

AORTIC COARCTATION IN INFANTS AND CHILDREN – DIAGNOSE, TREATMENT AND PROGNOSIS

Gabriela Doros^{1,2}, Anca Popoiu^{1,2}, Cristina Olariu², Alexandra Popescu²,
Ramona Olteanu², Raluca Isac², Mihai Gafencu^{1,2}

Abstract

Coarctation of the aorta is a congenital heart defect involving a narrowing of the descending aorta. Blood pressure is higher before the narrowing and lower past the narrowing, with 20 mmHg difference between upper and lower limbs, clinically expressed by absent femoral pulses. In infants, coarctation of the aorta is severe and represent a cardiological emergency, but in child, aortic coarctation is sometime under-diagnosed, patients presenting at the hospital for high blood pressure of unknown etiology, or complaining of headache, or lower limb pain in effort.

We want to highlight aortic coarctation as a cause of high blood pressure in children and to drawn attention that this kind of patients, even after aortic coarctation repair may remain with hypertension that has to be treated and patients followed up. We want to share our experience with some cases of surgical correction of aortic coarctation and complications such as recoarctation in special type of gothic aortic arch and also, recoarctation after stent implanting.

Key words: coarctation of the aorta, high blood pressure, children, infants, recoarctation

Introduction

Coarctation of the aorta (CoAo) or descending aorta narrowing is a relatively common defect that accounts 5-8% of all congenital heart defects, with a prevalence of 4 in 10 000 live birth. Coarctation of the aorta may occur at any point after the transverse arch, to the iliac bifurcation, but 98% occur just below the origin of the left subclavian artery at the origin of the ductus arteriosus (juxtaductal coarctation - Fig. 1); rarely, a coarcted segment is present in the lower thoracic or abdominal aorta. It may be as an isolated defect or in association with various other lesions, most commonly bicuspid aortic valve (may be seen in nearly two thirds of infants with coarctation of the aorta), hypoplasia of the aorta, ventricular septal defect, atrial septal defect, transposition of great arteries, and patent ductus arteriosus and complex lesions. Boys have the defect more commonly than girls and the ratio is 2:1, excepted Turner syndrome.

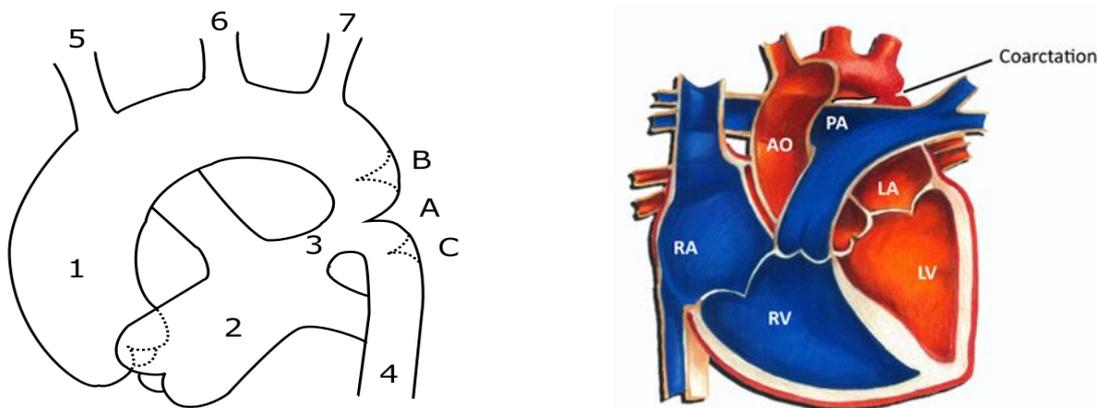


Fig 1. a. Schematic drawing of alternative locations of a coarctation of the aorta, in relation to the ductus arteriosus. A: Ductal coarctation, B: Preductal coarctation, C: Postductal coarctation. 1: Ascending aorta, 2: Pulmonary artery, 3: Ductus arteriosus, 4: Descending aorta, 5: Brachio-cephalic Trunk, 6: Common left carotid artery, 7: Left subclavian artery.; b. Coarctation of the aorta.

¹“Victor Babeş” University of Medicine and Pharmacy Timișoara, Romania

²“Louis Turcanu” Children’s Emergency Hospital Timișoara, IIIrdPediatric Clinic

E-mail: gdoros@gmail.com, apopoiu@umft.ro, olariu.ioanacristina@yahoo.com, popescu.mariaalexandra@yahoo.com, ramona_veronica_tudor@yahoo.com, ralu_isac@yahoo.com, mgafencu@umft.ro

Clinical presentation and examination

In *fetal life*, coarctation of the aorta can be diagnosed by fetal echo, but with no severe consequences on fetus, because of the ductus arteriosus circulation.

After birth, some of the newborns with aortic coarctation will remain asymptomatic due to a good antegrad aortic flow through the aortic isthmus and normal ductal flow, but about half of *newborns* with coarctation of the aorta will develop symptoms in the first days/weeks of life, after ductal closure. This neonates start to have hemodynamic deterioration, with poor feeding, poor weight gain, dyspnea, cyanosis in the lower part of the body, because the circulation in the lower part of the body is through the ductus arteriosus, right to left and soon after, in the time of ductal closure, they start to develop tachypnea, tachycardia, and cardiogenic shock, because of hypoperfusion of the abdominal organs, leading to severe metabolic acidosis and kidney failure. The diagnose include blood pressure (BP) discrepancies between the extremities, with high blood pressure in the upper part of the body and minimum 20 mmHg lower pressure in the lower part of the body and reduced or absent pulses at the femoral artery. Echocardiography will confirm the aortic coarctation. Neonates in this condition need urgent treatment for heart failure with short-acting inotropic agents, correction of acidosis, sometime intubation and mechanical ventilation before immediate surgical correction.

In *children*, aortic coarctation is generally asymptomatic and is incidental discovered and diagnosed. Children are sometime complaining of headache, leg pain after effort, claudication, epistaxis. At the clinical examination, they can be discovered with cardiac murmur and high blood pressure in the arms and absent or diminished femoral pulse. In coarctation of the aorta, blood pressure is higher before the narrowing of the aorta and lower past the narrowing, with a minimum 20 mmHg difference between upper and lower limbs, associated with absent femoral pulses. Symptoms depend on how much blood can flow in the lower part of the body, through the coarctation. During time, collateral circulation from aorta will develop in the upper part of the body, to feed the lower part of the body.

In milder cases, symptoms may not develop until the child has reached adolescence. Other symptoms include: chest pain, cold feet or legs, dizziness or fainting, decreased ability to exercise, failure to thrive, leg cramps with exercise, nosebleed, poor growth, pounding headache, shortness of breath.

Diagnose

It is based on clinical examination, electrocardiography (ECG), cardiopulmonary X-ray, echocardiography, angioCT with 3D reconstruction or angio MRI and if necessary catheterization.

Electrocardiography in newborn will show right axis deviation and right ventricular hypertrophy, much frequent than left ventricular hypertrophy; in children, ECG may be normal in 20% of cases, meaning large coarctation, or may

reveal left ventricular hypertrophy, meaning narrow coarctation.

Cardiopulmonary X-ray will show marked cardiomegaly with pulmonary edema or pulmonary venous congestion in newborns and on barium esophagogram the “E” shaped indentation or reversed figure of “3” sign configuration and rib notching in children older than 5 yo, due to collateral circulation.

Echocardiography, from suprasternal notch will show the location of the coarctation and with continuous Doppler will measure the gradient through the coarctation.

Angio CT with 3D reconstruction is the investigation that reveal the exact site and the size of the coarctation and the collateral circulation. The time for this investigation is very short and applicable in newborn and infants, but this method is irradiating.

Angio MRI is also a perfect investigation to reveal the aortic coarctation, non-irradiating, but with long time of sedation. It is perfect for older children, that cooperate and do not need sedation, or for small children that do not cooperate and need sedation.

Catheterisation is not a diagnostic tool now, but it is used in the treatment of aortic coarctation, a perfect method for interventional dilatation and stent implantation.

Treatment

Medical treatment

In symptomatic newborns, prostaglandin E1 has to be started to reopen the duct, to assure good blood flow to the kidney. In case of heart failure, short-acting inotropic agents as dopamine or dobutamine, diuretics and oxygen is needed before surgery.

Systemic hypertension has to be treated with beta blockers in older children. Lifelong prophylaxis of bacterial endocarditis is necessary in case of bicuspid aortic valve.

Surgical treatment

Surgical repair can be done by four techniques: resection of the coarctation and end to end anastomosis of the aorta, enlarging the coarcted zone by parching with Dacron, or subclavian flap repair, or conduit insertion in place of aortic coarctation if this is expressed on a large segment of the descending aorta, all done by left lateral thoracotomy. All four techniques have a high rate of recoarctation, especially if the correction is in newborn period or infants. The intervention by parching with Dacron is no longer used, because of a high rate of aneurysm formation. The flap repair needs the left subclavian artery to be legated because the proximal part is used to patch the coarcted segment and the patient is pulsless in the left arm, where the circulation will be assured by collaterals. If the symptomatic newborns are not operated, the mortality rate is around 90%.

Interventional treatment

Interventional balloon dilatation was introduced to treat recoarctation after surgery intervention, but soon after became the first option in the treatment of aortic coarctation in children. Rare complications can be mentioned as: aortic dissection, rupture and aneurysm formation. The age for

balloon dilatation depends on the center to center experience, but still remain the risk for recoarctation

Interventional stent implantation became the treatment of choice in aortic coarctation, with low rates of immediate complications. Despite these, recognized complications are: aneurysm formation, stent fractures and recoarctation, with a lower rate comparing with surgery. Reintervention and stent in stent implantation is possible. Angio CT is used to detect de post procedural aneurysm formation and the stent integrity.

Follow-up and Prognosis

Even after early surgery or interventional repair of aortic coarctation, approximately 30% of patients will be hypertensive by adolescence. 60% of adults, after correction of aortic coarctation in childhood will be hypertensive. This is the reason this kind of patients have to be followed up at 6-12 months. A part of the patients have normal rest blood pressure, but at exercise develop exaggerate blood pressure response, meaning the onset of overt hypertension. In time has to be followed complications regarding the bicuspid aortic valve, aneurysm formation, stent recoarctation and persistent hypertension that have to be medicated. Prognosis depends on complications.

Our experience in follow up complications

We want to highlight two children with aortic coarctation, a girl and a boy, both surgical operated at the age of 5 with end to end anastomosis. They were followed up yearly. In time, they started to develop recoarctation, with significant gradient, hypertension in the arms, headache and pain in the legs. Both were on medication for high blood pressure.

The first case, the girl, at the age of 10 performed angio CT with 3D reconstruction, confirming the recoarctation detected by echocardiography. She had an interventional stent implantation, with good result after implantation, becoming free of symptoms. This was the reason she didn't come for follow up evaluation during a 6 months period. After that time, high blood pressure and headache reappeared and she came for a cardiology check. Significant gradient was found at echocardiography and a new angio CT was done, confirming that the stent was not

complete expanded. She performed a new interventional dilatation of the stent and the gradient dropped to normal. Despite this, high blood pressure persisted, and antihypertensive treatment was continued. She is now in a regular follow up program, but free of symptoms.

The second case, a 12 yo boy started to develop high blood pressure and he was medicated. In time he developed severe headache associated with high blood pressure, despite the medication. At echocardiography he presented serial stenosis due to a gothic aortic arch and associated bicuspid aortic valve with no gradient, but with a subaortic restrictive ventricular septal defect. The gradient was not severe at the level of the surgical operated aortic coarctation, but in serial stenosis, echocardiography is not perfect in detecting the gradient. An angio CT was performed, with 3D reconstruction, reflecting the stenotic areas of the aortic root, due to the gothic aortic arch and the recoarctation. He performed a catheter exploration and balloon dilatation, because stenting was impossible in his aortic arch, due to the risk of obstructing the left common carotid artery. After balloon dilatation he was free of symptoms, but still on medication for high blood pressure. He is in a regular 3 mo follow up program, and in case of symptoms correlated with high gradient, he will perform a complete surgical correction in an experienced cardiovascular surgery center.

Conclusion

Coarctation of the aorta or aortic narrowing is the fifth most common defect that accounts for 5-8% of all live births with congenital heart defects. Absent femoral pulse associated with high blood pressure in upper extremities can suspect this diagnose of aortic coarctation. Newborns and infants who present early with severe coarctation of the aorta are seriously ill and require urgent medication and transfer to a pediatric cardiovascular center for balloon dilatation or surgical corection. Postsurgery re-coarctation has to be followed. Interventional balloon dilatation and stent implantation or surgical correction in selected cases has to be performed in children. Even so, patients have to be followed up because of complications such as recoarctation or persistence of high blood pressure that has to be medicated.

References

1. Ali Devrim Karaosmanoglu, Ranish Dedar Ali Khawaja, Mehmet Ruhi Onur, Mannudeep Karla. CT and MRI of Aortic Coarctation: Pre- and Postsurgical Findings. American Journal of Roentgenology. 2015, Vol. 204, Issue 3
2. Yuan SM., Raanani E. Late complications of coarctation of the aorta. *Cardiol J.* 2008;15(6):517-24
3. Chakrabarti S., Kenny D., Morgan G., Curtis SL., Hamilton MC., Wilde P., Tometzki AJ., Turner MS., Martin RP. Balloon expandable stent implantation for native and recurrent coarctation of the aorta--prospective computed tomography assessment of stent integrity, aneurysm formation and stenosis relief. *Heart.* 2010;96(15):1212-6. Epub 2009 Sep 10.
4. Budoff MJ., Shittu A., Roy S. Use of cardiovascular computed tomography in the diagnosis and management of coarctation of the aorta. *J Thorac Cardiovasc Surg.* 2013;146(1):229-32. Epub 2013 Feb 8.
5. Malčić I., Kniewald H., Jelić A., Sarić D., Bartoniček D., Dilber D., Belina D., Anić D. Coarctation of the aorta in children in the 10-year epidemiological study: diagnostic and therapeutic consideration. *Lijec Vjesn.* 2015;137(1-2):9-17.

- | | |
|--|---|
| <p>6. O'Brien P., Marshall AC. Coarctation of the aorta. <i>Circulation</i>. 2015;131(9):e363-5</p> <p>7. Dzsinič C., Vasziy M., Vallus G., Dzsinič M., Berek P., Barta L., Darabos G., Nyiri G., Nagy G., Pataki T., Szentpétery L. Late complications and treatment options of aortic coarctation operated in childhood. <i>Orv Hetil</i>. 2014; 27;155(30):1189-95.</p> | <p>8. Harris KC, Du W, Cowley CG, Forbes TJ, Kim DW. A prospective observational multicenter study of balloon angioplasty for the treatment of native and recurrent coarctation of the aorta. <i>Congenital Cardiac Intervention Study Consortium (CCISC)</i>. <i>Catheter Cardiovasc Interv</i>. 2014 ;1;83(7):1116-23. Epub 2013 Dec 4.</p> |
|--|---|

Correspondence to:

Gabriela Doros
"Victor Babeş" University of Medicine
and Pharmacy Timișoara
Eftimie Murgu, 2,
Timișoara
Romania
E-mail: gdoros@gmail.com