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A Loeys-Dietz Patient with a Trans-Atlantic Odyssey
Repeated Aortic Root Surgery ending with a Huge Left Main Coronary Aneurysm

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21 **Key words:** Loeys-Dietz syndrome – Aortic root replacement – Left main coronary
22 aneurysm – Graft infection

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24 **Summary**

25 We present a patient who required several aortic root procedures. A huge aneurysm of
26 the left main coronary artery required emergency surgery after failed transcatheter
27 procedure.

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30 **Introduction**

31 Repeated aortic root surgery may be a challenging procedure. In case of prosthetic
32 material infection, complete resection of the foreign material is mandatory in addition to
33 adequate antibiotic treatment. Aortic homograft is a good option to control hemostasis
34 and the infectious process. This Loeys-Dietz patient presented with a pseudoaneurysm
35 of the left main following multiple root procedures.

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38 The 25 year-old patient presented initially with aortic root aneurysm. Because of a very
39 asymmetric pseudobicuspid aortic valve (Sievers type I, subcategory L-R) and the strong
40 wish of the parents to avoid anticoagulation, he received a tissue valved conduit in Bern
41 in 2003 (Shelhigh Inc. NJ, USA – presently BioIntegral, Canada). Recovery was
42 uneventful but echocardiography at 6 months surprisingly demonstrated a limited
43 dehiscence at the proximal anastomosis. Re-exploration showed that the suture line was
44 not healed. Infection was excluded in presence of normal clinical and laboratory findings
45 and unsuspecting intraoperative inspection. A pericardial patch was used to reinforce
46 the suture. Evolution was favourable.

47 In 2008, he presented with a degenerated tissue valve requiring reoperation. The
48 procedure was performed at Johns Hopkins University. The bio-conduit was resected
49 and a mechanical composite graft implantated. The day after, the patient underwent
50 repair of pectus excavatum. Recovery was uneventful.

51 Yearly echocardiographies and/or MRI were normal. In 2014, he developed fever and
52 pneumonia was diagnosed. Blood cultures remained negative but staphylococcus was
53 revealed on PCR. Several weeks later, echocardiography and MRI showed a partially
54 thrombosed aneurysm of the left main coronary artery (Figure 1). PET-scan was not
55 conclusive. The parents were against a 4th operative procedure and asked several
56 opinion-leaders for a transcatheter technique. A covered stent was recommended to
57 exclude the aneurysm. However, stable positioning of the stent between the ostium and
58 the distal left main was not successful and obstruction of the left main occurred
59 requiring resuscitation. Percutaneous ECMO was installed. The patient was immediately
60 moved to the operation theater. Echocardiography showed a LV-EF of 10-15%).

61 Re-sternotomy was performed and ECMO switched to cardiopulmonary bypass through
62 central cannulation. Intraoperative inspection showed thrombotic material inside the
63 graft. To expose the aneurysm, pulmonary artery was transected above the valve,
64 thrombotic material was removed. Continuity of the left main was restored with a short
65 vein graft. Aortic homograft was implanted in the mini-root technique and pulmonary
66 artery re-anastomosed during reperfusion. ECMO support was necessary during 48
67 hours. The patient was discharged after 5 weeks with compromised LV-function (EF 25-
68 30%).

69 Antibiotics were continued for 6 weeks although intraoperative cultures remained
70 negative. 18 months postoperatively, LV-EF has stabilized at 40%. The patient is doing
71 well and findings are normal (Figure 2).

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75 **Comment**

76 Repeated aortic root surgery is a surgical challenge (1). Aneurysm of the left main
77 coronary artery is infrequent but may occur in patients with connective tissue disease or
78 as a result of infection.

79 This patient's history presents several critical decision-making points:

- 80 - Initial biological composite graft was a wrong decision. A mechanical composite-
81 graft would have been the best long-term option since in 2003, a valve sparing
82 procedure was not an established standard in young patients with connective
83 tissue disease and pseudobicuspid valve. The parents were strictly against
84 anticoagulation.
- 85 - Dehiscence of the suture observed early postoperatively is a problem that we and
86 other have described following implantation of this conduit (2,3).
- 87 - Etiology of the coronary aneurysm was uncertain. PET-scan was not conclusive
88 and intraoperative cultures remained negative.
- 89 - Transcatheter attempt to exclude the left main aneurysm was not a good decision.
90 Sudden coronary obstruction lead to resuscitation and required immediate ECMO
91 support.
- 92 - Transection of the main pulmonary artery greatly facilitated exposure of the left
93 main.

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97 Surgery was challenging because re-sternotomy was performed under resuscitation and
98 after repaired pectus excavatum. Aortic homograft was found the best option to control
99 intraoperative hemostasis (1). Re-attachment of the coronary arteries required short
100 vein graft interposition to avoid tension and exclude the aneurysm.

101 We conclude that indication for surgery should never be weakened because
102 transcatheter intervention seems theoretically possible. In this case, catheter
103 intervention turned the repair into an emergency situation and compromised
104 significantly the myocardial function. In addition, decision-making was challenged by
105 the opinion of non-expert relatives instead of a decision based on experience and
106 expertise on technical adequacy.

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124 **Figures**

125 **Figure 1**

126 A. (Pseudo)-aneurysm of the left main artery on conventional CT-scan (A) and 3D
127 reconstruction (B)

128 **Figure 2**

129 Postoperative 3-D reconstruction of the aortic homograft with separate interposition of
130 two short vein grafts for the left and right coronary arteries