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Changes in the Anterior Nasal Spine and the Alveolar Process of the Maxillae in Leprosy A Clinical Examination[1, 2]

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In 1952 I introduced the term *facies leprosa* to denote certain specific pathologic changes in the skeletal structures of the face with the following characteristics: atrophy of the anterior nasal spine (ANS); atrophy and recession of the alveolar process of the maxillae (APM) confined to the incisor region, beginning centrally at prosthion and resulting in loosening and/or loss of the corresponding teeth; endonasal inflammatory changes.

In the same year *facies leprosa* was found in five of the seven leprosy patients sent to the Pleiestiftelse No. 1 in Bergen ([9, 15, 16, 17]). Because this discovery was made in Bergen, it was suggested that the term the "Bergen Syndrome" be used to denote the clinical symptoms corresponding to *facies leprosa*

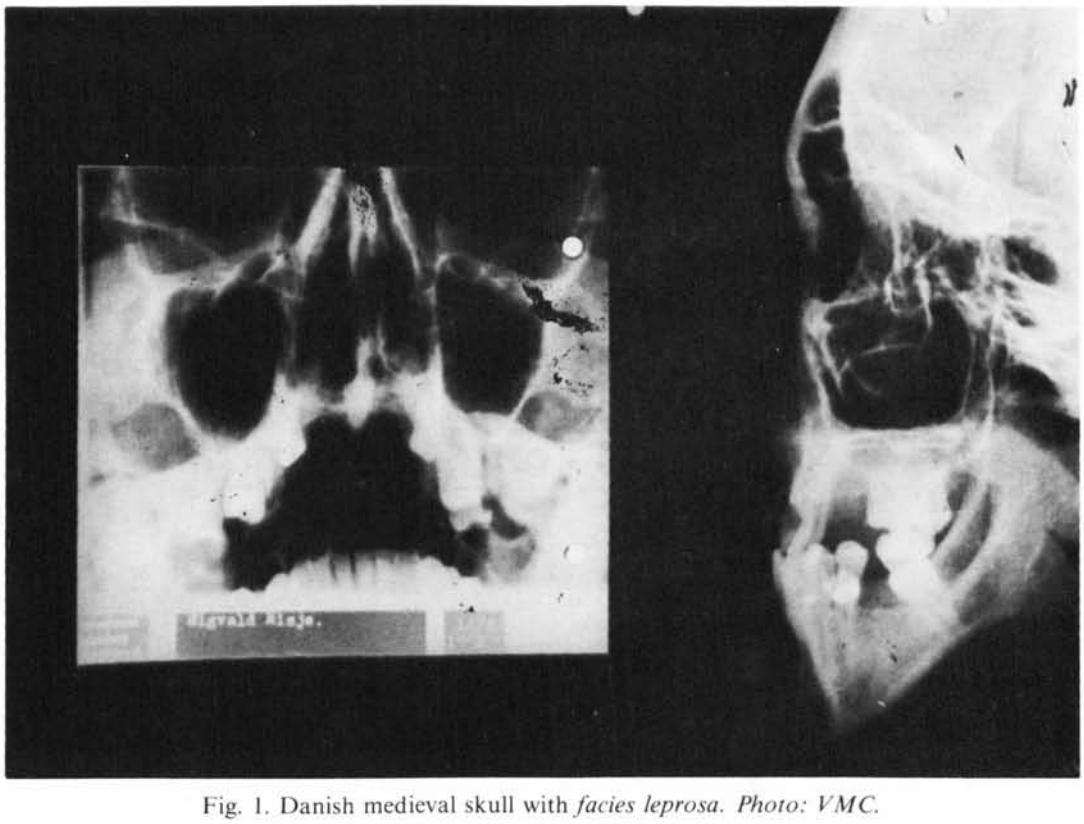

Fig. 1. Danish medieval skull with *facies leprosa. Photo: VMC.*

(Fig. 1). Since that time this syndrome has been discussed several times ([1, 2, 3, 4, 5, 6, 8 10, 11, 12, 14]). Subsequent studies led to an elaboration into Types I and II.

Inflammation of the nasal cavity occurred in 100% of the Danish skeletons examined which had leprosy changes in hands and/or

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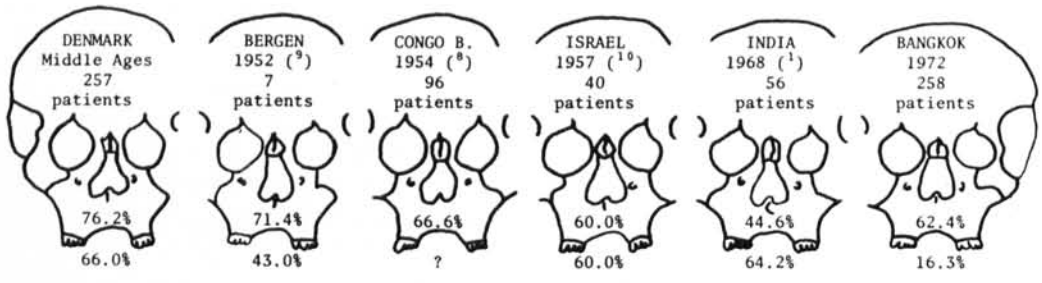

Fig. 2. Diagram showing the "Bergen Syndrome I and II" in cumulative studies. The upper percentage figure refers to changes in the anterior nasal spine (ANS), and the lower figure refers to changes in the alveolar process of the maxillae (APM).

TABLE 1. *Relative changes in 107 Danish skulls.*

No.	Male	Female	Atrophy of ANS	Atrophy of APM
107	44	63	77 (72.0%)	70 (63.4%)
44	male		28 (63.6%)	30 (68.1%)
63	female		49 (77.7%)	40 (63.5%)

feet. Figure 2 shows the percentages of pathologic changes in the skulls of 257 leprosy skeletons from the Danish Middle Ages. Atrophy of the anterior nasal spine was seen in 76%, atrophy of the alveolar process of the maxillae together with loosening and/or loss of teeth was seen in 66%, symmetrical occurrence of *usura orbitae (cribra orbitalia)* was seen in 63% of the cases ([12]), and typical deformities of hands and/or feet were seen in 85% of 115 skeletons having *facies leprosa*. These (Fig. 2) accentuate the differences between the occurrences of atrophy of the anterior nasal spine and atrophy of the alveolar process of the maxillae.

In the whole Danish group of skeletons there have been found *facies leprosa* coincident with leprosy changes in hands and/or feet in 74%, and *facies leprosa* as the only change in 2%. Leprosy changes in hands and/or feet alone or coincidental with *facies leprosa* occurred in 98%, and leprosy changes in hands and/or feet as the only leprosy change occurred in 26% of the cases.

The relative findings in a group of 107 skeletons with lepromatous leprosy are summarized in Table 1.

The diagram (Fig. 2) summarizes the estimated figures which have been derived from examinations of patients with lepromatous leprosy ([1, 8, 9, 10]).

In February-March 1972 a clinical examination of the anterior nasal spine and of the alveolar process of the maxillae, the two components of the Bergen Syndrome, was made in 333 inpatients at the Prapradaeng Leprosarium, Bangkok, Thailand; the results are here reported.

MATERIALS AND METHODS

Three hundred and thirty-three inpatients, aged 13 to 79 years, were examined and classified as to clinical type as detailed in Table 2.

The examination consisted of digital examination of the anterior nasal spine and the usual oral and dental inspection. As it was not feasible to make x-ray examina-

TABLE 2. *Clinical classification of leprosy types.*

Group	Male	Female	Total
Lepromatous	128	130	258
Borderline	7	12	19
Tuberculoid	37	19	56
Total	172	161	333

tions, the clinical oral and dental examinations concentrated on the establishment of loosening and/or loss of the central upper incisors.

Only those cases were registered where remembered oral or facial traumata could be excluded as well as general paradentosis or sequelae because of decay. Therefore, the recorded figures seem to be the absolute minimal. X-ray examination probably would have revealed more changes in both the ANS and the APM.

RESULTS

In the tuberculoid group there was only one case with changes of the APM. This case has to be disregarded because the type of leprosy is questionable.

In the borderline group there were three cases with changes of the APM, but because the diagnoses are questionable all these three cases also have to be disregarded. Therefore, only the lepromatous group consisting of 258 patients (128 male and 130 female) are tabulated in Table 3. Of these (Table 3) 161 patients (87 male, 74 female), 62.4% show pathologic changes of the anterior nasal spine, i.e., the leprosy nose changes representing the Bergen Syndrome I and 42 patients (24 male, 18 female), 16.3% showed pathologic changes of the alveolar process of the maxillae with manifest dental changes, i.e., leprosy loosening and/or loss of incisors, characteristic of Bergen Syndrome II.

Thirty-six patients (Table 4) (22 male, 14 female) or 22.4% showed pathologic changes of the alveolar process of the maxillae together with dental changes, that is Bergen Syndrome II, and 125 patients (65 male, 60 female) or 77.6% presented normal alveolar processes.

In the group of 42 patients with Bergen Syndrome II (pathologic changes of the alveolar process of the maxillae with dental changes) in Table 5: 36 patients (22 male, 14 female) or 85.6% showed Bergen Syndrome I, and 6 patients (2 male, 4 female) or 14.4% showed normal anterior nasal spines.

It has been observed that the ratio of Bergen Syndrome I and II is always lower in female groups.

DISCUSSION

The most fascinating leprosy patient with Bergen Syndrome I and II that I met in Thailand was Mrs. S., whom I first examined in 1959 and met again in 1972.

Her leprosy began when she was ten years old. Changes in her fingers developed seven years later. Her nose collapsed and changes

TABLE 3. *ANS and APM changes in Bangkok study.*

No.	Male	Female	Change of ANS Manifest	Change of ANS None	Changes of APM with dental changes Manifest	Changes of APM with dental changes DBS	Changes of APM with dental changes None
258	128	130	161 (62.4%)	97 (37.6%)	42 (16.3%)	29 (11.2%)	187 (72.5%)
128	male		87 (68.0%)	41 (32.0%)	24 (18.7%)	14 (10.9%)	90 (70.4%)
130	female		74 (57.0%)	56 (43.0%)	18 (13.9%)	15 (11.5%)	97 (74.6%)

TABLE 4. *Manifest changes of the anterior nasal spine in 161 patients.*

No.	Male	Female	Changes of APM with dental changes
161	87	74	36 (22.4%)
87	male		22 (25.5%)
74	female		14 (19.0%)

TABLE 5. *Manifest changes of APM with dental changes in 42 patients.*

No.	Male	Female	Changes of the anterior nasal spine
42	24	18	36 (85.6%)
24	male		22 (91.7%)
18	female		14 (77.8%)

in the toes appeared when she was 22 years old. She was admitted to the Prapradaeng leprosy hospital in 1959 at the age of 33, which is where I met her (Fig. 3). At that time she had observed looseness of the central upper incisors for about a year. Three years later she lost her loose incisors: 1 + 1.

Three months before I met her again in 1972, she observed loosening of her lateral frontal incisors: 2 + 2 (Fig. 4). The case of Mrs. S. indicates that leprosy changes of the nose are a relatively late occurrence developing after 12 years of the disease, and that the leprogenic alveolar changes had occurred still 11 years later, thus, after 23 years of disease duration.

FIG. 3. The 33 year old Mrs. Sorn as seen in 1959. *Photo: VMC.*

In five other cases from the group with lepromatous leprosy, the leprogenic changes of the alveolar process of the maxillae have been observed as occurring within one to nine years of disease duration. In one 20 year old woman, the loosening of the central incisors occurred within two years after a diagnosis of lepromatous leprosy was made.

FIG. 4. Mrs. Sorn as seen in 1972. *Photo: VMC.*

Usually, however, it may take several years before this symptom develops. X-ray examination of thousands of patients is necessary in order to arrive at a final solution of the problem.

SUMMARY

A brief review is presented of the Danish medieval skeletons with leprosy changes, particularly in the skulls, where the pathologic specific changes found are termed *facies leprosa.*

The clinical aspect of *facies leprosa* is termed Bergen Syndrome I (nasal leprosy) and Bergen Syndrome II (the leprogenic changes of the alveolar process of the maxilla superior with loosening and/or loss of the frontal incisors).

An account is given of the interim result of a clinical examination of 333 inpatients at the Prapradaeng Leprosarium in Bangkok, Thailand. In the group of 258 patients with lepromatous leprosy, 62.4% presented Bergen Syndrome I and 16.3% Bergen Syndrome II. These proportions differ clearly from the corresponding ratio found in the Danish medieval skulls.

RESUMEN

Se presenta una breve revisión de los esqueletos daneses del medioevo que tienen alteraciones debido a lepra, especialmente en la calavera, donde los hallazgos patológicos específicos encontrados se han denominado—*facies leprosa.*

El aspecto clínico de la *facies leprosa* se ha denominado Síndrome Bergen II (los cambios leprogénicos del proceso alveolar del maxilar superior con aflojamiento y/o desprendimiento de los incisivos frontales).

Se presenta un informe sobre el resultado provisional del examen clínico de 333 pacientes en el Prapradaeng Leprosarium, en Bangkok, Tailandia. En el grupo de 258 pacientes con lepra lepromatosa, 62,4% presentaron el Síndrome Bergen I y 16,3% el Sindrome Bergen II. Estas proporciones difieren claramente de la tasa correspondiente encontrada en las calaveras medioevales danesas.

RÉSUMÉ

On a brièvement passé en revue les squelettes danois du Moyen-Age présentant des modifications lépreuses, particulièrement au niveau de la face et du crane, où les modifications pathologiques spécifiques observées sont nommées *facies leprosa.*

L'aspect clinique de ce *facies leprosa* a reçu l'appellation Syndrome de Bergen I (lèpre nasale) et Syndrome de Bergen II (modifications léprogènes des alvéoles du maxillaire supérieur avec ballottement et/ou pertes des incisives frontales). On a donné les résultats partiels de l'examen clinique pratiqué chez 333 malades hospitalisés à la Léproserie de Prapradaeng à Bangkok, en Thaïlande. Dans un groupe de 258 malades atteints de lèpre lépromateuse, 62,4 pour cent ont présenté le Syndrome de Bergen I et 16,3 pour cent le Syndrome de Bergen II. Ces proportions sont nettement différentes des proportions correspondantes trouvées dans les cranes danois du Moyen-Age.

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