Article published online: 2024-06-25



# Bow Hunter's Syndrome with Rotational Atlantoaxial Instability: A Rare Association

Malini S.[1] Anu C. Thomas[1] Sajeev S. Vadakkedam[1] Parameswaran K.[2] Anand M.[3]

[1] Department of Neurosurgery, Indo American Hospital, Institute of Brain and Spine, Vaikom, Kerala, India
[2] Department of Neurology, Indo American Hospital, Institute of Brain and Spine, Vaikom, Kerala, India
[3] Department of Interventional Neuroradiology, Indo American Hospital, Institute of Brain and Spine, Vaikom, Kerala, India

**Address for correspondence** Malini S., DrNB, Indo American Hospital, Institute of Brain and Spine, Chemmanakary, Vaikom, Kottayam, Kerala 686143, India (e-mail: drmalini87@gmail.com).

Asian J Neurosurg 2024;19:572–575.

## Abstract

Bow Hunter's syndrome (BHS) is a very rare condition in which there is rotational vertebral artery (VA) insufficiency. The association of BHS with rotational atlantoaxial instability is extremely rare. We are reporting a case of pediatric BHS who presented with features of VA insufficiency on neck rotation. Careful evaluation revealed rotational C1-C2 instability. Provocative digital subtraction angiography and dynamic neck computed tomography were the mainstay of our diagnostic armamentarium. Our case emphasizes the fact that VA abnormalities need special consideration in young patients with craniovertebral junction instability and a high degree of suspicion is necessary in most instances for accurate diagnosis.

**Keywords**
- Bow Hunter's syndrome
- atlantoaxial rotatory subluxation
- C1-C2 fusion

## Introduction

Rotational vertebral artery (VA) syndrome or Bow Hunter's syndrome (BHS) is an uncommon cause of vertebrobasilar insufficiency. It results from mechanical compression of the VA during rotation/flexion/hyperextension of the head and neck. The compression can be caused by various factors including cervical osteophytes/bone spurs, trauma, disk herniation, spondylotic changes, and musculotendinous compression.[1] The condition was first reported in 1978 by B. F. Sorensen in an archer who developed modified Wallenberg syndrome during archery practice.[2]

Our case is unique in that BHS in association with atlantoaxial rotatory subluxation (AARS) is very rare and to the author's best knowledge only approximately three other cases have been reported so far.[3]

## Case Presentation

A 14-year-old girl, while practicing yoga, "Chakrasana" bend her body backward almost 180 degrees and on trying to get back up, had sudden loss of tone and became unconscious and fell down hitting the back of her head on the ground (►**Fig. 1**). She recovered after sometime. The patient sustained occipital bone fracture and interhemispheric subdural hematoma. She was admitted and treated at another hospital and discharged after 2 days. She presented to our institute for a second opinion regarding further care and prevention of such events in the future since she is a classical dancer and wished to continue her dancing practice.

A thorough analysis of history revealed loss of tone and consciousness after extreme hyperextension/rotation of neck. No neurological deficits were found during clinical

**article published online**
June 25, 2024

**DOI** https://doi.org/10.1055/s-0044-1787865.
**ISSN** 2248-9614.



Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India


**Fig. 1** Chakrasana yoga pose.

examination. There was also history of episodes of dizziness on turning head to the right side since the past several months. So we suspected rotational VA compression syndrome.

Magnetic resonance imaging (MRI) brain and MR angiography were normal (►**Fig. 2A, B**).

In view of high clinical suspicion, we did dynamic digital subtraction angiography (DSA) that is a provocative test for diagnosing BHS. There was near-complete occlusion of left VA on rotation to contralateral side and refilling while straightening the neck (►**Fig. 3A, B**).

There was also flow restriction to a lesser degree on the right side (►**Fig. 4A, B**) on contralateral head rotation.

Computed tomography (CT) cervical spine in neutral position and flexion-extension were normal. However, there was near total rotational subluxation of C1-C2 on rotation to either side (►**Figs. 5**–**7**).

In view of symptomatic vertebrobasilar insufficiency and presence of AARS, surgical intervention was decided upon. She underwent posterior cervical stabilization with C1 lateral mass and C2 pedicle screws and rods and fusion with rib

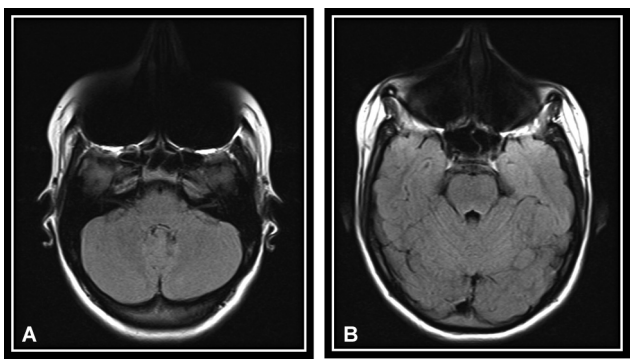


**Fig. 2** (**A**, **B**) Magnetic resonance imaging (MRI) brain axial sections showing no significant abnormalities.

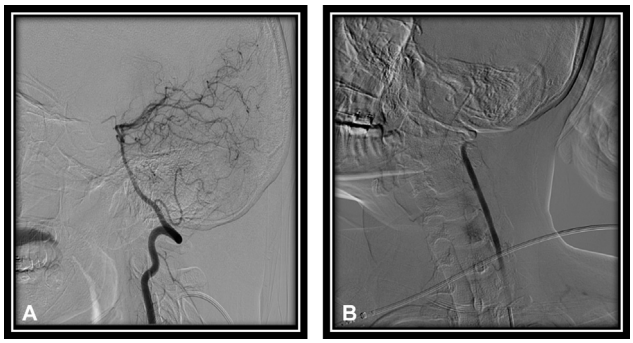


**Fig. 3** (**A**, **B**) Left vertebral artery (VA) filling in neutral position and complete occlusion at C1-C2 level on head rotation to the right side.

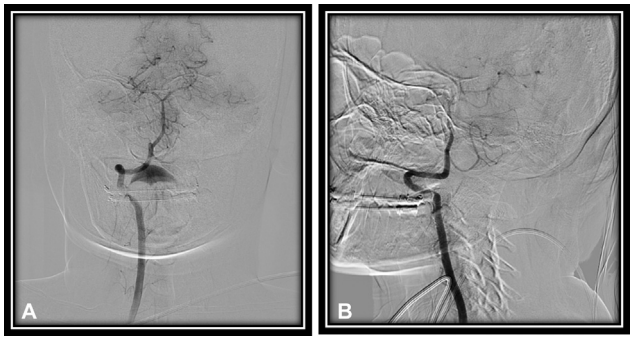


**Fig. 4** (**A**, **B**) Right vertebral artery (VA) filling in neutral position and reduced flow on head rotation to the left side.

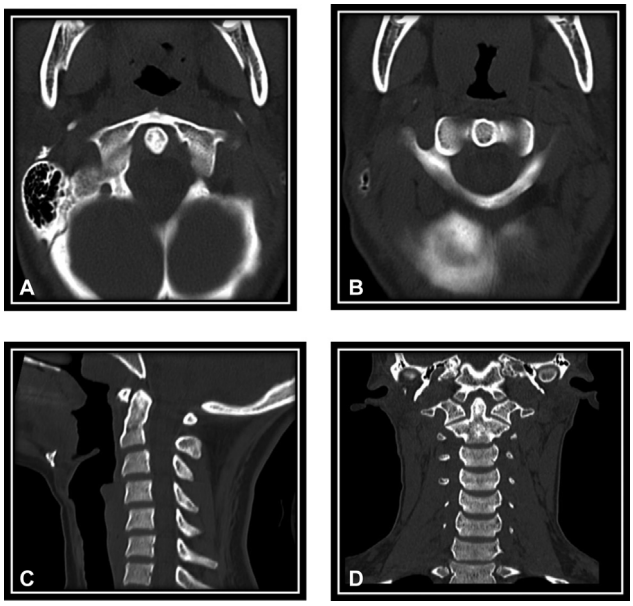


**Fig. 5** (**A**–**D**) Computed tomography (CT) cervical spine in neutral position was essentially normal.

grafting under neuromonitoring and neuronavigation guidance. The rotational C1-C2 subluxation was reduced (►**Fig. 8**).

She was discharged on eighth postoperative day. The patient is doing well during 1-year follow-up with complete relief of symptoms. She has resumed schooling and rejoined her dancing classes.

 

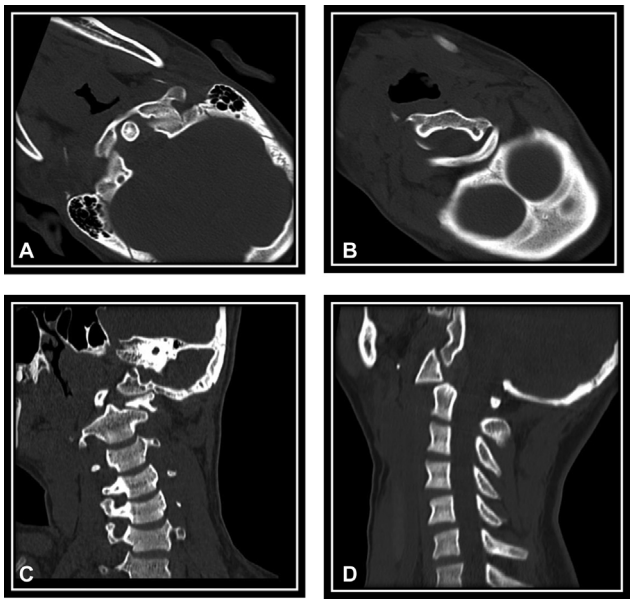


**Fig. 6** (A–D) Computed tomography (CT) C-spine in 45 degrees right lateral rotation axial, coronal, and sagittal images showing significant anterior translation of right C1-C2 joint.

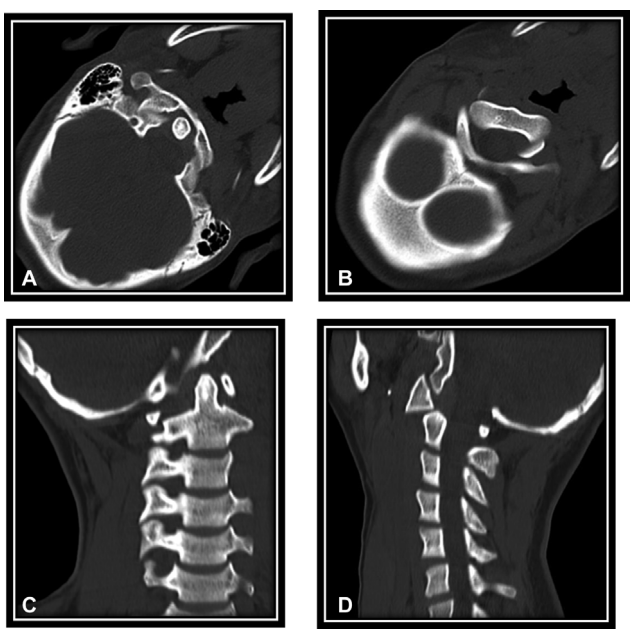


**Fig. 7** (A–D) Computed tomography (CT) C-spine in 45 degrees left lateral rotation axial, coronal, and sagittal images showing left C1-C2 subluxation.

## Discussion

BHS is a rare and potentially life-threatening condition in which there is VA insufficiency during rotation of the head within normal physiologic range. Reports of BHS in association with AARS are quite rare in literature, amounting to only approximately three cases or so, considering the literature evidence over the past three to four decades.[3,4] It is caused by mechanical compression of VA mostly in adults in fifth to seventh decades of life.[3]

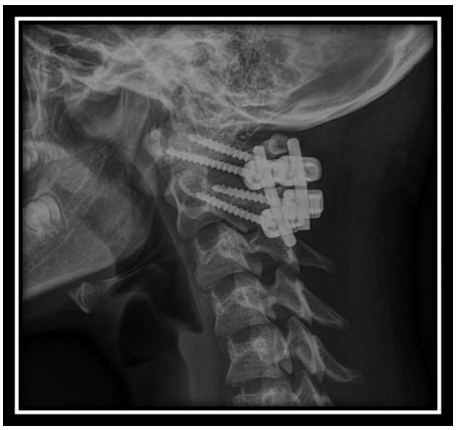

**Fig. 8** Postop image showing C1-C2 fusion.

Patients present with vertigo, nystagmus, and nausea with associated emesis, and ataxia during head rotation which is relieved in neutral position. Horner's syndrome, blurring of vision, syncope, drop attacks, and rarely motor or sensory deficits have also been reported.[2,5]

BHS can be classified into primary or acquired.[2] Primary BHS occurs due to bone spurs, osteophytes, disc herniation, spondylotic changes, hypertrophied ligaments, neck muscle hypertrophy, and instability. Acquired BHS occurs as a complication of surgery or trauma. Also, BHS was classified into three types by Cornelius et al—atlantoaxial, subaxial, and mixed type.[6] The causes for compression at the atlantoaxial level are atlas assimilation, ossified or thickened atlanto-occipital membrane, facetal hypertrophy, dural fold in the foramen magnum, or rheumatoid arthritis. In the subaxial level, compression can be due to bony spur or herniated disc. In mixed type compression can be at multiple levels and/or bilateral.

Dynamic or provocative DSA is the gold-standard diagnostic modality for suspected BHS. It is superior to other modalities by virtue of time factor (shorter duration of imaging), lesser radiation (as compared with CT angiography [CTA]), and patient safety (patient herself can control the degree of neck rotation). The effective radiation dose for CTA as a static study itself is more than DSA for cervicocerebral vessels.[7] Since patients usually become symptomatic with contralateral rotation of head causing compression of ipsilateral VA and the symptoms disappear in neutral position, traditional imaging of the head and neck usually fails to diagnose this condition. Hence, provocative DSA is of utmost importance in the diagnosis of rotational VA stenosis.

There are no strict guidelines regarding management of BHS since these cases are a rarity. The decision regarding management should be made after considering individual patient factors, severity of symptoms, and pathologic cause. Conservative management with anticoagulation, cervical collar or brace, as well as surgical vascular decompression and recently endovascular procedures including coil embolization of VA[8,9] has been described. Surgical management is adopted in case of bony spurs, herniated disc, and instability.

AARS is defined as a condition in which there is gross departure from normal rotational relationship between atlas and axis. The atlantoaxial motion segment is the most flexible

 

motion segment in the entire spine with regard to axial rotation. It allows for a bilateral range of motion of 80 degrees or more. Kinematics of C1 and C2 rotation can be understood in three distinct phases—single motion phase of first 23 degrees in which only C1 moves; double motion phase from 23 to 65 degrees in which both C1 and C2 rotate; and beyond 65 degrees head turns exclusively at subaxial spine with no rotation at C1-C2.[10] Rotational instability of C1-C2 joint can occur due to various reasons including trauma, infection, previous head and neck surgery, and conditions causing ligamentous laxity like Ehlers-Danlos syndrome, Marfan syndrome, Down syndrome, Morquio disease, rheumatoid arthritis, and congenital cervical anomalies.

The diagnostic imaging study of choice of AARS is dynamic CT with three-dimensional reconstructions. The C1-C2 angles (obtained by algebraically subtracting C2 angle from C1 angle) measured with CT imaging can be compared with normal C1-C2 rotation motion curve.[10] The type of subluxation can be assessed using grading systems like Fielding and Hawkins[11] or White and Punjabi.[12] MRI spine helps to assess ligamentous integrity. The treatment of AARS is based on type of subluxation and duration of symptoms. Chronic symptoms (more than 3 months) or failure of conservative methods like cervical collar, halter skeletal traction, or halo immobilization are indications for surgical intervention. Irreducible deformities also require surgical correction in the form of posterior C1-C2 fusion. The prognosis is usually good if the condition is identified early and treated promptly.

## Conclusion

BHS is a rare disorder resulting in rotational VA stenosis/occlusion, and even more rare to be associated with rotational instability at craniovertebral junction (CVJ). Our case reemphasizes the importance of ruling out VA anomalies/dissection/stenosis that may be associated with CVJ instability, especially in younger age group in which the risk of missing subtle neurological symptoms and signs is very high.

### Note

Bow Hunter's syndrome is a disorder resulting in rotational vertebral artery insufficiency. It is a rare but dangerous entity that in some cases may cause profound neurological deficits and may turn fatal if not recognized and treated promptly.

### Conflict of Interest

None declared.

## References


1 Choi K-D, Choi J-H, Kim J-S, et al. Rotational vertebral artery occlusion: mechanisms and long-term outcome. Stroke 2013;44 (07):1817–1824
2 Duan G, Xu J, Shi J, Cao Y. Advances in the pathogenesis, diagnosis and treatment of Bow Hunter's syndrome: a comprehensive review of the literature. Intervent Neurol 2016;5(1-2):29–38
3 Schulz R, Donoso R, Weissman K. Rotational vertebral artery occlusion ("Bow Hunter's syndrome"). Eur Spine J 2021;30(01): 1–12
4 Bando K, Okazaki T, Mure H, Korai M, Takagi Y. A juvenile case of Bow Hunter's syndrome caused by atlantoaxial dislocation with vertebral artery dissecting aneurysm. World Neurosurg 2020; 137:393–397
5 Rastogi V, Rawls A, Moore O, et al. Rare etiology of Bow Hunter's syndrome and systematic review of literature. J Vasc Interv Neurol 2015;8(03):7–16
6 Cornelius JF, George B, N'dri Oka D, Spiriev T, Steiger HJ, Hänggi D. Bow-hunter's syndrome caused by dynamic vertebral artery stenosis at the cranio-cervical junction–a management algorithm based on a systematic review and a clinical series. Neurosurg Rev 2012;35(01):127–135, discussion 135
7 Manninen A-L, Isokangas J-M, Karttunen A, Siniluoto T, Nieminen MT. A comparison of radiation exposure between diagnostic CTA and DSA examinations of cerebral and cervicocerebral vessels. AJNR Am J Neuroradiol 2012;33(11):2038–2042
8 Strickland BA, Pham MH, Bakhsheshian J, Russin JJ, Mack WJ, Acosta FL. Bow Hunter's syndrome: surgical management (video) and review of the literature. World Neurosurg 2017;103:953. e7–953.e12
9 Sugiu K, Agari T, Tokunaga K, Nishida A, Date I. Endovascular treatment for bow hunter's syndrome: case report. Minim Invasive Neurosurg 2009;52(04):193–195
10 Pang D. Atlantoaxial rotatory fixation. Neurosurgery 2010;66 (03):161–183
11 Fielding JW, Hawkins RJ. Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg Am 1977;59(01):37–44
12 White AA, Panjabi MM. Clinical Biomechanics of the Spine. 2nd ed. Philadelphia: JB Lippincott; 1990


 