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Visible Deformity in Childhood Leprosy—A 10-Year Study

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ABSTRACT

Deformity seen in children with leprosy has not often been studied, as the disease itself is less common in children. Deformity, being synonymous with the stigma of leprosy, is a definite social problem in children. In this study we have focused on the burden of deformity in children with leprosy, and various factors responsible for the deformities are discussed. We have observed an incidence of 10.5% of Grade II deformities in children with leprosy, which is very high compared to the community rate of 1.4%. Various factors which contributed significantly to the deformities in our study were: increasing age of children, delay in accessing health care, multiple skin lesions, multibacillary disease, smear positivity, multiple nerve involvement, and reaction at the time of presentation to the hospital. Logistic regression analysis showed that children with thickened nerve trunks had 6.1 times higher risk of developing deformities compared to those who did not have nerve enlargement. Children with the above risk factors should be followed up more frequently so as to detect any deformity as early as possible.

RÉSUMÉ

Les déformations observées chez les enfants souffrant de lèpre ne sont guère étudiées, peut-être parce que la maladie chez ces derniers est elle-même moins fréquente. Les déformations associées à la lèpre, synonymes de stigmatisation, sont accompagnées de problèmes sociaux bien définis chez les enfants. Dans cette étude, nous nous sommes attachés à définir la prévalence des déformations chez les enfants atteints de lèpre, et à discuter les divers facteurs associés à ces déformations. Nous avons observé une incidence de 10,5 % de déformations de grade II chez les enfants souffrant de la lèpre, ce qui est très élevé par rapport au taux de 1, 4 % pour la communauté. Les facteurs ayant contribué de façon significative aux déformations observées dans notre étude ont été une corrélation positive avec l'âge, le délai à accéder aux soins de santé, des lésions cutanées multiples, une forme multibacillaire, un examen bactérioscopique positif du suc dermique, des atteintes nerveuses multiples et des réactions immunopathologique au moment de la présentation à l'hôpital. Une analyse de régression logistique a montré que les enfants présentant des troncs nerveux épaissis avaient

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un risque 6,1 fois plus élevé de développer des déformations que les enfants sans épaississements des nerfs. Les enfants avec les facteurs de risque analysés plus haut devraient être suivi plus fréquemment afin de détecter les déformations le plus précocement possible.

RESUMEN

Las deformidades en los niños con lepra son poco estudiadas porque la lepra en sí, es menos común en los niños que en los adultos. La deformidad, sinónimo de estigma de la lepra, es un verdadero problema social entre los niños. En este estudio enfocado al estigma de la deformidad en los niños con lepra, se discuten varios factores responsables de la deformidad. Observamos una incidencia del 10.5% de deformidades del Grado II en los niños con lepra. Esta incidencia es muy alta comparada con la incidencia del 1.4% en la comunidad general. Varios factores contribuyeron significativamente a la alta incidencia de deformidades en nuestro estudio, entre ellos: incremento en la edad de los niños, retardo en la procuración de atención médica, lesiones múltiples en la piel, enfermedad multibacilar, positividad bacilar en la linfa cutánea, afectación nerviosa múltiple, y estados reaccionales al momento de su ingreso al hospital. Los análisis de regresión logística mostraron que los niños con troncos nerviosos engrosados tuvieron un riesgo 6.1 veces mayor de desarrollar deformidades que los niños sin engrosamiento de los nervios. Se concluye que los niños con los factores de riesgo antes mencionados deben vigilarse con mayor frecuencia con el fin de detectar cualquier deformidad tan pronto como aparezca.

Childhood leprosy is an indicator of endemicity of the disease ([13]). To the common man leprosy is well known because of the stigma that is still prevalent in the society. This stigma is almost synonymous with visible deformity. Major factors that contribute to deformity in various studies in adult leprosy patients are delay in diagnosis ([10, 20]), high bacillary load ([16]), multiple nerve enlargements ([16]), occurrence of reaction ([19]), and delay in provision of proper care of the disease. Therefore deformity is a preventable complication in the majority of patients. Deformity occurring in children is more distressing both socially and psychologically, as they have to live their whole life with stigma and in a hostile environment. This study was carried out to document the number of children with Grade II deformity as well as to identify the role of various risk factors contributing to its development.

MATERIALS AND METHODS

All the new childhood leprosy cases (<15 yrs) registering at Scheiffelin Leprosy Research & Training Center, Karigiri, for 10 yrs (from July 1994 through June 2003) were analyzed retrospectively. Details were collected concerning each patient on admission included sex, age, WHO classification ([2]), skin smear status, duration of disease, involvement of nerves, occurrence of reaction, presence and nature of deformities and the distribution of any deformities. For the purpose of this study, deformity was graded according to the WHO classification ([25]) and only patients with Grade II deformities were considered for analysis of risk factors. Nerve involvement was recorded according to the examining physician's perception of thickened nerves. Details of any previous therapy received, as well as subsequent treatment received from this institute, were also noted. All the patients received multidrug therapy as per WHO schedule ([26, 14]) and the progress of the disease was assessed in each patient. The odd's ratio was calculated with 95% CI for each parameter while analyzing relative risk. Logistic regression analysis was done to determine the significance of individual risk factors.

RESULTS

In all, 6031 new patients were registered during the 10 yr period, from July 1994 through June 2003, including 275 (4.5%) children with leprosy. Of these, 163 (59.2%) were male and 112 (41.8%) were females. In age distribution, 13 patients were below 4 yrs, 71 were between 5–9 yrs and 191 were between 10–15 yrs (Table 1). Comparing both the groups, i.e., those with visible deformities to those without visible deformities (Table 2), the majority of the deformities were in the age group of 10–15 yrs, followed by those in the age range of 5–9 yrs. Deformities affecting children

TABLE 1. *Distribution of children with leprosy by age and sex.*

Age Group	F (%)	M (%)	Total (%)
0–4	7 (96.2)	6 (3.6)	13 (4.7)
5–9	31 (27.7)	40 (24.6)	71 (25.8)
10–15	74 (66.1)	117 (71.8)	191 (69.5)
Total	112 (40.7)	163 (59.3)	275 (100)

10–15 yrs old was statistically significant when compared to the other age groups ($p < 0.05$). Considering disease classification, 238 children (86.5%) had paucibacillary (PB) disease whereas 37 (13.4%) had multibacillary (MB) disease. Of the 238 PB patients, 20 (8.4%) had deformities, whereas 9 of the 37 MB cases (24.3%) had deformities ($p = 0.007$). One hundred twelve children (40.7%) presented with single patch, 126 (45.8%) with 1–4 patches and the remaining 37 patients (13.4%) had multiple (≥5) patches.

Skin smears were negative at the time of presentation for 252 of the patients (91.6%), and 23 (8.4%) patients had a positive skin smear. Deformities were seen in 24 of 252 (9.5%) smear-negative patients, whereas 5 out of 23 (21.7%) smear-positive patients had deformities ($p = 0.06$).

The average age of onset of the disease among children was 9.49 ± 2.9 yrs. Average duration of disease at the time of diagnosis was (1.09 ± 0.8) yrs. Two hundred eleven (76.7%) children presented to the hospital within one year of onset of the disease, whereas 64 (23.2%) patients presented after one year of onset of the disease. The majority of the patients with deformities (58.6%) attended the hospital before one year of onset of the disease whereas 42.4% attended after one year of the onset of the disease. In the group without any deformities, 193 (78.4%) presented for health care before one year of onset of the disease and the rest 53 (21.5%) presented after one year of onset of the disease. This difference was statistically significant ($p = 0.03$)

The common peripheral nerve trunks involved in leprosy are the ulnar, radial, median, lateral popliteal, posterior tibial and the facial nerves ([17]). Thickened nerve trunks were present in 110 out of 275 patients (40%) at the time of presentation, out of which 60 (21.8%) had only one major nerve involved, 22 (8%) had thickening of two major nerve trunks, and 28 (8.3%) had multiple (≥3 nerves) nerve involvement. The ulnar nerve was the commonest nerve trunk involved in 93 patients (33.8%), followed by the lateral popliteal nerve in 51 (18.55%) and the posterior tibial nerve in 12 (4.3%). Of the 110 children who presented with thickened nerve trunks, 24 (21.8%) had deformities, whereas only 5 of 165 patients (3.0%) who presented without any nerve thickening had visible deformities ($p < 0.001$).

TABLE 2. *Risk factors for deformity in children. Odds rations of risk factors, adjusted and unadjusted by logistic regression.*

Risk Factors	Group	With Deformity	Without Deformity	Unadjusted O.R.	Unadjusted 95% C.I.	Adjusted by Logistic Regression O.R.	Adjusted by Logistic Regression Significance (P)
Age (years)	10–15	26	165	4.255	1.251–14.473	2.563	0.1570
	0–9	3	81				
Sex	M	16	147	0.829	0.382–1.799	0.757	0.5173
	F	13	99				
Duration	>1 yr	11	53	2.225	0.991–4.999	2.048	0.1137
	<1 yr	18	193				
MB/PB	MB	9	28	3.504	1.453–8.443	1.914	0.2124
	PB	20	218				
Smear	Skin Smear +	5	18	2.639	0.889–7.743	0.814	0.7457
	Smear –	24	228				
Reaction	Yes	11	44	2.806	1.238–6.357	1.546	0.3459
	No	18	202				
Nerve Enlargement	Yes	24	86	8.930	3.290–24.239	6.134	0.0009
	No	5	160				

A reaction was present in 55 (20%) of the children at the time of admission, and 11 (20%) of these patients had deformities compared to 18 of 220 patients (8.1%) without reaction at the time of presentation ($p = 0.02$). All but one of the children had Type I reactions.

Grade II deformity (per WHO grading ([1, 27])) was present in 29 patients (10.5%), of whom 16 (55.1%) were male and 13 (44.9%) were female, a male to female ratio of 1.2:1. There was not a single child with deformity below 4 yrs of age. Three children with deformities (10.3%) were in the 5–9 yr range and the remaining 26 (89.7%) were in the age range of 10–15 yrs. The average age of onset of the disease among the deformed children was 12.2 ± 2.3 yrs, and the average duration of disease was 1.5 ± 1.2 yrs. In 24 patients (82.7%), deformities involved the upper limbs whereas the lower limbs were involved in only 3 (10.3%). Deformities of the right hand were seen in 16 (55.1%) patients and left hand involvement was present in 10 of the patients (34.4%).

During the 10 yrs studied, the distribution of deformities had shown no significant change, although a slight downward trend can be seen in the new millennium (Table 3). This could be due to the increasing awareness of the public regarding leprosy.

TABLE 3. *Distribution of Grade II deformities in children by year of diagnosis.*

Year	Total number of children with leprosy	Number of children with deformities
1995	13	4
1996	22	3
1997	23	4
1998	42	4
1999	48	4
2000	34	2
2001	37	2
2002	16	1
Total	235	24

DISCUSSION

This study reveals that 4.5% of the new cases seen in the outpatient department of a referral hospital during the years 1994 to 2003 were children and that 10.5% of these had visible deformities. Deformities due to leprosy in children were observed more frequently in boys than in girls, although the difference was not statistically significant. It is well known that leprosy is prevalent more frequently among males than in females, on the order of 2:1. Other studies of leprosy in childhood have shown equal incidence, or a slight preponderance of males ([6, 22]). In the present study the male to female ratio in the deformity group is 1.2:1 which tallies well with the observations made by Meima ([10]), Kushwah ([8]), Nilkantha Rao ([15]) and Bravo ([2]).

The overall consensus among leprosy workers is that the deformity rate increases with an increase in age, as noted down in various studies by Bravo ([2]) and Noordeen ([12]), though this has not been studied specifically in children, and there is a greater chance of exposure to the disease process with advancing age. In this study the disability rates were observed to increase with increasing age of the children. There was no deformity in the 0–4 yr age group and the maximum frequency of deformity was observed in the 10–15 yrs age range, which agrees with the observations made in the previous studies.

Deformity affecting children with MB disease was significantly more frequent than in PB cases. Studies by Schreuder, *et al.* ([20]), Saunderson, *et al.* ([19]), and Saha, *et al.* ([18]) have also found a significantly higher incidence of nerve function impairment (NFI) in MB as compared to PB leprosy patients. High disability rates among MB patients is explained by widespread nerve damage after several years of exposure in lepromatous cases and due to extensive large nerve involvement in borderline cases compared to localized nerve involvement in PB cases. Children with MB leprosy are found to be at higher risk of reversal reactions as observed by Selvasekar, *et al.* ([22]) and require a regular follow-up and prompt interventions for the prevention of deformities.

A higher bacterial load increases the risk of reactions and nerve damage leading to deformities. Roche, *et al.* ([16]) has also shown high incidence of neuropathy in patients with high BI similar to those reported by Noordeen ([12]) and Zhang ([28]), and can be explained by the widespread and progressive nature of smear positive type of leprosy. Other types of leprosy are more local-

ized and have a shorter evolution. Therefore early detection of the disease and treatment along with health education are very important for disability control. However in our study smear positivity was not found to be a significant risk factor for development of deformities, which could be attributed to the very low number of smear-positive cases in the cohort.

In this study the deformity rate was observed to increase with increased duration of the disease. Studies by Meima ([10]), Schreuder ([20]), Wittenhorst ([25]), and Nichols ([11]) have reported on increased NFI in patients presenting late after developing skin lesions. In our study of children, the average duration of the disease in those with deformities was over one year, and 40% of the patients with deformities attended the hospital after one year of onset of their disease. Noordeen ([12]) and Thappa ([24]) made similar observations. Early detection and early access to the health care system would help to prevent such deformities.

Leprosy is a disease that particularly affects peripheral nerves. In our study peripheral nerve trunk involvement was seen in 40% of the patients at the time of presentation. Deformities seen among those with nerve trunk involvement were significantly more frequent compared to those without nerve trunk involvement. The risk of developing deformity in those with nerve enlargement in our study was 6.13 times greater than those who did not have nerve enlargement. The ulnar nerve, lateral popliteal nerve and the posterior tibial nerve are the common nerves involved in our patients. Saha ([18]) and Sharma ([23]) have reported similar observations. Saunderson and colleagues ([19]) in their study had reported a relative risk of 2.8 and 6.5 respectively in case of 1–5 and more than 5 thickened nerves. Roche, *et al*. ([16]) has also reported similar findings.

Reactional episodes are reportedly less frequent in children ([7]) but in our series 20% of the children presented with reversal reactions at the time of admission. This is less than the value of 30% reported by Hammond ([5]). In another study by Jain and colleagues ([6]), 29.7% of the children developed reaction. Patients with reaction at the time of presentation had more deformities compared to those without reactions. Bravo ([2]), Thappa ([24]) and Gupte ([4]) also noted reactions as significant risk factors in the development of deformities in their studies. Saunderson ([19]) has reported a relative risk of 14.7 to develop nerve function impairment in patients with reversal reaction. Reactions can be managed with prompt use of corticosteroids and hence effectively managing them would help to decrease the deformity load, athough steroid use in children is likely to lead to many complications and to growth retardation.

In our series the majority of the deformities involved the upper limbs, lower limbs being involved only in 10% of the patients. Sehgal ([21]) reported deformities of the hands in 85.7% of patients compared to deformities of the feet in 48.5%. Martinez-Dominiqueez ([9]) and Thappa ([24]) have found the hands and feet to be affected with equal frequency. However the high frequency of right hand involvement in our patients has a definite socioeconomic impact, as the dominant hand is disabled.

CONCLUSIONS

There is no previous study reporting the deformity rate in children with leprosy, and this study found an incidence of 10.5% of grade II deformity. This is high, compared to the overall deformity rate in the community ([3]), but the results of this hospital-based study may not be comparable with the outpatient community. The unadjusted results indicate an increase in risk of deformity for the following factors: increasing age of children, delay in accessing health care, multiple patches, multiple nerve involvement, and reaction at the time of presentation to the hospital. Logistic regression analysis, however, reveals that nerve enlargement is the most significant risk factor in children for the development of deformities. Children with these risk factors must be followed-up more closely so as to prevent deformities. Children presenting late, with stigmatizing deformity, indicates inadequate early case detection activities as well as reluctance on the part of children and their parents to come forward to access the health system. Promoting various ways to motivate parents to bring their children to the hospital at the earliest sign of leprosy is of utmost importance in the present situation.

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