Surgical Management of Aortic Stenosis in a Child

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Introduction

Aortic valve replacement and choice of prosthesis is a challenge in children as compared to adults. The situation is complicated by the problems of growth in children, leading to patient prosthesis mismatch and requirement of anticoagulation. In spite of variety of mechanical and biological valve designs, an ideal valve is still a distant dream [1]. An ideal valve prosthesis in children should offer good hemodynamics even in small sizes, avoid anticoagulation and should grow with the child. Small prosthesis which do not grow with the child become obstructive and require re-do surgery. The pulmonary autograft or Ross operation I, as an aortic valve replacement provides the solution to most of the problems, in which the aortic valve is replaced by the pulmonary valve of the patient and pulmonary valve is replaced by a homograft or a valved conduit [2]. We report our experience of the first case from armed forces.

Case Report

A sixteen year old patient, a known case of annular hypoplasia with severe aortic stenosis who underwent unsuccessful aortic balloon dilatation earlier, presented with the history of recurrent episodes of syncope since 11 years of age along with breathlessness New York Heart Association functional class II of three months duration. There was no history of cyanosis, palpitation, headache or chest pain. Her physical examination was normal and cardiovascular examination revealed sustained apical impulse in fifth intercostal space with an ejection systolic murmur grade V/VI in aortic area with radiation to carotids. Respiratory and abdominal examination was normal. Routine hematological and biochemical investigations were within normal limits. Electrocardiogram showed left ventricular hypertrophy with strain pattern. 2-D echocardiography revealed a bicuspid aortic valve with a gradient of 142 mmHg suggestive of severe aortic stenosis and an annulus of 16mm suggestive of small aortic root. There was no subaortic membrane or left

ventricular outflow tract hypertrophy. Patient was taken up for Ross operation. Standard cardiopulmonary bypass was established by aorto-bicaval cannulation. Heart was arrested by applying aortic cross clamp and giving cold blood potassium rich cardioplegia. Peroperatively pulmonary valve was 18 mm, tricuspid and competent, whereas the aortic root was narrow measuring 14mm. Aortic valve was bicuspid, thickened, dysmorphic with subaortic fibrosis and thickening resulting in severe valvular aortic stenosis (pinhole). Aortic valve was excised and subaortic region cleared. Pulmonary valve was enucleated and implanted in aortic position using root replacement technique (Fig.1,2). A bovine jugular vein conduit (contegra) of size 22 was implanted to restore right ventricle pulmonary artery continuity. Patient was gradually weaned off cardiopulmonary bypass. Heparin was reversed with protamine, decannulation and hemostasis was performed. Chest was closed leaving pericardial and retrosternal drains. Patient was shifted to intensive care unit in stable state. She had an uneventful postoperative recovery and was discharged home on 15th postoperative day with standard anti-failure medication and tablet ecosprin 75 mg for three months. Echocardiography before discharge and at six months of follow up showed both operated valves functioning well with no evidence of stenosis or insufficiency.

Discussion

In 1967, Ross, described replacement of the aortic valve (Ross I) as well as replacement of the mitral valve (Ross II) with a pulmonary autograft [2]. The original Ross procedure involved a subcoronary replacement of the aortic valve [2]. The procedure is now being performed almost universally as a root replacement. There was evidence that the initial subcoronary implantation technique, has higher early and midterm failure rates when adopted and practiced by many surgeons. The usefulness of the Ross operation as a root replacement in children is proven and many of these patients are not candidates for subcoronary Ross operation. The root replacement technique has the

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advantage of larger annulus diameters and greater aortic effective orifice area [3]. However, long-term dilatation of the root, if no reinforcement techniques are used during anastomoses is a potential drawback. The limitations of subcoronary implantation technique are the expertise of the surgeon, extensive root calcification, especially around the coronary ostia and a marked distortion of the annulus [4].

The use of conventional biologic prosthesis in children avoids anticoagulation, but their general use is limited owing to the disappointing durability of biologic valve tissue in children [5]. Due to higher metabolic rate in growing children these valves are prone to early calcification requiring re-replacements. Aortic valve replacement with cryopreserved aortic homografts have not been a convincing option in children [6].

The freedom from anticoagulation and freedom from frequent blood testing together with a complete absence of thromboembolic or hemorrhagic complications contribute to quality of life and possibility of a normal childhood, advantages that are not possible with mechanical prosthesis. Thus, the Ross operation offers a striking advantage as compared to the mechanical valve prosthesis in young female patients. Dore et al [7], have reported 14 pregnancies in eight young women after pulmonary autograft valve replacement, without valve related complications [7]. Somatic autograft growth is well recognized and the advantages have been supported by clinical observations [8] and experimental studies [9]. Pessotto et al [10], have reported that the pulmonary autograft continues to grow as a normal pulmonary root, even when placed in the aortic position.

Pulmonary homografts were the conduit of choice for reconstruction in Ross procedures for most of the institutions. Constraints of availability of homografts has forced us to use the new Contegra bovine jugular vein conduit. The Contegra (Medtronic Inc, Minneapolis, MN) xenograft conduit is a bovine jugular vein fixed with glutaraldehyde. Since its introduction in 1999 early and intermediate clinical results of its use for right ventricular outflow tract repair in congenital anomalies and Ross procedure are encouraging [11,12]. It is a biological conduit, obtained by fixing a valved segment of bovine jugular vein with low-concentration buffered glutaraldehyde at very low pressure which is then further sterilized. The conduit is available in 12 to 22-mm sizes and comes with natural integral tri-leaflet venous valve in the centre of the conduit. Freedom from graft dysfunction and reoperation at two, five and seven years was 88.9%, 87.6%, and 81.3% for all homografts and 94.0%, 90.7%, and 90.7% for Contegra grafts [13]. Late outcome for the Ross procedure is excellent in terms of survival and quality of life. Late root dilatation, autograft

regurgitation and homograft stenosis, however show an increasing prevalence with time. Technical modifications of the procedure, yearly aortic root imaging during follow up and early reintervention on the dilated neo-aortic root may further enhance the durability of the autologous pulmonary valve.

In conclusion, the Ross operation can be performed with good autograft function and low mortality in young patients resulting in a better quality of life unencumbered by systemic anticoagulation.

Conflicts of Interest

None identified

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