LETTER TO THE EDITOR

Intraoperative aortic dissection in a Turner syndrome patient

Aortic dissection is a rare but catastrophic complication of cardiac surgery with a mortality ranging from 24-43% (1). Turner syndrome (TS) patients are known to be at an increased risk of undergoing an insidious aortic dissection, however, little is known about their risk of undergoing iatrogenic dissection. In addition to airway management complications [due to retrognathia, micrognathia, limited neck mobility and short trachea (2)], these patients may also be a high risk population for experiencing an intraoperative aortic dissection particularly during cardiopulmonary bypass (CPB).

We experienced an ascending aortic dissection in a 64 year old TS patient with coronary artery disease who was admitted for elective coronary artery bypass grafting. This was noted after aortic unclamping (*Figure 1*), following successful weaning from CPB, and subsequently required reinstitution of CPB in order to repair the dissected proximal aorta with a prosthetic tube graft. The dissection area was limited and did not require repositioning of aortic cannulation. The patient was then again weaned from CPB with a 3 µg/min infusion of both epinephrine and norephinephrine (0.028 µg/kg/min of each). Strict blood pressure control was maintained throughout the surgery. The distal ascending aorta was cannulated opposite the innominate artery take-off, as per the surgeon's routine, and hypothermia to 32°F was utilized during the aortic repair. To our knowledge this is the first report of a dissection occurring intraoperatively in a TS patient despite their known higher risk.

TS occurs in about 1 in 2,500 females and aortic dissection is 6 times more likely in this population compared to the incidence of dissection in the general population (male and female) of 2.6-3.5 per 100,000 person-years (3). Because of this, echocardiography during diagnosis has become a standard of care screening for these

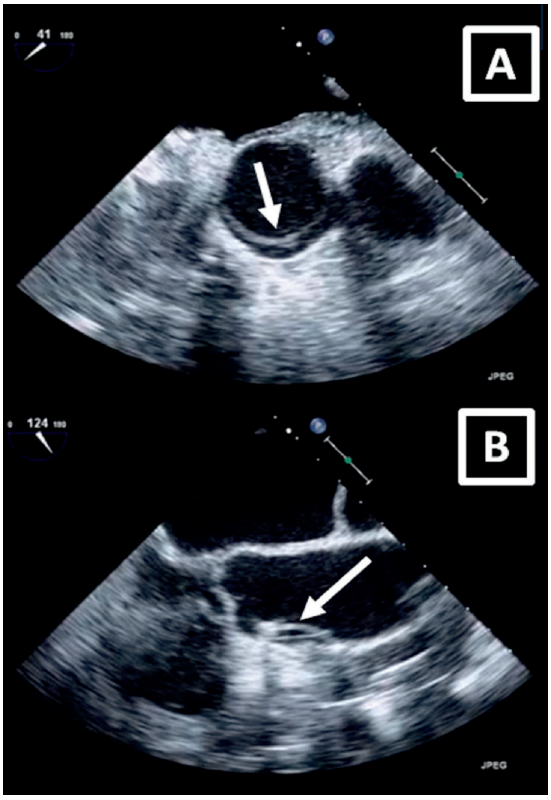

Figure 1 - *Ascending aortic dissection shown in short axis A and long axis B.*

patients and it is recommended that they undergo routine thoracic magnetic resonance imaging (MRI) at 10 years of age (4). Presumably the risk is increased in TS due to the connective tissue pathogenesis and hypertension attributed to the syndrome. This patient had hypertension as well as diabetes mellitus type II and hypercholesterolemia, all associated with TS. She also had a history of congestive heart failure secondary to diastolic dysfunction and sleep apnea. She was diagnosed at childhood and did not receive growth hormone therapy or undergo any prior aortic imaging with the exception of cardiac catheterization which noted no significant aortic dilatation. She was 150cm tall, weighed 108.5 kg, had a bilateral hearing loss with hearing aids, and was managed on ASA, furosemide, atorvastatin, metoprolol, valsartan, potassium, and calcium with vitamin D.

Prior to surgical incision we performed a basic transesophageal echocardiogram (TEE) exam which, upon aortic evaluation, showed no dissection and only a very small calcified plaque at the right sino-tubular junction (STJ). The effaced STJ measured 25 mm, the maximal ascending aortic diameter was 31 mm (yielding an aortic size index of 1.5 cm/m^2 given her body surface area of 2.13 m^2), and the arch and proximal descending aorta measured 18 mm. She had a tricuspid aortic valve with trace central aortic insufficiency, normal left and right systolic function, mild left ventricle hypertrophy, mild left and severe right atrial (RA) dilatations. There was no evidence of aortic coarctation or aortic flow acceleration at the proximal descending arch. She had an interrupted inferior vena cava (IVC), a vascular anomaly associated with TS, whereby venous return to the RA was through a dilated Azygos vein rather than the IVC. The dilated RA may have been related to altered flow dynamics since total RA flow is not increased with an interrupted IVC.

Many of the TS case reports in the International Turner Syndrome Aortic Dissection Registry (5) describe the complication of aortic dissection as developing insidiously. It is possible that this was an untoward intraoperative dissection unrelated to the patient's pathology but we believe that, on the contrary, the shear stress of aortic cannulation and decannulation during CPB connection precipitated dissection of a predisposed weak vessel. This hypothesis is more likely given that iatrogenic intraoperative ascending aortic dissections occurred in only 7 out of 15,144 consecutive general population patients following cardiac surgery (1) and also by the observation that TS patients with type A dissection had a mean ascending aortic size index of 2.7 ± 0.6 cm/m^2, significantly higher than our patient's (5). Furthermore, aortic pathology noted that the "layers of the aorta appear to be separating" in the 4.5x3x2 cm specimen with "segments of large artery" (histological study not performed). The dissected area included the area where the aortic root vent was placed - its contribution is possible - though we could not ascertain it. Due to the potential increased aortic wall friability in TS, we recommend that, in addition to routine TEE, surgeons practice extra caution during aortic manipulation, particularly with aortic cannulation in these patients. Surgical practices of upmost importance under this scenario include: avoidance of aortic cannula with stiff occluders, ensuring proper cannula location and application or removal of aortic cross-clamp only during reduction (or cessation) of CPB along with preference toward using the Chitwood clamp instead of balloon occlusion when minimally invasive cardiac surgery is performed.

REFERENCES

1. Narayan P, Angelini GD, Bryan AJ. Iatrogenic intraoperative type A aortic dissection following cardiac surgery. Asian Cardiovasc Thorac Ann. 2015; 23: 31-5.
2. Liu WC, Hwang CB, Cheng RK, Wu RS, Hui YL, Tan PP. Unexpected left endobronchial intubation in a case of Turner's syndrome. Acta Anaesthesiol Sin. 1997; 35: 253-6.
3. Bickerstaff LK, Pairolero PC, Hollier LH, Melton LJ, Van Peenen HJ, Cherry KJ, et al. Thoracic aortic aneurysms: a population-based study. Surgery. 1982;92:1103.
4. Turtle EJ, Sule AA, Webb DJ, Bath LE. Aortic dissection in children and adolescents with Turner syndrome: risk factors and management recommendations. Arch Dis Child. 2014; 0: 1-5.
5. Carlson M, Airhart N, Lopez L, Silberbach M. Moderate Aortic Enlargement and Bicuspid Aortic Valve Are Associated With Aortic Dissection in Turner Syndrome Report of the International Turner Syndrome Aortic Dissection Registry. Circulation. 2012; 126: 2220-6.

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