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Case Report
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Personalized External Aortic Root Support in Loeys Dietz Syndrome: Report of an Aortic Dissection at the Origin of the Right Coronary Artery

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Abstract

Personalized external aortic root support is a novel therapy designed to wrap the aortic root externally to prevent progressive aneurysmal dilation in patients with connective tissue disorders such as Marfan syndrome. Histological investigation has revealed that it becomes integrated into the aortic adventitia thus providing additional strength. This procedure has been performed in 325 patients worldwide. Our centers established a PEARS program in 2016 and have undertaken 14 cases to date. We report an iatrogenic aortic dissection at the right coronary ostium in case 9 in our series in a patient with Elhers Danlos. We herein report this case to assist other centers considering PEARS and to disclose the important learning points encountered through our experience.

Introduction

Personalized External Aortic Root Support (PEARS) is a novel technique designed to prevent aneurysmal dilation and subsequent dissection of the root and ascending aorta in patients with connective tissue disorders. A 3D printed model of the patients' aortic root is created from Computed Tomography (CT) images and an individualized polyethylene terephthalate mesh sleeve constructed to intimately fit the patient's aortic contours [1]. The size can be individually specified and as a default, 100% and 95% stents are supplied for each patient. Performed *via* a median sternotomy, the PEARS is placed around the aortic root and ascending aorta after off-pump dissection of the aortic root down to the valve annulus and underneath the coronary arteries with two defects to accommodate the coronary ostia and preserve flow [2]. It is reconstituted by a combination of continuous and interrupted sutures with typically 4/0 Ethibond (Ethicon). Animal studies suggest the mesh subsequently incorporates into the aortic wall to both thicken it and prevent any further aneurysmal change [3]. Compared with the traditional treatment; aortic root replacement, proposed benefits of PEARS include avoidance of lifelong anticoagulation in the case of a mechanical aortic valve implantation or fear of failure with valve conserving surgery [4]. The majority of PEARS to date have been carried out on patients with Marfan syndrome and a paucity of data regarding other connective tissue disorders exists [2].

Case Presentation

A 34-year-oldfemale with Loeys Dietz Syndrome (LDS) SMAD3 variant was referred for PEARS. She had a family history of sudden death in two young relatives two generations removed. Her daughter was found to carry the same genetic variant. Clinical examination demonstrated tall stature, bifid uvula, arachnodactyly and skin striae. Cardiac gated Computed Tomography (CT) of the aorta demonstrated aortic root dilation, 4.5 cm with normal coronary anatomy and a right dominant circulation (Figure 1). The patient was counseled regarding operative options including valve sparing root replacement, a Bentall and a PEARS and chose to undergo PEARS. The procedure was performed via using the 100% pears graft. Sternal closure resulted in ST changes in the inferior leads and hypotension. The sternum was reopened and a series of radial cuts in the Right Coronary Artery (RCA) orifice made to ensure no compression was occurring. A small bleb-like hematoma was noted to arise from the anterior aspect of the proximal RCA at this time. The patient entered ventricular fibrillation and cardiac arrest requiring cardioversion three times then stabilized. Closure was achieved and she was transferred to intensive care in a stable condition. CT on day one post-surgery demonstrated a small pseudoaneurysm close to the ostium of the RCA (Figure 2). The patient was recovering well and planned for discharge home when a further CT was

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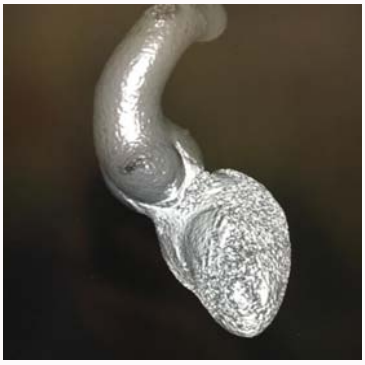

Figure 1: CT thoracic aorta pre-operatively demonstrating isolated aortic root dilation measuring 4.8 cm.

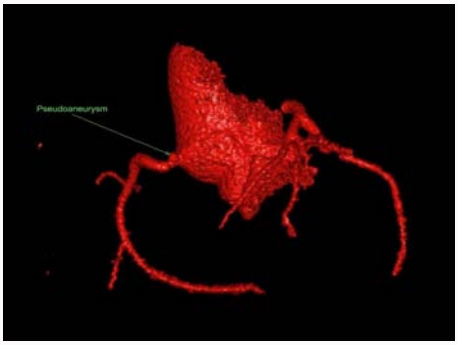

Figure 2: Surface rendered Cardiac CT day 1 post-surgery demonstrating a right coronary artery pseudoaneurysm. Left main stem is widely patent.

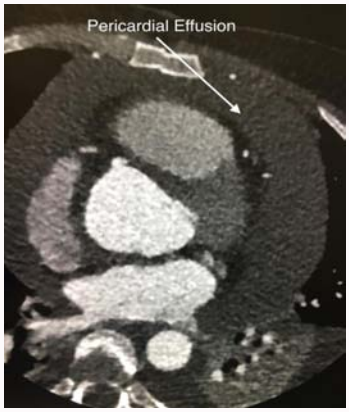

Figure 3: Significant pericardial effusion, small pseudoaneurysm at the origin of the RCA, moderate narrowing of the ostium is shown and represents and increases in size.

performed on day six which demonstrated a significant pericardial effusion and pseudoaneurysm of the right coronary measuring 3 cm × 3 cm (Figure 3).

Urgent reoperation was undertaken. A large amount of blood was drained from the pericardial cavity. Cardiopulmonary bypass was instituted and following cross-clamp and arrest a localized dissection identified at the site of concern on the RCA (Figure 4). The RCA was divided from the aorta and a pericardial patch applied with double layer 4-0 and 5-0 proline sutures used to cover the ostial defect. A section of the unused 95% PEARS graft was then taken to patch the defect in the original PEARS graft. Right Internal Mammary Artery

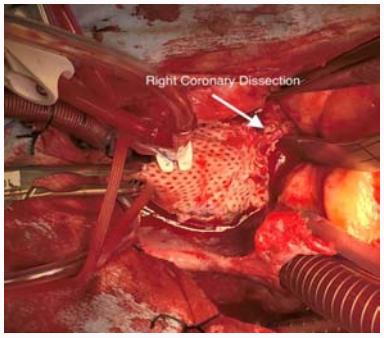

Figure 4: Ascending aorta at the time of emergency reoperation, the PEARS graft is shown and the disrupted RCA at the level of the ostium is shown.

(RIMA) was harvested and anastomosed to the mid RCA using 7-0 proline. The patient weaned from bypass without difficulty and made an uncomplicated recovery. She represented three weeks post-surgery with chest wall pain and was investigated. Cardiac investigations were normal and a further CT confirmed the RIMA-RCA graft was patent. She remains well on most recent follow up at one year.

Discussion

PEARS are a novel procedure and experience in patients with connective tissue diseases affecting the aortic root other than Marfan syndrome is limited. One previously documented PEARS case had a ventricular fibrillation arrest in intensive care and the emergency team released the closing sutures thus allowing restoration of output [2]. Coronary complications are recognized with PEARS but this case is the first reported localized aortic dissection. As a result, there is a number of learning points the authors feel are of value.

Firstly, the case confirms existing knowledge regarding tissue integrity in LDS is fragile and represents an aggressive and problematic issue peri-operatively [5]. LDS is an autosomal dominant connective tissue disorder characterized by aortic aneurysm, arterial tortuosity, hypertelorism and bifid uvula/cleft palate. The TGF-β super family mediates normal cellular growth and development. Derangement in LDS results in elastin disarray, loss of elastic fiber architecture and increased collagen expression in the arterial wall [6]. Significant consideration was given at the time of surgery to suture repair of the small hematoma arising from the right coronary artery. On balance it was felt this would likely result in further problems as there was no active bleeding from the site at that time. A case report describing a right coronary artery dehiscence and resultant pseudo-aneurysm arising in a patient with two successive aortic root replacements first for root enlargement and then for left coronary pseudo-aneurysm and dehiscence has been described [7]. This confirms the tissue fragility and proclivity for dissection and aneurysmal change in LDS. We made every reasonable effort to avoid applying the aortic cross clamp but ultimately felt this was unavoidable and did so with a larger broader clamp than usual. We elected to carry out a RIMA to RCA graft with closure of the RCA ostium by patching the aorta with autologous pericardium supported by a patch of PEARS as it was not felt possible to preserve the native vessel due to the degree of damage to the coronary arterial wall.

Authors suggest that the index of suspicion of coronary issues must be high in patients with connective tissue disorders undergoing PEARS especially in the setting of cardiac arrhythmia post procedure. The decision to undertake the second CT scan was based purely on

suspicion as opposed to any clear clinical indication. These patients are typically young and will have significant capacity for hemodynamic compensation in the context of a significant pericardial effusion. The patient was preparing for discharge and had in no way demonstrated clinical symptoms resultant from her pericardial hematoma and coronary disruption. There are currently no guideline documents pertaining to follow up imaging in the PEARS cohort, which is an issue that should be addressed going forward.

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