Childhood leprosy: giving an identity to a name

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The chief guest Dr. Dieter Rienel, Guest of honour Dr. Mrs. Ganga Sirimanne, the president of Sri Lanka College of Dermatologists Dr. Pushpa Karunasekara, Past presidents, Madam Vinitha Perera, members of the family of late Dr. W D H Perera, members of the council and members of the Sri Lanka College of Dermatologists, visiting members of the German Dermatological Society and Berlin Foundation for Dermatology, distinguished guests, ladies and gentlemen, It is a great honour and a privilege for me to deliver Dr. W D H Perera memorial oration this year and I thank the president and the council for according me this great honour.

Late Dr. W D H Perera had his education at Ananda College Colombo, where he excelled both in sports and studies. The picture shows him greeting late Mr. Javaharalal Neru, the first Prime minister of India. Subsequently he entered, and graduated from the University of Peradeniya. While at Peradeniya he met his future wife Dr. Vinitha Perera.

Having obtained his MRCP UK he returned to Sri Lanka and was appointed as consultant Dermatologist to General Hospital Ratnapura. Late Dr. W D H Perera was instrumental in developing dermatology in to a well recognized and respected specialty in Sri Lanka. He also played a key role in establishing South Asian Regional Association of Dermatologists. For this work he was awarded a life time achievement award posthumously, by the Prime minister of Nepal.

His longstanding friendship with late Dr. Gunter Shwenzer, paved the way for the long standing co-operation that exists between German Dermatological Society and Sri Lanka College of Dermatologists.

He was a person with many talents. His bird photography is a testimony for this.

I had the privilege of learning my ABC of dermatology under this great clinician. I consider him to be my mentor. I completed my postgraduate training with Dr. Ganga Sirimanne, an eminent dermatologist and a great, compassionate teacher. Both my teachers instilled in me a special interest in leprosy. Having gained invaluable knowledge in clinical dermatology locally I proceeded to Australia and was attached to the Department of Dermatology, University of Melbourne. While there, I trained under the supervision of Prof. Robin Marks and Prof. Rodney Sinclair.

On my return to Sri Lanka in early 2006, I was appointed as Dermatologist to the historic city of Anuradhapura. In the following year I was posted to Chilaw General Hospital, and subsequently to Ratnapura Provincial General Hospital. I continued to maintain my interest in leprosy at all these stations. Let me now show you some clinical cases that I have seen in these places.

(Borderline Tuberculoid Leprosy with a satellite lesion: Borderline Tuberculoid Leprosy with Lagophthalmos: Leonine Face of Lepromatous Leprosy)

There were even rare presentations like Histoid Leprosy.

Some unfortunate patients presented late with advanced disease having neurological disabilities. However majority of these patients were adults.

In 2012, I was appointed as a consultant dermatologist to the Lady Ridgeway Hospital for Children Colombo, the tertiary centre for paediatrics in Sri Lanka. Here, I was exposed to a wide array of unique childhood dermatoses, as you can see.

At the commencement, I was not familiar with the true picture of leprosy in children. Though rare in children, I still continued to maintain my interest in leprosy.

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Let me now take you through my experience and try to give you a clear picture of childhood leprosy in Sri Lanka, hence the title "Childhood leprosy: giving an identity to a name".

My presentation is based on my work in the leprosy study group of Lady Ridgeway Hospital for Children. The Study was initiated by Prof. Jayamini Seneviratne in 2001.

Let me quickly go through few basic aspects of this disease, which is caused by *Mycobacterium leprae.* Here I have outlined the mechanisms by which *mycobacterium leprae* causes damage.

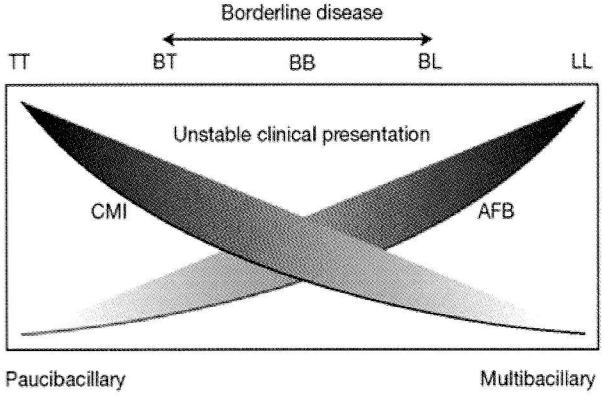
Direct infiltration and invasion.

Immunological mechanisms.

Peripheral nerve damage.

Psychological impact.

The Ridley Jopling classification of the disease which has withstood the test of time, is shown here.



Ridley Jopling classification

As you realize, host cell mediated immune responses determine the clinical spectrum of the disease. If the cell mediated immune responses are robust, patients develop tuberculoid leprosy and poor immune responses lead to dissemination of the organism. The latter is known as multibacillary or lepromatous leprosy.

Leprosy still is an important disease in the developing countries.

Let me now tell you few known facts about childhood leprosy. It is well known that majority of children who are exposed to the organism do not develop established disease. A fair number of early lesions also heal spontaneously. However in a minority, the disease will progress to an established form of the disease. It is important to note that leprosy in children has many health implications. It is indicative of both ongoing transmission in the population and the possibility of an undetected household contact. A diagnosis of leprosy still carries a considerable amount of psychological distress and stigma. Permanent peripheral nerve damage is a lifelong complication.

The child rate continues to trouble health care planners including the WHO (Globally 9.1%). These are some statistics relevant to Sri Lanka. Both adult and child new case detection rates continue in a stable manner over the years. This has cast a doubt about efficacy of Multi Drug therapy in preventing the transmission of the disease. As you know it is 32 years since Multi Drug Therapy was introduced in Sri Lanka. This graph highlights the point I raised earlier on.

At our clinic also, a similar observation is made. Approximately 15% of all children with leprosy are managed at the Children's Hospital.

We have studied 443 children with leprosy since 2001. Some of the patients are still been followed up after completion of treatment. However, as you can see leprosy remains a statistically insignificant entity at our clinic. (0.21% of all new cases) We studied all aspects of the disease. Both Ridley-Jopling and WHO classifications were used. A properly conducted contact examination has been carried out way back in 2007. The details of which, I will be showing later.

A simple slit skin smear was performed whenever indicated. A positive result classifies a patient as multibacillary disease irrespective of the number of skin lesions.

There was no sex preponderance. Majority were school children (84%). This is probably because of the long incubation period and possible cross protection from BCG vaccination which in Sri Lanka is given at birth. At presentation majority (67%) had single lesion leprosy. Deformities were rare (1%) compared to adults.

Single lesion leprosy could be due to indeterminate, tuberculoid or borderline tuberculoid types.

The presence of a thickened nerve in a suspected case, is strong evidence of the disease. This is especially important in children, as sensory testing is a difficult task in them. Attempts are being made to use a feather instead of a sharp object for sensory testing in children (Figure 1).

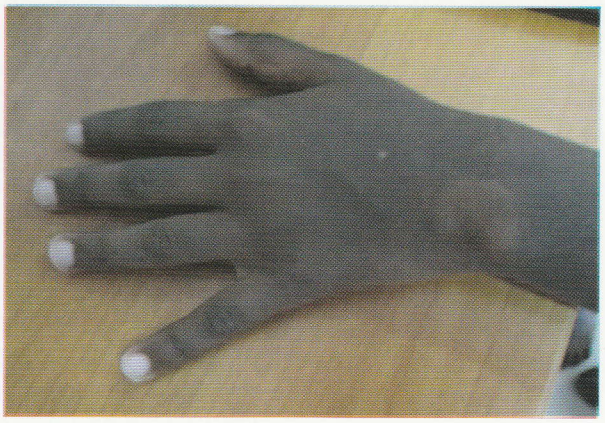


Figure 1. *Thickened nerve in the perilesional area.*

Here is a maculoanaesthetic lesion with Dactylitis and wasting of small muscles. The ulcer on the index finger is indicative of sensory impairment. Often leprous neuropathy is of mixed sensory motor type. You can also appreciate the fact that the distribution of the lesion is that of the median nerve, indicative of secondary spread to skin from the nerve (Figure 2).

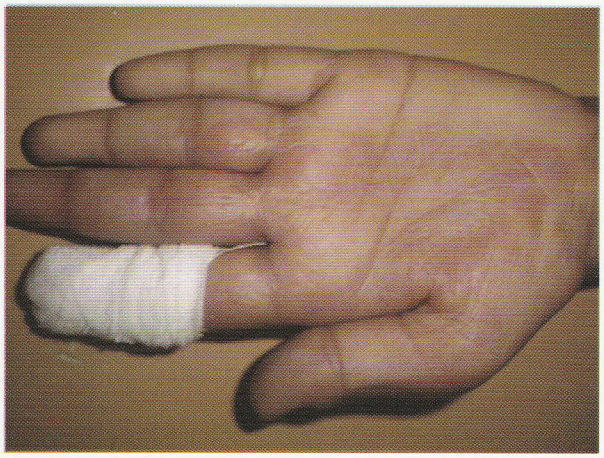


Figure 2. *Maculoanaesthetic lesion distributed over the area supplied by the median nerve with Dactylitis and wasting of small muscles.*

Single lesion leprosy with pseudopodia is shown here. These pseudopodia will subsequently become satellite lesions. As you can appreciate initial spread of the lesion is superficial as optimal temperature requirement of *Mycobacterium leprae* is around 30 C.

This is a large maculo anaesthetic lesion of borderline tuberculoid leprosy with satellite lesions. Such lesions are very vulnerable to develop type I lepra reaction (Figure 3).

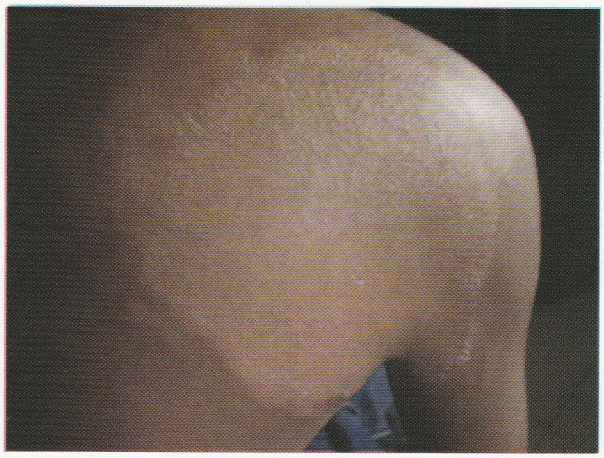


Figure 3. *Borderline tuberculoid with satellite lesions.*

This child presented with a large borderline tuberculoid lesion with psoriasiform appearance (Figure 4).

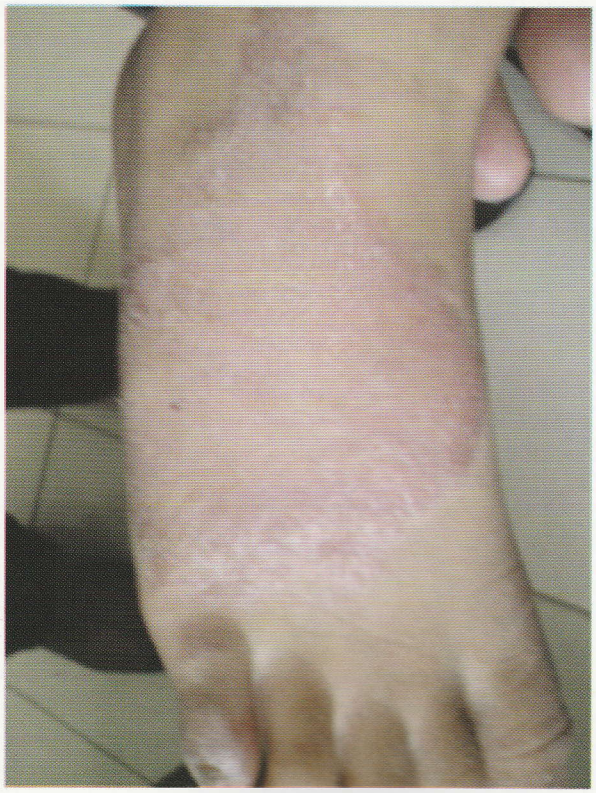


Figure 4. *Borderline tuberculoid lesion with psoriasiform appearance.*

For the purposes of treatment the WHO classification was used. The majority belonged to paucibacillary category (86%).

When Ridley-Jopling classification was used better stratification of patients could be achieved. Indeterminate, tuberculoid and borderline tuberculoid belong to paucibacillary leprosy and the rest belong to multibacillary leprosy.

Let me now show few other clinical types of leprosy. This is a child with mid borderline leprosy.

This type is characterized by large number of annular or oval shaped lesions. During our study we have not seen a child with a leonine face.

Children with leprosy often manipulate their lesions. This child presented with a hyperpigmented nodule as a result of self-demonstration of anaesthesia. A large maculoanaesthetic dry lesion is seen surrounding the nodule (Figure 5).

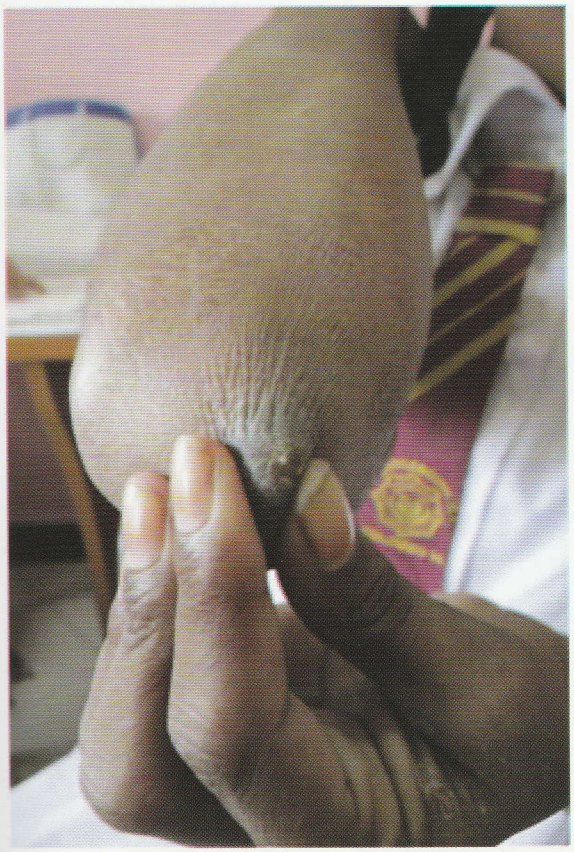


Figure 5. *Hyperpigmented nodule due to self-demonstration of anaesthesia*

Dactylitis is often an overlooked clinical feature of leprosy. Lesional involvement of the thumb is shown in this photograph (Figure 6).

Sauza Campos nodule of leprosy or nodular childhood leprosy is a rare type seen in South America. An infected grandmother with non-nodular lepromatous leprosy was the source of infection in this child (Figure 7).

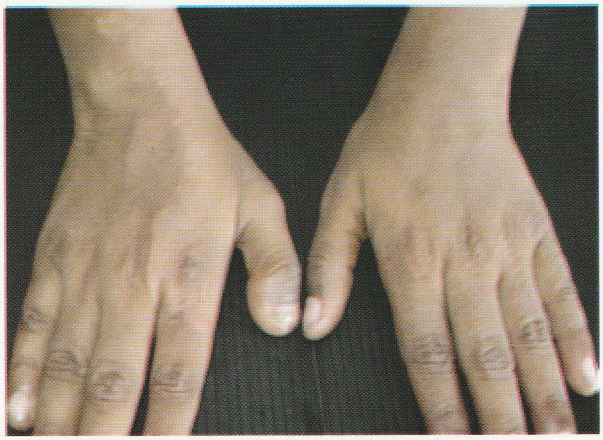


Figure 6. *Dactylitis.*

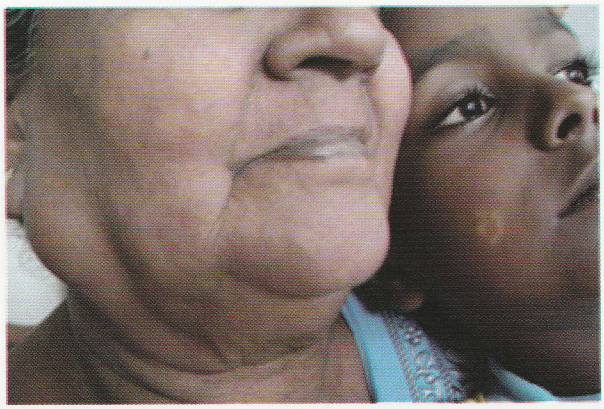


Figure 7. *Sauza Campos nodule of leprosy.*

I have now shown you the common and uncommon clinical types of leprosy. Now let me elaborate few other aspects of the disease.

Majority of our children had presented early (50% within 6 months). Still a significant number had presented late (13% after 24 months). Late presentation is known to be associated with deformities.

Let me now run through some photographs of deformities that I have seen over the years. Fortunately, such deformities are rare in children.

This is a child with Borderline Tuberculoid leprosy involving the area supplied by the ulnar nerve with a fissure at the root of little finger. Such fissures often promote secondary bacterial infections. You can

also appreciate the dryness of the lesion indicative of autonomic nerve dysfunction (Figure 8).

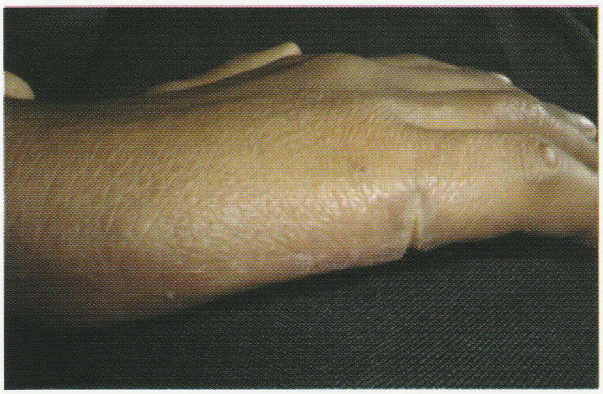


Figure 8. *Dryness and fissuring over the lesion.*

This unfortunate boy has wasting of small muscles of his dominant hand. This is bound to interfere with his writing and his education (Figure 9).

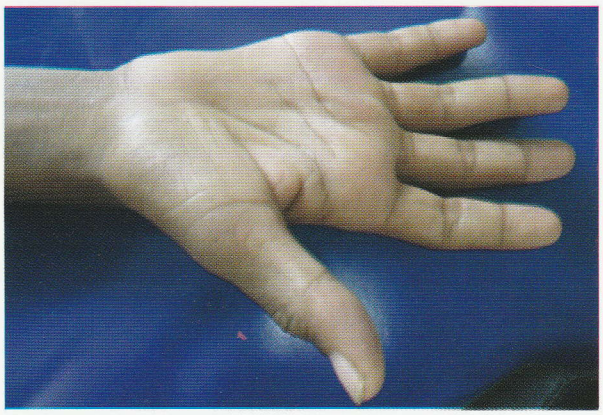


Figure 9. *Wasting of small muscles.*

At times the diagnosis of leprosy cannot be entertained purely on clinical grounds. In such a situation a skin biopsy is useful. Majority of our biopsies were compatible with a clinical diagnosis of leprosy. A negative result warrants a clinical review of the case. Once diagnosed children were treated with multidrug therapy as per WHO guidelines. Pretreatment assessment consisted of a full blood count, a liver profile and G6PD assay. The blue one is the Pauci bacillary pack and the brown one is the Multibacillary pack.

An analysis of treatment outcome carried out in 2012 showed good compliance. Compared to adults, defaulters (7%) were rare. Defaulting often results from adverse drug effects. However majority (98%) of children treated with multi drug therapy did not develop serious side effects.

Here is a child who developed Stevens Johnson Syndrome/ Toxic epidermal necrolysis overlap (Figure 10).

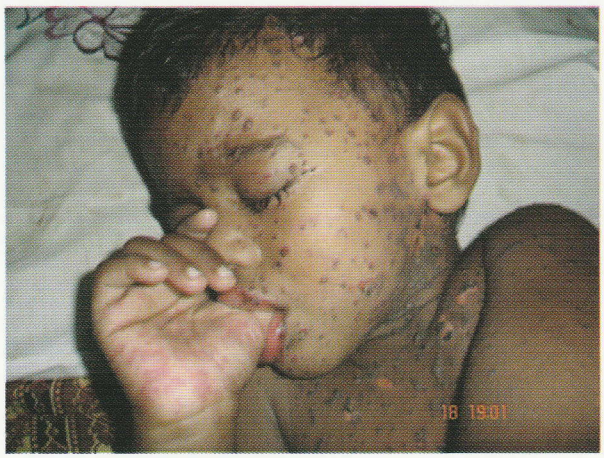


Figure 10. *Severe drug reaction.*

As can be seen here clofazimine induced pigmentation is often commented on by parents. This pigmentation is due to deposition of Clofazimine and accumulation of Ceroid-lipofuscin. Clofazimine pigmentation is often pronounced in leprosy involved skin. Clofazimine induced pigmentation is subsequently replaced by a patch of acquired ichthyosis.

Lepra reactions are troublesome acute inflammatory reactions often seen in the borderline spectrum of the disease. They are often said to be precipitated by multidrug therapy. However they may precede or follow multidrug treatment. Fortunately, lepra reactions are rare in children (Type I - 2%: Type II - 1%).

Type I reactions are characterized by acute inflammation of existing lesions. In addition lesions become thick, and show scaling. You can appreciate the marked epidermal changes. This is to show type I reaction in a borderline Tuberculoid lesion (Figure 11).

Type I reaction is often not limited to the skin. An involved nerve may become tender, enlarged and show loss of function.

Type II lepra reaction seen in the Multibacillary spectrum of the disease is an acute systemic vasculitis. As can be seen here, tender nodules may develop along the peripheral nerves. Apart from acute

systemic features, these children develop multiple indurated nodules, hence the term erythema nodosum leprosum (Figure 12). Both type I and Type II reactions are treated with oral steroids.

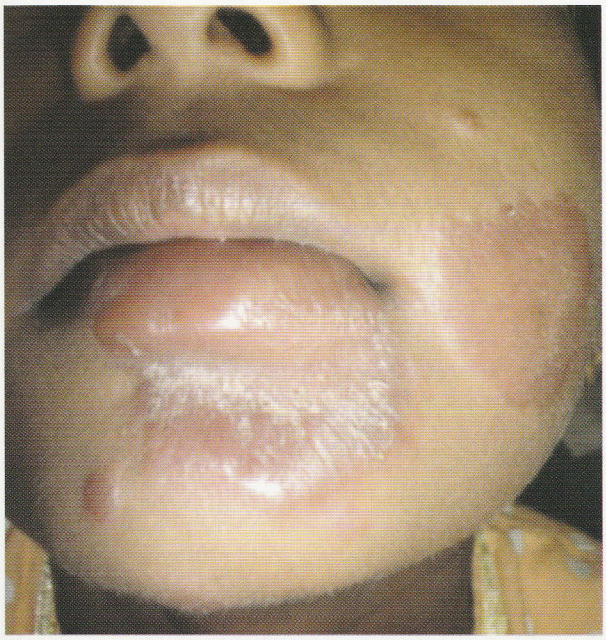


Figure 11. *Type I Lepra reaction.*

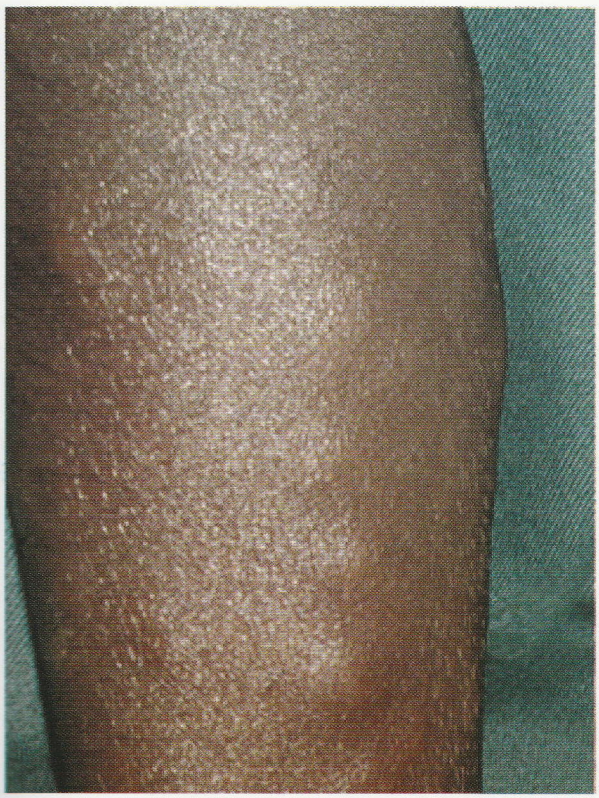
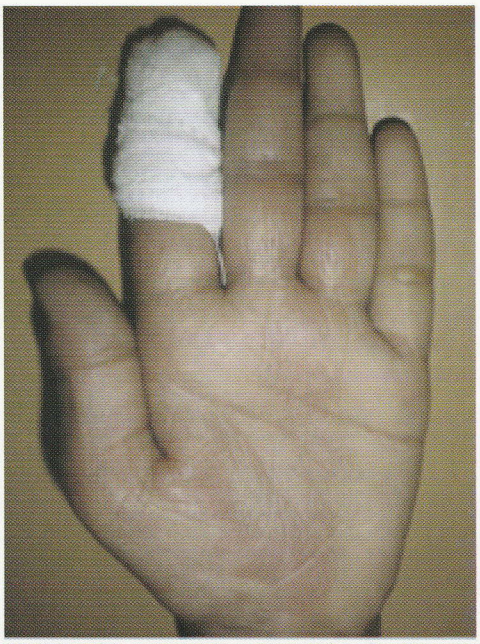


Figure 12. *Erythema nodosum leprosum.*

Long term follow up of our patients have revealed new information. The 2nd photograph below taken nearly after 10 years shows, development of palmar hyperlinearity of the affected hand (Figure 13).



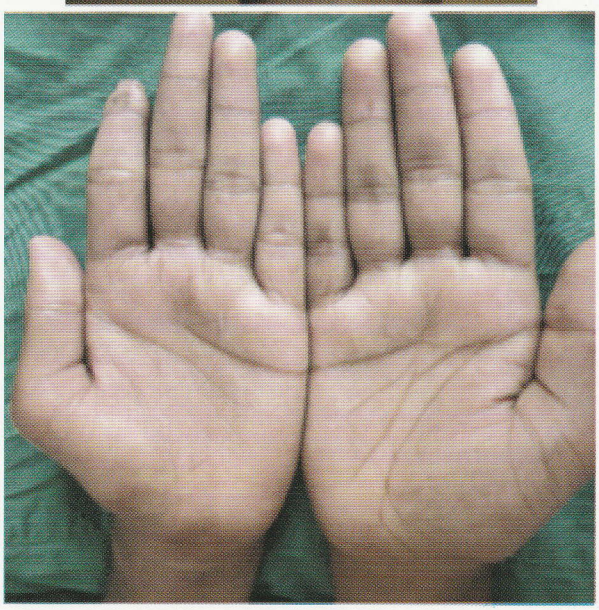


Figure 13. *Hyperlinearity of the affected area.*

This photograph shows development of skin atrophy after completion of multi drug therapy.

New case detection constitutes an important aspect of leprosy prevention. Simple questionnaire based interview at the time of diagnosis revealed that 5% had a known leprous contact.

A more detailed study with active screening of household contacts was carried out in 2007. This revealed a positive rate of 33% among household contacts. This study was a real eye opener. It is of importance to note that the majority (83%) of contacts had paucibacillary leprosy. This also raises issues about transmission. This work was published in the *Ceylon Medical Journal* in 2011.

Since then, routine screening of family members is carried out in our clinic. Analysis of the results for the last 5 years shows a persistent trend. As a result we continue to pick up a significant number of new cases among the contacts.

Our work was presented at the commonwealth session of the British Association of Dermatologists annual conference in June 2014. Same time, it was published in the *British Journal of Dermatology*

At the end of last year, an attempt was made to review all our previous patients since 2001. Out of a total of 443, 142 responded. Information was gathered by interviewing the patient and a parent, by reviewing available records, and by examination of patients. Let me now briefly go through the important information gathered.

The majority (95%) were from the three districts of the western province. Only a small minority (3%) had sought alternative medical therapy. An overwhelming majority (95%) of children had successfully completed the treatment. Most of our children (80%) did not show residual evidence of the disease. Similarly, they had not developed new deformities. But, those who had deformities at the time of presentation (3%) unfortunately continued to live with them. Similar results were observed for peripheral nerve involvement.

It was heartening to know that majority (98%) of parents and children have adjusted well to a diagnosis of leprosy. Since strict confidentiality is maintained about patient details stigmatization has remained low (5%).

Let me now outline the main conclusions of our studies.

1. Compared to adults children present early and therefore most of them do not develop complications.
2. Majority have single lesion leprosy.
3. Deformities and reactions are rare.
4. WHO multidrug therapy is effective, safe and acceptable in treating individual patient. Follow up is recommended.
5. Multi drug therapy which is in use since 1982, has not reduced the child rate significantly.
6. Nerve damage though rare is a significant complication of the disease.
7. Contact screening is an important tool in detecting new cases and therefore prevention of leprosy.

Ladies and Gentlemen I am sure that you agree that I have now given you a clear picture of Childhood Leprosy in Sri Lanka. In other words I feel that I have been able to establish an identity to this unique entity.

Finally let me conclude by quoting Mahathma Gandhi. I quote "Leprosy relief work is not merely medical relief; it is transforming frustration of life in to joy of dedication, personal ambition into selfless service". I unquote.

I pay my sincere gratitude to all my teachers for their invaluable guidance in all aspects of my carrier. I sincerely acknowledge Prof. Jayamini Seneviratna for incorporating me in ongoing work on leprosy at the Children's Hospital and also for the intellectual and moral support he extends to me always, Dr. Hasanga Gamage, Dr. Charuni Gamage and the medical officers in my clinic for the support extended in collecting data and analyzing them, Dr. Nayani Madarasinghe for permitting me to share the data in her study, Dr. Indira Kahawita for providing me with statistics on leprosy in Sri Lanka, all clinic staff at LRH, our patients and their parents for their cordial assistance, my wife Ayesha and daughter Isuri for their help in accomplishment of my work, and all of you in the audience for being with me in this evening.

Thank you.