

Icono-Diagnosis of Craniofacial Disorders: The Eye of the Plastic Surgeon

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As a craniofacial surgeon, it is not uncommon to receive the picture of a patient before the first consultation. It happens most often when the patient is abroad and he/she, or his referring physician wants to know whether such deformation can be treated surgically, before undertaking a possibly useless travel. More than any other specialists, a surgeon who has gained expertise in treating congenital anomalies such as clefts, oxycephaly, plagiocephaly, hypertelorism, Crouzon or Apert disease, or first branchial arch syndrome, can easily make a diagnosis on a simple photograph, as he could also diagnose, with a great percentage of certitude, diseases and tumors like teratomas or ameloblastomas, Recklinghausen neurofibromatosis, noma sequellae, fibrous-dysplasia, and several other disorders like facial palsy and posttraumatic facial deformity. When treating patients from the third world, these approximate diagnoses, made on pictures only, are essential for the handling and the cost of the therapy. It may determine whether a patient can be operated in a local hospital, during a temporary surgical mission, or if surgery can only be undertaken safely in a fully equipped medical center, with a multispecialized team, and an intensive care unit. Of course, like in medical archeology, these diagnoses, made remotely, are greatly enhanced by written medical histories, and X-rays if available.

THE CONCEPT OF ICONO-DIAGNOSIS

Anneliese Pontius, psychiatrist at Harvard Medical School, was interested in what art can teach about the diseases of nonhistorical civilizations. In an attempt to demonstrate the ancient presence of Crouzon's disease in the Cook Islands based on human representations of this cultural isolate, she introduced the term icono-diagnosis in 1983.¹ To apply her method, the researcher must have a good training in anatomy and physiology. He seeks to identify pathologic states whose bodily manifestations are very striking and emotionally charged to such an extent that these appearances are imposed on artists "at the expense of the representation of the banal body." For Anneliese Pontius, this method is reserved for prehistory, that is, civilizations without writing, and certain cultural isolates. Grmek and Gourevitch broadened this initial definition and used the same approach as we enter historical times, when the understanding of malformations and diseases were not based on scientific grounds.² The artist of Antiquity, even more than the "primitive," can and sometimes wants to represent unusual aspects of the human body without any real knowledge of the diseases they reflect. As a number of literary authors, the incentive of the artists is to produce or reproduce a condition that he judges abnormal, a figure which out

passes the frontiers of normality.³ The contributions to medical history of this retrospective diagnostic method, based on the study of images, complement those of the exegesis of ancient medical writings, the pathography of historical personages and palaeopathology, that is the study of human remains.

Before analyzing Cook Islands' prehistoric art to detect signs of Crouzon's malformations, Dr Pontius had studied several pictorial and sculptural artistic objects in isolate ethnical populations like in Easter Island,⁴ New Zealand (Maori) and in the most isolated area of New Guinea. Her approach is the one of a psychiatrist. She assumes that there is a decisive factor triggering the artists to represent "a stressful experience—a cognitive mismatch—of seeing the human body in a form different from its expected shape." "It is speculated that in prehistoric groups such traumatic experience as observing diseases and malformations without knowledge of their natural causes is one factor which triggered a creative leap toward the emerging of art depicting the human body, a kind of sublimation, analogous to that occurring in artistic individuals ever since." To evaluate the incidence of a hereditary congenital malformation or a widespread disease in a given culture, the researcher should find the same characteristics in a large number of items, which are repeated during a long time. As an example, in the 1.000 stone giants on Easter Island, Dr Pontius detected 12 specific signs of leprosy "in a reverse, overcorrected representation." She assumes that "the fright-evoking destruction of the socially most prominent body parts, face and hands, was 'undone' by the prehistoric artists. They depicted each of those body parts which were characteristically destroyed by leprosy as particularly strong and beautiful: the cartilaginous part of the nose, especially the nostrils; the fingers; the ear lobes."

As for the Crouzon malformations in Cook Islands, Dr. Pontius assimilates the wood carvings of the natives, known as "fishermen's gods," to cleidocraniopelvic dysostosis, an extremely rare syndrome, that Crouzon would have described in 1919, and which includes craniofacial synostosis, as we know it today. In any case, in both examples, the relation between the observed figures and the actual medical diagnosis seems to us very questionable. If leprosy was effectively present in Easter Island not long ago,⁵ and some rare hereditary malformations affected the population of the Cook Islands, it seems very doubtful that for generations the sculptors represented bodies in a distorted way "from actual model in their experience."

Undoubtedly, history, in particular medical history, is based on traces, written documents, vestiges, works of art, and remains of animal and human bodies. Since the discovery of numerous ancient terracotta and other artistic facial representations, the translation of several texts of dead languages like Egyptian or Cuneiform texts, the analysis of ancient skulls, the progress in DNA research and new technologies, the study of antique pathology in the craniofacial sphere arouse interest, not only from the historians, but also from various specialties, among them a few plastic surgeons. Eugen Holländer (1867–1932) was known for his pioneer operations on fat transplantation (1906), face lifting, and breast reduction, published in 1912 an illustrated mega-book entitled *Plastik und Medizin*,⁶ where he presents a series of ancient sculptures, terracotta, ex-voto,

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masks, skeletons and mummies from all over the world, illustrating health and disease. A number of those represent facial anomalies and sequels of diseases such as leprosy, syphilis, or possible noma. More oriented on the craniofacial malformations, similar studies have been published by plastic surgeon JJ Longacre on the pre-Columbian pottery of Central America,⁷ Saman, Gross, Ovchinsky, and Wood-Smith on cleft lip and palate in the arts,⁸ and ENT surgeon Wolfgang Pirsig on congenital craniofacial malformations or syndromes in visual arts in general.^{9,10} Moreover, several “case reports” on various pathologies have been reported in the recent years.^{11–14} However, we are indebted to the French medical historians Mirko Grmek and Danielle Gourevitch, in their book *Les Maladies dans l'Art antique (Diseases in Antic Art)*, for the most compelling analysis, review of the literature, and reproduction of numerous Greco-Roman images representing diseases in general.²

If all kinds of disability have been the subject of their scrutiny, portraiture of the face, torn, suffering, mutilated by accident, malformation or disease, often stimulated the artists to reproduce what they saw in the affected individuals.¹⁵ Coarser imaging of craniofacial disorders have also been produced and displayed in shrines and temples as ex-voto, to ask or to thank god for the cure of the diseased organ.¹⁶

When visiting and screening archeologic museums, one may find objects of art representing craniofacial disorders, mostly terracotta, sometimes statues, paintings or masks, and it is tempting to make icono-diagnoses on pathologic conditions well known in our surgical practice. However, one should be aware that the artists often like to represent personages of mythology, or monsters of pure imagination which have nothing to do with a human pathology or malformation. Grmek and Gourevitch also denounce the traps and the wrong interpretations of some quests, especially when people want to acquire such objects. The story of the German ophthalmologist Theodor Meyer-Steineg is exemplary. Already a known medical historian and collector, he visited Greece and the island of Cos in 1910, where he was very amiable with the local inhabitants, giving them care notably for trachoma, which was very common at that time. To thank him, the people found for him numerous pieces that he thought came from the famous sanctuary of Asclepion. In fact, today, it has been recognized that except for a few true ancient objects, most pieces in his collection were fake or fabricated for him.

CONGENITAL MALFORMATIONS

A child born with a lethal or nonlethal congenital craniofacial malformation has always been the subject of intense scrutiny, and the attitudes of the parents and society toward these errors of nature have often been fear and reject. No wonder, representations of such deformities are not uncommon in the ancient world, clefts lips being the more frequent. However, it is interesting to note that moldings or statues of individuals with clefts are exceptional in the Greco-Roman world and far more frequent in Central and South American ancient cultures¹⁷ (Fig. 1A). Is it due to the incidence of such malformation or to their religious or social significations? We do not know. But it is well recognized today that the incidence of clefts lips or palate is very variable between countries and ethnic groups, as it is for mid-facial encephaloceles, which are much more common in a few Asiatic countries than in the rest of the world.

On the contrary, one may find in ancient Greek and Roman terracotta a few figurines with various common malformations like prominent ears (Fig. 1B), baggy eyelids (Fig. 1C), strabismus and antimongoloid eye slants (Fig. 1D), asymmetrical face (plagiocephaly) (Fig. 1E), mandibular prognathism (Fig. 1F), and lower jaw hypoplasia indicting a possible malocclusion. Interestingly, the figure of the Greek Demosthenes (384–322 BC), who was considered to be the most eloquent speaker of his time, is often represented with a



FIGURE 1. (A) Pre-Columbian pottery-Lambayeke culture of Peru. Völkerkundemuseum, Berlin. (B) Prominent ears. Greek Terracotta. Art and History Museum, Geneva. (C) Roman lady. Museum of Vatican. (D) Roman head. Archeological National Museum Madrid. (E) Terracotta. Geneva Art Museum. (F) Marble statue of a Greek warrior (490 BC). Aegina. (G) Demosthenes sculpture. National Museum Roma. (H) Hellenistic sculpture. Benaki Museum. Athens.

malocclusion and hypoplastic mandibula, which corresponds to his known misarticulating of the Greek $\rho\omega(r)$ (Fig. 1G).

More complex craniofacial malformations have also been represented, like Down syndrome (Fig. 1H), oxycephaly (Fig. 2A). In the Archeological Museum of Limassol (Cyprus), we were able to photograph an exceptionally well-designed and preserved sculpture of a princess, with an obvious dish-face deformity, brachycephaly and exorbitism, typical of Apert or Crouzon’s disease (Fig. 2B). An interesting archaic Greek statuette (575 BC) found in Selinonte (Sicily) and exposed in the Art and History Museum in Geneva has been examined and published by several historians,^{2,18} as representing a typical patient with phoco- or ectro-melia, as both lower limbs and the left upper limb are missing (Fig. 2C). On closer examination of the face, however, one notices a deformity of the right eyelid slant and nose, which had been overlooked by medical historians who never had encountered such anomaly (Fig. 2D). For us, it represents a patient with Tessier no 3 cleft or naso-ocular dysplasia, as described by Van der Meulen et al.¹⁹ Another undated pottery, exposed in the Berlin Völkerkundemuseum, is even more striking, as its complex facial deformity evokes a nasomaxillary dysplasia or a severe craniofacial microsomia (Fig. 2E), comparable to the masks described by Hwang and Chung.¹⁴

TRAUMA

Several Greek painted vases illustrate the result of trauma to the face, particularly in athletes and boxers.²⁰ In some patients, nose-bleed is even visible on the painting (Fig. 2F). Broken or deviated noses have also been illustrated in statues of known personages of

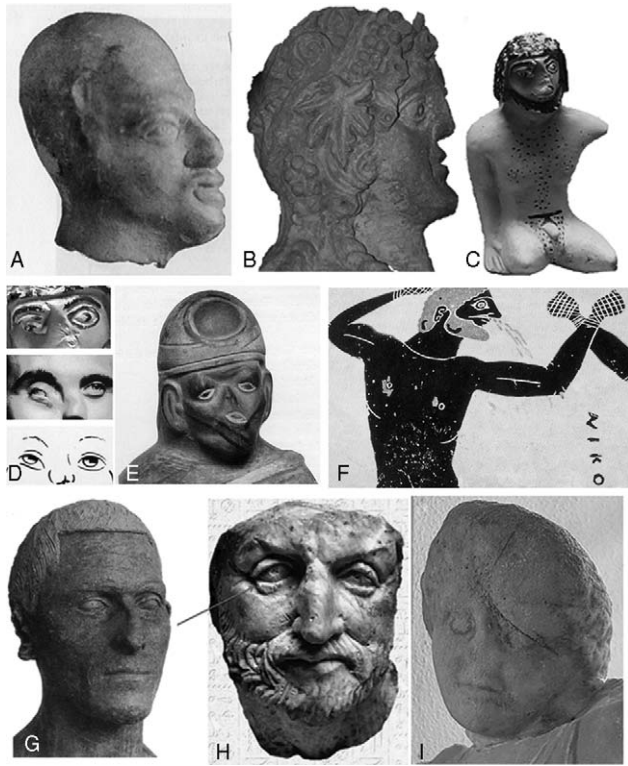


FIGURE 2. (A) Smyrna Terracotta. Louvre, Paris. (B) Hellenistic sculpture. Archeological Museum, Limassol Cyprus. (C) Archaic Greek terracotta. Art and History Museum, Geneva. (D) Archaic Greek terracotta. Facial detail. Art and History Museum, Geneva. (E) Pottery of South American origin. Berlin Völkerkundemuseum. (F) Painting on Atticus amphora (530 BC), Archeological museum, Athens. (G) Etrusc portrait, Villa Giulia, Roma. (H) Ivory sculpture. Archeological museum Thessaloniki. (I) Ex-voto. Asclepius Museum. Epidaurus.

Antiquity (Fig. 2G). The most interesting patient with trauma iconodiagnosis is the discovery of a small marble statue in Vergina (Northern Greece), representing the head of Philip II of Macedon, the father of Alexander the Great (Fig. 2H). On this very realistic piece of art, it is noticeable that the right malar bone is depressed and induces a moderate lower eyelid ectropion. This deformation corresponds to the wound Philip sustained during the battle of Methoni (355 BC), when he was hit by an arrow, penetrating the right orbit, as reported by several authors.^{21,22} Although the arrow was removed and the wound well treated by the famous army surgeon, Critobolus, the deformity remained visible. This has been confirmed, in spite of the body's cremation, by the discovery of the king's skeletal remains, showing healed wounds on the right malar and maxillary bones. The combination of literary accounts, iconodiagnosis and paleopathologic analysis is extremely rare, as in the case of Philip II, but representations of known personages, with a broken nose or other trauma sequels are more frequent.

An interesting statue is exposed in the archeologic Museum of Epidaurus on the Greek Peloponnese where the sanctuary was dedicated to healing and the gods Apollo and Asclepius. It represents certainly an individual who suffered a severe injury. The deep furrow crossing the left orbit and frontal bone reflects with no doubt the consequences of a forceful impact and possibly lethal trauma. We suppose that the statue has been placed as an ex-voto, to thank the gods for the survival of the person who was hit (Fig. 2I).

Burn sequels on the face might appear on a few ancient sculptures or paintings. However, the most striking neck burn retraction with its effect on the face is clearly represented on one of the wood Easter Island sculptures, the "moai miro" (Fig. 3A). Although historians



FIGURE 3. (A) Easter Island moai (wood carving). Barbier Müller Museum, Geneva. (B) Smyrna Terracotta National Museum of antiquities, Leyde. (C) Huaco statue. Berlin Völkerkundemuseum. (D) Pottery jug. Peru. Mochiga period. British Museum. (E) Limestone portrait of Akhenaton. c. 1365 BC. Aegyptus Museum. Berlin. (F) Terracotta Hellenistic period, Museum of art and history, Geneva. (G) Terracotta Hellenistic period. Louvre Museum, Paris. (H) Peruvian pottery. Völkerkundemuseum, Berlin.

have attributed the facial deformation represented on this small statue to a congenital anomaly or a hemiplegia, it seems clear for a plastic surgeon dealing with burns, that the severe right side neck burn retraction has pulled the soft tissues of the right face, including the lip, the cheek and the lower eyelid, as it is sometimes observed following severe untreated burns.

DISEASES AND TUMORS

The most common facial diseases represented in antic art are certainly the asymmetrical faces due to facial nerve palsies, whatever their origin: leprosy, tumor, stroke, or idiopathic. One may find such figures in the Greco-Roman world (Fig. 3B), as well as in South and Central America (Fig. 3C-D). Endocrinal disease such as acromegaly with enlargement of the jaws, nose, and forehead, is represented for the first time in a lime stone portrait of Akhenaton (Amenothep IV), Egypt 23rd dynasty, c. 1365 BC (Fig. 3E). But figurines with overgrowth of the maxillaries and forehead suggesting patients with acromegaly can also be found in a few Greek terracotta figurines (Fig. 3F-G).

Some diseases may leave severe mutilations of the face, particularly in the nasal and upper lip region. In Central and South America, people were often affected by the leishmania parasite. The cutaneous form of leishmaniasis may destroy the mid facial soft tissues that are the nose and upper lip. The disease is called "uta" in Peruvian Andes and may well have been represented in several small potteries (Figs. 3H and 4A-B). Alternatively, one may consider some of these lesions as typical mutilations of "noma disease" classified as type II sequel (Fig. 4C).



FIGURE 4. (A) Pottery vase. Chimbote, Peru. Mochica culture. Cambridge Museum of Archeology. (B) Pottery vase. Chimbote, Peru. Mochica culture. Völkerkundemuseum, Berlin. (C) South American Pottery. Cambridge Museum of Archeology. (D) Hellenistic head. Museum of Tarente, Italy. (E) Pottery. Nigeria. Nok culture. First century BC. (F) Smyrna terracotta. Louvre, Paris. (G) Smyrna terracotta. Louvre, Paris. (H) Smyrna terracotta. Louvre, Paris. (I) Ex-voto. Naples Museum. (J) Peruvian pottery. Völkerkundemuseum, Berlin.

Noma affects principally children of the Sub-Saharan African countries, but the disease is still endemic in poor South American regions.²³

Tumors on the face are not often represented in art. A good example of a possible teratoma of the orbit or a retinoblastoma has been sculpted on a Hellenistic head, probably as an ex-voto (Fig. 4D). Representations of tumor-like protuberances on a Nigerian sculpture seem to us that the person was affected by Recklinghausen neurofibromatosis (Fig. 4E).

The symptoms of tetanus were already described in Hippocratic treatises. The suffering of patients showing clenched teeth and backward heads' position, most probably infected by tetanus, is well depicted by artists in figurines of Smyrna (Fig. 4F-G). Signs of rhinoscleroma, rarely seen today are probably illustrated in another Smyrna terracotta (Fig. 4H). Leprosy may affect the face in various forms, and may have been represented as a cutaneous disease, as shown on a Greek ex-voto (Fig. 4I), or a nasal collapse and swelling of the upper lip as displayed in a South American pottery (Fig. 4J). Of course, most of these interpretations are conjectural; however, it is interesting to discover pathologies that are rarely seen nowadays, but have impressed our ancestors, although they had no idea about the origin of these diseases.

PALEO-PATHOLOGY

The study of ancient pathology is based on 3 elements: literary texts describing diseases, icono-diagnostics and paleopathology.²⁴ For the first 2, modern retrospective diagnosis is based on the narrative and the pathologic morphology perceived by the narrator or artist, without being conditioned or transformed or normalized by formal medical knowledge. Since antiquity, the notion of disease has fundamentally changed. Morbid entities and their nomenclature are recent creations, which do not necessarily correspond to the diseases described or illustrated in antiquity.

In contrast, paleopathology, that is, the study of human remains, mainly bones, provides much more precise information, especially since

the methods used, as in forensic analysis, have benefited from many discoveries in the life sciences, such as macro and microscopic morphology, osteometry, endoscopy, radiographic and magnetic resonance techniques, biochemical analyses, genetics, and densitometry.

Nevertheless, icono-diagnosis allows the identification of pathologies undetectable on fossils or cadaveric remains. As far as the craniofacial sphere is concerned, the expertise—the eye—of a plastic or a craniofacial surgeon can greatly contribute to the diagnosis and help the archaeologist or the medical historian to perceive or refine the type of pathology represented by the artist. Moreover, following the pioneer work of Eugen Holländer, bearing in mind the search for a new example of icono-diagnosis may become a fascinating game, while visiting an archeologic museum.

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