



Dynamic imaging of the craniovertebral junction is mandatory in patients with posterior circulation strokes

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Abstract

Introduction The course of the vertebral artery after exiting from the C1 foramen transversarium and prior to entering the dura lends itself to compression in C1–2 instability. However, atlantoaxial dislocation presenting with vertebrobasilar insufficiency and posterior circulation stroke (PCS) is rare.

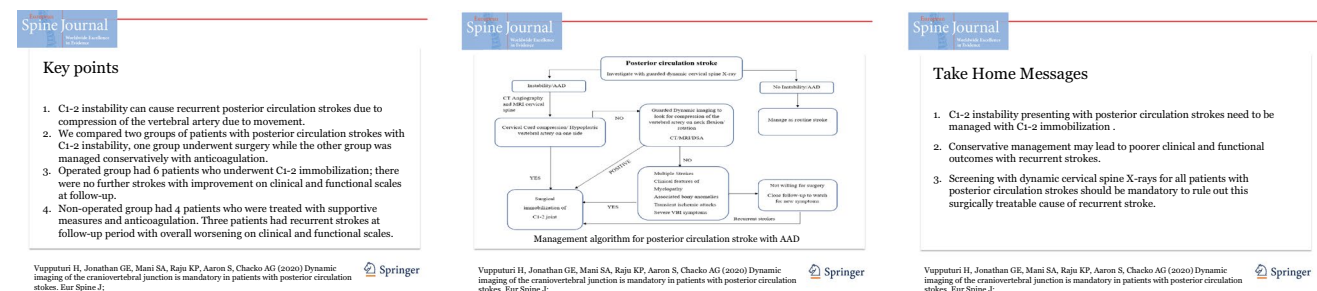
Methods In this retrospective study, we identified 96 patients with PCS who had complete radiological data. Ten (10.4%) patients had craniovertebral junction (CVJ) anomalies, of which six underwent surgery and four were managed conservatively. The clinical and functional outcomes were measured in the two groups.

Results Left-sided strokes were seen in 7/10 patients, the majority of whom had left dominant vertebral arteries. The mean age at presentation in those with CVJ anomalies was 27.2 ± 12.8 years that was significantly lower than those without CVJ anomalies, 52.2 ± 14.5 years ($p \leq 0.001$). The etiologies of PCS in those < 50 years were CVJ anomalies (30%), atherosclerosis (30%) and vasculitis (27%); however, the overwhelming majority of strokes in the > 50 year age group was atherosclerosis (91%). Postoperatively, there were no recurrent strokes in the operated patients, who also obtained significant clinical improvement on the modified Rankin Scale, Nurick Scale and modified McCormick Scale as compared to those who did not undergo surgery.

Conclusion Early diagnosis and surgical treatment of CVJ instability prevent recurrent strokes and improve outcomes in patients with PCS. Physicians and spine surgeons need to be sensitized regarding CVJ anomalies as a cause of PCS enabling early diagnosis with dynamic imaging particularly in the younger age group.

Graphic abstract

These slides can be retrieved under Electronic Supplementary Material.



Keywords Bow hunter's stroke · Posterior circulation strokes · Vertebral artery · Atlantoaxial dislocation · C1–2 fusion

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Extended author information available on the last page of the article

Introduction

Bow hunter's strokes, first described in archers, occur due to intermittent vertebral artery occlusion from extreme head rotation [1]. The tortuous horizontal portion of the V3 segment of the vertebral artery, between its exit from the C1 foramen transversarium and its dural entry at the posterior atlantooccipital membrane, makes it susceptible to dynamic compression [2]. In the presence of unilateral vertebral artery hypoplasia, head rotation to the ipsilateral side stretches and compromises the contralateral dominant artery and might result in a posterior circulation stroke (PCS) [3, 4]. Bony craniovertebral junction (CVJ) anomalies are known to compound this stroke risk as the added dynamic instability results in repeated intimal stress and thrombogenesis in the vertebral artery [5, 6]. Cerebral angiograms have confirmed vertebral artery occlusion with a shortened and stretched V3 segment of the dominant vertebral artery—the so called stretched loop sign during head rotation [7]. Although there are some data that surgical stabilization of the CVJ prevents recurrent strokes, conclusive proof in this regard is lacking [4, 8]. In this paper, we present ten patients with C1–2 instability who presented with PCS, six of whom underwent CVJ stabilization and four who were managed with anticoagulation alone.

Materials and methods

Between January 2012 and December 2018, 477 patients with posterior circulation stroke (PCS) presented to the Neurology Stroke Unit. All patients had an MR stroke protocol that identified the territory of the stroke, but only those who had neck pain, young patients <40 years of age, those who had recurrent strokes and if vasculitis was suspected

had a 3D-CT angiogram of the neck and head. Thus, 96 patients had complete radiological data including MR imaging and 3D-CT angiograms and were included in the study. Ten patients: six with acute strokes and four with recurrent strokes, were found to have craniovertebral junction anomalies leading to dynamic imaging with CT angiography/MRI angiography (Fig. 1). The six patients who presented to the Neurology Stroke Unit with an acute stroke were admitted for anticoagulation and supportive care. Following the acute treatment, they were discharged on a cervical collar with anticoagulation and were readmitted for surgery after 3 months. The four patients who had recurrent strokes were also started on anticoagulation, immobilized with a cervical collar and offered surgery directly. For those patients undergoing surgery, anticoagulation was stopped a week prior to the surgery. The type of surgery was decided on a case-to-case basis.

Clinical data were collected from the in-patient records. Radiological images were reviewed by a neuroradiologist on our picture archiving and communication system (GE-PACS V3.0). Atlantodental interval (ADI), clival canal angle (CCA) and Powers ratio were determined on the CT scans preoperatively [9]. Atlantoaxial dislocation (AAD) was classified based on a system using the Cartesian coordinates that takes into consideration the anteroposterior dislocation, rotational components and coronal tilt [10]. Adequate reduction was defined as ADI < 3 mm in adults and < 4.5 mm in children, CCA of > 150° and a Powers ratio < 1.0 [11].

The clinical outcome measures were number of strokes and interval between the strokes. The clinical outcome measures were number of strokes, interval between the strokes, change in modified Rankin scale (MRS) [12] and Patient Global Impression of Change (PGIC) scale [13]. Functional status was assessed by change in imNurick grade [14] and modified McCormick Scale [15] pre- and postoperatively.

Fig. 1 Case 3: 29-year-old man with an acute infarction of the right ventral-superior pons with a Type 1 AAD. **a** The magnetic resonance angiography (MRA) shows a dominant vertebral artery with a hypoplastic right vertebral artery (white arrow). **b** The MRA in flexion shows almost complete obliteration of the right vertebral artery (white arrows)

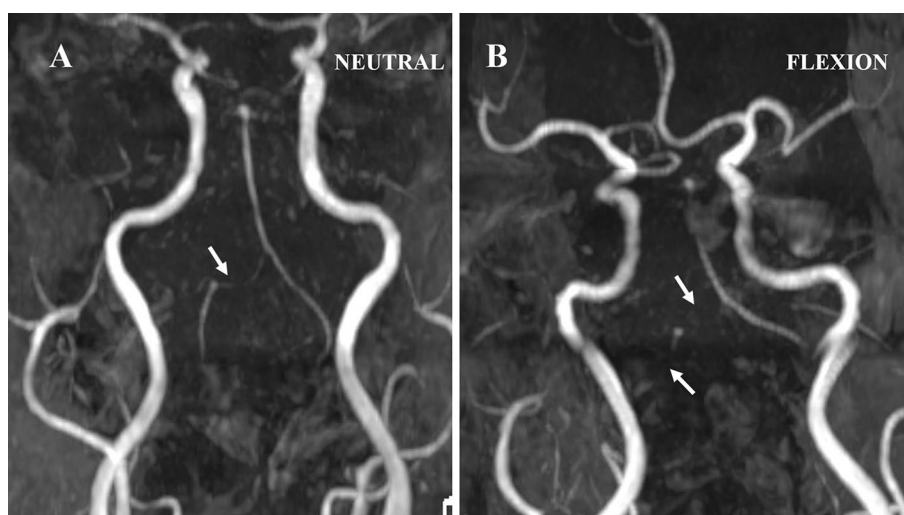


Table 1 Age-stratified etiology of posterior circulation stroke

Etiology	Age ≤ 50 years (n = 30)	Age > 50 years (n = 66)
Atherosclerosis	9 (30%)	60 (90.9%)
CVJ instability	9 (30%)	1 (1.5%)
Vasculitis	8 (26%)	2 (3%)
Vertebral artery dissection	2 (6%)	2 (3%)
Unilateral vertebral artery hypoplasia without CVJ instability	2 (6%)	1 (1.5%)

In addition, we assessed the neck disability index (NDI) in those undergoing surgery [16].

Four patients: two with acute strokes and two with recurrent strokes, refused surgery and were managed with a cervical collar, anticoagulation and physiotherapy. All patients were frequently followed up and their INR values titrated to therapeutic ranges. In the operated group, six patients, the anticoagulants were gradually tapered and stopped after 3 months. To determine the incidence of PCS in patients with CVJ anomalies, we searched our database for all CVJ anomalies undergoing surgery during the study period.

Statistical analysis was done by using SPSS software version 16.0 (SPSS Inc, Chicago IL; version 16.0). Fisher exact test, Mann–Whitney test and paired Student's *t* test were used for statistical analysis considering a *P* value of < 0.05 as significant.

This study was approved by the institutional ethics committee.

Results

Posterior circulation strokes

Of the 96 patients included in the study, 66 (68%) were > 50 years of age and 30 (32%) were ≤ 50 years of age. A left dominant vertebral artery was seen in 72 of the 96 patients (75%) in our series. Of the ten patients with PCS, seven were on the left side. The age-stratified distribution of causes of stroke is described in Table 1. CVJ anomalies causing PCS were significantly more common in the age group < 50 years (*p* < 0.001). During the same study period, a total of 153 cases of CVJ anomalies were treated surgically in our unit none of whom had prior PCS. Thus, the overall incidence of PCS in patients with CVJ anomalies was 10/163 (6.13%).

Atlantoaxial dislocation

There were ten patients (eight males and two females; M:F ratio = 4:1) with AAD, none of whom had a prior diagnosis of a CVJ anomaly. Table 2 summarizes the demographic

Table 2 Clinical and demographic profile of patients including the incidence of associated anomalies and the type of CVJ anomaly

	Age/sex	Vessel involved	VA dominance	No of strokes	Duration of symptoms (months)	Associated anomalies	Mean stroke interval (months)	Myelopathy	Neck pain	Type of CVJ anomaly
1	25/F	L-VA	Left	2	6	C1 complete assimilation, C2–3 Klippel–Feil	3	Yes	Yes	3
2	52/M	BA	Right	1	3	C1 incomplete assimilation, C2–3 Klippel–Feil, right vertebral artery stenosis	–	Yes	No	5
3	29/M	L-VA	Left	2	2	None	1	Yes	Yes	1
4	11/M	L-VA	Left	2	12	None	6	Yes	Yes	5
5	7/F	L-VA	Left	2	8	Right vertebral artery hypoplasia	4	Yes	No	5
6	18/M	L-VA	Left	2	24	C1 complete assimilation, C2–3/C4–5 Klippel–Feil	8	Yes	Yes	3
7 ⁺	30/M	R-SCA, B/L PICA	Right	3	6	C1 partial assimilation	2	Yes	No	3
8 ⁺	34/M	R-PICA	Left	1	18	None	–	Yes	No	1
9 ⁺	24/M	L-VA	Left	3	6	C2–3 Klippel–Feil	1.5	Yes	Yes	1
10 ⁺	42/M	Left PCA	Right	1	12	Left vertebral artery hypoplasia	–	No	No	3

M Male, F female, R right, L left, PCA posterior cerebral artery, AICA anterior inferior cerebellar artery, PICA posterior inferior cerebellar artery, SCA superior cerebellar artery, VA vertebral artery, BA basilar artery

⁺Indicates conservatively managed patients

and clinical profile of these ten patients. The mean age at presentation was $27.2 \text{ years} \pm 12.8$. The mean duration of symptoms was 9.7 ± 6.5 months. 9/10 (90%) had myelopathy at presentation. None of the patients had trauma. Only three patients had a single stroke, the remaining had 2 or more strokes prior to presentation, and the mean stroke interval was 3.6 months. There were no other risk factors for stroke in any of these patients. In one patient, a 7-year-old girl who had recurrent strokes in the left thalamus, AAD was demonstrated on plain X-rays and CT scan, but the 3D-CT angiogram did not show compression of the vertebral artery (Fig. 2). A dynamic vertebral DSA demonstrated a decrease in vessel caliber on neck flexion. She underwent a C1–2 fixation, and at her 16-month follow-up, the imaging showed good fixation and with no further strokes.

Type of atlantoaxial dislocation and associated anomalies (Table 2)

The most common type of AAD was the Type 3, in which four patients had horizontal translational dislocation with central dislocation (basilar invagination) associated with

C2–3 Klippel–Feil anomaly, C1 assimilation or vertebral artery hypoplasia. Three patients had a Type 1 AAD—translational dislocation alone, and only one patient had a C2–3 Klippel–Feil in addition. Three patients had a Type 5 AAD characterized by a central dislocation (BI), rotational dislocation and a coronal tilt, but only one patient had an associated C2–3 Klippel–Feil and C1 arch assimilation.

Surgical intervention

Six patients underwent CVJ fixation, while four patients refused any operative intervention and were managed with a cervical collar and anticoagulation alone. Table 3 summarizes the surgery done and changes in radiological parameters. Patients with Type 3 AAD were treated with C1 lateral mass–C2 pars screw, C1–2 distraction with spacer and a modified Gallie's fusion using an iliac autograft. Patients with Type 5 AAD were treated with C1–2 distraction and occipitocervical fusion. In Case 5, we limited ourselves to C1–2 distraction and modified Gallie's fusion as the patient had narrow pedicles and C2 pars interarticularis (Fig. 2). The mean preoperative ADI was 7.10 mm improving to a

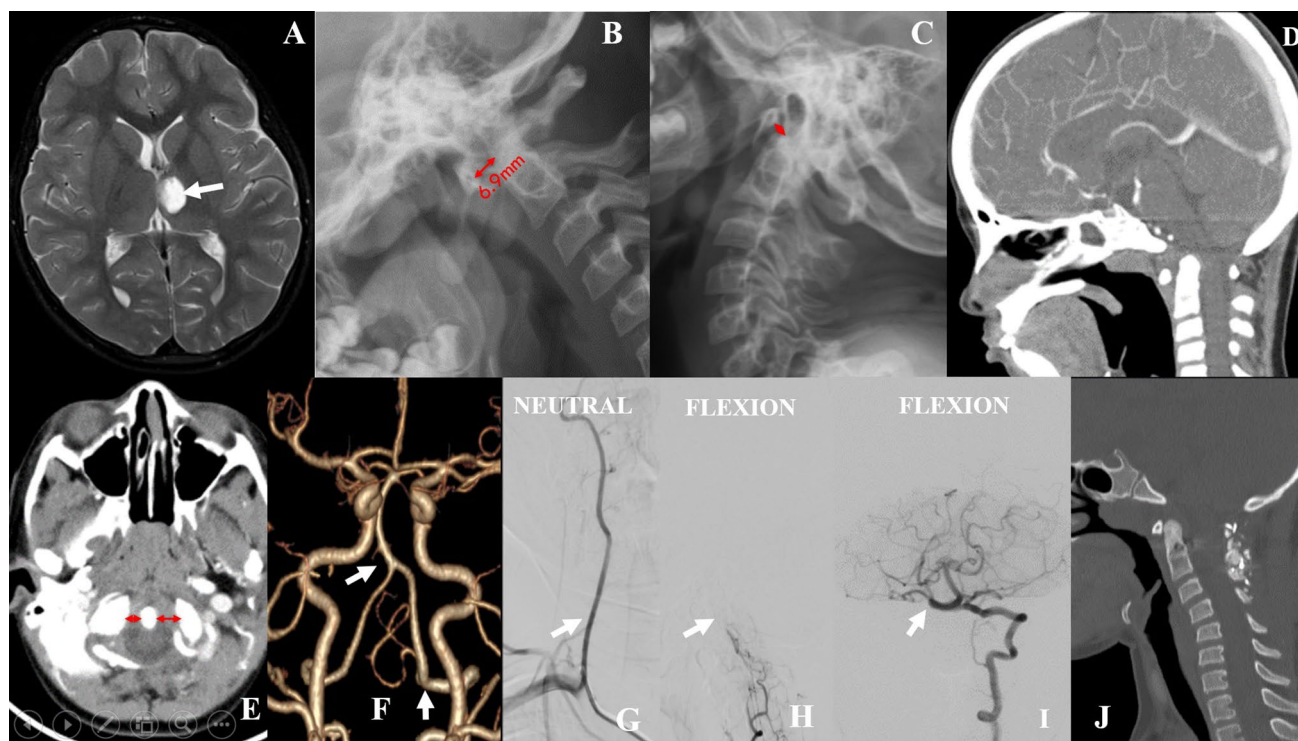


Fig. 2 Case 5: a 7-year-old girl with a left thalamic infarct who had a Type 5 AAD. **a** T2w MRI axial view shows a left thalamic infarction. Dynamic cervical spine X-rays in flexion (**b**) and extension (**c**) show a reducible AAD with an ADI of 6.9 mm. The CT cervical spine sagittal (**d**) and axial (**e**) views show a central AAD along with mild rotation (Type 5 AAD)—note that the odontoid is closer to the right lateral mass. 3D-CT angiogram reconstruction shows a left dominant

vertebral artery (white arrow) (**f**). DSA with a right vertebral injection in neutral (**g**) and flexion (**h**) shows complete occlusion of the artery in flexion (white arrows). A left vertebral artery injection in neck flexion shows no retrograde flow into the right vertebral artery (white arrow) (**i**). 1 year after C1–2 distraction with spacers and a modified Gallie's fusion CT sagittal view (**j**) shows complete reduction of the BI and AAD

Table 3 Surgical interventions and change in radiological parameters after surgery as measured with ADI, CCA and Powers ratio

S. no.	Surgery done	ADI (mm)		CCA		Powers ratio	
		Preop	Postop	Preop	Postop	Preop	Postop
1	C1–2 distraction and modified Gallie's fusion	8.8	2.83	141	154.2	1.044	0.88
2	C1–2 distraction, OC fusion, C0–C2 modified Gallie's fusion	7.6	6.01	138	144	1.099	0.91
3	FMD, C1–2 distraction and C0–2 modified Gallie's fusion	7.54	4.38	137	146	1.02	0.904
4	C1–2 distraction and fusion	7.3	2.73	143	150	0.95	0.72
5	C1–2 distraction, OC fusion, C0–C2 modified Gallie's fusion	6.9	1.72	126	142.2	0.98	0.88
6	Occiput–C3 fusion	4.5	1.52	144	152	1.02	0.82

OC Occipitocervical, ADI atlantodental interval, CCA clival canal angle

mean of 3.3 mm at follow-up (12 months). The mean CCA in the preoperative group was $138^\circ \pm 5.98^\circ$ and improved to $148^\circ \pm 4.32^\circ$ at follow-up.

Clinical and functional outcomes (Table 4)

All patients were followed up with a mean follow-up duration of 41.6 months (range 12–82 months) in the operated group and 18.5 months (range 13–29 months) in the non-operated group. None of the patients in the operated group had strokes after posterior fusion; however, three patients sustained recurrent strokes in the non-operated group despite patient being on anticoagulation and cervical immobilization on a collar.

MRS

Patients in the operated group reported significant improvement in the MRS (–30.7%) as compared to non-operated patients who reported steady decline in the neurological function (+12.5%) (*p* value 0.25).

PGIC scale

Those patients who were operated reported significant improvement in symptoms and quality of life on the PGIC scale, while those in the non-operated group did not experience any improvement in symptoms (*p* value 0.023).

Table 4 Comparison of clinical and functional outcomes between the operated and non-operated groups

S. no	Stroke	MRS		PGIC	imNurick grade		Modified McCormick Scale		NDI	Follow-up duration (months)
		Initial	At F/U		Initial	At F/U	Initial	At F/U		
Operated group										
1	No	3	2	1	3	2	4	2	12	38
2	No	3	2	2	3	1	4	3	13	82
3	No	4	4	3	5	4	4	4	6	38
4	No	4	1	4	4	0	4	1	7	64
5	No	1	1	4	1	0	1	1	15	16
6	No	1	1	2	1	0	1	1	15	12
Mean		2.6±1.24	1.8±1.06	2.6±1.1	2.8±1.46	1.16±1.46	3±1.41	2±1.15	11.3±3.52	41.6
Non-operated group										
7 ⁺	Yes (2)	2	3	6	1	2	2	3	0	16
8 ⁺	Yes	2	2	5	1	1	2	2	0	29
9 ⁺	Yes	2	2	5	2	2	2	2	0	13
10 ⁺	No	1	1	6	0	0	1	1	0	16
Mean		1.75±0.43	2±0.70	5.5±0.5	1±0.70	1.25±0.82	1.75±0.43	2±0.70	0	18.5
P values	0.1667	0.25		0.023	0.07		0.41	0.002		

MRS Modified Rankin Scale, F/U follow-up, PGIC patient global impression of change, NDI neck disability index

Patients marked with + were managed conservatively

Functional outcomes

The mean imNurick grade preoperatively was 2.8 ± 1.46 , and this improved to 1.16 ± 1.46 at follow-up ($p=0.07$), with a Nurick recovery rate of 58.5%. We noted a 33% improvement in the modified McCormick Scale in the operated group, while the non-operated group showed a steady decline in the neurological function (p value 0.41). As expected, patients undergoing surgery demonstrated a worsening of the neck disability index to a mean of 11.3 ± 3.52 .

Discussion

Pathology and pathogenesis

CVJ anomalies are more frequently seen in India as compared to the rest of the world with AAD being the most common malformation followed by assimilation of C1 and basilar invagination with a C2–3 Klippel–Feil anomaly [6, 17]. While the association between vertebrobasilar insufficiency and skeletal CVJ anomalies is well recognized [8], established PCS due to CVJ anomalies is rare and might even be

fatal [3, 18]. The V3 part of the vertebral artery, extending from the C2 foramen transversarium to the foramen magnum, has two parts: a vertical part ascending from C2 to the C1 lateral mass and a horizontal tortuous part over the posterior arch of C1. CVJ instability leads to stretching and kinking of the V3 horizontal segment. Repeated movements damage the intimal lining causing platelet aggregation and thromboembolism [4, 5, 19]. The left vertebral artery has been reported to be dominant in about 61–69% of the normal population [20, 21]. We had a slightly higher left-sided dominance of 75% that could explain our high incidence of left-sided strokes. Other etiologies include constriction of the vertebral artery by a fibrous band or thickened atlantooccipital membrane that can cause dynamic compression [4]. In addition, anomalous vertebral artery anatomy has been described in patients with CVJ anomalies [22] contributing to the higher PCS rates in these patients.

Incidence and screening

It is not possible to arrive at the exact incidence of CVJ anomalies in patients presenting with PCS since they are usually admitted under neurology and do not have a

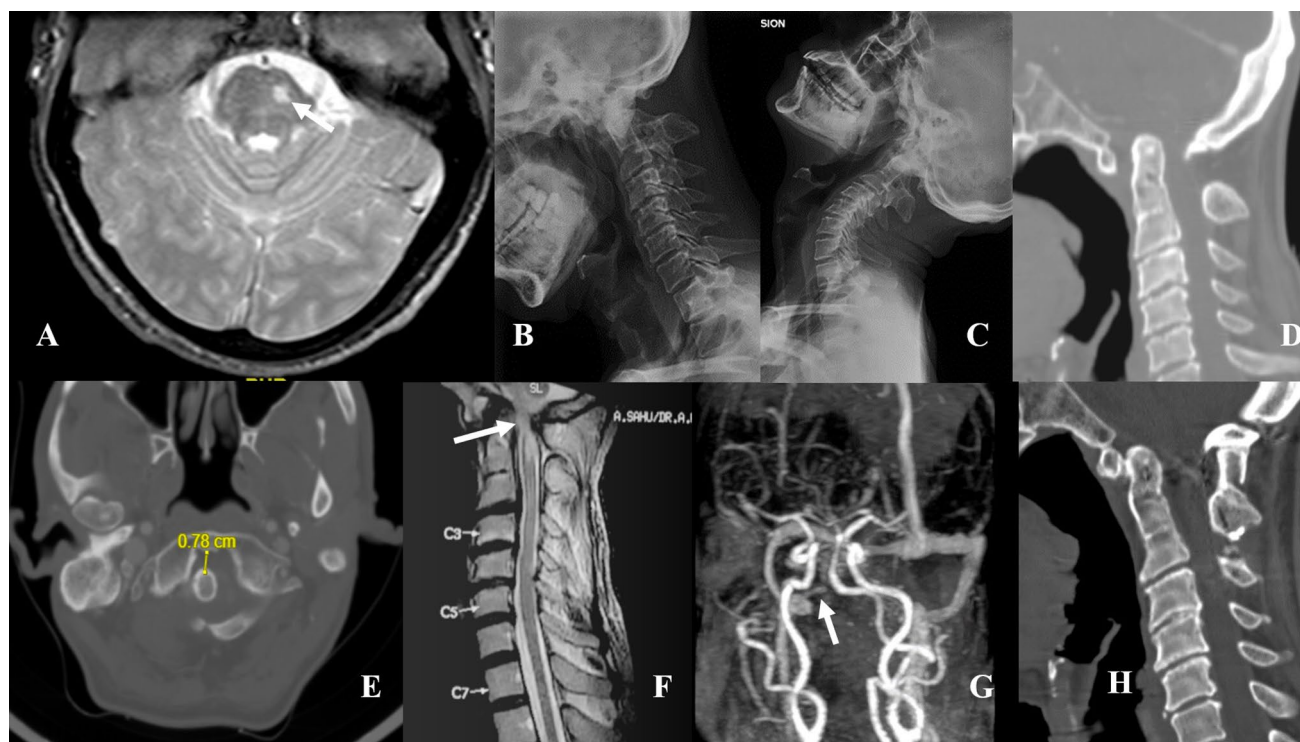


Fig. 3 A 52-year-old diabetic and hypertensive male with a left ventrolateral pontine infarct (a) and basilar invagination, occipitalized atlas and a rotatory AAD demonstrated on dynamic cervical imaging (b, c). CT cervical spine sagittal (d) and axial (e) show the central and rotatory AAD (Type 5) with an ADI of 7.8 mm. MRI cervical spine sagittal view (f) shows significant cord compression with T2w

intramedullary hyperintensities at the CVJ. 3D-CT angiogram shows a hypoplastic vertebral artery with narrowing of the basilar and carotid arteries (g) suggestive of atherosclerosis. Postoperative CT cervical spine 6 years after C1–2 fixation showed good reduction of the AAD and BI (h)

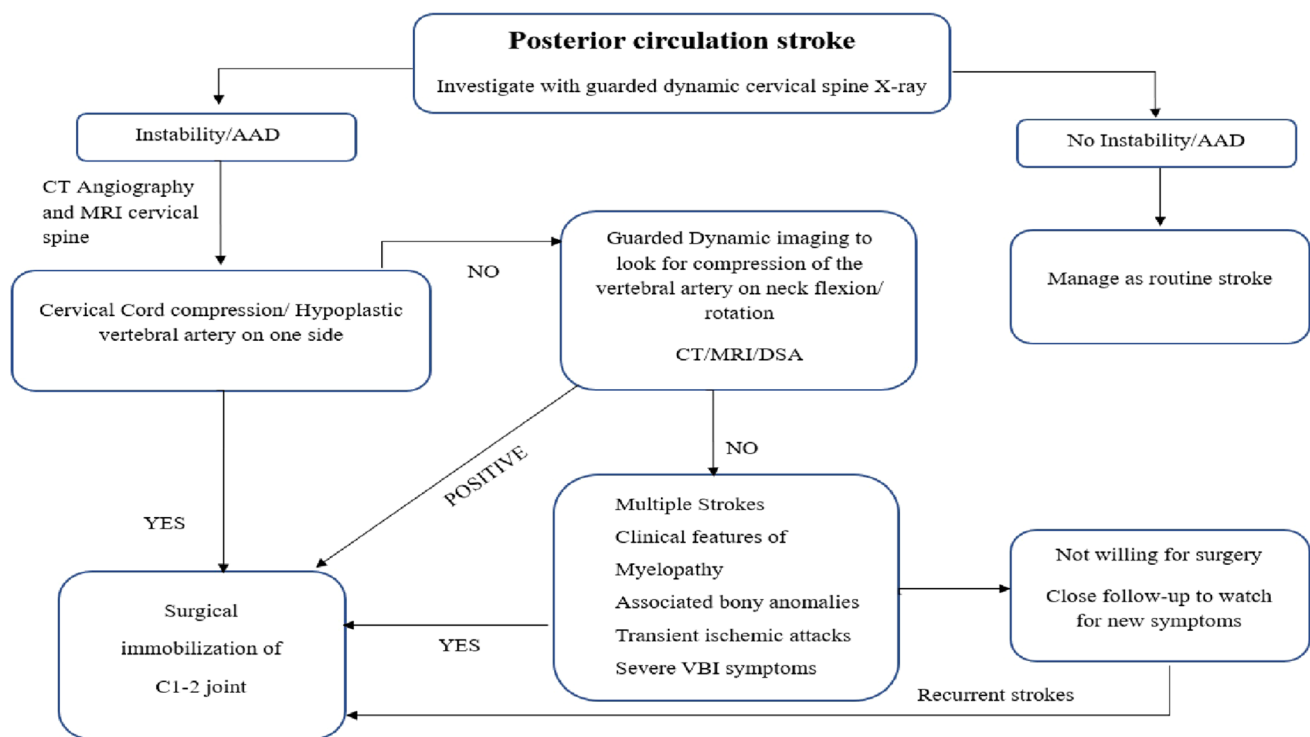


Fig. 4 Treatment algorithm for management of patients presenting with posterior circulation stroke

complete evaluation of the CVJ. However, our data suggest that CVJ anomalies as a cause of PCS are about 10% when patients with recurrent strokes or who have neck pain are evaluated with dynamic cervical X-rays and CT angiograms of the neck. On the other hand, data from surgical series reporting on outcomes of surgery for CVJ anomalies report a low incidence of PCS (2.6%) as a presenting symptom [17]. This difference can be explained by the fact that patients with CVJ anomalies usually present to the neurosurgeon with cervical myelopathy and are promptly treated with C1–2 immobilization thus protecting them from strokes [3, 4].

Since such a fatal condition can be easily identified by dynamic imaging [23], treating physicians should be sensitized to ensure that all patients with posterior circulation stroke regardless of age undergo screening with basic dynamic cervical spine X-rays to rule out an AAD. CVJ anomalies should be suspected even in elderly patients as atherosclerotic vertebral artery stenosis and dissection may coexist [24]. In our series, we had a 52-year-old, diabetic and hypertensive male with an AAD who presented with a basilar branch, left AICA and PICA infarct probably due to a precarious vertebrobasilar circulation acutely compromised by the AAD (Fig. 3).

Risk factors for PCS

AAD is the most common CVJ anomaly causing PCS in our series as noted by others [3, 5, 25]. Congenital vertebral artery hypoplasia and C1 assimilation are also known to increase the incidence of recurrent strokes [3]. There were two children in our series aged 7 and 11 years, both of whom had rotatory subluxation (AAD Type 5) that appears to be a significant risk factor for PCS and should be treated aggressively.

Management of PCS with AAD (Fig. 4)

Imaging

Patients presenting with a PCS are evaluated with dynamic X-rays of the CVJ. If no instability is noted and there is no evidence of any bony CVJ anomaly, no further imaging is recommended since all our patients with PCS had some bony CVJ anomaly detected on a dynamic plain cervical X-ray. A CT angiogram and MRI cervical spine are obtained if AAD or any other bony anomaly is detected. If vertebral artery hypoplasia or cervical cord compression is noted, no dynamic CT/MRI is required. On the other hand, if the CT angiogram shows no vertebral artery hypoplasia, a flexion/rotation study under supervision may be done the next day to

avoid exceeding the recommended safe daily dose of contrast administered. If the CT angiogram is negative in flexion/rotation, a DSA in flexion/rotation might identify a vertebral artery compromise, as seen in one of our patients (Fig. 2). CT has faster acquisition times as compared with MRI; however, it requires double contrast administration. MR angiography is more practical with a minimum neck holding time of 2.5 min and does not require contrast administration.

Medical management

Patients are started on anticoagulation and the neck immobilized in a Philadelphia collar. Those presenting with acute stroke are advised to return for surgery after 3 months to tide over the acute phase with supportive care and rehabilitation. Those presenting with old strokes are offered surgery immediately.

Surgery and follow-up

Reports on C1–2 immobilization protecting the vertebral artery from dynamic compression and subsequent strokes seem promising; however, no data are available for patients managed without surgery [3]. We confirm that surgery was protective against recurrent strokes and patients even demonstrated neurological improvement and improved quality of life on long-term follow-up, a consequence of CVJ stabilization improvement in myelopathy and prevention of recurrent strokes. With these benefits, the patient may have to accept the limitation of neck movements from fusion. In addition, surgery obviates the need for long-term anticoagulation and its attendant risks. On the contrary, those patients who refused surgery had repeated strokes with poor outcomes despite being on anticoagulation. The principal limitation in our study is its retrospective nature and the lack of complete CVJ imaging data on all patients with posterior circulation strokes.

Conclusion

Early identification of CVJ instability and timely C1–2 immobilization improves the clinical and functional outcomes in young patients presenting with posterior circulation stroke. Neurologists, physicians and spine surgeons need to be sensitized regarding the prevalence of this condition in PCS. Surgery helps in preventing further strokes and the need for long-term anticoagulation in these patients.

Author's contribution AGC, SA and KP contributed to the conception and design. HV and EJG helped in drafting the article. SAM contributed to the interpretation of radiology. AGC was the guarantor.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interests.

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