

A Clinical Pathway for Nystagmus?

**Parents and
children**

**Nystagmus
research in Wales**

**What can
optometrists do?**



Pictured: Shooting the forthcoming Nystagmus Network videos at Cardiff School of Optometry and Vision Science. One video will be about the impact of nystagmus and features BBC TV quiz show presenter Richard Osman as well as others who have the condition. The second video looks at treatment and research. Both videos are funded by the Giles Warman Foundation set up by the family, friends and colleagues of the late Giles Warman, whose grandson has nystagmus.

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Introduction from John Sanders, Guest Editor

Firstly, I'd like to thank WCB for giving me the opportunity to guest edit this issue of Roundup. Nystagmus (not to be confused with astigmatism) is more common than most people think. Even if you don't have nystagmus yourself, someone in your circle of family, friends and colleagues almost certainly does, although you may not know it. Several famous people have nystagmus. For instance BBC TV's Pointless presenter Richard Osman, Black Eyed Peas band member apl.de.ap, flute player Sir James Galway, Kenny Rogers and of course WCB director Owen Williams.

Secondly, even if you don't have nystagmus (also known as "wobbly eyes") I hope you find at least some of these articles relevant to you. One thing I've learnt is that we all face much the same challenges regardless of why we can't see very well or indeed at all. Most of us with a visual impairment have felt isolated or lonely at some point in our lives. Depending on when our visual impairment started, most of us faced difficulties in education and employment. Most of us wish we could drive or didn't have to give up driving. And most of us wonder 'Why me?'

Thirdly, I'd like to thank everyone who contributed to making this issue possible. We have brought together a unique collection of articles by clinical staff, researchers, people with nystagmus and the parents of children with nystagmus.

Finally, the next section provides a brief description of nystagmus as it affects individuals. Although lots of different words are used to describe nystagmus, the big question if you have it is how old you are when it starts. As a general rule, if nystagmus starts when you're very young you have

poor vision but a stable image most of the time. If nystagmus starts when you're an adult you may technically still have 6/6 vision, but you're more likely to see the world jumping around. And as anyone with late onset nystagmus will tell you, that can be very debilitating.

*John Sanders, executive manager Nystagmus Network,
Cardiff, September 2015*

A very quick guide to nystagmus

Nystagmus is an eye condition where the eyes move involuntarily. This nystagmus movement is often described as wobbling, flickering or fluttering. Since our eyes weren't designed to wobble involuntarily, nystagmus is not a good thing to have and it does result in visual impairment.

An estimated one in 1,000 people have nystagmus. How much nystagmus affects someone's vision varies a lot. It depends for instance on what causes the individual case of nystagmus and how old you are when it starts.

Infantile Nystagmus Syndrome (INS and also known as Congenital Nystagmus and Early Onset Nystagmus among other things) usually starts within the first six months of life and can affect vision in many ways. People with INS usually have vision that varies throughout the day, need more time to see, may have poor balance and depth perception and be light sensitive.

Distance vision is usually reduced in INS and the field of vision can be affected too. People with INS sometimes – especially when anxious or tired – experience the world moving. However, despite the eyes moving, they generally have a stable but poor image. INS is a lifelong condition and

there is currently no cure and only limited treatment options.

Acquired Nystagmus (AN or late onset nystagmus) typically starts in adulthood. People with AN often experience the world moving some or all of the time. They therefore find it difficult to walk, read a book, watch TV, etc. Again, their vision can vary throughout the day. An eye test may show their visual acuity to be unchanged - if given plenty of time to read letters from an eye chart. But their functional vision is considerably reduced.

INS is often caused by or associated with many eye conditions and syndromes. These include albinism, aniridia, Congenital Stationary Nightblindness (CSNB), optic nerve damage. Causes of AN include multiple sclerosis, stroke, head trauma and neurological disease.

The parent's tale: from envelopes to events

Carol Bashford was the very first person to call the NN (Nystagmus Network) helpline back in the spring of 1992. Carol, who lives in Swansea, is now a committee member and events organiser for the charity. Carol rang the helpline because, like many other parents, when the doctor said her daughter had nystagmus it was a long word she had never heard before and it meant absolutely nothing to her.

Much has happened in the last 23 years. That baby girl (Louise) is now 23, married, with a degree and two daughters of her own. On top of that Louise has competed very successfully at international level in disability swimming and athletics.

But it wasn't all plain sailing. Carol noticed early on that Louise's eyes seemed different but, being a new mum, didn't think much of it. The diagnosis of nystagmus came after a

routine six week check-up for Louise. That triggered a hectic series of hospital appointments and tests in and around London where Carol lived at the time - and much uncertainty for Carol and husband Bob.

A consultant ophthalmologist at Great Ormond Street Hospital finally confirmed that Louise could see and that she had nystagmus. Although the news that Louise could see came as a relief, Carol still had many questions: “To us nystagmus was a long word and we didn’t know what it meant. The health visitor handed us some literature and sent us on our way.”

Fortunately, one of the orthoptists in Great Ormond Street had given Carol the new helpline number for NN. When Carol got home, she rang the number and spoke to Geri Holloway, a retired social worker who has nystagmus herself and had volunteered to answer calls from her home.

Sunshine

Carol had never knowingly met anyone with nystagmus and there was no family history. She knew several blind colleagues in the bank where she worked as a PA. But listening to Geri talk about nystagmus Carol soon came to discover a whole new world of vision between 20/20 and total blindness. “Until then my world had been very bleak when it came to thinking about my daughter’s future. The sunshine came out when I spoke to Geri.

“I joined NN and had a lot of help from Geri. We also had a very good VI teacher.” Carol met other adults with nystagmus through NN and eventually responded to a letter asking for volunteers. She started by helping out in the office stuffing envelopes and doing photocopying, then moved on to looking after the charity’s database.

Carol also began talking to other parents of newly diagnosed children. That kind of peer group support is often one of the

most valuable things that a charity like NN can offer. And Carol, as the first parent to benefit from those phone conversations with Geri years earlier, knew that better than anyone. “NN had been helpful to me. It was great to meet professional people with nystagmus on the committee who had achieved things in life. And you’ve written loads of letters for Louise for school too. This was my way of giving something back.”

International organiser

By the time NN started planning for its first international research workshop in 2005, Carol was back in Swansea and – with her experience as a PA - a natural to get involved in organising an event 150 miles away near Oxford. Carol’s colleagues at NN were already well aware of her efficiency and flair for getting things done. So it came as no surprise when overseas delegates started addressing their emails “Dear Dr Carol”.

Carol has helped organise and run all three NN research workshops, a clinical training day and many of the charity’s annual open days. She laughs loudly at the thought of being addressed as Dr Carol, but (although she would deny it) she does know a lot about nystagmus.

That knowledge came in useful again in 2014 when Carol’s second grand-daughter was born with nystagmus. Everyone in the family knows nystagmus is not the end of the world – and even that there can be some positives from being visually impaired. They never have to queue in Disney and they benefit from discounts on cinema tickets for instance.

NN remains a big part of Carol’s life – she’s already planning ahead for the next research event. As a volunteer and committee member, Carol has made friends through the charity and would find it hard to imagine life without NN now. “It’s opened doors in schools. It’s also opened doors personally from being on the committee. As a member I learnt

a lot from the teachers' pack and from talking to people. As a family we've benefitted from NN. That's why I do so much."

Towards a Clinical Pathway for Nystagmus

Chris Harris, Royal Eye Infirmary, Plymouth; Julie Owen, Royal Eye Infirmary, Plymouth; & John Sanders, Nystagmus Network, UK

Why Do We Need a Clinical Pathway for Nystagmus?

People affected by nystagmus have three basic questions: what causes it, how will it affect them/their child and what can be done about it? The lucky ones leave the eye hospital with comprehensive answers to all three questions and a positive outlook. The unlucky ones come away with little or no information, confused and despondent, with the words "there's nothing we can do" ringing in their ears.

How many fall into the unlucky category is hard to say, but the steady flow of emails, phone calls and facebook posts to the Nystagmus Network suggests it's a lot, certainly hundreds every year. In fact, it may well be that the majority of people diagnosed do not get the basic support and information they need. As a result, they are at risk of performing poorly at school, not getting into employment, becoming depressed and isolated. Anecdotal evidence collected by NN UK suggests strongly that this is a worldwide problem.

There can clearly be a disconnection between what patients and their families expect, and what clinical teams deliver. Some centres provide exhaustive investigations, provide treatment where possible, and discuss the nystagmus with patients and their families at length. Others do not. The reason for these differences cannot just be blamed on time and money (although these are not insignificant issues).

There are other factors including training, experience, and attitudes towards nystagmus. Based on clinical experience and the experiences of NN, we have identified what we believe are the major underlying issues; both patients and clinicians need to be aware of these:

- 1) From a clinical perspective, nystagmus is a difficult subject, and no two patients are the same. There are many types of nystagmus that are difficult to distinguish. Very few centres have eye tracking facilities, and in any case, they require expertise that is generally not available. In spite of numerous textbooks discussing nystagmus (usually quite old), there is very little up-to-date guidance of how to approach the problem of identifying the different types of nystagmus. Moreover, most patients with nystagmus do not follow classic text-book pronunciations. Nystagmus is not common, so it takes many years to accumulate sufficient experience to be an expert in nystagmus. It is not surprising therefore, to find wide variations among centres and even between clinicians within a centre.
- 2) Overall, the causes and associations of nystagmus are wide, ranging from acute neurological disease to idiopathic nystagmus. The room for error is daunting, and most clinicians have a degree of trepidation when dealing with nystagmus. Rarely does the nystagmus pin-point the underlying condition, so the clinical priority is to find, or rule out, any associated condition. Thus, the nystagmus is seen as a clinical sign, and not as a clinical condition in itself.
- 3) Sometimes, no underlying cause can be found, and the label 'idiopathic nystagmus' is used (idiopathic = unknown cause), or 'isolated nystagmus'. In many medical conditions, an occasional case of idiopathy does not raise eyebrows. Indeed a few cases of acquired nystagmus in older patients remain unexplained. However, in infantile

nystagmus syndrome (INS) (formerly known as ‘congenital nystagmus’) a very high proportion end up being labelled as idiopathic! By definition idiopathy is a ‘diagnosis’ by exclusion, and depends on how hard one looks for a cause. It is well known that some underlying conditions (such as congenital stationary night blindness, ocular albinism, cone dysfunctions) are easily missed when using an ophthalmoscope. Many of these cases can be detected by electrophysiology (VEPs and ERGs), but this is not available (or not used) in many centres. Thus, what is ‘idiopathic’ in one centre is not in another. Even when electrophysiology is routinely used, about a third of INS cases are still idiopathic. Clearly, we are missing something important.

- 4) Failure to identify the underlying cause has consequences for both clinicians and patients alike. A) Diagnoses are incorrect B) It has become ‘acceptable’ to label a patient with idiopathic nystagmus. One cannot but wonder cynically whether such a high incidence would be acceptable if the nystagmus were a life-threatening condition.
- 5) Most patients will be unaware of the underlying differential diagnoses (as one would hope from a professional practice), and perhaps do not appreciate the relief of not finding some sinister cause. Nevertheless, their perceived priority is still the nystagmus itself. Nystagmus IS life-affecting, and even if there is no treatment, understanding what to expect and how to cope with nystagmus in real everyday life is their priority. A major source of complaint is that patients feel that their nystagmus has been dismissed and they are given little, if any, information.
- 6) What constitutes poor vision is a continuing source of confusion. In the clinic, how well one can see has become defined by measures of Visual Acuity (VA). It is assessed via a chart in bright light and the patient is given plenty of

time and encouragement to recognise the letters – a far cry from real life. The relationship between VA and nystagmus is complicated, but the crucial point is that VA does not take into account many of the problems confronting people with nystagmus including null regions and head postures, long visual response times ('time-to-see'), effects of stress, oscillopsia, difficulty seeing in fast moving and/or cluttered environments, poor depth perception and balance problems.

- 7) The focus on VA is also a continuing source of frustration for patients with idiopathic infantile nystagmus who are 'told' that they have normal or near-normal vision, when in fact, they have poor functional vision. It is particularly irksome for some families who are unable to get their child registered as sight impaired (CVI) because the ophthalmologist believes the child's VA is not sufficiently poor. Whereas, other ophthalmologists do recognise the broader visual impairment caused by nystagmus and register their patients regardless of VA. It is not uncommon for families to change ophthalmologist in order to get their child registered. Recently, McLean et al (2012) carried out a quality of life survey on patients with nystagmus (early-onset and acquired). They reported an overwhelmingly negative experience of living with nystagmus. They identified 6 domains with various subcategories, which we have reproduced verbatim in the Table.

8) Perhaps, the most significant problem is that clinical teams do not recognise that there is a problem! Whilst one would not expect hospital clinical teams to address all the psychological aspects of nystagmus, they should at least recognise the problems faced by people with nystagmus. At a recent NN nystagmus training day, attended by ophthalmologists, orthoptists, optometrists, and VI teachers, many of the above issues were discussed. At the end of the day, feedback revealed that many of delegates were unaware of the extent of the disconnection between the patient and the clinical team, and vowed to change their approach

We propose that if a clinical pathway were adopted across all centres, many of the issues would be reduced,

- **Visual function**
 - General visual deficit
 - Distances
 - Near
 - Faces
 - Focusing
 - Oscillopsia
 - Personal preening
 - Discomfort
- **Restriction of movement**
 - Driving
 - Education
 - Leisure
 - Occupation
 - Public transport
 - Relying on others
 - New environments
- **Standing out/not fitting in**
 - Being different
 - Comparisons to others
 - Others' failure to recognise
 - Others' perception
 - Eye contact
 - Visible aspects/cosmesis
 - Explaining to others
- **Feelings about inner self**
 - Confidence and self-esteem
 - Dwelling on problem
 - Guilt
 - Inferiority
 - Sadness/distress
- **Negatively regards future**
 - Giving up/not attempting
 - Abandonment/let down
 - Hopelessness
 - Isolation
- **Relationships**
 - Family
 - Friends
 - Education
 - New encounters

From McLean et al. (2012)

if not eliminated. A clinical pathway would be economical in the long run by actually saving the NHS money and time.

What is a Nystagmus Clinical Pathway?

A clinical pathway (CP) is a multi-disciplinary route by which patients with a common condition, such as nystagmus, follow a common path through various clinical investigations and interventions. The idea is to standardise healthcare so that all patients follow a similar journey through their medical care, regardless the actual hospital or medical specialists seen by any particular patient. This standardisation is supposed to optimise healthcare and to be evidence based.

There are a number of advantages for a NCP.

- Recognises nystagmus as a condition rather than as an incidental association
- Provides a minimum standard of care
- Provides a multi-disciplinary approach and is patient-centred.
- Is evidence-based
- Facilitates research since patients have had a consistent work-up

There is a danger that a nystagmus CP will itself become out of date, and inflexible. We propose that a nystagmus CP would need to be dynamic to take into account new information and procedures. Even today we can identify likely upcoming and potentially far-reaching research outcomes that will affect how we manage children (and adults) with nystagmus. We need “Placeholders” for these (see below).

We now discuss a proposed nystagmus CP.

1. Pathway Entrance

Patients of any age (infancy to adulthood) with nystagmus should enter the pathway. We recognise that nystagmus may not be the presenting sign and/or a patient may be under care for an associated condition. Nevertheless, their nystagmus is likely to be persistent, affect quality of life negatively, and also to be perceived by the patient as a major problem.

2. Identifying the Nystagmus

An explicit effort should be made to identify the nystagmus. This may be achieved with eye movement recording if available. At least an informed clinical examination is required. Identifying the nystagmus will narrow down underlying conditions and guide subsequent stages. If the nystagmus cannot be identified, then this should be stated explicitly. The current practice of repeatedly noting the existence of 'manifest nystagmus' should be avoided as this provides no information, and can in some instances lead to a false impression that the nystagmus has been identified.

3. Identifying and Managing the Underlying/associated Condition

Most types of nystagmus will fall into one of two broad categories:

- 1) Early-onset nystagmus (< 6 months), which includes infantile nystagmus syndrome (INS) (formerly congenital nystagmus) and latent nystagmus (LN). The majority of associations are sensory, and hence require ophthalmological investigations in the first instance. The 'diagnosis' of idiopathic nystagmus **cannot** be made without extensive investigations including electrophysiology (ERGs and VEPs).
- 2) Late-onset or 'acquired' (>6 months), and includes many types of nystagmus. The majority of cases have an underlying neurological cause (rather than sensory) and require neurological assessment.

These categories are only a guide. Some infants and young children will have 'acquired' nystagmus with an underlying neurological cause. Some adults will have a sensory cause (often undiagnosed INS or LN). So, an expedient mechanism for cross-referral between ophthalmology and neurology needs to be in place.

Nystagmus from both categories can be familial/genetic, and so an extensive and *recorded* family history is essential.

4. Managing the Nystagmus

In most cases, the nystagmus will not be 'cured' by managing the underlying condition. For example, cataract removal even in early infancy does not stop nystagmus.

In acquired nystagmus a variety of drug therapies have been described with variable success. Currently, the drug of choice depends on the type of nystagmus (such as downbeat, upbeat, acquired pendular, periodic alternating, and epileptic nystagmus). Diplopia can be managed by prismatic corrections

For INS and LN, there is no known treatment (in spite of many claims to the contrary). Spectacles and contact lenses should be worn to correct other eye problems, but will not correct the nystagmus.

We have been constantly surprised about the (apparent) benefits of a frank discussion about nystagmus with patient and family. For acquired nystagmus, adults are quite capable of understanding the basic neuroanatomy that gives rise to their nystagmus. They find the causal relationship between their underlying pathology, the nystagmus, and the visual consequences of the nystagmus (oscillopsia) strangely reassuring. For the developmental nystagmus (INS and LN), a brief discussion of developmental plasticity has a similar effect.

School-aged children should be referred to sensory support teachers, but it is important to explain that teachers are not familiar with nystagmus.

5. Pathway Exit

It is common practice to maintain follow-up visits for years. The reason for this is not clear. The nystagmus itself does not change much; it may decrease slightly in intensity over the first ten years but it does not worsen. Any progressive visual disorder or acute neurology should have been diagnosed, so there is no visual deterioration to expect. Refractive errors may change, but this in itself does not warrant eye clinic visits. Presumably, follow-up visits are an insurance plan in case something subsequently goes amiss. However, we are not aware of any evidence base to justify this *provided* the previous stages have been fulfilled.

The costs, on the other hand, are more tangible. In over-stretched clinics, filling NHS slots with follow-up visits is a substantial workload. From the patient's perspective, time off work and/or school and travelling to and from a clinic is not trivial for some. There is also the important issue that longitudinal visits unnecessarily maintain a high level of patient medicalization and unfulfilled expectations.

We propose that patients should be discharged from the pathway when there is no tangible benefit for continued follow-up. Indeed, having a pathway makes such discharge possible. Thus, when the nystagmus has been identified, underlying pathology has been positively identified, or all currently available investigations have been exhausted then it is feasible to discharge. However, it is crucial that

- a) Contact with the patient is maintained so that they can return at some later date if desired
- b) Full information is provided at discharge. As discussed above, lack of information is a major source of complaint.

- c) Refractive check-ups are available outside the eye clinic by opticians/optometrists who are familiar with nystagmus.

Place-Holder

In our view, a nystagmus CP needs to adapt to modern innovation and medical science. Two obvious upcoming place holders need to be reserved for OCT and genetics.

Over the last decade or so, it has become increasingly clear that OCT can provide clinically useful information about the morphology of the retina, and possible explanations for nystagmus (such as subtle foveal hypoplasia and optic nerve hypoplasia). Most eye clinics have OCT equipment already in use, so it is potentially useable today. Indeed some centres are already using OCT routinely for nystagmus. There are some issues such as different equipment and the problem of obtaining good images from patients whose eyes are not still (especially for the optic nerve head), but we expect these problems will be ironed out over time.

In many cases, the nystagmus is familial with a genetic origin. Even sporadic cases may reflect de novo mutations.

Currently, genetic testing is not routine, and costs need to be justified. However, the price of genetic testing continues to tumble dramatically with potentially huge ramifications. It seems highly likely that within the next decade the price will become so low that it would become difficult to justify **not** looking for nystagmus genes. Indeed, it seems feasible that testing for a bank of known nystagmus genes, or even sequencing a patient's whole genome will become the **primary** investigation – even before clinical/phenotypic investigations commence. However, while this would be very welcome for nystagmus patients, it should be recognised that such developments are likely (at least initially) to actually widen the differences among centres. A dynamic and up-to-date CP would hopefully smooth this transition.

Early-onset nystagmus is a sentence to a lifetime of visual impairment (in addition to any underlying sensory cause). Putative treatments have had minimal effect on vision (although they can reduce nystagmus intensity). One possibility is that this is because VA is an inappropriate measure of vision – it is the wrong outcome measure. There is now a strong move to look for ‘time-to-see’ measures, as these seem more sensitive to waveform manipulations. Whether these will be fruitful, and whether they can be translated into the clinic, remains to be seen. But if there is an evidence-base, then it should be adopted by a nystagmus CP in the future.

Conclusion

In our collective experience, the investigation of patients with nystagmus has changed little over the past 25 years. The charity Nystagmus Action Group (later to become Nystagmus Network) was formed in 1984 by patients and their families in response to a lack of information, wildly different experiences across centres, and a perceived lack of interest by clinicians. Judging from the flow of complaints in 2015 to Nystagmus Network, the disconnection between what clinical teams believe they provide, and what patients and families believe they receive has not gone away. We propose that a nystagmus clinical pathway would help close this gap by recognising that nystagmus is in itself a condition (not just a sign of an underlying pathology), by providing a consistent and minimal standard of care, and by emphasising the need to actually talk to patients and families about the impact nystagmus will have their lives.

School Survival

This article by Sophie Brown first appeared on her Living, Laughing & Loving with Wobbly Eyes blog on July 30, 2015

Starting a new school or school in general can be pretty nerve wracking for everyone; going into an unfamiliar environment without your parents, having to make friends with people you've never met before, and so on.

My parents were particularly concerned about how my nystagmus would affect both my education and socialising – would the other kids make fun of me? Would I be able to see the board okay?

After attending four schools, college and university; my personal answer is that nystagmus won't affect you if you don't let it!

Based on my own experience, here are some tips for school survival with wobbly eyes:

1 NEVER be ashamed!!!

Never ever EVER be ashamed, embarrassed or self-conscious of your nystagmus. When I was younger I believed that no one wanted to be my friend because I had wobbly eyes. I soon learnt that was NOT true – in fact, quite a few of my classmates thought it was cool!

2 Educate

You go to school to learn things, right? So **teach your classmates AND teachers about nystagmus** – none of my teachers had heard of it before

I've found that children and teenagers tend to **make fun of things they don't understand**

At the first primary school I went to, my mum and I held an assembly in front of all the staff and students. We explained about nystagmus – what it is, how it affects the individual etc.

Secondary schools can be a bit different – you can't fit the ENTIRE school in an assembly hall. Nevertheless the same tip applies, just go about it differently

Approach your form tutor and explain about nystagmus, ask them if you could talk to your form about it

If you don't like public speaking, mention it to people individually (I've used it as a conversation starter before!!!)

3 Use the NN's 'Yellow Card'

The Nystagmus Network have a card that informs teachers about the **effects of nystagmus and advice** to help students with nystagmus

I personally found this card more useful at secondary school because every teacher teaches 100+ students each day, whereas primary school teachers usually have a class of thirty odd

I often found that most of my secondary school teachers forgot that I was visually impaired, so they'd write on the whiteboard with green pen, or hand me a textbook to share. If this happens, and like me you don't want to make a scene, discreetly get their attention and show them the yellow card again

Get the Yellow Card here:

<http://www.nystagmusnet.org/cms/index.php/about-us/news/archive-2012/123-download-free-quick-guide-for-teachers>

4 Sit at the front

Always **opt for the front**. It's wayyyyyy better than constantly

squinting at the board from the back row – or worse, not being able to see at all!

If you're placed at the back, explain to the teacher that you can't see and they'll move you

Sometimes I was too afraid to ask because I was scared of the teacher – but they don't bite, I promise!!!!

5 Speak up

Same as tip 4, never be afraid to say if you're struggling with something!

6 Pre-visit the school

Ask your parents to **book an appointment** to visit the school with you before you start

This way you can map out any odd steps and judge the depth of the stairs

You'll hopefully feel more comfortable now you're not a complete stranger to the place, so when it comes to the school rush it won't be so bad!

7 Contact the schools disability support

Most schools now have some form of a disability support department. Ask your parents to contact them **prior** to you starting there

The extra support you may be entitled to from your **local educational authority** in the UK is sooooo worth it!

In primary school, I had a sloping desk so I wasn't craning my neck to see the text, magnifiers, and extra large print in my SAT papers

In secondary school I was even given binoculars for the first year so I could sit at the back – which I stopped using after a couple of weeks because it was less hassle just sitting at the

front

By year 10, my mum had fought my case to get me provided with a laptop and I was allowed to use one for my GCSEs – they forgot to turn the spell check off!!!

If you don't ask, you don't get!!!

You may be entitled to **extra time** for your exams

During my GCSEs, all us 'extra timers' were in a separate classroom rather than the school hall

Even though most of the time I found that I didn't need the extra time, I felt less pressurised and relaxed by being in a classroom

If I was in the school hall environment, my nystagmus would have increased due to stress = I'd get tired easily = I'd fall asleep during the exam!!!

8 Learn to 'touch-type'

Learning to touch type at primary school is the best thing I've ever done

I was pulled out of class once a week to practice

When I got my laptop in year 10 I was able to type as fast as the teacher was talking (**handy for taking notes**) **WHILST** the teacher was talking!

See Sophie's other blogs at

<https://nystagmusinanutshell.wordpress.com/>

What can optometrists do for people with nystagmus?

Dr Phil Jones PhD, BSc(hons), MCOptom

As you know, nystagmus is an extremely variable condition. Those who suffer with the condition have difficulties with many day to day tasks. Optometrists are well placed to provide eye care and advice for those with nystagmus.

Visual Acuity

Visual acuity (VA) shows a large variation in people with nystagmus, ranging from -0.3 to 2.0 LogMAR^{6,17}. Idiopathic nystagmus has a better average VA (0.35 LogMAR) than that associated with albinism and other visual abnormalities (0.67 and 0.55 LogMAR, respectively). In the case of albinism, this is because of the foveal hypoplasia.

The near normal VAs of better than 0.0 LogMAR obtained by some people with nystagmus is thought to be due to the presence of foveation periods in the nystagmus waveform. Foveation duration is also dependent on null position. Therefore, by association, so is VA. Patients with nystagmus often achieve their best VA when they are using their null position.

Environmental factors such as stress and anxiety have also been shown to affect nystagmus, resulting in shorter foveation. From this it would be expected that VA would suffer. However, recent research has show that VA is unaffected. The same research suggests that the time taken to see the stimulus is increased. This has implications for people with nystagmus in the real world for example, seeing bus numbers on a moving vehicle, or trying to find something or read something where time is restricted.

Driving and Nystagmus

The official DVLA standards for a standard driving licence state that drivers must be able to read a car number plate (made after 1st September 2001) at 20 metres. They must also have a VA of at least 6/12 with both eyes. As described above, some people with nystagmus may be able to achieve this level of vision. It should be remembered that nystagmus is variable dependant on the situation and so the time taken to see things changes. This could have implications for driving. Those with nystagmus that meet the vision standard should use their personal judgement as to whether they feel confident in their visual capabilities to drive.

Refractive Error

As with VA, there is a large variation in refractive error for those with nystagmus. The range of refractive error has been found to be between -16DS and +11DS. (The range of astigmatic errors between Plano and 6DC). The majority of astigmatism is "with the rule". A study investigating the development of astigmatic refractive error in the first 8 years of life showed that both the prevalence and size of astigmatic error increased with age. The higher prevalence of with the rule astigmatism in subjects with nystagmus is thought to be the result of the mechanical action of the lids on the cornea.

This wide variation shows that there is no systematic relationship between nystagmus and either hypermetropia or myopia. However, research has indicated that the shift away from the normative curve is indicative of poor emmetropization.

Optometric management of nystagmus

With any treatment of nystagmus, it is important to correct the refractive error. Once the appropriate refraction is in place, various approaches can be used to try to improve VA and visual capabilities. The main technique for helping those with nystagmus is to use prisms so that the null position can be utilised without having to use an abnormal head posture.

However, this is only practical for patients who have a null position which is close to the primary position. For those people who have a convergence null (nystagmus movements become less when looking at things up close), base out prisms can be put in the lenses of the glasses. This forces the person to converge and in essence reduces the movements of the eyes.

The use of contact lenses has been noted to be preferable to spectacles in improving VA for those with nystagmus. The research present on the use of contact lenses is largely contradictory. The largest study into the use of contact lenses shows no significant improvement of VA with soft or RGP contact lenses compared to spectacles. However, it cannot be ignored that some patients with nystagmus do report improvement when wearing contact lenses.

Presbyopia and Nystagmus

From the time we are born, the lens inside the eye begins to lose its flexibility. When we reach our mid 40s, this loss of flexibility begins to affect how we read. It is at this stage that reading spectacles become of use. There are many ways in which presbyopia can be managed with spectacles. People commonly use bifocal or varifocal lenses to give them the type of vision they had before presbyopia. However, these lenses have narrow areas for near vision and so may be difficult to use for those with nystagmus. In these situations it may be advisable to have separate pairs of spectacles for different tasks.

Another possibility for the correction of presbyopia is the use of multifocal contact lenses. This could provide a useful alternative to give simultaneous distance and near vision. However, it should be noted that these contact lenses still have a limited range of powers and so may not be suitable for all.

Conclusion

As can be seen above, there are many ways in which optometrists can help those with nystagmus. It is always advisable to discuss any possibilities with your optometrist to see if they can help.

We need to talk about Snellen

John Sanders explains why the problems for people with nystagmus go beyond visual acuity.

There is no doubt that it is useful to measure how far someone with Congenital Nystagmus (also known as Early Onset Nystagmus and Infantile Nystagmus among other things) can see. However, measuring visual acuity with a conventional eye test chart and scale like Snellen or LogMAR is only a starting point when it comes to complex conditions like nystagmus. Distance is not the only aspect of vision affected.

Here are just some of the ways that nystagmus can affect how and what people see:

1. Nystagmus varies throughout the day, so a single measure of visual acuity (VA) made under ideal conditions bears little resemblance to functional vision in everyday settings.
2. Response times are slower, so people with nystagmus need more time to see (www.ncbi.nlm.nih.gov/pubmed/24222308).
3. Many people with nystagmus have a null point, a direction of gaze where the eyes move least and vision is best. In all other gaze directions vision is worse. Therefore the field of vision is affected, although this may not show up on a conventional field test.

4. Nystagmus reduces depth perception so it's harder to see kerbs and steps when out and about.
5. Nystagmus can affect balance too, so people with nystagmus may appear clumsy or unsteady (especially with late onset or acquired nystagmus).
6. Cluttered, crowded, busy environments make it harder to see than a simple measure of visual acuity would suggest. As a result, most people with nystagmus struggle to scan a page of text for a particular word or to find one person in a large group.
7. People with nystagmus struggle to see and / or follow fast moving objects. Consequently they are at greater risk for instance in traffic than their VA would suggest.
8. People with nystagmus may have poor contrast sensitivity, so for instance anything printed in a similar shade or colour to the background will be hard to read.
9. Many with nystagmus are light sensitive (photophobic) – usually because of associated conditions such as albinism, achromatopsia and high myopia.
10. Although early onset nystagmus is not usually degenerative, normal changes to vision in middle age may have a greater - and often unrecognised - effect than on people with ordinary vision.

Social

In addition, nystagmus can and does have big social and emotional consequences. These occur broadly for two reasons and are often poorly recognised or understood.

Firstly, poor vision (as outlined above) means that people with nystagmus often miss out on everyday social cues, facial gestures and body language that people with ordinary vision

take for granted. As a result, those with nystagmus may appear socially awkward, appear to ignore instructions or even seem rude.

Secondly, adults with nystagmus are often aware that they look unusual due to their wobbling eyes, possible head turn or tilt and head nodding or shaking. Many find it hard to make or maintain eye contact – a crucial aspect of non-verbal communication. These factors in turn affect their self-confidence and ability to communicate. As a result, many report adverse effects on their education and employment opportunities and everyday relationships.

As a result, this combination of sensory, social and emotional effects can lead to a far greater adverse impact on the quality of life than a visual acuity measure suggests. For this reason, people with nystagmus should not be assessed or judged on visual acuity alone.

Nystagmus Surgery

By Maria Theodorou, consultant ophthalmologist at Moorfields Eye Hospital, London and NN medical adviser

Nystagmus is a constant oscillatory eye movement that has several possible causes. Sometimes the cause in a particular patient is not obvious. However, regardless of the cause, the nystagmus may vary in the gaze position (ie. depending on where the person is looking), or even when looking at something near (convergence). Essentially this means that the nystagmus movements can vary in frequency or size, and this results in what is known as a null point or zone, a position/area where the nystagmus is reduced and the vision better.

Not everyone with nystagmus has a null point, or uses their null point if they do have one. But if people with nystagmus

have null points/zones which significantly improve their vision, they may adopt a head position other than the normal straight-ahead position in order to utilise their null, thereby minimising the nystagmus and maximising vision. This is often done involuntarily (ie. without the patient being aware) to improve the vision. Null points/zones often become apparent by the age of 18 months of age. For children of school age who adopt an abnormal head posture to use their null point/zone, it is especially important that their teacher be made aware so that they can be seated in an appropriate position in the classroom.

Surgical intervention is not suitable for all people with nystagmus. A careful eye examination must be carried out before nystagmus surgery is even considered, particularly to correct refractive error (i.e. short/long sightedness and/or astigmatism) with spectacles or contact lenses. This may not only improve vision, but partly, or even completely, correct an underlying squint. It is also important to exclude a variable head turn with a variable null point before considering surgical intervention. Where a compensatory head position is present, it is often difficult to measure accurately, as this is often only adopted when maximum effort is made to read a test type in the clinic, and a full detailed assessment should be performed.

Types of surgery in nystagmus can be broadly subdivided into:

- squint surgery;
- surgery for an abnormal head postures;
- artificial divergence surgery;
- surgery to minimise the nystagmus.

Squint Surgery

Squint surgery is the most common type of surgery carried

out in people with nystagmus. This is a condition in which the eyes, rather than the head, deviate away from the straight-ahead position, and often occurs in people with / without nystagmus. Sometimes it is appropriate to carry out this surgery in early childhood to reduce the risk of amblyopia (lazy eye). It may have the additional effect of reducing the nystagmus.

Botulinum toxin therapy is an alternative to squint surgery and has the advantage of not requiring general anaesthesia, but does often need to be repeated at regular intervals. A small dose is injected into the muscle(s) to temporarily weaken it. It can be used as long term treatment or as pre-operative assessment (ie. to simulate the effects of surgery).

Surgery for abnormal head postures

Surgery for abnormal head postures aims to rotate the eyes toward the direction of the abnormal posture (commonly a face turn), and may be advised with large head postures, particularly when associated with neck discomfort/pain. However, it is generally agreed that the early response to surgery does wear off in some cases, with recurrence of the abnormal posture. The benefits of surgery are largely cosmetic, since best corrected acuity remains the same; however the null zone may become larger.

Botulinum toxin therapy may also be used an alternative as a long term management in older children/adults that are able to tolerate local anaesthesia, or as pre-operative assessment in children/adults to simulate the effects of surgery.

Artificial divergence surgery

In patients whose nystagmus significantly improves when looking at something near (convergence), artificial divergence surgery may help, but a trial of prisms and/or botulinum toxin therapy should be done beforehand to assess whether surgery is likely to be successful.

Surgery to minimise the nystagmus

Other types of surgery are used to minimise the nystagmus itself rather than to correct a squint, or move a null point, although these are not commonly carried out in the UK. This may involve moving the eye muscles back from their original insertions to weaken their effects and therefore reduce the nystagmus, or detaching then reattaching the muscles without actually moving them.

More recently the Sinskey procedure has gained a lot of attention in the US. In this procedure a larger amount (than is currently standard) of the horizontal muscles are removed so the eyes are unable to move as much. However, this procedure is not without risk and short/long term complications, so is not advocated in the UK.

The procedure

Surgery is usually carried out as a day-case procedure under a general anaesthetic. Small incisions are made into the conjunctiva (the transparent layer over the white surface of the eye) to access the muscles of the eye. The eye muscles are found a few millimetres from the edge of the coloured part of the eye, just underneath the conjunctiva. These muscles are then carefully detached (+/- part of the muscle removed depending on the type of surgery), and reattached at the desired position with stitches (often absorbable).

In adults and older children, eye muscle surgery may be carried out using an adjustable stitch technique in which the eyes are tied to an adjustable bow during surgery. This allows the surgeon to fine-tune the position of the eyes after the operation while the patient is awake. It may be used where there is a risk of post-operative double vision. Anaesthetic drops are used to numb the eye, allowing adjustment of the bow knot until the correct position is achieved.

Ideal age?

There is no lower/upper age limit to nystagmus surgery,

although ideally a child should be co-operative enough to perform a reasonably reliable examination pre-operatively. Certain types of squint operations need to be carried out in early childhood to reduce the risk of developing a lazy eye. However, surgery done for cosmetic reasons is often deferred until the child is older, as young children rarely complain of their appearance and there is no evidence that abnormal head posture produces significant long-term sequelae (complications).

Risks of Surgery

Eye muscle surgery, as with all types of operations, is not without risks. The eye will be red and slightly uncomfortable for up to a few weeks after the surgery, but most people are able to return to normal everyday activities within 2-3 days of their surgery. A combination of anti-inflammatory and antibiotic drops are administered to the eye for a few weeks after the surgery to help prevent infection and to reduce inflammation.

The most serious risks of eye muscle surgery are very rare. Perforation of the outer coat of the eye (sclera) occurs in 1-2% of operations. This does not usually lead to any long-term problems, but rarely it can result in an infection within the eyeball (endophthalmitis) or a retinal detachment, both of which require further treatment and in a worse-case scenario result in a permanent reduction in the vision of that eye. A muscle can become 'lost' if it is not tied securely to the outer coat of the eye; it can be very difficult to find it again and a second operation will be necessary. The risk of sight-threatening problems following eye muscle surgery is rare, probably in the region of 1 in 10-12,000.

In summary, squint surgery is the most common eye operation carried out in patients with nystagmus. The indications, procedure and risks are similar to those without nystagmus, the primary aim in childhood being to reduce the risk of a lazy eye. Other types of nystagmus surgery are less commonly performed in the UK.

Acquired nystagmus

Early onset (or congenital) nystagmus is often described as a Cinderella condition. Late onset nystagmus (more commonly known as Acquired Nystagmus or AN) is not just an orphan condition but also homeless. In medical terms it often falls between ophthalmology and neurology. In terms of numbers we have little idea how common AN is. And as a patient it can be difficult to get information about AN and its impact on everyday life.

AN can start at any time in life, but the diagnosis is generally understood to refer to nystagmus that develops in adulthood. Many things can cause AN including multiple sclerosis (MS), stroke, head injury and neurological diseases such as ataxia. In some cases the diagnosis is still idiopathic (no known cause).

AN causes the eyes to move involuntarily - horizontally, vertically and/or with a torsional element. People with AN typically experience oscillopsia (they see the world moving) in some cases all or most of the time, in other cases intermittently. As a result AN often results in balance problems and can severely restrict your ability to perform basic tasks such as walking, reading, watching TV or preparing a meal. As one caller to the NN helpline said:

“I am like a drunkard and zombie and often when I walk I can feel one foot clumping in front of the other. Shopping is difficult as in a supermarket one looks down the aisle and casts the eyes along the shelves, but I have to stand still and look straight ahead.”

Many (possibly most) people with AN have to give up work and driving. Many become isolated as they fear (or are unable) to leave home unaided. However, the impact of AN does vary widely. Some are able to carry on an active life,

albeit usually at a slower and more cautious pace.

In some cases AN is made worse by looking in certain directions, suggesting that as with CN, people with AN can have a null point or null zone. Unlike CN, visual acuity is generally not affected in clinical terms. In other words, given sufficient time to read down an eye test chart in ideal conditions, someone with AN would probably have the same visual acuity as before the onset of nystagmus. However, the VA score would give a very misleading indication of their functional vision, due largely to the debilitating impact of oscillopsia.

Drugs such as gabapentin, Baclofen and memantine can help reduce the impact of AN in some people. In others drugs bring little or no benefit, or any benefit is outweighed by side effects. Prisms are another option which can help in some cases. Glasses or contact lenses do not fix AN (just as with CN), but should be worn to correct any other eye problems. Botox (botulinum toxin) can offer some respite to some patients who have AN due to MS.

Surgery is not yet an option for treating or alleviating AN. However, two experimental procedures may hold out hope for some groups of AN patients in the future. A London based team is investigating the use of small medical magnets to slow unwanted AN eye movements. In the USA a Seattle based team is developing a Cochlear like implant which may help one small sub-group of AN patients.

Information

In the Nystagmus Network's experience, information is an often neglected but effective tool in the range of options for helping people with AN. Here are some typical comments from callers with AN to NN's helpline:

“Thank you so much for your reply! My nystagmus is acquired as I have only experienced it the past few years, I'll be 51

later this year. ... Thanks again, your response was very much unexpected and deeply appreciated.”

“This (conversation) has moved me on. I know how to go on to the next step.”

“The doctors look bored when I mention nystagmus. They seem to turn off. There was no mention of nystagmus in the notes to my GP. He thought my difficulty with walking was due to back problem, I had to tell him, no it’s because of the nystagmus.”

A common complaint from people with AN is that no-one understands how the condition affects them. Many go so far as to say that even close family and friends do not believe or recognise the problems they face as a result of oscillopsia. Simply talking to someone who does understand is a great relief. And receiving printed information makes them feel less isolated and helps them convince others of the reality and daily impact of AN.

Spreading the Wobbly word

For most people the first time they hear the word nystagmus is when someone they know is diagnosed with the condition. It may be because it’s a word that doesn’t trip off the tongue very easily. Or maybe it’s because affecting around 1 in 1,000 people is not enough for it to be well known, yet too many to be seen as rare.

One of the aims of Nystagmus Network is to **raise awareness of nystagmus**: They want health professionals to understand the consequences of having the condition so they can give helpful advice at the time of diagnosis. They are also helping people working in education to be aware of the way nystagmus affects learning.

For many years NN promoted itself and raised awareness of the condition through traditional methods: posters and leaflets in hospitals, sending news releases to the VI community when there was something important to say. All that changed in 2013 with the advent of **Wobbly Wednesday** – nystagmus awareness day. The **first Wednesday of November** is now embedded in the calendar as a day dedicated to telling the world what it's like to have wobbly eyes.

Jelly throwing

It doesn't matter what people do on Wobbly Wednesday, the important thing is that they are raising awareness. Using jelly is fast becoming a favourite way, especially at school where teachers actually volunteer to be targets in jelly throwing games and pupils sell pots of the wobbly stuff to friends at break-time. A more grown-up idea in 2014 was jelly laced with vodka.

The date is close to both Halloween and Bonfire Night so parties can be spooktacular! If the awareness campaign has a colour it is blue reflected in international support which saw Niagara Falls, the CN tower Toronto and our own Blackpool Tower all lit up blue especially for Wobbly Wednesday last year.

The nystagmus awareness campaign is supported by school and hospital staff. Fun things with jelly are augmented by children who have nystagmus themselves telling class-mates what it's like to have wobbly eyes. Eye clinic staff join the party with displays that show the work they do in the diagnosis of nystagmus and the valuable role they play in referring patients to Nystagmus Network.

Like any successful awareness day in the 21st century, social media has an important role in the Wobbly Wednesday strategy. The hashtag **#WobblyWednesday** helps supporters connect with each other on Twitter. The figures for 2014 are

impressive; through Facebook 54,318 were reached. A social media thunderclap saw 117,448 people simultaneously posting a message supporting Wobbly Wednesday. Interest in nystagmus on Wobbly Wednesday brought nearly four times as many people as usual to the NN website (967 and 250 respectively).

Speaking after Wobbly Wednesday 2014, Nystagmus Network chairman Richard Wilson, said, “Wobbly Wednesday is about raising awareness so that one day everybody will know that nystagmus is an eye condition. We particularly admire the courage that our younger supporters showed in explaining to their peers what it’s like to have nystagmus.”

The aim of Wobbly Wednesday is to raise awareness of nystagmus, However, the generosity of NN’s supporters meant that last year the charity was able to make an additional research award. A grant of £10,000 was given to Southampton University Hospital to develop a new way of assessing nystagmus. The new test will mean that eye health professionals can better understand the eye movements of babies and give more accurate advice to parents about treatment options.

It’s easy to be a part of Wobbly Wednesday (**4th November 2015**) and help raise awareness of nystagmus:

- Organise an event to spread awareness of nystagmus.
- Order a Wobbly Wednesday pack from info@nystagmusnet.org or call 0845 634 2630.
- Join the Wobbly Wednesday Facebook group; <https://www.facebook.com/groups/WobblyWednesday/>
- Download a free poster and activity sheet; <http://www.nystagmusnet.org/cms/index.php/component/phocadownload/category/17-wobbly-wednesday>

Newcastle and New Orleans

Support groups for people with nystagmus exist in several countries. The oldest is the UK Nystagmus Network <http://www.nystagmusnet.org/cms/> followed by the American Nystagmus Network (ANN) <http://nystagmus.org/new/index.php>. NN UK's aims are to provide support and information, encourage research and raise awareness.

One of the most effective ways to provide support is to bring people together so they can meet others in a similar situation. NN UK does this every year. In 2015 our annual Open Day was in Newcastle and next year it will be in Reading on Saturday May 7th. One adult who was one of around 170 people at this year's Newcastle Open Day in May commented:

“It felt so good being around people who have an understanding of what life is like for me, who have had similar experiences and know the impact. Being isolated has made things much worse for me than they needed to be, over the years.”

Among the speakers in Newcastle were actor Gerard McDermott who explained that having nystagmus, while presenting challenges, has never held him back in his acting career. Gerard trained at the Welsh College of Music and Drama in Cardiff where he soon became known to the staff because he had to use their photocopier to enlarge scripts.

US event

Black Eyed Peas band member apl.de.ap made a guest appearance at the American Nystagmus Network conference in New Orleans on August 1st. The singer, who has nystagmus himself, spoke about being teased as a youngster

and how his life slowly changed for the better. But nystagmus still affects him now. He checks out every stage before a performance to make sure he knows how to avoid any hazards when he's dancing.

Apl stayed the evening chatting to others with wobbly eyes and was a great inspiration to everyone - especially the younger delegates and parents of newly diagnosed children. He also met some of the clinicians in New Orleans for a nystagmus research workshop and even had an impromptu eye examination from a UK ophthalmologist.

Earlier in the conference several of the researchers gave an overview of their work and commented on how successful the workshop had been. One vision scientist told delegates how he still has to battle with some doctors who would rather believe something about nystagmus they read in a medical textbook than listen to what their patients have to say about living with the condition.

Others spoke about the fascinating challenges of studying nystagmus. For instance, if the eyes of people with congenital nystagmus move involuntarily all the time, how on earth is it that the brain generally gives people nystagmus a stable image? Yet why do people with acquired nystagmus mostly see the world moving around? Those are big questions to which we do not yet have answers and show how complex nystagmus is.

The Impact of Nystagmus on Quality of Life

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Quality of life is the perceived general well-being of an individual which includes the emotional, social and physical aspects of a person's life. Health related quality of life is the assessment of how a medical condition may, over time, affect an individual's quality of life. Impaired vision has a wide variety of consequences for quality of life as various eye conditions affect vision in different ways. In terms of nystagmus, studies have shown that the impact is significant with visual functioning scores in nystagmus being worse than other visual disease such as macular degeneration. In order to gain further insight into exactly how nystagmus impacts upon activities of daily living, the team at the University of Leicester conducted a study whereby adults with nystagmus were interviewed. Interview questions explored the impact, both good and bad, that nystagmus has on daily living. Those being interviewed had a range of characteristics such as age, gender and vision in order to capture as many different viewpoints as possible.

A number of areas of daily living were identified as being affected by nystagmus. The areas mentioned most frequently by the interviewees were vision tasks, driving, others noticing the visible aspects (eyes moving, head posture etc) and the failure of others to recognise what it is like to have nystagmus. Restriction of movement was also discussed by the majority of the interviewees. This included both the physical movement of getting from place to place and also social movement in terms of education, employment and

access to leisure activities. Issues with confidence, self-esteem, hopelessness and sadness were also highlighted. In most cases nystagmus leads to reduced vision and therefore it was expected that visual functioning would be a concern expressed during the interviews. Specifically distance and near vision was mentioned and also the ability to recognise faces and struggling in crowded places whereby images would blur into one. This reduced vision also ultimately means that the majority of people with nystagmus cannot drive which, in itself, was a problem but, not being able to drive, also had a knock on effect on life choices such as employment and location of home. It is often assumed that the reduced vision is the major concern of having nystagmus but these interviews showed that the impact on living is about much more than visual function. Another key area that was reported was the feeling of standing out/not fitting in. The cosmetic appearance of the nystagmus was of the utmost importance and the awareness that other people were noticing the eye moving. So much so that having nystagmus affects connecting with others including avoiding making eye contact, an action that is important for successful social interaction. This was a new finding that had not previously been identified and therefore when clinicians are considering treatment for nystagmus it is very probable that the cosmetic effects are underestimated.

Other people's failure to recognise what it is like to have nystagmus and misunderstanding/lack of knowledge about the condition was upsetting for many of those being interviewed. This lack of knowledge and understanding about nystagmus is likely to have implications for relationship building on both a personal and professional level. Studies have shown that a facial appearance that differs from the norm can have an effect on other people's opinions of social functioning such as employability, honesty, trustworthiness and effectiveness. In strabismus (a turn in the eye), another visual condition that affects the appearance of the eyes,

relationship building has been reported to be affected with a negative impact on the ability to obtain employment and the capability of finding a partner. As the occurrence of nystagmus in the population is far lower than that of strabismus, it is fair to assume that the awareness of nystagmus as a condition is minimal. This lack of awareness about nystagmus will most likely lead to a negative perception of individuals.

Throughout the interviews there was an overlying theme that was mentioned by each and every one of the people being interviewed and this was that nystagmus affects every aspect of everyday life. In addition almost half of the interviewees wept whilst being interviewed. Sadness and distress because of nystagmus were apparent themes throughout, along with feelings of hopelessness and abandonment (particularly by the medical community).

All that being said, the interviewees also had positive comments to make. Relationships with others could be stronger because of nystagmus with feelings of acceptance by others and, support given in order to help perform daily tasks, being acknowledged. Coping strategies to work around difficulties with things such as travelling to places, daily tasks such as shopping and meeting new people were also emphasized. And, perhaps most importantly, hope for the future in terms of educating others and generating awareness of nystagmus as a condition in order to improve understanding. Further optimism was expressed with regards to treatment possibilities that may help to reduce the movement and possibly improve vision.

This study has uncovered a number of areas that are affected by nystagmus such as cosmetic affects, social restrictions and psychological distress that had not been previously considered when describing the impact of nystagmus. The effect of nystagmus on quality of life can be vast and spans

across not only the visually functioning domains of life but also the social and psychological domains. The content of the interviews show that the impact of nystagmus is far more complex than was previously thought. In the words of one of the interviewees:

‘Take nystagmus seriously. It may only be a few dozen thousand of us who’ve got it and so on but we are severely affected by it. Return on investment could be remarkable because there are so many of us who could do so much more.’

The themes that were drawn from the interviews are now being used to produce a disease specific quality of life questionnaire for use in nystagmus in order to accurately measure the impact of nystagmus and to use as a potential outcome measure for treatment trials. For further information please contact Rebecca McLean at rjm19@le.ac.uk.

Infantile Nystagmus Research in Wales

Jonathan T. Erichsen, Matt J. Dunn & Lee McIlreavy, Cardiff University Research Unit for Nystagmus [RUN]

The Research Unit for Nystagmus [RUN] was established in the School of Optometry and Vision Sciences at Cardiff University over 10 years ago. Today, RUN remains the only such centre in Wales and has become one of the leading nystagmus research groups in the UK and indeed the world. Our purpose, then and now, was to determine the underlying causes of infantile nystagmus and help develop treatments for the condition. Until that longer-term goal is achieved, we remain dedicated to investigating the mechanisms that produce the continuous oscillation of the eyes and to arriving

at a better understanding of the impact of these eye movements on vision and the daily lives of those affected.

The key to success in our studies is to be able to measure, in real time, the eye movements in a variety of situations. Fortunately, the technology of eye tracking systems has improved dramatically over the last decade, allowing us to record accurately and non-invasively the nystagmus oscillations as well as other eye movements made in response to different visual scenes or stimuli. This enables us to investigate the interaction between moment-to-moment eye movements and visual perception. Thanks to the generous support of a number of charities, including the Nystagmus Network and Fight for Sight, we have three state-of-the-art eye tracking laboratories. In addition, we use a variety of large display screens, one of which is capable of three-dimensional (stereo) projection, allowing us to investigate how the eyes work together.

There is another very important kind of support that we receive, and that is our large cohort of over 90 people with nystagmus (both in Wales and the rest of the UK) who generously volunteer their time by travelling to Cardiff and participating in our studies. Our success in discovering more about nystagmus would simply not be possible without their help and enthusiasm. Again, the Nystagmus Network has been instrumental in advertising our studies and facilitating the recruitment of our volunteers.

So, what have we been up to and what have we found out about the impact of nystagmus eye movements on perception? As one example, consider the fact that judging whether or not an object is moving in the world requires the brain to compare what the eyes are seeing with how the eyes are moving. Any inaccuracy in these judgements should produce the powerful sensation that the world is moving, a symptom known as *oscillopsia*. Yet, despite their constant

involuntary to-and-fro eye movements, people with infantile nystagmus generally do not experience oscillopsia but report that the world appears stable. Quite how people with infantile nystagmus achieve this remarkable compensation remains unknown, and is just one of our ongoing investigations.

As another example, we already know that the 'intensity' of the nystagmus, which can involve both the size of the eye oscillations and how frequently they occur, varies considerably in people depending on where they are looking (e.g. the 'null zone') or on their emotional state (e.g. stress level). One might reasonably think that, if the eye movements are faster, this should make someone's vision worse. Indeed, many people with nystagmus tell us this. The obvious, conventional way of testing someone's vision is to measure the finest detail that can be seen. This is what an Optometrist does routinely by using an eye chart with letters that get smaller as you move down the chart. The smallest letters that can be seen define what is known as *visual acuity*. However, we have completed and published a number of studies clearly showing that, in any given individual, visual acuity is largely unaffected by even quite large changes in his or her nystagmus. Whether the changes are due to stress or moving the eyes away from the null zone, the result is still the same. Indeed, we have even used a briefly flashed stimulus to remove the effect of the eye movements altogether, and yet, visual acuity remains unchanged. Other nystagmus researchers around the world have confirmed our findings.

Aside from this being a rather surprising result, what are the implications for people with nystagmus? Well, first of all, the currently available treatments (involving surgery and/or medication) are all designed to slow the nystagmus oscillations in the belief that vision will be improved. However, our findings suggest that any slowing of the movements is unlikely to produce much improvement in visual acuity. Nonetheless, people with nystagmus do sometimes report that they can see

better and most will choose to look in a particular direction (i.e. use their null zone) to slow their eye movements, even if this means they need to adopt an unusual head posture. All of this suggests that their eye movements do affect some aspect of their vision, but what is it?

Most recently, we have been investigating whether nystagmus can have an effect on how long it takes to see something. Our most recent study indicates that people with nystagmus do not take longer to mentally process visual information. However, we have also found evidence that, due to their eye movements, people with nystagmus may need to look at things for longer than others to achieve their 'best' level of vision (i.e. their maximum visual acuity). How long it takes to find and/or discriminate objects or even people in different settings may have a profound impact on peoples' everyday lives, and so, we are now developing new tests that might better reflect how vision is affected by changes in nystagmus eye movements.

Another important issue with which we are concerned is how best to ensure that recently available eye tracking technology can become more widely available, especially to eye health professionals in the clinic. For one thing, proper diagnosis of infantile nystagmus sometimes requires a high quality recording of the eye movements to look for the characteristic oscillation pattern. For another, if we are to adequately assess the impact of treatments now and in the future, eye movement recordings will provide a rapid and precise quantitative record of any effect on a person's nystagmus. Ideally, we are also aiming to provide an integrated solution that includes both improved diagnosis and more suitable measurements of how, if at all, vision is improved as a result of changes to nystagmus eye movements.

The Nystagmus Network

Who We Are

The Nystagmus Network (NN) is a UK charity run by individuals and families affected by the eye condition nystagmus. We have three part-time members of staff - Development Manager John Sanders, Information Officer Kerry Pleasant and Sue Ricketts. They are supported by volunteers and committee members. The committee chair is Richard Wilson and our president is Vivien Jones.



John, who has nystagmus himself, is the main point of contact for NN's email support and phone helpline. If you want someone to do some training or give a talk about nystagmus, it will probably be John who will come along, although other speakers are available too.



Kerry, whose son Oliver has nystagmus, is the main point of contact for our online shop and membership. You can contact Kerry at kerry.pleasant@nystagmusnet.org.

Sue joined NN as development officer in August this year. Sue has a grown-up daughter with nystagmus and was previously a trustee and parent adviser.

We are always looking for volunteers to help us do more and keep NN vibrant. Contact John via john.sanders@nystagmusnet.org to find out more.

Helpline service:

Email: info@nystagmusnet.org

Telephone: 029 2045 4242 or 0845 634 2630 (whichever number is cheaper for you).

Wobbly Wednesday



Nystagmus Awareness Day

4th November 2015

What is nystagmus?

Nystagmus is a complex condition where the eyes move involuntarily. This makes it harder to see. Few people with nystagmus drive and most face difficulties in everyday life.

Who gets nystagmus?

Every year hundreds of children are born with nystagmus, often with no known family history. Adults can develop nystagmus too, due to accidents or illnesses like stroke or MS.

What does the Nystagmus Network (NN) do?

NN funds research into nystagmus, provides information and support, raises awareness and educates people about the daily impact of nystagmus.

www.nystagmusnet.org

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The Nystagmus Network is a UK registered charity (803448)