sheaths, alongside the internal jugular vein and the vagus nerve, from which they receive innervation [1]. The extracranial portion extends from the bifurcation to its entry through the carotid canal at the base of the skull [1]. In the absence of dolichoarteriopathies, the ICA typically passes approximately 2.5 cm from the tonsils [12] a detail of surgical significance that must be reported, especially when anatomical variants are present.

Etiology: The pathophysiology and natural history of dolichoarteriopathies are not fully understood [8]. It is hypothesized that these anomalies result from an increased arterial length between fixed points at the bifurcation and the petrous bone, caused by a disproportionate growth of the muscular layer relative to the adventitia [2,13,14]. Additional theories attribute these changes to physiological aging processes, added to conditions such as atherosclerosis, vasculitis, or fibromuscular dysplasia, which cause loss of arterial elasticity, which ultimately leads to arterial elongation and tortuosity [6,15]. Some researchers suggest a multifactorial origin, where congenital kinking can change with age, diminishing with growth, and reappearing during aging [16].

Classification: A classification system, developed by Paulsen and collaborators, divides dolichoarteriopathies of the extracranial portion of the internal carotid artery into four types based on the degree of deviation from the vertical axis: straight (less than 15°), curved (15° to 70°), looped (70° to 145°), and coiled types (360° deviation) [2,17]. However, the most widely accepted classification of dolichoarteriopathies was established in 1965 by Weibel and Fields, and they are categorized into three types: tortuosity, coiling, and kinking [18]. Notably, multiple types can coexist within the same artery, and bilaterally has been well described [8] (**Figure 1).**

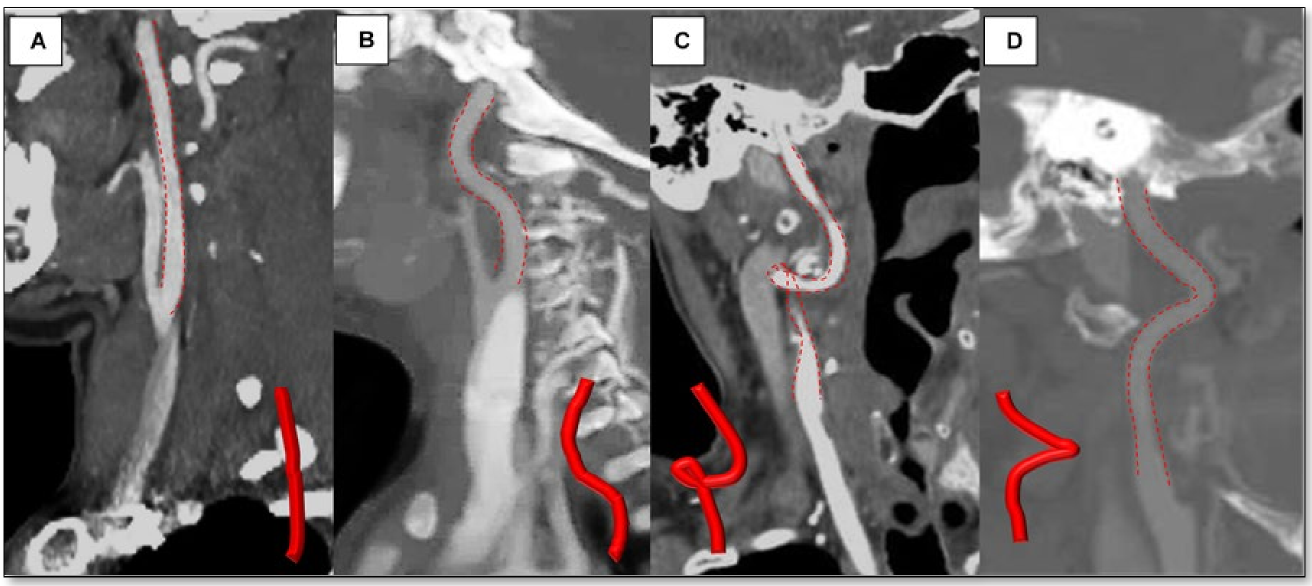

Figure 1. Morphological classification of the Internal Carotid Artery. A) Straight; B) Tortuosity; C) Coiling; D) Kinking.

Tortuosity: Tortuosity refers to elongation and C- or S-shaped curvature [6]. It is further subdivided into elongation, redundancy, and undulation [2,8]. For some authors tortuosity may encompass other types of dolichoarteriopathies, such as coiling and kinking [2].

Coiling: Coiling is defined as elongation of the extracranial portion of the internal carotid artery within a confined space, resulting in an exaggerated "C," "U," or "S" shape or a simple or multiple circular configuration [8,18,19] due to a complete 360° rotation of the arterial tract [6].

Kinking: Kinking is characterized by an acute angulation of the extracranial portion of the internal carotid artery of less than 90° [6]. Some authors believe kinking as a variation of coiling [8]. Kinking is further classified by the degree of curvature into three grades [8] (**Figure 2**).

CLINICAL IMPLICATIONS

Clinically, it may be asymptomatic or present as a pulsatile neck mass or pharyngeal bulge [8]. It may have a benign clinical course, or cause reduced cerebral blood flow, particularly in the presence of atheromatous plaques and stenosis [6,20]. Ischemia related to these anomalies is mainly attributed to two mechanisms. These include endothelial injury leading to thromboembolic events and hemodynamic alterations impairing cerebral perfusion [8]. Reported symptoms include pulsatile tinnitus of arterial origin, hemilingual spasm from hypoglossal nerve compression, cognitive decline in adults and children,

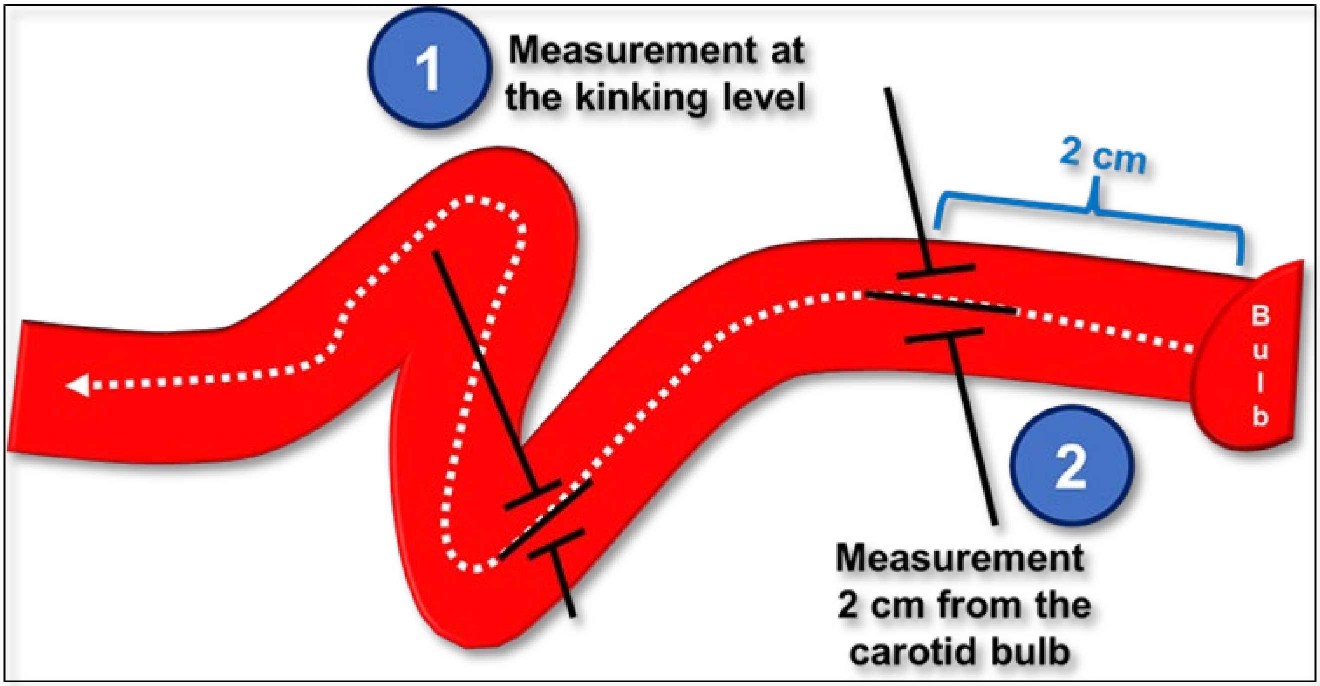

Figure 3. Schematic of the Internal Carotid Artery evaluation using spectral Doppler.

Peak Systolic Velocity is recorded at the kink or coil level and two centimeters proximal to the carotid bulb. The Kinking Ratio (PSVKR) is the ratio between both measurements. Based on Di Pino, 2020. This measurement correlates with the degree of angulation and the severity of obstruction of anatomic variants [6] offering functional insights into affected segments [6]. For instance, kinking severity has been linked to blood flow reductions: 40% in grade II and for grade III in 60% [8]. Additional factors influencing hemodynamic repercussions include intimal hyperplasia, atheromatous plaques, aneurysms, stenosis, and collateral cerebral circulation [8]. Ultrasound is advantageous for dynamic assessments accounting for physiological states such as hypotension during sleep, head hyperextension, and head rotation, which can affect flow dynamics. Also, it is particularly valuable for postoperative evaluation of internal carotid artery (ICA) surgery, aiding in assessment of vessel alignment and flow restoration. Despite its benefits, Doppler ultrasonography has limitations. It is operator-dependent, it requires a steep learning curve, and implies technical challenges that may arise in patients with small necks or certain body compositions [8].

CT angiography

CT angiography with contrast or MRI provides superior anatomical detail and is unaffected by ultrasound limitations [6,9]. CT is particularly useful for evaluating arterial trajectories and detecting alterations such as microaneurysms [8,20]. It enables a comprehensive assessment of supra-aortic trunks and branches, vessel calibers, and atheromatous plaques, aiding in preoperative planning [21]. Sagittal or coronal CT and MRI slices allow measurements based on Metz's classification and enable three-dimensional reconstructions for a more accurate assessment of tortuosity types [8] (**Figure 4)**.

One recommended protocol, consists in a simple scan that must be obtained from the aortic arch to the cranium vertex, followed by a contrast enhanced phase. 50 to 70 ml of non-ionic intravenous contrast is injected with a flow of 6 ml/s [22]. Measurement of kinking angle is determined by drawing two lines, one along the axis of the internal and common carotid arteries and another along the axis of the cervical segment of the internal carotid artery [8], however, another proposed measurement is drawing one line along the axis of the proximal portion of the vessel and other in the distal portion (**Figure 5)**.

The kinking angle is measured by drawing two lines: one along the axis of the cephalic portion of the kink and another along the caudal portion, following the typical direction of blood flow.

Figure 4. Evaluation of Internal Carotid Arteries using 3D Volume Rendering from Multidetector CT.

Figure 5. Evaluation of kinking with multidetector CT.

Magnetic resonance imaging

Magnetic resonance imaging (MRI) is particularly indicated in dolichoarteriopathies when there is an impairment of blood flow, as MR perfusion studies can provide valuable insights [8]. This non- invasive method offers multiple sequences to evaluate both the vessel pathway and its hemodynamic implications. Contrast-enhanced MRI is considered more effective for diagnosing dolichoarteriopathies [3,8] (**Figure 6**).

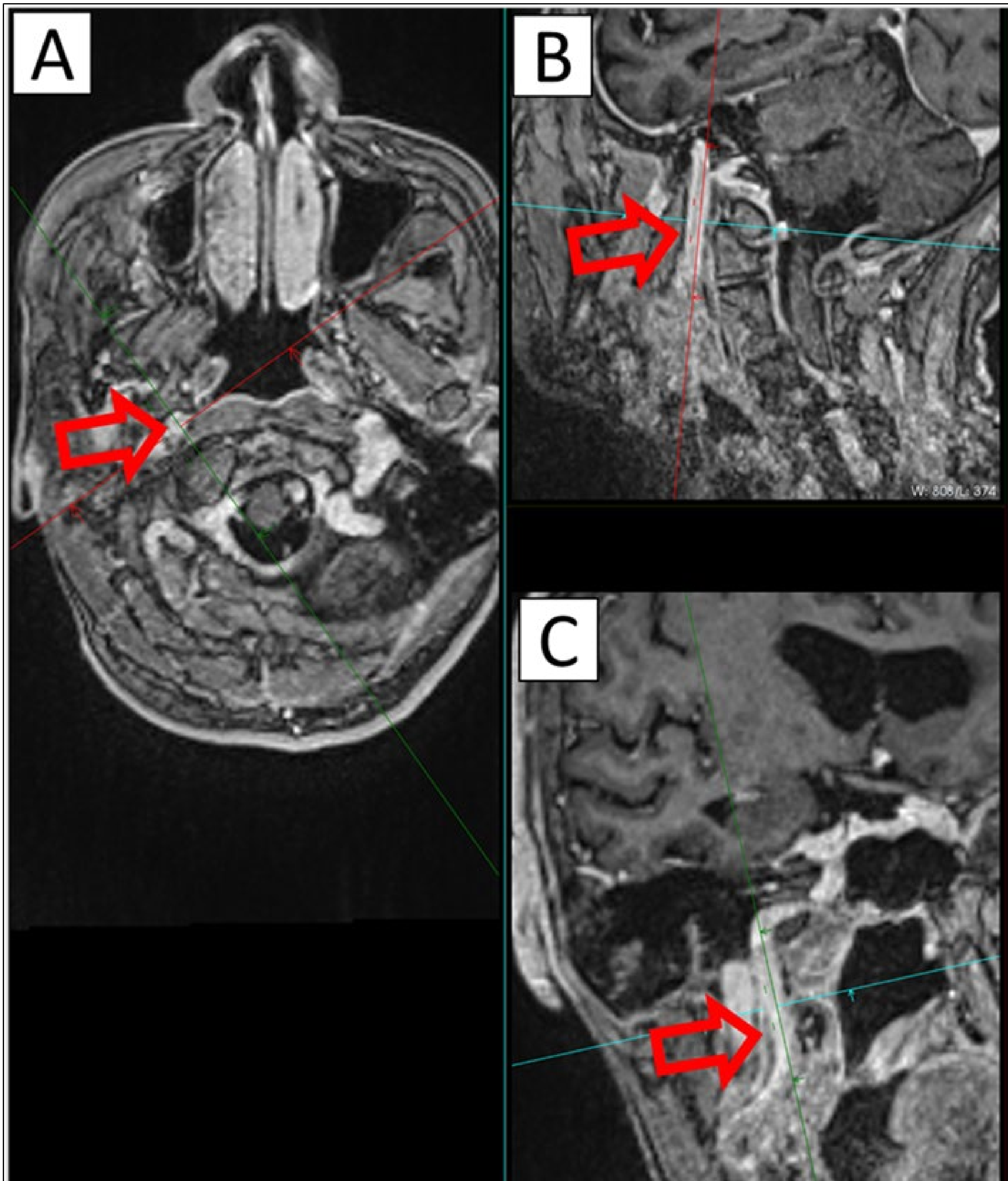

Figure 6. Multiparametric assessment of the right Internal Carotid Artery using Contrast Enhanced MRI T1 Sequence.

Alternatively, non-contrast Angio-MRI using Time-of-Flight (TOF) sequences is a viable option (**Figure 7**).

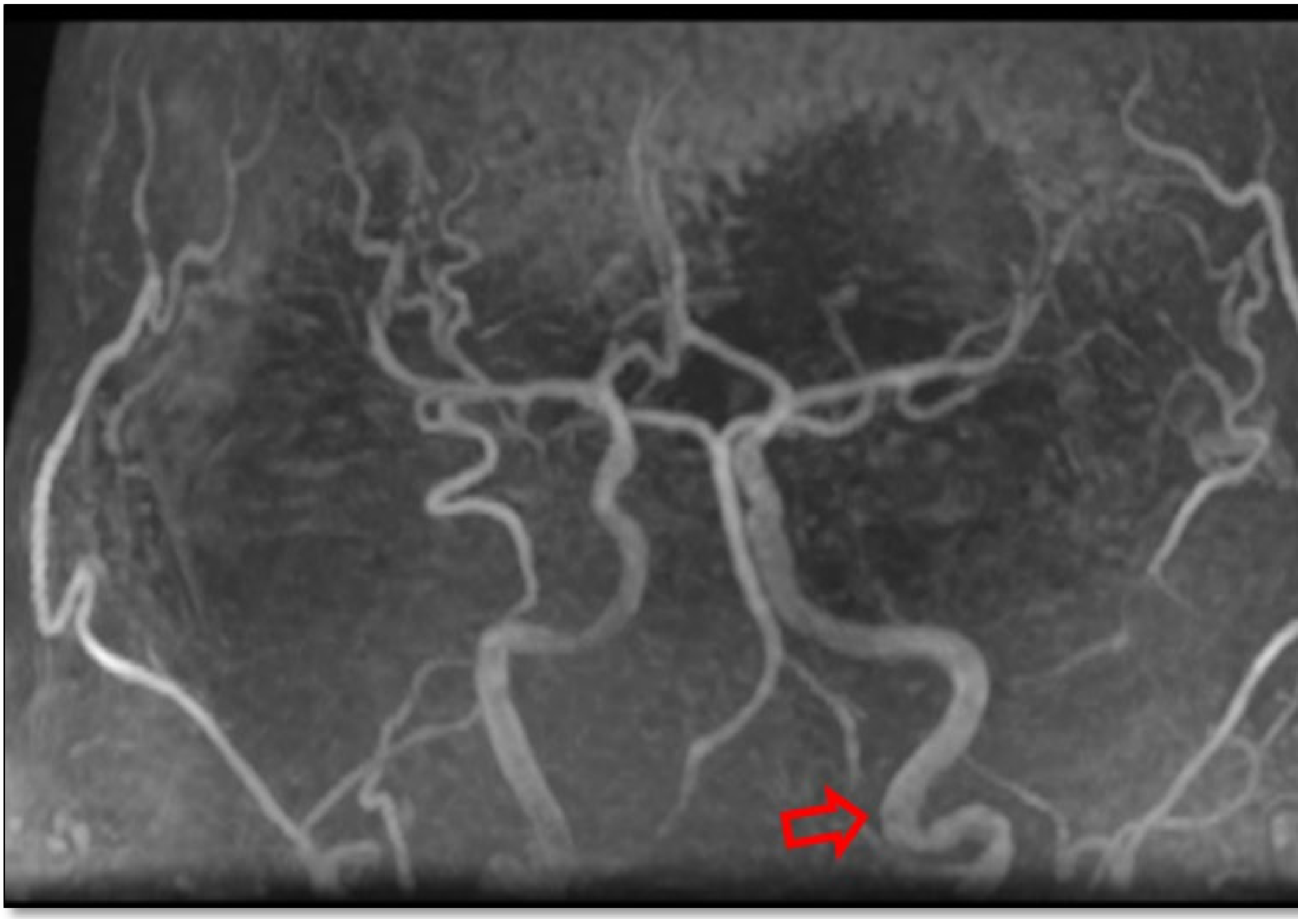

Figure 7. Evaluation of Internal Carotid Arteries with 3D TOF Magnetic Resonance Sequence.

Digital subtraction angiography

Digital subtraction Angiography (DSA) is a fluoroscopic technique for the visualization of blood vessels. It provides detailed arterial morphology, useful distinguishing morphologic subtypes [3] particularly through 3D reconstructions, and also has the advantage of providing real-time hemodynamic data concerning brain flow. Also, postprocessing, such as in the PACS system, can provide valuable tools for the measurement of dolichoarteriopathies [23]. However, it does not evaluate the arterial wall, unlike CT and MRI [8].

Nuclear imaging

Nuclear imaging with intravenous technetium-99m (99mTc) pertechnetate can provide perfusion data in symptomatic patients, this is useful, especially in the context of kinking for its association with blood flow reduction [8] (**Figure 8**).

MANAGEMENT AND PROGNOSIS

Surgical intervention may be considered in cases where significant hemodynamic impairment is documented, although the optimal treatment remains controversial [8]. Procedures include resection of redundant segments with vascular reconstruction [24] end-to-end anastomosis to a lower internal carotid segment or at the bifurcation, end-to-side anastomosis with the external or common carotid artery, endarterectomy with patching or imbrication of the common carotid artery, bypass grafting or eversion endarterectomy with excision of excess arterial length. The aim is to shorten the vascular trajectory, restore perfusion, and prevent recurrence [8,25]. For cerebrovascular insufficiency caused by kinking, surgical intervention can prevent further deterioration of perfusion. In cases of tortuosity, surgery is recommended only when atherosclerosis causes stenosis [8,26-30]. Post-surgical outcomes are generally favorable, with low morbidity and mortality rates. Follow-up studies have shown no recurrences over six years [8] and cognitive function and symptoms, such as ocular disturbances, often improve significantly after treatment [8].

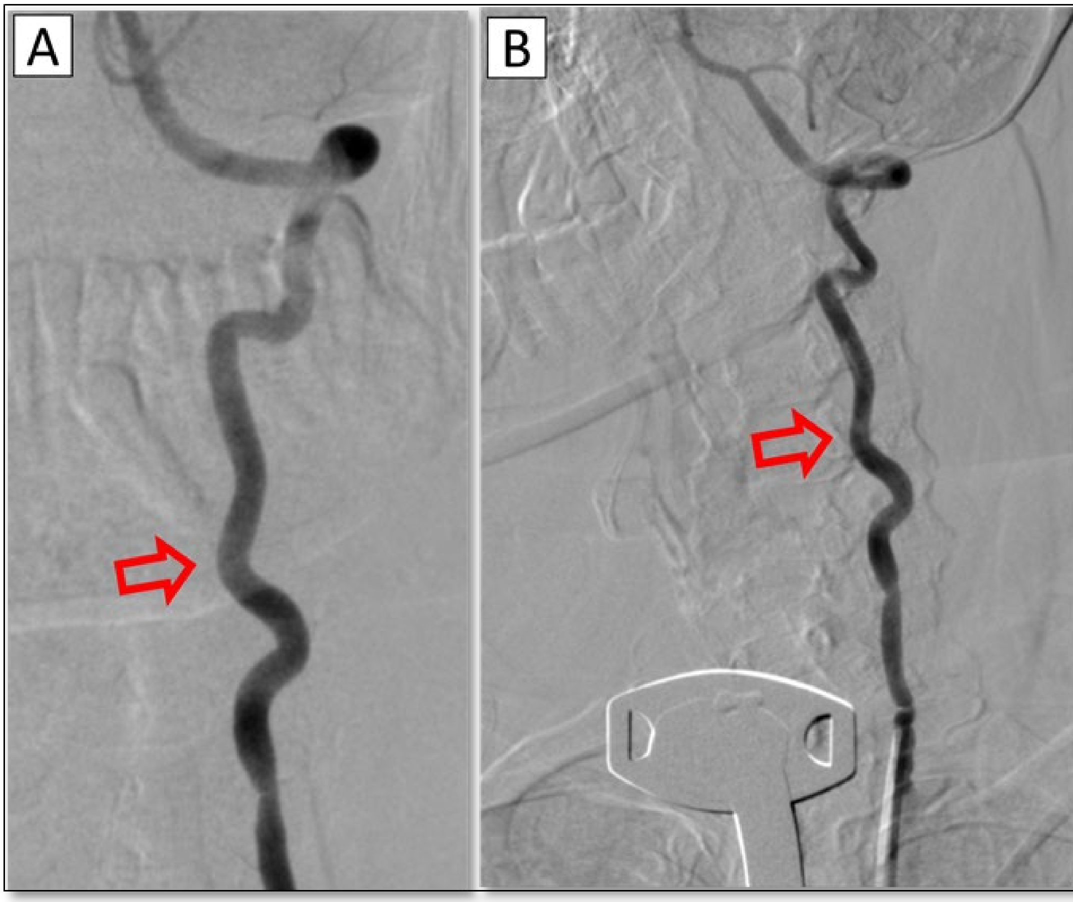

Figure 8. Digital Subtraction Angiography (DSA) showing tortuosity in the extracranial ICA pathway.

CONCLUSION

Vascular anomalies of the extracranial internal carotid artery have elicited attention due to their potential association with brain hypoxemia, and the increased surgical procedures they pose. As imaging techniques continue to evolve, the study of these variants must adapt to advancements. Accurate detection and routine reporting in imaging are crucial for improving patient care, facilitating surgical planning, preventing complications, and enhancing knowledge in these entities. This article provides a comprehensive review of the current state of imaging techniques used to evaluate dolichoarteriopathies and serves as a foundation for further studies.

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