

## Clinical Article

# The Incidence and Clinical Implications of Congenital Defects of Atlantal Arch

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**Objective :** Atlantal arch defects are rare. Few cadaveric and imaging studies have been reported on the variations of such anomalies. Our goal in this study was to examine the incidence and review the clinical implications of this anomaly.

**Methods :** A retrospective review of 1,153 neck or cervical spine computed tomography (CT) scans was performed to identify patients with atlantal arch defects. Neck CT scans were performed in 650 patients and cervical spine CT scans were performed in 503 patients. Posterior arch defects of the atlas were grouped in accordance with the classification of Currarino et al. In patients exhibiting this anomaly, special attention was given to defining associated anomalies and neurological findings.

**Results :** Atlantal arch defects were found in 11 (11/1153, 0.95%) of the 1,153 patients. The type A posterior arch defect was found in nine patients and the type B posterior arch defect was found in two patients. No type C, D, or E defects were observed. One patient with a type A posterior arch defect had an anterior atlantal-arch midline cleft (1/1153, 0.087%). Associated cervical spine anomalies observed included one C6-7 fusion and two atlantal assimilations. None of the reviewed patients had neurological deficits because of atlantal arch anomalies.

**Conclusion :** Most congenital anomalies of the atlantal arch are found incidentally during investigation of neck mass, neck pain, radiculopathy, and after trauma.

**KEY WORDS :** Arch · Atlas · Incidence · Congenital defect.

## INTRODUCTION

Congenital defects of the arch of the atlas, a developmental failure of chondrogenesis, are a rare anomaly. These defects are considered by some to be a benign variation; and indeed, almost all of them are discovered incidentally. However, detection of these anomalies is clinically important because they can cause acute neurologic deficits, which are closely associated with neck extension. Currarino et al.<sup>8)</sup> and Geipel<sup>16)</sup> reported the incidence of atlantal arch defects by a cervical radiographic study and a cadaveric study, respectively. Additionally, Senoglu et al.<sup>29)</sup> studied the anatomical, clinical, and imaging features as well as incidence of congenital defects of the atlas by evaluation of cervical spine computed tomography (CT) scans and cadaveric dissections.

In Korea, no cadaveric studies have been reported on the variations of such atlas arch defects. Only four case studies with atlantal arch defects have been reported<sup>1,18,19,24)</sup>. We therefore investigated the incidence of these congenital defects in cervical spine or neck CT studies. Imaging features of these defects and their clinical significances for neurosurgical practice were the focus of this study.

## MATERIALS AND METHODS

To evaluate the incidence of congenital defects of the atlas arch, we reviewed the institutional database and retrospectively evaluated consecutive neck (3 mm interval image from lower occiput to second thoracic vertebra) and cervical spine (2 mm interval image from lower occiput to second thoracic vertebra) CT scans. The number of patients evaluated was 1,153. These patients presented to our institution between January 2002 and May 2009. Two different models of CT scanner were used. The Siemens Somatom CT scanner (Siemens Somatom Volume Zoom 4 Channel; Siemens Medical Solutions, Germany) was used during the

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**Table 1.** Clinical characteristics of 11 patients with atlantal arch defects

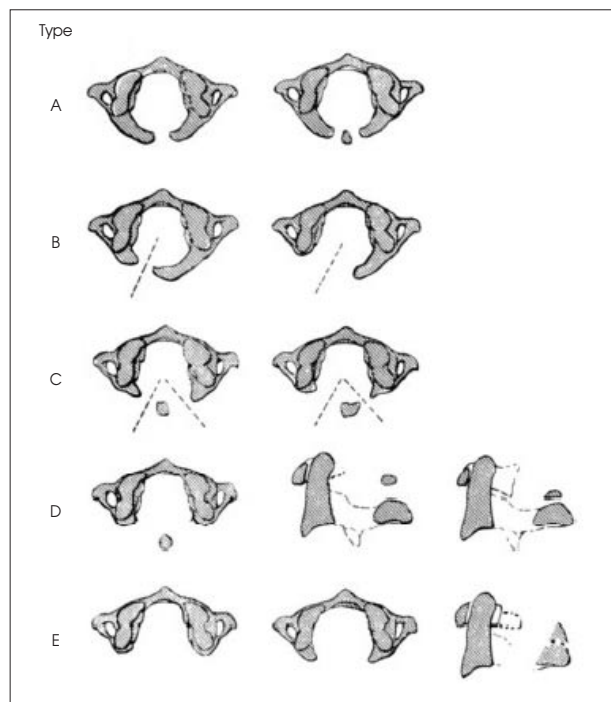
Patient	Sex	Age	Type of arch defect <sup>a</sup>	Associated anomaly	Symptoms	Diagnosis	Type of CT
1	M	34	A	No	Headache, neck pain because of falling	Cervical sprain	Toshiba c-spine CT
2	M	42	A	No	Quadriplegia because of TA	C7 fracture, spine epidural hematoma	Siemens c-spine CT
3	F	59	A	Anterior atlantal arch defect, C6-7 fusion	Neck pain, arm pain	Facet joint syndrome	Siemens c-spine CT
4	F	29	A	No	Palpable neck mass	Thyroid mass	Toshiba neck CT
5	M	23	A	No	Sore throat	Acute tonsillitis	Siemens neck CT
6	M	58	A	Atlantal assimilation	Palpable neck mass	Thyroid cancer	Siemens neck CT
7	F	32	A	Atlantal assimilation	Confused mentality because of TA	Traumatic SAH, DAI	Toshiba c-spine CT
8	M	35	A	No	Neck pain because of falling	Cervical sprain	Siemens neck CT
9	F	42	A	No	Neck pain because of trauma	Cervical sprain	Toshiba c-spine CT
10	M	55	B	No	Confused mentality because of falling	SDH, C5-6 fracture and dislocation	Siemens c-spine CT
11	M	30	B	No	Neck pain because of TA	Cervical sprain	Toshiba c-spine CT

Types of arch defects were grouped in accordance with the classification of Currarino et al.<sup>8)</sup>. CT : computed tomography, DAI : diffuse axonal injury, SAH : subarachnoid hemorrhage, SDH : subdural hematoma, TA : traffic accident

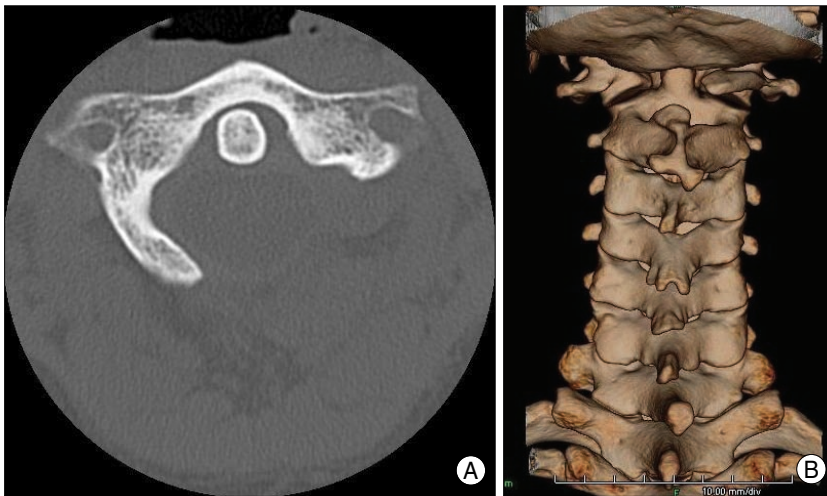
early period (January 2002 to April 2007) and the Toshiba CT scanner (Toshiba Aquilion TSX-101A 64 Channel; Toshiba Medical Systems, Tokyo, Japan) was used in the later period (May 2007 to May 2009). The Siemens Somatom CT scanner was used to perform 431 neck CT scans and 353 cervical spine CT scans. The Toshiba CT scanner was used to perform 219 neck CT scans and 150 cervical spine CT scans. The patients presented with various medical problems, including weakness, palpable neck mass, posterior neck pain, radiculopathy due to degenerative disease, arm pain, sore throat, posterior neck pain or radiculopathy after traffic accident. When a congenital defect of the atlas arch was identified on a CT scan, the patient's medical record was reviewed to determine his or her neurological status. An associated anomaly was searched by retrospective review of X-ray and CT findings. Posterior arch defects of the atlas were grouped in accordance with the classification of Currarino et al.<sup>8)</sup> (Fig. 1).

## RESULTS

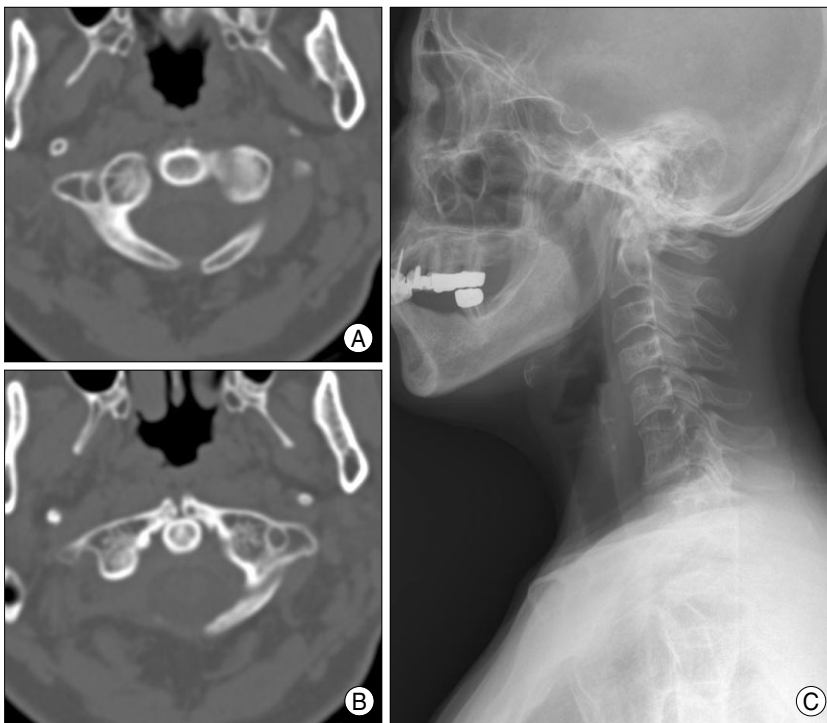
Overall, 11 patients (11/1153, 0.95%) with atlas arch anomalies were found in the 1,153 patients evaluated (650 neck CT scans, 503 cervical CT scans). The clinical characteristics of patients with congenital defects of the atlas arch are described in Table 1. Nine of these patients (9/1153, 0.78%) had a type A posterior arch defect and two patients (2/1153, 0.17%) had a type B posterior defect (Fig. 2). No type C, D, or E posterior arch defects were found. An anterior arch cleft was observed in only one patient with a



**Fig. 1.** Classification of posterior arch defects of the atlas. A : Failure of posterior midline fusion of the two hemiarches. Sometimes a small separate ossicle is seen within the gap. B : Unilateral cleft, ranging from a small gap to a complete absence of the hemiarch and posterior tubercle. C : Bilateral clefts of the lateral aspects of the arches with preservation of the most dorsal part of the arch. D : Complete absence of the posterior arch with a persistent isolated tubercle. This anomaly is conceivably a more extensive form of bilateral clefts. The lateral parts of the posterior arch are absent except for the posterior tubercle. E : Absence of the entire posterior arch including the tubercle. Reproduced with permission from Currarino G, Rollins N, Diehl JT : Congenital defects of the posterior arch of the atlas : a report of seven cases including an affected mother and son. AJNR Am J Neuroradiol 15 : 249-254, 1994<sup>8)</sup>.



**Fig. 2.** Computed tomography (CT) findings of type B defect (Patient 11). A : Axial CT scan of the atlas showing a congenital defect of the left posterior hemiarch and posterior tubercle. B : A three-dimensional reconstruction of CT demonstrating a congenital defect of the left posterior hemiarch and posterior tubercle.



**Fig. 3.** Radiological findings of type A defect with anterior atlantal arch defect (Patient 3). A : An axial computed tomography (CT) scan of the atlas revealing a posterior arch cleft. B : An axial CT scan of the atlas demonstrating an anterior arch cleft. C : A lateral cervical spine film showing fusion of the C<sub>6-7</sub> vertebral bodies.

type A posterior arch defect (1/1153, 0.087%). One patient (patient 3) who had a type A defect had a congenital cleft of the anterior arch of the atlas and a C<sub>6-7</sub> congenital fusion (Fig. 3). Two patients (patient 6 and 7) with type A posterior arch anomalies also had atlantal assimilation.

Based on their medical records, the 11 patients had no neurological deficits because of atlas arch defects.

dysgenesis, Klippel-Feil syndrome, Arnold-Chiari malformations, and Turner and Down syndrome<sup>28</sup>). In this study, atlantal arch defects were associated with C<sub>6-7</sub> congenital fusion in one patient and assimilation of the atlas in two patients.

#### Incidence of atlantal arch defects

The older classification of congenital clefts and defects of

## DISCUSSION

### Development of atlantal arch defects

Normally, three primary ossification centers of the atlas appear during the embryonic period. Although the anterior arch of the atlas is usually cartilaginous at birth, 20% of newborns have an ossification center at that location. The anterior center of the anterior tubercle and the anterior arch usually unite with the two lateral centers at 5-9 years of age. An anterior atlantal arch defect may occur in the absence of an anterior ossification center, which results in the lateral masses not fusing anteriorly or no fusion occurring between the two anterior ossification centers<sup>5</sup>.

At birth, the posterior arches of the atlas are nearly fused except for several millimeters of cartilage. The two centers of the lateral masses normally unite posteriorly by perichondral growth, giving rise to the posterior arch at 3-5 years of age<sup>20</sup>. Only rarely does a fourth ossification center appear, which results in the posterior tubercle of the atlas and unites with the lateral masses of the atlas<sup>4</sup>. Posterior atlantal arch defects are attributed to the defective or absent development of the cartilaginous preformation of the arch rather than a disturbance of the ossification<sup>16</sup>. This is supported by findings at autopsy or intra-operatively that connective tissue bridges the bony defect<sup>26</sup>.

Congenital anomalies of the atlantal arch frequently occur with various combinations of neural abnormalities, suggesting an interrelationship exists<sup>21</sup>. Common associations include gonadal

the posterior arch of the atlas has been recently revised<sup>8)</sup>. Depending on the extent to which these lateral masses fail to develop, the defect can be classified into five types: type A - failure of midline fusion of the two hemiarches, i.e., failure of the two lateral centers to unite posteriorly in the midline; type B - unilateral cleft ranging from a small defect to the complete absence of one hemiarch and posterior tubercle; type C - bilateral clefts of the lateral aspects with preservation of the most dorsal part of the arch; type D - absence of the posterior arch with a persistent posterior tubercle; and type E - absence of the entire arch including the tubercle (Fig. 1). Type A defects occur in 4% of the general population<sup>15)</sup>, whereas the other types (B-E) occur in 0.69% of the general population<sup>8)</sup>.

Geipel<sup>16)</sup> found clefts of the posterior arch in 4% of 1,613 cadaveric dissections, 97% of which were median clefts. Currarino et al.<sup>8)</sup> evaluated 1,440 lateral cervical radiographs and found that over 90% of defects were type A. They estimated that 0.69% of the general population harbor type B-E defects of the posterior arch of the atlas. Senoglu et al.<sup>29)</sup> reported that 3.35% (37/1354) of 1,354 CT scans and cadaveric studies had congenital defects of the posterior arch of the atlas.

An anterior cleft is observed in about 0.09-0.1% of the population<sup>14,22,29,34)</sup>. In most patients, the anterior cleft is associated with the anomalous development of the posterior arch<sup>12)</sup>. The rarest abnormality is an anterior defect alone, which has been reported in only a few patients<sup>32)</sup>. The association of anterior and posterior arch defects, called a bipartite atlas, has also been described<sup>6,9,11,17,23,25)</sup>. Clefts of the anterior arch are nearly always very narrow, and may be median or paramedian<sup>6)</sup>.

To the best of our knowledge, four patients with atlantal arch defects have been reported in Korea. Two patients with type A defects, one with a type B defect, and one with a type E defect have been reported<sup>1,18,19,24)</sup>. Of the two patients with type A defects, one patient had a bony inturning of the type A cleft presented with transient quadriplegia and the other patient had a type A defect associated with an anterior atlantal arch defect<sup>1,24)</sup>. In this study, the incidence of posterior atlantal arch defects was 0.95% (11/1153) and the incidence of anterior atlantal arch defects was 0.087% (1/1153). In our study, the incidence of posterior arch defects was lower than that of previous studies. However, the incidence of anterior arch defects of the atlas was similar to that of other studies. Because the cutting interval of images in the cervical spine and neck CT scans were 2 or 3 mm, most arch defects were identified in this study. Although the patients in this study do not represent the general Korean population, the results indicate that the

incidence of posterior arch defects in this study was lower than that of other countries.

### Clinical implications of atlantal posterior arch defects

Overall, atlantal posterior arch defects are considered benign anatomical variations. Indeed, in all our patients, the anomalies were found incidentally. And, our patients showed no cervical instability on radiograph. Although rare, acute neurological symptoms or deficits and atlanto-axial instability have been associated with these defects<sup>20,34)</sup>. The neurological symptoms described in the literature consistently include weakness in the four limbs<sup>26,30,33)</sup>. Sensory symptoms such as paresthesia in the four limbs, in the upper limbs only, or in the ipsilateral upper and lower limbs have also been described.

The presence of a posterior tubercle remnant (type C and D) is likely to cause transient quadriplegia after cervical trauma<sup>13,33)</sup>. In the classification of Currarino et al.<sup>8)</sup>, defect types C and D, manifesting as absence of the bilateral posterior arches and an isolated posterior tubercle, are clinically very important because these anomalies often cause acute neurologic deficits such as transient quadriplegia, paraparesis, Lhermitte's sign, chronic neck pain, and headache<sup>27)</sup>. It also appears that the bony inturning of the type A cleft defect may become symptomatic at an earlier stage without exacerbating factors such as chronic degenerative changes, atlanto-axial instability, or trauma<sup>1,7,10)</sup>. It is postulated that the bony expansion of the inturned posterior hemiarches results from traction of ligamentous and muscular structures that normally insert on the posterior tubercle of the atlas, or from abnormal stresses arising from other bony fusions<sup>7)</sup>.

Surgery is the treatment of choice in symptomatic compression. Excision of the posterior arch is curative. Once symptomatic, it is probably best treated as early as possible because a trivial trauma, such as a fall when walking, can precipitate severe neurologic deficits and even respiratory distress<sup>10)</sup>.

### Clinical implications of atlantal anterior arch defects

Atlas anterior arch clefts are usually incidental findings discovered with routine cervical radiography. There is rarely complete aplasia of the anterior arch, which is associated with craniocervical instability<sup>3)</sup>. However, because cervical radiography is often performed following trauma, careful differentiation between an acute burst fracture and a congenital defect is essential. CT is most helpful in evaluating the integrity of the atlas ring and differentiating acute injury from a developmental cleft. A CT scan of an atlas

arch defect can demonstrate a small defect in the arch with smooth corticated margins<sup>6</sup>.

A so-called split atlas is a very rare congenital malformation resulting in the nonfusion of anterior and posterior arches of the atlas. This unusual variant is usually an incidental finding, which is difficult to diagnose on plain radiographs. However, some split atlas can be associated with asymptomatic lateral atlantoaxial subluxation<sup>24</sup>. In our study, one patient (patient 3) with a type A defect also had a congenital cleft of the anterior arch of the atlas (Fig. 3). It is, however, extremely important to recognize a split atlas in case of cervical trauma<sup>2</sup>. A congenital anomaly of the atlas in which the anterior and posterior arches are not fused can create an image on X-rays similar to that of a Jefferson fracture. There are differences in these two conditions that enable us to distinguish one from the other. Generally, the lateral translation of the lateral masses with a congenital anomaly is 1 to 2 mm, and with a Jefferson fracture they are more than 3 mm<sup>6</sup>. In a young population (3 months to 4 years old), the lateral masses of the atlas commonly extend 1-3 mm beyond the margins of the axis, secondary to different growth patterns of the vertebral bodies. This is called pseudospread of the atlas<sup>31</sup>. The classic radiographic feature of a Jefferson fracture is a bilateral atlanto-axial lateral offset of 3-9 mm<sup>17</sup>. A lateral translation of more than 7 mm is an indication of transverse ligament damage, which causes instability in the upper cervical complex<sup>9</sup>.

## CONCLUSION

Most congenital anomalies of the atlantal arch are found incidentally during investigation of neck pain, radiculopathy, neck mass, and after trauma. Anomalies of the atlas arch are associated other cervical spine anomalies. The incidence of anomalies of the atlas arch in this study is lower than that of other countries.

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