

Acute Type A Dissection Repair in an Achondroplastic Dwarf

Anesthetic, Perfusion, and Surgical Concerns

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Abstract

In this report we present a 43-year-old male with achondroplastic dwarfism who presented with acute Type A aortic dissection with aortic insufficiency. The patient underwent successful Bentall and hemiarch repair. Anesthetic, perfusion-related, and surgical planning and execution are presented.

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Key Words

Dwarfism · Dissection

Introduction

Achondroplasia is the most common type of chondrodysplasia with an incidence of 1 in 25,000 to 40,000 births [1]. It occurs due to a mutation in fibroblast growth factor receptor 3 (FGFR3). To our knowledge, there is no association between achondroplastic dwarfism and aortic dissection.

To the best of our knowledge, this case represents the fifth case report of open heart surgery in a dwarf patient and the first one of aortic dissection repair [2–5]. Informed consent was obtained from the patient for publication of this case report with the accompanying images.

Case Presentation

A 43-year-old man with achondroplastic dwarfism and known hypertension presented with severe chest

pain migrating to his back. Computed tomography angiography (CTA) showed acute Type A aortic dissection, and the patient was brought to our institution for surgical management.

Past medical history was significant for achondroplastic dwarfism [height, 103 cm; weight, 37 kg; body surface area (BSA), 1.03 m²], hypertension, smoking, and chronic obstructive pulmonary disease. Specific complications of achondroplasia included thoracic kyphosis, spinal stenosis, and a surgically fused cervical spine.

Chest X-ray showed a wide mediastinum (Fig. 1) and CTA demonstrated an aortic root dissection flap that extended through the ascending aorta and aortic arch to the iliac bifurcation (Fig. 2A and 2B). The great vessels of the aortic arch communicated with the true lumen, as did the major abdominal branches.

Intraoperative Management

The patient was taken to the operating room for a Bentall procedure and replacement of the ascending aorta and hemiarch. General anesthesia was induced and intubation was performed by video laryngoscopy. Right internal jugular vein and left femoral artery lines were placed with ultrasound guidance. Intraoperative transesophageal echocardiogram (TEE) demonstrated a dissection flap in a dilated (52 mm) aortic root with dissection extending into the descending thoracic



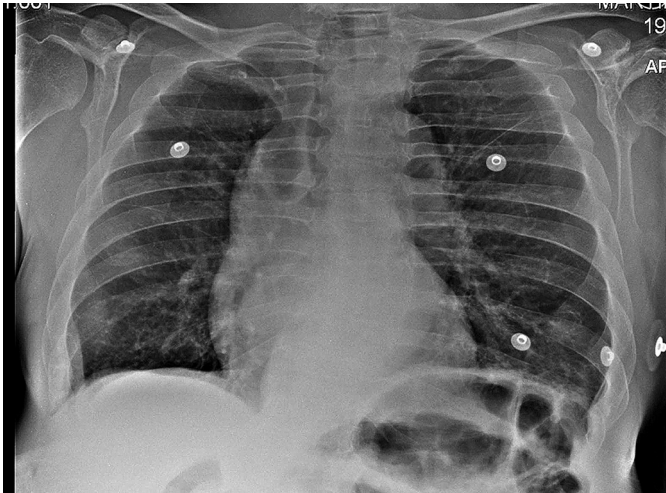


Figure 1. Preoperative chest X-ray.

aorta (Fig. 3). There was severe aortic insufficiency due to a failure of central coaptation.

A 6-mm graft was anastomosed to the right axillary artery using a subclavicular approach. Median sternotomy was performed, the patient was heparinized, and arterial and venous cannulation (28/30F, Medtronic, Minneapolis, MN, USA) were established. We utilized a standard-sized cardiopulmonary bypass (CPB) circuit. Retro-priming was used to remove 400 cc of prime volume. Flow rates maintained a cardiac index of 1.8-2.5 L/min/m². Hemo-concentration was performed throughout the procedure.

Cooling was commenced to a target temperature of 18°C. At this point, we were not sure whether we would be able to use antegrade selective cerebral perfusion (ASCP) due to the patient's small-caliber vessels. The aorta was cross-clamped and 1 L of cold retrograde cardioplegic solution was administered. The aortic aneurysm was opened and the dissection flap was identified at the right coronary sinus. The aorta was resected, the coronary buttons were detached, and the valve was excised. A composite graft constructed from a 26-mm conduit and a 23-mm mechanical bileaflet valve was inserted using 13 sutures of 2-0 Tycron reinforced with pledgets. At 18°C, the innominate artery was clamped and circulatory arrest was initiated with ASCP (12-15 cc/kg/min; mean pressure 39 mm Hg). Once we established adequate ASCP, we started to rewarm the patient. The aorta was transected just proximal to the innominate artery and a 22-mm Dacron graft was anastomosed end-to-end. The nondissected aortic tissue in the aortic arch was

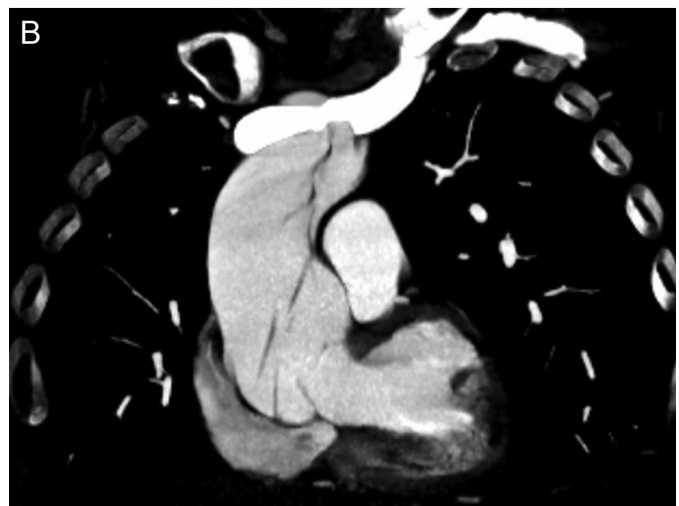
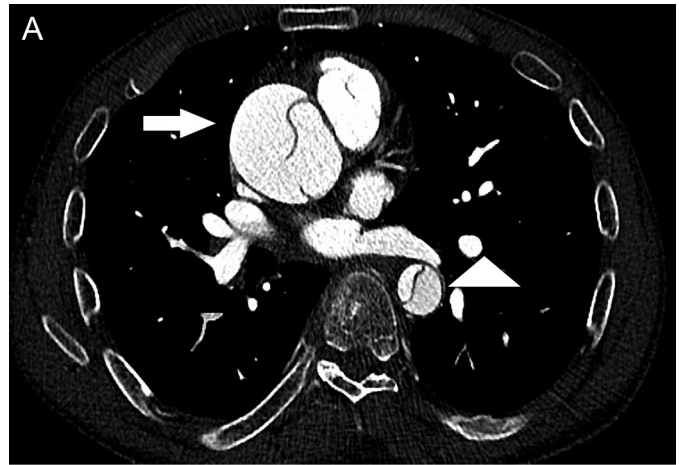


Figure 2. A and B. Computed tomography angiography (CTA) of the chest with contrast demonstrating the dissected ascending aorta (arrow) and the descending aorta (arrowhead).

robust with no obvious signs of aortopathy. The graft was clamped and CPB was reinitiated. The remaining proximal anastomosis and reimplantation of the coronary buttons were completed.

TEE confirmed a normal-functioning prosthetic valve. Global ventricular function was preserved and the patient was weaned from bypass without difficulty. With satisfactory hemostasis achieved, the sternum, presternal fascia, and skin were closed in routine fashion. Total CPB, cross-clamp, and circulatory arrest times were 154, 102, and 19 min, respectively. No blood products were given. Total pump balance including cardioplegia was +850 mL.

The patient's overall hemodynamic status was stable, and he remained neurologically intact. Postoperative TTE

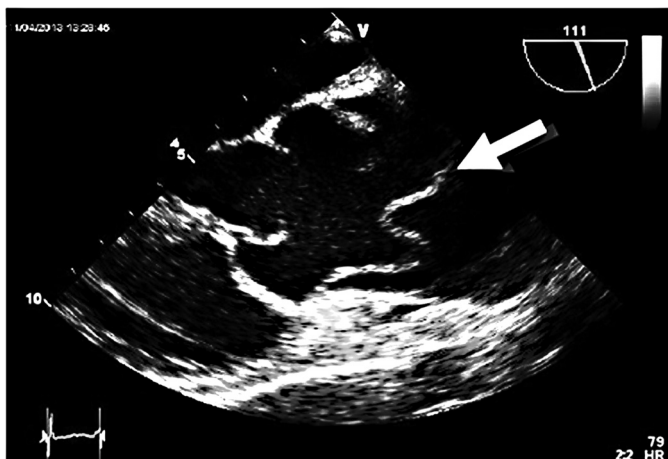


Figure 3. Intraoperative transesophageal echocardiogram (TEE) demonstrates the dissection tear (**arrow**) localized to the aortic root.

demonstrated normal biventricular function, and a well seated #23 mechanical aortic valve with a 6 mm Hg mean gradient and a 2.2 cm² effective orifice area. The patient was discharged to a rehabilitation facility on postoperative day 6. By six weeks postoperatively, he had near complete functional recovery (Fig. 4).

Discussion

Although still relatively rare, achondroplastic dwarfism is the most common type of dwarfism. To our knowledge, there is no association between achondroplastic dwarfism and aortic dissection. This, to the best of our knowledge, is the first report of an acute Type A aortic dissection in an achondroplastic dwarf. There are only four other accounts of cardiac surgeries and dwarfism [2–5]. For a coronary artery bypass graft procedure, the major concern is limited saphenous vein reserve, but otherwise, the procedure can be performed routinely [3]. For aortic valve replacement, aortic root enlargement was required for a very small aortic annulus [4].

Anesthetic Considerations

Achondroplasia can present some unique challenges for anesthetic management [6–8]. Difficult airway management may occur, possibly related to short kyphotic cervical spine, maxillary hypoplasia, and megalencephaly [6]; however, larynx visualization is usually uncomplicated. There may be stenosis of foramen magnum and hydrocephaly. Our patient had



Figure 4. Patient image at follow-up clinic visit.

short thyromental distance and remote history of surgical cervical spine intervention with spinal stenosis. Despite preparations for difficult airway, the trachea was intubated uneventfully with neutral neck position and a video laryngoscope.

Restrictive lung disease from thoracic kyphosis and rib hypoplasia may be present and can lead to pulmonary hypertension and cor pulmonale in advanced cases [6,8]. Our patient had a history of kyphosis but did not have any preexisting symptoms of cardiorespiratory dysfunction.

Finally, vascular access and positioning may be difficult because of reduced joint mobility, thoracic kyphosis, and small limb size [8]. Careful patient positioning using modified padding was required due to marked kyphosis and reduced extremity mobility. Ultrasound guidance was critical for central venous and femoral arterial vascular access.

Perfusion Considerations

The overall perfusion strategy employed was that of a pediatric patient. Every effort was taken to mini-

mize the static and dynamic prime volumes. We used retrograde autologous priming. Also, venous tubing was downsized from 1/2 inch to 3/8 inch and arterial axillary cannulation was accomplished with a 6 French graft without flow or drainage issues. Scafuri et al. [4] reported the use of pediatric cannulae both for the aorta (20F) and for the superior and inferior venae cavae (28F), but their patient was considerably smaller than ours (height 100 cm, weight 27 kg, BSA 0.87 m²). However, because ours is an adult cardiac center, pediatric cannulae were not readily available on such short notice. The CPB circuitry was optimized to reduce the number of shunts, surface area, and connectors. Although we did not have access to the small adult oxygenator with integrated filter, this would have further reduced our dynamic prime volume. Hemo-concentration was utilized throughout to minimize hemodilution. A hemo-concentrator can also be utilized for modified ultrafiltration post-CPB; however, this technique was not employed in our patient. Overall, the patient responded well to CPB.

Surgical Considerations

To our surprise and relief, the surgical management was rather uncomplicated as the patient's thorax was of relatively normal size. The right axillary cut-down was performed with the usual depth and anatomical landmarks. Due to small lower extremities, axillary cannulation is preferred because it avoids cannulation of hypoplastic femoral arteries. The sternum was moderately kyphoid, but sternotomy was uncomplicated. The aortic root was dilated, and although we did not have preop-

erative echocardiographic imaging, the CTA showed quite a large left ventricular outflow tract. Because the BSA was only 1.03 m², patient-prosthesis mismatch was unlikely to be a concern. We planned to implant either a 19- or a 21-mm valve but were pleased to find that a 23-mm valve could be implanted without difficulty. In the only other reported case of aortic valve replacement in a patient with dwarfism, a small aortic root required a Manouagian-type patch aortic root enlargement to fit a 16-mm mechanical Carbomedics valve (Sorin Group Company, Austin, TX, USA) in a 56-year-old female with a very small, 10-mm, aortic annulus [4].

Conclusion

This report, to the best of our knowledge, is the first to describe successful aortic dissection repair in a patient with achondroplastic dwarfism. The procedure was performed without the need for specialized cannulation equipment or aortic root enlargement procedures to facilitate a suitable valve prosthesis. This report adds to the experience of successful acquired and congenital cardiac surgery in this unique patient population.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question

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