

# OCCIPITO-CERVICAL SYNOSTOSIS: ITS OCCURANCE AND EMBRYOLOGICAL BASIS

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## ABSTRACT

**Introduction:** Occipito-cervical synostosis is a rare anomaly. It is also known as assimilation or occipitalization of atlas. It is defined as a congenital fusion of atlas vertebra with the base of occipital bone, it may be complete or partial. This anomaly results due to failure in segmentation and separation of last occipital sclerotome and first cervical sclerotome during development of foetus.

**Material and Methods:** The present study was conducted on 240 skull bones in the department of Anatomy King George Medical University, Lucknow, India. We observed the synostosis of atlas vertebrae with basilar part of occipital bones.

**Results:** Among 240 bones we noticed 7 craniums which were fused with the first cervical vertebra and the details of these are described in following article.

**Conclusion:** This type of anomaly may alter the dimensions of foramen magnum and may compress the spinal cord and vertebral artery. Therefore, improved knowledge on the fusion of atlas with the occipital bone is important for orthopedicians, neurosurgeons, radiologist, anaesthetist, physiotherapist and anatomist, as it shows multiple variations and combinations.

**KEY WORDS:** Atlas, occipital bone, synostosis, variation, sclerotome .

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## INTRODUCTION

The occipital bone forms the back and base of cranium and its inferior surface having two occipital condyles. Atlas is first cervical vertebra which supports the skull, so also called atlas (after the mythical Greek God who supported the globe) [1]. Atlas vertebra is ring shaped, without a body having an anterior arch, posterior arch and two lateral masses. Its superior articular facets articulate with the occipital condyles to form atlanto-occipital joint which is a synovial joint of ellipsoid variety hav-

ing movements of flexion, extension and slight lateral rotation [2]. The anterior arch articulates with dens of the axis to form pivot type synovial joint. The craniovertebral anomalies are common and are often seen in radiological studies, occipito-cervical synostosis is rare congenital anomaly seen at craniovertebral region [3]. It is defined as congenital fusion of atlas to the base of the occiput. The other terms for the same are occipitalization of atlas, assimilation of atlas, atlanto-occipital synostosis and atlantooccipital fusion. Motabagani & Surendra 2006 [4] quoted

that it was first described by Rokitansky in 1884 and first radiological report was published in 1911 by Suchuller. Its incidence is 1 in 109 adult skulls of Asian population. The fusion may be complete or incomplete.

## MATERIALS AND METHODS

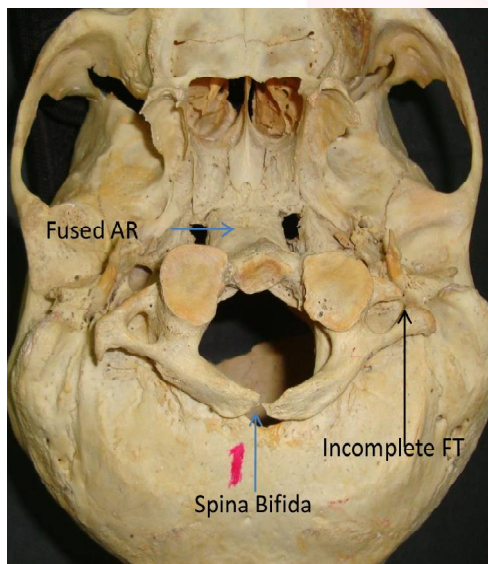
The present study was conducted on 240 skull bones in the department of Anatomy King George's Medical University, Lucknow, India in the period of 2014-2016. We observed the synostosis of atlas vertebra with basilar part of occipital bone. The bones were studied carefully and photograph was taken from the base of the cranium to see the fusion properly. Measurement of foramen magnum and inferior articular facets were done by Vernier calliper.

## RESULTS

Among 240 bones we found 7 skull bones with partial and asymmetrical synostosis of atlas vertebrae. The dimensions are depicted in Table 1 and the following observations were noted:-

### In skull 1(Fig 1)

**Fig. 1:** Skull 1: Showing Fused Anterior Arch(AR), Spina Bifida(SB)& incomplete Foramen transversarium(FT) on left side.

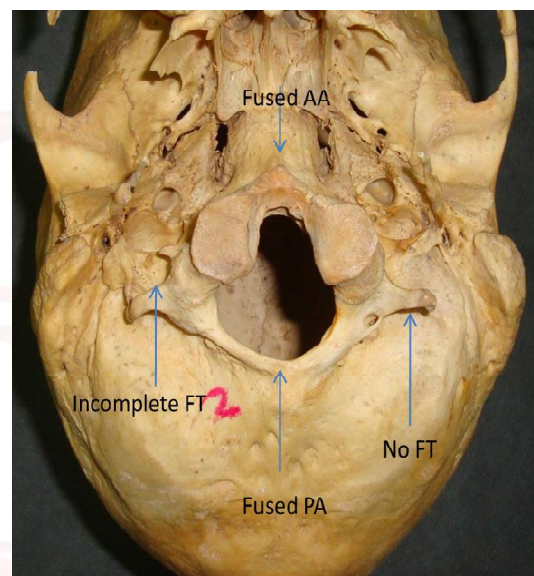


1. The fusion is not exactly in midline but slightly inclined to left side.
2. Anterior arch is completely fused with the anterior margin of foramen magnum.
3. The right half of posterior arch is fused leaving a gap between the basilar parts of occipital bone. Left half of posterior arch is not fused with posterior margin of foramen magnum.

4. Spina bifida, means each half of posterior arch of atlas have not fused with each other.
5. Left foramen transversarium is not complete, deficient anterolaterally and Right foramen transversarium is complete.
6. Both foramen transversarium are free.
7. Superior articular facets of atlas have completely fused with condylar facets of occipital bone.

### In skull 2(Fig 2)

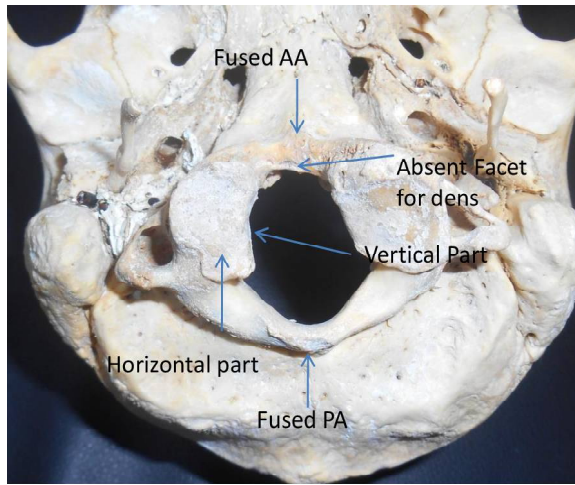
**Fig. 2:** Skull 2: Showing Fused Anterior Arch(AR), Fused posterior arch(PA)& incomplete Foramen transversarium(FT) on right side & no FT at left side.



1. The fusion is not exactly in midline but slightly inclined to right side.
2. Fusion of anterior arch of atlas with basilar part of occipital bone leaving a foramen which is situated left of midline.
3. Posterior arch is completely fused with posterior margin of foramen magnum.
4. Lateral masses and posterior arch are fused leaving 2 gaps which is situated below and behind the right and left lateral masses.
5. The left transverse processes doesn't bear foramen transversarium & right had incomplete foramen transversarium.
6. Left transverse process is free and right is attached with the jugular process of occipital bone via a spicule.
7. Superior articular facets of atlas have completely fused with condylar facets of occipital bone.

### In skull 3(Fig 3)

**Fig. 3:** Skull 3: Showing Fused Anterior Arch(AR), Fused posterior arch(PA), Absent dens facet & vertical & horizontal part of right inferior articular facet.



1. The fusion is in midline.
2. The anterior and posterior arches are completely fused with the anterior and posterior margins of foramen magnum.
3. A canal was observed bilaterally between atlas and occipital bone just behind the fused lateral masses which most probably gave passage to vertebral artery.
4. Both transverse processes appeared to be normal.
5. On left side, transverse process was free from the base of the cranium but on right side, a spicule of bone connected the costotransverse bar with jugular process due to which a foramen was created that most probably gave exit to first cervical nerve.
6. Superior articular facets of atlas were completely fused with occipital condyles.
7. There was absence of articular facet for dens on the anterior arch of atlas.
8. The right and left inferior articular facets of atlas were irregular in shape and the right facet was divided into two parts i.e. horizontal part and vertical part.

### In skull 4(Fig 4)

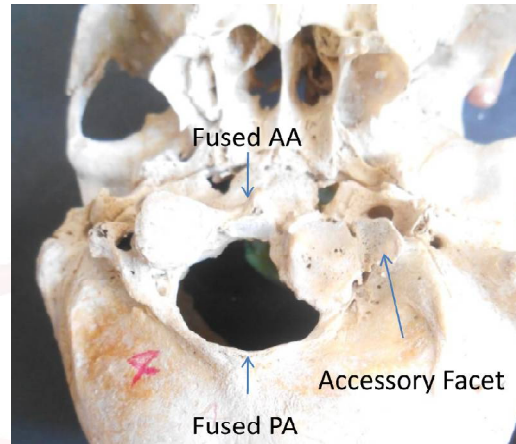
1. The fusion is not exactly in midline but slightly inclined to right side.
2. Anterior arch is completely fused with the anterior margin of foramen magnum.
3. Posterior arch is completely fused with the posterior margin of foramen magnum.

4. Right transverse process bear foramen transversarium but left doesn't have any foramen transversarium.

5. The left inferior articular facet is facing medially towards foramen magnum.

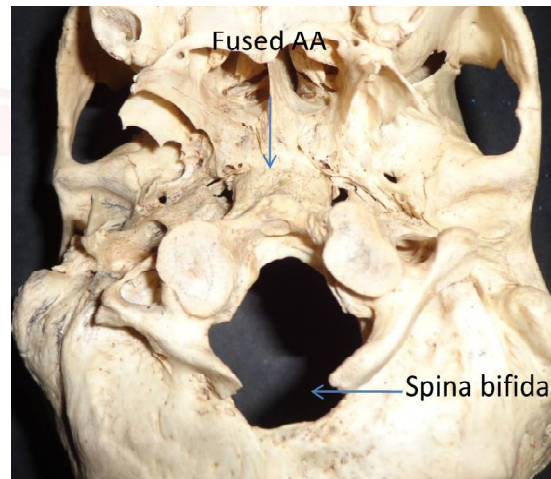
6. An accessory facet is also present just lateral to left inferior articular facet.

**Fig. 4:** Skull 4: Showing Fused Anterior Arch(AR), Fused posterior arch(PA), No Foramen transversarium on left side & Accessory facet.



### In skull 5(Fig 5)

**Fig. 5:** Skull 5: Showing Fused Anterior Arch(AR), Spina Bifida(SB) & complete Foramen transversarium(FT) on both side.

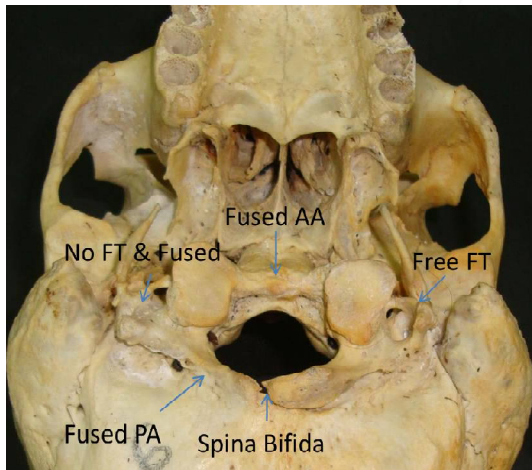


1. The fusion is inclined to left side.
2. The anterior arch is fused with the anterior margin of foramen magnum leaving a slit like gap.
3. Spina bifida is present.
4. Left 1/4<sup>th</sup> of the posterior arch is completely fused and right 1/4<sup>th</sup> is partially fused leaving a gap.
5. Foramen transversarium is present on both side but left foramen transversarium is larger as compared to right side.

6. Superior articular facets of atlas have completely fused with condylar facets of occipital bone.

#### In skull 6(Fig 6)

**Fig. 6:** Skull 6: Showing Fused Anterior Arch(AR), Fused posterior arch(PA), Spina Bifida, No Foramen transversarium(FT) on Right side(fused with occiput) & on left side complete FT which is not fused with occiput.

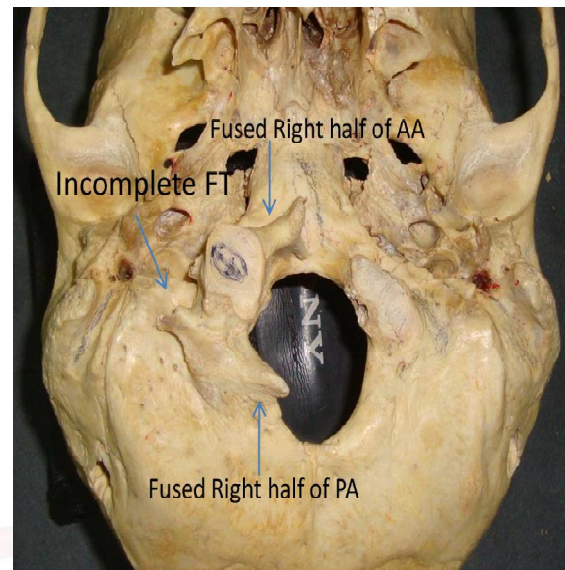


1. The fusion is inclined to right side.
2. Anterior arch is fused with the anterior margin of foramen magnum, leaving 2 small foramina between them which is situated to 3 and 4 mm left and right from anterior tubercle of atlas respectively.
3. The right half of posterior arch was completely fused whereas left half incompletely fused leaving a small gap.
4. Spina bifida of posterior arch of atlas.
5. Right transverse process doesn't bear foramen transversarium and fused with the basilar part of occipital bone.
6. Left transverse process is free and bears foramen transversarium.
7. Superior articular facets of atlas have completely fused with condylar facets of occipital bone.

#### In skull 7(Fig 7)

1. In this skull fusion was incomplete.
2. Right half of the atlas is completely fused with right half of anterior arch, right lateral mass and right half of posterior arch.
3. Right superior articular facet of atlas vertebra has completely fused with the right condylar facet of occipital bone.
4. Left half of atlas is not present.

**Fig. 7:** Skull 7: Showing Fused Right half of Anterior Arch(AR), Fused right half of posterior arch(PA), incomplete Foramen transversarium(FT) on right side & no fusion of atlas on left side.



**Table 1:** Comparison of diameters of foramen magnum, inferior articular facet, breadth and length of 7 skulls.

Parameters (mm)	Skull 1	Skull 2	Skull 3	Skull 4	Skull 5	Skull 6	Skull 7
Anteroposterior diameter of foramen magnum	30.21	29.11	26.32	27.18	30.11	12.73	15.24
Transverse diameter of foramen magnum	28.63	28.24	21.2	25.28	28.34	12.46	15.22
Anteroposterior diameter of right inferior articular facet	18.84	17.98	18.24	15.12	20.01	14.96	12.32
Transverse diameter of right inferior articular facet	16.12	11.35	12.12	13.18	13.18	10.99	8.4
Anteroposterior diameter of left inferior articular facet	18.84	17.41	22.18	18.24	21.34	14.97	-
Transverse diameter of left inferior articular facet	15.09	12.75	18.34	15.32	16.21	14.4	-
Maximum breadth of skull	122.68	126.47	127.21	124.34	128.3	126.7	112.5
Maximum length of skull	148.69	173.65	176.28	172.12	178.19	166.14	170.4

## DISCUSSION

At the time of development, the ventro-medial portion of somite forms sclerotome which surrounds the notochord and develops into vertebral body. The dorsal portion surrounds the neural tube and develops into posterior vertebral arch. Then the caudal half of each

sclerotome combine with the cranial half of the sclerotome below it. The cranial half of first cervical sclerotome combine with the caudal half of last occipital sclerotome to form the base of skull. While the caudal half of first cervical sclerotome combine with the cranial half of second cervical sclerotome to form the first cervical vertebra. The pattern continues in this way to form rest of the vertebra. The dens of second cervical vertebra is formed by the body of first cervical vertebra, thus the first cervical vertebra doesn't have body [5].

Jayanti et al. (2003) [6] have reported two cases of synostosis with spina bifida of atlas. In 1<sup>st</sup> case there was a complete fusion of only one of the transverse process with occipital bone & incomplete fusion of anterior arch of atlas. In 2<sup>nd</sup> one the anterior arch has fused. Nayak S et al (2005) [7] also reported about the fusion of atlas vertebra with the occipital bone, the two superior facets of atlas had completely fused with occipital condyles. The anterior and posterior arches had an incomplete fusion with the basilar part. Vandana et al (2013) [8] found two skulls of incomplete assimilation. Ranade et al (2007) [9] have examined 98 Indian human skulls for assimilation of atlas & noted two cases showing various degree of assimilation of atlas. Sani et al (2009) [10] have observed assimilation of atlas in 2 Indian skulls. In present study the finding of 1<sup>st</sup> & 5<sup>th</sup> skulls is similar to the finding of Jayanti's study & 2<sup>nd</sup>, 6<sup>th</sup> & 4<sup>th</sup> is similar to Vandana's study. The 3<sup>rd</sup> skull contains the horizontal & vertical part of right inferior articular facet & absent facet for dens. After observing this we can say the dens may be articulate with the vertical part of inferior articular facet which will leads to torticollis.

The synostosis of skull & atlas vertebra is most probably arise as a result of failure of segmentation and separation of most caudal occipital sclerotome and first cervical sclerotome during initial weeks of foetal life [11]. This variation may often be unnoticed, but incidentally found during radiological and operative procedure or during autopsy [12]. The onset of clinical symptoms can be sudden or may be precipitated by minor trauma and sudden death has also been reported [13]. It may produce narrowing of foramen magnum and spinal canal which may

compress spinal cord or brain stem. The standard dimensions of foramen magnum ranges between 28-38 mm for sagittal and 25-40 mm for transverse diameter [4] and the spinal cord compression occurs when sagittal diameter is less than 18 mm [14]. It can produce signs and symptoms which vary from simple headache to full blown neurological syndrome i.e. neck pain, occipital headache, numbness, abnormal head posture and convulsion. The vertebral artery and first cervical nerve are related with superior aspect of atlas, so there may be chances of their compression which can cause compromise blood flow to the brain leading to dizziness, seizure, syncope and neurological symptoms [6]. If cranial nerve is involved, there may be tinnitus and visual disturbance. Clinically, wide variety of other signs and symptoms may be associated like restricted or absent movement, ataxia, muscular spasticity and torticollis. Some associated malformations may commonly occur with occipito-cervical synostosis like fusion of C1 & C2, absence or malformation of transverse ligament, hyperplasia or aplasia of dens and anomalies of vertebral artery. The findings of the 3<sup>rd</sup> skull suggest that patient might have left torticollis with restricted neck movement and absent or malformed transverse ligament.

Gholve et al (2007) [15] classified the occipito-cervical synostosis into 4 types based on the zone of atlas that fused with occiput i.e.:

Zone -1 = Fusion of anterior arch

Zone -2 = Fusion of lateral masses

Zone -3 = Fusion of posterior arch

Zone -4 = Combination of zones

According to this classification, 7 skulls of the present study can be categorised into Zone 4.

## CONCLUSION

Occipito-cervical synostosis is a congenital anomaly & partial synostosis is more common. This anomaly should be checked in time especially by neurosurgeons and orthopaedic surgeons because this can cause severe neurological symptoms, convulsion, seizure, severe neck pain and even sudden death. As the transverse process is very important landmark for surgeons, the knowledge of fusion may be

important for head and neck surgeries. It may cause failure of cisternal puncture, therefore also important for anaesthetist. Physiotherapist dealing with neck pain and radiologist dealing with abnormalities of cervical spine, must also be aware of this condition. Because of the serious consequences of this type of osseous anomaly there is a need of thorough clinical assessment and evaluation of every patient.

**Conflicts of Interests: None**

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