Therapeutics, Pharmacology and Clinical Toxicology
Vol XIII, Number 4, December 2009
Pages 409-413

THE TREATMENT OF COARCTATION OF THE AORTA IN THE NEWBORN WITH HEART FAILURE

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Abstract. Coarctation of the aorta is a congenital heart defect, 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 newborns and the treatment options.
Keywords: coarctation of the aorta, neonates, heart failure, treatment.

Introduction

Coarctation of the aorta 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.

Coarctation of the aorta accounts for 6-8% of all congenital heart defects or 50 to 100000 of all viable births. It is more common in males (M/F=2/1). It is more commonly associated with other congenital heart defects such as: patent ductus arteriosus, bicuspid aortic valve, ventricular septal defect and mitral valve anomalies. Coarctation of the aorta 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 two groups: the infantile type (preductal) and the adult type (postductal). (Figure 1, Figure 2). This classification concerns the time of patients' presentation and the clinical presentation in child and adult.

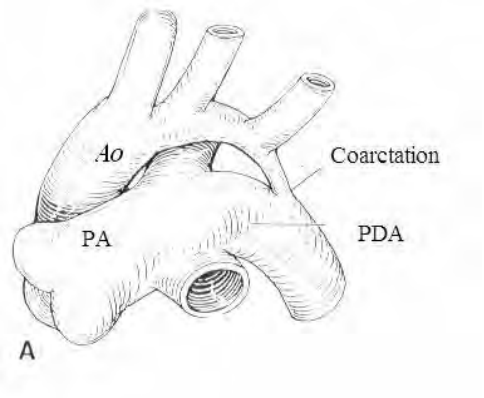

Figure 1. Coarctation of the aorta: the preductal type. The patent ductus arteriosus supplies most of the blood flow to the descending aorta. (after Mavroudis)[13].

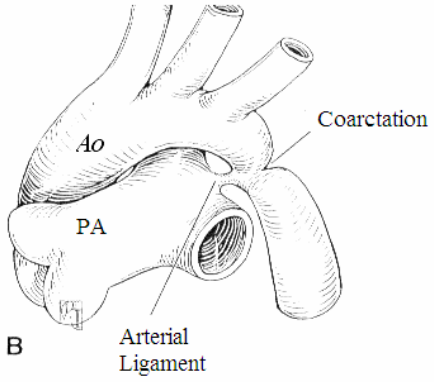

Figure 2. Coarctation of the aorta: the postductal type: The aria of narrowing is juxtaductal and consists of a posterior infolding localized in the lumen. The arterial duct is now closed (the arterial ligament) (after Mavroudis)[13].

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The surgical classification describes three groups: I. isolated; II. CoA associated with VSD; III. CoA associated with complex cardiac anomalies.

There are significant pathophysiologic differences between the adult with coarctation of the aorta and the child who depends on the patency of the arterial duct. The location, with regard to the arterial duct, establishes the pathophysiology, and the patency or closure of the arterial duct, changes in pulmonary vascular resistance, the presence of associated diseases and the expansion of collateral vessels. The more significant is the narrowing, the more ample are the collateral vessels.

In neonates (and children younger than 1 year of age): in the presence of arterial duct, the pulmonary vascular resistance maintains the systolic right-to-left shunt (ductal dependency of distal systemic circulation, right ventricle pressure overload); the reduction of pulmonary vascular resistance and/or the duct closure causes **poor perfusion of the lower body and inadequate kidney, liver, mesenteric perfusion (metabolic acidosis).**[2]

Neonates with severe coarctation and closed arterial duct present with signs of cardiac failure: tachypnea, cough, hepatomegaly, lower extremity edema, pulmonary rales, gallop rhythm. When treatment is lacking, the heart failure rapidly progresses, leading to acute pulmonary edema and consequent anuria. If associated anomalies are present, cyanosis can be observed as localized cyanosis in the lower body and normal color appearance in the upper body.

The diagnosis can be established on: blood pressure difference of more than 20 mmHg in arms than in legs, absent or diminished femoral pulse. Chest radiography may reveal cardiomegaly. Electrocardiography may reveal right ventricular hypertrophy in the first months of live. The 2-dimensional echocardiography and color Doppler may reveal lack of flow in the descending aorta, the anatomic location of coarctation, the size of the transversal aortic arch and other significant intracardiac anomalies. Cardiac catheterization helps evaluate the hemodynamic disturbance and the morphology of great vessels.[3,1]

Treatment

1. Treatment methods:

A. Isolated Coarctation

In isolated coarctation, surgery is indicated because the probability of survival and a normal blood pressure is greater postsurgery. Surgery is indicated when the diagnosis is established in neonates and infants with significant heart failure.

Resuscitation is initiated if coarctation is suspected in neonates with severe clinical presentation. This includes prostaglandin E1 (5 ng/kg/min) infusion, dopamine and diuretics. Ventilatory assistance is provided to patients with a more severe condition. Surgery is performed after heart failure stabilization.

Balloon angioplasty in the treatment of congenital coarctation is a controversial issue. There is a significant risk of coarctation recurrence and femoral vessels involvement, especially in neonates.

b. Coarctation of the aorta associated with VSD:

In this circumstance, patients undergoing repair of coarctation also undergo closing of VSD, using a single-stage repair. Another approach is to place a pulmonary artery band at the time of coarctation repair and VSD closure may be performed at a later time.

c. Coarctation of the aorta associated with major intracardiac anomalies:

In this circumstance, the decision on operating using a single-stage repair, two-stages repair or performing only the repair of coarctation is very difficult. The presence of a single ventricle, implies the decision on undergoing the repair of coarctation and the cardiac anomaly or performing the placement of pulmonary artery band only.[4,5]

2. Methods and means of treatment:

a. Medical therapy

The medical therapy is necessary for reduction of high blood pressure in the arms and for the control of heart failure symptomatology.

Diuretics and digoxin are used in neonates and infants with heart failure. Dopamine may be used in severe heart failure (for kidney function amelioration). Infusion of Prostaglandin E1 is used for opening the ductus arteriosus. (0,05-0,15 μg/Kg/min)

b. Interventional therapy:

This therapy consists of balloon angioplasty of the aortic isthmus, with or without stent implantation. This method is used in children younger than 1 year of age and in the less severe forms of disease. [6]

c. Surgical therapy:

This treatment is performed in neonates diagnosed with coarctation. The techniques used more often in the neonates are resection and end-to-end

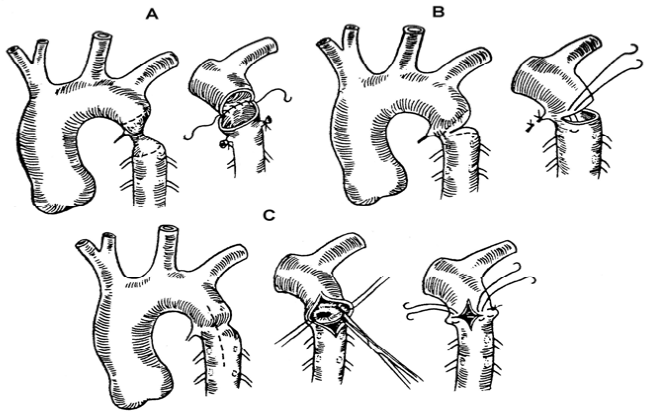

Figure 3. Resection and end-to-end anastomosis [15]

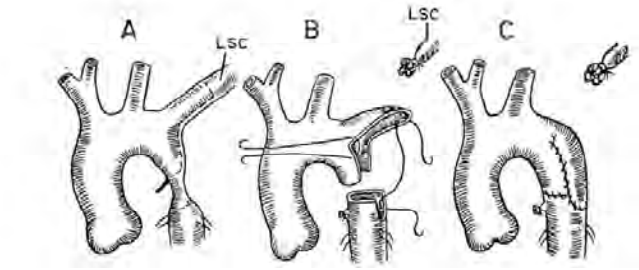

Figure 4. Left subclavian flap aortoplasty (Waldhausen method)[15]

anastomosis (Figure 3) and left subclavian flap aortoplasty (Figure 4).[7,8]

Excision of coarctation with restoration of continuity by end-to-end anastomosis.

Wedge excision with aortoplasty.

Aortoplasty and excision of membrane.

3. Comments:

Percutaneous balloon angioplasty has the advantage of being a less invasive method.

Disadvantages are: increased rate of restenosis, possible hemorrhagic complications. The following are late results: recurrent stenosis of coarctation in the following 2-8 years, aortic aneurysm formation (0-50%) in 1-2 years after intervention, increased risk of thrombosis of the femoral artery (because of the use of large-caliber angioplasty catheters; may respond well to the treatment with streptokinase). Paradoxical hypertension has not been encountered with this procedure.

Percutaneous balloon angioplasty can be used in the treatment of recoarctation because, in this case, surgery intervention is more difficult because of the presence of scar tissue and local adhesions. The stent placement is well tolerated and rarely leads to complications. Most of the stents can be redilated in 3 years after implantation, according to the growth stage.

The resection of coarctation with end-to-end anastomosis has the advantage of removing the ductal tissue entirely and the disadvantage of a laborious dissection and tension at the site of anastomosis.

Subclavian flap aortoplasty (Waldhausen method) consists of using the left subclavian artery (either by dissection or by reimplantation), to enlarge the area of coarctation. Disadvantages are: blood flow interruption to the upper left arm, residual ductal tissue, late postoperative aneurysm. [9]

The postoperative results depend on many parameters, the most important being: the patient's age at the time of surgery, the surgery technique and the presence or absence of associated anomalies. It is difficult to choose the „ideal" surgery technique. The early mortality rate in the neonate with or without patent ductus arteriosus is 2-10%.

Possible postoperative complications are: hemorrhages, post coarctectomy syndrome (sudden increase in blood pressure following 3-5 days post intervention), paradoxical hypertension, aortic aneurysm formation, recoarctation.[10]

The following tables 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%)
			571	124 (21%)	111 (19%)

Table I. Results of CoA resection with end-to-end anastomosis. (after Mavroudis)[7]

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%)
			663	60 (9%)	77 (12%)

Table II. Results of subclavian flap aortoplasty (after Mavroudis)[7]

Balloon Angioplasty compared with surgical intervention

Few randomized prospective studies compared the results of these two methods for the treatment of coarctation. Robert Shaddy and associates (in Salt Lake City) looked at 36 children with coarctation and found similar hemorrhagic complications rate and similar reintervention rate in both groups (20 patients underwent balloon angioplasty, 16 patients underwent surgical intervention). The risks of aneurysm formation and femoral artery thrombosis were higher in the balloon angioplasty group. Rao, 1994, compared the efficacy of balloon angioplasty with that of surgical intervention in infants older than 3 months. The author examined 29 infants between 1982-1992 (14 patients underwent surgical correction and 15 underwent balloon angioplasty). The author found lower morbidity rate and complications following balloon angioplasty than those seen with surgical therapy. The reintervention rate was similar in both groups. Paraplegia and paradoxical hypertension are very common following surgical repair and are rare following balloon angioplasty. The risk of aneurysm formation and the femoral artery thrombosis rate are higher following balloon angioplasty.

The risk of restenosis is higher in the newborns and infants in the first months of live who undergo balloon angioplasty, therefore this interventional procedure should be considered **palliative** and not a definite treatment. This procedure has been abandoned in newborns and infants, because of the high restenosis rate and femoral artery complications, excepting very ill patients in whom surgery has a higher risk. Other authors consider that restenosis lends itself to frequentative dilatation until the critical time disappears. The transductal or transumbilical approach represent alternatives in an attempt to avoid the use of femoral arteries. [11,12]

If we compare balloon angioplasty with surgical intervention, we can conclude that the mortality rates are similar (and these are probably related to the associated anomalies and not to the type of intervention the patients underwent), the morbidity rate and complications are lower following angioplasty.

Thus, until now, there is no well-defined algorithm treatment regarding the therapy of coarctation of the aorta in newborns and infants. The morphological features, the patient clinical status, the local institutional state, all these can influence the results of both procedures, especially the mortality and restenosis rate.

Many cardiologists prefer surgical intervention, whereas a few may opt for balloon angioplasty. Balloon angioplasty is useful in the treatment of

extremely ill neonates and infants with severe coarctation. Most surgeons prefer balloon angioplasty for the treatment of postsurgical recoarctations.[14]

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