

Research Article – Basic and Applied Anatomy

Multi-detector computed tomography in the evaluation of variants and anomalies of aortic arch and its branching pattern

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Abstract

Objective: Evaluate the prevalence of aortic arch anatomy and branching pattern variants and anomalies in 1359 patients by Multi-Detector Computed Tomography Angiography.

Methods: Retrospective multi-centric study including exams performed for various clinical issues with the same acquisition and injection protocols on 64-slices scanners. Multi-Planar reformations and Volume Rendering Images were available in all cases.

Results: A total of 965 patients (71%) had normal aortic arch anatomy and branching pattern. Anatomical variations and anomalies were present in the remaining 394 patients (29%). The most common variation was the common origin of the brachiocephalic and the left common carotid arteries (302/1359 cases, about 22%). The most common anomaly were aberrant right subclavian artery (4/1359, about 0.5%) and right aortic arch (4/1359 cases, about 0.5%).

Conclusions: Our multicentric series is, as far as we know, the largest reported to date for the definition of the prevalence of variation and anomalies of aortic arch anatomy and branching pattern. Knowledge of such findings may avoid both in the immediate and in the long term an injury to the patient, decreasing the rate of complications during surgery and interventional radiology procedures in the head and neck district.

Key words

Aortic arch anomalies, aortic arch variants, aortic anatomy, multi-detector computed tomography.

Introduction

Development of the aorta takes place during the third week of gestation; it is a complex process associated with the formation of the endothelial heart tube. During the embryonic development, six pairs of aortic arches connect the two primitive ventral and dorsal aortas, the fifth being rudimentary. The ventral aorta becomes the ascending aorta, the dorsal aorta originates the descending thoracic aorta and the fourth left aortic arch forms the normal aortic arch. The majority of the first, second,

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and fifth arches regress. The third aortic arches form the beginning of the internal carotid arteries. The right fourth arch involutes beyond the beginning of the subclavian artery. The right sixth arch disappears except a minor portion becoming part of the right pulmonary artery; the left sixth arch forms the ductus arteriosus. Finally, the seventh intersegmental arteries persist, migrate cephalically and form the subclavian arteries. Regression of the right dorsal aorta between the right subclavian artery and the descending aorta leaves the left aortic arch as normal condition (Sadler and Thomas, 2010).

The classic left aortic arch and descending thoracic aorta are seen in 70% individuals. The three main branches of the aortic arch are the brachiocephalic (or innominate) artery (dividing into the right subclavian and common carotid arteries), the left common carotid artery and the left subclavian artery. The incidence of this pattern is reported to be between 48% and 84% (Standring, 2008), with the variation largely attributable to differences between white and black individuals. Malformations of the aortic arch system can be explained by persistence of segments of the aortic arches that normally regress, disappearance of segments that normally remain, different growth rates in the various arteries, anomalous "migration" and "merging" of the branches (Standring, 2008; Sadler and Thomas, 2010).

Anomalies of the aortic arch rarely compromise health since childhood (Ramos-Duran et al., 2012). Sometimes they are associated with symptoms that occur in young adults or even in the elderly, of diverse nature and unpredictable entity depending on the vessel involved and on the disease. Much more frequently anomalies are completely asymptomatic, constituting an incidental finding in young adults or elderly during imaging studies performed for other reasons (Natsis et al., 2009).

The increasingly extensive use of multi-detector row computed tomography angiography (MDCTA, now widely recognized as the diagnostic gold standard in this field: Webb et al., 1982; Natsis et al., 2009; Jakanani and Adair, 2010) for the most varied clinical issues (e. g. cancer staging, polytrauma), has increased the number of incidentally found vascular variations and anomalies (Webb et al., 1982; Jakanani and Adair, 2010). The non-reporting of such findings, resulting most often from lack of knowledge rather than inattention of radiologists, may cause injury to the patient both immediately and in the long term, increasing the rate of complications during surgery and interventional radiology in the head and neck district (Berko et al., 2009). The present study describes the prevalence of variants and anomalies of the aortic arch branching pattern in a consecutive series of adult patients studied by MDCTA.

Materials and methods

We retrospectively reviewed 1383 consecutive CT angiograms (CTAs) of the chest performed in adult patients for the staging of malignancies ($n = 523$), polytrauma ($n = 371$), bronchopneumonia ($n = 118$), suspicion of aortic dissection ($n = 125$), haemoptysis ($n = 86$), dysphagia ($n = 40$), dyspnea ($n = 32$) and other issues ($n = 88$) between March 2010 and October 2012. All CTAs were performed on the 64-slices scanners at the Diagnostic Imaging Departments of the second University of Naples, Italy (Aquilion 64, Toshiba, Shah Alam, Malaysia), of the hospital "Vincenzo Monaldi", Naples, Italy (GE Lightspeed, General Electrics, Fairfield, CT), and of the

clinic "Pineta Grande", Castel Volturno, Italy (Somatom Sensation, Siemens, Berlin, Germany). The study protocol was the same in all cases, using a volumetric acquisition technique before and during the administration of a iodine-based medium contrast (Iomeron, Bracco Imaging Italia, Milano, Italy; 370-400 mg/mL - 2 mL/kg, 3 mL/sec). A bolus tracking technique was used placing the region of interest on the ascending aorta. Images acquired in an early arterial phase were available in all cases. The data were reformatted with the use of Multiplanar Reconstructions (MPR), Maximum Intensity Projections (MIP) and Volume Rendering (VR) techniques. No CTA was electrocardiogram-gated.

Twenty-four studies were technically limited and therefore were excluded. Hence, the study population was composed of the remaining 1359 CTAs (558 females, 801 males; median age 48.3 years; age range 17-89 years). The vast majority of the patients were caucasian (1196, 88%); 82 patients were black (6%), 43 Asian (3%) and 38 of mixed origin (3%).

Each case was jointly reviewed by three groups of radiologists experienced in reporting cardio-thoracic CT exams (S.C. and F.I. at the second University of Naples, G.R. and T.V. at the hospital "Vincenzo Monaldi", M.S. and G.S. at the clinic "Pineta Grande").

Attention was paid to aortic arch anatomy and branching pattern, documenting, when present, anatomical variants and anomalies. The latter terms were used on the basis of prevalence in the general population and functional significance: we have indicated as "anatomical variant" an anatomical situation different from what is observed in most subjects but not associated with clinically relevant symptoms, and as "anomaly" a marked deviation from normality, resulting from congenital or hereditary defects and susceptible to cause symptoms.

This study was approved by the institutional review boards of the three participating centres. Informed consent was not required for this retrospective review.

Upon determination of the prevalence of normal anatomy, anatomical variations and anomalies, we used Fisher's exact test for comparison of proportions and Mann-Whitney's U test for differences in median values. A value of $P < 0.05$ was considered statistically significant.

Results

A total of 965 patients (71%) had normal aortic arch anatomy and branching pattern. Anatomical variations and anomalies were present in the remaining 394 patients (29%). The most common variations were: common origin of brachiocephalic trunk and left common carotid artery (302/1359 cases, 22%, Fig. 1), direct origin of left vertebral artery from the aortic arch (45/1359 cases, 3%, Fig. 2), origin of left common carotid artery from the innominate artery (21/1359, 1.5%) and the presence of bilateral brachiocephalic arteries (BBA, 13/1359 cases, 1%).

The most common anomalies were: aberrant right subclavian artery (4/1359, 0.5%, Fig. 3, 4), aberrant left subclavian artery (/1359, 0.2%), right aortic arch (RAA, 4/1359 cases, 0.5%, Fig. 5), double aortic arch (DAA, 1/1359 cases, 0.1%, Fig. 6) and Kommerell's diverticulum (Fig. 7).

Of the patients with variations and anomalies, 156 (39%) were women and 238 (61%) were men, similar to the sex distribution in the overall patient population. The



Figure 1 – Maximum intensity projection coronal image of a case of common origin of the brachiocephalic and left common carotid arteries. The superior vena cava is the most opacified structure in this phase of the study, since the contrast medium was injected through an arm vein. Contrast enhancement of arteries is less intense.

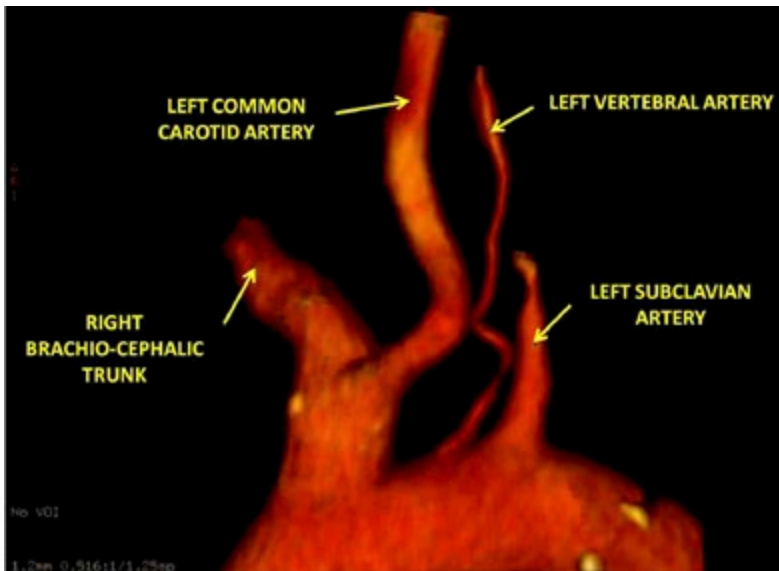


Figure 2 – Volume rendering image displaying a case of common origin of the brachiocephalic and left common carotid arteries and aberrant origin of the left vertebral artery. The aberrant vessel directly originates from the aortic arch, between the common origin of the right brachiocephalic and left common carotid arteries and the origin of the left subclavian artery.

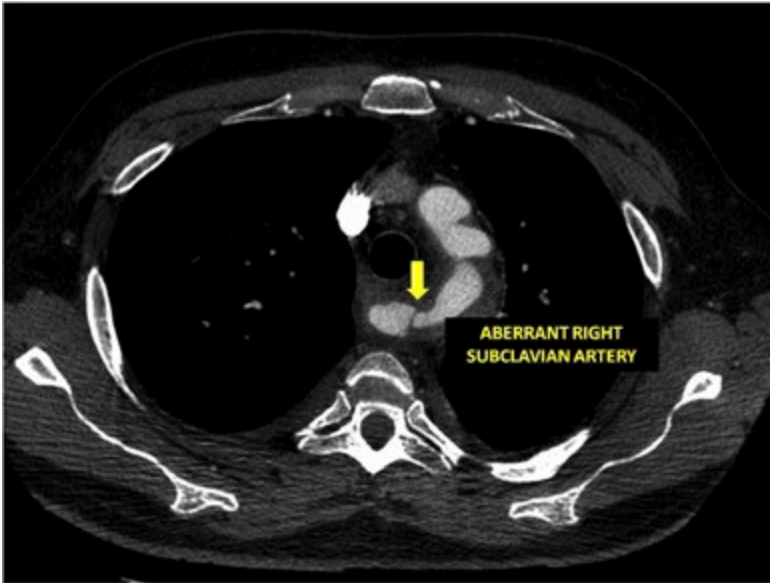


Figure 3 – Axial image from a case of aortic dissection involving an aberrant retroesophageal right subclavian artery.

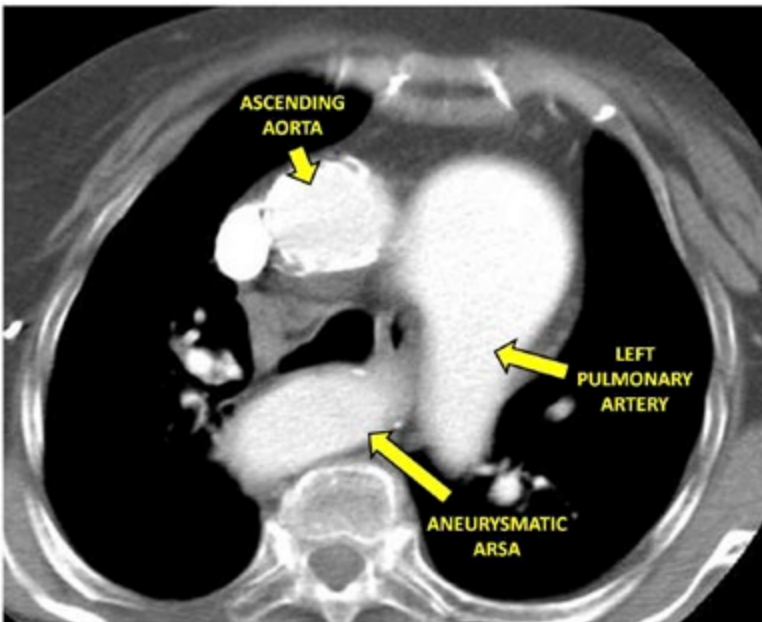


Figure 4 – Aneurysmatic retroesophageal aberrant right subclavian artery (ARSA): the patient complained of worsening dysphagia.

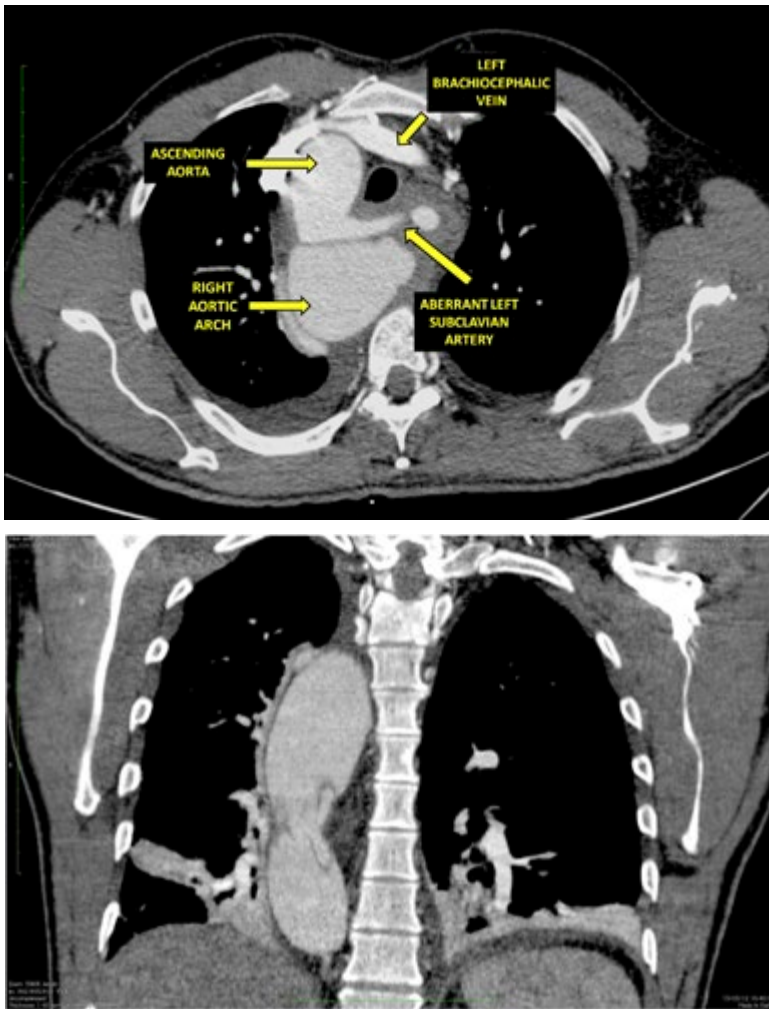


Figure 5 – Axial (a) and coronal (b) images of a dissected right aortic arch.

median age of patients with anomalies was 52.6 years (range 19-82 years), not significantly different from the median age in the overall population.

Discussion

Despite the superiority of MDCTA to evaluate variants and anomalies of the aortic arch and of its branching pattern (Webb et al., 1982; Jakanani and Adair, 2010), only a few studies have been published to date on this issue, most of which are limited by the small number of cases (Bhatia et al., 2005; Gupta and Sodhi, 2005; Mül-

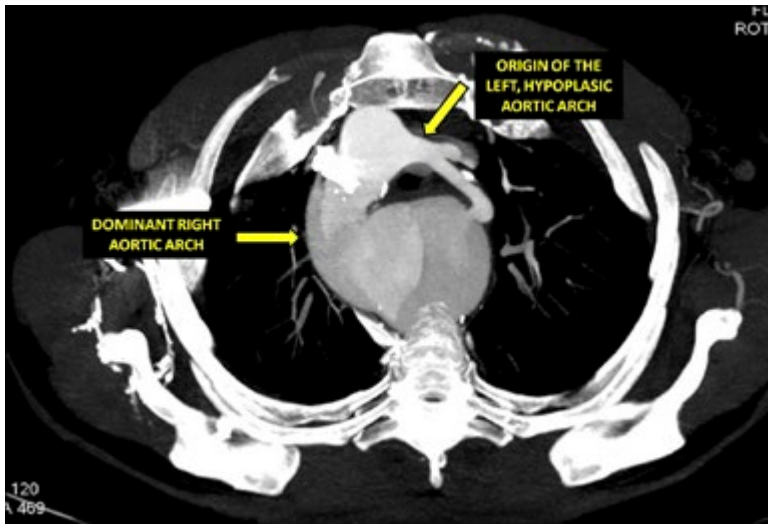


Figure 6 – Axial maximum intensity projection image in a case of double aortic arch (right arch dominant, left arch hypoplastic).

ler et al., 2011). Berko et al. (2009), in the only study significant from the statistical point of view, reviewed 1000 CTAs of the chest: abnormalities were found in 34% cases, with the bovine trunk reported as the most frequent (27% cases), similarly to what observed in our study. Our multicentric series is, as far as we know, the largest reported to date on this topic.

Normal anatomy and branching pattern of the aortic arch

The most common aortic arch branching pattern in humans consists of three great vessels originating from the arch. The first vessel is the innominate artery, which branches into the right subclavian and common carotid arteries; the second is the left common carotid artery; the last one is the left subclavian artery. Vertebral arteries normally originate from the ipsilateral subclavian arteries (Standing, 2008). The vast majority of the patients in our series ($n = 965$, 71%) had normal aortic arch anatomy and branching pattern.

The so-called “bovine arch”

A common origin of the brachiocephalic and the left common carotid arteries is the most common variant in the branching pattern of the aortic arch, occurring in 10-27% individuals (Layton et al., 2006; Faggioli et al., 2007; Berko et al., 2009; Hornick et al., 2012) and accounting for more than two thirds of all branching anomalies; this pattern is found more often in blacks, with previous cadaveric reports documenting occurrence in 25% blacks (Tranquilli and Elefteriades, 2012) and in only 8% whites (Layton et al., 2006). In our series, the incidence was 22% (302/1359 cases).

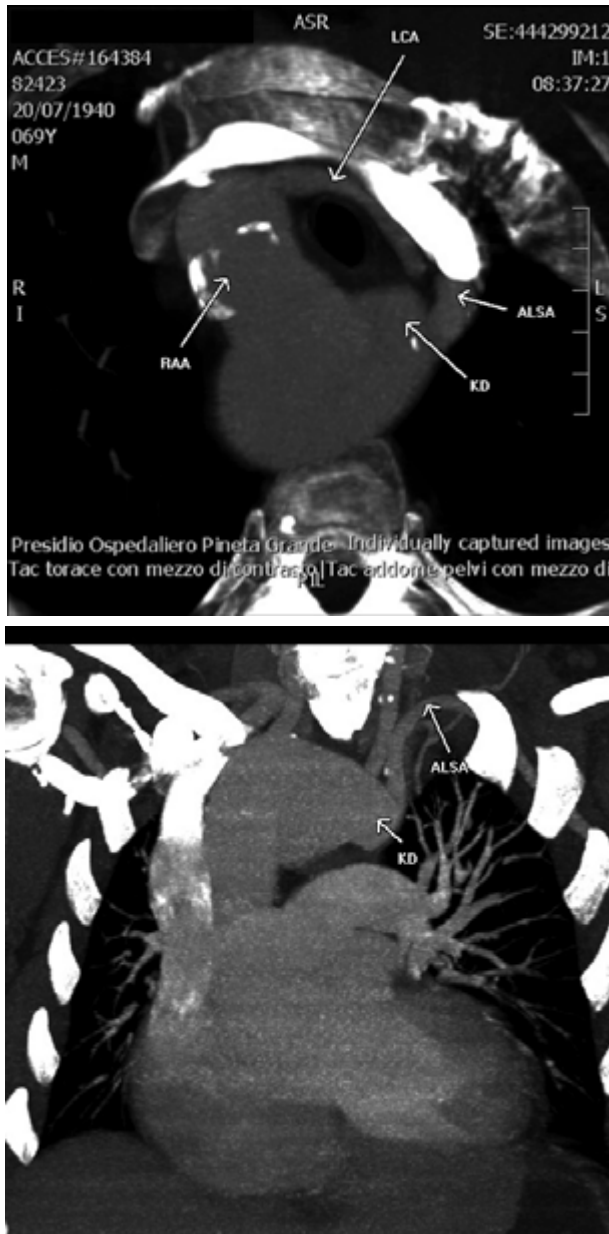


Figure 7 – Maximum intensity projection axial (a) and coronal (b) images obtained in a case of right aortic arch with Kommerell's diverticulum. In b the origin of the aberrant left subclavian artery from the diverticulum is well appreciable (RAA: right aortic arch, LCA: left carotid artery, ALSA: aberrant left subclavian artery, KD: Kommerell's diverticle, ASR: anterior-superior-right, PIL: posterior-inferior-left, RI: right-inferior, LS: left-superior). The superior vena cava and the left brachiocephalic vein are the most opacified structure in this phase of the study, since the contrast medium injected through an arm vein. Contrast enhancement of arteries is less intense.

Although this branching pattern is commonly referred to as “bovine arch”, this name is inappropriate because this is not the branching pattern found in cows (Layton et al., 2006). In cattle, in fact, a single great vessel originates from the aortic arch; this large brachiocephalic trunk gives rise to both subclavian arteries and a bicarotid trunk, which then bifurcates into the left and right common carotid arteries. The presence of a single long vessel originating from the aortic arch is a common occurrence in animals with deep chests: the general belief among veterinary anatomists is that the long distance from the aortic arch to the thoracic inlet explains why all the great vessels originate from the arch as a single vessel, the brachiocephalic trunk. A common origin of the innominate and left common carotid arteries, however, is found in several other species, including cats, dogs, and rabbits, leading to the suggestion that this branching pattern should more correctly be designated “feline”, “canine” or “rabbit” arch (Layton et al., 2006; Hornick et al., 2012).

A similar but less common variant, also erroneously referred to as “bovine arch”, occurs when the left common carotid artery originates directly from the innominate artery rather than as a common trunk (origin of the left common carotid artery from the innominate artery). The left common carotid artery originates off the brachiocephalic trunk at an average distance of less than 1 cm from the aortic arch, with the maximal distance being 2.5 cm. This variant also occurs more commonly in blacks (10%) than in whites (5%), with an overall rate of 9% in the general population (Layton et al., 2006; Hornick et al., 2012).

Aberrant origin of vertebral arteries

Although the left vertebral artery is classically described as the first branch of the ipsilateral subclavian artery, multiple variations in the origin of this vessel have been reported in the literature (Goray et al., 2005). This artery not infrequently arises directly from the aortic arch, with reported prevalence of 3.3-7.4%; incidence in our series was 3%. When it originates directly from the arch, the left vertebral artery usually enters the transverse foramen of the fourth or fifth, rather than of the sixth cervical vertebra (Albayram et al., 2002; Goray et al., 2005). The most frequent location of origin of a left vertebral artery is between the left common carotid and subclavian arteries (Standring, 2008). Less frequently, this vessel may arise from the common, internal or external carotid arteries or from subclavian branches, such as the thyrocervical trunk. Rarely, the proximal left vertebral artery is duplicated (Türkvatan et al., 2009). In this case, one part arises from the arch and the other from the left subclavian, or both originate from the aortic arch. While the direct origin of the left vertebral artery from the aortic arch is a not uncommon finding in clinical practice, the direct origin from aorta of the right vertebral artery is a rare anatomical variant. In such instances, the right vertebral artery generally arises distal to the supraortic trunks (Standring, 2008). In our series we never found this anomaly.

Bilateral brachiocephalic arteries

Bilateral brachiocephalic arteries, present in 1.2% to 2% cases of previous autopsy series (Standring, 2008; Natsis et al., 2009; Restrepo et al., 2012), was found in 1% patients of this series.

Aberrant right subclavian artery arising from a left aortic arch

An aberrant right subclavian artery is the most common vascular ring anomaly (with the latter term are indicated variants or anomalies of aortic arch anatomy and branching pattern leading to “encirclement” and compression of trachea and esophagus) in the aortic arch. Its prevalence in previous studies ranged from 0.4% to 2%, with the aberrant artery being the last branch of the left aortic arch in 1% individuals (Ka-Tak et al., 2007; Natsis et al., 2009; Guzman and Eagleton, 2012): its prevalence was 0.5% in the present series. The course of the aberrant artery, usually emerging from the descending aorta rather than from the aortic arch (from which the right common carotid artery, the left common carotid artery and the left subclavian artery originate in sequence), is usually retroesophageal (80% cases), as also occurred in every case in this series; less frequently the aberrant right subclavian artery may run between oesophagus and trachea, anterior to trachea or anterior to main bronchus (Ka-Tak et al., 2007; Guzman and Eagleton, 2012). With regard to the embryological basis of aberrant subclavian arteries, Edwards (1977) hypothesized an origin at an early stage of development when there is still a bilateral aortic arch and before the separation between the definitive aortic arch and ductus arteriosus takes place. In this model, the right carotid and subclavian arteries would arise from the right arch and the left carotid and subclavian arteries would originate from the left arch. The descending aorta is on the midline. Interruption of this arch system at different locations can explain the various aortic arch anomalies. The normal arch system results from interruption of the dorsal segment of the right arch between the right subclavian artery and descending aorta. An aberrant right subclavian artery occurs as a result of the interruption between the right carotid and the right subclavian arteries, while a right aortic arch with an aberrant left subclavian artery, a much rarer condition (0.2% in our series), results from interruption of the left arch between the left common carotid and left subclavian arteries in the developing double aortic arch (Ka-Tak et al., 2007; Guzman and Eagleton, 2012).

Right aortic arch

A right aortic arch has been reported to occur in approximately 0.1% of the population (Kanne and Godwin, 2012) and in 0.5% of our series; it results from the persistence of the right fourth aortic arch (Sadler and Thomas, 2010). The two main types of right aortic arch, classified according to branching patterns, include mirror image branching (left brachiocephalic, right common carotid and right subclavian arteries, usually associated with cyanotic congenital heart disease and present in 20% to 25% of patients with tetralogy of Fallot: Ramos-Duran et al., 2012) and aberrant left subclavian artery (the most common type of right aortic arch, rarely associated with congenital heart disease, in which left common carotid, right common carotid, right subclavian and left subclavian arteries successively emerge from the arch) (Kanne and Godwin, 2010). The latter variant is usually asymptomatic; symptoms, however, may arise from vascular ring formation (Natsis et al., 2009; Ramos-Duran et al., 2012).

Double aortic arch. This is a relatively rare congenital cardiovascular malformation (0.1% in our series) in which two aortic arches form a complete vascular ring that can compress the trachea and/or oesophagus (Natsis et al., 2009; Türkvatan et al.,

2009). Most commonly there is a larger (dominant) right arch behind and a smaller, hypoplastic left aortic arch in front of the tracheo-esophageal complex. The two arches join to form the descending aorta which is usually on the left side. In some cases the end of the smaller left aortic arch occludes (left atretic arch) and the vascular tissue becomes a fibrous cord. Although in these cases a complete ring of two patent aortic arches is not present, the term “vascular ring” is the accepted generic term even for this anomaly (Türkvatan et al., 2009).

Kommerell's diverticulum

This is a dilatation of the proximal portion of an aberrant subclavian artery near its origin from the aorta (Natsis et al., 2009; Türkvatan et al., 2009; Jakanani and Adair, 2010). It represents the remnant of the distal right aortic arch in cases of left aortic arch with aberrant right subclavian artery, and the remnant of the left dorsal aortic root in cases of right aortic arch with aberrant left subclavian artery (Sadler and Thomas, 2010). Kommerell's diverticulum is commonly (60%) associated with aberrant right subclavian artery.

More than twenty different aortic arch configurations have been described but those specifically described above are by far the most commonly encountered (Natsis et al., 2009; Jakanani and Adair, 2010; Restrepo et al., 2012). Identification of anomalies of the aortic arch anatomy and branching pattern may be important before vascular surgery in the head, neck and mediastinum regions, such as aortic arch surgery, in order to avoid inadvertent vascular injury and optimize access for vascular anastomosis during surgery and four-vessel cerebral angiography. Almost all the anomalies mentioned above may be associated with an increased rate of technical failure and neurological complications in carotid artery stenting procedures and their reporting in diagnostic tests (*e. g.* computed tomography angiography, magnetic resonance angiography, Doppler ultrasonography) or recognition during interventional angiography are key elements for therapeutic success and the welfare of the patient (Faggioli et al., 2007; Solomon et al., 2010; Dahm et al., 2011; Ventoruzzo et al., 2012; Barbiero et al., 2013).

Acknowledgements and conflict of interest statement

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