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ISSN: 2329-6607
Cardiovascular Pharmacology: Open Access

Opinion Article

Advancements and Outcomes in the Therapeutic Management of Aortic Root Aneurysm in Marfan Syndrome

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DESCRIPTION

A hereditary condition called Marfan syndrome damages the body's connective tissue. The emergence of aortic root aneurysms, a disease that can be fatal if not appropriately handled, is one of the most severe consequences linked to Marfan syndrome. The causes, signs, symptoms, diagnosis, and available treatments for aortic root aneurysms and Marfan syndrome will all be covered in this article. Genetic change Fibrillin-1, a protein necessary for the formation of the connective tissue in the body, is encoded by the FBN1 gene, which is where Marfan syndrome is brought on by a mutation [1]. The mutation causes abnormalities of connective tissue throughout the body, affecting the heart, blood vessels, joints, and other organs. Marfan syndrome can appear differently in different people, with some showing very moderate symptoms and others having more serious health issues [2]. The principal artery connecting the heart's supply of oxygen-rich blood to the body's tissues is the aorta. The portion of the aorta that links to the left ventricle of the heart closest to the heart is called the aortic root. Marfan syndrome can cause the aortic root to expand or dilate, which can result in an aneurysm [3]. The possibility of aortic dissection, a potentially fatal disorder in which the layers of the aorta wall separate, makes this condition extremely dangerous. In those with Marfan syndrome, a genetic connective tissue mutation is the main contributor of aortic root aneurysms. The aorta wall's structural integrity is compromised by this mutation, which increases the risk of aneurysm development and aortic dilatation. The continuous flow of blood from the heart into the aorta might erode the already vulnerable aortic wall. People with Marfan syndrome who are older are more likely to develop and grow aortic root aneurysms [4]. In Marfan syndrome, aortic root aneurysms are frequently asymptomatic until they grow to a dangerous size or cause problems [5]. An aortic dissection, a medical emergency, can cause quick, sharp chest pain that is frequently characterised as acute and ripping. The area between the shoulder blades may experience radiating pain. If the aneurysm compresses surrounding tissues or if it proceeds to aortic valve failure, breathing problems may develop [6].

Auscultation may reveal a cardiac murmur because of aberrant blood flow via the dilated aortic root. The aortic root, its size, and any aneurysms can be seen with diagnostic methods including echocardiogram, Computed Tomography (CT) scans, and Magnetic Resonance Imaging (MRI) [7]. It is important to confirm the existence of Marfan syndrome by genetic testing since it helps direct management and treatment. The goal of treating aortic root aneurysms in people with Marfan syndrome is to avoid consequences including aortic rupture and dissection. To lower blood pressure and lessen the strain on the aorta wall, doctors frequently give beta-blockers and Angiotensin Receptor Blockers (ARBs) [8]. The aortic root may need to be repaired or replaced surgically if it reaches a specific size or if there is evidence of dissection [9]. The Bentall method or valve-sparing root replacement are two alternatives. A healthcare team with expertise in cardiovascular care must regularly examine people with Marfan syndrome and aortic root aneurysms to gauge the aneurysm's growth and general heart health [10]. With improvements in treatment and surgery, the outlook for people with Marfan syndrome and aortic root aneurysms has dramatically improved. To properly manage the illness and avoid potentially fatal consequences, early diagnosis, continuous monitoring, and prompt action are essential. Aortic root aneurysms and Marfan syndrome are strongly related, with the genetic mutation that causes Marfan syndrome predisposing people to the development of aortic root dilatation and aneurysms. For people with Marfan syndrome to obtain the right care and treatment, it is important that they are aware of the symptoms, have frequent checkups, and undergo genetic testing. Medical research advances assure improved outcomes and a higher quality of life for people affected by this complex genetic disease, even if treating the illness requires determination.

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Received: 04-Sep-2023, Manuscript No. CPO-23-23448; **Editor assigned:** 06-Sep-2023, Pre QC No. CPO-23-23448(PQ); **Reviewed:** 20-Sep-2023, QC No. CPO-23-23448; **Revised:** 27-Sep-2023, Manuscript No. CPO-23-23448(R); **Published:** 04-Oct-2023, DOI: 10.35248/2329-6607.23.12.368

Citation: Kodogo K (2023) Advancements and Outcomes in the Therapeutic Management of Aortic Root Aneurysm in Marfan Syndrome. Cardiovasc Pharm. 12:368.

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