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Original Article

Management of mitral valve regurgitation in patients with ALCAPA

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Abstract
Introduction: *Anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital malformation. Immediate surgical correction on diagnosis with the aim of restoring a two-coronary system circulation is the current standard of treatment. However, the management of the mitral valve regurgitation in these patients is controversial.*
Methods: *Fifty one patients of ALCAPA underwent coronary reimplantation over a 10-year period. The mitral valve was not addressed (except one patient with an organic basis) at the time of coronary implantation even in patients with moderate to severe mitral regurgitation. These patients were followed postoperatively with with serial echocardiography at six months and at two years thereafter to assess the improvement in ventricular and mitral valvular function.*
Result: *A total of 53 cases were studied. Mitral valve regurgitation of varying severity was seen in 51 (96.5%) of the 53 patients preoperatively. There were 5 postoperative deaths (9.6%). Postoperatively, after 6 months, mitral regurgitation was none to mild in 38 patients (84.4%, n= 45). Moderate to severe mitral regurgitation was seen in 7 patients (15.6%). At 2 years follow up, 43 patients (95.5%, n= 45) had none to mild mitral regurgitation. Only two patients had moderate to severe mitral regurgitation.*
Conclusion: *Reimplantation of anomalous coronary artery to the aorta must be the procedure of choice in patients with ALCAPA. Mitral valve repair is not required even in severe regurgitation, except when there is an organic basis of regurgitation.*
Keywords: *ALCAPA, mitral regurgitation, coronary reimplantation.*

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly, first described by Brooks[1] in 1886. It is usually seen as an isolated lesion[2,3] and is present in one of 300,000 live births (0.25% to 0.5%[2,4,5]). It represents one of the most common causes of myocardial ischemia and infarction in children and if left untreated, results in mortality rate of up to 90% within the first year of life[6]. Immediate surgical correction on diagnosis with the aim of restoring a two-coronary system circulation is the current standard in patients with ALCAPA[3,7,8].

Mitral Regurgitation is a very common finding in patients with ALCAPA. It is related both to

ischemic LV dilatation with mitral annulus enlargement and to ischemic dysfunction of papillary muscles. As myocardial ischemia, ventricular dilatation and papillary muscle dysfunction improve after repair, associated MR may also improve[9].

In many series, moderate or even severe mitral insufficiency has not been addressed at the time of initial repair of ALCAPA, the primary goal of which has been coronary reperfusion and myocardial salvage. There are others who suggest that some intervention needs to be done on the mitral valve if there is an organic basis of mitral regurgitation or if the mitral regurgitation is severe.

Methods

Fifty one patients underwent coronary reimplantation from January 1999 to December 2008 during a 10-year period (median age, 4 months; range, 1 month to 14 years) at Pediatric Division, Deptt. of CVTS AIMS, Kochi during a period of 10 years from January 2000 to December 2009, with a median follow-up of 65 months. In all patients (except one), the mitral valve was not addressed at the time of coronary implantation, even in the presence of moderate or severe mitral regurgitation. In this one patient associated with severe MR, mitral valve repair (annuloplasty at posteromedial commisure, chordal reconstruction with pericardial strip of chorda of AML) was done.

Five patients could not be weaned off bypass at 1st attempt. They were put on support bypass for about 1-2 hours before being weaned off successfully.

Results

A total of 53 cases were studied. The median age at presentation was 4 months. 29 (54.7 %) patients were males and 24 (45.3 %) patients were females. The mean preoperative ejection fraction was 36.5 % (S.D. 10.74). Mitral valve regurgitation of varying severity was seen in 51 (96.5%) of the 53 patients. Of these, mild MR was present in 16 patients (30.2 %), moderate MR was present in 29 patients (54.7 %) and severe MR was present in 6 patients (11.6 %).

All patients underwent direct coronary reimplantation into the aorta. The mitral valve was not addressed at the initial surgery, even in presence of moderate or severe mitral regurgitation. In only one patient, mitral valve repair was performed, in whom there was, in addition to dilatation of the annulus, scarring of the papillary muscles. In this patient, annuloplasty was done with PTFE pledgets over PML commisure and anterior mitral leaflet chorda was constructed with a pericardial strip.

There were 5 postoperative hospital deaths with an overall mortality of 9.6%.

Postoperatively, after 6 months, mitral regurgitation was none to mild in 38 patients (84.4%, n= 45). Moderate to severe mitral regurgitation was seen in 7 patients (15.6%), (Fig 1).

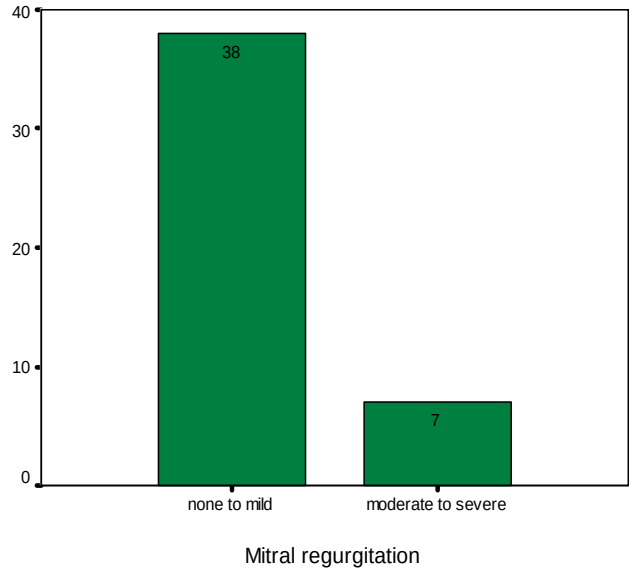

Figure 1. Bar Chart Showing Mitral Regurgitation At 6 Months Postoperatively

At 2 years follow up, 43 patients (95.5%, n= 45) had none to mild mitral regurgitation. Only two patients had moderate to severe mitral regurgitation (Fig 2).

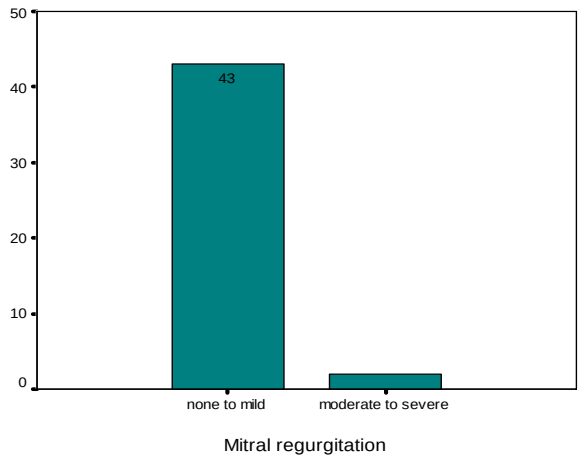

Figure 2. Bar Chart Showing Mitral Regurgitation At 2 Year Follow Up

At 6 months, 20 patients with moderate to severe mitral regurgitation preoperatively had none to mild mitral regurgitation. This change in the degree of mitral regurgitation is statistically significant ($p< 0.05$).

At 2 years follow up, there was further improvement in the degree of mitral regurgitation with 25 patients who had moderate to severe mitral regurgitation preoperatively having no MR or only a mild MR ($p< 0.05$).

Discussion

ALCAPA is a rare congenital anomaly that often causes myocardial ischemia or infarction within the first few weeks or months of life, leading to LV dysfunction, mitral valve incompetence, and congestive heart failure. Establishing a 2-coronary system by direct reimplantation of the anomalous coronary artery into the aorta with pulmonary artery repair has been the main form of surgery since last three decades. First reported by Neches and colleagues,[7] this procedure has been successfully used in infants and adults.

There is controversy regarding the management of the mitral regurgitation in patients with ALCAPA at the time of initial surgery. In many series, moderate or even severe mitral insufficiency has not been addressed at the time of initial repair of ALCAPA, the primary goal of which has been coronary reperfusion and myocardial salvage[10, 11]. They concluded that the mitral valve dysfunction in patients of ALCAPA is usually due to LV dilatation and dysfunction; rarely there might be papillary muscle ischemia with chordal elongation or papillary muscle infarction. Resolution of MR is intricately linked to myocardial reserve remodeling after establishment of a two coronary system. This is usually complete by about seven months postoperatively. Also, it has been seen that infants with myocardial infarcts retain the ability to regenerate lost myocardium. MR persists or progresses in a few, but it is not possible to identify these patients preoperatively whose MR will progress. They also believed that in critically ill patients with compromised myocardium, it is not prudent to prolong the cross clamp time, especially when most patients will have complete resolution of their MR. Presumably, the added ischemic time to perform mitral valve precedures in the setting of severely compromised ventricular function, is potentially more deleterious than helpful. Furthermore, the results from surgical treatment of ischemic MR in adults are relatively disappointing, and "overcorrection" of mitral annulus diameter is not an option in children. Other techniques for correction of ischemic regurgitation, such as Whooler annuloplasty, artificial chordae, chordal shortening, and others may not be applicable in newborns and infants. Early postoperative mild-to-moderate mitral insufficiency is usually acceptable and, after improvement of left ventricular function, persistently symptomatic or hemodynamically significant MR may be treated at a later time.

Some authors[12] recommend routine mitral valve repair at the time of coronary artery revascularization on the grounds that early postoperative cardiac output is improved and operative mortality is reduced. It remains speculative whether their excellent results would have been achieved without associated mitral valve procedures or if their patients were mostly infants, as in our study. Others advocate mitral annuloplastyonly in the presence of severe MR or if there is an organic basis of MR[13,14,15]. In our mostly infant patient population, the mitral valve

was not addressed at the initial repair. In only one patient, where there was an organic basis of MR, a mitral valve repair was done. When we analyzed our results and adverse outcomes, our philosophy of not repairing moderate or severe MR at the initial ALCAPA repair supports our approach. Out of 35 patients of moderate to severe MR, 3 patients (9.09%) who suffered early mortality had moderate to severe mitral regurgitation preoperatively (Fig. 19). 2 patients who died had none to mild mitral regurgitation preoperatively. The relationship between mortality and preoperative mitral incompetence was not found to be statistically significant (p= 1.0).

All 3 patients with severe preoperative MR also had severe LV dysfunction. All 3 infants survived ALCAPA repairs and in our opinion would have been better candidates for a definitive mitral valve repair or replacement if they had been older, had recovered from the myocardial ischemia caused by ALCAPA, and their left ventricles had improved or returned to normal function. The 4 early deaths in the 5 patients who presented with moderate MR were infants (<4 months old) with severe LV dysfunction (EF < 20%). One patient died of an arrhythmia, one died of severe sepsis and one patient died of multiple organ failure. We doubt that mitral valve repair would have changed these patients' outcomes.

Conclusion

The aim of the treatment in ALCAPA is to halt the process of myocardial ischemia and to restore the normal anatomy of the coronary arteries. Reimplantation of anomalous coronary artery to the aorta, with or without elongation techniques, must be the procedure of choice. There is still a controversy as to the best management of mitral valve regurgitation in patients with ALCAPA. Some have advocated leaving the mitral valve alone in all cases. Others have suggested that mitral valve repair is warranted in selected cases. Still others have advocated mitral valve repair in all patients with any degree of mitral regurgitation at the time of presentation. From the experience gained in our series, simultaneous repair of the mitral valve at the time of initial operation is probably not necessary, as a general rule. Although severe mitral regurgitation is a risk factor for death in some series, the added ischemia time for this procedure adds to the risk in an infant with severely compromised ventricular function.

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