

PALEOPATHOLOGICAL FINDINGS OF DRY SKULLS WITH PLAGIOCEPHALY

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Summary: In literature, plagiocephaly is defined as abnormal asymmetrical head shape with unilateral flattening of the anterior and/or posterior part of the skull. It is classified as synostotic or deformational. Synostotic plagiocephaly is the result of premature unilateral fusion of neurocranium sutures. Positional plagiocephaly is caused by asymmetrical intrauterine and/or postnatal external forces. The authors report findings of both types of plagiocephaly in the dry skull collection from pertaining Broumov Ossuary (13th – 18th century), Czech Republic. In the paper, the incidence of paleopathological asymmetrical skull deformities is discussed in the context with recent clinical data.

Key words: *Plagiocephaly; Craniosynostosis; Synostotic plagiocephaly; Unicoronal synostosis; Unilambdoid synostosis; Deformational plagiocephaly; Paleopathology*

Introduction

Plagiocephaly is a general term for abnormal asymmetrical cranium shape with unilateral flattening of the anterior or posterior part of the skull. The term meaning “oblique head” (“lopsided skull”) derives from the Greek word roots “plagios” and “kefalê”. Pathogenically, plagiocephaly can be classified as synostotic (organic) or deformational (positional, functional, postural, nonsynostotic) (21). Synostotic plagiocephaly is caused by premature unilateral fusion of the coronal suture (frontal or anterior synostotic plagiocephaly), lambdoid suture (occipital or posterior synostotic plagiocephaly, true occipital plagiocephaly) or lateral skull sutures (1,4,17,21). In the literature, the term “hemicranial plagiocephaly” is sometimes used for deformities associated with multiple unilateral sutural fusions (1). Deformational occipital (posterior) plagiocephaly is caused by intrauterine and/or postnatal external forces on the one side parieto-occipital region of the developing skull (13).

Morphological characteristics of the frontal plagiocephaly (Figs. 1, 2) are following (5,10,18,19): asymmetrical “rhomboid” skull shape (in vertex view), asymmetry of the skull base (predominantly of the anterior cranial fossa), ipsilateral flattening of the forehead and ipsilateral compensatory occipital bossing, contralateral compensatory bossing of the forehead (asymmetrical “pointed” forehead), asymmetry of the nose root (it is deviated towards fused coronal suture), asymmetry of the orbits (ipsilateral orbit is enlarged and elevated), and asymmetrical position of the ears (the ear of the affected side is situated anterior and superior). Three forms of frontal plagiocephaly are defined: mild form

(with minimal frontal flattening, symmetrical orbits, and a midline nose), moderate form (with ipsilateral flattening, compensatory contralateral bossing, and nasal root deviation), and severe form (with vertical orbital dystopia, severe ipsilateral flattening, contralateral bossing, and deviation of the nose root) (19). Occipital synostotic plagiocephaly (Fig. 1) is characterized by the asymmetrical “trapezoid” skull shape (in vertex view), ipsilateral flattening of the forehead and bulging of the mastoid process (it is pathognomonic for unilambdoid synostose), compensatory contralateral bossing of the forehead and occiput: “pointed” occiput, asymmetry of the ears (ipsilateral ear is situated posterior); facial asymmetry is not mostly severe (12,16,21). Occipital positional plagiocephaly (Fig. 2) is associated with asymmetrical “parallelogram” skull shape, unilateral parieto-occipital flattening, ipsilateral frontal bossing, contralateral frontal flattening and occipital bossing, asymmetry of external nose (nasal root is in a midline position but nasal bones and piriform aperture are deviated on the contralateral side), asymmetry of the orbits (contralateral orbit is situated low then ipsilateral orbit), asymmetry of the ear position (ipsilateral ear is situated anterior and superior) (9,11,13,16,21). Positional plagiocephaly can be classified as mild (without facial asymmetry) or severe (with facial asymmetry) (3).

Material and Methods

The incidence of the synostotic and/or deformational plagiocephaly was investigated in the collection of 745 dry skulls (647 skulls of adults of both sexes, 98 skulls of children) from “Broumov Ossuary” (13th–18th century), Czech

Republic. All skulls were minutely investigated by cranioscopy and craniometry. Positive findings of plagiocephaly were documented by the digital camera Camedia 3030 ZOOM, Olympus. In the part of them, X-ray and/or computed tomography (CT) examination was performed.

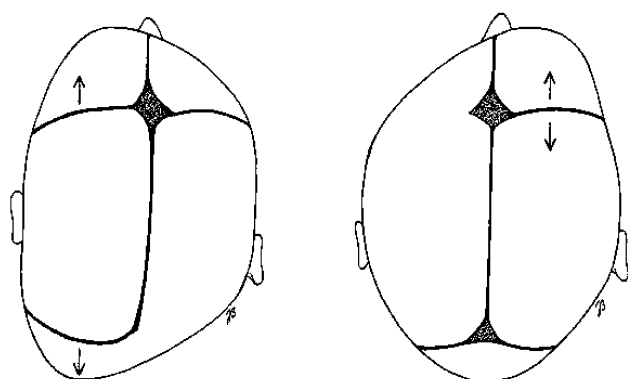


Fig. 1: Schematic picture of the skull shape deformities in the occipital (A) and frontal (B) synostotic plagiocephaly; arrows: compensational growth of the cranium.

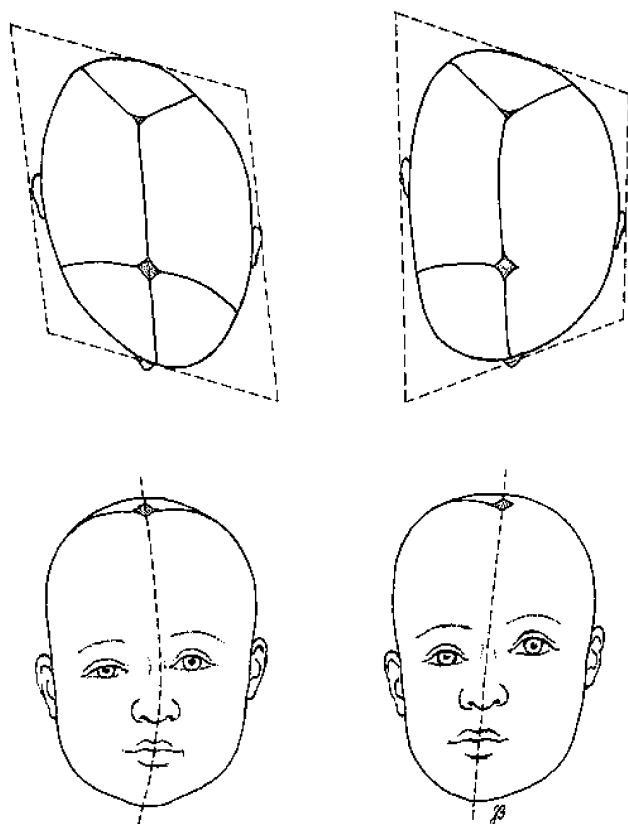


Fig. 2: Schematic drawing of morphological characterizations of the occipital positional plagiocephaly (A) and frontal synostotic plagiocephaly (B). Upper: skull shape (vertex view), lower: face asymmetry.

Results

Of the 745 examined dry skulls, 106 skulls (14.22 % – 87 adult skulls, 19 children's skulls) had evidence of plagiocephaly. From the total number of skulls, synostotic plagiocephaly (Figs. 3–7) was noticed in 13 (1.74 %) skulls (8 adult skulls, 5 children's skulls), occipital deformational plagiocephaly (Figs. 8–10) in 93 (12.48 %) skulls (79 adult skulls, 14 children's skulls). Frontal synostotic plagiocephaly with unicoronal fusion was seen in 2 (0.27 %) skulls (1 adult skull, 1 child's skull) (Fig. 3, 4). Occipital unilambdoid synostotic plagiocephaly was diagnosed in 1 (0.13 %) skull of adult individual. Synostotic plagiocephaly caused by asymmetrical obliterations of some skull sutures was observed in 7 (0.94 %) skulls (5 adult skulls, 2 children's skulls) (Fig. 5). Premature fusions of multiple cranium sutures associated with synostotic plagiocephaly and another form of synostotic skull deformity (scaphocephaly or brachycephaly) was found in 3 (0.40 %) skulls (1 adult skull, 2 children's skulls) (Figs. 6, 7).

Discussion

Presented paleopathological study is focused on the incidence of synostotic and/or deformational plagiocephaly in the dry skull collection from the Broumov Ossuary (13th– 18th century), Czech Republic. Although asymmetrical skull deformities are quite common today, not many cases have been described in the paleopathological literature (1).

In the literature, the craniosynostosis is defined as a premature fusion of one or more of cranial sutures (8). It can occur as an isolated defect (nonsyndromic) or as a part of the syndrome (e.g. Crouzon, Apert, Pfeifer, Cole-Carpenter). It may affect the coronal, sagittal, metopic, lambdoid and/or lateral sutures in isolation or in combinations (14). The skull growth is restricted predominantly in a direction perpendicular to the premature fused suture. In response to the closed suture, the remaining sutures undergo compensatory growth. Uncorrected craniosynostosis may be associated with deformity of the craniofacial skeleton and it can also cause serious constriction of the developing brain (craniostenosis), eyes (orbitostenosis), and facial viscera (faciostenosis) (14). Craniosynostosis is classified into two main categories: "simple" (or unisutural) when only one suture is involved (Fig. 11), and "complex" (compound) when two or more sutures are involved. It is usually more severe when more than one suture closes too early. The occurrence of the various types of simple craniosynostosis is approximately as follows: sagittal 50–55%, coronal 20–29%, metopic 4–10%, and lambdoid 2–4% (8).

Although the etiology of craniosynostosis is currently unknown, animal experiments and a recent interest in molecular biology point toward interplay between the dura mater and the underlying brain (14). The cranial suture fate is programmed by the dura derived paracrine signals. The dura

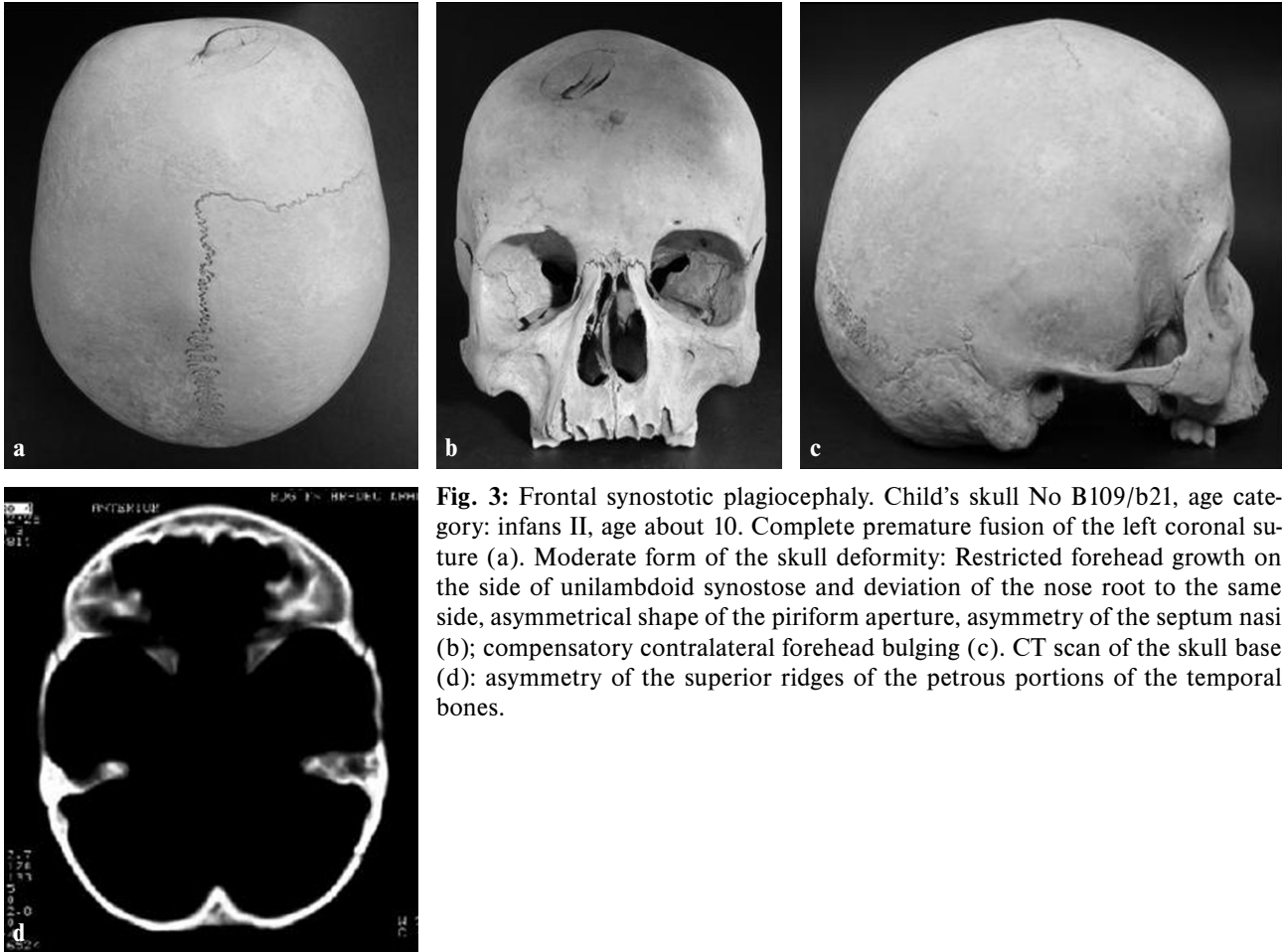


Fig. 3: Frontal synostotic plagiocephaly. Child's skull No B109/b21, age category: infans II, age about 10. Complete premature fusion of the left coronal suture (a). Moderate form of the skull deformity: Restricted forehead growth on the side of unilambdoid synostose and deviation of the nose root to the same side, asymmetrical shape of the piriform aperture, asymmetry of the septum nasi (b); compensatory contralateral forehead bulging (c). CT scan of the skull base (d): asymmetry of the superior ridges of the petrous portions of the temporal bones.

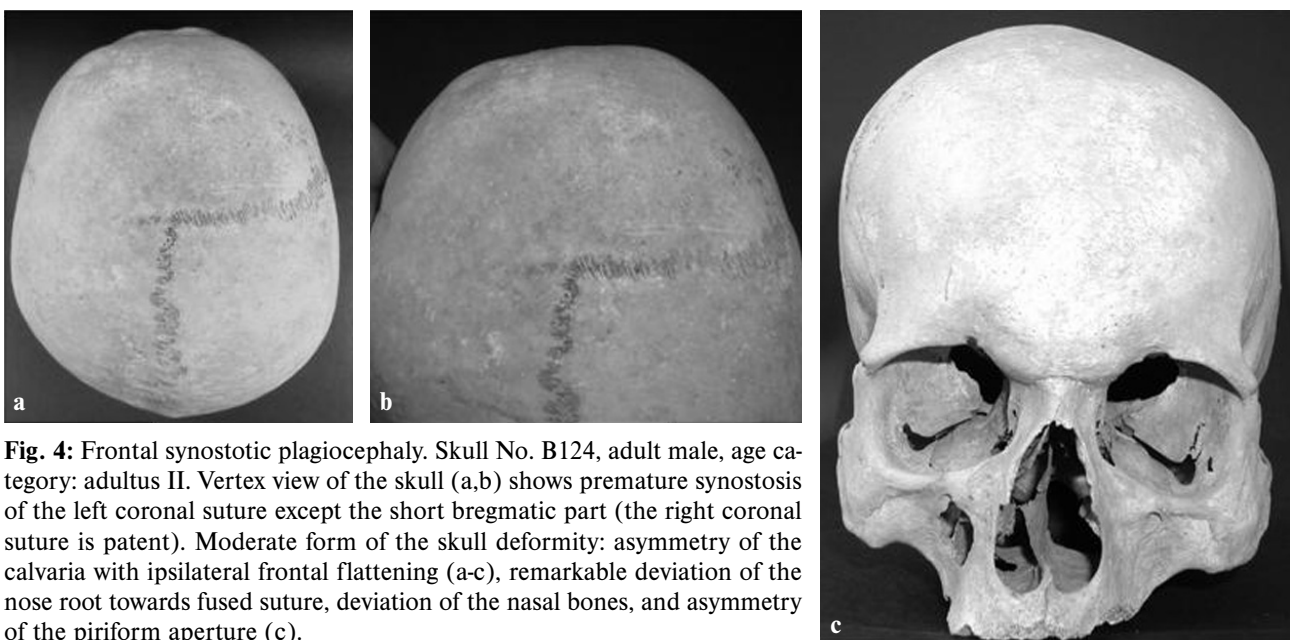


Fig. 4: Frontal synostotic plagiocephaly. Skull No. B124, adult male, age category: adultus II. Vertex view of the skull (a,b) shows premature synostosis of the left coronal suture except the short bregmatic part (the right coronal suture is patent). Moderate form of the skull deformity: asymmetry of the calvaria with ipsilateral frontal flattening (a-c), remarkable deviation of the nose root towards fused suture, deviation of the nasal bones, and asymmetry of the piriform aperture (c).

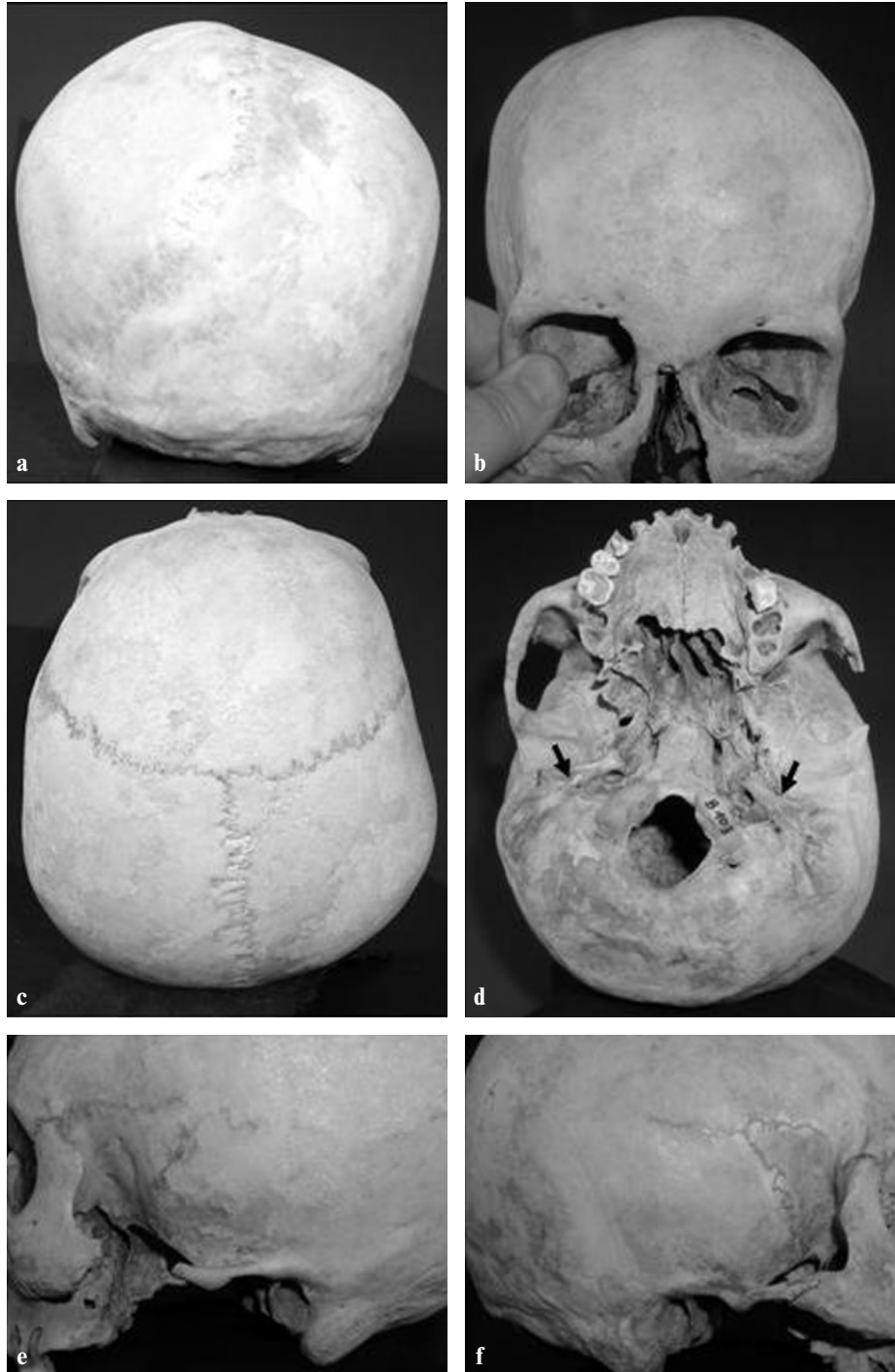


Fig. 5: Synostotic plagiocephaly associated with multiple premature sutural fusions. Skull No B103, adult male, age category: adultus II. Premature fusion of the right lambdoid suture (a), bilateral symmetrical fusion of the occipitomastoid sutures (d,e,f), bilateral partial and asymmetrical fusion of the squamous suture (e,f). Parietooccipital flattening at the side of obliterated lambdoid suture (a-d). Asymmetrical course of the sagittal suture (c) with deviation towards the affected side. Scoliosis of the external cranial base, asymmetrical position of the external acoustic meatus (d, arrows). Asymmetry of temporal fossae (e,f): on the right side, the surface of the greater wing of the sphenoid bone is shorter. Remarkable face asymmetry: asymmetrical shape of the orbits, deviation of the nose root to the side of the lambdoid synostose, asymmetrical shape of the piriform aperture (b).

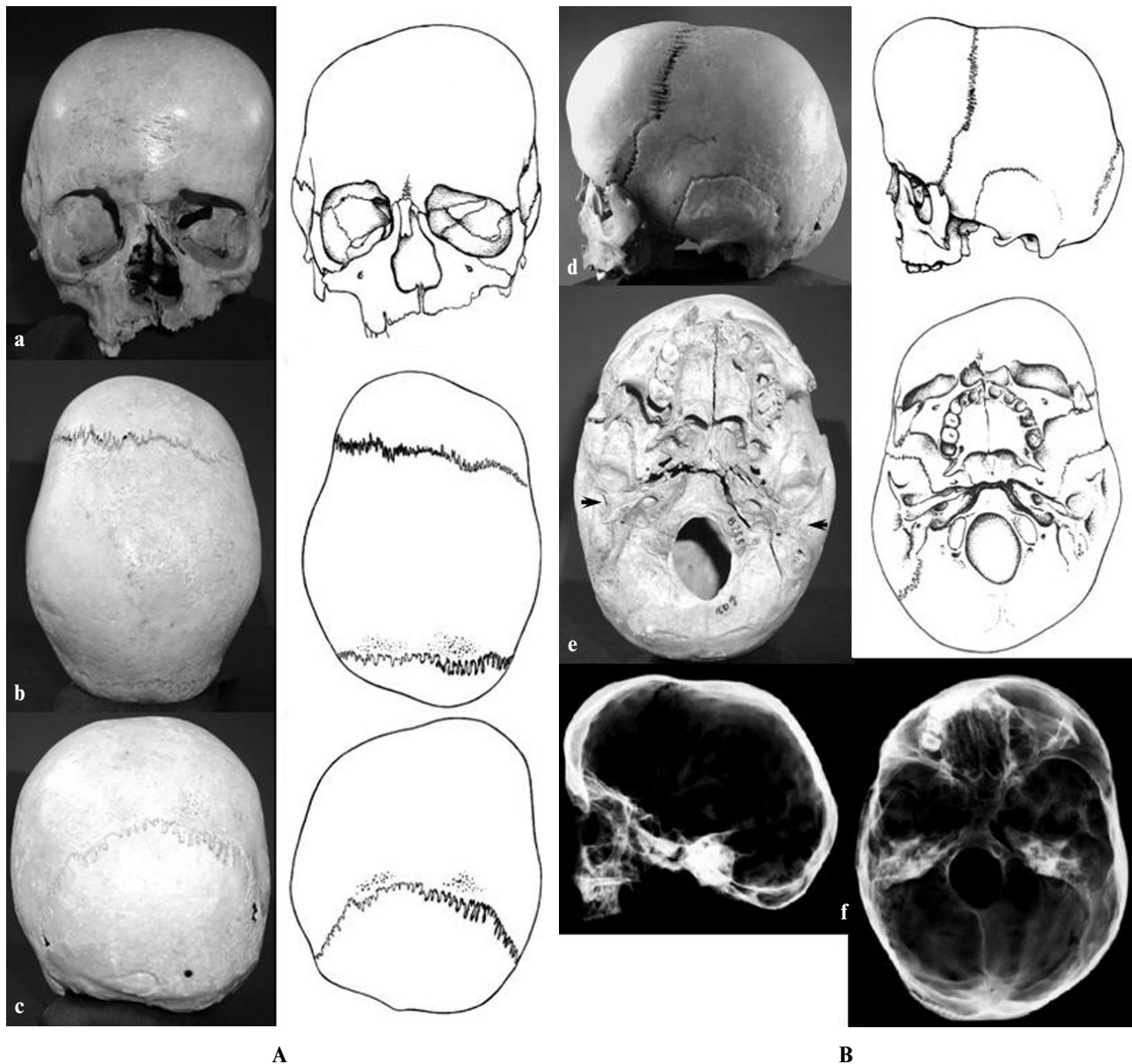
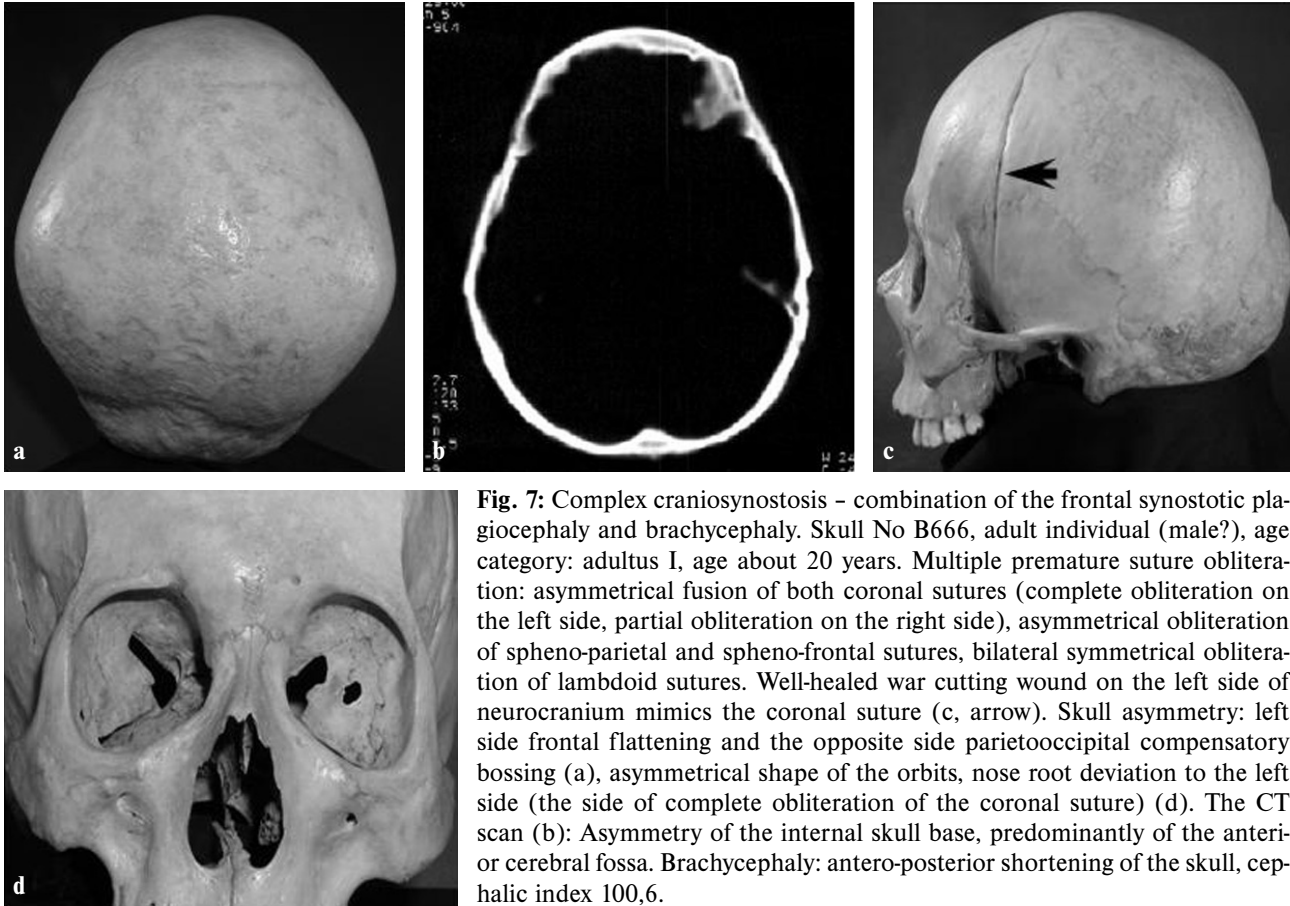


Fig. 6 A,B: Complex craniosynostosis with severe skull deformity: combination of the synostotic plagiocephaly with scaphocephaly Child's skull No B255/b92, age category: infans I, age about 5. Multiple premature suture synostosis: Partial obliteration of the right side coronal suture (A/b); complete obliteration of the sagittal suture (A/a), the left side sphenoparietal suture (B/d) and occipitomastoid suture (B/d,e); obliteration of the posterior part of the left side squamous suture (B/d). Asymmetrical deformity: Frontal flattening (A/a,b, B/e) and occipital bossing (B/e) on the side of fused coronal suture, compensatory frontal bossing on the opposite side (A/a,b, B/d,e), occipital flattening on the side of obliterated occipitomastoid and squamous sutures (A/c), asymmetry of the external acoustic meatus (B/e-arrows), asymmetrical shape of the foramen magnum (B/e); remarkably asymmetrical orbits – on the side of coronal synostosis, the orbit is enlarged and its upper margin is deviated upward (A/a); deviation of the nose root to the side of coronal fusion, asymmetry of the piriform aperture (A/a). Scaphocephaly: hyperdolichocephaly (A/b), cephalic index 74. Radiograph (B/f): long and narrow skull, asymmetry of the skull base. Distinctive impressioes digitatae are suspect from intracranial hypertension.



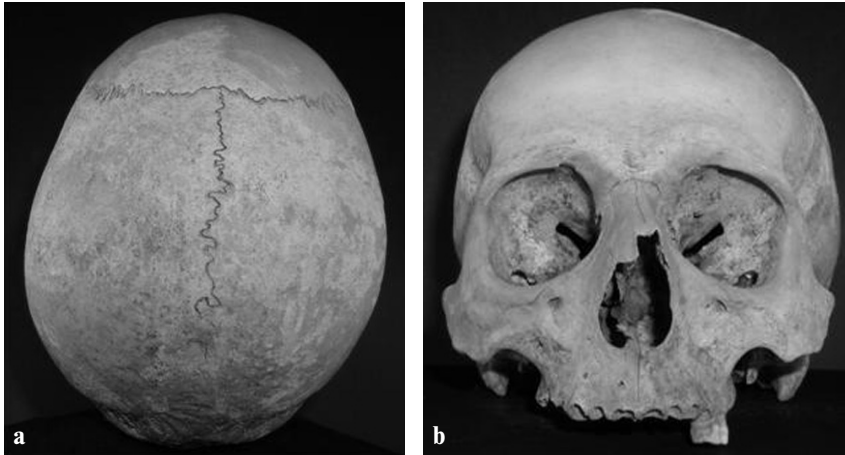


Fig. 9: Occipital positional plagiocephaly. Skull No B120, adult male, age category: adultus II. Parietooccipital skull flattening on the right side and ipsilateral compensatory frontal skull bossing. "Parallelogram" skull shape (a). Midline position of the nose root, deviation of the ossa nasalia to the left side (b).

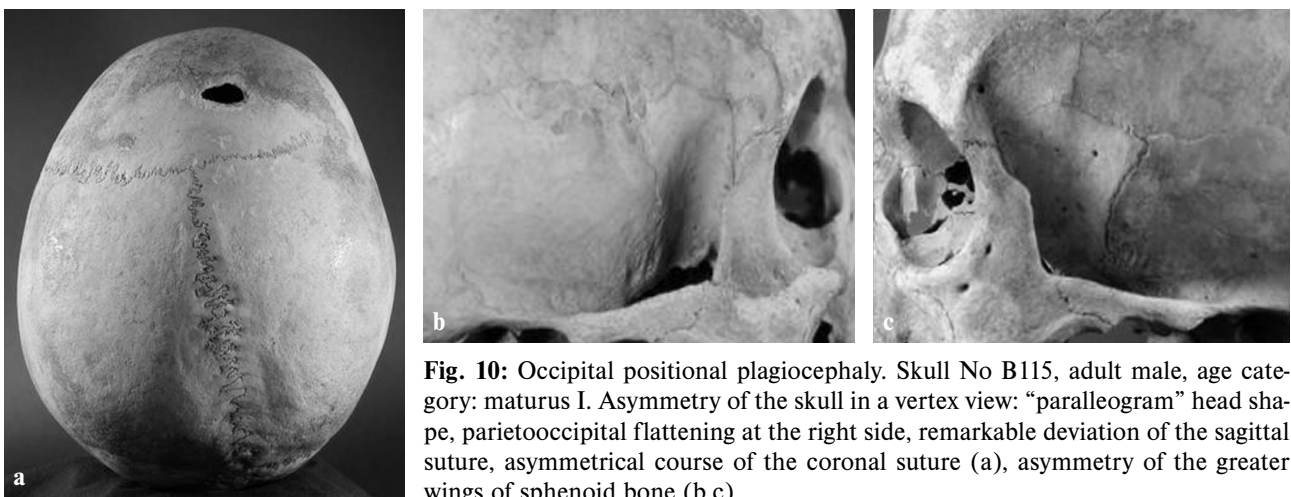


Fig. 10: Occipital positional plagiocephaly. Skull No B115, adult male, age category: maturus I. Asymmetry of the skull in a vertex view: "parallelogram" head shape, parietooccipital flattening at the right side, remarkable deviation of the sagittal suture, asymmetrical course of the coronal suture (a), asymmetry of the greater wings of sphenoid bone (b,c).

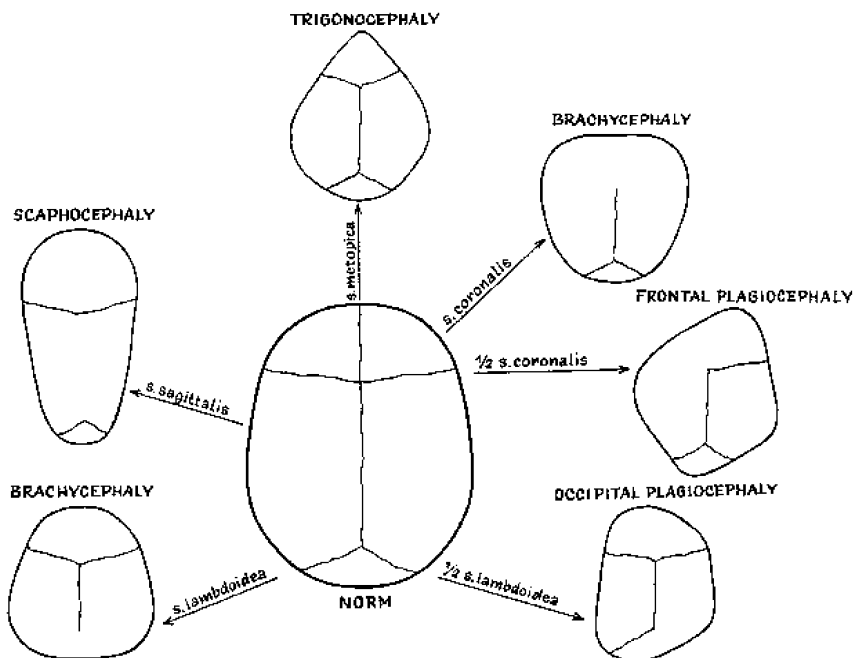


Fig. 11: Schematic drawing of simplex craniosynostosis: typical deformities of the skull.

acts as a regionally specialized endogenous tissue engineering and provides the osteogenic factors (e.g. fibroblast growth factors). These factors regulate fetal osteogenic growth and possibly influence fetal suture patency (20). Mutations in the gene coding for fibroblast growth factors cause some syndromes with craniosynostosis (2).

In the contemporaneous population, craniosynostosis has been reported to occur approximately once per 2,000 live births (8). Unicoronal craniosynostosis – scarce form of craniosynostosis – happens in approximately one out of every 2,500 live birth (2), rare unilambdoid craniosynostosis happens in approximately three out of every 100,000 births (6). In contradistinction to recent clinical studies, in Broumov historical populations dating to the 13th–18th centuries, the incidence of synostotic plagiocephaly was rather high (1.74 %): unicoronal craniosynostosis was present in 0.27 % skulls, unilambdoid craniosynostosis in 0.13 % skulls, synostotic plagiocephaly with asymmetrical fusions of some skull sutures in 0.94 % skulls, and combination of synostotic plagiocephaly with another form of synostotic skull deformity in 0.40 % skulls. Kabbani and Raghuvver (8) assume that the true incidence of craniosynostosis in population is higher than clinical studies assert. The present report supports the assumption of these authors.

Positional plagiocephaly is the most common cause of plagiocephaly (15). It results from asymmetrical cranial growth without craniosynostosis. The practice of artificial skull deformation by pressure to an infant's head dates back to 2000 BC when several ancient civilizations have used head-molding to produce desired skull shape. Positional occipital plagiocephaly is caused by constant external pressure on the one side parieto-occipital region of the developing skull (3). Risk factors associated with the positional head deformity are prolonged exposure to traditional “back” sleeping position, premature birth, congenital muscular torticollis, and intrauterine constraint (e. g. in multiple gestation or oligohydramnion) (13). Following the American Academy of Pediatrics recommendation (in 1992) that babies sleep in supine position to reduce the risk of Sudden Infant Death Syndrome, the incidence of deformational plagiocephaly has increased dramatically. Before 1992, the incidence was one of every 300 healthy born infants. Nowadays, the condition occurs in one of every 60 live birth (7). High positional plagiocephaly occurrence (12.48%) in skulls of Broumov historical populations was probably pre-

dominately caused by traditional back sleeping position of newborns and suckling infants in past centuries.

Acknowledgement

The work was partially supported by the Ministry of Education Grant Agency (Grant No. 2532/F3/02).

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Submitted November 2006.

Accepted December 2006.

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