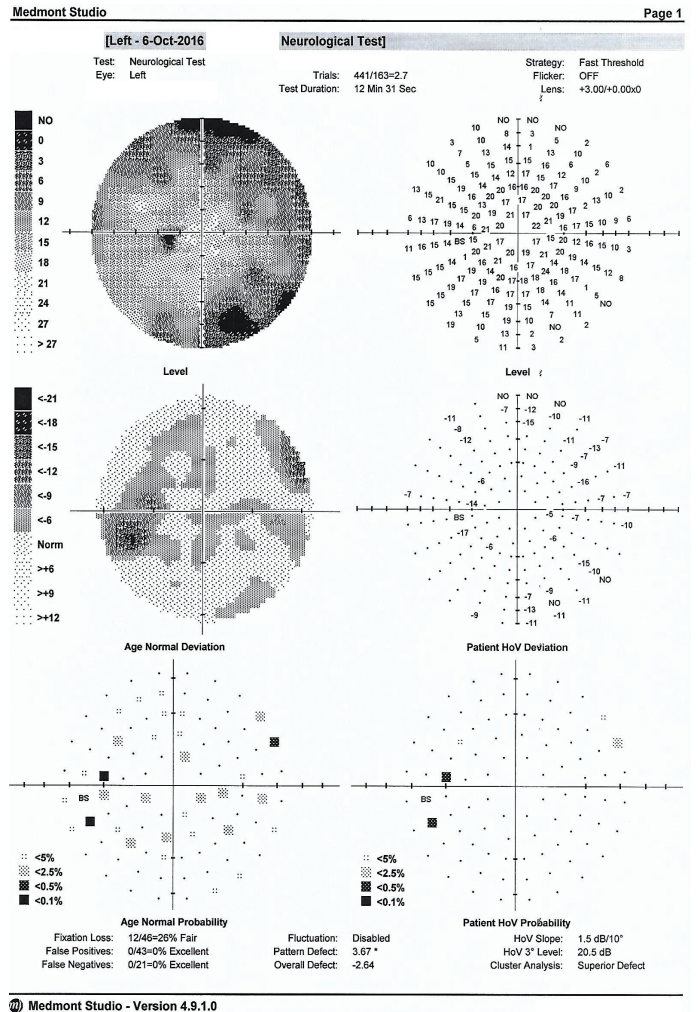
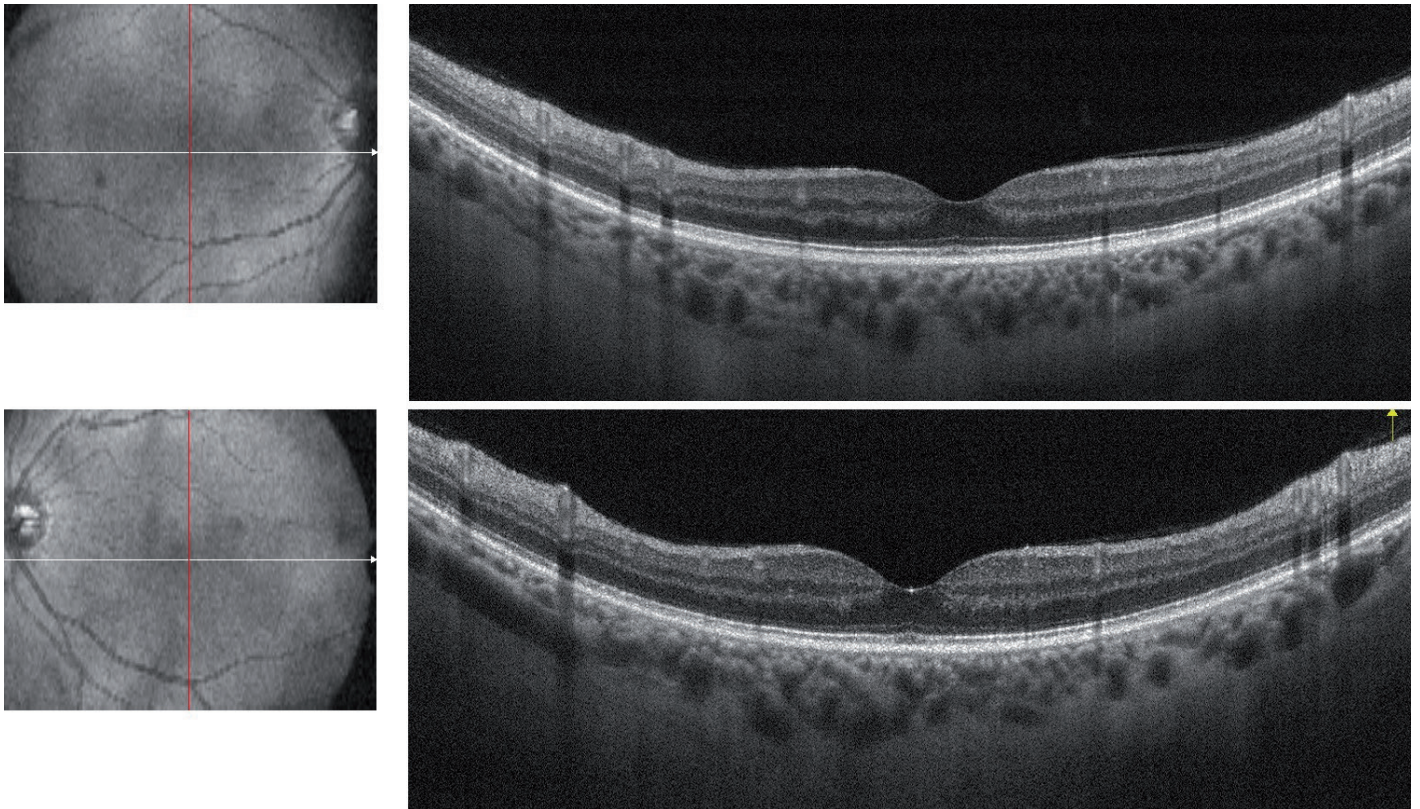


Figure 3. Medmont neurological visual field of the unaffected left eye at initial presentation.





**Figure 4.** Spectral-domain OCT of the right and left maculae, demonstrating a normal appearance.

Nine months after diagnosis, increasing metastases were detected, including bony progression and a new hepatic metastasis. The patient subsequently undertook three cycles of intravenous (IV) nab-paclitaxel chemotherapy, which was poorly tolerated. Whilst re-staging demonstrated improvement in the liver metastasis, there was progression of her bony disease. She was then switched to oral capecitabine chemotherapy and was continued on denosumab. A steady improvement in all areas of metastatic disease was demonstrated on subsequent imaging.

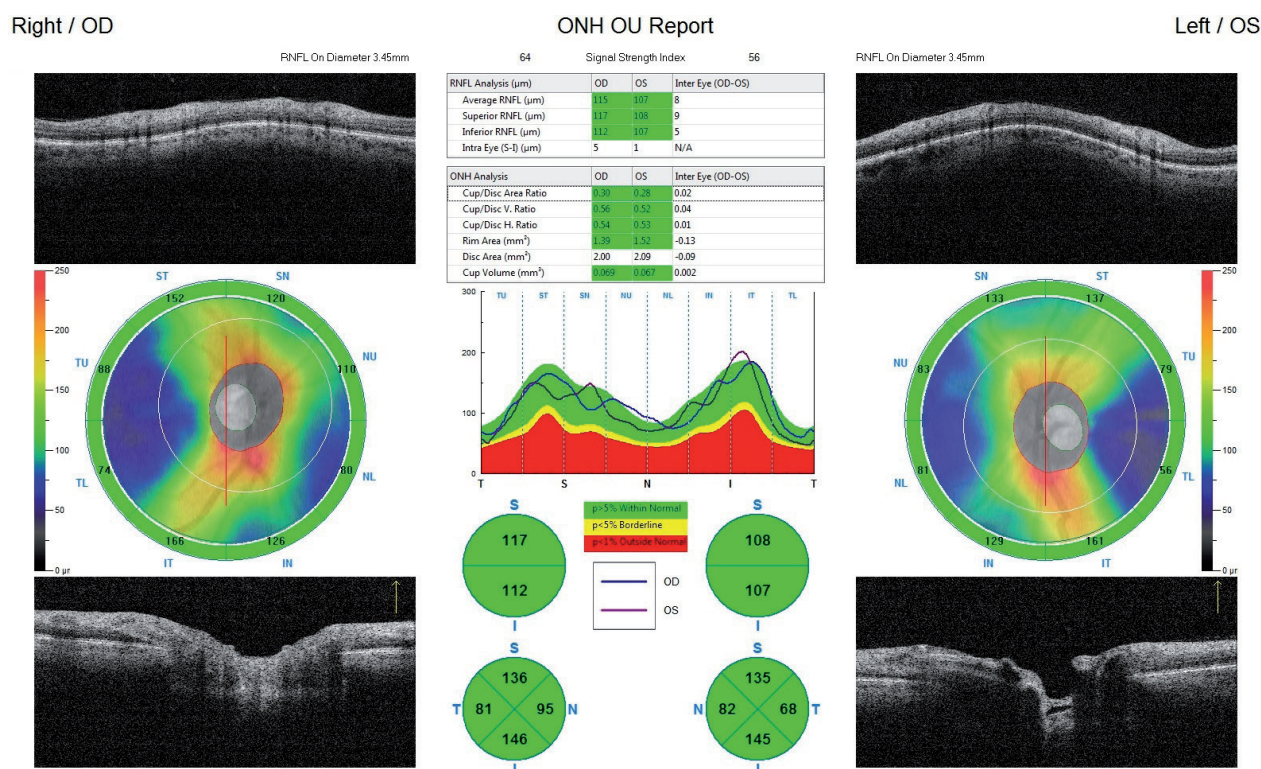
Oral capecitabine chemotherapy was continued for six months when new onset ocular and visual symptoms developed, as described above. Magnetic resonance imaging (MRI) of the brain and orbits revealed bilateral orbital inflammatory changes, left greater than right, not convincing for orbital metastases, but rather suggestive of orbital pseudo-tumour or an orbital inflammatory process (Figure 1). There was marked thickening of the left medial rectus and to a lesser extent, left lateral and inferior recti. However, this was not consistent with the symptomatic right-sided vision loss. There was no evidence of superior ophthalmic vein or cavernous sinus thrombosis. At this point, capecitabine was interrupted and ophthalmological opinion sought.

Upon first presentation to the ophthalmology clinic, best-corrected visual acuity (BCVA) was right 6/48 and left 6/6, using a rear-illuminated Bailey-Lovie LogMAR chart.

Medmont neurological visual field testing confirmed severe, widespread field loss in the right eye with only a small intact arc of vision superonasally (Figure 2). The left visual field was full (Figure 3). Dilated fundus examination was unremarkable in each eye. Spectral-domain optical coherence tomography (OCT) revealed both maculae (Figure 4) and retinal nerve fibre layer (RNFL) (Figure 5) to be normal. Anterior segment examination was unremarkable. On Ishihara colour vision testing, the patient was unable to detect any plates with the right eye but demonstrated normal colour vision in the left. Light-brightness sensitivity was reduced in the right eye and a right afferent pupil defect was present. Ocular movements were full and no diplopia was reported, with no apparent exophthalmos or nystagmus. A presumptive diagnosis of right retrobulbar optic neuritis was made and the patient went on to commence same-day pulsed IV methylprednisolone delivered over four days, followed by high dose oral prednisolone (60mg daily) which was subsequently weaned at home.

Upon follow-up examination four days post hospital discharge, right visual acuity had improved negligibly to 6/36(pt) with steroid treatment, however the patient still had no useful vision in this eye owing to the extensive field loss and need for significant head movement to achieve this acuity. The left visual acuity remained 6/6. Repeat visual field testing was performed and the results remained unchanged. Light-brightness sensitivity had improved





**Figure 5.** Bilateral OCT RNFL scan depicting RNFL thickness within normal limits, healthy optic nerve heads and normal inter-eye cup-to-disc symmetry.

but right colour vision remained affected. The right optic disc appearance was unremarkable and anterior segment examination was normal. The patient remained off oral capecitabine.

Repeat MRI conducted three weeks later revealed persistent, severe fusiform swelling of the left medial rectus with enhancement, and mild to moderate oedema within the intraconal fat bilaterally, left greater than right (Figure 6). There were skull base and frontal bone metastases with mild pachymeningeal thickening and enhancement, right greater than left, as well as subtle nodularities in the right frontoparietal and temporal lobe regions consistent with leptomeningeal carcinomatosis. The appearance was now more suggestive of underlying orbital metastases rather than an inflammatory process, although there remained no obvious focal lesion in either orbit. A two-week course of bilateral orbital and skull base radiotherapy was undertaken.

A diagnosis of diffuse skull base metastatic disease with extension particularly into the left orbit seemed fitting, albeit the right-sided visual impairment was not consistent with the MRI findings of disease greater on the left than the right and there was no obvious, identifiable right-sided orbital infiltration. A subjective improvement in vision was reported following radiotherapy, but no objective improvement was observed.

Repeat brain MRI was undertaken three months later to determine the patient's response to radiotherapy. Imaging revealed that the pre-existing orbital soft tissue oedema

was basically unchanged from earlier studies. There was persistent bilateral orbital inflammatory process, most severe on the left side with marked swelling and enhancement of the medial and inferior recti, as well as marked oedema in the intraconal fat and around the globe (Figure 7). There remained no focal lesion apparent in either orbit to suggest metastasis causing this appearance. No further orbital radiotherapy was indicated. MRI also identified more advanced leptomeningeal changes in the posterior cranial fossa as well as extensive nodular pachymeningeal thickening around the entire brain (Figure 8). The patient subsequently underwent a two-week course of radiotherapy to the whole brain, excluding the orbits and optic chiasm (30Gy in 10 fractions), managing this further radiotherapy without problems.

At one-day post radiation therapy, BCVA was 6/60 (single optotype) in the right eye and 6/6 in the left. However, at this visit there was right optic disc pallor not evident previously (Figure 9) and the left disc was now oedematous and haemorrhagic (Figure 10). Intraocular pressures were 17 and 16 mmHg in the right and left eyes, respectively.

Several weeks later, the patient was re-commenced on her capecitabine chemotherapy. By that stage her blood film morphology was consistent with extensive skeletal marrow infiltration, including falling haemoglobin and decreased platelet count. Sadly, she became increasingly transfusion dependent and not long after, succumbed to her battle with cancer.

DISCUSSION

This represented a rare case of severe, unilateral vision loss with accompanying pupil defect that could not be fully explained upon clinical examination or neuro-imaging studies. Whilst no definitive diagnosis was available, there remain several plausible aetiologies that cannot be excluded with certainty.

Drug toxicity to chemotherapy agents such as capecitabine is possible, given the temporal relationship between the commencement of oral chemotherapy and symptom onset. However, the use of combination therapy and previous exposure to other chemotherapy agents make it difficult to draw conclusions. Ocular toxicity induced by chemotherapy is not uncommon but is frequently under-reported in the literature. With respect to capecitabine use specifically, there exist only a few reported cases of adverse ocular effects.<sup>13,14</sup> Although rare, conjunctivitis is the most common ocular side effect, affecting 5% of patients receiving capecitabine.<sup>13</sup> There have also been two reported cases of corneal toxicity, severe ocular irritation and decreased vision associated with capecitabine use.<sup>14</sup> Both patients presented with superficial punctate keratitis and multiple white granular sub-epithelial corneal deposits.<sup>14</sup> Treatment was discontinued in both instances with resolution of the corneal deposits and ocular symptoms, including restoration of normal vision. Common to both cases was a past ocular history of keratoconjunctivitis sicca.

Several differences exist between these reported cases and that of our own. In the case of our patient, vision

loss was asymmetric and the left visual acuity remained unaffected. Biomicroscopy was normal in our patient and thus, not suggestive of ocular surface toxicity. Furthermore, symptoms did not improve with cessation of capecitabine treatment.

Our patient had had previous exposure to other chemotherapy agents that have known ocular side effects. Prior to commencing capecitabine, she had undergone three cycles of IV nab-paclitaxel chemotherapy. Taxane-based chemotherapeutic agents, including paclitaxel and docetaxel, have been associated with adverse ocular side effects.<sup>15-19</sup> Of note, cystoid macular oedema (CMO) has been reported, although uncommon.<sup>15-19</sup> In most presenting cases, dilated fundus examination revealed bilateral CMO which was then confirmed on OCT imaging.<sup>15-19</sup> This association was further supported by the reversibility of the CMO after discontinuation of chemotherapy in all cases.<sup>15-19</sup> It is important to highlight that there was no evidence of CMO in our patient. Macular OCT repeated over consecutive visits was unremarkable in both eyes. Ocular involvement was also unilateral in our patient. Nab-paclitaxel chemotherapy was poorly tolerated and there was bony progression on re-staging. As such, it was discontinued after just three cycles and the patient was switched to oral capecitabine. Ocular symptoms developed later and occurred six months after commencing capecitabine. Our patient discontinued capecitabine chemotherapy upon oncology recommendation and was off capecitabine for approximately eight months in total, without resolution or reversal of ocular symptoms.

Initial observations draw the attention towards possible

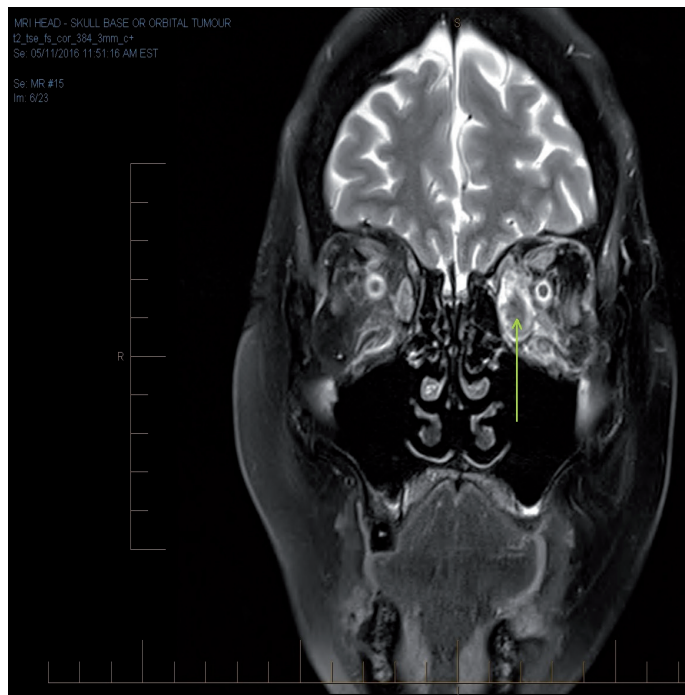


Figure 6. Coronal T2 fat suppressed MRI of the brain and orbits post steroid treatment showing persistent, severe swelling of the left medial rectus with enhancement.

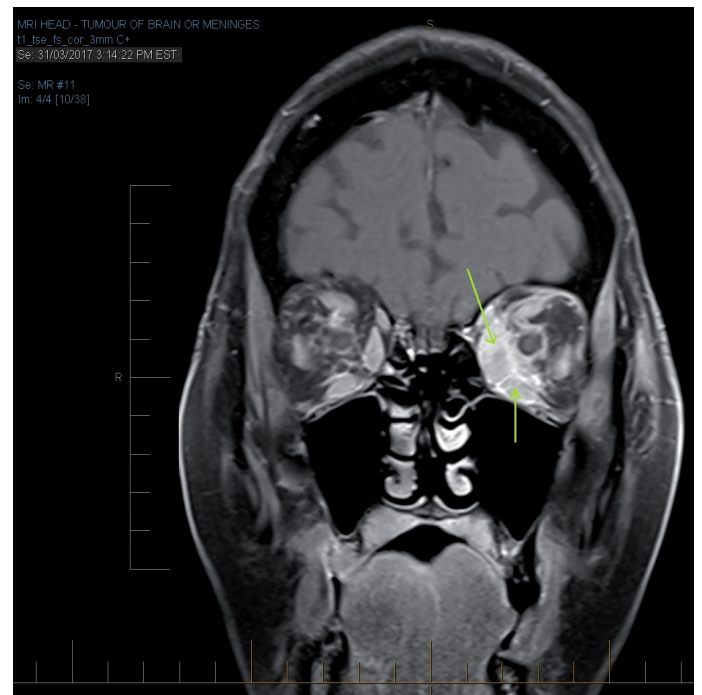
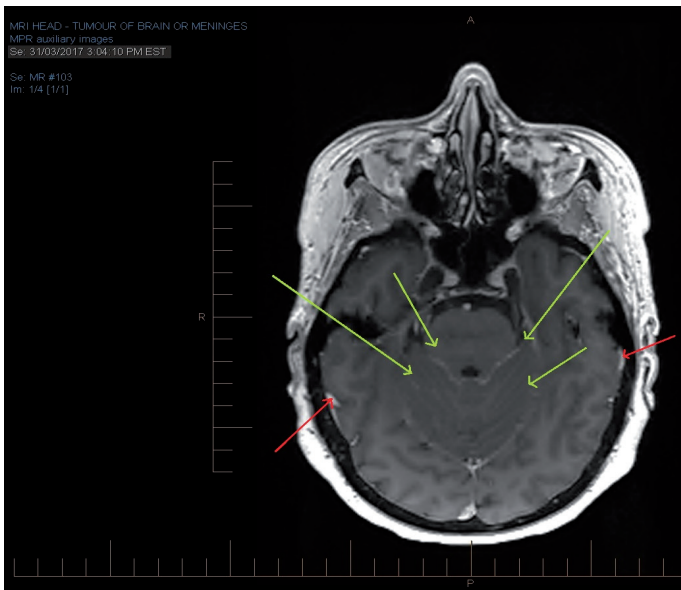


Figure 7. Coronal T1 fat suppressed MRI of the brain and orbits post radiotherapy showing persistent bilateral orbital inflammatory process (L>R). Marked swelling of the left medial and inferior recti is seen.





**Figure 8.** MRI brain (axial view) showing leptomeningeal tumour infiltration in the posterior fossa. Green arrows denote multiple linear hyperintensities representative of enhancing metastatic involvement of the cerebellar folia and brainstem. There is also nodular thickening of the pachymeninges consistent with metastatic infiltration (red arrows).

sub-clinical ophthalmic side effects associated with oral capecitabine therapy. However, this cannot be verified and to our knowledge, there are no reported cases in the literature of capecitabine cytotoxicity aside from those few cases discussed above with respect to ocular surface toxicity.<sup>13,14</sup>

In addition to drug toxicity, another diagnosis of consideration is infiltration of the right optic nerve sheath and/or partial compression of the right optic nerve. This is prudent to discuss given the presence of oedema within the intraconal fat and particularly in the setting of advanced metastatic disease. However, infiltrative disease or partial optic nerve compression were not readily apparent on imaging. The normal appearance of the right optic nerve head and lack of any associated findings such as exophthalmos, also failed to lend support to this diagnosis. There was late infiltration of the cavernous sinus and this could have contributed to compression of the optic chiasm or optic tract given its close anatomical proximity. However, given the partial decussation of nerve fibres at the chiasm, a chiasmal or post-chiasmal lesion would result in a visual field defect in the fellow eye which was not the case herein. The left visual field remained unaffected even late in the disease course. Moreover, there was no evidence of a posteriorly displaced chiasm which may have resulted in compression of the optic nerve before the chiasm secondary to a cavernous sinus lesion. The patient's initial symptom of pain on eye movement was thought to be due to the intraorbital oedema in the absence of any other significant findings.

One final causal possibility here is an autoimmune neuro-ophthalmic entity, paraneoplastic autoimmune optic



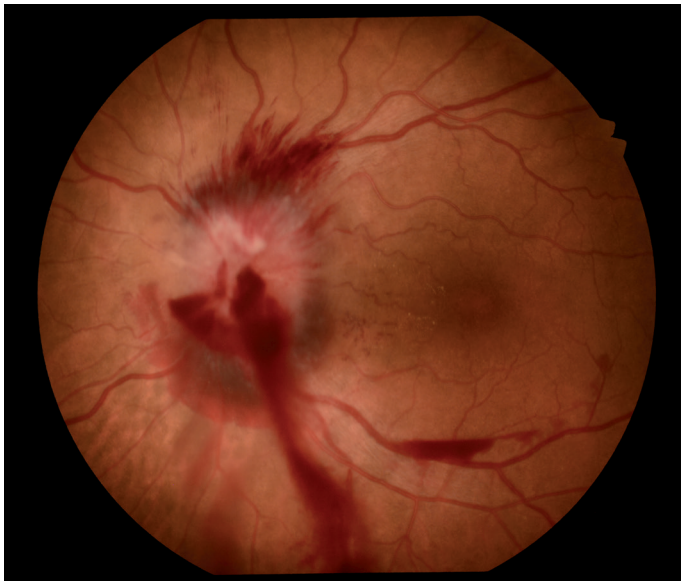
**Figure 9.** Fundus photograph of the right optic disc depicting disc pallor.

neuritis, which is defined by collapsin response mediated protein 5 (CRMP-5-IgG). There are several documented cases of this in the literature.<sup>20</sup> A case series of 16 patients who were sero-positive for the auto-antibody CRMP-5-IgG, and who had optic neuritis, reported that almost all cases presented with sub-acute, painless loss of vision over weeks to months, swollen and haemorrhagic optic discs, and anomalous visual fields.<sup>20</sup> However, unlike in our case, most of these patients had a history of small-cell lung carcinoma and vision loss was typically bilateral. To confirm this diagnosis, serology testing specific for CRMP-5-IgG is necessary, however this was not performed in our patient. As such, it remains unknown whether she was positive for this marker auto-antibody.

Our case of unexplained vision loss highlights the complexity of the aetiologies underlying vision loss in patients with cancer and that sometimes the cause remains unknown. Cancer patients should be advised to remain alert to any visual or ocular changes whilst undergoing treatment and to promptly report any such changes. Ophthalmic evaluation/referral is warranted in patients with new onset of symptoms. As a minimum, appropriate workup includes a detailed history, clinical examination and imaging studies. Biochemistry and electrophysiology testing may also help to improve diagnostic yield and inform treatment in instances where the aetiology is not overtly apparent.

## CONCLUSION

This represents a rare case of unexplained, unilateral vision loss. There remain several aetiologies that can be neither excluded nor concluded with certainty, including but not limited to: drug toxicity to chemotherapy agents; right-sided orbital infiltration or an undetected compressive lesion; and



**Figure 10.** Fundus photograph of the left optic disc depicting a grossly oedematous disc with haemorrhages.

paraneoplastic autoimmune optic neuritis. Whilst this case does not lend itself to a definitive diagnosis, it is important to document cases of unexplained vision loss or suspected ocular drug toxicity such that others who encounter patients with similar clinical presentation and medical history might collectively be able to help solve these diagnostic riddles.

## ACKNOWLEDGEMENTS

The authors would like to gratefully acknowledge the assistance of Drs David Blakey and Bob Dempster for their collaborative efforts as part of this work.

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## Teaching Orthoptics to Ophthalmology Residents: A Needs Assessment Study

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### ABSTRACT

**Aims:** This is a needs assessment study with the primary aim of examining the relevance of orthoptic tests to ophthalmology residents' practice, and their confidence in performing and interpreting the tests, to establish the need for further orthoptic education during residency.

**Method:** Participants were Years 1 to 5 ophthalmology residents from a tertiary hospital training program where no formal orthoptic training is offered. An online nine-question survey was conducted over four weeks to assess residents' perceptions of the relevance of orthoptic tests to their practice, their confidence in performing and interpreting ten common orthoptic tests, and preferences for curriculum content and delivery. Responses consisted of 5-point Likert scale options and selection of tests out of ten options. Data were analysed by descriptive statistics using median and range.

**Results:** Of 31 eligible residents, 23 (74%) responded. Relevance to practice was rated high among all respondents (median rating 4 out of 5, range 2 to 5) for all ten tests. Self-rated confidence in test performance was generally low (median rating 3, range 1 to 5). Confidence in test interpretation was higher (median rating 4, range 1 to 5). Respondents selected five tests for which they desired further training. Preference for e-learning was high, with 70% considering this modality 'very useful'.

**Conclusion:** Ophthalmology residents consider orthoptics to be relevant to their practice. Baseline self-reported confidence in test performance is low. They express a desire for further orthoptic training and e-learning is an acceptable teaching format.

**Keywords:** needs assessment, ophthalmology residents, orthoptic tests, e-learning, test confidence

### INTRODUCTION

Orthoptic competencies are not well-defined among the required competencies for ophthalmology residency graduation.<sup>1-4</sup> Orthoptics is a field that specialises in the evaluation of ocular misalignments and binocular functions. The ophthalmology residency program trains doctors who have completed their undergraduate medical school, in the field of ophthalmology. The Royal College of Ophthalmologists residency curriculum lists performing 'orthoptic assessment' as an important learning outcome without defining curricular requirements to achieve competency.<sup>1</sup> There is a need for ophthalmology residents to graduate with the competencies of both performing and interpreting orthoptic tests to enable best clinical treatment decisions.<sup>2,5</sup> In the orthoptists' training curriculum, orthoptic

topics are taught in multiple ways including didactics, hands-on skill practice with feedback and e-learning.<sup>5,6</sup> E-learning, defined as 'knowledge acquisition using electronic media and information technologies',<sup>7,8</sup> is already recommended for teaching in ophthalmology residency.<sup>9,10</sup> It has been demonstrated to be more effective and satisfying for learners than traditional didactic methods. This potentially translates into improved motivation and performance with better retention rates.<sup>7-11</sup> In addition, e-learning is asynchronous, can utilise multimedia including videos, and can be designed to be interactive using the Knowles model of self-directed learning.<sup>12</sup>

Little is known about how ophthalmology residents perceive their own need for orthoptic training. We therefore conducted a needs assessment among ophthalmology residents to determine its perceived relevance to their practice and to identify specific areas of learning need with the purpose of designing future curricula.<sup>13,14</sup>

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Accepted for publication: 20th August 2018

Our primary aim was to investigate the perceived relevance of common orthoptic tests to ophthalmology residents' practice, and their levels of confidence in performing and interpreting the tests. Our secondary aim was to explore their preferences for learning about different orthoptic tests and mode of curriculum delivery. We hypothesised that orthoptic topics would be perceived as relevant to ophthalmology residents, but confidence in test performance and result interpretation would be low.

## METHODS

### Setting and participants

Participants were Years 1 to 5 ophthalmology residents from a major tertiary hospital training program in Singapore, a Southeast Asian island nation with a population of 5.6 million. The 5-year residency program consists of three years of clinical training, followed by two senior years in general and subspecialty ophthalmology including oculoplastics, cornea, vitreo-retina, glaucoma, paediatric ophthalmology and neuro-ophthalmology.<sup>15</sup> The hospital assesses more than 300,000 patients per year in the Ophthalmology Department.<sup>16</sup> Thirty-one residents (15 males, 16 females) were available and eligible to participate in our study. Junior residents were defined as those in their first three years of residency while senior residents were those in the fourth and fifth years. Our institutional review board approved the study as exempt (CIRB reference number: 2017/2063).

### Study design

This is a descriptive cross-sectional online survey study. As there was no existing survey that addressed our research aim, we designed a nine-question survey using consensus agreement among a team of educators consisting of two expert orthoptists and a residency program director. We piloted the survey on two senior ophthalmologists who provided feedback that allowed further survey refinement. The survey asked questions in three domains relevant to orthoptic education and used a 5-point Likert scale from 1 to 5 with higher numbers indicating greater importance, relevance, confidence or usefulness. The three domains were: Relevance of orthoptic tests to practice and level of confidence in orthoptic test performance and result interpretation (four questions); Selection of orthoptic tests for future training (one question); Delivery mode and content (four questions). Questions 1 to 3 referred to ten groups of commonly performed orthoptic tests. They were: stereoacuity tests, cover/prism cover test, Hirschberg/Krimsky test, ocular movements, convergence/prism fusion range, accommodative amplitude/facility, visual acuity tests for children, Worth Four Dot test/Bagolini striated glasses test, double Maddox Rod, and Hess screen test/field of binocular single vision (BSV) test (Table 1). The survey also captured demographic information including year of residency.

Data was collected using an anonymised link administered by Google Forms®. The email link was sent out to all residents by a program administrator twice over four weeks; the second time as a reminder to encourage participation by non-respondents. The email invited residents to complete the survey to help educators understand their need for learning orthoptics, and their openness to learning by electronic modules.

Table 1. Needs assessment survey questions

Survey Domains	Number	Questions/format
1a. Relevance of orthoptic tests 1b. Level of confidence in orthoptic test performance and result interpretation	1	Please rate the following tests according to their relevance to your current or future practice 1 = least relevant, 2 = slightly relevant, 3 = relevant, 4 = fairly relevant, 5 = most relevant (5-point Likert scale)
	2	Please rate the following tests according to your level of confidence in performing them 1 = least confident, 2 = slightly confident, 3 = confident, 4 = fairly confident, 5 = most confident (5-point Likert scale)
	3	Please rate the following tests according to your level of confidence in interpreting their results 1 = least confident, 2 = slightly confident, 3 = confident, 4 = fairly confident, 5 = most confident (5-point Likert scale)
	4	How important do you think orthoptics is to your work? 1 = least important, 2 = slightly important, 3 = important, 4 = fairly important, 5 = most important (5-point Likert scale)
2. Selection of orthoptic tests	5	Which 5 tests would you most like to see featured in the teaching video(s)? (Checkboxes)
3. Delivery mode/content	6	How useful do you think the teaching videos will be for learning orthoptics? 1 = least useful, 2 = slightly useful, 3 = useful, 4 = fairly useful, 5 = most useful (5-point Likert scale)
	7	Select the orthoptic content you wish to see covered in video format (Checkboxes)
	8	What is your preferred mode of learning for orthoptic curricula and who do you think should teach it? (Open question)
	9	What is your opinion about learning via videos vs conventional lectures for orthoptics? (Open question)

Domain 1 (Questions 1 to 4) addressed the relevance and importance of orthoptics, as well as the residents' confidence in performing and interpreting tests. Domain 2 (Question 5) asked residents to select five from among ten commonly performed orthoptic tests that they would like to learn more about. Domain 3 (Questions 6 to 9) addressed the usefulness of teaching videos and invited respondents to select their preferred orthoptic content and mode of delivery. Respondents were asked to choose any number from a list of seven choices. The choices were 'Features of the orthoptic tests', 'Test techniques', 'Case studies', 'Real patient assessment', 'Patient management', 'Recording techniques', and 'Interpretation of test results'. Questions 8 and 9 were open-ended and residents typed in their responses to each question with no word limit imposed.

### Data analysis

Data captured on the 5-point Likert scale was exported into Excel format and data analysis was performed using descriptive statistics, reporting median and range. Subgroup analysis comparing junior versus senior respondents was performed via the Mann-Whitney U test using SPSS version 24.0 (IBM Corp, 2016).

The rating '1' was described on the survey as 'least confident' and a rating of '5' defined as 'most confident'. We used '3' ('confident') as the mid-point to define values above '3' as 'high' and values of '3' and below as 'low'.

Narrative responses for the last two open-ended questions were collated and independently read and interpreted by the two researchers, who then met and discussed emerging themes. The themes derived from these two questions were combined, summarised and described.

Our primary outcome was the responses to Domain 1 (Questions 1 to 4). Our secondary outcomes were the responses to Domains 2 and 3 (Questions 5 to 9).

## RESULTS

### Participants

Of 31 eligible residents, 23 (74%) responded to the online survey. The response rate was 48% (15 of 31 residents) for the first email invitation and increased to 74% at the second mailing. Seven (30%) were Year 3 resident, six (26%) from Year 1, four (17%) from Year 4, and three (13%) each from Years 2 and 5.

### Domain responses

#### Primary outcome (Domain 1):

For perceptions of the relevance of orthoptic tests to respondents' practice (Question 1), all ten listed orthoptic tests were highly relevant to practice (median rating 4 out of 5, range 2 to 5) (Figure 1).

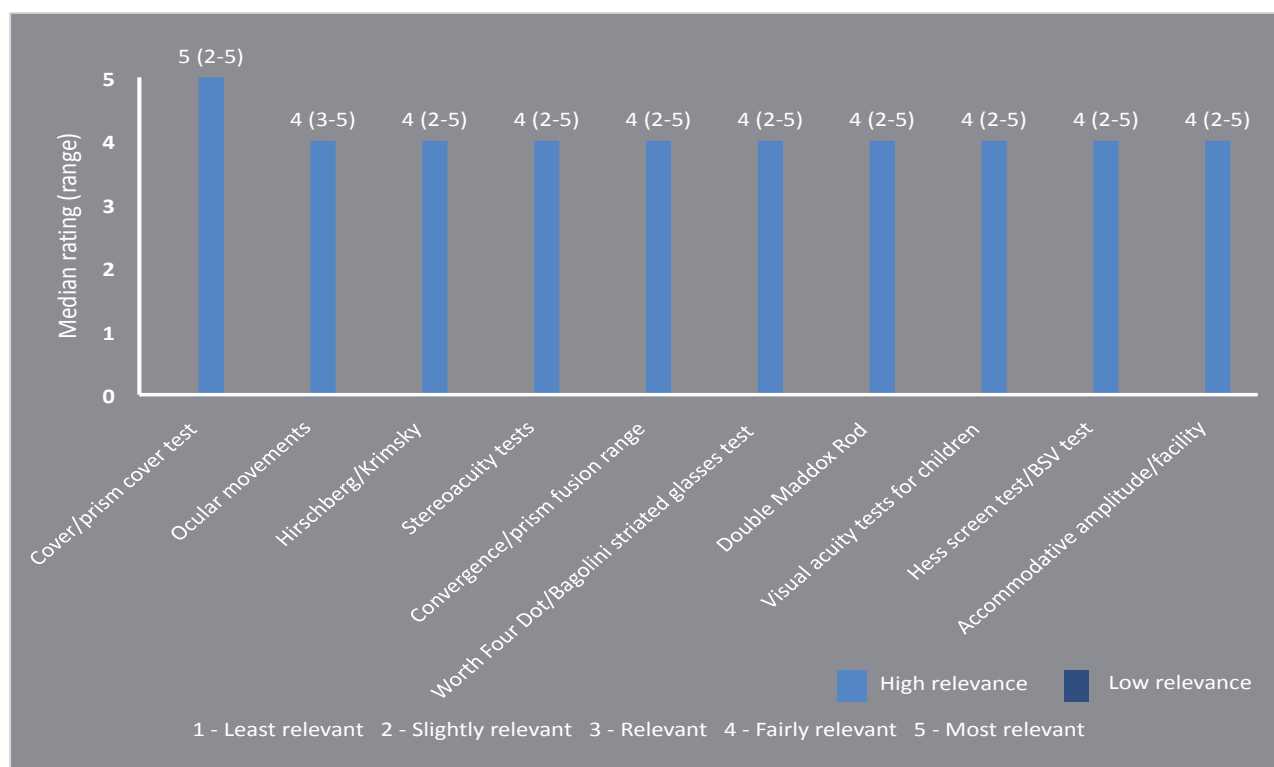


Figure 1. Ophthalmology residents' responses to 'Rate the relevance of orthoptic tests to your practice' (n=23) Singapore, 2017.

For orthoptic test performance (Question 2), respondents generally rated low confidence (median rating 3, range 1 to 5) across six out of ten tests (Figure 2). They expressed high confidence in performing four of the ten tests: stereoacuity tests, Hirschberg/Krimsky test, cover/prism cover test and ocular movements.

For test result interpretation (Question 3), respondents rated low confidence for only one test (median rating 4, range 1 to 5) (Figure 3). The test was accommodative amplitude/facility, which respondents also expressed low confidence in performing. They rated high confidence in interpretation for the other test results, while reporting low confidence in performing them.

Nineteen out of 23 respondents (83%) felt that orthoptics was 'very important' to their work while the remaining four responded that it was 'important' (Question 4).

Subgroup analysis was conducted between senior (n=7) and junior (n=16) respondents and a statistically significant difference in the perceptions of test relevance between them was found (p=0.005). The juniors perceived the orthoptic tests as more relevant than the seniors. No statistically significant differences were found for their levels of confidence in either test performance or interpretation (p=0.912, p=0.063, respectively).

**Secondary outcomes (Domains 2 and 3):**

For Question 5, the top five groups of orthoptic tests selected by residents for further training were: convergence/prism fusion range (74% of respondents), Worth Four Dot test /Bagolini striated glasses test (74%), double Maddox Rod (74%), accommodative amplitude/facility (74%) and Hess screen test/BSV test (57%).

For Question 6, all respondents reported that e-learning would either be 'very useful' (70%) or 'useful' (30%) for learning orthoptics.

For Question 7, respondents selected 'Interpretation of test results' (78%), 'Test techniques' (78%), and 'Case studies' (70%) as the top three orthoptic procedures that they would like to learn more about.

All 23 respondents provided narratives for the open-ended Questions 8 and 9. The length of each narrative ranged from 6 to 40 words. The researchers agreed on four common themes after independent coding. The most common theme that emerged was 'Orthoptics should be taught by hands-on clinical practice with patients, with observation and feedback'. The next was 'Orthoptics should be taught by orthoptic faculty and clinicians', followed by 'Videos on orthoptic tests allow me to learn at my own pace' and 'Blended learning with a combination of video demonstration and didactic lectures is the best strategy'.

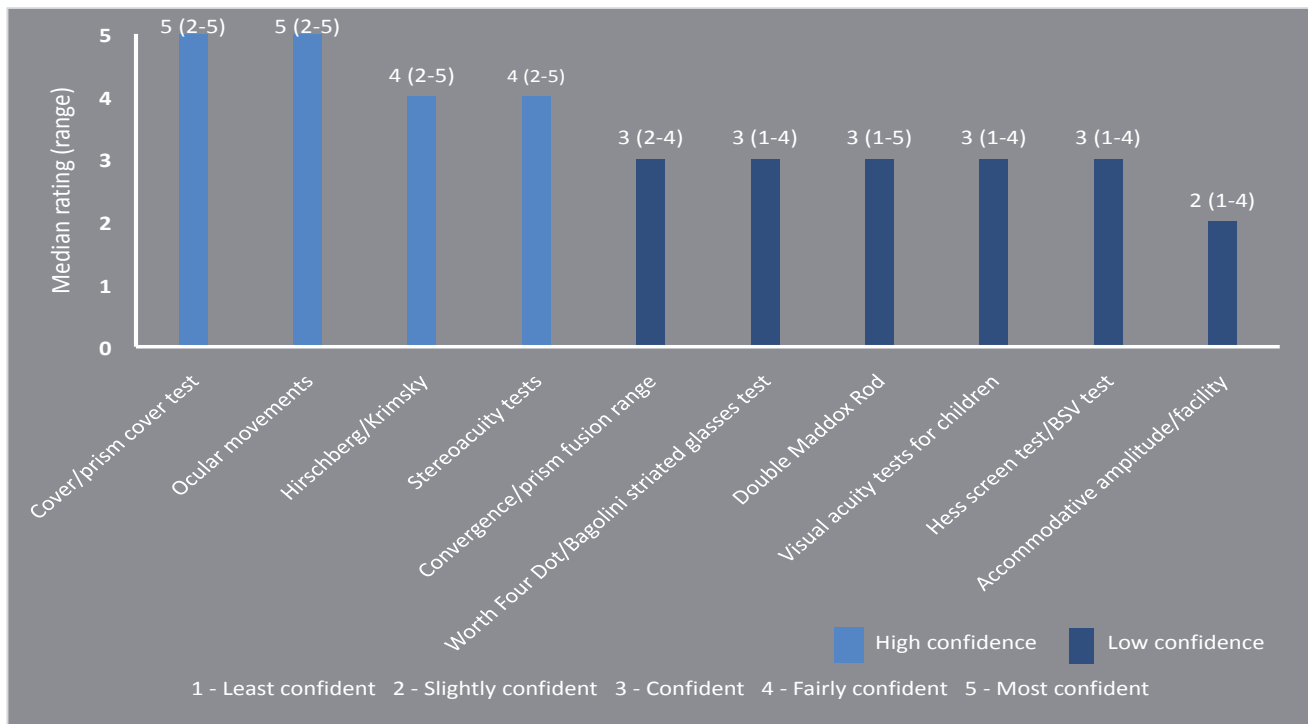


Figure 2. Ophthalmology residents' responses to 'Rate your level of confidence in performing orthoptic tests' (n=23) Singapore, 2017.



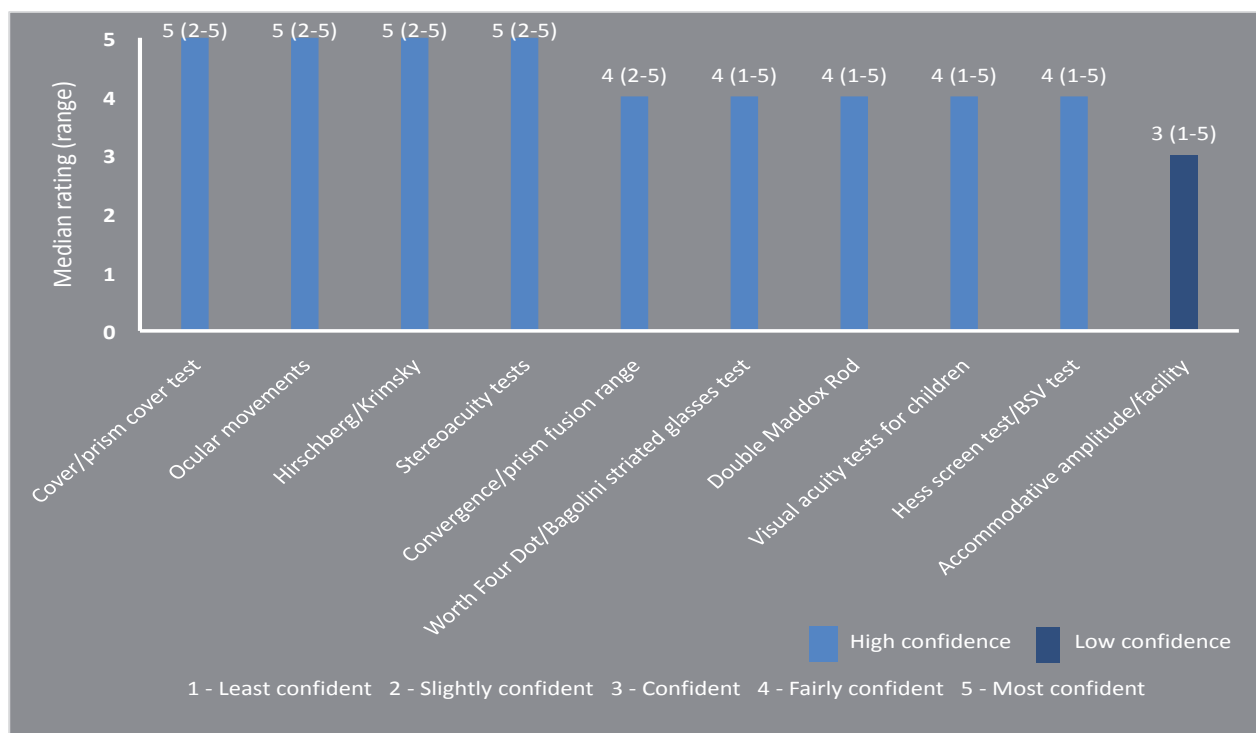


Figure 3. Ophthalmology residents' responses to 'Rate your level of confidence in interpreting orthoptic test results' (n=23) Singapore, 2017

## DISCUSSION

Needs assessment is an important aspect of curriculum development to generate baseline data to best meet learners' needs.<sup>13,14</sup> We conducted a learner needs assessment using an online survey, on ophthalmology residents from one program to determine their interest and need for orthoptic training. We hypothesised that all residents would perceive orthoptics as relevant to practice and we met this hypothesis. We found a mismatch between the residents' confidence in performing and in interpreting orthoptic tests. This could be attributed to their reliance on reading as a basis for learning about the tests, as opposed to performing the tests on patients themselves. This mismatch could be addressed by an interactive and visually-guided curriculum exposing them to specific test performance requirements, as well as by observed practice with feedback. Furthermore, it is not uncommon for learners to overestimate their own knowledge and abilities which can question the accuracy of self-assessments.<sup>17,18</sup>

Orthoptics as a subject has not been well addressed in residency curricula, despite its relevance to the practice of ophthalmology,<sup>2-4,15</sup> yet no published study has examined residents' attitudes toward learning orthoptics. Our study is unique in finding strong interest among all levels

of ophthalmology residents for orthoptic curriculum. Junior residents especially considered orthoptic tests to be highly relevant to their practice. This could result in greater acceptance of the curriculum designed for them.

We found that the top five of ten tests residents selected for greater exposure were those for which they expressed the lowest confidence in performing. Some were tests that they were less likely to be exposed to in their routine patient care such as the double Maddox Rod, tests of accommodation and Hess screen/BSV tests. This is not surprising, and the information allows planning and prioritising when designing future orthoptic curricula.

Using e-learning with videos as a teaching strategy to deliver curriculum has not been widely explored in the field of orthoptics. We are not surprised that the residents showed a preference for this modality because e-learning facilitates a learner-centred approach and interactive learning at their own pace, allowing them to monitor their progress via immediate feedback.<sup>19</sup> 'Case studies' was one of the top three content areas selected for presentation in an orthoptic curriculum. This reflects and supports residents' need to apply theoretical knowledge to practical application.<sup>20</sup>

The strengths of our study are the high response rate and representation by all levels of residents producing consistent findings. One study limitation is the small sample size

which did not have the power to compare responses across subgroups. This limitation is common to most ophthalmology residencies which tend to be small. Future studies will involve multiple programs to capture a larger sample size. A second limitation is that some residents may have had prior exposure to orthoptics through working with orthoptists and the variability in baseline knowledge may have confounded responses. However, the lack of significant differences found between junior and senior residents in terms of their confidence levels in performing and interpreting orthoptic tests suggests that any prior exposure had limited impact on their responses. It could be argued that not using a neutral grade might have contributed towards a more positive response. On the other hand, having a neutral grade might attract respondents who are undecided and it is harder for the ambivalent responses to be interpreted meaningfully. Finally, the survey reflects self-reported data; future studies should also address faculty-reported data on residents' knowledge and performance skills.

In conclusion, ophthalmology residents at all levels identify a need to learn more about orthoptics, especially around test performance, and are open to an e-learning curriculum presenting common orthoptic tests using a video format.

## ACKNOWLEDGEMENTS

No financial support was received for this study. We thank the ophthalmology residents for their participation. We are grateful to Professors Audrey Chia and Ian Yeo, and Dr Hla Myint Htoon for their technical and moral support.

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## Excluding Non-English Speaking People from Health Research Including Falls Research for Community-Dwelling Older People

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### ABSTRACT

The exclusion of people with limited or no English language skill from health research occurs often, due to logistical and financial constraints. Exclusion limits the generalisation of study outcomes especially in culturally and linguistically diverse populations. This includes falls research for community-dwelling older people. Reduced vision has been reported in the literature to be a significant independent risk factor for falls in this population. Excluding non-English speaking people from health research also impacts on eye research. The aims of this review are to increase the awareness of the issues arising from the exclusion of non-English speaking older people from health research and to encourage researchers to include this

vulnerable population in health research. English language skill is a valid indicator of health status and older people with limited English language skill have significantly poorer self-reported health status than those who speak English only. Despite this, guidelines governing inclusion of this population in health research are inconsistent. Resources and advocacy of inclusion will ensure ongoing equity of access to health care services for this population.

**Keywords:** non-English speaking, health research, falls, community-dwelling older people

### INTRODUCTION

People with limited or no English language skill are often excluded from health research.<sup>1-2</sup> This includes research into falls for community-dwelling older people.<sup>3-8</sup> Falls are a major health concern and the main reason for trauma-related hospital admission in people aged 65 years and older.<sup>9-10</sup> Reduced vision has been reported in the literature to be a significant independent risk factor for falls in this population.<sup>11</sup> When this minority group are included in health research and falls research for example, but the research variable of English language skill is not reported,<sup>12-17</sup> their outcomes remain unknown. Excluding non-English speaking people from health research is an area of interest and clinical relevance as it also impacts on eye research. The exclusion of this population from health research limits the generalisation of study outcomes especially in culturally and linguistically diverse populations. The aims of this narrative review are firstly to increase the awareness of the issues arising from the exclusion of non-English speaking older people from health research, both qualitative and quantitative,

including falls research in community-dwelling older people. Secondly, this review aims to target researchers and encourage inclusion of non-English speaking people in health research.

### DISCUSSION

The Australian guidelines by which human research is regulated, advocate equity of access and recognise the cultural diversity of Australia's population.<sup>18</sup> These guidelines include the values and principles of ethical conduct, and the ethical consideration of risk, benefit, consent, research methods, recruitment of participants and accountability.<sup>18</sup> The Department of Immigration and Border Protection<sup>19</sup> reported on this diversity, based on the statistics from the 2011 Australian Census. Twenty-seven percent of Australians aged 65 years and over, who spoke a language other than English, reported that they either did not speak English well or did not speak English at all. This particular statistic was not included in the 2016 Australian census. In other countries like the United States for example, the United States National Institutes of Health (NIH)<sup>20</sup> also have guidelines in place to ensure that minority groups such as those with limited English proficiency are adequately

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Accepted for publication: 6th September 2018

represented in clinical research. These guidelines suggest that minority groups must be included in all NIH-funded clinical research, unless inclusion is inappropriate due to the health of the participant or the purpose of the research.<sup>20</sup> Despite these guidelines, exclusion of people with limited or no English language skills has not declined, but in some cases has increased.<sup>21</sup>

### **Exclusion of non-English speaking people from health research**

The reasons for exclusion of people with limited or no English language skill in health research include logistical and financial constraints.<sup>1-2</sup> Glickman et al<sup>1</sup> reported that clinical researchers are not always encouraged by their governing body to include people with limited English proficiency. The authors reported on the differences in American Institutional Review Board (IRB) policies, regarding the consent process of people with limited English proficiency in clinical research projects. The sample was limited to academic IRBs and restricted to policies posted on the Internet. The consent requirements for all clinical research of the IRB of 134 American academic health centres were reviewed and the authors found eight statements that could discourage investigators from including people with limited English proficiency. For example, one IRB cautioned the researchers to carefully consider the ethical and legal implications (obtaining informed consent) of recruiting subjects with limited English proficiency, whilst another IRB warned investigators to carefully consider whether, even with a trained interpreter, a legal informed consent can be obtained. Equally, there were eight statements that encouraged investigators to include people with limited English proficiency, for example, that care must be taken to not exclude non-English speakers and another statement reminded investigators that non-English speakers must not be excluded unless there is an ethical or scientific reason agreed on by the IRB. It is encouraging that inclusion is supported but there needs to be a consistency which is not always seen. A consistent approach will ensure equity of access to health research for this minority group.

Frayne et al<sup>2</sup> also reported on the exclusion of non-English speaking people from health research. Unlike in Glickman et al<sup>1</sup> the investigators in this review were not discouraged from including non-English speaking people. The main reason for exclusion was oversight. To determine how often non-English speaking people are excluded from medical research, Frayne et al<sup>2</sup> surveyed the authors of publications on provider-patient relations about their exclusion criteria. In this study, 'provider-patient relations' refers to activities such as patient education, health education, or patient satisfaction. Of the 172 authors surveyed, sixty-eight (40%) excluded non-English speaking people. The most common reason for exclusion was not having considered it

(51%). The authors who did consider inclusion but decided to exclude non-English speaking people did so for a variety of reasons; 58% due to the absence of study instruments in languages other than English, 55% due to having to translate responses into English, 45% due to the expense of translating the study material and 45% due to problems recruiting bilingual staff. These percentages show that a large number of researchers understand the importance of including people who do not speak English. What is needed are resources targeted at these specific barriers to support researchers to include people who do not speak English.

Data quality concerns may also lead to the exclusion of non-English speaking people from health research. Ngo-Metzger et al<sup>22</sup> reported on data quality in health research by reviewing response rate and missing data. The authors compared response rates and missing data of telephone and mail surveys among Asian Americans with limited English proficiency. The authors concluded that their data quality was comparable to studies which had been conducted with English speaking subjects. This study is also an example of successfully including people who do not speak English in research using resources such as bilingual staff. In this particular study, 479 patients (mean age 44 years) were surveyed about quality of medical care. Eighty-three percent of the subjects did not speak English or did not speak English well. The survey contained 78 items and was delivered via two modes: a self-administered mail survey with telephone reminder and a telephone survey. Both survey modes were in the participant's native language, Cantonese, Mandarin or Vietnamese. An overall response rate of 67% (322 of 479) was achieved. There was a higher response rate to the telephone survey (75% of 240) compared with the mail survey (59% of 239) and the missing data was minimal with respondents completing over 90% of the survey questions. The conclusion made by Ngo-Metzger et al,<sup>22</sup> that their data quality is comparable with studies conducted with English speaking subjects is consistent with the findings of authors Sullivan et al<sup>23</sup> and Kerr et al,<sup>24</sup> who surveyed English speaking people about their health outcomes. Sullivan et al<sup>23</sup> surveyed 983 English speaking people with diabetes (mean age 60 years) on their health outcomes and reported an overall response rate of 70.9% (697 of 983) and a completion rate of over 84%. This particular survey was delivered via three modes; mail, handed out, and face to face interview. The overall response rates for Sullivan et al<sup>23</sup> and Ngo-Metzger et al<sup>22</sup> were similar as were the completion rates. More recently, Kerr et al<sup>24</sup> conducted a single mode, mail survey with 5,110 English speaking patients (mean age 72 years) on their satisfaction with hypotensive eye drops. The response rate was 50% (2,541 of 5,110) and the completion rate was over 99%. Once again, the completion rate reported by Ngo-Metzger et al<sup>22</sup> is similar to that achieved by Kerr et al,<sup>24</sup> and the response rate of 59% achieved by Ngo-Metzger et al<sup>22</sup> from the group



who received the survey via mail, compares with the response rate that Kerr et al<sup>24</sup> received for their survey which was also delivered via mail (50%). As response rates and missing data have been shown to be similar despite English language skill, the evidence does not support concerns about the data quality of subjects with limited English-language proficiency in health research.

### **Exclusion of non-English speaking older people from falls research**

In Australian research studies concerning falls for community-dwelling older people, it is common for non-English speaking people to be excluded.<sup>6-8</sup> Falls are a major health issue for older people aged 65 years and over and are caused by the complex interaction between multiple risk factors.<sup>25-26</sup> Examples of such risk factors for falls include poor vision, impaired cognition, impaired balance, previous falls, the use of more than four medications and use of psychoactive medications, that is medications used for treating depression or anxiety.<sup>26</sup> A fall can be defined as 'an unexpected event in which the participant comes to rest on the ground, floor or lower level'.<sup>12</sup> Around 43% of older people living in the community (ie living at home or independently in a retirement village) have one or more falls each year (43.5%,<sup>12</sup> 43.6%,<sup>27</sup> 43.2%<sup>6</sup>), with the fall rate increasing with age,<sup>1,17,28</sup> along with the rate of fall-related injuries requiring emergency medical attention and hospital admission.<sup>29-30</sup>

The common barriers to inclusion of non-English speaking older people from falls research are once again operational and financial, for example, a lack of validated assessments available (neuropsychological, anxiety and depression) in languages other than English, questionnaires and falls calendars which are only available in English, physical assessments which require comprehension of oral instructions to ensure reliability and an absence of funding for language interpreters (A Tiedemann, February 8, 2012, personal communication; S Lord, February 9, 2012, personal communication). The barriers to inclusion of non-English speaking older people experienced by researchers over a decade ago<sup>2</sup> are the same today (A Tiedemann, February 8, 2012, personal communication; S Lord, February 9, 2012, personal communication). This is surprising considering the focus on positive health outcomes for our diverse community.<sup>31-33</sup>

### **English language skill, self-reported poor health and falls**

English language skill is a useful variable for health research and a valid indicator of health status.<sup>34-35</sup> Older people with limited English language skill have significantly poorer self-reported health status than those who speak English only.<sup>35-36</sup> As self-reported poor health is a falls risk factor for community-dwelling older

people,<sup>37</sup> then faller status for non-English speaking older people may differ from English speaking older people. As non-English speaking people are often excluded from falls research, then this association remains unclear.

In two large American studies on linguistic disparities in health access and health status in older people, data was analysed from the 2001<sup>35</sup> and the 2007<sup>36</sup> California Health Interview Survey. Both studies were conducted in similar languages including English, Spanish, Cantonese, Mandarin, Korean and Vietnamese. The study by Ponce et al<sup>35</sup> also included Khmer. The sample size in the study by Ponce et al<sup>35</sup> was less than half that of Sentell and Braun<sup>36</sup> (n = 18,659 and n = 48,427, respectively) and also older (Ponce et al,<sup>35</sup> 53% aged 65 years and over; Sentell & Braun,<sup>36</sup> 11.8% aged 65 years and over). Despite this, each study had a similar percentage of respondents with limited English proficiency (Ponce et al,<sup>35</sup> 7%; Sentell and Braun,<sup>36</sup> 7.7%). Ponce et al<sup>35</sup> reported that the respondents with limited English language proficiency had 68% increased risk of poorer self-reported health (fair to poor health status) compared with the English only speakers (RR 1.68, 95% CI: 1.37 - 2.02, P < .001). Sentell and Braun<sup>36</sup> found that the adults with limited English proficiency were significantly more likely to self-report poor health status compared with those adults who were English proficient (42.9% versus 14.9%; OR 2.10, 95% CI: 1.7 - 2.58). These two large sample studies have highlighted the disparities in health status between older people with limited English language proficiency and older people who are English proficient. Both studies have shown that older people with limited English proficiency are more likely to self-report poor health than those who are proficient in English, even when questioned in their native language. Limited English proficiency is a major barrier to health care. The provision of health service interpreters is an example of how health care systems could reduce the linguistic barrier and improve access to health care, thus improving the health status of this vulnerable population.<sup>35-36</sup>

Gill et al<sup>37</sup> determined that self-reported poor health is a falls risk factor for community-dwelling older people. Their population-based survey, conducted in Australia, investigated a range of potential factors for falls defined in community dwelling older people. A definition of a fall was not included. Although language spoken at home was included as a demographic variable, English-language skill was not. One of the factors included in the survey was general health, self-reported as either poor, fair, good, very good or excellent. Two-thousand-six-hundred-and-nineteen older people responded to the survey (females n = 1,481) which was conducted via telephone. The respondents who self-reported poor or fair general health compared to good, very good or excellent general health were significantly more likely to have fallen in the previous 12 months (OR 1.34, 95% CI: 1.09-1.67, P < 0.001).<sup>37</sup>

The retrospective recall of falls data may have led to an under-reporting of falls, limiting the accuracy of the data analysis.

Also, it could not be established if the fall contributed to the self-reported poor health rating, that is whether it came before the poor health, or if self-reported poor health was a risk factor for the fall, or if both patterns occurred.

## CONCLUSION

People with limited or no English language skill are often excluded from health research including falls research due to logistical and financial constraints. There are guidelines in place governing human research which generally support the inclusion of participants with limited or no English language skill and concerns such as limited data quality are not substantiated in the literature. Exclusion of this group from health research may have a negative impact on the general health outcomes of this culturally and linguistically diverse population as they are also more likely to self-report poor health compared to older people who are English proficient. This includes community-dwelling older people from this group who may not have access to evidence-based health care services including falls prevention. As poor vision is associated with an increased risk in falls for older-people then exclusion also limits the generalisation of important clinical findings especially in culturally and linguistically diverse populations. Awareness of the issues arising from the exclusion of non-English speaking older people from health research including eye research, need to be considered to ensure ongoing equity of access to health care services for this population.

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# Functional Impact of Perifoveal Geographic Atrophy in Patients with Dry AMD: A Systematic Review

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## ABSTRACT

Conventional parameters such as best-corrected visual acuity (BCVA) often grossly underestimate the profound visual limitations experienced by patients with perifoveal geographic atrophy (GA) secondary to dry age-related macular degeneration (AMD). Foveal preservation in these patients means that BCVA is often only moderately impaired, despite significant challenges often reported in undertaking day-to-day vision-requiring tasks. BCVA may lead to a misrepresentation of the extent of real-world visual dysfunction in this clinical population and yet, is widely used as the gold standard measure in assessing patient eligibility for disability entitlements and driving. This systematic review investigated the relationship between microperimetry thresholds and performance on tests of functional vision and visual function in patients with perifoveal GA.

A systematic search of the Embase, CINAHL, Medline, PubMed and Web of Science electronic databases was conducted to identify all relevant studies published between

January 2002 and December 2017 in the English language and involving human participants. A search of the grey literature was also conducted. Ten relevant articles were found and a critical appraisal undertaken.

Only two of the 10 studies investigated functional deficits specifically in patients with perifoveal GA. The remaining eight studies were more broadly defined and failed to subclassify participants according to GA location. Microperimetry was found to represent a valuable tool in quantifying visual deficits in these patients and was much more sensitive than conventional measures, including BCVA and low-luminance visual acuity. This review highlighted the importance of a multimodal approach to assessment to better capture the real-world visual dysfunction experienced by patients with perifoveal GA.

**Keywords:** geographic atrophy, perifoveal, visual function, functional vision, microperimetry

## INTRODUCTION

Age-related macular degeneration (AMD) is the leading cause of severe, irreversible visual impairment and blindness among individuals over the age of 65 in developed countries.<sup>1</sup> Of its two principal forms – wet (neovascular; exudative) AMD and dry (atrophic; non-exudative) AMD – the latter is more common and constitutes 90% of all diagnosed cases.<sup>2</sup> Dry AMD is characterised by a gradual and progressive atrophy of the photoreceptors, retinal pigment epithelium (RPE) and choriocapillaris.<sup>1</sup> In contrast, neovascular AMD

is characterised by choroidal neovascularisation and involves leakage of blood including serous fluid and other constituents, into the sub-retinal and/or intra-retinal space.<sup>2</sup>

Vision loss owing to neovascular AMD can be delayed by anti-vascular endothelial growth factor (VEGF) therapy. However, no treatment exists for dry AMD. Current management is concentrated on delaying progression to advanced disease by controlling for known, modifiable risk factors such as dietary and smoking habits, body mass index, blood pressure regulation and glycaemic control.<sup>3-7</sup> Supplementation of dietary zinc, vitamins C and E<sup>8,9</sup> is recommended together with lutein and zeaxanthin for those with reduced dietary green and coloured vegetable consumption.<sup>9</sup>

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Accepted for publication: 8th October 2018



Hallmark findings in the early stages of dry AMD include whitish-yellow drusenoid deposits which can manifest with or without pigmentary abnormalities of the RPE (either hyper- or hypo-pigmentation). Drusen are accumulations of extracellular, amorphous debris subjacent to the basement membrane of the RPE.<sup>10,11</sup> Whilst these clinical findings alone do not typically cause overt central vision loss in patients, substantial functional decline can ensue especially with advancing disease.

Small, multifocal atrophic areas form and initially are only visible on ocular coherence tomography.<sup>12</sup> These gradually enlarge and coalesce to form clinically obvious geographic atrophy (GA)<sup>13,14</sup> that are visible as depigmented areas with prominent choroidal vessels and are hypofluorescent on autofluorescence imaging.<sup>15</sup> The areas of GA result in decreased threshold sensitivity or scotomata in the corresponding parts of the visual field.<sup>16</sup> Importantly, the percept is not a black 'hole' or 'gap' in the patient's visual field, but rather, the missing content is 'filled-in' based on information from the surrounding intact visual field in a 'filling-in phenomenon'.<sup>17,18</sup> The adapted area is inferred or extrapolated from visual information acquired in adjacent, intact parts of the visual field.<sup>19,20</sup> Impaired facial recognition, compromised reading ability, nyctalopia and fluctuating vision are often experienced by the patient.<sup>21,22</sup>

In a proportion of patients with dry AMD, the GA develops paracentrally and the fovea is spared, at least initially.<sup>13,14</sup> These atrophic regions sometimes develop in a 'U'-shaped horseshoe pattern and gradually coalesce to form a perifoveal ring of atrophy with the centremost fovea being preserved. The retinal atrophy produces an annular area of blindness in the patient's visual field surrounding the central fixation locus, referred to as a 'donut scotoma' or 'ring scotoma', but central acuity is preserved. Eventually, the fovea also becomes atrophic and the visual acuity may decline to 6/60 or worse.<sup>14</sup>

One of the greatest challenges to date lies in quantifying the profound visual limitations experienced by patients with perifoveal GA. To appreciate this, a distinction must first be made between two equally important yet fundamentally different aspects of vision loss – visual function and functional vision. Visual function describes how well the eye and visual system work.<sup>23</sup> It relates not just to the workings of the eye but the entire cortical visual pathway. Under visual function, tests such as visual acuity, perimetry, contrast sensitivity and colour vision are considered. Functional vision, as distinct from visual function, relates to how well a patient can perform vision-related activities, such as reading text, pouring liquids, and orientation and mobility.<sup>23</sup> This differentiation is important as the two concepts are not inextricably linked.<sup>24</sup> Good visual function does not necessarily translate into good functional vision. For patients with perifoveal GA secondary to dry AMD, a failure to appreciate this important distinction often leads

to a misunderstanding of the patient's condition by family, friends and practitioners alike.

Historically, the most commonly used measure of visual function in both clinical practice and ophthalmic research is best-corrected visual acuity (BCVA). BCVA is widely accepted as the gold standard measure of macular function. Clinical trials often assess BCVA to determine the efficacy of new therapeutic agents and treatments. Government and other regulatory authorities also recognise BCVA as the gold standard when assessing patient eligibility for concession entitlements and reimbursement programs.<sup>25-27</sup> However, in patients with perifoveal GA, BCVA is not an accurate representation of the true level of visual dysfunction as it fails to assess all nuances of human vision which is a complex phenomenon. Foveal preservation in these patients means that BCVA is often only moderately impaired, for example, in the order of 6/7.5 to 6/12. Despite this, these patients typically report profound difficulty with reading and recognising faces, over and above what would be predicted on the basis of their acuity alone. This is because a full word or face may not 'fit' in the small spared central area that is surrounded by the area of GA, with respect to the visual angle subtended and the retinal image size. In an attempt to acquire more visual information, the patient may scan their environment by moving their head or eyes. This strategy can aid visual processing but enhanced visual processing occurs at the expense of time. A BCVA of 6/12 or better does not give an indicator of the struggle one may have had in the process of achieving this level of acuity, inclusive of the time taken or increased scanning. The greatest difficulty is often experienced when the patient is required to localise kinetic objects or read a passage of text. Hence, when taken as a stand-alone measure, BCVA can lead to a gross overestimation of a patient's visual capabilities in the real world. To base clinical trial endpoints and eligibility criteria, for example 'legal blindness', exclusively on parameters of visual function reflects a misunderstanding of the important distinction between visual function and functional vision, and the pathogenesis of dry AMD.

In light of an aging population, the global prevalence of AMD is increasing. There is a critical need to define the functional impact of perifoveal GA in patients with dry AMD and to explore the use of other measures to gain a more accurate depiction of functional vision in this clinical population. One such test of increasing utilisation in ophthalmic practice is microperimetry.<sup>28</sup> Microperimetry is a psychophysical diagnostic technique correlating threshold sensitivity of individual points on the retina with ophthalmoscopic retinal appearance in real time.<sup>29</sup> Location of fixation sites at the fovea and macula enable accurate follow-up test-retest examination as stimuli are projected directly onto the retina and the same retinal point is monitored via eye-tracking.<sup>30-32</sup> During testing, stimuli are projected in random order to measure macular threshold sensitivity at pre-determined

retinal locations, typically over the central 10° of the retina.<sup>33</sup> Retinal locations are registered at baseline and the same locations are measured on repeat testing. Various microperimetry outputs are obtained, including average threshold sensitivity, fixation location and fixation stability. In the clinical setting, microperimetry is becoming increasingly popular as a means of detecting functional deficits arising from dry AMD.<sup>28</sup>

It is crucial to be able to accurately quantify the efficacy of new treatments and pharmacological agents, and to understand how these new-age therapies translate into functional real-world improvements, beyond patient performance in the examination chair. There is a need for ongoing and future clinical studies to use more sensitive and appropriate endpoints to monitor disease progression and evaluate potential treatment responses. Consequently, this systematic review aimed to investigate the relationship between microperimetry thresholds and performance on both functional vision and visual function testing in patients with GA secondary to dry AMD, specifically those with perifoveal atrophy. The significance and strength of any correlation may lend support for or against the clinical usefulness of microperimetry in assessing the degree of functional disability in this clinical population.

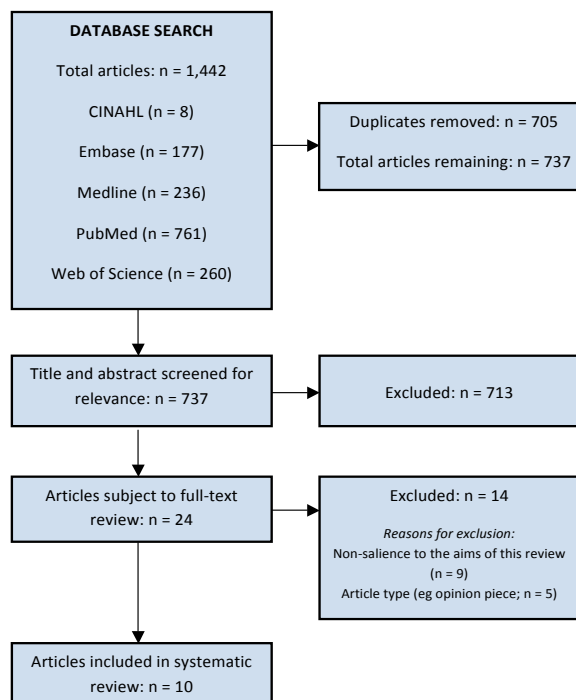
**METHOD**

A systematic search of the Embase, CINAHL, Medline, PubMed and Web of Science electronic databases was conducted to identify all relevant studies published between January 2002 and December 2017 that explored

the functional impact of perifoveal GA in patients with dry AMD using microperimetry. The database search was restricted by date as the first fully automatic microperimeter (Nidek MP-1; Nidek Technologies, Padua, Italy) was developed in 2002. Only full-text articles published in the English language and involving human participants were eligible for inclusion. No search restrictions were placed on study type, although editorial articles and opinion pieces were removed during screening by the reviewers.

All searches included a combination of key words: 'macular degeneration or age-related macular degeneration or age related macular degeneration or dry age related macular degeneration or atrophic age related macular degeneration or AMD or geographic atrophy or atrophy\* or atrophi\* or macular disease or degenerative macular disease or degenerative maculopath\* or foveal sparing or scotoma\* or ring scotoma\*' and 'microperimet\* or visual function or functional vision or visual disability or macular function or patient reported outcome\* or PROM\*' (where \* indicates truncation).

Titles and abstracts identified by the search were screened by the first author (JB) as well as a second, independent reviewer who otherwise had no involvement in this paper. Any articles that did not meet the eligibility criteria or that were deemed non-salient to the aims of this review were excluded. Duplicates were identified and removed. The full text of the article was retrieved for all relevant studies and read thoroughly, including when it was unclear whether the study met the inclusion criteria. The reference lists of included studies were reviewed to search for other relevant papers potentially missed in the database search.



**Figure 1.** Flow-chart of article selection process.

A search of the grey literature was also conducted, including unpublished ophthalmology conference proceedings and theses in the field.

All included papers were critically evaluated using the Critical Appraisal Skills Programme (CASP) Research Checklist for Cohort Studies<sup>34</sup> or the Joanna Briggs Institute (JBI) Critical Appraisal Checklist for Cross Sectional Studies<sup>35</sup> depending on study type. Each study was independently appraised by the two reviewers and the results cross-checked. In the event of a discrepancy, this was to be resolved by third party consultation with a senior investigator (WH).

## RESULTS

The initial database search revealed 1,442 abstracts (Figure 1). A total of 705 duplicates were identified and removed. All remaining articles were screened for relevance. Once duplicates and papers that did not address the aims of this review were removed, 24 articles remained for potential inclusion. Of these, a further 14 were excluded upon full-text review owing to article type (opinion piece;  $n = 5$ ) or non-pertinence to the topic ( $n = 9$ ). Upon searching the reference lists and grey literature, no additional works relevant to the topic were found. A total of 10 articles were included in this systematic review. Table 1 describes these 10 studies, detailing the methodology and research findings. A critical appraisal using the CASP<sup>34</sup> and JBI Checklists<sup>35</sup> revealed these papers to be of moderate quality. The most common factors affecting study quality were: lack of a control or failure to include the control group in the data analyses, small sample size, single centre recruitment, or lack of longitudinal follow-up. There was complete agreement between the two independent reviewers for study quality. Heterogeneity across the studies with respect to the methodological approaches, sample characteristics and outcomes precluded a meta-analysis from being performed

### Functional deficits measured using microperimetry in patients with perifoveal GA secondary to dry AMD

Only two of the 10 studies found in this review investigated functional deficits specifically in patients with perifoveal GA secondary to AMD.<sup>36,37</sup> The remaining eight studies were more broadly defined in terms of their sample and investigated patients with dry AMD, but did not further sub-classify participants according to the pattern of GA, that is foveal versus non-foveal involving.<sup>38-45</sup> As such, they did not conduct clinical subgroup analyses.

Of the two studies that specifically recruited participants with non-foveal GA secondary to AMD, both reported a significant reduction in mean retinal sensitivity outputs measured using the MAIA microperimeter in patients with AMD compared to healthy controls.<sup>36,37</sup> Moreover, they found that microperimetry was much more sensitive than visual

acuity measures, including BCVA and low-luminance visual acuity (LLVA), in detecting functional deficits in this clinical population. Longitudinal observation further revealed that microperimetry can detect subtle changes over a 12-month period even when no change is demonstrated on BCVA or LLVA testing.<sup>37</sup>

Wu, Ayton, Guymer and Luu<sup>36</sup> conducted a cross-sectional study of 179 eyes with a spectrum of dry AMD clinical severity (early, intermediate, and non-foveal GA sub-groups;  $n = 101, 65$  and  $13$  eyes, respectively) and 26 age-matched control eyes. They found that BCVA, LLVA and MAIA macular sensitivity were significantly reduced for all AMD groups when compared with controls, except for those with small drusen classified as between  $63-125 \mu\text{m}$  and no pigmentary anomalies. Low luminance deficit, calculated as the difference between LLVA and BCVA, was significantly different from controls only in the non-foveal GA group but not in any other AMD group. Macular sensitivity was found to be significantly correlated with LLVA, but not BCVA. A significant, strong positive correlation between macular sensitivity and low luminance deficit was reported, suggesting that LLVA may detect a greater extent of functional deficit than BCVA in eyes with increasingly poorer retinal sensitivity. Using linear regression models for macular sensitivity and BCVA, and macular sensitivity and LLVA, it was estimated by Wu et al<sup>36</sup> that when a reduction of two standard deviations (SDs) away from normal for BCVA and LLVA was measured, a much greater decline in macular sensitivity of 6.1 and 3.7 SDs, respectively, was apparent. Hence, their findings suggest that microperimetry is a much more sensitive measure than BCVA and LLVA in detecting functional deficits in dry AMD. However, a notable limitation of this study was that it lacked any longitudinal follow-up with the majority of participants having been assessed at a single visit only.

Wu, Ayton, Luu and Guymer<sup>37</sup> investigated longitudinal changes in microperimetry and LLVA over a 12-month period in patients with dry AMD. Forty-nine eyes of 49 patients with dry AMD ( $n = 8$  eyes with non-foveal GA, 41 eyes with intermediate AMD) and 10 eyes of 10 healthy controls underwent BCVA and LLVA testing, multimodal imaging, MAIA microperimetry and dilated fundus examination. Participants with AMD were assessed at three visits (baseline, 6 months and 12 months) and control participants seen at two visits (baseline and 12 months). Pathological progression was assessed in eyes with intermediate AMD by side-by-side comparison of coloured fundus photographs obtained at baseline and 12 months. Eyes were graded as 'stable', 'progressed/worsened', or 'improved'. No significant changes from baseline were detected in mean BCVA, mean LLVA or low luminance deficit in any group over the 12-month period. In eyes with non-foveal GA, a significant reduction in mean microperimetric point-wise sensitivity was detected at both 6 months and 12 months compared with baseline.



**Table 1. Summary of the methodological approaches and outcomes of studies included in the systematic review**

Study authors	Title of article	Study design	Primary aim/s	Methodology	Sample demographics	Key findings
Chandramohan et al (2016) <sup>38</sup>	Visual function measures in early and intermediate age-related macular degeneration	Prospective, controlled exploratory pilot study  Single centre (Duke Eye Centre, Durham, USA)	To evaluate the test-retest repeatability of computerised tests of LLVA, cone-specific contrast (CSCT), contrast sensitivity, and MAIA microperimetry in dry AMD.	Participants underwent BCVA, LLVA, contrast sensitivity, cone-specific contrast testing, and MAIA microperimetry assessment at baseline and at follow-up examination conducted one-month later ( $\pm$ 10 days).	<b>n = 30 eyes of 30 participants (20 AMD patients and 10 healthy controls)</b>  8 with early AMD (AREDS Stage 2) Mean age 67.5 $\pm$ 7.6 years 5 males, 3 females Snellen BCVA range (ETDRS letters): 20/13 (94) – 20/40 (72)  12 with intermediate AMD (AREDS Stage 3) Mean age 71.8 $\pm$ 6.8 years 7 males, 5 females Snellen BCVA range (ETDRS letters): 20/16 (90) – 20/40 (72)  10 healthy controls Mean age 69.2 $\pm$ 8.6 years 6 males, 4 females Snellen BCVA range (ETDRS letters): 20/13 (97) – 20/25 (83)	High test-retest repeatability was found at one month for all visual function metrics (LLVA, CSCT and MAIA microperimetry; intraclass correlations >0.7) with the exception of log contrast sensitivity (intraclass correlations 0.6).  Compared with age-matched controls, patients with intermediate AMD showed significant deficits on BCVA, LLVA, percent-reduced threshold on microperimetry and red CSCT but not on contrast sensitivity, green and blue CSCT.
Chen et al (2011) <sup>39</sup>	Nidek MP-1 is able to detect subtle decline in function in inherited and age-related atrophic macular disease with stable visual acuity	Retrospective exploratory review  Single centre (Moorfields Eye Hospital, London, UK)  No control	To investigate whether the Nidek MP-1 microperimeter can detect subtle functional decline in patients with progressive atrophic macular disease but stable VA.	Retrospective review of patient data collected at three routine clinical visits. Each patient had undergone three serial microperimetry tests at baseline, 6 months and 12 months. BCVA and fundus autofluorescence images were also performed at each visit.  Changes in the following outcome measures were analysed for each of the three time-points: BCVA, fixation characteristics on microperimetry, retinal sensitivity on microperimetry (both overall mean macular sensitivity and regional sensitivity based on specific topographical and functional areas), and fundus appearance on autofluorescence imaging.	<b>n = 9 eyes of 9 patients with atrophic macular disease</b> Median age 59 years (range 38 – 76 years) Median LogMAR BCVA at baseline 0.3 (range 0.18 – 0.78)  3 with ABCA4 retinopathy (macular dystrophy) Median age 45 years (range 38 – 55 years) 1 male, 2 females  3 with GA secondary to AMD Median age 73 years (range 60 – 76 years) 1 male, 2 females  3 with macular dystrophy of unknown genetic cause Median age 59 years (range 41 – 68 years) 2 males, 1 female	Nidek MP-1 microperimetry can detect significant regional sensitivity decline in patients with atrophic macular disease, stable VA, and progressive atrophy on fundus autofluorescence. In particular, sensitivity within the central macular region and at the edge of a dense scotoma showed statistically significant decline within 6 months or 12 months.  No significant change in overall mean macular sensitivity was observed over 12 months and no patient had a decline or improvement in overall mean MS beyond that which would be typical of test-retest variability (2.2 dB).
Dinc, Yenerel, Gorgun & Oncel (2008) <sup>40</sup>	Assessment of macular function by microperimetry in intermediate age-related macular degeneration	Retrospective review  Single centre (Yeditepe University Eye Hospital, Istanbul, Turkey)  Controlled	To evaluate retrospectively the central retinal function of patients with intermediate AMD using the MP-1 microperimeter.	Macular function was evaluated in all participants using the MP-1 microperimeter. Mean sensitivity (MS), mean defect (MD) parameters, fixation patterns, and fixation localisations were assessed. Testing was conducted during a single visit.	<b>n = 60 eyes of 60 participants (30 AMD patients and 30 healthy controls)</b>  30 eyes of 30 patients with intermediate AMD Mean age 67.7 $\pm$ 7.3 years (range 55 – 81 years) 16 males, 14 females Mean Snellen BCVA between 20/32 – 20/20  30 healthy eyes of 30 participants Mean age 68.7 $\pm$ 5.4 years (range 59 – 83 years) Gender breakdown not specified Mean Snellen BCVA between 20/25 – 20/20	When microperimetry findings of patients with intermediate AMD were compared to the control group, mean macular sensitivity was significantly decreased and mean defect significantly increased in the intermediate AMD group.  Fixation was predominantly central and stable in most participants, however a small number of patients in the intermediate AMD group demonstrated unstable fixation and extrafoveal localisation.
Hartmann et al (2011) <sup>41</sup>	Scanning laser ophthalmoscope imaging stabilized microperimetry in dry age-related macular degeneration	Observational cross-sectional study  Control group included but data from age-matched controls was not analysed  Single centre (Jacobs Retina Centre, University of California, California)  Testing conducted in a single visit. No longitudinal follow-up	To determine the effect of drusen and GA in dry AMD on focal retinal sensitivity using eye tracking SLO microperimetry, and to correlate retinal sensitivity with SD-OCT in these patients.	Retinal sensitivity was tested using the OPKO SLO microperimeter and structural fundus changes were measured with SD-OCT at precisely colocalised retinal points.  Threshold perimetry was performed over individual drusen or retinal features.  Drusen volume, diameter and height were graded, and inner segment/outer segment (IS/OS) junction integrity score calculated based on SD-OCT imaging.	<b>n = 44 eyes of 33 patients with drusen or GA secondary to dry AMD</b>  28 eyes of 22 patients with drusen secondary to dry AMD Age range 65 – 88 years 8 males, 14 females ETDRS BCVA better than 20/32  16 eyes of 11 patients with GA secondary to dry AMD Age range 62 – 90 years 7 males, 4 females ETDRS BCVA ranged between 20/80 – 20/20  25 age-matched control eyes from 16 patients Gender breakdown and age range not specified ETDRS BCVA better than 20/32 Data from this group was not included in the analysis	SLO microperimetry can detect changes in retinal sensitivity in AMD patients overlying drusen and at the margin of GA.  Retinal sensitivity overlying individual drusen was significantly reduced compared with the adjacent uninvolved retina. There was a significant correlation between retinal sensitivity and drusen volume, as well as IS/OS junction integrity.  IS/OS junction integrity was found to be the strongest predictor of retinal sensitivity over other predictive factors such as drusen volume, diameter and height.  In eyes with GA, an absolute scotoma was confirmed. Retinal sensitivity at the margin of GA was significantly decreased compared with the adjacent uninvolved tissue.
Meleth et al (2011) <sup>42</sup>	Changes in retinal sensitivity in geographic atrophy progression as measured by microperimetry	Prospective cohort study  Single centre (National Eye Institute, National Institutes of Health, Bethesda, Maryland, US)  Controlled	To characterise changes in macular sensitivity during GA progression over a 24-month period using microperimetry.	Retinal sensitivity of the central 20° of the macula was evaluated using MP-1 microperimetry every 6 months over a 24-month period. Microperimetric parameters of interest included number of scotomatous points, mean retinal sensitivity of responding points, and fixation stability. Autofluorescence imaging and coloured fundus photography was also obtained.	<b>n = 18 eyes of 9 patients with bilateral GA secondary to AMD</b>  Patients had been enrolled in an interventional Phase II drug trial in which one eye had been randomised to treatment and the fellow eye observed (control)  Mean age 76.8 $\pm$ 8.27 years (range 65 – 88 years) 3 males, 6 females Mean ETDRS BCVA at baseline 52 $\pm$ 17.6 letters (range 9 – 79 letters)	Mean number of scotomatous points increased significantly as a function of time (at a rate of 4.4 points per year).  Mean retinal sensitivities of all tested points, all responding retinal points, and all peri-lesional points (responding points just outside of a functional scotoma) all decreased significantly with time, as did fixation stability.  Growth of GA lesion area (based on fundus photography and autofluorescence imaging) was significantly associated with the observed changes in the number of scotomatous points but not with the changes in the other microperimetric parameters.

Midena et al (2007) <sup>43</sup>	Microperimetry and fundus autofluorescence in patients with early age-related macular degeneration	Cross-sectional study  Single centre (Medical Retina Clinic, Department of Ophthalmology, University of Padova)  No control  Testing conducted in a single visit. No longitudinal follow-up	To compare microperimetry and fundus autofluorescence of the macular in patients with drusen and pigment abnormalities secondary to early AMD.	Retinal sensitivity was assessed on all participants using the MP-1 microperimeter. Coloured fundus photography and autofluorescence imaging of the macular were recorded at the same visit. Microperimetry results were topographically superimposed over FAF images.	<b>n = 13 eyes of 13 patients with bilateral early AMD</b>  All eyes had presence of drusen and associated RPE changes, but no clinical evidence of GA  Mean age 76.2 ± 6.55 years  3 males, 10 females  ETDRS BCVA 20/20 in all eyes	A significant reduction in retinal sensitivity was observed over areas characterised by large soft drusen and/or pigment abnormalities.  Large drusen had a greater impact on retinal sensitivity than RPE pigment abnormalities alone. However, when both characteristics were present the reduction was greater than compared with either characteristic in isolation.
Wu, Ayton, Guymier & Luu (2014) <sup>44</sup>	Comparison between multifocal electroretinography and microperimetry in age-related macular degeneration	Cross-sectional study  Multi-centre (private ophthalmology clinics and RVEEH, Melbourne, Australia)  Controlled	To correlate and compare the magnitude of functional deficits obtained with mfERG testing versus MAIA microperimetry in eyes with intermediate AMD.	mfERG and MAIA microperimetry testing was performed on one eye of each participant. Thirteen hexagons in the central three rings of a 103-hexagon stimulus grid for mfERG and retinotopically matched points on microperimetry were chosen and converted into standard deviations away from that of control eyes (Z score) to represent the magnitude of measured functional deficit and allow a direct comparison of the two measures.	<b>n = 104 eyes from 104 participants</b>  <i>60 eyes of 60 patients with intermediate AMD</i>  Mean age 69.6 ± 7.5 years (range 51 – 89 years)  Mean logMAR BCVA 0.02 ± 0.10	<i>44 eyes of 44 control participants</i>  Age range 53 – 86 years  Mean logMAR BCVA -0.13 ± 0.10  No significant correlation was found between average Z scores of retinal sensitivity with the average Z scores of mfERG implicit time or response amplitudes.  Functional deficit measured using MAIA microperimetry was greater than that measured using mfERG (response amplitude and implicit time) for corresponding points at all three rings in eyes with intermediate AMD.
Wu, Ayton, Guymier & Luu (2014) <sup>36</sup>	Low-luminance visual acuity and microperimetry in age-related macular degeneration	Cross-sectional study  Multi-centre (private ophthalmology clinics & RVEEH, Melbourne, Australia)  Controlled	To compare the effectiveness of LLVA and microperimetry in assessing functional deficit in patients with dry AMD.	ETDRS BCVA, LLVA and MAIA microperimetry was performed on one eye of each participant. Low luminance deficit (LLD) was calculated as the difference between LLVA and BCVA.  The results of functional testing were compared across 6 clinical severity groups. The relationship and strength of any correlation between different functional parameters was evaluated and compared.	<b>n = 205 eyes of 205 participants</b>  <i>179 eyes of 179 patients with a clinical spectrum of different dry AMD severity (early, intermediate, and non-foveal GA)</i>  Mean age and BCVA dependent on clinical severity classification – refer Table 1 (p. 1614) and Figure 1A (p. 1615)  Gender breakdown not specified	<i>26 eyes of 26 control participants</i>  Mean age 65.5 ± 5.0 years  BCVA data – see box plot data Figure 1A (p. 1615)  Gender breakdown not specified  BCVA, LLVA and macular sensitivity were significantly reduced for all AMD clinical severity groups when compared with controls, except for those (in Group 2) with drusen between 63 and 125 µm.  LLD in AMD clinical severity groups was not significantly different from control participants, with the exception of those in the non-foveal GA group.  A significant positive relationship ( $R = 0.613$ ) between macular sensitivity and LLD, but not BCVA, was found, suggesting that LLVA may detect a greater extent of functional deficit than BCVA in eyes with increasingly poorer retinal sensitivity.
Wu, Ayton, Luu & Guymier (2015) <sup>37</sup>	Longitudinal changes in microperimetry and low luminance visual acuity in age-related macular degeneration	Prospective, longitudinal study  Single centre (Centre for Eye Research Australia, Melbourne, Australia)  Controlled	To investigate whether microperimetry and LLVA can detect functional changes over a 12-month period in patients with intermediate AMD.	Participants underwent ETDRS BCVA, LLVA, multimodal imaging (CFP, NIR, SAF, SD-OCT), MAIA microperimetry (mean point-wise sensitivity [PWS]) and clinical examination.  Participants with AMD were seen at three visits during a 12-month period at 6-month intervals (baseline, 6 months and 12 months) and all control participants were seen at two visits (baseline and 12 months).  Pathological progression was assessed in eyes with intermediate AMD by visual comparison of coloured fundus photographs obtained at baseline and 12 months. Eyes were graded as 'stable', 'progressed/worsened', or 'improved'.	<b>n = 49 eyes of 49 patients with AMD and 10 eyes of 10 healthy controls</b>  <i>41 eyes with intermediate AMD</i>  Mean age 68.8 ± 9.2 years (range 50 – 87 years)  Gender breakdown not specified  Baseline BCVA not specified	<i>8 eyes with non-foveal GA secondary to AMD</i>  Mean age 69.1 ± 6.8 years (range 58 – 79 years)  Gender breakdown not specified  Baseline BCVA not specified  <i>10 healthy controls</i>  Mean age 66.0 ± 3.5 years (range 60 – 72 years)  Gender breakdown not specified  Baseline BCVA not specified  No significant changes in BCVA or LLVA were detected in any of the groups over the 12-month period.  A significant reduction in mean microperimetric PWS was detected at 12 months compared with baseline in eyes with intermediate AMD graded as stable or worsened. A significant improvement in mean PWS was detected in eyes graded as improved.  A significant reduction in mean PWS was identified in eyes with non-foveal GA at both 6 months and 12 months compared with baseline.  No significant change in mean PWS was detected over a 12-month period in control eyes.
Wu, Guymier & Finger (2016) <sup>45</sup>	Low luminance deficit and night vision symptoms in intermediate age-related macular degeneration	Cross-sectional study  Multi-centre (private ophthalmology clinics & RVEEH, Melbourne, Australia)  No control	To investigate the relationship between self-reported night vision symptoms and visual function measures in bilateral intermediate AMD.	All participants underwent BCVA, LLVA and MAIA microperimetry at a single visit. The Night Vision Questionnaire (NVQ-10) was completed by all participants at the same visit and used to assess the degree of self-reported night vision difficulties experienced by the patient.  Low luminance deficit (LLD) was calculated as the difference between LLVA and BCVA.	<b>n = 200 eyes of 100 patients with bilateral intermediate AMD</b>  Mean age 69.8 ± 7.4 years (range 51 – 81 years)  26 males, 74 females Mean BCVA (logMAR) of better seeing eye -0.03 ± 0.09 (range -0.22 to 0.16)	Responses on the NVQ-10 significantly correlated with LLD but not BCVA, LLVA or MAIA microperimetry retinal sensitivity. Participants with the highest degree of self-reported night vision problems had significantly worse LLD than those with the least difficulty.

In eyes with intermediate AMD graded as either stable or worsened, a significant reduction in mean point-wise sensitivity was not identified by the second visit (6 months) compared with baseline, but was detected by the third visit (12 months) compared with baseline. In AMD eyes graded as improved, a significant improvement in mean point-wise sensitivity was detected at 12 months, but not 6 months. No significant change in mean point-wise sensitivity was detected over a 12-month period in control eyes. These findings demonstrate that microperimetry is capable of detecting subtle functional changes over a 12-month period in eyes with dry AMD, even when visual acuity outcomes remain seemingly unchanged.<sup>37</sup>

Collectively, the above two studies indicate that microperimetry represents a valuable tool in quantifying visual deficits in patients with non-foveal GA secondary to dry AMD and should be considered as a means of assessing the efficacy of novel interventions in patients.<sup>36,37</sup> However, an important methodological limitation of both studies was that the number of eyes in each non-foveal GA sub-group was small ( $n = 13$  and  $8$ , respectively). Larger studies are required to increase the generalisability of these findings. Additionally, both studies only correlated patient performance on microperimetry with visual acuity parameters. They did not investigate the relationship between microperimetry and other functional measures, such as reading speed or patient-reported outcome measures (PROMS) such as self-reported visual function and quality of life questionnaires, in this clinical population.

Patient-centred outcome measures often involve the participant reporting the degree of difficulty that they experience across a variety of different domains such as near function, distance function and colour appreciation.<sup>46</sup> Multi-factorial assessment is necessary in these patients because although psychophysical tests such as BCVA, LLVA and contrast sensitivity can reveal specific visual function deficits, they do not capture the entire range of effects that a disease might incur.<sup>46</sup> Additional studies incorporating a multimodal approach are warranted to more fully assess visual impairment in these patients and explore the relationship between different clinical endpoints.

### **Functional deficits in patients with dry AMD more generally**

The remaining eight studies included as part of this systematic review explored functional deficits in patients with dry AMD more broadly.<sup>38-45</sup> As mentioned, they failed to differentiate the sample demographic with respect to foveal versus perifoveal GA involvement. As such, it remains unclear as to whether each sample did in fact include patients in the population of interest to us. Notwithstanding, the findings of these eight studies have been discussed below in the broader context of dry AMD.

### **Visual acuity and microperimetry parameters**

Compared with controls, patients with dry AMD demonstrate significant deficits on BCVA and LLVA,<sup>38</sup> and microperimetry outputs including mean macular sensitivity<sup>40,42</sup> and percent-reduced threshold.<sup>38</sup> A longitudinal cohort study of nine patients (18 eyes) with bilateral GA secondary to AMD showed that mean retinal sensitivities of all tested points, all responding retinal points, and all peri-lesional points (responding points just outside of a functional scotoma) all decreased significantly with time.<sup>42</sup> Others, however, have failed to detect any significant change in overall mean macular sensitivity.<sup>39</sup> Chen et al<sup>39</sup> found no significant change in overall mean macular sensitivity measured using microperimetry over a 12-month period in their sample of nine patients (9 eyes) with progressive, atrophic macular disease but stable BCVA. Instead, they reported a significant regional sensitivity decline upon dividing the retina into specific topographical and functional areas, whereby sensitivity within the central macular region and at the border of dense scotomata showed significant decline within 6 months and 12 months of baseline. Another study similarly reported regional-based sensitivity changes, whereby retinal sensitivity at the margin of GA was significantly decreased compared with adjacent, uninvolved tissue.<sup>41</sup>

Incongruity in the above findings may be owing to differences in the methodology across studies. The retrospective review by Chen et al<sup>39</sup> was conducted at a single centre and included only nine eyes in the analyses. As such, the sample may not have been representative of the population and did not provide sufficient power to reach statistically valid conclusions. The other studies had small but somewhat larger sample sizes of 18 to 30 eyes.<sup>40,42</sup> Furthermore, the type of microperimeter used by the investigators differed across studies, with some using the Nidek MP-1 microperimeter<sup>39,40,42</sup> and others using the MAIA microperimeter<sup>38</sup> or OPKO SLO microperimeter.<sup>41</sup> This may have in turn led to differences in the microperimetric outcomes measured.

### **Correlation between microperimetry and patient-reported outcome measures (PROMs)**

Only one of the 10 papers in this systematic review included PROMs as a clinical endpoint.<sup>45</sup> No other study incorporated PROMs, nor correlated self-reported outcomes with other observed measures of visual function and functional vision. Wu, Guymer and Finger<sup>45</sup> examined the relationship between night vision symptoms measured using the 10-item Night Vision Questionnaire (NVQ-10) with BCVA, LLVA and microperimetry. In their cross-sectional study of 100 patients with bilateral intermediate dry AMD, they found low luminance deficit but not BVCA, LLVA or microperimetric retinal sensitivity (neither mean sensitivity nor central sensitivity) to be significantly associated with the

degree of self-reported night vision symptoms measured using the NVQ-10. Participants with the highest degree of self-reported night vision problems had significantly worse low luminance deficit than those with the least difficulty. These findings suggest that low luminance deficit may be a useful measure that can be readily implemented in clinical practice and may better capture the degree of night vision symptoms experienced by patients with intermediate AMD than the conventional measure of BCVA.<sup>45</sup>

The NVQ-10 implemented by Wu et al<sup>45</sup> uses a 10-item scale adopted from four items of the 25-item National Eye Institute Vision Functioning Questionnaire (NEI VFQ-25), and six items of a night vision symptom list. It is important to note that whilst the NVQ-10 has been used previously in other studies involving AMD participants,<sup>47,48</sup> it has not been formally validated. As such, its internal validity remains unknown. Also, in the study by Wu et al<sup>45</sup> it is not specified how the NVQ-10 was administered. If the questionnaire was interviewer-administered by study personnel who were also involved in the patients' care, it is possible that participant responses were at high risk of bias. The lack of a control group also prevents a comparison between the degree of self-reported night vision symptoms experienced by patients with AMD versus those of healthy, unaffected individuals. Future studies might also benefit from the inclusion of other visual function and functional vision parameters, such as contrast sensitivity and near visual acuity, to further explore the relationship between these measures and PROMs.

#### **Correlation between microperimetry and multi-focal electroretinography (mfERG)**

Multi-focal electroretinography (mfERG) is being increasingly utilised in the study of eyes with dry AMD,<sup>49-52</sup> but to date few studies have investigated the relationship between microperimetry and mfERG in quantifying visual deficits owing to dry AMD.<sup>44</sup> A controlled cross-sectional study by Wu, Ayton, Guymer and Luu<sup>44</sup> found that microperimetry might be better able to detect subtle differences in retinal function compared to mfERG in patients with intermediate dry AMD.

When considering retinotopically matched points, no significant correlation was found between the average z-scores of microperimetric retinal sensitivity and mfERG implicit time, nor response amplitudes. Furthermore, the magnitude of the measured functional deficit captured by microperimetry was significantly greater than that measured by mfERG parameters.<sup>44</sup> A comparison of the converted z-scores for mfERG and BCVA testing further revealed that there was no significant correlation between mfERG parameters and BCVA outcomes. The measured functional deficit in mfERG implicit time was not greater than BCVA, and the measured functional deficit of mfERG response amplitude was significantly less than BCVA.<sup>44</sup> At the outset, microperimetry appears a sensitive means of

assessing visual dysfunction experienced by patients with dry AMD, but further studies are necessary to ascertain its usefulness in evaluating visual deficits in patients with perifoveal GA specifically.

The current review was limited in that the data were non-synthesisable and heterogeneity in the methodological approaches and outcomes precluded a meta-analysis from being performed. Furthermore, the search was restricted to English language articles only and as such, non-English papers were not included in this review.

## **CONCLUSION**

This systematic review identifies a clear need for additional studies with a specific focus on the sub-population of dry AMD patients with perifoveal GA. Notwithstanding their limitations, the above studies provide a platform for further research in this field and point to the usefulness of employing a battery of tests when assessing functional deficits in patients with dry AMD, rather than one sole method of testing. For ongoing and future clinical studies of GA, a multimodal approach to assessment will facilitate greater understanding of the nature and progression of this disease and the real-life impact it has on patients, beyond performance in the examination chair. Indeed, existing models used to assess patient eligibility for disability support need to shift away from strictly unimodal or bimodal methods of testing, for example BCVA +/- standard automated perimetry. Adopting a multimodal approach to assessment is especially necessary in this clinical population given the unique pathogenesis of the disease. Doing so will allow a more comprehensive evaluation of visual function and functional vision that will ultimately better capture the very real and oppressing limitations often experienced by these patients. Seldom does a visual acuity of 6/12 or better provide a true account of a patient's visual dysfunction in the real world.

## **ACKNOWLEDGEMENTS**

This work was carried out whilst the first author was the recipient holder of a La Trobe University Postgraduate Research Scholarship.

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## Selected Abstracts from the Orthoptics Australia 75th Annual Scientific Conference held in Adelaide, 19th to 21st November 2018

### PATRICIA LANCE LECTURE ASSOCIATION AND PROFESSION - YOU CAN'T HAVE ONE WITHOUT THE OTHER

**Marion Rivers**

The presentation examined the relationships between profession and association. It investigated what makes a profession and why a profession needs an association. It also investigated what makes a professional and how that differs from a technical expert. We will continue the exploration of professionalism to incorporate modern professional associations and their governance, examining the co-dependence between association and profession.

The presentation examined the current state of Orthoptics Australia and where it might be heading in the future to best serve the needs of a diversified workforce. Heading into the future, Orthoptics Australia must provide the best possible member benefits to support the growth of the profession of orthoptics.

### THE PREVALENCE OF EYE CONDITIONS IN CHILDREN ADMITTED TO NEONATAL INTENSIVE CARE UNITS

**Felicia Adinanto**

**Purpose:** To compare the prevalence of eye conditions in children who had been admitted to Neonatal Intensive Care Units (NICU) and those who had not.

**Methods:** The Sydney Children's Eye Disease Study examined 2,446 children between six months and six years. All children underwent a comprehensive ocular examination including visual acuity, cover test and cycloplegic refraction.

**Results:** A total of 150 children were reported to have been admitted to NICU. Overall, 28.7% of children admitted to NICU presented with some form of eye condition and this did not differ from those who had not been admitted (23.5%,  $p=0.148$ ). However, there was a significantly higher prevalence of strabismus in children admitted to NICU (6.7%) compared to those not admitted (2.8%,  $p=0.007$ ). There was also a significant difference in the prevalence of refractive errors with a higher prevalence of myopia ( $\leq -0.50D$ , 6.8%,  $p=0.007$ ) and anisometropia (6.8%,  $p<0.0001$ ). The prevalence of ocular pathologies ( $<1.00D$ , 6.7%,  $p=0.46$ ) in children admitted to NICU was similar to those not admitted.

**Conclusions:** The overall prevalence of eye conditions with children admitted to NICU is similar to those who had not been admitted, however there was an increased risk of developing strabismus, myopia and anisometropia.

### ORTHOPTIC LED PAEDIATRIC DIABETIC RETINOPATHY SCREENING CLINIC IN SOUTH AUSTRALIA

**Gulsah Bakar**

This presentation outlined the paediatric diabetic retinopathy screening protocol developed and implemented by the orthoptists and ophthalmologists at Flinders Medical Centre.

The aim of the orthoptic led screening clinic is to improve clinic and

patient flow, as well as to provide better patient care to paediatric diabetic patients through a multidisciplinary approach involving orthoptists, ophthalmologists, endocrinologists and paediatricians.

### A IS FOR ATROPINE

**Louise Brennan**

It is quite remarkable that a drug that has been used in ophthalmology since the 1800s still remains at the forefront of eye care in paediatrics. Atropine is well known for its use in facilitating fundus examination and objective refraction. It is also used to treat uveitis, it can be helpful in the glasses adaptation of hypermetropes and more recently low dose atropine is being used to aid myopia control. Most commonly in the paediatric setting atropine is used to augment amblyopia treatment.

The cautions, downsides, benefits and tricks of using atropine as part of an amblyopia treatment plan were discussed.

### WHERE ORTHOPTICS AND OPTOMETRY DIVERGED IN THE NON-SURGICAL MANAGEMENT OF STRABISMUS

**Shayne Brown**

Orthoptics as practised by orthoptists evolved from ophthalmology. Ophthalmologists recognised that the time-consuming therapy required to restore binocular vision to patients with strabismus could be undertaken by 'lay people'. These 'lay people' became the first orthoptists. By the late 1920s orthoptics was in its early stages of development in London. Ophthalmic opticians (optometrists as they are known in Australia) also practised a form of orthoptics in 1920s London. This paper explored the theories underlying the difference in orthoptics as practised by orthoptists trained in the medical model compared with optometric practice. It was shown that orthoptics had its roots in the physiology of eye movements and binocular vision, especially in the understanding of depth perception. Whereas the optometrists based their practice on an understanding of eye movements and depth perception based on psychological theories. The fundamental difference in approach goes some way in explaining why ophthalmologists trained orthoptists to follow the physiological, and therefore the medical evidence path in preference to working with optometry which based its practice on a non-medical model.

### ORTHOPTIC LED VISION SCREENING SERVICE: ONE YEAR IN

**Jessica Collins**

This presentation continues on from the 74th OA Annual Conference where the design of the orthoptist led vision screening service was described. With limited resources comprising only three orthoptists and a small suitcase, the service is now in its first year of operation at the Women's and Children's Hospital in Adelaide. This presentation provided an update on the service's successes, pitfalls, barriers and solutions encountered along the way. It highlighted the importance of an orthoptist's role in the context of a demanding public health system.

## RETINOPATHY OF PREMATURITY IN RETROSPECT: TRENDS IN RETINOPATHY OF PREMATURITY OVER A 10-YEAR PERIOD

Renee Fernandez

**Abstract:** Retinopathy of prematurity (ROP) is a potentially blinding condition which affects the developing retinal blood vessels of infants born prematurely. Improvement in neonatal care over the years has led to increased survival of extremely premature infants, who are at particular risk of developing ROP. This retrospective study will report on the incidence of ROP in a population of premature infants admitted to a South Australian neonatal care unit over a 10-year period. It will further examine the relationship between gestational age, birthweight, incidence and severity of ROP.

## EYES ON THE HORIZON: PATHWAYS TO PRIVATE ORTHOPTIC PRACTICE

Julie Fitzpatrick

We live in an aging population and two-thirds of Australians with low vision are aged 65 years or over. Age-related macular degeneration, cataract, glaucoma and diabetic retinopathy (DR) can all cause vision loss to varying degrees, with DR on the increase.

Orthoptists are perfectly placed to deliver high quality and 'person-centred' low vision care to these individuals in need, or people at any stage of their lives. There is an emerging group of orthoptists setting up to provide fee-for-service based assessments. Additional funding opportunities through Veterans Affairs, NDIS, TAC and My Aged Care are making this an exciting time for both providers and clients.

The presentation discussed common challenges based on recent research, regarding the perception of low vision and uptake of services, and how these can be considered for better outcomes. With more independent practitioners in the arena, awareness of orthoptic services will only increase.

## A RETROSPECTIVE REVIEW OF RETINOBLASTOMA IN A TERTIARY SETTING

Katie Geering

Retinoblastoma is the commonest primary malignant tumour of childhood. The Children's Hospital at Westmead is the primary centre for treatment in NSW. A retrospective review has been conducted on patients who were diagnosed with retinoblastoma within the last five years at The Children's Hospital at Westmead. The presenting reason, patient's age, visual outcome and treatment type was discussed.

## CLOSING THE GAP OF VISION BY 2020 – WHERE ARE WE AT?

Rosamond Gilden

Aboriginal and Torres Strait Islander Australians still suffer higher rates of vision loss and reduced access to eye services than other Australians. The Roadmap to Close the Gap for Vision (2012) provides 42 sector-endorsed, evidence-based recommendations that address this inequity of vision and eye health. Significant progress is being made as evidenced in the National Eye Health Survey (2016), Roadmap Annual Update (2017), and Australian Institute of Health and Welfare (2018) reports. From the 2017 Roadmap Annual Update, 16/42 recommendations were fully implemented, with two-thirds of intermediate steps complete. Roadmap activity is occurring in more than 37 regions across the country, covering 60% of the Indigenous population. The 2018 report outlines further progress.

Findings from the NEHS and AIHW, showed blindness rates had halved from six times (in 2008) to three times, with increased rates of diabetic retinopathy screening and cataract surgery of Indigenous Australians. Although great progress has been made in improving eye health outcomes of Indigenous Australians, the same is also occurring for other Australians. This presentation provided an update on achievements and progress since the last Annual Update (2017) and what still remains to be done to Close the Gap of Vision by 2020.

## ORTHOPTICS AUSTRALIA WORKFORCE SURVEY

Mara Giribaldi

**Aim:** To present data collected from the Orthoptics Australia workforce survey 2016/17 about current demographics, academic qualification, employment patterns and current professional practice.

**Method:** Financial members of Orthoptics Australia as well as non-members were encouraged to participate in the online workforce survey. Data was collected from 1st July 2017 to December 2017 using the online survey tool, Survey Monkey.

**Results:** The presentation reported on the data collected surrounding gender, distribution, age, nationality, place of residence, location and status of employment and diverse clinical areas of practice.

**Conclusion:** This workforce survey provides information about the profession in 2016/17. The results are a platform for data analysis for use in the future.

## 'CROUCH, TOUCH, PAUSE, ENGAGE': USING A VISUAL TOOL TO DETECT CONCUSSION IN RUGBY UNION

Premkumar Gunasekaran

**Aim:** To determine the utility of the King-Devick test (K-D) in diagnosing concussion and identifying its incidence in semi-professional rugby players.

**Methods:** Forty male rugby players (mean age 23.48 ( $\pm$ 3.7 years) (22 forwards and 18 backs) who played in the top two divisions at the Randwick Rugby Club, Sydney, were recruited. Thirty-six players performed K-D test as a pre-season baseline. Twenty-eight were followed-up to repeat the test throughout 18 matches and following a concussion diagnosis.

**Results:** There were 112 injuries across the season with three direct ocular-related injuries, including one orbital fracture. Seven were diagnosed concussions (85.7% in forwards and 14.3% in backs) resulting in 9.72 concussions per 1,000 match hours. Baseline testing resulted in an average completion time of the K-D of 41.4 ( $\pm$ 7.89) seconds. Players that repeated the test throughout the season demonstrated significant improvements to their baseline; 40.25 ( $\pm$ 7.1) vs 36.41 ( $\pm$ 6.1) seconds,  $p < 0.001$ . Concussed athletes displayed average K-D scores that were significantly worse than baseline; 33.63 ( $\pm$ 5.4) vs 36.04 ( $\pm$ 6.0) seconds,  $p = 0.032$ , with a mean difference of 2.41. One player demonstrated a two-second improvement post-concussion.

**Conclusion:** The K-D test was useful in detecting concussion within this cohort. Results reflect over double the rate of concussion previously reported of 4.73 concussions per 1,000 match hours.



## YOUR ROLE AS AN ORTHOPTIST AND THE NDIS

**Laura Hartley**

Our role as an orthoptist has once again diversified since the introduction of NDIS. It has had a huge impact in the community and health sector. Many of us are still learning, not confident and quite unsure about NDIS. My presentation aimed to explain, educate other orthoptists and promote discussion regarding the NDIS.

- What is NDIS? who is eligible, when and how to refer a patient for NDIS services
- What is an NDIS plan? How does it come about? What to do with it
- Ability for participants to have choice (eg self-managed vs NDIS-managed) and control
- The impact the NDIS has had on low vision services such as fee for service and the change this has had on services
- The impact NDIS has had on our role as orthoptists. Our profession being recognised, skills acknowledged consequently given authority as assistive technology assessors.

## WORKING AS AN ORTHOPTIST AT THE CHILDREN'S HOSPITAL AT WESTMEAD

**Amy Huynh**

Working as an orthoptist in the hospital environment involves more than just working within eye clinic. There are frequent interactions with other allied health departments and low vision support services in order to diagnose and provide the best management care for our patients. An overview of the main services the eye clinic has been involved with were explored through some selected patient cases. In addition, I will share my roles and experiences as a new graduate orthoptist at CHW.

## WHAT IS THE IDEAL VISUAL ACUITY CUT-OFF FOR DETECTING OCULAR CONDITIONS IN PRESCHOOL CHILDREN?

**Mythili Ilango**

**Purpose:** Visual acuity (VA) cut-offs used in preschool vision screening programs are not universal. We aimed to establish the sensitivity and specificity for detection of refractive error, amblyopia and strabismus using different VA cut-offs.

**Method:** VA was measured using an electronic vision chart (single-surround HOTV) on 216, 4-year-old children. An orthoptic examination and cycloplegic autorefractometry (cyclopentolate 1%, Canon RK-F1) were conducted.

**Results:** Using a VA cut-off of 6/15 and 6/18, 100% sensitivity was achieved, however lower specificity (74.2% and 73.8%, respectively) meant a number of children with ocular conditions (25.8%, n=55 and 26.2%, n=56, respectively) would go undetected. At 6/9.5 and 6/12, higher specificity was achieved, particularly with 6/9.5 (sensitivity 84.6%, specificity 76.8%). Of the 203 children who achieved 6/9.5 or better VA, 47 were false negatives; mainly with hyperopia  $\geq 2.00D$  (63.8%), but 23.4% had strabismus and/or amblyopia. Cover test combined with 6/9.5 VA cut-off, improved sensitivity (89.5%) and specificity (79.2%).

**Conclusion:** There was a good sensitivity and specificity using a 6/9.5 VA cut-off, making it appropriate for vision screening programs. However, a number of children with amblyopia and/or strabismus passed at the 6/9.5 cut-off, which indicates that a cover test may improve detection of these childhood eye conditions.

## DEVELOPING THE ORTHOPTIST'S SCOPE OF PRACTICE IN A GENERAL OPHTHALMIC SETTING

**Sevag Ipradjian**

As demand and expectations of our patients continue to rise: we can meet them. As more orthoptists move away from ocular motility, traditional scope of practice boundaries are being tested. This presentation aimed to enlighten and empower orthoptists to initiate and assist in growth and development, not only in the work-place, but our profession. The management of many common conditions demonstrate the increase in dependence on orthoptists and our skills.

- Specialised dry eye treatment systems and how the orthoptic staff play a vital role for cataract and MGD patients
- Satellite outreach clinics and how the orthoptist can assist with volume and flow
- Postoperative care – a developing role for orthoptists
- Glaucoma monitoring and their six-monthly reviews.

We are increasingly sharing the workplace with optometrists, nurses and technicians – but have much to offer in the field of ophthalmic sciences. Meeting the demands of patients is a critical aspect of business development, and of course – patient confidence.

## THE SUCCESS AND PITFALLS OF INVERSE OCCLUSION

**Lindley Leonard**

The role of inverse occlusion in clinical practice was discussed. Case studies highlighted the perceived success and pitfalls when making clinical decisions for children who have eccentric fixation.

## ORTHOPTISTS HELPING TO IMPROVE ACCESS TO EYE HEALTH CARE

**Catherine Mancuso**

Since 2012 there have been a series of reforms of specialist outpatient clinics in Victorian Public Hospitals with the aim to improve access to care for patients. Despite significant efforts made by the various health services, the state government has recognised that more support is required to address current system constraints and gaps impacting on timely access. Over 2017 and 2018, Better Care Victoria and the Department of Health and Human Services initiated 'The Specialist Clinics Access Improvement Partnership' (SCAIP) which was formed with 11 Victorian health services. The Royal Victorian Eye and Ear Hospital (E+E) was one of those health services. Orthoptists have long been recognised as a highly skilled and versatile professional group at E+E and many different iterations of our scope of practice have been explored over the years. Three different projects were undertaken at E+E, all utilising the orthoptic workforce in different ways. These three projects were presented and the impact of the orthoptic workforce in the SCAIP will be discussed.

## VISION-RELATED QUALITY OF LIFE AS A PREDICTOR OF PROGRESSION TO LATE AGE-RELATED MACULAR DEGENERATION: SELF-REPORTED OUTCOMES FROM THE LEAD STUDY

**Myra McGuinness**

Despite major advances in the assessment of visual function and ocular structure during this century, there is no way of predicting exactly which patients with the earlier stages of age-related macular degeneration (AMD) will go on to develop later stage AMD. It has been hypothesised that patient-reported visual function may provide insight into physiological processes that are not yet clinically detectable. Participants of the Laser intervention in Early stages of Age-related macular Degeneration (LEAD) trial completed the 28-item Impact of Visual Impairment (IVI) Questionnaire and the 10-item Night Vision Questionnaire (NVQ) every year for three years in order to quantify their vision-related quality of life. The psychometric properties of these questionnaires were assessed via item response theory to validate their use among patients with intermediate AMD. Standardised scores were then analysed to assess the ability of the scales to reflect clinical measures of visual function, such as visual acuity and microperimetric sensitivity, and structural changes assessed via multimodal imaging. On average, the standardised scores decreased over the duration of the study and the risk of progression to late AMD, particularly geographic atrophy, was greater among participants who had lower questionnaire scores at baseline.

## THE REDEVELOPMENT OF THE KAY PICTURE PAEDIATRIC VISUAL ACUITY TEST

**Ashli Milling**

An accurate assessment of visual acuity (VA) is vital to inform diagnosis and management. Currently, there are a number of paediatric VA assessments available, of which few are validated yet are still in use. In the United Kingdom the Kay picture VA test, developed in the early 1980s, is one of the leading tests for pre-literate children in clinical practice. The test has since been redesigned with an aim to validate the updated optotypes to improve the resolution acuity, recognition, repeatability and to compare with other gold standard LogMAR acuity assessments.

**Methods:** To evaluate the redesign of the Kay Picture test, four stages were involved. In all phases the pictures were presented on a monitor as a single crowded optotype, with five optotypes at each VA level.

**Phase one:** Resolution acuity for 25 pictures, eight Landolt Cs and five ETDRS letters were assessed in adult subjects to ensure results were not impacted by varying cognitive abilities. Phase two: Recognition phase assessed children younger than 30 months to determine the most commonly identified pictures. Phase 3: Resolution acuity of a reduced number of pictures and the Landolt C was reassessed. Phase 4: The redesigned Kay Picture test was compared with LEA symbols and the ETDRS letters.

**Results:** Resolution acuity was assessed in 50 adults. Mean acuity scores ( $\pm$ SD) with the 25 pictures ranged from  $-0.123 (\pm 0.124)$  to  $-0.308 (\pm 0.105)$ . The mean acuity for the eight Landolt C orientations was  $-0.059 (\pm 0.120)$  and  $-0.128 (\pm 0.101)$  for the ETDRS letters. Three pictures were removed at this point. The recognition of the pictures was assessed in 420 children. Analysis resulted in removal of 10 further pictures based on the recognition. Resolution acuity was assessed in 42 adults with the remaining 12 pictures. Based on mean bias levels and further recognition data the picture selection was reduced to six. A further 113 adults were assessed with the new Kay picture test, the ETDRS and LEA symbol. The mean bias indicated similar results between the tests. The final phase evaluated the repeatability of the newly designed test and the ETDRS. Kay pictures test and the ETDRS were assessed in 100 adults, and no significant difference was found between either test (paired t-test,  $p=0.1$ ).

## EMERGING TOOLS IN THE MEASUREMENT OF TIME SPENT OUTDOORS

**Long Phan**

Time outdoors has been strongly associated with the prevention of childhood myopia in numerous studies. The proposed mechanism behind this light-mediated effect has also been supported in animal models. The implementation of this environmental modification into broad public health policies has been slow, as existing quantitative outcomes have been based upon subjective measures and accurate dose-response relationships have not been recognised. Recently, devices such as the Clouclip P2 light meter (Mirror Technology Co Ltd, Hangzhou) have emerged with the potential to precisely capture light exposures as well as near work; another significant contributor to myopia. This device can provide further detail of two important facets of time outdoors: the duration of exposure and the intensity of light required for a protective effect. Together with existing knowledge on other environmental factors such as intensive near work, education, socioeconomic status and geographic location, as well as pre-determined risk factors such as parental myopia and ethnicity, more effective intervention trials can be developed. This study investigated the validity of the Clouclip P2 as an objective device to more precisely measure outdoor time in combination with near work and compared the inherent differences between previously used illuminometers and questionnaires.

## PSYCHOSOCIAL IMPACT OF REPEATED INTRAVITREAL INJECTIONS ON PATIENTS WITH DIABETIC MACULAR OEDEMA

**Monique Rose**

Diabetic macular edema (DME) is caused by leakage of fluid from damaged blood vessels. Vascular endothelial growth factor (VEGF) is elevated in eyes with DME and drives vascular leakage. Centre-involving sight-affecting DME is currently treated with intravitreal anti-VEGF injections. It is a commonly performed procedure, which involves multiple injections every 4-8 weeks until the fluid is resolved and may be continued indefinitely to maintain vision, posing a high burden on patients. Patients differ in their personal need to undertake treatment. Patients evaluate clinician's advice and decide to follow it based on individual judgment and understanding of the illness and treatment. Theoretical models have been developed to increase understanding of treatment adherence behaviour. Horne and Weinmann developed the Necessity-Concerns Framework (NCF) to identify beliefs influencing patients' decisions to undertake medication/treatment. The NCF postulates that adherence is influenced by the necessity (personal need for the treatment) and concerns about potential adverse effects. A mixed method design (in-depth interview and self-administered questionnaires) was utilised to develop an understanding of treatment adherence in patients undergoing repeated intravitreal injection treatment for DME. The Belief of Medicines Questionnaire-Specific (BMQ-Specific) assessed patients' beliefs and adherence to intravitreal treatment and the Satisfaction with Information about Medicine Scale (SIMS) measured satisfaction with information received about treatment. The results will be presented by categorising public and private DME participants into the NCF and comparing treatment information satisfaction and qualitative reasons for treatment adherence. Enhanced awareness and understanding of nonadherence and patients' beliefs could assist in the development of interventions to improve adherence.

## DYNAMIC RETINAL VASCULAR ASSESSMENT: AN INNOVATIVE APPROACH FOR EARLY GLAUCOMA SCREENING

**Sahar Shariflou**

**Purpose:** Spontaneous venous pulsations (SVPs) are a potential biomarker for glaucomatous optic neuropathy, with reduced SVPs associated with thinner retinal nerve fibre layer and lower retinal ganglion cell (RGC) counts. We used a novel fundus imaging tool to investigate the association between SVPs and RGC estimates.

**Methods:** Forty-one participants [30 confirmed glaucoma, 74 ( $\pm 11$ ) years, 14 male; 11 suspects, 66 ( $\pm 10$ ) years, 5 male] had a 10-second video recording of venous circulation at the optic nerve head using a digital ophthalmoscope following dilation. RGC counts were estimated using established methodology (Humphrey Visual Field and Optical Coherence Tomography). SVP amplitudes were extracted from the videos using a custom written algorithm and a linear regression was applied to study the association between SVP amplitude and RGC counts.

**Results:** The mean percentile change in venous diameter (SVP amplitude) and RGC count was 38% ( $\pm 12$ ) and 635,455 ( $\pm 169,665$ ), respectively. We observed a positive association between SVP amplitude and RGC count ( $r=0.34$ ).

**Conclusion:** Our findings suggest that SVPs may be a quantitative measure of structural and functional changes in GON. This novel tool could be further developed for early screening in glaucoma.

## IMAGING IN RETINA

**Sally Steenbeck**

Sydney Eye Hospital runs a very busy uveitis clinic in conjunction with one of its medical retina clinics. The availability of multiple retinal imaging devices, such as Spectralis OCT and OPTOS, has made diagnosis both easier and faster. A few examples were presented to highlight this.

## ORTHOPTIC LED POSTOPERATIVE CLINIC

**Julie Taylor**

The current Surgical Ophthalmology Service clinic model at the Eye and Ear Hospital, Melbourne, is experiencing increased demand for postoperative cataract reviews. This demand has been generated from an increased level of surgical throughput required to support the hospital's funding agreement with the Victorian Department of Health & Human Services.

A trial of Orthoptic-led Surgical Postoperative (OSOP) Clinic was developed as an alternative sustainable model of care to support timely postoperative access for routine cataract patients. This presentation described the OSOP trial implementation process, including training requirements as well as the current state, impacts made, supporting data, and the potential future state.

## AUDIT OF CLINICAL DECISION MAKING IN AN OPHTHALMIC DIABETIC PHOTOGRAPHIC SCREENING CLINIC

**Danielle Thorburn**

The Austin Hospital, Melbourne, currently runs an Ophthalmic Diabetic Photographic Screening Clinic for patients diagnosed with diabetes. This is an orthoptist led clinic whereby patients are solely assessed by an orthoptist who screens for diabetic retinopathy. Despite the inclusion of orthoptists in this service, the patient's clinical pathway or clinical management is determined by the ophthalmology registrar upon reviewing the orthoptist's clinical notes at a later time.

The aim of this study is to investigate the agreement between orthoptists and a principal ophthalmologist, on the clinical management decision for patients with diabetes presenting to this clinic. There is scope for future extension of the orthoptists' role within this traditional service delivery model.

De-identified clinical notes and retinal photos from patients attending this clinic in 2016 were retrospectively reviewed by a senior orthoptist and principal ophthalmologist for agreement on diagnosis and treatment plan and timing. Results are currently being analysed. The first 200 eyes will be presented.

## UNBLOCKING THE SYSTEM: CHALAZION AND NASOLACRIMAL DUCT OBSTRUCTION PHONE CLINIC

**Faren Willett**

Lady Cilento Children's Hospital is the major specialist children's hospital with a large catchment area including the entire Queensland state and Northern New South Wales, receiving approximately 5,200 new referrals to Ophthalmology outpatients per year. These are then categorised by urgency according to the referral. There are numerous referrals for eye conditions such as chalazia and blocked tear ducts that may be conservatively managed at home, with improvement or complete resolution likely prior to their appointment. An orthoptist-led phone clinic has been implemented with a purpose to contact patients with these referrals to attempt to educate the family and resolve the issue before presentation. It is additionally an excellent screening tool to detect more serious pathology and ensure it is addressed in a timely manner. The phone call clinic has been in action for eighteen months, with positive impacts on waitlist numbers and waiting times.

## Named Lectures, Prizes and Awards of Orthoptics Australia

### THE PATRICIA LANCE LECTURE

1988	Elaine Cornell	Home exercises in orthoptic treatment
1989	Alison Pitt	Accommodation deficits in a group of young offenders
1990	Anne Fitzgerald	Five years of tinted lenses for reading disability
1992	Carolyn Calcutt	Untreated early onset esotropia in the visual adult
1993	Judy Seaber	The next fifty years in orthoptics and ocular motility
1995	David Mackey	The Glaucoma Inheritance Study in Tasmania (GIST)
1997	Robin Wilkinson	Heredity and strabismus
1998	Pierre Elmurr	The visual system and sports performance
1999	Kerry Fitzmaurice	Research: A journey of innovation or rediscovery?
2005	Kathryn Rose	The Sydney Myopia Study: Implications for evidence based practice and public health
2006	Frank Martin	Reading difficulties in children - evidence base in relation to aetiology and management
2008	Stephen Vale	A vision for orthoptics: An outsider's perspective
2009	Michael Coote	An eye on the future
2010	John Crompton	The pupil: More than the aperture of the iris diaphragm
2011	Neryla Jolly	On being an orthoptist
2012	Shayne Brown	A snapshot of orthoptics from the 1960s to 2000
2013	Sue Silveira	Finding the leader within
2014	Patricia Dunlop	A life in orthoptics
2015	Fiona Rowe	The spectrum of post-stroke visual impairment
2016	Linda Santamaria	50 years: The development of research and publication in the Australian Orthoptic Journal
2017	Sandra Staffieri	Delayed diagnosis of childhood strabismus: When does it matter?
2018	Marion Rivers	Association and profession - you can't have one without the other

### THE EMMIE RUSSELL PRIZE

1957	Margaret Kirkland	Aspects of vertical deviation
1959	Marion Carroll	Monocular stimulation in the treatment of amblyopia exanopsia
1960	Ann Macfarlane	A study of patients at the Children's Hospital
1961	Ann Macfarlane	A case history "V" Syndrome
1962	Adrienne Rona	A survey of patients at the Far West Children's Health Scheme, Manly
1963	Madeleine McNess	Case history: Right convergent strabismus
1965	Margaret Doyle	Diagnostic pleoptic methods and problems encountered
1966	Gwen Wood	Miotics in practice
1967	Sandra Hudson Shaw	Orthoptics in Genoa
1968	Leslie Stock	Divergent squints with abnormal retinal correspondence
1969	Sandra Kelly	The prognosis in the treatment of eccentric fixation
1970	Barbara Denison	A summary of pleoptic treatment and results
1971	Elaine Cornell	Paradoxical innervation
1972	Neryla Jolly	Reading difficulties
1973	Shayne Brown	Uses of fresnel prisms
1974	Francis Merrick	The use of concave lenses in the management of intermittent divergent squint
1975	Vicki Elliott	Orthoptics and cerebral palsy
1976	Shayne Brown	The challenge of the present
1977	Melinda Binovec	Orthoptic management of the cerebral palsied child
1978	Anne Pettigrew	
1979	Susan Cort	Nystagmus blocking syndrome
1980	Sandra Tait	Foveal abnormalities in ametropic amblyopia
1981	Anne Fitzgerald	Assessment of visual field anomalies using the visually evoked response
1982	Anne Fitzgerald	Evidence of abnormal optic nerve fibre projection in patients with dissociated vertical deviation: A preliminary report
1983	Cathie Searle	Acquired Brown's syndrome: A case report
	Susan Horne	Acquired Brown's syndrome: A case report
1984	Helen Goodacre	Minus overcorrection: Conservative treatment of intermittent exotropia in the young child
1985	Cathie Searle	The newborn follow up clinic: A preliminary report of ocular anomalies
1988	Katrina Bourne	Current concepts in restrictive eye movements: Duane's retraction syndrome and Brown's syndrome
1989	Lee Adams	An update in genetics for the orthoptist: A brief review of gene mapping



1990	Michelle Gallahe	Dynamic visual acuity versus static visual acuity: Compensatory effect of the VOR
1991	Robert Sparkes	Retinal photographic grading: The orthoptic picture
1992	Rosa Cingiloglu	Visual agnosia: An update on disorders of visual recognition
1993	Zoran Georgievski	The effects of central and peripheral binocular visual field masking on fusional disparity vergence
1994	Rebecca Duyshart	Visual acuity: Area of retinal stimulation
1995-7	Not awarded	
1998	Nathan Clunas	Quantitative analysis of the inner nuclear layer in the retina of the common marmoset callithrix jacchus
1999	Anthony Sullivan	The effects of age on saccades made to visual, auditory and tactile stimuli
2001	Monica Wright	The complicated diagnosis of cortical vision impairment in children with multiple disabilities
2005	Lisa Jones	Eye movement control during the visual scanning of objects
2006	Josie Leone	The prognostic value of the cyclo-swap test in the treatment of amblyopia using atropine
2007	Thong Le	What is the difference between the different types of divergence excess intermittent exotropia?
2008	Amanda French	Does the wearing of glasses affect the pattern of activities of children with hyperopic refractive errors?
2009	Amanda French	Wide variation in the prevalence of myopia in schools across Sydney: The Sydney Myopia Study
2010	Alannah Price	Vertical interline spacing and word recognition using the peripheral retina
2011	Amanda French	Comparison of the distribution of refraction and ocular biometry in European Caucasian children living in Northern Ireland and Sydney
2012	Melanie Cortes	Treatment outcomes of children with vision impairment detected through the StEPS program
2013	Jess Boyle	The accuracy of orthoptists in interpreting macular OCT images
2014	Allanah Cramer	Orthoptist-led clinics: investigating the effectiveness and efficiency of orthoptists in diabetic retinopathy screening and cataract assessment
2015	Jess Boyle	The psychological impact of repeated intravitreal injections on patients with neovascular age-related macular degeneration
2016	Gareth Lingham	Early life risk factors of amblyopia, strabismus and anisometropia in a young adult population
2017	Linden Chen	The twilight zone
2018	Premkumar Gunasekaran	'Crouch, touch, pause, engage': using a visual tool to detect concussion in rugby union

## PAEDIATRIC ORTHOPTIC AWARD

1999	Valerie Tosswill	Vision impairment in children
2000	Melinda Syminiuk	Microtropia - a challenge to conventional treatment strategies
2001	Monica Wright	The complicated diagnosis of cortical vision impairment in children with multiple disabilities
2005	Kate Brassington	Amblyopia and reading difficulties
2006	Lindley Leonard	Intermittent exotropia in children and the role of non-surgical therapies
2007	Jody Leone	Prevalence of heterophoria in Australian school children
2008	Jody Leone	Can visual acuity screen for clinically significant refractive errors in teenagers?
2009	Jody Leone	Visual acuity testability with the electronic visual acuity-tester compared with LogMAR in Australian pre-school children
2010	Fiona Gorski	Neurofibromatosis and associated ocular manifestations
2011	Suzy King	Understanding Sturge-Weber syndrome and the related ocular complications
2012	Jane Scheetz	Accuracy of orthoptists in the diagnosis and management of triaged paediatric patients
2013	Louise Brennan	Visual outcomes of children seen in the StEPS High Priority Clinic at The Children's Hospital at Westmead
2014	Nicole Carter	Understanding ocular motor apraxia
2015	Lindley Leonard	Long-term follow-up of a high priority referral clinic at The Children's Hospital at Westmead - beyond the clinic
2016	Cem Oztan	A novel method for measuring nystagmus
2017	Sarah Harkins	An audit of paediatric referrals of patients with suspected papilloedema made to The Children's Hospital at Westmead
2018	Renee Hernandez	Retinopathy of prematurity in retrospect: trends in retinopathy of prematurity over a 10-year period

## THE MARY WESSON AWARD

1983	Diana Craig (Inaugural)	1998	Not Awarded	2011	Zoran Georgievski)
1986	Neryla Jolly	2001	Heather Pettigrew	2014	Mara Giribaldi
1989	Not awarded	2004	Ann Macfarlane	2017	Keren Edwards
1992	Kerry Fitzmaurice	2008	Julie Barbour		
1995	Margaret Doyle	2010	Elaine Cornell		

## ZORAN GEORGIEVSKI MEDAL

2012	Neryla Jolly (Inaugural)	2015	Sue Silveira	2018	Catherine Mancuso
2013	Connie Koklanis	2016	Julie Barbour		
2014	Linda Santamaria	2017	Meri Vukicevic		

## Presidents of Orthoptics Australia

1945-7	Emmie Russell	1965-6	Beverly Balfour	1985-6	Geraldine McConaghy
1947-8	Lucy Willoughby	1966-7	Helen Hawkeswood	1986-7	Alison Terrell
1948-9	Diana Mann	1967-8	Patricia Dunlop	1987-9	Margaret Doyle
1949-50	E D'Ombra	1968-9	Diana Craig	1989-91	Leonie Collins
1950-1	Emmie Russell	1969-70	Jess Kirby	1991-3	Anne Fitzgerald
1951-2	R Gluckman	1970-1	Neryla Heard	1993-5	Barbara Walsh
1952-4	Patricia Lance	1971-2	Jill Taylor	1995-7	Jan Wulff
1954-5	Diana Mann	1972-3	Patricia Lance	1997-00	Kerry Fitzmaurice
1955-6	Jess Kirby	1973-4	Jill Taylor	2000-2	Kerry Martin
1956-7	Mary Carter	1974-5	Patricia Lance	2002-4	Val Tosswill
1957-8	Lucille Retalic	1975-6	Megan Lewis	2004-6	Julie Barbour
1958-9	Mary Peoples	1976-7	Vivienne Gordon	2006-8	Heather Pettigrew
1959-60	Patricia Lance	1977-8	Helen Hawkeswood	2008-10	Zoran Georgievski
1960-1	Helen Hawkeswood	1978-9	Patricia Dunlop	2010-13	Connie Koklanis
1961-2	Jess Kirby	1979-80	Mary Carter	2013-15	Meri Vukicevic
1962-3	Patricia Lance	1980-1	Keren Edwards	2015-16	Paul Cawood
1963-4	Leonie Collins	1981-82	Marion Rivers	2016-17	Julie Hall
1964-5	Lucy Retalic	1982-3	Jill Stewart	2017-18	Marion Rivers
		1983-5	Neryla Jolly		

## Editors and Reviewers of the Australian Orthoptic Journal

Vol 8 1966	Barbara Lewin & Ann Metcalfe	Vol 26 1990	Elanie Cornell	Vol 41 2009	Zoran Georgievski & Connie Koklanis
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Vol 11 1971	Neryla Heard & Helen Hawkeswood	Vol 29 1993	Julia Kelly	Vol 44 2012	Connie Koklanis & Linda Santamaria
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Vol 13 1973-74	Diana Craig	Vol 31 1995	Julie Green	Vol 46 2014	Linda Santamaria & Connie Koklanis
Vol 14 1975	Diana Craig	Vol 32 1996	Julie Green	Vol 47 2015	Connie Koklanis & Linda Santamaria
Vol 15 1977	Diana Craig	Vol 33 1997-98	Julie Green	Vol 48 2016	Linda Santamaria & Meri Vukicevic
Vol 16 1978	Diana Craig	Vol 34 1999	Julie Green	Vol 49 2017	Meri Vukicevic & Linda Santamaria
Vol 17 1979-80	Diana Craig	Vol 35 2000	Neryla Jolly & Nathan Moss	Vol 50 2018	Linda Santamaria & Meri Vukicevic
Vol 18 1980-81	Diana Craig	Vol 36 2001-02	Neryla Jolly & Kathryn Thompson		
Vol 19 1982	Diana Craig	Vol 37 2003	Neryla Jolly & Kathryn Thompson		
Vol 20 1983	Margaret Doyle	Vol 38 2004-05	Neryla Jolly & Kathryn Thompson		
Vol 21 1984	Margaret Doyle	Vol 39 2007	Zoran Georgievski & Connie Koklanis		
Vol 22 1985	Margaret Doyle	Vol 40 2008	Connie Koklanis & Zoran Georgievski		
Vol 23 1986	Elaine Cornell				
Vol 24 1987	Elaine Cornell				
Vol 25 1989	Elaine Cornell				

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Kerry Fitzmaurice	Natalia Kelly	Nicole Moore	Jane Scheetz	
Julie Fitzpatrick	Khoi Khuat	Julie Morrison	Jane Schuller	
Kamil Gorsky	Connie Koklanis	Vincent Nguyen	Maria Simos	
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Discipline of Orthoptics  
 School of Allied Health  
 La Trobe University  
 Bundoora, VIC 3086  
 T: 03 9479 5285  
[www.latrobe.edu.au/courses/orthoptics](http://www.latrobe.edu.au/courses/orthoptics)

#### SYDNEY

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Discipline of Orthoptics  
 Graduate School of Health  
 University of Technology  
 15 Broadway, Ultimo, NSW 2007  
 T: 02 9514 2000  
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