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Notes to Palaeopathological Diagnostics of Children's Diseases

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ABSTRACT

Children represent an important component of the population of every society, which is why all medical anthropological studies pay considerable attention to children's skeletons. This paper presents an overview of the current knowledge of the diagnosis and interpretation of symptoms of various disorders found on children's skeletons, with an emphasis on infancy. A detailed knowledge of the developmental stages of bone is required when analyzing children's skeletal remains in order to avoid mistaking developmental changes for pathological ones. In the past considerable deformities of children's skeletons were caused by metabolic diseases, such as rickets (vitamin D deficiency) or scurvy (vitamin C deficiency). Partial fractures are the most frequent trauma related changes, complete fractures less so, given the elasticity of children's bones. Injuries often cause disproportional bone growth while in some cases they heal almost without a trace. Inflammatory changes in children's bones are caused most frequently by tuberculosis or, later in life, by the congenital form of syphilis, though rarely by osteomyelitis. Sets of children's skeletons allows for a deeper study of some congenital skeletal defects, such as premature ossification of the cranial sutures and spina bifida. In order to interpret findings correctly, it is important to understand the living and climatic conditions of the population being investigated, the society's approach to child care and upbringing, characteristics of childhood diseases of the particular historic period, and the level of medical care.

1. Introduction

The human organism undergoes quite a number of significant changes in the course of ontogenetic development that are most notable in childhood. Consequently, there are morphological characteristics (form and size of individual organs and their topographical relations) as well as physiological characteristics (especially organs' efficiency changes) specific to age. A child's organism usually reacts to the influence of pathogenic factors in a different way than an adult, therefore several diseases occur only in children, or present differently between childhood and maturity.

The examination of pathological changes in children's skeletal remains brings specific difficulties, partially due to the fact that the more fragile bones of children can be more easily damaged post-mortem. This paper presents

an overview of current knowledge of diagnosis and the interpretation of symptoms of various diseases that manifest on children's skeletons, with an emphasis on infancy.

2. Material

More than 644 children's full and partial skeletons, from seven Moravian locations, and various historical periods, underwent medical anthropological analysis. The oldest skeletal set came from Blatec (9th–7th centuries BC) and from Líbivá u Břeclavi (5th–6th centuries AD). Other infant skeletons examined include remains from "Velký Špalíček" in Brno (14th–15th centuries AD), a set of bones from the ossuary in Křtiny (13th–18th centuries AD), remains from Veselí nad Moravou (16th–18th centuries AD), from the monastery cemetery of the Merciful Brothers Hospital in Brno (18th century AD), and from the former Municipal Cemetery in Malá Nová Street in Brno (1785–1883).

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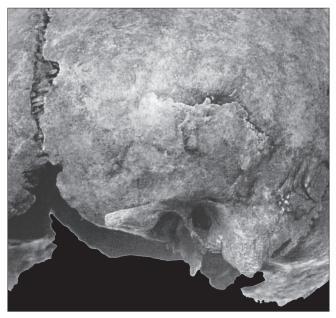


Figure 1. Premature obliteration of the squamous suture between the left temporal and parietal bones, approx. 14-years boy (K 261, 18th century, Cemetery of the Merciful Brothers Hospital in Brno).

3. Methods

Palaeopathological diagnostics were based on standard anthropological analysis of children's skeletal remains using standard morphoscopic and anthropometric methods that are outlined in the following literature: (Flecker 1932–33; Martin, Saller 1957; Stloukal, Hanáková 1978; Ubelaker 1987; Knussmann 1988; Florkowski, Kozlowski 1994; Stloukal *et al.* 1999).

The assessment of palaeopathological findings was based primarily on criteria used by Steinbock 1976; Zimmermann, Kelly 1982; Ortner, Putschar 1985; Aufderheide, Rodríguez-Martín 1998; Vyhnálek *et al.* 1998; Horáčková *et al.* 2004.

Palaeopathological diagnosis was based primarily on detailed macroscopic examination, radiological examination (plain X-ray or CT), and histological examination through light or electron microscope. Genetic examination (Mycobacterium tuberculosis DNA detection) was used where indicated.

4. Results and discussion

A detailed knowledge of the developmental stages of individual bones is the basic prerequisite for paleopathological analysis of children's skeletons. Development changes can be easily mistaken for pathological symptoms, if agerelated growth specifics are not taken into account. Though more detailed textbooks of anatomy and osteology (such as Borovanský *et al.* 1972, 57–161; Čihák 1987, 99–234; and Stloukal *et al.* 1999, 235–339) provide some data about gradual skeleton maturation, complete visual documentation

of individual bones at various developmental stages are largely unavailable. Pictures of a new-born's skeleton shown in detail in Crelin's atlas (1969, 256) represent an exception in this respect. Complete illustrations of individual bones in later stages of children's development are largely unavailable. The book written by Baker *et al.* (2005) is a rare, work on this topic, though not fully comprehensive.

As with adult skeletal remains, a number of diseases can also be noted on children's skeletal remains, such as congenital skeletal abnormalities, traumas, endocrinous and metabolic diseases including blood diseases, and specific as well as non-specific inflammations. Tumours and other less frequent diseases are rarely found on children's skeletons.

Congenital bone anomalies or malformations are defined as congenital abnormalities of bone shape that are beyond variability limits and are likely to cause health problems to afflicted individuals. They can be caused by heredity or damage to an embryo by physical, chemical or biological factors during intrauterine development. Changes of shape may affect one bone only or they may occur on several bones of one skeleton. Multiple anomalies are usually part of more serious, genetically based developmental syndromes. Taking into account that the frequency of these syndromes is relatively low in the population and that the afflicted individuals as a rule die in early childhood, discoveries of congenital bone anomalies are sporadic even in large osteological sets.

A prematurely closed left squamous suture on the skull of an approximately fourteen year old boy from the 18th century monastery cemetery of the Merciful Brothers Hospital in Brno (Figure 1) is one example of congenital bone anomaly. Individual cranial sutures close progressively in a certain time sequence as an individual matures. (Hajniš, Novák 1984, 112–124). Although considerable individual variability exists in the closing of cranial sutures, the closure of the squamous suture in particular occurs in persons over sixty years old, as described in all studies dealing with this matter. The child's skull, with a closed squamous suture, definitely represents a pathological condition. The cause of premature closure of cranial sutures has not yet been fully clarified. The craniosynostosis is described as a symptom of some rare congenital disorders, such as Apert's, Pfeiffer's, Crouzon's, and Carpenter's syndromes (Žižka 1994, 394) or as a symptom of some acquired syndromes (Cohen 1976, 372-382). Considering the monastery hospital's focus on mental patients (the Merciful Brothers Hospital in Brno was a branch of St. Ann's Provincial Public Hospital dedicated entirely to insane men according to the contract from 1804) it is highly probable that the premature closure of the cranial suture was related to some mental illness (Vargová, Zapletalová 2007, 128–137).

Signs of anaemia are the most common blood diseases found on children's skeletons. Many palaeopathologists (for example Moseley 1963, 121–130) are inclined to agree that porotic changes on the orbital roof referred to as cribra orbitalia, or usura orbitae (Møller-Christensen, Sandison 1963, 175–183) or hyperostosis spongiosa orbitae (Hengen 1971, 57–76) belong to symptoms of sideropenic anaemia.



These are characteristic for bone destruction and subsequent formation of new bone in the front part of the orbital roof, in the form of small areas with a porotic or spongious structure. The hypertrophic diploe in this case pushes a thin cortical layer of orbital roof into the orbit and the excessive spongious bone penetrates into it (Figure 2). Lesions of the orbital roof can be divided into three basic types according to the morphological appearance representing their different stages of development: porotic, cribrotic, and trabecular (Horáčková et al. 2004, 145-146). It is known from paleopathological research that the frequency of cribra orbitalia differs in individual historical populations, but usually hovers around 20% as a rule. Most population studies confirm the highest occurrence of cribra orbitalia occurs on children's skulls in particular, as was the case of the Early Modern skeletal remains from the former Municipal Cemetery in Malá Nová Street in Brno (Vargová, Horáčková 2006, 431–438). The frequent occurrence of children's anaemia probably results from iron deficient food (e.g., the consumption of iron-poor goat's milk) or from a disorder of iron metabolism due to various diseases, such as inflammation of stomach mucosa or intestinal mucosa, kidney inflammations and parasitical diseases.

A sufficient amount of iron in food, and a balance of other dietary components are important for the proper development of a growing child's skeleton. Vitamin D is one of the most important components as its deficiency causes rickets. This vitamin ensures sufficient absorption of calcium from food. Its absence leads to a decrease in the calcium level in blood, which is compensated for by the release of calcium and phosphorus from the bones. This results in a deficiency of inorganic substances in the intercellular matter of bone tissue and in loss of bone strength, leading to distinct bone deformities at the lower extremities that can bend even with normal physiological loads - under their own body weight for example (Bayer et al. 2002, 195-216). Traces of rickets were observed in as many as 80% of children under two years of age in industrial areas of developed countries at the end of the 19th century (Welch et al. 2000, 143-145). With its many textile factories Brno belonged to one of the most developed industrial towns of the Austro-Hungarian Empire in the 19th century. Symptoms of rickets were observed in two children's skeletons in the 19th century osteological set from the former Municipal Cemetery in Malá Nová Street in Brno – these skeletons belonged to a $2\frac{1}{2}$ –3 year old child from grave A848, and an approx. 6 year old child from grave A1857, Figure 3 (Vargová, Horáčková 2006, 431-438). Rickets symptoms in both afflicted skeletons showed the typical sagging of the middle parts of diaphyses of the long bones of the extremities, with a club-shaped widening of their metaphyseal ends (so-called Marfan's sign). The two typical examples of rickets found represent only 0.5% of the 440 children's skeletons examined from the Brno location. The poor preservation of afflicted children's remains may be considered the main reason for the low percentage of discovered cases. Moreover, rickets did not developed to the stage of bone changes in all cases of afflicted children. Some

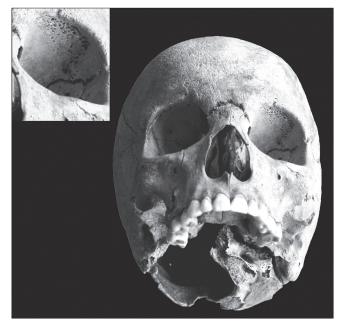


Figure 2. Porotic changes in the orbits (cribra orbitalia of IInd stage), child 14–15 years (H 1, 14th–15th centuries, Brno – Velký Špalíček).



Figure 3. Tibias of approx. 6 years-old child with remarkable bending of diaphyses (A 1857, 18th–19th centuries, Municipal Cemetery in the Malá Nová Street in Brno). The comparison of legs afflicted by rickets in the picture by Tillmanns (1895, 549).

cases of tooth enamel hypoplasia, as mentioned in the work of Vargová *et al.* (2008, 14–19) may be a possible symptom of this disease as well.

A deficiency of vitamin C may also cause marked pathological changes on a growing child's skeleton. This vitamin is necessary for the proper synthesis of collagen in connective tissues and its absence causes the disease called scurvy. Pathological changes manifest themselves by an enchondral ossification disorder, and bleeding to the muscles and hypodermis, and from the gums. Extensive



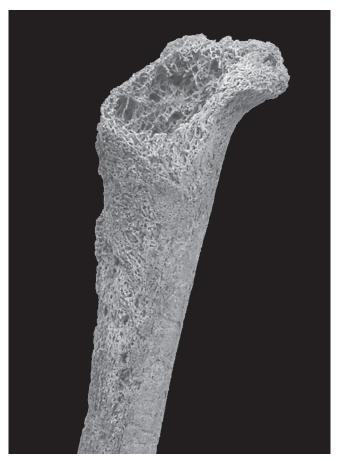


Figure 4. A detail of the left femur diaphysis with deposit of the new formed bone tissue – symptoms of scurvy; 1–1.5 years-old child (A 829, 18th–19th centuries, Municipal Cemetery in the Malá Nová Street in Brno).

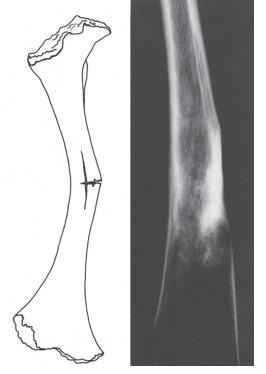


Figure 5. Scheme of a "green stick" fracture (ORTNER 2003, 123). X-ray of the infraction, the diaphysis of the right femur, child 14–16 years (K II 81, 13th–18th centuries, ossuary Křtiny).



Figure 6. A healed fracture of the right femur, 8–9 years-old child (Bt 36/99, 9th–7th centuries BC, Blatec).

depositions of newly formed bone tissue may appear at bone surfaces due to ossified subperiosteal haematomas. Fractures may be present as well. Possible traces of scurvy were observed in two children's skeletons, as described in the paleopathological analysis of skeletal remains from the Brno cemetery on Malá Nová Street (Vargová, Horáčková 2006, 431-438) (Figure 4). They were observed in a skeleton of a 4-5 year old child (grave No. A867) in the first case, where areas of newly formed bone tissue of various sizes were found on the surface of some bones (right ribs, left shoulderblade, left pelvic bone as well as the right tibia). A similar finding was also observed in diaphysis fragments of femurs from a 1-11/2 year old child from grave A829. Document sources indicate that the scurvy epidemic in the 19th century was almost worldwide, especially in orphanages (and in prisons in the case of adults) where poor sanitary conditions and food (or lack thereof) were the main reasons for the spread of this disease.



Traumas represent another group of disorders, the traces of which can be observed in some children's skeletons. The ratio between the organic component (ossein) and mineral substances changes in the bone tissue in the course of ontogenetic development. Ossein diminishes in favour of calcium salts with advancing age. Children's bones are therefore much more flexible and more resistant to fractures than bones of adults.

As a result a partial bone fracture often occurs without periost damage (subperiosteal fracture) (Figure 5) in childhood. This type of bone damage, in which the partially preserved periost holds the fragments close to each other, is also referred to in the literature as a "green stick" fracture. That is also why the dislocation of fragments in children's fractures occurs less frequently than in adults. The fracture of a femur diaphysis represents an exception, since the pull of strong thigh muscles shifts the peripheral fragment in the proximal direction distinctly and the fractures remain extremely dislocated. A well healed, dislocated, oblique fracture of the right femur diaphysis of an eight to nine year old child (Blatec, 9th–7th century BC) is an example of this (Figure 6).

Children's fractures usually heal fast and post-traumatic complications occur less often. The healing abilities of a growing child's body are better then those of an adult. It is not unusual after some childhood injuries that few traces of healing remain visible in adulthood at the original fracture site.

More significant traumatogenic skeletal deformities occur in instances of damage to the growth cartilages or bone ossification centres. When injured these parts lag behind in their development and considerable asymmetries may arise if the adjacent structures grow normally. In some cases, it is possible to estimate the age at which the fracture happened, even in adults, by the nature of the damage. A fracture of a right lower mandible of a 30–40 year old man from the Migration Period (Líbivá u Břeclavi, 4th–5th century) is an example of this. It is obvious from the distinct asymmetry that the injury happened in the childhood, in the period prior to the second acceleration of splanchnocranial growth, namely between 9th–12th year of life (Horáčková *et al.* 2001, 63–69) (Figure 7).

Literature sources suggest, however, that infectious diseases were one of main causes of death of children in various historical periods. Most of these serious diseases (for example plague, cholera, smallpox, encephalitis, and meningitis of diverse aetiologies) had a very fast course and did not affect bones. It is possible, however, to carry out genetic examinations of bone samples and to prove a pathogenic organism's DNA in some infectious diseases (despite numerous technical difficulties). Diagnosis of most infectious diseases in palaeopathology is difficult and the best results are achieved with mummified bodies.

We find traces of such infectious processes in skeletal remains, only if they are chronic (for example tuberculosis and congenital syphilis).

Tuberculosis may affect any bone of the body in childhood and in adulthood, but findings traces of tuberculous



Figure 7. The mandible of an adult man with asymetry of the both mandibular rami – result of a fracture of the right articular process between 8 and 11 years of age. (H 12, 5th-6th centuries, Líbivá u Břeclavi).

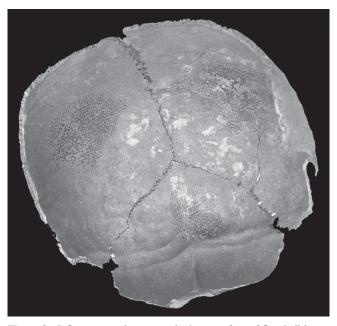


Figure 8. Inflammatory changes on the inner surface of flat skull bones (probably of a tuberculous origin), 6–7 years-old child (VnM 69, 16th–17th centuries, Veselí nad Moravou).

inflammation of meninges (tuberculous meningitis) are more frequent in children's skeletons. Pathological changes are usually characterised by small granular impressions or fine plates of newly formed bone tissue in the imprints of meningeal arteries, and sometimes these imprints are shaped like the thin branches of a small shrub (Figure 8).

The contagion of an individual with congenital syphilis occurs in the course of intrauterine development. The bone damage results from the destruction caused by a pathogenic organism, from trophic disorders of the growing skeleton, and from the subsequent repair process. Inflammatory changes may be located both at the skull (destruction of nasal and palate areas – a saddle-back nose, "caries sicca" in flat bones



Figure 9. An enamel hypoplasia of the first lower molar ("mulberry molar") – a symptom of the congenital syphilis, 6–7 years-old child (A 1897, 18th–19th centuries, Municipal Cemetery in the Malá Nová Street in Brno).

of cranial vault), and even at the long bones of extremities (periostitis, osteomyelitis); and the affliction of joints is no exception. Dental stigmas are some of the most typical symptoms of congenital syphilis. They include "Hutchinson's teeth", which are incisive teeth having barrel shaped crowns and crescent shaped incisal edges. The first molar is also quite often affected by the syphilitic process. The hypoplasia of enamel and dentin, which is most significant along the perimeter of occlusal area of the crown, changes the shape of the whole tooth. The molar looks like an unblossomed rosebud or mulberry flower (so-called Fournier's tooth or Moon's molar) (Figure 9).

5. Conclusion

No anthropological study of population is complete without a detailed paleopathological analysis of children's skeletal remains. The specifics of a child's age (morphological, physiological as well as specifics in the reactivity to pathogenic factors) significantly influence the work of anthropologists and palaeopathologists. Separated epiphyses must not be forgotten at exhumations of children's skeletons. The cleaning and handling of children's skeletons must always be done very carefully, so that these considerably fragile bones are not damaged further. A thorough knowledge of stages of skeletal development, the spectrum of diseases typical for children, and the given historical period is a matter-of-course. Anamnestic data can be substituted to some degree by basic demographic data, especially in determinations

of the mortality, because the age of the individual can be a significant clue to a correct diagnosis in the differential diagnostics. In some cases, it is also possible to ascertain the probable cause of elevated childhood mortality and of the level of health care for affected individuals in the given population in a given historical period, in addition to the diagnostics of diseases occurrence, and to completing epidemiological studies of the geographical distribution of illnesses.

Knowledge about children's health conditions based on medical anthropological analysis of children's skeletons, together with complementary data from literature and iconographical sources, contribute significantly to enriching and widening our knowledge about the life ways of populations.

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