1
What is blindness?

'So would you say I'm blind?' Sam asked.

This sounds like a straightforward question. Sam had been attending my low vision clinic for six years, so surely I would be able to answer her without a second thought?

Sam had an eye condition called Stargardt disease, which was slowly causing the cells in the central part of her retina to stop working. Just after starting high school when she was 11, she had found it difficult to see the whiteboard in some of her classrooms. Assuming she needed an eye test, her parents took her to a local optometrist who prescribed spectacles, but they didn't seem to make much difference to her sight. Her family realised there was something seriously wrong when Sam asked for the ketchup bottle to be passed, not seeing that it was right in front of her. A trip to her doctor led to a referral to an eye hospital, blood tests, scans, photographs and the unwelcome news that she had a serious, inherited and generally untreatable eye disease.

Sam remembers the news being broken: 'The consultant just said "there's not a lot we can do",' she told me. 'I felt a bit like he was washing his hands of me, although I'm really pleased he referred me to this place.' 'This place' was the low vision clinic we were sitting in, buried away at the back of the hospital. At her previous visits to the clinic I had prescribed Sam strong reading spectacles, given her various magnifying glasses and shown her how to set up her iPhone to make it easier to see. I'd spoken to her specialist teacher for visual impairment to make sure she had a relay system for the whiteboard at school, and had given her details for a group so she could meet other teenagers with sight loss. Since her first visit, Sam had changed from being a shy and slightly awkward girl to a rebellious teenager (an appearance not helped by the way that her vision

loss made it difficult to maintain eye contact), then a funny and engaging young adult. Now she had green hair and wore Doc Martens, a denim jacket and a 'Meat is Murder' T-shirt.

Sam's question about whether I would call her 'blind' may have been spurred on by the fact that her vision had clearly got worse. For the first time, she could no longer make out the letters on the top row of my sight chart, four metres away from her. When I wheeled the chart closer to Sam she could read the first few letters by moving her eyes around, sliding the blind area in the central part of her vision away from what she was looking at and using her peripheral retina to just about see.

Sam had told me that she'd got the grades she wanted in her A Levels and that she was very excited about moving to Leeds to study politics. She'd told me that her football team had won a tournament that summer and that she'd started a band with some of her college friends. She could travel independently, using apps on her phone to help when she couldn't see a bus number or platform sign. Her vision was too poor to have a driving licence, but she could cycle to band rehearsals. She'd had enlarged print and additional time in her exams, but she didn't read braille or have a guide dog. Would this active and successful teenager meet most people's idea of a blind person? The poet Stephen Kuusisto talks about people entering 'the planet of the blind',[1] but would Sam be welcome there?

The word 'blind' is emotionally charged and tends to be avoided by people working in eye clinics. In the UK, being 'registered blind' was replaced in the early 2000s with 'having a certificate of vision impairment'. In ophthalmology research, we even speak about 'double masked trials' rather than the 'double blind' studies used in other areas of medicine.

This coyness around the word 'blind' isn't universal. Many of the major sight loss charities use the word in their names, such as the UK's Royal National Institute of Blind People, the National Association for the Blind in India and New Zealand's Blind and Low Vision NZ. In 2011, the London-based Metropolitan Society for the Blind renamed themselves 'Blind Aid', which almost feels like reclaiming the word for people with vision impairment.

Writer Georgina Kleege uses the word 'blind' as she dislikes the alternatives: 'The word "impairment" implies impermanence … but my condition has no cure or treatment … I crave the simplicity of a single, unmodified adjective. Blind. Perhaps I could speak in relative terms, say I am blinder than some, less blind than others.'[2] Kleege has only come to embrace the word 'blind' after several decades of low vision. She writes

that as a teenager 'the most I would admit to was a "problem with my eyes", sometimes adding, "and they won't give me glasses", indicating that it was not me but the wilfully obstructionist medical establishment which was to blame for my failure to see as I should'.

When Sam asked if she was blind, I heard her father gently whistle at the gravity of the question. The room was so small that I thought I could feel his breath on the back of my neck. The background hum of the busy clinic around me seemed to drop, as if everyone was waiting for my response. Even as I spoke, I knew my answer was a cop out:

'The word "blind" means something different to nearly everyone I meet,' I told her. 'We prefer to say "severely sight impaired". It's true that if you were in America you'd be called "legally blind", but you're certainly someone who uses your eyes for most things, so I don't think "blind" would be the best word to describe you.'

'Legally blind,' Sam said, almost under her breath. I thought she was going to comment on this dramatic label, but she surprised me instead. 'We've been looking for a name for our band, and I think that might be it!'

How blind is blind?

Most people probably would not see Sam and think 'There goes a blind woman,' which raises the question: how well can someone see but still be classified as 'blind'? Would anyone whose vision is too poor to drive qualify as blind? What about someone with advanced tunnel vision, who can see small details but only in one tiny pinpoint of the world? What if someone is so sensitive to light that they can't leave the house in the daytime, even when wearing the darkest sunglasses? Or should the word 'blind' be reserved for people who don't see any light at all, who can't say whether a room light is switched on or off?

When I think about how well someone sees, I think about two factors: the smallest size of object they can see (their visual acuity) and how far they can see around them (their visual field).

Visual acuity

Visual acuity is usually measured with a Snellen chart: the common test seen in high street opticians' practices, doctor's surgeries and eye clinics – so familiar that it's even used as a shorthand for an eye examination in cartoons. The largest letter at the top of the chart is

10 times bigger than the standard of 'good vision', so somebody who can only see the top letter has a visual acuity about 10 times poorer than someone without eye disease. The letters towards the bottom of the chart are given the size '6', which means they can be read by someone with good sight from six metres. Someone reading these letters has a visual acuity of 6/6 in the UK, or 20/20 in the USA, where their dislike of the metric system means the chart is placed at 20 feet rather than six metres. Another person who can only see the top letter has a visual acuity of 6/60, or 20/200, meaning they can see from six metres objects of the same size that we would expect to be seen from 60 metres. Put another way, things need to be 10 times larger, or they need to be 10 times closer, for them to see them as well as someone with good sight.

Since the Snellen chart was invented in 1862 it has been used all over the world, with versions using pictures, numbers, Cyrillic, Japanese and Arabic print. The image below (Figure 1.1) shows a sight chart being used outside an eye clinic in Tanzania, with a 'tumbling E' being used for people who are unable to read: the person taking the test indicates the direction of each letter E by holding their hand in the corresponding direction.

Of course, some people can't see the top of the chart even when they're right in front of it. These people may be able to see movement,

Figure 1.1 A 'tumbling-E' Snellen chart being used to test vision in Tanzania. H Kuper. Attribution-NonCommercial 4.0 International (CC BY-NC 4.0). Image from: https://wellcomecollection.org/works/hzyjwfpq.

colours or lights and this vision can still be useful. The difference between being able to see daylight through a window and not seeing anything at all can have a huge impact on quality of life. Only a small proportion of people described as blind have no light perception at all, and even these people may sense light in different ways.

In the low vision clinic, we don't use Snellen charts, as they are not the most accurate way to measure visual acuity in people with vision impairment. There are very few letters towards the top of the chart with big gaps between the letter sizes, so quite large changes in vision cannot always be detected. The larger letters are shown on their own rather than as a row of characters, which makes them easier to identify, and it's quite easy to accidentally memorise the first few letters on the chart. Instead we tend to use logarithmic letter charts, which can be shown at any distance and easily moved to an appropriate place for the person to read them.

Sam read the top line of the chart when I wheeled it to half of its normal distance, meaning she had a visual acuity of 3/60, about 20 times poorer than mine. Would this mean she should be called 'blind'? The World Health Organization curates the International Classification of Diseases system, now in its eleventh iteration (the ICD-11).[3] This classification usually makes the news when new diseases are added to it, such as when hoarding disorder was added to the database, under the disease code 6B24. The ICD-11 classifies vision impairment in terms of visual acuity, ranging from 'no visual impairment' for people who see better than 6/12 (that is, anyone whose vision is good enough to allow them to drive, in most countries) through to 'blindness'. Sam's level of vision does – just – put her into the 'blind' category, so strictly speaking she would meet this definition. Table 1.1 shows the ICD-11 classification for different levels of visual acuity along with the threshold to receive a certificate of vision impairment in the UK, as sight impaired (which used to be called 'partially sighted') and severely sight impaired (formerly 'blind').[4]

As the table shows, there are people who have mild or moderate vision impairment but do not meet the threshold for registration as sight impaired. This is not unique to the UK, as the threshold for 'legal blindness' does not vary considerably around the world, with most countries using the same criterion as 'sight impairment' in the UK (although in India the level is equivalent to the British 'severely sight impaired' level).[5] Someone with moderate vision impairment normally would not be allowed to drive, may well use magnifiers to read and may need mobility training to navigate safely, but would not receive

Table 1.1 Levels of vision impairment defined by the ICD-11[1] and corresponding levels of UK certification of vision impairment.[2]

Level of vision	ICD-11 definition	Certificate of visual impairment
Better than 6/12	No vision impairment	Not eligible
6/12 to 6/18 *2–3 times poorer than good vision*	Mild vision impairment	Not eligible
6/18 to 6/60 *3–10 times poorer than good vision*	Moderate vision impairment	Not eligible
6/60 to 3/60 *10–20 times poorer than good vision*	Severe vision impairment	Sight impairment
Worse than 3/60, including light perception and no light perception; OR visual field of $<10^{o}$ radius	Blindness	Severe sight impairment

[1] World Health Organization. ICD-11: International Classification of Diseases (11th Revision). 2019. http://who.int.

[2] Department of Health. Certificate of Vision Impairment: Explanatory Notes for Consultant Ophthalmologists and Hospital Eye Clinic Staff in England, 2017. https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/637590/CVI_guidance.pdf.

the help and protection associated with legal registration. This group of people is sometimes said to have 'low vision' and they may struggle to communicate their vision loss to their teachers, employers and friends, partly as they don't have the shorthand of being able to say 'I'm registered as sight impaired' or 'I'm legally blind'.

Like many people with vision impairment who don't wear spectacles, Sam sometimes gets asked: 'Can't you get glasses to help you see better?' Leaving aside the ignorance of this question (do people really think that she wouldn't have thought of that?), it's important to note that for these purposes vision is measured with glasses on and with both eyes open. For the same reason, the people who try and empathise with Sam by saying 'I'm blind without my glasses' miss the point, as if they can see well with their glasses on and they have access to the correct spectacles (sadly not the case in much of the world, as we shall see later), they are not vision impaired.

Visual field

The other measurement I consider when I'm assessing how well someone sees is their visual field, which is how much of the world they can see at once, without moving their eyes. You can check this

yourself at home by staring at something straight ahead (something small, like a light switch, is ideal), holding your left arm out as if you're signalling to turn left on a bicycle, pushing your arm back as far as it will go, wiggling your fingers, then moving your arm forward slowly. You should first see your fingers moving when your arm is about level with your shoulder.

To quantify this measurement, you can think about the angle between the direction you are looking and the angle of your arm (Figure 1.2). This angle is about 90 degrees, so your visual field extends around 90 degrees to that side of your eye. If you do something similar with your arm in front of you, you can measure the vertical extent of your visual field. Someone with a full visual field can probably see about 80 degrees down without moving their eyes, which is useful for detecting obstacles and steps. The visual field does not extend quite so far upwards, usually reaching only about 70 degrees, a fact lamented by anyone who has banged their head on a low doorframe or cycled into an overhanging branch.

Measuring visual fields precisely is more difficult than asking someone to hold their arms out and wave them around as if they are directing traffic. You may have noticed it was difficult to keep your eyes still when you were doing this test, as it is natural to try and peek towards where your hand is. Modern visual field tests monitor the position of the eyes. These machines look like large bowls which the patient puts their head into, like a miniature planetarium. A spot of light will flash at various places around the bowl and the patient presses a button when the light is seen. It can be a tedious test for patient and practitioner alike. Kurt Vonnegut's *Slaughterhouse Five*[6] includes a scene where an

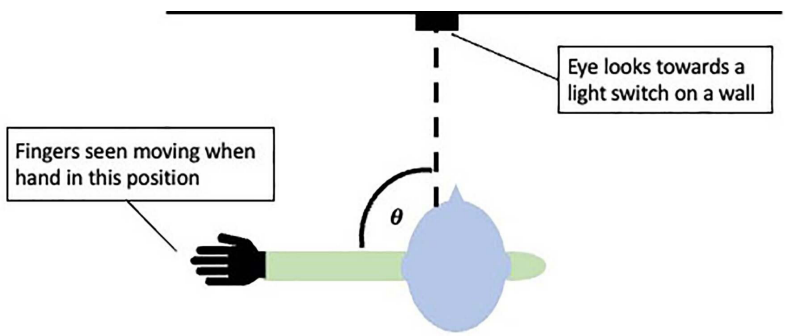

Figure 1.2 A top-down view of a way to approximately measure the visual field. Angle θ shows the extent of the visual field to the left. Image drawn by the author.

optometrist falls asleep in the middle of an eye examination (the only example of an optometrist hero in literature that I am aware of). I would be willing to bet this scenario happened when the lights were dimmed for a visual field test.

Visual field loss results from many eye diseases and can happen independently of visual acuity loss. It's not uncommon for me to meet people in the clinic who can read letters on the bottom line of the chart but who have a visual field of only a couple of degrees, as if they are looking at the world through a drinking straw. It may take them several seconds to find where the vision chart is, even in an uncluttered room, and they may need to be guided to the chair. If I ask them to look at my nose, they can't see my eyes or mouth without moving their eyes around and they tell me they are frequently surprised by someone suddenly appearing in front of them. They are likely to struggle with mobility and may well use a white cane or have a guide dog. People with severe visual field loss meet the image that most people have of 'a blind person' and the ICD-11 recognises this by classifying anyone with a visual field of less than 10 degrees as 'blind'.

The most common cause of visual field loss is glaucoma, a group of diseases where the eye's nerve fibres stop functioning, often because the pressure in the eye is too high. In glaucoma people tend to lose their peripheral vision over a period of years. This gradually increasing tunnel vision is very difficult for the person to notice, but once visual field is lost in glaucoma it normally cannot be recovered. This is one of the main reasons why it's important for people to have their eyes examined regularly, so the visual field can be checked to screen for glaucoma, the so-called 'thief of sight'. Glaucoma is more common in older people and in some populations, such as Black African people. Once diagnosed, the progression of visual field loss can usually be slowed or stopped with an operation or by using eye drops.

Tunnel vision is not the only type of field loss. Sam's eye condition affected her central retina so, although she could see well to the sides, she had a blind spot right in the middle of her vision, blocking her view of anything she looked at directly. Age-related macular disease causes a similar central field loss, whereas stroke and brain injury can cause hemianopia, which is an absence of exactly half of the visual field.

Although visual acuity and visual field are the most significant measures of vision, there are other factors that are relevant when thinking about how well someone sees. The ability to recognise colours and to see well under different light levels is also important. Contrast sensitivity is another key part of how well someone sees. People with reduced contrast

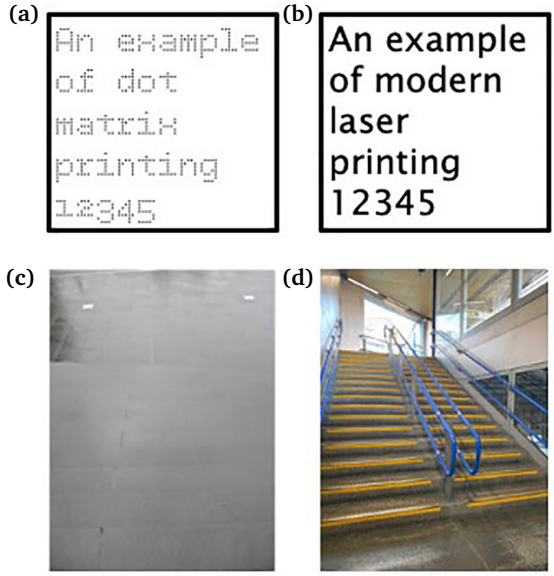

Figure 1.3 An example of low-contrast (left column) and high-contrast (right column) tasks. Images drawn/photographed by the author.

sensitivity can see black on white objects quite well but struggle with grey print on a grey background. These people often tell me that many websites are designed with 'young eyes' in mind, with thin grey writing against an off-white background. When I teach trainees about contrast sensitivity, I have stopped talking about dot matrix printers (they smile and nod at me like I did as a child when an older neighbour talked to me about the Second World War), but these printouts were particularly troublesome for people with poor contrast (Figure 1.3a and 1.3b).

Not being able to see subtle changes in contrast is also important for mobility. Identifying the edge of a concrete step in the rain on a grey day can be difficult even for people with perfect sight, but can be impossible for people with reduced contrast sensitivity (Figure 1.3c). This is why steps should have high-contrast markers at their edges, as found on the London Underground (Figure 1.3d).

There are other functional ways to describe vision loss. In 1920, the UK's Blind Persons Act used an employment-based definition, stating that people were blind if they were 'so blind as to be unable to perform any work for which eyesight is essential'.[7] A functional definition of low vision is to be 'unable to read a newspaper with conventional glasses'.[8] Almost every organisation seems to have its own preferred definition, each of which includes a different proportion of the world's population.

What causes blindness?

Only a small proportion of people are born with no sight at all, usually because the visual parts of the brain are severely damaged, or because they are born without fully developed eyes. Others are born with inherited conditions such as albinism, where vision is reduced but stable throughout life. Some, like Sam, start noticing poor vision in their teens, but the overwhelming majority of people with vision impairment lose their sight later in life.

More than half of the people with significant vision impairment in most high-income countries have age-related macular degeneration (AMD). The macula is the central part of the retina, which is the most sensitive to detail and which is used to look straight at something. In the most common 'dry' form of AMD, cells in the macula gradually die, reducing central vision over many years. In contrast, in the neovascular or 'wet' type of age-related macular degeneration, new blood vessels quickly grow into the macula and leak, leading to a sudden drop in vision. For about 10 years, wet AMD has been treatable with drugs injected into the eye, stopping the growth of these blood vessels and causing them to regress. Dry macular degeneration remains untreatable, although it may be slowed by lifestyle changes such as stopping smoking and by adopting a Mediterranean diet of vegetables, fruits, legumes, cereals, fish and moderate amounts of alcohol, while also limiting meat and dairy consumption. Age-related macular disease is remarkably common in older adults. One in eight people over 80 years old in the UK have lost vision due to AMD.[9]

Glaucoma is the second most common cause of severe sight impairment in Europe and is responsible for many cases of blindness in the Global South and middle-income countries, largely because of lack of access to treatment. However, the biggest cause of severe vision loss worldwide is cataract, where the lens within the eye becomes opaque. Usually associated with age (a former colleague used to tell his patients that everyone with grey hair has a bit of cataract), in high-income countries cataracts are treated with a routine 20-minute operation. Despite the relative simplicity of this procedure, it still requires a clean operating theatre, implantable lenses and a skilled surgeon. There are about 3,000 ophthalmologists in the UK for a population of 65 million, but in Somalia there is only one ophthalmologist for every 2.5 million people.[10] This inequality of access to surgery is a major reason why more than 10 million people around the world are blind from cataracts.[11]

Even more shocking is the number of people who simply don't have spectacles. More than 100 million people are visually impaired because they do not have access to the correct glasses.[12] My smartphone tells me that I can get spectacles made in more than 20 different opticians' practices within a mile of my office in central London. I can be confident that they will be prescribed correctly and, if I had low income, the NHS would pay for the eye examination and glasses. In much of the Global South, it is impossible to access any eye care at all, and even the US$3 cost of a cheap pair of glasses is prohibitive to many.[13]

Optometrists, ophthalmologists and others

I have already mentioned optometrists and ophthalmologists so it might be useful here to define the many different people who work in the eye clinic, nearly all of whom have a job title which starts with the letter O.

I am an optometrist, which literally means an 'eye-measurer'. The bulk of my optometry degree was spent studying the physics and biology of vision, alongside some clinical skills teaching. Most of my peers from university went on to work in high-street optometry practices, measuring vision, prescribing glasses and contact lenses, detecting serious eye disease and treating minor eye conditions. I didn't follow them into primary care, instead finishing my training in an eye hospital as part of a team working with people with severe eye disease. My PhD investigated changes in eye movements made by people with macular disease, then I specialised in working in the low vision clinic, where I see people like Sam and assess their vision using all the tests described above. Like my optometrist colleagues in the community, I prescribe spectacles, but also dispense low vision aids like high-power glasses, telescopes, magnifying glasses and specialist tinted lenses. I demonstrate high-tech electronic devices and give advice on techniques to use the eyes more effectively, such as how to make eye movements to overcome visual field loss. Perhaps most importantly, I make sure that people with vision impairment are supported by all the other services they need. I can refer them to a huge range of other professionals, from counsellors to specialist teachers, all within the same hospital. On my clinic days, I spend the morning in a children's low vision clinic and the afternoon working with adults, which means I might assess a four-year-old and someone who is 104 on the same day.

All the patients I see will also visit the clinic of an ophthalmologist, a medical doctor specialising in eye disease. Ophthalmology training is tough – it takes at least six years of specialist training after medical

school – and very competitive, perhaps as it is a nice specialism to work in, with few night shifts, generally healthy patients and the ability to have a big impact on people's quality of life.

I also work with orthoptists (specialists in eye movements and children's vision), ophthalmic nurses, dispensing opticians (who fit and make spectacles), ocularists (who make false eyes) and ophthalmic technicians. Beyond the jobs starting with the letter 'O', I work with specialist teachers of vision impairment, eye clinic liaison officers, counsellors, play specialists, paediatricians, medical photographers and clinic clerks. A colleague at Royal Dutch Visio (a specialist charity for people with sight loss) once told me that they employed people from 18 different professions to support their clients.

Sam didn't call her band 'Legally Blind' in the end. She is proud to define herself as a Londoner, a student, a vegan, a punk singer and a central midfielder, and she considers vision loss to be the least interesting thing about her.

Notes

1 Kuusisto, 1998.
2 Kleege, 1999.
3 World Health Organization, 2019.
4 Department of Health, 2017.
5 KV and Vijayalakshmi, 2020.
6 Vonnegut, 1970.
7 Dickinson, 1998.
8 Leat, 2011.
9 Owen et al., 2012.
10 Resnikoff et al., 2020.
11 Khairallah et al., 2015.
12 Adelson et al., 2021.
13 Durr et al., 2014.