

CONGENITAL DISEASES IN TENERIFE'S NORTHWEST: THE CASE OF ANEMIA

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Abstract. Traditionally, anemia has been diagnosed in paleopathology by the presence of cribotic lesions (cribotic syndrome) in the orbital root (*cribra orbitalia*), the external surface of the skull vault (porotic hyperostosis) and the upper metaphyses of the long bones (*cribra femorii*, *cribra humerii* and, even, *cribra fibulae*). Apart of anemia, other causes for these lesions have been argued (chronic infections, parasitism, malnutrition, etc). During the prehispanic period of Tenerife (be-

fore 1496), only less than 15 cases in a collection of 2000 individuals show these lesions. However, since the late 17th and 18th centuries something changed in the northwestern area of the island and the frequency of the cribotic lesions selected for this study (*cribra orbitalia*, porotic hyperostosis and *cribra femorii*) increased in an astonishing way. Description of the lesions, distribution by age, differential diagnosis and possible causes are discussed in this paper.

Keywords: Tenerife, Los Silos, *Cribra orbitalia*, Hyperostosis porotica, *Cribra femori*, anemia.

INTRODUCTION

After almost a century of fights, in 1496, the Spanish conquest of the Canary Islands ended when the Guanches of Tenerife surrendered to the Spaniards after the battles of La Laguna and Acentejo, both, - in the northern slope of the island - and were almost decimated by the epidemic known as “modorra” or “moquillo” (1494-1495) that most probably was the first influenza epidemic in the archipelago (Rodríguez-Martín, 1994; Rodríguez-Martín & Hernández-González, 2005, Rodríguez-Martín & Martín-Oval, 2009, Rodríguez-Martín & Martín-Oval, 2014).

Between the end of the 15th and the beginning of 17th centuries, an important number of European families – many of them with the aim to reach the Americas - settled in the island conforming the new population that was formed by elements of the aboriginal and the European populations that, in general, were mixed from the beginning. However, some places, like those of San Miguel de Abona in the South, Mountains of Anaga in the northeastern slope and Daute in the northwest, remained isolated of the rest of the island territory due to its orography that complicated communications by roads or paths (Berthelot, 1842; Rodríguez-Martín, 1994; Rodríguez-Martín, 2000, Rodríguez-Martín & Martín-Oval, 2009).

In general, Tenerife was well connected to the exterior thanks to the important ports of Garachico, in the north, and Santa Cruz, in the southeastern slope. When the eruption of Trevejo volcano (also called Montaña Negra or Arenas Negras) destroyed Garachico in 1706 the port of Santa Cruz was progressively increasing its importance until the city was independent of San Cristóbal de La Laguna in 1803 and nominated capital of the then unique province of the Canary Islands in 1833.

However, as we mentioned before, due to its orographical characteristics (high mountains, deep ravines and large forests) isolated areas existed in the Anaga mountains (northeast), San Miguel de Abona (south) and Daute demarcation (northwest). This isolation was the responsible of high rates of inbreeding conditioning the presence of congenital diseases in those areas showing a much higher prevalence than those of the Spanish Peninsula and other European regions. Many of these conditions were caused by local mutations occurred between the late 16th and the early 18th centuries.

CONGENITAL DISEASES IN THE PREHISPANIC POPULATION

An important number of congenital diseases (most of them observed in the skeleton, especially those of the spine), were present in the prehispanic period of Tenerife in the whole island (Rodríguez-Martín & Campillo, 1994, Rodríguez-Martín, 1995; Rodríguez-Martín & Martín-Oval, 2009). The conditions most commonly observed are the following:

- Spina bifida occulta, especially that involving the sacrum, shows a frequency of 10-20% depending of the geographic area.
- Sacralization of L5 was the second most common congenital malformation with a frequency between 7 and 10%.
- Lumbarization of S1 accounted for 4-6%.
- Vertebral block. This malformation is not so common as the previous (1-2%).

Less commonly appeared malformations of the skull (scaphocephaly, plagiocephaly) and limbs (clubfoot – Rodríguez-Martín & Isidro, 2003; Blount's disease – Barreda López, 1986) with a prevalence under 1% in all the cases.

UNCOMMON CONGENITAL DISEASES OBSERVED IN TENERIFE'S NORTHWEST IN RECENT YEARS

Severe congenital diseases related to inbreeding have been diagnosed in this part of the island in recent years. Their etiology seems to be a mutation occurred between four to two centuries ago. These diseases are:

From an hematological perspective

Two hematological diseases showing a surprisingly high frequency in comparison to other areas of the world were detected in the northwestern part of the island by the hematologists of the Health Canarian System. The frequency of both conditions is much higher than in any other part of Spain or Europe:

- A congenital coagulopathy consisting in deficiency of coagulation factor XIII leading to severe hemorrhagic disturbances (1.88% per year per patient). This disease is normally autosomal recessive and very uncommon and it is probably caused by inbreeding that could produce a mutation, as it is revealed by its high prevalence in this isolated area of Tenerife (García-Talavera, 2008).
- Congenital afibrinogenemia is a rare inherited blood disorder due to lack or malfunction of fibrinogen, the essential protein for coagulation.

From a cardiological perspective

- Dilated cardiomyopathy in families (related between them although they didn't know that before) with a common ancestor who suffered a mutation more than 200 years ago (García-Pavía et al. 2013). The disease involves the cardiac muscle that is unable to keep the cardiac pumping and may lead to sudden death (Cuenca et al., 2016). The condition is hereditary and can be transmitted by women or men but only men develop the disease.

THE ANCIENT CONVENT OF SAN SEBASTIÁN DE LOS SILOS

Located in the small village of Los Silos (municipality in Tenerife's northwest), the Convent of San Sebastián was founded in 1649. The exact chronology of its funerary space lasts since 1719 to 1885.

According to the Municipal Archives of the village, between 1719 and 1837 the nuns were buried there and between 1871 and 1885 the funerary space was used for civilians too, although many people (mainly newborns and fetuses as the archaeological excavation of the site demonstrated) were buried there along the complete period of use of the funerary area.

THE ARCHAEOLOGICAL EXCAVATION

The funerary space of the convent was excavated in 1996 by a team of Tenerife's Archaeological Museum during the works of restoration of the building in order to give it other uses. During the excavation hundreds of skeletal remains, many of them fetuses and newborns, that were buried there in ossuaries at the level of the chorus of the convent – with big amounts of quicklime around the bodies and bones (practice that was very usual at that time to prevent the smell and possible contagions, Rodríguez-Martín and Martín-Oval, 2017) appeared there. Individual coffins, mostly belonging to adult individuals, were also discovered and excavated “in-block”. (Fig. 1)

CRIBRA ORBITALIA, POROTIC HYPEROSTOSIS AND CRIBRA FEMORII IN THE CONVENT OF SAN SEBASTIÁN

After observing the presence of possible markers of anemia (especially on the skull) in some individuals buried in the funerary space, a sample of skulls



Fig. 1. Child burial with quicklime. 18th – 19th centuries. Convent of Los Silos (Tenerife).

and femora was selected arbitrarily for further pathological analysis in searching for *cribra orbitalia* (Fig. 2), *porotic hyperostosis* (Fig. 3) and *cribra femorii* (Figs. 4, 5), that – in general – shows peaks during childhood and adolescence decreasing later with age. The frequency of the lesions was very important as we can check here:

Skulls

Fifty skulls (12 adult males, 7 adult females and 31 subadults) were analyzed for observing the possible existence of *cribra orbitalia* and hyperostosis porotica.

- Adult skulls: 5 show *cribra orbitalia* (3 males and 2 females) and 1 shows both conditions (male). As we see, six of 12 skulls (50% of the adult population) show cribotic lesions.
- Subadult skulls: 12 show *cribra orbitalia*, 9 *cribra orbitalia* and porotic hyperostosis and only one case presents porotic hyperostosis.



Fig. 2. *Cribrá orbitalia*.



Fig. 3. *Porotic hiperostosis*.



Fig. 4. *Cribra femorii*.



Fig. 5. *Cribra femorii*.

The ages of the subadult individuals are:

- 0-2 years (18 individuals): 5 *cribra orbitalia*, 4 both conditions and one porotic hyperostosis.
- 3-4 years (10 individuals): 5 *cribra orbitalia* and 4 both conditions.
- > 5 years (3 individuals): 2 *cribra orbitalia* and 1 both conditions.

These data show that more than 70% of the subadult sample was affected by one or both conditions.

Femora

Cribra femorii seems to be more frequent in females and decreases with age in the general population. Regarding its presence in this sample, seven adults and 30 subadults were analyzed for observing the lesion.

- Adults: on the contrary to the skull sample, none of the seven adults show the condition.
- Subadults: 30
 - Perinatal (8 individuals): no signs of *cribra femorii*.
 - 0-3 months (4 individuals): all of them show the condition.
 - 4-6 months (5 individuals): 2 show *cribra femorii*.
 - 7-12 months (6 individuals): 5 show *cribra femorii*.
 - > 1 year (7 individuals): all of them show the condition.

18 subadult individuals of 30 selected in this sample show *cribra femorii* (60%) and if we discount the group of perinatal individuals in which the lesion does not appear, the frequency of the condition increases until almost 82%.

It is clear that the three conditions (*cribra orbitalia*, porotic hyperostosis and *cribra femorii*) show an astonishingly high frequency in the selected sample, especially among the subadult group.

Due to the fact that the skeletal remains come from an ossuary, it was impossible to relate skulls and femora.

In an interesting survey of porotic lesions on immature skeletons from Stara Torina (late medieval Serbia), Djuric et al (2008) found a prevalence rather similar to that of Los Silos, although the authors attributed the cause to infectious diseases that were very common in the area (thing that didn't happen in Tenerife's northwest at that moment).

ETIOLOGY OF THE POROTIC CONDITIONS

Porotic syndromes, especially *cribra orbitalia* and porotic hyperostosis are not always related and, according to Stuart-Macadam, 1989a and Rivera and Mirazon Lahr, 2017, they can reflect different underlying conditions. In fact, different causes have been argued to justify these cribrotic lesions being the most common etiology the following (Ortner & Putschar, 1985; Stuart-Macadam, 1989b; and Aufderheide & Rodríguez-Martín, 1998).

- Anemic syndromes:
 - Acquired (the most common type is that of iron-deficiency). However, Walker et al. (2009) consider that iron-deficiency *per se* cannot sustain the massive red blood cell production and expansion of the bone marrow.
 - Congenital.
- Malnutritional status.
- Different parasitic infections (giardia, ascaris, plasmodium, etc). In this case, its development is due to the typical anemia produced by the parasitic infections.
- Metabolic disturbances: scurvy, rickets.
- Trauma.

The general frequency of porotic lesions is rather variable in different populations around the world (between less than one per cent to 30% or even more according to Angel, 1966) and some authors have considered the anemic syndrome a biological mechanism of environmental adaptation and, even, protection, for decreasing the infectious diseases activity.

TYPES OF ANEMIA

According to different authors (Bunn, 1981; Díaz-Fernández, 1984; González-García, 2012), the most common types of anemias can be classified, in a very summarized form, in the following:

Most common types of acquired anemias

Iron - deficiency anemia (ferropenia), B 6 and B 12 vitamin deficiencies (megaloblastic anemia, that for Walker et al., 2009, is the most plausible cause of the porotic syndrome), extrinsic hemolytic anemia (paroxysmal nocturnal hemoglobinuria or PNH and autoimmune hemolytic anemia or AHA), anemia by chronic disturbances (aplasia, myelodysplasia).

Most common types of congenital anemias

The intrinsic hemolytic anemias include erythropathic anemia (spherocytosis, elliptocytosis), enzymatic deficit (G6PhD, pyruvate kinase), hemoglobinopathies (Beta Thalassemia Major or Cooley's Disease, Alfa Thalassemia, Sickle Cell Disease).

Fanconi's anemia is an uncommon hereditary and severe disease involving the three blood cell series: erythrocytes, leukocytes and platelets.

Of the above mentioned anemias, the most common universally is that produced by iron deficiency (ferropenia). However, iron-deficiency anemia is absent in neonatal individuals – in those individuals the most common causes of anemia are blood loss that can be related to coagulopathy, decrease in red cell production or hemolysis, besides some other physiological processes - and the iron – deficiency may appear months after birth. This datum is important for the cases observed in the Convent of Los Silos that points to a congenital cause.

Regarding congenital anemias, it is important to note the following data:

- Spherocytosis: it does not usually manifests before five or six years of life.
- Elliptocytosis: although extremely uncommon, it may appear in the newborn.
- Deficit of G6PhD or favism: it is the most common enzymatic deficit around the world and is linked to the X chromosome but does not appear in the newborn.
- Deficit of pyruvate kinase: it may appear, in exceptional cases, in the newborn as *hydrops fetalis*. Anemia appears few months after birth.
- Thalassemia: of the congenital anemias produced by abnormal hemoglobin the most severe is the Beta Thalassemia Major in which Beta globin is absent and this leads to fragility of the red cells and subsequent massive hemolysis in the spleen. Although universal, it is typical of the Mediterranean Basin.
- Sickle cell disease is a special hemoglobinopathy affecting black people almost exclusively. It causes hemolysis and skeletal disturbances.
- Fanconi's anemia: uncommon condition that develops gradually affecting the three blood cells series during the subadult age. It is not related to the presence of porotic syndrome.

CRIBRA ORBITALIA, POROTIC HYPEROSTOSIS AND CRIBRA FEMORII IN THE GUANCHE POPULATION OF TENERIFE

Only isolated cases, scattered through the island, were observed among the Guanche population. In total, the three conditions (*cribra orbitalia*, porotic hyper-

ostosis and *cribra femorii*) account for less than 15 cases with prevalence under 0.5% of the entire Guanche population, fundamentally affecting subadults (Rodríguez-Martín & Rodríguez-Maffiotte, 1986; Rodríguez-Martín, 1995; Rodríguez-Martín & Martín Oval, 2009).

EVALUATION OF THE PROBLEM

After evaluating the different possibilities for the very high prevalence of the porotic syndrome in the Convent of San Sebastián in Los Silos, we exclude metabolic diseases, malnutrition or infectious conditions due to the pathological picture observed in the sample and because those disturbances did not have an important prevalence during the postconquest history of this area of Tenerife. Therefore, we conclude that the most probable diagnosis is anemia. But the question is what type of anemia? In general, we think that the disease must be related to inbreeding and, most probably, the condition is one of the hemolytic types of anemia.

After reviewing the different types of congenital anemias, the probable etiology of the porotic lesions observed at the level of the orbits, skull vault and femur is an enzymatic deficit (common in the island of Tenerife). Of all possible enzymatic deficiencies we point out to that of pyruvate kinase due to the clinical picture in newborns and early infancy and because this type is the most common cause of anemia (of non spherocytic type) showing differences in its severity (the disease can produce death like seems to be the case).

The difference in the prevalence between prehispanic and postconquest (after the late 17th and early 18th centuries) populations of Tenerife is due, in our opinion, to a mutation occurred at that time in this area where inbreeding was constant during, at least, three centuries after the end of the Spanish conquest.

CONCLUSIONS

The differences observed between Guanche and historic populations regarding the porotic lesions on the skull (*cribra orbitalia* and porotic hyperostosis) and femur, in the whole island, but very especially in Tenerife's northwest, may indicate that around three centuries ago a mutation producing an anemic disease occurred there and affected the population since the earliest phases of life causing high mortality in that area of Tenerife. Of the possible causes of that anemic condition we

think that pyruvate kinase deficiency is the probable responsible although further studies to be carried out on the total number of the individuals buried in the funerary space of the convent of San Sebastián are needed.

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