



Leprosy and the eye

teaching set

Leprosy is a disease which is still endemic in most developing countries and also continues to be a significant cause of blindness in many communities. In 2009, 244,796 new patients with leprosy from 141 countries were detected worldwide.¹

The number of new leprosy patients is slowly decreasing. Over the past twenty years, more than 14 million patients have been 'released from treatment' (RFT) for leprosy² but it is estimated that between one and two million people currently have severe disabilities, including blindness, due to past and present leprosy.³

As the number of leprosy patients decreases, it becomes more difficult to ensure health workers have sufficient knowledge about this disease. This teaching set provides a practical tool for teaching health workers how to recognise and refer leprosy patients with eye disease. It also sets out the measures necessary for the treatment of eye disease in leprosy.

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1. Transmission of leprosy



What is leprosy and how is leprosy transmitted?

Leprosy is an infectious disease caused by a micro-organism, the bacillus *Mycobacterium leprae*. *M. leprae* is a slow-growing organism with a preference for cooler temperatures. As a result, the skin, nose, ear lobes, certain peripheral nerves, and the slightly cooler anterior part of the eye are more likely to be affected by leprosy.

Leprosy is transmitted by untreated patients with multibacillary leprosy (*see below*) through nasal discharges and sneezing. Incubation time is about five years, but may be longer (*see below*). The photograph on the right shows nasal discharge containing many red-stained bacilli. Multibacillary leprosy patients may develop ulcers in the nasal septum, with heavy loads of *M. leprae*. A septum perforation finally causes the collapse of the nose in such patients. Leprosy is not transmitted through infected wounds: these are infected by other common bacilli.

Because leprosy may cause severe deformities, there used to be a great fear of leprosy and leprosy patients were often forced into isolation. With modern anti-leprosy treatment, patients are made non-infectious almost immediately. Therefore, patients are treated as outpatients and isolation has become obsolete.

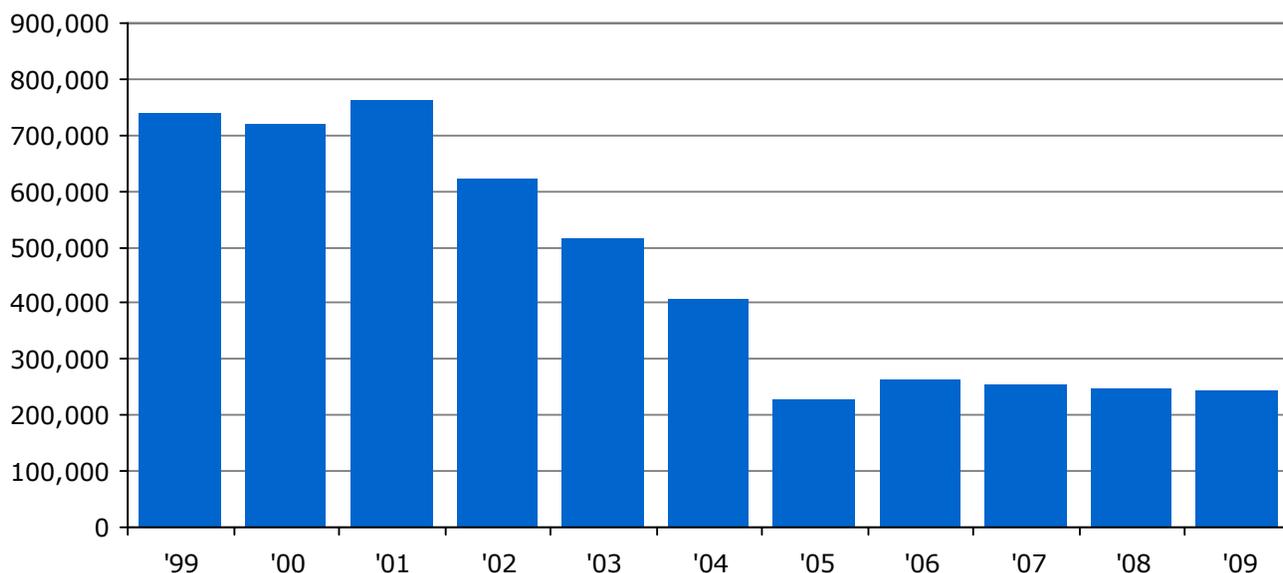
Most people have a natural immunity against leprosy. Health workers who work with leprosy patients are unlikely to contract leprosy.

Leprosy is sometimes called Hansen's disease, named after the doctor who discovered the bacillus, Dr G A Hansen.

2. Leprosy around the world

244,796 newly detected cases in 2009

Newly detected leprosy patients 1999 - 2009¹



How common is leprosy and in which countries does it occur?

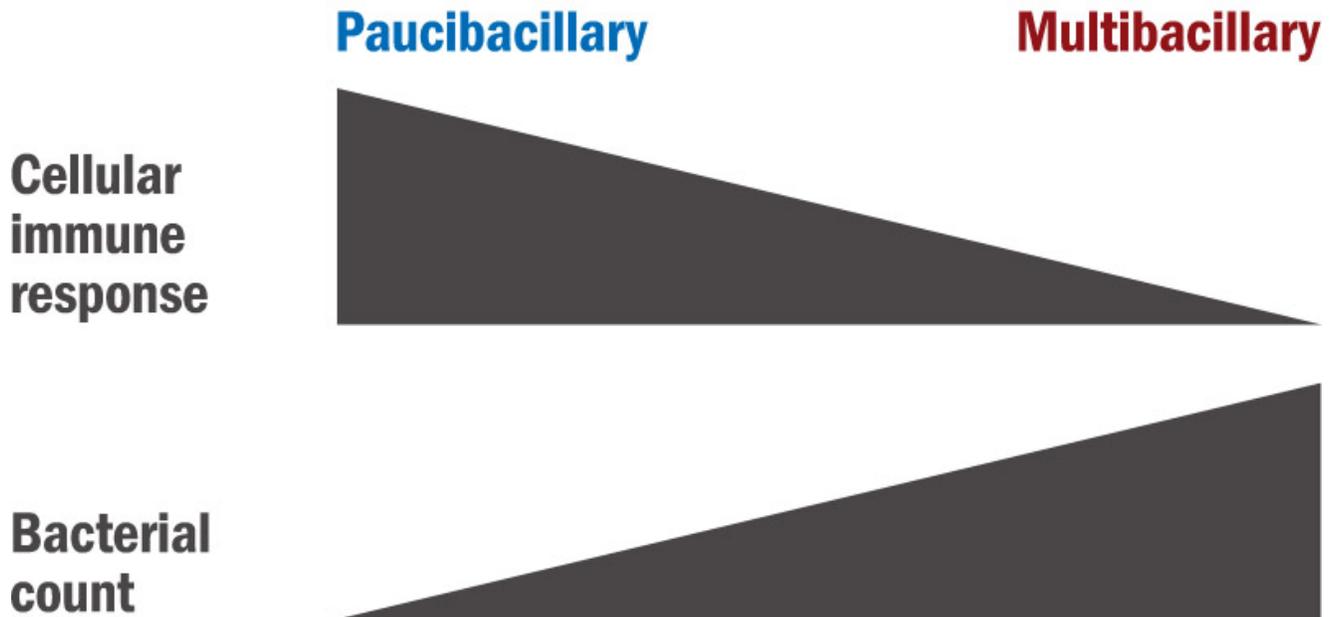
Note: The World Health Organization definition of a leprosy patient is a patient with leprosy in need of antibacterial treatment.

Leprosy is still endemic in many developing countries.

Great progress in leprosy control has been made over the past 25 years. In 1982, before the introduction of fixed duration multidrug therapy (MDT), more than 12 million patients were on treatment registers and treatment was lifelong. These patients have now been released from treatment. An estimated 1-2 million of them still suffer from severe disabilities, including blindness.³ These patients are not included in the usual prevalence and case detection statistics.

India, Brazil, Indonesia, Congo, Bangladesh, Ethiopia and Nepal reported the most new leprosy patients in 2009.¹

3. Clinical presentations



What determines the clinical picture of leprosy?

The clinical picture of leprosy is determined by the immune response of the individual.

If the immune response is high, the corresponding bacterial count will be low and so-called paucibacillary (PB) leprosy will develop. PB leprosy is subdivided into PB single lesion leprosy (one skin lesion) and PB two to five lesions leprosy (two to five skin lesions).

If the cellular immune response is low, the corresponding bacterial count will be high and so called multibacillary (MB) leprosy will develop (more than five skin lesions).

Type of leprosy

- PB leprosy: single skin lesion
- PB leprosy: 2-5 skin lesion
- MB leprosy: more than 5 skin lesions

MB leprosy patients may spread the disease. PB leprosy patients do not spread the disease.

Knowing the type of leprosy is important for both the treatment regimens and the type and pattern of systemic complications that may occur.

Note: Previously, PB leprosy was subdivided into Tuberculoid (TT) and Borderline Tuberculoid (BT) leprosy; MB leprosy was subdivided in Borderline Borderline (BB), Borderline Lepromatous (BL) and Lepromatous (LL) leprosy, as a continuous spectrum. In the new classification more previously PB patients are now classified as multibacillary.

4. Paucibacillary leprosy and multibacillary leprosy



What are the clinical presentations of PB & MB leprosy?

PB leprosy is characterised by skin lesions that are hypopigmented, well defined and anaesthetic (no sensation to touch). Any skin lesion that is anaesthetic to light touch is suspicious for leprosy. There is no itching.

Note: Lesions in the face usually do not show loss of sensation (photo: top left).

In PB leprosy less than 3 'body areas' (e.g., arm, leg, chest, etc.) are affected. Only a few nerves are enlarged on palpation. An example of enlarged greater auricular nerve is shown in the photo on the bottom left.

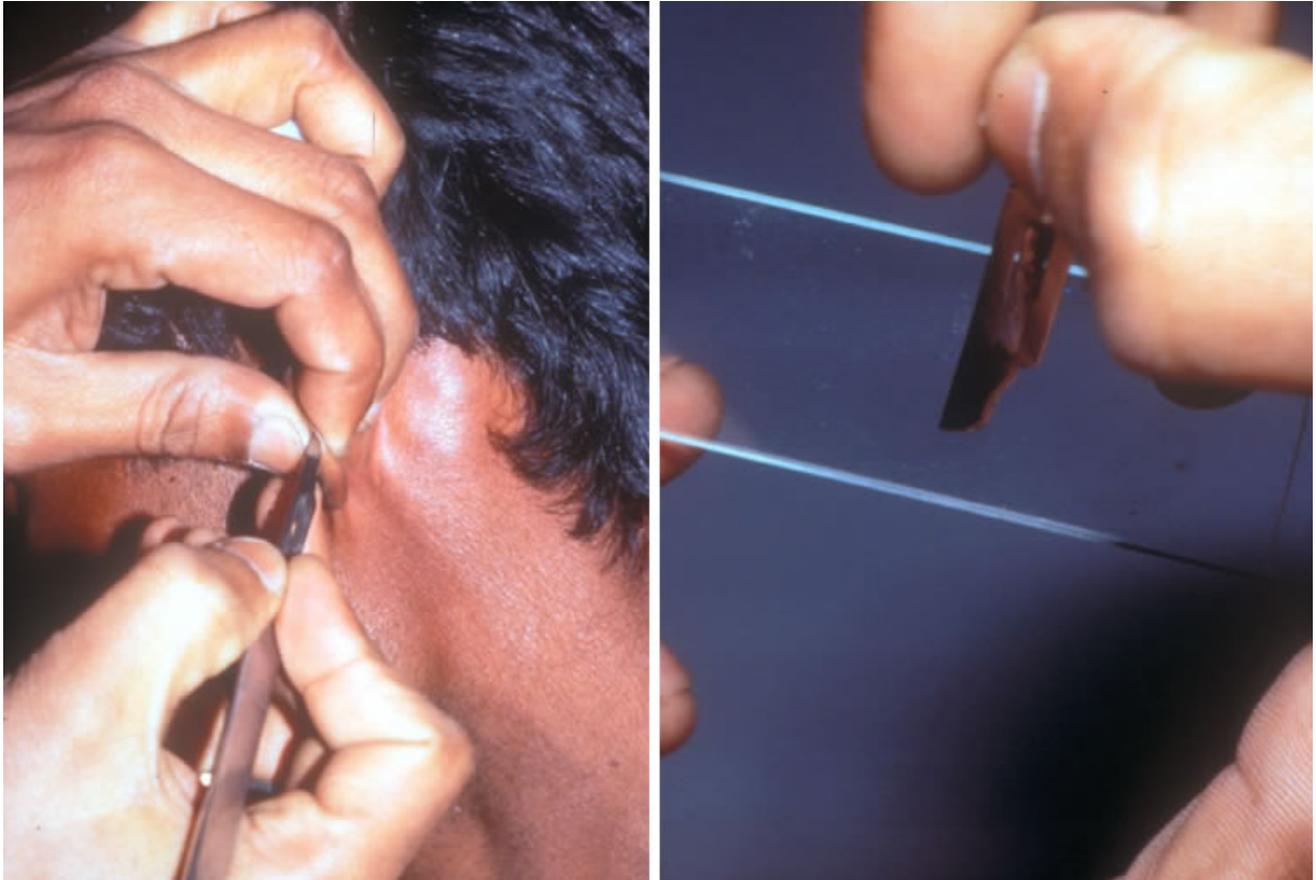
MB leprosy is characterised by numerous pale or reddish, poorly defined skin lesions or nodules (photo: top right) without loss of sensation and symmetrical enlargement of the peripheral nerves. There is no itching. In long-standing disease loss of eyebrows, collapse of the nose and nodules on the ears will develop.

Loss of sensation, for example of the hands and feet, results in insensitivity to repeated 'minor' injuries, which can lead on to ulceration. An example of a typical foot (plantar) ulcer is shown bottom right.

What is the differential diagnosis of the skin lesions?

Vitiligo, other depigmented lesions and birthmarks should be recognised in the differential diagnosis of leprosy. However, none of these lesions show loss of sensation. Fungal infections are usually itchy and also do not have any loss of sensation.

5. Skin smear in diagnosis



To confirm the diagnosis of leprosy, skin smears may be taken.

A small skin incision is made in one of certain preselected areas, for example ear lobes. A small scraping is made and the tissue is placed on a glass microscope slide. This is stained for *M. leprae* ('acid-fast' bacilli stained by the Ziehl-Neelsen method).

How may this test confirm the diagnosis of leprosy?

Paucibacillary leprosy patients have very few bacilli and therefore have negative skin smears. Multibacillary patients have high numbers of leprosy bacilli and therefore skin smears may be positive. However, because the definition of multibacillary patients (for control programmes) has changed to identification of more than 5 skin lesions, as few as around 12% of these patients are actually smear positive. Also, the skin smear results are very much dependent on the skills of the laboratory technician and are therefore not always reliable. Many leprosy control programmes rely on the clinical picture alone. Further, because of the emergence of HIV/AIDS, skin smears are less commonly taken.

Leprosy bacilli need special tissue for culture and will only culture satisfactorily in the mouse footpad. The nine banded armadillo is the only animal that is known to be a natural host to *M. leprae*.

6. Multidrug therapy (MDT)

What is the standard treatment for leprosy?

There are standard World Health Organization recommended multidrug therapy (MDT) regimens for the treatment of leprosy. Dapsone alone (monotherapy) is no longer used. MDT is free of charge and is provided in monthly blisterpacks through leprosy control services and in integrated programmes through general health posts.

Adults:

Paucibacillary leprosy (single skin lesion):

- rifampicin 600mg as a single dose
- ofloxacin 400mg as a single dose
- minocycline 100mg as a single dose

Paucibacillary leprosy (2-5 skin lesions):

- DDS (dapsone) 100mg unsupervised once daily
- rifampicin, 600mg supervised, once per month, 6 doses to be completed in a maximum of 9 months.

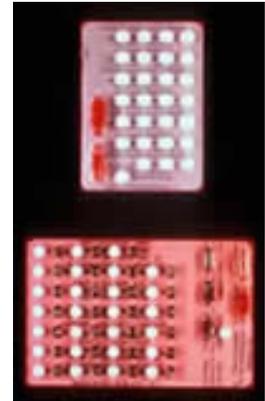
Multibacillary leprosy:

- DDS (dapsone) 100mg and clofazimine (lampoene) 50mg, unsupervised once daily.
- rifampicin 600mg with clofazimine 300mg, supervised, once per month, 12 doses to be completed in a maximum of 18 months.

Children: Should receive adjusted doses.

Leprosy patients are nowadays usually treated as outpatients through leprosy control programmes within the primary health care system. In case of severe reactions or serious ulcers, patients may need to be admitted into hospitals. Leprosy patients should no longer be isolated in leprosaria.

After completing the standard regimen, the patients are 'released from treatment' (RFT) and are no longer considered as 'leprosy patients' in the WHO statistics even if they are patients with severe disabilities and in need of rehabilitation or regular control, eg for ulcers. Recurrence of leprosy after RFT is rare.



7. Reactions (type 1 and type 2): massive infiltration with *M. leprae* and secondary atrophy



The photographs show (from left to right):

- Type 1: reversal reaction
- Type 2: ENL reaction
- Massive infiltration with *M. leprae*
- Secondary atrophy

How do the abnormalities found with leprosy develop?

Two mechanisms are responsible for nerve damage in leprosy and these may lead to various disabilities. These are A: reactions and B: atrophy due to massive infiltration with *M. leprae*.

A. Reactions

Reactions are sudden changes in the immune response. There are two types of reactions:

1) Reversal reactions (RR), also called Type I reactions

Reversal reactions occur as a result of a sudden increase in cellular immunity. There is acute redness and swelling in the skin (photo on far left) and painful swelling of the affected peripheral nerves. This may cause loss of both sensory and motor nerve function, leading to anaesthesia (loss of sensation) and muscle weakness. The ulnar, median and posterior tibial nerves are most commonly affected. For the eye, involvement of the facial nerve (N VII) and the ophthalmic branch of the trigeminal nerve (N V,1) are important: these may cause lagophthalmos and corneal hypoaesthesia (reduced sensation) respectively.

Reversal reactions are common and occur either before the diagnosis of leprosy, at the start of anti-leprosy treatment, during the first six months of anti-leprosy treatment or, less commonly, after release from treatment (RFT). A reversal reaction is often the reason why the patient seeks help.

2) Erythema nodosum leprosum (ENL) reactions, also called Type 2 reactions.

ENL reactions occur only in MB patients and are due to antigen - antibody reactions. ENL reactions are clinically evident by the onset of a sudden but often recurring syndrome, with fever, subcutaneous nodules, swelling of nerves and inflammation of certain organs (photo

second from left). Acute iritis and scleritis are manifestations of ENL in the eye and may lead to blindness.

ENL reactions occur in about 20% of MB patients and appear usually during treatment, or in untreated MB patients with a long history of disease.

B. Massive infiltration with *M. leprae* and secondary atrophy of the involved tissues

Massive infiltration (photo: second from right) and secondary atrophy (photo: far right) occur only in MB patients with long-standing disease. The skin of the face, the nasal septum mucosa and ear lobes can be heavily infiltrated with *M. leprae* and later show atrophy. This results in madarosis of the eyebrows and eyelashes, collapse of the nose, and thin ear lobes. Within the eye, infiltration followed by atrophy may affect the iris, resulting in iris atrophy.

Infiltration of nerves and secondary atrophy will result in diffuse hypo/anaesthesia over the limbs and muscle weakness.

The anaesthesia, due to reactions or nerve atrophy, may lead to repeated ulcers of hands and feet following neglected injuries.

Muscle paralysis may lead to clawing of the hands and feet and foot drop.

Severe deformities may develop, a common picture in late neglected leprosy patients.

8. Type I reaction and result after 48 hours of treatment with prednisolone



A Type 1 reversal reaction (left) and the same patient after 48 hours of treatment with prednisolone (right).

1. What is the treatment for reactions in leprosy?

The most important treatment for a reaction is a course of systemic steroids. Anti-leprosy drugs are meanwhile continued.

Treatment of reversal reactions

- Mild reversal reaction without nerve involvement: rest, with aspirin and chloroquine.
- Severe reversal reaction with nerve involvement: (new anaesthesia or muscle weakness) and any reversal reaction in the face: systemic prednisolone. The standard course is a starting dose of 40mg/day, slowly decreasing over a period of 12 weeks. If no improvement occurs within a short period (a few days) the starting dose may be increased.

The photographs show a Type 1 reversal reaction (left) and the same patient after 48 hours of treatment with prednisolone (right). Patients with such a facial reaction are at great risk of developing facial nerve damage and lagophthalmos. Timely treatment of a reaction in the face will prevent or diminish damage to the facial nerve.

Any recent nerve damage of less than 6 months' duration is treated by a course of systemic prednisolone. Nerve damage of more than 6 months' duration will usually not improve, even with prednisolone.

Treatment of ENL reactions

- Mild ENL reaction: rest with aspirin and chloroquine.
- Severe ENL reaction: short course of systemic steroids (e.g., prednisolone 40–80mg) and high doses of clofazimine (300 mg/day). The side-effects of such high doses of clofazimine are dark or orange discolouration of the skin and redness of the sclera. The latter should not be confused with scleritis or sclero-uveitis. The discolouration is reversible after discontinuing clofazimine.

The frequency and severity of ENL reactions have become less since low doses of clofazimine have been included in MDT for multibacillary leprosy. This has also diminished the manifestations of ENL inside the eye, such as acute iritis and episcleritis.

Note: the sometimes prolonged treatment with prednisolone for recurrent reactions should get priority over the possible side effect of secondary cataract, as the damage due to reactions will be definite. Cataract can be treated later, after the reactions have subsided.

2. What is the treatment for massive infiltration with *M. leprae* and secondary atrophy?

The complications of infiltration and secondary atrophy can only be prevented by early diagnosis of leprosy and effective multidrug anti-leprosy treatment.

9. Eye complications

Leprosy: potentially blinding lesions

1. Lagophthalmos:	whole spectrum
2. Corneal hypoaesthesia:	whole spectrum
3. Acute iritis and scleritis:	MB leprosy
4. Chronic iritis and iris atrophy:	MB leprosy
(Cataract, age related and secondary:	the whole spectrum of leprosy)

Why is good eyesight so important for leprosy patients?

A leprosy patient should be able to observe and avoid any object that could cause injury. A leprosy patient should also check, visually, anaesthetic hands and feet, so that ulceration following injury can be treated early and later deformities prevented. A blind or severely visually impaired leprosy patient cannot see or feel wounds and ulcers. Good eyesight is also of great importance for patients with poor mobility.

What are the eye complications in leprosy and how commonly do they occur?

In 2002 it was estimated that up to 100,000 persons were blind (<0.1) as a consequence of leprosy, including former leprosy patients who had been released from treatment (RFT). If age-related cataract and other non-leprosy causes are also considered, the total number of blind leprosy patients may be as many as 300,000.⁴

Eye complications in leprosy are fairly common. Leprosy patients can, of course, also develop other potentially blinding eye diseases not related to leprosy, such as cataract. The consequent visual loss is the same.

Eye complications due to leprosy are confined to the cooler anterior segment of the eye (cornea, sclera, iris, ciliary body, lens) and to the ocular adnexae. The posterior segment (retina, choroid, optic nerve) is not involved.

There is a close correlation between the type of leprosy and the eye complications that may occur.

Potentially blinding lesions:

Occurring in:

Lagophthalmos and exposure keratitis:	the whole spectrum of leprosy
Corneal hypoaesthesia:	the whole spectrum of leprosy
Acute iritis and episcleritis:	MB leprosy
Chronic iritis and iris atrophy:	MB leprosy
(Cataract, age related and secondary:	the whole spectrum of leprosy)

These lesions may progress and lead to severe visual impairment or blindness long after patients have been released from treatment (RFT).

Most eye complications are found in elderly patients with long-standing MB leprosy and multiple disabilities. These patients are mostly already RFT and often cluster in leprosy settlements.

Many young patients with a short history of disease still have normal eyes.

Thus there are large differences in eye complications, depending on where the study was done, either in the field, in leprosy departments of hospitals or in leprosy settlements or in new patients on MDT versus RFT patients with a long history of disease and initial possibly irregular dapsone monotherapy.

There are also large differences in reported complications, depending on which eye lesions were included, only the potentially blinding lesions, or all lesions in relation with leprosy (including madarosis of brows), or even lesions without a direct relation to leprosy.

10. Facial patches and lagophthalmos



Photograph on the left shows a pale patch over the area of the facial nerve. Such patches should be monitored for development of reaction. Patients should be instructed accordingly and present immediately.

Photograph on the right shows red and raised skin patch in reaction with early lagophthalmos. Note accompanying madarosis of left eye brow and eye lashes. A skin smear has just been taken from the margin of the patch, on the chin (small piece of cotton wool).

Facial reactions and lagophthalmos

Lagophthalmos is the most well known eye complication in leprosy.

Who is at risk of developing lagophthalmos?

Patients who have a pale patch in the face, over the area of the facial nerve (N VII) may develop a sudden Type I reaction over the area of the patch. Such reaction usually happens within the first 6 months of MDT, or may already be present at the time of first presentation and diagnosis of leprosy. Reactions may also occur later during MDT, or may recur, be it seldom.

If it develops, the underlying facial nerve may get damaged and orbicularis weakness and paralysis may develop, resulting in lagophthalmos. Depending on the location of the patch, either one side may be affected or, in midfacial patches, both sides.

Red and raised patches in the face should be treated promptly with a course of prednisolone, even before facial nerve damage develops, so that damage can be prevented.

Over time the skin reaction will subside and fade but, if left untreated, the nerve damage will have become permanent.

About 2 – 4 % of the leprosy patients will develop lagophthalmos before diagnosis or during

their course of MDT. 6-10% of the old and disabled leprosy patients have lagophthalmos.

Eye care personnel will usually see patients later, with an established lagophthalmos, after the patches have faded. Patients will usually remember a period of a red and raised skin, before the onset of lagophthalmos, if asked.

Lagophthalmos, often bilateral and with sagging lower lids, may also develop late and gradual in untreated MB disease, as a result of infiltration and secondary atrophy. Corneal sensation is often diminished in such cases.

Note: In leprosy endemic countries, leprosy should always be considered in the differential diagnosis of a 'Bell's palsy' (paralysis of the facial nerve causing drooping/sagging of one side of the face. Patients in general do not mention spontaneously that they are or have been treated for leprosy.

11. Assessment of lagophthalmos



Leprosy patients with lagophthalmos may complain about watering or a 'burning sensation', due to insufficient function of the lacrimal pump. This may be present in one eye only.

Step 1: Observe the frequency and extent of blinking: 'inadequate blinking' can be due either to incomplete eyelid closure in lagophthalmos and/ or abnormal corneal sensitivity

Step 2: Ask the patient to close the eyes 'as in sleep': observe if there is any eyelid gap (≤ 1 mm is normal). Measure or estimate the width of the eyelid gap in mm and note in particular if the cornea is exposed in mild closure. The photographs above show a patient with lagophthalmos and exposure keratitis affecting the left eye, assessed by asking him to close his eyes 'as in sleep'.

Step 3: Ask the patient to close the eyes tightly: in normal orbicularis muscle function the eye cannot be opened against resistance. In this way early orbicularis weakness may be discovered.

Additionally, on straight gaze, there may be sagging of the lower eyelid, with the sclera visible below the cornea, or obvious ectropion. The upper eyelid may show retraction, due to overaction of the levator muscle, as if the patient has hyperthyroidism.

12. Treatment of lagophthalmos

1. Duration of lagophthalmos < 6 months: prednisolone 40mg/day slowly reducing over 12 weeks
2. Duration of lagophthalmos > 6 months with eyelid gap < 6 mm: conservative treatment, e.g. sunglasses, 'think blink'
3. Duration of lagophthalmos > 6 months with eyelid gap \geq 6 mm: eyelid surgery

What is the treatment of lagophthalmos?

Treatment of lagophthalmos is dependent on:

1. Duration of the lagophthalmos.
2. Width of the eyelid gap, and exposure of the cornea.
3. Presence or absence of corneal hypoaesthesia.



Guidelines for treatment:

1. Duration of lagophthalmos < 6 months: systemic steroids as for reversal reaction.
2. Duration of lagophthalmos > 6 months and eyelid gap < 6 mm (without corneal exposure and with intact corneal sensitivity): conservative treatment:
 - a. Protection of the eye: (sun) glasses (see photo), hat or cap with broad rim, covering the face at night time with a clean cloth.
 - b. Blinking exercises to reinforce the orbicularis function: strong blinking exercises three times daily, each time 20 blinks.
 - c. 'Think blink': conscientious and regular strong blinking throughout the day. By making use of the Bell's phenomenon (upward turning of the eyes on attempted closure of the eyelids) the cornea is moistened behind the upper eyelids in a kind of 'reverse blinking'.

Note 1: Under field circumstances the use of artificial tears and eye ointment is less appropriate. Lagophthalmos is a lifelong condition: drugs are expensive and the supply may be irregular. Further, clean storage is not assured.

Note 2: eye lid surgery may of course also be considered for moderate lid gaps for cosmetic reasons.

3. Duration of lagophthalmos > 6 months and eyelid gap \geq 6 mm (with corneal exposure or marked corneal hypaesthesia): eyelid surgery to protect the cornea. Surgery is definitely indicated where exposure keratitis is present. At the other hand, it is often remarkable how long the cornea can remain clear and unaffected in spite of a wide lid gap, as long as corneal sensitivity is intact.

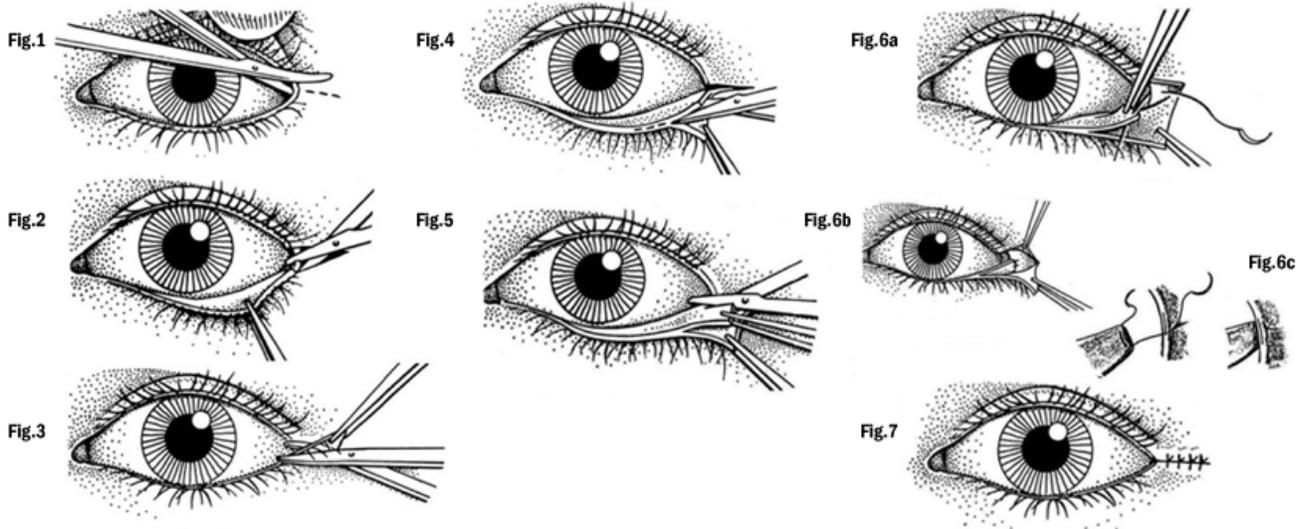
There are many surgical procedures. The simplest procedure is a 'permanent lateral tarsorrhaphy', but this is cosmetically less attractive. The loss of temporal field of vision as result of the tarsorrhaphy is detested by patients. Horizontal lid shortening may be a better option. This will correct ectropion as well. The presently recommended technique is the tarsal strip procedure with or without a medial tarsorrhaphy. In any lagophthalmos surgery the lid

gap should be assessed before and after operation. A lid gap of ≥ 5 mm after surgery is considered as poor result.

Some plastic surgeons advise Temporalis Muscle Transfer (TMT) surgery. In this operation, slips of the temporalis muscle tendon are channelled through the eyelid margins and fixed at the inner medial canthal ligament. In this way the patient can close his eye by chewing. The cosmetic results can be excellent. However, a normal blinking reflex cannot be created and the method is only applicable in cases of good corneal sensitivity and a strongly motivated patient. It requires an expert surgeon and good physiotherapy.

The health worker should provide good health education, seeking to protect the lagophthalmic eye, but must also recognise the indications for surgery and refer as soon as possible. Patients with lagophthalmos require lifelong follow-up.

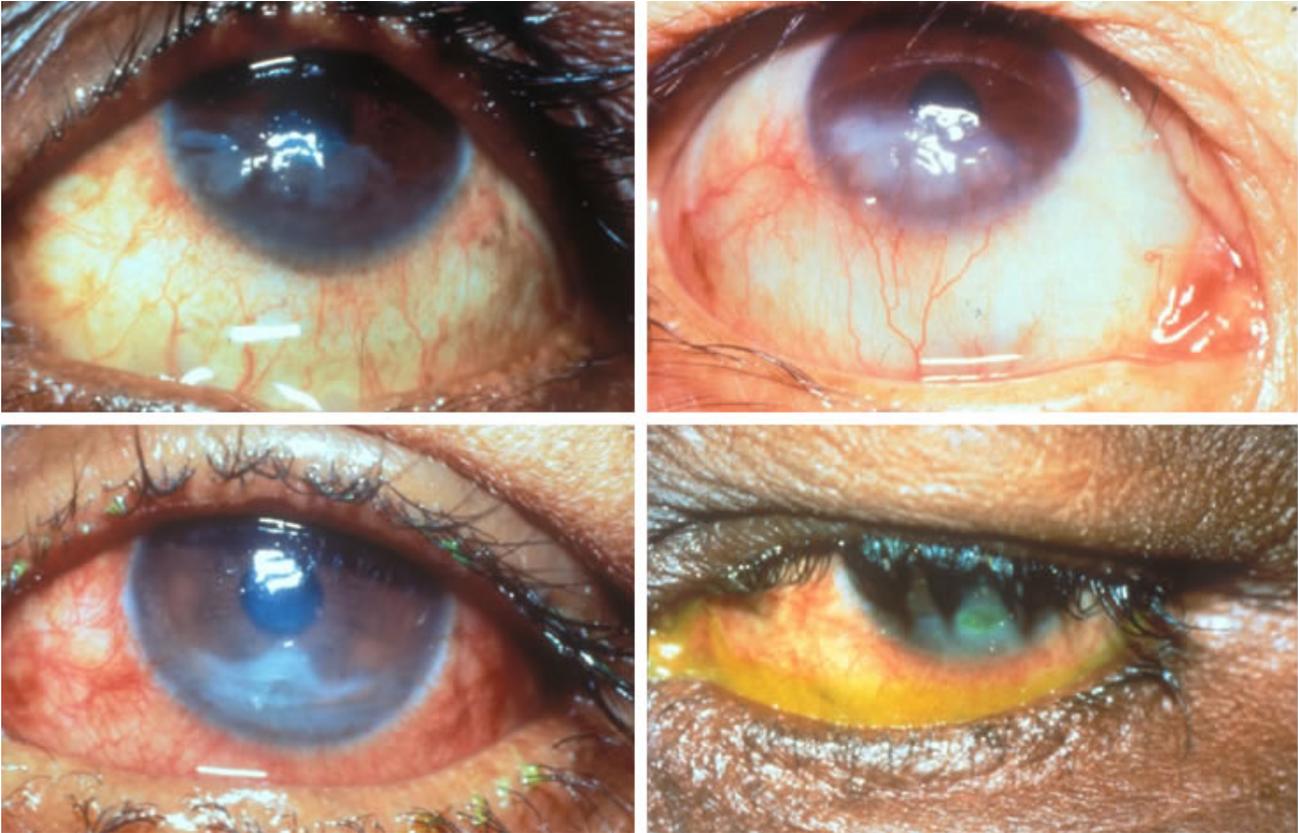
13. Tarsal strip procedure for lagophthalmos



How is the tarsal strip procedure carried out?⁵

1. Inject local anaesthetic (3-5 ml of xylocaine, 1% or 2 %, (with / without epinephrine 1: 100 000) into the lateral part of both upper and lower lids.
2. With scissors, make a cut to separate the upper from the lower lid (a lateral canthotomy). (Fig.1)
3. Direct the scissors downwards and laterally to cut the lower part of the lateral canthal tendon, thus separating the lower lid from its attachment to the bone (the lateral orbital rim). One or two cuts may be necessary to free the lower lid completely. (Fig.2)
4. Pull the lower lid laterally and upwards in the desired position so that the lower lid margin covers the lower part of the cornea by about 1-2 mm. Mild overcorrection is required.
5. Mark the excess skin (a triangular- shaped piece) and cut it off with scissors. (Fig.3)
6. Use the tarsus at the lateral end of the lower lid to fashion a new ligament (lateral tarsal strip). This is done by removing of the overlying skin and muscle, cutting away the lashes and their follicles, and scraping off the conjunctiva on the back surface of the tarsus. (Figures 4 & 5)
7. Suture the newly fashioned strip to the periosteum of the lateral orbital wall using a 5/0 preferably non-absorbable suture. (Figs. 6 a, b, c)
8. Use the same suture to close the layers of skin and muscle making up the new lateral canthus. (Fig.7)
9. Throughout the procedure, bleeding can usually be controlled with pressure. Using epinephrine 1:100,000 mixed with the local anaesthetic will markedly reduce the bleeding. The eye can be patched for 24 hours if needed.

14. Exposure keratitis



What is exposure keratitis and how should it be treated?

Exposure keratitis is damage to the lower, exposed part of the cornea, due to dryness. Initially a superficial punctate keratitis, that stains with fluorescein, may be seen with the slit-lamp. In eyes with lagophthalmos and corneal exposure a deeper corneal defect may develop any time. Such a corneal ulcer may become secondarily infected. This may lead to blindness by scarring or possible perforation.

Chronic exposure may lead to progressive scarring of the lower half of the cornea. The pupil may become obscured by the scar.

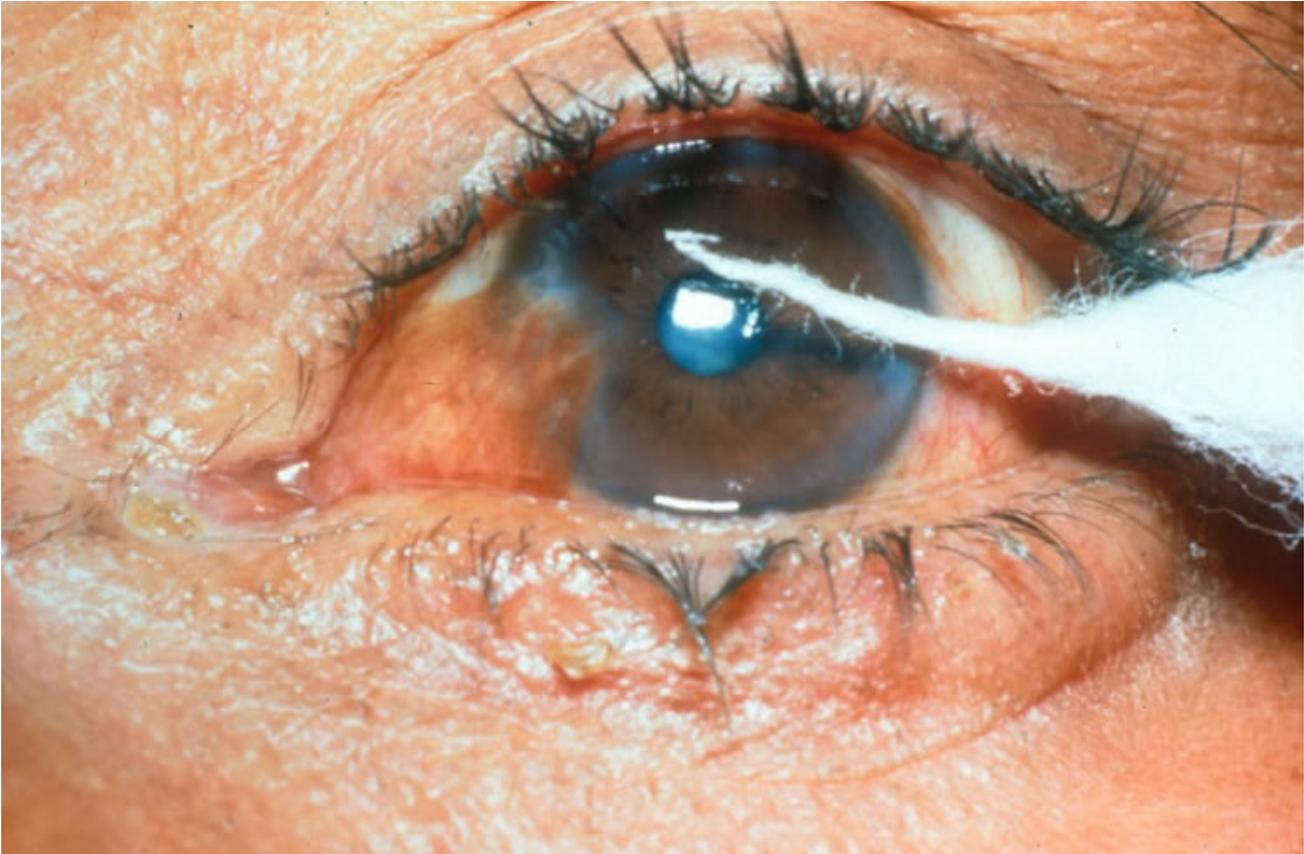
Any eye with significant lagophthalmos is at risk of damage by corneal foreign bodies and other injuries, with a subsequent corneal ulcer and potential scarring.

The photographs show examples of exposure keratitis each with inferior corneal scarring. The eye, bottom right has had fluorescein dye instilled to help demarcate any ulceration.

Treatment of an acute corneal ulcer in exposure keratitis is by antibiotic eye ointment and an eye shield. Do not close an eye which has an exposure ulcer with an eye pad, as the gauze may touch the cornea and cause further damage. It is better to close the eye with a temporary mattress suture.

An exposure ulcer is a definite indication for eyelid surgery.

15. Corneal hypoaesthesia



What are the causes of corneal hypoaesthesia and how important is this condition?

Corneal 'anaesthesia' is usually not as severe as, for example, corneal anaesthesia in herpes and should rather be called corneal 'hypoaesthesia'.

Causes of corneal hypoaesthesia:

1. As a result of reversal reaction in the trigeminal nerve (V cranial nerve).
2. As a result of exposure of the cornea in lagophthalmos.
3. As a result of severe scleritis and damage to the ciliary nerves (often bilateral).
4. As a result of bacterial infiltration and secondary atrophy of ciliary and corneal nerves (often bilateral).

Corneal hypoaesthesia may occur in combination with lagophthalmos, either as a result of a reversal reaction in the trigeminal nerve together with the facial nerve or secondary to exposure. Corneal hypoaesthesia may also occur as a result of scleritis with damage to the ciliary nerves in an ENL reaction and as a late complication of infiltration with the leprosy bacillus and secondary atrophy of ciliary and corneal nerves. In the latter cases it is usually bilateral. These patients will only rarely blink and may develop exposure keratitis without lagophthalmos!

Testing for corneal hypoaesthesia is done by observation of blinking; if regular, there is no reason for testing. If in doubt, or in the case of lagophthalmos, corneal sensitivity is tested with a whisp of clean cotton wool, touching the cornea from the side and observing the blink reflex (see photograph). A more standardized, but laborious, method is with the corneal

aesthesiometer of Crochet and Bonnet.

Corneal hypoaesthesia is dangerous as the patient will not be aware of corneal injuries. There is no cure for corneal hypoaesthesia. Patients should receive good health education: protection of the eyes with glasses or sunglasses, regular blinking exercises and regular inspection of the eyes by the patient and/or relative with immediate medical advice in any case of redness.

Corneal hypoaesthesia may be an indication for early eyelid surgery in lagophthalmos.

16. Type 2 reaction: acute iritis



What are the eye complications associated with an ENL reaction?

1. Acute iritis.
2. Acute episcleritis.
3. Acute scleritis.

What is acute iritis and how is it treated?

Acute iritis

Leprosy related acute iritis occurs only in MB patients and is considered to be evidence of ENL reaction inside the eye. Acute iritis may recur at any time, unrelated to disease activity or systemic ENL reaction.

The clinical signs and symptoms are the same as for acute iritis in general: redness, pain, photophobia, reduced visual acuity and a constricted pupil (photos: left and top right). In severe cases there may be haziness of the cornea and secondary glaucoma. Acute iritis may be unilateral or bilateral.

Treatment is the same as for acute iritis in general: atropine sulphate 1% twice daily, steroid eye drops 6 times daily and steroid ointment at night time. Systemic steroids are not required for acute iritis unless there is a concurring severe ENL reaction which then requires systemic prednisolone. If the iritis cannot be followed by slit-lamp examination, treatment should continue for about 6 weeks with a slowly decreasing dosage of topical steroids.

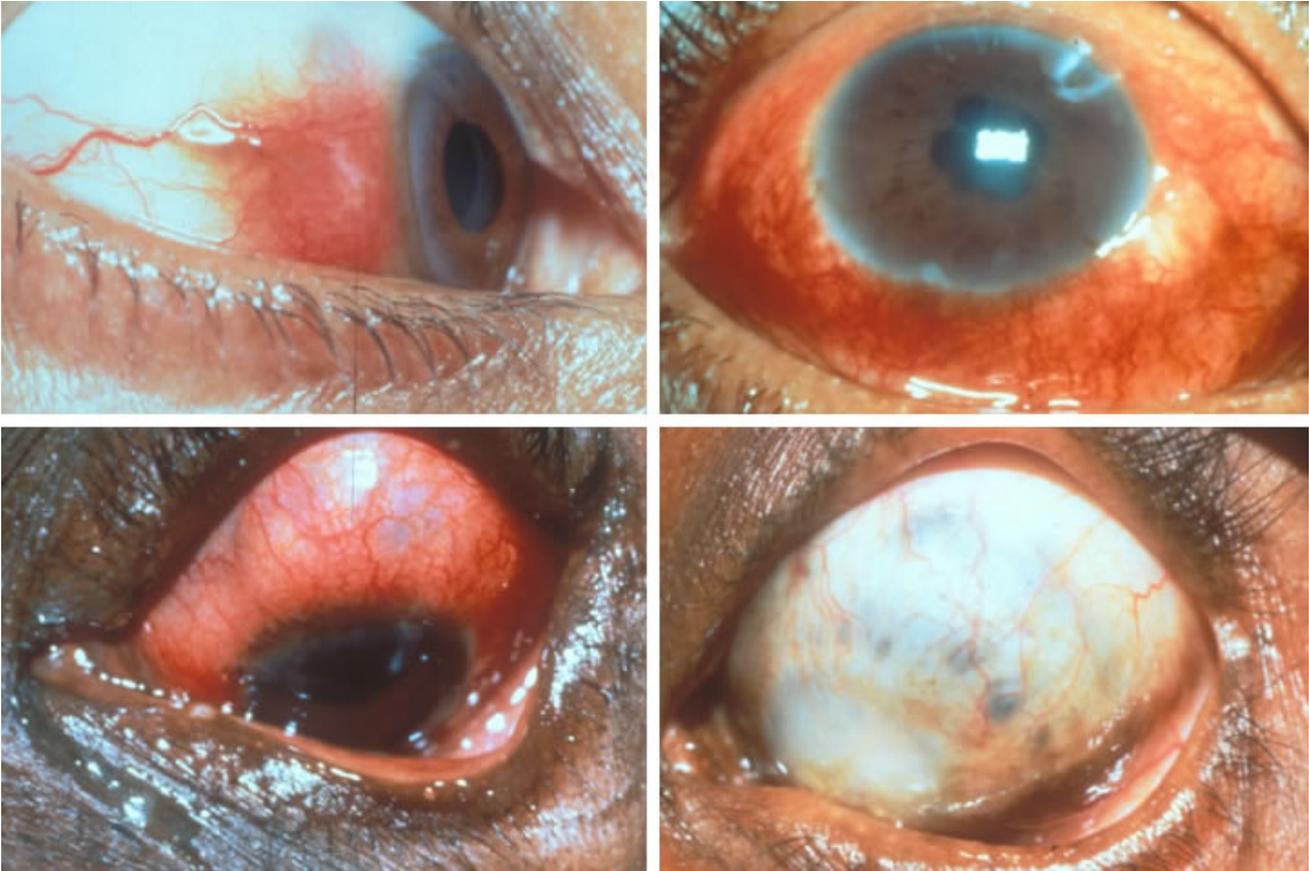
Under field conditions, the diagnosis of acute iritis can be confirmed by using a short acting

mydriatic eye drop, to demonstrate posterior synechiae (photo: bottom right), before instilling atropine.

In the differential diagnosis of acute red eye in a leprosy patient, all other common conditions causing an acute red eye should be considered. These include acute conjunctivitis (where a topical antibiotic may be used), corneal foreign body and injury, corneal ulcer and acute glaucoma. In addition, the use of high doses of clofazimine for ENL reaction may also cause red eyes.

Note: In leprosy endemic countries, leprosy should be considered in the differential diagnosis of causes of acute iritis. In the case of leprosy the patient will usually show signs of MB leprosy, such as (temporal) madarosis of eyebrows, early collapse of the nose or nodules on the ear or may give a history of treatment for leprosy, if asked . Patients usually do not mention spontaneously that they are or have been treated for leprosy.

17. Type 2 reaction: acute episcleritis and scleritis



What are acute episcleritis and scleritis and how are they treated?

Acute episcleritis and scleritis each have a direct relationship with ENL reactions.

Episcleritis

Episcleritis is a transient condition with pain and local redness, often as a precursor of a systemic ENL reaction, and resolves spontaneously and completely. To relieve symptoms and signs topical steroids may be used.

Scleritis

Acute scleritis may be a very serious condition, often bilateral and in combination with a severe ENL reaction. Scleritis may be nodular or diffuse and occurs with or without acute iritis and with or without sclerosing keratitis. In combination with iritis (sclero-uveitis) there may be haziness of the cornea and secondary glaucoma. The photographs show nodular scleritis (top left) and diffuse scleritis with uveitis (top right).

The eye is usually very painful. However, after repeated attacks, the ciliary nerves may become damaged and pain becomes less. After repeated attacks or longstanding severe scleritis, ciliary staphylomas may develop (bottom left), with subsequent scleral translucency and thinning of the sclera (bottom right). In sclerosing keratitis the whole cornea may become opaque.

Acute scleritis is treated topically with steroids (and atropine, in the case of accompanying acute iritis).

Treatment of the ENL reaction will require high doses of systemic steroids and clofazimine. Thalidomide may be necessary (in male patients or in females only if effective anti-conception can be assured, because of the teratogenic side effects on the unborn child).

Note: Subconjunctival injections of steroids are contra-indicated, as this injection may cause further thinning of the sclera. Oral non steroidal anti-inflammatory drugs (NSAID's), such as Ibuprofen (400 mg four times daily), or indomethacin (25 mg three times daily) may be tried systemically. However, be aware of the risk of gastrointestinal haemorrhage in combination with systemic steroids.

18. Massive bacillary infiltration: peri-orbital complications



Which abnormalities occur around and in the eye due to infiltration by bacilli and secondary atrophy?

These lesions occur only in patients with longstanding MB leprosy and may be divided into:

A: Lesions around the eye

B: Lesions within the eye

A. Lesions around the eye

- loss of eyebrows and lashes (madarosis)
- collapse of the nose and secondarily blocked lacrimal sac
- loose skinfold in the upper eyelids (blepharochalasis) and trichiasis

The photograph shows a patient with all these features.

Madarosis of the eyebrows is the result of infiltration of the eyebrows by bacilli and secondary atrophy. It starts on the temporal side of the eyebrows. It is a well known sign of MB leprosy and usually appears after at least 5 years of untreated disease. Cosmetic treatment may be by eyebrow pencil or, in cultures where loss of eyebrows is an important social stigma, by eyebrow transplant.

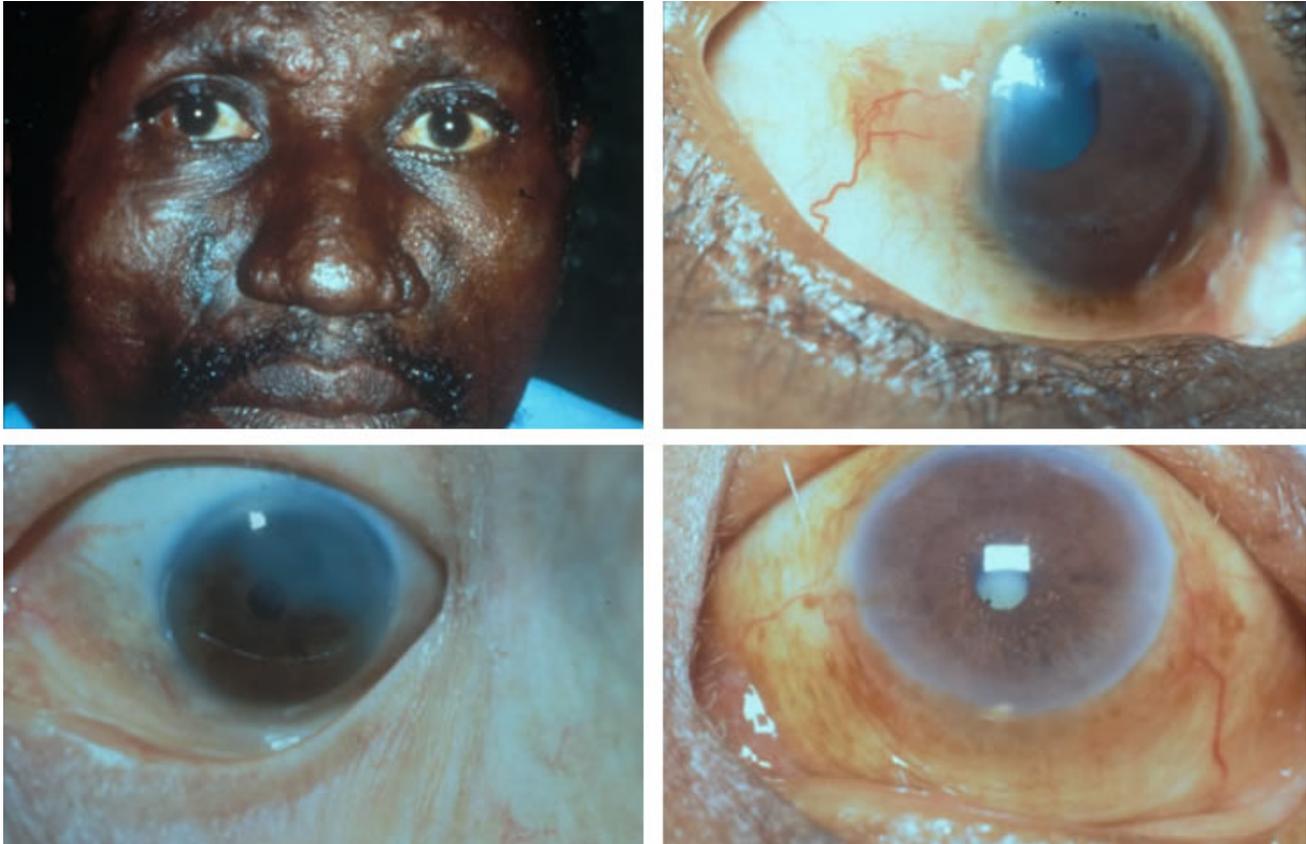
The eyelashes also become atrophic and scanty and finally there may also be complete absence of lashes.

Collapse of the nose is an obvious sign of MB leprosy, caused by a nasal septum defect. It is sometimes surgically corrected. A blocked lacrimal sac may be a complication of a collapsed nose, although this is not frequent.

Dacryocystorhinostomy is contra-indicated in such cases. A dacryocystectomy may be done, or regular massage of the sac if surgery is not available.

Blepharochalasis may lead to trichiasis of the upper eyelid and may need correction. However, the eyelashes are usually scanty and atrophic in such patients and the cornea may often show some hypoaesthesia. The trichiasis may therefore be asymptomatic and be left alone, although this situation must be watched carefully.

19. Massive bacillary infiltration: ocular infiltration



Which abnormalities occur within the eye due to ocular infiltration of bacilli into the eye?

A: Lesions around the eye

B: Lesions within the eye

B. Lesions within the eye

- Ocular leproma
- Superficial lepromatous keratitis and iris pearls
- Iris atrophy and pinpoint pupil
- Chronic iritis
- Atrophy of the ciliary body

M. leprae is believed to enter the eye via the ciliary body.

Lesions within the eye due to infiltration

Ocular leproma is a lepromatous nodule in the eye, usually in the region of the lateral limbus (photo: top right). Also, a ring of nodules may form around the limbus. They arise locally at the limbus or deep in the ciliary body. Lepromas are usually pinkish red or yellowish and fleshy and may resemble a conjunctival tumour. Unlike a nodule in episcleritis, a leproma

is completely painless. The pupil may be deformed and drawn in the direction of the leproma.

Ocular lepromas occur in MB patients treated irregularly by dapsone monotherapy, and are usually a sign of dapsone resistance. The condition has become rare in this era of MDT and is only found in war torn areas, where leprosy control programmes have been disrupted. Treatment is through supervised MDT treatment, on which lepromas will slowly resolve. Displacement of the pupil is permanent.

Some lepromas can become very large and may destroy the whole eye.

Punctate avascular leprous keratitis and iris pearls

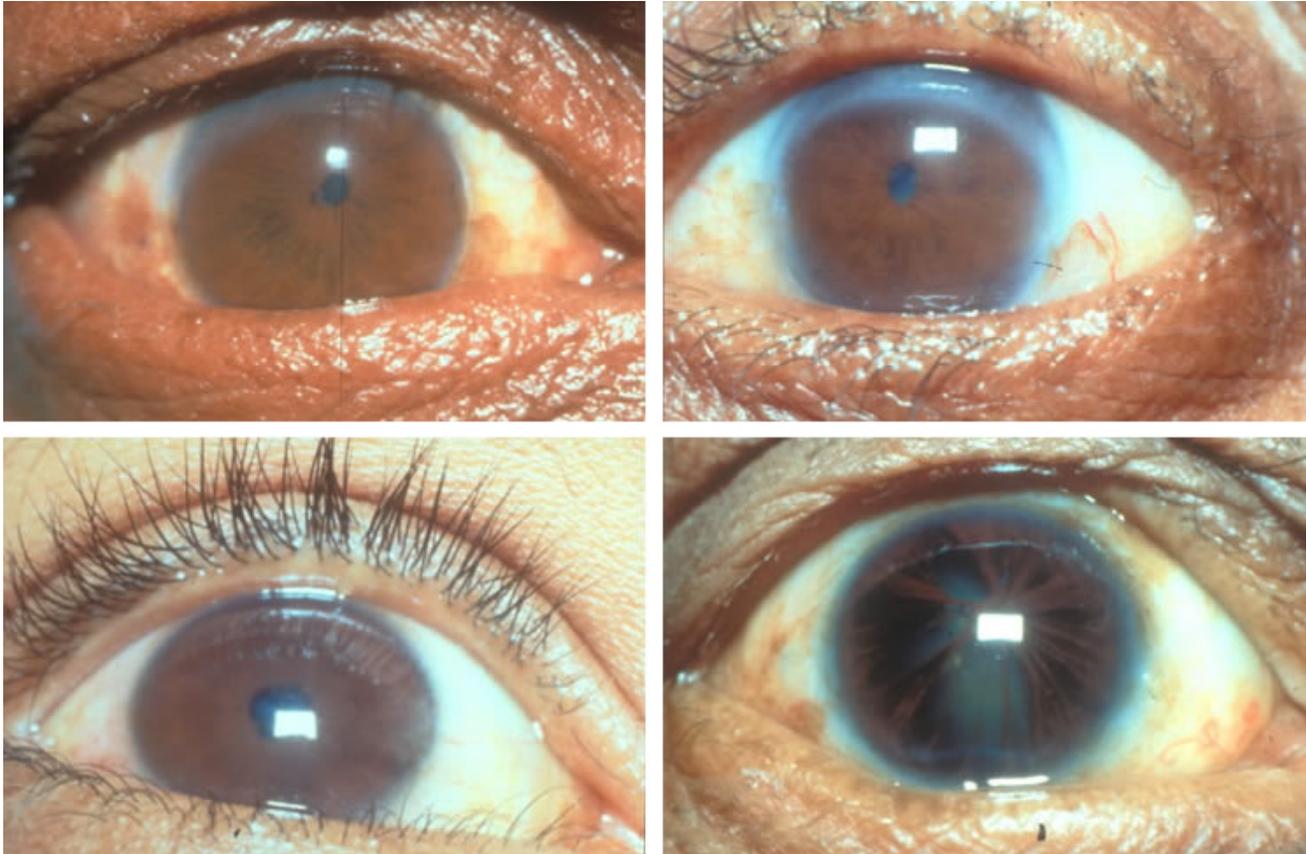
Punctate leprous keratitis appears as faint discrete superficial infiltrates in the upper outer quadrant of the cornea. They consist of clumps of bacilli-laden cells. Later they may become tiny white opacities, and finally coalesce in a diffuse haze (photo: bottom left). The condition is usually bilateral and asymptomatic.

Beading of the corneal nerves may be seen with high slit-lamp magnification and is pathognomonic (diagnostic) for leprosy.

Iris pearls are tiny white clumps (like grains of salt) extruding from the surface of the iris (photo: bottom right). On histology they consist of *M. leprae* cast in chalk and therefore are proof of invasion of the anterior uvea by bacilli. They are completely asymptomatic and are discovered by slit-lamp examination. They remain for a period of time on the surface of the iris and then dislodge into the lower angle of the anterior chamber, where they may be absorbed or give rise to anterior synechiae. Iris pearls are pathognomonic for leprosy.

These types of lesions are only seen in longstanding MB leprosy and have become rare since the wide use of effective MDT treatment.

20. Massive bacillary infiltration: ocular atrophic changes



Which abnormalities occur within the eye due to ocular atrophic changes?

Lesions within the eye due to atrophy

Iris atrophy and pin-point pupil. As a result of iris infiltration, iris atrophy may follow. The iris crypts flatten, the stroma thins and the deep pigmented layer of the iris may become visible. Finally full thickness iris holes may appear. Iris atrophy is usually most prominent in the stroma of the iris dilator muscle. As a result the pupil becomes very small and finally pin-point, and this may occur without posterior synechiae. The condition is usually bilateral and the patient will become 'night blind', as the pupils do not dilate in evening time.

The photographs illustrate these findings. Iris atrophy is particularly marked in the picture shown bottom right.

Particularly in eyes with pin-point pupils, central lens opacities, even if small, will result in very poor visual acuity. Attempts may be made to dilate the pupil with phenylephrine 2.5 - 5% or any other mydriatic, but usually fail. In the case of bilateral blindness a sector iridectomy can be carried out with a small radial cut in the inferior iris sphincter, to prevent updrawing of the pupil. However, due to the friable iris tissue, it is not easy to get a good grasp of the iris. The procedure may be combined with cataract extraction. Iris atrophy and pin-point pupil are typical manifestations of longstanding MB leprosy.

Chronic iritis. Chronic insidious iritis is a well known eye complication in longstanding MB leprosy. There is no redness but the eyes show chronic flare and cells in the anterior chamber, and small keratic precipitates. Greyish exudates may form along the pupillary margin, the pupil constricts and some posterior synechiae may form. Chronic iritis usually persists in spite of treatment with atropine and steroids. There is no relation with ENL reaction and there is often a combination with iris atrophy. Both may be the result of a

'neuroparalytic iritis', due to invasion of the small autonomic nerves in the iris and ciliary body by the bacilli.

Atrophy of the ciliary body. The ciliary body is believed to be the port of entry of *M. leprae* into the eye, from where the bacilli spread into the iris and cornea. Early loss of accommodation has been observed in MB leprosy patients. The intraocular pressure tends to be low in the late stages of MB leprosy, due to atrophy of the ciliary body. In an eye with intraocular pressure of < 8 mm Hg the eye may not recover from cataract surgery. However, chronic iritis with peripheral anterior synechiae formation may block the outflow of aqueous fluid and counteract the low production of aqueous.

21. Leprosy and cataract



How common is cataract in leprosy patients? Is there a need for special precautions in case of cataract surgery?

Cataract is nowadays the single most important cause of blindness in leprosy (see photo). Cataract is responsible for as much as 75% of the new cases of blindness in leprosy.

The causes of cataract are:

1. Age-related (by far the most important cause)
2. Secondary to the use of systemic steroids for reactions.
3. Secondary to acute or chronic iritis.

Secondary cataracts are more common among MB patients.

Precautions to be observed before cataract extraction:

- The patient must be on anti-leprosy treatment or released from treatment (RFT)
- Preferably no reactions should have occurred in the previous 6 months (because the operation might stimulate a reaction and there is more risk for postoperative uveitis)
- No dirty wounds
- A 'clean' eye with eyelids sufficiently functional to protect the cornea. A dacryocystitis should be treated first. Cataractous eyes in MB patients may show either posterior

synechiae and/or poor dilatation of the pupil, often associated with a friable iris, and, in that case, surgery will be more difficult.

Regarding wounds, it should be noted that a blind patient may get into a difficult situation because he or she may commonly get infected wounds. Patients with small and granulating wounds elsewhere, e.g., foot ulcer, should be considered for cataract surgery. Otherwise these patients may never qualify for surgery.

Lens extraction for age-related cataract in a leprosy patient should not be different to any other cataract extraction.

Good results are obtained with modern IOL surgery (including phako emulsification) in leprosy patients and this is the method of choice, if available.

Note: previously operated aphakic patients will still need aphakic glasses and may need replacement of broken or lost glasses.

Some detailed recommendations for cataract surgery in leprosy patients ⁶

1. In case of chronic iritis or occluded pupils a keyhole iridectomy, with or without sphincterotomies will be needed to obtain sufficient dilatation for capsulotomy.
2. Postoperative uveitis is more common in MB patients with active disease or who had uveitis in the past. This requires frequent steroid drops. Cycloplegics should be avoided as these may lead to pupillary capture of the IOL, unless the IOL has been surely placed in the bag.
3. In lagophthalmos with healthy corneas, it may be better to do cataract surgery first, before lid surgery for lagophthalmos. In case of severe lagophthalmos, with ectropion and exposure keratitis a temporary tarsorrhaphia may be done first, until the cornea heals. Thereafter cataract surgery, followed by definite lid surgery a few weeks later.
4. In case of previous Temporalis Muscle Transfer (TMT) surgery it may be necessary to release a tight tendon / graft slip(s) at the lateral canthus, to avoid squeezing during surgery as local anaesthesia will not paralyse the temporalis muscle.
5. For patients with thinned scleras as result of scleritis, a clear cornea incision with buried suture(s) is recommended.
6. After 1-3 years follow-up visual acuity may decrease as result of posterior capsular opacification (PCO) and Nd YAG laser or needle capsulotomy may be indicated.
7. Monitoring of outcome and long time follow-up is recommended.

22. Barriers to cataract surgery for leprosy patients



The photograph shows a field worker explains how a visual chart works to a leprosy patient in Viet Nam.

Why do many leprosy patients with blinding cataract remain without surgery?

- Due to stigma, disabled leprosy patients feel and often are unwelcome at general eye care services
- Leprosy settlements are often at great distance from the eye care facilities
- Many old and disabled patient have difficulties in travelling and lack a guardian
- Many patients are poor and dependent on subsidized surgery
- Funds for subsidized surgery are limited and may only be available for patients with finger count vision (< fc one metre)

Leprosy patients may have difficulty in accessing general eye care facilities for surgery, either due to poverty or stigma. Serious efforts should be made to facilitate access for these patients or to actively include them in outreach activities. In some situations it may be necessary to perform cataract surgery in leprosy settlements or other centres suitable for leprosy patients.

Note: There are still leprosy hospitals catering only for leprosy patients even though this is no longer medically necessary.

23. Examination of the eyes in leprosy

The photograph shows a patient with lagophthalmos and severe exposure keratitis. Note the wide lid gap and sagging of the lower eye lid, with sclera exposure.

How should the eyes be routinely examined?

1. Visual acuity

- in field programmes often recorded by finger counting at 6 metres: 6/60; if available by letter/ E chart at cut-off point of 0.3.

2. Face

- examine for pale and flat or red & raised patches

3. Eyelids

- observe blinking
- test mild and strong eyelid closure
- measure eyelid gap in mm and exposure of the cornea (if any)
- check for eyelid deformities and trichiasis

4. Lacrimal sac

- check for discharge on pressure

5. Check for pericorneal redness: differential diagnosis

- acute red eye
- limbal leproma (dd episcleritis)
- use of high dose of clofazimine (lamprene)

6. Cornea

- examine cornea for exposure keratitis
- test corneal sensitivity with a whisp of cotton wool (if abnormal blinking)

7. Pupil

- check size: normal/< 2 mm
- check shape: regular/irregular
- check position: central/displaced



8. Iris

- observe iris tissue for atrophy and holes

9. Lens: examine for cataract

- pupil black, grey or white; observation of red reflex with ophthalmoscope, if available
- aphakia; pseudophakia; posterior capsular opacification (PCO)

10. Globe

- normal
- hard eye (on palpation)
- soft eye (on palpation)
- staphyloma
- shrunken eye (phthisis) or absent eye

24. Disability grading

This is done in leprosy programmes as an indication of accessibility and quality of the service. It is usually done in three steps, but for eyes only two steps are used:

Disability grade 0:

No eye problems due to leprosy;

No evidence of visual loss ($VA \geq 0.3$)

Disability grade 2:

Eye problems due to leprosy present;

1. severe visual impairment (vision worse than 6/60; unable to count fingers at six metres);
2. lagophthalmos

Other eye conditions are considered difficult to diagnose reliably under field conditions.

Note: Each eye is to be assessed and classified separately. If disability grade 2 is found in the patient due to a cause other than leprosy, this must be recorded. Any patient with a visual acuity of $< 6/60$ should be referred for further examination and possible treatment (depending on programme policy, patients to be referred if VA is < 0.3).

25. Patients most at risk of severe eye complications and blindness



Which leprosy patients are most at risk of developing eye complications leading to severe visual impairment or blindness?

Elderly multiple disabled MB patients with a long history of disease are most at risk of developing severe eye complications and blindness. Many of these patients have been released from treatment and control, after completion of MDT. Those who already had some signs of intraocular involvement at the time of discharge are especially at risk.

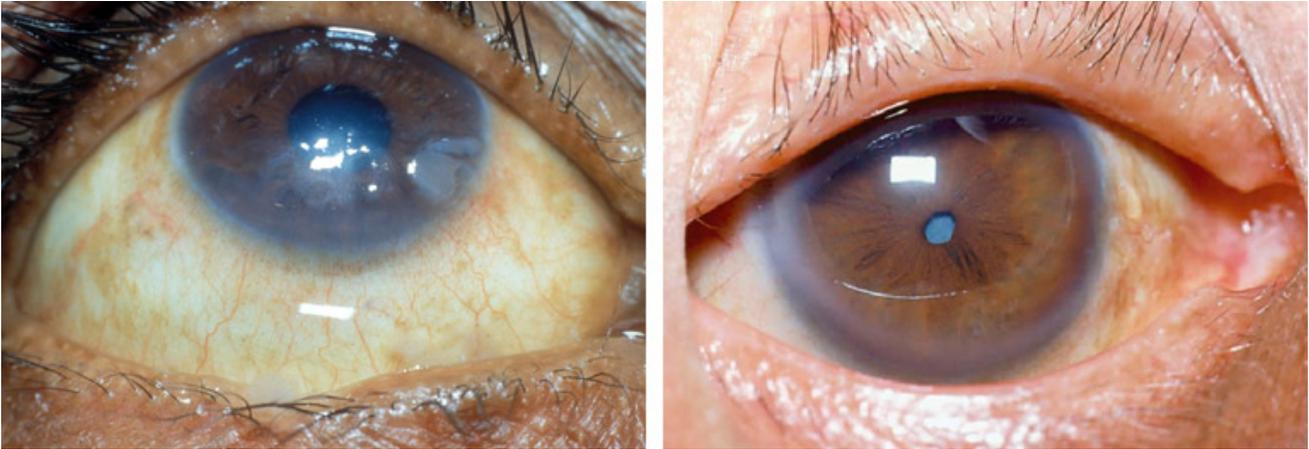
Exposure keratitis, due to lagophthalmos, may progress. Corneal anaesthesia may lead to corneal ulceration due to a neglected foreign body or other injury. Acute iritis may recur long after MDT has been completed. Immature cataracts will progress to blinding cataracts.

Disabled former leprosy patients tend to live together in settlements. Close cooperation with Leprosy Control Services and special programmes to visit and screen persons affected by leprosy (PALs) in such settlements at least once per year are highly recommended.

Some of these patients can greatly gain from reading glasses and aphakic and other spectacles. For some, low vision aids may be helpful.

For those who are incurably blind, due to leprosy, special rehabilitation services should be offered.

26. Prevention of blindness due to leprosy



Can visual impairment and blindness in leprosy be prevented?

Visual impairment and blindness in leprosy may be prevented by:

- Early diagnosis of leprosy and timely MDT treatment
- Early recognition of reactions and effective treatment of reactions with systemic steroids
- Regular eye examination, refraction and treatment of any complications
- Lagophthalmos surgery in patients with a eyelid gap of ≥ 6 mm
- Lens extraction with IOL implantation in any leprosy patients who develop blinding cataract

The photograph on the left shows an eye with exposure keratitis. The picture on the right shows an eye with a mature cataract behind a typical pinpoint pupil.

Good quality, well accepted leprosy services in themselves will cause a decrease in eye complications.

Leprosy services are nowadays mostly integrated in the general health services. Health staff should therefore be well trained in leprosy and in the recognition of eye complications and have at least an antibiotic eye ointment, and possibly short acting mydriatics, readily available.

Eye health care workers, such as eye nurses and ophthalmic assistants, should have good knowledge and interest in leprosy and eye complications in leprosy. Good co-operation between health staff and eye care staff is essential for successful prevention of visual impairment and blindness in leprosy.

The global VISION 2020 programme for the elimination of avoidable blindness, with its emphasis on developing National Prevention of Blindness programmes, offers a great opportunity to include leprosy patients in various programmes, e.g., for cataract surgery, for the correction of refractive errors and the provision of low vision aids.

Outreach programmes should include the leprosy settlements on at least an annual basis.

Remember: EYE CARE in LEPROSY is the responsibility of EYE CARE PROGRAMMES.

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Recommended reading

- Courtright, P. Lewallen S. Prevention of blindness in leprosy, 2nd ed. Available from ICEH as free download, <https://www.iceh.org.uk/x/9Y1V>

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