

Unilateral Congenital Aural Atresia from an Ychsma Group Burial at the Site of Pachacamac, Peruvian Central Coast

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Abstract

Congenital malformations of the human skeleton are a major area of palaeopathological and bioarchaeological interest, although our understanding of such conditions in the ancient world is hampered by their often extreme rarity, and inconsistent reporting in the field. A cranium from a group burial at the Peruvian site of Pachacamac was found to display almost complete absence of the right external auditory meatus, styloid process, vaginal process and tympanic plate of the right temporal bone. Following a differential diagnosis, it was determined that the skeletal pathology likely represents an instance of congenital aural atresia, a developmental anomaly resulting in the partial or complete aplasia of the external acoustic meatus. This condition is often associated with other congenital abnormalities and syndromes. However, as the postcranial remains of this individual could not be distinguished from other remains in the group burial, no further associated skeletal anomalies could be detected. This is the first instance of congenital aural atresia recovered from a defined funerary context in Peru, complementing other bioarchaeological reports of this rare congenital anomaly. The pathology of the condition is described, and compared with clinical and bioarchaeological data. The potential social implications concerning social attitudes of the Pachacamac population towards this individual are also considered, in reference to both historical and archaeological contexts.

Introduction

Congenital aural atresia has been frequently reported in the clinical record but has rarely been recorded in the archaeological literature. Several examples have been recovered from prehispanic Peruvian contexts (Hrdlička, 1933), with other reported cases in the United Kingdom (Knüsel & Bowman, 1996; Wells, 1962), Czech Republic (Vyhnánek & Kuzelka, 1998), United States (Hodges et al., 1990; Hrdlička, 1933), and more recently in Bulgaria, Canada, and Venezuela (Keenleyside, 2011; Swanston et al., 2011; van Duijvenbode et al., 2015). This article will explore the condition, the social context of archaeological

cases, and review a new discovery of congenital aural atresia in an Ychsma group burial at Pachacamac, Peru. It also provides information concerning how the Pachacamac population seem to have viewed this individual, whose condition does not seem to have precluded them from social acceptance insofar as burial context is concerned. This finding from the Ychsma is then contrasted with ethnohistorical information reflecting dissimilar behaviour in the subsequent Inka polity.



Figure 1: Map of Peru with the Location of Pachacamac (Eeckhout, 2021)

Congenital aural atresia

Congenital aural atresia is a birth defect causing malformation or absence of the external auditory canal, often associated with severe microtia (underdevelopment of the external ear) (Schuknecht, 1989; van Duijvenbode et al., 2015). It usually manifests unilaterally (Verma et al., 2016). Congenital aural atresia can be associated with facio-auriculo-vertebral spectrum (Goldenhar Syndrome), Pierre Robin Syndrome, Treacher Collins Syndrome, and diverse chromosomal abnormalities (Lo et al., 2014).

Pathological characteristics

Congenital aural atresia should not be confused with the atresia (narrowing) caused by external auditory exostoses, which is particularly associated with coastal populations from the Palaeolithic until the present day (Villotte and Knusel 2016; Trinkaus and Villotte, 2017). Characteristics of congenital aural atresia include partial or total aplasia of the external auditory canal, a

hypoplastic middle ear cavity, tympanic bone deformity (bony atresia plate), pinna (external ear) malformation, ossicular erosion and malformation, erosion of the tegmen tympani, anterior location of the descending facial nerve canal, bony atresia, as well as abnormalities of the jaw and alterations to levels of mastoid pneumatization (Tassano et al., 2015; Todd, 1994; Trojanowska et al., 2012; Verma et al., 2016). In practical terms, congenital aural atresia causes partial or complete deafness on the affected side; modern day interventions may include surgery, electronic implants, and/or auricular reconstruction (Lo et al., 2014).

Modern prevalence and distribution

Congenital aural atresia prevalence is estimated to be between 1:10,000 and 1:20,000 individuals (Karmody and Annino 1995; Lo et al., 2014). It is twice as common in males as females (Kelley & Scholes, 2007), affects the right ear more than the left (De la Cruz & Teufert, 2003), and is most common in Hispanic, Native American, and Western Asian populations (Ali et al., 2017). Prevalence of microtia/anotia (congenital absence of the ear) follows a similar pattern (Harris et al., 1996).

Archaeological Examples

Congenital aural atresia has been identified among archaeological human remains since the 1930s (Hrdlička, 1933). Of the cases summarised in Table 1, 55.6% are Amerindian, 38.8% are European and 5.5% are Western Asian; 81.2% affected the right side, and 75% of affected individuals were female. It is unlikely that these figures reflect ancient realities, as reporting has been inconsistent. Pachacamac has previously yielded several examples of congenital aural atresia (Hrdlička, 1933, p. 356), although these were surface finds without any archaeological context. This makes it impossible to map the frequency of the trait over time or to discuss the possible cultural attitudes towards this very visible disability. The current example is the first case of congenital aural atresia to be discovered in a culturally-specific context not only at Pachacamac, but the Andean area as a whole.

Table 1 Archaeological Cases of Congenital Aural Atresia.

Site	Country	Affiliation	Sex	Age	Side	Ref
Apollonia Pontica	Bulgaria	Classical	F	Adult	R	(Keenleyside, 2011)
<i>Mladá Boleslav</i>	Czech Republic	Middle Ages (13 th C)	F	Adult	?	(Vyhnanek and Kuzelka, 1998)
<i>Mladá Boleslav</i>	Czech Republic	Middle Ages (13 th C)	F	Adult	?	(Vyhnanek and Kuzelka, 1998)
Chapel Ossuary, Rain	Germany	Modern (1400-1800)	?	Subadult	L	(Panzer et al., 2008)
St Jacob's Chapel	Slovakia	Modern (AD 1500-1800)	?F	MA	R	(Masnicová and Benuš, 2001)
Caister-on-Sea	UK	Anglo-Saxon	F	YA	R	(Wells, 1962)
St Brides Church	UK	Modern (19 th C)	M	MA	L	(Knüsel and Bowman, 1996)
<i>Lachish</i>	Israel	Iron Age	?	Adult	R	(Lukaszek et al., 2010)
St Vitals Church	Canada	Modern (19 th C)	M	YA	R	(Swanston et al., 2011)
Chicama	Peru	Prehispanic	M	MA	R	(Hrdlička, 1933, p. 360)
Chicama	Peru	Prehispanic	F	YA/MA	R	(Hrdlička, 1933, p. 361)
Pachacamac	Peru	Prehispanic	?F	Subadult	R	(Hrdlička, 1933, p. 361)
Pachacamac	Peru	Prehispanic	F	YA/MA	R	(Hrdlička, 1933, p. 361)
Pachacamac	Peru	Prehispanic	F	YA	R	(Hrdlička, 1933, p. 361)
Pachacamac	Peru	Prehispanic	?M	MA	R	Current Paper
Arkansas	USA	-	F	YA	R	(Hrdlička, 1933, p. 361)
Carlsbad, NM	USA	-	F	YA	R	(Hrdlička, 1933, p. 361)
Jackson County	USA	Late Woodland	F	YA/MA	L	(Hodges et al., 1990)
Los Tamarindos	Venezuela	Prehispanic	M	Adult	R	(van Duijvenbode et al., 2015)

Materials and methods

Pachacamac (Figure 2) is a large multi-period, multi-polity monumental religious and burial site located 30km south of Lima (Figure 1). Founded in the 3rd century AD, Pachacamac rose to prominence as a habitation and ritual site under the Lima group, burgeoning to become the capital of the subsequent Ychsma polity (c. 1000-1476) during the Late Intermediate Period (LIP). It was subsequently conquered by the Inka (Late Period), becoming a prominent pilgrimage and religious site until its eventual conquest by European invaders in 1533 (Eeckhout, 2013; Eeckhout & Owens, 2008; Owens & Eeckhout, 2015).

The site is known for the Sacred Precinct, and its series of highly distinctive dynastic buildings ('pyramids with ramps') that are particularly associated with the Ychsma polity. These designs continued into the Late Period occupation, along with more typically Inka

constructions (including a Temple of the Sun, and an Acllahuasi [Temple of the Chosen Women]) associated with the site's important function as a religious focus for the Andean region. Considerably less is known about the residential areas of the site, most previous research having focused upon the monumental architecture of the administrative centre, and the Sacred Precinct.

The site also had a funerary function, with an estimated 80,000 interments spanning all periods of the site's occupation. Ychsma interments are noted for their variety, but range from single pit burials to group/communal tomb interments containing up to 130 individuals. Tomb architecture includes wood and reed supports, holding up a plant fibre roof. Bodies were usually buried wrapped in successive layers of textile and plant fibre, especially reeds. Group burials often follow a 'core and periphery' pattern, focused on older male individuals who are usually associated with the widest range of grave goods (Owens & Eeckhout, 2015).

Individual 2018 B15 (16/193) consists of a single cranium, recovered from a collective tomb containing the remains of over 30 individuals. The tomb was situated within the Sacred Precinct, adjacent to a small

temple that – unusually – was decorated with anthropomorphic and geometric designs. The tomb context was looted in antiquity and the human remains commingled, although the remains of prestige textiles,

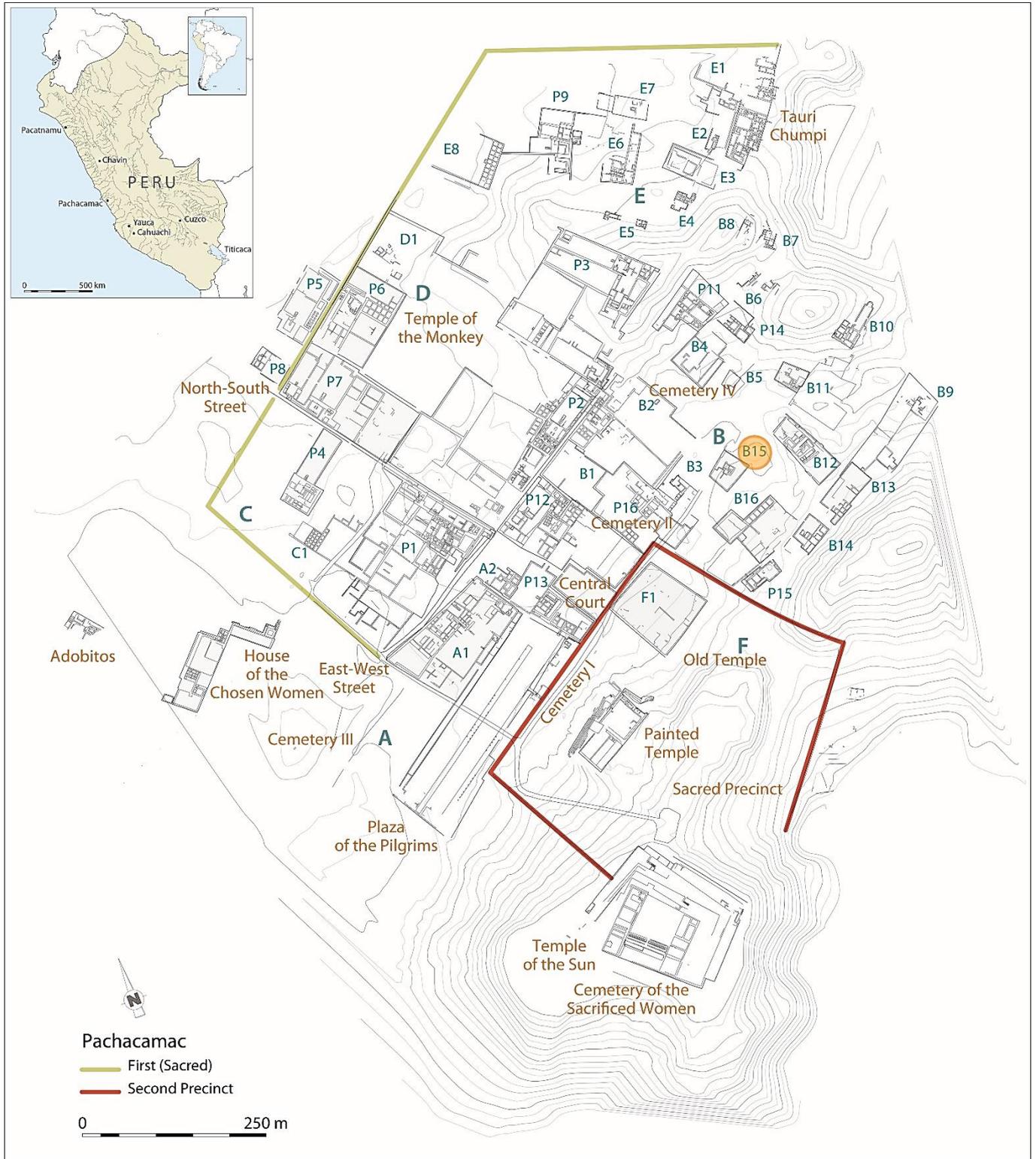


Figure 2: Site map of Pachacamac with the location of B15 (16/193) (Eeckhout, 2021)



Figure 3: Lateral view of the right temporal bone of B15 (16/193). (Owens, 2019)

feather garments, ceramics, and other grave goods were recovered. Ceramic analysis indicates a date in the Early Ychsma, around AD 1100-1200 (Eeckhout, Pers. Comm.). The cranium was not associated with a mandible, nor any postcranial remains. It was in good overall condition with no signs of breakage or extensive weathering.

The individual was analysed according to conventional methods for osteological sex estimation, consisting of observation of the glabella, supraorbital ridges, supraorbital margin, suprameatal crest, mastoid process, and nuchal crest (Acsádi & Nemeskéri, 1970; Bass, 2005; Buikstra & Ubelaker, 1994; Ferembach et al., 1980; Krogman & Iscan, 1986). Age estimation was necessarily restricted to dental wear and sutural closure (Buikstra & Ubelaker, 1994; Lovejoy, 1985), and thus provided only a broad estimation of age. A macroscopic analysis of the cranium was undertaken to examine oral and joint diseases, as well as any other pathological changes or developmental anomalies present.

Results and discussion

The individual was judged to be a probable male, based upon supraorbital anatomy, nuchal development and mastoid process morphology. Ageing was more tentative, given the absence of pelvic remains; tooth wear and sutur

al obliteration patterns suggested the individual was a middle/older adult (35-50+ years). In the right temporal region, the external auditory meatus was almost entirely absent, along with the styloid process of the temporal bone, the vaginal process and the tympanic plate (Figure 3, 4, 5). No abnormal irregular proliferative or destructive bone remodelling processes were observed in this region of the cranium, with the affected area consisting of smooth, uninterrupted cortical bone. The temporal region of the left side of the cranium appeared normal. Right side occipital condylar degeneration/lipping, septal deviation, bilateral cribra orbitalia, and slight asymmetry of the nasal aperture and orbits were also observed, although these are unlikely to be associated with the congenital aural



Figure 4: Right temporal region of B15 (16/193). (Owens, 2019)

atresia. Antemortem maxillary tooth loss was extensive, the molars being absent and the alveolar bone remodelled/remodelling. A partially healed dental abscess (left UM2) and a supernumerary tooth (adjacent to the right UI1) were also noted.

Differential diagnosis

A careful consideration of the exact appearance and distribution within the skeleton of skeletal abnormalities or changes is required to accurately determine which pathological processes may have been responsible, and to rule out others (Klaus, 2017). A differential diagnosis was undertaken using the suggested modified nomenclature of the Istanbul Protocol, as proposed by Appleby and colleagues (2015). The right temporal bone of Individual 2018 B15 (16/193) exhibited almost complete absence of the external auditory meatus, tympanic region, vaginal process, and styloid process. Differential diagnosis included pseudopathology, stenosis (narrowing) of the external auditory meatus (due to external auditory exostosis), acquired aural atresia (due to trauma, infection or neoplasm), and congenital aural atresia.

The excellent condition of the cranium indicated that the lesions were not consistent with pseudopathology. Partial stenosis of the external acoustic meatus can be caused by benign osseous growth within the canal, known as external auditory exostoses. However, the formation of exostoses within the auditory canal is not likely to result in complete occlusion of the external acoustic meatus and normally occurs bilaterally, and



Figure 5: Inferior view of the cranium of B15 (16/193). (Owens, 2019)

was, therefore, not consistent with the observed absence of the tympanic region, vaginal process, and styloid process of the temporal bone (House & Wilkinson, 2008).

The loss of anatomical features on the temporal bone can potentially be brought about by destruction and new bone formation as a result of trauma, infectious disease, or neoplastic disease, although such cases are rare (Bajin et al., 2015). A traumatic lesion causing severe deformity and structural absences without affecting surrounding areas was considered to be inconsistent with the observed pathology, due to the localised nature of the affected area. Similarly, infectious disease was unlikely to have been restricted solely to the auricular area, while the lack of neoplastic growth (in the event of a benign lesion) or perforative

pathology (suggestive of aggressive metastatic neoplastic processes) were entirely absent. The smooth, intact cortical surface and the lack of any associated abnormal irregular bone reaction, either proliferative or destructive, in this region of the cranium was thus not consistent with these pathological processes.

The absence of anatomical features is most likely to have occurred during the development of the temporal bone itself. The complex development and ossification of the ear and surrounding anatomical features of the temporal bone begin within the first few weeks of embryonic development (Cunningham et al., 2017, pp. 78–85). Thus, the condition observed was typical of a congenital disorder developing in utero.

Due to the complex nature of the anatomical structures of the ear, several developmental anomalies can affect this region. These variously affect the external ear (the pinna), the external acoustic meatus, and the middle and inner ears (Bartel-Friedrich & Wolke, 2007). The observed lack of development of the anatomical structures of the right ear is diagnostic of congenital aural atresia, this being the only condition that results in the complete absence of the external auditory canal, as well as tympanic bone deformity (Tassano et al., 2015; Todd, 1994; Trojanowska et al., 2012; Verma et al., 2016). Congenital aural atresia occurs most frequently in males, with a ratio of 2:1, and is three to five times more likely to occur unilaterally, with the right side most frequently affected (Liaw et al., 2017). This is consistent with our observations of individual 2018 B15 (16/193).

Although it can develop in isolation, congenital aural atresia is also often associated with other developmental anomalies and congenital syndromes (Lo et al., 2014). Malformation of the middle ear and auditory ossicles are often affected in instances of congenital aural atresia (Trojanowska et al., 2012). For example, CT scanning of an archaeological individual from pre-Columbian Venezuela with congenital aural atresia of the right ear demonstrated the fusion of the incus and malleus (van Duijvenbode et al., 2015). However, involvement of the middle ear was unobservable in individual 2018 B15 (16/193) due to

the lack of access to CT scanning equipment. Disturbance of the normal development and eruption of teeth has been clinically associated with congenital aural atresia (Boone et al., 2011); nothing abnormal was noted in the dentition of this individual, with the exception of the supernumerary tooth mentioned above.

Facio-auriculo-vertebral syndrome (or Goldenhar syndrome) can also affect the normal development of the ears, resulting in congenital aural atresia. This syndrome also affects the development of the maxilla, zygomatic arch, mandible and teeth, ranging from mild facial asymmetry to severe cranio-facial deformation. This can include skeletal changes such as cleft palate, agenesis of teeth, supernumary teeth, cleft spine and other spinal defects, microcephaly, club foot, radial hemimelia, thumb abnormalities, and various abnormalities of the internal organs (Bogusiak et al., 2017, Table. 1).

The single supernumerary tooth and the aural atresia are consistent with facio-auriculo-vertebral syndrome (D'Alessandro et al., 2006; Hoffman et al., 2019; Tasse et al., 2005). However, while this condition could not be ruled out as a cause of the aural atresia, none of the aforementioned clefting or other cranial abnormalities were found in the current individual (Martelli-Júnior et al., 2010). Further, none of the mandibles, radii, foot/ankle bones or first metacarpals recovered from the funerary context displayed any indications of facio-auriculo-vertebral syndrome.

Discussion

Patients with congenital aural atresia present with deafness on the affected side, cognitive and linguistic issues, and problems with directional hearing (Keenleyside, 2011). The physical deformity of the external ear may result in negative social outcomes in childhood (Friedman, 2009; Lipan & Eshraghi, 2011; Stanley, 2014), requiring social, financial, and emotional support networks (Ear Community, 2020). Archaeologically, ancient social attitudes towards affected individuals may be ascertained by considering pathology, demography, burial location, burial style and lifeway contextualisation (Keenleyside, 2011; Nystrom & Tilley, 2018). As it is not a life-threatening condition,

the 'Bioarchaeology of Care' concept (Tilley, 2012; Tilley & Oxenham, 2011) – used to determine the strength of social and caring networks in antiquity – would not seem to apply.

Significant deviations from standard burial practice have been argued to represent special – and often negative – status for the deceased in the eyes of the burying population (Tsaliki, 2008), yet unusual burials can also signal anthropodeic appeals, marks of honour, or gestures of affection as easily as a gesture of denigration or negativity (Gabelmann and Owens, 2020). Establishing the norm for a population and whether specific individuals fall within or outside it may thus be informative as to social mores regarding the perception of 'otherness' in that population (although the possibility of certain individuals being disposed of beyond the reach of archaeologists should also be considered). There are some signs that ancient peoples recognised – and made funeral format choices based upon – aural deformity and/or deafness.

An affected individual from prehispanic Venezuela (van Duijvenbode et al., 2015, p. 18) was interred with a pair of antlers located beside the head, a unique finding in the Venezuela/Caribbean region. Conversely, the Greek colonial case cited by Keenleyside (2011) was contextually unremarkable other than the head being turned so the affected ear was facing downwards. The prone body position of a deaf child at Romano-British Poundbury was shared by others at the site, while the grave was if anything more prestigious than many others (Roberts & Cox, 2003, p. 115-6). A series of deaf individuals buried together at the Amerindian site of Roffelsen was suggested to represent ancestral relationships (Spence et al., 2014), or as recognition of their condition.

There are no contextualised findings of ancient congenital aural atresia in the Andean area. However, contact-period sources provide Andeanists with unusual access to the social mores of the 16th and 17th centuries, although the cultural specificities (i.e. Inka) of these sources should be considered. The limited records dealing with this issue suggest a measure of negativity towards the profoundly deaf. De Molina notes that all whose "...ears were broken, and all

deformed persons" were kept away from ceremonial occasions, as the Inka believed that "they were in that state as a punishment for some fault [and] their ill luck might drive away some piece of good fortune" (De Molina, 2011 [1576], p. 21). At the very least, anyone whose ear "...orifice was broken through by any accident, the man to whom it happened was looked upon as unfortunate..." (De Molina, 2011 [1576], p. 46). Yet the profoundly deaf were not exempt from paying tribute, and seem to have been excluded from the Inka system of supporting those with severe disabilities who were unable to work (Vega, 2016, p. 354, 369, 383). We can therefore conclude that ear deformity 'mattered' to the Inka – which accords with the Inka desire for physical perfection in various of their rituals (Cobo, 1990 [1653]) – while not considering it to be sufficiently serious to merit the charity received by the more physically infirm. Ascertaining whether this policy of social exclusion applied to earlier groups is less certain, as the Ychsma – who preceded the Inka at Pachacamac – are beyond the reach of historical record, and in any case the Inka cannot be relied upon to accurately relate the social conventions of their predecessors. The current individual may be informative in this respect.

B15 (16/193) is likely to have been deaf in one ear, which would have been visibly deformed; yet this individual was interred in a prestigious Ychsma tomb in a key part of one of the Andean world's most important sites. Considering that the Inka discriminated against physical deformity both in the living (see above) and the dead (Cobo, 1990 [1653]), this key location in the Sacred Precinct is an unlikely burial spot for someone who would have been excluded from it in life. We would therefore suggest that the Ychsma did not follow these same precepts, and that the fact that they were buried in a tomb of notable (if not elite) importance suggests social acceptance and – by extension – that physical deformities did not determine social (or at least funeral) treatment in the Ychsma of this period.

Conclusion

This paper presents the first reported discovery of congenital aural atresia in Peru for over eighty years. The differential diagnoses precludes traumatic and infectious causes, and has determined a likely

congenital cause for this auricular deformity. Facio-auriculo-vertebral syndrome could not be ruled out, but seems to be unlikely owing to the absence of major cranial clefting, and while the cranium was admixed with other individuals, none of the postcrania recovered showed any signs of other pathology associated with the condition.

This is also the first Andean case of congenital aural atresia ever to be found in a contextualised funerary setting, and this has proven to be significant for examining the social attitudes of the Ychsma towards those with disabilities or deformities. Published sources from elsewhere in the world suggest that those suffering from the condition are often highlighted culturally in archaeological contexts, to either positively or negatively denote their differing status (see above). In the current case, however, the individual did not receive any such marker, and while the precise details of the interment are lacking owing to looting, the nature and location of the tomb and associated grave goods suggests that they were held in considerable regard by the burying population.

We suggest that the Ychsma therefore differ notably from the Inka of the Late Period. Non-normative burials of 'perfect' children and the social exclusion of those with physical deformities suggests a rather different social landscape among the Inka - given that the Ychsma only used non-normative burial to denote those who died in sacrificial ritual (Eeckhout & Owens, 2008), and seem to have privileged relatively elderly individuals - bearing various markers of trauma and physical degeneration - with the best-equipped burials. Children - by comparison - were marginalised and even buried en masse as grave goods with more wealthy (adult) burials (Owens and Eeckhout, 2015).

This is the first time that this approach has been utilised in analysis of the Ychsma polity; the seeming social acceptance of this individual by contemporary populations casts a positive light on how physically disadvantaged people were viewed in the Andean Late Intermediate Period. Future research should focus on refining and testing this hypothesis - using larger datasets and undisturbed contexts to examine both

pathology - and social attitudes towards it - in the ancient Andean world.

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