

Vision and care of the eyes



Eye problems which may occur in PWS

The chromosome abnormality that causes Prader-Willi syndrome leads to various problems in development, including a number of ocular features.

Squint (Strabismus)

Perhaps 3% of the general population have a squint (strabismus), but it occurs much more frequently in congenital conditions that involve chromosome abnormality.

The squint might be constant or, less commonly, intermittent. One eye may consistently be the straight one, or the eye which is the straight one may swap from one to the other (alternating).

About 60% of children with Prader-Willi syndrome have a squint, which is present at birth or occurs soon after. Almost all squints in PWS are of the crossed-eyes type (esotropia). When one eye looks straight ahead, the other eye turns inward towards the nose.

Wall eyes (exotropia), in which the eye not looking straight ahead turns outward away from the nose, is much less common as an original fault in PWS. However, it is a feature in some teenagers and adults as a later result of the surgical correction of crossed-eyes due to 'post-operative drift'.

The development of a squint at an early age in PWS could be due to the hypotonia (weak muscle tone) present at birth.

Another factor could be a restriction of eye muscle movement caused by the reduced space available in the eye socket, which is associated with the narrow face and almond shaped eyes seen in many people with PWS.

Frequency of eye examinations

Because eyes and vision in people with PWS appear to change throughout all stages of life, regular routine eye examinations are recommended.

During the period that an eye condition is being treated, the practitioner who is looking after the eyes will give advice on frequency of review.

Otherwise teenagers should be seen annually.



Lazy eye (amblyopia)

Firstly, one eye becomes the dominant sighting eye and the image from the other eye is switched off (suppressed). If one eye is consistently suppressed then the necessary visual pathways (rather like electrical circuits), leading from the image-receiving retina in the eye to the appropriate areas in the brain, will not be formed properly. The sharpness of vision (visual acuity) cannot then develop properly and a lazy eye (amblyopia) will be the inevitable consequence. This is present in about 40% of people with PWS.

It may be necessary to put a patch over the better eye (occlusion therapy) for a time, to prevent suppression and to encourage the vision in the weaker eye to develop. Occlusion therapy needs to be carried out during the visual development period, before the age of about 5 years. After this time it is much less effective.

Accommodation problems and convergence insufficiency

Other focussing and eye muscle/movement problems can affect teenagers and adults with PWS, even those who have generally straight eyes.

When we focus on a near object, it is necessary to shorten the focal length of the eye's optical system. Our eyes automatically do this, using focussing muscles inside the eye (accommodation). At the same time the eyes have to turn inwards so that each eye is directed at precisely the same near point (convergence).

Both accommodation and convergence can be insufficient in people with PWS, which can have the effect of making near work uncomfortable or blurred. These conditions can be helped by the use of spectacles - and/or eye exercises if learning difficulties are only moderate. Such assistance can help progress at school or enable better efficiency at work.



Spectacles/contact lenses

Correction of refractive errors - short and long-sightedness and astigmatism is perhaps better achieved with spectacles rather than contact lenses for most people with PWS.

Hygiene requirements and the need for care in contact lens handling could be too much for some to deal with. Even spectacle wear can sometimes be difficult, with lost or damaged spectacles being a common problem. However, for those with more than a slight refractive error, the improved quality of life achieved through better vision is worth the hassle.

Refractive error (hyperopia, myopia, astigmatism)

Long-sightedness (Hyperopia)

In long-sightedness, vision is often good, particularly for far distance, but some natural accommodation usually occurs to correct the hyperopia, keeping vision clear.

Almost all children with PWS are long-sighted to a greater or lesser extent. A low degree of long-sightedness, relatively equal in the two eyes, is normal in infancy and childhood.

However, if the degree of long-sightedness is quite different in the two eyes or is of large magnitude, the increased accommodation required, in keeping vision clear, can only occur along with increased convergence of the eyes. This will increase the possibility of squint and lazy eye developing and spectacles will be required to correct it.

Short-sightedness (Myopia)

In short-sightedness (myopia), near vision is good but far vision is blurred. Trying to use the power of accommodation to correct the short-sightedness only makes the vision more blurred and so, unlike long-sightedness, it cannot be corrected temporarily by natural means.



After the age of 16, more than 50% of people with PWS will be short-sighted, and 20-30% will be significantly short-sighted. It is not understood at present why so many do develop short-sightedness. It is associated both with inheritance and frequent close work but many people with PWS do not spend a lot of time in activities involving close vision after mid to late teens. Spectacles will usually improve the sight to a reasonably good standard.

Astigmatism

Astigmatism means that the lens of the eye does not have a perfectly spherical surface. The curvature is greater along one meridian than along the opposite meridian - a so-called toroidal surface. To help imagine this, a soccer ball has a spherical surface and a rugby ball has a toroidal surface.

Astigmatism is commonly associated with both long and short-sightedness. A correction for any astigmatism present is usually incorporated in the prescribed spectacle correction. People with PWS are often found to have significant degrees of astigmatism.



Pale coloured eyes and the effect of light

Many individuals with PWS have blonde hair and pale blue eyes. These individuals are likely to have a reduced amount of pigment within the coloured part of the eye (iris) and also in the light-sensitive membrane inside the eye (retina).

Because of the reduced amount of pigment, up to 25% of people with PWS will exhibit iris translucency, detected with the slit-lamp biomicroscope. When the light beam of the instrument is shone through the pupil and the beam reflected back from the retina is observed through the microscope, a glow of light can be seen not only in the pupil but also passing through the fibres of the iris, which would normally be opaque to light.

This means that more light than usual is entering the eyes, indoors and outside. This is unlikely to cause any problems indoors, but this may not be the case when outside in the sunlight.

The lens of a child's eye absorbs less ultra-violet than in adulthood, so more of these short wavelengths of light are able to pass through to the retina.

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Three dimensional vision (stereopsis)

Accurate three-dimensional vision (stereopsis) develops at around four to six months of age. If the eyes are not straight during this period, good stereopsis will not develop.



Almost all children with PWS are long-sighted to a greater or lesser extent.



Pale coloured eyes and the effect of light (continued)

For these individuals sunlight outside might be rather dazzling but, more importantly, in the long term, the passage of excessive amounts of ultra-violet light into the eye could be considered to be a potential hazard.

The use of a peaked cap is an easy way to reduce the amount of the sun's rays directly entering the eyes. Wearing sun spectacles outdoors with a tint to eliminate the ultra-violet light and reduce blue light would also be a sensible precaution.

Conjunctivitis and blepharitis

External eye conditions - conjunctivitis and blepharitis - are common in children and adults with PWS. These conditions are often only temporary but sometimes treatment is necessary. Blepharitis can be very persistent.

Conjunctivitis

The white of our eyes (sclera) is covered by a thin transparent membrane, the conjunctiva, which at the top, bottom and sides of the eyeball continues back as the lining of the eyelids. Tiny blood vessels thread through the membrane, but they are so small that they are not usually obvious.

The conjunctiva is sensitive to allergy and infection and, when this occurs, the little blood vessels become more prominent, making the eye appear red (conjunctivitis or 'pink eye'). If the eyes are also itchy, the problem is usually allergic in origin. This might be due to hayfever or perhaps to something rubbed into the eyes from hands or fingers.

A more prickly sensation usually means infection, and if this lasts for more than a day or two, particularly if accompanied by a discharge, a visit to the optometrist or GP is indicated.

Blepharitis

Inflammation of the eyelids, usually around the lid margin (edge) and eyelashes, ranging from very mild to severe, is referred to as blepharitis.



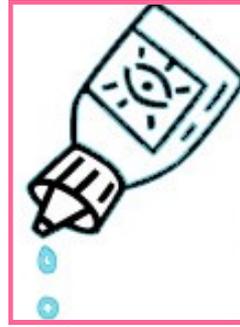
It is fairly common and can be due to a generalised skin condition (seborrhoeic blepharitis) or due to a low-grade bacterial infection (staphylococcal blepharitis). It is not contagious and cannot be passed from person to person.

Signs and symptoms include yellow or white flakes or scales crusted around the eyelashes, especially on waking, redness of the lid margins, a scratchy feeling and watering. It can be diagnosed by viewing the eyelids through a slit-lamp biomicroscope, an instrument found in the majority of optometrists' practices.

Abnormalities in the function of tiny oil glands in the lids may be a contributory cause to the development of blepharitis and lack of hygiene and eye rubbing can result in alterations to the bacterial colonies that normally live on the skin.

Crusty deposits on the eyelashes

If crusty deposits are present on the eyelashes, these need to be removed. A fresh piece of gauze dipped in a dilute solution of baby shampoo (1 part shampoo to 5 parts water), using cooled water that has been previously boiled, can be rubbed along the edges of the eyelids.



Alternatively, ready-made eyelid cleaners - tissue squares impregnated with a suitably formulated solution, are commercially available. This will usually be sufficient treatment to keep the condition at bay. If it is severe or persistent, antibiotic eyedrops and/or oral antibiotic medication are sometimes necessary.

Thank you

Many thanks to Peter Warren MSc. ClinOptom FC Optom for contributing this article.

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