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# MR imaging of aortic coarctation

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***Purpose.*** *The purpose of this paper is to analyse the contribution of MRI as diagnostic procedure in the preoperative diagnosis of aortic coarctation (CoA), in patients with clinical and echocardiographic suspicion for this disease.*

***Patients and methods.*** *During the period of three years, eight patients were examined, 5 (62.5%) male and 3 (37.5%) female patients with clinical echocardiographic suspicion of CoA. The ratio between male and female patients was 1.7 : 1. The youngest patient was 3 and the oldest 46 years (median age was 15 years). Without administration of contrast media and using body coil the examinations were performed with MR machine Magnetom 1.0 Tesla (»Siemens«), with the slice thickness of 6 mm, Fast spin-echo (FSE) T1W sequences, Cine gradient echo (GRE) sequence with slab 7 mm and time of flight (TOF) sequence with MIP reconstructions were applied. During the examinations the patients underwent also ECG gating. Examinations were done in axial, coronal and oblique sagittal projections with measuring of the dimensions of cardiovascular structures.*

***Results.*** *CoA was found in 8 (100%) patients. In 7 (87.5%) cases, coarctation developed at isthmus and in one case, coarctation was detected at the horizontal part of aortic arch, between the truncus arteriosus of the left carotid communis artery. Aortal insufficiency was found in 7 (87.5%) patients; in four of them (50%), bicuspidia was confirmed (bicuspid aortic valve), 7 (87.5%) patients had slightly expressed hypertrophy of the left ventricle. Two (25%) patients had dilatation of the ascendant aorta, six (75%) wider outgoing vessels of the aortic arch, four (50%) had well developed arterial collaterals and 2 (25%) patients rib notching. In 2 (25%) patients as side finding thymus persistent was found. Average diameter of coarctation was 10 mm. In one patient, CoA was accompanied with stenosis of pulmonary artery, in one with ventricular septal defect, and one with tricuspid insufficiency. The results of MRI 100% were in correlation with clinical and echocardiographic findings.*

***Conclusions.*** *MRI is a non-invasive method of investigation of the heart and large blood vessels and it is more and more an alternative to the invasive angiographic investigations, especially in paediatrics, because there is no radiation at all. It is complementary to the echocardiographic, intra-arterial digital subtraction angiography (IA DSA) and helical CT (SCT).*

*Key words: aortic coarctation-diagnosis; magnetic resonance imaging*

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## Introduction

Coarctation of aorta (CoA) is acongenital anomaly of the aortic arch as anomaly of the caliber.[1] It occurs as the obstruction of aortic isthmus, very frequently localized distally to the left subclavian artery, opposite to insertion of the ductus arteriosus.[2-4] The level of the collateral blood circulation through the subclavial and intercostal arteries depends on the level of narrowing of the aortic coarctation.[3] A stenosed lumen can be narrowed into the size of the knitting needle, but it can be very rarely completely occluded.[5] Pathohistologicaly this is an obstruction membrane in location of aortic isthmus.[6]

Based on the statistics of M. Abbott, we meet this anomaly in 8% of overall congenital heart malformations; it is three times more frequently in males than in females patients.[5] This is the third most frequent anomaly of cardiovascular system.[3]

There are two types of CoA: juvenile - the diffuse type (preductal tubular hypoplasia) and the adult type - stenosis of short segment (postductal and periductal).[2-5]

This anomaly can be accompanied with the so-called coarctation syndrome, which is a coartctation triad of previous ductus arteriosus and ventricular septal defect. It can also be accompanied with hypoplasia of aortal arch (small arch) bicuspodia of the aortal valve (25-50%), aneurysm of sinus valsavle, aneurysm at the place of coartctation, aneurysms of subclavia artery, ductus arteriosus or circulus Willis, anomalies of aortal arch as in many other congenital heart diseases (until 70%).[2,3,6,7] We meet it also in Turner's syndrome.

Coarctation of abdominal aorta is seen in 2% of cases.[2]

Radiologically similar to coarctation is so called pseudocoarctation. It is unusual asymptomatic variant of coarctation, in which the descendent end of aortal arch retracted sharply in front, at the place of insertion of the arteriosum ligament. The aortal arch above the bended part is abnormally high and convex and it can look similar to the tumour of back upper part of the mediastinum. Stenosis, deficit of pulse and rib notches do not exist in pseudocoarctation.[2,6,8]

The idea of some authors is to declare all mediastinal masses as vascular etiology, until it is not proved the otherwise.[6]

Two changes are regarded as repercussion to coarctation: development of arterial collaterals and hypertrophy of left ventricle.[2]

Coarctation of the aorta is a disease which very frequently escapes on the early detection and treatment, too. This may causes cardiovascular accidents and early death as the consequence of the coartctation increase with delaying the treatment.[9]

It was diagnosed in about 20% of our patients at adolescence for the first time. In a big series of patients with coarctation of aorta the mean age at diagnosis was 10 years. In younger patients most frequent sign (mean age 6 years) was a murmur, and in older patients (mean age 18 years), was systemic hypertension.[9]

Average survival of patients with the adult type of this malformation is 33 year; only 25% survive 40 years.[5] Death occurs because of the rupture of aorta (around 25%), infective endocarditis or aortitis (around 20%), heart insufficiency (around 20%), intracranial haemorrhage (around 10%), and rupture of the heart (1%).[5]

Beside murmurs and hypertension, other accompanying signs are the delay of pulse between radial and femoral artery, difference in pressure between upper and lower extremities or rib notches in the lower margins of ribs on the thoracic plain film.[9] Rib notches were anatomically found by Meckel in 1827 and radiologically by Resler in year 1928. They are almost always present in adults, especially we meet them between the 3$^{th}$ and 6$^{th}$ rib, some authors found them between the 4$^{th}$ and 8$^{th}$ rib, bilaterally, but not symmetrically.

Although, usually they do not occur in children younger then 7-8 years of age, they can be met even at child nine months old. According to Pugh they are not present in 1/3 of cases.[2,5]

Beside rib notches, thoracic plain film shows hypertrophy of the left ventricle, aorta ascendant very frequently forms the right edge of the heart, and rarely aortal button is missing. The examination of the esophageous with barium contrast showed the number three sign. Lung vascularisation is normal.[2,5]

Most frequent clinical feature is hypertension of upper extremities, while femoral pulse is usually not palpable.[6]

The treatment of coarctation is surgery with resection of coarctation and end-to-end anastomosis, patch isthmio-plastics with synthetic material or subclavian patch, balloon angioplasty, for stenosis of short segment.[2,6]

Beside the above mentioned clinical findings and the thoracic plain film, in the diagnostics of the coarctation, the conventional angiography, intra arterial digital subtraction angiography (IA DSA), helical CT (SCT), trans-oesophageal echocardiography (TEE) and magnetic resonance imaging (MRI) are used.[4,6,9] The last three methods are non-invasive and more and more used in the diagnostics of the conditions of the heart and vascular anomalies. MRI is especially interesting in establishing the diagnosis of coarctation of aorta and planning the treatment in the early phase because of its multiplanarity, no radiation and information which it can give about the present haemodinamic disturbances.

The purpose of our study is to analyse the MRI contribution in diagnostics of CoA.

## Patients and methods

In the period of three years eight patients are examined and all of them were suspect clinically and echo-cardiographycally for CoA. Among those patients 5 (62.5%) were male and 3 (37.5%) female. The rate between male (5) and female (3) patients was 1.7 : 1. The youngest patient was three years old and the oldest 46 (average age was 15 years). Examinations were performed with MR machine Magnetom 1.0 Tesla (»Siemens«). We did not use the contrast media because of missing the proper connector required by the size of gantry. During the examination, fast spin-echo (FSE) T1W (black blood) sequences were used, especially in the oblique saggittal plane because of determining the location and level of the stenosis, the spread of the coarctation and presence of collateral vessels in regard to the morphology of vascular structures and accurate orientation Cine-gradient echo (GRE) sequence. From GRE (bright blood) sequence, Cine sequence is used aiming to evaluate the disturbance of signal void in the part of coarctation and valve apparatus (place of stenosis). In other words the evaluation of the functional state of the aortal coarctation and time of the flight (TOF) sequence was performed, with the use of maximum intensity projection (MIP) reconstructions, to show the place of coarctation, outgoing vessels of the aortic arch, collaterals and collateral flow. During the examination, ECG gating was also used.

Examinations were performed in axial, coronal and oblique positions, in the direction and extend of aortic arch, with the slice thickness of 6 mm. In cases of improper cross-section because of the thickness of the slices in the region of the coarctation e.g. in case of small children, stratified slices were made to acquire the best scan. During the investigation, measurements of the heart wall and heart cavity were performed, especially of the left ventricle, ascendant aorta, aortic arch, descendent aorta and the zone of coarctation. Examinations were done preoperatively in order to evaluate the severity of the disease and decide about the preoperative plan. Body coils are not used because they were not available.

## Results

Among the examined patients, coarctation of the aorta was found in 8 patients. In 7 (87.5%) cases, the coarctation developed at the isthmus, and in one case at the horizontal part of the aortic arch. Aortal insufficiency was confirmed in 7 (87.5%) patients, in 4 (50%) of them bicuspidia was observed, while in 7 (87.5%) patients hypertrophy of the left ventricle was slightly expressed. Wider outgoing vessels from the aortic arch were found in 6 (75%) patients. In 2 (25%) patients dilatation of the ascending aorta was observed. In 4 patients (50%), collateral flow was well developed, and in 2 (25%) patients, rib notching in the plain thoracic radiographs was revealed.

In one patient coarctation was accompanied by stenosis of pulmonary artery, in one by ventricular septal defect and in other one with tricuspid insufficiency. Those findings were in correlation with echo cardio graphic findings. The oldest patient (46 years) had in medical history the heart infarction and dilated cardiomyopathy. Patient was with coarctation in the horizontal part of the aortic arch between truncus brachyocephalicus and the left carotid communis artery, and with knocking of the same in its back part (variant of pseudocoarctation). As accompanying finding, in 2 (25%) younger patients, thymus persistent was found.

In all cases spin-echo (SE) sequence gave an excellent morphologic presentation of the location of coarctation of the ascendant aorta, aortic arch, with outgoing vessels from the arch, descendent thoracic aorta, discontinuity of the collateral flow and mammary artery. An average diameter of the coarctation place was 10 mm; in one young female patient 13 aged, the diameter was 5 mm.

Cine sequence in the slab of 7 mm, though providing satisfactory morphologic presentation in real time, appears to be extraordinary sensitive to the velocity, thus clearly showing the signal void in the region of the valve apparatus and at the site of coarctation.

The image of aortal coarctation was complete after using the time of flight (TOF) sequence that contributed enormously to morphology, in addition to providing an image of the collateral flow and the mammary artery.

Using this set of sequences - about the morphology of the coarctation and circulatory disorders - important information was obtained in a non-invasive way, and without using the contrast media.

MR results were in 100% of cases in correlation with the ones gained by echocardiography when it was about the heart's morphology and the valve apparatus.

## Discussion

As it was presented in the introductory part, CoA is the most frequent anomaly of the cardiovascular system. Its diagnostics is relatively easy and for a long time it has been made only clinically, based on the discovery of heart murmur, hypertension, differences in pressure between upper and lower limbs and rib notches in the thoracic plain films.

In this investigation the most frequent clinical sign in young patients was heart murmur and the difference in pressure between the upper and lower limbs, and in older patients hypertension. Rib notches were found in a 1/4 of patients, whereas in literature there were found in 1/3. Here we have to bear in mind that half of our patients were younger then 8 years.

In this series the oldest child was 13 year old, and the 2 grown patients were 30 and 46 years old, respectively. In the larger series the mean age of patients at diagnosis was 10 years, and in our series it was 15 years. The ratio between male and female patients was 1.7 : 1 in our study, while in literature it was 3 : 1.[5]

In adults over 40 years CoA is observed in

only 20 % of cases, and in our small series 1 (25%) patient was over 40 years. Aortal insufficiency was found in 7 patients; in 50% of them bicuspidia was discovered, which is in line with the percentage reported in literature, where it ranges between 25-50%.[2] Correlation with other congenital anomalies in CoA is high and reaches 70 %; in our series, it was 37.5%. The obtained results were in line with the one gained from echocardiography. MR proved to be very sensitive to flow disturbances.

CoA falls in the group of curable congenital anomalies of the vascular system. The treatment of choice is surgery; therefore, it is necessary to establish a precise diagnose and to identify all accompanying anomalies.

Today, several excellent methods for analysis of the big blood vessels and Coal, like SCT, MR and TEE, are used.[3]

Conventional angiography has been, until recently, the golden standard. It allows measuring the pressure, but it is invasive, risky because of radiation, haemorrhage, vascular lesions, thrombosis and allergic reactions to the contrast media.

Echocardiography is non-invasive alternative which is widely used today. However, at times, it is very hard to acquire good visualization of the site of coarctation because of small acoustic window, long distance between transducer and the isthmus region, and because of non-cooperation of patient.

In recent years the use of MR imaging have become more intensive and proved to be excellent tool for non-invasive investigation of cardiovascular system in older and younger patients.[10]

In our series clinical and echocardiographic suspicions on CoA were confirmed in 100% of cases by MRI with the use of fast spin echo (FSE), Cine and TOF (time of flight) sequences, that allow an extraordinary morphologic presentation of the heart and big vascular structures, as zones of disturbed flow (Figures 1,2,3,4).

**Figure1.** Coarctation of aorta, spin-echo (SE) T1W (Black blood)

**Figure 2.** Coarctation of aorta on MRI with measurements

Drawbacks of this technique are its incapacity to measure the pressure in the zone of coarctation and the impossibility of applying of contrast media in the way as it is applied in contrast enhanced magnetic resonance angiography (CE MRA). Reasons for this were of technical nature. The shortage of the thoracic coil was, to a certain degree, substituted by proper sedation and preparation of patient before scanning.

**Figure 3.** Coarctation of aorta, Cine - gradient echo (GRE) (Bright blood - signal void: arrow).

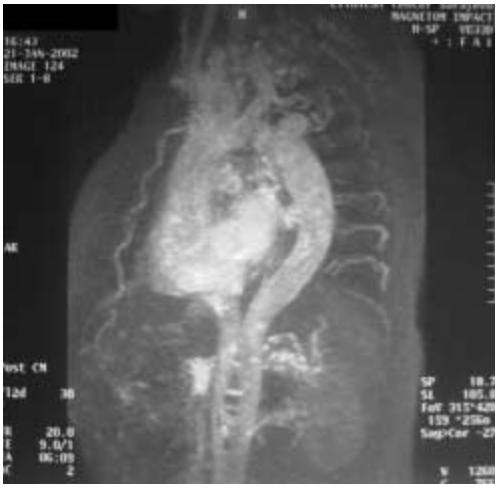

**Figure 4.** Coarctation of aorta, arterial collateral's time of flight (TOF).

According to literature MR, imaging is a powerful tool in the evaluation of full range of congenital and acquired diseases of the thoracic aorta. It is safe, accurate and can be repeated several times, giving extremely useful anatomic information and clearly detecting collateral vessels. It also provides information about the extraluminar relationship of aorta with the surrounding organs, no matter in which projections they are seen.[11]

In our investigation, this advantage was observed with the discovery of thymus in 2 younger patients with coarctation of aorta.

Today, in the diagnostics of aortal coarctation, FSE and angiographic gradient echo (Cine - GRE, TOF, PC and recently also CE 3D MR) sequence are used worldwide.

MRI SE sequences provide very useful anatomic details, while Cine MRI sequence can detect a turbulent jet of blood and the place of coarctation.

MR angiography (MRA) does not provide adequate information about the wall of the vessel; the intimal flap and parietal thrombus can be dizzy and hardly seen even with the help of MIP (maximum intensity projection) algorithm.

SE and GRE technique are complementary; the first takes a lot of time, especially when ECG triggering is used.

MRI is non-invasive and doesn't depend much on the experience of operator.

In children, sedation is needed, possibly general anaesthesia; echo navigator techniques have been developed to prevent image degradation.[12]

MRI can be used preoperatively to evaluate the severity of the disease and to decide about the operative plan.

Beside the analyses of morphologic abnormalities, it also allows to measure the size of vascular structures with the speed of the flow at the place of stenosis, in order to assess functional disturbances.[13]

It has also some limitations, as artefacts because of respiratory movements, saturation problems and long time of diagnostic orocedure.[14]

Basic (without contrast) MRA-GRE technique is time of flight (TOF) and phase-contrast (PC) sequences.15 Beside those the so-called contrast enhanced (CE) three-dimensional (3D) MRA sequence is also used today. CE 3D MRA and TSE sequence are used in the set for analyzing the morphology of aortal arch, vascular diameter, location and scope of

the abnormality of coarctation, its relationship to the outgoing vessels from aortic arch.[16] Measuring the dimensions of the aorta proved to be very useful before the surgical correction or balloon dilatation of the coarctation. A good correlation was observed between the measurement performed with MIP reconstruction and catheter angiography.[17]

3D reconstruction using the maximum intensity projection (MIP) technique is a reliable non-invasive technique which can replace the diagnostic catheter angiography, especially during postoperative controls of the coarctation. It provides the clinician with valuable information concerning further application of invasive procedures. In regard to decreasing the effect of altered flow dynamics, CE 3D MR angiography of thoracic aorta has several advantages over to spin-echo SE (black blood) and TOF (bright blood) MRI sequences. It is independent from ECG triggering, which is of special importance in patients with arrhythmias. This technique is proved to be reliable in the diagnosis of aortic coarctation.[15] ECG triggered breath-hold contrast material-enhanced magnetic resonance angiography sequence has been developed for imaging the thoracic aorta, aiming to decrease the respiratory motion artefacts and pulsation artefacts. 3D MRA is independent from that if the flow is laminar, turbulent or stagnating. Different congenital and acquired abnormalities are clearly identified with this technique.[18]

Breath hold contrast enhanced 3D MR angiography is faster and more accurate method in diagnostics of the diseases of thoracic aorta, but has limitations in the estimation of the aortal lumen. This is why black blood and bright blood MR sequences are necessary in the analysis of aortal wall, aortal valve and periaortic tissue.[19]

CE MRI is especially useful in small children who cannot keep their breath, which is required in 3D reconstructions.[10] Moreover; gadolinium has smaller nephrotoxicity and less allergic reactions as iodine based contrast medias.[12]

Aortal diameter, stenosis, aneurism, intimal calves or atherosclerotic plaques can be analyzed with 3D reconstruction using MIP technique.[17]

The role of MRI is also of special value for following the patient's with repaired cardiovascular anomalies and is also widely used for presenting the morphology of large blood vessels. MR flow mapping can be used for analyzing the flow in large vessels.[20]

3D CE MRA with 0.2 mmol/kg Gd. in IV bolus, more exactly 20 ml Gd was followed with 20 ml saline most frequently at a flow of 3ml/s with ECG gating and preceding bolus test may be necessary. With such proceeding it is obtains optimal quality of the image, made in the oblique saggittal plain with standard MIP/MPR reconstructions. It is important in case of artefacts from surgical clips which can disturb considerably the quality of the image.[21-23]

3D MRA (FISP) is used to measure the flow and elasticity of the vascular wall. With these techniques it is possible to have an adequate follow up of coarctation, meaning that, usually, IA DSA is not needed in defining the condition and size of the proximal and distal anastomosis and the morphology of the thoracic aorta.[23,24]

The so-called fluoroscopic triggering of centrically encoded 3D MR angiographic acquisitions have also been developed and have been reported as highly accurate methods of acquiring 3D MR angiograms with high spatial resolution.[25]

Because of the reasons mentioned earlier, the described 3D CE MRA sequences were not applied in this study.

## Conclusions

MRI is a non-invasive method of examination; it is becoming more and more an alter-

native to the invasive angiography, especially in paediatric patients, because it is radiation-free. It is complementary to the other methods, like US, DSA and SCT. MRI is an excellent technique for evaluating the morphologic anomalies of aortic isthmus before and after the operation or percutaneus treatment. It may be concluded that, by this method the coarctation of aorta can be followed up non-invasively.

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