



Case Study

A severe case of congenital aural atresia in pre-Columbian Venezuela

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ABSTRACT

A severe case of unilateral congenital aural atresia was observed in an adult male from the pre-Columbian archaeological site of Los Tamarindos in Venezuela. Macroscopic analysis of the right temporal showed a complete absence of an external auditory meatus. Further examination of the cranium using a micro-CT scanner revealed a fusion of the incus and malleus on the affected side. Modern clinical data suggest this would have resulted in hearing loss on the affected side and was likely accompanied by visible malformations of the external ear. The placement of antlers besides the head of this individual may represent a social acknowledgment of the physical impairment by this prehistoric community.

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1. Introduction

Reports on congenital aural atresia – a pathological condition involving malformation of the external and middle ear – are rare within the archaeological and palaeopathological literature. Cases of aural atresia include several prehistoric Amerindian crania from Peru and the United States of America (Hrdlička, 1933) and more recently from Anglo-Saxon England (Wells, 1962), the United States of America (Hodges et al., 1990), Slovakia (Masnicová and Beňuš, 2001), Bulgaria (Keenleyside, 2011), Canada (Swanston et al., 2011), and Peru (Kato et al., 2007). Additionally, the presence of congenital aural atresia as part of a syndrome has recently been reported in two archaeological case studies. Knüsel and Bowman (1996) reported atresia of the external auditory meatus as one of the symptoms of an individual with a potential diagnosis of von Recklinghausen's neurofibromatosis from St. Bride's Church in London. A probable case of Goldenhar syndrome, including unilateral aural atresia, is documented in an individual from Germany by Panzer and colleagues (2008).

A discrepancy exists between these scarce reports of the condition in prehistoric skeletal assemblages and the assertion in current clinical literature that the condition occurs on average in one per 10,000–20,000 newborns (Gill, 1969; Declau et al., 1999; Kösling et al., 2009). This article briefly reviews the clinical and palaeopathological literature on congenital aural atresia to support

the diagnosis of the condition in an adult male from the pre-Columbian site of Los Tamarindos in Venezuela and discusses the medical ramifications of this particular malformation. The social construction of impairment and disability in this society and the potential consequences for the treatment of this individual by other members of his group are investigated through an analysis of the burial context.

1.1. Congenital aural atresia

Congenital aural atresia is defined as the malformation or absence of the external auditory canal, usually in association with middle ear abnormalities and deformation of the pinna (the latter condition is also known as microtia) (Swartz and Faerber, 1985). In rare cases, involvement of the inner ear structures may also be present (Schuknecht, 1989; Declau et al., 1999; Kösling et al., 2009). Features associated with severe congenital aural atresia include a complete absence of the external auditory canal, a hypoplastic middle ear cavity, severe ossicular deformities, poor mastoid aeration, a bony atresia plate (tympanic bone deformity caused by lack of auditory canal development), a small tympanic cavity, anteriorly located descending facial nerve canal, and the absence of the oval window and/or round window (Swartz and Faerber, 1985). The majority of cases are unilateral, more commonly affecting the right ear. Furthermore, congenital aural atresia is observed more frequently in males than in females (Gill, 1969; Schuknecht, 1989; Declau et al., 1999; De La Cruz and Borne Teufert, 2003; Kösling et al., 2009).

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Unilateral congenital aural atresia results in varying degrees of hearing loss in the affected ear depending on the severity of the malformation, whereas the unaffected side functions normally. Individuals with unilateral hearing loss will experience difficulties in sound localization and speech differentiation over background noise (Declau et al., 1999; Lieu, 2004). The intellectual, speech, and language development of the affected individual will not be impacted in a substantial manner (Declau et al., 1999; Kelley and Scholes, 2007). Furthermore, it has been suggested that individuals develop adaptive strategies during life to compensate for the unilateral hearing loss (Borton et al., 2010).

Information on the epidemiology of aural atresia and microtia is scarce. Different methodologies hinder global and regional comparisons, but a certain trend can be seen in the limited data available. Several studies report a higher incidence of aural malformations in individuals of Amerindian or Asian descent (Jaffe, 1969; Wiet, 1979; Harris et al., 1996), suggesting a degree of genetic influence in the etiology of these defects.

1.2. The social construction of disability

Recent years have seen increased interest in research on the social context of illness and disability within bioarchaeological studies, in line with the growing significance of social identities within the field (Hubert, 2000; Roberts, 2000; Marsteller et al., 2011). Following the social model of disability, evidence of physical impairment found in the skeletal remains of an individual is no longer directly equated to socially constructed notions of disability. Likewise, what constitutes a disability and how disabled persons are treated within a group are dynamic and flexible notions that may change over time (Dettwyler, 1991; Hubert, 2000; Roberts, 2000; Cross, 2007; Marsteller et al., 2011; Tilley and Oxenham, 2011; Southwell-Wright, 2013).

A multidisciplinary approach, combining the archaeological evidence with medical, historical, or ethnographic accounts, is best suited to investigate disability in past societies and to gain more insight into the repercussions of physical impairment in individual cases (Shakespeare, 1999; Roberts, 2000; Marsteller et al., 2011). In the same vein, Tilley proposed the bioarchaeology of care framework designed to assess past levels of health care, where care may consist of direct support of the individual or accommodation of the impairment, based on individual case studies (Tilley and Oxenham, 2011; Tilley, 2012).

Valuable additional information from historical sources has successfully been incorporated in studies on various periods of European history, but such an approach is far more challenging for societies that did not produce written documents (but see Marsteller et al., 2011). In such cases, early colonial sources may provide some insights despite their inherent limitations such as the applicability of colonial documents in reconstructing prehistoric societies and the bias of the author (Tosh, 2010). Archaeological data on deviant or abnormal burial practices including one of more of the following – burial location, burial structure, posthumous treatment of the body, grave goods – may also aid in the reconstruction of social notions on disability (Roberts, 2000; Marsteller et al., 2011).

2. Materials and methods

The skeletal remains of 23 individuals from the site of Los Tamarindos are curated at the Peabody Museum of Archaeology and Ethnology, Harvard University (PMAE). These are being studied by one of the authors (AVD) as part of a larger research project on indigenous identities in the circum-Caribbean through an assessment of intentional cranial modification. During an examination of

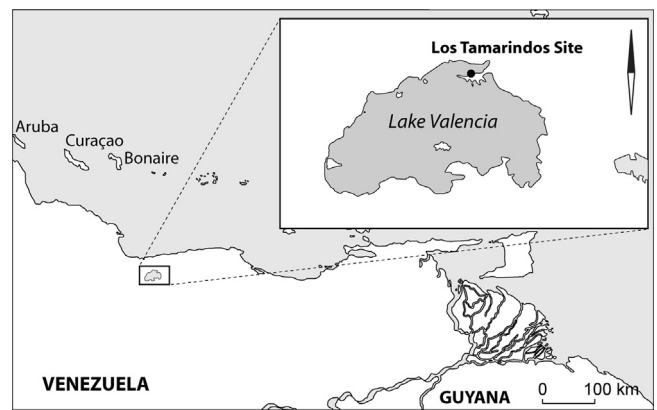


Fig. 1. Map of central Venezuela showing the location of Los Tamarindos.

the crania, the absence of an external auditory meatus was noted on the right temporal bone of PMAE 33-100-30/N895.0, originally numbered burial 27,¹ hereafter referred to as individual N895/27. Kidder's (1944) observations are the only published reference to this cranium and the malformation of the auditory canal.

The recovered skeletal remains of individual N895/27 consist solely of a cranium, none of the other skeletal elements visible in the burial drawing were shipped to the PMAE. There is minor postmortem damage to the right maxilla, the palate, and both zygomatic arches. An osteological assessment of sex and age was carried out using the standards outlined by Meindl and Lovejoy (1985) and Lovejoy (1985). High resolution computed tomography (CT) was used to evaluate the temporal bone at the Harvard University Center for Nanoscale Systems. An X-Tek micro-CT X-ray imaging system (model HMXST225) produced high resolution scans of both the affected and normal temporal bones, as well as a lower resolution scan of the entire preserved cranium.

The archaeological site of Los Tamarindos is located on the La Cabrera Peninsula, which projects into the North side of Lago Valencia in central Venezuela (see Fig. 1). It was excavated in 1933 and 1934 by A. Kidder II. Los Tamarindos is a settlement site containing burials within the habitation area with two distinct phases based on ceramic typology: La Cabrera (BC 550–AD 1000) and Valencia (AD 1000–1500). These dates are based on the relative chronology proposed by Rouse and Cruent (1958/1959, 1963), as no radiocarbon dates exist for the site of Los Tamarindos (Kidder, 1944; Rouse and Cruent, 1958/1959, 1963).

Kidder (1944) provides a drawing of the burial context of individual N895/27 – which can be seen in Fig. 2 – in addition to the following description:

“Sub-humus gravel. Adult male, flexed on back. Skull lacks all trace of auditory meatus, possibly explaining two antlers placed at this side of head.” (Kidder, 1944:52). Despite this assertion by Kidder, the antlers are depicted on the left side of the skull in the drawing. Unfortunately, the original field notes could not be located to resolve this discrepancy.

Burial N895/27 was encountered in the second meter of excavation. The primary nature of the burial and its context in the sub-humus layer indicate that the individual belongs to the earlier

¹ A note accompanying the skull identifies it as orig #122, bur #26, L-23, 1.10. However, Kidder (1944:52) clearly identifies burial 27 as the individual without the external auditory meatus in his publication. Burial 26 is described as a secondary burial of adult bones underneath a broken urn. The burial drawing does not show the presence of a complete skull, unlike the image for burial 27 (Kidder, 1944). In this case, the clear identification of burial 27 as the skull without an auditory meatus is accepted, as the accompanying note could have been accidentally switched at any point during shipment and subsequent storage of the cranium.

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