Double incomplete aortic arch and Kommerell’s Diverticulum as a cause of chronic cough


Head of Cardiology at American British Cowdray Medical Center, Santa Fe Campus, Mexico, D.F., Mexico
Pre-grade Intern of the American British Cowdray Medical Center, Mexico, D.F., Mexico
Staff Physician of Radiology and Molecular Imaging Department at American British Cowdray Medical Center, Mexico, D.F., Mexico
Staff Physician of Pediatrics Department at American British Cowdray Medical Center, Mexico, D.F., Mexico
Head of Cardiac Surgery at American British Cowdray Medical Center, Mexico, D.F., Mexico

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Abstract Vascular rings which can cause symptoms related the trachea and esophagus compression occur in less than 1% of all cardiovascular malformations. Double incomplete aortic arch with right-sided aorta and aberrant left subclavian artery is the rarest one, and its present in 0.04–0.1% of autopsy series. A case of this malformation with a Kommerell’s Diverticulum is presented. This diverticulum has risk of severe complications such as dissection and/or rupture.

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PALABRAS CLAVE
ARM-Contrastada; RMC; Divertículo de Kommerell; Doble Arco Aórtico Incompleto; Arco Aórtico a la Derecha
Vascular rings which can cause symptoms related the trachea and esophagus compression occur in less than 1% of all cardiovascular malformations and usually associates with others left sided ones, highlighting the importance of a comprehensive approach of the heart and the vascular structures in the same study, such as CMR to plan the surgical approach.

Double incomplete aortic arch (DIAoA) with right-sided aorta (RSAoA) and aberrant left subclavian artery (ALSA) is the rarest one, and its present in 0.04–0.1% of autopsy series.

We present a case of a one-year-old boy with chronic cough and difficulty for the feeding progression process, an out-site barium’s swallow reported extrinsic compression of the esophagus. Due to radiation-safety concerns, a CMR was

**Figure 1** CMR of the aorta. SSFP fixed cine image axial view at aortic arch level and just distal (A) shown a double incomplete aortic arch with right-sided aorta and a KD (B). An axial T2-W image showed compression of the trachea (T) and the esophagus (E) by vascular structures (C). CMR, cardiac magnetic resonance; SSFP, steady state free precession; T2-W, T2-weighted; CE-MRA, contrast-enhanced magnetic resonance angiography; MIP, maximum intensity projection; 3D VR, 3-dimensional volume rendering; Ao, aorta; DIAoA, double incomplete aortic arch; RSAoA, right sided aortic arch; ALSA, aberrant left subclavian artery; KD, Kommerell’s Diverticulum.

**Figure 2** CMR of the aorta. ALSA originates from the incomplete left aortic arch as shown in the CE-MRA on MIP (A) and 3D VR reconstructions (B). The KD originating from the descending aorta is shown in (B). A coronal SSFP view shows "V" shape of the double incomplete aortic arch (C). CMR, cardiac magnetic resonance; SSFP, steady state free precession; T2-W, T2-weighted; CE-MRA, contrast-enhanced magnetic resonance angiography; MIP, maximum intensity projection; 3D VR, 3-dimensional volume rendering; Ao, aorta; DIAoA, double incomplete aortic arch; RSAoA, right sided aortic arch; ALSA, aberrant left subclavian artery; KD, Kommerell’s Diverticulum.
performed; a DIAoA with RSAoA and a Kommerell’s Diver-
ticulum (KD) were seen, which showed compression of the
trachea and the esophagus by vascular structures. ALSA ori-
ginates from the incomplete left aortic arch.

The RSAoA develops when the fourth left aortic arch invo-
lute and the right one persists.\textsuperscript{3,4} When an ALSA exists, it
can create an aneurysmatic vascular dilatation, known as
KD, which can be concomitant to the double aortic arch
(DAA).\textsuperscript{2} The KD represents the persistency of the distal seg-
ment of DAA, generally the left one which proximal segment
is atretic or disappears.\textsuperscript{1} There are three KD types described,
the second one is the rarest one and it forms when the KD
coexists with RSAoA and ALSA (Figs. 1 and 2).\textsuperscript{3-6}

The KD has risk of severe complications such as dissection
and/or rupture.\textsuperscript{7}

Ethical responsibilities

Data confidentiality. The authors declare that they have
followed the protocols of the workplace on the publication
of patient data.

Right to privacy and informed consent. The authors
declare that no patient data appear in this article.

Protection of human subjects and animals in research.
The authors declare that no experiments were performed
on humans or animals for this study.

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