

**STICKLER SYNDROME
SUPPORT GROUP
(SSSG)
Registered Charity: 1060421**

**EYE INVOLVEMENT
WITHIN
STICKLER SYNDROME**

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1. INTRODUCTION

Individuals who have Stickler syndrome are predisposed to a number of different visual problems, the most devastating being detachment of the retina. Retinal detachment in Stickler syndrome is often complex in nature and may be difficult to manage, although there have been significant improvements in the management of this problem over the past four decades. This has been due mainly to two areas of advance:

- The medical developments in vitrectomy surgery and techniques and surgery for dealing with multiple breaks and very large retinal tear.
- Treatment to prevent or reduce the risk of retinal detachment occurring.

2. MAIN PROBLEM AREAS

2.1. MYOPIA

In contrast to myopia in the general population which often develops during the teenage years, myopia in Stickler syndrome is typically evident from birth. Myopia or short-sight is often progressive in the general population, but in Stickler syndrome, although it may be of high degree, it is not unusual for the myopia to remain reasonably stable over long periods of time. Although myopia is common, it is important to remember that patients who are not short-sighted can still have Stickler syndrome.

2.2. RETINAL DETACHMENTS

There is a very high risk of retinal detachment, frequently affecting both eyes. Both children and adults with Stickler syndrome are at risk.

2.3. VITREORETINAL DEGENERATION

The space filling the back of the eye between the lens and the retina is filled with a jelly-like substance called the vitreous humour. In people affected by Stickler syndrome this jelly has not been formed properly from birth, and this therefore provides a very useful clue to the diagnosis for the ophthalmologist. The resulting abnormal distribution of protein and membranes within the vitreous often produce 'floaters' in the vision.

2.4. TEARS AND BREAKS IN THE RETINA

These holes and tears occur in the inner lining of the eye (the retina) which is extremely thin. They are frequently large, multiple or both and can recur at any time.

2.5. CATARACT

This is the clouding or opacity of the lens located inside the eye and affects the passage of light-rays through to the retina. Patients with Stickler syndrome can be affected at an early age, and many show an unusual characteristically curved cortical or comma shape. Once removed there is the consolation that cataracts will not recur.

2.6. GLAUCOMA

This is a condition where the pressure in the eye increases or is too high. In persons affected by Stickler syndrome this usually occurs because of obstruction of the normal circulation and outflow of the aqueous humour. Thus pressure rises, which untreated leads to irreversible damage to the optic nerve resulting in visual impairment or ultimately even blindness.

3. SIGNS THAT SOMETHING IS GOING WRONG

3.1. RETINAL DETACHMENT

The first symptom that may alert a patient that something is wrong with the eyes is when they see new floaters – floating bits swimming around in their field of vision. This vitreous separation from the retina is a common event in the general population, usually occurring between the ages of 45 and 65 years. In a minority of patients, the vitreous does not separate ‘cleanly’ and tears the retina, this in turn leads on to a retinal detachment.

In Stickler syndrome the vitreous often separates at a much earlier age and has a much higher chance of retinal tear formation. Once the vitreous has separated without complication, then the risk of retinal detachment is very low indeed. The process of vitreous separation from the retina is often associated with a dim flash of light in the edge of the vision and this symptom is a useful indication to the patient that he or she needs urgent assessment of the retina by a vitreoretinal specialist.

When we talk of degeneration of the vitreous in Stickler syndrome, we mean that the fibres come to lie together and these are what the patient sees as ‘floaters’. Other symptoms of retinal detachments may include black dots, cobweb effects, or a sensation that a curtain or ‘something black’ or “watery” is falling or moving across the eye. These symptoms frequently occur in the very early stages of a retinal detachment, or when a break is developing in the retina.

It is essential to be alert for any changes in the vision, and to report them immediately. Failure to do so could result in loss of useful vision, so don’t delay.

3.2. MYOPIA

In Stickler syndrome myopia, as mentioned, is generally severe i.e. -8 dioptres where dioptres is the strength of the lens as measured by the optometrist. Up to -6 is called simple myopia, and anything over -6 is termed high or progressive myopia. Newborn babies are normally hyperopic (+1 dioptres or greater), therefore the finding of any degree of myopia in an 'at-risk' newborn – a baby who has Pierre-Robin Sequence or an affected parent – should be considered as an indication to consider the diagnosis of Stickler syndrome. In 40% of cases, myopia develops before the age of ten and in 75% of cases by the age of 20.

3.3. THE RETINA

The retina lines the inside of the back of the eye, and is made up of shaped cells called rods and cones. The cones are responsible for direct vision like reading and for receiving bright lights and colours. The rods are for side or peripheral vision and react particularly to low levels of light, but they do not distinguish colours. Each has its part to play; in daylight the cones function whilst in darkness the rods are more active. The focused image seen by the retina is transmitted to the brain along the optic nerve which acts like a television cable.

3.4. RETINAL DETACHMENT

Retinal detachment associated with Stickler syndrome is often complex to manage and can be extensive. Urgent expert medical attention is required immediately for without repair this dangerous situation may lead to blindness.

Stickler syndrome is the most common inherited cause of rheumatogenous (tears and breaks) retinal detachments in children. Detachments can occur spontaneously and can happen in both eyes. They frequently occur under the age

of 30 in Stickler syndrome patients, and again should lead an ophthalmologist to suspect Stickler syndrome within that family. These detachments are generally responsive to surgery, provided they are discovered early and the correct action taken.

I hope I am not painting too gloomy an outlook, but I do feel it is important for those affected by this disorder to be fully aware of the potential problems and the importance of seeking medical attention immediately things appear to be going wrong, even if it turns out to be a false alarm.

Remember, as I wrote earlier, the main signs people experience when a retina begins to detach are flashing lights, new floaters, black dots, cobwebs, or a sensation that a curtain shadow, or 'something black' or a 'watery' distortion is falling or moving across the eye. The most severe detachments can suddenly reduce the vision in the affected eye to almost nothing and that can be very frightening for the person concerned. Early surgery is vital.

A detachment left untreated will progress to complete blindness in a relatively short time. It is important to note that detachments related to Stickler syndrome are not trauma related. Therefore patients are at no greater risk of detachment through trauma than those of the general population. In other words, wearing eye protection (goggles) or protective head gear will not prevent a detachment in Stickler syndrome, only general eye trauma.

When a retina becomes detached, the light-sensitive rods and cones become separated from the tissue underneath. The fluid normally present inside the eye leaks behind the tear and separates the retina from the layer behind. One consultant described the retina to me as being like thin

two-ply paper tissue rubbing together causing a weakness or tear. Another referred to the 'Stickler' retina as a piece of priceless moth-eaten material having to be carefully preserved at all costs.

Diagnosing a retinal detachment is not an easy task for an ophthalmologist, but when he or she looks into the eye with an ophthalmoscope an obvious tear or even a fold in the retina may be seen where the patient has described seeing 'curtains'. A number of less obvious changes occur long before this dramatic stage is reached. Drops, inserted into the eye to dilate the pupil, make it possible for these changes to be seen more easily by the ophthalmologist.

The extent of the retinal detachment and the speed it progresses varies according to the position, size and number of tears in the retina. In some patients the detachment will progress quickly, within days, whereas in others, the progression is very slow, over a period of months or even years. In these cases the patient may be unaware that there is a problem and diagnosis is sometimes made during a routine examination.

3.5. SURGICAL REPAIR: RETINAL DETACHMENT

When a retina becomes detached the surgeon must, under general anaesthesia, seal the retina to the underlying layer, like sticking a piece of wallpaper back in place, to prevent any further loss of vision or blindness. The aim of surgery is to repair the tear or tears in the retina using biological 'adhesive'; in the form of cryotherapy (cryo – Greek word meaning cold, and therapy meaning cure) or laser. In practice the 'biological glue' takes several weeks to reach full strength and some form of splint is required to maintain the stability of the retina during this critical period as the retinal repair matures to full strength.

3.6. RETINAL REPAIR: USE OF SPLINTS

External Splint

- In many cases the tear in the retina can be repaired using a piece of silicone rubber or sponge sewn directly to the wall of the back of the eye producing an indent or 'buckle' which maintains closure of the retinal tear as healing takes place. This implant is called a scleral buckle and is not usually visible to the naked eye, since it is stitched to the eyeball's outside surface in precisely the correct place to seal the retinal tear.

Internal Splint

- In other patients, closure of the retinal tears using an external approach is not possible or appropriate. This may be for a wide variety of reasons, but usually because the tears are very numerous and/or very large so that even the largest external splint cannot close the tears effectively while the adhesive repair develops. Using microsurgical 'key-hole' instruments the vitreous gel filling the space in front of the retina is removed (an operation called vitrectomy) and replaced with gas or silicone oil which seals the retinal tears from the inside. If a gas is used this slowly disappears over the weeks following surgery. If the silicone oil splint is required then a second operation (stage II) may be planned at a later date to remove the silicone oil once the retina is stable.

Mixed Splints

- In some patients a combination of internal and external splints are required.

3.7. RETINAL REPAIR: POST OPERATIVE POSTURING

If the 'internal' splint of gas or silicone oil is used, you may be asked to posture in the early post-operative period so that your head is in a particular position. This allows the

internal splint to float and support that particular part of the retina which was torn and seal the retinal tear or tears. As no two retinal detachments are the same, you will be given specific instructions on the appropriate posturing position after your operation. Although it is perfectly safe for you to visit the bathroom, have a meal, etc, the healing process will benefit from continuing to posture at home for 10-14 days after discharge from hospital.

3.8. RETINAL REPAIR: POST OPERATIVE RECOVERY PERIOD

Although a retinal repair is a major operation for the eye, it is rarely very painful. During the first week the eyelids will be swollen and puffy, and the eye will be very watery. The puffiness of the eyelids can be exaggerated by posturing as the swelling tends to track to the front of the eye. The 'white' of the eye will look red.

These features are all part of the normal post-operative healing process. Tears contain natural antibiotic enzymes preventing infection, so that profuse watering is a healthy and natural response to surgery.

The surface stitches dissolve after they have fulfilled their purpose so that a gritty feeling in the eye is usual during the first month. The eyelid swelling, watering and redness will all gradually subside, usually in that order but it may take several months for the redness to fade to pink and finally the white of the eye to resume its normal colour.

Flying

- When a bubble of gas is used as an internal splint you must not fly in an aeroplane until the bubble is fully absorbed. This may take several weeks. The depressurisation of the aircraft will cause the gas bubble to expand and your eye will become extremely

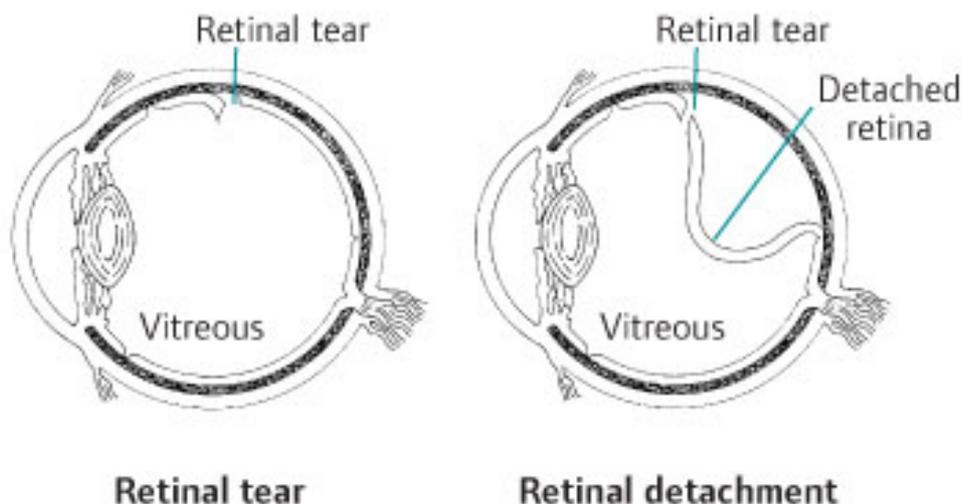
painful with increased pressure. If in doubt always ask the advice of your ophthalmologist.

NOTE: It is quite safe to fly if silicone oil has been used.

3.9. RECOVERY OF VISION

The retina is a very delicate film of nerve tissue and separation from its proper position affects its function considerably. When the retina is re-attached the recovery of function is gradual over several weeks. Further gradual improvement may continue for many months after surgery.

The focus of the eye is nearly always altered so that the final visual result may not be apparent until all drops have been discontinued and spectacles adjusted. It is important to note that if the 'reading' part of the retina (the macula) was detached then even with successful surgery the quality of the reading vision may be compromised by residual distortion due to microscopic wrinkling in the area of the retina that was detached.



This can improve with time, but rarely resolved completely to the 'pre-detachment' level. It is also important to remember that your eye-drops should be continued until your post-operative outpatient appointment when the frequency and dosage may be adjusted.

Detachments can be corrected surgically if recognised and dealt with early, although they can be very difficult to repair. Regular checks must be followed up to detect any early detachments. Therefore, the sooner they are diagnosed and operated on, the better the chance of saving useful sight. Early diagnosis is vital if blindness is to be prevented.

3.10. LATTICE DEGENERATION AND GIANT TEARS AND HOLES

Lattice degeneration occurs in about 8-11% of the general population in later life, and shows as a linear trail of fibrous vessels within the retina in a 'lattice pattern', but there seems to be a higher incidence of myopia in patients with this problem. These changes include degenerative thinning of the retina, particularly along the blood vessels, and positioned far back in the retina with characteristic pigmentation along these blood vessels, called 'pigmented paravascular lattice degeneration and these can be useful signs to aid the ophthalmologist in making the diagnosis.

Giant retinal tears (GRTs) or large horseshoe shaped tears are the commonest cause of retinal detachment in Stickler syndrome. In the very earliest stages laser or cryotherapy can be used to repair a tear that has already occurred, before the retina has started to detach. Both treatments work in an identical way, with laser generally being used for tears towards the back of the eye, and cryotherapy for tears nearer the front. The adhesion will take about 10-14 days to mature to full strength and is only effective for treatment of retinal holes or weak areas.

These procedures are usually performed under a local anaesthetic in an outpatient clinic, although some adults as well as children need a general anaesthetic. Since some tears are difficult to close and present long-term complications for the patient, your surgeon may decide to perform another type of operation.

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VITRECTOMY

Vitrectomy surgery in Stickler syndrome is usually performed under general anaesthesia. The vitreous humour is removed from the back of the eye by means of a suction-cutter, whilst at the same time the cavity is re-filled with an injection of a clear substance – saline fluid, air, gas, or silicone oil. Tiny stitches are used to close the wound and do not need to be removed.

Contrary to what people may tell you, the eye is never removed and replaced when any surgery is carried out. As soon as possible after surgery, and for at least three or four days after surgery, the patient is often nursed in a particular position, called posturing, as with a retinal detachment. The head is positioned in a certain way to enable the bubbles of gas, air or silicone oil to press against the retina to close the hole in the retina. For the first few days you must posture as directed although it is permissible to take breaks each hour to stretch and for meals, going to the toilet and washing etc. Once up and about the patient is told never to lie in the flat position as the substance in your eye may float to the front of your eye and away from the retina.

If gas has been used this will be gradually absorbed and disappear. The time this takes will depend upon the type of gas and the amount used.

If a silicone oil is used this remains in the eye until your ophthalmic surgeon makes a decision to remove it. In my particular case this operation took place six months after it had been used, and was most successful. Prior to the 'oil change' I had many complications with re-occurring detachments. Since then I have experienced few problems, with no new tears and the retina remaining flat. Because the silicone oil acts as a permanent splint, it gives some breathing space to be as sure as possible that the retina is secure before it is removed, or occasionally, if the retina is very unstable it may be left in as a permanent.

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REVENTIVE TREATMENT

Patients without a tear, detachment or new symptoms who see the vitreoretinal team at Addenbrooke's Hospital at Cambridge are now offered planned cryotherapy to the edge of the retina in order to reduce their risk of retinal detachment, although there are two schools of thought on this treatment. This method is not universally accepted by some vitreoretinal surgeons, but as with any treatment, the risks have to be balanced with other possible benefits.

The long term outlook can only be properly assessed after 10-20 years, but so far the studies look promising with about a ten-fold reduction in the incidence of retinal detachment over 15 years compared with patients who have not undergone preventative treatment. Only eight per cent of patients who receive prophylactic cryotherapy go on to have a retinal detachment, and even if these patients present with a retinal detachment after cryotherapy, it will often have limited the detachment making it easier to deal with, and the visual outlook for the patient is better.

Those who are interested in the vitreoretinal service at Cambridge should visit the website www.vitreoretinalservice.org

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THE VITREOUS ANOMALY

An ophthalmologist using a slit lamp to view the vitreous through the dilated pupil can clearly see the difference between the two types of Stickler syndrome, and this is a valuable tool for ophthalmologists when making a clinical diagnosis of Stickler syndrome.

Type 1 Vitreous Anomaly:

- In Type 1 Stickler syndrome there is an abnormal formation of the vitreous gel so that very little is produced and the vestigial remnant occupies the space immediately behind the lens leaving clear fluid in front of the retina.

Type 2 Vitreous Anomaly:

- In Type 2 the appearance is very different in that the gel fills the back of the eye but the structure is abnormal and has a different beaded fibrous structure from the general population.

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OTHER POINTS TO NOTE

Eye surgery has advanced considerably over the last thirty years, and surgery for a detached retina is no exception. Approximately 90% of retinal detachment operations are successful with a single operation. If the surgery is unsuccessful, or the patient develops a new tear re-operation is always a possibility and can tide the person over for many months or even years.

As already mentioned, if a retinal detachment operation has been successful you will be able to see although the retina will not regain full function immediately after the operation. Recovery of sight can be a gradual process,

often taking up to six months, but the quality of your vision may never be as good as previously, depending on the extent of the original problem.

If you are a driver, you must also remember that the law requires you to inform DVLA in Swansea and your insurance company of any changes in health or sight that is likely to affect the safety of your driving. The law requires you to read a number plate at 20.3 metres in good daylight, with spectacles if worn. You must also have a good field of vision.

You should never restart driving until you have had confirmation from your ophthalmic surgeon that your vision meets these standards. Failure to do so can put yourself and others at risk. Under such circumstances, you could also be faced with a situation where the insurance company declares your policy null and void with disastrous consequences financially and emotionally.

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ATARACTS

Early cataract can be another symptom of Stickler syndrome. This is a clouding of the lens of the eye associated with increased water content. People often describe the effects of a cataract as 'looking through a mist,' or 'a fine chiffon scarf' but it can also cause glare in sunlight or around headlights at night and occasionally lead to progression of short-sightedness.

An early cataract may have little effect on vision and you may be unaware of its existence until your consultant mentions it to you. Other times it will occur rapidly, (particularly after retinal detachment and giant tear) and the signs will be obvious. The main signs that a cataract is developing is a gradual blurring of vision, and as it progresses, the hole in the iris (pupil) looks white or

yellow, instead of black. Congenital cataracts from birth are usually wedged or comma-shaped in appearance, and are a very useful diagnostic sign, but are not usually visually significant to the patient.

As a cataract develops, you will notice a slight blurring of vision which can increase in bright light and decrease when dark glasses are worn. Headlights of cars appear like stars of light and when the sun is low in the sky it will cause dazzle. Usually there is 'ghosting' like a haze around objects, and occasionally blurring of the whole vision. If you do have to wait for an operation, there are a number of things which will help you to make the most of your sight:

- Wearing tinted lenses in your glasses will reduce the dazzle on a bright day.
- When out of doors wear a hat with a wide brim to shield your eyes from direct sunlight.
- Make sure indoor lighting is correct for your needs. Try to avoid bright overhead lights shining directly into your eyes.
- Try to choose a chair not facing a window which will dazzle your view. It can be most annoying to be in a room full of people and not see who is talking to you.
- When reading or doing any close work, have the light shining over your shoulder and positioned as near to the work as is practical. Daylight simulation light bulbs, which eliminate the 'orange' glow of conventional light bulbs, may help considerably.
- Immediately report any changes in your vision to your eye surgeon. Never suffer in silence; he or she has your well-being at heart.

Surgery is the only satisfactory treatment for a cataract and is usually performed at a time best suited to the patient. For example, some patients may be happy to

struggle along with relatively poor vision, others may be unable to continue in his or her occupation for very long, and therefore will welcome an operation sooner. It is a myth that a patient must wait until the cataract becomes 'ripe'.

However, for Stickler syndrome patients the surgeon may decide to delay a cataract operation because of other potential problems associated with the condition. The prime concern is to maintain as much useful vision as possible. The surgeon will tell you why, and you should always respect his or her reasoning.

The operation to remove a cataract can be performed under either local or general anaesthetic, depending on the type of cataract and your general health. There are several ways of dealing surgically with cataracts, and the whole procedure will take about half an hour. During the operation the lens is removed and must be replaced with an artificial lens.

There are three ways of replacing the lens: with special cataract glasses, a contact lens or a plastic lens implant, called an intra-ocular lens or I.O.L for short.

These I.O.L's are the most commonly used and lie either in the pupil or just behind it, to correct the focus of the eye for distance. This is not always suitable for patients with a high degree of short-sightedness (nearsightedness). The surgeon will decide and explain to the patient which is the best method. After the operation the operated eye is kept covered for 12 to 24 hours and it is advisable to wear sun glasses for a few days after the operation for comfort.

Because the eye is changing shape as it heals the focus is also changing so there is no point in being tested for glasses immediately. A wait of a few weeks after the

operation is the norm. This is a frustrating time but the wait is worthwhile. Unless there are other complicated problems you will need both distance and reading glasses, but if this is not the case, your surgeon will explain why.

Following a lens implant operation the vision is usually good but it will take a few weeks to stabilise and get your spectacle lens prescription brought up to date. Don't rush and don't worry about holding people up. Never be afraid to ask for help where needed. People are only too willing to help, if asked in the correct manner.

It will be difficult to judge distances, especially those of moving cars, so it is important to seek out and use a pedestrian crossing wherever necessary. It is far better to make a slight detour to the nearest crossing than run the risk of being knocked down crossing a busy road. It may take several months before you feel completely confident to go out alone into a busy street and traffic. On your first day out take someone with you, and build up your confidence slowly.

3.16.

LAUCOMA

Glaucoma can be a feature of Stickler syndrome, especially following complex retinal detachment. It develops when pressure in the eye rises because fluid cannot escape. This is quite different from blood pressure and the two should never be confused.

The normal eye pressure is about 15 mm Hg, but in the case of glaucoma it may increase to twice that level. My own pressure climbed to an alarming 34 mm Hg on one occasion. When the pressure rises it is more difficult for the blood to be pumped into the eye to supply vital nourishment to the optic nerve and the retina. The condition should be treated as soon as possible because

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glaucoma can cause vital nerve tissue to die, and once dead it cannot regenerate.

Secondary glaucoma - meaning it develops in response to other changes - is not uncommon in Stickler patients and is characterised by a very slow change of vision. The condition is usually painless, affects the side or peripheral vision and generally goes unnoticed until advanced. Fortunately, testing for glaucoma is a routine part of your hospital visit and should be diagnosed early before the loss of vision is too severe. Again, if left untreated it can lead to blindness.

The test is simple and painless. A gentle puff of air is blown against the eye, or a small instrument is placed gently against the eye after the eye has been numbed by drops. At the same time the ophthalmic consultant will look into the eye to see if the optic nerve is damaged and will also test for 'gaps' in your field of vision.

Treatment is usually by eye drops, tablets or both, and it is extremely important to use these as instructed. The aim of the medication is to lower the eye pressure to an acceptable level, but this will only happen as long as you are using the drops or taking the tablets. Successful management will not improve the level of your vision but it will prevent it from becoming worse.

Sometimes the treatment has unpleasant side effects which may be severe enough for a treatment to be ceased. Eye drops can cause side-effects such as blurring of vision, affecting breathing and pulse rate. Tablets may give rise to pins and needles in the fingers and toes and cause indigestion. If the symptoms persist the doctor's advice should be sought immediately and on no account should treatment be stopped until you are told to do so.

In some situations, particularly after a complex or especially recurrent retinal detachment, and despite strict adherence to drug treatment, the pressure will sometimes fluctuate or remain high. In such cases a surgeon may decide to try and reduce the pressure by surgery after which the surgeon may inform you that it is now possible to stop using your drops. Remember ALWAYS to continue to use your eye drops as prescribed until told that you can stop.

3.17. REGISTRATION: BLIND AND PARTIALLY SIGHTED

If your surgeon suggests that you should be registered as blind, this does not always mean total loss of vision. In fact only a tiny proportion of people on the blind register are unfortunate enough to be unable to see at all, and of these, many have been blind since birth. Also it does not mean that one day you will not be able to see at all.



According to the National Assistance Act of 1948, a person who is 'so blind as to be unable to perform any work for which sight is required' is classed as blind. A patient's sight is considered to have reached that stage if only the top letter on the test chart, called a Snellen chart (see diagram on left), can be seen by the patient

This is known as 3/60 vision, meaning the patient can see at three metres distance, whereas people with normal

vision would be able to see the letter at 60 metres. Sometimes a patient will be registered blind if rather more can be seen. It may be possible to read the top letter whilst sitting 6 metres distance away, but in these circumstances, 6/60 vision is considered as blindness because the field of vision is also limited. Generally people whose sight is considered to be substantially impaired are registered as partially sighted. This means that, although a person's sight is poor, it is not poor enough to be registered blind, because they can usually read the top letter, and sometimes, the next three lines.

Your ophthalmic surgeon will need to fill in a form, known as a Certificate of Visual Impairment (CVI), certifying that your vision is sufficiently poor to warrant registration. This is sent to your local social services department who will send someone around to discuss your situation. Usually the same procedure is followed for the registration of children, after a child has reached the age of four, and the vision has been corrected by glasses or contact lenses.

However, children younger than four who have an inherited eye condition such as Stickler syndrome, and whose sight is severely restricted, will be certified as partially sighted, unless they are obviously blind.

4. HOW YOU CAN REACH US

Write to:

**Stickler Syndrome Support Group
PO Box 3351
LITTLEHAMPTON
BN16 9GB**

Telephone:

01903 785771

Email:

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Visit our website:

www.stickler.org.uk

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