EDITIORIAL

Children with leprosy

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Introduction and epidemiology

It is a sad thing to see a young child with irreversible nerve impairment due to leprosy. We must do all we can to prevent leprosy in children and to prevent disability in leprosy-affected children. However, it is important to be confident of the diagnosis of leprosy before giving multi-drug therapy to avoid over-diagnosis and unnecessary treatment.

Leprosy in children (under 15 years old) is still common in countries where leprosy continues to be endemic. The global figures for 2012 show 21,349 new child cases, 9% of all new cases,\(^1\) with 76-5% of these residing in the South-East Asia region. The proportion of new cases which are in children varies between individual countries from 0-6% in Argentina to 41-3% in Micronesia. In India, 10 states have child proportions of over 10%, while in Daman and Diu it was 30%.\(^2\) Studies from well documented hospital series\(^3\)--\(^5\) have reported 4·5–14% of cases to be in children. Active population surveys give much higher proportions, for example 35% in Maharashtra and 32·5% in Agra.\(^6\),\(^7\) The number of child cases has decreased in line with a general reduction in case detection, but there is not necessarily a reduction in the proportion of child cases amongst new cases.\(^8\),\(^9\) The time has come to change the standard indicator (proportion of child cases amongst new cases\(^10\)) and in future to express the burden of child cases as an age-specific rate – the number of cases per 100,000 children under 15 years.

A recent study from Cebu shows that the age-specific new case detection rate in those under 15 years declined from around four cases per 100,000 to just under two per 100,000 between 2000 and 2011, while the proportion of child cases remained the same, at 11%. This study also showed that the mean age of children who developed leprosy remained static over the same period. The case detection rate in children in Norway fell from around 15 per 100,000 in 1851, to under two per 100,000 by 1890 and to less than 0·1 per 100,000 by 1920.\(^11\)

The age specific incidence of leprosy varies from place to place but it has been observed to have two peaks, in under 14s and in older adults\(^12\) and age is a risk factor independent of other factors. An increase in mean age at diagnosis has been observed,\(^13\) and was noted both in Japan over the period 1964-2008,\(^14\) and in China,\(^15\) coinciding with a fall in prevalence rate.

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Diagnosis and treatment

Children are more likely to present with paucibacillary (PB) leprosy than are adults. In Burma, amongst 610 cases enrolled for the BCG trial by Bechelli, 86% had a single skin lesion, (65% Tuberculoid, 35% Indeterminate) and there were no lepromatous cases. Diagnosis may be hampered by the inability of the child to cooperate with sensory testing. Presenting the test as a game with a feather is one useful technique, another is to demonstrate on the mother’s skin as he watches then ask the child to copy his mother. The differential diagnosis may be helped by biopsy (an undesirable procedure in the very young child) – in one study 35% of doubtful cases could be confirmed by histology. There are several reports from the pre-treatment era of self-healing of skin lesions in children. In a leprosarium in the Philippines, of 2000 children examined, 470 had symptoms of leprosy and amongst those with 6 years or more of follow up while untreated, 75% subsided. In Africa, a study of the natural history of leprosy revealed that, if observed over 2 years, a proportion of untreated lesions apparently self-healed: overall self-healing was seen in 33% of cases but in the 0–19 years age group, only 12% of untreated cases self-healed. In children with diagnostic signs it is usual to treat with MDT rather than to wait and observe, but in view of the uncertainty about prognosis, this may represent over treatment. In some countries, the risk from dapsone hypersensitivity syndrome or other adverse effects of MDT may be 2% and this may be a significant factor in deciding not to treat an early lesion when there is a chance of self-healing in this age group. In doubtful cases it is usually safe and acceptable to keep the child under observation (untreated) for a period of time. When re-examined after an interval the lesion may show more definitive features and the child may be more cooperative. With frightened young children one needs skill and patience for clinical examination; engaging the mother to do the body charting or even to do the sensory testing may be desirable. If a skin smear is considered essential it should be the last activity before the child leaves the clinic. The patient’s weight should always be recorded and used to determine the correct dosage of MDT, bearing in mind that in some endemic countries the standard ‘child pack’ contains doses too high for the majority of child cases. One aims for doses close to 1 mg/kg of rifampicin and 1–2 mg/day of dapsone.

Should a child have to discontinue dapsone after developing the hypersensitivity syndrome, the standard alternative therapy, as for adults, is a combination of clofazimine and monthly rifampicin, but one option for PB cases would be single dose rifampicin, ofloxacin and minocycline (ROM). There is a dearth of evidence for safety and efficacy of monthly ROM for children with MB leprosy.

Compliance with drug therapy in children requires a level of understanding and involvement of the carer. A study in West Bengal identified the need to empower parents of children affected by leprosy so that they could become knowledgeable and active partners in the successful management of the disease and its complications.

Prevention of disability

Prevention of impairment and disability (POID) needs special attention in children because of the life time impact of leprosy-related disability. Reconstructive surgery for those with established impairment is feasible even in young patients. The outcome of surgery for ulnar paralysis in 79 children aged 7–14 years is reported as satisfactory, and undue delay may
compromise the chance of a successful outcome. Too many children already have disability at diagnosis, although the proportion has decreased over time in some areas. Delay in diagnosis may contribute to the occurrence of disability and the reasons for this delay may differ in children compared with adults. The occurrence of reactions or neuritis is reported from 20–30% of child cases and is commonly the reason for initial presentation, but little has been published on the frequency, management or results of treatment of reaction or neuritis in children. Similarly, evidence is lacking on the specific difficulties of teaching self-care to children and their carers. When a child presents with new nerve function impairment or overt reaction, his/her body weight (not only age) should be used to decide on steroid dosage. One must be mindful of the specific risks of steroids in growing children as well as the well-known adverse effects which can occur at any age. For ENL reactions clofazimine (in appropriate doses) is well tolerated by children. Hospitalisation should be avoided if possible, but may be unavoidable for short periods, for example, if there is a nerve abscess needing incision and drainage.

Long-term studies of childhood leprosy, using tools such as the P-scale or a Quality of Life scale, to assess the impact on psycho-social functioning in adult life of a childhood affected by leprosy are needed.

Public health issues

Congenital leprosy appears to be rare, if it ever genuinely occurs, and transmission through breast milk may be possible. Rare reports of leprosy in infants (including cases confirmed by histology) are mostly from the pre-MDT era including a 3-week old child from Martinique. Leaving aside these unusual infant cases, a relatively high rate of leprosy amongst children is thought to indicate continuing transmission in the community. The source of infection in children is likely to be within the household since young children mix with fewer individuals outside the home compared with adults. Amongst child cases, a contact case was found 18-87% cases, and it was usually a family member. In Japan, this decreased over time. In the pre-MDT era, the higher attack rate within households of known cases, compared with general population, was recognised and it was noted that “apparent clustering of cases in close relatives could be explained by more intimate contact” although it is now recognised that the risk in blood relatives is higher than in not-related people living in the same household. Hence household contact surveys should be an especially effective way of case finding when the index case is a child. Conversely, children in a household where a new leprosy case has been diagnosed are at increased risk compared with the general population and even compared with adults in the household: the risk to children aged 1–14 years was much higher than in older people. However, even before MDT was available, only a small proportion of children exposed at home developed leprosy: 6% in the study in Culion.

If transmission to children is still occurring, largely from undiagnosed cases in the community, can it be interrupted? Can progression to leprosy disease be prevented in those contacts more exposed to infection or even in those already infected, but currently in a subclinical state? The COLEP trial in Bangladesh indicated that over the first 2 years a single dose of rifampicin given to close contacts, soon after the index case began taking MDT, reduced clinical leprosy by 57%. This effect was greater for subjects aged 10–14 and 20–29 years old, and in those who had received BCG in the past. Whether it is feasible and
cost-effective to offer such chemoprophylaxis to whole communities or only to close contacts needs investigation.\textsuperscript{32,33,34,36}

Enhanced immunity through vaccination with BCG alone may reduce the incidence of leprosy in the community: this was proposed over 60 years ago\textsuperscript{34} and the evidence base for this has expanded recently.\textsuperscript{35} BCG vaccination in infants is the policy in most leprosy–endemic countries (as part of tuberculosis control programmes) and might have an effect for 5–10 years, after which it wanes.\textsuperscript{36} Whether a second dose sustains the effect for longer is as yet uncertain:\textsuperscript{37} a trial is in progress comparing BCG alone versus combined BCG vaccination and single dose rifampicin for contacts of newly diagnosed leprosy cases but this trial excludes children under 5 years old.\textsuperscript{38}

Leprosy control programmes need to monitor the number of children (and their ages) being detected and to consider in each case the likely source of infection; ensuring household contact surveys (or, preferably ‘extended contact surveys’ which include near neighbours) are carefully conducted. The clinician’s response to a new child case should include not only prescribing MDT at appropriate doses, but also a careful assessment for existing nerve function impairment and risk of future impairment, and an assessment of the child’s and the family’s ability to respond to the diagnosis in a way that minimises the psychological impact and maximises successful self-care. The feasibility of routine provision of prophylaxis (chemo- or immune- or both) for healthy children in households of each newly diagnosed adult leprosy case needs further research.

Conclusion

In summary, child cases continue to present in substantial numbers and it is suggested that a new indicator be used, number of new cases of leprosy in children per 100,000 children. There are major diagnostic challenges when assessing a child with suggestive signs of leprosy, and if there is any doubt it is generally safer to keep the child under observation (maybe 2–3 months), then re-examine in the most favourable circumstances possible. For every confirmed new case, careful and sympathetic teaching of the parents is essential. This includes the risks of MDT, signs of reaction, and other topics (such as care of sensory-impaired limbs) according to the features of the individual case. In relation to each new child case, as for adult patients, household contact examinations need to be arranged. In future it may be appropriate for programme managers to introduce chemo/immuno-prophylaxis for those at greatest risk of contracting leprosy.

References
