

Reconstrutive surgery of regurgitating mitral valve in children

Operação reconstrutora da valva mitral regurgitante em crianças

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INTRODUCTION

Ever since the introduction of open heart surgery for valve operations, dilatation of the mitral annulus found in all patients with mitral valve regurgitation, has been treated conservatively. In 1956, several surgeons, almost simultaneously, started to correct mitral valve regurgitation using annular plicature, a surgical procedure which is still utilized today. After observing the anatomic alterations in patients with mitral valve regurgitation, Carpentier [1], the father of mitral valve reconstructive surgery, developed several techniques for the correction of mitral valve regurgitation, including annuloplasty using a prosthetic ring. After this, several centers around the world adopted his different techniques with excellent results. Antunes et al. [2], from South Africa, widely divulged its use in rheumatic patients very similar to many of our cases.

From 1979 to 1986 almost 100 patients were operated on in our service using Carpentier rings [3]. An absence of dilatation of the anterior portion of the mitral annulus between the two fibrous trigones was seen in all cases. However, in

all cases dilation was observed in the posterior section and more intensively in the proximal segment near the posteromedial commissure [4]. Since 1987, we have routinely employed an open prosthesis for mitral valve regurgitation repair in almost all patients who underwent conservative surgery [5]. Many surgeons in Brazil have utilized this prosthesis, including Carvalho et al. [6], who also recommended it in an inverted position for the repair of tricuspid regurgitation. Together with annuloplasty, reconstructive techniques have also been developed by us to repair mitral valve prolapse caused by the prolongation or rupture of tendinous cords, which is the basis of reconstructive surgical treatment in our service.

Due to the great number of patients with rheumatic valve disease in Brazil, many surgeons developed their own techniques of mitral annuloplasty, contributing very much to the development of reconstructive surgery.

It is important to highlight the great importance of preserving the native mitral valve with regurgitation especially when the patient is under 15 years of age, as the patient may require many more reoperations.

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Anatomic aspects of the mitral valve

In order that contraction of the heart muscles results in effective functioning, they need points of support. These are constituted by a fibrous skeleton in the heart, composed of the right and left fibrous trigones and of the rings of the aortic, mitral, tricuspid and pulmonary valves. All these valve rings are complete, except for the mitral valve ring, whose anterior portion, between the fibrous trigones does not exist with this portion occupied by the complete ring of the aortic valve [7]. The mitral valve is thus composed of an incomplete fibrous ring, a portion of the subjacent left ventricle in the ring and four leaflets (cusps), which are one anterior leaflet and three posterior leaflets (lateral, medial and posterior leaflets). The anterior one is an excursion cusp, while the posterior cusps are for contention. This structure includes the commissures which are nothing more than two other cusps which correspond to the point between the anterior cusp and the lateral and medial cusps (posterior).

This set of structures, in a normal heart, causes the ring has a shape similar to that of a kidney, as the greatest diameter is between the two trigones and corresponds to the anterior cusp. This diameter does not increase with enlargement of the left ventricle. Whatever the cause of the enlargement of the left ventricular (myocardiopathy, myocarditis or left ventricle volume overload) the mitral ring will dilate in the anteroposterior position – as Carpentier observed in his studies – with it being more pronounced on the right, near to the posterior-medial commissure, as previously described by Gregori Jr [8].

During ventricular systole, tendinous cords connected to the papillary muscles impede prolapse of the cusps thereby avoiding valve regurgitation. Tendinous cords can also be connected to the free wall of the left ventricle and can be marginal, intermediate or so-called structural, according to their insertion in the ventricular face of the cusps. The elongation or rupture of these cords always leads to valve prolapse with mitral valve regurgitation. Structural cords when torn cause significant mitral valve regurgitation.

The tendinous cords are a bundle of collagenous fibers composed of a central axis of dense collagen, encircled by loose collagen and elastin lined by endocardial cells. Histologically, tendinous cords present a structure similar to the cusps; with predominance of collagenous tissue, few elastic and muscle fibers covered by endothelial and endocardial cells. In the region of insertion, the structure of the cords is almost imperceptibly confused with the structure of the cusps.

The papillary muscles form two sets, one anterior and the other posterior. The anterior papillary set has two heads, one anterior and the other posterior. The posterior papillary

set frequently has three heads: one anterior, one intermediate and the other posterior. Tendinous cords attached to the anterior head are anchored to the anterior cusp, cords attached to the intermediate head are anchored to the commissures and the cords attached to the posterior head are anchored to the posterior cusp. The anterior papillary muscle receives blood irrigation from the anterior descending artery or the circumflex artery. The posterior papillary muscle is supplied by the right coronary artery or the circumflex artery and its branches.

Mitral annuloplasty

Annuloplasty consists in the repair of mitral valve annulus dilatation. It must be the last procedure of valve reconstruction, after the actuation of the cusps, tendinous cords and papillary muscles. In our experience, in only 15% of patients it constituted a procedure in isolation.

It has been performed, over the years, in several ways. To simplify understanding of the different methods, we propose a systematization of the annuloplasty according to the material used (sustentation tissues, prosthesis, etc.) and the form of actuation (posterior, circular), which can be seen in Table 1.

Plicatures

These were the first techniques utilized and generally, consist in the plicature of the mitral annulus near to the commissures, so that the posterior cusps are brought nearer to the anterior cusp, thereby repairing the central regurgitation. It may be symmetrical as suggested by Lillehei et al. [9] or asymmetric as proposed by Kay et al. [10], Wooler et al. [11] and Reed et al. [12], a technique which is still performed today. In the latter technique, sutures are passed through the mitral annulus near to the commissures, to restrain the posterior mitral annulus more intensively.

McGoon [13] and Merendino et al. [14] in the early 1960s, developed techniques to repair prolapse of the cusps. The technique of the latter should be considered an annuloplasty, as, after resection of the prolapsed portion of the posterior cusp, a plicature of the mitral ring is performed. More recently, other authors such as Burr et al. [15] proposed a circular restriction of the entire mitral annulus, a practice few adopted.

With sustentation tissues

In these cases, strips of synthetic materials such as Teflon, Dacron and polytetrafluoroethylene (PTFE) are applied to the posterior annulus, guiding the posterior cusps to the anterior cusp (Kay et al. [10], Pomerantzeff et al. [16], Salati et al. [17], Barbosa et al. [18]), as well as Hendren et al. [19] who uses bovine pericardium to substitute the synthetic materials.

Table 1. Mitral Annuloplasty

	POSTERIOR		CIRCULAR	
PLICATURES WITH SUTURES	- Lillehei	1957		
	- Kay	1958	- Burr	1976
	- Wooler	1962		
	- Reed	1965		
SUPPORT TISSUES	Teflon	- Kay 1958 - Jatene 1962	Dacron	- Cooley 1992
	PTFE	- Fundaró 1986 - Barbosa 1989	AP	- Victor 1993
	BP	- Hendren 1991	PTFE	- Warinsirikul 1999
	PROSTHESES	Rigid	- Gregori-Braile 1987	Rigid
Flexible		- Camilleri 1976	Flexible	- Duran 1976
		- Braile 1984		
		- Cosgrove 1993		- Puig-Massana 1980
		- Melo 1995		- Abdulmassih 1985
				- Shumway 1988
		- Sculptor 1990		
		- St. Jude-Biflex 1993		
		- Physio-ring 1994		

BP: Bovine pericardium, AP: autologous pericardium, PTFE: polytetrafluoroethylene

The involvement of all the perimeter of the mitral annulus was recommended by Cooley et al. [20] with Dacron rings and by Victor & Nayak [21] using autogenous pericardium treated in glutaraldehyde. Recently, Warinsirikul et al. [22] have utilized rings made during the operation in specific sizes for the case at hand which consist in a 3-mm diameter cylinder of PTFE, with a steel thread inside. This thread is initially tied and after the edges of the tube are brought together using silk stitches a “homemade-ring” with the ovoid format results, similar to the ring of Carpentier.

With posterior prosthesis

Rigid

The ring developed by us, denominated the Gregori-Braile ring [4], is a hemi-ring made of 316-medical stainless

steel, lined with a layer of silicone rubber and finally covered using Dacron. The absence of the anterior segment is based on the fact that the dilatation of the mitral annulus rarely or never occurs between the right and left fibrous trigones, the space occupied by the fibrous ring of the aortic valve. Its semicircular shape presents a rectification in its right half to repair the posterior dilatation of the mitral annulus more intensely to the right. It is produced in several sizes and can even be used in small children. The numbers vary from 24 to 36, according to the maximum opening. Each prosthesis corresponds to a metallic measure to be utilized, during the operation, to help to decide the correct size. This must be exclusively based on the distance between the projections of the commissures in the mitral annulus, without considering the anteroposterior diameter. The malleability

of the steel used in the prosthetic ring allows small modifications to its opening, which can be performed manually during the surgery, for a better prosthetic adjustment in the mitral annulus.

The anchoring of the prosthesis is achieved using 2-0 polyester thread initially passed in a "U-shape" in the mitral annulus followed by the external part of the prosthetic ring, usually using a total of seven to eight threads. With this prosthesis, the annular dilatation is repaired avoiding later complications of mitral stenosis in children and young patients, due to the restriction to the normal growth of the mitral annulus, observed when annuloplasties use circular prostheses.

Flexible posterior mitral annuloplasty

Camilleri et al. [23] have been using a linear reducer composed of a silicone polymer lined with a braid of Dacron and two copper threads in 20-centimeter segments. It is not possible to stretch or to constrict lengthways in order to achieve a desired reduction. It is, however, totally flexible in its width depending on the physiology of the mitral annulus.

Bovine pericardium prostheses described by Braile et al. [24] consist of a bovine pericardium tube treated in glutaraldehyde, which has a radiopaque polyester thread inside. Thus, the set, in spite of being elastic, is not subject to longitudinal deformation. After implantation using single 2-0 polyester sutures from trigone to trigone, the prosthesis is molded to the ring without deforming it, adequately bringing the posterior cusp closer to the anterior cusp.

Cosgrove et al. [25] developed a semicircular ring with a malleable strut and with a manual adjustment so that it can be regulated to the degree of reduction of the anteroposterior diameter, after its implantation. The strut is made of polyester to stimulate its rapid endothelialization. It is implanted using a rigid mold which is only removed when the prosthesis is totally anchored in the mitral annulus.

With circle prosthesis

Rigid prosthesis

In 1969 Carpentier [26] presented rigid circular rings made of stainless steel covered in Dacron in the shape of a kidney, similar to the normal mitral valve ring. With the implantation of this ring, the posterior cusp is brought closer to the anterior cusp in the anteroposterior direction, without interfering in the transversal diameter (inter-trigonal). It has the disadvantage of limiting the normal growth of the mitral annulus of the patient, when used in children. Additionally, obstruction of the left ventricle outflow tract due to the anterior movement of the mitral annulus during systole has been observed in patients who underwent annuloplasty for degenerative mitral

regurgitation [27], a fact rarely observed when open prosthesis are employed. It is important to stress that this is undoubtedly the most commonly used prosthesis in the world.

Flexible prosthesis

Among these, the most commonly used is the ring of Duran et al. [28]. It determines uniform circular restriction of the dilated mitral annulus, even the region between the fibrous trigones and demands the creativity of the surgeon so that the reduction of the anteroposterior diameter is adequate.

A series of prostheses were developed aiming at adjusting the annular constriction after their implantation as well as allowing the functional movement of the contraction of the mitral annulus which include the prostheses of Puig-Massana [29], Abdulmassih [30], Shumway [31], Sculptor [32] and St.Jude-Biflex [33].

More recently, Carpentier et al. [34] created the *Physio-ring*, which has the same format as the original ring, but with transverse flexibility in its posterior portion, allowing contractility of the correspondent part of the mitral annulus. As it is rigid lengthways, it avoids plicature and the constrictive effect in respect to its anchoring using sutures.

However, randomized, double blind studies with an adequate number of cases are necessary to come to a definitive conclusion in respect to the superiority of flexible rings in comparison to rigid prosthesis.

REPAIR OF MITRAL VALVE PROLAPSE

Mitral valve prolapse can be caused by elongation or rupture of the tendinous cords or by dysfunction or rupture of the secondary papillary muscles due to coronary regurgitation.

In Brazil, the commonest causes of injuries to the tendinous cords are rheumatic fever and myxomatous degeneration. In both, the mitral valve system is globally affected, specifically the tendinous cords. The technique to be used will be adequate depending on the degree of involvement and location of the tendinous cords. Posterior prolapse is generally treated with partial rectangular resection of the posterior cusp and plicature of the mitral annulus, as recommended by Merendino et al. [14]. Anterior prolapse was initially treated with cuneiform resections of the cusp, or even McGoon plicatures [13], both methods subsequently abandoned as the methods limit the mobility of the anterior cusp (excursion cusp). The same problem does not occur in the resection of the posterior cusp (contention cusp).

Several procedures were developed for the repair of anterior cusp prolapse.

Elongation of cords

Shortening of cords with intra-papillary burying, according to Carpentier [1]

This technique consists in first making a longitudinal incision in the head of the papillary muscle. Subsequently, a 5-0 polypropylene thread ties behind the cord or the bundle of elongated cords, with the needle exiting further down at the edge of the incision in the papillary muscle.

When it is tied, the cord is pulled down, thereby repairing, the lengthening of the cords.

The lower the needle emerges, the greater the shortening will be.

Shortening of cords with burying in the cusps, according to Gregori et al. [35]

This is very beneficial for children in whom the papillary muscles are thin and shortening is difficult.

An orifice of approximately 2 mm is performed in the anterior cusp of the mitral valve, near to the insertion of the elongated tendinous cord. Through this orifice the cord is pulled out of the valve plane, depending on the degree of shortening necessary. After this, raffia of the incision is started using 5-0 polypropylene, at the same time in which the cord, already shortened is fixed together with the orifice of the cusp, thereby repairing the prolapse. More than one shortening can be performed along the edge of the coaptation of the anterior cusp. Recently, this technique was also employed in a few cases to the posterior cusp giving satisfactory results.

Rupture of tendinous cords

Cord transference from the posterior cusp to the anterior cusp, according to Carpentier et al. [36]

This is a technique employed worldwide. A part of posterior cusp of the mitral valve, in contraposition to the prolapsed portion of the anterior cusp is removed. This piece, containing cusp and tendinous cords is displaced to the free border of the anterior cusp, near to the bundle of torn cords, and it is sutured there using 5-0 polypropylene, thus repairing the prolapse.

The borders of the divided posterior cusp are brought together by single 5-0 polypropylene sutures after plicature of the mitral annulus in the corresponding site.

Partial transference of the tricuspid valve to the mitral valve, according to Gregori et al. [37,38].

Two variant techniques are described. The first one, the most frequently used consists in the total removal of the posterior valve with all its elements, that is, cusp, tendinous cords and papillary muscle. This piece is then transferred to the mitral valve with torn cords, suturing the papillary muscle to the papillary muscle, utilizing 5-0 polypropylene "U-shaped" sutures anchored on Dacron pads. After, the cusp

of the graft is sutured to the free border of the anterior cusp of the mitral valve, thereby eliminating the possibility of long cords remaining that may maintain the prolapse and consequently the mitral valve regurgitation. The same care must be taken in the opposite case, leaving short cords, with retraction of the anterior cusp, impeding perfect coaptation with the posterior cusp.

The tricuspid ring is plicated using a 4-0 polypropylene "U-shaped" suture anchored to Dacron pads. The valve essentially becomes a bicuspid valve and possible functional tricuspid regurgitation is simultaneously repaired.

Anterior and posterior prolapse of the mitral valve can be simultaneously repaired dividing in half the cusp of the graft.

Neocord creation with a patch of the anterior cusp, according to Gregori et al. [39]

This is indicated, almost exclusively, in cases with anterior cusp of the mitral valve presenting with a well developed area (myxomatous degeneration or Barlow syndrome). A rectangular patch of the tissue of the anterior cusp measuring approximately 10 mm in length by 2 mm in width is removed. This patch is moved from the ring up to the free border of the anterior cusp, remaining attached only to the anterior cusp. Subsequently it is lowered to the fibrous head of the papillary muscle with ruptured cords where it is sutured, thus constituting a neocord. After a time, this rectangular tissue becomes thin, adopting the aspect of normal tendinous cord (a fact that was observed in a reoperated patient).

The orifice of the anterior cusp is sutured with single 5-0 polypropylene sutures and the anterior cusp of the mitral valve continues with an adequately size.

Artificial cords

Synthetic and biological materials have been utilized to substitute the tendinous cords.

David et al. [40] have used PTFE sutures since 1985 with excellent long-term results in patients with of mitral valve prolapse secondary to rupture or elongation of cords. They have been employed by several groups around the world.

Frater et al. [41] introduced the use of bovine pericardial strips to substitute the cords. The fear of calcification of this tissue has led surgeons not to use this method routinely.

OTHER RECONSTRUCTIVE TECHNIQUES

Removal of localized calcification, fenestration and sectioning of retractable cords, according to Carpentier [1]

Restriction in the mobility of the cusp is the result of commissural fusion, cord fusion, cord retraction or even hypertrophy of the cords. Sectioning of secondary cords can be performed mainly of those that originate in the left

ventricle posterior wall, thereby liberating the posterior cusp giving better coaptation with the anterior cusp. Also fenestration of the cords, which is sometimes necessary in rheumatic disease patients, by triangular resection of the fused cords, corrects subvalve stenosis, improving valve mobility. Localized calcifications can be removed with the resection of a wedge of the affected cusp, or even, by displacement of the calcium plaques.

Chance holes in these membranes can be corrected using autogenous pericardium or bovine pericardium patches, that have either been treated in glutaraldehyde or not.

Utilization of tissue patches

Autologous pericardium patches treated in glutaraldehyde, have been used to close post-endocarditis holes, for cusp enlargement or even substitution and after fibrous resections or calcifications [42].

Partial substitution of the mitral valve by autologous or homologous grafts

Hvass et al. [43] employed a technique similar to ours for cases of anterior cusp cord rupture, that is, for the partial transference of the tricuspid valve (posterior cusp) to the mitral valve, only in this case for the substitution of part of the valve after resection of calcification.

Revuelta et al. [44] demonstrated in an experimental work with sheep, the viability of using cryopreserved homografts for the repair of localized lesions of the mitral valve.

RESULTS IN CHILDREN

From May 1980 to November 2001, 117 under 15-year-old patients were submitted to conservative surgery of the mitral valve. All patients suffered from mitral valve regurgitation [45].

Their ages ranged from 1 to 15 years, with a mean of 10 years. Forty-three (36.8%) patients were male and 74 (63.2%) were female.

Eighty-seven (74.4%) patients suffered only from mitral valve regurgitation and 30 (25.5%) presented with associated stenosis. The etiology was rheumatic disease in 95 (81.2%) cases, congenital in 16 (13.7%), infectious endocarditis in five (4.3%) and myxomatous degeneration in one (9%) patient. All patients were classified in III and IV functional classes.

The patients were submitted to surgery with cardiopulmonary bypasses (CPB) and moderate hypothermia and myocardial protection was performed with intermittent clamping of the aorta at 15-minute intervals. The mean time of CPB was 68 minutes (22-158 min) and the mean myocardial ischemia time was 38 minutes (8-108 min).

In all patients the mitral valve annulus was remodeled.

Prostheses or support tissues were not utilized in only seven patients. In six patients (5.1%) bovine pericardial strips were used, in 35 patients (29.9%) Carpentier rings were used and in the other 69 patients (59.0%) Gregori-Braile rings were used. An isolated annuloplasty was performed in 22 patients (18.8%) and in 95 patients (81.2%) there was a necessity to operate on the valve cusps or on the subvalvar system.

Sixty-six patients (56.4%) were submitted to shortening of long cords, 30 patients (25.6%) to commissurotomy or papillectomy, 11 patients (9.4%) to the partial resection of the posterior cusp, nine patients (7.7%) to anterior cusp resection, six patients (5.1%) to posterior cusp plication, five patients (4.3%) to cord transposition, five patients (4.3%) to cord transference, five patients (4.3%) to retractable cords sectioning, four patients (3.4%) to suturing of orifices in the cusps and two patients (1.7%) to the removal of calcium on the anterior cusp.

The following associated procedures were performed: tricuspid valvuloplasty in six patients (5.1%), closing of an interatrial communication in three patients (2.6%), ligation of patent arterial ducts in two patients (1.7%), aortic valve replacement in 10 patients (8.5%), aortic valvuloplasty in six patients (5.1%), resection of a subaortic fibrous ring in one patient (0.9%), atrioventricular duct repair in one patient (0.9%) and Cox surgery without cryoablation in one patient (0.9%).

Thromboembolic phenomena did not occur.

One hundred and thirteen patients (96.6%) remain alive, 104 of them (88.9%) continue with their native valves and 99 (84.6%) are in I and II functional classes (NYHA).

The hospital mortality rate was one patient (0.9%) who suffered from uncontrolled cardiac insufficiency and the late mortality rate was three patients (2.6%), with two deaths occurring in reoperations.

CONCLUSIONS

Mitral valve reconstruction surgery has a great number of options and materials which can repair defects of the valve ring, cusps or subvalvar system, allowing adequate treatment of mitral valve regurgitation and consequent preservation of the valve. Thus, always when possible, this should be chosen, especially when treating children.

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