*Cardiology in the Young*

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# Merged bilateral arterial duct and circumflex retroesophageal right aortic arch in a fetus with normal intracardiac anatomy

Saïd Bichali[1,2], Romulus Cristian Grigorescu[3], Mathilde Lefebvre[3], Charlotte Oriot[4], Ferdinand Dhombres[5], Damien Bonnet[1,6] and Lucile Houyel[1,6]

[1]Pediatric Cardiology, Centre de Référence Malformations Cardiaques Congénitales Complexes – M3C, Necker Hospital for Sick Children, Assistance Publique des Hôpitaux de Paris, Paris, France; [2]Pediatric Cardiology, Hôpital Femme-Enfant-Adolescent, Centre Hospitalier Universitaire de Nantes, Nantes, France; [3]Foetopathology, Hôpital Armand-Trousseau, Assistance Publique des Hôpitaux de Paris, Paris, France; [4]Pediatrics, Hôpital Femme-Enfant-Adolescent, Centre Hospitalier Universitaire de Nantes, Nantes, France; [5]Obstetrics, Hôpital Armand-Trousseau, Assistance Publique des Hôpitaux de Paris, Paris, France and [6]Université Paris Descartes, Sorbonne Paris Cité, Paris, France

## Brief Report

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**Author for correspondence:**
Saïd Bichali, Hôpital Femme-Enfant-Adolescent, 38 Boulevard Jean Monnet, 44093 Nantes, 0033240083333, France.
E-mail: said_b91@hotmail.fr

**Abstract**

We report the case of a fetus with anamnios sequence and VACTERL syndrome, having a circumflex right aortic arch. Two arterial ducts join anteriorly to form a common vessel that connects to the pulmonary trunk with confluent pulmonary branches. Embryologically, the dorsal right 6th aortic arch did not disappear and the aortic arch development stopped in a symmetrical state with an exceptional "Y-shaped" merged bilateral arterial duct.

Bilateral ducts are not usually seen with confluent pulmonary arteries. We report, however, the case of a fetus with confluent pulmonary arteries and an exceptional merged bilateral duct associated with a retro-esophageal circumflex right aortic arch. The aortic arch encircles the trachea and oesophagus from the right with a left descending aorta.[1] We successively describe the case, discuss the findings, and suggest a developmental hypothesis to this aortic arch and its association with VACTERL syndrome.

## Clinical case

A woman aged 30 years old, who has had a normal child born from a first pregnancy, then an extra-uterine pregnancy, had a screening ultrasound scan at 23 weeks gestation that showed anamnios and bilateral renal agenesis, while the first trimester scan was considered normal. The parents chose to terminate pregnancy at 24 weeks.

The male fetus weighed 559 g, and pathological examination showed extra-cardiac malformations. These included facial dysmorphy, limbs and extremities malposition with anamnios sequence, single umbilical artery, thoracic vertebral abnormalities, anal imperforation, right kidney agenesis, left kidney multicystic dysplasia and hypoplasia, and pulmonary hypoplasia. These abnormalities suggested VACTERL syndrome.

## Results

Fetal cardiac examination showed a circumflex right aortic arch with a retro-esophageal segment and a left descending aorta (Fig 1). The right aortic arch gave successively a common carotid trunk dividing into a left and a right carotid artery, a right subclavian artery, and a distal aberrant left subclavian artery.

A bilateral arterial duct had a right branch originating from the posterior part of the transverse retro-esophageal aortic arch and a left branch originating from the left subclavian artery. These two branches merged to form a unique vessel connecting to the pulmonary trunk. This trunk was normally divided into confluent right and left pulmonary arteries.

In addition, there was an anomalous course of the left brachiocephalic vein, passing below the aortic arch (retro-aortic) instead of antero-superiorly.

The heart had a normal inner architecture and a normal volume.

## Discussion

This is a very rare description of a "Y-shaped" merged bilateral arterial duct, associated here with a circumflex right aortic arch.

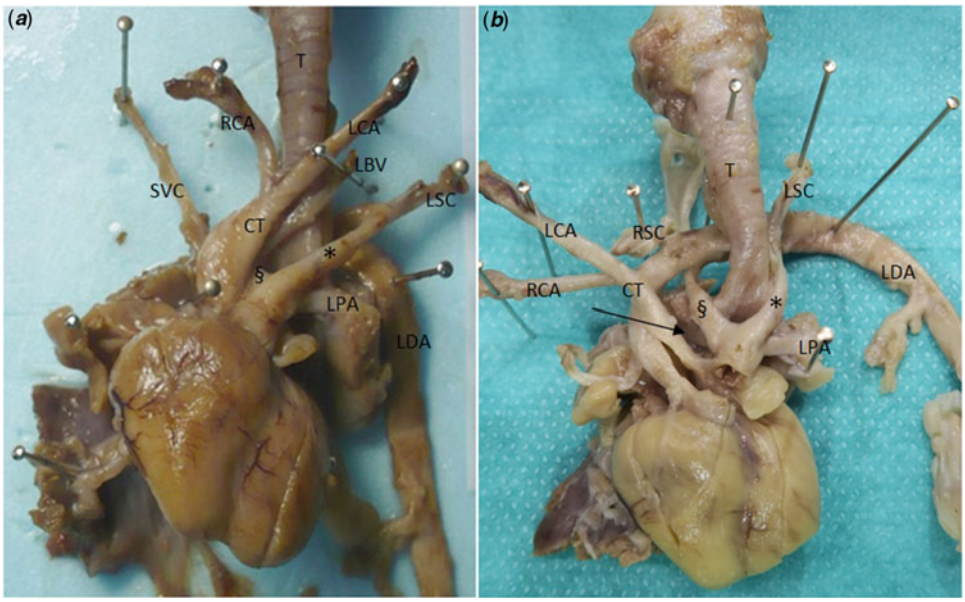


**Figure 1.** Anterior views of the heart and the great vessels before (***a***) and after (***b***) having reflected the aorta to show the right pulmonary artery (arrow) and the posterior insertion of the right arterial duct (§). Note the left arterial duct (*) joining the right one. CT = carotid trunk; LBV = left brachiocephalic vein; LCA = left carotid artery; LDA = left descending aorta; LPA = left pulmonary artery; LSC = left subclavian artery; RCA = right carotid artery; RSC = right subclavian artery; SVC = superior caval vein; T = trachea.

Bilateral arterial ducts are usually associated with non-confluent pulmonary arteries, sometimes isolated or most often in the setting of tetralogy of Fallot with pulmonary atresia.[1,2] These bilateral arterial ducts can be associated with left, right, or double aortic arch with or without isolated or aberrant brachiocephalic arterial trunk or subclavian artery. The aortic arch can also be interrupted.

Circumflex retro-esophageal right aortic arch is characterised by the right-sided arch crossing the midline behind the esophagus, with a left-sided descending aorta. Rarely, the opposite, or mirror-image, is found: left-sided aortic arch with a retro-esophageal segment and right descending aorta.[3] Usually in circumflex right aortic arch, the most proximal vessel arising from the aorta is the left carotid artery, then the right carotid artery, followed by the right subclavian artery. The most distal vessel is usually the left subclavian artery.[4] A particularity in our case is that the left and right carotid arteries originate from a common carotid trunk.

This malformation has been recently added to the original Stewart classification of congenital aortic arch abnormalities.[1] It is relatively rare among cases of right aortic arch particularly in those discovered during childhood.[5–7] Indeed, a circumflex aortic arch is frequently isolated, hence poorly diagnosed early in age, whereas a right aortic arch with mirror-imaged branching of the brachiocephalic vessels and without retro-esophageal segment is often associated with so-called "conotruncal defects" like tetralogy of Fallot with or without pulmonary atresia and common arterial trunk, with or without microdeletion 22q1.1.[6,8] Association between a circumflex aortic arch and an intra-cardiac lesion is rare but not exceptional: ventricular septal defect has been reported.[4,6]

A circumflex aortic arch can sometimes be discovered upon symptoms of esophageal and tracheo-bronchial compression, especially in adults.

Of note, a retro-aortic left brachiocephalic vein, as found in our case, is frequently associated with a right aortic arch.[9]

## Developmental hypothesis

Embryologically, the aortic arches arise from the aortic sac.[10] The most cranial is the 3rd aortic arch (which forms the two carotid arteries), then the 4th aortic arch (the right one usually disappears, and the left one forms the segment of the aortic arch between the left carotid artery and the left subclavian artery), then the 6th aortic arch. The 6th aortic arch forms the two arterial ducts. The right duct or right 6th arch artery normally disappears, while the right-sided dorsal aorta involutes.[10] The two pulmonary arteries originate from the caudal margin of the 6th aortic arch and then connect to the primitive intrapulmonary arteries and the pulmonary trunk. The pulmonary trunk and the ascending aorta derive from the distal part of the outflow tract and fuse with the aortic sac at the margins of the pericardial cavity. The rare anomaly found in our case could thus be explained on the basis of abnormal remodeling of the aortic sac: the caudal part of the aortic sac has retained its connection with the pulmonary trunk which then feeds directly the persisting 6th arch arteries and the right and left pulmonary arteries. This preserved connection provides a common origin for the 6th arch arteries themselves, the origins of the 6th and 4th arch arteries being separated. This symmetrical state, associated with the fusion of the two 3rd aortic arches in a common carotid trunk, is explained by the preservation of a stage of normal development, the right-sided structures retaining their size compared to the left-sided components. In addition, the left 4th aortic arch disappears (region 2 of the Edwards hypothetical double arch, Fig 2) and regions 3 and 4 persist, leading to aberrant left subclavian artery. Further abnormal remodelling is needed to explain the retroesophagal course of the aortic arch and the position of the descending aorta to the left.

In our case, the existence of at least vertebral, anal, and renal abnormalities confirmed VACTERL diagnosis. The etiology of VACTERL syndrome is heterogenous, and most often sporadic in humans, rare mutations, and mitochondrial dysfunction are described.[11] Ventricular septal defect is the most frequent cardiac anomaly in patients with VACTERL syndrome and also in patients with circumflex aortic arch.[12] In animal models, the association between trachea-esophagus and aortic arch abnormalities, including right aortic arch, has been published and the developmental sonic hedgehog pathway has been involved in 6th aortic arch formation.[13] This case is to our knowledge


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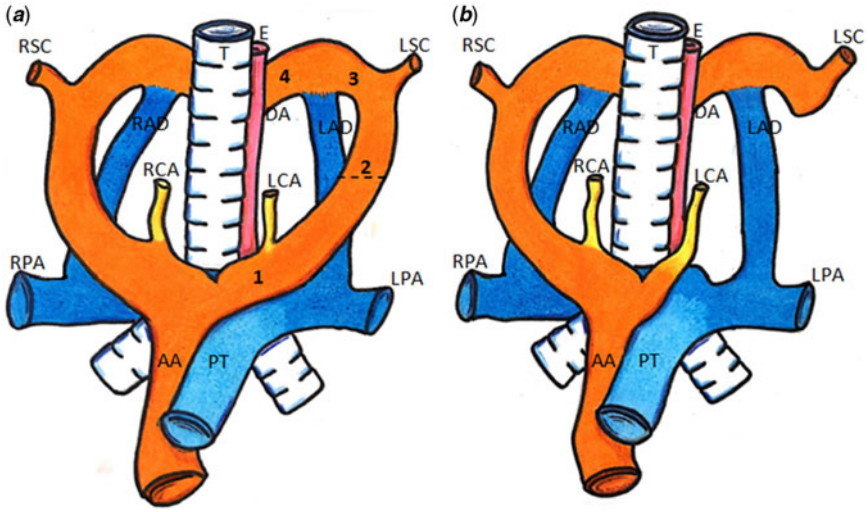


**Figure 2.** Diagram showing the arterial tree on the basis of the Edwards hypothetical double arch before (***a***) and after (***b***) partial disappearance of the left 4th aortic arch. The double arch is broken between the left carotid artery and the left subclavian artery (region 2, see dashed line), whereas regions 1, 3 and 4 persist. AA = ascending aorta; DA = descending aorta; E = esophagus; LAD = left arterial duct; LCA = left carotid artery; LPA = left pulmonary artery; LSC = left subclavian artery; PT = pulmonary trunk; RAD = right arterial duct; RCA = right carotid artery; RPA = right pulmonary artery; RSC = right subclavian artery; T = trachea.

the first human description of the association between VACTERL syndrome and circumflex right aortic arch.

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**Conflicts of Interest.** None.

**Ethical Standards.** The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the institutional committees.

**Author Contributions.** S.B. wrote the manuscript. R.C.G., M.L., and F.D. took part to data collection. C.O. realised the drawing. D.B. and L.H. reviewed intellectual content.

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