Surgical Approach To A Congenital Mitral Stenosis Case With Left Persistent Superior Vena Cava

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Citation

Abstract
Congenital mitral valve stenosis is a rare pathology.

In this study, we aimed to present our surgical approach and successful valve repair to a case with congenital mitral valve stenosis combined with left persistent superior vena cava.

Conservative surgery of the congenital mitral valve stenosis can be performed with acceptable early and midterm outcome in terms of mortality and reoperation rate. For this reason it is the procedure of choice for congenital mitral valve stenosis.

INTRODUCTION
Congenital malformations of the mitral valve are rare, complex, and frequently associated with other cardiac malformations(1). In congenital mitral anomalies, mostly because of the presence of the dysplastic leaflet group, the anatomy overlaps the functional groups and repair strategies can be identical(1). The current risk of mitral valve operation in the pediatric age group is low, and the long-term results are satisfactory, irrespective of severe deformation of the mitral valve apparatus and associated complex cardiac anomalies(2). The long-term results of conservative surgery are confirmed with a low incidence of reoperation except in mitral valve stenosis(3).

CASE PRESENTATION
Our case was a 5-year-old girl who was admitted to the Department of Pediatric Cardiology with complaints of fatigue. Echocardiographic investigations revealed severe mitral stenosis and she was hospitalized for surgical correction.

She was operated under endotracheal general anesthesia and in supine position. Following a median sternotomy, pericardium was opened longitudinally. After heparinization, extra-corporeal circulation was established between the venae cavae and the ascending aorta. A cross clamp was placed on aorta and by antegrade intermittent isothermic blood cardioplegia, cardiac arrest was established. Hypothermia was moderate (28ºc). A vent was placed via the right superior pulmonary vein. After standard cannulation, right atriotomy was performed revealing an excessive amount of blood draining from coronary sinus ostium. Persistent left superior vena cava was found and suspended and snared (Figure 1).

Figure 1
Figure 1

Subsequently, left atriotomy was done. The diameter of mitral annulus was compatible with her age. A severe mitral stenosis was seen due to commissural fusions. Suspensory
sutures were put on the midpoints of the edges of anterior and posterior leaflets. These leaflets were suspended superolaterally demonstrating that the leaflet structure was severely deformed (Figure 2).

**Figure 2**
Figure 2

Mitral commissurotomy was performed to the commissural areas not attached by the chordae (Figure 3).

**Figure 3**
Figure 3

We tested the valve competence after this step on observing valve closure while the left ventricular cavity was filled with saline solution. There wasn't any saline regurgitation (Figure 4).

**Figure 4**
Figure 4

Anterior and posterior leaflet structures were found to be competent. Opening of the valve was tested via Hegar dilators pointing that it was optimal (Figure 5).

**Figure 5**
Figure 5

No additional problem was seen postoperatively and she was discharged on 7th postoperative day with surgical cure and outpatient clinic follow-up was recommended. Postoperatively on the day of discharge and after 3 months an echocardiographic investigation revealed no regurgitation for the repaired mitral valve.

**DISCUSSION**

In the last few years, surgery of congenital mitral valve lesions has gained from echocardiography, which shows the exact function and anatomy of the mitral valve(3).

Conservative surgery is recognized as the best option(4).
Surgical management of congenital malformation of the mitral valve in the pediatric age group remains a therapeutic challenge for the wide spectrum of the morphological abnormalities and the high incidence of associated cardiac anomalies, and small patient size. Mitral valve (MV) conservative surgery is always advisable and its results are superior to MV replacement.

In the series of Prifti et al., between January 1990 and February 2001, 94 consecutive children with congenital MV disease underwent valve repair. The mean age was 5.2+/−3.3 years (range 20 days to 15 years). Twenty-five (26.6%) children were less than 1 year old. Isolated MV disease was found in 21 (22.4%) patients. MV stenosis was the predominant lesion in 21 (22.4%) patients. The hospital mortality was 8.5% (8 of 94). Actuarial survival and actuarial reoperation-free survival were 89.2% and 76.3%, respectively.

Early mitral valve repair saves the lives of patients with severe symptoms, particularly those with mitral stenosis.

MV reconstructive procedures in infants and children with congenital MV dysplasia may be effective and reliable with low mortality and low incidence of reoperation rate.

When definitive repair cannot be realized, effective palliation can be achieved to permit growth and subsequent implantation of larger prostheses in anatomic position.

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