



Visual disorders in Children with Down syndrome

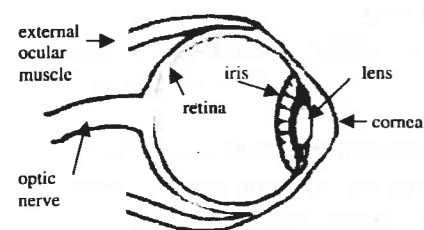
by Robyn Brady

This article, the first in a three part series discussing visual disorders in Down syndrome, looks at common ophthalmologic disorders. It is the most technical in the series because it discusses the anatomy of vision and attempts to explain some of the diagnoses parents may have been given. Subsequent articles in the series will change from the medical to the parent's perspective, looking at milestones in children's vision: what to watch for, and finally discussing practical issues with glasses and contact lenses.

Introduction

Aside from the characteristic appearances of the eyes of Down syndrome children, these children are particularly prone to a number of disorders that affect the eyes and vision. This article will attempt to describe how the eyeball gives images to the brain and common problems that may interfere in this process in Down syndrome.

How the eye works



The cornea, the windscreen of the eye, is a thin layer of transparent tissue continuously renewed by the special cells which make up its base. Special fluid called aqueous

humour lies between the cornea and the iris and lens. The coloured iris contains tiny muscle fibres which make its central hole, which we call the pupil, larger or smaller to allow more or less light to pass through it and the lens. The lens can become fatter (to focus closer) if the muscular ciliary body, which helps hold it in place, contracts.

After being focussed by the lens, light passes through the jelly like fluid behind the lens, called the vitreous humour, to the retina, a web of light sensitive nerve

cells which send the messages back to the brain along the optic nerve. The optic nerves from the two eyes exchange half their nerve fibres in the front part of the brain so that messages about what is seen on the right side of a person go to the left side of the brain, and vice versa. Messages from the centre of focus, however, are so detailed that they are delivered to both sides of the brain, and are marked for special attention. It is here in the occipital cortex of the brain that sense is made out of the messages, and they are related to other visual memories, and to other information that the brain has received about its environment (sounds, feel etc).

Good vision is more than just an image being tracked through to the retina and occipital cortex. It also involves the ability of the eyes to recognise and attend to something interesting, either close up like a face or far away like a flying seagull. Attending to something in a coordinated way involves six pairs of eye muscles so that the pictures from the two eyes match up in the occipital cortex, and the eyes are kept in alignment no matter what the direction of gaze. This may be difficult if the eye/s have limited vision or if it is hard for one or both eyes to focus, as focussing nearby causes the eyes to turn in. Having matching pictures from the centre of focus from each eye is essential to see things in three dimensions. (This develops in the first few years of life if vision is normal and is what you are relying on when you attempt to solve the 3D visual puzzles). And there are various other important and complex aspects of vision like colour sense and the ability to pick out images against differing backgrounds.

From front to back: eye problems or variations which may be present in children with Down syndrome:

1. Appearance of the eyes. Epicanthic folds are the smooth curves running from above to below the eye sockets and partially obscuring the inner angles of the eyes. Brushfield spots are the little speckles on the outer edges of the iris. These characteristics are not exclusive to Down syndrome and do not interfere with vision. Blepharitis, or dry scaly inflamed eyelid margins, is very common in Down syndrome and relates to a general tendency to dry, sensitive skin.
2. Corneal problems. Keratoconus occurs in 15% of



persons with Down syndrome. In this disorder the formation, by the basal cell layer of the cornea, of the thin protective transparent fibrous layer above it, is disturbed. The resultant irregularities may cause blurred vision or opaque scarring. This may be treated by corrective contact lenses, or if necessary, corneal grafting (replacing the damaged cornea with a donor cornea).

Corneal oedema may occasionally occur as a complication of a variety of problems, including keratoconus or glaucoma (high eyeball pressure), or after eye surgery. When this occurs the cornea gets acutely swollen and may appear cloudy. Sight is grossly disturbed and there is often pain, depending on the cause. It is an ophthalmologic emergency.

3. Refractive disorders (discrepancy between lens focussing power and the distance to the retina on which the image is focussed). These are very common in children with Down syndrome. Corrective lenses usually allow satisfactory compensation. The myopic child (short-sight, long eyes) can see close objects without much difficulty but his lens is too 'strong' for the length of his eyeball and a biconcave lens correction brings the distant images back to the focal plane of his retina. The hypermetropic (long-sighted, short eyeball) child has difficulty with close vision, as she must use all her focussing ability even to see things at the other side of the room. When she tries to look more closely, she may develop a squint when the brain gives up trying to match the two eye images, in order to see at least one more clearly. Biconcave lenses add to the child's existing focussing power, to bring the image 'forward' to the focal plane of the closer retina of the short eyeball. In astigmatism the optical power of the lens is irregular: different in different planes, so that a focussed image is not possible without correction.

4. Squint is when the two eyes are not looking parallel; ie when one appears to be turning in or out. This can occur because of focussing difficulties as above, or because one eye is not seeing properly, or the muscles controlling eye movements are not functioning properly. Recognising a squint is very important: partly in order to identify and correct whatever has caused it, and partly because if the brain is seeing two different images from the two eyes it will try to block out in the brain the image that is least useful, (the one from the turned eye) and this can cause problems with 3D vision and a lack of development of the part of the brain that receives the turned eye images.

5. Lens disorders. 15% of people with Down syndrome are born with or later develop cataracts which significantly block vision. A cataract is a hardening of the lens that makes an opaque area which cannot transmit light to the retina. They may be present at birth or may develop by themselves or as a result of a variety of upsets to the eye. Small cataracts may still allow adequate vision through the rest of the lens but when the central focus of vision is unable to be seen the lens will have to be removed. When possible this will be replaced with a synthetic lens within the eye but in some circumstances an external lens will be used.

6. Cortical problems (attention/interpretation) The brain is responsible for making sense out of what is seen. Visual analysis allows a child to extract the most important clues out of the visual world in front of him, to scan that world so that significant objects near and far may be considered, and to distinguish between similar patterns or pictures with similar contrast (bright/darkness). Many children with Down syndrome have some limitation occurring at this level even if the eyes themselves transmit images perfectly. This may mean for example that a child may tend to focus in nearby space, rather than scanning the distance, even though she can actually see well in the distance once her attention is directed there. This may increase her susceptibility environmental dangers, for example crossing a road. Another aspect which may be less finely developed is the ability to pick out relevant images from a background, especially if the important feature is similar to the background. This may be important when choosing storybooks for children with Down syndrome: simple outlines against brightly contrasting backgrounds will be easiest to make sense of at first. The Miffy books by Dick Bruna are a classic example.

The above descriptions are a short run through some of the more common visual disorders occurring in children with Down syndrome. Vision is a complex subject and the reader is referred to library texts for further information and illustrations of the above principles and problems. In the next article I plan to look at the steps of normal development of vision, and what to watch for to confirm adequate vision.

Many thanks to Robyn for these articles.