

References

1. Gittenberger-De Groot AC. Persistent ductus arteriosus: Most probably a primary congenital malformation. *Br Heart J* 1977; 39: 610-8
2. Campbell M. Natural history of persistent ductus arteriosus. *Br Heart J* 1968; 30: 4-13.
3. Patel AL, Shaikh WA, Deshpande PV, Malaviya AP, Dighe PS, Dangi UB. Patent ductus arteriosus with a left to right shunt in a 64-year-old asymptomatic male. *J Assoc Physicians India* 2004; 52: 753-5.
4. Zarich S, Leonardi H, Pippin J, Tuthill J, Lewis S. Patent ductus arteriosus in the elderly. *Chest* 1988; 94: 1103-5.
5. Kouris NT, Sifaki MD, Kontogianni DD, Zaharos I, Kalkandi EM, Grassos HE, et al. Patent ductus arteriosus endarteritis in a 40-year old woman, diagnosed with Transesophageal Echocardiography. A case report and a brief review of the literature. *Cardiovasc Ultrasound* 2003; 24; 1: 2.
6. Çay S, Öztürk S, Korkmaz S, Türkvatın A. Asymptomatic patent ductus arteriosus in a 41-year old woman. *Int J Cardiovasc Imaging* 2006; 22: 283-5.
7. Goitein O, Fuhrman CR, Lacomis JM. Incidental finding on MDCT of patent ductus arteriosus: use of CT and MRI to assess clinical importance. *Am J Roentgenol* 2005; 184: 1924-31.
8. Shyu KG, Lai LP, Lin SC, Chang H, Chen JJ. Diagnostic accuracy of transesophageal echocardiography for detecting patent ductus arteriosus in adolescents and adults. *Chest* 1995; 108: 1201-5.
9. Hoffmann MH, Shi H, Manzke R, Schmid FT, De Vries L, Grass M, et al. Noninvasive coronary angiography with 16-detector row CT: effect of heart rate. *Radiology* 2005; 234: 86-97.

Surgical repair of supra-avalvular aortic stenosis in Williams syndrome

Williams sendromlu bir olguda supra-avalvüler aort stenozunun cerrahi tedavisi

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Introduction

Williams syndrome is a rare genetic condition which occurs in 1/7500 births, and causes cardiovascular, developmental and intellectual problems. The syndrome is characterized by varying degrees of left ventricular outflow tract obstruction beginning superior to the aortic valve, hypercalcemia, elfin faces, and mental retardation (1).

Case report

In April 2007, a 17-year-old boy was referred to our clinic because of a cardiac murmur. He had moderate mental retardation documented with WISC-R test. Aortography, coronary angiography, echocardiography and 3-D computed tomography were performed before the operation, and all demonstrated a typical hour-glass type stenosis just above the aortic valve (Fig. 1-2). Computed tomography revealed minimal stenosis of left pulmonary artery (Fig. 3). The preoperative peak systolic pressure gradient between the left ventricle and ascending aorta was 110 mmHg. There was no coronary stenosis, other cardiac anomalies or renal artery stenosis.

We performed successful surgical repair of extended aortoplasty using Doty's method (two sinus reconstructions) for congenital supra-avalvular aortic stenosis. After standard cardiopulmonary bypass, bi-sinusal inverted Y incision was made between noncoronary cusp and right coronary cusp; and the ridge was removed with sharp dissection above the left coronary sinus. Resulting defect on the aorta was then repaired with Dacron patch (Fig. 4). The systolic pressure gradient between the left ventricle and ascending aorta improved to

normal postoperatively. Peroperative gradient between the main pulmonary artery and pulmonary artery was 22 mmHg on the right and 10 mmHg on the left and did not require surgical correction. The patient was extubated 7 hours following surgery, and was discharged at the 5th postoperative day. Six months after the procedure, the patient was fine and had no complaints.

Discussion

Supra-avalvular aortic stenosis is an unusual form of obstruction of the left ventricle outflow tract, which occurs in 3% to 6% of cases of various types of aortic obstruction. It may be due to the presence of a discrete fibrous membrane, an hourglass narrowing, or a diffuse narrowing. The anatomical variants of supra-avalvular aortic stenosis affect post surgery results and survival rates, e.g. the hourglass type of obstruction is the most common, whereas the diffuse type of stenosis mostly results in death (2). The syndrome was first described in 1930 by Mencarelli, and its association with other facial defects and psychomotor retardation was established in 1961 (3). In 1965, Logan described patients who had a familial form of supra-avalvular aortic stenosis with normal face and normal mental development, which may delay diagnosis (4, 6). Supra-avalvular aortic stenosis is a familial autosomal dominant disorder with variable expression and affects both sexes equally. The vascular pathology of familial supra-avalvular aortic stenosis and Williams syndrome results from mutations involving the elastin gene on chromosome 7q11.23 (5).

In 1961, McGoon et al. proposed widening the aorta diameter with a synthetic graft as a treatment option. In 1977, Doty used his



Figure 1. Computed tomography image of supralvalvular aortic stenosis



Figure 2. Aortography image showing supralvalvular aortic stenosis

well-known technique, which involves an inverted Y patch for the first time with success. When the stenosis is very close to the valves and the coronary ostia, the use of the Doty technique may be preferred (7). There are several variations of the surgical technique for correcting this type of defect. In a study concerning the efficacy of several techniques, Hazekamp et al., did not find any significant differences in change in valve function, and found the efficacy of reducing the pressure gradient was similar and acceptable in all techniques (8). In the cases of recurrent serious stenosis, an alternative is to use a valved conduit between the free wall of aortic root and the stenosis free aorta (3). Similarly, autologous arterial graft from the pulmonary artery, as described by Al-Halees et al, can be used as another option not only for cases of recurring stenosis but also for complex cases with diffuse stenosis (9).

Conclusion

In conclusion, supralvalvular aortic stenosis is a complex and uncommon disease that requires careful preoperative evaluation and proper selection of surgical treatment method. We conclude that Doty's inverted Y patching method may be an effective and suitable choice for correction of the supralvalvular aortic stenosis.

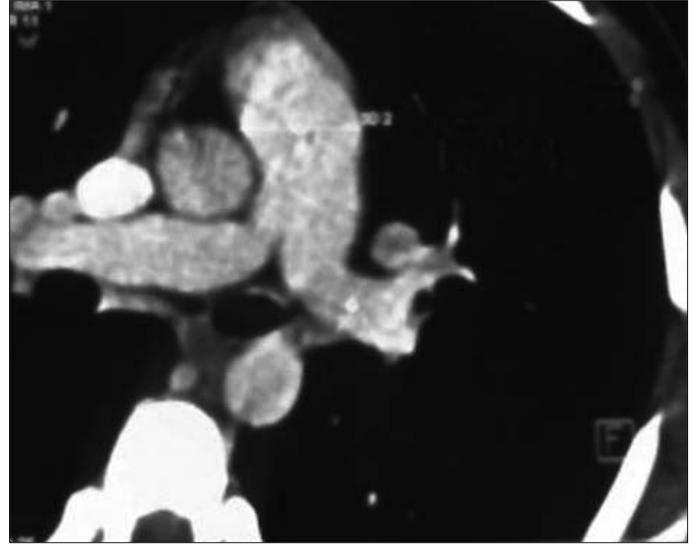


Figure 3. Computed tomography image of left pulmonary artery stenosis.

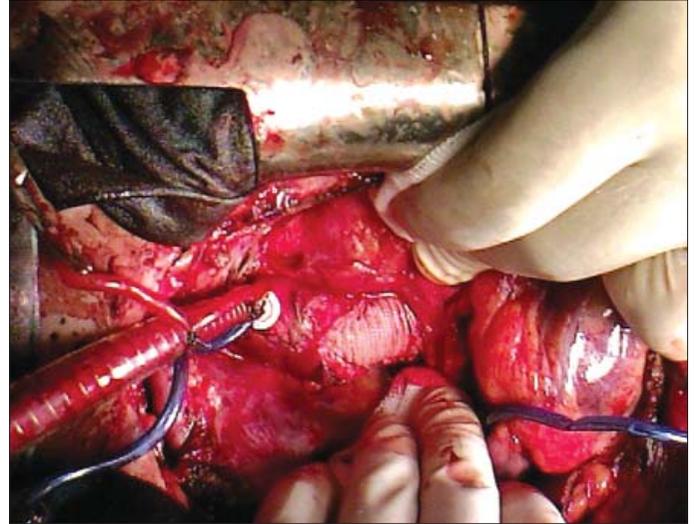


Figure 4. Doty's procedure

References

1. Garc'a RE, Friedman WF, Kaback MM, Rowe RD. Idiopathic hypercalcemia and supralvalvular aortic stenosis. Documentation of a new syndrome. *N Engl J Med* 1964; 271: 117-25.
2. Keane JF, Fellows KE, LaFarge G, Nadas AS, Bernhard WF. The surgical management of discrete and diffuse supralvalvular aortic stenosis. *Circulation* 1976; 54: 112-6.
3. Williams JCP, Barrat-Boyes BG, Lowe JB. Supralvalvular aortic stenosis. *Circulation* 1961; 24: 1311-8.
4. Logan WFW, Jones E Wyn, Walker E, Coulshed N, Epstein EJ. Familial supralvalvular aortic stenosis. *Br Heart J* 1965; 27: 547-59.
5. Curran ME, Atkinson DL, Ewart AK, Morris CA, Leppert MF, Keating MT. The elastin gene is disrupted by a translocation associated with the supralvalvular aortic stenosis. *Cell* 1993; 73: 159-68.
6. Bostan ÖM, Çil E. Supralvalvüler aort stenozlu dokuz vakanın değerlendirilmesi. *Türk Kardiyol Dern Arş* 2000; 28: 752-6.
7. Doty DB, Polansky DB, Jenson GB. Supralvalvular aortic stenosis. Repair by extended aortoplasty. *J Thorac Cardiovasc Surg* 1977; 74: 362-71.
8. Hazekamp MG, Kappetein AP, Schoof PH, Ottenkamp J, Witsenburg M, Huysmans HA, et al. Brom's three-patch technique for repair supralvalvular aortic stenosis. *J Thorac Cardiovasc Surg* 1999; 118: 252-8.
9. Al-Halees Z, Prabhakar G, Galal O. Reconstruction of supralvalvular aortic stenosis with autologous pulmonary artery. *Ann Thorac Surg* 1998; 65: 532-4.