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Mini Review

Aortic Disease: The Evolution and the Thoracic Aorta Surgical Treatment

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Introduction

Despite the advances in diagnostic methods and techniques for surgical treatment in the last two decades, aortic diseases remain a major cause of mortality and cardiovascular morbidity, challenging physicians and molecular biologists. It is believed that about 600 million years ago, during the Cambrian period, variant forms of life appeared, among them were the oxygen-producing cyano bacteria. The progressive enrichment of oxygen in the atmosphere continued during this period. Because most specimens were adapted to a preexisting atmosphere without oxygen, many have disappeared. New mutations, allowed the surviving specimens to adapt to aerobic forms of life [1].

Vertebrates have been successful in the colonization of all ecological niches in this journey. Their extraordinary ability to survive was related to the development of a system that allowed them to maximize their cells access to oxygen. An elastic tissue was essential in developing lungs and cardiovascular system to capture and carry oxygen to the most remote cells of beings with increasing complexity. Some authors consider a coincidence that the elastin gene arose during this period. Elastin, in evolutionary terms, is much newer than most of the collagens. The phyla genetic development of elastin has been crucial for vertebrates to develop efficient respiratory and circulatory systems. However, these highly specialized systems suffer malfunctioning related to the aging process by gene determination and mutations, causing severe diseases in humans. It seems that creation of new genes is related to a mechanism to empower each individual species for reproductive success and not to prolong life. Aortic diseases contribute to the wide spectrum of arterial diseases including aortic aneurysms, acute aortic syndromes, atherosclerotic, inflammatory affections, genetic diseases, like Marfan, Loyes-Dietz Syndrome and others, and also congenital abnormalities such as aortopathy in bicuspid aortic valve disease and coarctation of the aorta. In healthy adults, thoracic aortic diameter does not exceed 40mm and tapers gradually downstream. It is variably influenced by different factors including age, gender body surface area and blood pressure. In this regard, aortic expansion is about 0.9mm in men and 0.7mm in women for each decade of life [2].

The middle layer of the aorta in humans is made up of four basic elements: elastic fibers, collagen fibers, smooth muscle cells and amorphous substance, arranged in lamellar units. Each unit consists of two lamellar elastic fibers parallel with smooth muscle cells in between. This basic pattern is present throughout the length of the vessel, although there are quantitative and qualitative differences between the thoracic and abdominal segments [3]. The middle layer of elastic arteries has an important role in maintaining the architecture of the vascular wall in response to deformation caused by the pulse wave determined by cardiac systole. It is also important to note that humans lose the ability to produce elastic fiber early in life, and short synthesis of elastin can be detected in the aortic wall after infancy. Moreover, the number of lamellar units is constant in the aorta of mammals, except humans [4].

Elective surgical treatment of the thoracic aorta is highly recommended to the detriment of emergency treatment, with five-year survival after elective surgery of 85%, while successful emergency operations survival in the same period is only 37% after a rupture or dissection. Evidences show that genetic mutation in a single gene may predispose individuals to aneurysms and dissections of the thoracic aorta and its branches. New protocols propose image diagnosis management and ideal timing for a precocious operation, avoiding emergence procedures with higher morbidity and mortality. Genetic mutations may be syndrome, with phenotypic identification, or may be of a familial nature and, in such cases; the familial occurrence of genetic mutation must be deepened through family history, imaging scans and, recently, genetic markers. The best recognized genetic mutation is the Marfan syndrome, dominant auto somal characteristic, a mutation of the FBN-1 gene. This gene is responsible for the production of fibrillin-1 protein, which is a component of the micro fibrils that make up the fibrous skeleton of the heart and the elastic tissue of the tunica media of the thoracic aorta. These patients rarely present intracranial aneurysms [5].

Recently recognized, the Loyes-Dietz syndrome is associated with the mutation of genes TGFBR1 and TGFBR2, also dominant autosomal character, leading to the loss of function of the TGF-ß

(transforming growth factor ß) signaling activity in the maintenance of the contractile function of smooth muscle cells of the tunica media. Especially when it comes to the TGFBR2 mutation, some authors recommend surgery of the ascending aorta with minimal dilations, around 42 mm in adults, due to the high risk of dissection and rupture. However, aneurysms and dissections may affect the thoracic aorta and other vessels, including intracranial vessels [6].

Also in the category of family disease, mutation of the SMAD3 gene, called aneurysm-osteoarthritis, affects different vessels in addition to the thoracic aorta, the abdominal segment, iliac arteries and intracranial branches, having osteoarthritis as an early symptom. Patients with this syndrome should undergo imaging tests for the entire vascular bed, including the intracranial vessels [7]. Finally, bicuspid aortic valve disease (BAV), with a prevalence of 1-2% in the general population, which results from the fusion of the valve leaflets during valvulo genesis is associated to coarctation of the aorta, spontaneous dissection of the carotid arteries, and also associated with incompletely characterized aortopathy leading to aneurysm and dissection. The fragility of the thoracic aorta seems to be associated with morphological variation of the aortic valve and even individuals with normally functioning BAV may have aortic disease. Molecular analysis has demonstrated fibrillin-1 deficiency of the great vessels in these patients. Deficient fibrillin-1 content in the vasculature of patients with BAV might trigger matrix metalloproteinase production, leading to matrix disruption and dilatation, especially in young males with aortic regurgitation and root dilation [8].

Recent publication has suggested three distinct phenotypes of BAV associated aortopathy. Patient characteristics and also valvular dysfunction vary by phenotype, suggesting that the location of aortic pathology could be related to the underlying patho physiology of this disease. It is indispensable that experts from different areas came together to guide surgeons to treat aortic catastrophes and prolong human life. After all, nature evolution happens too slowly [9].

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