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REVIEW

Anatomical Basis and Clinical Significance of Atlas Stenosis: a Systematic Review

Christoforos SERIFIS[a], Leonora SOLOMOU[a], Dimitrios FILIPPOU[a]

[a]Department of Anatomy, Medical School, National and Kapodestrian University of Athens, Athens, Greece

ABSTRACT

Aim: *In this intriguing review, the authors dive into the complex world of atlas stenosis and its clinical impact on cervical myelopathy. The authors shed light on the origins of atlas stenosis, highlighting how congenital abnormalities of the atlas are a result of failures in embryological development. Furthermore, diagnosis and treatment methods are described.*

Materials and method: *Utilizing a detailed search of the literature in the PubMed database, the authors have compiled a comprehensive analysis of the anatomical and biological basis of this condition. After applying the exclusion criteria, 25 papers were deemed appropriate for the present review.*

Results: *With the help of computed tomography (CT) and magnetic resonance imaging (MRI) scans, diagnosis becomes possible, but the authors emphasize the importance of using special tests for a more accurate assessment. When it comes to treatment, surgical decompression with laminectomy or laminoplasty is the suggested approach. This highlights the severity of atlas stenosis and the need for intervention to alleviate the symptoms and prevent further neurological damage. What sets this review apart is its focus on the clinical profile of atlas stenosis. The authors discuss how the obstruction of cerebrospinal fluid flow can lead to neurological disorders and headaches. This sheds light on the broader impact this condition can have on patients' lives beyond simply the anatomical implications.*

Conclusion: *In the conclusion, the authors emphasize the susceptibility of the spinal cord to injuries even with mild mechanical pressure or instability due to aging in patients with atlas stenosis. Additionally, the instabilities caused by anatomical abnormalities of the atlas further highlight the importance of considering atlas stenosis in cases of cervical trauma. Overall, this review provides a fascinating insight into the connection between atlas stenosis and cervical myelopathy. By exploring the anatomical, biological, diagnostic, and treatment aspects of this condition, the authors have contributed to the understanding of this complex and often overlooked topic.*

Keywords: cervical spinal stenosis, atlas hypoplasia, atlas stenosis, atlas, stenosis, hypoplasia, cervical myelopathy.

Address for correspondence:
Christoforos Serifis
16 Lydias str, GR-11527 Athens, Greece
Tel: +30.6976.080683; email: koserifis10@gmail.com, smd2100130@uoa.gr

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INTRODUCTION

In this expert review, the authors take a deep dive into the rare but significant condition of spinal canal stenosis at the C1 level, also known as atlas hypoplasia (1). The review highlights the two main causes of this condition, including congenital abnormalities (0.69-4% (2) and acquired abnormalities, and provides valuable insights into the mechanisms and consequences of this stenosis. The authors present a comprehensive overview of the topic, discussing the impact of reduced diameter at the C1 vertebrae on the spinal cord and dura mater, and its correlation to symptomatic spinal canal stenosis. Spinal canal stenosis is one of the most common mechanisms that leads to cervical myelopathy (3). The rarity of the condition, with its occurrence below the C2 level, is intriguing and stimulates further interest in understanding its etiology. The review delves into the numerous factors contributing to the compression of the spinal cord and dura mater, including ossification of ligaments, osteophytes, and other degenerative factors. The authors effectively communicate the potential outcomes of these compressions, such as cervical myelopathy and circulation problems of cerebrospinal fluid, which engages in the pathomechanisms of the headache (4). The authors highlight the discrepancy between the prevalence of pain and the expression of other symptoms in patients with cervical myelopathy caused by C1 stenosis, providing valuable insights for physicians in diagnosing and treating this condition. Remarkably, 50% of patients with atlas stenosis report pain (5), while spasticity and walking issues seem to be the main symptoms (6). The brief mention of positive surgical outcomes for C1 stenosis patients adds further significance to the importance of timely diagnosis and intervention.

One of the strengths of this expert review lies in the use of relevant references from the literature, primarily case reports. This contributes to the credibility of the information presented. However, the limited availability of such literature further emphasizes the rarity of this condition and underscores the need for further research.

The review concludes by emphasizing the significance of considering C1 stenosis as a potential diagnosis in patients with myelopathy symptoms or mild neck trauma (7). The authors' suggestion to perform a thorough literature search while referencing normal anatomy and development of the atlas enhances the clinical applicability of the present review. Overall, this expert review offers a well-researched and comprehensive overview of C1 spinal canal stenosis and its implications for cervical myelopathy. ❑

MATERIAL AND METHODS

A literature search in the PubMed database was conducted in order to investigate the topic at hand. The authors diligently applied exclusion criteria to ensure that all articles selected for analysis were of high quality and relevance. By excluding articles about animals, those with paid full-text access, and those focusing solely on surgical techniques, the researchers ensured that only the most pertinent information would be considered. Additionally, articles related to genetic syndromes and C1 stenosis as part of C2 pathology were excluded, further narrowing down the scope of the review, focusing on relevant aspects of the topic. The researchers only included literature sources published during the past five years. However, publication date as an exclusion criterion limited the available references to such a large extent that a detailed review and of the topic would be impossible. Con-

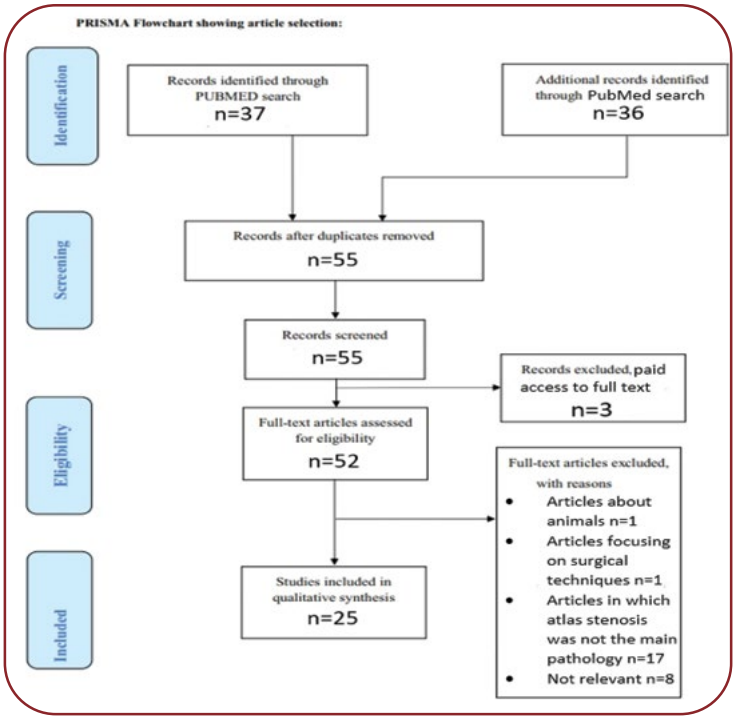

FIGURE 1. Flow chart

sequently, publication date was not considered an exclusion criterion.

The researchers implemented a two-step search process using specific keywords. The first search yielded 31 results, but upon closer examination, 11 cases were excluded for not meeting the established criteria. Subsequently, a second search was conducted using different keywords, resulting in 36 initial results. However, after eliminating duplicates and applying the exclusion criteria once more, a total of 25 relevant references were ultimately included in the review. By selecting only the most relevant articles and focusing on key sections such as the conclusion and discussion, the reviewers aimed to provide valuable insights and perspectives. The searching method is briefly shown in Figure 1. ❑

DISCUSSION

Embryological development. Congenital atlas anomalies can occur as a result of failures during embryogenesis. Understanding the normal developmental process of the atlas is crucial in comprehending the mechanisms behind these anomalies. During the seventh week of intrauterine life, three ossification centers give rise to the atlas (3, 6, 8-13). These centers form the anterior tubercle, anterior arch, posterior arch, and posterior tubercle. The posterior arch is nearly complete at birth, with only a small amount of cartilage remaining. Fusion of the posterior arch is typically completed between the ages of 3 and 10 years, while fusion of the anterior and lateral centers occurs between the ages of 5 and 9 years. In a small percentage of the population, a fourth ossification center forms the posterior tubercle. The presence of this fourth center, alongside failures in cartilage formation, can lead to anomalies in the posterior arch (12).

Posterior arch hypoplasia, or underdevelopment, is often caused by premature fusion or insufficient lengthening of the lateral center. This results in a smaller posterior arch in which two segments may fail to fuse, causing a midline cleft. In other cases, the atlas may be underdeveloped but still have a complete arch, leading to compression of the spinal cord. Mesenchymal defects during development can also lead to cartilage failure and various deformities. However, it is important to note that posterior arch defects are primarily a result of abnormalities in cartilage formation rather than ossification defects. The most common defect is a midline split of the posterior arch, which accounts for more than 90% of all posterior arch defects (8). Interestingly, these clefts may actually increase the available space for the spinal cord and may not always result in myelopathy. Connective tissue fills the gap and provides stability to the arch. The exact reasons for these defects are still unknown, but genetic factors are believed to play a significant role (8, 13). Further research is needed to fully understand the mechanisms behind these anomalies and to develop effective treatment.

Diameter variations at the C1 level. Atlas stenosis, a condition characterized by narrowing of the spinal canal at the C1 level, requires a complete understanding of the normal and pathologic diameters of the canal. The spinal canal is naturally larger at the C1 level, becoming smaller at lower levels before widening again at the C6 and C7 levels. However, the C4 level is considered the most critical for spinal canal stenosis (1).

The "Rule of Thirds," proposed by Steel in 1968 (3), suggests that the spinal cord, the dens and the cerebrospinal fluid space should each occupy one-third of the spinal canal's diameter at the C1 level. The average dural sac diameter is 10-12 mm at this level, while the normal sagittal spinal canal diameter ranges from 16-25 mm. A sagittal diameter below 14 mm can cause severe cord compression, and below 10 mm can result in symptoms of myelopathy (10, 11, 14-16). These findings are supported by Yamahata *et al* (16), who measured the spinal canal diameter in 213 patients and found a mean diameter of 18.6±1.9 mm in males and 17.6 ± 1.6 mm in females. Yamahata *et al* (16) also discovered that the cross-sectional area (CSA) of C1 (639±75 mm^2 in males and 574±60 mm^2 in females) and the inner anteroposterior diameter (IAP) of C1(30.7±2.0 mm in males and 28.5±1.6 mm in females) could indicate C1 smallness. The authors noted that C1 size increased with height and weight, emphasizing the importance of considering physiological data in determining normal and pathologic diameters. Kelly *et al* (17) conducted a cadaveric study with 543 specimens and found an inner sagittal diameter of 30.8±2.4 mm. They defined hypoplasia as an inner sagittal diameter below 28.1 mm, representing the lower 2.5% of measurements after considering size and radiographic magnification.

These results closely align with the previously mentioned findings. Kelly *et al* (17) also observed that, in 10% of cases, the dens occupied more than 40% of the C1 spinal canal, which was violating the "Rule of Thirds". Interestingly, Musa *et al* (7) reported different findings, with a sagittal inside diameter of 37.1±2.6 mm in males and 34.4±2.4 mm in females. They defined hypoplasia as a measurement two standard deviations below the standard. However, the measurements performed by Kelly *et al* (17) and the bone window CT scans conducted by Yamahata *et al* (16) appear to be more accurate due to their direct measurements in bony specimens.

Atlas anomalies associated with stenosis. Anomalies in C1 development are often leading to anatomical variations in C1, which can potentially be harmful to the individual. These abnormalities are more commonly found in the posterior arch, with 4% occurring in the posterior arch and 1% in the anterior arch, based on anecdotal cases (18). These variations in C1 do not typically affect the stability of the C1 joints and are mostly asymptomatic. However, Pacheco *et al* (2) propose a new classification of C1 abnormalities, continuing Currarino's classification and highlighting two types. Type A is characterized by hypertrophy of the posterior tubercle, where a fourth ossification center is present, alongside with premature fusion of the lateral sections of the posterior arch. This results in the formation of a hypoplastic C1, with the hypertrophied posterior arch possibly causing compression of the spinal cord (19). Type B is the most common defect in the posterior arch, occurring in 3-4% of the general population and represents over 90% of posterior arch defects (10). It is characterized by a midline cleft where the two hemiarches fail to fuse. Although this defect is generally asymptomatic (18), the abnormal thickening and knobby edges of the incomplete bone can lead to cord compression by incurring towards the spinal canal (20). This can be caused by the traction of muscles and ligaments attached to C1 or a possible pseudarthrosis. Types C and D are associated with unilateral and bilateral clefts, respectively. On the other hand, type E is the complete absence of the posterior arch, with only the posterior tubercle remaining, and type F is the absence of both the posterior arch and the posterior tubercle. Types E and F rarely cause compression as they tend to increase the available space for the cord. However, isolated hypoplasia is quite rare and can be accompanied by syndromic diseases such as Down syndrome, Turner syndrome, Morquio disease, achondroplasia, and degenerative changes due to aging such as cysts and ligament ossifications. Moreover, stenosis at the C1 level can occur due to abnormal fusion between the odontoid process of C2 and the anterior tubercle of C1. However, this condition is considered to be an abnormality of the C2 vertebra and is beyond the scope of this review.

Diagnosis. The diagnosis of C1 stenosis can be easily made by skilled doctors using MRI to detect possible compression of the spinal cord at the C1 level. Additionally, CT scans can be performed to identify any bone defects, such as enlargements or cracks, which may contribute to the condition. Oshima *et al* (3) proposed a screening test for C1 stenosis that involves assessing the position of the ventral plate of the atlas in relation to the line of the spinal plate. This test has demonstrated high sensitivity (100%) and specificity (80%) in detecting tight spaces that can be treated with wiring. Therefore, it is regarded as an accurate method for diagnosing possible C1 stenosis using standard lateral radiographs.

Symptoms of C1 stenosis are often similar to those experienced in cervical spondylopathy due to compression of the spinal cord at lower levels of the spine. These symptoms tend to be neurological in nature and include sensory and motor disturbances, unsteadiness, difficulty walking, and sphincter dysfunction (3). It is important to note that even mild mechanical compression associated with C1 stenosis can increase the risk of spinal cord injury (7). In some cases, patients with C1 stenosis may also experience spasticity, dizziness, paresthesia (tingling or numbness) of the limbs, and difficulties with attention. Remarkably, there have been reports of tetraplegia in children with this condition, underscoring the severity of the impact on neurological function (21). Additionally, Gutmann *et al* (4) reported cases of headaches related to dural compression of the C1 short posterior arch. The mechanical stenosis can hinder the flow of cerebrospinal fluid, leading to rapid and irregular changes in intracranial pressure, which can cause headaches. In a study conducted by Liliang *et al* (21), biomechanical abnormalities were identified in 119 patients, with 19 cases featuring combined abdominothoracic compression and

11 cases exhibiting C1 posterior arch shortening out of the total abnormalities observed. These findings highlight the significance of identifying and addressing these abnormalities early on to prevent further complications.

Treatment principles. In the realm of treatment principles, surgical intervention is the recommended course of action for individuals experiencing symptomatic C1 stenosis. The two primary treatments for this condition are C1 laminectomy and C1 laminoplasty (22). C1 laminectomy involves the complete removal of the lamina in the area of compression. This traditional technique has proven effective in creating additional space for the spinal cord and achieving notable neurological improvement. However, it is important to note that C1 laminectomy is only suitable when there is no or minimal atlantoaxial subluxation. This is due to the potential risk of exacerbating joint instability, which can further complicate matters. In cases where atlantoaxial subluxation is a contributing factor (as is often the case with symptomatic atlas canal stenosis), C1 laminoplasty emerges as a more efficient technique. This operation involves cutting and opening the lamina like a door, thereby creating more space for the spinal cord. To support this door-like structure, titanium miniplates are utilized. Additionally, autogenous iliac bone grafts are placed in the opening. It is worth noting that C1 laminoplasty is specifically performed in the cervical part of the spine to relieve neurological symptoms. In the majority of instances, C1 stenosis necessitates a combined approach of decompression and stabilization. As such, C1-C2 fusion is required in conjunction with C1 laminoplasty. However, achieving C1-C2 fusion poses challenges when using C1 laminectomy, as there is often insufficient bony material available for fusion.

To support the efficacy of C1 laminoplasty and C1-C2 fusion, a study conducted by Chen *et al* (22) demonstrated positive outcomes. The study included 16 patients who underwent these procedures, ultimately leading to full decompression of the cord and a notable decrease in neck pain. Notably, the inner sagittal diameter increased from 26.3±2.6 mm to 34.9±2.9 mm after the operation. With an average follow-up period of 24.9 months, all patients experienced significant improvement in neurological symptoms.

In summary, when addressing symptomatic C1 stenosis, surgical intervention is crucial for preventing the aggravation of neurological symptoms. While C1 laminectomy provides substantial neurological improvement, it is essential to consider atlantoaxial subluxation and potential joint instability. In cases where subluxation is present, C1 laminoplasty offers a more efficient treatment option. Moreover, the combination of C1-C2 fusion and C1 laminoplasty is crucial for achieving decompression and stabilization. The study conducted by Chen *et al* (22) further supports the positive outcomes of these procedures, showcasing the significant improvement in neurological symptoms among patients. ❑

CONCLUSIONS

In the fascinating world of anatomy, the atlas takes center stage as a crucial vertebra that plays a vital role in supporting the weight of our skull and protecting our delicate spinal cord. But just like in any great production, sometimes errors occur during the development process, leading to interesting abnormalities in this ring-shaped bone. Embryonic mishaps and endochondral hyperplasia can cause the atlas to exhibit conditions such as hyperossification, fissures, or even loss of certain parts. While many of these abnormalities may go unnoticed, there are cases where they can have a significant impact on our health. When the spinal canal's diameter is greatly reduced due to these abnormalities, the spinal cord can become compressed, resulting in the development of myelopathy symptoms. But that is not the only twist in this tale; abnormal changes in intraventricular pressure can also lead to pounding headaches, adding yet another layer of complexity.

Luckily, modern medical advancements offer us a way to unlock the secrets of C1 stenosis. Through the power of magnetic resonance imaging (MRI) and computed tomography (CT), healthcare professionals can examine soft tissue compression and bony irregularities and diagnose atlas stenosis. When it comes to treatment, innovative techniques such as C1 laminoplasty with C1-C2 fusion take the stage. This tried and tested method has proven its effectiveness in improving neurological symptoms, offering hope to those affected by C1 stenosis. ❑

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References

1. **Yamahata H, Niiro T, Mori M, et. al.** Is the atlas size associated with the pathophysiology of symptomatic spinal canal stenosis at the C1 level? *J Clin Neurosci* 2018;57:58-62.
2. **Junior MGP, Dos Santos NADSQ, Ribeiro RT, et al.** Hypoplasia of C1's posterior arch: Is there an ideal anatomical classification? *Surg Neurol Int* 2021;12:623.
3. **Joaquim AF, Baum G, Tan LA, et al.** C1 Stenosis – An Easily Missed Cause for Cervical Myelopathy. *Neurospine* 2019;16:456-461.
4. **Gutmann G, Roesner J.** The subforaminal stenosis headache. *Acta Neurochir* (Wien). 1979;50:201-215.
5. **Desai SK, Vadivelu S, Patel AJ, et al.** Isolated cervical spinal canal stenosis at C-1 in the pediatric population and in Williams syndrome. *J Neurosurg Spine* 2013;18:558-563.
6. **Pereira Duarte M, Camino Willhuber G, Petracchi M, et al.** Symptomatic Atlas Hypoplasia in a Latin-American Patient: Case Report and Literature Review. *J Am Acad Orthop Surg Glob Res Rev* 2021;5:e21.00041.
7. **Musha Y, Mizutani K.** Cervical myelopathy accompanied with hypoplasia of the posterior arch of the atlas: case report. *J Spinal Disord Tech* 2009;22:228-232.
8. **Atasoy C, Fitoz S, Karan B, et al.** A rare cause of cervical spinal stenosis: posterior arch hypoplasia in a bipartite atlas. *Neuroradiology* 2002;44:253-255.
9. **Chang H, Cho WJ, Park JB, et al.** Cervical myelopathy secondary to atlas hypoplasia – reports of 3 adult cases. *Asian Spine J* 2007;1:48-52.
10. **Hsu YH, Huang WC, Liou KD, et al.** Cervical spinal stenosis and myelopathy due to atlas hypoplasia. *J Chin Med Assoc* 2007;70:339-344.
11. **Phan N, Marras C, Midha R, et al.** Cervical myelopathy caused by hypoplasia of the atlas: two case reports and review of the literature. *Neurosurgery* 1998;43:629-633.
12. **Chung SB, Yoon SH, Jin YJ, et al.** Anteroposterior spondyloschisis of atlas with incurving of the posterior arch causing compressive myelopathy. *Spine* (Phila Pa 1976) 2010;35:E67-E70.
13. **Urasaki E, Yasukouchi H, Yokota A.** Atlas hypoplasia manifesting as myelopathy in a child--case report. *Neurol Med Chir* (Tokyo) 2001;41:160-162.
14. **Nishikawa K, Ludwig SC, Colón RJ, et al.** Cervical myelopathy and congenital stenosis from hypoplasia of the atlas: report of three cases and literature review. *Spine* (Phila Pa 1976) 2001;26:E80-E86.
15. **Sato K, Kubota T, Takeuchi H, et al.** Atlas hypoplasia associated with non-traumatic retro-odontoid mass. *Neurol Med Chir* (Tokyo) 2006;46:202-205.
16. **Yamahata H, Hirano H, Yamaguchi S, et al.** What Is the Most Representative Parameter for Describing the Size of the Atlas? CT Morphometric Analysis of the Atlas with Special Reference to Atlas Hypoplasia. *Neurol Med Chir* (Tokyo) 2017;57:461-466.
17. **Kelly MP, Oshima Y, Yeom JS, et al.** Defining hyoplasia of the atlas: a cadaveric study. *Spine* (Phila Pa 1976) 2014;39:E1243-E1247.
18. **Devi BI, Shenoy SN, Panigrahi MK, et al.** Anomaly of arch of atlas--a rare cause of symptomatic canal stenosis in children. *Pediatr Neurosurg* 1997;26:214-217; discussion 217-218.
19. **Kasliwal MK, Traynelis VC.** Hypertrophic posterior arch of atlas causing cervical myelopathy. *Asian Spine J* 2012;6:284-286.
20. **Connor SE, Chandler C, Robinson S, et al.** Congenital midline cleft of the posterior arch of atlas: a rare cause of symptomatic cervical canal stenosis. *Eur Radiol* 2001;11:1766-1769.
21. **Liliang PC, Lui CC, Cheng MH, et al.** Atlantal stenosis: a rare cause of quadriparesis in a child. *Case report. J Neurosurg* 2000;92(2 Suppl):211-213.
22. **Chen L, Zhu X, He B, et al.** Effect of C1 Single-door Laminoplasty on Symptomatic Atlas Canal Stenosis. *Orthop Surg* 2022;14:2757-2765.
23. **Lampropoulou-Adamidou K, Athanassacopoulos M, Karampinas PK, et al.** Congenital variations of the upper cervical spine and their importance in preoperative diagnosis. A case report and a review of the literature. *Eur J Orthop Surg Traumatol* 2013;23 Suppl 1:S101-S105.
24. **Matsunaga S, Imakiire T, Koga H, et al.** Occult spinal canal stenosis due to C-1 hypoplasia in children with Down syndrome. *J Neurosurg* 2007;107(6 Suppl):457-459.
25. **Yunoki M.** A surgical case of C1 arch stenosis: A case report and review of literature. *Surg Neurol Int* 2021;12:71.