Discrete Subvalvular Aortic Stenosis With Recurrent Patent Ductus Arteriosus

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Abstract

The majority of the congenital malformations need surgical treatment and can be corrected anatomically with a low operative risk. Patients with subaortic stenosis represent a heterogeneous group. Associated anomalies are common. In this study, we aimed to present our successful surgical technique under the light of the literature applied to a 26-year-old female who had undergone closure of patent ductus arteriosus when she was 7 years old. A combined pathology of patent ductus arteriosus and discrete subvalvular aortic stenosis was found recently.

The prevalence of recurrent patent ductus arteriosus due to insufficient ligation is about 1%. Also discrete subaortic stenosis is a rare, late complication of the surgical repair of several congenital heart defects. Its open aortic resection is safer and more likely to achieve a better haemodynamic result.

INTRODUCTION

Hemodynamical classification, general distribution and surgical considerations of the most common congenital malformations of the heart are described (1). The first successful ligation of patent ductus arteriosus had been carried out by Robert Gross in 1939 (2). At Children’s Memorial Hospital in Chicago, during the investigation of a series of 1108 cases throughout 46 years, the frequency of recurrent PDA due to insufficient ligation as the major complication causing reoperation was calculated as 0.09% (3). At Viet Duc Hospital in Vietnam, among 100 PDA cases that underwent ligation between 1960 and 1979, the recurrence rate was 5% (4). Discrete subaortic stenosis is a rare, late complication of the surgical repair of several congenital heart defects (5). Because of the unexpected finding of discrete membranous subaortic stenosis in the infants and young children who had undergone surgery for a large patent ductus arteriosus and because of the treacherous worsening of the effects of the discrete membranous subaortic stenosis as childhood progressed, it is important that those patients with a persistent systolic murmur after ductal ligation not be discharged from cardiac follow-up as cured. Serial cardiac catheterization during the growing years appears to be the most accurate way of detecting worsening discrete membranous subaortic stenosis, so that the membrane can be excised before severe complications occur (6).

CASE PRESENTATION

Our case was a 26-year-old female. Her chief complaint was palpitation going on for the last 2 years. Her medical history revealed that she had undergone an operation of PDA closure when she was 7 years old at another center. Her transthoracic echocardiography showed severe aortic stenosis (mean gradient of 62 mm Hg) and an image compatible with a membrane at subaortic region. Moreover, images compatible with mild aortic regurgitation and a recurrent PDA showing a shunting via Color Doppler (Figure 1). Transesophageal echocardiography performed afterwards revealed similar findings.
Figure 1
Figure 1: Shunting through recurrent PDA shown by Color Doppler of transthoracic echocardiography performed preoperatively.

Aortography showed severe aortic stenosis and aortic regurgitation as well as high output recurrent PDA (Figure 2).

Figure 2
Figure 2: Recurrent PDA seen in aortography of our case.

SURGICAL TECHNIQUE
Standard median sternotomy was performed. Right subclavian artery was the site for arterial cannulation. Using total circulatory arrest and retaining the flow rate at 500 cc/min aortic cross-clamp was put below the innominate artery. Afterwards pulmonary arteriotomy was performed to explore the ostium of large PDA (0.5 millimeters in diameter) (Figure 3).

Figure 3
Figure 3: Transpulmonary view of recurrent PDA.

Closure of recurrent PDA was carried out by using Dacron patch with the support of Teflon©-pledgeted sutures (Figure 4). Total circulatory arrest lasted 11 minutes.

Figure 4
Figure 4: View of recurrent PDA closed by Dacron patch.

Afterwards, commissurotomy was carried out to the commissure between right and noncoronary cusps via aortotomy. Discrete fibrous membrane was resected that was located mainly under right coronary leaflet (Figure 5).
Pulmonary arteriotomy and aortotomy incisions were closed and cardiopulmonary bypass was ended without facing a problem perioperative transesophageal echocardiography revealed mild aortic regurgitation and operation was finished successfully (Figure 6).

The patient was discharged on 7th postoperative day with total recovery. In the first postoperative month a control transthoracic echocardiography was carried out showing an insignificant aortic regurgitation and no flow through PDA (Figure 7, 8 and 9). Follow-up of this case still continues.
**DISCUSSION**

Congenital heart defects are the most common birth defects and represent an increasing proportion of adolescent and adult patients followed by cardiologists. While many of these patients have undergone successful palliative or corrective surgery with excellent functional results, most of them still require careful follow-up. Further, even complex lesions may first be diagnosed in adolescence and adulthood. Therefore, cardiologists caring for adults need to become more familiar with these defects. In addition, the catheterization laboratory remains a critical venue for diagnosis (3).

Discrete subaortic stenosis is a rare, late complication of the surgical repair of several congenital heart defects (3). The three anatomic types of obstruction found are the thin membranous type, the fibromuscular collar type and the tunnel type (3). The obstruction is usually severe and progressive obstruction with an increasing gradient is documented(3). Significant associated cardiac defects, often masked the typical clinical and cardiac catheterization features of subaortic stenosis. The stenosis is often not discovered until after surgery for the associated defect(3). In study of Penkoske et al., 21 children required surgical treatment for subaortic stenosis (3). Associated anomalies were common and patent ductus arteriosus occurred in three patients. In the report of Tokel et al, presented nine patients who had no significant left ventricular-aortic obstruction at initial cardiac catheterization or echocardiographic examination, but later developed significant subvalvular aortic stenosis. Associated lesions included patent ductus arteriosus in two and ventricular septal defect and patent ductus arteriosus in one case. Nine patients were diagnosed with subvalvular aortic stenosis 18 months to eight years after surgical correction. Eight of the patients required surgery for subvalvular obstruction (3). The criteria for operability of discrete subaortic stenosis should be the angiographic demonstration of a discrete subvalvular diaphragm and the presence of a resting left ventricular to aortic systolic pressure gradient of 40 mm Hg or more (3). Early resection and additional procedures can be performed with a low mortality rate and can eliminate aortic insufficiency in many cases (3).

The attempt of closure of patent ductus arteriosus (PDA) with traditional methods may cause life-threatening bleeding if done when other abnormalities coexist or in cases when left pleural membranes are adherent (3). In this type of cases, under cardiopulmonary bypass and deep hypothermia with low flow rates or under total circulatory arrest PDA can be closed via transpulmonary approach (3). After constituting low flow rate with deep hypothermia or total circulatory arrest, PDA can be reached via transpulmonary approach and PDA can be closed by using Dacron patch or teflon pledged interrupted sutures (3). Deep hypothermic total circulatory arrest provides options to close aortopulmonary connection without causing an irreversible neurologic deficit. The advantage of this technique is that it provides bloodless and comfortable surgical field and it secures the treatment of accompanying cardiac abnormalities in the same session (3). In this type of patients, total circulatory arrest usually lasts short and it is mostly sufficient to enter total circulatory arrest intermittantly. Even if accompanying pathology exists, it does not prolong the time of total circulatory arrest. The correction of coexisting abnormalities can be performed during warming up period after total circulatory arrest (3).

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