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# Are Kinking and Coiling of Carotid Artery Congenital or Acquired?

Ricardo Beigelman, MD, PhD, Andrés M. Izaguirre, MD, Martín Robles, MD, Daniel R. Grana, VMD, Giuseppe Ambrosio, MD, PhD, FACC, FESC, FAHA, and José Milei, MD

Dolichoarteriopathies consist of tortuosity, kinking, or coiling of the extracranial carotid arteries. Some authors consider these alterations a consequence of atherosclerotic vessel remodeling, while others ascribe them to anatomical variations of embryological origin. The objective was to establish whether carotid dolichoarteriopathies belonged to a congenital origin or to acquired conditions. Color Doppler ultrasonography of neck vessels was performed in 885 participants, whose age ranged from 1-day-old infants to 90-year-old adults. Prevalence of kinking and coiling was evaluated, and it was related to the presence of cardiovascular risk

factors. Prevalence of either kinking or coil of carotid arteries showed no increase with age, as it was comparable across all ages; furthermore, frequency of these alterations showed no relationship to cardiovascular risk factors nor to the presence of atheromatous plaques. These findings suggest that carotid dolichoarteriopathies are a result of alterations in embryological development rather than vascular remodeling secondary to aging and/or atherosclerosis.

**Keywords:** hypertension; cerebrovascular alterations; ultrasound; risk factors; carotid artery

## Introduction

Atherosclerosis is the most frequent cause of extracranial carotid artery disease.<sup>1</sup> However, although atheromatous pathology of the carotid bulb and bifurcation is a major cause of stroke, other causes of carotid disease may also cause vessel occlusion, such as fibrodysplasia, trauma (with subsequent dissection of carotid arteries), aortic arch pathology as in Takayasu disease, and aortic dissection.<sup>2</sup> Among nonatheromatous alterations of the carotid arteries, interest has long been placed on specific anatomical abnormalities called dolichoarteriopathies. Carotid

dolichoarteriopathies can be classified into 3 different types<sup>3</sup> (Figure 1). Type 1: tortuosity—a nonrectilinear stretch of an artery with an angulation  $>90^\circ$ ; type 2: loop—a  $360^\circ$  angulation of an artery on its transverse axis (“coil” configuration); type 3: kinking—the inflection of 2 or more segments of an artery with an internal angle of  $\leq 90^\circ$ .

Dolichoarteriopathies of carotid arteries are frequent, ranging between 10% and 45%.<sup>4</sup> For type 3, a prevalence of 5% to 25% has been described.<sup>5,6</sup>

Published studies have reached disparate conclusions with regard to the origin or cause of carotid dolichoarteriopathies, as well as their hemodynamic and prognostic significance.<sup>4-9</sup> Mukherjee and Inahara<sup>9</sup> proposed that carotid kinking would induce turbulent flow, thus favoring intimal ulceration, platelet deposition, and distal thrombus embolism. Other investigators similarly believe that a causal connection exists between cerebral flow alteration and severe carotid dolichoarteriopathies, to the point of proposing surgical correction of kinking and coiling to prevent stroke.<sup>10-12</sup> Conversely, other authors consider carotid

From the Instituto de Investigaciones Cardiológicas “Prof. Dr. Alberto C. Taquini,” School of Medicine, University of Buenos Aires—CONICET, Buenos Aires, Argentina (RB, AMI, MR, DRG, JM); and Division of Cardiology, University of Perugia School of Medicine, Perugia, Italy (GA).

Address correspondence to: José Milei, Instituto de Investigaciones Cardiológicas “Prof. Dr. Alberto C. Taquini” (ININCA) UBA-CONICET, Marcelo T de Alvear 2270 (C1122AAJ), Ciudad Autónoma de Buenos Aires, Argentina; e-mail: [ininca@fmed.uba.ar](mailto:ininca@fmed.uba.ar).

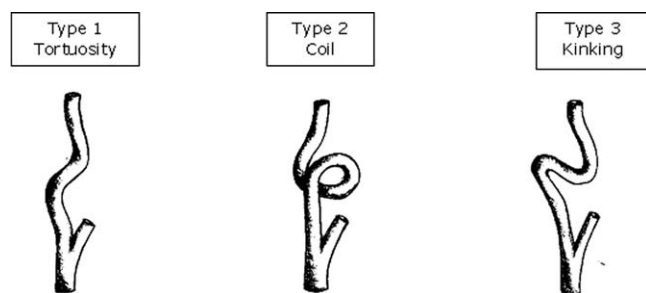


Figure 1. Types of dolichoarteriopathies, according to the definition of Weibel and Fields.<sup>3</sup>

dolichoarteriopathies as a mere anatomical variety, devoid of clinical consequences.<sup>13</sup>

Establishing the clinical impact of dolichoarteriopathies is further complicated by the fact that the mechanisms responsible for their formation are still debated. One theory maintains that they are pathological alterations caused by arterial aging and/or changes induced by atherosclerotic remodeling, which would cause the vessel to bend,<sup>4,14-17</sup> while other reports do not support an association between dolichoarteriopathies and cardiovascular risk status.<sup>13,18,19</sup> Alternatively, it has therefore been postulated that they have an embryological origin. Indeed, Kelly<sup>20</sup> observed that carotid arteries may be kinked or show loops at some point during intrauterine development, when the descent toward the mediastinum occurs enabling the union of the third aortic arch with the dorsal aorta.

Obviously, as these 2 etiological theories are so different from each other, they may also entail different implications both clinically and prognostically. Part of the uncertainty derives from the fact that previous observations report conflicting results, due to the fact that they had been made in small studies or in selected populations.<sup>13-19</sup> Therefore, it would be important to establish which theory has more solid basis.

The aim of the current study was to get additional insights into whether kinking- and coiling-type carotid dolichoarteriopathies are related to anomalies in embryological development or whether they develop as a consequence of atherosclerosis and/or in relation to cardiovascular risk factors. To this goal, we performed ultrasonographic investigation of the carotid territory in a large population of individuals who spanned across all ages, from newborn babies to >90 years, and in whom risk factors for atherosclerotic diseases were assessed. We reasoned that an embryological malformation would be detectable already in early childhood and be unrelated to

cardiovascular risk factors; conversely, if carotid dolichoarteriopathies are a manifestation of atherosclerotic vessel remodeling, their prevalence should increase with age and be associated with the presence of cardiovascular risk factors.

## Materials and Methods

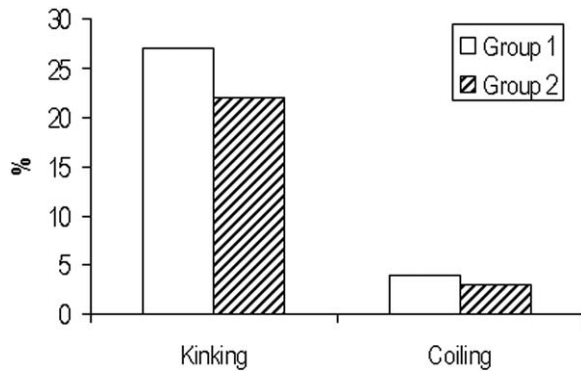
### Patient Population

This was an observational study, conducted on 885 participants of either sex, whose age ranged from newborn babies (4 hour 30 minutes) to 90 years. Patients were divided into 2 groups (G): G1 (control, healthy participants)  $n = 245$ . It consisted of infants, children, and adolescents up to 15 years of age (mean  $6 \pm 3$  years) from Marcos Paz, a town of 43 000 inhabitants just outside of Buenos Aires; these children participated in a voluntary screening health program, approved by their parents, under the patronage of the municipality of Marcos Paz that was performed in hospital and schools. Group 2 (G2;  $n = 640$ ) consisted of patients from 16 to 90 years of age (mean  $57 \pm 8$  years) in whom diagnostic color Doppler ultrasonography investigation of neck vessels had been requested for clinical suspicion of atherosclerotic disease. Patients were assessed with regard to presence of cardiovascular risk factors (hypertension, dyslipidemia, smoking). Presence of atheromatous plaques in the regions affected by dolichoarteriopathies was also evaluated.

### Methodology

Patients were studied in dorsal decubitus, with the head slightly turned toward the opposite side of the carotid artery under examination. An ATL Apogee CX echocardiographic machine, equipped with a 7.5-MHz transducer, was used. A 3.5-MHz transducer was used in case of high carotid bifurcations, to be able to examine the full stretch of the internal carotid artery.

Carotid artery study started by placing the transducer at the base of the neck and moving it up to the jaw. Conventional images were acquired on the longitudinal axis and then on the transverse axis; then, color Doppler mapping was performed; the study of each segment ended with pulsed Doppler. Images were stored on videotape and printed on black and white and color photographic paper. An



**Figure 2.** Prevalence of kinking (left bars) and of coiling (right bars) in G1 (control participants, open bars) and in G2 patients (hatched bars).

experienced investigator unaware of patients' clinical characteristics performed image analysis.

### Statistical Analysis

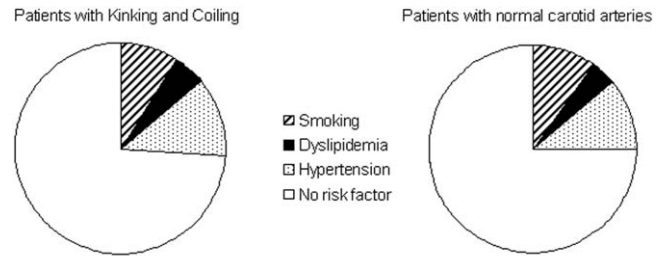
To assess statistical significance, univariate and multivariate analysis was performed using  $\chi^2$  test (Statistica 7 for Windows). Data are expressed as mean  $\pm$  standard deviation.

## Results

A total of 885 participants enrolled in the study, whose age ranged from 4.5 hours (youngest control participant) to 90 years (oldest patient). Participants in G1 ( $n = 245$ ; 52% males) averaged  $6 \pm 3$  years of age, while patients in G2 ( $n = 640$ ; 49% males)  $57 \pm 8$  years of age.

Coiling prevalence was similar in healthy participants (G1 4%;  $n = 10$ ) and in patients (G2 3%;  $n = 19$ ; NS; Figure 2). At the same time, kinking prevalence in G1 was 27% ( $n = 67$ ), and it was 22% ( $n = 143$ ) in G2, which did not show any statistically significant association either (Figure 2). Atheromatous plaques intrakinking were only observed in 3 G2 patients (0.47%). In this group, 56.2% of patients presented carotid atherosclerotic disease.

Within G2 patients, prevalence of cardiovascular risk factors evaluated individually was similar when patients were divided according to presence or absence of kinking and/or coiling (Figure 3).



**Figure 3.** Prevalence of cardiovascular risk factors in G2 patients, with (left chart) or without (right chart) carotid dolichoarteriopathies.

## Discussion

In this study, we report on the prevalence of kinking and coiling of carotid arteries (ie, dolichoarteriopathies) in a large number of participants whose age spanned from 1 day to 90 years, who underwent an echographic evaluation of extracranial carotid arteries, and in whom assessment of cardiovascular risk factors was performed. We observed these alterations with similar frequency across all ages, from newborn infants to elderly individuals. Furthermore, their prevalence was unrelated to the presence of cardiovascular risk factors or of frank atherosclerotic pathology of carotid artery. Collectively, these findings suggest that carotid dolichoarteriopathies are a result of alterations in embryological development rather than of vascular remodeling secondary to aging and/or atherosclerosis.

### Causes of Carotid Dolichoarteriopathies: Remodeling Versus Embryology

Reports on the prevalence of carotid dolichoarteriopathies in the general population show some variability (between 10% and 45%)<sup>6-8,13</sup> according to different diagnostic methodology and patients' inclusion criteria, but in aggregate they concur to indicate that this is a rather frequent finding. In contrast, lack of consensus exists over the clinical and prognostic significance of these alterations or even about their etiology. Much of the controversy revolves around a "nature-or-nurture" type of issue, that is, whether carotid dolichoarteriopathies develop late in life as a manifestation of vessel remodeling, particularly in individuals at risk of atherosclerosis, or rather they originate from alterations of embryological development.

With regard to the possibility that dolichoarteriopathies may be the result of a degenerative process,

over the years several hypotheses have been put forward trying to explain how they may develop. Some of these seem quite unlikely, such as the possibility that carotid kinking could be due to kyphosis or lordosis of the spine, which might deviate the carotid axis,<sup>21</sup> or that inflammation of the tissues around the carotid arteries would cause them to retract.<sup>22</sup> Other investigators have proposed that arterial hypertension would produce alterations in the wall over time, which would favor its weakening with subsequent kinking of the artery,<sup>14,16</sup> while other authors hypothesized a relationship between aging and arterial anatomical abnormalities.<sup>4,17</sup> However, those observations refer to early papers reporting small case series.

Data gathered in modern times also do not help in establishing firm conclusions. Two rather large reports by Ghilardi et al<sup>16</sup> and Del Corso et al<sup>23</sup> described a great prevalence of hypertension and atherosclerosis in patients with carotid dolichoarteriopathies; however, both studies lack a group of normal participants, and deal with a population of patients selected for vascular pathology, and in whom predominance of such cardiovascular risk factors is in fact expected. More recently, Pancera et al<sup>18</sup> have reported an association between carotid artery kinking and age and with hypertension as well. However, in that study, prevalence of carotid abnormalities was actually identical across the age groups from 60 to >80 years, which represented 87% of their cohort: younger patients, in whom prevalence was apparently lower, were instead too few to make a solid comparison. This same reasoning applies to the effect of hypertension reported in that study, which was actually based on about a dozen patients.<sup>18</sup>

The possibility that carotid dolichoarteriopathies may have an embryological origin had also been suggested in the past. Again, however, those reports were not conclusive. Sometimes, this was because of the small numbers of cases studied. In this respect, Weibel and Fields<sup>3</sup> described at angiography 14 cases of anatomical abnormalities in patients aged between 1 and 20 years, while Sarkari et al<sup>24</sup> reported 8 children (aged 9 months to 16 years) with symptomatic carotid kinkings and coilings. In a substantially larger study, there may have been a selection bias<sup>25</sup>; in that case, 282 angiographies of neck vessels were obtained in patients aged between 6 months and 82 years. The authors found that prevalence of carotid abnormalities in adults was 24% and even greater (43%) in children. Although that finding may seemingly support the view that in fact there is no association with aging

and that dolichoarteriopathies have an embryological origin, the significance of such a high frequency in children might have been restricted to the peculiar population studied, as it is quite conceivable that children who were subjected to an invasive procedure such as angiography underwent it because of a high clinical suspicion of carotid abnormalities. More recently, through noninvasive Doppler ultrasonography assessment, carotid dolichoarteriopathies were observed in 24.6% of a consecutive patient series, with no apparent relation between carotid alterations and cardiovascular risk factors.<sup>13</sup> That report is in agreement with our findings. However, it should be emphasized that, unlike the current study, there was no precise identification of patients' age (younger or older than 60 years); furthermore, there was no control group of otherwise healthy participants.

To our knowledge, the data presented in the current study are the first systematic description of carotid dolichoarteriopathies in a rather large cohort that included healthy individuals and was performed on participants of various ages, from young babies to very elderly individuals. As such, our findings exclude a degenerative mechanism as the etiology of carotid alterations and support the hypothesis that carotid dolichoarteriopathies represent embryological malformations.

### Possible Embryological Mechanisms

In 1924, Cairney<sup>26</sup> had already reported autoptic findings in fetuses from the fifth month in whom morphological carotid arteries abnormalities were observed. In this respect, it is important to notice that the vascular wall in fetus develops from mesenchymal cells islets; in any artery, the tunica muscularis develops first in the main trunk and later in its branches. The proximal portion of the internal carotid artery originates from the third aortic arch, while the more distal parts originate from the left dorsal aorta.<sup>21</sup> Harrison and Dávalos<sup>21</sup> suggested that development of carotid arteries and of skeletal system might be asynchronous, the different velocity explaining the tortuous path. Ochsner et al<sup>27</sup> proposed that fibromuscular dysplasia occurring during fetal life, located in a sector of the carotid artery, would be responsible for subsequent weakening and kinking of the wall at that level. Contrary to this specific hypothesis, however, is the fact that presence of fibromuscular dysplasia in areas with dolichoarteriopathies is very rare.<sup>28-30</sup>

## Conclusions

According to the results of this research, carotid dolichoarteriopathies would have an embryological origin. This is based on the similar prevalence in newborn infants, children, adolescents, and adults. This conclusion is also supported by the lack of correlation with cardiovascular risk factors. Thus, their definition as true dysembryoplasias seems to be justified.

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