

Thai J Neurological Surg 2026: 17(1): 46 - 54

รายงานผู้ป่วย/Case Report

Modified DCER technique for Basilar invagination and atlantoaxial dislocation in neurofibromatosis type 1: A Case Report

Sirawich Paokunta, Todsapon Praphanuwat, Chumpon Jetjumnong

Neurosurgery Unit, Department of surgery, Faculty of Medicine,
Chiang Mai University, Thailand

Abstract

Introduction: Spinal deformities are common in Neurofibromatosis Type 1 (NF-1), a rare autosomal dominant disorder, however, craniovertebral junction anomalies such as atlantoaxial dislocation (AAD) and basilar invagination (BI) are uncommon. Surgical management is indicated in symptomatic cases. Although combined anterior-posterior fixation remains the standard approach, the distraction, compression, and extension reduction (DCER) technique offers a safe and effective alternative in selected patients.

Case presentation: A 21-year-old woman with clinical features of Neurofibromatosis Type 1 (NF-1) presented with sudden-onset quadriparesis following a fall to the ground. Neuroimaging revealed atlantoaxial dislocation (AAD) and basilar invagination (BI). She was initially managed with cervical traction, followed by a single-stage posterior fixation and fusion using a modified distraction, compression, and extension reduction (DCER) technique. Postoperatively, spinal alignment and stability were restored, and the patient demonstrated marked neurological improvement.

Conclusion: Surgical management of atlantoaxial dislocation (AAD) and basilar invagination (BI) in Neurofibromatosis Type 1 (NF-1) is particularly challenging due to the presence of dystrophic anatomy. The posterior-only distraction, compression, and extension reduction (DCER) technique offers an effective, patient-specific approach for deformity correction and achieving optimal outcomes.

Keywords: Neurofibromatosis Type 1 (NF-1), Atlantoaxial Dislocation (AAD), Basilar Invagination (BI), DCER technique, Craniovertebral Junction

Received: November 17, 2025 | Revised: January 31, 2026 | Accepted: January 31, 2026

*Corresponding Author: Todsapon Praphanuwat

Email: todsapon.p@cmu.ac.th

Introduction

Neurofibromatosis type 1 (NF-1) is a rare autosomal dominant genetic disorder characterized by multiple café-au-lait macules, axillary or inguinal freckling, iris Lisch nodules, and pathogenic variants of the NF1 gene located on chromosome 17.¹ The prevalence of spinal abnormalities is significantly higher in individuals with NF-1 compared to the general population², most commonly including dural ectasia, scoliosis, and neuroforaminal tumors. However, cervical spine involvement is comparatively uncommon. Atlantoaxial dislocation (AAD) and basilar invagination (BI) are exceedingly rare manifestations in NF-1, with only ten cases reported in the literature between 1983 and 2019.³

The instability is typically congenital in origin. When patients present with neural compression or myelopathy, surgical correction remains the mainstay of treatment. According to current literature⁴, achieving sagittal realignment and near-anatomical stabilization through combined anterior and posterior approaches is considered the standard surgical procedure. Nevertheless, this strategy carries several drawbacks, including an increased risk of vertebral artery injury, cerebrospinal fluid leakage, and higher hospitalization costs. To address these challenges, Chandra et al. (2013) proposed the posterior-only distraction, compression, and extension reduction (DCER) technique, which has been shown to be a safe, effective, and reproducible alternative.⁵

Case presentation

A 21-year-old woman presented to the emergency department with sudden-onset quadriplegia following a fall in which the occiput struck the ground. Her medical history revealed a three-year history of gait disturbance that had not been previously investigated.

Physical examination revealed a conscious and oriented patient with generalized café au lait spots, multiple cutaneous neurofibroma, freckling at axillary and multiple iris Lisch nodules (Figure 1). These findings were compatible with NF-1, according to revised diagnostic criteria in 2021.¹ Motor examination showed 3 out of 5 scores muscle power of bilateral deltoid and bicep muscles; 4 out of 5 scores muscle power in the remaining muscles of upper extremities; and 2 out of 5 scores muscle power in the lower extremities. Deep tendon reflexes were hyperreflexia in both upper and lower extremities. Babinski signs were positive, with impairment of bowel and bladder function.

Computed tomography (CT) of the cervical spine revealed scoliosis, widening of the Condyle–C1 interval (CCI) (3.2 mm on the right and 3.1 mm on the left), and an increased atlantodental interval (ADI) of 7.5 mm between the anterior arch of the atlas and the odontoid process. The odontoid tip projected superiorly into the foramen magnum (Figure 2). Magnetic resonance imaging (MRI) demonstrated marked spinal canal narrowing with increased T2 signal intensity in the spinal cord at the craniocervical junction, consistent with traumatic myelopathy following an extension injury in the setting of preexisting basilar invagination. Diffuse dural ectasia with lateral neuroforaminal meningoceles involving the entire cervical spine and retroclival region resulted in pressure-induced remodeling of the posterior vertebral elements (Figure 3). A 3.4 × 2.2 × 6.1 cm infiltrative soft-tissue mass involving the dens of C2 was also identified, associated with widening of the ADI and inferior extension into the C2–C4 prevertebral and left cervical regions, forming a plexiform configuration. The lesion exhibited mild hyperintensity on both T1- and T2-weighted sequences, findings consis-

tent with a plexiform neurofibroma (**Figure 4**). Computed tomography angiography (CTA) of the neck showed no evidence of vascular injury, vascular malformation, or arteriovenous shunting.

The patient was advised to undergo surgical intervention to prevent further progression of myelopathy resulting from atlantoaxial instability and basilar invagination. Preoperative radiographs obtained after the application of 5 kilograms of Gardner-Wells skull traction (**Figure 5**) demonstrated partial reducibility of the deformity. Following informed consent, a single-stage posterior approach was performed. The patient was positioned prone using a Mayfield skull clamp to maintain the neck in a neutral position. A standard midline incision was made, and subperiosteal dissection was carried out to achieve wide exposure from the occiput to C4. Under the operating microscope, both atlantoaxial joints were fully exposed by dividing the C2 nerve roots. The joint spaces were temporarily distracted with a curette, after which approximately 8 × 8 × 8 mm autologous iliac bone grafts were packed into the bilateral spaces. This maneuver achieved substantial vertical reduction of basilar invagination and later served as a fulcrum for the compression maneuver. The occipital plate was then secured with screws. The Mayfield skull clamp was unlocked, and the patient's head

was stabilized by an assistant. Under continuous fluoroscopic guidance, the compression was applied between the occipital plate and the inferior aspect of the C2 spinous process in conjunction with a slight extension of the head. This compression–extension technique resulted in successful reduction of anterior atlantoaxial subluxation and eliminated spinal cord compression. A temporary rod was then placed to maintain craniovertebral alignment.⁵ Because bone remodeling from dural ectasia precluded placement of standard posterior cervical screws at C1 and C2, a right C3 pedicle screw and left C2/3 and C3/4 transfacet screws were inserted under image-guided navigation. Definitive rods were then contoured, placed, and secured. The final radiograph demonstrated the near complete correction of AAD and BI as well as cord compression (**Figure 6**). An autologous rib graft was harvested and applied to achieve posterior fusion from the occiput to the C2 lamina.

The postoperative period was uncomplicated, with steady neurological improvement. At one month, lower-limb strength had increased to 3/5–4/5, and upper-limb strength as well as bowel and bladder function had also improved. By the 1-year follow-up, she exhibited marked neurological recovery and was walking independently.



Figure 1. Numerous café-au-lait macules measuring approximately 0.5 to 5 cm in diameter are scattered over the trunk and extremities. Multiple soft, skin-colored to light-brown dome-shaped nodules are also observed on the trunk and upper limbs, consistent with cutaneous neurofibromas.

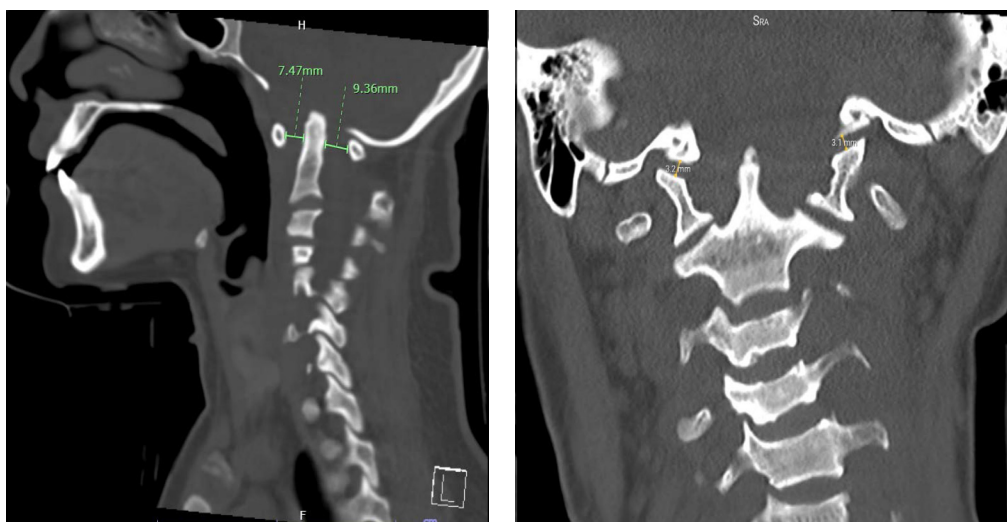


Figure 2. Computed tomography of the craniovertebral junction. (Left) Sagittal image showing an increased anterior atlantodental interval (ADI) of 7.5 mm with a posterior atlantodental interval (PADI) of 9.4 mm. (Right) Coronal image demonstrating widening of the condyle-C1 interval (CCI) measuring 3.2 mm on the right and 3.1 mm on the left.



Figure 3. Magnetic resonance imaging (MRI) of the cervical spine. (Left) Sagittal T1-weighted image and (Right) sagittal T2-weighted image demonstrate a short-segment intramedullary T2 hyperintensity extending from the craniocervical junction to the C2–C3 level, consistent with myelopathy. Dural ectasia is evident, causing remodeling and thinning of the posterior elements of the cervical vertebrae and the clivus.

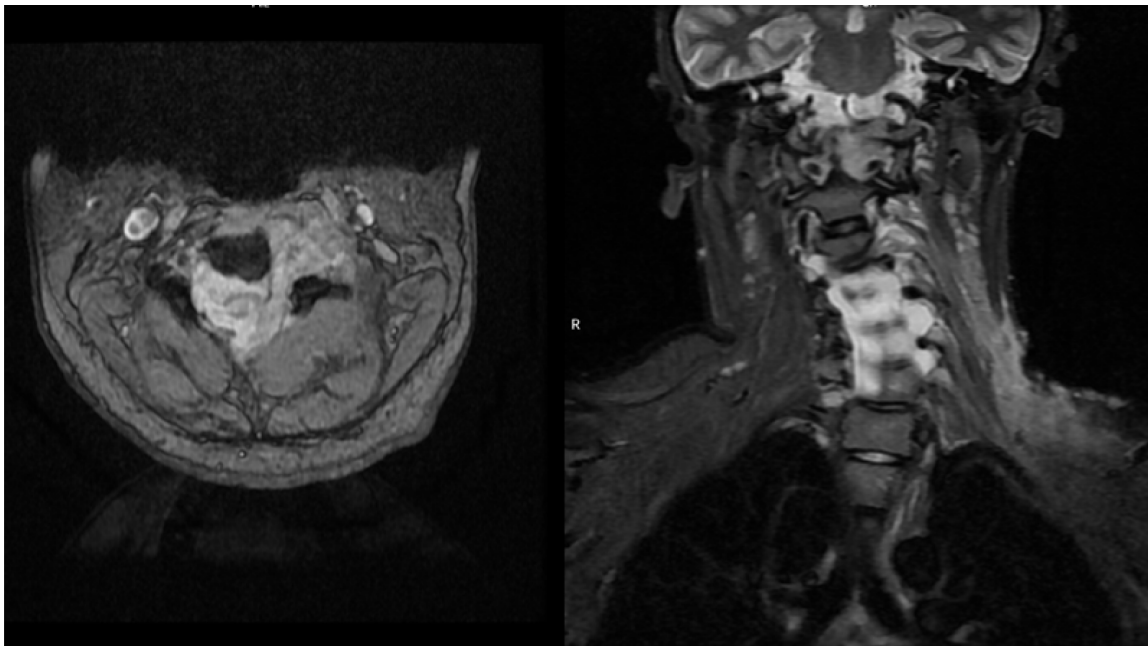


Figure 4. Magnetic resonance imaging (MRI) of the cervical spine. T2-weighted images: (Left) axial and (Right) coronal views demonstrate an infiltrative soft-tissue mass involving the dens of C2, extending inferiorly and laterally into the left cervical region, consistent with a plexiform neurofibroma.

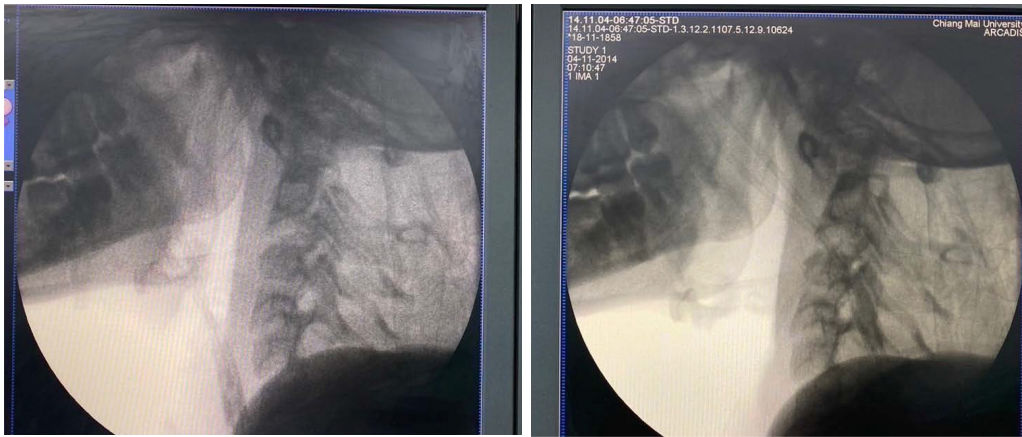


Figure 5. Preoperative application of Gardner-Wells skull traction. (Left) Traction with 5 kilograms and (Right) with 2 kilograms demonstrate partial reduction of the craniovertebral malalignment.

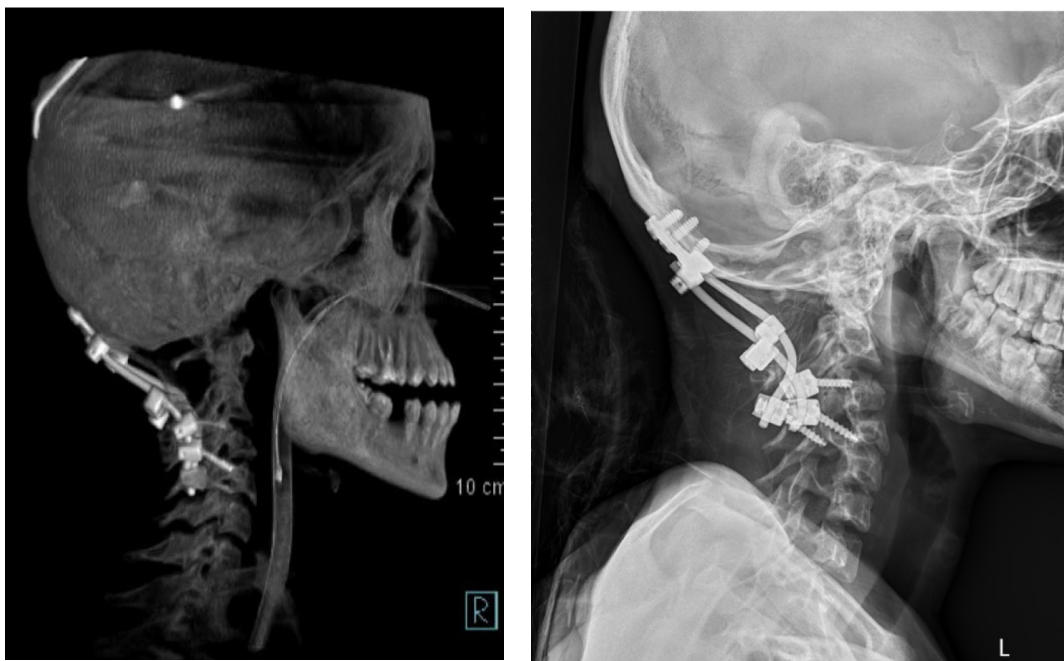


Figure 6. Computed tomography (CT) and lateral cervical spine radiograph. (Left) Intraoperative CT scan demonstrating reduction and realignment of the craniovertebral junction. (Right) Lateral cervical spine radiograph obtained 1 month postoperatively shows maintenance of correction of atlantoaxial dislocation (AAD) and basilar invagination (BI).

Table 1. Pre-and postoperative radiographic parameters showing improvement in craniovertebral alignment.

Index Measured	Preoperative value	Postoperative value
ADI	7.47 mm	
Chamberlain line	5.70 mm above the line	2.03 mm below the line
Wackenheim line	6.18 mm above the line	6.07 mm below the line
McRae line	0 mm below the line	11.23 mm below the line
Modified Ranawat line	20.84 mm	26.19 mm
Sagittal inclination	115°	103°
Coronal inclination	102°	127°
Craniocervical tilt	66°	67°

Discussion

Epidemiology and Rarity

To date, only 10 cases of atlantoaxial dislocation (AAD) and basilar invagination (BI) in patients with Neurofibromatosis Type 1 (NF-1) have been reported in the literature from 1983 to 2019³, highlighting the rarity of these craniovertebral junction anomalies in the context of NF-1. Due to the rarity of such cases, formulating consistent diagnostic and management protocols remains difficult.

Pathophysiology

Currently, the exact pathophysiology of craniovertebral abnormalities in NF-1 are complex and unclear. The mutation on chromosome 17 causes a defect in the NF-1 gene, resulting in problems with skeleton development.⁶ Patients often have underlying skeletal dysplasia and ligamentous laxity, both of which can contribute to instability at the craniovertebral junction. These structural abnormalities make the craniovertebral junction particularly susceptible to gradual deformity and sudden instability, even after minor trauma.

Radiological Criteria

Computed tomography (CT) and magnetic resonance imaging (MRI) are crucial in evaluating the alignment of cervical spine, the position of odontoid process, and any bony changes associated with dysplasia. The atlantodental interval and the relationship of the odontoid to Chamberlain's and McRae's lines are used to confirm diagnosis and guide treatment planning.

Treatment Principles

The previous treatment strategy of atlantoaxial dislocation (AAD) and basilar invagination (BI) consist of both non-operative treatment and surgical procedure for symptomatic AAD. In irreducible dislocation, the transoral odontoidectomy is indicated. The posterior surgical approach is used alone for reducible AAD or combined with anterior transoral decompression in irreducible cases.⁷

The management of AAD and BI in patients with neurofibromatosis type 1 (NF-1) is particularly challenging due to the presence of abnormal bone morphology, dural ectasia, and inherent ligamentous laxity, which predispose to instability and complicate fixation. The Distraction, Compression,

Extension, and Reduction (DCER) technique, as introduced by Chandra et al.⁸, represents a novel posterior-only surgical strategy that enables reduction of dislocation or invagination, decompression of the neural elements, and stabilization of the craniovertebral junction (CVJ) while minimizing surgical morbidity. In this patient, where the sagittal inclination of atlantoaxial joint was 115 degrees, a joint remodeling was performed as type II surgery of DCER.⁵

However, in syndromic or dysplastic conditions such as NF-1, conventional C1–C2 fixation may be precluded by fragile or malformed osseous structures. In such scenarios, a modified DCER can be employed, maintaining the same biomechanical principle of vector-based reduction while shifting the fixation anchors to the occiput and sub-axial vertebrae. Through controlled compression and extension, this modification achieves indirect odontoid descent and adequate realignment without the need for anterior decompression. Importantly, it preserves the fundamental advantages of the posterior-only approach while accommodating the complex and atypical craniovertebral anatomy characteristic of NF-1.

Outcome

The patient's neurological symptoms showed clinical improvement prior to hospital discharge. At long-term follow-up, the patient achieved a Nurick

grade of 1. This outcome is consistent with the findings reported by Chandra et al. (2013), in which the mean postoperative Nurick score at one year was 1.3 ± 0.53 .

Radiographic evaluation was performed only with an immediate intra-operative CT scan, which demonstrated improvement in most parameters except for the Modified Ranawat line. Due to the patient's dysmorphic anatomy, standard reference values for these parameters⁹ were not appropriate for comparison. Furthermore, the absence of follow-up imaging limited the assessment of postoperative bone fusion.

Limitation

This case report has some limitations. As a single-patient observation, the findings may not be generalizable to all patients with similar conditions. Long-term follow-up was limited, preventing clinical and radiographic evaluation of potential delayed outcomes.

Conclusion

Surgical management of AAD and BI in patients with NF-1 remains controversial and the surgical planning should be customized to each patient due to dystrophic vertebral anatomy. The DCER technique, a posterior only approach, is the useful surgical plan to correct the deformities and achieve desired clinical and radiographic outcomes.

References

1. Legius E, Messiaen L, Wolkenstein P, Pancza P, Avery RA, Berman Y, et al. Revised diagnostic criteria for neurofibromatosis type 1 and Legius syndrome: an international consensus recommendation. *Genet Med* 2021;23(8):1506–13. doi:10.1038/s41436-021-01170-5
2. Well L, Careddu A, Stark M, Farschtschi S, Bannas P, Adam G, et al. Phenotyping spinal abnormalities in patients with Neurofibromatosis type 1 using whole-body MRI. *Sci Rep*. 2021;11(1):16889.doi:10.1038/s41598-021-96310-x

3. Ding C, Guo Y, Wu T, Wang B, Huang K, He J, et al. Atlantoaxial Dislocation Associated with Type 1 Neurofibromatosis: Case Report and Review of the Literature. *World Neurosurg.* 2020;143:261-7. doi:10.1016/j.wneu.2020.07.176
4. Yang SY, Boniello AJ, Poorman CE, Chang AL, Wang S, Passias PG. A review of the diagnosis and treatment of atlantoaxial dislocations. *Global Spine J.* 2014;4(3):197-210. doi:10.1055/s-0034-1376371
5. Chandra PS, Bajaj J, Singh PK, Garg K, Agarwal D. Basilar Invagination and Atlantoaxial Dislocation: Reduction, Deformity Correction and Realignment Using the DCER (Distraction, Compression, Extension, and Reduction) Technique With Customized Instrumentation and Implants. *Neurospine.* 2019;16(2):231-50. doi:10.14245/ns.1938194.097
6. Kossler N, Stricker S, Rödelberger C, Robinson PN, Kim J, Dietrich C, et al. Neurofibromin (Nf1) is required for skeletal muscle development. *Hum Mol Genet.* 2011;20(14):2697-709. doi:10.1093/hmg/ddr149
7. Yang SY, Boniello AJ, Poorman CE, Chang AL, Wang S, Passias PG. A review of the diagnosis and treatment of atlantoaxial dislocations. *Global Spine J.* 2014;4(3):197-210. doi:10.1055/s-0034-1376371
8. Chandra PS, Kumar A, Chauhan A, Ansari A, Mishra NK, Sharma BS. Distraction, compression, and extension reduction of basilar invagination and atlantoaxial dislocation: a novel pilot technique. *Neurosurgery.* 2013;72(6):1040-53. doi:10.1227/NEU.0b013e31828bf342
9. Chandra PS, Prabhu M, Goyal N, Garg A, Chauhan A, Sharma BS. Distraction, Compression, Extension, and Reduction Combined with Joint Remodeling and Extra-articular Distraction: Description of 2 New Modifications for Its Application in Basilar Invagination and Atlantoaxial Dislocation: Prospective Study in 79 Cases. *Neurosurgery.* 2015;77(1):67-80. doi:10.1227/NEU.0000000000000737