

Cardiac Imaging

Reoperation for Ascending Aorta and Total Arch Replacement Combined with Patent *Ductus Arteriosus* Closure in a Child with Marfan Syndrome

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A 14-year-old girl diagnosed with Marfan syndrome was operated on at the age of 4 for severe aortic regurgitation and ascending aortic aneurysm. The operation consisted of a composite valve graft (Carbomedics 25 mm) and reimplantation of the coronary arteries (Bentall procedure). Reoperation was required ten years later because of progression of the dilatation of the native ascending aorta. Magnetic resonance imaging (MRI) of the thoracic aorta revealed a maximum diameter of 50 mm just distal to the composite valve graft, 46 mm at the brachiocephalic artery and 33 mm at the left common carotid artery. It also showed a 29 mm long patent *ductus arteriosus* (PDA) with a diameter reaching 3 mm at the pulmonary and 35 mm at the aortic end (Figures 1A, 2A). The transverse slice of the reimplanted right coronary artery (RCA) in a “time of flight” MR angiography showed severe calcification of the composite valve graft (a very rare finding in MRI, especially at this age) extending to the dilated ostium of the RCA, which reached a diameter of 7 mm (Figure 1B). The reoperation was performed under hypothermic circulatory arrest and intermittent antegrade cerebral perfusion. First, the PDA was recessed and ligated at its pulmonary end. Then, the brachiocephalic vessels were removed *en bloc* from the aortic arch and reattached to a prosthetic tube graft that was

sewn directly to the descending aorta and closed blindly at its proximal end. This 24 mm wide prosthetic graft was anastomosed end-to-side with a second prosthetic graft, which was subsequently attached directly to the composite valve graft (Figure 2B). The patient recovered uneventfully with no neurological deficits and is in stable clinical condition.

The most common cardiovascular features in Marfan syndrome are mitral valve prolapse and dilatation of the sinuses of Valsalva with subsequent mitral and aortic valve regurgitation.^{1,2} In a recent report from LeMaire et al,³ in 300 patients undergoing cardiovascular surgery for confirmed or suspected Marfan syndrome no case of concomitant repair for congenital cardiac defects was reported. To our knowledge this is the first report of a combined staged thoracic aorta repair combined with closure of a PDA in a child operated on for Marfan syndrome.

References

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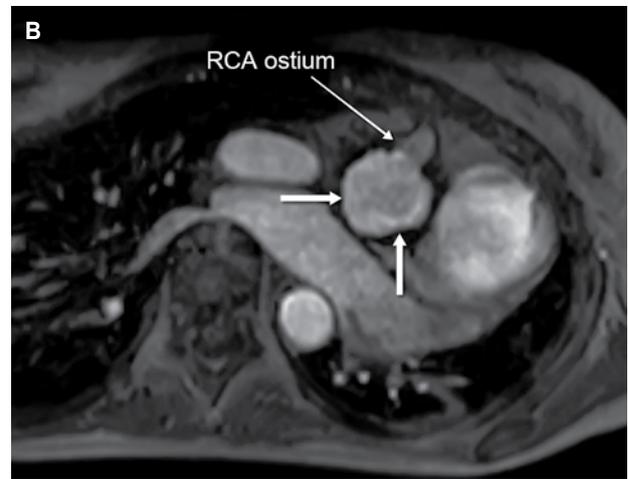
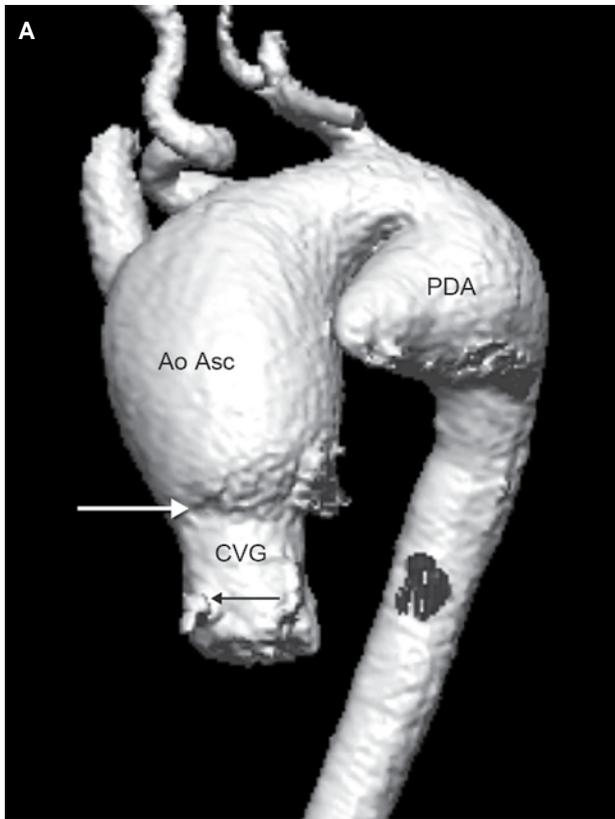


Figure 1. A. contrast medium-enhanced three-dimensional magnetic resonance angiogram showing the patent *ductus arteriosus* (PDA) and the native ascending aorta (Ao Asc) aneurysm adjacent to the composite valve graft (CVG) (anastomosis site, white arrow; reimplanted ostium of the right coronary artery, black arrow); B. “time of flight” magnetic resonance angiography shows the severe calcification of the CVG (thick arrows) extending into the dilated, calcified ostium of the right coronary artery (RCA), which reaches a diameter of 7 mm (thin arrow).

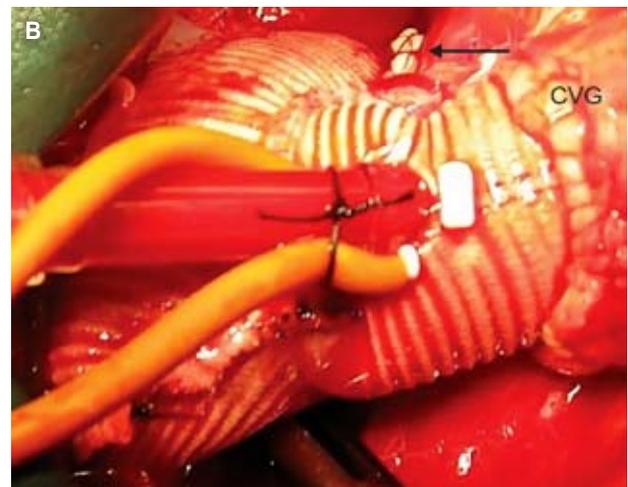
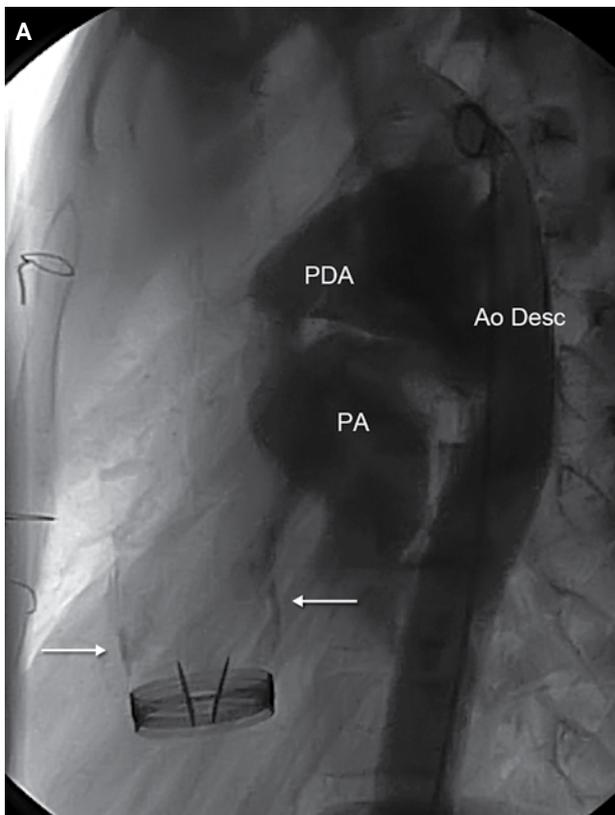


Figure 2. A: aortogram shows the patent *ductus arteriosus* (PDA) and the severely calcified composite valve graft (white arrows). Ao Desc – descending aorta; PA – pulmonary artery; B: operative site, revealing the final result with the oversewn pulmonic end of the PDA (black arrow) and the two prosthetic tube grafts used for replacement of the aortic arch and ascending aorta sutured with the composite valve graft (CVG).