Everything you need to know about MDCT Angiography (MDCTA) in the diagnosis of aortic pathology in pediatric patients

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Learning objectives

To describe the semiology of different aortic pathologies in children and their findings at Multidetector Computarized Tomography-Angiography (MDCTA).

To know the indications of this technique and discuss its advantages and disadvantages in the evaluation of the aorta abnormalities.
Background

The aortic pathology in the children is usually of congenital origin and it can present itself as an anomaly isolated or associated with complex congenital heart diseases. The anomalies of the aortic arch with all its variants, aberrant vessels, coarctation of the aorta, pseudocoarctation and interruption of the aortic arch are the most frequent isolated anomalies. In other cases the aortic pathology is associated with complex congenital heart diseases: Tetralogy of Fallot, great vessel transposition, double outlet right ventricle, Williams syndrome... The acquired aortic pathology is rare in the childhood, being able to affect to the aorta the traumatisms, the infections or the inflammatory of connective tissue diseases.

Echocardiography is useful for the evaluation of the aortic valve, but it can be limited to value the ascending aorta, the aortic arch, the anatomy of the aberrant vessels, the supraaortic vessels, the pseudocoarctation of the aorta and the descending aorta.

Magnetic Resonance Angiography (MR-A) is a useful tool in the diagnosis image of aortic pathology, but frequently it is not accessible, it supposes major exploration time, needs anesthesia or it is contra-indicated.

Conventional Arteriography presents limitations for its nature in 2D and difficulties in the simultaneous evaluation of the pulmonary and aortic vascular system. Also involves the administration of abundant volume of intravascular contrast medium, needs general anesthesia in most of cases and implies an important radiation dose.

MDCTA is a relatively new and considered currently the first image technique in the diagnosis and characterization of the aortic pathologies in adults and also in children. MDCTA studies the aorta in a non-invasive fast way and accurately. In the well-known or suspected complex congenital heart diseases TCMDA can be used for imaging of the aortic arch and great vessels and for the postoperative evaluation, allowing the detection of complications post-treatment. The combination of axial images, 2D, MIP and 3D reformatted images allows to define the anatomical relationships.

- MDCTA ADVANTAGES:
  1. Global assessment, including lungs, airways and other regional structures
  2. Accessible technique
  3. Short time examination and usually well tolerated
  4. It is not operator-dependent (contrary to cardiac ultrasound)
5. It does not have the contra-indications of MR
6. Its multiplanar and three-dimensional capacity
7. Lower cost than MR or arteriography
8. It sometimes requires sedation but rarely anesthesia

- **MDCTA DISADVANTAGES:**

1. Radiation; although there are systems for dose reduction and protectors against lead and bismuth.
2. Contrast media. The risk of major adverse reactions (spasm of the airways and cardiovascular collapse) to iodinated contrast media is extremely low in children.

- **TECHNIQUE AND PARAMETERS of pediatric MDCTA:**

It is important to pay attention to the technique of the TCMDA since if not, the ability to diagnose is limited.

Pediatric MDCTA is less standardized than in adults.

The examination must be directed to obtain the most diagnostic efficiency with the less radiation dose.

The movement of the patient must be controlled, for what sometimes the *sedation* is necessary in children less than 5-year-old.

The technique will depend on different factors:

- Adjustment of the number of detectors
- Thickness of the detectors
- Rotation time
- Tube current (mA)
- Peak voltage (Kv)

In general, for a MDCT of 64 detectors the parameters to be used will be:

- *Scan field of view:* small
- *Detector thickness:* 64 x 0,625
- *Slice thickness and interval of reconstruction:* 2,5 mm and interval of reconstruction of 2,5 mm
- *Tube current:* 60mA (as low as possible)
- *Peak voltage:* 80kVp for a patient of less than 45 kg, and more for heavier patients
- *Gantry (rotation time):* 0,5 seg
Bismuth protectors reduces the radiation dose on the breast with the same quality of the image.

Modulation of dose and iterative reconstructions reduce the dose, so they should be used if available.

The images are acquired during the phase of arterial opacification, for the standard study of the aorta.

In a TCMDA of the thoracic aorta, the acquisition is from the low neck to the thoracoabdominal union; the study of thoracoabdominal aorta will finish at the pelvis.

Pre-contrast studies are only performed in cases of stent patients carriers.

In children and young people, the studies of the aorta are performed without cardiac synchronism, so that they have a sensibility of 82 % for the visualization of the origin of the coronary arteries.

TCMDA is acquired in inspiration in those patients who collaborate. In sedated patients, the study is performed during a calm breathing.

The acquired axial images are sent to a workstation for further analysis and review. A combination of "Multiplanar reformats" (MPR), "Maximum Intensity Projection" (MIP) and "Volume Rendering" (VR) is used to evaluate the aorta and its branches. The multiplanar reformats 2D can be generated in any plane, with a resolution comparable to that of the axial images. Coronal and sagittal images provide information on cardiovascular structures that may not be apparent on axial images.

- CONTRAST MEDIA in pediatric CTMDA

Hypo-osmolar nonionic contrasts (fewer side effects, less nausea, greater comfort) are used.

- Iodine concentration: 300 mg / ml
- Dose: 1.5 to 2 ml / kg, up to 3ml / kg and up to an overall maximum of 125 ml
- Flow rate: 1.5 ml / sec (angiocatheter 24); 2 to 2.5 ml / sec (angiocatheter 22); 3-4 ml / sec (angiocatheter 20).
• **Automatic injector**, with double track and possibility of mixing contrast and saline.

In low weight newborns and infants, automatic mixing of 50% of contrast and saline (dilution 1:1) can be used to increase the injected volume. For example, in patients 3 kg weight can be used 6cc of contrast and 6 cc of saline, which would reduce the possibility of artifacts by the contrast medium.

It is recommended to use a "Test Contrast" or "Bolus tracking" in which images are acquired during the arterial phase, with automatic detection of the maximum peak enhancement in the ascending thoracic aorta (threshold of 150 HU).

- **INDICATIONS of pediatric CTMDA:**

The coming of TCMDA has revolutionized the vascular image. Thus, the role of TCMDA in the diagnosis of vascular disease has increased thanks to the very fast scanning, greater anatomical coverage, reconstructions of high quality and thin collimations. Many times the TCMDA serves as a noninvasive study prior to final diagnosis.

In general, when there is suspicion of aortic pathology or complex cardiac disease associated with aortic pathology, echocardiography is the first study to be performed; however, it may be limited in the assessment of the ascending aorta, arch, supra-aortic trunks, descending aorta or collateral circulation, in which case the study should be completed with MR-angiography or TCMDA. Other times, the TCMDA or MRI will be subsequently referred to echocardiography when there is a need to assess structures or extracardiac anomalies in the pre- or post-operative.

The TCMDA should be performed when:

- Contra-indication or unavailability of MR
- Suspect of vascular rings
- Assessment of stents
- Assessment of airways and lungs
- Assessment of extra-cardiac structures
- Suspect of collateral circulation, specially in patients with right ventricular outflow tract obstruction
- Suspect of traumatic or iatrogenic aortic lesion
- Suspect of aneurysm or pseudoaneurysm
Findings and procedure details

PEDiatric AortA Abnormalities

The diagnosis of abnormalities of the aorta will depend largely on the understanding of the normal anatomy, anatomical relationships and variations.

Thoracic aorta can be divided into five segments (Fig. 1 on page 15):

2. Ascending aorta (from the aortic root to the brachiocephalic trunk)
3. Proximal aortic arch (up to the left subclavian artery)
4. Distal aortic arch or isthmus (from the left subclavian artery to the ligamentum arteriosum)
5. Descending aorta (from the ligamentum arteriosum to the diaphragm)

We will divide the pathology of pediatric aorta in:

- Congenital cause
- Acquired cause
- Associated with complex congenital heart diseases

A. CONGENITAL AORTIC PATHOLOGY

A. 1. ABNORMALITIES OF THE AORTIC ARCH

The abnormalities of the aortic arch and cervical vessels are relatively common.

The first three have little or no clinical relevance.

a) Left sided aortic arch with aberrant subclavian artery (Fig. 2 on page 15)

It is the most common abnormality of the aortic arch and it has normally no clinical impact.

The aberrant artery arises distal to the left subclavian artery and passes posterior to the esophagus to form an incomplete vascular ring.

It may arise from an outpouching, known as diverticulum of Kommerell.
b) Bovine arch (Fig. 3 on page 16)

Occurs when the brachiocephalic artery shares a common origin with the left common carotid artery.

c) Ectopic origin of the left vertebral artery from the aortic arch (Fig. 4 on page 16)

d) Double aortic arch (Fig. 5 on page 17)

It is an actual vascular ring, in which the ascendent aorta divides into two aortic arches that converge in a sole descendent aorta.

e) Right sided aortic arch with aberrant left subclavian artery (Fig. 6 on page 17)

It can be found in 1/1000 people. It usually produces no symptoms and is associated with congenital heart defects in only 12% of the cases.

f) Right sided aortic arch with mirror image branching

It is associated with congenital heart diseases in 90% of the cases (tetralogy of Fallot and patent truncus arterious, mainly)

There is a right sided aortical arch with mirror image branching: 1st left brachiocephalic trunk, 2nd right carotid artery and 3rd right subclavian artery (Fig. 7 on page 18).

A. 2. PATENT DUCTUS ARTERIOSUS

It is a vascular structure that normally connects the proximal descending aorta with the left pulmonary artery.

In term newborns functionally the ductus closes at 18-24 hours of life and anatomically closes at one month of age. If it remains permeable three months after birth, is considered a patent ductus arteriosus. Symptoms depend on the magnitude of the shunt, which depend on the size, diameter and configuration of the ductus. (Fig. 8 on page 19, Fig. 9 on page 20 and Fig. 10 on page 20)

Echocardiography is the standard technique for diagnosis. MRI or TCMDA are used to define the anatomy in complicated cases and guide toward treatment (ductus measures and degree of pulmonary hypertension).
A. 3. AORTIC HYPOPLASIA

The gradual narrowing of the distal aortic arch and the isthmus is considered pathological after the first 3 months of life.

The proximal segment of the aortic arch is defined as hypoplastic when its external diameter is <60% of the ascendent aorta's diameter, <50% of the distal aortic arch's diameter and <40% of the isthmus' diameter. (Fig. 11 on page 20 and Fig. 12 on page 21)

It is associated with other forms of obstruction or restriction to aortic flow, mainly aortic coarctation (Fig. 13 on page 21).

MDCTA provides information about the location and size of the hypoplastic segment, and also of the path of collateral vessels and other associated findings. (Fig. 14 on page 22)

A. 4. COARCTATION OF THE AORTA

It is defined as a narrowing of the aorta in the area of the ligamentum arteriosum.

There are two types:

1. Preductal (or infantile): proximal to the origin of the left subclavian artery. It is associated with a decrease in caliber of a long segment of the aortic arch and other congenital heart disease. Collateral circulation is normally absent. It is diagnosed in the first six months of life with symptoms of heart failure.

2. Postductal (or adult): distal to the origin of the left subclavian artery. It is associated with the dilation of the proximal aorta and with the development of the collateral circulation.

It can be associated with patent ductus arteriosus (80%), bicuspid aortic valve (30-60%), ventricular septal defect (10%), aortic arch hypoplasia (Fig. 13 on page 21) or brain aneurysms (10%).

The diagnosis is made during the first 7-14 days of life (when the ductus closes) in those cases of critical coarctation with symptoms of heart failure and poor systemic perfusion. In those cases of non-significant stenosis or good collateral circulation, the diagnosis is made in patients with ages 1-36. (Fig. 15 on page 23)
Echocardiography is used for the diagnosis of coarctation in childhood. MRI and MDCTA provide anatomical details of coarctation in older children and adults (who often have limited acoustic window in ultrasound), allowing accurate assessment of the aortic root, ascending aorta, aortic arch, coarctation, descending aorta and existing collateral circulation. (Fig. 16 on page 23)

Its treatment can be surgical repair, implantation of an endovascular prosthesis or balloon dilation.

MDCTA is useful after treatment to detect any complications such as residual stenosis, restenosis, aneurysm formation, dissection and rarely rupture. (Fig. 17 on page 24)

A. 5. PSEUDOCOARCTATION OF THE AORTA

It consists of an elongation and kinking of the aortic arch with secondary narrowing of the aorta at the level of the isthmus. It does not produce collateral circulation. It usually presents no symptoms and appears by chance.

A. 6. INTERRUPTED AORTIC ARCH

It is a rare anomaly consisting of complete luminal and anatomical discontinuity between the ascending and the descending aorta.

There are three types:

1. Distal to the left subclavian artery.
2. Between the left common carotid artery and the left subclavian artery.
3. Between the brachiocephalic trunk and the left carotid artery.

The patent and dilated ductus irrigates the distal aorta to the interruption.

Patients present in the neonatal period with cyanosis, distress and congestive heart failure. It may be an isolated anomaly, although it is usually associated with other cardiac disorders (interventricular septal defect, truncus arteriosus, transposition of great vessels, etc.).

Echocardiography is used for diagnosis and MDCTA is used to monitor postoperative patients and to study other associated abnormalities. (Fig. 18 on page 24)
It requires surgical treatment.

A. 7. MID-AORTIC DYSPLASTIC SYNDROME OR MIDDLE AORTIC SYNDROME

It is a very rare syndrome of unknown origin, although many researchers suggest that this is a noninflammatory congenital hypoplasia of the thoraco-abdominal aorta. It usually manifests in the 2nd decade of life with hypertension, abdominal pain, weakness or absence of femoral pulses.

The stenosis is usually restricted to the thoracoabdominal junction, but it can be extended to the abdominal aorta and the visceral branches.

MDCTA values the location and extent of the stenosis, as well as the presence of collateral circulation (Fig. 19 on page 25). It is also very useful in planning or monitoring of treatment (Fig. 20 on page 25).

Treatment consists in revascularization, either by stenting or vascular grafts.

B. ACQUIRED AORTIC PATHOLOGY

B. 1. TAKAYASU'S ARTERITIS

It's a rare condition, more frequently found in asian patients. It is a chronic inflammatory arteritis of the largest blood vessels. Aorta is the most commonly affected vessel, it affects the abdominal aorta in the 60-75% of cases and the thoracic aorta in the 40-50%.

The diagnosis is based on the symptoms, blood count and imaging tests.

Common findings at MRI and MDCTA are: thickening or enhancement of the wall, lumen dilation, stenosis or occlusion of the affected blood vessels. Both MRI and MDCTA are helpful to value the progression or stability of vascular anomalies and their complications (Fig. 21 on page 26).

B. 2. AORTIC ANEURYSMS
Aortic aneurysms are very rare in children and are usually related to other pathologies or previous trauma history.

a) Aneurysms associated with connective tissue diseases

- Marfan syndrome: Disease of the connective tissue with dominant autosomal inheritance, it may associate cardiovascular anomalies such as mitral valve prolapse, aortic root dilation or descending aorta aneurysms. More than a 80% of the patients present prolapse of the mitral valve and dilation of the aortic root before they are 18 years old. The most significant complications are aortic dissection and rupture of the aorta. The diagnosis is based on echocardiography and MRI / MDCTA (also useful for follow-up). (Fig. 22 on page 26)

- Ehlers-Danlos syndrome: Group of connective tissue disorders, affecting the skin, ligaments, joints, blood vessels and visceral organs. The most frequent cardiovascular associated abnormalities are prolapse of the mitral valve and others like dilated aortic root, aneurysm, and aortic dissection or rupture. Marfan aneurysm differs from that in the Ehlers Danlos, because in Ehlers Danlos they are fusiform and occur in different locations of the thoracic and abdominal aorta.

b) Aneurysms associated with noninfectious aortitis

- Takayasu's arteritis (previously explained)

c) Infectious aortic aneurysms

Acute infectious aortitis in children is usually caused by bacterial septicemia resulting from infected catheters or intravascular devices, valve endocarditis or direct extension. It usually affects patients with risk factors such as congenital heart disease or immunosuppression Staphylococcus or Streptococcus spp are the most frequent ones, and fungi for immunosuppressed patients.

The virulent organisms can adhere and invade the aortic wall, which can lead to a suppurative necrosis that weakens the wall and predispose to the formation of an aneurysm or pseudoaneurysm. Finally, it can cause rupture of the aneurysm or pseudoaneurysm, which is the most serious complication of infectious aortitis.

The diagnosis can be made with MDCTA or MRI, although MDCTA is preferred for its greater speed and accessibility (Fig. 23 on page 26).

d) Posttraumatic aortic aneurysms
They appear as after effects of a trauma or as complications subsequent to heart surgery or manipulation. They are false aneurysms, held by the adventitial layer (Fig. 20 on page 25).

B. 3. AORTIC DISSECTION

It is rare in children and is usually secondary to weakness of the media associated with coarctation, hereditary disorders (Marfan syndrome, Ehlers-Danlos syndrome, Takayasu) or manipulation (Fig. 24 on page 27).

Classic findings of acute aortic dissection are two lumen filled with contrast (true and false lumen) separated by the intimal flap. The true lumen is recognized for its similar enhanced related to the not dissected aortic lumen, smaller diameter and usually located in the inner side of the aorta. The false lumen is usually placed on the outer curvature of the aorta and usually has greater diameter than the true lumen.

B. 4. AORTIC POSTOPERATIVE COMPLICATIONS

MDCTA is often used to detect complications after aortic stent placement. The most common are endoleaks, migration, collapse, thrombosis of the prosthesis and "kinking". Other rare complications are pseudoaneurysm caused by infection of the prosthesis, dissection, embolism, etc. (Fig. 17 on page 24 and Fig. 24 on page 27). Images of the MDCTA compared with MRI in the control of aortic stent are less susceptible to artifacts and can demonstrate patency of stents and its relation to the great vessels.

C. AORTIC PATHOLOGY ASSOCIATED WITH COMPLEX CONGENITAL HEART DISEASES

In general, MDCTA has a limited role in the diagnosis of untreated cyanotic heart disease as it appears during the first days of life and it is diagnosed by echocardiography.

In those cases the indications are:

1. Evaluation and measure of the confluence of the pulmonary arteries
2. Evaluation of coronary arteries anomalies associated
3. Post-surgical evaluation of the permeability of palliative shunts and residual cardiac defects (Fontan, Glenn, ascending aorta - main pulmonary artery shunt, etc.)
4. Assessment of anatomy pre and post-surgical procedure (Fig. 25 on page 28, Fig. 26 on page 28, Fig. 27 on page 28 and Fig. 28 on page 29).
5. Assessment of post-surgical complications
6. Noninvasive method of study prior to definitive diagnosis

Congenital heart diseases can be associated with an anomalous origin of the coronary arteries. Most common are an anomalous origin of the left coronary artery from the pulmonary cone and an anomalous origin of the non-coronary sinus or opposed-sinus. In pediatric patients, anomalous origin of coronary arteries is associated with complex congenital heart diseases such as transposition of the great vessels, Fallot tetralogy and unique ventricle, but they can also be found alone.

There are four possible coronary artery anomalies:

1. Origin of the right coronary artery from the left sinus.
2. Origin of the left coronary artery from the right sinus.
3. Origin of anterior descending and circumflex arteries from the right coronary sinus.
4. Origin of left or right coronary artery from the non-coronary sinus.

Anomalous right coronary artery with interarterial course has the worst prognosis; the others have a better prognosis and are considered benign (Fig. 29 on page 30).
Fig. 1: 1. Aortic root (a. Aortic annulus, b. Sinuses of Valsalva, c. Sinotubular junction) 2. Ascending aorta (from the aortic root to the brachiocephalic trunk) 3. Proximal aortic arch (up to the left subclavian artery) 4. Distal aortic arch or isthmus (from the left subclavian artery to the ligamentum arteriosum) 5. Descending aorta (from the ligamentum arteriosum to the diaphragm)

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**Fig. 2:** 20-day-old male, operated on for interrupted aortic arch. He presents an abnormality of the supra-aortic trunks; carotid arteries arising from a single trunk (arrowhead) and aberrant right subclavian artery (arrows).

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**Fig. 3:** 7-day-old newborn with suspected Scimitar syndrome, based on echocardiography. The MIP and 3D images of coronary reconstructions show an abnormal origin of the supra-aortic trunks; a common trunk from which the left subclavian artery and both common carotid arteries arise (arrow) and the origin of the left vertebral artery in the aortic arch (arrowhead), proximal to the origin of the left subclavian artery.

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**Fig. 4:** 25-day-old male, with Down Syndrome and pulmonary hypertension. He is O2 dependent. The echocardiography reveals an atrial septal defect and a ventricular septal defect. The MDCTA, coronal MIP and 3D reconstruction, show an ectopic origin of the left vertebral artery, from the distal aortic arch (arrows).

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**Fig. 5:** 1-month-old female with clinical signs and symptoms of stridor. The axial MDCTA images (a, b) show a complete vascular ring (arrowheads) around the trachea and the esophagus (nasogastric intubation in lumen). (c, d) The 3D reconstructions show two arches (arrowheads) arising from the ascendent aorta and converging at a lower level in a sole descendent aorta.

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Fig. 6: 13-year-old male without symptoms. The axial MDCTA image - performed for other reasons- shows a right sided aortic arch (arrow) and an aberrant left subclavian artery (arrowhead).

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**Fig. 7:** Right sided aortical arch (arrows) with mirror image branching: 1 left brachiocephalic trunk, 2 right carotid artery and 3 right subclavian artery (numbers).

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**Fig. 8:** 2-month-old breastfed male with clinical signs and symptoms of heart failure. MDCTA - sagittal (a) and 3D oblique sagittal (b) reconstructions - reveal a patent ductus arteriosus (arrowheads) of 6 mm in length that connects the proximal segment of the descending aorta with the main pulmonary artery, which is close to the bifurcation. (c) Furthermore, the MinIP coronal reconstruction shows a mosaic perfusion pattern in both lung parenchyma caused by pulmonary hypertension and a tracheal bronchus as anatomical variant (arrow).
Fig. 9: 3-month-old breastfed male. In the sagittal reconstructions (a) and 3D images (b), a large-caliber ductus arteriosus (arrow) with moderate aortic ectasia at that level and immediately proximal kinking (arrowhead) is observed. Enlarged cardiac cavities secondary to left-right shunt. The patient was treated with surgical closure of the ductus.

Fig. 10: Blood vessel that normally connects the proximal descendent aorta with the left pulmonary artery. It usually closes shortly after birth. If it remains opened 3 months after birth, it is considered patent ductus arteriosus (arrows)
**Fig. 11:** 4-year-old male with Williams syndrome. (a) The axial MDCTA image shows pulmonary arteries with small interior diameter (arrows). The MIP and 3D oblique sagittal reconstructions show a narrowing of the aortic arch too and of the descendent thoracic aorta (arrowheads). This narrowing is related to hypoplasia.

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**Fig. 12:** 4-day-old newborn with anomalous right pulmonary venous return (the inferior pulmonary vein drains the innominate vein through a collecting duct). Besides, the CT images - coronal and oblique sagittal MIP (a,b) and 3D (c) - show hypoplasia in the distal aortic arch (arrowheads) and patent ductus arteriosus (arrows).

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**Fig. 13:** 3-week-old male. (a) The sagittal MIP image shows stenosis (related to coarctation) distal aorta on its way out of the left carotid artery (arrowhead). (b) The 3D reconstruction shows a segment of aorta of 7 mm in length, between the origin of the left carotid and subclavian arteries, with a narrowing of the interior diameter; hipoplastic (arrows). The patient underwent surgical treatment (removal of the coarctated segment).

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**Fig. 14:** 8-year-old female with previous history of interventricular communication and patent ductus repaired. MDCTA shows an aortic isthmus stenosis (arrows) and abdominal aorta hypoplasia, more significant distal to the origin of the renal arteries (arrowheads).
Fig. 15: 16-year-old male with no symptoms. A MDCTA revealed the presence of a coarctation of the aorta (arrowheads) associated with an aneurysm of the ductus arteriosum (arrows).
**Fig. 16:** 8-year-old male. MDCTA shows a severe coarctation at the level of the aortic isthmus (periductal) (arrows) and a bicuspid aortic valve. The dilation of the left subclavian artery and the presence of extensive collateral circulation (arrowheads) are significant.

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**Fig. 17:** 15-year-old female with already treated coarctation of the aorta. MDCTA was performed to check the permeability of the stent (arrows) and the absence of complications.

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**Fig. 18:** 20-day-old male, operated on for interrupted aortic arch (reparation with left subclavian artery). MDCTA evidences the association with (a) an aortic arch hypoplasia (arrow) and (b) a critical stenosis in the origin of the left pulmonary artery (arrowhead). The patient subsequently suffered from a restenosis of the aortic arch that required balloon dilation.

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**Fig. 19:** 2-month-old male hospitalized due to cardiogenic shock. The frontal (a) and lateral oblique (b) MIP images and the 3D images (c,d) show stenosis of the juxta-renal aorta (arrows), as well as an extensive collateral circulation that connects the thoracic with the abdominal aorta through the internal mammary arteries, superior epigastric arteries, inferior epigastric arteries and external iliac arteries (arrowheads). Renal arteries receive blood through collateral circulation from the posterior intercostal arteries, lumbar arteries and renal arteries.

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Fig. 20: Already treated middle aortic syndrome. MDCTA - performed after the placement of two stents (arrowheads) in the stenotic segment of the abdominal aorta - shows a separation between both stents (initially placed next to each other), as well as an aneurysmal dilation of the aorta between both stents.

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Fig. 21: Male diagnosed with Marfan syndrome. MDCTA shows an aortic root aneurysm. The ascending aorta, the aortic arch and the descending aorta have a normal interior diameter. The aortic valve is tricuspid.

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Fig. 22: Marfan syndrome. (a,b,c) MDCTA reveals a fusiform aneurysm of the descendent aorta (70 x 70 x 92 mm) arising from the aortic root, which is typical of the aneurysms associated with this syndrome. (d) The patient underwent surgery and the aortic root was replaced by an artificial valve prosthesis.

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Fig. 23: 2-year-old woman with heart transplant. MDCTA shows the presence of two false aneurysms in the left subclavian artery (arrows) and in the ascending thoracic aorta (arrowhead). The blood cultures showed bacteraemia due to Pseudomonas. The patient underwent surgical treatment for both aneurysms.

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Fig. 24: Complication after endovascular treatment of middle aortic syndrome. The control arteriography performed to the patient showed focal dissection of the intima
(arrowheads) at the level of the aneurysmal dilation present between both stents (arrows). Conservative treatment of the complication was decided.

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Fig. 25: Operated complex heart disease assessment. 8-year-old woman, operated on for TGA and VSD with the Rastelli procedure. A pulmonary valved conduit was implanted. MDCTA was requested in order to assess the situation of the pulmonary artery (arrows) and the aorta (arrowhead) with respect to the sternum to perform sternotomy in a new operation

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Fig. 26: Scimitar syndrome. 7-year-old female. MDCTA reveals the existence of: (a) partial anomalous pulmonary venous return with the right inferior pulmonary vein draining towards the inferior vena cava (yellow arrowheads), anomalous blood supply from the aorta to the right inferior lobe of the lung (orange arrowheads), (b) right pulmonary hypoplasia (yellow arrows) and (c) isthmus of pulmonary tissue connecting the posterior-basal segments of both lungs (horseshoe lung) (orange arrows).

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**Fig. 27:** 9-day-old male with pulmonary artery atresia and ventricular septal defect. He presents an abnormality in the origin of the pulmonary arteries, which originate in major aortopulmonary collateral arteries (MAPCAS). Axial MDCTA images (a,b,c) and oblique sagittal MIP images (d,e,f) show a branch that supplies the right superior lobe of the lung (orange arrowheads) in the proximal aortic arch. A trunk (arrow) arises from the distal aortic arch, goes backwards and bifurcates in the main right and left pulmonary arteries (yellow arrowheads).

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**Fig. 28:** 9-month-old female with operated truncus and protein-losing enteropathy. MDCTA showed several arteriovenous malformations that depend on the inferior
mesenteric artery (arrowheads). The abdominal aorta has a small interior diameter, which is probably a consequence of steal syndrome.

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Fig. 29: 2-year-old woman operated on for tetralogy of Fallot. MDCTA reveals a false aneurysm of the right ventricular outflow tract (arrows). There is also a coronary anomaly with an origin of the anterior descendent artery (yellow arrowhead) from the right coronary artery (orange arrowhead).

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Conclusion

Aortic pathology is rare in children, however, a quick diagnosis contributes to a correct therapeutic planning.

MDCTA is a useful and very quick imaging modality for the morphological assessment of the aorta. A combination of axial images with 2D multiplanar reconstructions and 3D images can be used to diagnose aortic abnormalities and evaluate in an accurate manner its anatomical relationships.
References


