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LETTER TO THE EDITOR

A Crouzon syndrome from the Classic period of Maya civilization?

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Abstract
This article describes a retrospective diagnosis through an artistic representation of a pre-Columbian Central America bowl figuring a child with clinical characteristics of Crouzon syndrome. The report also highlights the importance of icono-diagnosis for a better description of the existing diseases into ancient societies.

Keywords Cranial deformities · Crouzon syndrome · Paleopathology · Maya civilization · Iconodiagnosis

Most of the information about the classic period of Maya civilization, its history and culture, come from the remains of their architecture and art, including stone carvings and inscriptions on their buildings and monuments. In this period, they made paper from tree bark and wrote in books using a system of hieroglyphs. The use of icono-diagnosis increases knowledge about health and diseases in ancient societies besides to be a great contribution to medical learning [2].

Icono-diagnosis was first used by the physician Anneliese Pontius of Harvard University in 1983 when she described the presence of Crouzon syndrome in the human representations of Cook's Island [8]. More recently, Pachajoa and Rodriguez described two possible representations of the disease in a ceramic artifact from pre-Hispanic populations in South America [7]. Here, the authors report a possible art representation of Crouzon syndrome in a ceramic bowl from a French museum of anthropology and extra-occidental art.

We carried out a direct examination of a small statue conserved at the Musée du quai Branly-Jacques Chirac, Paris (Fig. 1). The figurine is a bowl shape made of polychrome ceramic from Quiché department (region of Nejab) in Guatemala, dating of the classic period of Maya civilization (600–800 AD) (N° Inv. 71.1965.28.1-2). Retrospective diagnosis was given accordingly.

The anthropomorphic image is that of a child or adolescent presenting a typical head, bilateral exophthalmos, ocular hypertelorism, strabismus, prominent nose, facial hypoplasia, micrognathism, caused by a possible craniosynostosis (Fig. 2). This corresponds totally to what the French neurologist Octave Crouzon, has described as "Crouzon syndrome" in 1912: a rare genetic disorder involving craniofacial skeleton development, characterized by a triad of cranial deformities, facial anomalies (midfacial hypoplasia), and exophthalmia [3, 4].

The diagnosis of Crouzon syndrome in this anthropomorphic image followed, in the foreground, the semiological reasoning from the changes of the body forms. A second analysis was carried out through the possible diagnoses found in the society which belongs to the image according to medical and cultural anthropology.

The previous published cases were described in adult individuals [7, 8]. A person affected by Crouzon syndrome could eventually reach adulthood in prehistoric population, but in cases of few deformities and probably by the absence of cranial hypertension and clinical aspects would be mild. According to our experience, Fisherman's god statues described by Pontius as representation of Crouzon syndrome in Cook Islands do not fulfill clinical criteria to diagnose that syndrome [8]. Nevertheless, the abnormalities found vary greatly from case to case including variations between affected members of the same family. The order and rate of fusion of the suture determine the degree of deformity and inability [1].

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Fig. 1 Frontal view of the bowl (Guatemala, Department of Quiché, Nebaj. 600–800 AD) (picture P. Charlier)

The main clinical features noticed in the statue were proptosis and head deformity. Proptosis or exophthalmos describes a condition where the eyeball protrudes from the eye socket, making it appear to bulge. It can affect one or both eyes. Head deformity can be caused by craniosynostosis, which is a congenital malformation resulting in premature fusion of one or more sutures of the human skull. The most frequent syndrome associated with exophthalmos and craniosynostosis is Crouzon syndrome [6, 9].

Its clinical features include deformation of the skull, proptosis (exophthalmos), strabismus, maxillary hypoplasia, underdeveloped upper jaw, ogival palate, dental problems, opening in the lip (cleft lip), and a parrot-beaked nose. Affected individuals exhibit a variety of signs and symptoms, which differ in severity. People with Crouzon syndrome may have hearing loss, average intelligence. Crouzon syndrome has a prevalence of 1 in 25,000 live births worldwide, and it is approximately 4.8% of all craniosynostosis [5].

The iconographic signs present in the ceramic artwork suggested the presence of Crouzon syndrome among pre-Hispanic populations of Central America. The artwork showed in this report testifies that Crouzon syndrome was represented by Maya's civilization long before the Europeans.

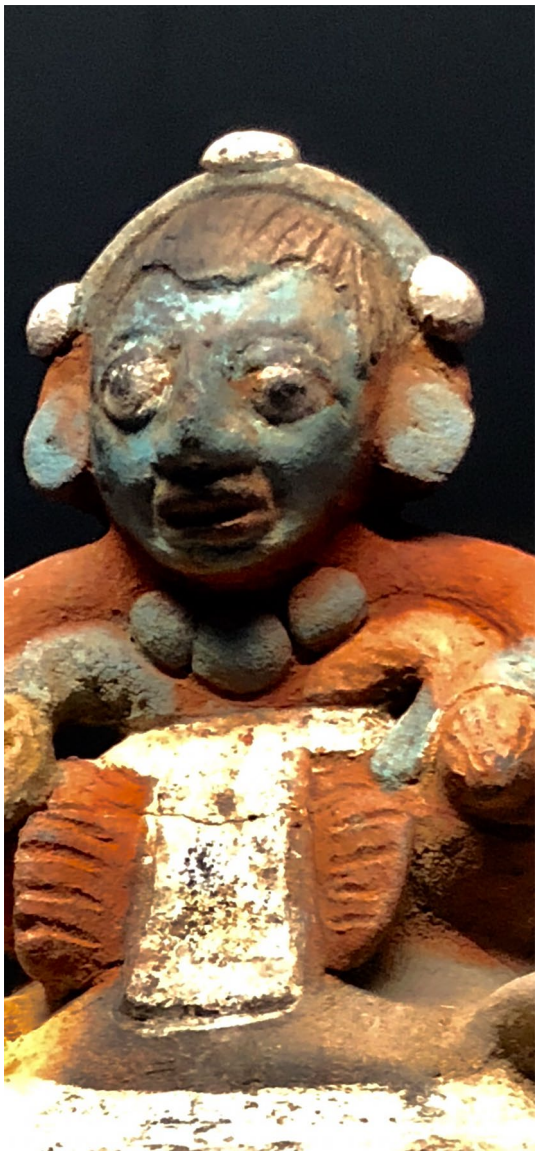

Fig. 2 Detail of the head of the ceramic (picture P. Charlier)

Moreover, the study uses anthropomorphic images, which are mostly of unknown authorship, but who has probably witnessed the dysfunction which he depicted [8], brings us a great deal of socio-cultural information from past or distant societies such as those displayed in the Musée du quai Branly's collection in Paris, France.

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Compliance with ethical standards

Conflict of interest Both authors declare that they have no conflict of interest related to the subject of this article.

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