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CRANIOFACIAL ANOMALIES AND PATHOLOGY

ABSTRACT — The skull owned by the Mozarteum shows two uncommon anomalies: a frontal dysmorphism and the print of an epidural haematoma in process of resolution. The first is noticeable on Mozart's portraits, the second probably went unnoticed at first but because of complications, might have contributed to the premature death of the musician.

KEY WORDS: — Mozart Skull owned by the Mozarteum — Early closure of the metopic suture — Temporo-parietal fracture — Print of epidural haematoma — Calcified chronic epidural haematoma.

INTRODUCTION

How, more than ten years after Mozart's death, could Constance Weber, Mozart's widow, assert to Griesinger, a friend of Haydn, the composer, that, if necessary, she would identify Mozart's skull among a thousand (11)? Which detail, which deformity could help her in the process of identifying him?

How can one affirm that the skull owned by the Mozarteum is authentic? We know that it cannot be a crude forgery since its morphological study confirms the sex, the age and the race. In the same way the superposition of the skull on the profiles

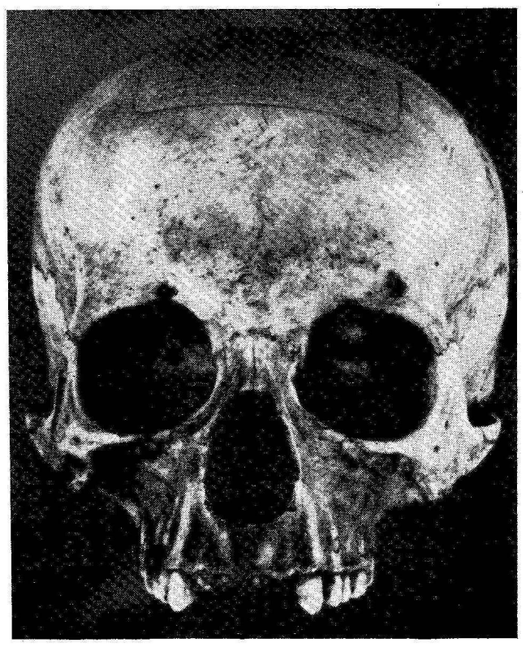
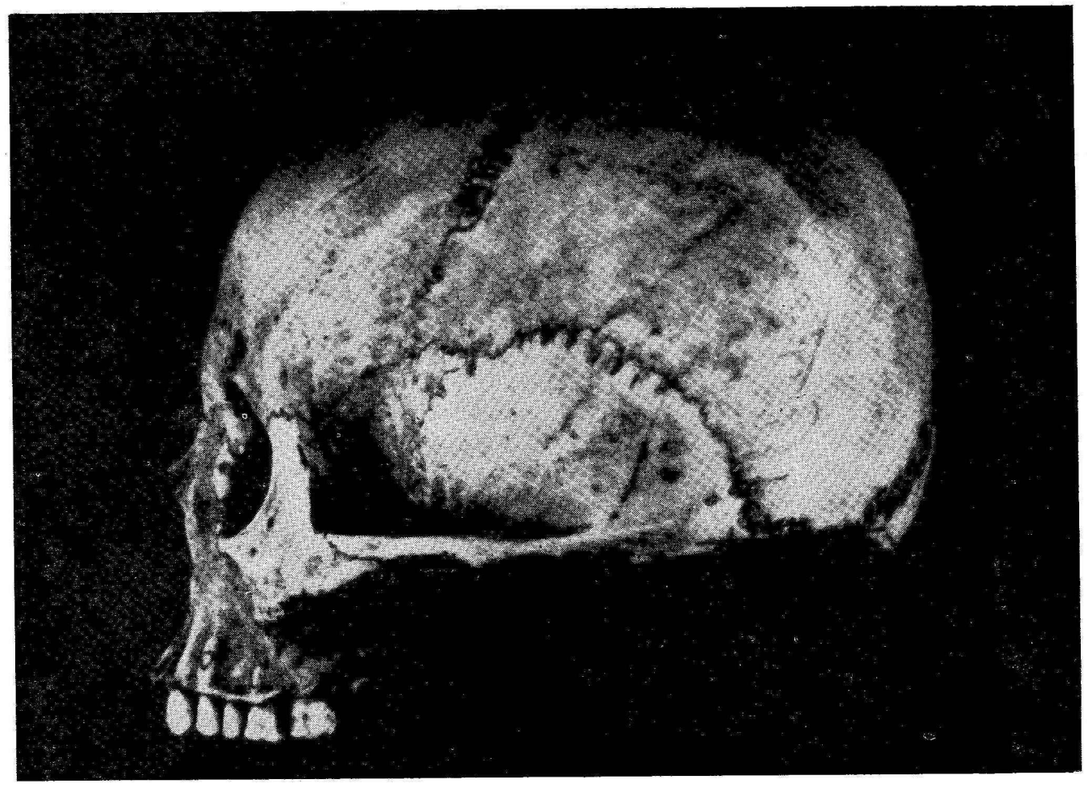


FIGURE 1. *"The grave-digger Joseph Rothmayer remembered the place where he had buried Mozart and he preserved this skull in 1801 at the time of the disinterment and replanning of the graves. Then he gave it to his successor Joseph Ratchoff, who in turn gave it to my brother Jacques in 1842. HYRTL". Certified handwritten inscription by Joseph Hyrtl, a Viennese anatomist. Middle frontal supraglabellar protuberance and grooves of numerous front emissary veins situated on either side of the midline. Metopic frontal area on the medial part of the frontal bone showing small grooves forming an intricate chevron pattern from the nasion to Hyrtl's inscription.*



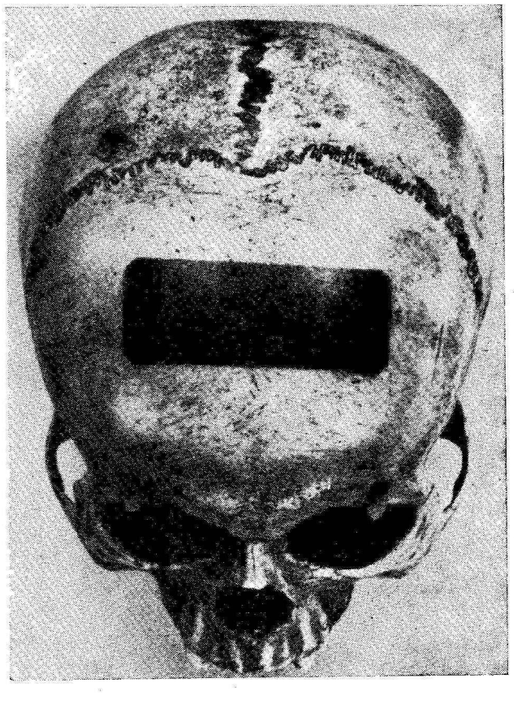


FIGURE 2. *Left profile of the skull. The supraglabellar protuberance is conspicuous. A marked alveolar prognatism is clearly shown by the direction of the nasoalveolar clivus. The forehead is straight and the supraorbital rims are reduced. The fracture is starting from the left parietal eminence and radiating anteriorly and downwards, towards the base with a bayonet-like shape at the parietal temporal junction.*

of Mozart is particularly striking. The historical inquiry, the probability of the facts, the characters of the different owners, strengthen our conviction that it can only be a genuine specimen (18). We can say that there are strong probabilities but the whole of the argument does not constitute an absolute proof.

As we were identifying the skull, we discovered two rare anomalies (12) one of which could well correspond to the individual physical feature which we were looking for, since it changes the middle part

FIGURE 3. *Oblique top view of the skull. The upper shifting of the bregma has displaced the upper limbs of the coronal suture. Both coronal and sagittal sutures are overbroad. The supraglabellar sulcus is divided in the midline by a small protuberance. The surrounding bone surface of the frontal midline is covered with many small superficial furrows.*

of Mozart's forehead in a remarkable way. The second anomaly is a chronic epidural haematoma in process of resolution.

The simultaneous presence of these completely different anomalies and the transposition of this skull pathology to Mozart's physical features has compelled us to divide our report into four parts: The study of the skull, the premature synostosis with the elements of diagnosis and the diagnosis, then the head injury and its consequences.

THE SKULL OWNED BY THE MOZARTEUM

(*Material*)

The history of this skull is known since the middle of the last century. The handwritten inscription by the anatomist Joseph Hyrtl on the upper part of the forehead indicates that it comes from Saint Marx Cemetery in Vienna. Jacob Hyrtl, Joseph's brother, an engraver and musician, obtained it from the grave-diggers of the above mentionned cemetery in 1842. It became Joseph's property after Jacob's death in January 1868. Then he sawed it and told his friends about it. At the time when the inheritance left by Joseph Hyrtl's widow was being settled in 1899, a judicial investigation confirmed that the skull was the one which was supposedly that of Mozart's and that the writing on the top of the skull was Hyrtl's (*Fig. 1, 2,* the skull). On the sixth of October 1901, the City of Salzburgnas become the new owner of the skull and still is up to now (18).

On the inner side of the calvaria, this skull which is of a dirty yellow colour, displays an irregular brown pigmentation at different levels, due to post-mortem sedimentation of endocranial elements. In places there are some organic residues of collagen, of meninges near the sutures and small vascular pedicles emerging from the foramina of the bone. Therefore its organic residues must have been

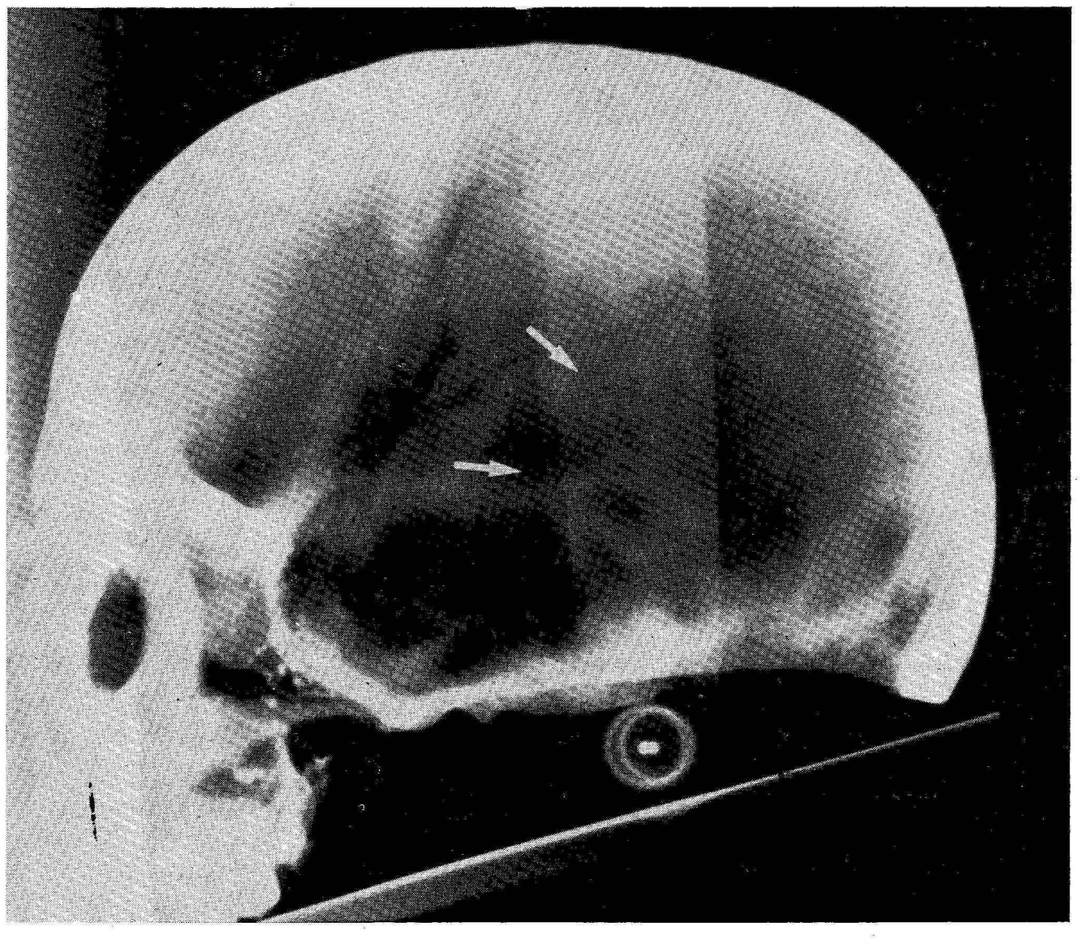


FIGURE 4. *The lateral standard X-ray reveals a temporoparietal fracture.*

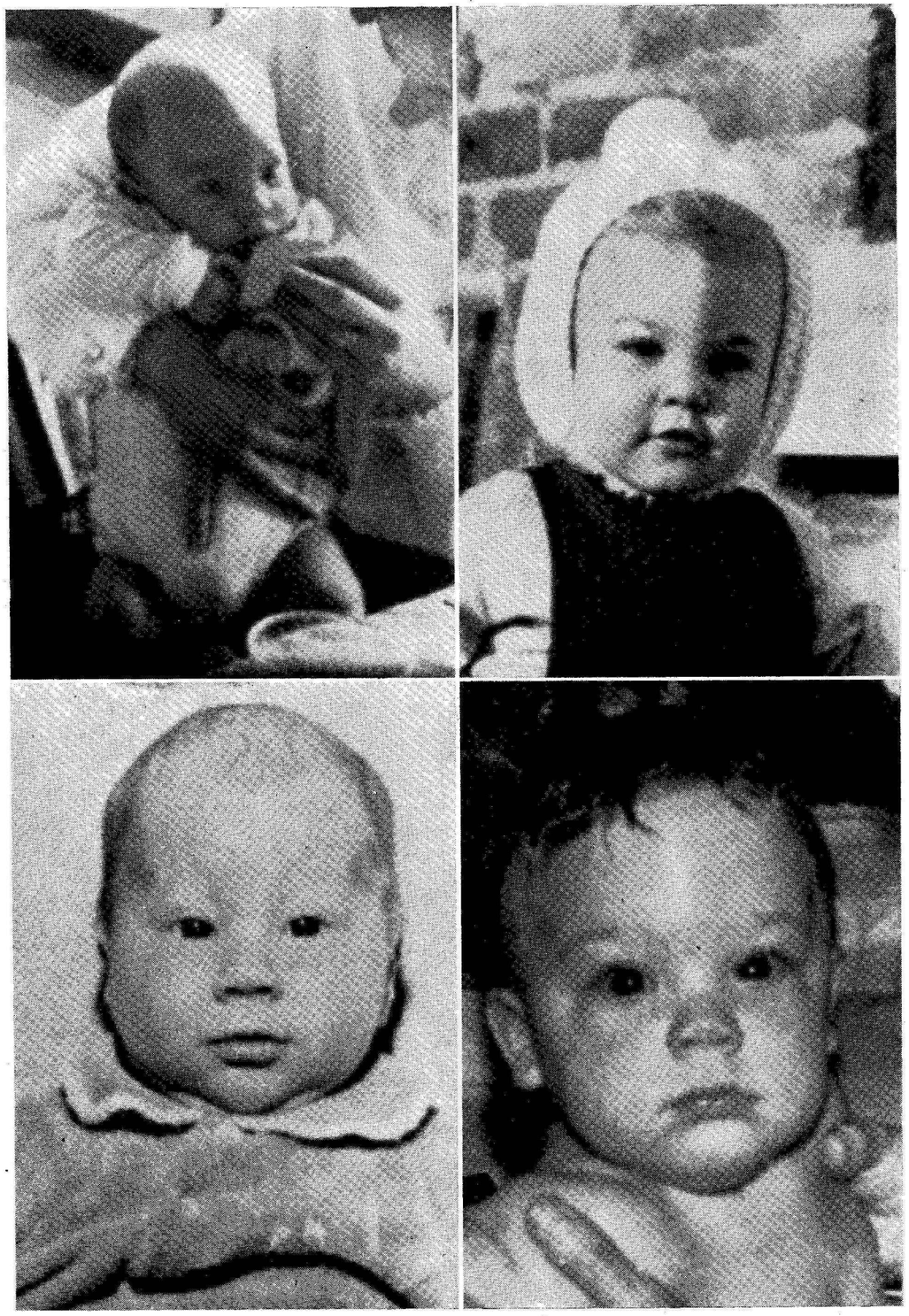


FIGURE 5. *Three young patients with a mild form of PSMS. The two top snapshots represent a 4 week, then an 18 month old patient. Underneath on the left a child whose X-ray clichés are in the publication by Dominguez. The other child, at the bottom, on the right shows a glabellar pseudohemangioma that often occurs with cranisynostosis. It will disappear as well as the frontal crest.*

cleaned off the skull before their complete degradation with time. The slashes left by a sharp tool on the outer surface of the vault and on the nasal bones, suggest that it has been stripped of its flesh or scoured clean with this type of tool. The skull is not complete because, as we have mentioned previously, Hyrtl sawed it along a line parallel to the Frankfort horizontal plane, across the external auditory meatus. It was probably at the time of this sawing that the body of the sphenoid disappeared. We cannot tell whether the orbits, the nasal bones and the ethmoid were damaged at the same time. The sawed off base has been lost, as well as the mandibula which still existed in the last century.

The skull is that of a young man, an ultrabrachycephalic Central European Caucasian, with widely separated frontal eminences, a vertical frontal bone, with a nasomaxillary protrusion and reduced orbits. Its cranial capacity is estimated to be 1,585 cc (13).

The outer surface of the frontal bone displays a series of small venous grooves forming an intricate chevron pattern, on both sides of the midline ridge, together with many small osseous foramina in the very bottom of these prints or all around them (*Fig. 3*, skull from oblique top). In that median region the protrusions of both brow-ridges meet above the nasion and form a medial superciliary protuberance. The trace of the metopic suture is missing. The parietal bones are widely developed. The orbits are as high as wide. The curve of the coronal suture is inverted at the level of the bregma. Therefore the bregma comes into the normal space of the frontal bone.

The sockets of the front teeth are protruding as they are anterior to a line perpendicular to the Frankfort horizontal plane, drawn down from the Glabella. The palate is wide of yet very shallow. The anterior diastema of the incisors and canines are not secondary to a microdontia but to a dentomaxillary disharmony. The first left molar is decayed on about one third of its coronal volume, and its pulpis also affected, the second left molar is slightly decayed. The third molar on the left has been lost postmortally. The general degree of dental attrition indicates that the skull belongs to an individual between 25 and

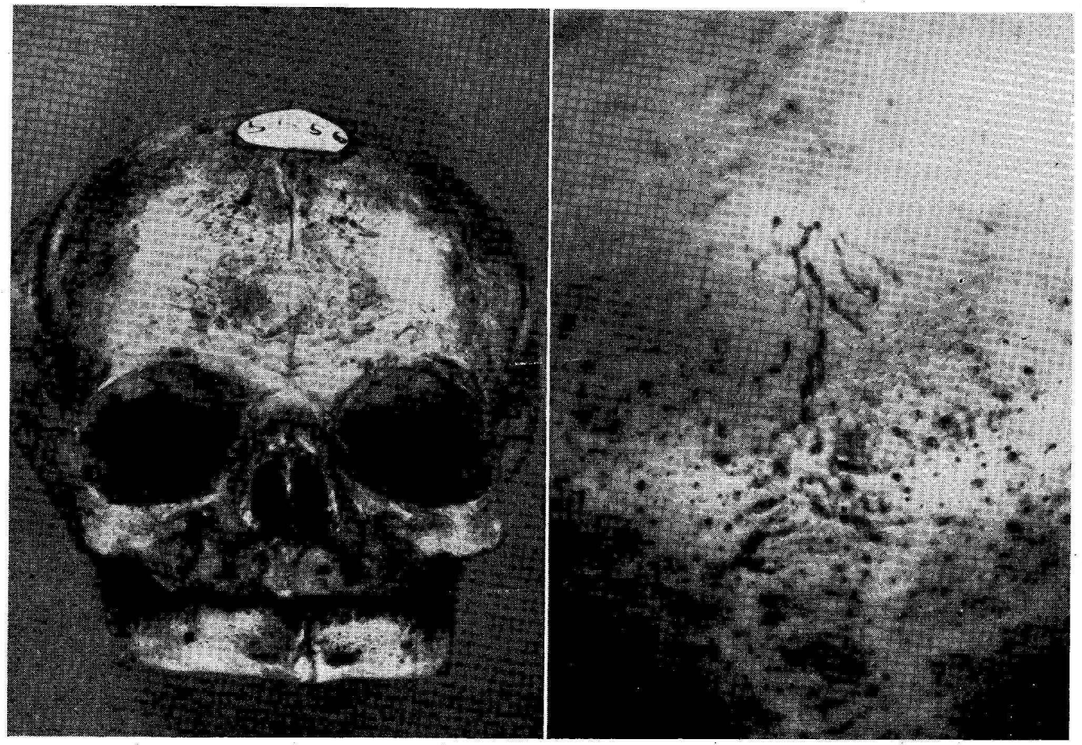


FIGURE 6. *On the left. Skull of a newborn child with a ECMS. The incomplete obliteration of the metopic suture is located at the middle of the suture, numerous vascular grooves on either side of the suture. Gall Collection. MUSEE DE L'HOMME. On the right, glabellar and supraglabellar protuberance of the skull owned by the Mozarteum with under periosteal passages of emissary veins, deep inside each furrow many foramina testify of the presence of emissary veins.*

40, an age confirmed by the closure of the apex of the roots of the wisdom teeth.

The standard X-ray of the skull reveals a linear fracture in the temporoparietal area. It starts at the level of the left parietal protuberance and develops forwards and downwards to the basis, covering a distance of ten centimeters (*Fig. 4* Lateral view of the skull). The vault of the skull, because of its overall thinness, presents patches which are extremely thin and which, in places, make the bone translucent. Because of this lack of thickness and of the age of the individual, the middle meningeal vascular patterns are stongly marked on the inner surface of the parietal bones (15). At the level of the left parietal, the posterior branch of the middle meningeal vessels is completely obliterated because of an oval osseous print on the underside of the fracture; it corresponds to an epidural haematoma in process of resolution.

THE METOPIC CREST OR THE PREMATURE SYNOSTOSIS OF THE METOPIC SUTURE OR THE EARLY CLOSURE OF THE METOPIC SUTURE

(The elements of the diagnosis)

The premature synostosis of the metopic suture (PSMS) is a benign craniosynostosis. It affects only the frontal bone. It was described in its major form for the first time in 1862 by Welcker and can be divided into two types: one isolated type, which is more or less dysmorphic and which goes without any other anomaly; the second one is of a more complex sort and occurs together with a series of deformities, even a chromosome aberration (21). In our study we are only dealing with the isolated type of craniosynostosis.

The unusual premature closure of the mediofrontal or metopic suture will stop the broadening and shaping of the faetal forehead during the growth of the head, thus entailing a facial dysmorphism from birth. The main feature of the PSMS is a deformity of the frontal bone, ranging from a mere vertical mediofrontal crest to a triangular forehead. This severe unaesthetic deformity is known as trigonocephaly.

The frequency of this crest, noticeable to the touch, is that of 0.3 out of 1,000 newly-born infants, but if minor forms are included, this frequency is certainly much higher (17). This deformity varies in its extent and evolution. Since this anomaly resolves spontaneously, it is by six times inferior in the neurosurgery statistics bearing on one or two year old baby-patients. The moderate or mild forms that diminish or even disappear with time are thereafter discarded. In these forms, the more appropriate term of early closure of the metopic suture (ECMS) should be used.

The more severe types of this anomaly must be opposed to the minor ones. Indeed, the former (PSMS) cause a real aesthetic prejudice lasting throughout adult age, may prevent both anterior hemispheres from expanding correctly during the first two years of life and require surgery, whereas the latter (ECMS) allow for an acceptable aesthetic aspect in course of time without any further complication (1, 2, 3).

The complete phenotype of craniosynostosis in infants is constituted by:

— a pointed forehead with the maximum frontal protuberance in the supraglabellar area. The top of the pointed forehead displays a metopic midline ridge, often palpable from the nasion to the bregma which constitutes the main feature of the anomaly (*Fig. 5*). The frontal eminences disappear in the severer types.
— the "surprised look" of the child due to unusually high eyebrows which constitute a secondary sign to the vertical ovalisation of the orbits (*Fig. 5*).
— a supraexternal and supraorbital depression, causing a lack of orbital cover and marked external concavity of the supraorbital region (*Fig. 6*).
— a depression at the level of the temporal canthus which is a sign of the narrowness of the anterior frontal bone (*Fig. 5*).
— an ocular exorbitism owing to a reduced orbital capacity; more marked in the outer area of the orbits due to the lateral depression of the outer orbital rims (*Fig. 5*).
— and lastly, quite often, a chubby face with falling cheeks.

FIGURE 7. *Mozart at the age of 27, by his brother in law Joseph Lange. Conspicuous superglabellar protuberance, with exorbitism. Photo Mozarteum.*

FIGURE 8. *Mozart at the age of 33, by Doris Stock. A straight forehead with anteriorly placed eyes, marked cheek bones and supraglabellar protuberance.*

Other signs are conspicuous either on the dry skull, or on the peroperative skull, or on X-rays:

— a hypotelorism, visible on standard skull films, softened on the face by an epicanthus which aesthetically widens the interorbital space.
— a genuine dilated venous plexus on the very periosteum, in the affected middle region of the frontal bone, together with small emissary veins connecting this venous plexus to the superior intracranial sagittal sinus and marking the external osseous surface on a short distance under the periosteum (Fig. 6, on the left) (8).
— the altered curve of the coronal suture, secondary to the hypoplasia of the prematurely closed frontal bone. The compensating growth of the parietal bone pushes the coronal suture forward and sends the bregma into the frontal bone area. The phenomenon is all the more visible as it is situated closer to the metopic suture (at the level of the bregma).
— the uncommon depth of the cribriform lamina of the ethmoid bone between the vertical inner orbital walls, secondary to the vertical orbital ovalisation. The widening process of the frontal bone is slowed down but the frontal bone continues to develop in height and compensates for the loss in width.

The anomaly does not have the same aspect in adults, because of its spontaneous resolution. This transformation is secondary to the compensating growth of the vault at the level of the nearest sutures unaffected by a premature synostosis, and to the reshaping of the frontal bone by apposition-resorption phenomena and the forward pressure of the brain. In the milder types (ECMS) there remain only the glabellar protuberance, the vertical forehead and a variable occurence of exorbitism.

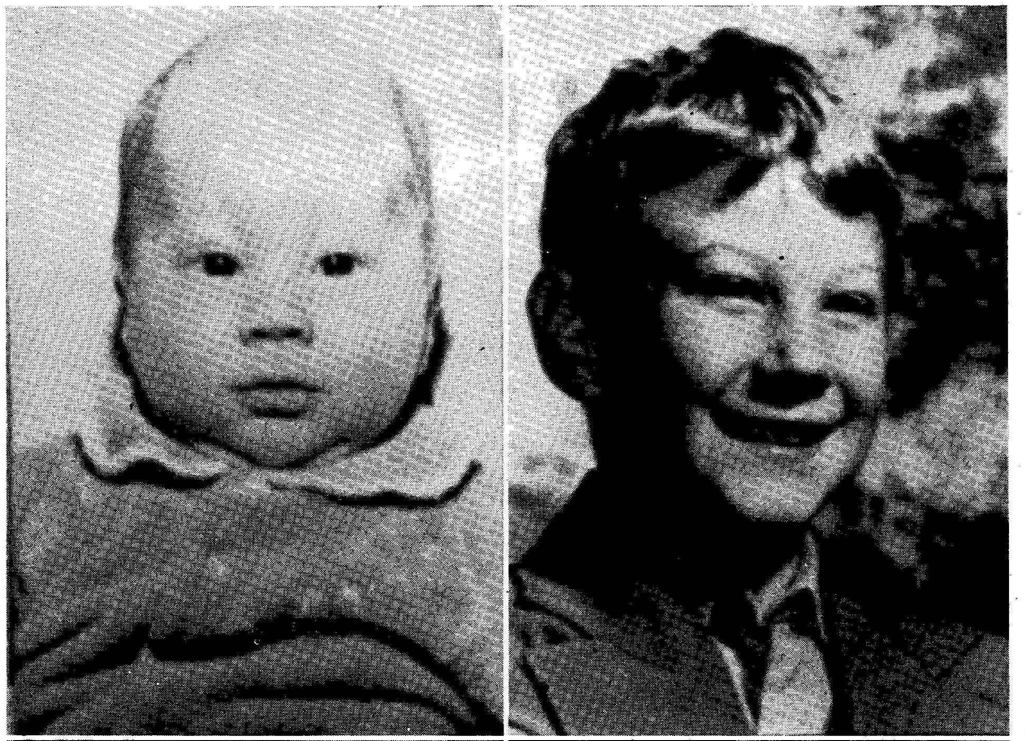
THE ECMS ON THE SKULL OWNED BY THE MOZARTEUM AND ON MOZART'S PORTRAITS

(The diagnosis of ECMS, results and discussion)

THE SKULL

The grooves of numerous emissary veins which form an intricate chevron pattern are visible in the median region of the frontal bone. On the site of the glabella a median prominence is joined by the superciliary arches on either side. These superciliary

FIGURE 9. *A detail from the painting: Mozart attends a concert at the Palace of Schonbrunn. Supraglabellar protuberance exorbitism, external supraorbital depression and chubby cheeks.*



arches are very lightly marked on their outer rims. The beginning of the metopic suture which is usually marked on other skulls of the same age, has disappeared. The curve of the coronal suture is inverted from the temporal crest to the bregma, which pushes the bregma forward (*Fig. 3*). The orbits, equal in breadth and height, have an oval shape with supranternal summit. The restored orbital capacity is of 22.5 cc for the right eye and of 21.5 cc for the left one, which is substantially below the normal figure. The supraexternal orbital region displays an anteriorly and outwardly concave shape.

MOZART'S PORTRAITS

It is most interesting to trace the various stigmas of the ECMS and the anomalies previously noticed on the skull, in Mozart's portraits. Among the portraits painted by his contemporaries, none really gave a very flattering physical representation of the composer. For Mozart as a child we have chosen a portrait of him, in prime infancy, as he was sitting between the archbishop prince of Salzburg and his father, during a concert at the Viennese Palace of Schonbrunn, as well as a portrait of Mozart sitting at the piano, at 14 (*Fig. 9, 10*). Among the portraits of Mozart at an adult age, we have discarded both the apocryphal portraits as well as those painted after his death, and we have chosen the unfinished portrait painted in 1783 by Joseph Lange, his brother in law, together with the lead pencil drawing of Mozart's profile made in 1789, by Dorothy Stock (*Fig. 7, 8*).

On the later portraits, the forehead is vertical, with a sharp nasofrontal angle (*Fig. 8*). Because of the alveolar prognathism, a nasomaxillary protrusion is noticeable. The missing supraglabellar depression was noticed and painted by Lange, Mozart's brother in law, and by Dora Stock. The composer's eyes on a level with the orbital rims are exorbited. Mozart was neither suffering from severe myopia nor hyperthyrodism nor from a bilateral orbital tumor. That exopthalmic aspect cannot be found either in his father, mother or sister. Most probably it is secondary to a reduced orbital volume. In connection with that exophthamos it is possible to notice a fold of the skin at the level of the upper eyelids. Starting from the superior palpebral fold, it runs to the inner canthus, and even lower down. This fold is what is left from a tarsalis epicanthus. On either side of the face a depression can be noticed at the level of the outer palpebral commissure, that is a hollow just before the temporal fossa area.

The frontal dysmorphism is more conspicuous in Mozart's portraits as a child than in his later portraits. Indeed, he was painted with chubby, falling cheeks, high eyebrows, an epicanthus, an evident mediofrontal prominence in the glabellar and supraglabellar area. The outer canthus are also clearly

FIGURE 10. *Photos at the top: ECMS of a few months old child and of the same child at the age of 13 (Dominguez). (Photos at the bottom: portraits of Mozart at the age of six and at the age of 14.)*

FIGURE 11. *Mozart's mother, marked frontal crest. Photo Mozarteum.*

visible (*Fig. 9*, Mozart as a child at the theatre). The presence of a phenotype and the evolution of the dysmorphism are obvious if the children's pictures showing a mild and regressive form of ECMS and belonging to the publication of Dominguez (3) are compared to the composer's portraits (*Fig. 10*).

Hereditary forms do exist in all craniosynostoses. In pediatric neurosurgery and in 10% cases, we did observe family varieties yet with different occurences. Out of 40 surgical cases we counted two twin-sisters, a father and his daughter, and a father and his son (2). A close study of the portraits of the composer's mother reveals a midline ridge on the forehead as well (*Fig. 11*, portrait of the mother).

200 years after Mozart's death and only on the visual basis of his portraits it is impossible for us to absolutely lay down a diagnosis of ECMS. However, the frontal modifications of the skull which are also to be found in the portraits may be considered as a form of ECMS inasmuch as there exists no other argument that could indicate a comparable affection and could account for the dysmorphism of Mozart's forehead and eyes. The midline ridge or the keel-like forehead that are noticeable among Eskimoes and some African ethnies offer a morphology quite similar to that of PSMS; however the racial characteristics of such keel-like forehead entail us to discard them from our debate (13).

THE CONSEQUENCE OF THE HEAD INJURY
(*The epidural haematoma. Results and discussion*)

The skull owned by the Mozarteum shows a temporoparietal fracture, on the left, and the print of an epidural haematoma (*Fig. 1, 4*).

The left temporoparietal fracture is 10 cm long. It starts from the left parietal eminence and radiates anteriorly and downwards towards the temporal fossa. This fracture is consolidated on all its length and is scattered with small foramina along its line.

On the inner surface of the skull and on the underside of the parietal fossa which is the impact area of the injury and the fracture — though without displacement — it is possible to notice the print of an epidural haematoma with an irregular thickening of the bone (*Fig. 12*, the print). On the inner calvarial surface this thickening of the bone leaves an oval print with polycyclic edges of 80×60 mm, obliterating the posterior branch of the middle meningeal vessels. In the middle of this print, in the central area (*Fig. 13*), the grooves of meningeal vessels are still visible. These facts clearly prove that at the time of the death, the haematoma was not entirely resolved yet, since it still presented a residual liquid layer preventing the adherence of the dura-mater to the inner table of the skull and preventing the inflammatory ossification-inducing phenomena.

On the left, at the level of the temporal squama underneath the print of the haematoma, the markings of the cerebral convolutions of the temporal bone together with the Sylvian crest are completely obliterated whereas they are clearly visible on the right. The adherence of the brain to the inner table of the skull seems to have been modified in this area, in consequence of the compression of the parietal and left temporal convolutions on the underside of the haematoma. At the time of the death the markings of the cerebral convolutions on the dome could not have been formed yet. We cannot tell whether the head injury entailed only one lesion, consequently there is no certainty as to whether the compression of the brain was due to the hematoma only.

All these factors lead us to date the head injury back to several months at least before the death

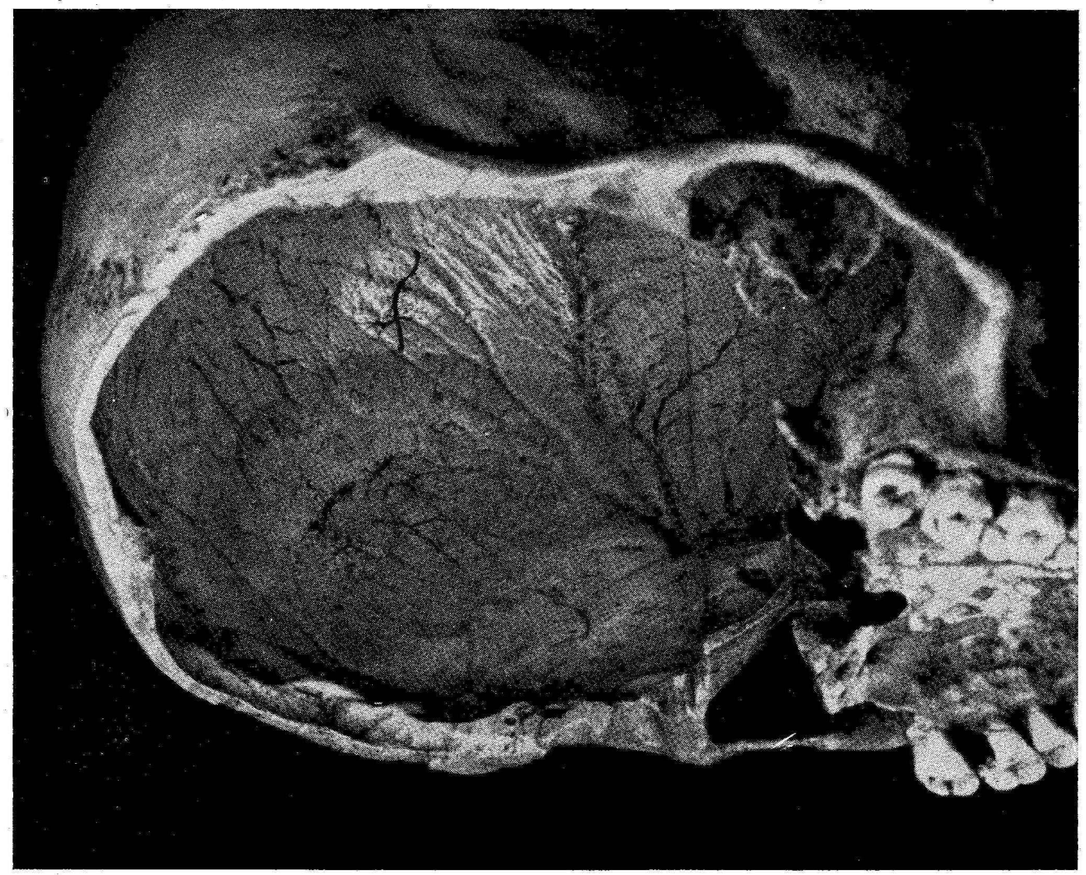


FIGURE 12. *Print of the EHD on the inner table.*

FIGURE 13. *Central region and central bony deposits of the print.*

since the fracture was clearly consolidated and the haematoma not yet resolved. The one-side left obliteration of the markings of the convolutions on the bony dome indicates that the brain was compressed and that the two hemispheres could not resume their usual placement and fall into their initial grooves. Therefore, we may conclude that at the time of the death all the phenomena were not yet resolved.

The head injury and the epidural haematoma cannot be the cause of the death since both the healing and resolution of the haematoma had started their process. Since the injury was situated in the left temporoparietal region both on the edge and behind the central sulcus of cerebrun (Rolandi sulcus) and at the level of the Sylvian fissure, it entailed only very few signs of brain suffering, all the more so in a right-handed individual (as was the case with Mozart). Though part of the haematoma lies on the underside of the language areas, language troubles in that sort of injury are most uncommon.

We do not exactly know the reason for Mozart's death. Even the most detailed description of his last days as reported by his close family and the doctors who cared for him must be investigated in the light of these facts. The head injury that we have noticed might have been caused by a simple backward fall. This sort of injury accompanied by a fracture and an EDH which may remain unnoticed or be initially benign is not considered unusual. Nowadays with CT Scans we know that epidural haematomas do exist without any neurological signs, and simply associated with headaches (5, 9, 10, 14). After a period of about two weeks with helmet headaches and sometimes vomiting fits, the signs diminish (16, 19, 20). In the spring of 1790, Mozart suffered from migraines, and some of his friends saw him with bandages around his head (7). In November 1791 Mozart suffered from paralysing troubles, vomiting fits and violent headaches. At that time, Doctor Closset who was Mozart's doctor, feared meningitis (7). Later, Mozart suffered from oedemas, high fever and skin eruptions; he died after a short one hour coma. Later and according to recent translations,the death register mentionned a miliary fever (hitzigen Friesel Fieber) but according to certain translations from the last century it probably was a high fever called cerebral fever (22). According to Harman, a German doctor who lived at the beginning of the 19th century, the miliary eruption was already connected to a variety of fevers, as well as other fever and sweat inducing illnesses. It consists in an eruption of transparent blisters over a rosy surface which only leaves the face clear (4). The

literal translation of the "hitziges Friesel Fieber" would be a high (or elevated) fever accompanied by vesiculate eruption.

Some aspects of the medical art at the end of the 18th century may appear incomprehensible to us; because of the doctors' habits, so unlike ours, of viewing and classifying the illnesses according to their symptoms, because of the words which describe the clinical signs and have different meanings from those we understand now, and lastly, because many long illnesses, fatal at that time, quite naturally led to an organic exhaustion preceding death. The final cachexic stage, if prolonged, necessarily induced infections which could not be mastered, with the secondary infectious centres (kidneys, livers, lungs and brain) complicating the initial semiology. A lot of ailments ended in hyperthermia, septicemia, infection of the lungs and kidneys and skin eruptions. All that would explain the oedemas, and the signs of infection of which Mozart suffered before his death and the variety of interpretations which have been forwarded since. Undoubtedly there have been some neurological complications but as the major symptoms were hyperthermia and skin eruption, this ailment could only be classified among the high fevers in the 18th century.

CONCLUSION

The pathology of the frontal bone of the skull owned by the Mozarteum which combines a dysmorphism of the metopic area, with orbits presenting a reduced volume and altered rims together with a nasoalveolar prognatism, proves to be an essential element in the identification of the skull, since we have always traced these characteristics in Mozart's portraits, no matter what his age was.

The bleak year of 1790, recognized for Mozart as a year of misery and of weak creative production, seems to coincide with the presence of an epidural haematoma. Although the subject survived and the haematoma resolved, the neurological complications which appeared later may have played a significant role in the premature death of the composer. This theory is appealing since Mozart's close family members reported that the musician suffered, among other problems, from pertinacious headaches and neurological troubles. This theory will be the subject matter of further reports.

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