Kinks, Coils, and Carotids: A Review

BY BINDU DESAI, M.B., B.S., AND JAMES F. TOOLE, M.D.

Abstract:
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Kinking and coiling of the internal carotid artery (ICA) sometimes may result in symptomatic cerebrovascular disease, but indisputable evidence linking the two conditions is lacking. However, there is enough evidence to warrant careful consideration of surgical correction in patients who have features of the carotid artery syndrome and kinking of the ICA as shown on angiography. Kinking or buckling of the artery is due to atherosclerosis and is to be distinguished from coiling, which is ascribed to embryological causes.

Definite recommendations regarding the advisability of surgery for infants who are discovered to have coils cannot be made, but coiling is generally asymptomatic. Adults with kinks in their carotid arteries who have recurrent transient ischemic attacks (TIAs) benefit most from surgical correction, particularly if symptoms are aggravated on head rotation, which may cause the kink to obstruct.

Additional Key Words
internal carotid artery
cerebrovascular insufficiency
head rotation
anatomical variability
surgical treatment

The association between kinking of the internal carotid artery (ICA) and cerebrovascular insufficiency was first noted in 1951. Since then several reports have dealt with the clinical relationship between carotid elongation and kinking and cerebrovascular disease. Although conclusive evidence linking the two is still lacking, certain inferences can now be drawn. In this review we will critically examine previous reports of this anomaly and thereby derive useful clinical parameters for physicians who must decide whether surgical treatment is indicated in patients with angiographical evidence of a kinked carotid and symptoms of cerebrovascular insufficiency. As the incidence of this abnormality in the total population has been estimated to be as high as 16%, the importance of its correct diagnosis and treatment cannot be overemphasized.

History
The anatomical variability of the ICA has been clearly recognized for decades. Its relationship to the tonsil was of special interest to otolaryngologists who at first thought the vessel might be the ascending pharyngeal. Detailed reports later confirmed that the ICA when tortuous "becomes a directly lateral relation of the tonsil, [with] only the superior constrictor intervening." The hazard of fatal hemorrhage during tonsillectomy was recorded and subsequently resulted in a timely warning graphically stated by Skillern: "Before operating, the surgeon should Stop, Look, and Listen. A thorough ocular and digital exploration of the pharynx for arterial pulsations should never be omitted." Recognition of the association between cerebrovascular disease and looping and kinking of the ICA is comparatively recent.

Pathology
Definition of Coils and Kinks
Determining the precise nature of the carotid artery abnormality through arteriography is crucial in deciding whether surgical intervention is indicated. In this regard the definition of coils and kinks given by Weibel and Fields is very helpful. We applied it in our review of the case reports considered later. According to this definition, coiling is elongation and redundancy of the ICA resulting in an exaggerated S-shaped curvature or in a circular configuration. Kinking is described as angulation of one or more segments of the ICA associated with stenosis in the affected segment.

Embryology and Symptomatology
Looping and kinking of the ICA has been observed in infants and even in fetuses. The cause of these loops is related to embryological development. The vessel is formed from the third aortic arch and from the dorsal aorta; hence, in the embryo it is normally kinked. Straightening occurs when the fetal heart and large vessels recede in the thoracic cavity. If the embryological state persists, it produces different kinds of undulations, loops, and kinks.

This anomaly generally does not become symptomatic until later in life. There appears to be no relationship between the severity of the kinking and either rising blood pressure or increasing age. The role of degenerative changes in the vessel wall remains uncertain. If this factor were important, then kinks should be more severe in older individuals because degenerative changes increase with age; however, this is not generally the case. At best the cause for looping and kinking of the carotid artery may be ascribed

From the Department of Neurology, Bowman Gray School of Medicine, Wake Forest University, Winston-Salem, North Carolina 27103 (reprint requests).
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partly to embryological development and partly as secondary to atherosclerotic changes.

**Carotid Artery Syndrome**

By definition, in the carotid artery syndrome the offending carotid artery is held responsible only for the following features: (1) contralateral hemiparesis, which may be transient, or hemiplegia, with or without sensory deficit; (2) ipsilateral monocular visual dysfunction; (3) ipsilateral frontal headache; (4) homonymous field defects; and (5) language defect (partial or complete) only when the dominant hemisphere is involved.

Symptoms such as syncope, confusion, roaring in the ears, and memory loss when cited as being due to ipsilateral carotid artery disease are considered only when the total angiographical report permits a clinicopathological correlation.

**Occurrence in Infants and Children**

Few reports deal with the association between infantile hemiplegia and kinking and coiling of the carotid artery. Sarkari's series\(^6\) consisted of nine infants with cerebrovascular disease who exhibited unilateral or bilateral looping or kinking. Theorizing that the vascular anomaly may have been responsible for the hemiplegia, they suggested surgical correction in such patients. The severe neurological deficits in these infants, due to looping of the carotid artery, were difficult to explain. A combination of events (sudden obstruction, neck rotation, and microemboli) could have been responsible for the cerebral ischemia; however, because no pathological studies of the vessel involved were performed, we do not have evidence for emboli formation. We believe that kinking of the vessel could cause eddy currents and microemboli formation due to slowing of blood flow at the site. These emboli would not result in any change in the vessel wall itself but could cause cerebral ischemia.

Parrish and Byrne\(^7\) reported five cases of hemiparesis and seizures in children who had angiographical evidence of kinking of the ICA. Two of these also had occluded middle cerebral arteries but were surgically treated and clinical improvement was noted.

At this stage we recommend carotid angiography in infantile hemiplegia but are unable to give definite criteria for surgical correction of loops or kinks. This must await further study.

**Occurrence in Adults**

Several reports deal with this condition in adults (Table 1); we will consider them each in turn.

Gass’ study\(^1\) included one patient with recurrent attacks of left hemiparesis and left hemisensory deficit. The patient was treated surgically but three months later had an occluded ICA without any resulting symptoms. Although Gass reported on some cases with sudden loss of consciousness, detailed angiographical evidence linking the symptom to the vascular anomaly is lacking. Therefore, we cannot comment on these cases.

In the study by Derrick et al.,\(^2\) no specific correlation of symptomatology and arteriographical abnormalities is found. The symptoms were rather broad-based and not strictly due to the carotid artery syndrome. Interestingly, 19 of these patients experienced symptoms associated with some extreme movement of the head.

**Table 1**

<table>
<thead>
<tr>
<th>Study</th>
<th>No. of pts.</th>
<th>Correlation factor*</th>
<th>No. treated surgically</th>
<th>Period of follow-up</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gass(^1)</td>
<td>6</td>
<td>Yes(^†)</td>
<td>1</td>
<td>3 months</td>
<td>Improved</td>
</tr>
<tr>
<td>Derrick(^2)</td>
<td>58</td>
<td>No</td>
<td>58</td>
<td>2–6 years (28)(‡)</td>
<td>13—no recurrence of symptoms; 12—slight return of symptoms; 1—unimproved; 2—died of unrelated causes</td>
</tr>
<tr>
<td>Quattlebaum(^3)</td>
<td>138</td>
<td>No</td>
<td>138</td>
<td>1–14 years (109)(‡)</td>
<td>39—excellent; 61—good; 14—fair; 24—poor</td>
</tr>
<tr>
<td>Sanger(^4)</td>
<td>5</td>
<td>Yes</td>
<td>5</td>
<td>None</td>
<td>All improved immediately</td>
</tr>
<tr>
<td>Randels(^5)</td>
<td>8</td>
<td>Yes</td>
<td>5</td>
<td>13 months</td>
<td>3 improved</td>
</tr>
<tr>
<td>Harrison(^6)</td>
<td>46</td>
<td>No</td>
<td>35</td>
<td>3 years</td>
<td>Improvement in 6 of 9 with hemiplegia, 9 of 12 with stroke in evolution, 11 of 14 TIAs</td>
</tr>
<tr>
<td>Freeman(^7)</td>
<td>44</td>
<td>No</td>
<td>44</td>
<td>Not stated</td>
<td>40 improved</td>
</tr>
<tr>
<td>Najafi(^8)</td>
<td>15</td>
<td>Yes</td>
<td>15</td>
<td>5 months–4 years</td>
<td>11 improved</td>
</tr>
</tbody>
</table>

*Individual cases correlated in terms of clinical symptoms and angiographical abnormality.

\(^†\) Sudden contralateral hemiparesis or loss of consciousness in four cases; repeated leftsided transient attacks in one case; bruit over left mastoid in one case.

\(^‡\) Number in parentheses indicates number of patients included in follow-up.
REVIEW OF KINKS, COILS AND CAROTIDS

Quattlebaum et al.\textsuperscript{13} reported that patients with a history of transient ischemic attacks (TIAs) who were operated on did well after surgery (82%), whereas those with symptoms of a more diffuse nature did not. All of the patients studied by Sanger et al.\textsuperscript{14} had the classical carotid artery syndrome. Unfortunately, no follow-up on these cases is available.

The study by Rundles and Kimbell\textsuperscript{16} included three cases with symptoms more likely to be vertebrobasilar than cerebrovascular in origin. The follow-up period is too short for any conclusions to be drawn.

Harrison and Davalos\textsuperscript{18} treated nine patients with hemiplegia and 12 with stroke in evolution. Because no specific correlation of angiographical abnormalities and symptoms is available, these cases are difficult to assess.

Freeman and Lippitt\textsuperscript{17} reported a 92% incidence of relief in their patients, but no follow-up is available.

Najafi et al.\textsuperscript{18} followed their cases for a period of five months to four years. Three of the 15 cases cannot be strictly included in the carotid artery syndrome.

Discussion

As can be noted from table 1, a fairly large number of cases with kinking or coiling of the carotid artery have been operated on and followed for various intervals. Symptomatic improvement following surgery is noted in nearly all the reports but no standardized criteria to judge the improvement have been applied.

None of the authors claim to imply that kinks and coils of the carotid artery are invariably symptomatic.\textsuperscript{11–18} The evidence suggests that coiling whether unilateral or bilateral rarely results in symptoms unless there is also atherosclerotic occlusive disease in the carotid, vertebral, or basilar arteries.\textsuperscript{7} When symptoms occur, they may be secondary to disease of intracranial vessels not adequately visualized by routine arteriography. Kinking of the ICA alone may occasionally cause symptoms of cerebrovascular disease, but several other factors such as variation in blood pressure, alterations in head and neck positions, and extracranial or intracranial occlusive diseases contribute significantly to the production of cerebral symptoms. Sometimes "development of symptoms depends on the fortuitous positioning of the head in such a way that the vessels are kinked rather than merely elongated."\textsuperscript{113} (p 576) Recurrent episodes of cerebrovascular insufficiency are more common in patients with kinks than in controls though this is just outside the conventional 5% level of significance.\textsuperscript{5} Indisputable evidence for any connection between kinking of the ICA and cerebrovascular symptoms does not exist. However, the evidence is suggestive enough to prompt careful consideration of surgical excision in patients with recurrent cerebrovascular episodes who have a kink and in whom no other adequate cause can be found in the cerebrovascular or cardiovascular systems. Surgery should be especially considered in those cases in which rotation of the neck produces symptoms.\textsuperscript{2}

According to Culligan,\textsuperscript{19} buckling of the great vessels at the base of the neck does not require surgical correction. The circulatory dynamics in the kinked carotid artery have been studied experimentally,\textsuperscript{20} and an inverse relationship between the severity of the kink and the blood pressure and blood flow distal to the kink has been noted; however, not everyone agrees that cerebrovascular insufficiency is directly related to the degree of angulation of the kinked segment.\textsuperscript{7}

It is gratifying that patients with TIAs and a contralateral looped or kinked carotid artery generally improved or had no further symptoms following surgery.\textsuperscript{2,13} This is probably the strongest indication for surgical correction of a kinked carotid. Since the surgery on these patients involved a generous resection of the common carotid artery,\textsuperscript{21} it also required removing the most common site of atherosclerotic plaque formation. Therefore, straightening of the ICA may not be the only reason for the improvement.

Several patients were treated surgically for symptoms (vertigo, blackout, progressive mental deterioration) not strictly attributable to carotid artery disease.\textsuperscript{13} Loss of balance\textsuperscript{22} and tinnitus\textsuperscript{15} are more properly considered as resulting from vertebrobasilar insufficiency. Patients with completed strokes\textsuperscript{16} or spastic hemiparesis\textsuperscript{23} should not be considered suitable surgical candidates.

Our own experience includes seven patients with TIAs and angiographical evidence of tortuosity with or without kinking of the ICA. Five had symptoms of vertebrobasilar insufficiency. The sixth had transient right-sided weakness and marked tortuosity of the left ICA. She was not treated surgically. Two years later she complained of generalized weakness but had no neurological deficit.

The last patient was a 77-year-old man with recurrent transient bouts of leftsided weakness. Angiography revealed a loop in the midcervical portion of the ICA with a 2-mm posterior wall ulcer near the carotid bifurcation (fig. 1). Head rotation did not result in occlusion of the vessel. The patient was initially anticoagulated with coumadin, but this was discontinued when he had another episode of cerebrovascular insufficiency, this time resulting in a mild left hemiparesis. Surgical correction of the carotid was then done, involving right carotid endarterectomy, with the redundant ICA resected and then reimplanted into the common carotid via end-to-end anastomosis. Blood flow in the ICA, which was 90 to 110 ml before surgery, increased to 700 ml following the operation. Pathological examination revealed atherosclerotic intima and media with evidence of fresh hemorrhage in the atherosclerotic plaque. This case is of interest for several reasons. Though head rotation did not result in occlusion of the blood vessel, blood flow...
Loop in the midcervical portion of the ICA with a 2-mm posterior wall ulcer near the carotid bifurcation.

increased markedly following surgery. Also the fresh hemorrhage seen on pathological examination could have resulted from anticoagulation.

Perhaps the best explanation for exploring the possible relationship between kinking of the ICA and cerebrovascular disease was given by Metz et al: "It is always difficult to be certain of the relevance of any factor to the production of cerebrovascular symptoms. Cessation of symptoms after removal of a factor does not establish the existence of a causal relation between the two. It is well recognized, for example, that transient ischemic episodes may cease spontaneously; hence, the disappearance or non-recurrence of symptoms following the excision of a kink in the internal carotid artery must be interpreted with caution. Nevertheless, the present unsatisfactory state of therapy for cerebrovascular disease demands that each potential method of treatment be thoroughly assessed." (p 424)

Conclusions
Infants with hemiplegia may show kinking and looping of the carotid arteries on angiography.
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