Craniosynostosis in skeletal material from Graubünden, Switzerland: case studies

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1. Introduction

Craniosynostosis is a condition in which some or all of the sutures in the skull of an infant close prematurely causing skull deformities and brain growth problems. Craniosynostosis can occur as an isolated defect related to certain risk factors like preterm birth, maternal drug and alcohol use or as part of a syndrome e.g. Crouzon, Apert (Kabbani & Raghuveer 2004, Shashi & Hart 2002, Lajeunie et al. 2001). Trauma of the head in the uterus may also cause premature suture closure (Lajeune et al. 2005). The prevalence of craniosynostosis is 1 per 2200 births (Kabbani & Raghuveer 2004). There are different forms of craniosynostosis depending on the sutures involved (Fig. 1-3):

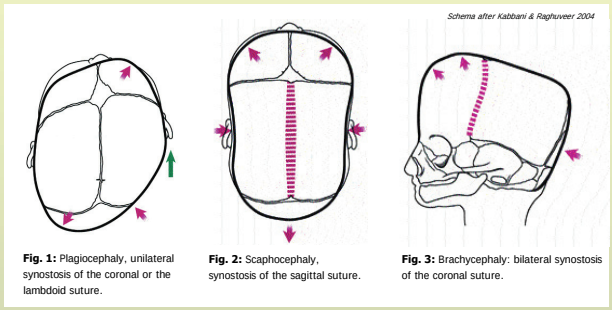


Fig. 1: Plagiocephaly, unilateral synostosis of the coronal or the lambdoid suture.

Fig. 2: Scaphocephaly, synostosis of the sagittal suture.

Fig. 3: Brachycephaly: bilateral synostosis of the coronal suture.

2. Material and methods

- Ossuary of Poschiavo, 637 crania, 17th-19th c. AD, Graubünden, Switzerland.
- Cemetery of Tomils, 404 skeletons (345 skulls), 11th-15th c. AD, Graubünden, Switzerland.
- Macroscopic and radiological examination (Univ. Clinic Balgrist, Zurich, Switzerland).

3. Results and discussion

Provenance	Cranium no.	Age[1]	Sex[1]	Closed suture	Type
Poschiavo, Switzerland, 17th-19th c. AD	18	60+	M	right coronal	plagiocephaly
	131	40-50	M	right coronal	plagiocephaly
	190	40-50	I	bicoronal	brachycephaly
	245	20-25	F	right coronal	plagiocephaly
	380	5-7	I	sagittal, squamosal, occipitomastoid	*multiple*
	443	20-25	F	left lamboid	plagiocephaly
	556	20-25	F	sagittal	scaphocephaly
Tomils, Switzerland, 11th-15th c. AD	473	25-30	M	left coronal	plagiocephaly

[1] Determination of age and sex after Ferembach et al. 1979; for age determination the pathologic sutures were excluded; (M=male, F=female, I=indeterminate).

Table 1: Individuals with craniosynostosis found in the ossuary of Poschiavo and the medieval cemetery of Tomils. Seven individuals (1.1%) from Poschiavo and one individual (0.3%) from Tomils exhibit craniosynostosis.

• **Plagiocephaly** was the most common type, observed in five cases. In modern populations among all types, plagiocephaly is found in 20-30% (Cohen 2005). Extremely rare is the case of lambdoid synostosis present in 3 out of 100.000 births (Kabbani & Raghuveer 2004). No differences in the frequency are reported between males and females (Cohen 2005).

• **Brachycephaly** was observed in one case. Bilateral coronal synostosis is commonly linked to a syndrome e.g. Apert, Crouzon, Pfeiffer, Bear-Stevenson and Jackson-Weiss (Blaser 2008).

• **Scaphocephaly** is found in Poschiavo in one female individual; according to clinical data sagittal synostosis is the most common type found in 40-60% and is more frequent in males than in females (Cohen 2005).

• **Multiple synostosis** was found in a child showing premature closure of the sagittal and of both occipitomastoidea and squamosal sutures. The closure of the squamosal suture is very rare (Duncan & Stojankowski 2008); experimental studies in mice link the condition to intrauterine constraint by artificially delaying birth (Moffet & Moffet 1989). Multiple suture closure is usually linked to a syndrome (Cohen 2005).

Differential diagnosis: deformational plagiocephaly (positional lambdoid flattening) and torticollis (tilted head) may also cause skull deformities, but in these cases no true suture closure is involved.

Suture closed	Location and references
Sagittal	Andes, 6.000 BC (Gerszten et al. 1998); Pakistan, 2.600-1.900 BC (Kennedy et al. 1993); Southwest USA (Bennett 1967); Illinois, USA (Kohn et al. 1994); Anatomic collection Museum Vrolik, Netherlands (Oostra et al. 2005); Eastern USA (Reichs 1989); Kansas, USA, ca. 1300 AD (Eisley & Aisling 1944); Egypt, Roman Period (Wells 1964); Münsingen-Rain, Switzerland, 475-250 BC (Kutterer & Alt 2008).
Coronal	Nebraska, USA 1780-1800 AD (Pederson & Anton 1998); Europe (Kreiborg & Bjork 1981); Anatomic collection, Museum Vrolik, Netherlands (Oostra et al. 2005).
Sphenofrontal	Switzerland, Münsingen-Rain, 475-250 BC (Kutterer & Alt 2008).
Lamboid	Spain, Middle Pleistocene 500.000-350.000 (Gracia et al. 2009).
Multiple sutures	Anatomic collection Museum Vrolik, Netherlands (Oostra et al. 2005); Georgia, USA, 16th century AD (Duncan & Stojankowski 2008).

Table 2: Types of craniosynostosis found in archaeological populations.

Archaeological record: a review of the literature (Table 2) shows the presence of the condition in archaeological populations, scaphocephaly and plagiocephaly are again the types most frequently reported, whereas multiple sutures closure is rare.

Fig. 4: Right coronal synostosis present at an adult male individual from Poschiavo (#18). The arrows point to the fused suture. The individual demonstrates asymmetry of the forehead with flattening of the right frontal bone, prominence of the left orbit. The intersection with the right coronal and the sagittal suture is deviated to the left; superior view.

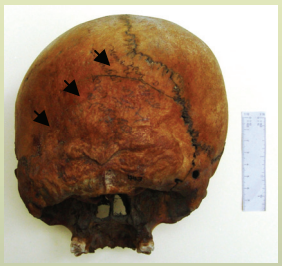
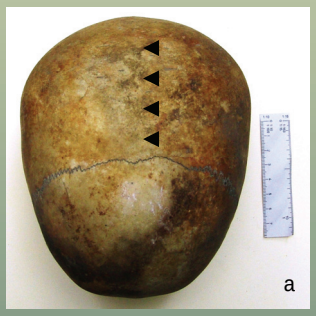


Fig. 5: Left lambdoid synostosis present at an adult female individual from Poschiavo (#443). The arrows point to the fused suture. The individual demonstrates flattening of the right occipital side, slight prominence of the right (contralateral) forehead; posterior view.



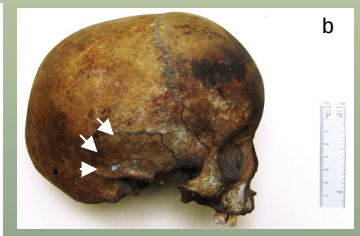


Fig. 6a, b: Sagittal synostosis (black arrows) and partial fusion of the squamous (white arrows) and the occipitomastoid sutures at a 5-7 years old child from Poschiavo (#380). The arrows point to the fused sutures. The individual demonstrates elongated skull shape associated with narrow biparietal diameter and frontal prominence; (a) superior view, (b) lateral view.

Fig. 7: Bilateral coronal synostosis present at an adult individual from Poschiavo (#190). The arrows point to the fused suture. The individual demonstrates short cranium, bilateral superior orbital rim retrusion and narrow skull base; lateral view.

4. Main outlook

• High incidence of craniosynostosis and especially of some rare types was found in historic populations from Canton Graubünden. The aetiology of the condition is prohibited by the lack of post-cranial skeletal material and soft tissues. In some cases the multiple suture closure may be linked to specific syndromes. This may highlight the high inbreeding rate observed in these regions (Wettstein 1902, Hägler 1941).

• Regarding the rarity of the condition according to clinical data and the few reports on the bioarchaeological record the presentation of such cases offers a great insight into the biological history of the alpine regions.

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*(All references related to **Table 2** on request from the author: christina.papageorgopoulou.ch)*

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Acknowledgements

The authors would like to thank the Archaeological Service of Canton Graubünden (Chur, Switzerland) and especially the director Urs Clavadetscher for the financial support as well as the University Clinic Balgrist (Zurich, Switzerland).