



# Percutaneous full endoscopic C1 laminectomy for developmental atlantal stenosis with myelopathy: a case report of three cases and review of the literature

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**Background:** Developmental atlantal stenosis with myelopathy (DASM) in adults is a rare disease that only sporadic cases have been reported over the years. C1 laminectomy (C1L) is one of the most common operations for its treatment. However, as an open surgery, it has shortcomings such as large trauma and slow postoperative rehabilitation, and minimally invasive spine surgery (MISS) offers alternative treatment options with advantages. MISS instruments expand the technical capabilities of surgeons, which allows safer and more effective therapeutics for difficult and complicated diseases. This case report presents a new minimally invasive approach; percutaneous full endoscopic C1 laminectomy (PFEC1L), for the treatment of DASM, and to consolidate the current literature on the condition to summarize its etiologies, clinical manifestations, diagnostic criteria, surgical management, and prognoses.

**Case Description:** The patient in Case 1 presented with neck pain and numbness and weakness in the limbs. The patient in Case 2 presented with numbness in the extremities and the patient in Case 3 presented with bilateral hand numbness and left lower limb weakness. They were all diagnosed with DASM and underwent PFEC1L treatment to maintain the enlargement and decompression of the atlantal canal, which achieved favorable outcomes without complications during the postoperative follow-up visit.

**Conclusions:** DASM is rare but potentially dangerous. Its diagnosis is made based on clinical manifestations combined with radiological imaging examinations, especially computed tomography (CT) scan and magnetic resonance imaging (MRI). While C1L is the most common surgical method, PFEC1L is a new feasible and safe therapeutic option with comparable good outcomes and the advantage of being minimally-invasive. To our knowledge this is the first report that PFEC1L was applied for DASM treatment.

**Keywords:** Endoscopic spine surgery; C1 laminectomy (C1L); cervical myelopathy; spinal canal stenosis; case report

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## Introduction

Despite its low prevalence, atlas deformity associated with spinal cord compression constitutes a potentially devastating condition, which may lead to quadriplegia and even death (1,2). In 1989, Sawada *et al.* (3) first reported the use of C1 laminectomy (C1L) for a case of canal stenosis at the level of the atlas. Subsequently, in 1994, seven cases of cervical myelopathy caused by aplasia of the posterior arch of the atlas (PAA) were reported by Currarino (4). Shah *et al.* (5) and Bhattacharjee *et al.* (6) also described similar cases of abnormal development of the C1 posterior arch which led to stenosis of the spinal canal. However, almost all of the relevant articles were case reports. Recently, Wang *et al.* (7) found the pathophysiological mechanism of developmental atlantal stenosis with myelopathy (DASM) was complex and multifactorial, and the clinical symptoms were diverse in a series of 15 cases.

Surgery is the most effective treatment for DASM, and can significantly relieve spinal cord compression, ameliorate myelopathy symptoms, and improve spinal cord function. At present, the most common surgical method for DASM is C1L with or without atlantoaxial fixation and fusion (8-11). Although this open surgery can improve neurological symptoms, it requires wide surgical exposure that may introduce remarkable operative trauma, and the operation risk is relatively high, especially in elderly populations with comorbidities.

Minimally invasive spine surgery (MISS) offers alternative treatment options with advantages. Endoscope systems expand the technical capabilities of surgeons, which allows safer and more effective therapeutics for difficult and complicated diseases (12,13). In the past decade, endoscopic spine surgery has been widely used for lumbar, thoracic, and cervical degenerative disease (14,15). Here we present a novel approach for the treatment of DASM by percutaneous full endoscopic C1 laminectomy (PFEC1L) to take over the disadvantage of C1L, such as soft tissue damage and massive hemorrhage as an open operation. To the best of our knowledge, it has not been previously reported. In addition, we review and summarize the relevant literature and discuss the etiology, pathophysiology, and diagnostic and treatment strategies for DASM. We present the following article in accordance with the CARE reporting checklist (available at <https://atm.amegroups.com/article/view/10.21037/atm-22-2282/rc>).

## Case presentation

### Ethics

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

### Case 1

A 66-year-old male patient presented to our hospital with complaints of neck pain and numbness and weakness in the limbs for more than 7 months. His symptoms were aggravated in the past 2 months and included walking instability and difficulty grasping and holding objects. Neurological signs including Hoffman, Babinski and ankle clonus were positive, and muscle strength of the upper and lower extremities was grade IV. Magnetic resonance imaging (MRI) demonstrated spinal cord compression, as well as high signal change on T2-weighted imaging (T2WI) at the level of the atlas (*Figure 1*). A preoperative sagittal computed tomography (CT) scan showed a 7.5-mm diameter of the C1 space available for the cord (SAC), and preoperative flexion and extension cervical radiographs revealed good atlantoaxial stability. A cervical CT angiography (CTA) examination was performed to specifically assess the position of the vertebral artery (VA) and estimate the distance between the VA and PAA. While the patient was treated conservatively in the clinic for 8 weeks before being hospitalized, the symptoms were not notably relieved or even aggravated. The preoperative modified Japanese Orthopaedic Association (mJOA) score was 8 points (8/17), and the visual analogue scale (VAS) was 5 points (5/10). The patient finally underwent robot-assisted PFEC1L after excluding contraindications on December 2018.

The operative time was 65 minutes and blood loss was minimal. No drainage, painkillers, or antibiotics were needed postoperatively, and the patient was discharged on the fifth postoperative day without problem. The 3-month postoperative mJOA score was 11 points (11/17), and 2-year postoperative was 14 points (14/17), at which time the



**Figure 1** A 66-year-old male with DASD who underwent PFEC1L. (A) Preoperative MRI. The arrow indicated the compression of the spinal cord, a high signal intensity of the central cord at C1 level and central herniation of the C3–6 disc with signs of spinal compression. (B) Preoperative cervical CTA. The arrow indicated an abnormally enlarged left VA which was closer to the left PAA. (C) Robot-assisted establishment of the working channel. (D) Robotic axial director profile was placed to the tubercle of the PAA. (E,F) AP and lateral views of the working tube. (G) Adequate decompression of the spinal cord after PFEC1L. (H) Postoperative MRI. The arrow indicated a complete neural decompression at the level of C1. (I) Postoperative CT three-dimensional reconstruction showing the resection of the PAA is about 1.9 cm. (J) Postoperative midline sagittal CT. The arrow indicated that the PAA was removed and the spinal canal of the atlas was apparently enlarged. DASD, developmental atlantal stenosis with myelopathy; PFEC1L, percutaneous full endoscopic C1 laminectomy; MRI, magnetic resonance imaging; CTA, computed tomography angiography; VA, vertebral artery; PAA, posterior arch of the atlas; AP, anteroposterior; CT, computed tomography.

patient reported slight limbs numbness without weakness or pain, and his mobility has basically returned to normal at 30-month postoperative follow-up. Postoperative MRI

showed significant improvement of spinal cord compression and the three-dimensional CT showed the removed section of the PAA was about 1.9 cm (*Figure 1*).

### Case 2

An 83-year-old woman experienced numbness in the extremities for 2 years. She had slight asthenia in the right upper limb, and the clinical manifestations had progressed in the past 6 months to where she could not walk without difficulty. The patient had a history of coronary heart disease, hypertension, type 2 diabetes mellitus, osteoporosis, and hyperlipidemia. Neurological examination of the sensory system revealed disturbance of deep sensation, which manifested as hyperreflexia of the patellar and Achilles tendons and a positive Romberg's test. The tongue and palate moved normally without atrophy or fasciculation, and there was neither atrophy nor wasting of the sternocleidomastoid muscles, although slight muscular wasting was observed in the right arms and forearms. Preoperative CT scan of the cervical spine revealed a markedly narrow canal at the level of the atlas, where the SAC was 7.0-mm. Preoperative MRI showed the deformed PAA compressing the spinal cord, and spinal cord compression was also observed at the location of C5–6 (*Figure 2*). Preoperative X-ray did not reveal obvious atlanto-axial subluxation or instability, and the mJOA score was 8 points (8/17) and VAS score 3 points (3/10). The patient was treated surgically by robot-assisted PFEC1L and percutaneous full endoscopic C5/6 decompression on October 2019, with a total operative time of 118 min, of which 62 min was for PFEC1L. The patient's neck pain was alleviated at the early postoperative stage, and she was discharged 3 days after surgery. At 2-year after surgery, her weakness and numbness of the extremities were alleviated, the motor examination showed improvement, and the postoperative mJOA score was 13 points (13/17) and VAS score 1 point (1/10). Postoperative MRI revealed spinal cord decompression was sufficiently attained, and CT scan demonstrated the PAA was partial removed (*Figure 2*).

### Case 3

A 52-year-old female with bilateral hand numbness and left lower limb weakness for 7 years complained of neck pain at the upper cervical spine level exacerbated with extension and rotation to either side. The patient had a history of severe anaemia and osteopenia. Physical examination revealed a short neck, hyperreflexic bilateral triceps, brachioradialis, patellar tendon, and Achilles tendon reflexes, and positive Romberg, Hoffman, and Babinski signs. The left lower limbs, including hip flexors, knee

extensors, ankle dorsiflexors, and ankle plantar flexors, showed grade IV strength. Initial MRI showed spinal cord narrowing thickness and heterogeneous signal at the level of C1, and the spinal cord compression due to abnormal PAA was particularly severe on the left side. A preoperative CT scan revealed a markedly narrow canal at the level of the atlas, and the SAC was 7.0 mm (*Figure 3*), and the mJOA scored 9 points (9/17) and VAS 4 points (4/10). Conservative treatment was commenced and continued for 3 months with virtually no improvement, and the patient accepted PFEC1L in July 2020.

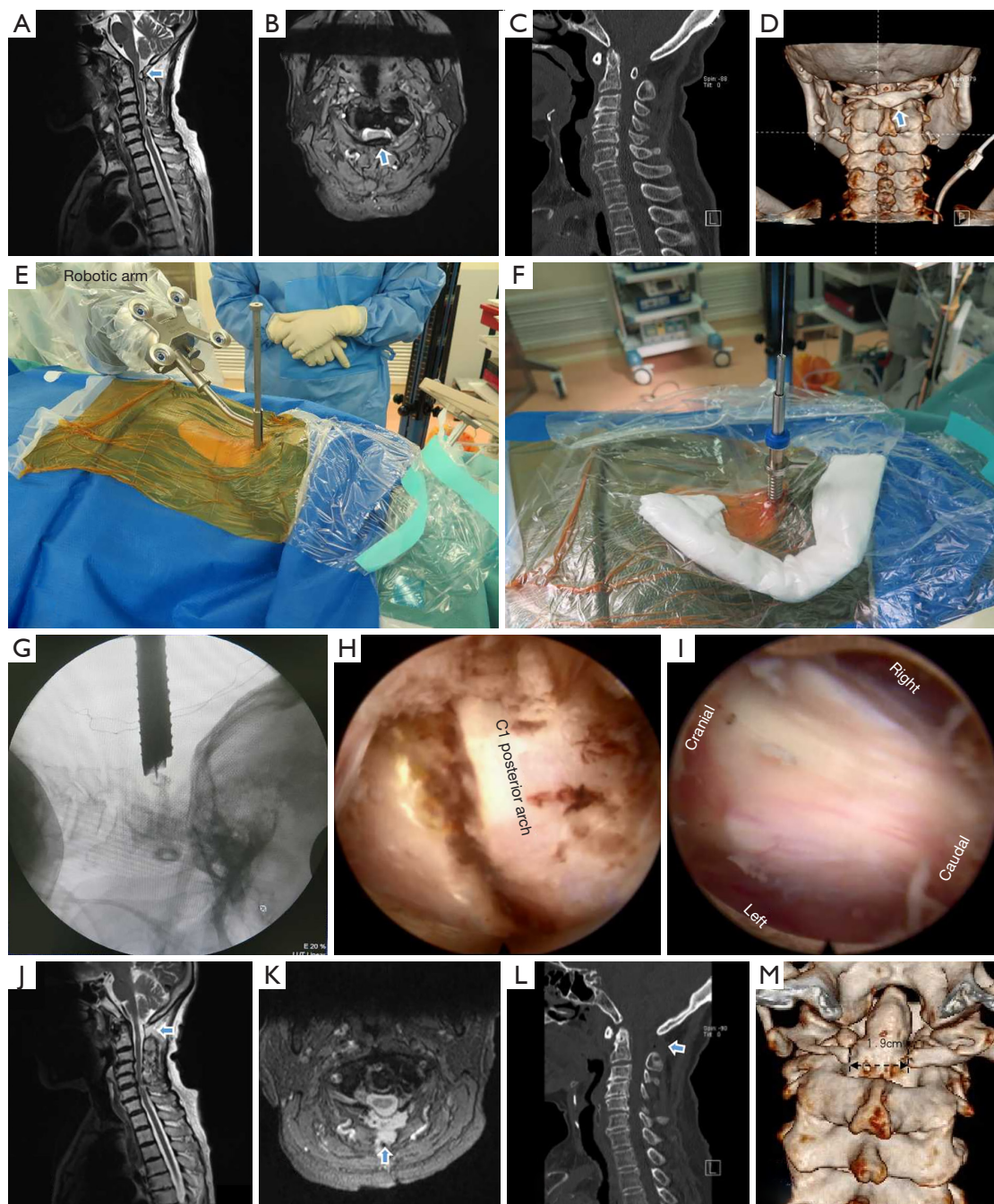
The operation took 70 minutes to complete, after which she was able to ambulate on the 1st postoperative day and was discharged well 4 days post-operation. The patients' symptoms were comparably improved significantly 1-year after surgery, and the mJOA was 14 points (14/17) and VAS score 1 point (1/10).

The whole process of diagnosis and treatment of DASM by PFEC1L in three cases was outlined in *Figure 4*.

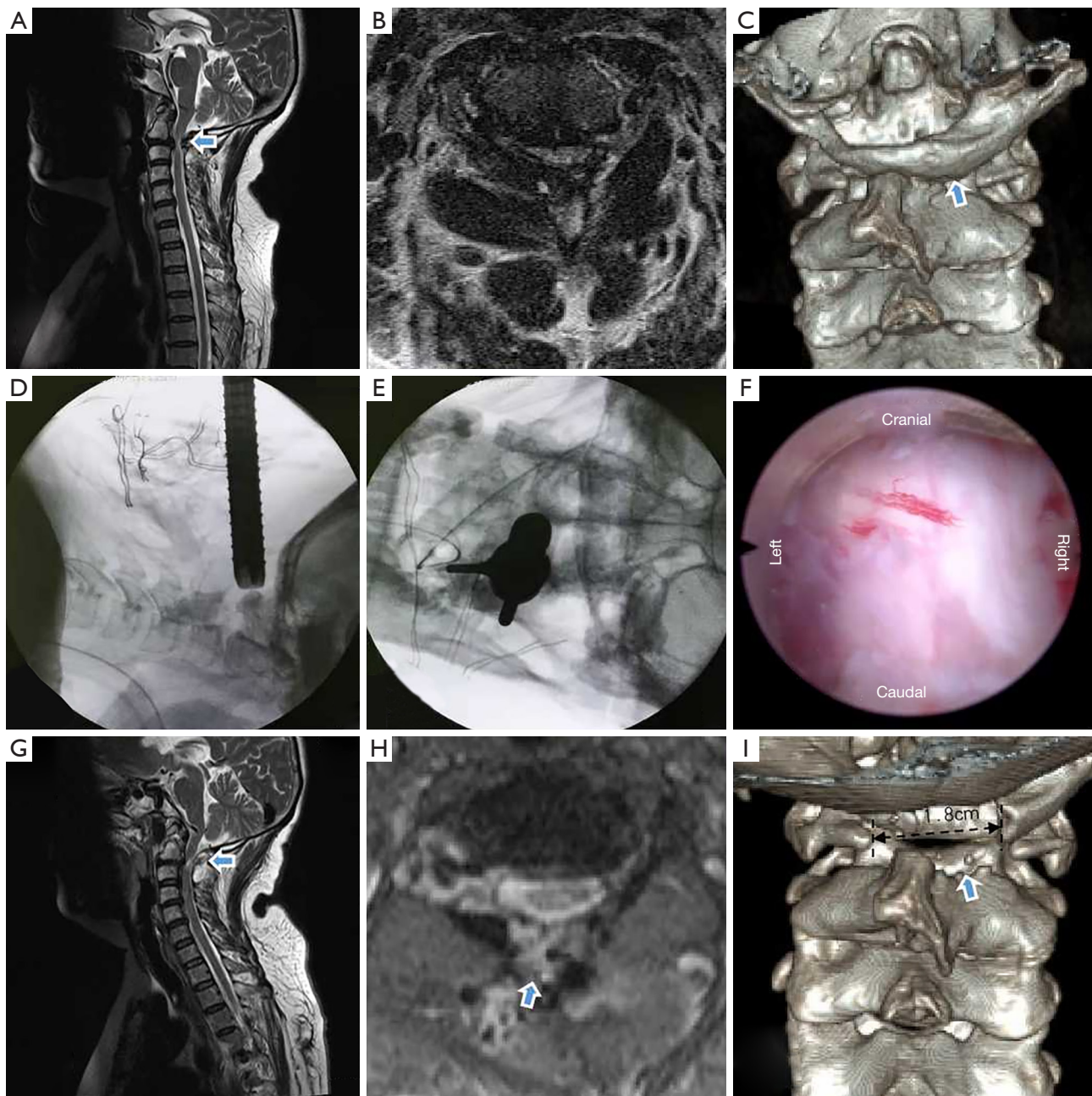
### Method of PFEC1L

The procedure was performed under general anesthesia with patients placed in the prone position. To begin with, planning of the working-channel endoscope trajectories was performed on the robotic workstation (*Figure 5*). With the help of the robot, a Kirschner wire was accurately established to the posterior tubercle of the atlas, then the dilating tube and working channel were rapidly placed along the wire. All subsequent steps were performed using the endoscope and non-bacteriostatic saline irrigation, including removal of the PAA and decompression of the spinal cord.

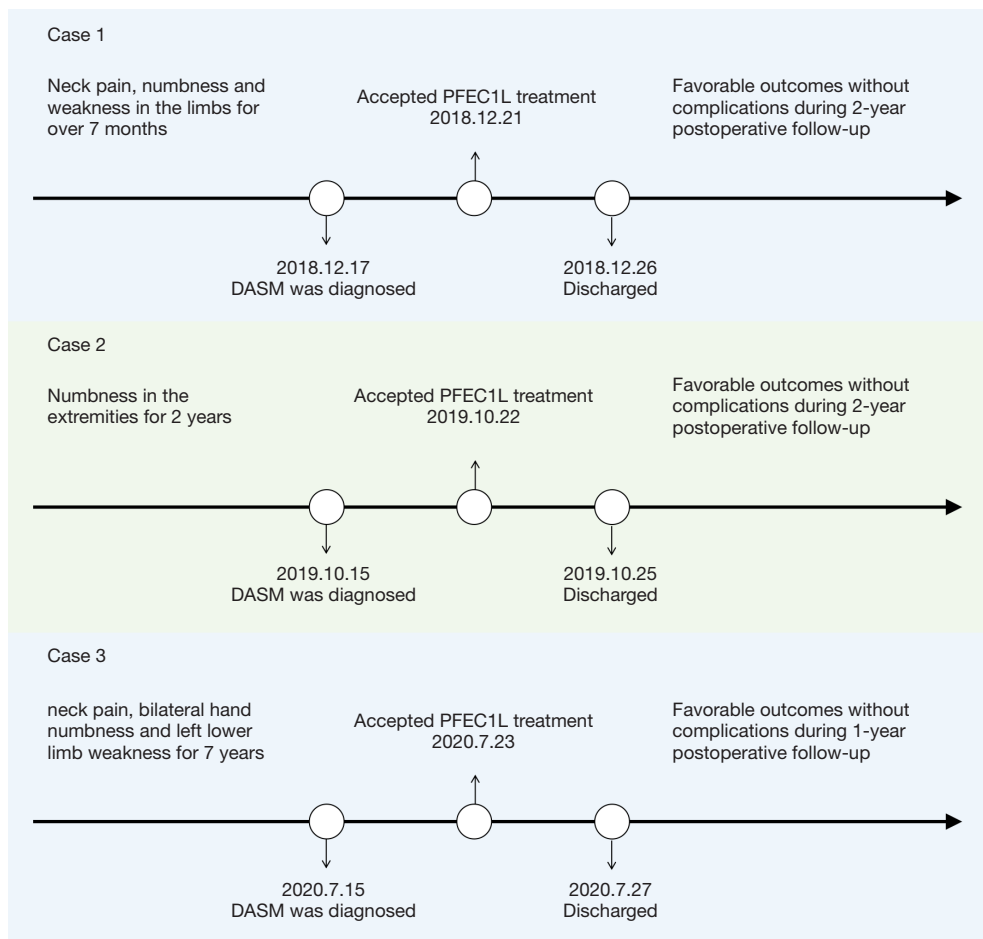
Under the endoscope, soft tissue around the PAA was cleaned until its upper and lower boundaries were clearly distinguishable, with the posterior tubercle taken as the center and expanded to the left and right sides by 1 cm each. The external cortical bone and internal cancellous bone were then removed using a high-speed drill, and the internal cortical bone was temporarily preserved. The internal cortical bone was carefully removed using a Kerrison rongeur, followed by exposure of the intact spinal cord by careful removal of the dorsal ligament. After adequate decompression of the spinal cord was achieved, a bipolar RF device was used for hemostasis, the endoscope and working tube were exited, and the wound was closed using surgical glue.



**Figure 2** An 83-year-old female with DASM and C5–6 CSM who underwent PFEC1L and percutaneous full endoscopic C5–6 decompression. (A) Preoperative sagittal T2-weighted MRI. The arrow indicated the obvious compression of the spinal cord at the level of C1 and C5–6. (B) Preoperative axial T2-weighted MRI. The arrow indicated the malformations of the PAA had compressed the spinal cord. (C) Preoperative sagittal CT scan, the SAC was 7.0-mm. (D) Preoperative three-dimensional CT. The arrow indicated the abnormal left-right asymmetry of the PAA. (E–G) The creation of an appropriate endoscopic surgical corridor with robotic assistance. (H) The PAA under the endoscopic view. (I) No sign of spinal cord compression after PFEC1L. (J,K) Postoperative MRI. The arrow indicated the spinal cord compression was relieved, especially at the level of the atlas. (L,M) Postoperative CT scan. The arrow indicated that the PAA was removed. DASM, developmental atlantal stenosis with myelopathy; CSM, cervical spondylotic myelopathy; PFEC1L, percutaneous full endoscopic C1 laminectomy; MRI, magnetic resonance imaging; PAA, posterior arch of the atlas; CT, computed tomography; SAC, space available for the cord; PAA, posterior arch of the atlas.



**Figure 3** A 52-year-old female with DASM who accepted PFEC1L and percutaneous full endoscopic partial hemilaminectomy at the upper half of C2. (A) Preoperative mid-sagittal MRI. The arrow indicated the compression of the spinal cord at the level of C1–2. (B) Preoperative axial MRI showing deformity of PAA as a cause of spinal cord compression. (C) Preoperative CT scan. The arrow indicated the developmental malformation of the PAA. (D,E) Intraoperative fluoroscopy to confirm the location of the working channel. (F) Full spinal cord decompression can be observed at endoscopy. (G,H) Postoperative MRI. The arrow indicated that the atlas canal was enlarged and the spinal cord compression was relieved. (I) Postoperative CT three-dimensional reconstruction. The arrow indicated that the PAA and the right C2 part of lamina was removed and the resection of the PAA was about 1.8-cm. DASM, developmental atlantal stenosis with myelopathy; PFEC1L, percutaneous full endoscopic C1 laminectomy; MRI, magnetic resonance imaging; PAA, posterior arch of the atlas; CT, computed tomography.



**Figure 4** Timeline for diagnosis and treatment of DASM by PFEC1L. DASM, developmental atlantal stenosis with myelopathy; PFEC1L, percutaneous full endoscopic C1 laminectomy.

### Literature search

PubMed, Ovid MEDLINE, and EMBASE were searched for relevant articles. The search terms were: [(“C1 stenosis” or “atlas stenosis” or “atlantal stenosis” or “atlantoaxial stenosis” or “atlanto-axial stenosis”) AND (“C1 hypoplasia” or “C1 malformation” or “atlas hypoplasia” or “atlas malformation”)]. All articles of any study design discussing DASM were considered for inclusion, while experimental or animal studies and non-English language studies were excluded. After an initial screen of abstracts and article titles, full text articles of all potential studies were obtained.

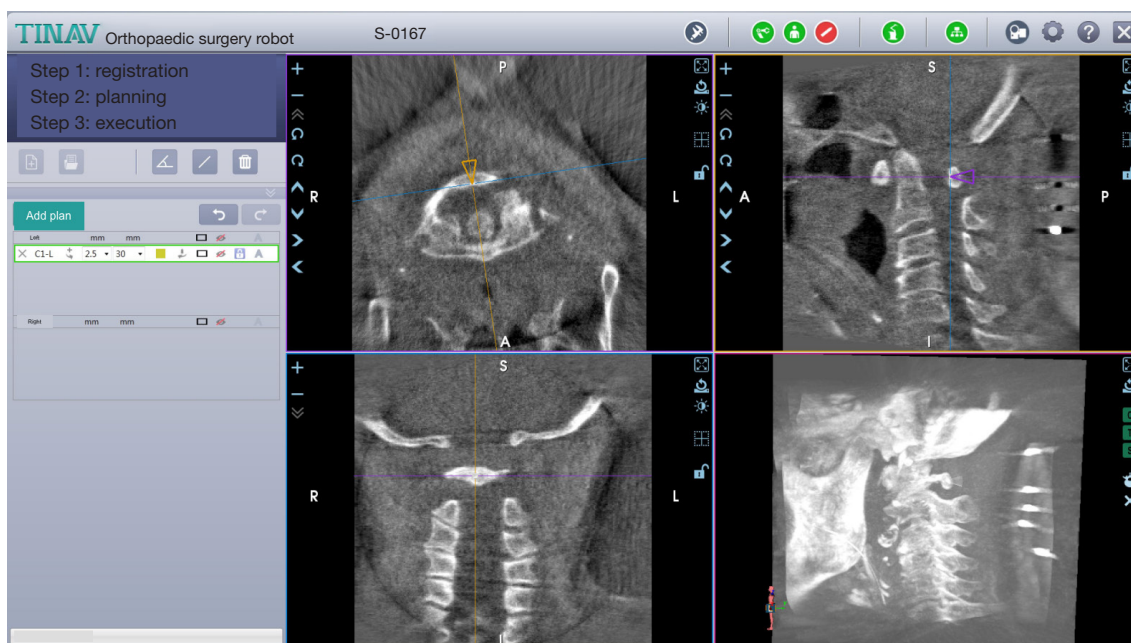
A total of 232 studies were identified from the initial search, then 27 duplicates, non-English, and animal studies were removed. Titles and abstracts of the 205 remaining studies were screened according to the pre-defined inclusion criteria, and 161 studies were excluded, leaving 44 articles

for critical review and consolidation for this literature review. *Figure 6* shows the flowchart.

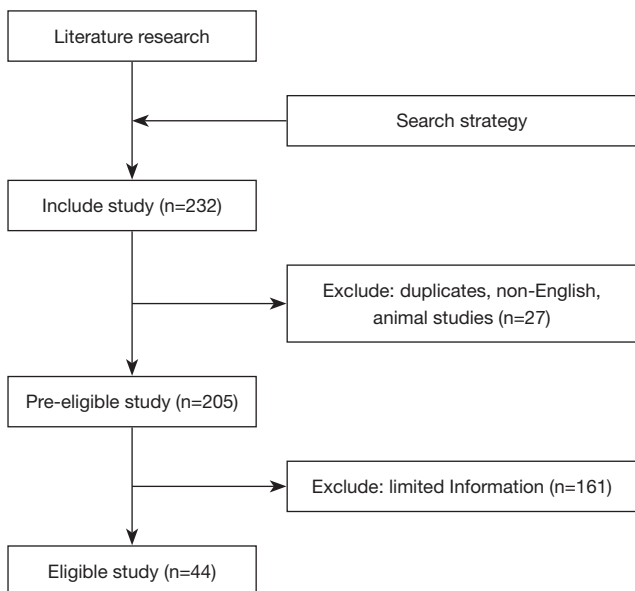
### Discussion

#### *Etiology and pathophysiology of DASM*

The appearance of DASM is highly unusual in adults, and its pathogenesis remains poorly understood. The development of atlantal stenosis involves several factors, among which a genetic cause is one of the most important (16). The occurrence of atlas developmental malformation is more common in patients with Chiari malformation, Down syndrome, Klippel-Feil syndrome, Noonan syndrome, Williams syndrome, diffuse idiopathic skeletal hyperostosis (DISH), atlanto-axial instability, and atlantoaxial subluxation with or without os odontoideum



**Figure 5** Planning of the puncture path and the anchoring point of the Kirschner-wire were performed on the robotic platform. Pre-operative planning can be observed on the sagittal, cross-sectional, coronal imaging, and three-dimensional reconstructions of the CT. CT, computed tomography.



**Figure 6** Study flowchart.

(4,5,17-24). Stenosis secondary of C1 without trauma and atlantoaxial instability is a possible pathophysiologic factor, which may be secondary to atlas hypoplasia, ossification of the posterior atlantoaxial membrane (PAAM) (20), or

ossification of the transverse atlantal ligament (OTAL) (25) that lead to consistent space reduction of the spinal cord. The developmentally narrowed spinal canal of the atlas in minors is generally displayed as the inturning of a bifid posterior arch, partial atlas agenesis, or absence of the PAA (4,26-29). However, in adults, the main deformities are usually an asymmetric structure, abnormal bone hyperplasia, and inborn spinal canal stenosis with complete PAA (30,31). On rare occasions, it can also show dysmorphic features with failure of fusion of the two hemi-arches with a midline cleft as reported by Shah *et al.* (5) in a 44-year-old female. We speculate DASM may be related to genes and age, and the deformity develops incrementally with age. According to the limited existing literature available, DASM is more commonly found in middle-aged and elderly individuals over 50 years old (8).

**Subtypes of DASM**

Depending on the etiology, atlantal stenosis can be classified as developmental or secondary (6,32,33). DASM is a congenital developmental deformity of the atlas resulting in reduction of the spinal canal diameter and spinal cord compression. Atlas developmental deformities mainly



manifest at three sites, including the anterior arch, posterior arch, and atlantoaxial lateral masses. Among these, the prevalence of posterior arch malformation is the highest (7) and can lead to a congenitally small spinal canal and subsequent myelopathy. It was reported that 4% of 1,613 autopsies showed posterior arch deformity of the atlas, which can be divided into two types: median fissure or hypoplasia (34,35). In another study, hypoplasia of the PAA was divided into spondyloepiphyseal dysplasia congenita and an idiopathic type (36).

Currarino *et al.* (4) divided congenital anomalies of the posterior arch into five types according to different morphologies of the PAA, including (I) failure of posterior midline fusion of the two hemiarches, (II) unilateral clefts, (III) bilateral clefts, (IV) absence of the posterior arch with persistent posterior tubercle, and (V) absence of the entire arch including the posterior tubercle, which is a classification method mainly based on abnormal morphological changes of the PAA in minors. According to the morphological analysis of CT image data, Wang *et al.* (7) divided DASM into four subgroups, including (I) small size atlas, (II) hypertrophy of PAA, (III) incurved of PAA, and (IV) hypertrophy of the odontoid. Among these, type I is relatively common, type II and III are relatively rare, and type IV are the least common.

### ***Clinical manifestations of DASM***

While anomaly of the PAA is an easily overlooked cause of symptomatic cervical myelopathy, its clinical symptoms can be very severe. The symptoms of DASM are complex and diverse, including occipital neuralgia, occipitocervical pain, paresthesia and weakness of limbs, gait disturbance, poor fine motor skills and difficulties in holding or grasping, urinary incontinence, ataxia, and apnea (5-8,10,21,31,33,37). Certain cervical positions can induce or exacerbate these symptoms, and movement restrictions in the upper cervical spine can be observed in some patients. Furthermore, breathing difficulty is observed in patients with severe worsening of the condition (6).

### ***Imaging characteristics of DASM***

Yamahata *et al.* (38) measured the internal anterior posterior diameter (IAPD) at the C1 level of 213 cases by CT scan, and the results showed the IAPD was 30.7 mm in males and 28.7 mm in females. Musha *et al.* (8) found the IAPD was 34.4 mm in Japanese females and 37.1 mm in males, and

that atlantal stenosis was confirmed when the IAPD less than 29.4 mm in females and 30.5 mm in males. In another study of 543 cadaveric cervical spine specimens (39), the mean C1 inner sagittal diameter was 30.8 mm, and a value below 26.1 mm was defined as atlas dysplasia. Although malformations of the spinal canal of the atlas can be defined by the IAPD, the SAC in relation to the dens and the C1 inner sagittal diameter are more important and sensitive indicators to show the useful activity space of the spinal cord.

In accordance with the results of Hinck *et al.* (40), the normal range of the SAC is 15 to 20 mm. It is generally assumed that atlas dysplasia can be defined when the SAC is less than 14 mm (41) and clinical signs and symptoms will develop when it is less than 10 mm (42). Consistent with the results of the above studies, the SAC of the cases in the present study was 7 to 7.5 mm. It should be emphasized that MRI provides an important basis for the preoperative diagnosis and treatment strategy, and the antero-posterior diameter of the spinal cord on the sagittal section measured by T1-weighted MRI would be even smaller than that measured by CT. In addition, the T2WI can clearly show the cord compression and signal change of the spinal cord.

### ***Diagnostic criteria of DASM***

At present, there is no literature to standardize the diagnostic criteria of DASM. We contemplate that the diagnosis of DASM is mainly based on cervical CT scan, MRI examinations, and clinical symptoms, in which case the following aspects must be taken into account: (I) the SAC is less than 10 mm; (II) MRI reveals spinal cord compression and signal changes, or even spinal cord edema; (cervical CT myelography is useful to show the location and severity of spinal cord compression if the patient is unable to obtain an MRI image); (III) clinical manifestations of spinal cord injury, which are primarily referable to a loss of sensory or motor function in limbs with positive pyramidal sign and pain with limitation of movement in the cervical spine; (IV) cases in which symptoms and signs cannot be explained by intraspinal compression in other locations such as the lower cervical and thoracic spine, or other central nervous system disorders.

### ***Surgical options and outcomes of DASM***

The principles of treatment for DASM are to prevent sudden death from neurological compromise and improve

neurological status and the quality of life. Wang *et al.* (7) recommended the appropriate surgical treatment for development spinal canal stenosis at atlas should be selected according to the pathologies, clinical manifestations, and imaging findings. They devised four primary surgical modalities, including posterior arch osteotomy, posterior arch resection and replantation, occipital cervical fixation and fusion, and odontoid reduction and atlantoaxial fixation by a transoral approach. Kim *et al.* (43) introduced a technique of C1 double-door laminoplasty augmented with an allograft spacer and a titanium miniplate, which allows bone grafting, decompression, and fusion to be performed without disruption of the C1 posterior arch. However, the use of internal fixation increases the cost and the risk of implant loosening and requires sacrificing the function of craniocervical and cervical motion. Hott *et al.* (44) noted that posterior C1–2 fusion can lead to more than a 50% rotational restriction of the upper cervical spine.

Indeed, an increasing number of reports have suggested a favorable outcome could be achieved by decompression alone for DASM without C1–2 instability and atlantoaxial dislocation. Kawabori *et al.* (18) reported a case of unique C1 posterior tubercle impingement and myelopathy caused by DISH, in which the patient underwent laminectomy from C1–3, and the myelocompression and myelopathy were improved after operative intervention. Overall, resection of the PAA without fixation and fusion is the most common surgical procedure according to the published literature.

It is worth mentioning that DASM in elderly patients often complicates with subaxial cervical stenosis, which present difficulties for precision diagnosis and a surgical plan. Although major surgeries, such as laminoplasty or laminectomy from C1 to C7, can complete spinal cord decompression in a single procedure, the surgical trauma and risk of complications should not be underestimated. As indicated by Yamahata *et al.* (45) the pathophysiology of spinal canal stenosis should be considered separately at the C1 and the subaxial cervical spine, and staged surgery is our coping strategy for this kind of situation if the duration of surgery is longer than 3 hours. The segment with the most severe spinal cord compression should be treated firstly by minimally invasive techniques, and a second surgery should be performed at least 6 months after the first if required. The benefit of staged surgery is that it can decrease anesthesia and operating time and reduce the risk of complications. In this study, all three patients obtained satisfactory results through a single operation, avoiding a second.

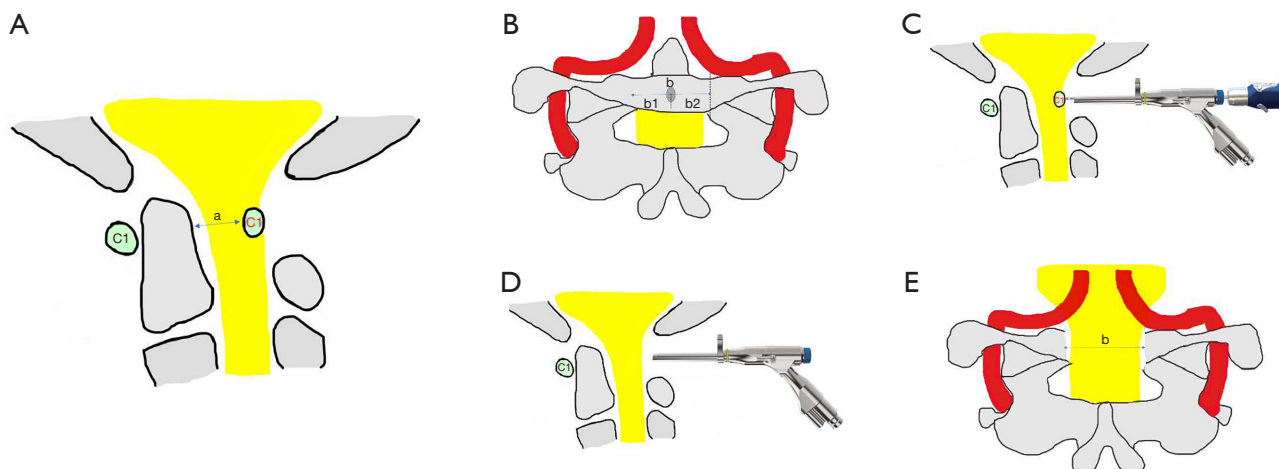
Although conventional C1L is the most common

procedure for DASM, it has the disadvantage of soft tissue damage and massive hemorrhage as an open operation. Notani *et al.* (35) reported a case of dynamic paraspinal muscle impingement after C1L which led to postoperative neurological deficits and eventually required a revision surgery of occiput to C3 fixation with instrumentation. In addition, Yeom *et al.* (46) reported extensive dissection and retraction for articular fusion may cause postoperative occipital neuralgia.

### *Feasibility of endoscopic surgery for DASM*

The surgical plan should also depend on the patient's comorbid conditions and bone quality. For frail elderly individuals comorbid with cardiovascular and cerebrovascular diseases, diabetes, and severe osteopetrosis, MIS can promote rapid recovery after surgery (47,48). In contrast to conventional open procedures, endoscopic spine surgery does not require extensive soft-tissue stripping, resulting in minimal occurrence of postoperative muscle impingement (49). It also has the advantages of fewer incisions, less pain, less bleeding, and less scarring (49,50).

In the cases reported in the present study, full endoscopic spine surgery was confirmed to be an effective method for treating DASM. However, the following details need to be brought to attention. The anatomical position of the atlas is deep and adjacent to several delicate vital organs such as the spinal cord, VA, and nerve roots. Therefore, how to establish the working channel on the surface of the PAA without bringing complications is the first key factor to consider. For DASM, the risk of VA injury is relatively high due to anatomical variations. Careful review of pre-procedural radiological imaging should be performed by the surgeon before planning the procedure, and the CTA superimposed on a CT three-dimensional reconstruction is especially helpful to estimating abnormalities of the VA. The posterior tubercle is an important anatomical landmark which can be taken as the starting point from where to remove the PAA to both sides until there is no compression on the spinal cord (*Figure 7*). According to an anatomical study from Cacciola *et al.* (51) the distance between the most medial edge of the VA groove over the PAA and the posterior tubercle of atlas vertebrae is about 14.6 mm. Consequently, removal of the PAA on each side of the posterior tubercle should not be extended beyond this span. Garrido *et al.* (52) also advocated that the posterior aspect of the PAA should not be exposed over 15 mm. In our experience, resection about 9 to 10 mm of the PAA from the midline is substantial enough to provide



**Figure 7** Schematic representation of the DASM and PFEC1L. (A) “a” represents SAC. It often requires operative intervention when “a” is less than 10-mm. (B) “b” represents the posterior tubercle of the PAA, which is the important anatomical landmark for PFEC1L. Taking “b” as a starting point to remove the PAA to both sides ( $b_1=b_2=1$  cm) and achieve adequate decompression of the spinal cord. (C) The working channel must be established precisely close to the PAA and is a prerequisite for successful PFEC1L. (D,E) After decompression, normal morphology of the spinal cord could be observed endoscopically. DASM, developmental atlantal stenosis with myelopathy; PFEC1L, percutaneous full endoscopic C1 laminectomy; SAC, space available for the cord; PAA, posterior arch of the atlas.

adequate decompression.

#### *Advantages of robot assistance*

Robot-assisted orthopaedic surgery has been proven successful for accurate surgical planning in pedicle screw placement and spine tumor surgery (53-56), and recent literature suggests incorporating robotic guidance in MISS provides a high degree of accuracy and reduction of radiation exposure (53,57,58). The challenging anatomy of the C1 mini lamina demands highly precise surgical maneuvers (59,60), especially for the percutaneous full endoscopic technique. With the help of robotic precise planning and navigation, the Kirschner wire and working sleeve can be quickly and safely anchored at the midpoint of the posterior tubercle of the atlas, and the anchor point left by the wire can also be used as an important reference for recognizing endoscopic anatomical boundaries.

#### *Indications and limitations of PFEC1L*

It is mainly applicable to the stenosis of the atlas canal caused by the posterior arch deformity of the atlas in a single segment. It is not suitable for the lesions of multiple segments, and the instability or dislocation of the atlas. For PFEC1L operation, surgeons are required to have skilled

experience in cervical endoscopy, and carefully identify the variant posterior arch of atlas to prevent spinal cord injury.

#### *Strengths and limitations of this study*

This study is the first to present the new surgery method of PFEC1L for the treatment of DASM and summarizes the relevant literature to discuss the pathological mechanism, diagnosis, and treatment strategy for this condition (*Table 1*). However, the limitation of our study is its low number of cases without a control group.

#### **Conclusions**

In summary, DASM is rare but potentially dangerous. Its diagnosis is made mainly based on clinical manifestations combined with radiological imaging examinations, especially CT scan and MRI. While C1L is the most common surgical method, PFEC1L is a new therapeutic option that has the advantage of being minimally-invasive, and the robot-assisted system could improve the accuracy of the working-channel placement.

#### **Acknowledgments**

*Funding:* This work was supported by the National Natural

**Table 1** Summary of adult patients with DASM reported in the literature

No.	References	Sex	Age (years)	MSCD (mm)	Symptoms	Therapy	Using fixation	Results
1	Tokiyoshi <i>et al.</i> (11)	M	55	8	Gait disturbance, numbness of the fingers, toes and the proximal parts of the limbs. Left shoulder pain after flexing the neck	C1L and dorsal opening of the foramen magnum	No	Normal gait and no sensory disturbance
2	Phan <i>et al.</i> (9)	M	80	8	Bilateral hand numbness and leg stiffness, urinary frequency	C1L and removal of the superior part of the lamina of C2	No	Postoperative superficial wound infection. However, the symptoms were partially improved
3	Phan <i>et al.</i> (9)	M	75	7	Weakness, numbness, and stiffness in extremities. Walking and initiating micturition difficulty	C1L	No	The symptoms improved remarkably
4	Hsu <i>et al.</i> (10)	M	38	6.23	Tingling sensations in the abdomen and perineum when flexing the neck, and numbness of both hands	C1L with duraplasty	No	The numbness and the abnormal tingling sensations improved
5	Kasliwal <i>et al.</i> (30)	F	26	–	Posterior cervical headaches with tingling and numbness involving right arm, trunk, and leg	Hemilaminectomy of the atlas preserving the C1/C2 joint	No	Patient had an uneventful postoperative course
6	Musha <i>et al.</i> (8)	F	50	8	Gait disturbance, numbness in the bilateral upper and lower limbs	C1L and occipito-cervical fusion (Occ-C2–3)	Yes	Numbness disappeared, loss of manual dexterity and spastic gait alleviated
7	Musha <i>et al.</i> (8)	M	75	9.5	Occipital and neck pain, numbness of both hands and feet, weakness of both legs and gait disturbance	C1L and Occ-C2–3 fusion, and expansive laminoplasty at C4–5	Yes	Motor function improved; urinary incontinence disappeared. sensory abnormalities significantly relieved
8	Kawabori <i>et al.</i> (18)	M	75	–	Numbness in both lower extremities, gait disturbance, disturbed precise motion of the hands, and urinary disturbance	Prophylactic posterior decompression between C1 and C3	No	Finger motion became smooth and the urinary disturbance disappeared, but dysesthesia still evident
9	Iki <i>et al.</i> (20)	M	81	9	Numbness and weakness of extremities and gait disturbance, needing a wheelchair	Laminectomy of C1 and partial C2	No	Neurological status improved 1 year postoperatively
10	Yunoki (37)	M	74	9	Gait disturbance, and clumsy hands	C1L	No	An uneventful postoperative course

**Table 1** (*continued*)

Table 1 (continued)

No.	References	Sex	Age (years)	MSCD (mm)	Symptoms	Therapy	Using fixation	Result
11	Tsuruta <i>et al.</i> (59)	F	79	8	Occipitalgia, gait impaired, requiring a cane	C1L	No	Occipitalgia disappeared, hemiparesis improved, able to walk without a cane
12	Bokhari <i>et al.</i> (60)	F	68	–	Walking and gait unbalanced, quadriparesis, weakness on the right side, hypoesthesia of the upper extremities	C1L	No	Able to walk with mild assistance 6 months postoperatively
13	Tang <i>et al.</i> (31)	F	58	5.5	Neck pain and limitation of neck rotation. numbness of all four limbs and disturbance of gait	C1L	No	Significant improvement in limb numbness and gait disturbance
14	Sawada <i>et al.</i> (3)	M	38	7	Right forearm and weakness of the right upper and both lower limbs, walking difficulty	C1L	No	Weakness and spasticity of the extremities were alleviated
15	Shah <i>et al.</i> (5)	F	44	6.36	Persistent neck pain for 6 weeks, which had gradually progressed to radiate into the right half of the body, associated with tingling and numbness in the right half of body	<i>En-bloc</i> wide excision of anomalous arch with occipito-cervical fusion	Yes	Excellent clinical outcome without any obvious complaints or disability 2 years later

DASM, developmental atlantal stenosis with myelopathy; MSCD, mid-sagittal spinal canal diameter; C1L, C1 laminectomy; SAC, space available for the cord.

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### Footnote

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**Ethical Statement:** The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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