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Cloverleaf Skull

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With 6 Figures

Summary

The authors report 3 cases of "Cloverleaf" skull, and discuss the nosological framework of this congenital deformity of the skull.

Among the premature closures of the cranial sutures that result in craniostenosis there is a rare form of deformity—the cloverleaf skull. There have been three such cases in the total number of 87 cases of craniostenosis admitted to the Clinic of Neurosurgery, Bucharest, between 1936 and 1970. In the medical literature up to 1970 (Duggan et al.) only 19 of these cases had been reported by different authors, and no more than two cases by the same author.

Case 1. B. E. Female, aged 3 weeks. Nothing relevant in the heredito-collateral and personal antecedents. At birth the child exhibited a monstrous deformity of the craniofacial extremity, a "cloverleaf" skull. General examination: weight 3200 g, with only slight turgor, weak muscular system, congenital "club foot". Thoraco-abdominal organs, normal. Neurological examination: normal. Head: trilobate, cloverleaf skull (Fig. 1), closed anterior fontanelle, ears displaced downwards and inwards, depressed root of the nose, marked exophthalmos, congested bulbar conjunctiva, divergent strabismus; fundi flat but pale, normal retinal vessels; macroglossia, prognathism. Because of nasal obstruction the child was obliged to breath through the mouth.

X-ray of the skull: closed cranial sutures, clover-like appearance. The anteroposterior view showed a constriction along the border between the frontal and temporal regions, on both sides. This constriction overlay two abnormal endocranial bone crests, starting from the posterior parietal region and running obliquely forward and downward, thus giving the false impression that the endocranium is formed of three compartment (a frontal and two lateral temporal compartments). The lateral radiograph showed a series of thin bone crests in the posterior parietal and occipital region with a

honey-comb like aspect (Fig. 2). The base of the skull was depressed with relative enlargement of the anterior fossa and much diminished posterior fossa. The orbits had very thin, asymmetrical walls and were not clearly outlined; ocular hypertelorism.

Pneumoencephalography: the air did not enter the ventricular system and outlined only the basalcisterns and interhemispheric space. Cerebrospinal fluid: normal. Panangiography: performed by catheterization of the

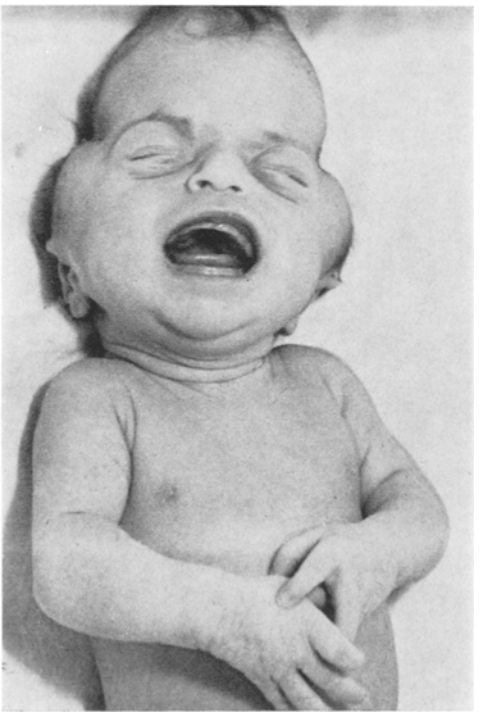


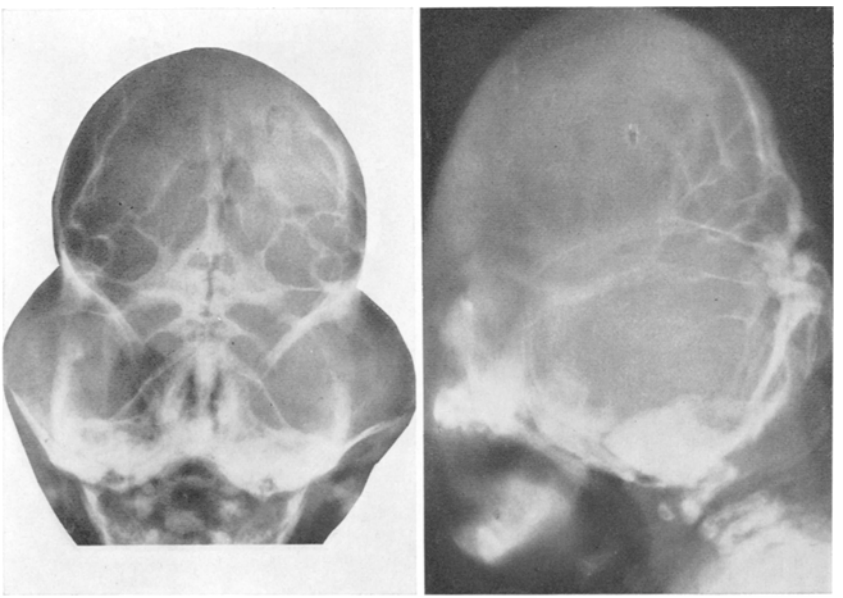
Fig. 1. Photograph of case 1

right humeral artery showed the existence of a single endocranial cavity, with a normal vascular network which displayed marked internal hydrocephalus. Following angiography the child's condition deteriorated and it died. Necropsy revealed severe internal hydrocephalus with extreme reduction of the cerebral parenchyma (the brain appeared to be transformed into a pouch with clear cerebrospinal fluid); severe hepatic dystrophy; intense hyperaemia in all the viscera; bilateral bronchopneumonia; acute dilatation of the stomach.

Case 2. F. I. Female, aged 6 years. Nothing relevant in the hereditocollateral and personal antecedents. Cloverleaf appearance of the skull since birth. Since birth the mother had noted discharge of cerebrospinal fluid from the right nostril which persisted until one week before admission, corresponding to the onset of splitting headaches. During the last two

weeks the child could not see well and bumped against the furniture in the house.

General examination: height 1.10 m, weight 20 kg, afebrile, vomiting. Head: cloverleaf appearance (Fig. 3), closed anterior fontanelle; bitemporal cranial perimeter 42 cm, viscero- and neurocranial height 22 cm; coronal diameter at the level of the orbits 18 cm. Hyperostosis of the sutures. Evident collateral venous circulation. The ears displaced downwards and inwards. Nasal obstruction, the child breathing through the open mouth.



a *b*

Fig. 2. Skull. a Note three endocranial compartments, one median superior frontal and two lateral temporal; b lateral view: in the parieto-occipital region there is a honeycomb like aspect; the base of the skull depressed; an upper and a lower compartment may be observed

Neurological examination: intense diffuse headaches, predominantly frontal and in the vertex, the child crying out with pain; difficult deglutition, speech disturbances due to malformation of the buccal cavity (arched vault, small buccal cavity, macroglossia, dental malposition). She showed a good understanding of what was spoken to her and could perform complicated orders. The psychical examination was difficult because of the severe headaches; mentally she appeared normal for her age. Exophthalmos of both eyes, convergent strabism in both eyes; photomotor reflex present; she could count the fingers of the hand at a distance of 1 meter. The fundi showed primary optic atrophy. X-ray of the skull (front and posteroanterior view) revealed a trilobate cloverleaf skull with the characteristic appearance given by the two lateral crests that appear to separate the cranium into three compartments: superior frontal and two lateral temporal (Fig. 4). Irregular

orbits with thin, asymmetrical edges, hypertelorism. The lateral view showed obvious digital markings in the posterior region, from the edges of which small endocranial bone crests arose, giving a honeycomb aspect. From the occipital and posterior parietal region two clearly visible thin lateral crests arose appearing to separate the cranial cavity into an upper and lower level. The middle fossa of the skull was flattened, diminished posterior fossa with disappearance of Welker's angle. Chromosomal formula:

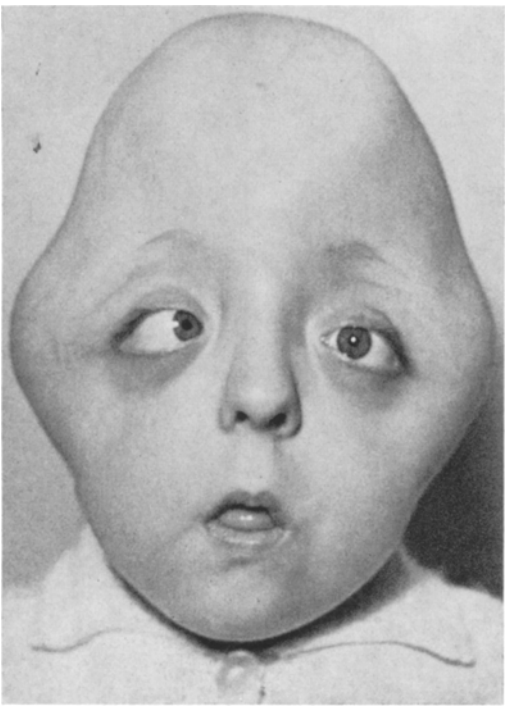
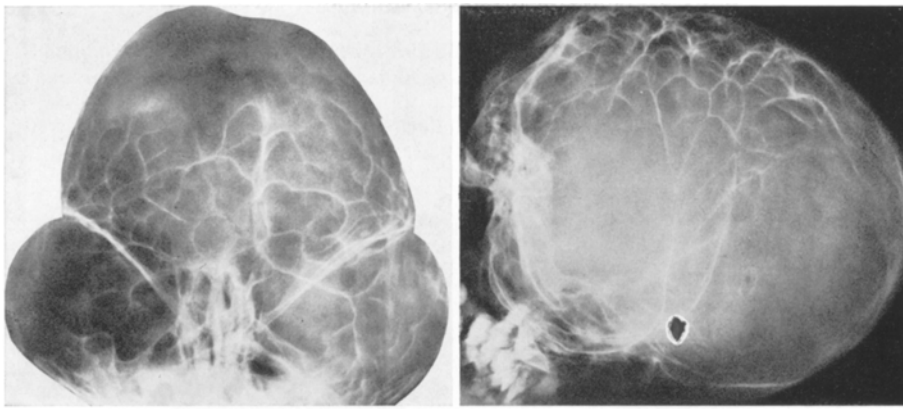


Fig. 3. Photograph of case 2

nothing pathological. EEG: low voltage tracing with dominant theta waves, and polymorphic delta waves on the anterior leads, without asymmetry. On this background, bursts of sharp, sinusoid delta waves. Ultrasound encephalography: normal; pneumoencephalography: only interhemispheric and subarachnoid penetration of the air. Normal cerebrospinal fluid. The child was operated by the Bagdasar-Arseni procedure (linear craniotomy along the coronal, lambdoid and parietotemporal sutures), in two stages at an interval of 4 weeks. Good postoperative course. The headaches disappeared. The clinical control performed after 3 months showed a good general condition, the child was bright, well oriented temporospatially, with adequate answers for her age. X-ray of the skull showed that the sutures were maintained open postcraniotomy.



a b

Fig. 4. X-ray of the skull. a Anteroposterior view of the cloverleaf skull with two lateral crests giving the false impression of a compartmented skull (one upper frontal and two lateral temporal). b Lateral view evident digitation in the parieto occipital region, honeycomb like aspect

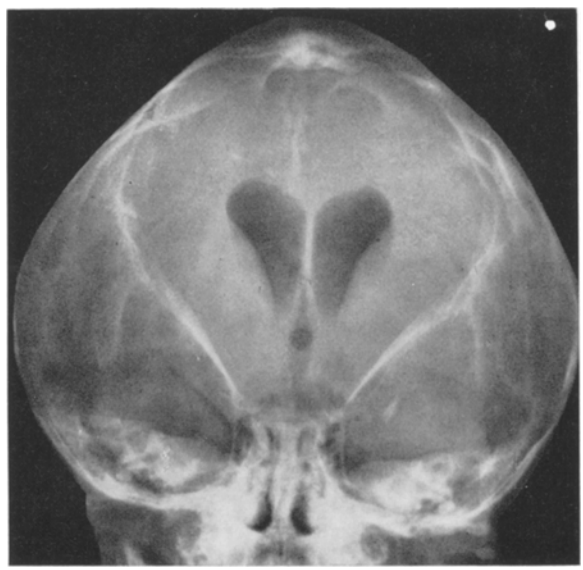


Fig. 5. Pneumoencephalography showing symmetrical hydrocephalus, cloverleaf skull

Case 3. T. A. Male, aged 3 months. Nothing relevant in the hereditocollateral and personal antecedents. Cloverleaf skull, exophthalmos of both eyes.

Presenting signs: good general condition, no fever, 6,000 g body weight. Normally coloured skin and mucosae, good turgor. Nothing else clinically significant. Head: cloverleaf skull, bitemporal perimeter 38 cm; sutures closed by hyperostosis, anterior fontanelle closed from birth. Ears displaced downwards and inwards. Partial nasal obstruction, arched roof of the mouth, macroglossia.

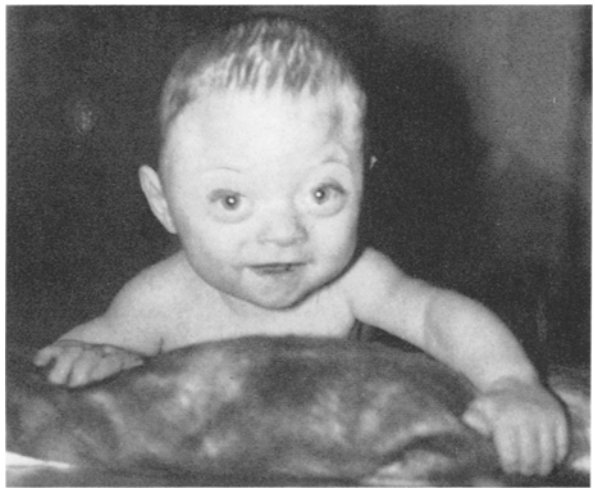


Fig. 6. The child two months after the operation

Neurological findings: the child was very agitated he could not hold up his head or follow any movement with his eyes; difficult deglutition, exophthalmos both eyes, lagophthalmos. Photomotor reflex present, normal ocular motility. Eye fundi: myopia forte (-15D) both eyes.

Ultrasound encephalography: normal. EEG tracing: medium voltage with dominant, polymorphic and monomorphic delta rhythm upon which rapid frequencies were superimposed in all leads. Cerebrospinal fluid—normal. X-ray of the lungs and articulations (elbow, shoulder, hip, knee) normal. X-ray of the cranium: characteristic of cloverleaf skull. Pneumoencephalography showed symmetrical hydrocephalus (Fig. 5). Operated by the Bagdasar-Arseni procedure in two stages, at seven weeks interval. Normal postoperative course. The child was much quieter, gained weight, swallowed easily and could follow what was happening around him; disappearance of lagophthalmos; he got hold of objects and brought them to his mouth, had an adequate mimicry, held his head up but could not sit up (at 5 months) (Fig. 6).

Discussion

The three cases reported should be classified among the craniofacial deformities known as cloverleaf skull. In this caudation there is intrauterine synostosis of the cranial sutures and early ossification anomalies

at the base of the skull (basilar region). In consequence the skull is trilobate, with an elongated narrow fronto-parietal part constituting the upper level which is separated from the lower by a constriction; the lower level is much enlarged by the obvious bulging of the temporal fossae. The surface of the posterior cranial fossa is reduced, the middle level of the skull is flattened and broader. The consequence may be a narrowing of the occipital foramen, hindering circulation of the cerebrospinal fluid, with secondary hydrocephalus.

These monstrous cranial deformities have been called in a plastic manner "Kleeblattschädel" by Holtmüller and Wiedemann (1960) and "cloverleaf skull" by Comings (1965). Because 13 out of 19 cases in the literature were associated with deformities of the bones of the achondroplastic type, the malformation was classed as a form of chondrodystrophy. That is why Duggan et al. classified it as a secondary craniostenosis. Holtmüller and Wiedemann did not accept "cloverleaf skull" as part of a chondrodystrophy and considered that it should be a separate entity—the cloverleaf skull syndrome. They define this syndrome as follows: an extremely rare congenital deformity with marked hydrocephalus of the neurocranium, and a typical configuration, eventually combined with other accompanying malformations. The features are:

1. Cloverleaf skull as already described.
2. Viscerocranial alterations of the orbit, nose and mandible.
3. Micromelia of all the limbs and defects in the development of the spinal column (neither are obligatory).
4. Characteristic X-ray of the skull and pneumoencephalography with marked dyscrania and dysencephaly, i.e. hydrocephalus (hydrocephalus irregularis permagnus).
5. Poor prognosis due to progressive raised intracranial pressure.

Angle et al. emphasized the deformities of the bones—complete ankylosis of both elbows, with short, broad bones (in all 4 cases in literature). They drew attention to hypopituitarism phenomena, probably secondary to hydrocephalus in the two cases they studied.

Büttner described multiple ankylosis and bilateral club feet fixed in a supine position. Bernard following macroscopic and histological examination of the infant's brain which died with Kleeblattschädel syndrome showed that the cerebral lesions were secondary to the deformities of the skull and to hydrocephalus Hydrocephalus is caused either by skull deformities (more frequently) or by vascular malformation in the pontine region (less frequently). Angiomas too were described in other regions of the skull (posterior fontanelle) by Holtermüller et al.

Our three cases exhibited the features of the cloverleaf skull syndrome, the 1st and 2nd case with phenomena of obstructive hydrocephalus.

In the first case internal hydrocephalus was revealed by angiography and confirmed at necropsy. In the second case the hydrocephalus had been compensated for six years by the cerebrospinal fistula since birth; raised intracranial pressure resulted from the spontaneous closure of the fistula. In the third case there was a moderate, communicating internal hydrocephalus.

X-rays of the bones showed no signs of chondrodystrophy.

In the first case, where the child died after the panangiography, the prognosis was probably unfavourable in any event. In the other two cases the postoperative prognosis was good. Case 3 showed obvious psychomotor improvement, whilst the syndrome of raised intracranial pressure was completely rehered in Case 2.

Since these cases should be included in the group of craniostenosis we consider that early surgery is indicated, as it always is in craniostenosis (proved by case 3 with a good vital, psychomotor and aesthetic prognosis).

If there is marked progressive hydrocephalus, a ventriculocardiac or a ventriculo-peritoneal shunt should be carried out.

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