INTERNATIONAL JOURNAL OF LEPROSY

Volume 69, Number 2 (Suppl.)
Printed in the U.S.A.
(ISSN 0148-916X)

OCULAR LEPROSY

Dr. Margreet Hogeweg
Consultant Ophthalmologist
Netherlands Leprosy Relief
Department of Ophthalmology
Leiden University Medical Centre
P.O. Box 9600
2300 RC Leiden, The Netherlands

This is the first time that ocular leprosy has been included in the series of keynote lectures in an International Leprosy Congress. An overview of the present outstanding issues is presented.

Blindness in leprosy. In 1998, Courtright estimated a total number of 350,000–400,000 blind leprosy patients, including PALs ([1]). This was based on the assumption that 1.5%–2% of the blindness is directly due to leprosy and another 2% due to nonleprosy causes, mainly age-related cataract. Blindness by WHO standards is a visual acuity (VA) of <3/60: "unable to count fingers at a distance of 3 meters," with the better eye. It is not "complete blindness" such as no perception of light. The Indian cut-off point for "blindness" is VA <6/60: "unable to count fingers at 6 meters," with the better eye. The same VA <6/60 is used as the cut-off point for disability grade 2.

Surveys done before 1980, before the introduction of multidrug therapy (MDT), reported eye complications in as much as 50%–90% of the leprosy patients and blindness in up to 50%. It was in such a leprosarium that I, many years ago, personally got interested in ocular leprosy. In very remote places, such percentages may still exist.

In control programs, after implementation of MDT, potentially sight threatening lesions (PST) are reported in 15%–20%; blindness, in 1%–3%. That is about double the level of blindness in the general population, in poor developing countries, of 0.8%–1%.

Surveys on eye complications and blindness in leprosy are prone to methodological problems: 1) Definition of patient: only active cases or including PALs.

2) Location of survey: There are large differences in eye complications and blindness, depending on the location of the study: a) in field programs; b) in leprosy hospitals and ulcer wards; c) in leprosy settlements, where disabled, elderly patients with long history of disease cluster. Eye complications will be comparatively low in field programs, but high in leprosy settlements.

3) Inclusion of eye conditions: a) only PST lesions due to leprosy; b) also nonblinding lesions due to leprosy; c) nonleprosy eye lesions. Example: inclusion of madarosis or not? Other nonblinding nonleprosy conditions? This will make large differences.

4) Prevalence (existing lesions), or incidence (new lesions within a certain period in time). One study ([2]) shows that blind leprosy patients have a 4.8-fold excess risk of dying compared to nonblind leprosy patients of the same age. This is one reason why we see comparatively few blind leprosy patients.

Eye complications. Eye complications are caused by the same mechanisms that cause complications in general in leprosy: a) type 1 reaction: lagophthalmos and corneal anesthesia; b) Type 2 reaction: acute iritis and scleritis; c) infiltration and secondary atrophy: a series of extra- and intra-ocular lesions. The latter two are only seen in multibacillary (MB) patients.

Potentially sight-threatening lesions in leprosy (PST lesions). While studying ocular leprosy, it is important to distinguish between potentially blinding versus nonblinding and less important lesions. Therefore, the term potentially sight-threatening lesions (PST lesions) has been coined.

Lesions such as lagophthalmos and exposure keratitis, corneal hypesthesia, acute

and chronic anterior uveitis (iridocyclitis), and (secondary) cataract are located in the anterior part of the eye, up to the level of the ciliary body and lens. Eye lesions in leprosy are therefore comparatively easy to diagnose with a normal torchlight, cotton wool and a short-acting dilating eye drop to demonstrate posterior synechiae (adhesions between iris and lens) as a result of anterior uveitis.

Visual acuity and patient card. Assessment of visual acuity is the single most important examination in ophthalmology. It should be realized that loss of vision is the same handicap to the patient, whether due to leprosy or to other causes not related to leprosy. Severe visual impairment or blindness may hamper or preclude self-care and is, therefore, more disabling in leprosy patients than in the general population.

Only 56% of the ILEP-supported programs reported to measure VA in a POD survey in 1995 ([3]). One of the problems is that most patient cards actually do not require assessment of visual acuity. For eyes, only facial nerve function and "red eye" are routinely assessed. In order to improve eye care in leprosy, routine assessment of VA, at least at intake and at release from treatment (RFT), should be included. In "care after cure patients" older than 50 years, VA should be assessed annually.

The cut-off point for referral and for disability grade 2 is VA <6/60 (unable to see the upper line on the letter- or E card or unable to count fingers at 6 meters).

Disability grading in eyes (1987–1997). In the WHO reporting system only disability grade 2 is of importance. For eyes, up to 1997, only VA <6/60 was considered disability grade 2. Since VA was not routinely assessed, the result was definitely an underestimation of eye complications in leprosy in the official WHO statistics.

Since 1997 the grading system for eyes has been changed and—in line with the general disability grading system—"visible deformities" of the eye, such as lagophthalmos, iritis and corneal opacity, have been included in grade 2, apart from VA <6/60. This should have led to more reporting of disability grade 2 for eyes and, hopefully, more attention to eye care, but no data regarding this have been published.

Cataract. As a result of increasing life expectancy, age-related cataract has become the most important cause of blindness worldwide. Age-related cataract is also the most important cause of blindness in leprosy nowadays, in particular among PALs. For a cataract-blind patient self-care becomes impossible, as he or she cannot avoid, or take care of, injuries and ulcers. In a recent study from Korea ([4]), cataract was responsible for 87% of the new cases of blindness in an 11-year follow-up study.

Leprosy patients, especially MB patients, have an extra risk of cataract due to the use of systemic steroids for reactions or secondary to iritis. It should be noted that steroid-induced cataract is not a reason to stop steroid treatment for reactions. Cataract can be operated successfully at a later stage, whereas the nerve damage caused by reactions cannot be repaired.

A study from Uganda ([5]) has shown that small pupils, as a result of chronic iritis in MB patients, increase the risk of blindness due to cataract threefold, because even small central lens opacities will greatly influence visual acuity in patients with small pupils.

Poverty and stigma lead to difficulties in access to cataract surgical services. Cataract-blind leprosy patients have therefore less chance of getting surgery than the cataract-blind in general.

Cataract surgery. Apart from the time proven ICCE with spectacles and ECCE with spectacles or artificial lens implantation (IOL), new techniques have recently been developed, such as phako-emulsification and small incision sutureless non-phako surgery, both with IOL implantation. A collapsed nose is an extra factor in favor of IOL surgery because of the lack of support for heavy spectacles. The same applies to severely damaged hands. causing difficulty in handling the spectacles.

Several comparatively small studies have shown that cataract surgery with IOL implantation can give good results in leprosy patients under favorable conditions. In age-related cataract, without intra-ocular complications, there should be no difference in outcome of IOL surgery compared with the general population.

The risk of intra-ocular infection in the case of co-existing lagophthalmos, or in case of ulcers elsewhere, may have increased, but no data are known.

Studies on cataract surgical coverage, barriers to cataract surgery for the patient and outcome of surgery are important to community health interested ophthalmologists and can also be applied to leprosy patients.

Cataract surgical coverage. Cataract surgical coverage is a "measure of service." In this case, it is the cataract surgery actually performed compared to the need for surgery. In other words, operated aphakic or pseudophakic patients (after IOL surgery) in the numerator, divided by the total number of cataract patients, including operated patients, in the denominator.

$$\frac{\text{service}}{\text{need}} = \frac{\text{aphakic and pseudophakic patients}}{\text{cataract blind + aphakic and pseudophakic patients}} \times 100\%$$

For example, if in the study population there are 50 operated patients and no patients with blinding cataract, the cataract surgical coverage is 50/50 = 100%. If, however, there are 20 operated patients and 30 patients with blinding cataract, cataract surgical coverage is 20/(30 + 20) = 20/50 = 40%: 40% of all cataract patients have been operated. The same type of calculation can be applied to other necessary surgery in leprosy.

To assess surgical coverage, a population-based survey is needed. In case of cataract in people older than 50 years, since this age group is most at risk for cataract blindness, such a survey is feasible in particular in a confined catchment area such as a leprosy settlement or, for example, within one's own control program, including RFT patients.

Under very favorable and exceptional circumstances, with a well-equipped eye department and an ophthalmologist especially assigned to a leprosy program, free surgery and short distances to the hospital, cataract surgical coverage was 80% in a study in Korea ([6]). It would be highly interesting to repeat this study in other settings. Most probably, the outcome will be different.

Barriers to cataract surgery. Distance, poverty and stigma play an important role in the access PALs have to cataract surgery.

Outreach or "camp" surgery will provide the lowest barriers to patients, since it is near the patient's home and low-cost or free, but the quality of the outcome should be strictly monitored.

Another barrier is surgical capacity. Although in India "blindness" is defined as VA <6/60, and disability grade 2 for leprosy also as VA <6/60, the criteria for free cataract surgery in outreach or eye camps is often only at the level of a bilateral VA of <1/60 (unable to count fingers at one meter). The patient has to be almost completely blind before qualifying for surgery. By that time, the patient has lost any earning capacity he/she may have had. The reason is that the service can otherwise not cope with the large number of patients.

Certainly, leprosy patients with loss of sensation in hands and feet should be operated earlier. Leprosy programs should establish cooperation with the local eye care services for referral of patients. Charity and service organizations. and in India District Blindness Control Societies (DCBS), can be approached for funding, possibly including IOLs.

Other reasons for not having cataract surgery: a) The leprosy staff may be unaware of poor VA in a patient since VA is not routinely measured and patients do not present because they may think that the leprosy services cannot do anything for eye problems. b) Rural people may not feel the need for cataract surgery at the 6/60 level, but only if VA drops to <3/60. c) There may be medical reasons for not having surgery. However, in case of ulcers it should be emphasized to the eye surgeon that a blind leprosy patient is in a vicious circle: exactly because he is blind, he cannot avoid injuries and ulcers. Such patients will always have ulcers. Cataract blindness in leprosy patients should be restored with priority, even in the presence of clean wounds. d) Finally, patients may be afraid of poor outcome, and the surgery may be more difficult in case of small pupils and previous iritis. Leprosy patients should only be operated by eye services with a good reputation, and the postoperative results should be monitored.

Visual outcome after cataract surgery. Visual outcome in age-related cataract in leprosy patients should be comparable to the outcome in the general population since they are the great majority of cataract-blind eyes.

Visual outcome in case of complicated cataracts, mainly in MB patients, is not well known, and even less known in the case of

IOL surgery in complicated cataract. Several authors have reported more complications during and after surgery, in particular in relation with uveitis and in darker, more pigmented eyes. Small pupils, as may exist in long-standing MB leprosy, will make cataract surgery more difficult because the pupil cannot be properly dilated during surgery.

The outcome of surgery in complicated cases in outreach or camp surgery is not known either. A well-controlled multicenter study is urgently needed on the outcome of IOL surgery in MB patients compared to IOL surgery in the general population, in field circumstances, near to where the patients live, and with sufficient follow up ([6]).

Lagophthalmos. Preliminary results from the "LOSOL study" on incidence of eye complications in leprosy under MDT indicate that all new cases of lagophthalmos appear in the first 6–12 months of MDT, with an overall incidence of about 2%. Alternatively, lagophthalmos is already present at the time of first presentation.

From two studies on patches, reactions and facial nerve damage in paucibacillary (PB) as well as in MB patients ([8, 9]) it becomes clear that almost all lagophthalmos is the result of "significant" facial patches around the eye in type 1 reaction and subsequent damage to the underlying facial nerve. Only a maximum 10% of the borderline patients show such patches.

Lagophthalmos is by and large preventable by the timely use of prednisolone for facial patches in reaction. Lagophthalmos of recent onset, usually with still visible reactive facial patches, can often be improved or cured, and vice versa, patients without facial patches are at almost no risk of developing lagophthalmos.

Lagophthalmos is much less common in lepromatous than in borderline leprosy patients and, in the above study ([9]), was seen almost exclusively in long-standing lepromatous disease. The mechanism of lagophthalmos in lepromatous leprosy is not clear.

For the prevention of lagophthalmos, health workers should concentrate on patients with facial patches. Health education to these patients, careful examination for reaction and early m. orbicularis weakness, with timely prescription of corticosteroids, should prevent or considerably reduce facial nerve damage.

In the POD survey of 1995, 91% of all ILEP-supported programs reported to routinely check lid closure. Contrary to VA testing, this is required on the patient card as a part of routine testing of the motor nerve function However, only 47% offered any lid surgery.

Treatment of lagophthalmos. As in nerve damage elsewhere, prednisolone is most effective in facial nerve damage of less then 6 months' duration.

Cut-off points for conservative treatment versus surgical treatment in lagophthalmos are arbitrary. No long-term prospective studies on the development of exposure keratitis in relation to lid gap have been published. Generally, cut-off points of 5–6 mm lid gap in mild closure are used as an indication for lid surgery. These values more or less coincide with exposure of the lower part of the cornea. However, values ranging from 4 mm to 10 mm have also been mentioned.

Conservative treatment consists of protection by sunglasses, blinking exercises and "think blink." It can be supported by artificial tears. Constraints in conservative treatment include: a) constant watering, due to malfunctioning of the tear pump and laxity of the lower eyelid, or ectropion; b) foreign body feeling because of dryness of the lower part of the cornea; c) any lagophthalmos is a cosmetic blemish; d) sunglasses may break, get lost or be sold; and e) artificial tears may be too costly in the long run and therefore not feasible. However, the eye itself is usually not at risk in lid gaps in mild closure of <5–6 mm, provided corneal sensation is not impaired.

Which is the best surgical method? Remarkably few studies have been done on the effectiveness of lid surgery. Various methods are in use but mostly on small samples with inadequate follow up and not compared to "no surgery." There is no "golden standard" to compare with. The most commonly performed surgical method in lagophthalmos, up to today, is temporal tarsorraphia because it is the simplest.

Different patterns of lagophthalmos with, for example, different grades of laxity of the lower lid, need different types of surgery. Surgery should best be individually geared.

Static lid surgery primarily aims at corneal protection. Tarsorraphia can be a

cosmetic blemish, in particular in unilateral cases. Extensive tarsorraphia causes a troublesome loss of temporal field of vision for the patient. Other techniques include various wedge excisions of the lower eyelid, medial tarsorraphia and tarsal strip procedures. Also gold weights in the upper lid are used in order to narrow the lid gap and enhance orbicularis function, but this is usually too costly. Often multiple procedures are necessary. Even so, a considerable residual lagophthalmos may remain.

In dynamic lagophthalmos surgery, a well done temporalis muscle transfer (TMT) can give excellent cosmetic results. Patients can blink, although usually no spontaneous blink habit develops. To be successful, it needs an excellent surgeon, a well-motivated patient and, preferably, good corneal sensitivity. The main disadvantages are the level of surgical skills required, often by a plastic and reconstructive surgeon, the intensive physiotherapy needed, rather long admissions and the potential for complications, such as entropion, ectropion or narrowing of the lid gap.

Lagophthalmos surgical coverage. For lagophthalmos also surgical coverage can be assessed: lagophthalmos patients operated with satisfactory result, divided by patients in need of lagophthalmos surgery (or in need of repeat surgery), plus the successfully operated patients.

In the Korean study ([10]) with the same favorable conditions as for the cataract surgery, the surgical coverage was 57%, if calculated with "any patients ever operated for lagophthalmos" as the numerator. Surgical coverage was only 26%, if taken into account that 53% of the operated patients still had a residual lid gap of >5 mm and, therefore, in fact still needed additional surgery.

Outcome of lagophthalmos surgery. As mentioned, 53% of the operated patients in the Korean study still had a >5-mm lid gap after lagophthalmos surgery through various methods, showing how rather unsatisfactory the result of lid surgery often is; 28% were not satisfied with the result themselves and 20% would not recommend lagophthalmos surgery to others.

It would be interesting to design such outcome studies, based on residual lid gap, elsewhere. Problems in cross-sectional population-based studies of outcome of lagophthalmos surgery include: a) no data on the surgical method available; b) no data on presence of exposure keratitis at time of initial surgery; and c) no data on visual acuity at the time of initial surgery. To answer these problems, long-term prospective studies on well-documented patients, including surgical technique, are needed and highly recommended.

The barriers to lagophthalmos surgery in the Korean study were: no knowledge about the possibility of surgery, costs, distance, service perceived as poor quality, and no need for surgery felt.

Recommendations for lagophthalmos surgery are: a) semi-standardized criteria for selection of patients for lid surgery for health workers are needed, such as mm lid gap in mild closure or presence of exposure keratitis; b) success of surgery to be monitored on residual lid gap; and c) each leprosy program should offer lagophthalmos surgery as part of the POD program, either themselves or through the ophthalmic services.

ENL, clofazimine and uveitis. It is a definite clinical impression that acute iritis and acute scleritis are less common since the introduction of MDT. This is attributed to the routine use of clofazimine which has led to a reduction in the frequency of erythema nodosum leprosum (ENL) reactions as well. Binding bilateral scleritis, with secondary glaucoma, in recurrent severe ENL used to be among the most important causes of blindness in leprosy. Nowadays this has become a rare complication.

Uveal disease, with keratic precipitates, cells and flare, and pupil shape abnormalities, continues in MB patients, in spite of MDT. According to preliminary LOSOL findings, cumulative incidence of any uveitis is about 5% at the end of MDT and increases to about 13% 2 years after RFT.

Corneal hypesthesia or anesthesia. Corneal hypesthesia is difficult to quantify unless measured by the Cochet-Bonnet monofilament esthesiometer. It may be overdiagnosed at times or confused with incomplete blinking, as in the case of lagophthalmos. It can sometimes be seen in lepromatous patients with a long history of disease. In that case it is probably the result of infiltration and secondary atrophy of the corneal and ciliary nerves and comparable to glove and stocking anesthesia. In these

cases there can be severe bilateral corneal hypesthesia without lagophthalmos.

In combination with lagophthalmos, one can imagine two mechanisms: direct damage to the trigeminal nerve at the time of facial nerve damage, or indirect damage as a result of long-standing exposure. No studies have been published on corneal hypesthesia occurring in a case with a reactive facial patch or in recent lagophthalmos. It is not known if corneal hypesthesia may recover by systemic steroids.

Progression of eye disease. The final results of the LOSOL study on incidence of eye complications and long-term outcome in MDT are still to be published.

In another study ([2]), progression of eye disease over a period of 11 years in RFT patients initially free of eye involvement happened in 14.7%, either as keratitis, synechiae or lagophthalmos. In addition, from those initially free of cataract, 5.7% developed bilateral blinding cataract. Of the incident cases of new blindness, 87% was due to cataract. It is thus important that "care after cure" disabled leprosy patients receive routine eye examinations, including visual acuity, at least once a year.

Research priorities. Research priorities should include: a) Cataract: studies on surgical coverage, outcome with / without IOL, and barriers to cataract surgery for leprosy patients in different settings are highly recommended. Lagophthalmos: studies are needed on indications and cut-off points for surgery, best technique, and long-term outcome, in combination with lagophthalmos surgical coverage, and barriers to lid surgery. c) Operational research: studies on best implementation of eye care in POD activities and integration of eye care for leprosy patients into the general eye care services.

Recommendations.

- The leprosy services should be the "watch dogs" for any sight-threatening eye disease.
- Facial patches in reaction and recent lagophthalmos should be treated with a course of prednisolone.
- Each leprosy program should have a collaborative agreement with a nearby eye care service for referral of patients who need specialist help, in particular for surgery.
- A policy giving priority to leprosy patients for cataract surgery should be developed in collaboration with the local eye care services.
- Eye care services and leprosy services in collaboration should provide training in eye care in leprosy to leprosy staff as well as to eye care staff and, together, should provide guidelines for treatment.

REFERENCES

1. COURTRIGHT, P. and LEWALLEN, S. Ocular manifestations of leprosy. In: *The Epidemiology of Eye Disease*. Johnson, Minassian & Weale, ISBN 0 412 64310 3 (HB), Chapman & Hall Medical.
2. COURTRIGHT, P., KIM, S. H., LEE, H. S., *ET AL.* Excess mortality associated with blindness in leprosy patients in Korea. Lepr. Rev. **68** (1997) 326–330.
3. ILEP MEDICAL COMMISSION. ILEP Medical Commission Survey: Prevention of disability in projects sponsored by ILEP members. London: ILEP, 1995.
4. LEWALLEN S., TUNGPAKORN, N. C., KIM, S. H. and COURTRIGHT P. Progression of eye disease in 'cured' leprosy patients: implications for understanding the pathophysiology of ocular disease and for addressing eyecare needs. Br. J. Ophthalmol. **84** (2000) 817–821.
5. Waddel, K. and SAUNDERSON, P. R. Is leprosy blindness avoidable? The effect of disease type, duration and treatment on eye damage from leprosy in Uganda. Br. J. Ophthalmol. **79** (1995) 250–256.
6. COURTRIGHT, P., LEWALLEN, S., TUNGPAKORN, N., CHO, B.-H., *ET AL.* Cataract in leprosy patients: cataract surgical coverage, barriers to acceptance of surgery and outcome of surgery in a population-based survey in Korea (submitted for publication).
7. WADDELL, K. Intra-ocular lens implantation for cataract following leprosy. Lepr. Rev. **70** (1999) 74–75.
8. HOGEWEG, M., KIRAN, K. U. and SUNEETHA, S. The significance of facial patches and type 1 reaction for the development of facial nerve damage in leprosy; a retrospective study among 1226 paucibacillary leprosy patients Lepr. Rev. **62** (1991) 143–149.
9. HOGEWEG, M., MUZZAFFARULLAH, S., REDDY, R. G. and SUNEETHA, S. Facial patches, type 1 reaction and facial nerve damage: a retrospective study among 1178 MB leprosy patients. Oral presentation at Asian Leprosy Congress, Agra, India, 2000.
10. COURTRIGHT, P., LEWALLEN, S., TUNGPAKORN, N., *ET AL.* Lagophthalmos surgery in leprosy: findings from a population based study in Korea (submitted for publication).