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Bovine Aortic Arch with an Aberrant Left Vertebral Artery in a 3-Year-Old Boy with VACTERL Association: A Case Report

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Male, 3-year-old
Final Diagnosis: VACTERL association
Symptoms: No clinical manifestations or complications
Clinical Procedure: —
Specialty: Cardiology

Objective: Congenital defects/diseases

Background: The VACTEREL association is an acronym that includes vertebral malformations (V), anal atresia (A), cardiac defects (C), tracheoesophageal fistula (TE), renal defects (R), and limb malformations (L). The aortic arch is the section between the ascending aorta and the descending aorta, where some variants have been described, such as the right aortic arch and bovine aortic arch, among others. A rare presentation in the Natsis classification is the "type X" where a bovine aortic arch and anomalous origin of the left vertebral artery are present. Several structural cardiac malformations have been described in the VACTEREL association. Still, there is no bovine arch or an anomalous left vertebral artery.

Case Report: Our patient was a 3-year-old boy with a diagnosis of VACTEREL association (type III esophageal atresia, congenital hip dislocation, scoliosis, bilateral clubfoot, and grade IV biliary ureteral reflux). Echocardiographic findings showed changes in the aortic arch, and angiotomography and magnetic resonance angiography showed a bovine aortic arch and an anomalous left vertebral artery. At the time of diagnosis, there were no clinical manifestations or complications due to the anomalous origin of the left vertebral artery.

Conclusions: This is the first description of a bovine type X arch according to the Natsis classification in a VACTEREL association. In general, knowledge of the anatomical variants of the aortic arch and the origin and course of the vertebral arteries is of great clinical and interventional importance, mainly because of the risk of cerebral ischemia.

Keywords: Case Reports • Aorta, Thoracic • Aortic Arch Syndromes

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Background

The aortic arch is the area between the ascending and descending aorta where variations can occur depending on the origin and number of blood vessels [1,2]. The aortic arch is the area between the ascending and descending aorta. The most common anatomical variant in humans is the left aortic arch with 3 vessels: (1) The brachiocephalic trunk from which the right subclavian and right common carotid arteries arise, (2) The left common carotid artery, and (3) The left subclavian artery. This variant is known as type I aortic arch and occurs in 74.0-89.4% in various populations [3,4] (**Figure 1A**).

Bovine aortic arch, or type II, is the second most common anatomical variant of aortic arch branching with origin in 2 branches: (1) The brachiocephalic trunk or innominate artery, which divides into 3 vessels (right subclavian artery, right common carotid artery, and left common carotid artery); and (2) The left subclavian artery. The prevalence of this variant varies from 11% to 27% (**Figure 1B**). There is another alternative variant of the bovine aortic arch, in which the origin of the innominate artery or brachiocephalic trunk is distal to the left common carotid artery, with a prevalence of 9% [3,4] (**Figure 1C**). Another rare variant described in the classification of Natsis et al is type X (bovine aortic arch and anomalous left vertebral artery) [4,5], in which 3 main arteries are observed: (1) The brachiocephalic trunk, from which the right subclavian, right common carotid, and left common carotid arteries arise, (2) The left vertebral artery, and (3) The left subclavian artery [3,5] (**Figure 2**).

The VACTEREL association is a group of congenital malformations of multicausal etiology. It was first described in 1972 as the VATER association as a combination of 3 or more of these malformations: vertebral, anal atresia, tracheoesophageal fistula, and radial or renal dysplasia. Later, additional features were added, such as cardiac and limb (L) malformations. Finally, the pathology received the name VACTERL association, which is the most widely used term [6,7]. Of the cardiac malformations described in the VACTERL association, there are no reports of bovine aortic arch or aberrant left vertebral artery. We present a case of a young boy with VACTERL association, in which the association of a bovine aortic arch and anomalous origin of the left vertebral artery was an incidental finding.

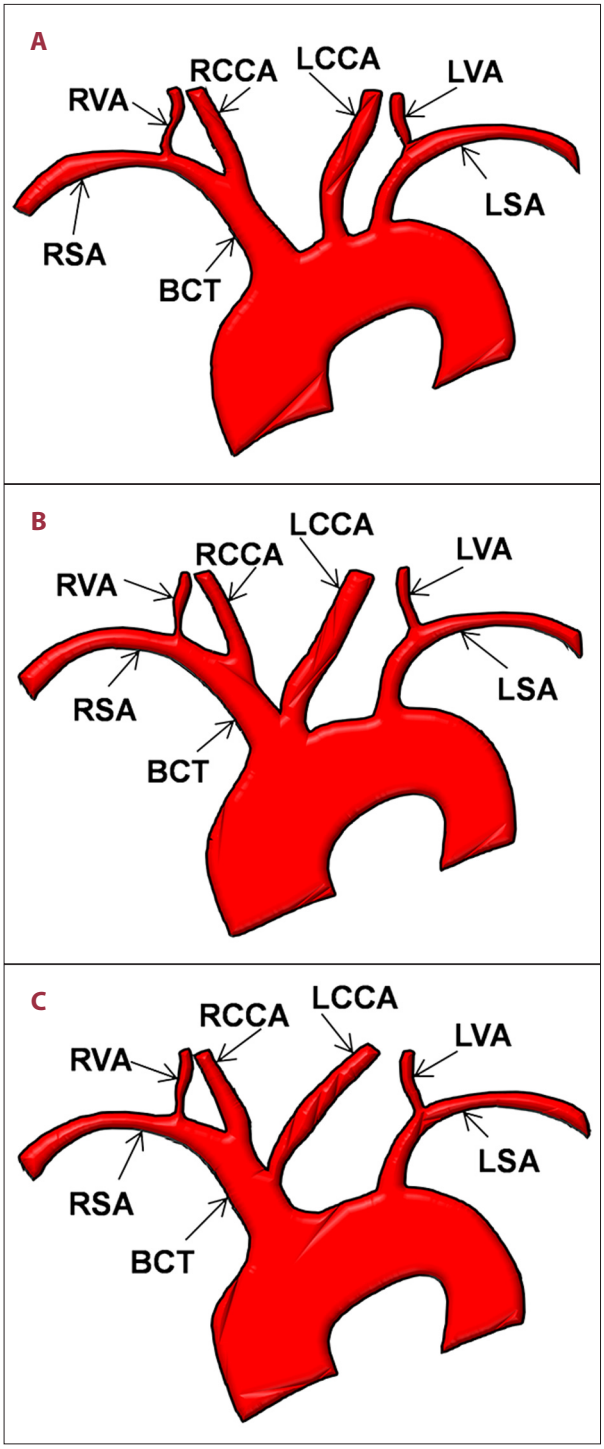

Figure 1. (**A**); Type I Aortic arch. (**B**); Type II bovine aortic arch and (**C**); Type II bovine aortic arch variant. BCT – brachiocephalic trunk; RSA – right subclavian artery; RVA – right vertebral artery; RCCA – right common carotid artery; LCCA – left common carotid artery; LVA – left vertebral artery; LSA – left subclavian artery (adapted from Goldsher et al [5]).

Case Report

Our patient was 3-year-old first-born. His birth was uncomplicated and his mother received adequate prenatal care. He was born at 38 weeks of gestation by cesarean section, without neonatal hypoxia, with a weight of 2.5 kg and a length of 49 cm. After birth, VACTERL was found to be associated with type III

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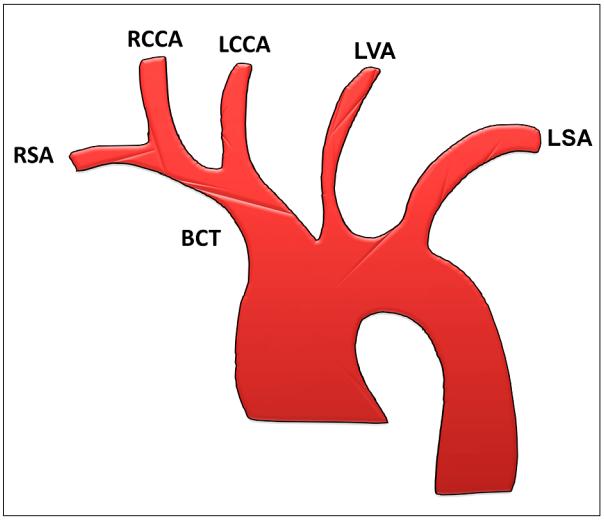

Figure 2. Bovine aortic arch type X (Natsis classification). BCT – brachiocephalic trunk; RSA – right subclavian artery; RCCA – right common carotid artery; LCCA – left common carotid artery; LVA – left vertebral artery; LSA – left subclavian artery (adapted from Natsis et al [4]).

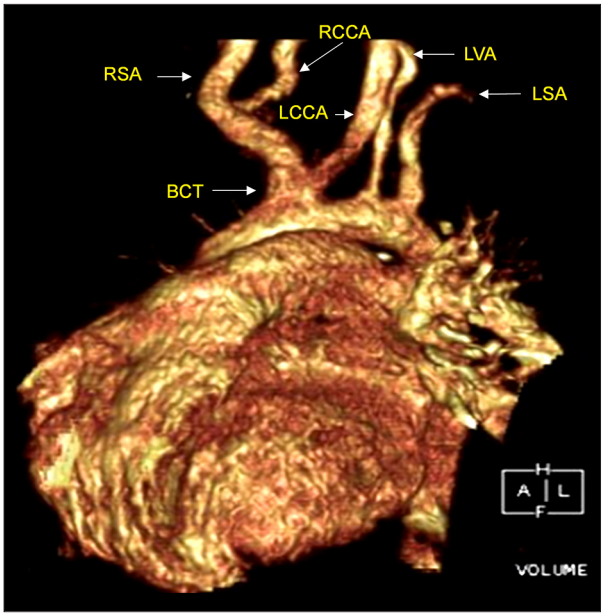

Figure 3. Reconstructed three-dimensional computed tomography showing the bovine aortic arch type X (Natsis classification). BCT – brachiocephalic trunk; RSA – right subclavian artery; RCCA – right common carotid artery; LCCA – left common carotid artery; LVA – left vertebral artery; LSA – left subclavian artery (adapted from Natsis et al [4]).

esophageal atresia, congenital hip dislocation, scoliosis, bilateral clubfoot, and grade IV vesicoureteral reflux. The esophageal atresia was surgically corrected with terminal esophagoplasty at 72 hours of life and vesicostomy with closure at 1 year of age.

The patient had a history of recurrent respiratory infections, so esophageal-gastroduodenal series and bronchoscopy were performed according to the study protocol, which reported mild tracheobronchitis, and gastroesophageal reflux (CARR score 7). Incidental findings included right bronchial malacia and pulsatile stenosis of the middle third of the trachea and

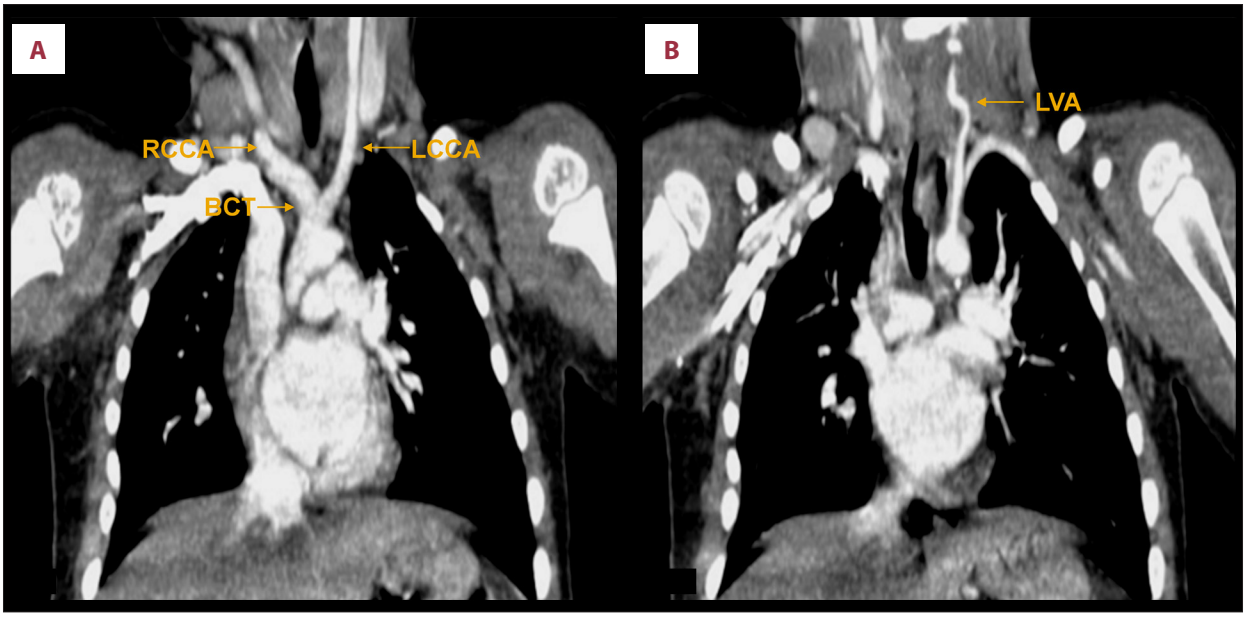

Figure 4. Reconstructed three-dimensional computed tomography angiography showing the bovine aortic arch type X (Natsis classification). (**A**) The origin of the right common carotid artery (RCCA) and the left common carotid artery (LCCA) of the brachiocephalic trunk (BCT) is observed. (**B**) Left vertebral artery (LVA) originating in the aortic arch.

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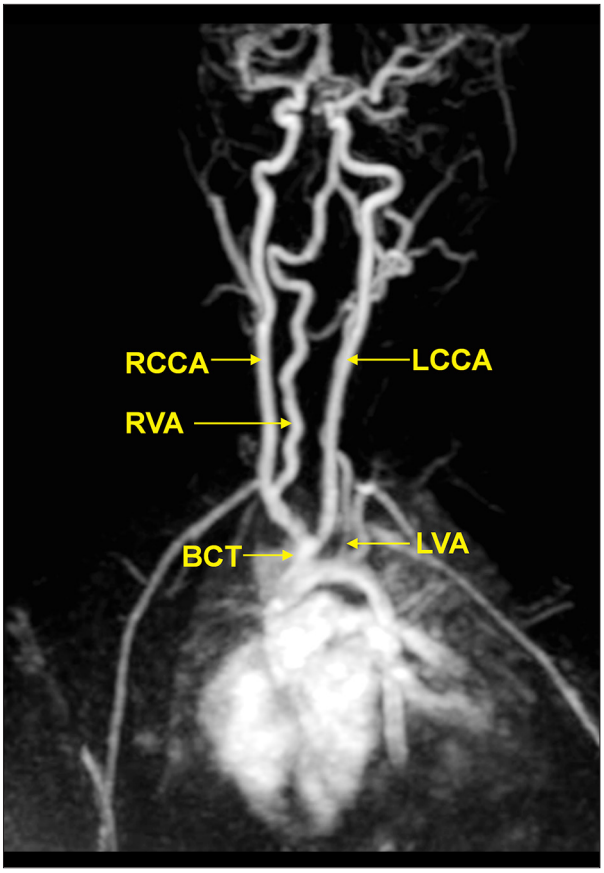

Figure 5. Magnetic resonance angiography demonstrates bovine type X aortic arch (Natsis classification). Origin of the right common carotid artery (RCCA) and left common carotid artery (LCCA) from the brachiocephalic trunk (BCT) and left vertebral artery (LVA) with origin in the aortic arch, not from the left subclavian artery.

left main bronchus. Because of the suspicion of aortic arch changes, an echocardiogram was performed, which showed a structurally normal heart without valvular defects and a bovine aortic arch with an aberrant artery. Computed tomography with three-dimensional reconstruction of the aortic arch (**Figure 3**), computed tomography, and magnetic resonance angiography of the aortic arch (**Figures 4, 5**) showed the presence of a type X bovine aortic arch and an anomalous origin of the left vertebral artery from the aortic arch and not from the left subclavian artery.

Discussion

In general, anatomical variations in the position of the aortic arch and the origin of the arteries are usually asymptomatic, and the vast majority are diagnosed incidentally. Dysphagia and dyspnea are the main symptoms of esophageal and tracheal compression, respectively, in patients with abnormal right aortic arch and right subclavian artery [8].

Under normal conditions, the left vertebral artery originates from the left subclavian artery, crosses the transverse foramen of C6, passes through the transverse processes of the upper cervical vertebrae to enter the foramen magnum, and at the end and, during its course, perfuses the posterior cervical musculature, brain, and spinal cord [9].

Anomalous origin of the left vertebral artery directly from the aortic arch is the most common variant, with a prevalence of 2.5% to 5.8%, in which case there are 4 vessels in the aortic arch: (1) the brachiocephalic trunk, (2) the left common carotid artery, (3) the left vertebral artery, and (4) the left subclavian artery [10,11]. Müller and coworkers [12] analyzed 2033 contrast-enhanced thoracic tomograms to evaluate the aortic arch's structural variations and the origin of the supra-aortic arteries. Anatomical variations of the aortic arch were present in 270 (13.3%), and bovine or type II aortic arch was observed in 163 cases (8%). In 86 patients (4.2%), the origin of the left vertebral artery came directly from the aortic arch proximal to the left subclavian artery; in 1 case, the origin of the left vertebral artery was observed distal to the left subclavian artery. In 20 cases (1.0%), patients had an aberrant right subclavian artery. In another study, Berko et al [13] retrospectively reviewed 1005 adult computed tomography angiograms to assess the prevalence of aortic arch coarctation. In 659 cases (65.9%), the aortic arch and the origin of the arteries were normal (ie, type I aortic arch). In 274 cases (27.4%), there was a bovine or type II aortic arch, 66 patients (6.6%) had an aberrant origin of the left vertebral artery, 61 (6.1%) were proximal to the left subclavian artery, and of these cases 16 (1.6%) also had a bovine aortic arch. These last described cases were similar to our patient, who presented a bovine type X aortic arch according to the classification of Natsis et al, in which the origin of the left vertebral artery is direct from the aortic arch and proximal to the left subclavian artery [4].

Defects in the position of the aortic arch or the origin of the vertebral arteries and their morphology or trajectory can affect the results of angiographic and surgical interventions if not considered. Some authors suggest that the anomalous origin of the vertebral arteries can cause intracerebral disorders by altering vascular hemodynamics, as they are exposed to turbulent blood flow from the aortic arch, which increases the risk of ischemia, thrombosis, aneurysm, dissection, occlusion, and atherosclerosis [9-12]. A recent study by Clerici et al evaluated the prevalence of anatomical variants of the aortic arch by Doppler ultrasound in 742 fetuses, as well as the hemodynamic differences between fetuses with normal aortic arch and fetuses with bovine aortic arch variants. They identified 697 (93.94%) fetuses with normal or classical aortic arch.

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In contrast, 45 (6.06%) had a bovine or type II aortic arch. We identified 697 (93.94%) fetuses with a classic or normal aortic arch. In comparison, 45 (6.06%) had a bovine or type II aortic arch, one associated with a ventricular septal defect and another with an aberrant right vertebral artery. Hemodynamic evaluation was possible in only 39 fetuses, and significant differences were observed between the study groups [13].

Congenital cardiac malformations have been described in 40-80% of patients with VACTERL association. They can range from structural defects (patent ductus arteriosus, atrial septal defect, ventricular septal defect, tetralogy of Fallot, double outlet right ventricle, and anomalous venous drainage of the pulmonary arteries) to defects in the emergence of the aortic arch arteries. García-Guzmán et al reported a series of 14 cases with VACTERL association; 85.7% (n=12) had cardiac malformations, mainly atrial septal defect in 9 patients, 3 had aortic arch malformations (right aortic arch, aortic protrusion, and 1had an anomalous origin of the left coronary artery) [14]. Moras et al evaluated the prevalence and distribution of cardiovascular malformations in 396 patients with anorectal malformations, including 79 patients with VACTERL association [15]. The prevalence of congenital heart disease in this group was 73.4% (58 cases), with only 1 patient with an anomalous left subclavian artery and 2 cases with anomalous origin of the coronary artery. The remaining congenital heart defects were septal defects (atrial and ventricular), tetralogy of Fallot, coarctation of the aorta, vascular ring in the right aortic arch, pulmonary stenosis, and single ventricle. No cases of association of VACTERL with bovine aortic arch and anomalous left vertebral artery have been reported.

Conclusions

The importance of our case lies in the description of a patient with VACTERL associated with a rare bovine arch malformation and anomalous origin of the left vertebral artery, which according to the Natsis classification is type X. Knowledge of the anatomical variants of the aortic arch and the origin and course of the vertebral arteries is of great clinical importance, especially for planning aortic arch surgery or endovascular interventions.

Acknowledgments

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Ethical Statement

The study was approved by the Ethics Committee of the Women's Hospital, Secretariat of Health, Culiacán, Sinaloa, México (No. 202310-20).

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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