

# THE EYES AND EB

Ms Gillian SIM Ophthalmic Nurse Specialist

Mr Malcolm Kerr-Muir Consultant Ophthalmologist

## Anatomy

The eye is a globe situated in the bony orbit/eye socket on the face. It is approximately 1" /2.5cm in diameter, and contains various tissues and fluids which together enable a person to see. A large nerve called the optic nerve is situated at the back of the globe and passes directly to the brain, carrying impulses, which are converted in the brain to produce the images we see around us.

Only the front/anterior aspect of the eye is visible, and is protected by the eyelids. The white surface of the globe is covered by a thin skin (Conjunctiva), like the rest of the body, but this epithelium is transparent over the coloured part (iris and pupil). This is called the cornea, and its function is to allow light to pass into the eye, and through the lens (like a camera lens), which is situated behind the iris.

The conjunctiva and cornea are very sensitive, and as well as being a physical protective layer, they also contain cells and glands which produce part of the tear film which continually washes over the surface of the eye. A gland (lacrimal) is situated under the upper, outer aspect of the eyelid near the eyebrow, and this produces the water content of the tear film, it also produces extra tears when we cry or if the eye has been traumatised.

The surface of the eye also contains many sensory nerve endings, with a much greater concentration in the cornea. These nerves act as a protective mechanism for the eye, alerting the person to foreign objects (e.g. eyelashes, sand/grit), or trauma (e.g. scratches from sharp objects such as fingernails), which may affect the tissues of the eye, and therefore the ability to see.

## Tears

Tears form a thin layer that covers the cornea and conjunctival epithelium (skin). The functions of this ultrathin layer are:

1. To wet and protect the delicate surface of the epithelium.
2. To inhibit infection by mechanical flushing with each blink and the action of chemical components within the tear film.
3. To provide the cornea with necessary nutrients.
4. To make the cornea a smooth surface for light to pass undistorted into the eye.

The tear film is composed of three layers.

- a. A superficial oily layer derived from the Meibomian glands situated along the eyelid margins.
- b. The middle aqueous (water) layer supplied by the main lacrimal gland and minor lacrimal glands (Krause and Wolfring located in the conjunctiva superiorly).
- c. A deep mucous layer derived from Goblet cells scattered throughout the conjunctiva.

The oily layer helps prevent rapid evaporation of the tears, and the mucous layer helps the tear film to adhere to the surface of the eye.

Changes to the tear film components, and or the surface of the eye cause instability resulting in what is called dry eye syndrome.

## Eyelids

The eyelids serve to protect the delicate structures of the eye. They are made up of skin, muscle and fibrous tissue. The skin of the eyelids is amongst the thinnest anywhere on the body.

The skin of the eyelids joins the conjunctiva at the lid margins, which also contain the Meibomian glands for the tear film, and the eyelashes. At the inner aspect of the upper and lower lid margins (towards the nose), there are small ducts, which drain away excess tears into the back of the nose.

The eyelids close over the eye (blinking) many times per minute, but can also be closed voluntarily. Blinking aids the flow of tears across the eye, and the lids can be closed to form an added protective layer at times of potential trauma. The eyelids are closed over the eye at night to help reduce the evaporation of tears.

Disruption of the eyelid structures can result in problems with the tearfilms' efficiency/production, and can cause continued irritation to the eye itself.

## Conjunctiva

The conjunctiva is the thin, transparent mucous membrane covering the posterior (back) surface of the lids and the anterior (front) surface of the eye. It contains the glands and cells necessary for the production of tears. It also acts as a protective layer of the eye. It is a delicate layer, easily traumatised, and inflamed (conjunctivitis).

## Cornea

The cornea is a transparent structure, which can be compared in size and form to the crystal of a small wristwatch. It is composed of five different layers and is approximately 0.5mm in thickness. The surface layer of epithelium is very delicate and can be disturbed very easily, although healing is rapid (often a new layer of cells is apparent within 24 hours of injury).

Its functions are to provide a barrier to prevent entrance of infections into the eye, and provide a 'window' through which the light rays pass.

The cornea contains many pain fibers, and therefore any disruption to the tissues causes intense pain, which continues until the structure has healed.

## Eye symptoms in EB

Many patients with EB suffer with eye problems, especially those with Dystrophic EB.

Like the rest of the body, the skin (epithelium) of the eye is prone to blistering. This can be spontaneous or caused by direct trauma, such as rubbing of the eye, or scratches caused by foreign objects, or a disruption of the tearfilm causing drying of the eye, and resultant friction.

Problems with the eyes are often distressing since, as well as pain there is often a temporary disruption of the vision. This can have an effect on daily activities, and patients, especially children can become quite anxious.

Care of the eyes is quite specific when blistering occurs, but it is important to look at the overall care of the eyes even when there is no blistering.

It should be remembered that patients might also suffer with other eye problems not associated with EB, such as cataracts and refractive errors (e.g. short sightedness requiring glasses correction). These are best dealt with separately, although the patient should ensure the clinician is aware they have EB.

## Examination

One of the anxieties encountered by EB patients is having the eye examined. Very often a good history from the patient can point to a diagnosis, so the patient is advised to have information available, such as what has happened (sudden pain and watering of the eye), when (this morning on waking), past history (happened three times before in same eye) etc. It may help to write it down, and some patients even keep a diary of events if they have re-occurring eye problems.

There are several simple rules to follow when examining the eyes of a patient with EB. Most ophthalmologists are very skilled in the care of patients with problems such as EB, but if the patient is in doubt, the following advice may prove helpful.

If at all possible a no touch technique should be used. In the adult patient this is easier since the patient is often able to assist the clinician. In the case of a child, distraction with toys or lights can be useful to promote opening of the eyes.

Application of a local anaesthetic drop such as Proxymetacaine Hydrochloride, which does not sting on application as much as say Benoxinate Hydrochloride, may be useful if the patient has pain causing them to be unable to open the affected eye.

The most important factor is that the lids should not be forced open, since this can result in further trauma to either the lids themselves or the conjunctiva/cornea. The lids can be very gently moved to examine areas of the conjunctiva not in view when the patient has their eyes open, but again touch should be very light, and if the patient is not able to co-operate, it is best to discontinue.

Examination by torch or slit lamp (if available), should be done obliquely, since shining the light directly into the eye causes spasm of the iris and resultant pain, photophobia, and closure of the eye.

Instilling a drop of Fluorescein (also available combined with Proxymetacaine, requiring only one drop application for anaesthesia and examination), will show any epithelial defects that may not be visible on simple light examination.

## Corneal blisters (erosions)

The surface of the eye can blister, just as the skin does. If a small blister occurs on the cornea (clear part of the eye) this can be very painful. It is known as a corneal erosion. The problem may first appear as a baby, but some patients do not have any episodes until adulthood. Usually the attacks have no predictable pattern, but a large erosion may well follow several relatively minor episodes.

The erosions heal spontaneously, usually within 24 hours, depending on the size of the area blistered. They usually heal without scarring, but if infection occurs, or the erosions are large and frequent, scarring can occur leading to visual problems later on.

A general area of haze is apparent in patients with Dystrophic EB, at the edges (limbus) of the cornea, usually inferiorly (lower edge). This is called Broad Limbus, and appears to have no significant affect on the function of the eye.

Treatment of corneal erosions for child and adult is basically the same.

Once the blister has occurred, the main treatment is for the severe pain that can accompany the episode. Oral analgesics such as Paracetamol may well be of help, but if oral medication is difficult, a drop such as Voltarol Ophtha (Diclofenac sodium 0.1%) may be of benefit.

Quiet rest, with the eyes shut is also helpful, since the eyelids form a 'bandage' over the eye, and the reduced movement of the eye itself reduces further friction of the lids on the affected area.

Anaesthetic eye drops such as Benoxinate should not be used as a treatment, since they delay the healing process, and the anaesthetic effect increases the danger of further trauma without the patient realising it. Anaesthetic drops are used for examination of the eye, and in certain special cases, under the supervision of a clinician, can be used to help the patient carry out a specific task. For instance, if a nervous child/adult is due to go to theatre, and has a severe episode of corneal blistering, local anaesthetic drops may help them see, as they go down to theatre, since lack of vision may increase anxiety further. However, the anaesthetist and operating staff must be made aware that drops have been instilled.

Steroid eye drops should only be used under strict supervision from a hospital expert, since they can have adverse affects.

Instillation of lubricants at the acute stage is also beneficial. Gels and ointments such as Lacri-Lube or Simple Eye Ointment are preferable since their effect lasts longer than drops, and reduces the need for frequent re-application. However, if it is impossible to open the eye to instill the lubricants, then it is best to abandon the treatment, otherwise further trauma can occur from the action of trying to prise open the lids.

If after 24 hours the erosion has not improved and there is a suspicion of infection, an ointment containing an antibiotic such as Fucithalamic gel, or Chloramphenicol ointment can be added under the supervision of a clinician.

Padding the affected eye can help with symptoms such as pain, but does not necessarily shorten the healing time. Keeping the pad in place is a problem, since tape cannot be used. So, using Tubifast, Netelast, or conforming bandage is recommended, although regular checks that the pad is correctly in place is necessary, since the pad itself can cause friction and therefore further damage to the affected eye.

Wearing sunglasses during an episode of corneal erosion reduces the problem of photophobia (pain on looking at bright light). When the eye is damaged, the iris (coloured part) of the eye goes into spasm, and as a result when the patient is in bright sunlight/ brightly lit room, the pupil constricts causing further spasm of the iris and resultant pain. If this pain is severe, instillation of a dilating drop such as Mydrilate (Cyclopentolate hydrochloride 0.5%/1%), 'relaxes' the iris and reduces the pain.

It has been shown that prophylactic (preventative) treatment of the eyes of patients with EB, reduces the incidence of corneal erosions.

Routine application of lubricant gels or ointment every night whether there are eye problems or not, appears to reduce the incidence of erosions considerably. Lubricant drops for use during the day (ointments tend to cause transient blurring of the vision), as necessary, also appear to help.

## Lids and Conjunctival problems

Like the cornea, the lids and the conjunctiva are susceptible to blistering. The eyelids can form blisters at any time, but usually it happens as a result of rubbing the eyes, especially in children. Blisters are best treated in the same way as other areas of the body, but again dressings are a problem. Instillation of lubricants, especially if the blistering is at the lid margin is recommended, since distortion of the lid edges can cause disruption of the tearfilm, drying of the eye and resultant friction, possibly causing corneal or conjunctival blistering also.

Conjunctival blisters are treated in the same way as corneal erosions, although they tend not to be as painful, and photophobia is less.

Unfortunately, recurrent conjunctival and lid blistering can result in scarring, especially in the Dystrophic EB patients.

This can result in Symblephron, where the inside of the lid fuses to the conjunctiva. This causes problems with closure of the eyes and the tear film is disrupted since the surface of the eye becomes irregular. If the eyelids become scarred as a result of recurrent blistering, incomplete closure, and inefficient blinking increase the incidence of corneal erosions. Lagophthalmos (inability of the lids to close properly) results in nocturnal exposure (eyelids remain open during sleep) which again causes drying of the eyes and possible blistering. Exposure of as little as 1-2mm appears to have an effect. It is often the parents or partner that become aware of the problem first since the patient is asleep and unaware.

Again, it appears that routine application of lubricants at night reduces the incidence of erosions of the cornea and conjunctiva. If the scarring and resultant distortion of the lids is severe, corrective surgery can be performed. This is best discussed with an ophthalmic/plastic surgeon experienced in the treatment of EB patients.

## Tearfilm problems

The tearfilm is easily upset by any irregularities of the surface of the eye, and this is a common problem in EB especially dystrophic EB, where scarring may have occurred.

The irregularities cause pooling of the tears in some areas, and drying of the tears in other areas. Overall the effectiveness of the tearfilm is diminished and as a result there is increased friction of the lids over the eye. This is especially noticeable on waking in the morning, when the lids, on opening, can cause trauma to the surface of the eye resulting in blistering.

Tearfilm efficiency can be affected by other factors such as environment (e.g. air conditioning), and reduced blinking (e.g. when concentrating on looking at a computer screen). Hayfever, and other allergic reactions may also have an effect, and these should be considered.

Treatment is routine lubricants. Ointment (e.g. Simple Eye Ointment), or Gels (e.g. Lacri-Lube) are used at night, and drops (e.g. Viscotears or Hypromellose) are administered as necessary during the day. These lubricants do not contain any medication, so the frequency of administration is not a problem. Some patients however, may develop sensitivity to the preservatives in the preparations, e.g. Benzalkonium Chloride, and if this is so, preservative free preparations are available.

There are at least 15 different types of lubricant available, and although the most commonly used are Viscotears, Lacri-Lube, Hypromellose and Simple Eye Ointment, the patient may try several different types before finding one that suits them best. This can be discussed with the clinician or pharmacist.

Washing the eye using eye baths is not recommended, since this will only further disrupt the tear film, and although giving some initial relief, will only be transient. However, if there is a foreign body in the eye (e.g. sand), flushing the eye with a solution such as Normasol (Sodium Chloride 0.9% in sachets), or even boiled, cooled water is recommended as an initial treatment, followed by a drop of lubricant once the foreign body has gone.

## Eyecare of the Child

Parents of children with EB have many priorities when caring for their child, and eye care may not be one of them. However, a child with an eye problem is very distressing to all concerned. So, anything that may help reduce the incidence of blistering to the eyes is worth considering.

All children, whether EB sufferers or not get caught up in the rough and tumble of life. This inevitably results in an eye problem of some sort, whether it is a fingernail scratch in the eye during a game, or sand in the eyes from the sandpit/playing on the beach.

Children (of any age!) tend to rub their eyes when tired, or distressed, and infections such as conjunctivitis are a common problem.

Any of the above incidents, and many others, can result in blistering of the eyes. Children should not be discouraged from joining in with activities as they are able, but the parents awareness of potential problems, and knowledge of prompt treatment, is invaluable.

Most children do not like having their eyes examined, or medication instilled. Interestingly, children who are born with blocked tear ducts and therefore have sticky watery eyes, seem to be much more amenable to examination and treatment. This may be due to the fact that since birth, they have become accustomed to regular cleaning, examination and drop treatment. Parents incorporate eye care into the daily routine, and babies soon learn that it is normal and non-threatening. Sometimes they even seem to enjoy the process. Examination of these babies in the clinic is relatively easy.

Although not scientifically proven, it may be that if the child's first encounter with eye care is when they have a painful problem causing anxiety in both child and parent, they may be fearful of further care in the future. Clearly not all children are the same, and some are more sensitive than others. But, a routine of gentle eye care from early infancy may help reduce problems when blistering does occur.

A school of thought feels that children with severe types of EB, especially dystrophic, should be prescribed routine lubricants from an early age. Gentle cleansing of the eyelids using boiled cooled water or saline solution becomes part of the daily routine.

Children should be discouraged from rubbing their eyes. Unfortunately, the dressings that the children may have on their hands, becomes an ideal tool for rubbing the eye!

If this is a problem especially with babies, soft cotton mitts can be put loosely over the dressings reducing the direct friction of the dressings on the eyelids.

If blistering occurs, do not attempt to prise open the eyelids to examine or instill lubricants. If the child is able to open the affected eye itself, a lubricant ointment or gel can be instilled, and painkillers can be given. If they are unable to open the eye, lie the child down flat, with their eyes closed and apply a generous amount of ointment or gel to the closed lids along the lashes and lid margins. Over a few minutes this will melt and seep into the eye. A gauze pad moistened with boiled cooled water or saline and laid over the closed eyes may help to make the eye more comfortable. An eyepad can be applied but it is not a necessity. The child should be encouraged to rest and not move the eyes as much as possible. If the blistering is minor, watching television is ideal, although computer games are not recommended since the concentration required usually reduces blinking time and it would therefore be better if the eye was padded.

If the affected eye is not more comfortable within 24 hours or has got worse, expert advice should be sought from an ophthalmologist.

Sunglasses are now available for all children and even the smallest babies. They are very helpful if a child has photophobia following a blister in the eye, or when playing outside with other children. Inexpensive, supple, brightly coloured, trendy sunglasses made by Tommy Tippee especially for babies and children are widely available in chemists.

Care should be taken that they are the right size, and that they do not cause rubbing behind the ears or on the bridge of the nose.

Children should have the usual routine eye tests throughout infancy and childhood, and squints and any other disorders such as lid problems should be referred to an ophthalmologist specialising in care of EB patients.

If a child needs to wear corrective glasses, parents should ensure that a qualified ophthalmic optician fits them correctly. Some sports companies now make frames that are supple and light and these seem to help reduce the rubbing and potential blistering behind the ears and bridge of the nose. Wire ear pieces are not recommended, so if the child is having difficulty keeping the glasses on, special bands made from a neoprene type material can be obtained from most sports/surf shops and some larger opticians. These brightly coloured bands cover the earpieces of the frames and are secured at the back of the head with a bead or tie.

## Eyecare of the adult

In adulthood, eye problems appear to diminish, but some patients continue to suffer recurrent erosions and may have scarring from earlier erosions, which cause further problems.

Patients should be encouraged to take responsibility for their own eye care. If they are able to write and comb their hair, most should be able to instill their own eye drops.

Some patients decide to discontinue their lubricant eye drops when their eyes feel better. This is discouraged, since the lubricants do not cure the problem, but maintain the tearfilm and therefore help reduce the incidence of blistering.

If patients' work involves using a computer for long periods of time, care should be taken that the concentration involved does not affect the eyes. Regular breaks from the computer, when the patient should blink with complete eye closure for about a minute. This will help combat the affects of reduced blinking and drying of the eyes which can occur.

Although not scientifically proven, it appears that stress and tiredness affect the tearfilm, especially at night. Many patients have stated that following a stressful/tiring period they have had more episodes of blistering than usual. Therefore, it is recommended that patients instill lubricants at night during those periods, if they are not routinely taking them.

For cosmetic reasons, or intolerance of glasses (persistent blistering of ears and bridge of nose) some patients may wish to wear contact lenses. They would need to discuss this with an ophthalmologist, experienced in patients with EB.