Congenital abnormalities of aortic artery. 
Assessment in neonates and early childhood with multislice tomography

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Abstract

Congenital abnormalities of aortic artery. Assessment in neonates and early childhood with multislice tomography

In the evaluation of aortic artery congenital abnormalities, the echocardiography and the plain X ray are the traditionally used imaging methods. Multislice angiography appears as an important method in diagnosis of these different diseases allowing evaluate these entities in a non invasive, fast and accurate form, giving to cardiovascular surgeons very important information to delineate the surgical strategy.

In this article, we review the applications of multislice angiography in the evaluation of most frequent congenital anomalies of aorta artery, performed in neonates and early childhood.

Key words: Aorta artery. Congenital anomalies. Neonates. Multislice angiography.

INTRODUCTION

The multislice angiotomography (MDCTA) presents several properties and is utilized with more frequency every time by childhood cardiovascular surgeons as a chosen method of imaging in the anatomic and pathological scan of different abnormalities of the aortic arch. The multiplanar capacity and three-dimensional reconstructions in particular are easily to interpret, as well as being extremely fast studies, with short anesthetic time and low contrast volume to administer.

The objective of the present study has been to review the findings enabled by multislice computer tomography (MDCT) of the aortic arch’s development most frequently found in our institution.

MATERIALS AND METHODS

Angiotomography studies (CTA) performed between October 2006 and November 2008 to patients who presented congenital abnormalities of aortic artery, were evaluated in a retrospective way. Total patients included in this study were neonates and little children in an age range between 6 days and 11 months. Echocardiography and/or thorax X ray, together with the symptoms, enabled a presumptive diagnosis (Table 1).

The studies were performed with a 64 detector CT (Aquilion, Toshiba Medical Systems, Tokio, Japan) using a software of radiation modulation doses (Sure Exposure) with effective doses between 0.6 to 1.4 mSv. Images taken every 0.5 mm thickness were obtained with a 0.3 mm reconstruction interval, a factor pitch of 0.828 and 0.5 sec rotation tube.

Between 1 and 1.5 ml/kg of non ionic intravenous contrast (Iopamiron 370°, Schering) were injected under anesthesia and cardiac monitoring, with a 1.5 to 2.5 ml/sec flow injection pump (Medrad, Stellant).

Three-dimension reconstructions were performed in a workstation (Vitrea, Vital Images). All patients were symptomatic and had surgery, which enabled to correlate the information observed by different imaging methods with intra-surgery findings, assumed as certain definitive diagnosis.

In 11 cases, the presumptive diagnosis was confirmed by angiography. In one patient, the angio-computed tomography showed a non suspected pathology that was further confirmed by surgery.

Embryology review

Blood vessels start to develop from the mesoderm that covers the yolk sac at 18 days of the embryo’s age. On day 24th, the cardiovascular system is composed
of two cardiac tubes, arterial and venous vessels. Subsequently, both cardiac tubes fuse to form a tubular heart that develops four dilatations named cavernous sinus, atrium, ventriculum and cardiac bulb.

The heart begins a process of folding and septation. Subsequently, the cardiac bulb divides in three parts: proximal, one intermediate called truncus arteriosus and distal, called aortic sac. The two big arteries that are born in the heart are not originated from preexisting vessels, but from the truncus arteriosus, which first forms a partition and then divides to form two independent vessels: the ascending aortic artery and the pulmonary artery trunk.

From the aortic sac there 6 pairs of aortic arches connecting the ventral and dorsal aortic sketches are born. Some of them usually regress with development, persisting at the end of the fifth week only third, fourth and sixth arcs, which will later originate the great vessels 1).

RESULTS

In the present study, twelve symptomatic patients were included, all of them with a previous echocardiography. One patient with coarctation diagnosis by echocardiography, was interpreted by CTA as an interruption of aortic arch type A, which was confirmed by surgery. In the other 11 patients, there was a coincidence between the echo-cardiography and the CTA. None of our patients underwent MRI.

Clinical cases

Coarctation

This is the most common entity in our series, with 5 patients studied with preoperative CTA (41.6%).

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender</th>
<th>Age</th>
<th>Diagnosis</th>
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<tbody>
<tr>
<td>DV</td>
<td>M</td>
<td>5 months</td>
<td>Incomplete aortic ring</td>
</tr>
<tr>
<td>LI</td>
<td>M</td>
<td>3 weeks</td>
<td>Truncus arteriosus type II</td>
</tr>
<tr>
<td>MM</td>
<td>F</td>
<td>2 weeks</td>
<td>Coarctation</td>
</tr>
<tr>
<td>PM</td>
<td>F</td>
<td>11 months</td>
<td>Aberrant subclavian right artery</td>
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<tr>
<td>PS</td>
<td>F</td>
<td>6 days</td>
<td>Interruption of aortic arch type B</td>
</tr>
<tr>
<td>SA</td>
<td>F</td>
<td>2 months</td>
<td>Coarctation</td>
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<tr>
<td>RM</td>
<td>F</td>
<td>11 months</td>
<td>Coarctation</td>
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<tr>
<td>VG</td>
<td>M</td>
<td>7 days</td>
<td>Coarctation</td>
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<tr>
<td>LC</td>
<td>F</td>
<td>5 months</td>
<td>Interruption of aortic arch type A</td>
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<tr>
<td>PM</td>
<td>F</td>
<td>6 months</td>
<td>Interruption of aortic arch type A</td>
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<td>SN</td>
<td>M</td>
<td>1 month</td>
<td>Coarctation</td>
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<tr>
<td>PA</td>
<td>M</td>
<td>1 month</td>
<td>Truncus arteriosus type III</td>
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<tr>
<td>SN</td>
<td>M</td>
<td>1 mes</td>
<td>Coartación</td>
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<tr>
<td>PA</td>
<td>M</td>
<td>1 mes</td>
<td>Tronco arterioso tipo III</td>
</tr>
</tbody>
</table>

Table 1

Fig. 1. Male patient, with 6 days of life. 3D reconstruction. Sagital plane. Coarctation of typical localization in descending aorta, distal to the appearance of left subclavian artery (arrow). Relative hipoplasia of transverse arch portion (arrowhead).

Fig. 2. Female patient, with 14 days of life. Signs of heart failure, pulse discrepancies and dorsal systolic murmur. Sagital plane. Maximum intensity reconstruction. Severe coarctation (arrow) with continuity to descending aorta through a filiform flow and intercostals collateral circulation (arrowhead). A.M.I: Internal mammary artery. In surgery, the diagnosis is proved. Resection was performed for coarctation with end to end anastomosis and ligation of the ductus, which was permeable (not visible in ACT).
Fig. 3. a) Female patient, 2 months old. Coronal plane: Situs inversus totalis plus coarctation (arrow). A.S.D.: Right subclavian artery. A.I: Left atrium. A right toracotomía is performed, coarctation section and end to end anastomosis. b) 3D reconstruction. Posterior PLANE. Coarctation (arrow). A.S.D: Right subclavian artery. T: trachea.

Fig. 4. a) Female patient. Interruption of aortic arch type A. Sagital plane. Maximum intensity reconstruction. Thoracic artery is interrupted in the posterior sector of the arch (arrow), in distal position to the left subclavian artery (ASI). The descending aorta receives flow from the pulmonary aorta (AP) through a permeable ductus (arrowhead). b) 3D reconstruction. Anterior oblique left plane. Ao. A.: Ascending aorta. Ao. D.: Descending aorta. AP: Pulmonary artery. ASI: Left subclavian artery. Interruption of aortic arch (arrow). c) 3D reconstruction. Posterior oblique plane. APD: Right pulmonary artery. API: Left pulmonary artery. ACPI: Left primitive carotid artery. ASI: Left subclavian artery. Ao. D.: Descending aorta. In the surgery, the ductus is joint. En la cirugía se liga el ductus, se reseca la zona interrumpida con posterior anastomosis que involucra el arco aórtico, la boca de la arteria subclavia y la aorta descendente.

Scheme 1: Illustrative scheme of the interruption of aortic arch type A.
VD: Right ventricle.
VI: Left ventricle.
AP: Pulmonary artery.
A Ao Asc: Ascending aortic artery.
A Ao Desc: Descending aortic artery.
Tr Bc: Brachiocephalic arterial trunk.
ACPI: Left primitive carotid artery.
ASI: Left subclavian artery.

It is a congenital abnormality characterized by narrowing of the aortic lumen. Stenosis is often found in the proximal descending aorta after the emergence of left subclavian artery. Considering its connection to the ductus arteriosus, they are classified in preductal, postductal or yuxtaductal (2, 3).

Other cardiovascular abnormalities are usually presented, among them, bicuspid aortic valve, hypoplastic transverse arch portion, dilatation of the ascending aorta, persistent ductus and aberrant right subclavian artery (4) (Fig. 1).

There is hypertension of superior extremity, which
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Congenital abnormalities of aortic artery tends to be offset by collateral development of intercostals, epigastrics and mediastinals (Fig. 2).

Infants with coarctation, particularly when a persisting ductus arteriosus exists, presented with signs of congestive heart failure (tachycardia and dyspnea). A dorsal systolic murmur is often listened, pulses are verified and differential pressures in the lower limbs.

Diagnosis is based on the clinical findings and on imaging methods (echocardiography, angiography, angiotomography and magnetic resonance).

The CT scan gives us information about the site and extension of stenosis, aspect of collaterals and the presence of associated abnormalities, currently an extremely useful modality in the following of postpercutaneous or surgery treatment (Fig 3).

**Interruption of aortic arch**

It is defined as the loss of anatomic continuity between the ascending and the descending aorta, and it is considered as an extreme form of coarctation. The flow towards the descending aorta is given by the ductus, which remains permeable and provides continuity between the trunk of the pulmonary artery and the distal aorta to the interruption. Despite this continuity, the blood that arrives to the abdomen and lower limbs is only partially oxygenated. In most of the cases, other cardiovascular abnormalities are associated, such as interventricular communication (IVC), obstruction in the exit tract of left ventricle, bicuspid aortic valve and persisting arterial ductus.

Patients evolve fast after birth with severe congestive heart failure, shock tendency and, if not treated, they often pass away in the neonatal period at few days of birth.
Its incidence is low, 3 cases at a million of born alive approximately, and represents the 1% of all cardiovascular abnormalities. In our study, there were 3 of the 12 studied patients.

They have been classified in three types according to the place where the interruption is:

Type A: Distal to the left subclavian artery.
Type B: Between the common left carotid artery and the left subclavian artery.
Type C: Between the brachiocephalic arterial trunk and the left subclavian artery.

Type B is the most frequent one (53%), followed by type A (43%) and type C (4%) (6).

The ACT clearly shows the interruption, its connection to the supra-aortic trunks, as well as the presence of the ductus, that provides continuity between the pulmonary artery and the descending aorta (Schemes 1 and 2) (Fig. 4, 5, 6).

Truncus arteriosus

The truncus arteriosus (TA) is characterized by the presence of a single large artery which originates in the base of the heart with a single anulus, that originates coronary, pulmonary and systemic arteries.

In most of the cases there is an interventricular communication and the trunk usually originates over this septal defect. It is the result of a failure in the early embryonic development due to a lack of division of the primary arterial precursor (truncus arteriosus) in aortic and pulmonary arteries, eventuality that must happen in the normal fetus at approximately 8 weeks of gestation. It is a pathology with low incidence, and represents approximately the 0.7% of cardiovascular congenital defects. It can be associated to other alterations, among them, the aortic arch to the right, coarctation and interruption of the aortic arch (association which is present in the 14% of TA), and also to extracardiac abnormalities, particularly Di George syndro-

me (facial anomalies, cleft palate hypoplastic thymus and hipocalcemia) (7).

There are two anatomic classification systems that basically consider the origin of pulmonary arteries: Collet-Edwards and Van Praagh.

Collet-Edwards classification

Type I. There is a small trunk that is separated. Right and left branches of the pulmonary artery are originated from it.

Type II. Both arteries are born from the posterior side of TA, separated by a short distance.

Type III. Each artery is born independently of the TA's lateral side, one away from the other.

Type IV. One branch is born from the trunk and the other, from the descending aorta or the ductus.

Van Praagh questions Collet’s classification, specially type IV, and introduces some modifications considering the presence or absence of interventricular communication and interruption of aortic arch.

He uses the prefix A to indicate the presence of IVC, or B to indicate the intact septum. On the other hand, he includes in type IV the TA associated to the interruption of the arch (10). There is a pulmonary flow increased with a mixture of systemic and pulmonary blood.

Most of the children arrive with cyanosis in early stages due to the mixture of oxygenated and deoxidized blood and with signs of congestive heart failure. The prognosis, if a surgery correction is not performed, is bad, with a survival of 25% at 6 months (11) (Schemes 3 and 4) (Fig. 7, 8).

Vascular rings

They belong to abnormal configurations of aortic arch, which are the result of an incomplete regression of one of the six present bronchial arches during the embryonic development. Vascular structures or arterial ligament surround and compress the trachea and esophagus, generating symptoms such as stridor, breathing difficulty, air trapping and apnea; the esophagus compression causes vomits, swallowing difficulty and dysphagia. They predispose to the appearance of aspiration pneumonias (12).

They can be complete, forming a continuous ring around the trachea and the esophagus, or incomplete, in such cases the symptoms are usually minor or even absent (13).

There are several forms of vascular rings:

Syndrome of the anomalous innominate artery.

The brachiocephalic arterial trunk originates from the aortic arch but more to the left than usual, being the reason for which it crosses diagonally ahead the trachea and can have certain degree of compression on its anterior side.

Double aortic arch. It can exist as complete aortic arch (functional), in which there is persistence and permeability of right and left arch, or as double arch with partial atresia of one of them (usually, the left one), where the continuity is given by a remaining
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Supra-aortic trunks appear separately, without a brachiocephalic arterial trunk, this is, subclavia artery and right carotid, subclavia artery and left carotid, which is named sign of the four vessels. When the minor arch is atresic, the fibrous cord generally appears distal to the appearance of left subclavian artery. In these cases, it can sometimes be misinterpreted as an aortic arch to the right; however, the sign of the four vessels is usually seen. On the other hand, on the atresic side, a small permeable segment previous to the fibrous cord usually remains, manifested as a diverticulum posterior to the esophagus (Scheme 5) (Fig. 9).

Aortic arch to the right with aberrant left subclavian artery. The aberrant left subclavian artery is the last trunk to branch off. The presence of the arterial ligament completes a true ring. They usually are symptomatic at early age.

DISCUSSION

Traditionally, congenital abnormalities of aortic arch were evaluated with echo-cardiography or conventional radiology and, in some instances, with conventional angiography. In recent years, magnetic resonance (MR) and ACT, specially with multislice technology, gained relevance in the diagnosis and pre-surgery scan of different abnormalities [14-16].

Each one of these modalities presents advantages and disadvantages that must be considered in choosing one.

Neonate patients and little infants present a heart rate that oscillates between 100 and 160 heart beats a minute. Therefore, it is essential to count with equip-

Figure 4: Illustrative scheme of truncus arteriosus type II.

VD: Right ventricle.
VI: Left ventricle.
CIV: Ventricular septal defect.
TA: Truncus arteriosus.
APD: Right pulmonary artery.
API: Left pulmonary artery.


anesthesia. Studies take between 20 – 40 minutes, longer than ACT.

The decision of which modality is to be used, depends on the availability and experience of each particular imaging service.

In our opinion, CT presents the advantage of being extremely fast and easy to program, requiring less complex anesthetic procedures.

CONCLUSION

Computed tomography angiography appears as an extremely useful complementary method in the diagnosis of congenital abnormalities of the aortic arch in neonates and early childhood patients. It is a fast and non invasive procedure, which takes short anesthetic time and low charge of intravenous contrast. In our series, it enabled to diagnose all the studied anomalies in an efficient and precise way, with excellent correlation to the surgery findings reported by surgeons.
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Bibliography