Total arch replacement procedure in a child with Loeys–Dietz syndrome

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I read with great interest this article describing a total aortic root and arch replacement in a patient with Loeys–Dietz syndrome (LDS) [1]. There is no doubt that this case was a monumental effort and a tribute to their cardiac surgical programme.

There is already a sufficient body of literature validating valve-sparing root replacements in children with connective tissue disorders [2–4], so the value of the article lies in their description of the arch replacement. This raises two related questions regarding the arch repair: the indication for total arch repair and the technique of arch repair.

The available literature reports very few cases of arch repair for patients with LDS [5, 6]. This implies that aneurysmal involvement of the arch is rare or is a later part of the nature history not yet described. The aortic arch in this case is minimal-ly involved on the superior aspect, leaving the possibility that more conventional resection of the underneath side of the arch with the ascending aortic graft bevelled with a single anastomosis to the arch would have been sufficient. It remains unknown if this more straightforward approach would have been sufficient.

The approach used by the authors has a clear advantage of complete resection of all aortic arch tissue at the expense of a more extensive procedure (270 min of cardiopulmonary bypass) and preventing any growth potential of the arch. Flow to the distal arch is limited by a 10 mm diameter section of Dacron graft. As our experience with LDS grows, the indications for complete resection of the aortic arch, as described in this case report, will hopefully be clarified.

REFERENCES


