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Editorial

Coarctation of the Aorta: A Generalized Arteriopathy Involving the Precoarctation Site

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Editorial

Coarctation of the Aorta (CoA) represents 5% - 8% of all congenital heart defects having an incidence of 4/1000 live births [1,2]. The stenosis is usually located opposite to the origin of the arterial duct, but it can be proximally or distally to its location. CoA can present as an isolated anomaly or co-exist with other congenital heart defects. Although CoA is usually a localized stenosis at the isthmus, it may present as tubular hypoplasia of the aortic arch, with several variations between the two extremes [2]. The most common extra cardiac anomaly of CoA patients is the development of berry aneurysms of the circle of Willis, which usually become manifest in the fourth to fifth decade of life [3,4].

CoA is a disease that often remains undiagnosed for many years without significant progress in timely diagnosis over time, despite the specific findings on physical examination and abnormal Blood Pressure (BP) values [5,6]. CoA was once considered a discrete narrowing of the isthmus that could be permanently cured after surgical excision of the stenosed region [1,2]. However, is has become widely acceptable that this condition may affect the aortic arch in a variable degree and that it represents a diffuse aortopathy of the vascular tree proximally to the stenosis, taking into consideration the abnormal histology of the arterial wall at this part of the aortic arch and the association of CoA with cardiovascular events later in life [1,2,7].

The aetiology of the regional arteriopathy in CoA has been attributed to the effects of the abnormal haemodynamic conditions due to the stenosis. Weather this represents a response to the decreased blood flow at the upper part of the body or the increased intraluminal pressure remains unknown [2]. Emerging evidence from morphological and molecular biological investigations suggest that the area of CoA is characterized by phenotypic modulation of smooth muscle cells, intimal thickening, and impaired elastic fiber formation. These changes extend to the pre-and post-stenotic aorta and impair arterial elasticity [7]. Indeed, adult patients after successful CoA repair have impaired endothelial function in the forearm circulation, increased intima/media thickness, decreased distensibility in the carotid arteries and increased levels of pro inflammatory cytokines and adhesion molecules than healthy controls. These results may partly explain the high incidence of coronary artery disease in patients with repaired CoA [8].

Also, normotensive repaired adult patients without recoarctation have aortic wall inflammation, as assessed by increased 18F-Fluoro Deoxy Glucose (FDG) uptake at Positron Emission Tomography/ Computed Tomographic imaging compared with age- and sex-matched controls. This finding suggests that despite successful CoA repair, haemodynamic burden early in life may have led to activation of pro inflammatory signaling in the aortic wall and sustained increased inflammatory cell activity [9].

Asymptomatic subjects are usually diagnosed due to upper limb hypertension and adult patients are less often identified due to rupture of berry aneurysms of the circle of Willis or left ventricular hypertrophy associated with heart failure [1,2]. If left untreated, according to the classic study of the natural history of CoA by Campbell M, patients who survive up to the first 2 years of life have 50% mortality at 30 years and 90 % at 58 years of life [10]. However, early and midterm outcome of treated patients is excellent and early mortality is less than 2%. Although early repair may delay the development of hypertension, about 30% of patients will be hypertensive in puberty [11]. Early surgical intervention decreases the incidence of hypertension; however it remains unknown if it just delays its development [11]. Yearly BP measurement at all four limbs and regular echocardiographic imaging of the heart and aortic arch are paramount [2]. Patient follow-up should also include 24 hour BP recordings and exercise testing, as exercise hypertension is a negative prognostic factor for development of hypertension in future and 1/3 are hypertensive on exercise (systolic >200 mmHg) [11,12]. The causes of hypertension are not clear and may involve dysfunction of the autonomic nervous system, arterial stiffness and upregulation of the renin-angiotensin system [11].

CoA has long been associated with aneurysmal disease which if left untreated is usually lethal. Repaired CoA is associated with aneurysms at the site of the previous intervention and in remote segments of the aorta and branch arteries [13]. Aneurysm formation in CoA has been attributed to genetic factors, persistence of ductal tissue in the wall, increased wall tension in hypertensive patients and weakening of the aortic wall by resection of the intimal ridge or compliance mismatch between the patch and the elastic aortic wall in operated CoA [13,14]. The co-existence of a bicuspid aortic valve which has been reported in 50% to 80% of patients with CoA increases the risk of cystic medial necrosis and aneurysm formation [1,2]. Aneurysms in CoA patients have also been reported in the left subclavian artery; innominate artery, intercostal artery, and abdominal aorta Therefore, patients merit periodic imaging studies with magnetic resonance imaging or computerized tomographic angiography to detect occult aneurysms [2].

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Although there has been significant progress in the management of CoA significant effort and research is needed to prevent chronic hypertension and thus minimize long-term morbidity [1,2]. Early intervention is crucial, as greater age at repair is associated with increased risk of hypertension, aneurysm formation and long term mortality. As CoA represents a generalized aortopathy, patients merit regular assessment and aggressive management in case of re-coarctation. Cardiovascular complications such aortic arterial and cerebral aneurysms, aortic root dilatation, premature atherosclerosis and coronary artery disease can become manifest decades after the initial repair [1,2]. These complications may occur despite effective and timely management and thus CoA patients require life-long regular follow-up.

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