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Leprogenic odontodysplasia: new evidence from the St. Jørgen's medieval leprosarium cemetery (Odense, Denmark)

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Abstract Leprogenic odontodysplasia (LO), also known as dens leprosus, consists of anomalous root development of the permanent upper incisors. This dental anomaly was first reported by Danielsen in 1968 among Danish juvenile skeletons from medieval leprosaria cemeteries. As yet, no clinical cases have been documented and the etiological and epidemiological significance of the condition are poorly understood. The aim of this study is to discuss a case of LO found amongst the skeletons from the St. Jørgen's leprosarium cemetery (13th–16th/17th centuries), housed in the ADBOU (Anthropological Database of Odense University), Southern Denmark University. A juvenile individual presents a disarticulated maxillary right central incisor possessing a short root that shows a groove caused by marked constriction beginning approximately 1.5 mm above the neck. From this groove, the diameter decreases considerably until the apex. Atrophy of the anterior alveolar maxillary process, extending laterally from the central incisors to the canines, is also apparent. This individual exhibits additional rhinomaxillary lesions (e.g. absorption of the piriform margin including the anterior nasal spine) and foot changes (including phalangeal acro-osteolysis) compatible with a diagnosis of lepromatous leprosy. This case contributes to the debate about the significance of this rare condition, particularly in terms of its presence in Scandinavian skeletons from medieval leprosaria cemeteries. Possible interpretations are discussed, including the pathognomonic value of the specific lesion and whether it indicates early childhood onset of leprosy during the Middle Ages. The understanding of LO epidemiology and its relationship with leprosy will benefit from future clinical and skeletal studies.

Key words: Hansen's disease, leprosy, paleopathology, tooth abnormalities, dens leprosus

Introduction

Leprogenic odontodysplasia (LO), also known as dens leprosus (Danielsen, 1968), consists of a developmental anomaly, identifiable as a concentric constriction groove in the root of the upper permanent incisors (Danielsen, 1968; Andersen, 1969; Danielsen, 1970; Roberts, 1986; Ortner, 2003). This malformation was first observed in an 8–9 year old individual exhumed from the St. Jørgen's medieval leprosarium cemetery in Næstved, Denmark, excavated in 1966 by Vilhelm Møller-Christensen and Knud Danielsen (Danielsen, 1970). Danielsen (1970) also described this condition in three additional juveniles, with ages at death ranging between 10 and 11 years old.

The co-occurrence of LO with facies leprosa in those juveniles led Danielsen to conclude that this condition was "caused by … low-resistance leprosy in childhood" (Danielsen, 1970, p. 19), currently known as lepromatous leprosy. Surprisingly, during four decades of studies no other cases were noticed (Matos and Santos, 2011). Recently, Kjellström (2012) reported LO in a juvenile skeleton from medieval Sigtuna in Sweden. Despite these archeological cases, no clinical cases have been described and the etiological and paleoepidemiological significance of LO are poorly understood.

This study aims to present a new case of LO from the medieval Danish archeological record, and will discuss the diagnostic challenges and pathological significance underlying this rare condition.

Material and Methods

The St. Jørgen's leprosarium cemetery at Odense was used between 13th and 16th/17th centuries AD (Arentoft, 1999). Its archeological excavation took place in 1980 and 1981 and more than 1200 individuals were unearthed (Arentoft, 1999; Boldsen, 2001). Currently, these remains are housed in the Anthropological Database of Odense University (ADBOU), at the Southern Denmark University.

The skeleton identified as number 572, presented in this paper, is one of 191 individuals (43 younger than 20 years old and 148 adults) with rhinomaxillary bones well preserved, studied by Matos (2009). This individual was

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inspected macroscopically, and is estimated to have been between 13 and 19 years old at the time of death based on epiphyseal fusion (Coqueugniot and Weaver, 2007; Cardoso, 2008a, b) and dental eruption (Ubelaker, 1989; Cardoso, 2005).

Results

Individual 572 has a relatively well-preserved skeleton (Figure 1), including the facial bones (Figure 2A–C), which show the classic rhinomaxillary changes described by Andersen and Manchester (1992), namely the enlargement and destructive remodelling of the piriform margin (Figure 2A), including resorption of the anterior nasal spine (Figure 2A, B); pitting and new bone formation on the nasal surface of the palatine process, and an 'empty nose' due to paleopathological destruction of the intranasal structures.

Twenty-four teeth were inspected, 21 within their respective alveoli. The upper right central incisor, detached from the corresponding alveolus, presents a shortening of the root length in around one-third of total tooth length. Moreover, this incisor shows a concentric constricted groove 1.5 mm above the neck towards the apex (Figure 3) and with the diameter decreasing considerably toward the apex. The left central upper incisor is absent, and thus a comparison cannot be made. The alveoli of the upper incisors are almost completely reabsorbed (Figure 2C) due to the destructive remodelling of the anterior maxillary alveolar process (Figure 2A, C) extending laterally from the central incisors to the canines.

The postcranial skeleton does not present any visible lesions in the following preserved bones: clavicles, scapulae, upper limb bones, vertebrae, ribs, os coxae, femora, patellae, 19 hand bones (both lunates, left capitate, left hamate, right scaphoid, both 2nd metacarpals, left 4th metacarpal, 5 fragments of metacarpals diaphyses, 2 proximal, 2 intermediate, and 2 distal phalanges). The diaphyses of both tibiae and fibulae show a mixture of woven and compact bone. Among the 22 foot bones preserved (both calcanei, tali, naviculars, cuboids, intermediate cuneiforms and right lateral cuneiform, 10 metatarsals and 1 proximal phalanx), 9 present the following skeletal changes: enlarged foramina and woven bone at the medial surface of the calcanei, woven bone on

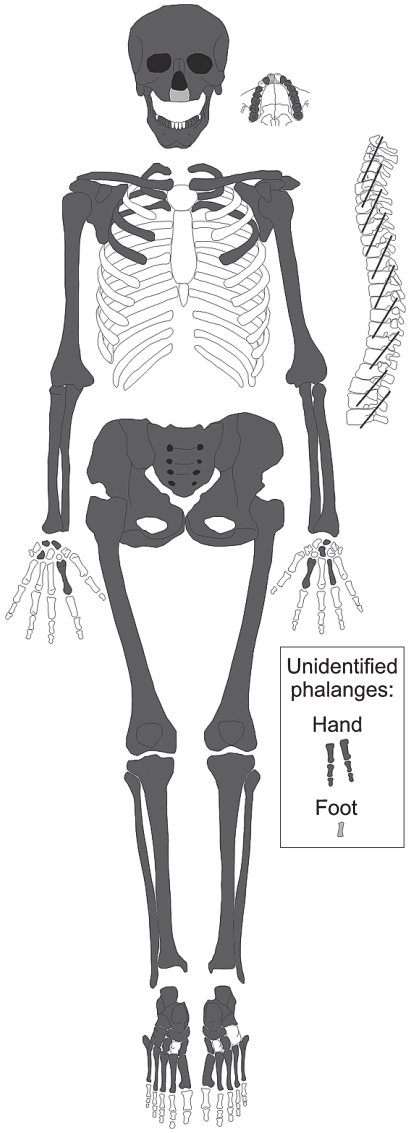

Figure 1. Visual inventory of skeletal elements and pathological lesions of individual 572. Preserved skeletal elements (in dark gray and oblique lines) and lesions in the rhinomaxillary area, maxillary anterior tooth with leprogenic odontodysplasia, and a foot phalange with acro-osteolysis (in light gray).

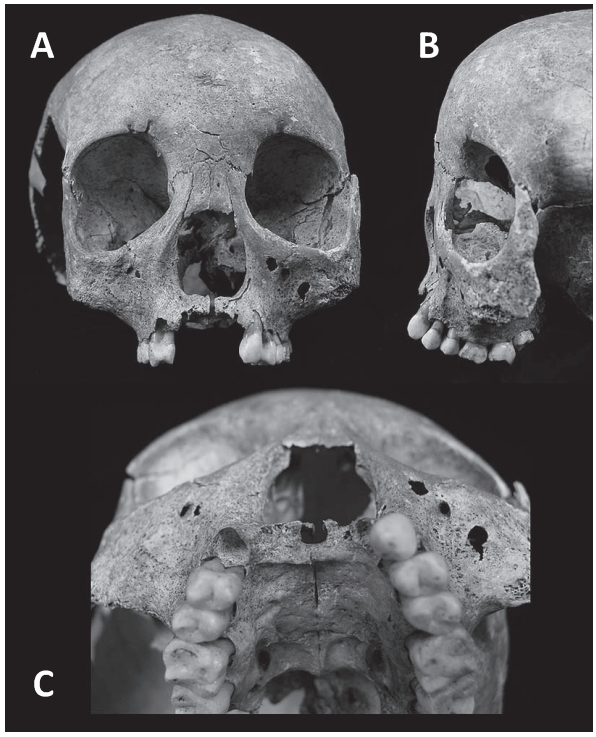

Figure 2. Cranium of the individual 572 presenting rhinomaxillary changes. A, B, and C are the anterior, left lateral and inferior views, respectively.

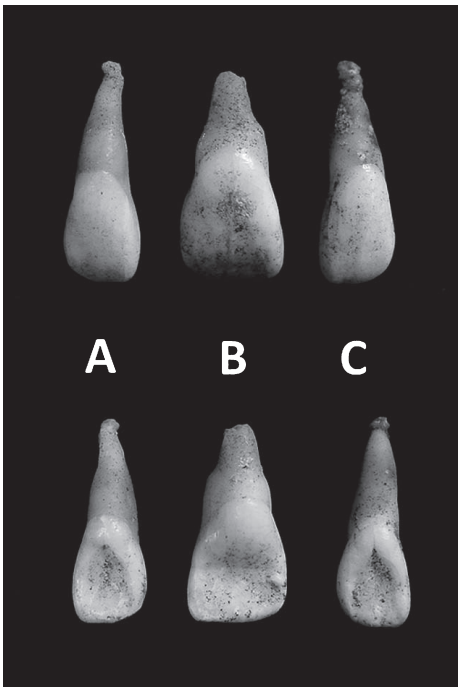

Figure 3. Labial (top) and lingual views (bottom) of the maxillary incisors. Lateral incisors (A and C) possess longer, regular root cones when compared to the right central incisor (B), whose smaller, constricted, and blunt root is compatible with leprogenic odontodysplasia.

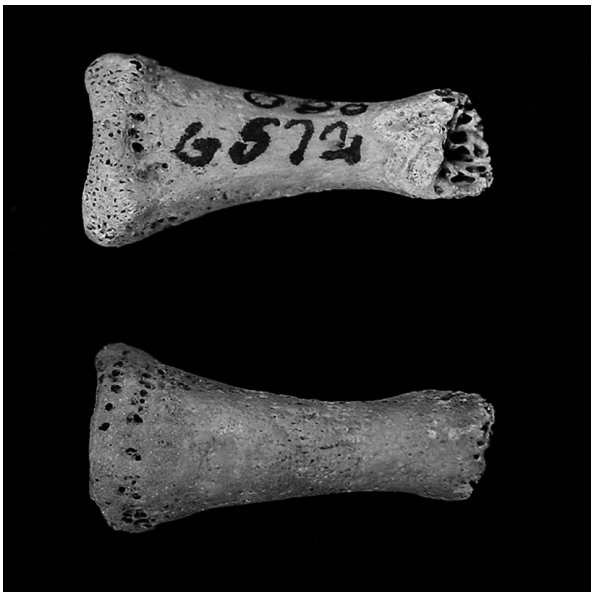

Figure 4. Proximal phalange of the foot showing distal acro-osteolysis in plantar (A) and dorsal view (B).

the diaphysis of the left 1st to 3rd and right 2nd to 4th metatarsals, and acro-osteolysis of the proximal phalanx, with nicking of its distal end (Figure 4).

Discussion and Conclusions

In the St. Jørgen's skeletal sample analyzed, only one (0.5%) juvenile was identified with a root anomaly, namely in the upper right central incisor (Figure 3), that is consistent with the description of LO made by Danielsen in 1968. Moreover, this individual exhibits a combination of lesions, namely rhinomaxillary changes and acro-osteolysis in a foot phalanx. None of these lesions by itself is pathognomonic of leprosy. For instance, rhinomaxillary changes can also be caused by treponemal diseases, cutaneous tuberculosis, leishmanioses, malignant neoplasms, among others (Hackett, 1976; Cook, 2002) and phalangeal acro-osteolysis occurs in diseases such as diabetes, Raynaud's syndrome, frostbite and treponematosis (Ortner, 2003; Powell and Cook, 2005). Nevertheless, the onset of skeletal involvement on these diseases is more common in adult individuals. None of the above-mentioned conditions presents simultaneously rhinomaxillary changes and phalangeal destruction. Thus, when these lesions coexist the most plausible diagnosis is lepromatous leprosy (Ortner, 2008a, b; Waldron, 2009), such as in this case.

The paleopathological evidence of LO was described by Danielsen (1968, 1970) in Danish skeletons and by Kjellström (2012) in a 11 or 12 year old individual from the Swedish medieval cemetery of Sigtuna. As yet, no other cases have been found in cemeteries associated with leprosaria such as the St. James and St. Mary hospital in Chichester, UK (Ortner, 2008b), and there are no clinical descriptions. Moreover, in the 300 clinical files studied from the archive of Portuguese National Leprosarium Rovisco Pais, LO was never mentioned by the physicians (Matos, 2009). In addition, this tooth anomaly is not mentioned in leprosy textbooks (Carayon and Dharmendra, 1985; Hastings, 1989; Gelber, 2006; Sehgal, 2006) or in a recent revision (Freiman et al., 2009) regarding the dental manifestations related to dermatological conditions. Due to its rarity, the etiological and epidemiological significance of the condition is poorly understood.

LO is characterized by the presence of the above-mentioned concentric constriction groove that represents a morphological feature distinct from other short root anomalies. The condition designated as short root anomaly or SR anomaly (Lind, 1972) occurs mostly in maxillary central incisors (Apajalahti et al., 2002) and is described in modern clinical literature (de Man, 1979; Apajalahti et al., 2002; Roinioti and Stefanopoulos, 2007). According to Turp and Alt (1998), variations in the root size and form of permanent teeth are determined by separate hereditary factors; however, tooth germ distortion or injury by trauma is also a possible origin of short roots (Hillson, 2005). Other possible etiologies of root shortening have been discussed in detail elsewhere (see Šikanjić and Meštrović, 2006).

The development of LO takes place when the onset of skeletal involvement due to leprosy begins at the age of dental root formation of the affected tooth (Roberts, 1986; Roberts and Manchester, 2005; Ortner, 2008b). The etiology of LO and the atrophy of the anterior maxillary alveolar process are, according to Danielsen (1970), thought to be caused by bacterial invasion of the pulp cavity and may occur in lepromatous leprosy patients (Sakai and Matsumoto,

1968). Both areas possess the lower body temperature essential for *Mycobacterium leprae* survival and multiplication (Rendall et al., 1976).

Based on the current state of knowledge, LO occurrence is concomitant with rhinomaxillary changes, and has only been found in juvenile individuals from Scandinavian medieval leprosaria cemeteries. Thus, important questions still need to be addressed: (i) Is LO pathognomonic of leprosy? (ii) If so, does LO indicate childhood onset of clinical leprosy in medieval Scandinavia, especially before 6 years of age, suggested by the age of development for upper incisor roots between 5–6 years and 10–11 years (Smith, 1991)? (iii) Why is LO only documented in juveniles? Does it mean that those individuals who contracted leprosy in early ages and developed LO were unlikely to reach adulthood? Is it possible that the shortened root paired with alveolar resorption could predispose the tooth to antemortem loss, such that LO is never found in association with older remains, even if it was present during development?

Juvenile 572 from the Odense St. Jørgen's leprosarium cemetery represents a significant paleopathological case not only because LO is an extremely rare finding, but the changes attributed to rhinomaxillary syndrome (Andersen and Manchester, 1992) or facies leprosa (Møller-Christensen, 1961) are uncommon in juveniles (Lewis, 2002, 2008). The rarity of rhinomaxillary changes in juveniles can be explained in light of the epidemiology of leprosy. Firstly, the peak incidence of leprosy usually occurs in older adolescents and young adults (Noorden, 1989; Irgens et al., 1990). This trend is attested by the proportion of juveniles among new leprosy cases in 2010, ranging from 0.85% in Argentina to 44.55% in the Marshall Islands (World Health Organization, 2011). Secondly, and most importantly, the onset of skeletal involvement in leprosy is often a later event (Paterson, 1961; Mallac, 1966; Møller-Christensen, 1974). As the early epidemiological study by Paterson (1961) reveals, bone changes were much more common in those patients for whom the onset of leprosy occurred five or more years before the detection of skeletal damage.

LO seems to represent a challenge to both paleopathology and current medical knowledge. The six cases so far reported have all occurred in permanent upper incisors of juvenile individuals. These teeth are often underrepresented in archeological skeletons because single rooted teeth easily become detached from the alveoli and are lost post mortem. Additionally, maxillary bone resorption is common in those individuals who suffered from leprosy (Andersen, 1969; Andersen and Manchester, 1992; Roberts and Manchester, 2005; Ortner, 2008b; Matos, 2009) and consequently anterior teeth are more prone to be lost ante mortem (Møller-Christensen, 1965; Núñez-Martí et al., 2004; Ogden and Lee, 2008), a phenomenon that would be expedited by a shortened root.

Among archeological skeletons, only a systematic radiographic assessment of root size and shape in situ would allow a more accurate estimation of LO prevalence in individuals from leprosaria cemeteries and other cemeteries. A search for clinical evidence, namely from current patients from endemic countries, would also help to ascertain if LO is definitely a rare condition or an underdiagnosed and potentially pathognomonic trait of leprosy. If so, LO could prove useful to diagnose early-onset leprosy in the absence of other visible skeletal pathological lesions known to be associated with the disease.

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References

Andersen J. (1969) Studies in the mediaeval diagnosis of leprosy in Denmark: an osteological, historical, and clinical study. Danish Medical Bulletin, 16: 1–142.

Andersen J. and Manchester K. (1992) The rhinomaxillary syndrome in leprosy: a clinical, radiological and palaeopathological study. International Journal of Osteoarchaeology, 2: 121–129.

Apajalahti S., Holtta P., Turtola L., and Pirinen S. (2002) Prevalence of short-root anomaly in healthy young adults. Acta Odontologica Scandinavica, 60: 56–59.

Arentoft E. (1999) De spedalskes hospital: udgravning af Sankt Jørgensgården i Odense. Udgivet af Odense Bys Museer, Odense.

Boldsen J.L. (2001) Epidemiological approach to the paleopathological diagnosis of leprosy. American Journal of Physical Anthropology, 115: 380–387.

Carayon A. and Dharmendra S. (1985) Bone and joint changes in leprosy. In: Dharmendra S. (ed.), Leprosy. Samant and Company, Bombay, pp. 872–885.

Cardoso H.F.V. (2005) Patterns of growth and development of the human skeleton and dentition in relation to environmental quality: a biocultural analysis of a 20th century sample of portuguese documented subadult skeletons. Ph.D. thesis, McMaster University, Hamilton, Ontario, Canada.

Cardoso H.F.V. (2008a) Age estimation of adolescent and young adult male and female skeletons II, epiphyseal union at the upper limb and scapular girdle in a modern Portuguese skeletal sample. American Journal of Physical Anthropology, 137: 97–105.

Cardoso H.F.V. (2008b) Epiphyseal union at the innominate and lower limb in a modern Portuguese skeletal sample, and age estimation in adolescent and young adult male and female skeletons. American Journal of Physical Anthropology, 135: 161–170.

Cook D.C. (2002) Rhinomaxillary syndrome in the absence of leprosy: an exercise in differential diagnosis. In: Roberts C., Lewis, M., and Manchester K. (eds.), The Past and Present of Leprosy: Archaeological, Historical, Palaeopathological and Clinical Approaches. Archaeopress—BAR International Series 1054, Oxford, pp. 81–88.

Coqueugniot H. and Weaver T.D. (2007) Brief communication: infracranial maturation in the skeletal collection from Coimbra, Portugal: new aging standards for epiphyseal union. American Journal of Physical Anthropology, 134: 424–437.

Danielsen K. (1968) Leprogenic odontodysplasia ('dens leprosus') [abstract]. International Journal of Leprosy, 36: 666–667.
Danielsen K. (1970) Odontodysplasia leprosa in Danish mediaeval skeletons. Tandlaegebladet, 74: 603–625.
de Man K. (1979) Abnormal root development, probably due to erythema multiforme (Stevens–Johnson syndrome). International Journal of Oral Surgery, 8: 381–385.
Freiman A., Borsuk D., Barankin B., Sperber G.H., and Krafchik B. (2009) Dental manifestations of dermatologic conditions. Journal of the American Academy of Dermatology, 60: 289–298.
Gelber R.H. (2006) Leprosy (Hansen's disease). In: Kasper D.L. (ed.), Harrison's Principles of Internal Medicine. McGraw-Hill, Blacklick, OH, pp. 966–972.
Hackett C.J. (1976) Diagnostic criteria of syphilis, yaws and trepanarid (treponematosis) and some other diseases in dry bone (for use in osteo-archaeology). Springer Verlag, Berlin.
Hastings R.C. (1989) Leprosy. Churchill Livingstone, Longman Group, Edinburgh.
Hillson S. (2005) Teeth. Cambridge University Press, Cambridge.
Irgens L.M., Caeiro F.M., and Lechat M.F. (1990) Leprosy in Portugal 1946–80: epidemiologic patterns observed during declining incidence rates. Leprosy Review, 61: 32–49.
Kjellström A. (2012) Possible cases of leprosy and tuberculosis in medieval Sigtuna, Sweden. International Journal of Osteoarchaeology, 22: 261–283.
Lewis M.E. (2002) Infant and childhood leprosy: present and past. In: Roberts C., Lewis M., and Manchester K. (eds.), The Past and Present of Leprosy: Archaeological, Historical, Palaeopathological and Clinical Approaches. Archaeopress—BAR International Series 1054, Oxford, pp. 163–170.
Lewis M.E. (2008) The children. In: Magilton J., Lee F., and Boylston A. (eds.), 'Lepers Outside the Gate': Excavations at the Cemetery of the Hospital of St. James and St. Mary Magdalene, Chichester, 1986–87 and 1993. Council for British Archaeology, Chichester excavations 10. CBA research report 158, York, pp. 174–187.
Lind V. (1972) Short root anomaly. European Journal of Oral Sciences, 80: 85–93.
Mallac M.J. (1966) Onset and pattern of deformity in leprosy. Leprosy Review, 37: 71–91.
Matos V. (2009) O diagnóstico retrospectivo da lepra: complementaridade clínica e paleopatológica no arquivo médico do Hospital-Colónia Rovisco Pais (Século XX, Tocha, Portugal) e na colecção de esqueletos da leprosaria medieval de St. Jørgen's (Odense, Dinamarca). Ph.D. thesis, University of Coimbra, Coimbra.
Matos V. and Santos A.L. (2011) Leprogenic odontodysplasia: new evidence of a rare and poorly understood malformation from the St. Jorgen's medieval leprosarium cemetery (Odense, Denmark). American Journal of Physical Anthropology, Suppl. 52: 208. Poster avalilable at http://www.uc.pt/en/cia/grupos/app/Posters/Matos_Santos_2011#Matos_Santos_2011
Møller-Christensen V. (1961) Bone Changes in Leprosy. Munksgaard, Copenhagen.
Møller-Christensen V. (1965) New knowledge of leprosy through paleopathology. International Journal of Leprosy, 33: 603–610.
Møller-Christensen V. (1974) Changes in the anterior nasal spine and the alveolar process of the maxillae in leprosy: a clinical examination. International Journal of Leprosy, 42: 431–435.
Noorden S.K. (1989) The epidemiology of leprosy. In: Hastings R.C. (ed.), Leprosy. Churchill Livingstone, Longman Group, Edinburgh, pp. 15–30.
Núñez-Martí J.M., Bagán J.V., Scully C., and Peñarrocha M. (2004) Leprosy: dental and periodontal status of the anterior maxilla in 76 patients. Oral Diseases, 10: 19–21.
Ogden A. and Lee F. (2008) Dental health and disease. In: Magilton J., Lee F., and Boylston A. (eds.), 'Lepers Outside the Gate': Excavations at the Cemetery of the Hospital of St. James and St. Mary Magdalene, Chichester, 1986–87 and 1993. Council for British Archaeology, Chichester excavations 10. CBA research report 158, York, pp. 188–197.
Ortner D.J. (2003) Identification of Pathological Conditions in Human Skeletal Remains. Academic Press, New York.
Ortner D.J. (2008a) Differential diagnosis of skeletal lesions in infectious diseases. In: Pinhasi R. and Mays S. (eds.), Advances in Human Palaeopathology. John Wiley & Sons, Chichester, pp. 57–76.
Ortner D.J. (2008b) Skeletal manifestations of leprosy. In: Magilton J., Lee F., and Boylston A. (eds.), 'Lepers Outside the Gate': Excavations at the Cemetery of the Hospital of St. James and St. Mary Magdalene, Chichester, 1986–87 and 1993. Council for British Archaeology, Chichester excavations 10. CBA research report 158, York, pp. 198–207.
Paterson D.E. (1961) Bone changes in leprosy: their incidence, progress, prevention, and arrest. International Journal of Leprosy, 29: 393–422.
Powell M.L. and Cook D.C. (2005) Treponematosis: inquiries into the nature of a protean disease. In: Powell M.L. and Cook D.C. (eds), The Myth of Syphilis: The Natural History of Treponematosis in North America. University Press of Florida, Gainsville, FL, pp. 9–62.
Rendall J.R., McDougall A.C., and Willis L.A. (1976) Intra-oral temperatures in man with special reference to involvement of the central incisors and premaxillary alveolar process in lepromatous leprosy. International Journal of Leprosy and Other Mycobacterial Disease, 44: 462–468.
Roberts C. (1986) Leprogenic odontodysplasia. In: Cruwys E. and Foley R.A. (eds.), Teeth and Anthropology. BAR International Series 291, Oxford, pp. 137–147.
Roberts C. and Manchester K. (2005) The Archaeology of Disease. Sutton Publishing. London.
Roinioti T.D. and Stefanopoulos P.K. (2007) Short root anomaly associated with Rothmund–Thomson syndrome. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology & Endodontics, 103: e19–22.
Sakai T. and Matsumoto S. (1968) Electron microscope study of the dental pulp of lepromatous patients. International Journal of Leprosy and Other Mycobacterial Disease, 36: 309–317.
Sehgal A. (2006) Leprosy. Chelsea House Publishers, Philadelphia.
Šikanjić P.R. and Meštrović S. (2006) A case of short-root anomaly in a female from medieval Istria. International Journal of Osteoarchaeology, 16: 177–180.
Smith B.H. (1991) Standards of human tooth formation and dental age assessment. In: Kelley M. and Larsen C.S. (eds.), Advances in dental anthropology. Wiley-Liss, New York, pp. 143–168.
Turp J.C. and Alt K.W. (1998) Anatomy and morphology of human teeth. In: Alt K.W, Rösing F.W., and Teschler-Nicola M. (eds.), Dental Anthropology: Fundamentals, Limits, and Prospects. Springer Verlag, Vienna, pp. 71–94.
Ubelaker D. (1989) Human Skeletal Remains: Excavation, Analysis, Interpretation. Taraxacun, Washington D.C.
Waldron T. (2009) Palaeopathology. Cambridge University Press, Cambridge.
World Health Organization (2011) Global leprosy situation, 2011. Weekly Epidemiological Record, 86: 389–400.