

Optimisation of the diagnosis in coarctation of the aorta in children and adolescents

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ABSTRACT

Coarctation of the aorta is a congenital heart defects, involving a narrowing of the descending thoracic aorta (hemodynamically significant), most likely located distal from the left subclavian artery, at the origin of the arterial ligament. This article discusses the characteristics of the coarctation of the aorta in children and also the possibilities of treatment.

Key words: coarctation of the aorta, children, hypertension, treatment

INTRODUCTION

Coarctation of the aorta (CoA) is a congenital narrowing of the descending aorta (hemodynamically significant), most likely located distal to the origin of the left subclavian artery, at the origin of the arterial ligament.

CoA accounts for 6-8% of all congenital heart defects or 5 to 10.000 of all viable births. It is more common in males (M/F=2/1). It is more common associated with other congenital heart defects such as: patent ductus arteriosus, bicuspid aortic valve, ventricular septal defect, and mitral valve anomalies. CoA is the most common cardiac defect associated with Turner syndrome (35%). It is 7 times more common in white people than in Asians (1).

In 1903, Bonnet classified coarctation into 2 groups: the infantile type (preductal) and the adult type (postductal). (FIGURE 1, FIGURE 2).

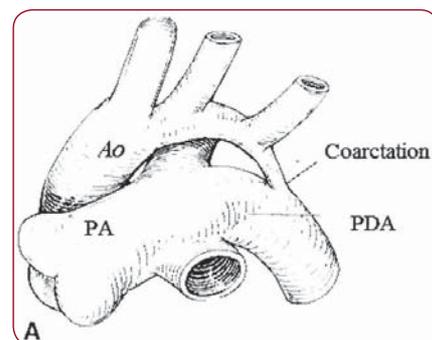


FIGURE 1. Coarctation of the aorta: the preductal type. The patent ductus arteriosus supplies most of the blood flow to the descending aorta. (after Mavroudis) (3)

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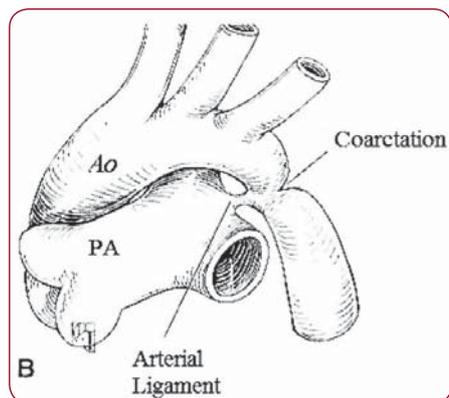


FIGURE 2. Coarctation of the aorta: the postductal type: The area of narrowing is juxtaductal and consists of a posterior infolding localized in the lumen. The arterial duct is now closed (the arterial ligament) (after Mavroudis) (3)

This classification concerns the specific clinical presentation in child and adolescent-adult (2).

In contrast with coarctation of the aorta in the newborn, when the hemodynamic pattern is dominated by the heart failure syndrome, the hemodynamic pattern in the child-adolescent is dominated by the arterial hypertension syndrome. This syndrome develops above the coarcted segment and is associated with poststenotic arterial hypotension, which leads to hypoperfusion of the lower half of the body. The perfusion disturbance favors the development of collateral vessels. The most frequent involved arteries in collateral vessels are the branches of the left subclavian artery and the intercostal arteries, pair number 3 and 4. The cause of hypertension is not only mechanical. Activation of the renin-angiotensin system may interfere (this is released if the renal perfusion is reduced-ischemia) (4).

With regard to the mortality, in USA, the patients with coarctation of the aorta survive to age 35 years. 20% of the patients survive to age 50 years. If the CoA is repaired before the age of 14 years, the 20-year survival rate is 90%. If the CoA is repaired after the age of 14 years, the 20-year survival rate is 75% (5).

If the follow-up care of the patients is not adequate, the diagnosis will be made when arterial hypertension occurs or following a major complication (intracranial hemorrhage, aortic dissection, endocarditis) (6). The treatment methods are well known, but the diagnosis is many times delayed if patients are not carefully evaluated. □

THE DIAGNOSIS

Clinical diagnosis

In children, the coarctation of the aorta is most often asymptomatic and is noted following a routine examination (7). The children may have a normal general development. The clinical presentation may be due to arterial hypertension (headache, dizziness and nose-bleed) or lower body hypoperfusion (muscle weakness, leg cramps, cold feet) (8). The following are noted: absent or diminished femoral pulse, hypertension in the upper extremities, a blood pressure difference of more than 20 mmHg in arms than in legs, intercostal pulsations, systolic murmur best heard in the second left intercostal space parasternal and interscapulothoracic. When the coarctation of the aorta is not diagnosed, complications may occur such as: intracranial hemorrhage, stroke or heart failure (9). Beyond the specific clinical manifestations due to hypertension, the child is brought to the doctor for other congenital disorders or intercurrents.

Imaging studies

- a. Chest radiography. In children the findings are: enlargement of the left heart border, rib notching C3-C8 in children older than 5 years (due to hypertrophied intercostal arteries), the shape of „3” number observed at the thoracic descending aorta (due to the traction of the arterial ligament). In some cases, barium esophagram shows the compression of the esophagus with dilatation of the pre and poststenotic segment (FIGURE 3) (2,10).
- b. Electrocardiogram reveals signs of left ventricular hypertrophy, left atrium dilatation, and incomplete right bundle branch block.
- c. Echocardiography excludes or confirms other associated significant intracardiac anomalies (bicuspid aortic valve 85%, mitral valve anomalies, subaortic stenosis) and allows assessment of left ventricle size. Doppler echocardiography assesses the site of obstruction and the severity of hemodynamic consequences; it shows a pressure gradient more than 20 mmHg (20 mmHg – mild clinical form, 70-80 mmHg – severe form of disease). The blood velocity measured by continuous wave Doppler, at the point before the coarctation, must be accounted for

in measuring the pressure gradient across the aortic arch (4,7). Normally, the blood flow in descending aorta has a fast ascending velocity signal during systole and a short retrograde blood flow during early diastole. In coarctation there is reduced systolic ascending velocity signal and continuous antero-gradate blood flow during diastole.

In some patients, coarctation may be difficult to diagnose by surface echocardiography. For these patients, transesophageal echocardiography, MRI or cardiac catheterization with angiogram may be necessary to make the diagnosis.

The positive diagnosis of coarctation can be made many times by physical examination only such as: arterial hypertension in child and adolescent, the pulse difference in arms and legs, intercostal pulsations, interscapulothoracic systolic murmur.

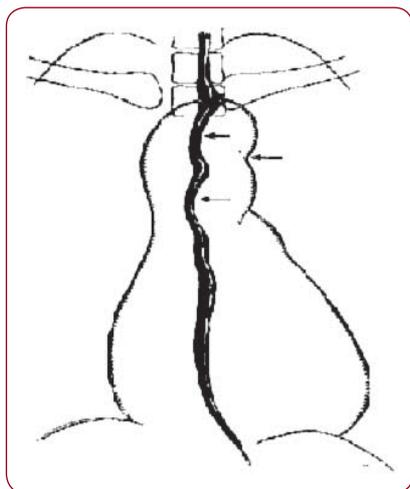


FIGURE 3. Compression of the esophagus with dilatation of the pre- and poststenotic segment (2)

TREATMENT

A. Medical therapy

The medical therapy is necessary for reduction of high blood pressure in the arms.

Preoperative hypertension can be treated using beta-blockers (propranolol 1-3 mg/kg/day, atenolol 1-3 mg/kg/day). Postoperative hypertension can be treated using beta-blockers and vasodilators (sodium nitroprusside). ACE inhibitors may be added if hypertension persists despite beta-blocker therapy.

B. Interventional therapy

This therapy consists of balloon angioplasty of the aortic isthmus, with or without stent implantation. The mechanism of dilatation consists of 1-3 mm tears of the tunica media, which heals in 8 weeks. This therapy is recommended in the mild forms of disease.

This therapy has the advantage of being a less invasive method.

The disadvantages are: increased rate of restenosis, possible hemorrhagic complications. The following are late results: recurrent stenosis of coarctation in the next 2-8 years, aortic aneurysm formation (0-50%) in 1-2 years after intervention, increased risk of thrombosis of the femoral artery. Paradoxical hypertension has not been encountered with this procedure (2,4).

C. Surgical therapy

The surgical therapy is recommended once the diagnosis of significant coarctation of the aorta with or without symptoms has been established (11). The preferred method depends on anatomy of the lesion and institutional experience. The techniques used more often are: resection and end-to-end anastomosis, left subclavian flap aortoplasty, patch aortoplasty, bypass graft repair (12).

The postoperative results depend on many parameters, the most important are: the patient age at the time of the surgery, the surgery technique and the presence or absence of associated anomalies. The early mortality rate in children is 1%. The 1-month survival rate is 98%, the 10-year survival rate is 91%, the 25-year survival rate is 81%. The mortality rate is lower following surgical repair compared with interventional therapy, but is higher than in general population (2).

Possible postoperative complications are: hemorrhages, postcoarctectomy syndrome, paradoxical hypertension, aortic aneurysm formation, recoarctation (13).

Percutaneous balloon angioplasty may be used in the treatment of recoarctation. Intravascular stent placement is used recently, with good results, although there are no long-term studies.

The following TABLES 1 and 2 present the studies results on the mortality and recoarctation rate. □

Author	Age	Year	Patients	Mortality	Recoarctation
Williams et al.	< 1 year	1980	176	66 (38%)	39 (33%)
Cobanoglu et al.	<3 months	1985	55	16 (29%)	3 (8%)
Korfer et al.	<4 months	1985	55	2 (4%)	3 (6%)
Ziemer et al.	<1 month	1986	24	8 (33%)	4 (25%)
Brouwer et al.	<2 years	1991	32	2 (6%)	4 (13%)
Kappetein et al.	<3 years	1994	48	5 (10%)	41 (86%)
Van Heurn et al.	<3 months	1994	42	5 (10%)	11 (30%)
Quaegebeur et al.	<1 month	1994	139	20 (14%)	6 (4%)
TOTAL			571	124 (21%)	111 (19%)

TABLE 1. Results of CoA resection with end-to-end anastomosis (after Mavroudis) (14).

Author	Age	Year	Patients	Mortality	Recoarctation
Metzdorff et al.	<2 months	1985	60	11 (18%)	10 (17%)
Ziemer et al.	< 1 month	1986	70	8 (11.4%)	9 (15%)
Ehrhardt; Walker	<1 month	1989	45	14 (31%)	7 (23%)
Milliken et al.	<1 month	1990	123	11 (9%)	20 (16%)
Van Heurn et al.	<3 months	1994	15	1 (7%)	6 (42%)
Quaegebeur et al.	<1 month	1994	112	9 (8%)	12 (12%)
Allen et al.	<3 months	2000	53	0	2 (4%)
Jahangiri et al.	<1 year	2000	185	6 (3%)	11 (6%)
TOTAL			663	60 (9%)	77 (12%)

TABLE 2. Results of subclavian flap aortoplasty (after Mavroudis) (14)

CONCLUSION

1. Coarctation of the aorta is a frequent congenital disorder which remains many times undiagnosed due to an incomplete clinical examination.
2. The treatment techniques and methods are well established. The surgical therapy is indicated at the moment of diagnosis. Balloon angioplasty is considered palliative and can be performed in the mild forms of disease or if restenosis occurs.
3. The main concern is the diagnosis. This can be most of the time a clinical diagnosis made by measuring arterial blood pressure in arms and legs, with hypertension, blood pressure difference of more than 20 mmHg in arms than in legs and absent or diminished pulse in legs. □

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