Coarctation of the aorta

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Submitted: 11 February 2012
Accepted: 15 February 2012

Arch Med Sci 2012; 8, 1: 14-16
DOI: 10.5114/aoms.2012.27274

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Ding *et al.* report in the current number of the AMS [1] a 12 year old patient with abdominal coarctation of the aorta above the renal arteries treated successfully with percutanous transluminal angioplasty. This report is a useful reminder that hypertension maybe the sole manifestation of aortic coarctation that has an estimated incidence of 1 in 2500 live births [2] and may vary from a single constriction to a tubular hypoplasia of the aorta. The localization of the aortic constriction in this patient was in the abdominal aorta, above the renal arteries, and this variety or coarctation is rare (0.2-2.0%). Constriction of the aorta in this area is frequently associated, but not in this patient, with stenosis of renal, celiac or mesenteric arteries, causing the "middle-aortic dysplastic syndrome" or "middle-aortic syndrome" (MAS) that is a feature of several congenital and acquired clinical conditions [3]. The possibility of genetic factors is suggested by the report of the disease in monocygotic twins [4] and autosomal dominant inheritance in familial cases [5]. Gridlock mutations in the hey2 gen induce in the zebrafish changes similar to aortic coarctation that may be corrected with induced upregulation of vascular endothelial growth factor [6], but the relevance of these studies to human disease remain to be demonstrated. Other possible causes include the local accumulation of constricting fibrous tissue similar to that in arterial duct resulting from increased collagen gene expression induced locally by hemodynamic abnormalities in the distribution of the blood flow in the aorta [7-9].

Hypertension with absent pulses in the lower extremities was the clinical presentation of the patient described by Ding *et al.* [1] and the diagnosis was confirmed by computed tomography (CT) angiography. It is not unusual that patients are not diagnosed until adolescence or adulthood but it should be emphasized that an early diagnosis is important because the development of left ventricular hypertrophy and heart failure worsen the prognosis. Prenatal diagnosis of aortic coarctation is difficult and relays in serial fetal echocardiographic determinations of isthmal-ductal ratios [10] but neonatal diagnosis is possible and balloon dilatation has been used successfully in neonates weighing less than 2500 g with significant reduction of transcoarctation gradient [11]. However, restenosis is common and may develop rapidly. Repair of coarctation can be made by surgical correction (end to end anatomosis or subclavian flap repair to increase the size of the aorta) or by balloon angioplasty, with or without stent placement. In the patient reported by Ding *et al.* [1] the location and limited size of the constriction made it particularly amenable to angioplasty. Coarctation repair by balloon angioplasty has gained widespread acceptance

despite a significant incidence of restenosis. Pooled data from the literature reviewed by Rao and Chopra [12] indicate that angioplasty has less rate of complications than surgical repair with respect to initial (7% vs. 23%) and late (2% vs. 25%) mortality and restenosis (11% vs. 18%). A particular concern in subclavian flap repair is the aneurysm formation that, if uncorrected, will rupture within 15 years [13]. Reported results from experienced groups indicate that 5-9 years after balloon angioplasty repair of the aortic coarctation located in the aortic arch, restenosis after occurs in approximately 80% of of neonates, two-thirds of infants and less than 10% in older children [14].

In the patient reported by Ding *et al.* [1] normal blood pressure was maintained more than 4 years after the angioplasty but longer follow up is mandatory since hypertension may reappear many years afterwards. Recent reports indicate that only half of the patients are normotensive two decades after repair procedures [15]. The lack of long-term success in the correction of hypertension is incompletely understood and abnormal compliance of aortic baroreceptors may play an important role [16].

The increased morbidity and mortality associated with late diagnosis of aortic coarctation requires increased awareness of the disease and the routine determination of blood pressure in neonates, infants and children. Magnetic resonance imaging is the most cost-effective diagnostic procedure in infants and older patients [17] and should be combined with neurological imaging to detect cerebral aneurysms that are present in 10% of the children with aortic coarctation [18]. Repair of aortic constriction is required when the transcatheter systolic coarctation gradient (TSCG) is > 20 mmHg as it was the case in this patient. The American Heart Association has established guidelines for the use of angioplasty in pediatric patients with and without stent placement [19]. Balloon angioplasty of native coarctation may be reasonable in patients beyond 4 to 6 months of age when associated with a TSCG > 20 mmHg and suitable anatomy. It may also indicated in patients with complex coarctation anatomy or systemic conditions such as connective tissue disease or Turner syndrome but decision should be made on a case-by-case basis. The placement of stents that may be expanded to adult size should be given consideration if there is a long segment of coarctation as well as in patients in whom balloon angioplasty has failed.

It should be kept in mind that medical treatment is necessary in most patients before surgery and in many patients after surgery. The use of ramipril and atorvastatin reverses the impaired endothelial function and decreases the expression of proinflammatory cytokines and adhesion molecules in patients with aortic coarctation [20, 21]. A specific complication occurring sometimes after successful surgery repair is paradoxical hypertension. This complication is not due to activation of the renin angiotensin system and responds better to suppression of sympathetic activity with metoprolol [22].

The paper of Digh *et al.* [1] emphasizes the need to consider coarctation of abdominal aorta coarctation, a potentially curable condition, in the differential diagnosis of hypertension in young individuals.

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